Revised Data Definitions Manual

PC4 software v3.0

This document defines each data element a user may need to enter into the registry. Please note that many items are only relevant for a subset of patients; such qualifiers are noted in the "Displayed if" section of the description. If the item is marked as required for case closure, it only applies to the subset of cases for which it is relevant.

Sites can opt out of sending any or all of the following fields to the registry:

- Medical record number
- Social security number / Social insurance number / National ID
- Patient first name
- Patient last name
- Patient DOB
- Mother's first name
- Mother's last name
- Birth zip code
- Residential zip code

Please work with your local compliance office to determine the data you can submit.

Wherever possible, fields are shared across registries and/or use common International Pediatric and Congenital Cardiac Code (IPCCC) definitions.

Data Definitions

Patient Information

MRN Seq Num: 120

Required for case closure: No *Submission is optional*

Registry field: [Demographics].[MedRecN]

Shared with PAC3

Description: Indicate the patient's medical record number at the hospital where the encounter occurred. This field should be collected in compliance with state/local privacy laws.

Social security number / Social insurance number/National ID Seq Num: 180

Required for case closure: No *Submission is optional*

Registry field: [Demographics].[NationalID]

Shared with PAC3

Description: Indicate the nine-digit Patient's Social Security Number (SSN). Although this is the Social Security Number in the USA, other countries may have a different National Patient Identifier Number. For example in Canada, this would be the Social Insurance Number. This field should be collected in compliance with state/local privacy laws.

Last name Seq Num: 140

Required for case closure: No *Submission is optional*

Registry field: [Demographics].[PatLName]

Shared with PAC3

Description: Indicate the patient's last name as documented in the medical record. This field should be collected in compliance with state/local privacy laws.

First name Seq Num: 160

Required for case closure: No *Submission is optional*

Registry field: [Demographics].[PatFName]

Shared with PAC3

Description: Indicate the patient's first name as documented in the medical record. This field should be collected in compliance with state/local privacy laws.

DOB Seq Num: 200

Required for case closure: Yes *Submission is optional*

Registry field: [Demographics].[DOB]

Shared with PAC3

Description: Indicate the patient's date of birth using 4-digit format for year. This field should be collected in compliance with state/local privacy laws.

Gender

Seq Num: 220

Required for case closure: Yes

Registry field: [Demographics].[Gender]

Shared with PAC3

Description: Indicate the patient's gender at birth as male, female or ambiguous.

Values	*Code*	*Text*
	1	Male
	2	Female
	3	Ambiguous

Mother's name known

Seq Num: 375

Required for case closure: Yes

Registry field: [Demographics].[MothKnown]

Shared with PAC3

Description: Indicate whether the name of patient's biological mother at time of patient's birth is known. If the patient is adopted and the name of the patient's biological mother is not known, indicate whether the name of the patient's adopted mother is known.

Values	*Code*	*Text*
	1	Yes
	0	No

Mother's last name

Seq Num: 380

Required for case closure: No *Submission is optional*

Registry field: [Demographics].[MothLName]

Shared with PAC3

Description: Indicate the last name of patient's biological mother at time of patient's birth, if it is known. If the patient is adopted, if the last name of the patient's biological mother is known, please enter the last initial of the patient's biological mother. If the patient is adopted, if the last name of the patient's biological mother is not known, please enter the last name of the patient's adopted mother. This field should be collected in compliance with state/local privacy laws.

Mother's first name

Seq Num: 400

Required for case closure: No *Submission is optional*

Registry field: [Demographics].[MothFName]

Shared with PAC3

Description: Indicate the first name of patient's biological mother at time of patient's birth, if it is known. If the patient is adopted, if the first name of the patient's biological mother is known, please enter the first initial of the patient's biological mother. If the patient is adopted, if the first name of the patient's biological mother is not known, please enter the first name of the patient's adopted mother. This field should be collected in compliance with state/local privacy laws.

Race documented

Seq Num: 230

Required for case closure: Yes

Registry field: [Demographics].[RaceDocumented]

Shared with PAC3

Description: Indicate whether race is documented.

Values	*Code*	*Text*
	1	Yes
	0	No
	3	Patient declined to disclose

Caucasian

Seq Num: 240

Required for case closure: Yes

Registry field: [Demographics].[RaceCaucasian]

Shared with PAC3

Description: Indicate whether the patient's race, as determined by the patient or family, includes Caucasian. This includes a person having origins in any of the original peoples of Europe, the Middle East, or North Africa. Definition source: Standards for Maintaining, Collecting, and Presenting Federal Data on Race and Ethnicity: The minimum categories for data on race and ethnicity for Federal statistics, program administrative reporting, and civil rights compliance reporting. (www.whitehouse.gov/omb/fedreg/1997standards.html)

Values	*Code*	*Text*
	1	Yes
	0	No

Black/African American

Seq Num: 260

Required for case closure: Yes

Registry field: [Demographics].[RaceBlack]

Shared with PAC3

Description: Indicate whether the patient's race, as determined by the patient or family, includes Black / African American. This includes a person having origins in any of the black racial groups of Africa. Terms such as "Haitian" or "Negro" can be used in addition to "Black or African American." Definition source: Standards for Maintaining, Collecting, and Presenting Federal Data on Race and Ethnicity: The minimum categories for data on race and ethnicity for Federal statistics, program administrative reporting, and civil rights compliance reporting. (www.whitehouse.gov/omb/fedreg/1997standards.html)

Values	*Code*	*Text*
	1	Yes
	0	No

Asian

Seq Num: 280

Required for case closure: Yes

Registry field: [Demographics].[RaceAsian]

Shared with PAC3

Description: Indicate whether the patient's race, as determined by the patient or family, includes Asian. This includes a person having origins in any of the original peoples of the Far East, Southeast Asia, or the Indian subcontinent including, for example, Cambodia, China, India, Japan, Korea, Malaysia, Pakistan, the Philippine Islands, Thailand, and Vietnam. Definition source: Standards for Maintaining, Collecting, and Presenting Federal Data on Race and Ethnicity: The minimum categories for data on race and ethnicity for Federal statistics, program administrative reporting, and civil rights compliance reporting. (www.whitehouse.gov/omb/fedreg/1997standards.html)

Values	*Code*	*Text*
	1	Yes
	0	No

Native American

Seq Num: 300

Required for case closure: Yes

Registry field: [Demographics].[RaceNativeAm]

Shared with PAC3

Description: Indicate whether the patient's race, as determined by the patient or family, includes American Indian / Alaskan Native. This includes a person having origins in any of the original peoples of North and South America (including Central America), and who maintains tribal affiliation or community attachment. Definition source: Standards for Maintaining, Collecting, and Presenting Federal Data on Race and Ethnicity: The minimum categories for data on race and ethnicity for Federal statistics, program administrative reporting, and civil rights compliance reporting. (www.whitehouse.gov/omb/fedreg/1997standards.html)

Values	*Code*	*Text*
	1	Yes
	0	No

Native Pacific Islander

Seq Num: 320

Required for case closure: Yes

Registry field: [Demographics].[RaceNativePI]

Shared with PAC3

Description: Indicate whether the patient's race, as determined by the patient or family, includes Native Hawaiian / Pacific Islander. This includes a person having origins in any of the original peoples of Hawaii, Guam, Samoa, or other Pacific Islands. Definition source: Standards for Maintaining, Collecting, and Presenting Federal Data on Race and Ethnicity: The minimum categories for data on race and ethnicity for Federal statistics, program administrative reporting, and civil rights compliance reporting. (www.whitehouse.gov/omb/fedreg/1997standards.html)

Values	*Code*	*Text*
	1	Yes
	0	No

Other race

Seq Num: 340

Required for case closure: Yes

Registry field: [Demographics].[RaceOther]

Shared with PAC3

Description: Indicate whether the patient's race, as determined by the patient or family, includes any other race.

Values	*Code*	*Text*
	1	Yes
	0	No

Hispanic or Latino ethnicity Seq Num: 360

Required for case closure: Yes

Registry field: [Demographics].[Ethnicity]

Shared with PAC3

Description: Indicate if the patient is of Hispanic or Latino ethnicity as determined by the patient / family. Hispanic or Latino ethnicity includes patient report of Cuban, Mexican, Puerto Rican, South or Central American, or other Spanish culture or origin, regardless of race.

Values	*Code*	*Text*
	1	Yes
	0	No
	3	Not documented

Premature birth Seq Num: 417

Required for case closure: Yes

Registry field: [Demographics].[Preterm]

Shared with PAC3

Description: If the patient age is <= 1 year, indicate whether patient was born prematurely as defined by a gestational period of less than 37 weeks.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Gestational age at birth known Seq Num: 420

Required for case closure: Yes

Registry field: [Demographics].[GestAgeKnown]

Shared with PAC3

Description: If the patient age is <= 1 year, indicate whether the patient's gestational age at birth is known

Values	*Code*	*Text*
	1	Yes
	0	No

Gestational age – weeks Seq Num: 440

Required for case closure: Yes

Registry field: [Demographics].[GestAgeWks]

Shared with PAC3

Description: If the gestational age is known, indicate the patient's estimated gestational age at birth in weeks.

Birth weight known Seq Num: 460

Required for case closure: Yes

Registry field: [Demographics].[BirthWtKnown]

Shared with PAC3

Description: If the patient age is <= 30 days, indicate whether the patient's birth weight is known.

Values	*Code*	*Text*
	1	Yes
	0	No

Birth weight (kg) Seq Num: 480

Required for case closure: Yes

Registry field: [Demographics].[BirthWt]

Shared with PAC3

Description: If the birth weight is known, indicate the weight in kilograms of the patient at birth.

Birth length (cm) Seq Num: 500

Required for case closure: No

Registry field: [Demographics].[BirthLen]

Shared with PAC3

Description: If the patient age is <= 30 days, indicate the length in centimeters of the patient at birth.

Birth head circumference (cm) Seq Num: 520

Required for case closure: No

Registry field: [Demographics].[BirthHCircum]

Shared with PAC3

Description: If the patient age is <= 30 days, indicate the head circumference in centimeters of the patient at birth.

Birth country Seq Num: 600

Required for case closure: No

Registry field: [Demographics].[BirthCountry]

Shared with PAC3

Description: If the patient age is <= 30 days, indicate the country in which patient was born. For a list of values, see the "Country of Residence" field in the Episode of Care section.

Values

Code	*Text*
USA	UNITED STATES OF AMERICA
AFG	AFGHANISTAN
ALA	ÅLAND ISLANDS
ALB	ALBANIA
DZA	ALGERIA
ASM	AMERICAN SAMOA
AND	ANDORRA
AGO	ANGOLA
AIA	ANGUILLA
ATG	ANTIGUA AND BARBUDA
ARG	ARGENTINA
ARM	ARMENIA
ABW	ARUBA
AUS	AUSTRALIA
AUT	AUSTRIA
AZE	AZERBAIJAN
BHS	BAHAMAS
BHR	BAHRAIN
BGD	BANGLADESH
BRB	BARBADOS
BLR	BELARUS
BEL	BELGIUM
BLZ	BELIZE
BEN	BENIN
BMU	BERMUDA
BTN	BHUTAN
BOL	BOLIVIA (PLURINATIONAL STATE OF)
BES	BONAIRE, SAINT EUSTATIUS AND SABA
BIH	BOSNIA AND HERZEGOVINA
BWA	BOTSWANA
BRA	BRAZIL
VGB	BRITISH VIRGIN ISLANDS

BRN	BRUNEI DARUSSALAM
BGR	BULGARIA
BFA	BURKINA FASO
BDI	BURUNDI
KHM	CAMBODIA
CMR	CAMEROON
CAN	CANADA
CPV	CAPE VERDE
CYM	CAYMAN ISLANDS
CAF	CENTRAL AFRICAN REPUBLIC
TCD	CHAD
CHL	CHILE
CHN	CHINA
COL	COLOMBIA
COM	COMOROS
COG	CONGO
COK	COOK ISLANDS
CRI	COSTA RICA
CIV	CÔTE D'IVOIRE
HRV	CROATIA
CUB	CUBA
CUW	CURAÇAO
CYP	CYPRUS
CZE	CZECH REPUBLIC
PRK	DEMOCRATIC PEOPLE'S REPUBLIC OF KOREA
COD	DEMOCRATIC REPUBLIC OF THE CONGO
DNK	DENMARK
DJI	DJIBOUTI
DMA	DOMINICA
DOM	DOMINICAN REPUBLIC
ECU	ECUADOR
EGY	EGYPT
SLV	EL SALVADOR
GNQ	EQUATORIAL GUINEA
ERI	ERITREA
EST	ESTONIA
ETH	ETHIOPIA
FRO	FAEROE ISLANDS
FLK	FALKLAND ISLANDS (MALVINAS)
FJI	FIJI

FIN	FINLAND
FRA	FRANCE
GUF	FRENCH GUIANA
PYF	FRENCH POLYNESIA
GAB	GABON
GMB	GAMBIA
GEO	GEORGIA
DEU	GERMANY
GHA	GHANA
GIB	GIBRALTAR
GRC	GREECE
GRL	GREENLAND
GRD	GRENADA
GLP	GUADELOUPE
GUM	GUAM
GTM	GUATEMALA
GGY	GUERNSEY
GIN	GUINEA
GNB	GUINEA-BISSAU
GUY	GUYANA
HTI	HAITI
VAT	HOLY SEE
HND	HONDURAS
HKG	CHINA, HONG KONG SPECIAL ADMINISTRATIVE REGION
HUN	HUNGARY
ISL	ICELAND
IND	INDIA
IDN	INDONESIA
IRN	IRAN (ISLAMIC REPUBLIC OF)
IRQ	IRAQ
IRL	IRELAND
IMN	ISLE OF MAN
ISR	ISRAEL
ITA	ITALY
JAM	JAMAICA
JPN	JAPAN
JEY	JERSEY
JOR	JORDAN
KAZ	KAZAKHSTAN
KEN	KENYA

KIR	KIRIBATI
KWT	KUWAIT
KGZ	KYRGYZSTAN
LAO	LAO PEOPLE'S DEMOCRATIC REPUBLIC
LVA	LATVIA
LBN	LEBANON
LSO	LESOTHO
LBR	LIBERIA
LBY	LIBYA
LIE	LIECHTENSTEIN
LTU	LITHUANIA
LUX	LUXEMBOURG
MAC	CHINA, MACAO SPECIAL ADMINISTRATIVE REGION
MDG	MADAGASCAR
MWI	MALAWI
MYS	MALAYSIA
MDV	MALDIVES
MLI	MALI
MLT	MALTA
MHL	MARSHALL ISLANDS
MTQ	MARTINIQUE
MRT	MAURITANIA
MUS	MAURITIUS
MYT	MAYOTTE
MEX	MEXICO
FSM	MICRONESIA (FEDERATED STATES OF)
MCO	MONACO
MNG	MONGOLIA
MNE	MONTENEGRO
MSR	MONTSERRAT
MAR	MOROCCO
MOZ	MOZAMBIQUE
MMR	MYANMAR
NAM	NAMIBIA
NRU	NAURU
NPL	NEPAL
NLD	NETHERLANDS
NCL	NEW CALEDONIA
NZL	NEW ZEALAND

NIC	NICARAGUA
NER	NIGER
NGA	NIGERIA
NIU	NIUE
NFK	NORFOLK ISLAND
MNP	NORTHERN MARIANA ISLANDS
NOR	NORWAY
PSE	OCCUPIED PALESTINIAN TERRITORY
OMN	OMAN
PAK	PAKISTAN
PLW	PALAU
PAN	PANAMA
PNG	PAPUA NEW GUINEA
PRY	PARAGUAY
PER	PERU
PHL	PHILIPPINES
PCN	PITCAIRN
POL	POLAND
PRT	PORTUGAL
PRI	PUERTO RICO
QAT	QATAR
KOR	REPUBLIC OF KOREA
MDA	REPUBLIC OF MOLDOVA
REU	RÉUNION
ROU	ROMANIA
RUS	RUSSIAN FEDERATION
RWA	RWANDA
SHN	SAINT HELENA
KNA	SAINT KITTS AND NEVIS
LCA	SAINT LUCIA
SPM	SAINT PIERRE AND MIQUELON
VCT	SAINT VINCENT AND THE GRENADINES
BLM	SAINT-BARTHÉLEMY
MAF	SAINT-MARTIN (FRENCH PART)
WSM	SAMOA
SMR	SAN MARINO
STP	SAO TOME AND PRINCIPE
SAU	SAUDI ARABIA
SEN	SENEGAL
SRB	SERBIA

SYC	SEYCHELLES
SLE	SIERRA LEONE
SGP	SINGAPORE
SXM	SINT MAARTEN (DUTCH PART)
SVK	SLOVAKIA
SVN	SLOVENIA
SLB	SOLOMON ISLANDS
SOM	SOMALIA
ZAF	SOUTH AFRICA
SSD	SOUTH SUDAN
ESP	SPAIN
LKA	SRI LANKA
SDN	SUDAN
SUR	SURINAME
SJM	SVALBARD AND JAN MAYEN ISLANDS
SWZ	SWAZILAND
SWE	SWEDEN
CHE	SWITZERLAND
SYR	SYRIAN ARAB REPUBLIC
TJK	TAJIKISTAN
THA	THAILAND
MKD	THE FORMER YUGOSLAV REPUBLIC OF MACEDONIA
TLS	TIMOR-LESTE
TGO	TOGO
TKL	TOKELAU
TON	TONGA
TTO	TRINIDAD AND TOBAGO
TUN	TUNISIA
TUR	TURKEY
TKM	TURKMENISTAN
TCA	TURKS AND CAICOS ISLANDS
TUV	TUVALU
UGA	UGANDA
UKR	UKRAINE
ARE	UNITED ARAB EMIRATES
GBR	UNITED KINGDOM OF GREAT BRITAIN AND NORTHERN IRELAND
TZA	UNITED REPUBLIC OF TANZANIA
VIR	UNITED STATES VIRGIN ISLANDS
URY	URUGUAY

UZB	UZBEKISTAN
VUT	VANUATU
VEN	VENEZUELA (BOLIVARIAN REPUBLIC OF)
VNM	VIET NAM
WLF	WALLIS AND FUTUNA ISLANDS
ESH	WESTERN SAHARA
YEM	YEMEN
ZMB	ZAMBIA
ZWE	ZIMBABWE
OTH	Other

Birth zip code/postal code

Seq Num: 620

Required for case closure: No *Submission is optional*

Registry field: [Demographics].[BirthZip]

Shared with PAC3

Description: If the patient age is <= 30 days and the patient was born in the United States or Canada, indicate the zip/postal code of residence at birth. This field should be collected in compliance with state/local privacy laws.

Antenatal diagnosis of congenital heart disease

Seq Num: 540

Required for case closure: Yes

Registry field: [Demographics].[AntenatalDiag]

Shared with PAC3

Description: If the patient age is <= 30 days, indicate whether a cardiac anomaly was diagnosed antenatally (e.g., fetal ultrasound)

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

In-vitro Fertilization Seq Num: 560

Required for case closure: No

Registry field: [Demographics].[BirthIVF]

Shared with PAC3

Description: If the patient age is <= 30 days, indicate if there is any notation that the patient was conceived through in-vitro fertilization

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

In- or out-born Seq Num: 580

Required for case closure: No

Registry field: [Demographics].[BornLoc]

Shared with PAC3

Description: If the patient age is <= 30 days, indicate Yes if the patient was born at this hospital or one in the immediate vicinity and affiliated with the PC4/PAC3 institution (e.g. Brigham and Women's Hospital and Boston Children's).

Values	*Code*	*Text*
	1	Yes
	0	No

Fundamental diagnosis

Seq Num: 640

Required for case closure: Yes

Registry field: [Demographics].[FundDiagnosis]

Shared with PAC3

Description: The fundamental diagnosis is a diagnosis that is carried with a patient throughout life, through all operations and hospitalizations. The fundamental diagnosis is the most complex cardiac anomaly or condition (congenital or acquired) of the patient. This may be the same as or differ from the patient's encounter cardiothoracic diagnosis. For example, a patient with HLHS admitted from home with new onset systemic AV valve regurgitation would have a fundamental diagnosis of "Hypoplastic Left Heart Syndrome" but an encounter cardiothoracic diagnosis of "Tricuspid regurgitation". The appropriate arrhythmia diagnosis should be listed as the fundamental diagnosis for patients with no other forms of structural heart disease that would be otherwise listed as a fundamental diagnosis.

Values	*Code*	*Text*	
	10	PFO	A small interatrial communication (or potential communication) confined to the region of the oval fossa (fossa ovalis) characterized by no deficiency of the primary atrial septum (septum primum) and a normal limbus with no deficiency of the septum secundum (superior interatrial fold).
	20	ASD, Secundum	A congenital cardiac malformation in which there is an interatrial communication confined to the region of the oval fossa (fossa ovalis), most commonly due to a deficiency of the primary atrial septum (septum primum) but deficiency of the septum secundum (superior interatrial fold) may also contribute.
	30	ASD, Sinus venosus	A congenital cardiac malformation in which there is a caval vein (vena cava) and/or pulmonary vein (or veins) that overrides the atrial septum or the septum secundum (superior interatrial fold) producing an interatrial or anomalous venoatrial communication. Although the term sinus venosus atrial septal defect is commonly used, the lesion is more properly termed a sinus venosus communication because, while it functions as an interatrial communication, this lesion is not a defect of the atrial septum.
	40	ASD, Coronary sinus	A congenital cardiac malformation in which there is a deficiency of the walls separating the left atrium from the coronary sinus allowing interatrial communication through the coronary sinus ostium.
	50	ASD, Common atrium (single atrium)	Complete absence of the interatrial septum. "Single atrium" is applied to defects with no associated malformation of the atrioventricular valves. "Common atrium" is applied to defects with associated malformation of the atrioventricular valves.
	71	VSD, Type 1 (Subarterial) (Supracristal) (Conal septal defect) (Infundibular)	A VSD that lies beneath the semilunar valve(s) in the conal or outlet septum.
	73	VSD, Type 2 (Perimembranous)	A VSD that is confluent with and involves the membranous

73	(Paramembranous) (Conoventricular)	septum and is bordered by an atrioventricular valve, not including type 3 VSDs.
75	VSD, Type 3 (Inlet) (AV canal type)	A VSD that involves the inlet of the right ventricular septum immediately inferior to the AV valve apparatus.
77	VSD, Type 4 (Muscular)	A VSD completely surrounded by muscle.
79	VSD, Type: Gerbode type (LV-RA communication)	A rare form of VSD in which the defect is at the membranous septum; the communication is between the left ventricle and right atrium.
80	VSD, Multiple	More than one VSD exists. Each individual VSD may be coded separately to specify the individual VSD types.
100	AVC (AVSD), Complete (CAVSD)	Indicate if the patient has the diagnosis of “AVC (AVSD), Complete (CAVSD)”. An “AVC (AVSD), Complete (CAVSD)” is a “complete atrioventricular canal” or a “complete atrioventricular septal defect” and occurs in a heart with the phenotypic feature of a common atrioventricular junction. An “AVC (AVSD), Complete (CAVSD)” is defined as an AVC with a common AV valve and both a defect in the atrial septum just above the AV valve (ostium primum ASD [a usually crescent-shaped ASD in the inferior (posterior) portion of the atrial septum just above the AV valve]) and a defect in the ventricular septum just below the AV valve. The AV valve is one valve that bridges both the right and left sides of the heart. Balanced AVC is an AVC with two essentially appropriately sized ventricles. Unbalanced AVC is an AVC defect with two ventricles in which one ventricle is inappropriately small. Such a patient may be thought to be a candidate for biventricular repair, or, alternatively, may be managed as having a functionally univentricular heart. AVC lesions with unbalanced ventricles so severe as to preclude biventricular repair should be classified as single ventricles. Rastelli type A: The common superior (anterior) bridging leaflet is effectively split in two at the septum. The left superior (anterior) leaflet is entirely over the left ventricle and the right superior (anterior) leaflet is similarly entirely over the right ventricle. The division of the common superior (anterior) bridging leaflet into left and right components is caused by extensive attachment of the superior (anterior) bridging leaflet to the crest of the ventricular septum by chordae tendineae. Rastelli type B: Rare, involves anomalous papillary muscle attachment from the right side of the ventricular septum to the left side of the common superior (anterior) bridging leaflet. Rastelli type C: Marked bridging of the ventricular septum by the superior (anterior) bridging leaflet, which floats freely (often termed a "free-floater") over the ventricular septum without chordal attachment to the crest of the ventricular septum.
110	AVC (AVSD), Intermediate (transitional)	An AVC with two distinct left and right AV valve orifices but also with both an ASD just above and a VSD just below the AV valves. While these AV valves in the intermediate form do form two separate orifices they remain abnormal valves. The VSD is often restrictive.
120	AVC (AVSD), Partial (incomplete) (PAVSD) (ASD, primum)	An AVC with an ostium primum ASD (a usually crescent-shaped ASD in the inferior (posterior) portion of the atrial septum just above the AV valve) and varying degrees of

120		malformation of the left AV valve leading to varying degrees of left AV valve regurgitation. No VSD is present.
140	AP window (aortopulmonary window)	Indicate if the patient has the diagnosis of “AP window (aortopulmonary window)”. An “AP window (aortopulmonary window)” is defined as a defect with side-to-side continuity of the lumens of the aorta and pulmonary arterial tree, which is distinguished from common arterial trunk (truncus arteriosus) by the presence of two arterial valves or their atretic remnants. (In other words, an aortopulmonary window is a communication between the main pulmonary artery and ascending aorta in the presence of two separate semilunar [pulmonary and aortic] valves. The presence of two separate semilunar valves distinguishes AP window from truncus arteriosus. Type 1 proximal defect: AP window located just above the sinus of Valsalva, a few millimeters above the semilunar valves, with a superior rim but little inferior rim separating the AP window from the semilunar valves. Type 2 distal defect: AP window located in the uppermost portion of the ascending aorta, with a well-formed inferior rim but little superior rim. Type 3 total defect: AP window involving the majority of the ascending aorta, with little superior and inferior rims. The intermediate type of AP window is similar to the total defect but with adequate superior and inferior rims. In the event of AP window occurring in association with interrupted aortic arch, code “Interrupted aortic arch + AP window (aortopulmonary window)”, and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and AP window separately to provide further documentation about the individual interrupted arch and AP window types.)
150	Pulmonary artery origin from ascending aorta (hemitruncus)	One pulmonary artery arises from the ascending aorta and the other pulmonary artery arises from the right ventricle. DOES NOT include origin of the right or left pulmonary artery from the innominate artery or the aortic arch via a patent ductus arteriosus or collateral artery.
160	Truncus arteriosus	Indicate if the patient has the diagnosis of “Truncus arteriosus”. A truncus arteriosus is also known as a common arterial trunk and is defined as a heart in which a single arterial trunk arises from the heart, giving origin to the coronary arteries, the pulmonary arteries, and the systemic arterial circulation. In the majority of instances there is a ventricular septal defect and a single semilunar valve which may contain two, three, four, or more leaflets and is occasionally dysplastic. Often, the infundibular septum is virtually absent superiorly. In most instances the truncal valve overrides the true interventricular septum (and thus both ventricles), but very rarely the truncal valve may override the right ventricle entirely. In such instances, there may be no ventricular septal defect or a very small ventricular septal defect, in which case the left ventricle and mitral valve may be extremely hypoplastic.
170	Truncal valve insufficiency	Functional abnormality - insufficiency - of the truncal valve. May be further subdivided into grade of insufficiency (I, II, III, IV or mild, moderate, severe).
2010	Truncus arteriosus + Interrupted	Indicate if the patient has the diagnosis of “Truncus

2010	aortic arch	arteriosus + Interrupted aortic arch". {A truncus arteriosus is also known as a common arterial trunk and is defined as a heart in which a single arterial trunk arises from the heart, giving origin to the coronary arteries, the pulmonary arteries, and the systemic arterial circulation. In the majority of instances there is a ventricular septal defect and a single semilunar valve which may contain two, three, four, or more leaflets and is occasionally dysplastic. The infundibular septum is virtually absent superiorly. In most instances the truncal valve overrides the true interventricular septum (and thus both ventricles), but very rarely the truncal valve may override the right ventricle entirely. If in such case there is no ventricular septal defect, then the left ventricle and mitral valve may be extremely hypoplastic.} {Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.}
180	Partial anomalous pulmonary venous connection (PAPVC)	Some, but not all of the pulmonary veins connect to the right atrium or to one or more of its venous tributaries. This definition excludes sinus venosus defects with normally connected but abnormally draining pulmonary veins (the pulmonary veins may drain abnormally into the right atrium via the atrial septal defect).
190	Partial anomalous pulmonary venous connection (PAPVC), scimitar	The right pulmonary vein(s) connect anomalously to the inferior vena cava or to the right atrium at the insertion of the inferior vena cava. The descending vertical vein resembles a scimitar (Turkish sword) on frontal chest x-ray. Frequently associated with: hypoplasia of the right lung with bronchial anomalies; dextroposition and/or dextrorotation of the heart; hypoplasia of the right pulmonary artery; and anomalous subdiaphragmatic systemic arterial supply to the lower lobe of the right lung directly from the aorta or its main branches.
200	Total anomalous pulmonary venous connection (TAPVC), Type 1 (supracardiac)	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium. In Type 1 (supracardiac) TAPVC, the anomalous connection is at the supracardiac level and can be obstructed or nonobstructed.
210	Total anomalous pulmonary venous connection (TAPVC), Type 2 (cardiac)	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium. In Type 2 (cardiac) TAPVC, the anomalous connection is to the heart, either to the right atrium directly or to the coronary sinus. Most patients with type 2 TAPVC are nonobstructed.
220	Total anomalous pulmonary venous connection (TAPVC), Type 3 (infracardiac)	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left

220		atrium. In Type 3 (infracardiac) TAPVC, the anomalous connection is at the infracardiac level (below the diaphragm), with the pulmonary venous return entering the right atrium ultimately via the inferior vena cava. In the vast majority of patients infracardiac TAPVC is obstructed.
230	Total anomalous pulmonary venous connection (TAPVC), Type 4 (mixed)	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium. In Type 4 (mixed) TAPVC, the anomalous connection is at two or more of the above levels (supracardiac, cardiac, infracardiac) and can be obstructed or nonobstructed.
250	Cor triatriatum	In the classic form of cor triatriatum a membrane divides the left atrium (LA) into a posterior accessory chamber that receives the pulmonary veins and an anterior chamber (LA) that communicates with the mitral valve. In differentiating cor triatriatum from supravalvar mitral ring, in cor triatriatum the posterior compartment contains the pulmonary veins while the anterior contains the left atrial appendage and the mitral valve orifice; in supravalvar mitral ring, the anterior compartment contains only the mitral valve orifice. Cor triatriatum dexter (prominent venous valve producing obstruction of the IVC and tricuspid valve) is to be coded as a systemic venous obstruction, not as a form of cor triatriatum.
260	Pulmonary venous stenosis	Any pathologic narrowing of one or more pulmonary veins. Can be further subdivided by etiology (congenital, acquired-postoperative, acquired-nonpostoperative) and extent of stenosis (diffusely hypoplastic, long segment focal/tubular stenosis, discrete stenosis).
2480	Pulmonary venous stenosis, Acquired	
2490	Pulmonary venous stenosis, Spontaneous	
270	Systemic venous anomaly	Anomalies of the systemic venous system (superior vena cava (SVC), inferior vena cava (IVC), brachiocephalic veins (often the innominate vein), azygos vein, coronary sinus, levo-atrial cardinal vein) arising from one or more anomalies of origin, duplication, course, or connection. Examples include abnormal or absent right SVC with LSVC, bilateral SVC, interrupted right or left IVC, azygos continuation of IVC, and anomalies of hepatic drainage. Bilateral SVC may have, among other configurations: 1) RSVC draining to the RA and the LSVC to the LA with completely unroofed coronary sinus, 2) RSVC draining to the RA and LSVC to the coronary sinus which drains (normally) into the RA, or 3) RSVC to the coronary sinus which drains (abnormally) into the LA and LSVC to LA. Anomalies of the inferior vena caval system include, among others: 1) left IVC to LA, 2) biatrial drainage, or 3) interrupted IVC (left or right) with azygos continuation to an LSVC or RSVC.
280	Systemic venous obstruction	Obstruction of the systemic venous system (superior vena cava (SVC), inferior vena cava (IVC), brachiocephalic veins (often the innominate vein), azygos vein, coronary sinus, levo-atrial cardinal vein) arising from congenital or acquired stenosis or occlusion. Cor triatriatum dexter (prominent

280		venous valve producing obstruction of the IVC and tricuspid valve) is to be coded as a systemic venous obstruction, not as a form of cor triatriatum.
290	TOF	Indicate if the patient has the diagnosis of “TOF”. Only use this diagnosis if it is NOT known if the patient has one of the following four more specific diagnoses: (1). “TOF, Pulmonary stenosis”, (2). “TOF, AVC (AVSD)”, (3). “TOF, Absent pulmonary valve”, (4). “Pulmonary atresia, VSD (Including TOF, PA)”, or (5). “Pulmonary atresia, VSD-MAPCA (pseudotruncus)”.{“TOF” is “Tetralogy of Fallot” and is defined as a group of malformations with biventricular atrioventricular alignments or connections characterized by anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta. Hearts with tetralogy of Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, and aortic override; hearts with tetralogy of Fallot will most often have right ventricular hypertrophy.} (An additional, often muscular [Type 4] VSD may be seen with TOF and should be coded separately as a secondary diagnosis as “VSD, Type 4 (Muscular)”. Pulmonary arteries may be diminutive or there may be an absent left or right pulmonary artery; additional coding for pulmonary artery and/or branch pulmonary artery stenoses may be found under RVOT obstruction. Abnormal coronary artery distribution may also be associated with tetralogy of Fallot and may be coded separately under coronary artery anomalies. The presence of associated anomalies such as additional VSD, atrial septal defect, right aortic arch, left superior vena cava, and coronary artery anomalies must be subspecified as an additional or secondary diagnosis under the primary TOF diagnosis. TOF with absent pulmonary valve or TOF with associated complete atrioventricular canal are NOT to be secondary diagnoses under TOF - they are separate entities and should be coded as such. Controversy surrounds the differentiation between TOF and double outlet right ventricle [DORV]; in the nomenclature used here, DORV is defined as a type of ventriculoarterial connection in which both great vessels arise predominantly from the right ventricle. TOF with pulmonary atresia is to be coded under "Pulmonary atresia-VSD.")
2140	TOF, Pulmonary stenosis	Indicate if the patient has the diagnosis of “TOF, Pulmonary stenosis”. Use this diagnosis if the patient has tetralogy of Fallot and pulmonary stenosis. Do not use this diagnosis if the patient has tetralogy of Fallot and pulmonary atresia. Do not use this diagnosis if the patient has tetralogy of Fallot and absent pulmonary valve. Do not use this diagnosis if the patient has tetralogy of Fallot and atrioventricular canal. {Tetralogy of Fallot is defined as a group of malformations with biventricular atrioventricular alignments or connections characterized by anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta. Hearts with tetralogy of

2140		Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, and aortic override; hearts with tetralogy of Fallot will most often have right ventricular hypertrophy. (An additional, often muscular [Type 4] VSD may be seen with TOF and should be coded separately as a secondary diagnosis as “VSD, Type 4 (Muscular)”. Pulmonary arteries may be diminutive or there may be an absent left or right pulmonary artery; additional coding for pulmonary artery and/or branch pulmonary artery stenoses may be found under RVOT obstruction. Abnormal coronary artery distribution may also be associated with tetralogy of Fallot and may be coded separately under coronary artery anomalies. The presence of associated anomalies such as additional VSD, atrial septal defect, right aortic arch, left superior vena cava, and coronary artery anomalies must be subspecified as an additional or secondary diagnosis under the primary TOF diagnosis. TOF with absent pulmonary valve or TOF with associated complete atrioventricular canal are NOT to be secondary diagnoses under TOF - they are separate entities and should be coded as such. Controversy surrounds the differentiation between TOF and double outlet right ventricle [DORV]; in the nomenclature used here, DORV is defined as a type of ventriculoarterial connection in which both great vessels arise predominantly from the right ventricle. TOF with pulmonary atresia is to be coded under "Pulmonary atresia-VSD.")}
300	TOF, AVC (AVSD)	TOF with complete common atrioventricular canal defect is a rare variant of common atrioventricular canal defect with the associated conotruncal abnormality of TOF. The anatomy of the endocardial cushion defect is that of Rastelli type C in almost all cases.
310	TOF, Absent pulmonary valve	Indicate if the patient has the diagnosis of “TOF, Absent pulmonary valve”. “TOF, Absent pulmonary valve” is “Tetralogy of Fallot with Absent pulmonary valve” and is defined as a malformation with all of the morphologic characteristics of tetralogy of Fallot (anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta), in which the ventriculo-arterial junction of the right ventricle with the main pulmonary artery features an atypical valve with rudimentary cusps that lack the anatomical semi-lunar features of normal valve cusps and which functionally do not achieve central coaptation. The physiologic consequence is usually a combination of variable degrees of both stenosis and regurgitation of the pulmonary valve. A developmental accompaniment of this anatomy and physiology is dilatation of the main pulmonary artery and central right and left pulmonary arteries, which when extreme, is associated with abnormal arborization of lobar and segmental pulmonary artery branches and with compression of the trachea and mainstem bronchi. One theory holds that absence of the arterial duct or ductal ligament (which is a nearly constant finding in cases of tetralogy of Fallot with absent pulmonary valve) in

310		combination with pulmonary `valve stenosis and regurgitation, comprise the physiologic conditions which predispose to central pulmonary artery dilatation during fetal development. (Tetralogy of Fallot with Absent Pulmonary Valve Syndrome is a term frequently used to describe the clinical presentation when it features both circulatory alterations and respiratory distress secondary to airway compression.)
320	Pulmonary atresia	Pulmonary atresia defects which do not readily fall into pulmonary atresia-intact ventricular septum or pulmonary atresia-VSD (with or without MAPCAs) categories. These may include complex lesions in which pulmonary atresia is a secondary diagnosis, for example, complex single ventricle malformations with associated pulmonary atresia.
330	Pulmonary atresia, IVS	Pulmonary atresia (PA) and intact ventricular septum (IVS) is a duct-dependent congenital malformation that forms a spectrum of lesions including atresia of the pulmonary valve, a varying degree of right ventricle and tricuspid valve hypoplasia, and anomalies of the coronary circulation. An RV dependent coronary artery circulation is present when coronary artery fistulas (coronary sinusoids) are associated with a proximal coronary artery stenosis. Associated Ebstein's anomaly of the tricuspid valve can be present; the tricuspid diameter is enlarged and the prognosis is poor.
340	Pulmonary atresia, VSD (Including TOF, PA)	Pulmonary atresia (PA) and ventricular septal defect (VSD) is a heterogeneous group of congenital cardiac malformations in which there is lack of luminal continuity and absence of blood flow from either ventricle (in cases with ventriculo-arterial discordance) and the pulmonary artery, in a biventricular heart that has an opening or a hole in the interventricular septum (VSD). The malformation forms a spectrum of lesions including tetralogy of Fallot with pulmonary atresia. Tetralogy of Fallot with PA is a specific type of PA-VSD where the intracardiac malformation is more accurately defined (extreme underdevelopment of the RV infundibulum with marked anterior and leftward displacement of the infundibular septum often fused with the anterior wall of the RV resulting in complete obstruction of blood flow into the pulmonary artery and associated with a large outlet, subaortic ventricular septal defect). In the vast majority of cases of PA-VSD the intracardiac anatomy is that of TOF. The pulmonary circulation in PA-VSD is variable in terms of origin of blood flow, presence or absence of native pulmonary arteries, presence or absence of major aortopulmonary collateral arteries (MAPCA(s)), and distal distribution (pulmonary parenchymal segment arborization) abnormalities. Native pulmonary arteries may be present or absent. If MAPCAs are present this code should not be used; instead, Pulmonary atresia, VSD-MAPCA (pseudotruncus) should be used.
350	Pulmonary atresia, VSD-MAPCA	MAPCA(s) are large and distinct arteries, highly variable in number, that usually arise from the descending thoracic aorta, but uncommonly may originate from the aortic arch or the subclavian, carotid or even the coronary arteries. MAPCA(s) may be associated with present or absent native pulmonary arteries. If present, the native pulmonary

350		arteries may be hypoplastic, and either confluent or nonconfluent. Systemic pulmonary collateral arteries have been categorized into 3 types based on their site of origin and the way they connect to the pulmonary circulation: direct aortopulmonary collaterals, indirect aortopulmonary collaterals, and true bronchial arteries. Only the first two should be considered MAPCA(s). If MAPCA(s) are associated with PA-VSD or TOF, PA this code should be used.
360	MAPCA(s) (major aortopulmonary collateral[s]) (without PA-VSD)	Rarely MAPCA(s) may occur in patents who do not have PA-VSD, but have severe pulmonary stenosis. The intracardiac anatomy in patients who have MAPCA(s) without PA should be specifically coded in each case as well.
370	Ebstein's anomaly	Indicate if the patient has the diagnosis of “Ebstein's anomaly”. Ebstein's anomaly is a malformation of the tricuspid valve and right ventricle that is characterized by a spectrum of several features: (1) incomplete delamination of tricuspid valve leaflets from the myocardium of the right ventricle; (2) downward (apical) displacement of the functional annulus; (3) dilation of the "atrialized" portion of the right ventricle with variable degrees of hypertrophy and thinning of the wall; (4) redundancy, fenestrations, and tethering of the anterior leaflets; and (5) dilation of the right atrioventricular junction (the true tricuspid annulus). These anatomical and functional abnormalities cause tricuspid regurgitation (and rarely tricuspid stenosis) that results in right atrial and right ventricular dilatation and atrial and ventricular arrhythmias. With increasing degrees of anatomic severity of malformation, the fibrous transformation of leaflets from their muscular precursors remains incomplete, with the septal leaflet being most severely involved, the posterior leaflet less severely involved, and the anterior leaflet usually the least severely involved. Associated cardiac anomalies include an interatrial communication, the presence of accessory conduction pathways often associated with Wolff-Parkinson-White syndrome, and dilation of the right atrium and right ventricle in patients with severe Ebstein's anomaly. (Varying degrees of right ventricular outflow tract obstruction may be present, including pulmonary atresia in some cases. Such cases of Ebstein's anomaly with pulmonary atresia should be coded with a Primary Diagnosis of “Ebstein’s anomaly”, and a Secondary Diagnosis of “Pulmonary atresia”.) (Some patients with atrioventricular discordance and ventriculoarterial discordance in situs solitus [congenitally corrected transposition] have an Ebstein-like deformity of the left-sided morphologically tricuspid valve. The nature of the displacement of the septal and posterior leaflets is similar to that in right-sided Ebstein's anomaly in patients with atrioventricular concordance and ventriculoarterial concordance in situs solitus. These patients with “Congenitally corrected TGA” and an Ebstein-like deformity of the left-sided morphologically tricuspid valve should be coded with a Primary Diagnosis of “Congenitally corrected TGA”, and a Secondary Diagnosis of “Ebstein's anomaly”.)
380	Tricuspid regurgitation, non-	Non-Ebstein's tricuspid regurgitation may be due to

380	Ebstein's related	congenital factors (primary annular dilation, prolapse, leaflet underdevelopment, absent papillary muscle/chordae) or acquired (post cardiac surgery or secondary to rheumatic fever, endocarditis, trauma, tumor, cardiomyopathy, iatrogenic or other causes).
390	Tricuspid stenosis	Tricuspid stenosis may be due to congenital factors (valvar hypoplasia, abnormal subvalvar apparatus, double-orifice valve, parachute deformity) or acquired (post cardiac surgery or secondary to carcinoid, rheumatic fever, tumor, systemic disease, iatrogenic, or other causes).
400	Tricuspid regurgitation and tricuspid stenosis	Tricuspid regurgitation present with tricuspid stenosis may be due to congenital factors or acquired.
410	Tricuspid valve, Other	Tricuspid valve pathology not otherwise specified in diagnosis definitions 370, 380, 390 and 400.
420	Pulmonary stenosis, Valvar	Pulmonary stenosis, Valvar ranges from critical neonatal pulmonic valve stenosis with hypoplasia of the right ventricle to valvar pulmonary stenosis in the infant, child, or adult, usually better tolerated but potentially associated with infundibular stenosis. Pulmonary branch hypoplasia can be associated. Only 10% of neonates with Pulmonary stenosis, Valvar with intact ventricular septum have RV-to-coronary artery fistula(s). An RV dependent coronary artery circulation is present when coronary artery fistulas (coronary sinusoids) are associated with a proximal coronary artery stenosis; this occurs in only 2% of neonates with Pulmonary stenosis, Valvar with IVS.
430	Pulmonary artery stenosis (hypoplasia), Main (trunk)	Indicate if the patient has the diagnosis of “Pulmonary artery stenosis (hypoplasia), Main (trunk)”. “Pulmonary artery stenosis (hypoplasia), Main (trunk)” is defined as a congenital or acquired anomaly with pulmonary trunk (main pulmonary artery) narrowing or hypoplasia. The stenosis or hypoplasia may be isolated or associated with other cardiac lesions. Since the narrowing is distal to the pulmonic valve, it may also be known as supravalvar pulmonary stenosis.
440	Pulmonary artery stenosis, Branch, Central (within the hilar bifurcation)	Indicate if the patient has the diagnosis of “Pulmonary artery stenosis, Branch, Central (within the hilar bifurcation)”. “Pulmonary artery stenosis, Branch, Central (within the hilar bifurcation)” is defined as a congenital or acquired anomaly with central pulmonary artery branch (within the hilar bifurcation involving the right or left pulmonary artery, or both) narrowing or hypoplasia. The stenosis or hypoplasia may be isolated or associated with other cardiac lesions. Coarctation of the pulmonary artery is related to abnormal extension of the ductus arteriosus into a pulmonary branch, more frequently the left branch.
450	Pulmonary artery stenosis, Branch, Peripheral (at or beyond the hilar bifurcation)	Indicate if the patient has the diagnosis of “Pulmonary artery stenosis, Branch, Peripheral (at or beyond the hilar bifurcation)”. “Pulmonary artery stenosis, Branch, Peripheral (at or beyond the hilar bifurcation)” is defined as a congenital or acquired anomaly with peripheral pulmonary artery narrowing or hypoplasia (at or beyond the hilar bifurcation). The stenosis or hypoplasia may be isolated or associated with other cardiac lesions.
470	Pulmonary artery, Discontinuous	Indicate if the patient has the diagnosis of “Pulmonary

470		artery, Discontinuous". Pulmonary artery, Discontinuous" is defined as a congenital or acquired anomaly with discontinuity between the branch pulmonary arteries or between a branch pulmonary artery and the main pulmonary artery trunk.
490	Pulmonary stenosis, Subvalvar	Subvalvar (infundibular) pulmonary stenosis is a narrowing of the outflow tract of the right ventricle below the pulmonic valve. It may be due to a localized fibrous diaphragm just below the valve, an obstructing muscle bundle or to a long narrow fibromuscular channel.
500	DCRV	The double chambered right ventricle is characterized by a low infundibular (subvalvar) stenosis rather than the rare isolated infundibular stenosis that develops more superiorly in the infundibulum, and is often associated with one or several closing VSDs. In some cases, the VSD is already closed. The stenosis creates two chambers in the RV, one inferior including the inlet and trabecular portions of the RV and one superior including the infundibulum.
510	Pulmonary valve, Other	Other anomalies of the pulmonary valve may be listed here including but not restricted to absent pulmonary valve.
530	Pulmonary insufficiency	Pulmonary valve insufficiency or regurgitation may be due to congenital factors (primary annular dilation, prolapse, leaflet underdevelopment, etc.) or acquired (for example, post cardiac surgery for repair of tetralogy of Fallot, etc.).
540	Pulmonary insufficiency and pulmonary stenosis	Pulmonary valve insufficiency and pulmonary stenosis beyond the neonatal period, in infancy and childhood, may be secondary to leaflet tissue that has become thickened and myxomatous. Retraction of the commissure attachment frequently creates an associated supravalvar stenosis.
550	Aortic stenosis, Subvalvar	Subaortic obstruction can be caused by different lesions: subaortic membrane or tunnel, accessory mitral valve tissue, abnormal insertion of the mitral anterior leaflet to the ventricular septum, deviation of the outlet septum (seen in coarctation of the aorta and interrupted aortic arch), or a restrictive bulboventricular foramen in single ventricle complexes. The Shone complex consists of subvalvar aortic stenosis in association with supravalvar mitral ring, parachute mitral valve, and coarctation of aorta. Subvalvar aortic stenosis may be categorized into two types: localized subvalvar aortic stenosis, which consists of a fibrous or fibromuscular ridge, and diffuse tunnel subvalvar aortic stenosis, in which circumferential narrowing commences at the annular level and extends downward for 1-3 cm. Idiopathic hypertrophic subaortic stenosis (IHSS) is also known as hypertrophic obstructive cardiomyopathy (HOCM), and is characterized by a primary hypertrophy of the myocardium. The obstructive forms involve different degrees of dynamic subvalvar aortic obstruction from a thickened ventricular wall and anterior motion of the mitral valve. Definitive nomenclature and therapeutic options for IHSS are listed under cardiomyopathy.
2500	Aortic Stenosis, Subvalvar, Discrete	
2510	Aortic Stenosis, Subvalvar, IHSS	
2520	Aortic Stenosis, Subvalvar, Tunnel-	

2520	like	
560	Aortic stenosis, Valvar	Valvar aortic stenosis may be congenital or acquired. In its congenital form there are two types: critical (infantile), seen in the newborn in whom systemic perfusion depends on a patent ductus arteriosus, and noncritical, seen in infancy or later. Acquired valvar stenosis may be seen after as a result of rheumatic valvar disease, or from stenotic changes of an aortic valve prosthesis. Congenital valvar stenosis may result: (1) from complete fusion of commissures (acommissural) that results in a dome-shaped valve with a pinpoint opening (seen most commonly in infants with critical aortic valve stenosis); (2) from a unicommissural valve with one defined commissure and eccentric orifice (often with two raphes radiating from the ostium indicating underdeveloped commissures of a tricuspid aortic valve); (3) from a bicuspid aortic valve, with leaflets that can be equal in size or discrepant, and in left-right or anterior-posterior position; and finally (4) from a dysplastic tricuspid valve, which may have a gelatinous appearance with thick rarely equal in size leaflets, often obscuring the commissures. The dysplastic, tricuspid or bicuspid form of aortic valve deformity may not be initially obstructive but may become stenotic later in life due to leaflet thickening and calcification.
570	Aortic stenosis, Supravalvar	Congenital supravalvar aortic stenosis is described as three forms: an hourglass deformity, a fibrous membrane, and a diffuse narrowing of the ascending aorta. The disease can be inherited as an autosomal dominant trait or part of Williams-Beuren syndrome in association with mental retardation, elfin facies, failure to thrive, and occasionally infantile hypercalcemia. Supravalvar aortic stenosis may involve the coronary artery ostia, and the aortic leaflets may be tethered. The coronary arteries can become tortuous and dilated due to elevated pressures and early atherosclerosis may ensue. Supravalvar aortic stenosis may also be acquired: (1) after a neoaortic reconstruction such as arterial switch, Ross operation, or Norwood procedure; (2) at a suture line from a previous aortotomy or cannulation; and (3) from a narrowed conduit.
590	Aortic valve atresia	Aortic valve atresia will most often be coded under the Hypoplastic left heart syndrome/complex diagnostic codes since it most often occurs as part of a spectrum of cardiac malformations. However, there is a small subset of patients with aortic valve atresia who have a well-developed left ventricle and mitral valve and a large VSD (nonrestrictive or restrictive). The diagnostic code "Aortic valve atresia" enables users to report those patients with aortic valve atresia and a well-developed systemic ventricle without recourse to either a hypoplastic left heart syndrome/complex diagnosis or a single ventricle diagnosis.
600	Aortic insufficiency	Congenital aortic regurgitation/insufficiency is rare as an isolated entity. There are rare reports of congenital malformation of the aortic valve that result in aortic insufficiency shortly after birth from an absent or underdeveloped aortic valve cusp. Aortic insufficiency is more commonly seen with other associated cardiac

600		anomalies: (1) in stenotic aortic valves (commonly stenotic congenital bicuspid aortic valves) with some degree of aortic regurgitation due to aortic leaflet abnormality; (2) in association with a VSD (especially in supracristal or conal type I VSD, more commonly seen in Asian populations); (3) secondary to aortic-left ventricular tunnel; (4) secondary to tethering or retraction of aortic valve leaflets in cases of supravalvar aortic stenosis that may involve the aortic valve; and similarly (5) secondary to encroachment on an aortic cusp by a subaortic membrane; or (6) turbulence caused by a stenotic jet can create progressive aortic regurgitation. Aortic insufficiency may also result from: (1) post-procedure such as closed or open valvotomy or aortic valve repair, VSD closure, balloon valvotomy, or diagnostic catheterization; (2) in the neo-aorta post arterial switch, pulmonary autograft (Ross) procedure, homograft placement, Norwood procedure, or Damus-Kaye-Stansel procedure; (3) as a result of endocarditis secondary to perforated or prolapsed leaflets or annular dehiscence; (4) secondary to annulo-aortic ectasia with prolapsed or noncoapting leaflets; (5) secondary to trauma, blunt or penetrating; or (6) as a result of aortitis, bacterial, viral or autoimmune. Aortic regurgitation secondary to prosthetic failure should be coded first as either conduit failure or prosthetic valve failure, as applicable, and secondarily as aortic regurgitation secondary to prosthetic failure (perivalvar or due to structural failure). The underlying fundamental diagnosis that led to the initial conduit or valve prosthesis placement should also be described.
610	Aortic insufficiency and aortic stenosis	Aortic insufficiency is often seen in association with stenotic aortic valve, commonly the stenotic congenital bicuspid aortic valve. The degree of aortic regurgitation is due to the severity of the aortic leaflet abnormality.
620	Aortic valve, Other	This diagnostic subgroup may be used to delineate aortic valve cusp number (unicuspid, bicuspid, tricuspid, more than three cusps), commissural fusion (normal, partially fused, completely fused), and valve leaflet (normal, thickened, dysplastic, calcified, gelatinous), annulus (normal, hypoplastic, calcified), or sinus description (normal, dilated). Note that any extensive descriptors chosen within those made available by a vendor will be converted, at harvest, to Aortic valve, Other.
630	Sinus of Valsalva aneurysm	The sinus of Valsalva is defined as that portion of the aortic root between the aortic root annulus and the sinotubular ridge. A congenital sinus of Valsalva aneurysm is a dilation usually of a single sinus of Valsalva. These most commonly originate from the right sinus (65%-85%), less commonly from the noncoronary sinus (10%-30%), and rarely from the left sinus (<5%). A true sinus of Valsalva aneurysm presents above the aortic annulus. The hierarchical coding system distinguishes between congenital versus acquired, ruptured versus nonruptured, sinus of origin, and chamber/site of penetration (right atrium, right ventricle, left atrium, left ventricle, pulmonary artery, pericardium). A nonruptured congenital sinus of Valsalva aneurysm may vary from a mild dilation of a single aortic sinus to an extensive windsock

630		deformity. Rupture of a congenital sinus of Valsalva aneurysm into an adjacent chamber occurs most commonly between the ages of 15-30 years. Rupture may occur spontaneously, after trauma, after strenuous physical exertion, or from acute bacterial endocarditis. Congenital etiology is supported by the frequent association of sinus of Valsalva aneurysms with VSDs. Other disease processes are also associated with sinus of Valsalva aneurysm and include: syphilis, endocarditis, cystic medial necrosis, atherosclerosis, and trauma. Acquired sinus of Valsalva aneurysms more frequently involve multiple sinuses of Valsalva; when present in multiple form they are more appropriately classified as aneurysms of the aortic root.
640	LV to aorta tunnel	The aortico-left ventricular tunnel (LV-to-aorta tunnel) is an abnormal paravalvular (alongside or in the vicinity of a valve) communication between the aorta and left ventricle, commonly divided into 4 types: (1) type I, a simple tunnel with a slit-like opening at the aortic end and no aortic valve distortion; (2) type II, a large extracardiac aortic wall aneurysm of the tunnel with an oval opening at the aortic end, with or without ventricular distortion; (3) type III, intracardiac aneurysm of the septal portion of the tunnel, with or without right ventricular outflow obstruction; and (4) type IV, a combination of types II and III. Further differentiation within these types may be notation of right coronary artery arising from the wall of the tunnel. If a LV-to-aorta tunnel communicates with the right ventricle, many feel that the defect is really a ruptured sinus of Valsalva aneurysm.
650	Mitral stenosis, Supravalvar mitral ring	Supravalvar mitral ring is formed by a circumferential ridge of tissue that is attached to the anterior mitral valve leaflet (also known as the aortic leaflet) slightly below its insertion on the annulus and to the atrium slightly above the attachment of the posterior mitral valve leaflet (also known as the mural leaflet). Depending on the diameter of the ring orifice, varying degrees of obstruction exist. The underlying valve is usually abnormal and frequently stenotic or hypoplastic. Supravalvar mitral ring is commonly associated with other stenotic lesions such as parachute or hammock valve (subvalvar stenosis), papillary muscle fusion (subvalvar stenosis), and double orifice mitral valve (valvar stenosis). Differentiation from cor triatriatum focuses on the compartments created by the supravalvar ring. In cor triatriatum the posterior compartment contains the pulmonary veins; the anterior contains the left atrial appendage and the mitral valve orifice. In supravalvar mitral ring, the posterior compartment contains the pulmonary veins and the left atrial appendage; the anterior compartment contains only the mitral valve orifice. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
660	Mitral stenosis, Valvar	Valvar mitral stenosis may arise from congenital (annular and / or leaflet) or acquired causes, both surgical (after mitral valve repair or replacement or other cardiac surgery)

660		and non-surgical (post rheumatic heart disease, infective endocarditis, ischemia, myxomatous degeneration, trauma, or cardiomyopathy). Mitral valve annular hypoplasia is distinguished from severe mitral valve hypoplasia and mitral valve atresia, which are typically components of hypoplastic left heart syndrome. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
670	Mitral stenosis, Subvalvar	Congenital subvalvar mitral stenosis may be due to obstructive pathology of either the chordae tendineae and / or papillary muscles which support the valve leaflets. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
680	Mitral stenosis, Subvalvar, Parachute	In parachute mitral valve, all chordae are attached to a single papillary muscle originating from the posterior ventricular wall. When the interchordal spaces are partially obliterated valvar stenosis results. This defect also causes valvar insufficiency, most commonly due to a cleft leaflet, a poorly developed anterior leaflet, short chordae, or annular dilatation. This lesion is also part of Shone's anomaly, which consists of the parachute mitral valve, supravalvar mitral ring, subaortic stenosis, and coarctation of the aorta. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
695	Mitral stenosis	Stenotic lesions of the mitral valve not otherwise specified in the diagnosis definitions 650, 660, 670, and 680.
700	Mitral regurgitation and mitral stenosis	Mitral regurgitation and mitral stenosis may arise from congenital or acquired causes or after cardiac surgery. Additional details to aid in coding specific components of the diagnosis are available in the individual mitral stenosis or mitral regurgitation field definitions. When coding multiple mitral valve lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
710	Mitral regurgitation	Mitral regurgitation may arise from congenital (at the annular, leaflet or subvalvar level) or acquired causes both surgical (after mitral valve repair or replacement, subaortic stenosis repair, atrioventricular canal repair, cardiac transplantation, or other cardiac surgery) and non-surgical (post rheumatic heart disease, infective endocarditis, ischemia (with chordal rupture or papillary muscle infarct), myxomatous degeneration including Barlow's syndrome, trauma, or cardiomyopathy). Congenital lesions at the annular level include annular dilatation or deformation (usually deformation is consequent to associated lesions). At the valve leaflet level, mitral regurgitation may be due to a cleft, hypoplasia or agenesis of leaflet(s), excessive leaflet tissue, or a double orifice valve. At the subvalvar level, mitral regurgitation may be secondary to chordae tendineae anomalies (agenesis, rupture, elongation, or

710		shortening as in funnel valve), or to papillary muscle anomalies (hypoplasia or agenesis, shortening, elongation, single-parachute, or multiple-hammock valve). When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
720	Mitral valve, Other	Mitral valve pathology not otherwise coded in diagnosis definitions 650 through 710.
730	Hypoplastic left heart syndrome (HLHS)	Hypoplastic left heart syndrome (HLHS) is a spectrum of cardiac malformations characterized by a severe underdevelopment of the left heart-aorta complex, consisting of aortic and/or mitral valve atresia, stenosis, or hypoplasia with marked hypoplasia or absence of the left ventricle, and hypoplasia of the ascending aorta and of the aortic arch with coarctation of the aorta. Hypoplastic left heart complex is a subset of patients at the favorable end of the spectrum of HLHS characterized by hypoplasia of the structures of the left heart-aorta complex, consisting of aortic and mitral valve hypoplasia without valve stenosis or atresia, hypoplasia of the left ventricle, hypoplasia of the left ventricular outflow tract, hypoplasia of the ascending aorta and of the aortic arch, with or without coarctation of the aorta.
2080	Shone's syndrome	Shone's syndrome is a syndrome of multilevel hypoplasia and obstruction of left sided cardiovascular structures including more than one of the following lesions: (1) supravalvar ring of the left atrium, (2) a parachute deformity of the mitral valve, (3) subaortic stenosis, and (4) aortic coarctation. The syndrome is based on the original report from Shone [1] that was based on analysis of 8 autopsied cases and described the tendency of these four obstructive, or potentially obstructive, conditions to coexist. Only 2 of the 8 cases exhibited all four conditions, with the other cases exhibiting only two or three of the anomalies [2]. [1] Shone JD, Sellers RD, Anderson RG, Adams P, Lillehei CW, Edwards JE. The developmental complex of "parachute mitral valve", supravalvar ring of left atrium, subaortic stenosis, and coarctation of the aorta. Am J Cardiol 1963; 11: 714–725. [2]. Tchervenkov CI, Jacobs JP, Weinberg PM, Aiello VD, Beland MJ, Colan SD, Elliott MJ, Franklin RC, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G. The nomenclature, definition and classification of hypoplastic left heart syndrome. Cardiology in the Young, 2006; 16(4): 339–368, August 2006. Please note that the term "2080 Shone's syndrome" may be the "Fundamental Diagnosis" of a patient; however, the term "2080 Shone's syndrome" may not be the "Primary Diagnosis" of an operation. The term "2080 Shone's syndrome" may be a "Secondary Diagnosis" of an operation.
740	Cardiomyopathy (including dilated, restrictive, and hypertrophic)	Cardiomyopathy is a term applied to a wide spectrum of cardiac diseases in which the predominant feature is poor myocardial function in the absence of any anatomic abnormalities. Cardiomyopathies can be divided into three relatively easily distinguishable entities: (1) dilated,

740		characterized by ventricular dilatation and systolic dysfunction; (2) hypertrophic, characterized by physiologically inappropriate hypertrophy of the left ventricle; and (3) restrictive, characterized by diastolic dysfunction, with a presentation often identical to constrictive pericarditis. Also included in this diagnostic category are patients with a cardiomyopathy or syndrome confined to the right ventricle, for example: (1) arrhythmogenic right ventricular dysplasia; (2) Uhl's syndrome (hypoplasia of right ventricular myocardium, parchment heart); or (3) spongiform cardiomyopathy.
750	Cardiomyopathy, End-stage congenital heart disease	Myocardial abnormality in which there is systolic and/or diastolic dysfunction in the presence of structural congenital heart disease without any (or any further) surgically correctable lesions.
760	Pericardial effusion	Inflammatory stimulation of the pericardium that results in the accumulation of appreciable amounts of pericardial fluid (also known as effusive pericarditis). The effusion may be idiopathic or acquired (e.g., postoperative, infectious, uremic, neoplastic, traumatic, drug-induced).
770	Pericarditis	Inflammatory process of the pericardium that leads to either (1) effusive pericarditis with accumulation of appreciable amounts of pericardial fluid or (2) constrictive pericarditis that leads to pericardial thickening and compression of the cardiac chambers, ultimately with an associated significant reduction in cardiac function. Etiologies are varied and include idiopathic or acquired (e.g., postoperative, infectious, uremic, neoplastic, traumatic, drug-induced) pericarditis.
780	Pericardial disease, Other	A structural or functional abnormality of the visceral or parietal pericardium that may, or may not, have a significant impact on cardiac function. Included are absence or partial defects of the pericardium.
790	Single ventricle, DILV	A congenital cardiac malformation in which both atria connect to a single, morphologically left ventricle. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of

790		atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
800	Single ventricle, DIRV	A congenital cardiac malformation in which both atria connect to a single, morphologically right ventricle The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH

800		(editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
810	Single ventricle, Mitral atresia	A congenital cardiac malformation in which there is no orifice of mitral valve The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
820	Single ventricle, Tricuspid atresia	A congenital cardiac malformation in which there is no orifice of tricuspid valve. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary

820		circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
830	Single ventricle, Unbalanced AV canal	Single ventricle anomalies with a common atrioventricular (AV) valve and only one completely well developed ventricle. If the common AV valve opens predominantly into the morphologic left ventricle, the defect is termed a left ventricular (LV)–type or LV-dominant AV septal defect. If the common AV valve opens predominantly into the morphologic right ventricle, the defect is termed a right ventricular (RV)–type or RV-dominant AV septal defect. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle,

830		congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart".
840	Single ventricle, Heterotaxia syndrome	"Heterotaxia syndrome" is synonymous with "heterotaxy", "visceral heterotaxy", and "heterotaxy syndrome". Heterotaxy is defined as an abnormality where the internal thoraco-abdominal organs demonstrate abnormal arrangement across the left-right axis of the body. By convention, heterotaxy does not include patients with either the expected usual or normal arrangement of the internal organs along the left-right axis, also known as 'situs solitus', nor patients with complete mirror-imaged arrangement of the internal organs along the left-right axis also known as 'situs inversus'. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
850	Single ventricle, Other	If the single ventricle is of primitive or indeterminate type, other is chosen in coding. It is recognized that a

850		considerable variety of other structural cardiac malformations (e.g., biventricular hearts with straddling atrioventricular valves, pulmonary atresia with intact ventricular septum, some complex forms of double outlet right ventricle) may at times be best managed in a fashion similar to that which is used to treat univentricular hearts. They are not to be coded in this section of the nomenclature, but according to the underlying lesions. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
851	Single Ventricle + Total anomalous pulmonary venous connection (TAPVC)	Indicate if the patient has the diagnosis of "Single Ventricle + Total anomalous pulmonary venous connection (TAPVC)". In the event of Single Ventricle occurring in association with Total anomalous pulmonary venous connection (TAPVC), code "Single Ventricle + Total anomalous pulmonary venous connection (TAPVC)", and then use additional (secondary) diagnostic codes to describe the Single Ventricle and the Total anomalous pulmonary venous connection (TAPVC) separately to provide further documentation about the Single Ventricle and Total anomalous pulmonary venous

851		connection (TAPVC) types. {"Total anomalous pulmonary venous connection (TAPVC)" is defined as a heart where all of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium.} The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
870	Congenitally corrected TGA	Indicate if the patient has the diagnosis of “Congenitally corrected TGA”. Congenitally corrected transposition is synonymous with the terms ‘corrected transposition’ and ‘discordant atrioventricular connections with discordant ventriculo-arterial connections’, and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and

870		classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.
872	Congenitally corrected TGA, IVS	Indicate if the patient has the diagnosis of "Congenitally corrected TGA, IVS". "Congenitally corrected TGA, IVS" is "Congenitally corrected transposition with an intact ventricular septum", in other words, "Congenitally corrected transposition with no VSD". (Congenitally corrected transposition is synonymous with the terms 'corrected transposition' and 'discordant atrioventricular connections with discordant ventriculo-arterial connections', and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)
874	Congenitally corrected TGA, IVS-LVOTO	Indicate if the patient has the diagnosis of "Congenitally corrected TGA, IVS-LVOTO". "Congenitally corrected TGA, IVS-LVOTO" is "Congenitally corrected transposition with an intact ventricular septum and left ventricular outflow tract obstruction", in other words, "Congenitally corrected transposition with left ventricular outflow tract obstruction and no VSD". (Congenitally corrected transposition is synonymous with the terms 'corrected transposition' and 'discordant atrioventricular connections with discordant ventriculo-arterial connections', and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)

876	Congenitally corrected TGA, VSD	Indicate if the patient has the diagnosis of “Congenitally corrected TGA, VSD”. “Congenitally corrected TGA, VSD” is “Congenitally corrected transposition with a VSD”. (Congenitally corrected transposition is synonymous with the terms ‘corrected transposition’ and ‘discordant atrioventricular connections with discordant ventriculo-arterial connections’, and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)
878	Congenitally corrected TGA, VSD-LVOTO	Indicate if the patient has the diagnosis of “Congenitally corrected TGA, VSD-LVOTO”. “Congenitally corrected TGA, VSD-LVOTO” is “Congenitally corrected transposition with a VSD and left ventricular outflow tract obstruction”. (Congenitally corrected transposition is synonymous with the terms ‘corrected transposition’ and ‘discordant atrioventricular connections with discordant ventriculo-arterial connections’, and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)
880	TGA, IVS	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with an intact ventricular septum. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).

890	TGA, IVS-LVOTO	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with an intact ventricular septum and associated left ventricular obstruction. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
900	TGA, VSD	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with one or more ventricular septal defects. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
910	TGA, VSD-LVOTO	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with one or more ventricular septal defects and left ventricular outflow tract obstruction. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
930	DORV, VSD type	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In double outlet right ventricle, VSD type, there is an associated subaortic or doubly-committed VSD and no pulmonary outflow tract obstruction. Subaortic VSD's are located beneath the aortic valve. Doubly-committed VSD's lie beneath the leaflets of the aortic and pulmonary valves (juxtaarterial). In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
940	DORV, TOF type	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or

940		predominantly from the right ventricle. In double outlet right ventricle, TOF type, there is an associated subaortic or doubly-committed VSD and pulmonary outflow tract obstruction. Subaortic VSD's are located beneath the aortic valve. Doubly-committed VSD's lie beneath the leaflets of the aortic and pulmonary valves (juxtaarterial). DORV can occur in association with pulmonary atresia, keeping in mind in coding that in the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles (in this situation DORV is coded as a primary diagnosis). Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate Single ventricle listing.
950	DORV, TGA type	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In double outlet right ventricle, TGA type, there is an associated subpulmonary VSD. Most frequently, there is no pulmonary outflow tract obstruction (Taussig-Bing heart). The aorta is usually to the right and slightly anterior to or side-by-side with the pulmonary artery. Associated aortic outflow tract stenosis (subaortic, aortic arch obstruction) is commonly associated with the Taussig-Bing heart and if present should be coded as a secondary diagnosis. Rarely, there is associated pulmonary outflow tract obstruction. In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
960	DORV, Remote VSD (uncommitted VSD)	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In double outlet right ventricle, Remote VSD type, there is a remote or noncommitted VSD. The VSD is far removed from both the aortic and pulmonary valves, usually within the inlet septum. Many of these VSD's are in hearts with DORV and common atrioventricular canal/septal defect. In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
2030	DORV + AVSD (AV Canal)	Indicate if the patient has the diagnosis of “DORV + AVSD (AV Canal)”. In the event of DORV occurring in association with AVSD (AV Canal), code “DORV + AVSD (AV Canal)”, and

2030		then use additional (secondary) diagnostic codes to describe the DORV and the AVSD (AV Canal) separately to provide further documentation about the DORV and AVSD (AV Canal) types. {"DORV" is "Double outlet right ventricle" and is defined as a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle.} In this case, the DORV exists in combination with an atrioventricular septal defect and common atrioventricular junction guarded by a common atrioventricular valve.
975	DORV, IVS	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In the rare case of double outlet right ventricle with IVS the ventricular septum is intact. In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connections with DORV are to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
980	DOLV	Double outlet left ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the left ventricle. In the nomenclature developed for DOLV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DOLV is to be coded under congenitally corrected TGA. DOLV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
990	Coarctation of aorta	Indicate if the patient has the diagnosis of "Coarctation of aorta". A "Coarctation of the aorta" generally indicates a narrowing of the descending thoracic aorta just distal to the left subclavian artery. However, the term may also be accurately used to refer to a region of narrowing anywhere in the thoracic or abdominal aorta.
1000	Aortic arch hypoplasia	Hypoplasia of the aortic arch is hypoplasia of the proximal or distal transverse arch or the aortic isthmus. The isthmus (arch between the left subclavian and insertion of the patent ductus arteriosus / ligamentum arteriosum) is hypoplastic if its diameter is less than 40% of the diameter of the ascending aorta. The proximal transverse arch (arch between the innominate and left carotid arteries) and distal transverse arch (arch between the left carotid and left subclavian arteries) are hypoplastic if their diameters are less than 60% and 50%, respectively, of the diameter of the ascending aorta.
92	VSD + Aortic arch hypoplasia	A ventricular septal defect, any type, associated with hypoplasia of the aortic arch. (See diagnosis definition 1000 for a definition of hypoplasia of the aortic arch.)
94	VSD + Coarctation of aorta	Indicate if the patient has the diagnosis of "VSD +

94		Coarctation of aorta". In the event of a VSD occurring in association with Coarctation of aorta, code "VSD + Coarctation of aorta", and then use additional (secondary) diagnostic codes to describe the VSD and the Coarctation of aorta separately to provide further documentation about the individual VSD and Coarctation of aorta types. {A "VSD" is a "Ventricular Septal Defect" and is also known as an "Interventricular communication". A VSD is defined as "a hole between the ventricular chambers or their remnants". (The VSD is defined on the basis of its margins as seen from the aspect of the morphologically right ventricle. In the setting of double outlet right ventricle, the defect provides the outflow from the morphologically left ventricle. In univentricular atrioventricular connections with functionally single left ventricle with an outflow chamber, the communication is referred to by some as a bulboventricular foramen.)} {A "Coarctation of the aorta" generally indicates a narrowing of the descending thoracic aorta just distal to the left subclavian artery. However, the term may also be accurately used to refer to a region of narrowing anywhere in the thoracic or abdominal aorta.}
1010	Coronary artery anomaly, Anomalous aortic origin of coronary artery (AAOCA)	Anomalous aortic origins of the coronary arteries include a spectrum of anatomic variations of the normal coronary artery origins. Coronary artery anomalies of aortic origin to be coded under this diagnostic field include: anomalies of take-off (high take-off), origin (sinus), branching, and number. An anomalous course of the coronary artery vessels is also significant, particularly those coronary arteries that arise or course between the great vessels.
1020	Coronary artery anomaly, Anomalous pulmonary origin (includes ALCAPA)	In patients with anomalous pulmonary origin of the coronary artery, the coronary artery (most commonly the left coronary artery) arises from the pulmonary artery rather than from the aorta. Rarely, the right coronary artery, the circumflex, or both coronary arteries may arise from the pulmonary artery.
1030	Coronary artery anomaly, Fistula	The most common of coronary artery anomalies, a coronary arteriovenous fistula is a communication between a coronary artery and either a chamber of the heart (coronary-cameral fistula) or any segment of the systemic or pulmonary circulation (coronary arteriovenous fistula). They may be congenital or acquired (traumatic, infectious, iatrogenic) in origin, and are mostly commonly seen singly, but occasionally multiple fistulas are present. Nomenclature schemes have been developed that further categorize the fistulas by vessel of origin and chamber of termination, and one angiographic classification scheme by Sakakibara has surgical implications. Coronary artery fistulas can be associated with other congenital heart anomalies such as tetralogy of Fallot, atrial septal defect, ventricular septal defect, and pulmonary atresia with intact ventricular septum, among others. The major cardiac defect should be listed as the primary diagnosis and the coronary artery fistula should be as an additional secondary diagnoses.
1040	Coronary artery anomaly, Aneurysm	Coronary artery aneurysms are defined as dilations of a coronary vessel 1.5 times the adjacent normal coronaries. There are two forms, saccular and fusiform (most common),

1040		and both may be single or multiple. These aneurysms may be congenital or acquired (atherosclerotic, Kawasaki, systemic diseases other than Kawasaki, iatrogenic, infectious, or traumatic) in origin.
2420	Coronary artery anomaly, ostial atresia	
1050	Coronary artery anomaly, Other	Coronary artery anomalies which may fall within this category include coronary artery bridging and coronary artery stenosis, as well as secondary coronary artery variations seen in congenital heart defects such as tetralogy of Fallot, transposition of the great arteries, and truncus arteriosus (with the exception of variations that can be addressed by a more specific coronary artery anomaly code).
1070	Interrupted aortic arch	Indicate if the patient has the diagnosis of “Interrupted aortic arch”. Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.
2020	Interrupted aortic arch + VSD	Indicate if the patient has the diagnosis of “Interrupted aortic arch + VSD”. In the event of interrupted aortic arch occurring in association with VSD, code “Interrupted aortic arch + VSD”, and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and the VSD separately to provide further documentation about the individual interrupted aortic arch and VSD types. {Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.} {A "VSD" is a "Ventricular Septal Defect" and is also known as an "Interventricular communication". A VSD is defined as "a hole between the ventricular chambers or their remnants". (The VSD is defined on the basis of its margins as seen from the aspect of the morphologically right ventricle. In the setting of double outlet right ventricle, the defect provides the outflow from the morphologically left ventricle. In univentricular atrioventricular connections with functionally single left ventricle with an outflow chamber, the communication is referred to by some as a bulboventricular foramen.)}
2000	Interrupted aortic arch + AP window (aortopulmonary window)	Indicate if the patient has the diagnosis of “Interrupted aortic arch + AP window (aortopulmonary window)”. In the event of interrupted aortic arch occurring in association with AP window, code “Interrupted aortic arch + AP window (aortopulmonary window)”, and then use additional (secondary) diagnostic codes to describe the interrupted

2000		aortic arch and the AP window separately to provide further documentation about the individual interrupted aortic arch and AP window types. {Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.} {An "AP window (aortopulmonary window)" is defined as a defect with side-to-side continuity of the lumens of the aorta and pulmonary arterial tree, which is distinguished from common arterial trunk (truncus arteriosus) by the presence of two arterial valves or their atretic remnants. (In other words, an aortopulmonary window is a communication between the main pulmonary artery and ascending aorta in the presence of two separate semilunar [pulmonary and aortic] valves. The presence of two separate semilunar valves distinguishes AP window from truncus arteriosus. Type 1 proximal defect: AP window located just above the sinus of Valsalva, a few millimeters above the semilunar valves, with a superior rim but little inferior rim separating the AP window from the semilunar valves. Type 2 distal defect: AP window located in the uppermost portion of the ascending aorta, with a well-formed inferior rim but little superior rim. Type 3 total defect: AP window involving the majority of the ascending aorta, with little superior and inferior rims. The intermediate type of AP window is similar to the total defect but with adequate superior and inferior rims. In the event of AP window occurring in association with interrupted aortic arch, code "Interrupted aortic arch + AP window (aortopulmonary window)", and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and AP window separately to provide further documentation about the individual interrupted arch and AP window types.)}
1080	Patent ductus arteriosus	Indicate if the patient has the diagnosis of "Patent ductus arteriosus". The ductus arteriosus (arterial duct) is an essential feature of fetal circulation, connecting the main pulmonary trunk with the descending aorta, distal to the origin of the left subclavian artery. In most patients it is on the left side. If a right aortic arch is present, it may be on the right or the left; very rarely it is bilateral. When luminal patency of the duct persists post-natally, it is referred to as patent ductus arteriosus (patent arterial duct). The length and diameter may vary considerably from case to case. The media of the ductus consists mainly of smooth muscle that is arranged spirally, and the intima is much thicker than that of the aorta. (A patent ductus arteriosus is a vascular arterial connection between the thoracic aorta and the pulmonary artery. Most commonly a PDA has its origin from the descending thoracic aorta, just distal and opposite the origin of the left subclavian artery. The insertion of the ductus is most commonly into the very proximal left

1080		pulmonary artery at its junction with the main pulmonary artery. Origination and insertion sites can be variable, however.)
1090	Vascular ring	The term vascular ring refers to a group of congenital vascular anomalies that encircle and compress the esophagus and trachea. The compression may be from a complete anatomic ring (double aortic arch or right aortic arch with a left ligamentum) or from a compressive effect of an aberrant vessel (innominate artery compression syndrome).
1100	Pulmonary artery sling	In pulmonary artery sling, the left pulmonary artery originates from the right pulmonary artery and courses posteriorly between the trachea and esophagus in its route to the left lung hilum, causing a sling-like compression of the trachea.
1110	Aortic aneurysm (including pseudoaneurysm)	An aneurysm of the aorta is defined as a localized dilation or enlargement of the aorta at any site along its length (from aortic annulus to aortoiliac bifurcation). A true aortic aneurysm involves all layers of the aortic wall. A false aortic aneurysm (pseudoaneurysm) is defined as a dilated segment of the aorta not containing all layers of the aortic wall and may include postoperative or post-procedure false aneurysms at anastomotic sites, traumatic aortic injuries or transections, and infectious processes leading to a contained rupture.
1120	Aortic dissection	Aortic dissection is a separation of the layers of the aortic wall. Extension of the plane of the dissection may progress to free rupture into the pericardium, mediastinum, or pleural space if not contained by the outer layers of the media and adventitia. Dissections may be classified as acute or chronic (if they have been present for more than 14 days).
1130	Lung disease, Benign	Lung disease arising from any etiology (congenital or acquired) which does not result in death or lung or heart-lung transplant; examples might be non-life threatening asthma or emphysema, benign cysts.
1140	Lung disease, Malignant	Lung disease arising from any etiology (congenital or acquired, including pulmonary parenchymal disease, pulmonary vascular disease, congenital heart disease, neoplasm, etc.) which may result in death or lung or heart-lung transplant.
1160	Tracheal stenosis	Tracheal stenosis is a reduction in the anatomic luminal diameter of the trachea by more than 50% of the remaining trachea. This stenosis may be congenital or acquired (as in post-intubation or traumatic tracheal stenosis).
2430	Tracheomalacia	
1170	Airway disease	Included in this diagnostic category would be airway pathology not included under the definition of tracheal stenosis such as tracheomalacia, bronchotracheomalacia, tracheal right upper lobe, bronchomalacia, subglottic stenosis, bronchial stenosis, etc.
1430	Pleural disease, Benign	Benign diseases of the mediastinal or visceral pleura.
1440	Pleural disease, Malignant	Malignant diseases of the mediastinal or visceral pleura.

1450	Pneumothorax	A collection of air or gas in the pleural space.
1460	Pleural effusion	Abnormal accumulation of fluid in the pleural space.
1470	Chylothorax	The presence of lymphatic fluid in the pleural space secondary to a leak from the thoracic duct or its branches. Chylothorax is a specific type of pleural effusion.
1480	Empyema	A collection of purulent material in the pleural space, usually secondary to an infection.
1490	Esophageal disease, Benign	Any benign disease of the esophagus.
1500	Esophageal disease, Malignant	Any malignant disease of the esophagus.
1505	Mediastinal disease	Any disease of the mediastinum awaiting final benign/malignant pathology determination.
1510	Mediastinal disease, Benign	Any benign disease of the mediastinum.
1520	Mediastinal disease, Malignant	Any malignant disease of the mediastinum.
1540	Diaphragm paralysis	Paralysis of diaphragm, unilateral or bilateral.
1550	Diaphragm disease, Other	Any disease of the diaphragm other than paralysis.
2160	Rib tumor, Benign	Non-cancerous tumor of rib(s) (e.g., fibrous dysplasia)
2170	Rib tumor, Malignant	Cancerous tumor of rib(s)- primary (e.g., osteosarcoma, chondrosarcoma)
2180	Rib tumor, Metastatic	Cancerous tumor metastasized to rib(s)from a different primary location
2190	Sternal tumor, Benign	Non-cancerous tumor of sternum (e.g., fibrous dysplasia)
2200	Sternal tumor, Malignant	Cancerous tumor of sternum - primary (e.g., osteosarcoma, chondrosarcoma)
2210	Sternal tumor, Metastatic	Cancerous tumor metastasized to sternum from a different primary location
2220	Pectus carinatum	Pectus carinatum represents a spectrum of protrusion abnormalities of the anterior chest wall. Severe deformity may result in dyspnea and decreased endurance. Some patients develop rigidity of the chest wall with decreased lung compliance, progressive emphysema, and increased frequency of respiratory tract infections.
2230	Pectus excavatum	Pectus excavatum is a congenital chest wall deformity in which several ribs and the sternum grow abnormally, producing a concave, or caved-in, appearance in the anterior chest wall. Pectus excavatum is the most common type of congenital chest wall abnormality. It occurs in an estimated 1 in 300-400 births, with male predominance (male-to-female ratio of 3:1). The condition is typically noticed at birth, and more than 90% of cases are diagnosed within the first year of life. Worsening of the chest's appearance and the onset of respiratory symptoms are usually reported during rapid bone growth in the early teenage years.
2240	Thoracic outlet syndrome	Thoracic outlet syndrome (TOS) is caused by compression at the superior thoracic outlet wherein excess pressure is placed on a neurovascular bundle passing between the anterior scalene and middle scalene muscles. It can affect the brachial plexus (nerves that pass into the arm from the neck), the subclavian artery, and - rarely - the vein, which does not normally pass through the scalene hiatus. TOS may

2240		occur due to a positional cause - for example, by abnormal compression from the clavicle (collarbone) and shoulder girdle on arm movement. There are also several static forms, caused by abnormalities, enlargement, or spasm of the various muscles surrounding the arteries, veins, and/or brachial plexus, a fixation of a first rib, or a cervical rib. The most common causes of thoracic outlet syndrome include physical trauma from a car accident, repetitive injuries from a job such as frequent non-ergonomic use of a keyboard, sports-related activities, anatomical defects such as having an extra rib, and pregnancy.
1180	Arrhythmia	Any cardiac rhythm other than normal sinus rhythm.
2440	Arrhythmia, Atrial, Atrial fibrillation	
2450	Arrhythmia, Atrial, Atrial flutter	
2460	Arrhythmia, Atrial, Other	
2050	Arrhythmia, Junctional	Indicate if the patient has the diagnosis of ‘‘Arrhythmia, Junctional’’. “Arrhythmias arising from the atrioventricular junction; may be bradycardia, tachycardia, premature beats, or escape rhythm [1]. [1]. Jacobs JP. (Editor). 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, Volume 18, Supplement S2, pages 1 –530, December 9, 2008, page 379.
2060	Arrhythmia, Ventricular	Indicate if the patient has the diagnosis of ‘‘Arrhythmia, Ventricular’’. “Arrhythmia, Ventricular’’ ROOT Definition = Abnormal rhythm originating from the ventricles [1]. [1]. Jacobs JP. (Editor). 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, Volume 18, Supplement S2, pages 1 –530, December 9, 2008, page 393.
1185	Arrhythmia, Heart block	Atrioventricular block may be congenital or acquired, and may be of varying degree (first, second, or third degree).
1190	Arrhythmia, Heart block, Acquired	Atrioventricular block, when acquired, may be post-surgical, or secondary to myocarditis or other etiologies; the block may be first, second or third degree.
1200	Arrhythmia, Heart block, Congenital	Atrioventricular block, when congenital, may be first, second or third degree block.
2530	Short QT syndrome	
2540	Long QT Syndrome (Ward Romano syndrome)	
2550	Wolff-Parkinson-White syndrome (WPW syndrome)	
1250	Aneurysm, Ventricular, Right (including pseudoaneurysm)	An aneurysm of the right ventricle is defined as a localized dilation or enlargement of the right ventricular wall.
1260	Aneurysm, Ventricular, Left (including pseudoaneurysm)	An aneurysm of the left ventricle is defined as a localized dilation or enlargement of the left ventricular wall.

1270	Aneurysm, Pulmonary artery	An aneurysm of the pulmonary artery is defined as a localized dilation or enlargement of the pulmonary artery trunk and its central branches (right and left pulmonary artery).
1280	Aneurysm, Other	A localized dilation or enlargement of a cardiac vessel or chamber not coded in specific fields available for aortic aneurysm, sinus of Valsalva aneurysm, coronary artery aneurysm, right ventricular aneurysm, left ventricular aneurysm, or pulmonary artery aneurysm.
1290	Hypoplastic RV	Small size of the right ventricle. This morphological abnormality usually is an integral part of other congenital cardiac anomalies and, therefore, frequently does not need to be coded separately. It should, however, be coded as secondary to an accompanying congenital cardiac anomaly if the right ventricular hypoplasia is not considered an integral and understood part of the primary congenital cardiac diagnosis. It would rarely be coded as a primary and/or isolated diagnosis.
1300	Hypoplastic LV	Small size of the left ventricle. This morphological abnormality usually is an integral part of other congenital cardiac anomalies and, therefore, frequently does not need to be coded separately. It should, however, be coded as secondary to an accompanying congenital cardiac anomaly if the left ventricular hypoplasia is not considered an integral and understood part of the primary congenital cardiac diagnosis. It would rarely be coded as a primary and/or isolated diagnosis.
1310	Mediastinitis	Inflammation/infection of the mediastinum, the cavity between the lungs which holds the heart, great vessels, trachea, esophagus, thymus, and connective tissues. In the United States mediastinitis occurs most commonly following chest surgery.
1320	Endocarditis	An infection of the endocardial surface of the heart, which may involve one or more heart valves (native or prosthetic) or septal defects or prosthetic patch material placed at previous surgery.
1325	Rheumatic heart disease	Heart disease, usually valvar (e.g., mitral or aortic), following an infection with group A streptococci
1340	Myocardial infarction	A myocardial infarction is the development of myocardial necrosis caused by a critical imbalance between the oxygen supply and demand of the myocardium. While a myocardial infarction may be caused by any process that causes this imbalance it most commonly results from plaque rupture with thrombus formation in a coronary vessel, resulting in an acute reduction of blood supply to a portion of the myocardium. Myocardial infarction is a usual accompaniment of anomalous left coronary artery from the pulmonary artery (ALCAPA).
1350	Cardiac tumor	An abnormal growth of tissue in or on the heart, demonstrating partial or complete lack of structural organization, and no functional coordination with normal cardiac tissue. Commonly, a mass is recognized which is distinct from the normal structural components of the heart. A primary cardiac tumor is one that arises directly

1350		from tissues of the heart, (e.g., myxoma, fibroelastoma, rhabdomyoma, fibroma, lipoma, pheochromocytoma, teratoma, hemangioma, mesothesioloma, sarcoma). A secondary cardiac tumor is one that arises from tissues distant from the heart, with subsequent spread to the otherwise normal tissues of the heart, (e.g., renal cell tumor with caval extension from the kidney to the level of the heart or tumor with extension from other organs or areas of the body (hepatic, adrenal, uterine, infradiaphragmatic)). N.B., in the nomenclature system developed, cardiac thrombus and cardiac vegetation are categorized as primary cardiac tumors.
1360	Pulmonary AV fistula	An abnormal intrapulmonary connection (fistula) between an artery and vein that occurs in the blood vessels of the lungs. Pulmonary AV fistulas may be seen in association with congenital heart defects; the associated cardiac defect should be coded as well.
1370	Pulmonary embolism	A pulmonary embolus is a blockage of an artery in the lungs by fat, air, clumped tumor cells, or a blood clot.
1385	Pulmonary vascular obstructive disease	Pulmonary vascular obstructive disease (PVOD) other than those specifically defined elsewhere (Eisenmenger's pulmonary vascular obstructive disease, primary pulmonary hypertension, persistent fetal circulation). The spectrum includes PVOD arising from (1) pulmonary arterial hypertension or (2) pulmonary venous hypertension or (3) portal hypertension, or (4) collage vascular disease, or (5) drug or toxin induced, or (6) diseases of the respiratory system, or (7) chronic thromboembolic disease, among others.
1390	Pulmonary vascular obstructive disease (Eisenmenger’s)	"Eisenmenger syndrome" could briefly be described as "Acquired severe pulmonary vascular disease associated with congenital heart disease (Eisenmenger)". Eisenmenger syndrome is an acquired condition. In Eisenmenger-type pulmonary vascular obstructive disease, long-term left-to-right shunting (e.g., through a ventricular or atrial septal defect, patent ductus arteriosus, aortopulmonary window) can lead to chronic pulmonary hypertension with resultant pathological changes in the pulmonary vessels. The vessels become thick-walled, stiff, noncompliant, and may be obstructed. In Eisenmenger syndrome, the long-term left-to-right shunting will reverse and become right to left. Please note that the specific heart defect should be coded as a secondary diagnosis.
1400	Primary pulmonary hypertension	Primary pulmonary hypertension is a rare disease characterized by elevated pulmonary artery hypertension with no apparent cause. Two forms are included in the nomenclature, a sporadic form and a familial form which can be linked to the BMPR-II gene.
1410	Persistent fetal circulation	Persistence of the blood flow pattern seen in fetal life, in which high pulmonary vascular resistance in the lungs results in decreased blood flow to the lungs. Normally, after birth pulmonary pressure falls with a fall in pulmonary vascular resistance and there is increased perfusion of the lungs. Persistent fetal circulation, also known as persistent pulmonary hypertension of the newborn, can be related to

	1410		lung or diaphragm malformations or lung immaturity.
	1420	Meconium aspiration	Aspiration of amniotic fluid stained with meconium before, during, or after birth can lead to pulmonary sequelae including (1) pneumothorax, (2) pneumomediastinum, (3) pneumopericardium, (4) lung infection, and (5) meconium aspiration syndrome (MAS) with persistent pulmonary hypertension.
	2250	Kawasaki disease	Kawasaki disease, also known as Kawasaki syndrome, is an acute febrile illness of unknown etiology that primarily affects children younger than 5 years of age. it was first described in Japan in 1967, and the first cases outside of Japan were reported in Hawaii in 1976. It is characterized by fever, rash, swelling of the hands and feet, irritation and redness of the whites of the eyes, swollen lymph glands in the neck, and irritation and inflammation of the mouth, lips, and throat. Serious complications of Kawasaki disease include coronary artery dilatations and aneurysms, and Kawasaki disease is a leading cause of acquired heart disease in children in the United States. The standard treatment with intravenous immunoglobulin and aspirin substantially decreases the development of coronary artery abnormalities.
	1560	Cardiac, Other	Any cardiac diagnosis not specifically delineated in other diagnostic codes.
	1570	Thoracic and/or mediastinal, Other	Any thoracic and/or mediastinal disease not specifically delineated in other diagnostic codes.
	1580	Peripheral vascular, Other	Any peripheral vascular disease (congenital or acquired) or injury (from trauma or iatrogenic); vessels involved may include, but are not limited to femoral artery, femoral vein, iliac artery, brachial artery, etc.
	2400	Trauma, Blunt	Injury (ies) sustained from blunt force, caused by motor vehicle accidents, falls, blows or crush injuries
	2410	Trauma, Penetrating	Injury (ies) sustained as a result of sharp force, including cutting or piercing instruments or objects, bites, or firearm injuries from projectiles.
	2560	Cardio-respiratory failure not secondary to known structural heart disease	
	2570	Myocarditis	
	2580	Common AV valve insufficiency	
	2590	Protein-losing enteropathy	
	2600	Plastic bronchitis	
	7000	Normal heart	Normal heart.
	7777	Miscellaneous, Other	Any disease (congenital or acquired) not specifically delineated in other diagnostic codes.
Retired	2040	Arrhythmia, Atrial	Indicate if the patient has the diagnosis of ‘‘Arrhythmia, Atrial’’. ‘‘Arrhythmia, Atrial’’ ROOT Definition = Non-sinus atrial rhythm with or without atrioventricular conduction. [1]. [1]. Jacobs JP. (Editor). 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients

Retired	2040	with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, Volume 18, Supplement S2, pages 1 –530, December 9, 2008, page 373.

Extracardiac anomaly

Seq Num: 760

Required for case closure: Yes

Registry field: [ExtracardAnomaly].[ExtracardAnom]

Shared with PAC3

Description: Select all of the noncardiac congenital anatomic abnormalities identified in the patient. If none, select "None."

Values	*Code*	*Text*	
	5	None	No known major noncardiac abnormality
	80	Major abnormality of head, Choanal atresia	A congenital anomaly in which a bony or membranous occlusion blocks the passageway between the nose and pharynx. The condition, caused by the failure of the nasopharyngeal septum to rupture during embryonic development, may result in ventilation problems in the neonate and requires surgical correction.
	90	Major abnormality of head, Cleft lip	A congenital anomaly consisting of one or more clefts in the upper lip that result from the failure of the maxillary and median nasal processes to close during embryonic development. Treatment is surgical repair in infancy.
	100	Major abnormality of head, Cleft palate	A congenital fissure in the roof of the mouth, resulting from incomplete fusion of the palate during embryonic development. It may involve only the uvula or extend through the entire palate.
	440	Major abnormality of head, Craniosynostosis	
	450	Major abnormality of head, Macrocephaly	Macrocephaly is defined as a head circumference which is greater than 2 standard deviations larger than the average for a given age and sex. It refers to an abnormally large head inclusive of the scalp, cranial bone and intracranial contents. Macrocephaly may be due to megalencephaly (true enlargement of the brain) or due to other conditions such as hydrocephalus or cranial thickening.
	460	Major abnormality of head, Microcephaly	Microcephaly is defined as smaller than normal circumference of the head because the cerebral cortex has not developed properly or has stopped growing. Microcephaly can be present at birth or may develop in the first few years of life.
	470	Major abnormality of head, Micrognathia	
	120	Major abnormality of brain, Hydrocephalus	Hydrocephalus is excessive CSF accumulation in the brain creating potentially harmful pressure. It may be congenital or acquired. Congenital hydrocephalus is present at birth and may be caused by either events or influences that occur during fetal development, or genetic abnormalities. Acquired hydrocephalus develops at the time of birth or at some point afterward. This type of hydrocephalus can affect individuals of all ages and may be caused by injury or disease.
	480	Major abnormality of brain, Tuberous sclerosis	

160	Major abnormality of spinal cord, Myelomeningocele	Developmental defect of the central nervous systemprotrude through a gap in the vertebral column; frequently accompanied by hydrocephalus and mental retardation. A hernial sac containing a portion of the spinal cord, its meninges, and cerebrospinal fluid protrudes through a congenital cleft in the vertebral column. The defect is covered by a thin membrane or skin.
170	Major abnormality of spinal cord, Spina bifida	Characterized by defective closure of the vertebral canal with herniation of the spinal cord and/or meninges. May cause skull enlargement due to an accumulation of cerebrospinal fluid. In its most severe form, termed spinal rachischisis, the entire spinal canal is open, exposing the spinal cord and nerves. More commonly, the abnormality appears as a localized mass on the back that is covered by skin or by the meninges.
660	Major abnormality of spinal cord, Tethered cord	
190	Major abnormality of spine, Scoliosis	Scoliosis is a lateral (side-to-side) curve in the spine, usually combined with a rotation of the vertebrae. "Most commonly presents as idiopathic (90%) but can present as a congenital or acquired defect.
640	Major abnormality of vertebra, Hemi-vertebrae	
650	Major abnormality of vertebra, Butterfly vertebrae	
490	Major abnormality of larynx - trachea - or bronchus, Laryngeal cleft	
210	Major abnormality of larynx - trachea - or bronchus, Laryngomalacia	Abnormal laxity of the laryngeal support cartilage resulting in excessive inward collapse and collapse of the lumen with inspiration during spontaneous ventilation. Characterized by inspiratory stridor.
220	Major abnormality of larynx - trachea - or bronchus, Congenital tracheal stenosis	Primary Tracheal narrowing at any level between the larynx and carina with significantly smaller than expected luminal diameter (not secondary to trauma or prolonged intubation). Frequently related to complete cartilagenous tracheal rings.
230	Major abnormality of larynx - trachea - or bronchus, Tracheomalacia	Abnormal laxity of the tracheal supporting structures resulting in inward collapse of the lumen during expiration during spontaneous ventilation. Characterized by expiratory stridor. May extend down into bronchi (tracheobronchial malacia).
70	Major abnormality of larynx - trachea - or bronchus, Tracheoesophageal fistula (TEF)	Presence of any type of patent communication below the larynx connecting the tracheo-bronchial tree to the esophagus. May be associated with other anomalies, including VATER, VACTERL and tracheal clefts. Typically congenital, but may occur due to trauma or pressure necrosis.
240	Major abnormality of larynx - trachea - or bronchus, Bronchomalacia	A deficiency in the cartilaginous wall of the bronchus that may lead to atelectasis or obstructive emphysema.
500	Major abnormality of chest wall, Pectus carinatum	

510	Major abnormality of chest wall, Pectus excavatum	
520	Major abnormality of lung, Alveolar capillary dysplasia	
260	Major abnormality of lung, Congenital lobar emphysema (CLE)	A developmental anomaly of the lower respiratory tract characterized by isolated hyperinflation of a lobe in the absence of extrinsic bronchial obstruction
270	Major abnormality of lung, Cystic congenital adenomatous malformation of the lung (CAM)	Cystic congenital adenomatous malformation of the lung (CAM): A spectrum of cystic and solid lesions of the lung that result from abnormal embryogenesis and typically present with symptoms of respiratory distress in newborns and infants.
280	Major abnormality of lung, Cystic fibrosis	Cystic fibrosis (also known as CF or mucoviscidosis) is an autosomal recessive genetic disorder affecting most critically the lungs, and also the pancreas, liver, and intestine. It is characterized by abnormal transport of chloride and sodium across an epithelium, leading to thick, viscous secretions
530	Major abnormality of lung, Hypoplastic lung	
290	Major abnormality of lung, Pulmonary lymphangiectasia	Pulmonary lymphangiectasia (PL) is a rare developmental disorder involving the lung characterized by pulmonary subpleural, interlobar, perivascular and peribronchial lymphatic dilatation. PL presents at birth with severe respiratory distress, tachypnea and cyanosis, with a very high mortality rate at or within a few hours of birth. Secondary PL may be caused by a cardiac lesion.
20	Major abnormality of diaphragm, Congenital diaphragmatic hernia (CDH), Bochdalek hernia	A developmental defect of the diaphragm that allows abdominal viscera to herniate into the chest. The volume of herniated contents may be small or large enough to contain most of the gut, spleen, or liver.
30	Major abnormality of abdominal wall, Gastroschisis	A congenital defect characterized by a defect in the anterior abdominal wall through which the intestines protrude. There is no sac covering the intestines. The defect is usually located to the right of the umbilicus.
60	Major abnormality of abdominal wall, Omphalocele	A defect in the medial anterior abdominal wall through which intraabdominal contents are extruded. The defect is covered by amnion and peritoneum and usually occurs at the base of the umbilical cord. The abdominal herniation usually includes small bowel and may include large bowel and/or liver.
540	Major abnormality of gastrointestinal system, Esophageal atresia	
550	Major abnormality of gastrointestinal system, Pyloric stenosis	
310	Major abnormality of gastrointestinal system, Biliary atresia	Biliary atresia is characterized by absence or discontinuity of the extrahepatic biliary system, resulting in obstruction to bile flow.
320	Major abnormality of gastrointestinal system, Duodenal	Congenital absence or closure of a portion of the duodenum.

320	atresia	
330	Major abnormality of gastrointestinal system, Duodenal stenosis	Stricture or narrowing of a portion of the duodenum.
340	Major abnormality of gastrointestinal system, Jejunal atresia	The congenital absence or closure of the middle section of the small intestine.
350	Major abnormality of gastrointestinal system, Jejunal stenosis	A constriction or narrowing of the middle section of the small intestine.
360	Major abnormality of gastrointestinal system, Ileal atresia	Congenital absence or closure of a portion of the ileum.
370	Major abnormality of gastrointestinal system, Ileal stenosis	Stricture or narrowing of a portion of the ileum.
50	Major abnormality of gastrointestinal system, Intestinal malrotation	Abnormal placement and fixation of intestines.
40	Major abnormality of gastrointestinal system, Hirschsprung's disease (Congenital aganglionic megacolon)	A disorder of the enteric nervous system characterized by an absence of ganglion cells in the distal colon resulting in a functional obstruction.
380	Major abnormality of gastrointestinal system, Stenosis of large intestine	A constriction or narrowing of the distal portion of the intestine, extending from its junction with the small intestine to the anus and comprising the cecum, colon, rectum, and anal canal.
390	Major abnormality of gastrointestinal system, Atresia of large intestine	Colonic atresia is usually segmental, most often involving the ascending colon, and may be accompanied of the small intestine, rectum, or anal canal.
400	Major abnormality of gastrointestinal system, Atresia of rectum	Congenital absence or closure of a portion of the rectum. Atresia of the rectum proper, or a portion of the rectum, is very rare. It can occur with or without anomalies of the small intestine, colon, or anal canal.
410	Major abnormality of gastrointestinal system, Stenosis of rectum	A constriction or narrowing of the terminal portion of the large intestine, extending from the sigmoid flexure to the anal canal.
10	Major abnormality of gastrointestinal system, Anal Atresia (imperforate anus)	Anal atresia, or imperforate anus, is a specific type of what are commonly referred to as anorectal malformations. Atresia of the anal canal occurs with or without a fistulous opening to an ectopic location on the perineum, within the urinary system, or into the vaginal vestibule.
560	Major abnormality of genitalia, Ambiguous genitalia	
570	Major abnormality of genitalia, Hypospadiasis	
580	Major abnormality of genitalia, Rectovaginal fistula	
590	Major abnormality of genitalia, Undescended testis	
600	Major abnormality of kidney, Horseshoe kidney	

610	Major abnormality of kidney, Hydronephronsis	
620	Major abnormality of kidney, Polycystic kidney	
630	Major abnormality of kidney, Single kidney	
990	Other	Other major noncardiac abnormality
110	Major abnormality of head (RETIRED)	
130	Major abnormality of brain, Macrocephaly (RETIRED)	
140	Major abnormality of brain, Microcephaly (RETIRED)	
150	Major abnormality of brain (RETIRED)	
180	Major abnormality of spinal cord (RETIRED)	
200	Major abnormality of spine (RETIRED)	
250	Major abnormality of larynx - trachea - or bronchus (RETIRED)	
300	Major abnormality of lung (RETIRED)	
420	Major abnormality of gastrointestinal system (RETIRED)	
430	Major abnormality of kidney - ureter - or bladder (RETIRED)	

Extracardiac anomaly - specify

Seq Num: 765

Required for case closure: No

Registry field: [ExtracardAnomaly].[ExtracardAnomSpec]

Shared with PAC3

Description: Indicate the other major extracardiac abnormality.

Chromosomal abnormality

Seq Num: 700

Required for case closure: Yes

Registry field: [ChromAnom].[ChromAnom]

Shared with PAC3

Description: Select all of the chromosomal anomalies identified in the patient. If none, select "No chromosomal abnormality identified."

Values	*Code*	*Text*
	5	No chromosomal or genetic abnormality identified
	490	Known Mosaicism
	360	1p36 del
	370	1q21.1 del
	380	1q21.1 dup
	70	1q42.1
	100	2p21
	110	3p22
	400	3q dup
	150	4p16
	410	4q del
	420	5p15.2 del
	430	5p15.33 del
	170	6p12
	180	7q11
	440	7q11.23 del
	450	7q11.23 dup
	200	7q32
	210	7q34
	460	8p23.1 del
	470	8p23.1 dup
	220	8q12
	480	9q34.3 del
	10	11p15.5
	20	11q
	30	12p1.21
	40	12p12.1
	50	12q24
	320	15q11.2 del
	60	15q21.1
	330	16p11.2 del
	340	17p11.2 del

350	17q21.31 del
80	20p12
90	22q11 deletion
390	22q11.2 dup
120	45X0
130	47,XXY
230	Monosomy X
250	Trisomy 08
260	Trisomy 09
270	Trisomy 13
280	Trisomy 18
290	Trisomy 21
310	Other chromosomal or genetic abnormality
140	4p (RETIRED)
160	5p (RETIRED)
190	7q11.23 (RETIRED)
240	TGFBR1 or 2 (RETIRED)

Chromosomal abnormality - specify

Seq Num: 705

Required for case closure: No

Registry field: [ChromAnom].[ChromAnomSpec]

Shared with PAC3

Description: Indicate the other chromosomal abnormality.

Syndromes

Seq Num: 820

Required for case closure: Yes

Registry field: [Syndromes].[Syndrome]

Shared with PAC3

Description: Select all of the syndromes or syndromic abnormalities identified in the patient. A “syndrome” is defined as a group of signs and symptoms that occur together, and characterize a particular abnormality. (Tchervenkov CI, Jacobs JP, Weinberg PM, Aiello VD, Beland MJ, Colan SD, Elliott MJ, Franklin RC, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G. The nomenclature, definition and classification of hypoplastic left heart syndrome. Cardiology in the Young, 2006; 16(4): 339–368, August 2006.) If none, select "No syndromic abnormality identified."

Values

Code	*Text*	
5	No syndromic abnormality identified	This patient has no syndromic abnormality identified.
680	1p36 deletion syndrome	
690	1q21.1 duplicaiton syndrome	
710	3q duplication syndrome	
720	4q deleltion syndrome	
730	7q11.23 duplication syndrome	
740	8p23.1 deletion syndrome	
660	15q11.2 deletion syndrome	
670	16p11.2 deletion syndrome	
700	22q11.2 duplication syndrome	
750	Adams-Oliver syndrome	
10	Alagille syndrome (intrahepatic biliary duct agenesis)	Alagille syndrome, or Alagille-Watson syndrome, is an autosomal dominant condition [mapped to 20p12 & 1p13-p11] of intrahepatic biliary duct agenesis or arteriohepatic dysplasia. Incidence is 1:70,000 births. The 20-year predicted life expectancy is 75% for all patients, 80% for those not requiring a liver transplant, and 60% for those requiring a liver transplant. Typical manifestations include intrahepatic cholestasis, distinctive facies, anterior chamber abnormalities of the eye, and butterfly hemiverterbrae. The most common cardiovascular abnormality is peripheral pulmonary artery stenosis. Additional defects include ASD, VSD, coarctation of the aorta and TOF. .
760	Alstrom syndrome	
580	Alveolar Capillary Dysplasia Syndrome	
20	Apert syndrome	Apert syndrome, also known as Apert-Crouzon disease or Vogt cephalodactyly, is an autosomal dominant condition [mapped to 10q26] of acrocephalosyndactyly. Incidence is 1:65,000-88,000 births; it occurs in strong association with advanced paternal age at conception. Apert syndrome is similar to Crouzon and Pfeiffer syndromes. Cardiovascular abnormalities include pulmonic stenosis, VSD, overriding aorta, and endocardial fibroelastosis.

770	Baller-Gerold Syndrome	
780	Bardet-Biedl syndrome	
790	Beckwith-Wiedemann syndrome	
30	Brugada syndrome (Sudden unexplained nocturnal death syndrome) (SUNDS)	Brugada syndrome, also known as sudden unexplained nocturnal death syndrome (SUNDS), is an autosomal dominant condition [mapped to 3p21, 3p22.3, 12p13.3 & 10p12], occurring in 1:2000 births. Brugada syndrome is associated with the risk of sudden cardiac death. Mean age of sudden death is approximately 40 years. Symptoms include right bundle branch block and ST segment elevation on ECG, idiopathic ventricular fibrillation, and cardiac arrest. Brugada syndrome, in its typical form is sinus rhythm with anterior raised ST segment in V1 and V2 due to a genetic ion-channel defect involving a sodium-channel defect isolated to SCN5A gene. Brugada syndrome is a type of “Channelopathy”. A ventricular tachycardia due to a genetic ion-channel defect is also known as a "Channelopathy" or "Ion channelopathy". This diagnosis is most commonly Long QT syndrome, but also includes Brugada syndrome, Jervell and Lange-Nielsen syndrome, Romano-Ward syndrome, Andersen syndrome, etc.
800	Brugada/Timothy Syndrome	
810	Cantu syndrome	
40	Cardiofaciocutaneous syndrome	Cardiofaciocutaneous syndrome (CFC) is a sporadic condition [mapped to 7q34] affecting the heart, face, skin and hair. Incidence is 1:333,000-500,000 births. CFC is similar to Noonan and Costello syndromes. Cardiovascular abnormalities include pulmonary valve stenosis, ASD and hypertrophic cardiomyopathy.
50	Carpenter syndrome	Carpenter syndrome is an autosomal recessive condition [mapped to 6p11] of acrocephalopolysyndactyly, type II. Incidence is 1:1,000,000 births. Cardiovascular abnormalities in 50% of cases include ASD, VSD, pulmonic stenosis, TOF, TGA and PDA.
60	Cat-eye syndrome	The cat-eye syndrome, or Schmid-Fraccaro syndrome, is an autosomal dominant condition [mapped to 22q11], associated with coloboma of the iris. Incidence is 1:50,000-150,000 births. The classic pattern of malformations includes mild mental deficiency, hypertelorism, down-slanting palpebral fissures, iris coloboma, pre-auricular pits or tags, and anal and renal malformations. Cardiovascular abnormalities in 40% of cases include TAPVC, ASD, VSD, persistent left superior vena cava, TOF, interruption of the inferior vena cava, and tricuspid atresia.
590	Caudal Regression Syndrome	
830	Char syndrome	
70	CHARGE Association	CHARGE syndrome, or Hall-Hittner syndrome, is an autosomal dominant condition [mapped to 8q12.1 & 7q21.11]; some sporadic cases have been reported. Incidence is 1:8500-10,000 births. CHARGE syndrome is a nonrandom association of congenital anomalies which may include Coloboma, Heart defects, Atresia choanae, Retarded growth and development and/or central nervous system

70		anomalies, Genital anomalies and/or hypogonadism and Ear anomalies and/or deafness. Diagnosis is made if 4/6 major (or 3 major & 3 minor) defects are present. Heart defects are present in 75% to 80% of cases. Of those with heart defects, most have conotruncal anomalies (TOF, DORV, truncus arteriosus) and aortic arch anomalies (vascular ring, aberrant subclavian artery, IAA, coarctation of the aorta, right aortic arch, aortic valve stenosis). Other cardiovascular abnormalities include PDA, AVSD, VSD, and ASD.
600	Chiari I Malformation	
840	Chromosome 17q12 deletion syndrome	
850	Coffin Lowry syndrome	
860	Coffin Siris Syndrome	
80	Cornelia de Lange syndrome	Cornelia de Lange syndrome (CDLS), also known as de Lange or Brachmann-de Lange syndrome, is an autosomal dominant condition [mapped to 5p13.1, Xp11.22-11.21 & 10q25]; some X-linked and sporadic cases have been reported. Incidence is 1:10,000-30,000 births. Cardiovascular abnormalities in 25% of cases most commonly include VSD and ASD.
90	Costello syndrome	Costello syndrome is an autosomal dominant condition [mapped to 12p12.1 & 11p15.5]; some sporadic cases have been reported. Incidence is 1:1,000,000 births. Cardiovascular abnormalities include ASD, VSD, pulmonic stenosis, mitral valve prolapse, hypertrophic cardiomyopathy and arrhythmias.
870	Cranioectodermal dysplasia (Sensenbrenner syndrome)	
100	Cri-du-chat syndrome	Cri-du-chat (cat cry), or LeJeune syndrome, is a chromosome deletion syndrome [mapped to 5p15.2]. Incidence is 1:20,000-50,000 births. Cri-du-chat refers to the distinctive cry of children with this disorder, caused by abnormal larynx development. Cardiovascular abnormalities in 30% of cases most commonly include VSD and ASD. Rare defects include TOF and AVSD.
610	Dandy Walker Malformation	
110	Deletion 10p syndrome	Deletions on the short arm of chromosome 10 are associated with septal defects, particularly ASDs, and DiGeorge/velocardiofacial 2 syndrome.
120	Deletion 8p syndrome	Deletions on the short arm of chromosome 8 are associated with ASD, AVSC, conotruncal abnormalities, pulmonic valve stenosis and Tetralogy of Fallot.
130	DiGeorge syndrome (velocardiofacial syndrome) (conotruncal anomaly face syndrome) (22q11 deletion)	DiGeorge syndrome, also known as Shprintzen, Takao, velocardiofacial, or conotruncal anomaly face syndrome, is an autosomal dominant condition [mapped to 22q11.2]. Incidence is 1:4000 births. Cardiovascular anomalies are seen in association with hypoplasia or aplasia of the thymus and parathyroid gland, which are derivatives of pharyngeal pouches III and IV, and which can result in abnormalities of the immune system and calcium metabolism respectively. Cardiovascular abnormalities include conotruncal or

130		outflow tract defects of the heart, such as tetralogy of Fallot, truncus arteriosus, and interrupted aortic arch, particularly type B IAA. Additional defects include VSD, right aortic arch, aberrant right subclavian artery, and PDA.
880	Distinct disorder	
140	Down syndrome (Trisomy 21)	Down syndrome, or Trisomy 21, is the most frequent chromosomal abnormality. Incidence is 1:600-1000 live births. Sporadic cases of Down syndrome occur in strong association with advanced maternal age at conception. Affected individuals have an extra (or third) copy of chromosome 21. Cardiovascular abnormalities in 40-50% of cases, in decreasing order of frequency, include AVSD, VSD, TOF and PDA. Left-sided obstructive defects, such as coarctation and aortic valve stenosis, are rare.
890	Duane Radial Ray (Okihiro) syndrome	
620	Duchenne Muscular Dystrophy	
150	Edwards syndrome (Trisomy 18)	Edwards syndrome, or Trisomy 18, is a chromosomal abnormality. Incidence is 1:3000-5000 births. Sporadic cases of Edwards syndrome occur usually in association with advanced maternal age at conception. Affected individuals have an extra (or third) copy of chromosome 18. Approximately 50% of infants with Trisomy 18 die within the first week of life, approximately 40% die within the first month of life, only 5-10% survive beyond the first year. Cardiovascular abnormalities in more than 50% of cases include VSD, ASD and PDA; bicuspid aortic and/or pulmonary valves, nodularity of valve leaflets, pulmonic stenosis, coarctation of the aorta in 10-50% of cases; and anomalous coronary artery, TGA, TOF, dextrocardia and aberrant subclavian artery in less than 10% of cases.
570	Ehlers-Danlos Syndrome	Ehlers-Danlos syndrome is a group of inherited disorders marked by extremely loose joints, hyperelastic skin that bruises easily, and easily damaged blood vessels. A variety of gene mutations involve collagen of the skin, bone, blood vessels, and internal organs. The abnormal collagen leads to the symptom which can include rupture of internal organs or abnormal heart valves.
160	Ellis-van Creveld syndrome	Ellis-van Creveld syndrome, or chondroectodermal dysplasia, is an autosomal recessive condition [mapped to 4p16] of skeletal dysplasia. Incidence is 1:60,000-200,000 births. Major features include short stature of prenatal onset (short limbs), hypoplastic nails and dental anomalies, postaxial polydactyly, narrow thorax, and cardiac defects. Cardiovascular abnormalities in more than 50% of cases most commonly include ASD or common atrium. Additional defects include PDA, persistent left superior vena cava, hypoplastic left heart defects, coarctation of the aorta, TAPVC, and TGA.
900	Familial atrial septal defects	
910	Familial CHD	
920	Familial non-syndromic CHD	
165	Fetal alcohol syndrome (FAS)	Indicate whether the patient has a history of Fetal alcohol

165		syndrome (FAS). Fetal alcohol syndrome (FAS) is a condition that results from prenatal alcohol exposure. FAS is a group of problems that can include mental retardation, birth defects, abnormal facial features, growth problems, problems with the central nervous system, trouble remembering and/or learning, vision or hearing problems, and behavior problems. Mothers who consume large quantities of alcohol during pregnancy may have babies who are born with Fetal Alcohol Syndrome (or FAS). A diagnosis of FAS is based on three factors: 1) prenatal and postnatal growth retardation; 2) central nervous system abnormalities, and, 3) abnormalities of the face. Many of these children display significant disabilities, learning disorders, and emotional problems as they mature.
166	Fetal drug exposure	Indicate whether the patient has a history of Fetal drug exposure. Fetal drug exposure can lead to numerous problems including low birth weight, prematurity, small for Gestational Age (SGA), failure to Thrive (FTT), neurobehavioral symptoms, infectious diseases, and Sudden Infant Death Syndrome (SIDS).
380	Fetal rubella syndrome (Congenital rubella syndrome)	Indicate whether the patient has a history of maternal rubella virus infection during first trimester of pregnancy. Fetal rubella syndrome is associated with PDA, peripheral pulmonary stenosis, fibromuscular and intimal proliferation of medium and large arteries, VSD and ASD.
930	Fragile X	
170	Goldenhar syndrome	Goldenhar syndrome, also known as hemifacial microsomia, oculoauriculovertebral dysplasia or spectrum, and facioauriculovertebral sequence, is an autosomal dominant condition [mapped to 14q32]. Incidence is 1:3000-5000 births. Cardiovascular abnormalities include VSD, PDA, TOF and coarctation
180	Heterotaxy syndrome, Unknown if asplenia or polysplenia	Heterotaxy is synonymous with 'visceral heterotaxy' and 'heterotaxy syndrome'. Heterotaxy is defined as an abnormality where the internal thoraco-abdominal organs demonstrate abnormal arrangement across the left-right axis of the body. By convention, heterotaxy does not include patients with either the expected usual or normal arrangement of the internal organs along the left-right axis, also known as 'situs solitus', nor patients with complete mirror-imaged arrangement of the internal organs along the left-right axis also known as 'situs inversus'.
190	Heterotaxy syndrome, Asplenia syndrome	"Asplenia syndrome" can be defined as a subset of heterotaxy with components of bilateral right-sidedness, usually associated with absence of the spleen.
200	Heterotaxy syndrome, Polysplenia syndrome	"Polysplenia syndrome" can be defined as a subset of heterotaxy with components of bilateral left-sidedness, usually associated with multiple spleens.
210	Holt-Oram syndrome	Holt-Oram, or heart hand, syndrome is an autosomal dominant condition [mapped to 12q24.1]. Incidence is 1:100,000 births. Holt-Oram syndrome was first described in 1960 by Holt and Oram who noted the association of radial anomalies with atrial septal defects. Cardiovascular abnormalities in 75% of cases most commonly include ASD.

210		Additional defects include first degree AV block, bradycardia, fibrillation, AVSD, VSD, HLHS and PDA.
220	Jacobsen syndrome	Jacobsen syndrome is a chromosome deletion syndrome [mapped to 11q23]. Incidence is 1:100,000 births. Associated cardiovascular abnormalities include VSD and ASD.
940	Joubert syndrome	
230	Kabuki syndrome	Kabuki, or Niikawa-Kuroki, syndrome is an autosomal dominant condition. Incidence is 1:32,000 births. Affected individuals have a facial appearance similar to Japanese Kabuki theatre actors. Cardiovascular abnormalities in 50% of cases include ASD, VSD, coarctation of the aorta, bicuspid aortic valve, mitral valve prolapse, TOF, single ventricle with common atrium, DORV, TGA, and pulmonic, aortic and mitral valve stenoses.
240	Kartagener syndrome (Siewert syndrome) (Primary ciliary dyskinesia)	Kartagener syndrome, also known as Siewert syndrome or primary ciliary dyskinesia, is an autosomal recessive condition [mapped to 9p21-p13]. Incidence is 1:30,000 births. Features include situs inversus and asplenia. Cardiovascular abnormalities include dextrocardia.
950	Kleefstra Syndrome	
250	Klinefelter syndrome (XXY Syndrome)	Klinefelter, or 47XXY syndrome, is a sporadic chromosomal abnormality in which males have at least two X chromosomes and at least one Y chromosome. Incidence is 1:500 males or 1:1000 births. Klinefelter syndrome occurs usually in association with advanced maternal age at conception. It is the most common sex chromosome disorder and the most common cause of hypogonadism and infertility. Cardiovascular abnormalities in more than 50% of cases include mitral valve prolapse, varicose veins and deep venous thrombosis.
960	Koolen-De Vries Syndrome	
260	LEOPARD syndrome	LEOPARD is an acronym for multiple Lentigines, Electrocardiographic conduction abnormalities, Ocular hypertelorism, Pulmonic stenosis, Abnormal genitalia, Retardation of growth, and sensorineural Deafness. LEOPARD syndrome is an autosomal dominant condition [mapped to 12q24.1 & 3p25]. Cardiovascular abnormalities include pulmonic stenosis in 40% of cases, and hypertrophic cardiomyopathy in 20% of cases. Additional defects include subaortic stenosis, complete heart block, bundle branch block, prolonged P-R and QRS, and abnormal P waves.
270	Loeys-Dietz syndrome	Loeys-Dietz syndrome is an autosomal dominant condition [mapped to 3p22 & 9q22]. Cardiovascular abnormalities include aortic and arterial aneurysms/dissections with tortuosity of the arteries, PDA, ASD, bicuspid aortic and pulmonic valves, and mitral valve prolapse.
290	Marfan syndrome	Marfan syndrome is an autosomal dominant condition [mapped to 15q21.1]. Incidence is 1:5000 births. Marfan syndrome is the most common connective tissue disorder, and is associated with the risk of sudden cardiac death. Cardiovascular abnormalities include aortic root dilation, aortic dissection and rupture, aortic regurgitation, ascending aortic aneurysm, mitral valve prolapse, mitral

290		regurgitation, tricuspid valve prolapse, premature calcification of the mitral annulus, pulmonary artery dilatation and CHF.
300	Marfan-like syndrome	Marfan-like syndrome is a connective tissue disorder, resembling Marfan syndrome.
970	McKusick-Kaufman syndrome	
980	Meckel-Gruber syndrome	
990	Microphthalmia syndromic 9	
1000	Mowat Wilson Syndrome	
310	Mucopolysaccharidosis type IH (Hurler syndrome)	Hurler syndrome, also known as mucopolysaccharidosis type IH (MPS IH), is an autosomal recessive condition [mapped to 4p16.3]. Incidence is 1:100,000 births. MPS is a lysosomal storage disease. Affected individuals appear normal at birth; subtle changes may be evident during the first 6 months. Survival beyond 10 years of age is unusual. Cardiovascular abnormalities include valve anomalies, coronary artery narrowing, and mitral and atrial regurgitation.
320	Mucopolysaccharidosis type IH/S (Hurler-Scheie syndrome)	Hurler-Scheie syndrome, also known as mucopolysaccharidosis type IH/S (MPS IH/S), is an autosomal recessive disorder [mapped to 4p16.3]. Incidence is 1:500,000 births. MPS is a lysosomal storage disease. Onset of symptoms occurs between ages 3 and 8 years. Survival to adulthood is typical. Cardiovascular abnormalities include mitral valve anomalies.
330	Mucopolysaccharidosis type II (Hunter syndrome)	Hunter syndrome, also known as mucopolysaccharidosis type II (MPS 2), is an X-linked recessive disorder [mapped to Xq28]. Incidence is 1:100,000-170,000 births. MPS is a lysosomal storage disease. Individuals with Hunter syndrome appear normal at birth. Symptoms emerge between ages 2 and 4. Life expectancy is 10-20 years. Cardiovascular abnormalities include valve anomalies, ischemic heart disease, ventricular hypertrophy and CHF.
340	Mucopolysaccharidosis type IS (Scheie syndrome)	Scheie syndrome, also known as mucopolysaccharidosis type IS (MPS IS), is an autosomal recessive disorder [mapped to 4p16.3], which occurs in 1:500,000 births. Scheie syndrome is a lysosomal storage disease. Survival to a late age is typical. Cardiovascular abnormalities include aortic regurgitation, aortic and mitral valve abnormalities.
1010	Nance Horan syndrome	
1020	Nephronophthisis	
1030	Neurofibromatosis	
1040	Non-syndromic CHD	
350	Noonan syndrome	Noonan syndrome is an autosomal dominant condition [mapped to 12q24.1]. Incidence is 1:1000-2500 births. Major features include short stature, seen in about half, mental retardation (usually mild), characteristic facial features, a shield chest deformity, cubitus valgus, and a short webbed neck. Cardiovascular abnormalities occur in at least 50% of cases and include pulmonary valve stenosis (75%) secondary to a dysplastic pulmonary valve with thickened valve leaflets, ASD (30%) usually associated with

350		pulmonary stenosis, PDA (10%), VSD (10%), and hypertrophic cardiomyopathy (10-20%) that can involve both ventricles. Rare lesions include TOF, coarctation of the aorta, subaortic stenosis, and Ebstein malformation. Hypertrophic cardiomyopathy is observed in 10% to 20% and can involve both ventricles.
1050	Oculofaciocardiodental	
1060	Oral-facial-digital syndromes (types I-XVI and unclassified)	
360	Patau syndrome (Trisomy 13)	Patau or Bartholin-Patau syndrome, or Trisomy 13, is a chromosomal abnormality. Incidence is 1:5000-10,000 births. Sporadic cases occur usually in association with advanced maternal age at conception. Affected individuals have an extra (or third) copy of chromosome 13. More than 90% of individuals with Trisomy 13 die within their first days or weeks of life. Only 5-10% survive beyond 1 year of age. Cardiovascular abnormalities in 80% of cases include VSD, PDA, ASD; dextrocardia in more than 50% of cases; and anomalous pulmonary venous connection, overriding aorta, pulmonary stenosis, hypoplastic aorta, mitral valve atresia, aortic valve atresia, and bicuspid aortic valve in fewer than 50% of cases.
1070	Peter’s Plus syndrome	
540	Pierre Robin syndrome	Pierre Robin Syndrome is characterized by an unusually small mandible (micrognathia), posterior displacement or retraction of the tongue (glossoptosis), and upper airway obstruction. Incomplete closure of the roof of the mouth (cleft palate) is present in the majority of patients, and is commonly U-shaped.
1080	Polycystic Kidney Disease	
1090	Primary ciliary dyskinesia (PCD)	
530	Prune Belly Syndrome	Prune belly syndrome, also known as Eagle-Barrett syndrome, is characterized by three main features: Anterior abdominal wall musculature ("stomach muscles") deficient or absent, urinary tract anomalies (such as a very large bladder) and bilateral cryptorchidism (two undescended testicles.) The incidence of prune belly syndrome is about 1 in 40,000 births; 95% of cases occur in males. It is thought that prune belly syndrome is a multisystem disease complex that derives from a primary defect in mesodermal development at about 8 weeks' gestation. The major prognostic factor is the degree of dilation of the urinary tract; 20% of patients are stillborn, 30% die of renal failure or urosepsis within the first two years of life, and the remaining 50% have varying degrees of urinary pathology.
370	Rethore syndrome (Trisomy 9)	Trisomy 9, or Rethore syndrome, is a rare chromosomal abnormality, which is a frequent cause of first trimester spontaneous abortions. Incidence is 1:100,000 births. Affected individuals have an extra (or third) copy of chromosome 9. Most affected individuals die during infancy or early childhood. Cardiovascular abnormalities occur in 75% of cases and include VSD, ASD, PDA, valve defects, DORV, persistent left SVC, and endocardial fibroelastosis.

1100	Roberts syndrome	
1110	Robinow syndrome	
390	Rubinstein-Taybi syndrome	Rubinstein-Taybi or Rubinstein syndrome is an autosomal dominant condition [mapped to 16p13.3 & 22q13]. Incidence is 1:100,000-125,000 births. Cardiovascular abnormalities occur in 30% of cases and include ASD, VSD and PDA.
1120	Saethre Chotzen syndrome	
1130	Short Rib Polydactyly Type I	
1140	Short rib thoracic dysplasias including Jeune chondrodysplasia, Saldino Mainzer	
550	Sickle cell disease	Sickle-cell disease (SCD), or sickle-cell anemia (SCA) is an autosomal recessive genetic blood disorder with overdominance, characterized by red blood cells that assume an abnormal, rigid, sickle shape. Sickling decreases the cells' flexibility and results in a risk of various complications. The sickling occurs because of a mutation in the hemoglobin gene. Sickle-cell disease occurs more commonly in people (or their descendants) from parts of tropical and sub-tropical regions where malaria is or was common.
560	Sickle cell trait	Sickle cell trait describes a condition in which a person has one abnormal allele of the hemoglobin beta gene (is heterozygous), but does not display the severe symptoms of sickle cell disease that occur in a person who has two copies of that allele (is homozygous). Those who are heterozygous for the sickle cell allele produce both normal and abnormal hemoglobin (the two alleles are co-dominant). Sickle cell disease is a blood disorder in which the body produces an abnormal type of the oxygen-carrying substance hemoglobin in the red blood cells. Sickling and sickle cell disease also confer some resistance to malaria parasitization of red blood cells, so that individuals with sickle-cell trait (heterozygotes) have a selective advantage in some environments.
1150	Sifrim-Hitz-Weiss syndrome (SIHIWES)	
1160	Simpson-Golabi-Behmel syndrome	
410	Situs inversus	Situs inversus is defined as an abnormality where the internal thoraco-abdominal organs demonstrate mirror-imaged atrial arrangement across the left-right axis of the body.
1170	Smith Magenis syndrome	
420	Smith-Lemli-Opitz syndrome	Smith-Lemli-Opitz syndrome is an autosomal recessive condition [mapped to 11q12-q13]. Incidence is 1:20,000-40,000 births. Cardiovascular abnormalities include VSD, ASD, coarctation of the aorta, and PDA.
1180	Sotos syndrome	
630	Spinal Muscular Atrophy, Type II	
1190	Sporadic and familial CHD	

1200	Syndromic CHD	
1210	TAR syndrome	
640	Thalassemia – Major	
650	Thalassemia – Minor	
1220	Townes-Brocks syndrome	
1230	Trisomy 13	
1240	Trisomy 18	
1250	Trisomy 21	
430	Turner syndrome (45XO)	Turner syndrome (45XO) is a chromosomal deletion abnormality, which occurs in 1:5000 live female births. Although common in first trimester, most 45XO conceptuses are spontaneously aborted. Affected individuals are missing one X chromosome. The major features include short stature, primary amenorrhea due to ovarian dysgenesis, webbed neck, congenital lymphedema, and cubitus valgus. Cardiovascular abnormalities occur in 20-40% of cases, the most common of which is coarctation of the aorta (70%). Additional defects include bicommissural aortic valve, aortic stenosis, a spectrum of left-sided obstructive defects and/or hypoplastic defects, hypoplastic left heart syndrome; aortic dilation, dissection, and rupture.
440	VACTERL syndrome (VACTER/VATER/VATERR syndrome)	VACTERL syndrome is a nonrandom association of defects, including Vertebral anomalies, Anal atresia, Cardiovascular anomalies, Tracheoesophageal fistula, Esophageal atresia, Renal and/or Radial anomalies, and Limb anomalies. Diagnosis is made if 3/7 defects are present. Incidence is 1:6000 births. Cardiovascular malformations include VSD, TOF, TGA and PDA.
450	VACTERL-H syndrome (VATER association with hydrocephalus) (Briard-Evans syndrome)	VACTERL-H association is also known as VATER association with hydrocephalus, Briard-Evans syndrome, David-O'Callaghan syndrome (autosomal recessive type), and Hunter-MacMurray syndrome (X-linked type) [mapped to 10q23.31 & Xp22.31]. VACTERL-H is an autosomal recessive condition; some X-linked cases have been reported. VACTERL-H is a nonrandom association of defects, including Vertebral anomalies, Anal atresia, Cardiac malformations, TracheoEsophageal fistula, Renal anomalies, Limb anomalies and Hydrocephalus. Diagnosis is made if 3/7 defects are present with hydrocephalus. Cardiovascular abnormalities include VSD, TOF, TGA and PDA.
520	von Willebrand disease (vWD)	Von Willebrand disease (vWD) is the most common hereditary coagulation abnormality described in humans, although it can also be acquired as a result of other medical conditions. It arises from a qualitative or quantitative deficiency of von Willebrand factor (vWF), a multimeric protein that is required for platelet adhesion. There are three forms of vWD: inherited, acquired and pseudo or platelet type. There are three types of hereditary vWD: vWD Type I, vWD Type II and vWD III. Within the three inherited types of vWD there are various subtypes. Platelet type vWD is also an inherited condition. vWD Type I is the most common type of the disorder and those that have it

520		are typically asymptomatic or may experience mild symptoms such as nosebleeds although there may be severe symptoms in some cases. There are various factors that affect the presentation and severity of symptoms of vWD such as blood type.
460	Warkany syndrome (Trisomy 8)	Trisomy 8, or Warkany syndrome, is a chromosomal abnormality, which is a frequent cause of first trimester spontaneous abortions. Complete Trisomy 8 is usually an early lethal disorder. Incidence is 1:25,000-50,000 births. Affected individuals have an extra (or third) copy of chromosome 8. Cardiovascular abnormalities include septal defects and great vessel anomalies.
470	Williams syndrome (Williams-Beuren syndrome)	Williams syndrome, or Williams-Beuren syndrome, is an autosomal dominant condition [mapped to 7q11.23]. Incidence is 1:7500 births. Williams syndrome was initially described by Williams and colleagues in four unrelated children with mental retardation, an unusual facial appearance, and supravalvar stenosis. Cardiovascular abnormalities occur in at least 50% of cases and include supravalvar aortic stenosis, bicuspid aortic valve, mitral valve prolapse, mitral regurgitation, coronary artery stenosis, pulmonary valve stenosis, ASD, VSD and peripheral pulmonary artery stenosis. Supravalvar aortic stenosis is the most frequent single defect, but any of the systemic or pulmonary arteries can be affected.
490	Wolf-Hirschhorn syndrome	Wolf-Hirschhorn syndrome is a chromosome deletion syndrome [mapped to 4p16.3]. Incidence is 1:96,000 births. Affected individuals have a 35% risk of mortality prior to age 2. Cardiovascular abnormalities include ASD and VSD.
1260	X-linked heterotaxy	
510	Other syndromic abnormality	This patient has other syndromic abnormality(ies) that are not on this list.
280	Long QT syndrome (Ward Romano syndrome) (RETIRED)	
400	Short QT syndrome (RETIRED)	
480	Wolff-Parkinson-White syndrome (WPW syndrome) (RETIRED)	

Syndrome - specify

Seq Num: 825

Required for case closure: No

Registry field: [Syndromes].[SyndromeSpec]

Shared with PAC3

Description: Indicate the other “Syndrome” or “Syndromic abnormality”.

Deceased Seq Num: 840

Required for case closure: No

Registry field: [Demographics].[Deceased]

Shared with PAC3

Description: Indicate Yes if the patient is known to be deceased

Values	*Code*	*Text*
	0	No
	1	Yes
	9	Unk

DOD Seq Num: 860

Required for case closure: No

Registry field: [Demographics].[DOD]

Shared with PAC3

Description: If the patient is deceased, indicate the date of death

EDC known Seq Num: 418

Retired in version 1.0

Required for case closure: No

Registry field: [Demographics].[EDCKnown]

Description: If the patient age is <= 1 year, is the estimated date of confinement known

Values	*Code*	*Text*
Retired	0	No
Retired	1	Yes

Estimated date of confinement Seq Num: 419

Retired in version 1.0

Required for case closure: Yes

Registry field: [Demographics].[EDCDt]

Description: Estimated date of confinement

Episode of Care (Hospitalization)

Hospital name

Seq Num: 911

Required for case closure: Yes

Registry field: [Hospitalization].[HospName]

Shared with PAC3

Description: Indicate the full name of the facility in which the patient was hospitalized. Values should be full, official hospital names with no abbreviations or variations in spelling for a single hospital. Values should also be in mixed-case.

Hospital admit date

Seq Num: 980

Required for case closure: Yes

Registry field: [Hospitalization].[HospAdmitDt]

Shared with PAC3

Description: Indicate the date the patient was admitted to the hospital. For those patients who originally enter the hospital in an out-patient capacity (i.e., catheterization), but then are not discharged, the admit date is the date of the patients entry into the hospital.

Patient age at hospital admission

Seq Num: 982

Required for case closure: Yes

Registry field: [Hospitalization].[HospAdmitAgeD]

Shared with PAC3

Description: The patient's age in days at hospital admission, calculated by the DOB and hospital admit date.

Country of residence

Seq Num: 920

Required for case closure: No

Registry field: [Hospitalization].[ResCountry]

Shared with PAC3

Description: Indicate the patient's country of permanent residence at time of hospital admission.

Values	*Code*	*Text*
	USA	UNITED STATES OF AMERICA
	AFG	AFGHANISTAN
	ALA	ÅLAND ISLANDS
	ALB	ALBANIA
	DZA	ALGERIA
	ASM	AMERICAN SAMOA
	AND	ANDORRA
	AGO	ANGOLA
	AIA	ANGUILLA
	ATG	ANTIGUA AND BARBUDA
	ARG	ARGENTINA
	ARM	ARMENIA
	ABW	ARUBA
	AUS	AUSTRALIA
	AUT	AUSTRIA
	AZE	AZERBAIJAN
	BHS	BAHAMAS
	BHR	BAHRAIN
	BGD	BANGLADESH
	BRB	BARBADOS
	BLR	BELARUS
	BEL	BELGIUM
	BLZ	BELIZE
	BEN	BENIN
	BMU	BERMUDA
	BTN	BHUTAN
	BOL	BOLIVIA (PLURINATIONAL STATE OF)
	BES	BONAIRE, SAINT EUSTATIUS AND SABA
	BIH	BOSNIA AND HERZEGOVINA
	BWA	BOTSWANA
	BRA	BRAZIL
	VGB	BRITISH VIRGIN ISLANDS

BRN	BRUNEI DARUSSALAM
BGR	BULGARIA
BFA	BURKINA FASO
BDI	BURUNDI
KHM	CAMBODIA
CMR	CAMEROON
CAN	CANADA
CPV	CAPE VERDE
CYM	CAYMAN ISLANDS
CAF	CENTRAL AFRICAN REPUBLIC
TCD	CHAD
CHL	CHILE
CHN	CHINA
COL	COLOMBIA
COM	COMOROS
COG	CONGO
COK	COOK ISLANDS
CRI	COSTA RICA
CIV	CÔTE D'IVOIRE
HRV	CROATIA
CUB	CUBA
CUW	CURAÇAO
CYP	CYPRUS
CZE	CZECH REPUBLIC
PRK	DEMOCRATIC PEOPLE'S REPUBLIC OF KOREA
COD	DEMOCRATIC REPUBLIC OF THE CONGO
DNK	DENMARK
DJI	DJIBOUTI
DMA	DOMINICA
DOM	DOMINICAN REPUBLIC
ECU	ECUADOR
EGY	EGYPT
SLV	EL SALVADOR
GNQ	EQUATORIAL GUINEA
ERI	ERITREA
EST	ESTONIA
ETH	ETHIOPIA
FRO	FAEROE ISLANDS
FLK	FALKLAND ISLANDS (MALVINAS)
FJI	FIJI

FIN	FINLAND
FRA	FRANCE
GUF	FRENCH GUIANA
PYF	FRENCH POLYNESIA
GAB	GABON
GMB	GAMBIA
GEO	GEORGIA
DEU	GERMANY
GHA	GHANA
GIB	GIBRALTAR
GRC	GREECE
GRL	GREENLAND
GRD	GRENADA
GLP	GUADELOUPE
GUM	GUAM
GTM	GUATEMALA
GGY	GUERNSEY
GIN	GUINEA
GNB	GUINEA-BISSAU
GUY	GUYANA
HTI	HAITI
VAT	HOLY SEE
HND	HONDURAS
HKG	CHINA, HONG KONG SPECIAL ADMINISTRATIVE REGION
HUN	HUNGARY
ISL	ICELAND
IND	INDIA
IDN	INDONESIA
IRN	IRAN (ISLAMIC REPUBLIC OF)
IRQ	IRAQ
IRL	IRELAND
IMN	ISLE OF MAN
ISR	ISRAEL
ITA	ITALY
JAM	JAMAICA
JPN	JAPAN
JEY	JERSEY
JOR	JORDAN
KAZ	KAZAKHSTAN
KEN	KENYA

KIR	KIRIBATI
KWT	KUWAIT
KGZ	KYRGYZSTAN
LAO	LAO PEOPLE'S DEMOCRATIC REPUBLIC
LVA	LATVIA
LBN	LEBANON
LSO	LESOTHO
LBR	LIBERIA
LBY	LIBYA
LIE	LIECHTENSTEIN
LTU	LITHUANIA
LUX	LUXEMBOURG
MAC	CHINA, MACAO SPECIAL ADMINISTRATIVE REGION
MDG	MADAGASCAR
MWI	MALAWI
MYS	MALAYSIA
MDV	MALDIVES
MLI	MALI
MLT	MALTA
MHL	MARSHALL ISLANDS
MTQ	MARTINIQUE
MRT	MAURITANIA
MUS	MAURITIUS
MYT	MAYOTTE
MEX	MEXICO
FSM	MICRONESIA (FEDERATED STATES OF)
MCO	MONACO
MNG	MONGOLIA
MNE	MONTENEGRO
MSR	MONTSERRAT
MAR	MOROCCO
MOZ	MOZAMBIQUE
MMR	MYANMAR
NAM	NAMIBIA
NRU	NAURU
NPL	NEPAL
NLD	NETHERLANDS
NCL	NEW CALEDONIA
NZL	NEW ZEALAND

NIC	NICARAGUA
NER	NIGER
NGA	NIGERIA
NIU	NIUE
NFK	NORFOLK ISLAND
MNP	NORTHERN MARIANA ISLANDS
NOR	NORWAY
PSE	OCCUPIED PALESTINIAN TERRITORY
OMN	OMAN
PAK	PAKISTAN
PLW	PALAU
PAN	PANAMA
PNG	PAPUA NEW GUINEA
PRY	PARAGUAY
PER	PERU
PHL	PHILIPPINES
PCN	PITCAIRN
POL	POLAND
PRT	PORTUGAL
PRI	PUERTO RICO
QAT	QATAR
KOR	REPUBLIC OF KOREA
MDA	REPUBLIC OF MOLDOVA
REU	RÉUNION
ROU	ROMANIA
RUS	RUSSIAN FEDERATION
RWA	RWANDA
SHN	SAINT HELENA
KNA	SAINT KITTS AND NEVIS
LCA	SAINT LUCIA
SPM	SAINT PIERRE AND MIQUELON
VCT	SAINT VINCENT AND THE GRENADINES
BLM	SAINT-BARTHÉLEMY
MAF	SAINT-MARTIN (FRENCH PART)
WSM	SAMOA
SMR	SAN MARINO
STP	SAO TOME AND PRINCIPE
SAU	SAUDI ARABIA
SEN	SENEGAL
SRB	SERBIA

SYC	SEYCHELLES
SLE	SIERRA LEONE
SGP	SINGAPORE
SXM	SINT MAARTEN (DUTCH PART)
SVK	SLOVAKIA
SVN	SLOVENIA
SLB	SOLOMON ISLANDS
SOM	SOMALIA
ZAF	SOUTH AFRICA
SSD	SOUTH SUDAN
ESP	SPAIN
LKA	SRI LANKA
SDN	SUDAN
SUR	SURINAME
SJM	SVALBARD AND JAN MAYEN ISLANDS
SWZ	SWAZILAND
SWE	SWEDEN
CHE	SWITZERLAND
SYR	SYRIAN ARAB REPUBLIC
TJK	TAJIKISTAN
THA	THAILAND
MKD	THE FORMER YUGOSLAV REPUBLIC OF MACEDONIA
TLS	TIMOR-LESTE
TGO	TOGO
TKL	TOKELAU
TON	TONGA
TTO	TRINIDAD AND TOBAGO
TUN	TUNISIA
TUR	TURKEY
TKM	TURKMENISTAN
TCA	TURKS AND CAICOS ISLANDS
TUV	TUVALU
UGA	UGANDA
UKR	UKRAINE
ARE	UNITED ARAB EMIRATES
GBR	UNITED KINGDOM OF GREAT BRITAIN AND NORTHERN IRELAND
TZA	UNITED REPUBLIC OF TANZANIA
VIR	UNITED STATES VIRGIN ISLANDS
URY	URUGUAY

UZB	UZBEKISTAN
VUT	VANUATU
VEN	VENEZUELA (BOLIVARIAN REPUBLIC OF)
VNM	VIET NAM
WLF	WALLIS AND FUTUNA ISLANDS
ESH	WESTERN SAHARA
YEM	YEMEN
ZMB	ZAMBIA
ZWE	ZIMBABWE
OTH	Other

Residential zip code

Seq Num: 940

Required for case closure: No *Submission is optional*

Registry field: [Hospitalization].[ResZip]

Shared with PAC3

Description: If the patient resides in the USA or Canada, indicate the zip/postal code of patient's permanent residence at time of hospital admission. This field should be collected in compliance with state/local privacy laws.

Primary insurance type

Seq Num: 960

Required for case closure: No

Registry field: [Hospitalization].[InsPrimType]

Description: Indicate the primary insurance type at the beginning of this hospitalization.

Values	*Code*	*Text*	
	1	Public	Includes Medicare, Medicaid, Military Health Care (e.g., TriCare), State-Specific Plan, and Indian Health Service.
	2	Private	Includes all indemnity (fee-for-service) carriers, Preferred Provider Organizations (PPOs), and Health Maintenance Organizations (HMOs).
	3	Non-U.S. insurance	Includes all non-U.S. insurance
	4	None / Self	No insurance was used by the patient to pay for this admission.

Initial weight

Seq Num: 1000

Required for case closure: Yes

Registry field: [Hospitalization].[HospAdmitWt]

Shared with PAC3

Description: Indicate the weight of the patient in kilograms at hospital admission

Initial length/height

Seq Num: 1020

Required for case closure: No

Registry field: [Hospitalization].[HospAdmitLen]

Shared with PAC3

Description: Indicate the length/height of the patient in centimeters at hospital admission. If not available, use length/height +/- one month of the hospital admit date

Tracheostomy at hospital admit

Seq Num: 1021

Required for case closure: Yes

Registry field: [Hospitalization].[HospAdmitTrach]

Shared with PAC3

Description: Indicate Yes if the patient had an actively cannulated tracheostomy present at hospital admission.
*
Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Home respiratory support at hospital admit

Seq Num: 1022

Required for case closure: Yes

Registry field: [Hospitalization].[HospAdmitResp]

Shared with PAC3

Description: Indicate Yes if the patient was on any home respiratory support -- invasive or non-invasive (CPAP or BiPAP) -- during all or part of the day/night at the time of hospital admission. This does not include supplementary oxygen only.
*
Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values

Code	*Text*
1	Yes
0	No
9	Unk

On heart transplant list at hospital admission

Seq Num: 1023

Required for case closure: Yes

Registry field: [Hospitalization].[HospListedAdmit]

Shared with PAC3

Description: Was the patient on the heart transplant list at the time of hospital admission? Code Yes if the patient is listed at any status, including hold status due to any contraindication.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Listed during admission

Seq Num: 1024

Required for case closure: Yes

Registry field: [Hospitalization].[HospListedNew]

Shared with PAC3

Description: If the patient was not listed at hospital admission, code Yes if he/she was either placed on the transplant list during this hospitalization or referred to another facility (as a direct transfer) with the expressed purpose of transplant evaluation or listing.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Date listed

Seq Num: 1025

Required for case closure: Yes

Registry field: [Hospitalization].[HospListedNewDt]

Shared with PAC3

Description: Document the date the patient was placed on the list. Use the date of hospital discharge if the patient was referred to another facility with the expressed purpose of transplant evaluation or listing.

Existing PPM/AICD at hospital admission

Seq Num: 1026

Required for case closure: Yes

Registry field: [Hospitalization].[HospPPMadmit]

Shared with PAC3

Description: Indicate Yes if the patient had a permanent implanted pacemaker or AICD at the time of hospital admission.

*

This includes wearable defibrillators such as LifeVest.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

PPM/AICD placed during admission

Seq Num: 1027

Required for case closure: Yes

Registry field: [Hospitalization].[HospPPMnew]

Shared with PAC3

Description: If no device was present on admission, was a PPM or AICD placed during this hospitalization?

*

This includes a new wearable defibrillator such as LifeVest.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Date PPM/AICD placed

Seq Num: 1028

Required for case closure: Yes

Registry field: [Hospitalization].[HospPPMnewDt]

Shared with PAC3

Description: Document the earliest date a PPM or AICD was placed during this hospital admission.

New dx diaphragm dysfunction Seq Num: 1029

Required for case closure: Yes

Registry field: [Hospitalization].[HospDiaphragm]

Shared with PAC3

Description: Did the patient receive a new diagnosis of diaphragm dysfunction during the hospital admission? Diaphragm dysfunction is defined as the presence of an elevated hemi-diaphragm on chest radiograph in conjunction with evidence of weak, immobile or paradoxical movement assessed by ultrasound or fluoroscopy.

*

This field is intended to capture acquired diaphragm dysfunction. A new diagnosis of congenital diaphragmatic hernia (CHD) may be coded in the extracardiac anomalies (#760) but should NOT be coded here.

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Diaphragm dysfunction dx date Seq Num: 1030

Required for case closure: Yes

Registry field: [Hospitalization].[HospDiaphragmDt]

Shared with PAC3

Description: If the patient had new diaphragm dysfunction, document the date it was first diagnosed during the hospital admission.

New dx vocal cord dysfunction Seq Num: 1031

Required for case closure: Yes

Registry field: [Hospitalization].[HospVocalCord]

Shared with PAC3

Description: Did the patient receive a new diagnosis of vocal cord dysfunction during the hospital admission? Vocal cord dysfunction is defined as the presence of poor or no vocal cord movement assessed by endoscopy. Patient may or may not have stridor, hoarse voice, or poor cry, in conjunction with endoscopic findings.

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Vocal cord dysfunction dx date Seq Num: 1032

Required for case closure: Yes

Registry field: [Hospitalization].[HospVocalCordDt]

Shared with PAC3

Description: If the patient had vocal cord dysfunction, document the date it was first diagnosed during the hospital admission.

Ever had a chest tube during this hospital admission Seq Num: 1033

Required for case closure: Yes

Registry field: [Hospitalization].[HospChestTube]

Shared with PAC3

Description: Did the patient ever have a chest tube in place during this hospital admission?
*
This field is intended to capture drains inserted into the pleural or intrapleural space.

Pericardial effusions that require drainage during the CICU encounter should be captured in the Complication section (#6540).

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Final chest tube date Seq Num: 1034

Required for case closure: Yes

Registry field: [Hospitalization].[HospChestTubeFinalDt]

Shared with PAC3

Description: Document the date the final chest tube was removed. If the patient died or was discharged to another facility with a tube in place, use the hospital discharge date.

Ever on cardiac acute care attending service Seq Num: 1036

Required for case closure: Yes

Registry field: [Hospitalization].[HospAcuteCare]

Description: During this hospital admission, was the patient ever cared for in a non-ICU setting where the cardiac acute care team was the primary attending service?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Hospital discharge date

Seq Num: 1040

Required for case closure: Yes

Registry field: [Hospitalization].[HospDischDt]

Shared with PAC3

Description: Indicate the date the patient was discharged from this hospital. If the patient died in-hospital, use the date of death.

Discharge weight

Seq Num: 1100

Required for case closure: No

Registry field: [Hospitalization].[HospDischWt]

Description: Indicate the weight of the patient in kilograms at hospital discharge

Mortality status at hospital discharge

Seq Num: 1060

Required for case closure: Yes

Registry field: [Hospitalization].[HospDischStat]

Shared with PAC3

Description: Indicate whether the patient was alive or deceased at hospital discharge.

Values	*Code*	*Text*
	1	Alive
	2	Deceased

Hospital discharge location

Seq Num: 1080

Required for case closure: Yes

Registry field: [Hospitalization].[HospDischLoc]

Shared with PAC3

Description: If the patient was alive at hospital discharge, indicate the location to which the patient was discharged.

Values	*Code*	*Text*
	1	Home
	2	Other acute care setting
	3	Other chronic care setting

Tube feeding at hospital discharge

Seq Num: 1120

Required for case closure: Yes

Registry field: [Hospitalization].[HospDischTube]

Description: Indicate Yes if patient was receiving any enteral feedings by temporary or permanent feeding tube at hospital discharge

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

New permanent feeding tube

Seq Num: 1140

Required for case closure: Yes

Registry field: [Hospitalization].[HospDischTubePerm]

Description: If the patient was tube fed at hospital discharge, indicate Yes if a new permanent feeding tube was placed during this hospital admission. This includes both placement of a new permanent feeding tube or revision of a previous permanent feeding tube to a different type (e.g. gastric tube changed to a gastro-jejunal tube).

*

The replacement of an existing tube with a similar tube (i.e., a larger tube, or the replacement of a defective tube) does not count as a new tube.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

30-day hospital readmission

Seq Num: 1240

Required for case closure: No

Registry field: [Hospitalization].[HospReadm]

Shared with PAC3

Description: Indicate whether the patient was readmitted within thirty days of discharge.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

30-day mortality status

Seq Num: 1260

Required for case closure: No

Registry field: [Hospitalization].[Hosp30DStat]

Shared with PAC3

Description: If the patient was alive at hospital discharge, Indicate whether the patient was alive or deceased on the 30th day post hospital discharge.

Values	*Code*	*Text*
	1	Alive
	2	Deceased
	3	Unknown

Beta-blockers on admit

Seq Num: 1180

Retired in version 2.0

Required for case closure: No

Registry field: [Hospitalization].[HospAdmitBeta]

Description: If the patient is >= 18 years of age and had cardiothoracic surgery during this episode of care (hospitalization), indicate Yes if the patient was on beta-blockers at hospital admission or received beta blockers within 24 hours preceding surgery, or No if this did not occur. Select "contraindicated" if beta blocker was contraindicated. The contraindication must be documented in the medical record by a physician, nurse practitioner, or physician assistant.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	2	Contraindicated
Retired	9	Unk

Beta-blockers at discharge

Seq Num: 1200

Retired in version 2.0

Required for case closure: No

Registry field: [Hospitalization].[HospDischBeta]

Description: If the patient is >= 18 years of age and had cardiothoracic surgery during this episode of care (hospitalization), indicate Yes if the patient was discharged on beta blockers, or no if this was not the case. Select "contraindicated" if beta blocker was contraindicated. The contraindication must be documented in the medical record by a physician, nurse practitioner, or physician assistant.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	2	Contraindicated
Retired	9	Unk

Cardiothoracic Surgery

Cardiothoracic surgery

Seq Num: 1160

Required for case closure: Yes

Registry field: [Hospitalization].[CardSurg]

Shared with PAC3

Description: Select Yes if the patient had any cardiac or thoracic surgery during this episode of care (hospitalization).

*

If the hospitalization meets criteria for inclusion in PC4, capture every cardiothoracic surgery or cardiac catheterization during the hospitalization, regardless of venue. For example, surgeries or catheterizations that happen in the NICU or acute care ward should all be captured.

Patients who undergo a hybrid procedure should have both a cardiac surgery procedure and a cardiac cath procedure (#1220) recorded.

Values	*Code*	*Text*
	1	Yes
	0	No

Cardiac surgery date

Seq Num: 1320

Required for case closure: Yes

Registry field: [Operative].[CardSurgDt]

Shared with PAC3

Description: If the patient underwent cardiac or thoracic surgery, indicate the date of surgery which equals the date the patient enters the OR or equivalent.

OR entry time

Seq Num: 1325

Required for case closure: Yes

Registry field: [Operative].[OREntryT]

Description: Indicate to the nearest minute (using 24-hour clock) the time the patient entered the OR. If the procedure was performed in a location other than the OR, record the time when the sterile field was set up. Recording time of last inpatient vital signs prior to procedure is also acceptable for sites not importing directly from their STS database.

Surgical Age in days Seq Num: 1340

Required for case closure: Yes

Registry field: [Operative].[CardSurgAgeD]

Shared with PAC3

Description: If the patient underwent cardiac or thoracic surgery, calculate the patient's age in days at the time of the surgery procedure. The patient's age will be calculated by the software from the date of birth and the date of surgery.

Weight (kg) at surgery Seq Num: 1360

Required for case closure: Yes

Registry field: [Operative].[SurgWtKg]

Shared with PAC3

Description: Indicate the weight of the patient in kilograms at the time of surgery

Number of prior cardiothoracic operations Seq Num: 1520

Required for case closure: Yes

Registry field: [Operative].[PrevOpCount]

Shared with PAC3

Description: Indicate, prior to this admission's surgical procedure, how many cardiothoracic (heart or great vessels) surgical procedures were performed with or without cardiopulmonary bypass (CPB). Also include lung procedures utilizing CPB or tracheal procedures utilizing CPB.

PGE infusion at time of surgery Seq Num: 1660

Required for case closure: Yes

Registry field: [Operative].[PGEsurg]

Shared with PAC3

Description: For patients age <= 30 days, indicate Yes if the patient was on PGE infusions at the time of surgery.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Planned surgery

Seq Num: 1525

Required for case closure: Yes

Registry field: [Operative].[OpPlanned]

Description: Record Yes if the surgery was the planned operative repair or part of a multi-stage palliative strategy determined prior to the first intervention (surgery or catheterization) during the hospitalization. Delayed sternal closure, ECMO decannulation, VAD decannulation, and removal of Broviac catheter should always be coded as Planned = Yes.

This field is intended to capture unplanned reinterventions following the patient's initial intervention (surgical or cath) for this hospitalization. Therefore, the first surgery or interventional catheter (whichever comes first) should always be coded as Planned, even if emergent. This includes ECMO.

Examples:
1. Hybrid Stage I followed by Norwood procedure in the same hospitalization. If the Norwood procedure was planned prior to the hybrid (i.e., there was no plan to perform comprehensive stage II), code the Norwood as Planned = Yes.

2. If the initial surgery was complete repair of AVSD and patient develops mitral regurgitation necessitating mitral valve repair as second operation, code the mitral valve repair as Planned = No.

3. If the initial intervention was balloon aortic valvuloplasty for critical aortic stenosis and the patient required a Ross procedure for aortic insufficiency, code the Ross procedure as Planned = No.

Additional examples:
4. A patient's sternum was left open after surgery, although the initial plan had been to close the chest at the end of the operation. The delayed sternal closure should still be coded as Planned.

5. A patient with HLHS underwent her Norwood surgery and had a complicated postoperative course. The original plan was to discharge her prior to her Glenn, but instead she remained in the hospital through her Stage 2 postop recovery. The Glenn should be coded as planned because it was part of her planned palliative course at the time of the original intervention.

6. A patient who requires emergent ECMO support prior to undergoing the originally planned surgical intervention, who then recovers and undergoes the originally planned intervention, should have both the ECMO cannulation (as the first intervention of the hospitalization) and the eventual surgical repair coded as “planned”.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Procedure diagnosis

Seq Num: 1440

Required for case closure: Yes

Registry field: [SurgDiag].[SurgDiag]

Shared with PAC3

Description: Indicate all diagnoses noted at the time of the surgical procedure or documented by preoperative studies. This entry may duplicate the Fundamental Diagnosis. (Please see the Fundamental Diagnosis field in the Patient Information section for available values)

Values	*Code*	*Text*	
	10	PFO	A small interatrial communication (or potential communication) confined to the region of the oval fossa (fossa ovalis) characterized by no deficiency of the primary atrial septum (septum primum) and a normal limbus with no deficiency of the septum secundum (superior interatrial fold).
	20	ASD, Secundum	A congenital cardiac malformation in which there is an interatrial communication confined to the region of the oval fossa (fossa ovalis), most commonly due to a deficiency of the primary atrial septum (septum primum) but deficiency of the septum secundum (superior interatrial fold) may also contribute.
	30	ASD, Sinus venosus	A congenital cardiac malformation in which there is a caval vein (vena cava) and/or pulmonary vein (or veins) that overrides the atrial septum or the septum secundum (superior interatrial fold) producing an interatrial or anomalous venoatrial communication Although the term sinus venosus atrial septal defect is commonly used, the lesion is more properly termed a sinus venosus communication because, while it functions as an interatrial communication, this lesion is not a defect of the atrial septum.
	40	ASD, Coronary sinus	A congenital cardiac malformation in which there is a deficiency of the walls separating the left atrium from the coronary sinus allowing interatrial communication through the coronary sinus ostium.
	50	ASD, Common atrium (single atrium)	Complete absence of the interatrial septum. "Single atrium" is applied to defects with no associated malformation of the atrioventricular valves. "Common atrium" is applied to defects with associated malformation of the atrioventricular valves.
	2150	ASD, Postoperative interatrial communication	A surgically created communication between the atria.
	71	VSD, Type 1 (Subarterial) (Supracristal) (Conal septal defect) (Infundibular)	A VSD that lies beneath the semilunar valve(s) in the conal or outlet septum.
	73	VSD, Type 2 (Perimembranous) (Paramembranous) (Conoventricular)	A VSD that is confluent with and involves the membranous septum and is bordered by an atrioventricular valve, not including type 3 VSDs.
	75	VSD, Type 3 (Inlet) (AV canal type)	A VSD that involves the inlet of the right ventricular septum immediately inferior to the AV valve apparatus.

77	VSD, Type 4 (Muscular)	A VSD completely surrounded by muscle.
79	VSD, Type: Gerbode type (LV-RA communication)	A rare form of VSD in which the defect is at the membranous septum; the communication is between the left ventricle and right atrium.
80	VSD, Multiple	More than one VSD exists. Each individual VSD may be coded separately to specify the individual VSD types.
100	AVC (AVSD), Complete (CAVSD)	Indicate if the patient has the diagnosis of “AVC (AVSD), Complete (CAVSD)”. An “AVC (AVSD), Complete (CAVSD)” is a “complete atrioventricular canal” or a “complete atrioventricular septal defect” and occurs in a heart with the phenotypic feature of a common atrioventricular junction. An “AVC (AVSD), Complete (CAVSD)” is defined as an AVC with a common AV valve and both a defect in the atrial septum just above the AV valve (ostium primum ASD [a usually crescent-shaped ASD in the inferior (posterior) portion of the atrial septum just above the AV valve]) and a defect in the ventricular septum just below the AV valve. The AV valve is one valve that bridges both the right and left sides of the heart. Balanced AVC is an AVC with two essentially appropriately sized ventricles. Unbalanced AVC is an AVC defect with two ventricles in which one ventricle is inappropriately small. Such a patient may be thought to be a candidate for biventricular repair, or, alternatively, may be managed as having a functionally univentricular heart. AVC lesions with unbalanced ventricles so severe as to preclude biventricular repair should be classified as single ventricles. Rastelli type A: The common superior (anterior) bridging leaflet is effectively split in two at the septum. The left superior (anterior) leaflet is entirely over the left ventricle and the right superior (anterior) leaflet is similarly entirely over the right ventricle. The division of the common superior (anterior) bridging leaflet into left and right components is caused by extensive attachment of the superior (anterior) bridging leaflet to the crest of the ventricular septum by chordae tendineae. Rastelli type B: Rare, involves anomalous papillary muscle attachment from the right side of the ventricular septum to the left side of the common superior (anterior) bridging leaflet. Rastelli type C: Marked bridging of the ventricular septum by the superior (anterior) bridging leaflet, which floats freely (often termed a "free-floater") over the ventricular septum without chordal attachment to the crest of the ventricular septum.
110	AVC (AVSD), Intermediate (transitional)	An AVC with two distinct left and right AV valve orifices but also with both an ASD just above and a VSD just below the AV valves. While these AV valves in the intermediate form do form two separate orifices they remain abnormal valves. The VSD is often restrictive.
120	AVC (AVSD), Partial (incomplete) (PAVSD) (ASD, primum)	An AVC with an ostium primum ASD (a usually crescent-shaped ASD in the inferior (posterior) portion of the atrial septum just above the AV valve) and varying degrees of malformation of the left AV valve leading to varying degrees of left AV valve regurgitation. No VSD is present.
140	AP window (aortopulmonary window)	Indicate if the patient has the diagnosis of “AP window (aortopulmonary window)”. An “AP window

140		(aortopulmonary window)" is defined as a defect with side-to-side continuity of the lumens of the aorta and pulmonary arterial tree, which is distinguished from common arterial trunk (truncus arteriosus) by the presence of two arterial valves or their atretic remnants. (In other words, an aortopulmonary window is a communication between the main pulmonary artery and ascending aorta in the presence of two separate semilunar [pulmonary and aortic] valves. The presence of two separate semilunar valves distinguishes AP window from truncus arteriosus. Type 1 proximal defect: AP window located just above the sinus of Valsalva, a few millimeters above the semilunar valves, with a superior rim but little inferior rim separating the AP window from the semilunar valves. Type 2 distal defect: AP window located in the uppermost portion of the ascending aorta, with a well-formed inferior rim but little superior rim. Type 3 total defect: AP window involving the majority of the ascending aorta, with little superior and inferior rims. The intermediate type of AP window is similar to the total defect but with adequate superior and inferior rims. In the event of AP window occurring in association with interrupted aortic arch, code "Interrupted aortic arch + AP window (aortopulmonary window)", and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and AP window separately to provide further documentation about the individual interrupted arch and AP window types.)
150	Pulmonary artery origin from ascending aorta (hemitruncus)	One pulmonary artery arises from the ascending aorta and the other pulmonary artery arises from the right ventricle. DOES NOT include origin of the right or left pulmonary artery from the innominate artery or the aortic arch via a patent ductus arteriosus or collateral artery.
160	Truncus arteriosus	Indicate if the patient has the diagnosis of "Truncus arteriosus". A truncus arteriosus is also known as a common arterial trunk and is defined as a heart in which a single arterial trunk arises from the heart, giving origin to the coronary arteries, the pulmonary arteries, and the systemic arterial circulation. In the majority of instances there is a ventricular septal defect and a single semilunar valve which may contain two, three, four, or more leaflets and is occasionally dysplastic. Often, the infundibular septum is virtually absent superiorly. In most instances the truncal valve overrides the true interventricular septum (and thus both ventricles), but very rarely the truncal valve may override the right ventricle entirely. In such instances, there may be no ventricular septal defect or a very small ventricular septal defect, in which case the left ventricle and mitral valve may be extremely hypoplastic.
170	Truncal valve insufficiency	Functional abnormality - insufficiency - of the truncal valve. May be further subdivided into grade of insufficiency (I, II, III, IV or mild, moderate, severe).
2470	Truncal valve stenosis	
2010	Truncus arteriosus + Interrupted aortic arch	Indicate if the patient has the diagnosis of "Truncus arteriosus + Interrupted aortic arch". {A truncus arteriosus is also known as a common arterial trunk and is defined as a heart in which a single arterial trunk arises from the heart,

2010		giving origin to the coronary arteries, the pulmonary arteries, and the systemic arterial circulation. In the majority of instances there is a ventricular septal defect and a single semilunar valve which may contain two, three, four, or more leaflets and is occasionally dysplastic. The infundibular septum is virtually absent superiorly. In most instances the truncal valve overrides the true interventricular septum (and thus both ventricles), but very rarely the truncal valve may override the right ventricle entirely. If in such case there is no ventricular septal defect, then the left ventricle and mitral valve may be extremely hypoplastic.} {Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.}
180	Partial anomalous pulmonary venous connection (PAPVC)	Some, but not all of the pulmonary veins connect to the right atrium or to one or more of its venous tributaries. This definition excludes sinus venosus defects with normally connected but abnormally draining pulmonary veins (the pulmonary veins may drain abnormally into the right atrium via the atrial septal defect).
190	Partial anomalous pulmonary venous connection (PAPVC), scimitar	The right pulmonary vein(s) connect anomalously to the inferior vena cava or to the right atrium at the insertion of the inferior vena cava. The descending vertical vein resembles a scimitar (Turkish sword) on frontal chest x-ray. Frequently associated with: hypoplasia of the right lung with bronchial anomalies; dextroposition and/or dextrorotation of the heart; hypoplasia of the right pulmonary artery; and anomalous subdiaphragmatic systemic arterial supply to the lower lobe of the right lung directly from the aorta or its main branches.
200	Total anomalous pulmonary venous connection (TAPVC), Type 1 (supracardiac)	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium. In Type 1 (supracardiac) TAPVC, the anomalous connection is at the supracardiac level and can be obstructed or nonobstructed.
210	Total anomalous pulmonary venous connection (TAPVC), Type 2 (cardiac)	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium. In Type 2 (cardiac) TAPVC, the anomalous connection is to the heart, either to the right atrium directly or to the coronary sinus. Most patients with type 2 TAPVC are nonobstructed.
220	Total anomalous pulmonary venous connection (TAPVC), Type 3 (infracardiac)	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium. In Type 3 (infracardiac) TAPVC, the anomalous connection is at the infracardiac level (below the diaphragm), with the pulmonary venous return entering the

220		right atrium ultimately via the inferior vena cava. In the vast majority of patients infracardiac TAPVC is obstructed.
230	Total anomalous pulmonary venous connection (TAPVC), Type 4 (mixed)	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium. In Type 4 (mixed) TAPVC, the anomalous connection is at two or more of the above levels (supracardiac, cardiac, infracardiac) and can be obstructed or nonobstructed.
250	Cor triatriatum	In the classic form of cor triatriatum a membrane divides the left atrium (LA) into a posterior accessory chamber that receives the pulmonary veins and an anterior chamber (LA) that communicates with the mitral valve. In differentiating cor triatriatum from supravalvar mitral ring, in cor triatriatum the posterior compartment contains the pulmonary veins while the anterior contains the left atrial appendage and the mitral valve orifice; in supravalvar mitral ring, the anterior compartment contains only the mitral valve orifice. Cor triatriatum dexter (prominent venous valve producing obstruction of the IVC and tricuspid valve) is to be coded as a systemic venous obstruction, not as a form of cor triatriatum.
260	Pulmonary venous stenosis	Any pathologic narrowing of one or more pulmonary veins. Can be further subdivided by etiology (congenital, acquired-postoperative, acquired-nonpostoperative) and extent of stenosis (diffusely hypoplastic, long segment focal/tubular stenosis, discrete stenosis).
2480	Pulmonary venous stenosis, Acquired	
2490	Pulmonary venous stenosis, Spontaneous	
270	Systemic venous anomaly	Anomalies of the systemic venous system (superior vena cava (SVC), inferior vena cava (IVC), brachiocephalic veins (often the innominate vein), azygos vein, coronary sinus, levo-atrial cardinal vein) arising from one or more anomalies of origin, duplication, course, or connection. Examples include abnormal or absent right SVC with LSVC, bilateral SVC, interrupted right or left IVC, azygos continuation of IVC, and anomalies of hepatic drainage. Bilateral SVC may have, among other configurations: 1) RSVC draining to the RA and the LSVC to the LA with completely unroofed coronary sinus, 2) RSVC draining to the RA and LSVC to the coronary sinus which drains (normally) into the RA, or 3) RSVC to the coronary sinus which drains (abnormally) into the LA and LSVC to LA. Anomalies of the inferior vena caval system include, among others: 1) left IVC to LA, 2) biatrial drainage, or 3) interrupted IVC (left or right) with azygos continuation to an LSVC or RSVC.
280	Systemic venous obstruction	Obstruction of the systemic venous system (superior vena cava (SVC), inferior vena cava (IVC), brachiocephalic veins (often the innominate vein), azygos vein, coronary sinus, levo-atrial cardinal vein) arising from congenital or acquired stenosis or occlusion. Cor triatriatum dexter (prominent venous valve producing obstruction of the IVC and tricuspid valve) is to be coded as a systemic venous obstruction, not

280		as a form of cor triatriatum.
290	TOF	Indicate if the patient has the diagnosis of “TOF”. Only use this diagnosis if it is NOT known if the patient has one of the following four more specific diagnoses: (1). “TOF, Pulmonary stenosis”, (2). “TOF, AVC (AVSD)”, (3). “TOF, Absent pulmonary valve”, (4). “Pulmonary atresia, VSD (Including TOF, PA)”, or (5). “Pulmonary atresia, VSD-MAPCA (pseudotruncus)”.{“TOF” is “Tetralogy of Fallot” and is defined as a group of malformations with biventricular atrioventricular alignments or connections characterized by anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta. Hearts with tetralogy of Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, and aortic override; hearts with tetralogy of Fallot will most often have right ventricular hypertrophy.} (An additional, often muscular [Type 4] VSD may be seen with TOF and should be coded separately as a secondary diagnosis as “VSD, Type 4 (Muscular)”. Pulmonary arteries may be diminutive or there may be an absent left or right pulmonary artery; additional coding for pulmonary artery and/or branch pulmonary artery stenoses may be found under RVOT obstruction. Abnormal coronary artery distribution may also be associated with tetralogy of Fallot and may be coded separately under coronary artery anomalies. The presence of associated anomalies such as additional VSD, atrial septal defect, right aortic arch, left superior vena cava, and coronary artery anomalies must be subspecified as an additional or secondary diagnosis under the primary TOF diagnosis. TOF with absent pulmonary valve or TOF with associated complete atrioventricular canal are NOT to be secondary diagnoses under TOF - they are separate entities and should be coded as such. Controversy surrounds the differentiation between TOF and double outlet right ventricle [DORV]; in the nomenclature used here, DORV is defined as a type of ventriculoarterial connection in which both great vessels arise predominantly from the right ventricle. TOF with pulmonary atresia is to be coded under "Pulmonary atresia-VSD.")
2140	TOF, Pulmonary stenosis	Indicate if the patient has the diagnosis of “TOF, Pulmonary stenosis”. Use this diagnosis if the patient has tetralogy of Fallot and pulmonary stenosis. Do not use this diagnosis if the patient has tetralogy of Fallot and pulmonary atresia. Do not use this diagnosis if the patient has tetralogy of Fallot and absent pulmonary valve. Do not use this diagnosis if the patient has tetralogy of Fallot and atrioventricular canal. {Tetralogy of Fallot is defined as a group of malformations with biventricular atrioventricular alignments or connections characterized by anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta. Hearts with tetralogy of Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, and aortic override;

2140		hearts with tetralogy of Fallot will most often have right ventricular hypertrophy. (An additional, often muscular [Type 4] VSD may be seen with TOF and should be coded separately as a secondary diagnosis as “VSD, Type 4 (Muscular)”. Pulmonary arteries may be diminutive or there may be an absent left or right pulmonary artery; additional coding for pulmonary artery and/or branch pulmonary artery stenoses may be found under RVOT obstruction. Abnormal coronary artery distribution may also be associated with tetralogy of Fallot and may be coded separately under coronary artery anomalies. The presence of associated anomalies such as additional VSD, atrial septal defect, right aortic arch, left superior vena cava, and coronary artery anomalies must be subspecified as an additional or secondary diagnosis under the primary TOF diagnosis. TOF with absent pulmonary valve or TOF with associated complete atrioventricular canal are NOT to be secondary diagnoses under TOF - they are separate entities and should be coded as such. Controversy surrounds the differentiation between TOF and double outlet right ventricle [DORV]; in the nomenclature used here, DORV is defined as a type of ventriculoarterial connection in which both great vessels arise predominantly from the right ventricle. TOF with pulmonary atresia is to be coded under "Pulmonary atresia-VSD.")}
300	TOF, AVC (AVSD)	TOF with complete common atrioventricular canal defect is a rare variant of common atrioventricular canal defect with the associated conotruncal abnormality of TOF. The anatomy of the endocardial cushion defect is that of Rastelli type C in almost all cases.
310	TOF, Absent pulmonary valve	Indicate if the patient has the diagnosis of “TOF, Absent pulmonary valve”. “TOF, Absent pulmonary valve” is “Tetralogy of Fallot with Absent pulmonary valve” and is defined as a malformation with all of the morphologic characteristics of tetralogy of Fallot (anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta), in which the ventriculo-arterial junction of the right ventricle with the main pulmonary artery features an atypical valve with rudimentary cusps that lack the anatomical semi-lunar features of normal valve cusps and which functionally do not achieve central coaptation. The physiologic consequence is usually a combination of variable degrees of both stenosis and regurgitation of the pulmonary valve. A developmental accompaniment of this anatomy and physiology is dilatation of the main pulmonary artery and central right and left pulmonary arteries, which when extreme, is associated with abnormal arborization of lobar and segmental pulmonary artery branches and with compression of the trachea and mainstem bronchi. One theory holds that absence of the arterial duct or ductal ligament (which is a nearly constant finding in cases of tetralogy of Fallot with absent pulmonary valve) in combination with pulmonary `valve stenosis and regurgitation, comprise the physiologic conditions which

310		predispose to central pulmonary artery dilatation during fetal development. (Tetralogy of Fallot with Absent Pulmonary Valve Syndrome is a term frequently used to describe the clinical presentation when it features both circulatory alterations and respiratory distress secondary to airway compression.)
320	Pulmonary atresia	Pulmonary atresia defects which do not readily fall into pulmonary atresia-intact ventricular septum or pulmonary atresia-VSD (with or without MAPCAs) categories. These may include complex lesions in which pulmonary atresia is a secondary diagnosis, for example, complex single ventricle malformations with associated pulmonary atresia.
330	Pulmonary atresia, IVS	Pulmonary atresia (PA) and intact ventricular septum (IVS) is a duct-dependent congenital malformation that forms a spectrum of lesions including atresia of the pulmonary valve, a varying degree of right ventricle and tricuspid valve hypoplasia, and anomalies of the coronary circulation. An RV dependent coronary artery circulation is present when coronary artery fistulas (coronary sinusoids) are associated with a proximal coronary artery stenosis. Associated Ebstein's anomaly of the tricuspid valve can be present; the tricuspid diameter is enlarged and the prognosis is poor.
340	Pulmonary atresia, VSD (Including TOF, PA)	Pulmonary atresia (PA) and ventricular septal defect (VSD) is a heterogeneous group of congenital cardiac malformations in which there is lack of luminal continuity and absence of blood flow from either ventricle (in cases with ventriculo-arterial discordance) and the pulmonary artery, in a biventricular heart that has an opening or a hole in the interventricular septum (VSD). The malformation forms a spectrum of lesions including tetralogy of Fallot with pulmonary atresia. Tetralogy of Fallot with PA is a specific type of PA-VSD where the intracardiac malformation is more accurately defined (extreme underdevelopment of the RV infundibulum with marked anterior and leftward displacement of the infundibular septum often fused with the anterior wall of the RV resulting in complete obstruction of blood flow into the pulmonary artery and associated with a large outlet, subaortic ventricular septal defect). In the vast majority of cases of PA-VSD the intracardiac anatomy is that of TOF. The pulmonary circulation in PA-VSD is variable in terms of origin of blood flow, presence or absence of native pulmonary arteries, presence or absence of major aortopulmonary collateral arteries (MAPCA(s)), and distal distribution (pulmonary parenchymal segment arborization) abnormalities. Native pulmonary arteries may be present or absent. If MAPCAs are present this code should not be used; instead, Pulmonary atresia, VSD-MAPCA (pseudotruncus) should be used.
350	Pulmonary atresia, VSD-MAPCA	MAPCA(s) are large and distinct arteries, highly variable in number, that usually arise from the descending thoracic aorta, but uncommonly may originate from the aortic arch or the subclavian, carotid or even the coronary arteries. MAPCA(s) may be associated with present or absent native pulmonary arteries. If present, the native pulmonary arteries may be hypoplastic, and either confluent or nonconfluent. Systemic pulmonary collateral arteries have

350		been categorized into 3 types based on their site of origin and the way they connect to the pulmonary circulation: direct aortopulmonary collaterals, indirect aortopulmonary collaterals, and true bronchial arteries. Only the first two should be considered MAPCA(s). If MAPCA(s) are associated with PA-VSD or TOF, PA this code should be used.
360	MAPCA(s) (major aortopulmonary collateral[s]) (without PA-VSD)	Rarely MAPCA(s) may occur in patents who do not have PA-VSD, but have severe pulmonary stenosis. The intracardiac anatomy in patients who have MAPCA(s) without PA should be specifically coded in each case as well.
370	Ebstein's anomaly	Indicate if the patient has the diagnosis of “Ebstein's anomaly”. Ebstein's anomaly is a malformation of the tricuspid valve and right ventricle that is characterized by a spectrum of several features: (1) incomplete delamination of tricuspid valve leaflets from the myocardium of the right ventricle; (2) downward (apical) displacement of the functional annulus; (3) dilation of the "atrialized" portion of the right ventricle with variable degrees of hypertrophy and thinning of the wall; (4) redundancy, fenestrations, and tethering of the anterior leaflets; and (5) dilation of the right atrioventricular junction (the true tricuspid annulus). These anatomical and functional abnormalities cause tricuspid regurgitation (and rarely tricuspid stenosis) that results in right atrial and right ventricular dilatation and atrial and ventricular arrhythmias. With increasing degrees of anatomic severity of malformation, the fibrous transformation of leaflets from their muscular precursors remains incomplete, with the septal leaflet being most severely involved, the posterior leaflet less severely involved, and the anterior leaflet usually the least severely involved. Associated cardiac anomalies include an interatrial communication, the presence of accessory conduction pathways often associated with Wolff-Parkinson-White syndrome, and dilation of the right atrium and right ventricle in patients with severe Ebstein's anomaly. (Varying degrees of right ventricular outflow tract obstruction may be present, including pulmonary atresia in some cases. Such cases of Ebstein's anomaly with pulmonary atresia should be coded with a Primary Diagnosis of “Ebstein’s anomaly”, and a Secondary Diagnosis of “Pulmonary atresia”.) (Some patients with atrioventricular discordance and ventriculoarterial discordance in situs solitus [congenitally corrected transposition] have an Ebstein-like deformity of the left-sided morphologically tricuspid valve. The nature of the displacement of the septal and posterior leaflets is similar to that in right-sided Ebstein's anomaly in patients with atrioventricular concordance and ventriculoarterial concordance in situs solitus. These patients with “Congenitally corrected TGA” and an Ebstein-like deformity of the left-sided morphologically tricuspid valve should be coded with a Primary Diagnosis of “Congenitally corrected TGA”, and a Secondary Diagnosis of “Ebstein's anomaly”.)
380	Tricuspid regurgitation, non-Ebstein's related	Non-Ebstein's tricuspid regurgitation may be due to congenital factors (primary annular dilation, prolapse, leaflet underdevelopment, absent papillary

380		muscle/chordae) or acquired (post cardiac surgery or secondary to rheumatic fever, endocarditis, trauma, tumor, cardiomyopathy, iatrogenic or other causes).
390	Tricuspid stenosis	Tricuspid stenosis may be due to congenital factors (valvar hypoplasia, abnormal subvalvar apparatus, double-orifice valve, parachute deformity) or acquired (post cardiac surgery or secondary to carcinoid, rheumatic fever, tumor, systemic disease, iatrogenic, or other causes).
400	Tricuspid regurgitation and tricuspid stenosis	Tricuspid regurgitation present with tricuspid stenosis may be due to congenital factors or acquired.
410	Tricuspid valve, Other	Tricuspid valve pathology not otherwise specified in diagnosis definitions 370, 380, 390 and 400.
420	Pulmonary stenosis, Valvar	Pulmonary stenosis, Valvar ranges from critical neonatal pulmonic valve stenosis with hypoplasia of the right ventricle to valvar pulmonary stenosis in the infant, child, or adult, usually better tolerated but potentially associated with infundibular stenosis. Pulmonary branch hypoplasia can be associated. Only 10% of neonates with Pulmonary stenosis, Valvar with intact ventricular septum have RV-to-coronary artery fistula(s). An RV dependent coronary artery circulation is present when coronary artery fistulas (coronary sinusoids) are associated with a proximal coronary artery stenosis; this occurs in only 2% of neonates with Pulmonary stenosis, Valvar with IVS.
430	Pulmonary artery stenosis (hypoplasia), Main (trunk)	Indicate if the patient has the diagnosis of “Pulmonary artery stenosis (hypoplasia), Main (trunk)”. “Pulmonary artery stenosis (hypoplasia), Main (trunk)” is defined as a congenital or acquired anomaly with pulmonary trunk (main pulmonary artery) narrowing or hypoplasia. The stenosis or hypoplasia may be isolated or associated with other cardiac lesions. Since the narrowing is distal to the pulmonic valve, it may also be known as supravalvar pulmonary stenosis.
440	Pulmonary artery stenosis, Branch, Central (within the hilar bifurcation)	Indicate if the patient has the diagnosis of “Pulmonary artery stenosis, Branch, Central (within the hilar bifurcation)”. “Pulmonary artery stenosis, Branch, Central (within the hilar bifurcation)” is defined as a congenital or acquired anomaly with central pulmonary artery branch (within the hilar bifurcation involving the right or left pulmonary artery, or both) narrowing or hypoplasia. The stenosis or hypoplasia may be isolated or associated with other cardiac lesions. Coarctation of the pulmonary artery is related to abnormal extension of the ductus arteriosus into a pulmonary branch, more frequently the left branch.
450	Pulmonary artery stenosis, Branch, Peripheral (at or beyond the hilar bifurcation)	Indicate if the patient has the diagnosis of “Pulmonary artery stenosis, Branch, Peripheral (at or beyond the hilar bifurcation)”. “Pulmonary artery stenosis, Branch, Peripheral (at or beyond the hilar bifurcation)” is defined as a congenital or acquired anomaly with peripheral pulmonary artery narrowing or hypoplasia (at or beyond the hilar bifurcation). The stenosis or hypoplasia may be isolated or associated with other cardiac lesions.
470	Pulmonary artery, Discontinuous	Indicate if the patient has the diagnosis of “Pulmonary artery, Discontinuous”. Pulmonary artery, Discontinuous” is defined as a congenital or acquired anomaly with

470		discontinuity between the branch pulmonary arteries or between a branch pulmonary artery and the main pulmonary artery trunk.
490	Pulmonary stenosis, Subvalvar	Subvalvar (infundibular) pulmonary stenosis is a narrowing of the outflow tract of the right ventricle below the pulmonic valve. It may be due to a localized fibrous diaphragm just below the valve, an obstructing muscle bundle or to a long narrow fibromuscular channel.
500	DCRV	The double chambered right ventricle is characterized by a low infundibular (subvalvar) stenosis rather than the rare isolated infundibular stenosis that develops more superiorly in the infundibulum, and is often associated with one or several closing VSDs. In some cases, the VSD is already closed. The stenosis creates two chambers in the RV, one inferior including the inlet and trabecular portions of the RV and one superior including the infundibulum.
510	Pulmonary valve, Other	Other anomalies of the pulmonary valve may be listed here including but not restricted to absent pulmonary valve.
530	Pulmonary insufficiency	Pulmonary valve insufficiency or regurgitation may be due to congenital factors (primary annular dilation, prolapse, leaflet underdevelopment, etc.) or acquired (for example, post cardiac surgery for repair of tetralogy of Fallot, etc.).
540	Pulmonary insufficiency and pulmonary stenosis	Pulmonary valve insufficiency and pulmonary stenosis beyond the neonatal period, in infancy and childhood, may be secondary to leaflet tissue that has become thickened and myxomatous. Retraction of the commissure attachment frequently creates an associated supravalvar stenosis.
2130	Shunt failure	Indicate if the patient has the diagnosis of “Shunt failure”. This diagnostic subgroup includes failure of any of a variety of shunts (“Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS)”, “Shunt, Systemic to pulmonary, Central (from aorta or to main pulmonary artery)”, “Shunt, Systemic to pulmonary, Other”, and “Sano Shunt”), secondary to any of the following etiologies: shunt thrombosis, shunt occlusion, shunt stenosis, shunt obstruction, and shunt outgrowth. This diagnosis (“Shunt failure”) would be the primary diagnosis in a patient with, for example, “Hypoplastic left heart syndrome (HLHS)” who underwent a “Norwood procedure” with a “Modified Blalock-Taussig Shunt” and now requires reoperation for thrombosis of the “Modified Blalock-Taussig Shunt”. The underlying or fundamental diagnosis in this patient is “Hypoplastic left heart syndrome (HLHS)”, but the primary diagnosis for the operation to be performed to treat the thrombosis of the “Modified Blalock-Taussig Shunt” would be “Shunt failure”. Please note that the choice “2130 Shunt failure” does not include “520 Conduit failure”.
520	Conduit failure	Indicate if the patient has the diagnosis of “Conduit failure”. This diagnostic subgroup includes failure of any of a variety of conduits (ventricular [right or left]-to-PA conduits, as well as a variety of other types of conduits [ventricular {right or left}-to-aorta, RA-to-RV, etc.]),

520		secondary to any of the following etiologies: conduit outgrowth, obstruction, stenosis, insufficiency, or insufficiency and stenosis. This diagnosis ("Conduit failure") would be the primary diagnosis in a patient with, for example, "Truncus arteriosus" repaired in infancy who years later is hospitalized because of conduit stenosis/insufficiency. The underlying or fundamental diagnosis in this patient is "Truncus arteriosus", but the primary diagnosis for the operation to be performed during the hospitalization (in this case, "Conduit reoperation") would be "Conduit failure". Please note that the choice "520 Conduit failure" does not include "2130 Shunt failure".
550	Aortic stenosis, Subvalvar	Subaortic obstruction can be caused by different lesions: subaortic membrane or tunnel, accessory mitral valve tissue, abnormal insertion of the mitral anterior leaflet to the ventricular septum, deviation of the outlet septum (seen in coarctation of the aorta and interrupted aortic arch), or a restrictive bulboventricular foramen in single ventricle complexes. The Shone complex consists of subvalvar aortic stenosis in association with supravalvar mitral ring, parachute mitral valve, and coarctation of aorta. Subvalvar aortic stenosis may be categorized into two types: localized subvalvar aortic stenosis, which consists of a fibrous or fibromuscular ridge, and diffuse tunnel subvalvar aortic stenosis, in which circumferential narrowing commences at the annular level and extends downward for 1-3 cm. Idiopathic hypertrophic subaortic stenosis (IHSS) is also known as hypertrophic obstructive cardiomyopathy (HOCM), and is characterized by a primary hypertrophy of the myocardium. The obstructive forms involve different degrees of dynamic subvalvar aortic obstruction from a thickened ventricular wall and anterior motion of the mitral valve. Definitive nomenclature and therapeutic options for IHSS are listed under cardiomyopathy.
2500	Aortic Stenosis, Subvalvar, Discrete	
2510	Aortic Stenosis, Subvalvar, IHSS	
2520	Aortic Stenosis, Subvalvar, Tunnel-like	
560	Aortic stenosis, Valvar	Valvar aortic stenosis may be congenital or acquired. In its congenital form there are two types: critical (infantile), seen in the newborn in whom systemic perfusion depends on a patent ductus arteriosus, and noncritical, seen in infancy or later. Acquired valvar stenosis may be seen after as a result of rheumatic valvar disease, or from stenotic changes of an aortic valve prosthesis. Congenital valvar stenosis may result: (1) from complete fusion of commissures (acommissural) that results in a dome-shaped valve with a pinpoint opening (seen most commonly in infants with critical aortic valve stenosis); (2) from a unicommissural valve with one defined commissure and eccentric orifice (often with two raphes radiating from the ostium indicating underdeveloped commissures of a tricuspid aortic valve); (3) from a bicuspid aortic valve, with

560		leaflets that can be equal in size or discrepant, and in left-right or anterior-posterior position; and finally (4) from a dysplastic tricuspid valve, which may have a gelatinous appearance with thick rarely equal in size leaflets, often obscuring the commissures. The dysplastic, tricuspid or bicuspid form of aortic valve deformity may not be initially obstructive but may become stenotic later in life due to leaflet thickening and calcification.
570	Aortic stenosis, Supravalvar	Congenital supravalvar aortic stenosis is described as three forms: an hourglass deformity, a fibrous membrane, and a diffuse narrowing of the ascending aorta. The disease can be inherited as an autosomal dominant trait or part of Williams-Beuren syndrome in association with mental retardation, elfin facies, failure to thrive, and occasionally infantile hypercalcemia. Supravalvar aortic stenosis may involve the coronary artery ostia, and the aortic leaflets may be tethered. The coronary arteries can become tortuous and dilated due to elevated pressures and early atherosclerosis may ensue. Supravalvar aortic stenosis may also be acquired: (1) after a neoaortic reconstruction such as arterial switch, Ross operation, or Norwood procedure; (2) at a suture line from a previous aortotomy or cannulation; and (3) from a narrowed conduit.
590	Aortic valve atresia	Aortic valve atresia will most often be coded under the Hypoplastic left heart syndrome/complex diagnostic codes since it most often occurs as part of a spectrum of cardiac malformations. However, there is a small subset of patients with aortic valve atresia who have a well-developed left ventricle and mitral valve and a large VSD (nonrestrictive or restrictive). The diagnostic code "Aortic valve atresia" enables users to report those patients with aortic valve atresia and a well-developed systemic ventricle without recourse to either a hypoplastic left heart syndrome/complex diagnosis or a single ventricle diagnosis.
600	Aortic insufficiency	Congenital aortic regurgitation/insufficiency is rare as an isolated entity. There are rare reports of congenital malformation of the aortic valve that result in aortic insufficiency shortly after birth from an absent or underdeveloped aortic valve cusp. Aortic insufficiency is more commonly seen with other associated cardiac anomalies: (1) in stenotic aortic valves (commonly stenotic congenital bicuspid aortic valves) with some degree of aortic regurgitation due to aortic leaflet abnormality; (2) in association with a VSD (especially in supracristal or conal type I VSD, more commonly seen in Asian populations); (3) secondary to aortic-left ventricular tunnel; (4) secondary to tethering or retraction of aortic valve leaflets in cases of supravalvar aortic stenosis that may involve the aortic valve; and similarly (5) secondary to encroachment on an aortic cusp by a subaortic membrane; or (6) turbulence caused by a stenotic jet can create progressive aortic regurgitation. Aortic insufficiency may also result from: (1) post-procedure such as closed or open valvotomy or aortic valve repair, VSD closure, balloon valvotomy, or diagnostic catheterization; (2) in the neo-aorta post arterial switch, pulmonary autograft (Ross) procedure, homograft placement,

600		Norwood procedure, or Damus-Kaye-Stansel procedure; (3) as a result of endocarditis secondary to perforated or prolapsed leaflets or annular dehiscence; (4) secondary to annulo-aortic ectasia with prolapsed or noncoapting leaflets; (5) secondary to trauma, blunt or penetrating; or (6) as a result of aortitis, bacterial, viral or autoimmune. Aortic regurgitation secondary to prosthetic failure should be coded first as either conduit failure or prosthetic valve failure, as applicable, and secondarily as aortic regurgitation secondary to prosthetic failure (perivalvar or due to structural failure). The underlying fundamental diagnosis that led to the initial conduit or valve prosthesis placement should also be described.
610	Aortic insufficiency and aortic stenosis	Aortic insufficiency is often seen in association with stenotic aortic valve, commonly the stenotic congenital bicuspid aortic valve. The degree of aortic regurgitation is due to the severity of the aortic leaflet abnormality.
620	Aortic valve, Other	This diagnostic subgroup may be used to delineate aortic valve cusp number (unicuspid, bicuspid, tricuspid, more than three cusps), commissural fusion (normal, partially fused, completely fused), and valve leaflet (normal, thickened, dysplastic, calcified, gelatinous), annulus (normal, hypoplastic, calcified), or sinus description (normal, dilated). Note that any extensive descriptors chosen within those made available by a vendor will be converted, at harvest, to Aortic valve, Other.
630	Sinus of Valsalva aneurysm	The sinus of Valsalva is defined as that portion of the aortic root between the aortic root annulus and the sinotubular ridge. A congenital sinus of Valsalva aneurysm is a dilation usually of a single sinus of Valsalva. These most commonly originate from the right sinus (65%-85%), less commonly from the noncoronary sinus (10%-30%), and rarely from the left sinus (<5%). A true sinus of Valsalva aneurysm presents above the aortic annulus. The hierarchical coding system distinguishes between congenital versus acquired, ruptured versus nonruptured, sinus of origin, and chamber/site of penetration (right atrium, right ventricle, left atrium, left ventricle, pulmonary artery, pericardium). A nonruptured congenital sinus of Valsalva aneurysm may vary from a mild dilation of a single aortic sinus to an extensive windsock deformity. Rupture of a congenital sinus of Valsalva aneurysm into an adjacent chamber occurs most commonly between the ages of 15-30 years. Rupture may occur spontaneously, after trauma, after strenuous physical exertion, or from acute bacterial endocarditis. Congenital etiology is supported by the frequent association of sinus of Valsalva aneurysms with VSDs. Other disease processes are also associated with sinus of Valsalva aneurysm and include: syphilis, endocarditis, cystic medial necrosis, atherosclerosis, and trauma. Acquired sinus of Valsalva aneurysms more frequently involve multiple sinuses of Valsalva; when present in multiple form they are more appropriately classified as aneurysms of the aortic root.
640	LV to aorta tunnel	The aortico-left ventricular tunnel (LV-to-aorta tunnel) is an abnormal paravalvular (alongside or in the vicinity of a valve) communication between the aorta and left ventricle,

640		commonly divided into 4 types: (1) type I, a simple tunnel with a slit-like opening at the aortic end and no aortic valve distortion; (2) type II, a large extracardiac aortic wall aneurysm of the tunnel with an oval opening at the aortic end, with or without ventricular distortion; (3) type III, intracardiac aneurysm of the septal portion of the tunnel, with or without right ventricular outflow obstruction; and (4) type IV, a combination of types II and III. Further differentiation within these types may be notation of right coronary artery arising from the wall of the tunnel. If a LV-to-aorta tunnel communicates with the right ventricle, many feel that the defect is really a ruptured sinus of Valsalva aneurysm.
650	Mitral stenosis, Supravalvar mitral ring	Supravalvar mitral ring is formed by a circumferential ridge of tissue that is attached to the anterior mitral valve leaflet (also known as the aortic leaflet) slightly below its insertion on the annulus and to the atrium slightly above the attachment of the posterior mitral valve leaflet (also known as the mural leaflet). Depending on the diameter of the ring orifice, varying degrees of obstruction exist. The underlying valve is usually abnormal and frequently stenotic or hypoplastic. Supravalvar mitral ring is commonly associated with other stenotic lesions such as parachute or hammock valve (subvalvar stenosis), papillary muscle fusion (subvalvar stenosis), and double orifice mitral valve (valvar stenosis). Differentiation from cor triatriatum focuses on the compartments created by the supravalvar ring. In cor triatriatum the posterior compartment contains the pulmonary veins; the anterior contains the left atrial appendage and the mitral valve orifice. In supravalvar mitral ring, the posterior compartment contains the pulmonary veins and the left atrial appendage; the anterior compartment contains only the mitral valve orifice. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
660	Mitral stenosis, Valvar	Valvar mitral stenosis may arise from congenital (annular and / or leaflet) or acquired causes, both surgical (after mitral valve repair or replacement or other cardiac surgery) and non-surgical (post rheumatic heart disease, infective endocarditis, ischemia, myxomatous degeneration, trauma, or cardiomyopathy). Mitral valve annular hypoplasia is distinguished from severe mitral valve hypoplasia and mitral valve atresia, which are typically components of hypoplastic left heart syndrome. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
670	Mitral stenosis, Subvalvar	Congenital subvalvar mitral stenosis may be due to obstructive pathology of either the chordae tendineae and / or papillary muscles which support the valve leaflets. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.

680	Mitral stenosis, Subvalvar, Parachute	In parachute mitral valve, all chordae are attached to a single papillary muscle originating from the posterior ventricular wall. When the interchordal spaces are partially obliterated valvar stenosis results. This defect also causes valvar insufficiency, most commonly due to a cleft leaflet, a poorly developed anterior leaflet, short chordae, or annular dilatation. This lesion is also part of Shone's anomaly, which consists of the parachute mitral valve, supravalvar mitral ring, subaortic stenosis, and coarctation of the aorta. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
695	Mitral stenosis	Stenotic lesions of the mitral valve not otherwise specified in the diagnosis definitions 650, 660, 670, and 680.
700	Mitral regurgitation and mitral stenosis	Mitral regurgitation and mitral stenosis may arise from congenital or acquired causes or after cardiac surgery. Additional details to aid in coding specific components of the diagnosis are available in the individual mitral stenosis or mitral regurgitation field definitions. When coding multiple mitral valve lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
710	Mitral regurgitation	Mitral regurgitation may arise from congenital (at the annular, leaflet or subvalvar level) or acquired causes both surgical (after mitral valve repair or replacement, subaortic stenosis repair, atrioventricular canal repair, cardiac transplantation, or other cardiac surgery) and non-surgical (post rheumatic heart disease, infective endocarditis, ischemia (with chordal rupture or papillary muscle infarct), myxomatous degeneration including Barlow's syndrome, trauma, or cardiomyopathy). Congenital lesions at the annular level include annular dilatation or deformation (usually deformation is consequent to associated lesions). At the valve leaflet level, mitral regurgitation may be due to a cleft, hypoplasia or agenesis of leaflet(s), excessive leaflet tissue, or a double orifice valve. At the subvalvar level, mitral regurgitation may be secondary to chordae tendineae anomalies (agenesis, rupture, elongation, or shortening as in funnel valve), or to papillary muscle anomalies (hypoplasia or agenesis, shortening, elongation, single-parachute, or multiple-hammock valve). When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
720	Mitral valve, Other	Mitral valve pathology not otherwise coded in diagnosis definitions 650 through 710.
730	Hypoplastic left heart syndrome (HLHS)	Hypoplastic left heart syndrome (HLHS) is a spectrum of cardiac malformations characterized by a severe underdevelopment of the left heart-aorta complex, consisting of aortic and/or mitral valve atresia, stenosis, or hypoplasia with marked hypoplasia or absence of the left ventricle, and hypoplasia of the ascending aorta and of the aortic arch with coarctation of the aorta. Hypoplastic left

730		heart complex is a subset of patients at the favorable end of the spectrum of HLHS characterized by hypoplasia of the structures of the left heart-aorta complex, consisting of aortic and mitral valve hypoplasia without valve stenosis or atresia, hypoplasia of the left ventricle, hypoplasia of the left ventricular outflow tract, hypoplasia of the ascending aorta and of the aortic arch, with or without coarctation of the aorta.
2080	Shone's syndrome	Shone's syndrome is a syndrome of multilevel hypoplasia and obstruction of left sided cardiovascular structures including more than one of the following lesions: (1) supravalvar ring of the left atrium, (2) a parachute deformity of the mitral valve, (3) subaortic stenosis, and (4) aortic coarctation. The syndrome is based on the original report from Shone [1] that was based on analysis of 8 autopsied cases and described the tendency of these four obstructive, or potentially obstructive, conditions to coexist. Only 2 of the 8 cases exhibited all four conditions, with the other cases exhibiting only two or three of the anomalies [2]. [1] Shone JD, Sellers RD, Anderson RG, Adams P, Lillehei CW, Edwards JE. The developmental complex of "parachute mitral valve", supravalvar ring of left atrium, subaortic stenosis, and coarctation of the aorta. Am J Cardiol 1963; 11: 714–725. [2]. Tchervenkov CI, Jacobs JP, Weinberg PM, Aiello VD, Beland MJ, Colan SD, Elliott MJ, Franklin RC, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G. The nomenclature, definition and classification of hypoplastic left heart syndrome. Cardiology in the Young, 2006; 16(4): 339–368, August 2006. Please note that the term "2080 Shone's syndrome" may be the "Fundamental Diagnosis" of a patient; however, the term "2080 Shone's syndrome" may not be the "Primary Diagnosis" of an operation. The term "2080 Shone's syndrome" may be a "Secondary Diagnosis" of an operation.
740	Cardiomyopathy (including dilated, restrictive, and hypertrophic)	Cardiomyopathy is a term applied to a wide spectrum of cardiac diseases in which the predominant feature is poor myocardial function in the absence of any anatomic abnormalities. Cardiomyopathies can be divided into three relatively easily distinguishable entities: (1) dilated, characterized by ventricular dilatation and systolic dysfunction; (2) hypertrophic, characterized by physiologically inappropriate hypertrophy of the left ventricle; and (3) restrictive, characterized by diastolic dysfunction, with a presentation often identical to constrictive pericarditis. Also included in this diagnostic category are patients with a cardiomyopathy or syndrome confined to the right ventricle, for example: (1) arrhythmogenic right ventricular dysplasia; (2) Uhl's syndrome (hypoplasia of right ventricular myocardium, parchment heart); or (3) spongiform cardiomyopathy.
750	Cardiomyopathy, End-stage congenital heart disease	Myocardial abnormality in which there is systolic and/or diastolic dysfunction in the presence of structural congenital heart disease without any (or any further) surgically correctable lesions.
760	Pericardial effusion	Inflammatory stimulation of the pericardium that results in

760		the accumulation of appreciable amounts of pericardial fluid (also known as effusive pericarditis). The effusion may be idiopathic or acquired (e.g., postoperative, infectious, uremic, neoplastic, traumatic, drug-induced).
770	Pericarditis	Inflammatory process of the pericardium that leads to either (1) effusive pericarditis with accumulation of appreciable amounts of pericardial fluid or (2) constrictive pericarditis that leads to pericardial thickening and compression of the cardiac chambers, ultimately with an associated significant reduction in cardiac function. Etiologies are varied and include idiopathic or acquired (e.g., postoperative, infectious, uremic, neoplastic, traumatic, drug-induced) pericarditis.
780	Pericardial disease, Other	A structural or functional abnormality of the visceral or parietal pericardium that may, or may not, have a significant impact on cardiac function. Included are absence or partial defects of the pericardium.
790	Single ventricle, DILV	A congenital cardiac malformation in which both atria connect to a single, morphologically left ventricle. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH

790		(editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
800	Single ventricle, DIRV	A congenital cardiac malformation in which both atria connect to a single, morphologically right ventricle The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
810	Single ventricle, Mitral atresia	A congenital cardiac malformation in which there is no orifice of mitral valve The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary

810		circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
820	Single ventricle, Tricuspid atresia	A congenital cardiac malformation in which there is no orifice of tricuspid valve. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart".

820		Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
830	Single ventricle, Unbalanced AV canal	Single ventricle anomalies with a common atrioventricular (AV) valve and only one completely well developed ventricle. If the common AV valve opens predominantly into the morphologic left ventricle, the defect is termed a left ventricular (LV)–type or LV-dominant AV septal defect. If the common AV valve opens predominantly into the morphologic right ventricle, the defect is termed a right ventricular (RV)–type or RV-dominant AV septal defect. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart".
840	Single ventricle, Heterotaxia syndrome	"Heterotaxia syndrome" is synonymous with "heterotaxy", "visceral heterotaxy", and "heterotaxy syndrome". Heterotaxy is defined as an abnormality where the internal thoraco-abdominal organs demonstrate abnormal arrangement across the left-right axis of the body. By convention, heterotaxy does not include patients with either the expected usual or normal arrangement of the internal organs along the left-right axis, also known as ‘situs solitus’, nor patients with complete mirror-imaged arrangement of the internal organs along the left-right axis also known as ‘situs inversus’.

840		The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
850	Single ventricle, Other	If the single ventricle is of primitive or indeterminate type, other is chosen in coding. It is recognized that a considerable variety of other structural cardiac malformations (e.g., biventricular hearts with straddling atrioventricular valves, pulmonary atresia with intact ventricular septum, some complex forms of double outlet right ventricle) may at times be best managed in a fashion similar to that which is used to treat univentricular hearts. They are not to be coded in this section of the nomenclature, but according to the underlying lesions. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a

850		spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
851	Single Ventricle + Total anomalous pulmonary venous connection (TAPVC)	Indicate if the patient has the diagnosis of "Single Ventricle + Total anomalous pulmonary venous connection (TAPVC)". In the event of Single Ventricle occurring in association with Total anomalous pulmonary venous connection (TAPVC), code "Single Ventricle + Total anomalous pulmonary venous connection (TAPVC)", and then use additional (secondary) diagnostic codes to describe the Single Ventricle and the Total anomalous pulmonary venous connection (TAPVC) separately to provide further documentation about the Single Ventricle and Total anomalous pulmonary venous connection (TAPVC) types. {"Total anomalous pulmonary venous connection (TAPVC)" is defined as a heart where all of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium.} The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the

851		systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
870	Congenitally corrected TGA	Indicate if the patient has the diagnosis of “Congenitally corrected TGA”. Congenitally corrected transposition is synonymous with the terms ‘corrected transposition’ and ‘discordant atrioventricular connections with discordant ventriculo-arterial connections’, and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.
872	Congenitally corrected TGA, IVS	Indicate if the patient has the diagnosis of “Congenitally corrected TGA, IVS”. “Congenitally corrected TGA, IVS” is “Congenitally corrected transposition with an intact ventricular septum”, in other words, “Congenitally corrected transposition with no VSD”. (Congenitally corrected transposition is synonymous with the terms ‘corrected transposition’ and ‘discordant atrioventricular connections with discordant ventriculo-arterial connections’, and is defined as a spectrum of cardiac

872		malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)
874	Congenitally corrected TGA, IVS-LVOTO	Indicate if the patient has the diagnosis of "Congenitally corrected TGA, IVS-LVOTO". "Congenitally corrected TGA, IVS-LVOTO" is "Congenitally corrected transposition with an intact ventricular septum and left ventricular outflow tract obstruction", in other words, "Congenitally corrected transposition with left ventricular outflow tract obstruction and no VSD". (Congenitally corrected transposition is synonymous with the terms 'corrected transposition' and 'discordant atrioventricular connections with discordant ventriculo-arterial connections', and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)
876	Congenitally corrected TGA, VSD	Indicate if the patient has the diagnosis of "Congenitally corrected TGA, VSD". "Congenitally corrected TGA, VSD" is "Congenitally corrected transposition with a VSD". (Congenitally corrected transposition is synonymous with the terms 'corrected transposition' and 'discordant atrioventricular connections with discordant ventriculo-arterial connections', and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and

876		Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)
878	Congenitally corrected TGA, VSD-LVOTO	Indicate if the patient has the diagnosis of "Congenitally corrected TGA, VSD-LVOTO". "Congenitally corrected TGA, VSD-LVOTO" is "Congenitally corrected transposition with a VSD and left ventricular outflow tract obstruction". (Congenitally corrected transposition is synonymous with the terms 'corrected transposition' and 'discordant atrioventricular connections with discordant ventriculo-arterial connections', and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)
880	TGA, IVS	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with an intact ventricular septum. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
890	TGA, IVS-LVOTO	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with an intact ventricular septum and associated left ventricular obstruction. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
900	TGA, VSD	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with one or more ventricular septal defects.

900		There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
910	TGA, VSD-LVOTO	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with one or more ventricular septal defects and left ventricular outflow tract obstruction. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
930	DORV, VSD type	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In double outlet right ventricle, VSD type, there is an associated subaortic or doubly-committed VSD and no pulmonary outflow tract obstruction. Subaortic VSD's are located beneath the aortic valve. Doubly-committed VSD's lie beneath the leaflets of the aortic and pulmonary valves (juxtaarterial). In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
940	DORV, TOF type	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In double outlet right ventricle, TOF type, there is an associated subaortic or doubly-committed VSD and pulmonary outflow tract obstruction. Subaortic VSD's are located beneath the aortic valve. Doubly-committed VSD's lie beneath the leaflets of the aortic and pulmonary valves (juxtaarterial). DORV can occur in association with pulmonary atresia, keeping in mind in coding that in the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles (in this situation DORV is coded as a primary diagnosis). Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate

940		Single ventricle listing.
950	DORV, TGA type	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In double outlet right ventricle, TGA type, there is an associated subpulmonary VSD. Most frequently, there is no pulmonary outflow tract obstruction (Taussig-Bing heart). The aorta is usually to the right and slightly anterior to or side-by-side with the pulmonary artery. Associated aortic outflow tract stenosis (subaortic, aortic arch obstruction) is commonly associated with the Taussig-Bing heart and if present should be coded as a secondary diagnosis. Rarely, there is associated pulmonary outflow tract obstruction. In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
960	DORV, Remote VSD (uncommitted VSD)	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In double outlet right ventricle, Remote VSD type, there is a remote or noncommitted VSD. The VSD is far removed from both the aortic and pulmonary valves, usually within the inlet septum. Many of these VSD's are in hearts with DORV and common atrioventricular canal/septal defect. In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
2030	DORV + AVSD (AV Canal)	Indicate if the patient has the diagnosis of “DORV + AVSD (AV Canal)”. In the event of DORV occurring in association with AVSD (AV Canal), code “DORV + AVSD (AV Canal)”, and then use additional (secondary) diagnostic codes to describe the DORV and the AVSD (AV Canal) separately to provide further documentation about the DORV and AVSD (AV Canal) types. {“DORV” is “Double outlet right ventricle” and is defined as a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle.} In this case, the DORV exists in combination with an atrioventricular septal defect and common atrioventricular junction guarded by a common atrioventricular valve.
975	DORV, IVS	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In the rare case of double outlet right ventricle with IVS the ventricular septum is intact. In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant

975		atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connections with DORV are to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
980	DOLV	Double outlet left ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the left ventricle. In the nomenclature developed for DOLV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DOLV is to be coded under congenitally corrected TGA. DOLV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
990	Coarctation of aorta	Indicate if the patient has the diagnosis of “Coarctation of aorta”. A “Coarctation of the aorta” generally indicates a narrowing of the descending thoracic aorta just distal to the left subclavian artery. However, the term may also be accurately used to refer to a region of narrowing anywhere in the thoracic or abdominal aorta.
1000	Aortic arch hypoplasia	Hypoplasia of the aortic arch is hypoplasia of the proximal or distal transverse arch or the aortic isthmus. The isthmus (arch between the left subclavian and insertion of the patent ductus arteriosus / ligamentum arteriosum) is hypoplastic if its diameter is less than 40% of the diameter of the ascending aorta. The proximal transverse arch (arch between the innominate and left carotid arteries) and distal transverse arch (arch between the left carotid and left subclavian arteries) are hypoplastic if their diameters are less than 60% and 50%, respectively, of the diameter of the ascending aorta.
92	VSD + Aortic arch hypoplasia	A ventricular septal defect, any type, associated with hypoplasia of the aortic arch. (See diagnosis definition 1000 for a definition of hypoplasia of the aortic arch.)
94	VSD + Coarctation of aorta	Indicate if the patient has the diagnosis of “VSD + Coarctation of aorta”. In the event of a VSD occurring in association with Coarctation of aorta, code “VSD + Coarctation of aorta”, and then use additional (secondary) diagnostic codes to describe the VSD and the Coarctation of aorta separately to provide further documentation about the individual VSD and Coarctation of aorta types. {A "VSD" is a "Ventricular Septal Defect" and is also known as an "Interventricular communication". A VSD is defined as "a hole between the ventricular chambers or their remnants". (The VSD is defined on the basis of its margins as seen from the aspect of the morphologically right ventricle. In the setting of double outlet right ventricle, the defect provides the outflow from the morphologically left ventricle. In univentricular atrioventricular connections with functionally single left ventricle with an outflow chamber, the communication is referred to by some as a bulboventricular foramen.)} {A “Coarctation of the aorta” generally indicates

94		a narrowing of the descending thoracic aorta just distal to the left subclavian artery. However, the term may also be accurately used to refer to a region of narrowing anywhere in the thoracic or abdominal aorta.}
1010	Coronary artery anomaly, Anomalous aortic origin of coronary artery (AAOCA)	Anomalous aortic origins of the coronary arteries include a spectrum of anatomic variations of the normal coronary artery origins. Coronary artery anomalies of aortic origin to be coded under this diagnostic field include: anomalies of take-off (high take-off), origin (sinus), branching, and number. An anomalous course of the coronary artery vessels is also significant, particularly those coronary arteries that arise or course between the great vessels.
1020	Coronary artery anomaly, Anomalous pulmonary origin (includes ALCAPA)	In patients with anomalous pulmonary origin of the coronary artery, the coronary artery (most commonly the left coronary artery) arises from the pulmonary artery rather than from the aorta. Rarely, the right coronary artery, the circumflex, or both coronary arteries may arise from the pulmonary artery.
1030	Coronary artery anomaly, Fistula	The most common of coronary artery anomalies, a coronary arteriovenous fistula is a communication between a coronary artery and either a chamber of the heart (coronary-cameral fistula) or any segment of the systemic or pulmonary circulation (coronary arteriovenous fistula). They may be congenital or acquired (traumatic, infectious, iatrogenic) in origin, and are mostly commonly seen singly, but occasionally multiple fistulas are present. Nomenclature schemes have been developed that further categorize the fistulas by vessel of origin and chamber of termination, and one angiographic classification scheme by Sakakibara has surgical implications. Coronary artery fistulas can be associated with other congenital heart anomalies such as tetralogy of Fallot, atrial septal defect, ventricular septal defect, and pulmonary atresia with intact ventricular septum, among others. The major cardiac defect should be listed as the primary diagnosis and the coronary artery fistula should be as an additional secondary diagnoses.
1040	Coronary artery anomaly, Aneurysm	Coronary artery aneurysms are defined as dilations of a coronary vessel 1.5 times the adjacent normal coronaries. There are two forms, saccular and fusiform (most common), and both may be single or multiple. These aneurysms may be congenital or acquired (atherosclerotic, Kawasaki, systemic diseases other than Kawasaki, iatrogenic, infectious, or traumatic) in origin.
2420	Coronary artery anomaly, Ostial Atresia	
1050	Coronary artery anomaly, Other	Coronary artery anomalies which may fall within this category include coronary artery bridging and coronary artery stenosis, as well as secondary coronary artery variations seen in congenital heart defects such as tetralogy of Fallot, transposition of the great arteries, and truncus arteriosus (with the exception of variations that can be addressed by a more specific coronary artery anomaly code).
1070	Interrupted aortic arch	Indicate if the patient has the diagnosis of “Interrupted aortic arch”. Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending

1070		aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.
2020	Interrupted aortic arch + VSD	Indicate if the patient has the diagnosis of “Interrupted aortic arch + VSD”. In the event of interrupted aortic arch occurring in association with VSD, code “Interrupted aortic arch + VSD”, and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and the VSD separately to provide further documentation about the individual interrupted aortic arch and VSD types. {Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.} {A "VSD" is a "Ventricular Septal Defect" and is also known as an "Interventricular communication". A VSD is defined as "a hole between the ventricular chambers or their remnants". (The VSD is defined on the basis of its margins as seen from the aspect of the morphologically right ventricle. In the setting of double outlet right ventricle, the defect provides the outflow from the morphologically left ventricle. In univentricular atrioventricular connections with functionally single left ventricle with an outflow chamber, the communication is referred to by some as a bulboventricular foramen.)}
2000	Interrupted aortic arch + AP window (aortopulmonary window)	Indicate if the patient has the diagnosis of “Interrupted aortic arch + AP window (aortopulmonary window)”. In the event of interrupted aortic arch occurring in association with AP window, code “Interrupted aortic arch + AP window (aortopulmonary window)”, and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and the AP window separately to provide further documentation about the individual interrupted aortic arch and AP window types. {Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.} {An “AP window (aortopulmonary window)” is defined as a defect with side-to-side continuity of the lumens of the aorta and pulmonary arterial tree, which is distinguished from common arterial trunk (truncus arteriosus) by the presence of two arterial valves or their atretic remnants. (In other words, an aortopulmonary

2000		window is a communication between the main pulmonary artery and ascending aorta in the presence of two separate semilunar [pulmonary and aortic] valves. The presence of two separate semilunar valves distinguishes AP window from truncus arteriosus. Type 1 proximal defect: AP window located just above the sinus of Valsalva, a few millimeters above the semilunar valves, with a superior rim but little inferior rim separating the AP window from the semilunar valves. Type 2 distal defect: AP window located in the uppermost portion of the ascending aorta, with a well-formed inferior rim but little superior rim. Type 3 total defect: AP window involving the majority of the ascending aorta, with little superior and inferior rims. The intermediate type of AP window is similar to the total defect but with adequate superior and inferior rims. In the event of AP window occurring in association with interrupted aortic arch, code “Interrupted aortic arch + AP window (aortopulmonary window)”, and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and AP window separately to provide further documentation about the individual interrupted arch and AP window types.)}
1080	Patent ductus arteriosus	Indicate if the patient has the diagnosis of “Patent ductus arteriosus”. The ductus arteriosus (arterial duct) is an essential feature of fetal circulation, connecting the main pulmonary trunk with the descending aorta, distal to the origin of the left subclavian artery. In most patients it is on the left side. If a right aortic arch is present, it may be on the right or the left; very rarely it is bilateral. When luminal patency of the duct persists post-natally, it is referred to as patent ductus arteriosus (patent arterial duct). The length and diameter may vary considerably from case to case. The media of the ductus consists mainly of smooth muscle that is arranged spirally, and the intima is much thicker than that of the aorta. (A patent ductus arteriosus is a vascular arterial connection between the thoracic aorta and the pulmonary artery. Most commonly a PDA has its origin from the descending thoracic aorta, just distal and opposite the origin of the left subclavian artery. The insertion of the ductus is most commonly into the very proximal left pulmonary artery at its junction with the main pulmonary artery. Origination and insertion sites can be variable, however.)
1090	Vascular ring	The term vascular ring refers to a group of congenital vascular anomalies that encircle and compress the esophagus and trachea. The compression may be from a complete anatomic ring (double aortic arch or right aortic arch with a left ligamentum) or from a compressive effect of an aberrant vessel (innominate artery compression syndrome).
1100	Pulmonary artery sling	In pulmonary artery sling, the left pulmonary artery originates from the right pulmonary artery and courses posteriorly between the trachea and esophagus in its route to the left lung hilum, causing a sling-like compression of the trachea.
1110	Aortic aneurysm (including	An aneurysm of the aorta is defined as a localized dilation

1110	pseudoaneurysm)	or enlargement of the aorta at any site along its length (from aortic annulus to aortoiliac bifurcation). A true aortic aneurysm involves all layers of the aortic wall. A false aortic aneurysm (pseudoaneurysm) is defined as a dilated segment of the aorta not containing all layers of the aortic wall and may include postoperative or post-procedure false aneurysms at anastomotic sites, traumatic aortic injuries or transections, and infectious processes leading to a contained rupture.
1120	Aortic dissection	Aortic dissection is a separation of the layers of the aortic wall. Extension of the plane of the dissection may progress to free rupture into the pericardium, mediastinum, or pleural space if not contained by the outer layers of the media and adventitia. Dissections may be classified as acute or chronic (if they have been present for more than 14 days).
1130	Lung disease, Benign	Lung disease arising from any etiology (congenital or acquired) which does not result in death or lung or heart-lung transplant; examples might be non-life threatening asthma or emphysema, benign cysts.
1140	Lung disease, Malignant	Lung disease arising from any etiology (congenital or acquired, including pulmonary parenchymal disease, pulmonary vascular disease, congenital heart disease, neoplasm, etc.) which may result in death or lung or heart-lung transplant.
1160	Tracheal stenosis	Tracheal stenosis is a reduction in the anatomic luminal diameter of the trachea by more than 50% of the remaining trachea. This stenosis may be congenital or acquired (as in post-intubation or traumatic tracheal stenosis).
2430	Tracheomalacia	
1170	Airway disease, Other	Included in this diagnostic category would be airway pathology not included under the definition of tracheal stenosis such as tracheomalacia, bronchotracheomalacia, tracheal right upper lobe, bronchomalacia, subglottic stenosis, bronchial stenosis, etc.
1430	Pleural disease, Benign	Benign diseases of the mediastinal or visceral pleura.
1440	Pleural disease, Malignant	Malignant diseases of the mediastinal or visceral pleura.
1450	Pneumothorax	A collection of air or gas in the pleural space.
1460	Pleural effusion	Abnormal accumulation of fluid in the pleural space.
1470	Chylothorax	The presence of lymphatic fluid in the pleural space secondary to a leak from the thoracic duct or its branches. Chylothorax is a specific type of pleural effusion.
1480	Empyema	A collection of purulent material in the pleural space, usually secondary to an infection.
1490	Esophageal disease, Benign	Any benign disease of the esophagus.
1500	Esophageal disease, Malignant	Any malignant disease of the esophagus.
1505	Mediastinal disease	Any disease of the mediastinum awaiting final benign/malignant pathology determination.
1510	Mediastinal disease, Benign	Any benign disease of the mediastinum.
1520	Mediastinal disease, Malignant	Any malignant disease of the mediastinum.

1540	Diaphragm paralysis	Paralysis of diaphragm, unilateral or bilateral.
1550	Diaphragm disease, Other	Any disease of the diaphragm other than paralysis.
2160	Rib tumor, Benign	Non-cancerous tumor of rib(s) (e.g., fibrous dysplasia)
2170	Rib tumor, Malignant	Cancerous tumor of rib(s)- primary (e.g., osteosarcoma, chondrosarcoma)
2180	Rib tumor, Metastatic	Cancerous tumor metastasized to rib(s)from a different primary location
2190	Sternal tumor, Benign	Non-cancerous tumor of sternum (e.g., fibrous dysplasia)
2200	Sternal tumor, Malignant	Cancerous tumor of sternum - primary (e.g., osteosarcoma, chondrosarcoma)
2210	Sternal tumor, Metastatic	Cancerous tumor metastasized to sternum from a different primary location
2220	Pectus carinatum	Pectus carinatum represents a spectrum of protrusion abnormalities of the anterior chest wall. Severe deformity may result in dyspnea and decreased endurance. Some patients develop rigidity of the chest wall with decreased lung compliance, progressive emphysema, and increased frequency of respiratory tract infections.
2230	Pectus excavatum	Pectus excavatum is a congenital chest wall deformity in which several ribs and the sternum grow abnormally, producing a concave, or caved-in, appearance in the anterior chest wall. Pectus excavatum is the most common type of congenital chest wall abnormality. It occurs in an estimated 1 in 300-400 births, with male predominance (male-to-female ratio of 3:1). The condition is typically noticed at birth, and more than 90% of cases are diagnosed within the first year of life. Worsening of the chest's appearance and the onset of respiratory symptoms are usually reported during rapid bone growth in the early teenage years.
2240	Thoracic outlet syndrome	Thoracic outlet syndrome (TOS) is caused by compression at the superior thoracic outlet wherein excess pressure is placed on a neurovascular bundle passing between the anterior scalene and middle scalene muscles. It can affect the brachial plexus (nerves that pass into the arm from the neck), the subclavian artery, and - rarely - the vein, which does not normally pass through the scalene hiatus. TOS may occur due to a positional cause - for example, by abnormal compression from the clavicle (collarbone) and shoulder girdle on arm movement. There are also several static forms, caused by abnormalities, enlargement, or spasm of the various muscles surrounding the arteries, veins, and/or brachial plexus, a fixation of a first rib, or a cervical rib. The most common causes of thoracic outlet syndrome include physical trauma from a car accident, repetitive injuries from a job such as frequent non-ergonomic use of a keyboard, sports-related activities, anatomical defects such as having an extra rib, and pregnancy.
1180	Arrhythmia	Any cardiac rhythm other than normal sinus rhythm.
2440	Arrhythmia, Atrial, Atrial fibrillation	
2450	Arrhythmia, Atrial, Atrial flutter	
2460	Arrhythmia, Atrial, Other	

2050	Arrhythmia, Junctional	Indicate if the patient has the diagnosis of ‘‘Arrhythmia, Junctional’’. “Arrhythmias arising from the atrioventricular junction; may be bradycardia, tachycardia, premature beats, or escape rhythm [1]. [1]. Jacobs JP. (Editor). 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, Volume 18, Supplement S2, pages 1 –530, December 9, 2008, page 379.
2060	Arrhythmia, Ventricular	Indicate if the patient has the diagnosis of ‘‘Arrhythmia, Ventricular’’. “Arrhythmia, Ventricular’’ ROOT Definition = Abnormal rhythm originating from the ventricles [1]. [1]. Jacobs JP. (Editor). 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, Volume 18, Supplement S2, pages 1 –530, December 9, 2008, page 393.
1185	Arrhythmia, Heart block	Atrioventricular block may be congenital or acquired, and may be of varying degree (first, second, or third degree).
1190	Arrhythmia, Heart block, Acquired	Atrioventricular block, when acquired, may be post-surgical, or secondary to myocarditis or other etiologies; the block may be first, second or third degree.
1200	Arrhythmia, Heart block, Congenital	Atrioventricular block, when congenital, may be first, second or third degree block.
1220	Arrhythmia, Pacemaker, Indication for replacement	Indications for pacemaker replacement may include end of generator life, malfunction, or infection.
2530	Short QT syndrome	
2540	Long QT Syndrome (Ward Romano syndrome)	
2550	Wolff-Parkinson-White syndrome (WPW syndrome)	
1230	Atrial Isomerism, Left	In isomerism, both appendages are of like morphology or structure; in left atrial isomerism both the right atrium and left atrium appear to be a left atrium structurally.
1240	Atrial Isomerism, Right	In isomerism, both appendages are of like morphology or structure; in right atrial isomerism both the right atrium and left atrium appear to be a right atrium structurally.
2090	Dextrocardia	Indicate if the patient has the diagnosis of ‘‘Dextrocardia’’. “Dextrocardia’’ is most usually considered synonymous with a right-sided ventricular mass, whilst ‘‘dextroversion’’ is frequently defined as a configuration where the ventricular apex points to the right. In a patient with the usual atrial arrangement, or situs solitus, dextroversion, therefore, implies a turning to the right of the heart [1]. [1]. Jacobs JP, Anderson RH, Weinberg P, Walters III HL, Tchervenkov CI, Del Duca D, Franklin RCG, Aiello VD, Béland MJ, Colan SD, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Elliott MJ. The nomenclature, definition and classification of cardiac structures in the setting of

2090		heterotaxy. In 2007 Supplement to Cardiology in the Young: Controversies and Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Anderson RH, Jacobs JP, and Wernovsky G, editors. Cardiology in the Young, Volume 17, Supplement 2, pages 1–28, doi: 10.1017/S1047951107001138, September 2007.
2100	Levocardia	Indicate if the patient has the diagnosis of ‘‘Levocardia’’. ‘‘Levocardia’’ usually considered synonymous with a left-sided ventricular mass, whilst ‘‘levoversion’’ is frequently defined as a configuration where the ventricular apex points to the left [1]. [1]. Jacobs JP, Anderson RH, Weinberg P, Walters III HL, Tchervenkov CI, Del Duca D, Franklin RCG, Aiello VD, Béland MJ, Colan SD, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Elliott MJ. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. In 2007 Supplement to Cardiology in the Young: Controversies and Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Anderson RH, Jacobs JP, and Wernovsky G, editors. Cardiology in the Young, Volume 17, Supplement 2, pages 1–28, doi: 10.1017/S1047951107001138, September 2007.
2110	Mesocardia	Indicate if the patient has the diagnosis of ‘‘Mesocardia’’. “Mesocardia’’ is most usually considered synonymous with the ventricular mass occupying the midline [1]. [1]. Jacobs JP, Anderson RH, Weinberg P, Walters III HL, Tchervenkov CI, Del Duca D, Franklin RCG, Aiello VD, Béland MJ, Colan SD, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Elliott MJ. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. In 2007 Supplement to Cardiology in the Young: Controversies and Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Anderson RH, Jacobs JP, and Wernovsky G, editors. Cardiology in the Young, Volume 17, Supplement 2, pages 1–28, doi: 10.1017/S1047951107001138, September 2007.
2120	Situs inversus	Indicate if the patient has the diagnosis of “Situs inversus” of the atrial chambers. The development of morphologically right-sided structures on one side of the body, and morphologically left-sided structures on the other side, is termed lateralization. Normal lateralization, the usual arrangement, is also known as ‘‘situs solitus’’. The mirror-imaged arrangement is also known as ‘‘situs inversus’’. The term ‘‘visceroatrial situs’’ is often used to refer to the situs of the viscera and atria when their situs is in agreement. The arrangement of the organs themselves, and the arrangement of the atrial chambers, is not always the same. Should such disharmony be encountered, the sidedness of the organs and atrial chambers must be separately specified [1]. [1]. Jacobs JP, Anderson RH, Weinberg P, Walters III HL, Tchervenkov CI, Del Duca D, Franklin RCG, Aiello VD, Béland MJ, Colan SD, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Elliott MJ. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. In 2007 Supplement to Cardiology in the Young: Controversies and

2120		Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Anderson RH, Jacobs JP, and Wernovsky G, editors. Cardiology in the Young, Volume 17, Supplement 2, pages 1–28, doi: 10.1017/S1047951107001138, September 2007.
1250	Aneurysm, Ventricular, Right (including pseudoaneurysm)	An aneurysm of the right ventricle is defined as a localized dilation or enlargement of the right ventricular wall.
1260	Aneurysm, Ventricular, Left (including pseudoaneurysm)	An aneurysm of the left ventricle is defined as a localized dilation or enlargement of the left ventricular wall.
1270	Aneurysm, Pulmonary artery	An aneurysm of the pulmonary artery is defined as a localized dilation or enlargement of the pulmonary artery trunk and its central branches (right and left pulmonary artery).
1280	Aneurysm, Other	A localized dilation or enlargement of a cardiac vessel or chamber not coded in specific fields available for aortic aneurysm, sinus of Valsalva aneurysm, coronary artery aneurysm, right ventricular aneurysm, left ventricular aneurysm, or pulmonary artery aneurysm.
1290	Hypoplastic RV	Small size of the right ventricle. This morphological abnormality usually is an integral part of other congenital cardiac anomalies and, therefore, frequently does not need to be coded separately. It should, however, be coded as secondary to an accompanying congenital cardiac anomaly if the right ventricular hypoplasia is not considered an integral and understood part of the primary congenital cardiac diagnosis. It would rarely be coded as a primary and/or isolated diagnosis.
1300	Hypoplastic LV	Small size of the left ventricle. This morphological abnormality usually is an integral part of other congenital cardiac anomalies and, therefore, frequently does not need to be coded separately. It should, however, be coded as secondary to an accompanying congenital cardiac anomaly if the left ventricular hypoplasia is not considered an integral and understood part of the primary congenital cardiac diagnosis. It would rarely be coded as a primary and/or isolated diagnosis.
2070	Postoperative bleeding	Indicate if the patient has the diagnosis of “Postoperative bleeding”.
1310	Mediastinitis	Inflammation/infection of the mediastinum, the cavity between the lungs which holds the heart, great vessels, trachea, esophagus, thymus, and connective tissues. In the United States mediastinitis occurs most commonly following chest surgery.
1320	Endocarditis	An infection of the endocardial surface of the heart, which may involve one or more heart valves (native or prosthetic) or septal defects or prosthetic patch material placed at previous surgery.
1325	Rheumatic heart disease	Heart disease, usually valvar (e.g., mitral or aortic), following an infection with group A streptococci
1330	Prosthetic valve failure	Indicate if the patient has the diagnosis of “Prosthetic valve failure”. This diagnosis is the primary diagnosis to be entered for patients undergoing replacement of a previously placed valve (not conduit) prosthesis, whatever

1330		type (e.g., bioprosthetic, mechanical, etc.). Failure may be due to, among others, patient somatic growth, malfunction of the prosthesis, or calcification or overgrowth of the prosthesis (e.g., pannus formation). Secondary or fundamental diagnosis would relate to the underlying valve disease entity. As an example, a patient undergoing removal or replacement of a prosthetic pulmonary valve previously placed for pulmonary insufficiency after repair of tetralogy of Fallot would have as a primary diagnosis "Prosthetic valve failure", as a secondary diagnosis "Pulmonary insufficiency", and as a fundamental diagnosis "Tetralogy of Fallot".
1340	Myocardial infarction	A myocardial infarction is the development of myocardial necrosis caused by a critical imbalance between the oxygen supply and demand of the myocardium. While a myocardial infarction may be caused by any process that causes this imbalance it most commonly results from plaque rupture with thrombus formation in a coronary vessel, resulting in an acute reduction of blood supply to a portion of the myocardium. Myocardial infarction is a usual accompaniment of anomalous left coronary artery from the pulmonary artery (ALCAPA).
1350	Cardiac tumor	An abnormal growth of tissue in or on the heart, demonstrating partial or complete lack of structural organization, and no functional coordination with normal cardiac tissue. Commonly, a mass is recognized which is distinct from the normal structural components of the heart. A primary cardiac tumor is one that arises directly from tissues of the heart, (e.g., myxoma, fibroelastoma, rhabdomyoma, fibroma, lipoma, pheochromocytoma, teratoma, hemangioma, mesothesioloma, sarcoma). A secondary cardiac tumor is one that arises from tissues distant from the heart, with subsequent spread to the otherwise normal tissues of the heart, (e.g., renal cell tumor with caval extension from the kidney to the level of the heart or tumor with extension from other organs or areas of the body (hepatic, adrenal, uterine, infradiaphragmatic)). N.B., in the nomenclature system developed, cardiac thrombus and cardiac vegetation are categorized as primary cardiac tumors.
1360	Pulmonary AV fistula	An abnormal intrapulmonary connection (fistula) between an artery and vein that occurs in the blood vessels of the lungs. Pulmonary AV fistulas may be seen in association with congenital heart defects; the associated cardiac defect should be coded as well.
1370	Pulmonary embolism	A pulmonary embolus is a blockage of an artery in the lungs by fat, air, clumped tumor cells, or a blood clot.
1385	Pulmonary vascular obstructive disease	Pulmonary vascular obstructive disease (PVOD) other than those specifically defined elsewhere (Eisenmenger's pulmonary vascular obstructive disease, primary pulmonary hypertension, persistent fetal circulation). The spectrum includes PVOD arising from (1) pulmonary arterial hypertension or (2) pulmonary venous hypertension or (3) portal hypertension, or (4) collage vascular disease, or (5) drug or toxin induced, or (6) diseases of the respiratory system, or (7) chronic thromboembolic disease, among

1385		others.
1390	Pulmonary vascular obstructive disease (Eisenmenger's)	"Eisenmenger syndrome" could briefly be described as "Acquired severe pulmonary vascular disease associated with congenital heart disease (Eisenmenger)". Eisenmenger syndrome is an acquired condition. In Eisenmenger-type pulmonary vascular obstructive disease, long-term left-to-right shunting (e.g., through a ventricular or atrial septal defect, patent ductus arteriosus, aortopulmonary window) can lead to chronic pulmonary hypertension with resultant pathological changes in the pulmonary vessels. The vessels become thick-walled, stiff, noncompliant, and may be obstructed. In Eisenmenger syndrome, the long-term left-to-right shunting will reverse and become right to left. Please note that the specific heart defect should be coded as a secondary diagnosis.
1400	Primary pulmonary hypertension	Primary pulmonary hypertension is a rare disease characterized by elevated pulmonary artery hypertension with no apparent cause. Two forms are included in the nomenclature, a sporadic form and a familial form which can be linked to the BMPR-II gene.
1410	Persistent fetal circulation	Persistence of the blood flow pattern seen in fetal life, in which high pulmonary vascular resistance in the lungs results in decreased blood flow to the lungs. Normally, after birth pulmonary pressure falls with a fall in pulmonary vascular resistance and there is increased perfusion of the lungs. Persistent fetal circulation, also known as persistent pulmonary hypertension of the newborn, can be related to lung or diaphragm malformations or lung immaturity.
1420	Meconium aspiration	Aspiration of amniotic fluid stained with meconium before, during, or after birth can lead to pulmonary sequelae including (1) pneumothorax, (2) pneumomediastinum, (3) pneumopericardium, (4) lung infection, and (5) meconium aspiration syndrome (MAS) with persistent pulmonary hypertension.
2250	Kawasaki disease	Kawasaki disease, also known as Kawasaki syndrome, is an acute febrile illness of unknown etiology that primarily affects children younger than 5 years of age. it was first described in Japan in 1967, and the first cases outside of Japan were reported in Hawaii in 1976. It is characterized by fever, rash, swelling of the hands and feet, irritation and redness of the whites of the eyes, swollen lymph glands in the neck, and irritation and inflammation of the mouth, lips, and throat. Serious complications of Kawasaki disease include coronary artery dilatations and aneurysms, and Kawasaki disease is a leading cause of acquired heart disease in children in the United States. The standard treatment with intravenous immunoglobulin and aspirin substantially decreases the development of coronary artery abnormalities.
1560	Cardiac, Other	Any cardiac diagnosis not specifically delineated in other diagnostic codes.
1570	Thoracic and/or mediastinal, Other	Any thoracic and/or mediastinal disease not specifically delineated in other diagnostic codes.
1580	Peripheral vascular, Other	Any peripheral vascular disease (congenital or acquired) or

1580		injury (from trauma or iatrogenic); vessels involved may include, but are not limited to femoral artery, femoral vein, iliac artery, brachial artery, etc.
2260	Complication of cardiovascular catheterization procedure	Unspecified complication of cardiovascular catheterization procedure
2270	Complication of cardiovascular catheterization procedure, Device embolization	Migration or movement of device introduced during a cardiac catheterization procedure to an unintended location
2280	Complication of cardiovascular catheterization procedure, Device malfunction	Malfunction of a device introduced during a cardiac catheterization procedure
2290	Complication of cardiovascular catheterization procedure, Perforation	Perforation or puncture caused by a device introduced during a cardiac catheterization procedure
2300	Complication of interventional radiology procedure	Unspecified complication of interventional radiology procedure
2310	Complication of interventional radiology procedure, Device embolization	Migration or movement of device introduced during an interventional radiology procedure to an unintended location
2320	Complication of interventional radiology procedure, Device malfunction	Malfunction of a device introduced during an interventional radiology procedure
2330	Complication of interventional radiology procedure, Perforation	Perforation or puncture caused by a device introduced during an interventional radiology procedure
2340	Foreign body, Intracardiac foreign body	Presence of a foreign body within the heart
2350	Foreign body, Intravascular foreign body	Presence of a foreign body within an artery or vein
2360	Open sternum with closed skin	Sternotomy edges not re-approximated prior to closure of skin incision
2370	Open sternum with open skin (includes membrane placed to close skin)	Sternotomy and skin incision left open following surgery, covered with a membrane or dressing
2380	Retained sternal wire causing irritation	Surgically placed wire causing soft tissue irritation, pain or swelling (not infected)
2390	Syncope	A transient, self-limited loss of consciousness with an inability to maintain postural tone that is followed by spontaneous recovery. The term syncope excludes seizures, coma, shock, or other states of altered consciousness.
2400	Trauma, Blunt	Injury (ies) sustained from blunt force, caused by motor vehicle accidents, falls, blows or crush injuries
2410	Trauma, Penetrating	Injury (ies) sustained as a result of sharp force, including cutting or piercing instruments or objects, bites, or firearm injuries from projectiles.
2560	Cardio-respiratory failure not secondary to known structural heart disease	
2570	Myocarditis	
2580	Common AV valve insufficiency	

	2590	Protein-losing enteropathy	
	2600	Plastic bronchitis	
	7000	Normal heart	Normal heart.
	7777	Miscellaneous, Other	Any disease (congenital or acquired) not specifically delineated in other diagnostic codes.
Retired	2040	Arrhythmia, Atrial	Indicate if the patient has the diagnosis of ''Arrhythmia, Atrial''. ''Arrhythmia, Atrial'' ROOT Definition = Non-sinus atrial rhythm with or without atrioventricular conduction. [1]. [1]. Jacobs JP. (Editor). 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, Volume 18, Supplement S2, pages 1 –530, December 9, 2008, page 373.

Primary diagnosis indicator

Seq Num: 1460

Required for case closure: Yes

Registry field: [SurgDiag].[SurgDiagPrim]

Shared with PAC3

Description: Indicate the diagnosis of primary importance at the time of this surgical procedure. Example: fundamental diagnosis of Tetralogy of Fallot. The current Diagnoses are both pulmonary insufficiency and residual ventricular septal defect. In this case, pulmonary insufficiency will be flagged as the primary diagnosis.

Values	*Code*	*Text*
	1	Yes
	0	No

Surgical procedure(s)

Seq Num: 1400

Required for case closure: Yes

Registry field: [Procedures].[ProcName]

Shared with PAC3

Description: If the patient underwent cardiac or thoracic surgery, indicate ALL procedures that were performed during this surgical procedure.

Values	Code	Text	
	10	PFO, Primary closure	Suture closure of patent foramen ovale (PFO).
	20	ASD repair, Primary closure	Suture closure of secundum (most frequently), coronary sinus, sinus venosus or common atrium ASD.
	30	ASD repair, Patch	Patch closure (using any type of patch material) of secundum, coronary sinus, or sinus venosus ASD.
	40	ASD repair, Device	Closure of any type ASD (including PFO) using a device.
	2110	ASD repair, Patch + PAPVC repair	Patch closure (using any type of patch material) of secundum, coronary sinus, or sinus venosus ASD plus PAPVC repair, any type
	50	ASD, Common atrium (single atrium), Septation	Septation of common (single) atrium using any type patch material.
	60	ASD creation/enlargement	Creation of an atrial septal defect or enlargement of an existing atrial septal defect using a variety of modalities including balloon septostomy, blade septostomy, or surgical septectomy. Creation may be accomplished with or without use of cardiopulmonary bypass.
	70	ASD partial closure	Intentional partial closure of any type ASD (partial suture or fenestrated patch closure).
	80	Atrial septal fenestration	Creation of a fenestration (window) in the septum between the atrial chambers. Usually performed using a hole punch, creating a specifically sized communication in patch material placed on the atrial septum.
	85	Atrial fenestration closure	Closure of previously created atrial fenestration using any method including device, primary suture, or patch.
	100	VSD repair, Primary closure	Suture closure of any type VSD.
	110	VSD repair, Patch	Patch closure (using any type of patch material) of any type VSD.
	120	VSD repair, Device	Closure of any type VSD using a device.
	130	VSD, Multiple, Repair	Closure of more than one VSD using any method or combination of methods. Further information regarding each type of VSD closed and method of closure can be provided by additionally listing specifics for each VSD closed. In the case of multiple VSDs in which only one is closed the procedure should be coded as closure of a single VSD. The fundamental diagnosis, in this case, would be "VSD, Multiple" and a secondary diagnosis can be the morphological type of VSD that was closed at the time of surgery.
	140	VSD creation/enlargement	Creation of a ventricular septal defect or enlargement of an existing ventricular septal defect.

150	Ventricular septal fenestration	Creation of a fenestration (window) in the septum between the ventricular chambers. Usually performed using a hole punch, creating a specifically sized communication in patch material placed on the ventricular septum.
170	AVC (AVSD) repair, Complete (CAVSD)	Repair of complete AV canal (AVSD) using one- or two-patch or other technique, with or without mitral valve cleft repair.
180	AVC (AVSD) repair, Intermediate (Transitional)	Repair of intermediate AV canal (AVSD) using ASD and VSD patch, or ASD patch and VSD suture, or other technique, with or without mitral valve cleft repair.
190	AVC (AVSD) repair, Partial (Incomplete) (PAVSD)	Repair of partial AV canal defect (primum ASD), any technique, with or without repair of cleft mitral valve.
2300	Valvuloplasty, Common atrioventricular valve	Common AV valve repair, any type
2250	Valvuloplasty converted to valve replacement in the same operation, Common atrioventricular valve	Common AV valve repair attempted, converted to valve replacement with prosthetic valve during the same operation
2230	Valve replacement, Common atrioventricular valve	Replacement of the common AV valve with a prosthetic valve
210	AP window repair	Repair of AP window using one- or two-patch technique with cardiopulmonary bypass; or, without cardiopulmonary bypass, using transcatheter device or surgical closure.
220	Pulmonary artery origin from ascending aorta (hemitruncus) repair	Repair of pulmonary artery origin from the ascending aorta by direct reimplantation, autogenous flap, or conduit, with or without use of cardiopulmonary bypass.
230	Truncus arteriosus repair	Truncus arteriosus repair that most frequently includes patch VSD closure and placement of a conduit from RV to PA. In some cases, a conduit is not placed but an RV to PA connection is made by direct association. Very rarely, there is no VSD to be closed. Truncal valve repair or replacement should be coded separately (Valvuloplasty, Truncal valve; Valve replacement, Truncal valve), as would be the case as well with associated arch anomalies requiring repair (e.g., Interrupted aortic arch repair).
240	Valvuloplasty, Truncal valve	Truncal valve repair, any type.
2290	Valvuloplasty converted to valve replacement in the same operation, Truncal valve	Truncal valve repair attempted, converted to valve replacement with prosthetic valve during the same operation
250	Valve replacement, Truncal valve	Replacement of the truncal valve with a prosthetic valve.
2220	Truncus + Interrupted aortic arch repair (IAA) repair	Truncus arteriosus repair usually includes patch VSD closure and placement of a conduit from RV to PA. In some cases, a conduit is not placed but an RV to PA connection is made by direct association. (Very rarely, there is no VSD) plus repair of interrupted aortic arch
260	PAPVC repair	PAPVC repair revolves around whether an intracardiac baffle is created to redirect pulmonary venous return to the left atrium or if the anomalous pulmonary vein is translocated and connected to the left atrium directly. If there is an associated ASD and it is closed, that procedure should also be listed.
270	PAPVC, Scimitar, Repair	In scimitar syndrome, PAPVC repair also revolves around whether an intracardiac baffle is created to redirect

270		pulmonary venous return to the left atrium or if the anomalous pulmonary vein is translocated and connected to the left atrium directly. If there is an associated ASD and it is closed, that procedure should also be listed. Occasionally an ASD is created; this procedure also must be listed separately. Concomitant thoracic procedures (e.g., lobectomy, pneumonectomy) should also be included in the procedures listing.
2120	PAPVC repair, Baffle redirection to left atrium with systemic vein translocation (Warden) (SVC sewn to right atrial appendage)	An intracardiac baffle is created to redirect pulmonary venous return to the left atrium and SVC sewn to right atrial appendage)
280	TAPVC repair	Repair of TAPVC, any type. Issues surrounding TAPVC repair involve how the main pulmonary venous confluence anastomosis is fashioned, whether an associated ASD is closed or left open or enlarged (ASD closure and enlargement may be listed separately), and whether, particularly in mixed type TAPVC repair, an additional anomalous pulmonary vein is repaired surgically.
2200	TAPVC repair + Shunt - systemic-to-pulmonary	Repair of TAPVC, any type plus a systemic to pulmonary shunt creation
290	Cor triatriatum repair	Repair of cor triatriatum. Surgical decision making revolves around the approach to the membrane creating the cor triatriatum defect, how any associated ASD is closed, and how any associated anomalous pulmonary vein connection is addressed. Both ASD closure and anomalous pulmonary venous connection may be listed as separate procedures.
300	Pulmonary venous stenosis repair	Repair of pulmonary venous stenosis, whether congenital or acquired. Repair can be accomplished with a variety of approaches: sutureless, patch venoplasty, stent placement, etc.
310	Atrial baffle procedure (non-Mustard, non-Senning)	The atrial baffle procedure code is used primarily for repair of systemic venous anomalies, as in redirection of left superior vena cava drainage to the right atrium.
330	Anomalous systemic venous connection repair	With the exception of atrial baffle procedures (harvest code 310), anomalous systemic venous connection repair includes a range of surgical approaches, including, among others: ligation of anomalous vessels, reimplantation of anomalous vessels (with or without use of a conduit), or redirection of anomalous systemic venous flow through directly to the pulmonary circulation (bidirectional Glenn to redirect LSVC or RSVC to left or right pulmonary artery, respectively).
340	Systemic venous stenosis repair	Stenosis or obstruction of a systemic vein (most commonly SVC or IVC) may be relieved with patch or conduit placement, excision of the stenotic area with primary reanastomosis or direct reimplantation.
350	TOF repair, No ventriculotomy	Tetralogy of Fallot repair (assumes VSD closure and relief of pulmonary stenosis at one or more levels), without use of an incision in the infundibulum of the right ventricle for exposure. In most cases this would be a transatrial and transpulmonary artery approach to repair the VSD and relieve the pulmonary stenosis. If the main pulmonary artery incision is extended proximally through the

350		pulmonary annulus, this must be considered "transannular" and thus a ventricular incision, though the length of the incision onto the ventricle itself may be minimal.
360	TOF repair, Ventriculotomy, Nontransanular patch	Tetralogy of Fallot repair (assumes VSD closure and relief of pulmonary stenosis at one or more levels), with use of a ventriculotomy incision, but without placement of a trans-pulmonary annulus patch. If the main pulmonary artery incision is extended proximally through the pulmonary annulus, this must be considered "transannular" and thus a ventricular incision, though the length of the incision onto the ventricle itself may be minimal.
370	TOF repair, Ventriculotomy, Transanular patch	Tetralogy of Fallot repair (assumes VSD closure and relief of pulmonary stenosis at one or more levels), with use of a ventriculotomy incision and placement of a trans-pulmonary annulus patch. If the main pulmonary artery incision is extended proximally through the pulmonary annulus, this must be considered "transannular" and thus a ventricular incision, though the length of the incision onto the ventricle itself may be minimal.
3330	TOF repair, Ventriculotomy, Transanular patch, plus native valve reconstruction	
3340	TOF repair, Ventriculotomy, Transanular patch, with monocusp or other surgically fashioned RVOT valve	
380	TOF repair, RV-PA conduit	Tetralogy of Fallot repair (assumes VSD closure and relief of pulmonary stenosis at one or more levels), with placement of a right ventricle-to-pulmonary artery conduit. In this procedure the major components of pulmonary stenosis are relieved with placement of the RV-PA conduit.
390	TOF - AVC (AVSD) repair	Tetralogy of Fallot repair (assumes VSD closure and relief of pulmonary stenosis at one or more levels), with repair of associated AV canal defect. Repair of associated atrial septal defect or atrioventricular valve repair(s) should be listed as additional or secondary procedures under the primary TOF-AVC procedure.
400	TOF - Absent pulmonary valve repair	Repair of tetralogy of Fallot with absent pulmonary valve complex. In most cases this repair will involve pulmonary valve replacement (pulmonary or aortic homograft, porcine, other) and reduction pulmonary artery arterioplasty.
420	Pulmonary atresia - VSD (including TOF, PA) repair	For patients with pulmonary atresia with ventricular septal defect without MAPCAs, including those with tetralogy of Fallot with pulmonary atresia, repair may entail either a tetralogy-like repair with transannular patch placement, a VSD closure with placement of an RV-PA conduit, or an intraventricular tunnel VSD closure with transannular patch or RV-PA conduit placement. To assure an accurate count of repairs of pulmonary atresia-VSD without MAPCAs, even if a tetralogy-type repair or Rastelli-type repair is used, the pulmonary atresia-VSD code should be the code used, not Rastelli procedure or tetralogy of Fallot repair with transannular patch.
2700	Pulmonary atresia - VSD - MAPCA	1-stage repair that includes bilateral pulmonary

2700	repair, Complete single stage repair (1-stage that includes bilateral pulmonary unifocalization + VSD closure + RV to PA connection [with or without conduit])	unifocalization + VSD closure + RV to PA connection [with or without conduit])
2710	Pulmonary atresia - VSD - MAPCA repair, Status post prior complete unifocalization (includes VSD closure + RV to PA connection [with or without conduit])	VSD closure + RV to PA connection [with or without conduit])
2720	Pulmonary atresia - VSD - MAPCA repair, Status post prior incomplete unifocalization (includes completion of pulmonary unifocalization + VSD closure + RV to PA connection [with or without conduit])	Completion of pulmonary unifocalization + VSD closure + RV to PA connection [with or without conduit])Pulmonary atresia - VSD - MAPCA repair, Status post prior incomplete unifocalization
2730	Unifocalization MAPCA(s), Bilateral pulmonary unifocalization - Complete unifocalization (all usable MAPCA[s] are incorporated)	Complete unifocalization , all usable MAPCA[s] are incorporated
2740	Unifocalization MAPCA(s), Bilateral pulmonary unifocalization - Incomplete unifocalization (not all usable MAPCA[s] are incorporated)	Incomplete unifocalization, not all usable MAPCA[s] are incorporated
2750	Unifocalization MAPCA(s), Unilateral pulmonary unifocalization	MAPCA(s), Unilateral pulmonary unifocalization (one side)
440	Unifocalization MAPCA(s)	Anastomosis of aortopulmonary collateral arteries into the left, right, or main pulmonary artery or into a tube graft or other type of confluence. The unifocalization procedure may be done on or off bypass.
450	Occlusion of MAPCA(s)	Occlusion, or closing off, of MAPCAs. This may be done with a transcatheter occluding device, usually a coil, or by surgical techniques.
460	Valvuloplasty, Tricuspid	Reconstruction of the tricuspid valve may include but not be limited to a wide range of techniques including: leaflet patch extension, artificial chordae placement, and papillary muscle translocation with or without detachment. Annuloplasty techniques that may be done solely or in combination with leaflet, chordae or muscle repair to achieve a competent valve include: eccentric annuloplasty, Kay annular plication, purse-string annuloplasty (including semicircular annuloplasty), sliding annuloplasty, and annuloplasty with ring placement. Do not use this code if tricuspid valve malfunction is secondary to Ebstein's anomaly; instead use the Ebstein's repair procedure code.
2280	Valvuloplasty converted to valve replacement in the same operation, Tricuspid	Tricuspid valve repair attempted, converted to valve replacement with prosthetic valve during the same operation
465	Ebstein's repair	To assure an accurate count of repairs of Ebstein's anomaly of the tricuspid valve, this procedure code was included. Repair of Ebstein's anomaly may include, among other techniques, repositioning of the tricuspid valve, plication of the atrialized right ventricle, or right reduction atrioplasty. Often associated ASD's may be closed and arrhythmias

465		addressed with surgical ablation procedures. These procedures should be entered as separate procedure codes.
470	Valve replacement, Tricuspid (TVR)	Replacement of the tricuspid valve with a prosthetic valve.
480	Valve closure, Tricuspid (exclusion, univentricular approach)	In a functional single ventricle heart, the tricuspid valve may be closed using a patch, thereby excluding the RV. Tricuspid valve closure may be used for infants with Ebstein's anomaly and severe tricuspid regurgitation or in patients with pulmonary atresia-intact ventricular septum with sinusoids.
490	Valve excision, Tricuspid (without replacement)	Excision of the tricuspid valve without placement of a prosthetic valve.
500	Valve surgery, Other, Tricuspid	Other tricuspid valve surgery not specified in procedure codes.
510	RVOT procedure	Included in this procedural code would be all RVOT procedures not elsewhere specified in the nomenclature system. These might be, among others: resection of subvalvar pulmonary stenosis (not DCRV type; may be localized fibrous diaphragm or high infundibular stenosis), right ventricular patch augmentation, or reduction pulmonary artery arterioplasty.
520	1 1/2 ventricular repair	Partial biventricular repair; includes intracardiac repair with bidirectional cavopulmonary anastomosis to volume unload a small ventricle or poorly functioning ventricle.
530	PA, reconstruction (plasty), Main (trunk)	Reconstruction of the main pulmonary artery trunk commonly using patch material. If balloon angioplasty is performed or a stent is placed in the main pulmonary artery intraoperatively, this code may be used in addition to the balloon dilation or stent placement code. If MPA reconstruction is performed with PA debanding, both codes should be listed.
540	PA, reconstruction (plasty), Branch, Central (within the hilar bifurcation)	Reconstruction of the right or left branch (or both right and left) pulmonary arteries (within the hilar bifurcation) commonly using patch material. If balloon angioplasty is performed or a stent is placed in the right or left (or both) pulmonary artery intraoperatively, this code may be used in addition to the balloon dilation or stent placement code. If, rarely, branch PA banding (single or bilateral) was performed in the past and reconstruction is performed associated with debanding, both codes should be listed.
550	PA, reconstruction (plasty), Branch, Peripheral (at or beyond the first lobar branch)	Reconstruction of the peripheral right or left branch (or both right and left) pulmonary arteries (at or beyond the hilar bifurcation) commonly using patch material. If balloon angioplasty is performed or a stent is placed in the right or left (or both) peripheral pulmonary artery intraoperatively, this code may be used in addition to the balloon dilation or stent placement code.
3350	PA, reconstruction (plasty), Branch, Peripheral (at or beyond the first lobar branch, proximal to first segmental branch)	
3360	PA, reconstruction (plasty), Branch, Peripheral (at or beyond the first lobar branch, beyond the first	

3360	segmental branch)	
570	DCRV repair	Surgical repair of DCRV combines relief of the low infundibular stenosis (via muscle resection) and closure of a VSD when present. A ventriculotomy may be required and is repaired by patch enlargement of the infundibulum. VSD closure and patch enlargement of the infundibulum, if done, should be listed as separate procedure codes.
3370	RV Rehabilitation, Endocardial Resection	
590	Valvuloplasty, Pulmonic	Valvuloplasty of the pulmonic valve may include a range of techniques including but not limited to: valvotomy with or without bypass, commissurotomy, and valvuloplasty.
2270	Valvuloplasty converted to valve replacement in the same operation, Pulmonic	Pulmonic valve repair attempted, converted to valve replacement with prosthetic valve during the same operation
600	Valve replacement, Pulmonic (PVR)	Replacement of the pulmonic valve with a prosthetic valve. Care must be taken to differentiate between homograft pulmonic valve replacement and placement of a homograft RV-PA conduit.
630	Valve excision, Pulmonary (without replacement)	Excision of the pulmonary valve without placement of a prosthetic valve.
640	Valve closure, Semilunar	Closure of a semilunar valve (pulmonic or aortic) by any technique.
650	Valve surgery, Other, Pulmonic	Other pulmonic valve surgery not specified in procedure codes.
610	Conduit placement, RV to PA	Placement of a conduit, any type, from RV to PA.
620	Conduit placement, LV to PA	Placement of a conduit, any type, from LV to PA.
1774	Conduit placement, Ventricle to aorta	Placement of a conduit from the right or left ventricle to the aorta.
1772	Conduit placement, Other	Placement of a conduit from any chamber or vessel to any vessel, valved or valveless, not listed elsewhere.
580	Conduit reoperation	Conduit reoperation is the code to be used in the event of conduit failure, in whatever position (LV to aorta, LV to PA, RA to RV, RV to aorta, RV to PA, etc.), and from whatever cause (somatic growth, stenosis, insufficiency, infection, etc.).
660	Valvuloplasty, Aortic	Valvuloplasty of the aortic valve for stenosis and/or insufficiency including, but not limited to the following techniques: valvotomy (open or closed), commissurotomy, aortic valve suspension, leaflet (left, right or noncoronary) partial resection, reduction, or leaflet shaving, extended valvuloplasty (freeing of leaflets, commissurotomy, and extension of leaflets using autologous or bovine pericardium), or annuloplasty (partial - interrupted or noncircumferential sutures, or complete - circumferential sutures).
2240	Valvuloplasty converted to valve replacement in the same operation, Aortic	Aortic valve repair attempted, converted to valve replacement with prosthetic valve during the same operation
2310	Valvuloplasty converted to valve replacement in the same operation,	Aortic valve repair attempted, converted to valve replacement with a pulmonary autograft and replacement

2310	Aortic – with Ross procedure	of the pulmonary valve with a homograft conduit during the same operation
2320	Valvuloplasty converted to valve replacement in the same operation, Aortic – with Ross-Konno procedure	Aortic valve repair attempted, converted to Konno aortoventriculoplasty using a pulmonary autograft root for the aortic root replacement.
670	Valve replacement, Aortic (AVR)	Replacement of the aortic valve with a prosthetic valve (mechanical, bioprosthetic, or homograft). Use this code only if type of valve prosthesis is unknown or does not fit into the specific valve replacement codes available. Autograft valve replacement should be coded as a Ross procedure.
680	Valve replacement, Aortic (AVR), Mechanical	Replacement of the aortic valve with a mechanical prosthetic valve.
690	Valve replacement, Aortic (AVR), Bioprosthetic	Replacement of the aortic valve with a bioprosthetic prosthetic valve.
700	Valve replacement, Aortic (AVR), Homograft	Replacement of the aortic valve with a homograft prosthetic valve.
715	Aortic root replacement, Bioprosthetic	Replacement of the aortic root (that portion of the aorta attached to the heart; it gives rise to the coronary arteries) with a bioprosthesis (e.g., porcine) in a conduit, often composite.
720	Aortic root replacement, Mechanical	Replacement of the aortic root (that portion of the aorta attached to the heart; it gives rise to the coronary arteries) with a mechanical prosthesis in a composite conduit.
730	Aortic root replacement, Homograft	Replacement of the aortic root (that portion of the aorta attached to the heart; it gives rise to the coronary arteries) with a homograft.
735	Aortic root replacement, Valve sparing	Replacement of the aortic root (that portion of the aorta attached to the heart; it gives rise to the coronary arteries) without replacing the aortic valve (using a tube graft).
740	Ross procedure	Replacement of the aortic valve with a pulmonary autograft and replacement of the pulmonary valve with a homograft conduit.
750	Konno procedure	Relief of left ventricular outflow tract obstruction associated with aortic annular hypoplasia, aortic valvar stenosis and/or aortic valvar insufficiency via Konno aortoventriculoplasty. Components of the surgery include a longitudinal incision in the aortic septum, a vertical incision in the outflow tract of the right ventricle to join the septal incision, aortic valve replacement, and patch reconstruction of the outflow tracts of both ventricles.
760	Ross-Konno procedure	Relief of left ventricular outflow tract obstruction associated with aortic annular hypoplasia, aortic valvar stenosis and/or aortic valvar insufficiency via Konno aortoventriculoplasty using a pulmonary autograft root for the aortic root replacement.
770	Other annular enlargement procedure	Techniques included under this procedure code include those designed to effect aortic annular enlargement that are not included in other procedure codes. These include the Manouguian and Nicks aortic annular enlargement procedures.
780	Aortic stenosis, Subvalvar, Repair	Subvalvar aortic stenosis repair by a range of techniques

780		including excision, excision and myotomy, excision and myomectomy, myotomy, myomectomy, initial placement of apical-aortic conduit (LV to aorta conduit replacement would be coded as conduit reoperation) , Vouhé aortoventriculoplasty (aortic annular incision at commissure of left and right coronary cusps is carried down to the septum and RV infundibulum; septal muscle is resected, incisions are closed, and the aortic annulus is reconstituted), or other aortoventriculoplasty techniques.
2100	Aortic stenosis, Subvalvar, Repair, With myectomy for IHSS	Subvalvar aortic stenosis repair including excision and myectomy
790	Aortic stenosis, Supravalvar, Repair	Repair of supravalvar aortic stenosis involving all techniques of patch aortoplasty and aortoplasty involving the use of all autologous tissue. In simple patch aortoplasty a diamond-shaped patch may be used, in the Doty technique an extended patch is placed (Y-shaped patch, incision carried into two sinuses), and in the Brom repair the ascending aorta is transected, any fibrous ridge is resected, and the three sinuses are patched separately.
800	Valve surgery, Other, Aortic	Other aortic valve surgery not specified in other procedure codes.
3380	Extended Ventricular Septoplasty (modified Konno, VSD creation and patch enlargement of LVOT, sparing aortic valve) for tunnel type sub aortic stenosis	
810	Sinus of Valsalva, Aneurysm repair	Sinus of Valsalva aneurysm repair can be organized by site of aneurysm (left, right or noncoronary sinus), type of repair (suture, patch graft, or root repair by tube graft or valved conduit), and approach used (from chamber of origin (aorta) or from chamber of penetration (LV, RV, PA, left or right atrium, etc.). Aortic root replacement procedures in association with sinus of Valsalva aneurysm repairs are usually for associated uncorrectable aortic insufficiency or multiple sinus involvement and the aortic root replacement procedure should also be listed. Additional procedures also performed at the time of sinus of Valsalva aneurysm repair include but are not limited to VSD closure, repair or replacement of aortic valve, and coronary reconstruction; these procedures should also be coded separately from the sinus of Valsalva aneurysm repair.
820	LV to aorta tunnel repair	LV to aorta tunnel repair can be accomplished by suture, patch, or both, and may require reimplantation of the right coronary artery. Associated coronary artery procedures should be coded separately from the LV to aorta tunnel repair.
830	Valvuloplasty, Mitral	Repair of mitral valve including, but not limited to: valvotomy (closed or open heart), cleft repair, annuloplasty with or without ring, chordal reconstruction, commissuorotomy, leaflet repair, or papillary muscle repair.
2260	Valvuloplasty converted to valve replacement in the same operation, Mitral	Mitral valve repair attempted, converted to valve replacement with prosthetic valve during the same operation
840	Mitral stenosis, Supravalvar mitral	Supravalvar mitral ring repair.

840	ring repair	
850	Valve replacement, Mitral (MVR)	Replacement of mitral valve with prosthetic valve, any kind, in suprannular or annular position.
860	Valve surgery, Other, Mitral	Other mitral valve surgery not specified in procedure codes.
870	Norwood procedure	The Norwood operation is synonymous with the term 'Norwood (Stage 1)' and is defined as an aortopulmonary connection and neoaortic arch construction resulting in univentricular physiology and pulmonary blood flow controlled with a calibrated systemic-to-pulmonary artery shunt, or a right ventricle to pulmonary artery conduit, or rarely, a cavopulmonary connection. When coding the procedure “Norwood procedure”, the primary procedure of the operation should be “Norwood procedure”. The second procedure that is coded as part of the Norwood (Stage 1) operation (Procedure 2 after the Norwood procedure) must then document the source of pulmonary blood flow and be chosen from the following eight choices: 1. Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS) 2. Shunt, Systemic to pulmonary, Central (from aorta or to main pulmonary artery) 3. Shunt, Systemic to pulmonary, Other 4. Conduit placement, RV to PA 5. Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn) 6. Glenn (unidirectional cavopulmonary anastomosis) (unidirectional Glenn) 7. Bilateral bidirectional cavopulmonary anastomosis (BBDCPA) (bilateral bidirectional Glenn) 8. HemiFontan
880	HLHS biventricular repair	Performed in patients who have small but adequately sized ventricles to support systemic circulation. These patients usually have small, but not stenotic, aortic and/or mitral valves. Primary biventricular repair has consisted of extensive aortic arch and ascending aorta enlargement with a patch, closure of interventricular and interatrial communications, and conservative approach for left ventricular outflow tract obstruction (which may include mitral stenosis at any level, subaortic stenosis, aortic stenosis, aortic arch hypoplasia, coarctation, or interrupted aortic arch). Concurrent operations (e.g., coarctation repair, aortic valve repair or replacement, etc.) can be coded separately within the database.
3390	LV Endocardial Fibroelastosis resection	
2755	Conduit insertion right ventricle to pulmonary artery + Intraventricular tunnel left ventricle to neoaorta + Arch reconstruction (Rastelli and Norwood type arch reconstruction) (Yasui)	
2160	Hybrid Approach "Stage 1", Application of RPA & LPA bands	A “Hybrid Procedure” is defined as a procedure that combines surgical and transcatheter interventional approaches. The term “Hybrid approach” is used somewhat

2160		differently than the term “Hybrid Procedure”. A “Hybrid approach” is defined as any of a group of procedures that fit into the general silo of procedures developed from the combined use of surgical and transcatheter interventional techniques. Therefore, not all procedures classified as “Hybrid approach” are truly “Hybrid Procedures”.
2170	Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA)	A “Hybrid Procedure” is defined as a procedure that combines surgical and transcatheter interventional approaches. The term “Hybrid approach” is used somewhat differently than the term “Hybrid Procedure”. A “Hybrid approach” is defined as any of a group of procedures that fit into the general silo of procedures developed from the combined use of surgical and transcatheter interventional techniques. Therefore, not all procedures classified as “Hybrid approach” are truly “Hybrid Procedures”.
2180	Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA) + application of RPA & LPA bands	A “Hybrid Procedure” is defined as a procedure that combines surgical and transcatheter interventional approaches. The term “Hybrid approach” is used somewhat differently than the term “Hybrid Procedure”. A “Hybrid approach” is defined as any of a group of procedures that fit into the general silo of procedures developed from the combined use of surgical and transcatheter interventional techniques. Therefore, not all procedures classified as “Hybrid approach” are truly “Hybrid Procedures”.
2140	Hybrid approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Aortic arch repair (Norwood [Stage 1] + Superior Cavopulmonary anastomosis(es) + PA Debanding)	A “Hybrid Procedure” is defined as a procedure that combines surgical and transcatheter interventional approaches. The term “Hybrid approach” is used somewhat differently than the term “Hybrid Procedure”. A “Hybrid approach” is defined as any of a group of procedures that fit into the general silo of procedures developed from the combined use of surgical and transcatheter interventional techniques. Therefore, not all procedures classified as “Hybrid approach” are truly “Hybrid Procedures”. It should be acknowledged that a Hybrid approach "Stage 2" (Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding, with or without Aortic arch repair) gets its name not because it has any actual hybrid elements, but because it is part of a planned staged approach that is typically commenced with a hybrid procedure.
2150	Hybrid approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Without aortic arch repair	A “Hybrid Procedure” is defined as a procedure that combines surgical and transcatheter interventional approaches. The term “Hybrid approach” is used somewhat differently than the term “Hybrid Procedure”. A “Hybrid approach” is defined as any of a group of procedures that fit into the general silo of procedures developed from the combined use of surgical and transcatheter interventional techniques. Therefore, not all procedures classified as “Hybrid approach” are truly “Hybrid Procedures”. It should be acknowledged that a Hybrid approach "Stage 2" (Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding, with or without Aortic arch repair) gets its name not because it has any actual hybrid elements, but because it is part of a planned staged approach that is typically commenced with a hybrid procedure.

2760	Hybrid Approach, Transcardiac balloon dilation	
2770	Hybrid Approach, Transcardiac transcatheter device placement	
890	Transplant, Heart	Heart transplantation, any technique, allograft or xenograft.
900	Transplant, Heart and lung	Heart and lung (single or double) transplantation.
910	Partial left ventriculectomy (LV volume reduction surgery) (Batista)	Wedge resection of LV muscle, with suturing of cut edges together, to reduce LV volume.
920	Pericardial drainage procedure	Pericardial drainage can include a range of therapies including, but not limited to: pericardiocentesis, pericardiostomy tube placement, pericardial window creation, and open pericardial drainage (pericardiotomy).
930	Pericardiectomy	Surgical removal of the pericardium.
940	Pericardial procedure, Other	Other pericardial procedures that include, but are not limited to: pericardial reconstruction for congenital absence of the pericardium, pericardial biopsy, pericardial mass or cyst excision.
950	Fontan, Atrio-pulmonary connection	The atrio-pulmonary Fontan is a type of Fontan with connection of the atrium to the pulmonary artery. “The Fontan” is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart.
960	Fontan, Atrio-ventricular connection	The atrio-ventricular Fontan is a type of Fontan with atrio-ventricular connection, either direct or with RA-RV conduit, valved or nonvalved. "The Fontan" is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart.
970	Fontan, TCPC, Lateral tunnel, Fenestrated	The lateral tunnel Fontan is a TCPC type of Fontan Procedure created with anastomosis of SVC and right atrium to the branch pulmonary artery and an intra-atrial baffle to direct IVC flow to pulmonary artery. “The Fontan” is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart. A “TCPC” is a Fontan where both the superior caval vein and the inferior caval vein are connected to the pulmonary circulation through separate connections that are either direct connections or tubular pathways. A fenestration of a Fontan is defined as a communication that is created to allow flow of blood between the systemic and pulmonary venous chambers.
980	Fontan, TCPC, Lateral tunnel, Nonfenestrated	The lateral tunnel Fontan is a TCPC type of Fontan Procedure created with anastomosis of SVC and right atrium to the branch pulmonary artery and an intra-atrial baffle to direct IVC flow to pulmonary artery. “The Fontan” is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart. A “TCPC” is a Fontan where both the superior caval vein and the inferior caval vein are connected

980		to the pulmonary circulation through separate connections that are either direct connections or tubular pathways. A fenestration of a Fontan is defined as a communication that is created to allow flow of blood between the systemic and pulmonary venous chambers.
1000	Fontan, TCPC, External conduit, Fenestrated	The external conduit Fontan is a TCPC type of Fontan operation created with anastomosis of SVC to the branch pulmonary artery a conduit outside of the heart to connect the infradiaphragmatic systemic venous return to the pulmonary artery. "The Fontan" is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart. A "TCPC" is a Fontan where both the superior caval vein and the inferior caval vein are connected to the pulmonary circulation through separate connections that are either direct connections or tubular pathways. A fenestration of a Fontan is defined as a communication that is created to allow flow of blood between the systemic and pulmonary venous chambers.
1010	Fontan, TCPC, External conduit, Nonfenestrated	The external conduit Fontan is a TCPC type of Fontan operation created with anastomosis of SVC to the branch pulmonary artery a conduit outside of the heart to connect the infradiaphragmatic systemic venous return to the pulmonary artery. "The Fontan" is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart. A "TCPC" is a Fontan where both the superior caval vein and the inferior caval vein are connected to the pulmonary circulation through separate connections that are either direct connections or tubular pathways. A fenestration of a Fontan is defined as a communication that is created to allow flow of blood between the systemic and pulmonary venous chambers.
2780	Fontan, TCPC, Intra/extracardiac conduit, Fenestrated	The TCPC with Intra/extracardiac conduit is a TCPC type of Fontan operation created with a tube where the tube is attached to the inferior caval vein inside of the heart, and then the tube passes outside of the heart and is attached to the pulmonary artery outside of the heart. "The Fontan" is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart. A "TCPC" is a Fontan where both the superior caval vein and the inferior caval vein are connected to the pulmonary circulation through separate connections that are either direct connections or tubular pathways. A fenestration of a Fontan is defined as a communication that is created to allow flow of blood between the systemic and pulmonary venous chambers.
2790	Fontan, TCPC, Intra/extracardiac conduit, Nonfenestrated	The TCPC with Intra/extracardiac conduit is a TCPC type of Fontan operation created with a tube where the tube is attached to the inferior caval vein inside of the heart, and then the tube passes outside of the heart and is attached to the pulmonary artery outside of the heart. "The Fontan" is defined as an operation or intervention that results in caval

2790		flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart. A “TCPC” is a Fontan where both the superior caval vein and the inferior caval vein are connected to the pulmonary circulation through separate connections that are either direct connections or tubular pathways. A fenestration of a Fontan is defined as a communication that is created to allow flow of blood between the systemic and pulmonary venous chambers.
3310	Fontan, TCPC, External conduit, Hepatic veins to pulmonary artery, Fenestrated	
3320	Fontan, TCPC, External conduit, Hepatic veins to pulmonary artery, Nonfenestrated	
1025	Fontan revision or conversion (Re-do Fontan)	“Fontan revision or conversion (Re-do Fontan)” is defined as an operation where a previously created Fontan circuit is either modified or taken down and changed into a different type of Fontan. “The Fontan” is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart. A “TCPC” is a Fontan where both the superior caval vein and the inferior caval vein are connected to the pulmonary circulation through separate connections that are either direct connections or tubular pathways.
1030	Fontan, Other	Other Fontan procedure not specified in procedure codes. May include takedown of a Fontan procedure. “The Fontan” is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart.
2340	Fontan + Atrioventricular valvuloplasty	“Fontan + Atrioventricular valvuloplasty” is defined as an operation to repair the systemic atrioventricular valve combined with a Fontan operation. Please also code the type of Fontan operation performed as the second procedure of this operation. “The Fontan” is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart.
1035	Ventricular septation	Creation of a prosthetic ventricular septum. Surgical procedure used to septate univentricular hearts with two atrioventricular valves. Additional procedures, such as resection of subpulmonic stenosis, should be listed separately.
1050	Congenitally corrected TGA repair, Atrial switch and ASO (double switch)	Repair of congenitally corrected TGA by concomitant atrial switch (Mustard or Senning) and arterial switch operation. VSD closure is usually performed as well; this should be coded separately.
1060	Congenitally corrected TGA repair, Atrial switch and Rastelli	Repair of congenitally corrected TGA by concomitant atrial switch (Mustard or Senning) and VSD closure to the aortic valve with placement of an RV-to-PA conduit.
1070	Congenitally corrected TGA repair,	Repair of congenitally corrected TGA by VSD closure only.

1070	VSD closure	
1080	Congenitally corrected TGA repair, VSD closure and LV to PA conduit	Repair of congenitally corrected TGA by VSD closure and placement of an LV-to-PA conduit.
1090	Congenitally corrected TGA repair, Other	Any procedures for correction of CCTGA not otherwise specified in other listed procedure codes.
1110	Arterial switch operation (ASO)	Arterial switch operation is used for repair of transposition of the great arteries (TGA). The pulmonary artery and aorta are transected and translocated so that the pulmonary artery arises from the right ventricle and the aorta from the left ventricle. Coronary artery transfer is also accomplished.
1120	Arterial switch operation (ASO) and VSD repair	Arterial switch operation is used for repair of transposition of the great arteries (TGA). The pulmonary artery and aorta are transected and translocated so that the pulmonary artery arises from the right ventricle and the aorta from the left ventricle. Coronary artery transfer is also accomplished. The VSD is closed, usually with a patch.
1123	Arterial switch procedure + Aortic arch repair	Concomitant arterial switch operation and repair of the aortic arch in patients with transposition of the great arteries with intact ventricular septum and associated coarctation of the aorta or interrupted aortic arch.
1125	Arterial switch procedure and VSD repair + Aortic arch repair	Concomitant arterial switch operation with VSD closure and repair of aortic arch in patients with transposition of the great arteries with VSD and associated coarctation of the aorta or interrupted aortic arch.
1130	Senning	Atrial baffle procedure for rerouting of venous flow in TGA resulting in a “physiological repair”. The caval flow is directed behind the baffle to the mitral valve, left ventricle and pulmonary artery while the pulmonary venous flow is directed in front of the baffle to the tricuspid valve, right ventricle, and aorta. The Senning procedure uses atrial wall to construct the baffle.
1140	Mustard	Atrial baffle procedure for rerouting of venous flow in TGA resulting in a “physiological repair”. The caval flow is directed behind the baffle to the mitral valve, left ventricle and pulmonary artery while pulmonary venous flow is directed in front of the baffle to the tricuspid valve, right ventricle, and aorta. The Mustard procedure uses patch material to construct the baffle.
1145	Atrial baffle procedure, Mustard or Senning revision	Revision of a previous atrial baffle procedure (either Mustard or Senning), for any reason (e.g., obstruction, baffle leak).
1150	Rastelli	Most often used for patients with TGA-VSD and significant LVOTO, the Rastelli operation consists of an LV-to-aorta intraventricular baffle closure of the VSD and placement of an RV-to-PA conduit.
1160	REV	The Lecompte (REV) intraventricular repair is designed for patients with abnormalities of ventriculoarterial connection in whom a standard intraventricular tunnel repair cannot be performed. It is also suitable for patients in whom an arterial switch procedure with tunneling of the VSD to the pulmonary artery cannot be performed because of pulmonary (left ventricular outflow tract) stenosis. A right ventriculotomy incision is made. The infundibular (conal)

1160		septum, located between the two semilunar valves, is aggressively resected if its presence interferes with the construction of a tunnel from the VSD to the aorta. The VSD is then tunneled to the aorta. The decision to perform or not to perform the Lecompte maneuver should be made at the beginning of the operation. If the Lecompte maneuver is not performed the pulmonary artery is translocated to the right ventricular outflow tract on the side of the aorta that provides the shortest route. (When the decision to perform the Lecompte maneuver has been made, the great vessels are transected and this maneuver is performed at the beginning of the operation.) The pulmonary artery orifice is then closed. The aorta, if it had been transected during the performance of the Lecompte maneuver, is then reconstructed. A vertical incision is made on the anterior aspect of the main pulmonary artery. The posterior margin of the pulmonary artery is sutured to the superior aspect of the vertical right ventriculotomy incision. A generous patch of autologous pericardium is used to close the inferior portion of the right ventriculotomy and the anterior portion of the pulmonary artery. A monocusp pericardial valve is inserted extemporaneously.
2190	Aortic root translocation over left ventricle (Including Nikaidoh procedure)	
2210	TGA, Other procedures (Kawashima, LV-PA conduit, other)	
3400	Double root translocation	
1180	DORV, Intraventricular tunnel repair	Repair of DORV using a tunnel closure of the VSD to the aortic valve. This also includes the posterior straight tunnel repair of Kawashima
3410	DORV repair, No Ventriculotomy	
3420	DORV repair, Ventriculotomy, Nontransannular patch	
3430	DORV repair, Ventriculotomy, Transannular patch	
3440	DORV repair, RV-PA conduit	
3450	DORV - AVC (AVSD) repair	
1200	DOLV repair	Because of the morphologic variability of DOLV, there are many approaches to repair, including: intraventricular tunnel repair directing the VSD to the pulmonary valve, the REV procedure, or the Rastelli procedure. In the case of DOLV use this code for tunnel closure to the pulmonary valve. If the REV or Rastelli procedures are performed then use those respective codes.
1210	Coarctation repair, End to end	Repair of coarctation of aorta by excision of the coarctation segment and end-to-end circumferential anastomosis of the aorta.
1220	Coarctation repair, End to end, Extended	Repair of coarctation of the aorta by excision of the coarctation segment and end-to-end anastomosis of the oblique ends of the aorta, creating an extended anastomosis.

3460	Coarctation repair, Descending aorta anastomosed to Ascending aorta	
1230	Coarctation repair, Subclavian flap	Repair of coarctation of the aorta by ligating, dividing, and opening the subclavian artery, incising the coarctation site, and folding down the subclavian artery onto the incision in the aorta, suturing the subclavian "flap" in place, creating a roof over the area of the previous coarctation.
1240	Coarctation repair, Patch aortoplasty	Repair of coarctation of the aorta by incising the coarctation site with placement of a patch sutured in place longitudinally along the aortotomy edge.
1250	Coarctation repair, Interposition graft	Repair of coarctation of the aorta by resection of the coarctation segment and placement of a prosthetic tubular interposition graft anastomosed circumferentially to the cut ends of the aorta.
3470	Coarctation repair, Extra-anatomic Bypass graft	
1260	Coarctation repair, Other	Any repair of coarctation not specified in procedure codes. This may include, for example, a combination of two approaches for coarctation repair or extra-anatomic bypass graft, etc.
1275	Coarctation repair + VSD repair	Coarctation of aorta repair, any technique, and simultaneous VSD repair, any type VSD, any type repair.
1280	Aortic arch repair	Aortic arch repair, any technique.
1285	Aortic arch repair + VSD repair	Aortic arch repair, any technique, and simultaneous VSD repair, any type VSD, any type repair. This includes repair of IAA with VSD.
1290	Coronary artery fistula ligation	Coronary artery fistula repair using any technique. If additional technique information may be supplied by another procedure code, please list separately (e.g., bypass graft).
1291	Anomalous origin of coronary artery from pulmonary artery repair	Repair of anomalous origin of the coronary artery (any) from the pulmonary artery, by any technique (ligation, translocation with aortic implantation, Takeuchi operation, or bypass graft). If additional technique information may be supplied by another procedure code, please list separately (for example, bypass graft).
1300	Coronary artery bypass	Coronary artery bypass graft procedure, any technique (with or without CPB, venous or arterial graft, one or more grafts, etc.), for any coronary artery pathology (coronary arterial fistula, aneurysm, coronary bridging, atresia of left main, acquired coronary artery disease, etc.).
1305	Anomalous aortic origin of coronary artery from aorta (AAOCA) repair	
1310	Coronary artery procedure, Other	Any coronary artery procedure not specifically listed.
1320	Interrupted aortic arch repair	Repair of interrupted aortic arch (any type) by any technique (direct anastomosis, prosthetic graft, etc.). Does not include repair of IAA-VSD.
1330	PDA closure, Surgical	Closure of a PDA by any surgical technique (ligation, division, clip) using any approach (i.e., thoracotomy, thoracoscopic, etc.).

1340	PDA closure, Device	Closure of a PDA by device using transcatheter techniques.
1360	Vascular ring repair	Repair of vascular ring (any type, except pulmonary artery sling) by any technique.
1365	Aortopexy	Surgical fixation of the aorta to another structure (usually the posterior aspect of the sternum) to relieve compression on another vessel or structure (e.g., trachea).
1370	Pulmonary artery sling repair	Pulmonary artery sling repair by any technique.
1380	Aortic aneurysm repair	Aortic aneurysm repair by any technique.
1390	Aortic dissection repair	Aortic dissection repair by any technique.
1400	Lung biopsy	Lung biopsy, any technique.
1410	Transplant, lung(s)	Lung or lobe transplantation of any type.
1420	Lung procedure, Other	Included in this procedure code would be any lung procedure other than transplant, such as, but not limited to: pneumonectomy (left or right), lobectomy (any lobe), bilobectomy (two lobes), segmental lung resection (any segment), or wedge resection.
1440	Tracheal procedure	Any tracheal procedure, including but not limited to relief of tracheal stenosis (any means including pericardial graft, autograft insertion, homograft insertion, resection with reanastomosis, rib cartilage insertion, or slide tracheoplasty). Tracheal stent placement or balloon dilation should be coded separately.
2800	Muscle flap, Trunk (i.e. intercostal, pectus, or serratus muscle)	A trunk muscle flap (intercostal, pectus, or serratus muscle) is rotated to buttress or augment a suture line, anastomosis or fill the pleural space.
2810	Muscle flap, Trunk (i.e. latissimus dorsi)	A trunk muscle flap (latissimus dorsi) is rotated to buttress or augment a suture line, anastomosis or fill the pleural space.
2820	Removal, Sternal wire	Excision of wire used to approximate sternum, previous sternotomy
2830	Rib excision, Complete	Complete excision of rib(s)
2840	Rib excision, Partial	Partial excision of rib(s)
2850	Sternal fracture - open treatment	Repair of a sternal fracture with sutures, wires, plates or bars.
2860	Sternal resection, Radical resection of sternum	Involves removal of the sternum with complex reconstructive requirements for either a tumor or severe sternal infection.
2870	Sternal resection, Radical resection of sternum with mediastinal lymphadenectomy	Involves resection of the sternum and mediastinal lymph node dissection.
2880	Tumor of chest wall - Excision including ribs	Excision of ribs and attached muscles for a benign or malignant tumor of the chest wall. When three or less ribs are taken or if the defect is covered by the scapula, reconstruction may not be necessary.
2890	Tumor of chest wall - Excision including ribs, With reconstruction	Resection of the chest wall tumor with reconstruction of the defect, usually with plastic mesh (marlex, prolene), methylmethracralate/mesh sandwich or a muscle flap.
2900	Tumor of soft tissue of thorax - Excision of deep subfascial or	Excision of a deep chest wall tumor that involves the muscles but not the ribs. These would usually be benign

2900	intramuscular tumor	tumors such as a fibroma or a deep lipoma.
2910	Tumor of soft tissue of thorax - Excision of subcutaneous tumor	Excision of tumor in the skin/fat of the chest wall-typically a lipoma.
2920	Tumor of soft tissue of thorax - Radical resection	En-bloc, radical excision of a cancer of the chest wall muscles, involving the skin, fat and muscles. Typically it would be a desmoid tumor or a sarcoma malignant fibrous histiocytoma, rhabdomyosarcoma.
2930	Hyoid myotomy and suspension	Typically done as a suprahyoid laryngeal release to reduce tension on a cervical tracheal resection anastomosis. The hyoid bone is cut laterally on both sides to allow it to drop down and thus lower the larynx and trachea.
2940	Muscle flap, Neck	A neck muscle flap is rotated to buttress or augment a suture line, anastomosis or fill a space. Commonly used neck muscles are strap muscles, sternocleidomastoid muscle, levator scapulae.
2950	Procedure on neck	Unlisted procedure of the neck
2960	Tumor of soft tissue of neck - Excision of deep subfascial or intramuscular tumor	Excision of a tumor that involves the muscles of the neck. These would usually be benign tumors such as a fibroma or a deep lipoma.
2970	Tumor of soft tissue of neck - Excision of subcutaneous tumor	Excision of a tumor in the skin/fat of the neck-typically a lipoma.
2980	Tumor of soft tissue of neck - Radical resection	A surgical procedure in which the fibrofatty contents of the neck are removed for the treatment of cervical lymphatic metastases. Neck dissection is most commonly used in the management of cancers of the upper aerodigestive tract. It is also used for malignancies of the skin of the head and neck area, the thyroid, and the salivary glands.
2990	Pectus bar removal	Removal of a previously implanted chest wall bar
3000	Pectus bar repositioning	Repositioning of a previously implanted chest wall bar
3010	Pectus repair, Minimally invasive repair (Nuss), With thoracoscopy	Placement of a Nuss transverse chest wall bar to push the sternum forward to repair a pectus deformity, with thoracoscopy
3020	Pectus repair, Minimally invasive repair (Nuss), Without thoracoscopy	Placement of a Nuss transverse chest wall bar to push the sternum forward to repair a pectus deformity, without thoracoscopy
3030	Pectus repair, Open repair	Resection of several costal cartilages, a partial osteotomy of the sternum, and often placement of a temporary bar for stabilization of pectus chest wall deformity
3040	Division of scalenus anticus, With resection of a cervical rib	Repair of Thoracic Outlet Syndrome variant where the scalenus anticus muscle or a band from it impinges on the brachial plexus along with resection of the abnormal cervical rib
3050	Division of scalenus anticus, Without resection of a cervical rib	Repair of Thoracic Outlet Syndrome variant where the scalenus anticus muscle or a band from it impinges on the brachial plexus along without resection of the abnormal cervical rib
3060	Rib excision, Excision of cervical rib	Removal of the first rib or a cervical rib for treatment of Thoracic Outlet Syndrome
3070	Rib excision, Excision of cervical rib, With sympathectomy	Removal of the first rib or a cervical rib and sympathectomy for treatment of Thoracic Outlet Syndrome

3080	Rib excision, Excision of first rib	Removal of the first rib
3090	Rib excision, Excision of first rib, With sympathectomy	Removal of the first rib and sympathectomy
3100	Procedure on thorax	Unlisted procedure on thorax
1450	Pacemaker implantation, Permanent	Implantation of a permanent pacemaker of any type (e.g., single-chamber, dual-chamber, atrial antitachycardia), with any lead configuration or type (atrial, ventricular, atrial and ventricular, transvenous, epicardial, transmural), by any technique (sternotomy, thoracotomy etc.).
1460	Pacemaker procedure	Any revision to a previously placed pacemaker system including revisions to leads, generators, pacemaker pockets. This may include explantation of pacemakers or leads as well.
2350	Explantation of pacing system	Removal of pacemaker generator and wires
1470	ICD (AICD) implantation	Implantation of an (automatic) implantable cardioverter defibrillator system.
1480	ICD (AICD) ([automatic] implantable cardioverter defibrillator) procedure	Any revision to a previously placed AICD including revisions to leads, pads, generators, pockets. This may include explantation procedures as well.
1490	Arrhythmia surgery - atrial, Surgical Ablation	Surgical ablation (any type) of any atrial arrhythmia.
1500	Arrhythmia surgery - ventricular, Surgical Ablation	Surgical ablation (any type) of any ventricular arrhythmia.
2500	Cardiovascular catheterization procedure, Diagnostic	Invasive diagnostic procedure involving the heart and great vessels
2520	Cardiovascular catheterization procedure, Diagnostic, Angiographic data obtained	Invasive diagnostic procedure involving the heart and great vessels using angiography
2550	Cardiovascular catheterization procedure, Diagnostic, Electrophysiology alteration	
2540	Cardiovascular catheterization procedure, Diagnostic, Hemodynamic alteration	Invasive diagnostic procedure involving pressure or flow alteration in the cardiovascular system
2510	Cardiovascular catheterization procedure, Diagnostic, Hemodynamic data obtained	Invasive diagnostic procedure involving pressure and flow assessment of the heart and great vessels
2530	Cardiovascular catheterization procedure, Diagnostic, Transluminal test occlusion	
2410	Cardiovascular catheterization procedure, Therapeutic	Invasive therapeutic procedure involving the heart and great vessels
2670	Cardiovascular catheterization procedure, Therapeutic, Adjunctive therapy	
1540	Cardiovascular catheterization procedure, Therapeutic, Balloon dilation	Invasive therapeutic procedure involving balloon dilatation of a cardiovascular structure
2590	Cardiovascular catheterization procedure, Therapeutic, Balloon	Invasive therapeutic procedure involving balloon dilatation of a valve

2590	valvotomy	
1580	Cardiovascular catheterization procedure, Therapeutic, Coil implantation	Invasive therapeutic procedure involving implantation of a coil
1560	Cardiovascular catheterization procedure, Therapeutic, Device implantation	Invasive therapeutic procedure involving implantation of a device
3110	Cardiovascular catheterization procedure, Therapeutic, Device implantation attempted	Invasive therapeutic procedure involving attempted but unsuccessful implantation of a device
2690	Cardiovascular catheterization procedure, Therapeutic, Electrophysiological ablation.	Invasive therapeutic procedure involving Catheter based creation of lesions in the heart with radiofrequency energy, cryotherapy , or ultrasound energy to cure or control arrhythmias
3120	Cardiovascular catheterization procedure, Therapeutic, Intravascular foreign body removal	Invasive therapeutic procedure involving removal of an intravascular foreign body
2640	Cardiovascular catheterization procedure, Therapeutic, Perforation (establishing interchamber and/or intervessel communication)	Invasive therapeutic procedure establishing interchamber and/or intervessel communication
2580	Cardiovascular catheterization procedure, Therapeutic, Septostomy	Invasive therapeutic procedure establishing an intracardiac septal communication
1550	Cardiovascular catheterization procedure, Therapeutic, Stent insertion	Invasive therapeutic procedure involving implantation of a stent
2630	Cardiovascular catheterization procedure, Therapeutic, Stent re-dilation	Invasive therapeutic procedure involving dilatation of a previously implanted stent
2650	Cardiovascular catheterization procedure, Therapeutic, Transcatheter Fontan completion	
2660	Cardiovascular catheterization procedure, Therapeutic, Transcatheter implantation of valve	Invasive therapeutic procedure involving deployment/ implantation of a valve
1590	Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS)	Placement of a tube graft from a branch of the aortic arch to the pulmonary artery with or without bypass, from any approach (thoracotomy, sternotomy).
1600	Shunt, Systemic to pulmonary, Central (shunt from aorta)	A direct anastomosis or placement of a tube graft from the aorta to the pulmonary artery with or without bypass, from any approach (thoracotomy, sternotomy).
3130	Shunt, Systemic to pulmonary, Central (shunt from aorta), Central shunt with an end-to-side connection between the transected main pulmonary artery and the side of the ascending aorta (i.e. Mee shunt)	Creation of a central shunt with an end-to-side connection between the transected main pulmonary artery and the side of the ascending aorta
3230	Shunt, Systemic to pulmonary, Potts - Smith type (descending aorta to pulmonary artery)	

1610	Shunt, Systemic to pulmonary, Other	Placement of any other systemic-to-pulmonary artery shunt, with or without bypass, from any approach (thoracotomy, sternotomy) that is not otherwise coded. Includes classic Blalock-Taussig systemic-to-pulmonary artery shunt.
1630	Shunt, Ligation and takedown	Takedown of any shunt.
2095	Shunt, Reoperation	Revision or replacement of a previously created shunt
1640	PA banding (PAB)	Placement of a pulmonary artery band, any type.
1650	PA debanding	Debanding of pulmonary artery. Please list separately any pulmonary artery reconstruction required.
3200	PA band adjustment	
1660	Damus-Kaye-Stansel procedure (DKS) (creation of AP anastomosis without arch reconstruction)	In the Damus-Kaye-Stansel procedure the proximal transected main pulmonary artery is connected by varying techniques to the aorta.
1670	Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn)	Superior vena cava to pulmonary artery anastomosis allowing flow to both pulmonary arteries with an end-to-side superior vena-to-pulmonary artery anastomosis.
1680	Glenn (unidirectional cavopulmonary anastomosis) (unidirectional Glenn)	Superior vena cava to ipsilateral pulmonary artery anastomosis (i.e., LSVC to LPA, RSVC to RPA).
1690	Bilateral bidirectional cavopulmonary anastomosis (BBDCPA) (bilateral bidirectional Glenn)	Bilateral superior vena cava-to-pulmonary artery anastomoses (requires bilateral SVCs).
1700	HemiFontan	A HemiFontan is an operation that includes a bidirectional superior vena cava (SVC)-to-pulmonary artery anastomosis and the connection of this “SVC-pulmonary artery amalgamation” to the atrium, with a “dam” between this “SVC-pulmonary artery amalgamation” and the atrium. This operation can be accomplished with a variety of operative strategies including the following two techniques and other techniques that combine elements of both of these approaches: (1) Augmenting both branch pulmonary arteries with a patch and suturing the augmented branch pulmonary arteries to an incision in the medial aspect of the superior vena cava. (With this approach, the pulmonary artery patch forms a roof over the SVC-to-pulmonary artery anastomosis and also forms a “dam” between the SVC-pulmonary artery amalgamation and the right atrium.) (2) Anastomosing both ends of the divided SVC to incisions in the top and bottom of the right pulmonary artery, and using a separate patch to close junction of the SVC and the right atrium.
2330	Superior cavopulmonary anastomosis(es) (Glenn or HemiFontan) + Atrioventricular valvuloplasty	
2130	Superior Cavopulmonary anastomosis(es) + PA reconstruction	
3300	Takedown of superior cavopulmonary anastomosis	
3140	Hepatic vein to azygous vein	

3140	connection, Direct	
3150	Hepatic vein to azygous vein connection, Interposition graft	
3160	Kawashima operation (superior cavopulmonary connection in setting of interrupted IVC with azygous continuation)	
1710	Palliation, Other	Any other palliative procedure not specifically listed.
2360	ECMO cannulation	Insertion of cannulas for extracorporeal membrane oxygenation
2370	ECMO decannulation	Removal of cannulas for extracorporeal membrane oxygenation
1910	ECMO procedure	Any ECMO procedure (cannulation, decannulation, etc.).
1900	Intraaortic balloon pump (IABP) insertion	Insertion of intraaortic balloon pump by any technique.
1920	Right/left heart assist device procedure	Any right, left, or biventricular assist device procedure (placement, removal etc.).
2390	VAD explantation	Removal of ventricular assist device
2380	VAD implantation	Insertion of a ventricular assist device
3170	VAD change out	Removal of previously inserted ventricular assist device and insertion of a new device
2420	Echocardiography procedure, Sedated transesophageal echocardiogram	Procedural sedation for echocardiogram
2430	Echocardiography procedure, Sedated transthoracic echocardiogram	Procedural sedation for echocardiogram, transthoracic
2435	Non-cardiovascular, Non-thoracic procedure on cardiac patient with cardiac anesthesia	Anesthesia provided by cardiac anesthesiologist for patient with congenital heart disease undergoing a non-cardiovascular, non-thoracic procedure
2440	Radiology procedure on cardiac patient, Cardiac Computerized Axial Tomography (CT Scan)	A patient with congenital heart disease undergoing cardiac CT scan
2450	Radiology procedure on cardiac patient, Cardiac Magnetic Resonance Imaging (MRI)	A patient with congenital heart disease undergoing cardiac MRI
2460	Radiology procedure on cardiac patient, Diagnostic radiology	A patient with congenital heart disease undergoing a diagnostic radiology procedure
2470	Radiology procedure on cardiac patient, Non-Cardiac Computerized Tomography (CT) on cardiac patient	A patient with congenital heart disease undergoing a non-cardiac CT scan
2480	Radiology procedure on cardiac patient, Non-cardiac Magnetic Resonance Imaging (MRI) on cardiac patient	A patient with congenital heart disease undergoing non-cardiac MRI
2490	Radiology procedure on cardiac patient, Therapeutic radiology	A patient with congenital heart disease undergoing a therapeutic radiology procedure
1720	Aneurysm, Ventricular, Right, Repair	Repair of right ventricular aneurysm, any technique.
1730	Aneurysm, Ventricular, Left, Repair	Repair of left ventricular aneurysm, any technique.

1740	Aneurysm, Pulmonary artery, Repair	Repair of pulmonary artery aneurysm, any technique.
1760	Cardiac tumor resection	Resection of cardiac tumor, any type.
1780	Pulmonary AV fistula repair/occlusion	Repair or occlusion of a pulmonary arteriovenous fistula.
1790	Ligation, Pulmonary artery	Ligation or division of the pulmonary artery. Most often performed as a secondary procedure.
1802	Pulmonary embolectomy, Acute pulmonary embolus	Acute pulmonary embolism (clot) removal, through catheter or surgery.
1804	Pulmonary embolectomy, Chronic pulmonary embolus	Chronic pulmonary embolism (clot) removal, through catheter or surgery.
1810	Pleural drainage procedure	Pleural drainage procedure via thoracocentesis, tube thoracostomy, or open surgical drainage.
1820	Pleural procedure, Other	Other pleural procedures not specifically listed; may include pleurodesis (mechanical, talc, antibiotic or other), among others.
1830	Ligation, Thoracic duct	Ligation of the thoracic duct; most commonly for persistent chylothorax.
1840	Decortication	Decortication of the lung by any technique.
1850	Esophageal procedure	Any procedure performed on the esophagus.
1860	Mediastinal procedure	Any non-cardiovascular mediastinal procedure not otherwise listed.
1870	Bronchoscopy	Bronchoscopy, rigid or flexible, for diagnostic, biopsy, or treatment purposes (laser, stent, dilation, lavage).
1880	Diaphragm plication	Plication of the diaphragm; most often for diaphragm paralysis due to phrenic nerve injury.
1890	Diaphragm procedure, Other	Any diaphragm procedure not specifically listed.
1930	VATS (video-assisted thoracoscopic surgery)	Video-assisted thoracoscopic surgery utilized; this code should be used in addition to the specific procedure code (e.g., if PDA ligated using VATS technique, PDA ligation should be primary procedure, VATS should be secondary procedure).
1940	Minimally invasive procedure	Any procedure using minimally invasive technique; this code should be used in addition to the specific procedure code (e.g., if ASD closed using minimally invasive technique, ASD repair should be primary procedure, minimally invasive procedure should be listed additionally).
1950	Bypass for noncardiac lesion	Use of cardiopulmonary bypass for noncardiac lesion; this code may be used in addition to the specific procedure code if one is available (e.g., tracheal procedures may be done using CPB - the tracheal procedure should be the primary procedure and use of cardiopulmonary bypass for noncardiac lesion should be listed additionally).
1960	Delayed sternal closure	Sternal closure effected after patient has left operating room with sternum open, either because of swelling or electively after complex heart procedures. This procedure should be operative type No CPB Cardiovascular.
1970	Mediastinal exploration	Mediastinal exploration, most often for postoperative control of bleeding or tamponade, but may be exploration to assess mediastinal mass, etc.

1980	Sternotomy wound drainage	Drainage of the sternotomy wound.
3180	Intravascular stent removal	Removal of a previously placed intravascular stent
3220	Removal of transcatheter-delivered device from heart	
3210	Removal of transcatheter-delivered device from blood vessel	
1990	Thoracotomy, Other	Any procedure performed through a thoracotomy incision not otherwise listed.
2000	Cardiotomy, Other	Any procedure involving an incision in the heart that is not otherwise listed.
2010	Cardiac procedure, Other	Any cardiac procedure, bypass or non-bypass, that is not otherwise listed.
2020	Thoracic and/or mediastinal procedure, Other	Any thoracic and/or mediastinal procedure not otherwise listed.
2030	Peripheral vascular procedure, Other	Any peripheral vascular procedure; may include procedures such as femoral artery repair, iliac artery repair, etc.
2040	Miscellaneous procedure, Other	Any miscellaneous procedure not otherwise listed.
2050	Organ procurement	Procurement of an organ for transplant (most likely, heart, lungs, or heart and lungs).
7777	Other procedure	Any procedure on any organ system not otherwise listed.
7800	Operation canceled before skin incision	Surgical procedure canceled after patient enters the operating room but prior to skin incision
7810	Operation aborted after skin incision	Surgical procedure canceled after skin incision made
3240	Attempted fetal intervention, percutaneous trans-catheter directed at interatrial septum (RETIRED)	
3250	Attempted fetal intervention, percutaneous trans-catheter directed at aortic valve (RETIRED)	
3260	Attempted fetal intervention, percutaneous trans-catheter directed at pulmonic valve (RETIRED)	
3270	Attempted fetal intervention “open” (maternal laparotomy with hysterotomy), directed at interatrial septum (RETIRED)	
3280	Attempted fetal intervention “open” (maternal laparotomy with hysterotomy), directed at aortic valve (RETIRED)	
3290	Attempted fetal intervention “open” (maternal laparotomy with hysterotomy), directed at pulmonic valve (RETIRED)	

Primary procedure indicator

Seq Num: 1405

Required for case closure: Yes

Registry field: [Procedures].[PrimProc]

Shared with PAC3

Description: Indicate whether this procedure is considered the PRIMARY procedure performed during this operation.

Values	*Code*	*Text*
	1	Yes
	0	No

Procedure location

Seq Num: 1480

Required for case closure: Yes

Registry field: [Operative].[ProcLoc]

Shared with PAC3

Description: Indicate the location where the operation/procedure was performed.

Values	*Code*	*Text*	
	9	Cardiac OR	Indicate if the operation/procedure was performed in the following location: Cardiac OR (Cardiac Operating Room).
	10	General OR	Indicate if the operation/procedure was performed in the following location: General OR (General Operating Room).
	3	Hybrid Suite	Indicate if the operation/procedure was performed in the following location: Hybrid Suite. A "Hybrid Suite" is defined as a room that is designed for both surgical procedure s and transcatheter interventional procedures. A "Hybrid Procedure" is defined as a procedure that combines surgical and transcatheter interventional approaches. The term "Hybrid approach" is used somewhat differently than the term "Hybrid Procedure". A "Hybrid approach" is defined as any of a group of procedures that fit into the general silo of procedures developed from the combined use of surgical and transcatheter interventional techniques. Therefore, not all procedures classified as "Hybrid approach" are truly "Hybrid Procedures".
	2	Cath lab	Indicate if the operation/procedure was performed in the following location: Cath lab (Cardiac catheterization laboratory).
	11	ICU	Indicate if the operation/procedure was performed in the following location: ICU (Intensive Care Unit).
	4	CVICU	Indicate if the operation/procedure was performed in the following location: CVICU (CardioVascular Intensive Care Unit).
	5	NICU	Indicate if the operation/procedure was performed in the following location: NICU (Neonatal Intensive Care Unit).
	6	PICU	Indicate if the operation/procedure was performed in the following location: PICU (Pediatric Intensive Care Unit).
	7	SICU	Indicate if the operation/procedure was performed in the following location: SICU (Surgical Intensive Care Unit).
	12	Radiology Suite	Indicate if the operation/procedure was performed in the following location: Radiology Suite
	13	Procedure Room	Indicate if the operation/procedure was performed in the following location: Procedure Room
	8	Other	Indicate if the operation/procedure was performed in the following location: Other (Any location not contained in this list).

Cardiac surgery type

Seq Num: 1500

Required for case closure: Yes

Registry field: [Operative].[CardSurgType]

Shared with PAC3

Description: If the patient underwent cardiac or thoracic surgery, indicate the type of primary surgical procedure performed (e.g., CPB, No CPB cardiovascular, thoracic, etc.)

*

AICDs, permanent pacemakers, and loop recorders should all generally be code as 2 - No CPB cardiovascular. However, please assure that the classification recorded in PC4 matches the classifications coded in STS.

Values	*Code*	*Text*	
	1	CPB Cardiovascular	If cardiopulmonary bypass is used, this must be chosen as the case category whether the procedure is thoracic (e.g., tracheal reconstruction) or cardiovascular in nature.
	2	No CPB Cardiovascular	If the procedure is cardiovascular, but cardiopulmonary bypass is not used, this must be chosen as the case category. This includes any procedure that includes the heart, great vessels, or any of the branches from the great vessels, where CPB is not used. Examples include but are not limited to: coarctation of the aorta repair, creation of a systemic-to-pulmonary artery shunt, patent ductus arteriosus ligation. A delayed sternal closure is included in this category. If a pericardial window done for cancer, it should be classified as a Cardiac Operation (Operation type = No CPB Cardiovascular).
	9	CPB Non-Cardiovascular	Procedures that are done with bypass support that do not involve a concomitant cardiovascular procedure. For example, tracheal surgery, neurosurgical procedures, resuscitation and rewarming of drowning victims.
	3	ECMO	If ECMO cannulation or decannulation is the primary procedure performed, this category must be chosen. However, if ECMO is initiated for support at the end of another type procedure (i.e., CPB, No CPB Cardiovascular), that procedure takes precedence and the category code would not be ECMO.
	4	Thoracic	If a procedure is performed on a structure within the chest cavity but does not involve the cardiac chambers or vessels, it would be a Thoracic category case (for example, lobectomy, pectus excavatum/carinatum repair, anterior spine exposure). There will be thoracic cases that require cardiopulmonary bypass (e.g., some types of tracheal reconstructions). In those cases, the use of cardiopulmonary bypass takes precedence and the case would not be Thoracic, but CPB.
	5	Interventional Cardiology	If an interventional device (e.g., occluder, stent) is placed in the operating room as the primary procedure performed, this category must be chosen. However, if in the course of another type procedure (i.e., CPB, No CPB Cardiovascular), an interventional device is placed in addition to the other procedure, the other category takes precedence and the

5		case would not be Interventional Cardiology.
6	VAD Operation Done With CPB	Ventricular Assist Device procedure done with CPB. This includes operations to insert the VAD or to remove the VAD.
7	VAD Operation Done Without CPB	Ventricular Assist Device procedure done without CPB. This includes operations to insert the VAD, to remove the VAD, or any procedure performed while on the VAD.
8	Non-cardiac, Non-thoracic procedure on cardiac patient with cardiac anesthesia	Any non-cardiac or non-thoracic procedure such as a general surgical procedure with anesthesia provided by cardiac anesthesiology because of the patient's underlying cardiac physiology.
777	Other	All other procedures that do not fall within the above definitions should be coded as category Other. This would include but not be limited to supportive minor procedures (e.g., line placements)

Cardiopulmonary bypass time

Seq Num: 1540

Required for case closure: Yes

Registry field: [Operative].[CPBTm]

Description: Indicate the total number of minutes that systemic return is diverted into the cardiopulmonary bypass (CPB) circuit and returned to the systemic system. This time period (Cardiopulmonary Bypass Time) includes all periods of cerebral perfusion and sucker bypass. This time period (Cardiopulmonary Bypass Time) excludes any circulatory arrest and modified ultrafiltration periods. If more than one period of CPB is required during the surgical procedure, the sum of all the CPB periods will equal the total number of CPB minutes. Enter zero if cardiopulmonary bypass technique was not used.

Multiple bypass runs

Seq Num: 1541

Required for case closure: No

Registry field: [Operative].[CPBmult]

Description: Indicate whether multiple bypass runs occurred during this surgery.

*

This field is meant to capture patients who separate from bypass and then require re-initiation of bypass for any reason. A patient who undergoes circulatory arrest during an operative procedure should not be coded as having multiple bypass runs solely due to the period of circulatory arrest.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Aortic cross-clamp time Seq Num: 1560

Required for case closure: Yes

Registry field: [Operative].[XClampTm]

Description: Indicate the total number of minutes that the coronary circulation is mechanically isolated from systemic circulation, either by an aortic cross clamp or systemic circulatory arrest. This time period (Cross Clamp Time) includes all intervals of intermittent or continuous cardioplegia administration. If more than one cross clamp period is required during this surgical procedure, the sum of the cross clamp periods is equal to the total number of cross clamp minutes. Enter zero if the coronary circulation was never mechanically isolated from systemic circulation, either by an aortic cross clamp or systemic circulatory arrest. For the following two operations: (1) "Transplant, Heart", and (2) "Transplant, Heart and lung", the field "Cross Clamp Time" will be defined as the cross clamp time of the donor heart. Therefore, these two operations represent the only operations where the field "Cross Clamp Time" can be greater than the field "Cardiopulmonary Bypass Time".

Circulatory arrest time Seq Num: 1580

Required for case closure: Yes

Registry field: [Operative].[DHCATm]

Description: Indicate the total number of minutes of complete cessation of blood flow to the patient. This time period (Circulatory Arrest Time) excludes any periods of cerebral perfusion. If more than one period of circulatory arrest is required during this surgical procedure, the sum of these periods is equal to the total duration of circulatory arrest. Enter zero if circulatory arrest technique was not used.

Cerebral perfusion used Seq Num: 1600

Required for case closure: Yes

Registry field: [Operative].[CPerfUtil]

Description: Indicate whether cerebral perfusion was performed.

Values	*Code*	*Text*
	1	Yes
	0	No

Cerebral perfusion time Seq Num: 1620

Required for case closure: Yes

Registry field: [Operative].[CPerfTm]

Description: If cerebral perfusion was used, indicate the total number of minutes cerebral perfusion was performed. This would include antegrade or retrograde cerebral perfusion strategies.

Ultrafiltration performed after CPB

Seq Num: 1621

Required for case closure: Yes

Registry field: [Operative].[Ultrafiltration]

Description: Indicate whether ultrafiltration was performed after CPB.

Values	*Code*	*Text*
	0	No
	2	Yes, Modified ultrafiltration (MUF)
	3	Yes, Conventional ultrafiltration (CUF)
	4	Yes, MUF and CUF
	9	Unk

Cross Clamp Time - No CPB

Seq Num: 1640

Required for case closure: Yes

Registry field: [Operative].[XClampTmNC]

Description: If the surgery type is "No CPB cardiovascular", indicate the total number of minutes the aorta is completely cross-clamped during this surgical procedure. Enter zero if no cross-clamp was used.

Endotracheal intubation

Seq Num: 1670

Required for case closure: Yes

Registry field: [Operative].[OpIntubate]

Description: Indicate whether an endotracheal intubation was performed.

This includes (1) patients intubated for this procedure; (2) those on preoperative invasive ventilation who remained on support until the surgical start time.
*
Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Extubated in OR/on arrival

Seq Num: 1671

Required for case closure: Yes

Registry field: [Operative].[OpExtubate]

Description: Indicate Yes if the patient (1) had the endotracheal tube removed in OR or PACU and arrived to the inpatient unit with a natural airway or (2) had the endotracheal tube removed in the inpatient unit by the anesthesia team shortly after arrival with no course of mechanical ventilation. (Bag-mask ventilation does not qualify as mechanical ventilation.)

This includes patients with a tracheostomy who are never mechanically ventilated but remain cannulated with their tracheostomy tube.
*

A patient who fails extubation in the OR and is then reintubated before arriving at the CICU should be coded as No.

Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Date/time of ICU/PACU arrival

Seq Num: 1680

Required for case closure: Yes

Registry field: [Operative].[ICUPACUAdmitDtTm]

Shared with PAC3

Description: Indicate the date/time the patient arrived in the ICU/PACU following this surgery.

If the patient was first admitted to CICU care following this surgery, this will be the same as the CICU start date/time. If, however, the patient was under CICU care during the pre-operative period, this will differ from the CICU start date/time.

If the procedure was performed at the bedside, use the procedure date/time.

For PAC3: If the patient did not go to the CICU or PACU after leaving the OR, record the OR exit time.

Do not modify this field without consulting the PC4 data champion because it impacts inotrope/vasopressor infusion calculations in the PC4 version 3 case report form (# 9921-10018).

*

For a patient who undergoes more than one procedure before entering the CICU, please add one minute to this value for the second procedure so that we can determine the sequencing in analysis.

Please enter a time for this field. If the time is left blank, the software will default to midnight. If the actual time value is midnight, please code as 00:01 so we know it is not a default value.

Glucose check

Seq Num: 1700

Retired in version 2.0

Required for case closure: No

Registry field: [Operative].[GlucoseCheck]

Description: If the patient age is >= 18 years and had cardiothoracic surgery during this episode of care (hospitalization), indicate Yes if the patient's glucose checked on the morning of postop day 1 and postop day 2. (Postop day 1 begins at midnight of the day following surgery).

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

VTE prophylaxis Seq Num: 1720

Retired in version 2.0

Required for case closure: No

Registry field: [Operative].[VTEProph]

Description: If the patient age is >= 18 years and had cardiothoracic surgery during this episode of care (hospitalization), indicate Yes if any VTE prophylaxis was used following this surgery.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Pharmacologic Seq Num: 1740

Retired in version 2.0

Required for case closure: No

Registry field: [Operative].[VTEProphPharm]

Description: If any VTE prophylaxis was used following this surgery, indicate Yes if it included pharmacologic prophylaxis (i.e., heparin or enoxaparin).

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Non-pharmacologic Seq Num: 1760

Retired in version 2.0

Required for case closure: No

Registry field: [Operative].[VTEProphNonPharm]

Description: If any VTE prophylaxis was used following this surgery, indicate Yes if it included non-pharmacologic prophylaxis

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Cardiac Catheterization

Cardiac Catheterization

Seq Num: 1220

Required for case closure: Yes

Registry field: [Hospitalization].[CardCath]

Shared with PAC3

Description: Indicate Yes if the patient had a cardiac catheterization -- either diagnostic or interventional -- during this hospitalization. This should include hybrid procedures and bedside balloon septostomy.

*

Patients who undergo a hybrid procedure should have both cardiac cath procedure and a cardiac surgery procedure (#1160) recorded.

Septostomies not captured by IMPACT, regardless of the venue in which they took place, should be captured in the cardiac cath module.

Patients receiving fluoroscopy only should NOT be captured in the cardiac cath module.

A sheath that remains in place after the catheterization should also be coded as a CVL.

Values	*Code*	*Text*
	1	Yes
	0	No

Cardiac Cath Date

Seq Num: 1810

Required for case closure: Yes

Registry field: [CardiacCath].[CardCathDt]

Shared with PAC3

Description: If cardiac catheterization was performed, indicate the date of the procedure.

Diagnostic Cath Seq Num: 1830

Required for case closure: Yes

Registry field: [CardiacCath].[ProcDxCath]

Shared with PAC3

Description: Select Yes if a diagnostic cath was performed. Diagnostic cardiac catheterization is the process of introducing a catheter into veins and/or arteries from which it is advanced to the right and/or left sides of the heart. Once the catheters are positioned the pressure of the blood in various chambers of the heart can be measured, blood samples can be taken, and dye (radiographic contrast material) can be injected (a process called angiography) to allow x-ray visualization.

Values	*Code*	*Text*
	1	Yes
	0	No

ASD closure Seq Num: 1850

Required for case closure: Yes

Registry field: [CardiacCath].[ProcASD]

Shared with PAC3

Description: Select Yes if an ASD closure was performed. Atrial septal defect (ASD) is a congenital heart defect in which the wall that separates the upper heart chambers (atria) does not close completely. During the procedure, a catheter is threaded to the heart's septum. The device is then pushed out of the catheter and positioned so that it plugs the hole between the atria.

Values	*Code*	*Text*
	1	Yes
	0	No

Aortic coarctation procedure Seq Num: 1870

Required for case closure: Yes

Registry field: [CardiacCath].[ProcCoarc]

Shared with PAC3

Description: Select Yes if an aortic coarct intervention was performed. Coarctation of the aorta is a congenital heart defect involving a narrowing of the aorta. To repair the aortic coarctation, a catheter is inserted and balloon inflated through the narrowed section of the aorta to stretch the area open. A stent may also be placed in the narrowed area after the balloon dilation to keep the aorta open.

Values	*Code*	*Text*
	1	Yes
	0	No

Aortic valvuloplasty Seq Num: 1890

Required for case closure: Yes

Registry field: [CardiacCath].[ProcAorticValv]

Shared with PAC3

Description: Select Yes if an aortic valvuloplasty was performed. Aortic stenosis is a narrowing of the aortic valve. Aortic valvuloplasty is the repair of a stenotic aortic valve using a balloon catheter inside the valve. The balloon is then inflated in an effort to increase the opening size of the valve and improving blood flow.

Values	*Code*	*Text*
	1	Yes
	0	No

Electrophysiology ablation procedure Seq Num: 1960

Required for case closure: Yes

Registry field: [CardiacCath].[ProcEPAblation]

Shared with PAC3

Description: Select Yes if an ablation was performed. Catheter ablation is a minimally invasive procedure in which flexible tubes, called catheters, are placed into superficial blood vessels and advanced into the heart, or into the pericardial space around the heart, where the substrate of heart rhythm disorders can be localized and eradicated using heat or cold energy delivered at the tip of the catheter. Includes endocardial and epicardial catheter ablation

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Electrophysiology cath Seq Num: 1955

Required for case closure: Yes

Registry field: [CardiacCath].[ProcEPCath]

Shared with PAC3

Description: Select Yes if a diagnostic electrophysiology cath was performed. One or more catheters capable of recording and pacing are placed in one or more of the cardiac chambers. The catheters may be used to measure conduction of the impulse from the sinus node to the ventricle; induce a tachycardia; and/or localize (map) the location where the tachycardia originates

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

PDA closure Seq Num: 1930

Required for case closure: Yes

Registry field: [CardiacCath].[ProcPDA]

Shared with PAC3

Description: Select Yes if a PDA closure was performed. Patent ductus arteriosus (PDA) is the persistence of a normal fetal structure between the left pulmonary artery and the descending aorta. Persistence of this fetal structure beyond 10 days of life is considered abnormal. A transcatheter device closure is a minimally invasive procedure where the doctor passes a small metal coil or other blocking device through the catheter to the site of the PDA. This corrects the congenital defect by blocking blood flow through the vessel.

Values	*Code*	*Text*
	1	Yes
	0	No

Proximal PA Stent Seq Num: 1950

Required for case closure: Yes

Registry field: [CardiacCath].[ProcProxPAStent]

Shared with PAC3

Description: Select Yes if a proximal PA stent was performed. Pulmonary artery stenosis is a narrowing (stenosis) that occurs in the pulmonary artery, a large artery that sends oxygen-poor blood into the lungs to be enriched with oxygen. Pulmonary artery stenting consists of moving a balloon dilation catheter into the narrowed area of the artery. Stent placement is accomplished by positioning the balloon dilatation catheter and stent across the narrowed segment of the artery. The balloon is inflated to its recommended pressure, expanding the stent and anchoring it in place.

Values	*Code*	*Text*
	1	Yes
	0	No

Pulmonary valvuloplasty Seq Num: 1910

Required for case closure: Yes

Registry field: [CardiacCath].[ProcPulmonaryValv]

Shared with PAC3

Description: Select Yes if a pulmonary valvuloplasty was performed. Pulmonary stenosis is a narrowing of the pulmonary valve. Pulmonary valvuloplasty is the repair of a stenotic pulmonary valve using a balloon catheter inside the valve. The balloon is then inflated in an effort to increase the opening size of the valve and improving blood flow.

Values	*Code*	*Text*
	1	Yes
	0	No

Transcatheter pulmonary valve replacement

Seq Num: 2120

Required for case closure: Yes

Registry field: [CardiacCath].[ProcPVplace]

Shared with PAC3

Description: Select Yes if a Transcatheter pulmonary valve replacement (TPVR) was performed. TPVR is a percutaneous replacement of a dysfunctional pulmonary valve for pulmonary regurgitation and right ventricular outflow tract obstruction in selected patients. The device is introduced through the femoral vein and advanced into the right side of the heart and put into place at the site of the pulmonary valve.

Values	*Code*	*Text*
	1	Yes
	0	No

Endomyocardial biopsy

Seq Num: 2210

Required for case closure: Yes

Registry field: [CardiacCath].[ProcBiopsy]

Shared with PAC3

Description: Select Yes if an endomyocardial biopsy was performed

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Pulmonary hypertension eval

Seq Num: 2180

Required for case closure: Yes

Registry field: [CardiacCath].[ProcPHTNeval]

Shared with PAC3

Description: Select Yes if a pulmonary hypertension eval was performed.

*

In this field, we are looking for cath lab procedures that include formal right heart and pulmonary artery hemodynamic catheterization in multiple conditions. These include testing of right heart and pulmonary artery pressures at rest/baseline conditions, and with the addition of supplemental oxygen and/or inhaled/intravenous pulmonary vasodilators.

Values	*Code*	*Text*
	1	Yes
	0	No

Transvenous pacemaker placement Seq Num: 2200

Required for case closure: Yes

Registry field: [CardiacCath].[ProcPPMplace]

Shared with PAC3

Description: Select Yes if a transvenous pacemaker placement was performed

*

This includes AICD placed in the cath lab by EP physicians.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Other cardiac cath procedure Seq Num: 2250

Required for case closure: Yes

Registry field: [CardiacCath].[ProcOther]

Shared with PAC3

Description: Select Yes if another cardiac cath procedure was performed

*

When importing a procedure from IMPACT that is not specifically listed, you must manually select this option in order to activate the "Cath Planned Yes/No" question.

Values	*Code*	*Text*
	1	Yes
	0	No

Other cath - specify Seq Num: 2251

Required for case closure: No

Registry field: [CardiacCath].[ProcOtherSpec]

Shared with PAC3

Description: Specify the other cath procedure(s) performed

Cath procedure

Seq Num: 1817

Required for case closure: No

Registry field: [CathProc].[CathProc]

Description: Indicate all procedures performed while the patient was in the cath lab.

*

You must list every cardiac cath (diagnostic or interventional) during the hospital stay, and you must answer all the Yes/No questions for various procedure types. If a procedure began but was subsequently aborted, please do capture that procedure.

If the patient had an intervention that is not included as one of the Yes/No questions, you must answer Yes to "Other cardiac cath procedure" (#2250). Please also put something in the "Other cath – specify" (#2251) field to help us understand what procedure took place.

Regardless of how you answer the Yes/No fields, you do not need to select all the individual procedures from this dropdown. This field is primarily intended for sites whose ACC-IMPACT and PC4 databases are linked. At these sites, the individual procedures will be automatically imported from IMPACT. If you do leave this field blank, you will get a warning during data verification/submission, but the warning will not prevent you from submitting the case.

Values

Code	*Text*
5	Adjunctive therapy - Adenosine
10	Adjunctive therapy - Beta blockade
15	Adjunctive therapy - Rapid pacing
20	Adjunctive therapy - Rapid pacing: Endocardial
25	Adjunctive therapy - Rapid pacing: Epicardial
40	Balloon dilation - Conduit: LA to LV
45	Balloon dilation - Conduit: LV to aorta
50	Balloon dilation - Conduit: LV to PA
55	Balloon dilation - Conduit: Other
60	Balloon dilation - Conduit: RA to PA
65	Balloon dilation - Conduit: RA to PA-pulmonary trunk
70	Balloon dilation - Conduit: RA to RV
75	Balloon dilation - Conduit: RV to aorta
80	Balloon dilation - Conduit: RV to PA
30	Balloon dilation - Conduit: Sano modification (RV to PA valveless conduit)
35	Balloon dilation - Conduit: Sano modification-with valve (RV to PA valved conduit)
85	Balloon dilation - Conduit: Shunt -

85	systemic-to-pulmonary
90	Balloon dilation - Intracardiac/septum: Atrial baffle S/P atrial switch
95	Balloon dilation - Intracardiac/septum: Atrial septum (Static balloon dilation [without pullback])
100	Balloon dilation - Intracardiac/septum: Fontan Baffle
105	Balloon dilation - Intracardiac/septum: Fontan fenestration
110	Balloon dilation - Intracardiac/septum: Ventricular septum
1185	Balloon dilation - Pulmonary artery, Peripheral, Lobar
1190	Balloon dilation - Pulmonary artery, Peripheral, Lobar, Left lingula PA
1195	Balloon dilation - Pulmonary artery, Peripheral, Lobar, Left lower PA
1200	Balloon dilation - Pulmonary artery, Peripheral, Lobar, Left upper PA
1205	Balloon dilation - Pulmonary artery, Peripheral, Lobar, Right lower PA
1210	Balloon dilation - Pulmonary artery, Peripheral, Lobar, Right middle PA
1215	Balloon dilation - Pulmonary artery, Peripheral, Lobar, Right upper PA
1220	Balloon dilation - Pulmonary artery, Peripheral, Sublobar = Segmental
1225	Balloon dilation - Pulmonary artery, Peripheral, Sublobar = Segmental, Left
1230	Balloon dilation - Pulmonary artery, Peripheral, Sublobar = Segmental, Right
1235	Balloon dilation - Pulmonary artery, Proximal, Left
1240	Balloon dilation - Pulmonary artery, Proximal, Right
115	Balloon dilation - Pulmonary artery: Central (Proximal left and/or proximal right pulmonary artery including the pulmonary artery bifurcation)
120	Balloon dilation - Pulmonary artery: Main (Trunk)

125 Balloon dilation - Pulmonary artery: Peripheral

130 Balloon dilation - Pulmonary artery: Proximal

135 Balloon dilation - Pulmonary vein: Left (Left pulmonary vein [LPV])

140 Balloon dilation - Pulmonary vein: Left lower (Left lower pulmonary vein [LLPV])

145 Balloon dilation - Pulmonary vein: Left upper (Left upper pulmonary vein [LUPV])

150 Balloon dilation - Pulmonary vein: Lingula (Lingular pulmonary vein)

155 Balloon dilation - Pulmonary vein: Pulmonary venous confluence

160 Balloon dilation - Pulmonary vein: Pulmonary venous confluence with left atrium

165 Balloon dilation - Pulmonary vein: Right (Right pulmonary vein [RPV])

170 Balloon dilation - Pulmonary vein: Right lower (Right lower pulmonary vein [RLPV])

175 Balloon dilation - Pulmonary vein: Right middle (Right middle pulmonary vein [RMPV])

180 Balloon dilation - Pulmonary vein: Right upper (Right upper pulmonary vein [RUPV])

1245 Balloon dilation - Systemic artery, Aorta, Abdominal aorta, Coarctation

1250 Balloon dilation - Systemic artery, Aorta, Abdominal aorta, Native (Primary) coarctation

1255 Balloon dilation - Systemic artery, Aorta, Abdominal aorta, Recurrent coarctation

1260 Balloon dilation - Systemic artery, Aorta, Thoracic aorta, Ascending aorta

1265 Balloon dilation - Systemic artery, Aorta, Thoracic aorta, Coarctation, Native (Primary) coarctation

1270 Balloon dilation - Systemic artery, Aorta, Thoracic aorta, Coarctation, Recurrent coarctation

1275 Balloon dilation - Systemic artery, Aorta, Thoracic aorta, Descending thoracic aorta

1280 Balloon dilation - Systemic artery, Aorta, Thoracic aorta, Transverse arch

1285 Balloon dilation - Systemic artery, Systemic artery other than aorta, Coronary artery

1290 Balloon dilation - Systemic artery, Systemic artery other than aorta, Femoral artery

1295 Balloon dilation - Systemic artery, Systemic artery other than aorta, Iliac artery

1300 Balloon dilation - Systemic artery, Systemic artery other than aorta, Innominate artery

1305 Balloon dilation - Systemic artery, Systemic artery other than aorta, Renal artery

1310 Balloon dilation - Systemic artery, Systemic artery other than aorta, Subclavian artery

185 Balloon dilation - Systemic artery: Aorta

190 Balloon dilation - Systemic artery: Systemic artery other than aorta

1175 Balloon dilation - Systemic vein

1180 Balloon dilation - Systemic vein, Caval vein, Superior vena cava

1315 Balloon dilation - Systemic vein, Non-Caval vein, Femoral vein

1320 Balloon dilation - Systemic vein, Non-Caval vein, Iliac vein

1325 Balloon dilation - Systemic vein, Non-Caval vein, Innominate (Brachiocephalic)

1330 Balloon dilation - Systemic vein, Non-Caval vein, Subclavian vein

195 Balloon dilation - Systemic vein: Caval vein

200 Balloon dilation - Systemic vein: Non-Caval vein

205 Balloon valvotomy - Aortic valve

210 Balloon valvotomy - Mitral valve

215 Balloon valvotomy - Pulmonic valve

220 Balloon valvotomy - Tricuspid valve

225 Biopsy - RV not S/P heart transplant

230 Biopsy - RV post heart transplant

235 Biopsy - Site not RV

1400	Cardiovascular catheterization procedure, Therapeutic, Perforation (establishing interchamber and/or intervessel communication) - Systemic artery, Systemic artery other than aorta, Iliac artery
240	Coil implantation - Atrial septal defect (ASD)
1335	Coil implantation - Coil implantation
255	Coil implantation - Conduit: LA to LV
260	Coil implantation - Conduit: LV to aorta
265	Coil implantation - Conduit: LV to PA
270	Coil implantation - Conduit: Other
275	Coil implantation - Conduit: RA to PA
280	Coil implantation - Conduit: RA to PA-pulmonary trunk
285	Coil implantation - Conduit: RA to RV
290	Coil implantation - Conduit: RV to aorta
295	Coil implantation - Conduit: RV to PA
245	Coil implantation - Conduit: Sano modification (RV to PA valveless conduit)
250	Coil implantation - Conduit: Sano modification-with valve (RV to PA valved conduit)
300	Coil implantation - Conduit: Shunt - systemic-to-pulmonary
305	Coil implantation - Coronary artery fistula
310	Coil implantation - Fontan fenestration
315	Coil implantation - Intracardiac baffle leak
320	Coil implantation - Patent ductus arteriosus (PDA)
325	Coil implantation - Perivalvar leak
330	Coil implantation - Pulmonary arteriovenous malformation
335	Coil implantation - Systemic arteriovenous malformation
1340	Coil implantation - Systemic artery
340	Coil implantation - Systemic artery to pulmonary artery collateral

345 Coil implantation - Systemic artery: Aorta

350 Coil implantation - Systemic artery: Systemic artery other than aorta

1345 Coil implantation - Systemic vein

355 Coil implantation - Systemic vein to pulmonary vein collateral

360 Coil implantation - Systemic vein: Caval vein

365 Coil implantation - Systemic vein: Non-caval vein

368 Data - Angiographic data obtained

367 Data - Hemodynamic data obtained

370 Device implantation - Aortopulmonary window (AP window)

375 Device implantation - Atrial septal defect (ASD)

1355 Device implantation - Conduit

390 Device implantation - Conduit: LA to LV

395 Device implantation - Conduit: LV to aorta

400 Device implantation - Conduit: LV to PA

405 Device implantation - Conduit: Other

410 Device implantation - Conduit: RA to PA

415 Device implantation - Conduit: RA to PA-pulmonary trunk

420 Device implantation - Conduit: RA to RV

425 Device implantation - Conduit: RV to aorta

430 Device implantation - Conduit: RV to PA

380 Device implantation - Conduit: Sano modification (RV to PA valveless conduit)

385 Device implantation - Conduit: Sano modification-with valve (RV to PA valved conduit)

435 Device implantation - Conduit: Shunt - systemic-to-pulmonary

440 Device implantation - Coronary artery fistula

445 Device implantation - Fontan

445	fenestration
450	Device implantation - Intracardiac baffle leak
455	Device implantation - Patent ductus arteriosus (PDA)
460	Device implantation - Patent Foramen Ovale (PFO)
465	Device implantation - Perivalvar leak
470	Device implantation - Pulmonary arteriovenous malformation
475	Device implantation - Pulmonary artery
480	Device implantation - Systemic arteriovenous malformation
1360	Device implantation - Systemic artery
485	Device implantation - Systemic artery to pulmonary artery collateral
490	Device implantation - Systemic artery: Aorta
495	Device implantation - Systemic artery: Systemic artery other than aorta
1365	Device implantation - Systemic vein
500	Device implantation - Systemic vein to pulmonary vein collateral
1370	Device implantation - Systemic vein, Caval vein, Inferior vena cava
1375	Device implantation - Systemic vein, Caval vein, Superior vena cava
505	Device implantation - Systemic vein: Caval vein
510	Device implantation - Systemic vein: Caval vein (Superior vena cava - Right)
515	Device implantation - Systemic vein: Non-Caval vein
520	Device implantation - Ventricular septal defect (VSD)
1350	Diagnostic - Transluminal test occlusion
525	Electrophysiology alteration - Atrial stimulation
530	Electrophysiology alteration - Ventricular stimulation
533	Hemodynamic alteration - Fuild bolus challenge
532	Hemodynamic alteration - Inotropy

532	test
531	Hemodynamic alteration - Oxygen/nitric test
1165	Hybrid Approach - Transcardiac balloon dilation
1170	Hybrid Approach - Transcardiac transcatheter device placement
1140	Hybrid Approach "Stage 1" - Application of RPA & LPA bands
1145	Hybrid Approach "Stage 1" - Stent placement in arterial duct (PDA)
1150	Hybrid Approach "Stage 1" - Stent placement in arterial duct (PDA) + application of RPA& LPA bands
1155	Hybrid approach "Stage 2" - Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Aortic arch repair (Norwood [Stage 1] + Superior Cavopulmonary anastomosis(es) + PA Debanding)
1160	Hybrid approach "Stage 2" - Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Without aortic arch repair
1380	Intravascular foreign body removal - Intravascular foreign body removal
535	Other invasive procedures/interventional techniques - Pericardiocentesis - elective
540	Other invasive procedures/interventional techniques - Pericardiocentesis - emergent
545	Other invasive procedures/interventional techniques - Pleuracentesis - elective
550	Other invasive procedures/interventional techniques - Pleuracentesis - emergent
555	Other invasive procedures/interventional techniques - Snare foreign body
560	Perforation (establishing interchamber and/or intervessel communication) - Atretic aortic

560 valve

565 Perforation (establishing interchamber and/or intervessel communication) - Atretic pulmonary valve

570 Perforation (establishing interchamber and/or intervessel communication) - Atrial septum

585 Perforation (establishing interchamber and/or intervessel communication) - Conduit: LA to LV

590 Perforation (establishing interchamber and/or intervessel communication) - Conduit: LV to aorta

595 Perforation (establishing interchamber and/or intervessel communication) - Conduit: LV to PA

600 Perforation (establishing interchamber and/or intervessel communication) - Conduit: Other

605 Perforation (establishing interchamber and/or intervessel communication) - Conduit: RA to PA

610 Perforation (establishing interchamber and/or intervessel communication) - Conduit: RA to PA-pulmonary trunk

615 Perforation (establishing interchamber and/or intervessel communication) - Conduit: RA to RV

620 Perforation (establishing interchamber and/or intervessel communication) - Conduit: RV to aorta

625 Perforation (establishing interchamber and/or intervessel communication) - Conduit: RV to PA

575 Perforation (establishing interchamber and/or intervessel communication) - Conduit: Sano modification (RV to PA valveless conduit)

580 Perforation (establishing interchamber and/or intervessel communication) - Conduit: Sano modification-with valve (RV to PA valved conduit)

630 Perforation (establishing interchamber and/or intervessel communication) - Conduit: Shunt - systemic-to-pulmonary

635 Perforation (establishing interchamber and/or intervessel communication) - Fontan Baffle

1385 Perforation (establishing interchamber and/or intervessel communication) - Perforation (establishing interchamber and/or intervessel communication)

1390 Perforation (establishing interchamber and/or intervessel communication) - Systemic artery

1395 Perforation (establishing interchamber and/or intervessel communication) - Systemic artery, Systemic artery other than aorta, Femoral artery

640 Perforation (establishing interchamber and/or intervessel communication) - Systemic artery: Aorta

645 Perforation (establishing interchamber and/or intervessel communication) - Systemic artery: Systemic artery other than aorta

1405 Perforation (establishing interchamber and/or intervessel communication) - Systemic vein

1420 Perforation (establishing interchamber and/or intervessel communication) - Systemic vein, Non-Caval vein, Femoral vein

1425 Perforation (establishing interchamber and/or intervessel communication) - Systemic vein, Non-Caval vein, Iliac vein

650 Perforation (establishing interchamber and/or intervessel communication) - Systemic vein: Caval vein

655 Perforation (establishing interchamber and/or intervessel communication) - Systemic vein: Non-Caval vein

660 Perforation (establishing interchamber and/or intervessel communication) - Ventricular septum

1410 Perforation (establishing interchamber and/or intervessel communication), Systemic vein, Caval vein, Inferior vena cava - Systemic vein, Caval vein, Inferior vena cava

1415	Perforation (establishing interchamber and/or intervessel communication), Systemic vein, Caval vein, Superior vena cava - Systemic vein, Caval vein, Superior vena cava
665	Septostomy - Balloon atrial septostomy by pullback (Rashkind) (BAS)
670	Septostomy - Blade atrial septostomy
1430	Septostomy - Septostomy
685	Stent insertion - Conduit: LA to LV
690	Stent insertion - Conduit: LV to aorta
695	Stent insertion - Conduit: LV to PA
700	Stent insertion - Conduit: Other
705	Stent insertion - Conduit: RA to PA
710	Stent insertion - Conduit: RA to PA-pulmonary trunk
715	Stent insertion - Conduit: RA to RV
720	Stent insertion - Conduit: RV to aorta
725	Stent insertion - Conduit: RV to PA
675	Stent insertion - Conduit: Sano modification (RV to PA valveless conduit)
680	Stent insertion - Conduit: Sano modification-with valve (RV to PA valved conduit)
730	Stent insertion - Conduit: Shunt - systemic-to-pulmonary
735	Stent insertion - Intracardiac/septum: Atrial baffle S/P atrial switch
740	Stent insertion - Intracardiac/septum: Atrial septum
745	Stent insertion - Intracardiac/septum: Fontan Baffle
750	Stent insertion - Intracardiac/septum: Fontan fenestration
755	Stent insertion - Intracardiac/septum: Ventricular septum
760	Stent insertion - PDA
1435	Stent insertion - Pulmonary artery, Peripheral, Lobar
1440	Stent insertion - Pulmonary artery,

1440 Peripheral, Lobar, Left

1445 Stent insertion - Pulmonary artery, Peripheral, Lobar, Right

1450 Stent insertion - Pulmonary artery, Peripheral, Sublobar = Segmental

1455 Stent insertion - Pulmonary artery, Peripheral, Sublobar = Segmental, Left

1460 Stent insertion - Pulmonary artery, Peripheral, Sublobar = Segmental, Right

1465 Stent insertion - Pulmonary artery, Proximal, Left

1470 Stent insertion - Pulmonary artery, Proximal, Right

765 Stent insertion - Pulmonary artery: Central (Proximal left and/or proximal right pulmonary artery including the pulmonary artery bifurcation)

770 Stent insertion - Pulmonary artery: Main (Trunk)

775 Stent insertion - Pulmonary artery: Peripheral

780 Stent insertion - Pulmonary artery: Proximal

785 Stent insertion - Pulmonary vein: Left (Left pulmonary vein [LPV])

790 Stent insertion - Pulmonary vein: Left lower (Left lower pulmonary vein [LLPV])

795 Stent insertion - Pulmonary vein: Left upper (Left upper pulmonary vein [LUPV])

800 Stent insertion - Pulmonary vein: Lingula (Lingular pulmonary vein)

805 Stent insertion - Pulmonary vein: Pulmonary venous confluence

810 Stent insertion - Pulmonary vein: Pulmonary venous confluence with left atrium

815 Stent insertion - Pulmonary vein: Right (Right pulmonary vein [RPV])

820 Stent insertion - Pulmonary vein: Right lower (Right lower pulmonary vein [RLPV])

825 Stent insertion - Pulmonary vein: Right middle (Right middle pulmonary vein [RMPV])

830 Stent insertion - Pulmonary vein: Right upper (Right upper pulmonary vein [RUPV])

1480 Stent insertion - Systemic artery, Aorta, Abdominal aorta

1485 Stent insertion - Systemic artery, Aorta, Abdominal aorta, Coarctation

1490 Stent insertion - Systemic artery, Aorta, Abdominal aorta, Native (Primary) coarctation

1495 Stent insertion - Systemic artery, Aorta, Abdominal aorta, Recurrent coarctation

1500 Stent insertion - Systemic artery, Aorta, Thoracic aorta

1505 Stent insertion - Systemic artery, Aorta, Thoracic aorta, Ascending aorta

1510 Stent insertion - Systemic artery, Aorta, Thoracic aorta, Coarctation, Native (Primary) coarctation

1515 Stent insertion - Systemic artery, Aorta, Thoracic aorta, Coarctation, Recurrent coarctation

1520 Stent insertion - Systemic artery, Aorta, Thoracic aorta, Descending thoracic aorta

1525 Stent insertion - Systemic artery, Aorta, Thoracic aorta, Transverse arch

1530 Stent insertion - Systemic artery, Aorta, Thoracoabdominal aorta

1535 Stent insertion - Systemic artery, Systemic artery other than aorta, Coronary artery

1540 Stent insertion - Systemic artery, Systemic artery other than aorta, Femoral artery

1545 Stent insertion - Systemic artery, Systemic artery other than aorta, Iliac artery

1550 Stent insertion - Systemic artery, Systemic artery other than aorta, Renal artery

1555 Stent insertion - Systemic artery, Systemic artery other than aorta, Subclavian artery

1560 Stent insertion - Systemic artery, Systemic artery other than aorta, Systemic pulmonary vessel connection

835 Stent insertion - Systemic artery: Aorta

840 Stent insertion - Systemic artery: Systemic artery other than aorta

1475 Stent insertion - Systemic vein, Caval vein, Superior vena cava

845 Stent insertion - Systemic vein: Caval vein

850 Stent insertion - Systemic vein: Non-Caval vein

1565 Stent insertion - Transcatheter implantation of valve

865 Stent re-dilation - Conduit: LA to LV

870 Stent re-dilation - Conduit: LV to aorta

875 Stent re-dilation - Conduit: LV to PA

880 Stent re-dilation - Conduit: Other

885 Stent re-dilation - Conduit: RA to PA

890 Stent re-dilation - Conduit: RA to PA-pulmonary trunk

895 Stent re-dilation - Conduit: RA to RV

900 Stent re-dilation - Conduit: RV to aorta

905 Stent re-dilation - Conduit: RV to PA

855 Stent re-dilation - Conduit: Sano modification (RV to PA valveless conduit)

860 Stent re-dilation - Conduit: Sano modification-with valve (RV to PA valved conduit)

910 Stent re-dilation - Conduit: Shunt - systemic-to-pulmonary

915 Stent re-dilation - Intracardiac/septum: Atrial baffle S/P atrial switch

920 Stent re-dilation - Intracardiac/septum: Atrial septum

925 Stent re-dilation - Intracardiac/septum: Fontan Baffle

930 Stent re-dilation - Intracardiac/septum: Fontan fenestration

935 Stent re-dilation - Intracardiac/septum: Ventricular septum

940 Stent re-dilation - PDA

945 Stent re-dilation - Pulmonary artery: Central (Proximal left and/or

945 proximal right pulmonary artery including the pulmonary artery bifurcation)

950 Stent re-dilation - Pulmonary artery: Main (Trunk)

955 Stent re-dilation - Pulmonary artery: Peripheral

960 Stent re-dilation - Pulmonary artery: Proximal

965 Stent re-dilation - Pulmonary vein: Left (Left pulmonary vein [LPV])

970 Stent re-dilation - Pulmonary vein: Left lower (Left lower pulmonary vein [LLPV])

975 Stent re-dilation - Pulmonary vein: Left upper (Left upper pulmonary vein [LUPV])

980 Stent re-dilation - Pulmonary vein: Lingula (Lingular pulmonary vein)

985 Stent re-dilation - Pulmonary vein: Pulmonary venous confluence

990 Stent re-dilation - Pulmonary vein: Pulmonary venous confluence with left atrium

995 Stent re-dilation - Pulmonary vein: Right (Right pulmonary vein [RPV])

1000 Stent re-dilation - Pulmonary vein: Right lower (Right lower pulmonary vein [RLPV])

1005 Stent re-dilation - Pulmonary vein: Right middle (Right middle pulmonary vein [RMPV])

1010 Stent re-dilation - Pulmonary vein: Right upper (Right upper pulmonary vein [RUPV])

1015 Stent re-dilation - Systemic artery: Aorta

1020 Stent re-dilation - Systemic artery: Systemic artery other than aorta

1025 Stent re-dilation - Systemic vein: Caval vein

1030 Stent re-dilation - Systemic vein: Non-Caval vein

1035 Transcatheter Fontan completion - Completion of total cavopulmonary connection (TCPC) using transcatheter covered stent

1040 Transcatheter implantation of valve - Not systemic or pulmonary outflow

1045 Transcatheter implantation of valve - Pulmonary outflow position

1570 Transcatheter implantation of valve - pulmonary ventricular inflow position

1050 Transcatheter implantation of valve - Systemic outflow position

1575 Transcatheter implantation of valve - Systemic ventricular inflow position

1065 Transluminal test occlusion - Conduit: LA to LV

1070 Transluminal test occlusion - Conduit: LV to aorta

1075 Transluminal test occlusion - Conduit: LV to PA

1080 Transluminal test occlusion - Conduit: Other

1085 Transluminal test occlusion - Conduit: RA to PA

1090 Transluminal test occlusion - Conduit: RA to RV

1095 Transluminal test occlusion - Conduit: RV to aorta

1100 Transluminal test occlusion - Conduit: RV to PA

1055 Transluminal test occlusion - Conduit: Sano modification (RV to PA valveless conduit)

1060 Transluminal test occlusion - Conduit: Sano modification-with valve (RV to PA valved conduit)

1105 Transluminal test occlusion - Conduit: Shunt - systemic-to-pulmonary

1110 Transluminal test occlusion - Fontan fenestration

1115 Transluminal test occlusion - Interatrial communication

1120 Transluminal test occlusion - Systemic artery: Aorta

1125 Transluminal test occlusion - Systemic artery: Systemic artery other than aorta

1130 Transluminal test occlusion - Systemic vein: Caval vein

1135 Transluminal test occlusion - Systemic vein: Non-Caval vein

Planned cath intervention

Seq Num: 2270

Required for case closure: Yes

Registry field: [CardiacCath].[CathPlanned]

Description: Record Yes if the cath was the planned intervention or part of a multi-stage palliative strategy determined prior to the first intervention (surgery or catheterization) during the hospitalization. For example, patient undergoing hybrid stage I palliation has initial ductal stent placement and branch PA banding. In a second cath procedure, an atrial septal stent is placed. Code the stent procedure as Planned = Yes.

*

This field is intended to capture unplanned reinterventions following the patient's initial intervention (surgical or cath) for this hospitalization. Therefore, the first surgery or interventional catheter (whichever comes first) should always be coded as Planned, regardless of the circumstances.

Please note: All interventional caths (not diagnostic-only) must have the "Planned/unplanned" field answered. That field is only enabled if you have answered Yes to at least one of the intervention Yes/No fields, **including Other (#2250)**.

(Diagnostic caths, biopsies, PHTN evals, and EP studies are all considered 'diagnostic'.)

Values

Code	*Text*
1	Yes
0	No
9	Unk

Invasive ventilation during this cath

Seq Num: 2272

Required for case closure: Yes

Registry field: [CardiacCath].[CathIntub]

Description: Indicate whether an endotracheal intubation was performed. This includes (1) patients intubated for this procedure; (2) those on pre-procedure invasive ventilation who remained on support until the cath start time.

*

Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Extubated in cath lab / on arrival

Seq Num: 2273

Required for case closure: Yes

Registry field: [CardiacCath].[CathExtub]

Description: Indicate Yes if the patient (1) had the endotracheal tube removed in cath lab or PACU and arrived to the inpatient unit with a natural airway or (2) had the endotracheal tube removed in the inpatient unit by the anesthesia team shortly after arrival with no course of mechanical ventilation. (Bag-mask ventilation does not qualify as mechanical ventilation.)

This includes patients with a tracheostomy who are never mechanically ventilated but remain cannulated with their tracheostomy tube.
*

If a patient has an unplanned or failed extubation in the cath lab and is then reintubated, code this field as No.

If a patient returns from the cath lab receiving manual ventilation and never gets connected to a ventilator in the CICU prior to extubation, code this field as Yes.

Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Cath end date/time

Seq Num: 2280

Required for case closure: Yes

Registry field: [CardiacCath].[CathEndDtTm]

Description: Indicate the date/time the procedure ended. For sites directly importing from IMPACT, this will be the time at which the operator breaks scrub at the end of the procedure All other sites should use the time of the first post-cath vitals in the inpatient unit.

Fenestration closure

Seq Num: 1970

Retired in version 2.0

Required for case closure: Yes

Registry field: [CardiacCath].[ProcFenClose]

Description: Select Yes if a fenestration closure was performed. This includes coil and device implantations.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Fenestration creation/enlargement Seq Num: 1990

Retired in version 2.0

Required for case closure: Yes

Registry field: [CardiacCath].[ProcFenCreate]

Description: Select Yes if a fenestration creation or enlargement was performed. This includes balloon dilation, perforation (establishing interchamber and/or intervessel communication), stent insertion, and stent re-dilation.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Balloon septostomy Seq Num: 2000

Retired in version 2.0

Required for case closure: Yes

Registry field: [CardiacCath].[ProcBAS]

Description: Select Yes if a balloon atrial septostomy (BAS) was performed.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Balloon dilation - Systemic-to-pulmonary shunt Seq Num: 2010

Retired in version 2.0

Required for case closure: Yes

Registry field: [CardiacCath].[ProcBallSyst]

Description: Select Yes if a balloon dilation of the systemic-to-pulmonary shunt was performed

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Balloon dilation - RV-PA conduit Seq Num: 2020

Retired in version 2.0

Required for case closure: Yes

Registry field: [CardiacCath].[ProcBallRVPA]

Description: Select Yes if a balloon dilation of the RV-PA conduit was performed

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

ASD creation/enlargement Seq Num: 2040

Retired in version 2.0

Required for case closure: Yes

Registry field: [CardiacCath].[ProcASDCreate]

Description: Select Yes if an ASD creation or enlargement was performed. This includes perforation (establishing interchamber and/or intervessel communication), blade septostomy, stent insertion, and stent re-dilation.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Stent placed in duct Seq Num: 2060

Retired in version 2.0

Required for case closure: Yes

Registry field: [CardiacCath].[ProcStentDuct]

Description: Select Yes if a stent was placed in a duct.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Stent placed in shunt Seq Num: 2080

Retired in version 2.0

Required for case closure: Yes

Registry field: [CardiacCath].[ProcStentShunt]

Description: Select Yes if a stent was placed in a shunt. This includes Sano modification (RV to PA valveless conduit), Sano modification-with valve (RV to PA valved conduit), and systemic-to-pulmonary shunts.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Stent placed in other location Seq Num: 2100

Retired in version 2.0

Required for case closure: Yes

Registry field: [CardiacCath].[ProcStentOth]

Description: Select Yes if a stent was placed in any other location (i.e., not a duct or shunt).

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Device closure of VSD Seq Num: 2140

Retired in version 2.0

Required for case closure: Yes

Registry field: [CardiacCath].[ProcVSDdevice]

Description: Select Yes if a device closure of a VSD was performed

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

RFA and balloon for pulmonary atresia

Seq Num: 2160

Retired in version 2.0

Required for case closure: Yes

Registry field: [CardiacCath].[ProcRFAPA]

Description: Select Yes if an RFA and balloon for pulmonary atresia was performed

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

CICU Encounter

Admission date/time

Seq Num: 2310

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUStartDtTm]

Description: Date and time a CICU attending assumes primary responsibility for patient care in an ICU setting. Patient may be in any ICU setting as long as the CICU team is responsible for care.

Patient age at start of CICU Encounter

Seq Num: 2380

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUPatAgeStartD]

Description: The patient's age in days at the start of the CICU encounter, calculated by the DOB and CICU admit date/time.

Critical care end date

Seq Num: 2341

Required for case closure: Yes

Registry field: [CICUEncounter].[CritCareEndDt]

Description: Date the CICU attending physician deems the patient medically ready to leave CICU service.

For hospitals with a single inpatient service model, this should correspond to a change from critical care status to another inpatient status. If the patient's status returns to critical care, you must start a new encounter.
*

This may differ from the CICU Discharge Date (#2360), which is the date the CICU service is no longer primarily responsible for the patient's care.

If your hospital has a single inpatient service model and therefore, by design, does not transfer patients from a critical care unit to an acute care unit, please use the Critical Care End Date for the CICU Discharge Date.

When a question asks whether a line, complication, therapy, etc. was present at CICU discharge, we are referring to the CICU Discharge Date/Time (#2360), not this field.

Please note that temporarily leaving the CICU for a procedure (e.g., to the OR or cath lab) does not end the encounter.

CICU discharge date/time

Seq Num: 2360

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUPhysEndDtTm]

Description: Indicate the date/time that the patient is transferred from the CICU attending service. For hospitals with a single inpatient service model, this should correspond to a change from critical care status to another inpatient status. If the patient's status returns to critical care, you must start a new encounter.

*

This may differ from the CICU Critical Care End Date (#2341), which is when the CICU attending physician deems the patient medically ready to leave CICU service.

If your hospital has a single inpatient service model and does not transfer patients from a critical care unit to an acute care unit, please use the Critical Care End Date for the CICU Discharge Date.

When a question asks whether a line, complication, therapy, etc. was present at CICU discharge, we are referring to the date in/time this field, not the Critical Care End Date (# 2341) .

Please note that temporarily leaving the CICU for a procedure (e.g., to the OR or cath lab) does not end the encounter.

Patient age at end of CICU Encounter

Seq Num: 2400

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUPatAgeEndD]

Description: The patient's age in days at the end of the CICU encounter, calculated by the DOB and CICU discharge date/time.

Planned CICU encounter

Seq Num: 2420

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUSched]

Description: Indicate whether this CICU encounter was planned or unplanned. Planned CICU encounters are those where the CICU was aware of the definite need for CICU admission during morning bed meeting. In general, planned admissions include those where a patient was admitted directly from the cardiac operating room and those situations where a baby is born in the PC4 institution with prenatally diagnosed congenital heart disease. Patients transferred from other operating room/procedure suite environments and transfers from other hospitals are likely to be more evenly split between planned and unplanned status. Conversely, patients transferred to the CICU from another unit in the hospital will most likely not be planned admissions

Values	*Code*	*Text*	
	10	Planned	
	20	Unplanned	
Retired	1	Scheduled	Admission was non-emergent and/or planned
Retired	2	Unscheduled	Admission was emergent and/or unplanned

Reason for encounter

Seq Num: 2460

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUReason]

Description: Indicate the primary reason the patient was initially cared for by the CICU team. **This represents the reason identified at time of admission regardless what ultimately happened to the patient.**

For example, a patient may have been admitted for decompensated heart failure and evaluation for heart transplant and eventually went on to have a transplant. This patient should be coded as Medical Condition as the need for surgery was not determined at the start of the encounter.

* Additional clarifications are below between asterisks. *

Values	*Code*	*Text*	
	1	Preop cardiothoracic surgery	CICU team began care prior to anticipated cardiothoracic surgery, regardless of whether surgery occurred. * If a patient is admitted to the CICU with a plan to undergo cardiac surgery but that plan changes during his stay, 1 - Preop cardiothoracic surgery was still the reason for admission. This code is ONLY for patients admitted with a plan in place for surgery. If a patient is admitted to the CICU to determine whether or not they will require surgery, code those patients as 7 - Evaluation of structural heart disease. *
	2	Postop cardiothoracic surgery	CICU team began care immediately after cardiac surgery. Patient was either cared for by another service prior to surgery (e.g., NICU) or patient went immediately to OR upon hospital admit (e.g., elective surgery from home). * This may include CICU patients who had their cardiothoracic surgery and immediate postop care at an outside hospital, if the patient is receiving post op care and that was the purpose of the transfer. However, if the primary reason for transfer to your CICU is not post-op care but rather to diagnose/treat a medical condition, they are more appropriately coded as 3 - Medical condition. *
	3	Medical condition	CICU care for medical condition; no surgery was anticipated at the time CICU assumed care. * This category includes patients admitted preoperatively for non-cardiothoracic surgery. *
	4	Non-cardiothoracic postop	CICU team began care immediately following non-cardiac surgery. If there is an active medical condition and the patient would have otherwise gone to a non-ICU recovery area, code Medical Condition.
	8	Pre cardiac cath	CICU team began care prior to anticipated cardiac cath, regardless of whether cath occurred
	5	Post cardiac cath	CICU team began care immediately after cardiac

5		catheterization for routine post-procedural monitoring and care. If there is an active medical condition and the patient would have otherwise gone to a non-ICU recovery area, code Medical Condition.
6	Evaluation of structural heart disease	Patient admitted with known or suspected congenital/structural heart disease for the purpose of a diagnostic/therapeutic trial to determine if intervention is needed. Examples of this scenario would be a) patient with suspected coarctation of the aorta who is observed without PGE infusion for signs of aortic narrowing, or b) patient with Tetralogy of Fallot who is observed to determine whether oxygen saturations are appropriate for discharge to home. If a patient is admitted with one of these conditions, but requires treatment of a medical diagnosis (e.g. respiratory failure, acute cardiogenic shock) at the time of admission, then they should NOT be coded with this reason and instead coded as a "Medical" Encounter with the correct accompanying diagnosis. If a patient is admitted for "Evaluation of Structural Heart Disease" and eventually goes on to have cardiac surgery, the reason for encounter should remain "Eval for Structural Heart Disease. * When the purpose of admission is to determine whether or not a patient requires surgery, use this code, regardless of whether surgery ultimately occurs. Patients requiring acute medical care should be coded as 3 - Medical condition. *
9	Post non-cardiac procedure	CICU team began care immediately following anesthesia for a non-cardiac procedure such as interventional radiology or MRI (not including catheterization, cardiac surgery or non-cardiac surgery). If there is an active medical condition and the patient would have otherwise gone to a non-ICU recovery area, code Medical Condition.
10	Anticoagulation	CICU care for initiation of anticoagulation
7	ICU overflow	Patient without any cardiac disease, or any cardiac medical condition, is admitted to the CICU under the care of the CICU attending physician. NOTE: These patients do NOT need to be added to the registry if an institution chooses not to do so. * This option is intended to allow sites to track patients who were cared for in the CICU only because a bed was unavailable in another ICU. These records can be submitted to the PC4 registry, but they will be excluded from all dashboard reports and analyses. *

Non-cardiothoracic surgery

Seq Num: 2480

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUNCSurg]

Description: If the reason for the CICU encounter is "Non-cardiothoracic postop", select the type of surgery that immediately preceded CICU care.

Values

Code	*Text*
20	ENT - Cervical tracheoplasty
40	ENT - Cleft lip and palate repair
210	ENT - Incision and drainage of peritonsilar or retropharyngeal abscess
30	ENT - Laryngoplasty
10	ENT - Tracheostomy
420	ENT - Tympanoplasty
7000	ENT - Other
170	General surg - Abdominal laparoscopic procedure
360	General surg - Cholecystectomy
260	General surg - Correction of malrotation
250	General surg - Gastric restrictive procedure
240	General surg - Gastrostomy or gastrojejunostomy tube placement, open
230	General surg - Gastrostomy or gastrojejunostomy tube placement, percutaneous
280	General surg - Hernia repair
220	General surg - Nissen procedure with or without gastrostomy tube placement
100	General surg - Pectus excavatum repair
330	General surg - Proctectomy
270	General surg - Reduction of volvulus, intussusception, or internal hernia
200	General surg - Repair of diaphragmatic hernia
180	General surg - Repair of hiatal hernia
390	General surg - Repair of omphalocele or gastroschisis

320	General surg - Repair of perforated colon
310	General surg - Repair of perforated small intestine
190	General surg - Repair of tracheoesophageal fistula
300	General surg - Resection of colon
340	General surg - Resection of liver
290	General surg - Resection of small intestine
160	General surg - Splenectomy
7050	General surg - Other
410	Neurosurg - Craniectomy or craniotomy
50	Neurosurg - Creation or revision of ventriculo-peritoneal or –pleural shunt
60	Neurosurg - Repair of myelomeningocele
7100	Neurosurg - Other
90	Oncologic - Excision of abdominal tumor
70	Oncologic - Excision of benign mass
80	Oncologic - Excision of chest wall tumor
7150	Oncologic - Other
140	Ortho - Fasciotomy
120	Ortho - Open repair of fracture, joint dislocation
130	Ortho - Other open orthopedic procedure
110	Ortho - Spinal fusion, insertion of spinal fixation device, or removal of spinal hardware
7200	Ortho - Other
380	Transplant - Kidney
350	Transplant - Liver
7250	Transplant - Other
400	Urology - Bladder, uterine, or ovarian surgery
370	Urology - Resection of kidney
7300	Urology - Other
150	Vasc - Repair of systemic artery with or without graft
430	Vasc - Tunneled venous catheter placement

440	Vasc - Tunneled venous catheter removal
7350	Vasc - Other
7777	Other non-cardiothoracic surgery

Non-cardiothoracic surgery - specify

Seq Num: 2481

Required for case closure: No

Registry field: [CICUEncounter].[CICUNCSurgSpec]

Description: Specify the type of non-cardiothoracic surgery

Current surgical status

Seq Num: 2490

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUsurgStatus]

Description: Select the category that best represents the status of a patient's surgical palliation at the beginning of this CICU encounter. For example, a patient status post Glenn/hemiFontan who undergoes tricuspid valve repair at the beginning of the encounter should be coded as s/p stage II palliation. **NOTE: Please keep in mind that we are trying to describe a patient's current status, not necessarily the most recent surgery they had. Generally speaking, focus on the patient's intracardiac anatomy and their dominant physiology.**

For example, a patient with a bidirectional Glenn and a completely septated heart is best described as s/p 2V surgery.

In contrast, in a patient with PA/IVS, a bidirectional Glenn, and an open atrial septum, s/p stage II may be more appropriate.

Please also note that this you can change this designation at a future date. For example, at the time of the patient's first surgery they are thought to be a 2V but then actually goes down a single ventricle pathway, you can change the status to correspond to their ultimate physiology.

* Additional clarifications are below between asterisks.*

Values

Code	*Text*	
0	Never had cardiothoracic surgery	
1	S/P stage I palliation - Norwood	Status post Norwood stage I palliation
2	S/P stage I palliation - Hybrid	Status post hybrid stage I palliation * Includes single ventricle patients on prostins with PA bands. *
3	S/P stage II palliation	Status post stage II palliation (bi-directional Glenn, hemi-Fontan or Kawashima procedure) * Includes: - A patient status post Glenn/hemiFontan who undergoes tricuspid valve repair at the beginning of the encounter - A single ventricle patient who has a Kawashima procedure*
4	S/P stage III palliation	Status post stage III palliation (fenestrated or non-fenestrated Fontan procedure)
5	S/P aortopulmonary shunt	Status post aortopulmonary shunt (including MBTS, RVPAS or central shunt) for 1V or 2V palliation
6	S/P other 1V surgery	Patient with single ventricle anatomy status post other surgery * Includes PA bands with 1V anatomy but NOT on PGE *
7	S/P 2V surgery	Patient with two ventricle anatomy status post other palliative or reparative surgery

7		* Includes vascular ring repair, permanent pacemaker or AICD placement, and PA bands with 2V anatomy. *
9	S/P heart transplant	Patient status post heart transplant
8	S/P thoracic surg (never had cardiac surg)	Patient never had cardiac surgery; status post thoracic surgery, including tracheal reconstruction, with or without CPB. * Includes VAD procedures. *

Encounter cardiothoracic diagnosis

Seq Num: 2540

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUDiag]

Description: Indicate the primary cardiothoracic anatomy/physiology requiring care in the CICU at the start of this encounter. This may be the same as or differ from the patient's fundamental diagnosis. For example, a patient with HLHS admitted from home with new onset systemic AV valve regurgitation presenting with respiratory insufficiency would have a fundamental diagnosis of "Hypoplastic Left Heart Syndrome" but an encounter cardiothoracic diagnosis of "Tricuspid regurgitation" and an encounter medical diagnosis of "Respiratory insufficiency." For CICU readmissions during the same hospitalization, this may differ from the initial encounter cardiothoracic diagnosis. For example, and infant with complete AVSD who presents for surgery at his initial CICU encounter would have both a fundamental diagnosis and an initial encounter cardiothoracic diagnosis of "AVC (AVSD), Complete." The patient is discharged from the CICU and develops mitral stenosis requiring readmission for respiratory insufficiency. At this readmission, his encounter cardiothoracic diagnosis is "Mitral stenosis" and his encounter medical diagnosis is "Respiratory insufficiency."

Values

Code	*Text*	
10	PFO	A small interatrial communication (or potential communication) confined to the region of the oval fossa (fossa ovalis) characterized by no deficiency of the primary atrial septum (septum primum) and a normal limbus with no deficiency of the septum secundum (superior interatrial fold).
20	ASD, Secundum	A congenital cardiac malformation in which there is an interatrial communication confined to the region of the oval fossa (fossa ovalis), most commonly due to a deficiency of the primary atrial septum (septum primum) but deficiency of the septum secundum (superior interatrial fold) may also contribute.
30	ASD, Sinus venosus	A congenital cardiac malformation in which there is a caval vein (vena cava) and/or pulmonary vein (or veins) that overrides the atrial septum or the septum secundum (superior interatrial fold) producing an interatrial or anomalous venoatrial communication Although the term sinus venosus atrial septal defect is commonly used, the lesion is more properly termed a sinus venosus communication because, while it functions as an interatrial communication, this lesion is not a defect of the atrial septum.
40	ASD, Coronary sinus	A congenital cardiac malformation in which there is a deficiency of the walls separating the left atrium from the coronary sinus allowing interatrial communication through the coronary sinus ostium.
50	ASD, Common atrium (single atrium)	Complete absence of the interatrial septum. "Single atrium" is applied to defects with no associated malformation of the atrioventricular valves. "Common atrium" is applied to defects with associated malformation of the atrioventricular valves.
2150	ASD, Postoperative interatrial	A surgically created communication between the atria.

2150	communication	
71	VSD, Type 1 (Subarterial) (Supracristal) (Conal septal defect) (Infundibular)	A VSD that lies beneath the semilunar valve(s) in the conal or outlet septum.
73	VSD, Type 2 (Perimembranous) (Paramembranous) (Conoventricular)	A VSD that is confluent with and involves the membranous septum and is bordered by an atrioventricular valve, not including type 3 VSDs.
75	VSD, Type 3 (Inlet) (AV canal type)	A VSD that involves the inlet of the right ventricular septum immediately inferior to the AV valve apparatus.
77	VSD, Type 4 (Muscular)	A VSD completely surrounded by muscle.
79	VSD, Type: Gerbode type (LV-RA communication)	A rare form of VSD in which the defect is at the membranous septum; the communication is between the left ventricle and right atrium.
80	VSD, Multiple	More than one VSD exists. Each individual VSD may be coded separately to specify the individual VSD types.
100	AVC (AVSD), Complete (CAVSD)	Indicate if the patient has the diagnosis of “AVC (AVSD), Complete (CAVSD)”. An “AVC (AVSD), Complete (CAVSD)” is a “complete atrioventricular canal” or a “complete atrioventricular septal defect” and occurs in a heart with the phenotypic feature of a common atrioventricular junction. An “AVC (AVSD), Complete (CAVSD)” is defined as an AVC with a common AV valve and both a defect in the atrial septum just above the AV valve (ostium primum ASD [a usually crescent-shaped ASD in the inferior (posterior) portion of the atrial septum just above the AV valve]) and a defect in the ventricular septum just below the AV valve. The AV valve is one valve that bridges both the right and left sides of the heart. Balanced AVC is an AVC with two essentially appropriately sized ventricles. Unbalanced AVC is an AVC defect with two ventricles in which one ventricle is inappropriately small. Such a patient may be thought to be a candidate for biventricular repair, or, alternatively, may be managed as having a functionally univentricular heart. AVC lesions with unbalanced ventricles so severe as to preclude biventricular repair should be classified as single ventricles. Rastelli type A: The common superior (anterior) bridging leaflet is effectively split in two at the septum. The left superior (anterior) leaflet is entirely over the left ventricle and the right superior (anterior) leaflet is similarly entirely over the right ventricle. The division of the common superior (anterior) bridging leaflet into left and right components is caused by extensive attachment of the superior (anterior) bridging leaflet to the crest of the ventricular septum by chordae tendineae. Rastelli type B: Rare, involves anomalous papillary muscle attachment from the right side of the ventricular septum to the left side of the common superior (anterior) bridging leaflet. Rastelli type C: Marked bridging of the ventricular septum by the superior (anterior) bridging leaflet, which floats freely (often termed a "free-floater") over the ventricular septum without chordal attachment to the crest of the ventricular septum.
110	AVC (AVSD), Intermediate (transitional)	An AVC with two distinct left and right AV valve orifices but also with both an ASD just above and a VSD just below the

110		AV valves. While these AV valves in the intermediate form do form two separate orifices they remain abnormal valves. The VSD is often restrictive.
120	AVC (AVSD), Partial (incomplete) (PAVSD) (ASD, primum)	An AVC with an ostium primum ASD (a usually crescent-shaped ASD in the inferior (posterior) portion of the atrial septum just above the AV valve) and varying degrees of malformation of the left AV valve leading to varying degrees of left AV valve regurgitation. No VSD is present.
140	AP window (aortopulmonary window)	Indicate if the patient has the diagnosis of "AP window (aortopulmonary window)". An "AP window (aortopulmonary window)" is defined as a defect with side-to-side continuity of the lumens of the aorta and pulmonary arterial tree, which is distinguished from common arterial trunk (truncus arteriosus) by the presence of two arterial valves or their atretic remnants. (In other words, an aortopulmonary window is a communication between the main pulmonary artery and ascending aorta in the presence of two separate semilunar [pulmonary and aortic] valves. The presence of two separate semilunar valves distinguishes AP window from truncus arteriosus. Type 1 proximal defect: AP window located just above the sinus of Valsalva, a few millimeters above the semilunar valves, with a superior rim but little inferior rim separating the AP window from the semilunar valves. Type 2 distal defect: AP window located in the uppermost portion of the ascending aorta, with a well-formed inferior rim but little superior rim. Type 3 total defect: AP window involving the majority of the ascending aorta, with little superior and inferior rims. The intermediate type of AP window is similar to the total defect but with adequate superior and inferior rims. In the event of AP window occurring in association with interrupted aortic arch, code "Interrupted aortic arch + AP window (aortopulmonary window)", and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and AP window separately to provide further documentation about the individual interrupted arch and AP window types.)
150	Pulmonary artery origin from ascending aorta (hemitruncus)	One pulmonary artery arises from the ascending aorta and the other pulmonary artery arises from the right ventricle. DOES NOT include origin of the right or left pulmonary artery from the innominate artery or the aortic arch via a patent ductus arteriosus or collateral artery.
160	Truncus arteriosus	Indicate if the patient has the diagnosis of "Truncus arteriosus". A truncus arteriosus is also known as a common arterial trunk and is defined as a heart in which a single arterial trunk arises from the heart, giving origin to the coronary arteries, the pulmonary arteries, and the systemic arterial circulation. In the majority of instances there is a ventricular septal defect and a single semilunar valve which may contain two, three, four, or more leaflets and is occasionally dysplastic. Often, the infundibular septum is virtually absent superiorly. In most instances the truncal valve overrides the true interventricular septum (and thus both ventricles), but very rarely the truncal valve may override the right ventricle entirely. In such instances, there may be no ventricular septal defect or a very small

160		ventricular septal defect, in which case the left ventricle and mitral valve may be extremely hypoplastic.
170	Truncal valve insufficiency	Functional abnormality - insufficiency - of the truncal valve. May be further subdivided into grade of insufficiency (I, II, III, IV or mild, moderate, severe).
2470	Truncal valve stenosis	
2010	Truncus arteriosus + Interrupted aortic arch	Indicate if the patient has the diagnosis of "Truncus arteriosus + Interrupted aortic arch". {A truncus arteriosus is also known as a common arterial trunk and is defined as a heart in which a single arterial trunk arises from the heart, giving origin to the coronary arteries, the pulmonary arteries, and the systemic arterial circulation. In the majority of instances there is a ventricular septal defect and a single semilunar valve which may contain two, three, four, or more leaflets and is occasionally dysplastic. The infundibular septum is virtually absent superiorly. In most instances the truncal valve overrides the true interventricular septum (and thus both ventricles), but very rarely the truncal valve may override the right ventricle entirely. If in such case there is no ventricular septal defect, then the left ventricle and mitral valve may be extremely hypoplastic.} {Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.}
180	Partial anomalous pulmonary venous connection (PAPVC)	Some, but not all of the pulmonary veins connect to the right atrium or to one or more of its venous tributaries. This definition excludes sinus venosus defects with normally connected but abnormally draining pulmonary veins (the pulmonary veins may drain abnormally into the right atrium via the atrial septal defect).
190	Partial anomalous pulmonary venous connection (PAPVC), scimitar	The right pulmonary vein(s) connect anomalously to the inferior vena cava or to the right atrium at the insertion of the inferior vena cava. The descending vertical vein resembles a scimitar (Turkish sword) on frontal chest x-ray. Frequently associated with: hypoplasia of the right lung with bronchial anomalies; dextroposition and/or dextrorotation of the heart; hypoplasia of the right pulmonary artery; and anomalous subdiaphragmatic systemic arterial supply to the lower lobe of the right lung directly from the aorta or its main branches.
200	Total anomalous pulmonary venous connection (TAPVC), Type 1 (supracardiac)	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium. In Type 1 (supracardiac) TAPVC, the anomalous connection is at the supracardiac level and can be obstructed or nonobstructed.
210	Total anomalous pulmonary venous connection (TAPVC), Type 2	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries.

210	(cardiac)	None of the pulmonary veins connect normally to the left atrium. In Type 2 (cardiac) TAPVC, the anomalous connection is to the heart, either to the right atrium directly or to the coronary sinus. Most patients with type 2 TAPVC are nonobstructed.
220	Total anomalous pulmonary venous connection (TAPVC), Type 3 (infracardiac)	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium. In Type 3 (infracardiac) TAPVC, the anomalous connection is at the infracardiac level (below the diaphragm), with the pulmonary venous return entering the right atrium ultimately via the inferior vena cava. In the vast majority of patients infracardiac TAPVC is obstructed.
230	Total anomalous pulmonary venous connection (TAPVC), Type 4 (mixed)	All of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of the pulmonary veins connect normally to the left atrium. In Type 4 (mixed) TAPVC, the anomalous connection is at two or more of the above levels (supracardiac, cardiac, infracardiac) and can be obstructed or nonobstructed.
250	Cor triatriatum	In the classic form of cor triatriatum a membrane divides the left atrium (LA) into a posterior accessory chamber that receives the pulmonary veins and an anterior chamber (LA) that communicates with the mitral valve. In differentiating cor triatriatum from supravalvar mitral ring, in cor triatriatum the posterior compartment contains the pulmonary veins while the anterior contains the left atrial appendage and the mitral valve orifice; in supravalvar mitral ring, the anterior compartment contains only the mitral valve orifice. Cor triatriatum dexter (prominent venous valve producing obstruction of the IVC and tricuspid valve) is to be coded as a systemic venous obstruction, not as a form of cor triatriatum.
260	Pulmonary venous stenosis	Any pathologic narrowing of one or more pulmonary veins. Can be further subdivided by etiology (congenital, acquired-postoperative, acquired-nonpostoperative) and extent of stenosis (diffusely hypoplastic, long segment focal/tubular stenosis, discrete stenosis).
2480	Pulmonary venous stenosis, Acquired	
2490	Pulmonary venous stenosis, Spontaneous	
270	Systemic venous anomaly	Anomalies of the systemic venous system (superior vena cava (SVC), inferior vena cava (IVC), brachiocephalic veins (often the innominate vein), azygos vein, coronary sinus, levo-atrial cardinal vein) arising from one or more anomalies of origin, duplication, course, or connection. Examples include abnormal or absent right SVC with LSVC, bilateral SVC, interrupted right or left IVC, azygos continuation of IVC, and anomalies of hepatic drainage. Bilateral SVC may have, among other configurations: 1) RSVC draining to the RA and the LSVC to the LA with completely unroofed coronary sinus, 2) RSVC draining to the RA and LSVC to the coronary sinus which drains (normally) into the RA, or 3) RSVC to the coronary sinus which drains

270		(abnormally) into the LA and LSVC to LA. Anomalies of the inferior vena caval system include, among others: 1) left IVC to LA, 2) biatrial drainage, or 3) interrupted IVC (left or right) with azygos continuation to an LSVC or RSVC.
280	Systemic venous obstruction	Obstruction of the systemic venous system (superior vena cava (SVC), inferior vena cava (IVC), brachiocephalic veins (often the innominate vein), azygos vein, coronary sinus, levo-atrial cardinal vein) arising from congenital or acquired stenosis or occlusion. Cor triatriatum dexter (prominent venous valve producing obstruction of the IVC and tricuspid valve) is to be coded as a systemic venous obstruction, not as a form of cor triatriatum.
290	TOF	Indicate if the patient has the diagnosis of “TOF”. Only use this diagnosis if it is NOT known if the patient has one of the following four more specific diagnoses: (1). “TOF, Pulmonary stenosis”, (2). “TOF, AVC (AVSD)”, (3). “TOF, Absent pulmonary valve”, (4). “Pulmonary atresia, VSD (Including TOF, PA)”, or (5). “Pulmonary atresia, VSD-MAPCA (pseudotruncus)”.{“TOF” is “Tetralogy of Fallot” and is defined as a group of malformations with biventricular atrioventricular alignments or connections characterized by anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta. Hearts with tetralogy of Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, and aortic override; hearts with tetralogy of Fallot will most often have right ventricular hypertrophy.} (An additional, often muscular [Type 4] VSD may be seen with TOF and should be coded separately as a secondary diagnosis as “VSD, Type 4 (Muscular)”. Pulmonary arteries may be diminutive or there may be an absent left or right pulmonary artery; additional coding for pulmonary artery and/or branch pulmonary artery stenoses may be found under RVOT obstruction. Abnormal coronary artery distribution may also be associated with tetralogy of Fallot and may be coded separately under coronary artery anomalies. The presence of associated anomalies such as additional VSD, atrial septal defect, right aortic arch, left superior vena cava, and coronary artery anomalies must be subspecified as an additional or secondary diagnosis under the primary TOF diagnosis. TOF with absent pulmonary valve or TOF with associated complete atrioventricular canal are NOT to be secondary diagnoses under TOF - they are separate entities and should be coded as such. Controversy surrounds the differentiation between TOF and double outlet right ventricle [DORV]; in the nomenclature used here, DORV is defined as a type of ventriculoarterial connection in which both great vessels arise predominantly from the right ventricle. TOF with pulmonary atresia is to be coded under "Pulmonary atresia-VSD.")
2140	TOF, Pulmonary stenosis	Indicate if the patient has the diagnosis of “TOF, Pulmonary stenosis”. Use this diagnosis if the patient has tetralogy of Fallot and pulmonary stenosis. Do not use this diagnosis if the patient has tetralogy of Fallot and pulmonary atresia.

2140		Do not use this diagnosis if the patient has tetralogy of Fallot and absent pulmonary valve. Do not use this diagnosis if the patient has tetralogy of Fallot and atrioventricular canal. {Tetralogy of Fallot is defined as a group of malformations with biventricular atrioventricular alignments or connections characterized by anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta. Hearts with tetralogy of Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, and aortic override; hearts with tetralogy of Fallot will most often have right ventricular hypertrophy. (An additional, often muscular [Type 4] VSD may be seen with TOF and should be coded separately as a secondary diagnosis as “VSD, Type 4 (Muscular)”. Pulmonary arteries may be diminutive or there may be an absent left or right pulmonary artery; additional coding for pulmonary artery and/or branch pulmonary artery stenoses may be found under RVOT obstruction. Abnormal coronary artery distribution may also be associated with tetralogy of Fallot and may be coded separately under coronary artery anomalies. The presence of associated anomalies such as additional VSD, atrial septal defect, right aortic arch, left superior vena cava, and coronary artery anomalies must be subspecified as an additional or secondary diagnosis under the primary TOF diagnosis. TOF with absent pulmonary valve or TOF with associated complete atrioventricular canal are NOT to be secondary diagnoses under TOF - they are separate entities and should be coded as such. Controversy surrounds the differentiation between TOF and double outlet right ventricle [DORV]; in the nomenclature used here, DORV is defined as a type of ventriculoarterial connection in which both great vessels arise predominantly from the right ventricle. TOF with pulmonary atresia is to be coded under "Pulmonary atresia-VSD.")}
300	TOF, AVC (AVSD)	TOF with complete common atrioventricular canal defect is a rare variant of common atrioventricular canal defect with the associated conotruncal abnormality of TOF. The anatomy of the endocardial cushion defect is that of Rastelli type C in almost all cases.
310	TOF, Absent pulmonary valve	Indicate if the patient has the diagnosis of “TOF, Absent pulmonary valve”. “TOF, Absent pulmonary valve” is “Tetralogy of Fallot with Absent pulmonary valve” and is defined as a malformation with all of the morphologic characteristics of tetralogy of Fallot (anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta), in which the ventriculo-arterial junction of the right ventricle with the main pulmonary artery features an atypical valve with rudimentary cusps that lack the anatomical semi-lunar features of normal valve cusps and which functionally do not achieve central coaptation. The physiologic consequence is usually a combination of variable degrees of

310		both stenosis and regurgitation of the pulmonary valve. A developmental accompaniment of this anatomy and physiology is dilatation of the main pulmonary artery and central right and left pulmonary arteries, which when extreme, is associated with abnormal arborization of lobar and segmental pulmonary artery branches and with compression of the trachea and mainstem bronchi. One theory holds that absence of the arterial duct or ductal ligament (which is a nearly constant finding in cases of tetralogy of Fallot with absent pulmonary valve) in combination with pulmonary `valve stenosis and regurgitation, comprise the physiologic conditions which predispose to central pulmonary artery dilatation during fetal development. (Tetralogy of Fallot with Absent Pulmonary Valve Syndrome is a term frequently used to describe the clinical presentation when it features both circulatory alterations and respiratory distress secondary to airway compression.)
320	Pulmonary atresia	Pulmonary atresia defects which do not readily fall into pulmonary atresia-intact ventricular septum or pulmonary atresia-VSD (with or without MAPCAs) categories. These may include complex lesions in which pulmonary atresia is a secondary diagnosis, for example, complex single ventricle malformations with associated pulmonary atresia.
330	Pulmonary atresia, IVS	Pulmonary atresia (PA) and intact ventricular septum (IVS) is a duct-dependent congenital malformation that forms a spectrum of lesions including atresia of the pulmonary valve, a varying degree of right ventricle and tricuspid valve hypoplasia, and anomalies of the coronary circulation. An RV dependent coronary artery circulation is present when coronary artery fistulas (coronary sinusoids) are associated with a proximal coronary artery stenosis. Associated Ebstein's anomaly of the tricuspid valve can be present; the tricuspid diameter is enlarged and the prognosis is poor.
340	Pulmonary atresia, VSD (Including TOF, PA)	Pulmonary atresia (PA) and ventricular septal defect (VSD) is a heterogeneous group of congenital cardiac malformations in which there is lack of luminal continuity and absence of blood flow from either ventricle (in cases with ventriculo-arterial discordance) and the pulmonary artery, in a biventricular heart that has an opening or a hole in the interventricular septum (VSD). The malformation forms a spectrum of lesions including tetralogy of Fallot with pulmonary atresia. Tetralogy of Fallot with PA is a specific type of PA-VSD where the intracardiac malformation is more accurately defined (extreme underdevelopment of the RV infundibulum with marked anterior and leftward displacement of the infundibular septum often fused with the anterior wall of the RV resulting in complete obstruction of blood flow into the pulmonary artery and associated with a large outlet, subaortic ventricular septal defect). In the vast majority of cases of PA-VSD the intracardiac anatomy is that of TOF. The pulmonary circulation in PA-VSD is variable in terms of origin of blood flow, presence or absence of native pulmonary arteries, presence or absence of major aortopulmonary collateral arteries (MAPCA(s)), and distal distribution (pulmonary parenchymal segment arborization)

340		abnormalities. Native pulmonary arteries may be present or absent. If MAPCAs are present this code should not be used; instead, Pulmonary atresia, VSD-MAPCA (pseudotruncus) should be used.
350	Pulmonary atresia, VSD-MAPCA	MAPCA(s) are large and distinct arteries, highly variable in number, that usually arise from the descending thoracic aorta, but uncommonly may originate from the aortic arch or the subclavian, carotid or even the coronary arteries. MAPCA(s) may be associated with present or absent native pulmonary arteries. If present, the native pulmonary arteries may be hypoplastic, and either confluent or nonconfluent. Systemic pulmonary collateral arteries have been categorized into 3 types based on their site of origin and the way they connect to the pulmonary circulation: direct aortopulmonary collaterals, indirect aortopulmonary collaterals, and true bronchial arteries. Only the first two should be considered MAPCA(s). If MAPCA(s) are associated with PA-VSD or TOF, PA this code should be used.
360	MAPCA(s) (major aortopulmonary collateral[s]) (without PA-VSD)	Rarely MAPCA(s) may occur in patents who do not have PA-VSD, but have severe pulmonary stenosis. The intracardiac anatomy in patients who have MAPCA(s) without PA should be specifically coded in each case as well.
370	Ebstein's anomaly	Indicate if the patient has the diagnosis of “Ebstein's anomaly”. Ebstein's anomaly is a malformation of the tricuspid valve and right ventricle that is characterized by a spectrum of several features: (1) incomplete delamination of tricuspid valve leaflets from the myocardium of the right ventricle; (2) downward (apical) displacement of the functional annulus; (3) dilation of the "atrialized" portion of the right ventricle with variable degrees of hypertrophy and thinning of the wall; (4) redundancy, fenestrations, and tethering of the anterior leaflets; and (5) dilation of the right atrioventricular junction (the true tricuspid annulus). These anatomical and functional abnormalities cause tricuspid regurgitation (and rarely tricuspid stenosis) that results in right atrial and right ventricular dilatation and atrial and ventricular arrhythmias. With increasing degrees of anatomic severity of malformation, the fibrous transformation of leaflets from their muscular precursors remains incomplete, with the septal leaflet being most severely involved, the posterior leaflet less severely involved, and the anterior leaflet usually the least severely involved. Associated cardiac anomalies include an interatrial communication, the presence of accessory conduction pathways often associated with Wolff-Parkinson-White syndrome, and dilation of the right atrium and right ventricle in patients with severe Ebstein's anomaly. (Varying degrees of right ventricular outflow tract obstruction may be present, including pulmonary atresia in some cases. Such cases of Ebstein's anomaly with pulmonary atresia should be coded with a Primary Diagnosis of “Ebstein’s anomaly”, and a Secondary Diagnosis of “Pulmonary atresia”.) (Some patients with atrioventricular discordance and ventriculoarterial discordance in situs solitus [congenitally corrected transposition] have an Ebstein-like deformity of the left-

370		sided morphologically tricuspid valve. The nature of the displacement of the septal and posterior leaflets is similar to that in right-sided Ebstein's anomaly in patients with atrioventricular concordance and ventriculoarterial concordance in situs solitus. These patients with "Congenitally corrected TGA" and an Ebstein-like deformity of the left-sided morphologically tricuspid valve should be coded with a Primary Diagnosis of "Congenitally corrected TGA", and a Secondary Diagnosis of "Ebstein's anomaly".)
380	Tricuspid regurgitation, non-Ebstein's related	Non-Ebstein's tricuspid regurgitation may be due to congenital factors (primary annular dilation, prolapse, leaflet underdevelopment, absent papillary muscle/chordae) or acquired (post cardiac surgery or secondary to rheumatic fever, endocarditis, trauma, tumor, cardiomyopathy, iatrogenic or other causes).
390	Tricuspid stenosis	Tricuspid stenosis may be due to congenital factors (valvar hypoplasia, abnormal subvalvar apparatus, double-orifice valve, parachute deformity) or acquired (post cardiac surgery or secondary to carcinoid, rheumatic fever, tumor, systemic disease, iatrogenic, or other causes).
400	Tricuspid regurgitation and tricuspid stenosis	Tricuspid regurgitation present with tricuspid stenosis may be due to congenital factors or acquired.
410	Tricuspid valve, Other	Tricuspid valve pathology not otherwise specified in diagnosis definitions 370, 380, 390 and 400.
420	Pulmonary stenosis, Valvar	Pulmonary stenosis, Valvar ranges from critical neonatal pulmonic valve stenosis with hypoplasia of the right ventricle to valvar pulmonary stenosis in the infant, child, or adult, usually better tolerated but potentially associated with infundibular stenosis. Pulmonary branch hypoplasia can be associated. Only 10% of neonates with Pulmonary stenosis, Valvar with intact ventricular septum have RV-to-coronary artery fistula(s). An RV dependent coronary artery circulation is present when coronary artery fistulas (coronary sinusoids) are associated with a proximal coronary artery stenosis; this occurs in only 2% of neonates with Pulmonary stenosis, Valvar with IVS.
430	Pulmonary artery stenosis (hypoplasia), Main (trunk)	Indicate if the patient has the diagnosis of "Pulmonary artery stenosis (hypoplasia), Main (trunk)". "Pulmonary artery stenosis (hypoplasia), Main (trunk)" is defined as a congenital or acquired anomaly with pulmonary trunk (main pulmonary artery) narrowing or hypoplasia. The stenosis or hypoplasia may be isolated or associated with other cardiac lesions. Since the narrowing is distal to the pulmonic valve, it may also be known as supravalvar pulmonary stenosis.
440	Pulmonary artery stenosis, Branch, Central (within the hilar bifurcation)	Indicate if the patient has the diagnosis of "Pulmonary artery stenosis, Branch, Central (within the hilar bifurcation)". "Pulmonary artery stenosis, Branch, Central (within the hilar bifurcation)" is defined as a congenital or acquired anomaly with central pulmonary artery branch (within the hilar bifurcation involving the right or left pulmonary artery, or both) narrowing or hypoplasia. The stenosis or hypoplasia may be isolated or associated with other cardiac lesions. Coarctation of the pulmonary artery is related to abnormal extension of the ductus arteriosus into a pulmonary branch, more frequently the left branch.

450	Pulmonary artery stenosis, Branch, Peripheral (at or beyond the hilar bifurcation)	Indicate if the patient has the diagnosis of "Pulmonary artery stenosis, Branch, Peripheral (at or beyond the hilar bifurcation)". "Pulmonary artery stenosis, Branch, Peripheral (at or beyond the hilar bifurcation)" is defined as a congenital or acquired anomaly with peripheral pulmonary artery narrowing or hypoplasia (at or beyond the hilar bifurcation). The stenosis or hypoplasia may be isolated or associated with other cardiac lesions.
470	Pulmonary artery, Discontinuous	Indicate if the patient has the diagnosis of "Pulmonary artery, Discontinuous". Pulmonary artery, Discontinuous" is defined as a congenital or acquired anomaly with discontinuity between the branch pulmonary arteries or between a branch pulmonary artery and the main pulmonary artery trunk.
490	Pulmonary stenosis, Subvalvar	Subvalvar (infundibular) pulmonary stenosis is a narrowing of the outflow tract of the right ventricle below the pulmonic valve. It may be due to a localized fibrous diaphragm just below the valve, an obstructing muscle bundle or to a long narrow fibromuscular channel.
500	DCRV	The double chambered right ventricle is characterized by a low infundibular (subvalvar) stenosis rather than the rare isolated infundibular stenosis that develops more superiorly in the infundibulum, and is often associated with one or several closing VSDs. In some cases, the VSD is already closed. The stenosis creates two chambers in the RV, one inferior including the inlet and trabecular portions of the RV and one superior including the infundibulum.
510	Pulmonary valve, Other	Other anomalies of the pulmonary valve may be listed here including but not restricted to absent pulmonary valve.
530	Pulmonary insufficiency	Pulmonary valve insufficiency or regurgitation may be due to congenital factors (primary annular dilation, prolapse, leaflet underdevelopment, etc.) or acquired (for example, post cardiac surgery for repair of tetralogy of Fallot, etc.).
540	Pulmonary insufficiency and pulmonary stenosis	Pulmonary valve insufficiency and pulmonary stenosis beyond the neonatal period, in infancy and childhood, may be secondary to leaflet tissue that has become thickened and myxomatous. Retraction of the commissure attachment frequently creates an associated supravalvar stenosis.
2130	Shunt failure	Indicate if the patient has the diagnosis of "Shunt failure". This diagnostic subgroup includes failure of any of a variety of shunts ("Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS)", "Shunt, Systemic to pulmonary, Central (from aorta or to main pulmonary artery)", "Shunt, Systemic to pulmonary, Other", and "Sano Shunt"), secondary to any of the following etiologies: shunt thrombosis, shunt occlusion, shunt stenosis, shunt obstruction, and shunt outgrowth. This diagnosis ("Shunt failure") would be the primary diagnosis in a patient with, for example, "Hypoplastic left heart syndrome (HLHS)" who underwent a "Norwood procedure" with a "Modified Blalock-Taussig Shunt" and now requires reoperation for thrombosis of the "Modified Blalock-Taussig Shunt". The underlying or fundamental diagnosis in this patient is "Hypoplastic left heart syndrome (HLHS)", but the primary

2130		diagnosis for the operation to be performed to treat the thrombosis of the “Modified Blalock-Taussig Shunt” would be “Shunt failure”. Please note that the choice “2130 Shunt failure” does not include “520 Conduit failure”.
520	Conduit failure	Indicate if the patient has the diagnosis of “Conduit failure”. This diagnostic subgroup includes failure of any of a variety of conduits (ventricular [right or left]-to-PA conduits, as well as a variety of other types of conduits [ventricular {right or left}-to-aorta, RA-to-RV, etc.]), secondary to any of the following etiologies: conduit outgrowth, obstruction, stenosis, insufficiency, or insufficiency and stenosis. This diagnosis (“Conduit failure”) would be the primary diagnosis in a patient with, for example, “Truncus arteriosus” repaired in infancy who years later is hospitalized because of conduit stenosis/insufficiency. The underlying or fundamental diagnosis in this patient is “Truncus arteriosus”, but the primary diagnosis for the operation to be performed during the hospitalization (in this case, “Conduit reoperation”) would be “Conduit failure”. Please note that the choice “520 Conduit failure” does not include “2130 Shunt failure”.
550	Aortic stenosis, Subvalvar	Subaortic obstruction can be caused by different lesions: subaortic membrane or tunnel, accessory mitral valve tissue, abnormal insertion of the mitral anterior leaflet to the ventricular septum, deviation of the outlet septum (seen in coarctation of the aorta and interrupted aortic arch), or a restrictive bulboventricular foramen in single ventricle complexes. The Shone complex consists of subvalvar aortic stenosis in association with supravalvar mitral ring, parachute mitral valve, and coarctation of aorta. Subvalvar aortic stenosis may be categorized into two types: localized subvalvar aortic stenosis, which consists of a fibrous or fibromuscular ridge, and diffuse tunnel subvalvar aortic stenosis, in which circumferential narrowing commences at the annular level and extends downward for 1-3 cm. Idiopathic hypertrophic subaortic stenosis (IHSS) is also known as hypertrophic obstructive cardiomyopathy (HOCM), and is characterized by a primary hypertrophy of the myocardium. The obstructive forms involve different degrees of dynamic subvalvar aortic obstruction from a thickened ventricular wall and anterior motion of the mitral valve. Definitive nomenclature and therapeutic options for IHSS are listed under cardiomyopathy.
2500	Aortic Stenosis, Subvalvar, Discrete	
2510	Aortic Stenosis, Subvalvar, IHSS	
2520	Aortic Stenosis, Subvalvar, Tunnel-like	
560	Aortic stenosis, Valvar	Valvar aortic stenosis may be congenital or acquired. In its congenital form there are two types: critical (infantile), seen in the newborn in whom systemic perfusion depends

560		on a patent ductus arteriosus, and noncritical, seen in infancy or later. Acquired valvar stenosis may be seen after as a result of rheumatic valvar disease, or from stenotic changes of an aortic valve prosthesis. Congenital valvar stenosis may result: (1) from complete fusion of commissures (acommissural) that results in a dome-shaped valve with a pinpoint opening (seen most commonly in infants with critical aortic valve stenosis); (2) from a unicommissural valve with one defined commissure and eccentric orifice (often with two raphes radiating from the ostium indicating underdeveloped commissures of a tricuspid aortic valve); (3) from a bicuspid aortic valve, with leaflets that can be equal in size or discrepant, and in left-right or anterior-posterior position; and finally (4) from a dysplastic tricuspid valve, which may have a gelatinous appearance with thick rarely equal in size leaflets, often obscuring the commissures. The dysplastic, tricuspid or bicuspid form of aortic valve deformity may not be initially obstructive but may become stenotic later in life due to leaflet thickening and calcification.
570	Aortic stenosis, Supravalvar	Congenital supravalvar aortic stenosis is described as three forms: an hourglass deformity, a fibrous membrane, and a diffuse narrowing of the ascending aorta. The disease can be inherited as an autosomal dominant trait or part of Williams-Beuren syndrome in association with mental retardation, elfin facies, failure to thrive, and occasionally infantile hypercalcemia. Supravalvar aortic stenosis may involve the coronary artery ostia, and the aortic leaflets may be tethered. The coronary arteries can become tortuous and dilated due to elevated pressures and early atherosclerosis may ensue. Supravalvar aortic stenosis may also be acquired: (1) after a neoaortic reconstruction such as arterial switch, Ross operation, or Norwood procedure; (2) at a suture line from a previous aortotomy or cannulation; and (3) from a narrowed conduit.
590	Aortic valve atresia	Aortic valve atresia will most often be coded under the Hypoplastic left heart syndrome/complex diagnostic codes since it most often occurs as part of a spectrum of cardiac malformations. However, there is a small subset of patients with aortic valve atresia who have a well-developed left ventricle and mitral valve and a large VSD (nonrestrictive or restrictive). The diagnostic code "Aortic valve atresia" enables users to report those patients with aortic valve atresia and a well-developed systemic ventricle without recourse to either a hypoplastic left heart syndrome/complex diagnosis or a single ventricle diagnosis.
600	Aortic insufficiency	Congenital aortic regurgitation/insufficiency is rare as an isolated entity. There are rare reports of congenital malformation of the aortic valve that result in aortic insufficiency shortly after birth from an absent or underdeveloped aortic valve cusp. Aortic insufficiency is more commonly seen with other associated cardiac anomalies: (1) in stenotic aortic valves (commonly stenotic congenital bicuspid aortic valves) with some degree of aortic regurgitation due to aortic leaflet abnormality; (2) in association with a VSD (especially in supracristal or conal

600		type I VSD, more commonly seen in Asian populations); (3) secondary to aortic-left ventricular tunnel; (4) secondary to tethering or retraction of aortic valve leaflets in cases of supravalvar aortic stenosis that may involve the aortic valve; and similarly (5) secondary to encroachment on an aortic cusp by a subaortic membrane; or (6) turbulence caused by a stenotic jet can create progressive aortic regurgitation. Aortic insufficiency may also result from: (1) post-procedure such as closed or open valvotomy or aortic valve repair, VSD closure, balloon valvotomy, or diagnostic catheterization; (2) in the neo-aorta post arterial switch, pulmonary autograft (Ross) procedure, homograft placement, Norwood procedure, or Damus-Kaye-Stansel procedure; (3) as a result of endocarditis secondary to perforated or prolapsed leaflets or annular dehiscence; (4) secondary to annulo-aortic ectasia with prolapsed or noncoapting leaflets; (5) secondary to trauma, blunt or penetrating; or (6) as a result of aortitis, bacterial, viral or autoimmune. Aortic regurgitation secondary to prosthetic failure should be coded first as either conduit failure or prosthetic valve failure, as applicable, and secondarily as aortic regurgitation secondary to prosthetic failure (perivalvar or due to structural failure). The underlying fundamental diagnosis that led to the initial conduit or valve prosthesis placement should also be described.
610	Aortic insufficiency and aortic stenosis	Aortic insufficiency is often seen in association with stenotic aortic valve, commonly the stenotic congenital bicuspid aortic valve. The degree of aortic regurgitation is due to the severity of the aortic leaflet abnormality.
620	Aortic valve, Other	This diagnostic subgroup may be used to delineate aortic valve cusp number (unicuspid, bicuspid, tricuspid, more than three cusps), commissural fusion (normal, partially fused, completely fused), and valve leaflet (normal, thickened, dysplastic, calcified, gelatinous), annulus (normal, hypoplastic, calcified), or sinus description (normal, dilated). Note that any extensive descriptors chosen within those made available by a vendor will be converted, at harvest, to Aortic valve, Other.
630	Sinus of Valsalva aneurysm	The sinus of Valsalva is defined as that portion of the aortic root between the aortic root annulus and the sinotubular ridge. A congenital sinus of Valsalva aneurysm is a dilation usually of a single sinus of Valsalva. These most commonly originate from the right sinus (65%-85%), less commonly from the noncoronary sinus (10%-30%), and rarely from the left sinus (<5%). A true sinus of Valsalva aneurysm presents above the aortic annulus. The hierarchical coding system distinguishes between congenital versus acquired, ruptured versus nonruptured, sinus of origin, and chamber/site of penetration (right atrium, right ventricle, left atrium, left ventricle, pulmonary artery, pericardium). A nonruptured congenital sinus of Valsalva aneurysm may vary from a mild dilation of a single aortic sinus to an extensive windsock deformity. Rupture of a congenital sinus of Valsalva aneurysm into an adjacent chamber occurs most commonly between the ages of 15-30 years. Rupture may occur spontaneously, after trauma, after strenuous physical

630		exertion, or from acute bacterial endocarditis. Congenital etiology is supported by the frequent association of sinus of Valsalva aneurysms with VSDs. Other disease processes are also associated with sinus of Valsalva aneurysm and include: syphilis, endocarditis, cystic medial necrosis, atherosclerosis, and trauma. Acquired sinus of Valsalva aneurysms more frequently involve multiple sinuses of Valsalva; when present in multiple form they are more appropriately classified as aneurysms of the aortic root.
640	LV to aorta tunnel	The aortico-left ventricular tunnel (LV-to-aorta tunnel) is an abnormal paravalvular (alongside or in the vicinity of a valve) communication between the aorta and left ventricle, commonly divided into 4 types: (1) type I, a simple tunnel with a slit-like opening at the aortic end and no aortic valve distortion; (2) type II, a large extracardiac aortic wall aneurysm of the tunnel with an oval opening at the aortic end, with or without ventricular distortion; (3) type III, intracardiac aneurysm of the septal portion of the tunnel, with or without right ventricular outflow obstruction; and (4) type IV, a combination of types II and III. Further differentiation within these types may be notation of right coronary artery arising from the wall of the tunnel. If a LV-to-aorta tunnel communicates with the right ventricle, many feel that the defect is really a ruptured sinus of Valsalva aneurysm.
650	Mitral stenosis, Supravalvar mitral ring	Supravalvar mitral ring is formed by a circumferential ridge of tissue that is attached to the anterior mitral valve leaflet (also known as the aortic leaflet) slightly below its insertion on the annulus and to the atrium slightly above the attachment of the posterior mitral valve leaflet (also known as the mural leaflet). Depending on the diameter of the ring orifice, varying degrees of obstruction exist. The underlying valve is usually abnormal and frequently stenotic or hypoplastic. Supravalvar mitral ring is commonly associated with other stenotic lesions such as parachute or hammock valve (subvalvar stenosis), papillary muscle fusion (subvalvar stenosis), and double orifice mitral valve (valvar stenosis). Differentiation from cor triatriatum focuses on the compartments created by the supravalvar ring. In cor triatriatum the posterior compartment contains the pulmonary veins; the anterior contains the left atrial appendage and the mitral valve orifice. In supravalvar mitral ring, the posterior compartment contains the pulmonary veins and the left atrial appendage; the anterior compartment contains only the mitral valve orifice. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
660	Mitral stenosis, Valvar	Valvar mitral stenosis may arise from congenital (annular and / or leaflet) or acquired causes, both surgical (after mitral valve repair or replacement or other cardiac surgery) and non-surgical (post rheumatic heart disease, infective endocarditis, ischemia, myxomatous degeneration, trauma, or cardiomyopathy). Mitral valve annular hypoplasia is distinguished from severe mitral valve hypoplasia and mitral

660		valve atresia, which are typically components of hypoplastic left heart syndrome. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
670	Mitral stenosis, Subvalvar	Congenital subvalvar mitral stenosis may be due to obstructive pathology of either the chordae tendineae and / or papillary muscles which support the valve leaflets. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
680	Mitral stenosis, Subvalvar, Parachute	In parachute mitral valve, all chordae are attached to a single papillary muscle originating from the posterior ventricular wall. When the interchordal spaces are partially obliterated valvar stenosis results. This defect also causes valvar insufficiency, most commonly due to a cleft leaflet, a poorly developed anterior leaflet, short chordae, or annular dilatation. This lesion is also part of Shone's anomaly, which consists of the parachute mitral valve, supravalvar mitral ring, subaortic stenosis, and coarctation of the aorta. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
695	Mitral stenosis	Stenotic lesions of the mitral valve not otherwise specified in the diagnosis definitions 650, 660, 670, and 680.
700	Mitral regurgitation and mitral stenosis	Mitral regurgitation and mitral stenosis may arise from congenital or acquired causes or after cardiac surgery. Additional details to aid in coding specific components of the diagnosis are available in the individual mitral stenosis or mitral regurgitation field definitions. When coding multiple mitral valve lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
710	Mitral regurgitation	Mitral regurgitation may arise from congenital (at the annular, leaflet or subvalvar level) or acquired causes both surgical (after mitral valve repair or replacement, subaortic stenosis repair, atrioventricular canal repair, cardiac transplantation, or other cardiac surgery) and non-surgical (post rheumatic heart disease, infective endocarditis, ischemia (with chordal rupture or papillary muscle infarct), myxomatous degeneration including Barlow's syndrome, trauma, or cardiomyopathy). Congenital lesions at the annular level include annular dilatation or deformation (usually deformation is consequent to associated lesions). At the valve leaflet level, mitral regurgitation may be due to a cleft, hypoplasia or agenesis of leaflet(s), excessive leaflet tissue, or a double orifice valve. At the subvalvar level, mitral regurgitation may be secondary to chordae tendineae anomalies (agenesis, rupture, elongation, or shortening as in funnel valve), or to papillary muscle anomalies (hypoplasia or agenesis, shortening, elongation, single-parachute, or multiple-hammock valve). When coding multiple mitral valvar lesions the predominant defect

710		causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.
720	Mitral valve, Other	Mitral valve pathology not otherwise coded in diagnosis definitions 650 through 710.
730	Hypoplastic left heart syndrome (HLHS)	Hypoplastic left heart syndrome (HLHS) is a spectrum of cardiac malformations characterized by a severe underdevelopment of the left heart-aorta complex, consisting of aortic and/or mitral valve atresia, stenosis, or hypoplasia with marked hypoplasia or absence of the left ventricle, and hypoplasia of the ascending aorta and of the aortic arch with coarctation of the aorta. Hypoplastic left heart complex is a subset of patients at the favorable end of the spectrum of HLHS characterized by hypoplasia of the structures of the left heart-aorta complex, consisting of aortic and mitral valve hypoplasia without valve stenosis or atresia, hypoplasia of the left ventricle, hypoplasia of the left ventricular outflow tract, hypoplasia of the ascending aorta and of the aortic arch, with or without coarctation of the aorta.
2080	Shone's syndrome	Shone's syndrome is a syndrome of multilevel hypoplasia and obstruction of left sided cardiovascular structures including more than one of the following lesions: (1) supravalvar ring of the left atrium, (2) a parachute deformity of the mitral valve, (3) subaortic stenosis, and (4) aortic coarctation. The syndrome is based on the original report from Shone [1] that was based on analysis of 8 autopsied cases and described the tendency of these four obstructive, or potentially obstructive, conditions to coexist. Only 2 of the 8 cases exhibited all four conditions, with the other cases exhibiting only two or three of the anomalies [2]. [1] Shone JD, Sellers RD, Anderson RG, Adams P, Lillehei CW, Edwards JE. The developmental complex of "parachute mitral valve", supravalvar ring of left atrium, subaortic stenosis, and coarctation of the aorta. Am J Cardiol 1963; 11: 714–725. [2]. Tchervenkov CI, Jacobs JP, Weinberg PM, Aiello VD, Beland MJ, Colan SD, Elliott MJ, Franklin RC, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G. The nomenclature, definition and classification of hypoplastic left heart syndrome. Cardiology in the Young, 2006; 16(4): 339–368, August 2006. Please note that the term "2080 Shone's syndrome" may be the "Fundamental Diagnosis" of a patient; however, the term "2080 Shone's syndrome" may not be the "Primary Diagnosis" of an operation. The term "2080 Shone's syndrome" may be a "Secondary Diagnosis" of an operation.
740	Cardiomyopathy (including dilated, restrictive, and hypertrophic)	Cardiomyopathy is a term applied to a wide spectrum of cardiac diseases in which the predominant feature is poor myocardial function in the absence of any anatomic abnormalities. Cardiomyopathies can be divided into three relatively easily distinguishable entities: (1) dilated, characterized by ventricular dilatation and systolic dysfunction; (2) hypertrophic, characterized by physiologically inappropriate hypertrophy of the left ventricle; and (3) restrictive, characterized by diastolic

740		dysfunction, with a presentation often identical to constrictive pericarditis. Also included in this diagnostic category are patients with a cardiomyopathy or syndrome confined to the right ventricle, for example: (1) arrhythmogenic right ventricular dysplasia; (2) Uhl's syndrome (hypoplasia of right ventricular myocardium, parchment heart); or (3) spongiform cardiomyopathy.
750	Cardiomyopathy, End-stage congenital heart disease	Myocardial abnormality in which there is systolic and/or diastolic dysfunction in the presence of structural congenital heart disease without any (or any further) surgically correctable lesions.
760	Pericardial effusion	Inflammatory stimulation of the pericardium that results in the accumulation of appreciable amounts of pericardial fluid (also known as effusive pericarditis). The effusion may be idiopathic or acquired (e.g., postoperative, infectious, uremic, neoplastic, traumatic, drug-induced).
770	Pericarditis	Inflammatory process of the pericardium that leads to either (1) effusive pericarditis with accumulation of appreciable amounts of pericardial fluid or (2) constrictive pericarditis that leads to pericardial thickening and compression of the cardiac chambers, ultimately with an associated significant reduction in cardiac function. Etiologies are varied and include idiopathic or acquired (e.g., postoperative, infectious, uremic, neoplastic, traumatic, drug-induced) pericarditis.
780	Pericardial disease, Other	A structural or functional abnormality of the visceral or parietal pericardium that may, or may not, have a significant impact on cardiac function. Included are absence or partial defects of the pericardium.
790	Single ventricle, DILV	A congenital cardiac malformation in which both atria connect to a single, morphologically left ventricle. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used

790		whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
800	Single ventricle, DIRV	A congenital cardiac malformation in which both atria connect to a single, morphologically right ventricle The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
810	Single ventricle, Mitral atresia	A congenital cardiac malformation in which there is no orifice of mitral valve

810		The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
820	Single ventricle, Tricuspid atresia	A congenital cardiac malformation in which there is no orifice of tricuspid valve. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically

820		include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
830	Single ventricle, Unbalanced AV canal	Single ventricle anomalies with a common atrioventricular (AV) valve and only one completely well developed ventricle. If the common AV valve opens predominantly into the morphologic left ventricle, the defect is termed a left ventricular (LV)–type or LV-dominant AV septal defect. If the common AV valve opens predominantly into the morphologic right ventricle, the defect is termed a right ventricular (RV)–type or RV-dominant AV septal defect. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally

830		univentricular heart".
840	Single ventricle, Heterotaxia syndrome	"Heterotaxia syndrome" is synonymous with "heterotaxy", "visceral heterotaxy", and "heterotaxy syndrome". Heterotaxy is defined as an abnormality where the internal thoraco-abdominal organs demonstrate abnormal arrangement across the left-right axis of the body. By convention, heterotaxy does not include patients with either the expected usual or normal arrangement of the internal organs along the left-right axis, also known as 'situs solitus', nor patients with complete mirror-imaged arrangement of the internal organs along the left-right axis also known as 'situs inversus'. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
850	Single ventricle, Other	If the single ventricle is of primitive or indeterminate type, other is chosen in coding. It is recognized that a considerable variety of other structural cardiac malformations (e.g., biventricular hearts with straddling atrioventricular valves, pulmonary atresia with intact ventricular septum, some complex forms of double outlet

850		right ventricle) may at times be best managed in a fashion similar to that which is used to treat univentricular hearts. They are not to be coded in this section of the nomenclature, but according to the underlying lesions. The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
851	Single Ventricle + Total anomalous pulmonary venous connection (TAPVC)	Indicate if the patient has the diagnosis of "Single Ventricle + Total anomalous pulmonary venous connection (TAPVC)". In the event of Single Ventricle occurring in association with Total anomalous pulmonary venous connection (TAPVC), code "Single Ventricle + Total anomalous pulmonary venous connection (TAPVC)", and then use additional (secondary) diagnostic codes to describe the Single Ventricle and the Total anomalous pulmonary venous connection (TAPVC) separately to provide further documentation about the Single Ventricle and Total anomalous pulmonary venous connection (TAPVC) types. {"Total anomalous pulmonary venous connection (TAPVC)" is defined as a heart where all of the pulmonary veins connect anomalously with the right atrium or to one or more of its venous tributaries. None of

851		the pulmonary veins connect normally to the left atrium.} The version of the IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS uses the term "single ventricle" as synonymous for the "functionally univentricular heart". The term "functionally univentricular heart" describes a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term "functionally univentricular heart". Reference: Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16, Supplement 1: 9 - 21, February 2006.
870	Congenitally corrected TGA	Indicate if the patient has the diagnosis of “Congenitally corrected TGA”. Congenitally corrected transposition is synonymous with the terms ‘corrected transposition’ and ‘discordant atrioventricular connections with discordant ventriculo-arterial connections’, and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners

870		and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.
872	Congenitally corrected TGA, IVS	Indicate if the patient has the diagnosis of "Congenitally corrected TGA, IVS". "Congenitally corrected TGA, IVS" is "Congenitally corrected transposition with an intact ventricular septum", in other words, "Congenitally corrected transposition with no VSD". (Congenitally corrected transposition is synonymous with the terms 'corrected transposition' and 'discordant atrioventricular connections with discordant ventriculo-arterial connections', and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)
874	Congenitally corrected TGA, IVS-LVOTO	Indicate if the patient has the diagnosis of "Congenitally corrected TGA, IVS-LVOTO". "Congenitally corrected TGA, IVS-LVOTO" is "Congenitally corrected transposition with an intact ventricular septum and left ventricular outflow tract obstruction", in other words, "Congenitally corrected transposition with left ventricular outflow tract obstruction and no VSD". (Congenitally corrected transposition is synonymous with the terms 'corrected transposition' and 'discordant atrioventricular connections with discordant ventriculo-arterial connections', and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)
876	Congenitally corrected TGA, VSD	Indicate if the patient has the diagnosis of "Congenitally corrected TGA, VSD". "Congenitally corrected TGA, VSD" is "Congenitally corrected transposition with a VSD". (Congenitally corrected transposition is synonymous with

876		the terms ‘corrected transposition’ and ‘discordant atrioventricular connections with discordant ventriculo-arterial connections’, and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)
878	Congenitally corrected TGA, VSD-LVOTO	Indicate if the patient has the diagnosis of “Congenitally corrected TGA, VSD-LVOTO”. “Congenitally corrected TGA, VSD-LVOTO” is “Congenitally corrected transposition with a VSD and left ventricular outflow tract obstruction”. (Congenitally corrected transposition is synonymous with the terms ‘corrected transposition’ and ‘discordant atrioventricular connections with discordant ventriculo-arterial connections’, and is defined as a spectrum of cardiac malformations where the atrial chambers are joined to morphologically inappropriate ventricles, and the ventricles then support morphologically inappropriate arterial trunks [1]. [1] Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Jacobs JP, Wernovsky G, Gaynor JW, and Anderson RH (editors). Cardiology in the Young, Volume 16 (Supplement 3): 72-84, September 2006.)
880	TGA, IVS	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with an intact ventricular septum. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
890	TGA, IVS-LVOTO	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with an intact ventricular septum and

890		associated left ventricular obstruction. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
900	TGA, VSD	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with one or more ventricular septal defects. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
910	TGA, VSD-LVOTO	A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with one or more ventricular septal defects and left ventricular outflow tract obstruction. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
930	DORV, VSD type	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In double outlet right ventricle, VSD type, there is an associated subaortic or doubly-committed VSD and no pulmonary outflow tract obstruction. Subaortic VSD's are located beneath the aortic valve. Doubly-committed VSD's lie beneath the leaflets of the aortic and pulmonary valves (juxtaarterial). In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
940	DORV, TOF type	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In double outlet right ventricle, TOF type, there is an associated subaortic or doubly-committed VSD and pulmonary outflow tract

940		obstruction. Subaortic VSD's are located beneath the aortic valve. Doubly-committed VSD's lie beneath the leaflets of the aortic and pulmonary valves (juxtaarterial). DORV can occur in association with pulmonary atresia, keeping in mind in coding that in the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles (in this situation DORV is coded as a primary diagnosis). Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate Single ventricle listing.
950	DORV, TGA type	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In double outlet right ventricle, TGA type, there is an associated subpulmonary VSD. Most frequently, there is no pulmonary outflow tract obstruction (Taussig-Bing heart). The aorta is usually to the right and slightly anterior to or side-by-side with the pulmonary artery. Associated aortic outflow tract stenosis (subaortic, aortic arch obstruction) is commonly associated with the Taussig-Bing heart and if present should be coded as a secondary diagnosis. Rarely, there is associated pulmonary outflow tract obstruction. In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
960	DORV, Remote VSD (uncommitted VSD)	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In double outlet right ventricle, Remote VSD type, there is a remote or noncommitted VSD. The VSD is far removed from both the aortic and pulmonary valves, usually within the inlet septum. Many of these VSD's are in hearts with DORV and common atrioventricular canal/septal defect. In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DORV is to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
2030	DORV + AVSD (AV Canal)	Indicate if the patient has the diagnosis of “DORV + AVSD (AV Canal)”. In the event of DORV occurring in association with AVSD (AV Canal), code “DORV + AVSD (AV Canal)”, and then use additional (secondary) diagnostic codes to describe the DORV and the AVSD (AV Canal) separately to provide further documentation about the DORV and AVSD

2030		(AV Canal) types. {"DORV" is "Double outlet right ventricle" and is defined as a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle.} In this case, the DORV exists in combination with an atrioventricular septal defect and common atrioventricular junction guarded by a common atrioventricular valve.
975	DORV, IVS	Double outlet right ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. In the rare case of double outlet right ventricle with IVS the ventricular septum is intact. In the nomenclature developed for DORV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connections with DORV are to be coded under congenitally corrected TGA. DORV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
980	DOLV	Double outlet left ventricle is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the left ventricle. In the nomenclature developed for DOLV, there must be usual atrial arrangements and concordant atrioventricular connections, and normal or near-normal sized ventricles. Discordant atrioventricular connection with DOLV is to be coded under congenitally corrected TGA. DOLV associated with univentricular atrioventricular connections, atrioventricular valve atresia, or atrial isomerism is to be coded under the appropriate single ventricle listing.
990	Coarctation of aorta	Indicate if the patient has the diagnosis of "Coarctation of aorta". A "Coarctation of the aorta" generally indicates a narrowing of the descending thoracic aorta just distal to the left subclavian artery. However, the term may also be accurately used to refer to a region of narrowing anywhere in the thoracic or abdominal aorta.
1000	Aortic arch hypoplasia	Hypoplasia of the aortic arch is hypoplasia of the proximal or distal transverse arch or the aortic isthmus. The isthmus (arch between the left subclavian and insertion of the patent ductus arteriosus / ligamentum arteriosum) is hypoplastic if its diameter is less than 40% of the diameter of the ascending aorta. The proximal transverse arch (arch between the innominate and left carotid arteries) and distal transverse arch (arch between the left carotid and left subclavian arteries) are hypoplastic if their diameters are less than 60% and 50%, respectively, of the diameter of the ascending aorta.
92	VSD + Aortic arch hypoplasia	A ventricular septal defect, any type, associated with hypoplasia of the aortic arch. (See diagnosis definition 1000 for a definition of hypoplasia of the aortic arch.)
94	VSD + Coarctation of aorta	Indicate if the patient has the diagnosis of "VSD + Coarctation of aorta". In the event of a VSD occurring in association with Coarctation of aorta, code "VSD + Coarctation of aorta", and then use additional (secondary)

94		diagnostic codes to describe the VSD and the Coarctation of aorta separately to provide further documentation about the individual VSD and Coarctation of aorta types. {A "VSD" is a "Ventricular Septal Defect" and is also known as an "Interventricular communication". A VSD is defined as "a hole between the ventricular chambers or their remnants". (The VSD is defined on the basis of its margins as seen from the aspect of the morphologically right ventricle. In the setting of double outlet right ventricle, the defect provides the outflow from the morphologically left ventricle. In univentricular atrioventricular connections with functionally single left ventricle with an outflow chamber, the communication is referred to by some as a bulboventricular foramen.)} {A “Coarctation of the aorta” generally indicates a narrowing of the descending thoracic aorta just distal to the left subclavian artery. However, the term may also be accurately used to refer to a region of narrowing anywhere in the thoracic or abdominal aorta.}
1010	Coronary artery anomaly, Anomalous aortic origin of coronary artery (AAOCA)	Anomalous aortic origins of the coronary arteries include a spectrum of anatomic variations of the normal coronary artery origins. Coronary artery anomalies of aortic origin to be coded under this diagnostic field include: anomalies of take-off (high take-off), origin (sinus), branching, and number. An anomalous course of the coronary artery vessels is also significant, particularly those coronary arteries that arise or course between the great vessels.
1020	Coronary artery anomaly, Anomalous pulmonary origin (includes ALCAPA)	In patients with anomalous pulmonary origin of the coronary artery, the coronary artery (most commonly the left coronary artery) arises from the pulmonary artery rather than from the aorta. Rarely, the right coronary artery, the circumflex, or both coronary arteries may arise from the pulmonary artery.
1030	Coronary artery anomaly, Fistula	The most common of coronary artery anomalies, a coronary arteriovenous fistula is a communication between a coronary artery and either a chamber of the heart (coronary-cameral fistula) or any segment of the systemic or pulmonary circulation (coronary arteriovenous fistula). They may be congenital or acquired (traumatic, infectious, iatrogenic) in origin, and are mostly commonly seen singly, but occasionally multiple fistulas are present. Nomenclature schemes have been developed that further categorize the fistulas by vessel of origin and chamber of termination, and one angiographic classification scheme by Sakakibara has surgical implications. Coronary artery fistulas can be associated with other congenital heart anomalies such as tetralogy of Fallot, atrial septal defect, ventricular septal defect, and pulmonary atresia with intact ventricular septum, among others. The major cardiac defect should be listed as the primary diagnosis and the coronary artery fistula should be as an additional secondary diagnoses.
1040	Coronary artery anomaly, Aneurysm	Coronary artery aneurysms are defined as dilations of a coronary vessel 1.5 times the adjacent normal coronaries. There are two forms, saccular and fusiform (most common), and both may be single or multiple. These aneurysms may be congenital or acquired (atherosclerotic, Kawasaki, systemic diseases other than Kawasaki, iatrogenic,

1040		infectious, or traumatic) in origin.
2420	Coronary artery anomaly, Ostial Atresia	
1050	Coronary artery anomaly, Other	Coronary artery anomalies which may fall within this category include coronary artery bridging and coronary artery stenosis, as well as secondary coronary artery variations seen in congenital heart defects such as tetralogy of Fallot, transposition of the great arteries, and truncus arteriosus (with the exception of variations that can be addressed by a more specific coronary artery anomaly code).
1070	Interrupted aortic arch	Indicate if the patient has the diagnosis of “Interrupted aortic arch”. Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.
2020	Interrupted aortic arch + VSD	Indicate if the patient has the diagnosis of “Interrupted aortic arch + VSD”. In the event of interrupted aortic arch occurring in association with VSD, code “Interrupted aortic arch + VSD”, and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and the VSD separately to provide further documentation about the individual interrupted aortic arch and VSD types. {Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.} {A "VSD" is a "Ventricular Septal Defect" and is also known as an "Interventricular communication". A VSD is defined as "a hole between the ventricular chambers or their remnants". (The VSD is defined on the basis of its margins as seen from the aspect of the morphologically right ventricle. In the setting of double outlet right ventricle, the defect provides the outflow from the morphologically left ventricle. In univentricular atrioventricular connections with functionally single left ventricle with an outflow chamber, the communication is referred to by some as a bulboventricular foramen.)}
2000	Interrupted aortic arch + AP window (aortopulmonary window)	Indicate if the patient has the diagnosis of “Interrupted aortic arch + AP window (aortopulmonary window)”. In the event of interrupted aortic arch occurring in association with AP window, code “Interrupted aortic arch + AP window (aortopulmonary window)”, and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and the AP window separately to provide further documentation about the individual interrupted aortic arch and AP window types. {Interrupted aortic arch is defined as

2000		the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.} {An "AP window (aortopulmonary window)" is defined as a defect with side-to-side continuity of the lumens of the aorta and pulmonary arterial tree, which is distinguished from common arterial trunk (truncus arteriosus) by the presence of two arterial valves or their atretic remnants. (In other words, an aortopulmonary window is a communication between the main pulmonary artery and ascending aorta in the presence of two separate semilunar [pulmonary and aortic] valves. The presence of two separate semilunar valves distinguishes AP window from truncus arteriosus. Type 1 proximal defect: AP window located just above the sinus of Valsalva, a few millimeters above the semilunar valves, with a superior rim but little inferior rim separating the AP window from the semilunar valves. Type 2 distal defect: AP window located in the uppermost portion of the ascending aorta, with a well-formed inferior rim but little superior rim. Type 3 total defect: AP window involving the majority of the ascending aorta, with little superior and inferior rims. The intermediate type of AP window is similar to the total defect but with adequate superior and inferior rims. In the event of AP window occurring in association with interrupted aortic arch, code "Interrupted aortic arch + AP window (aortopulmonary window)", and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and AP window separately to provide further documentation about the individual interrupted arch and AP window types.)}
1080	Patent ductus arteriosus	Indicate if the patient has the diagnosis of "Patent ductus arteriosus". The ductus arteriosus (arterial duct) is an essential feature of fetal circulation, connecting the main pulmonary trunk with the descending aorta, distal to the origin of the left subclavian artery. In most patients it is on the left side. If a right aortic arch is present, it may be on the right or the left; very rarely it is bilateral. When luminal patency of the duct persists post-natally, it is referred to as patent ductus arteriosus (patent arterial duct). The length and diameter may vary considerably from case to case. The media of the ductus consists mainly of smooth muscle that is arranged spirally, and the intima is much thicker than that of the aorta. (A patent ductus arteriosus is a vascular arterial connection between the thoracic aorta and the pulmonary artery. Most commonly a PDA has its origin from the descending thoracic aorta, just distal and opposite the origin of the left subclavian artery. The insertion of the ductus is most commonly into the very proximal left pulmonary artery at its junction with the main pulmonary artery. Origination and insertion sites can be variable, however.)

1090	Vascular ring	The term vascular ring refers to a group of congenital vascular anomalies that encircle and compress the esophagus and trachea. The compression may be from a complete anatomic ring (double aortic arch or right aortic arch with a left ligamentum) or from a compressive effect of an aberrant vessel (innominate artery compression syndrome).
1100	Pulmonary artery sling	In pulmonary artery sling, the left pulmonary artery originates from the right pulmonary artery and courses posteriorly between the trachea and esophagus in its route to the left lung hilum, causing a sling-like compression of the trachea.
1110	Aortic aneurysm (including pseudoaneurysm)	An aneurysm of the aorta is defined as a localized dilation or enlargement of the aorta at any site along its length (from aortic annulus to aortoiliac bifurcation). A true aortic aneurysm involves all layers of the aortic wall. A false aortic aneurysm (pseudoaneurysm) is defined as a dilated segment of the aorta not containing all layers of the aortic wall and may include postoperative or post-procedure false aneurysms at anastomotic sites, traumatic aortic injuries or transections, and infectious processes leading to a contained rupture.
1120	Aortic dissection	Aortic dissection is a separation of the layers of the aortic wall. Extension of the plane of the dissection may progress to free rupture into the pericardium, mediastinum, or pleural space if not contained by the outer layers of the media and adventitia. Dissections may be classified as acute or chronic (if they have been present for more than 14 days).
1130	Lung disease, Benign	Lung disease arising from any etiology (congenital or acquired) which does not result in death or lung or heart-lung transplant; examples might be non-life threatening asthma or emphysema, benign cysts.
1140	Lung disease, Malignant	Lung disease arising from any etiology (congenital or acquired, including pulmonary parenchymal disease, pulmonary vascular disease, congenital heart disease, neoplasm, etc.) which may result in death or lung or heart-lung transplant.
1160	Tracheal stenosis	Tracheal stenosis is a reduction in the anatomic luminal diameter of the trachea by more than 50% of the remaining trachea. This stenosis may be congenital or acquired (as in post-intubation or traumatic tracheal stenosis).
2430	Tracheomalacia	
1170	Airway disease, Other	Included in this diagnostic category would be airway pathology not included under the definition of tracheal stenosis such as tracheomalacia, bronchotracheomalacia, tracheal right upper lobe, bronchomalacia, subglottic stenosis, bronchial stenosis, etc.
1430	Pleural disease, Benign	Benign diseases of the mediastinal or visceral pleura.
1440	Pleural disease, Malignant	Malignant diseases of the mediastinal or visceral pleura.
1450	Pneumothorax	A collection of air or gas in the pleural space.
1460	Pleural effusion	Abnormal accumulation of fluid in the pleural space.

1470	Chylothorax	The presence of lymphatic fluid in the pleural space secondary to a leak from the thoracic duct or its branches. Chylothorax is a specific type of pleural effusion.
1480	Empyema	A collection of purulent material in the pleural space, usually secondary to an infection.
1490	Esophageal disease, Benign	Any benign disease of the esophagus.
1500	Esophageal disease, Malignant	Any malignant disease of the esophagus.
1505	Mediastinal disease	Any disease of the mediastinum awaiting final benign/malignant pathology determination.
1510	Mediastinal disease, Benign	Any benign disease of the mediastinum.
1520	Mediastinal disease, Malignant	Any malignant disease of the mediastinum.
1540	Diaphragm paralysis	Paralysis of diaphragm, unilateral or bilateral.
1550	Diaphragm disease, Other	Any disease of the diaphragm other than paralysis.
2160	Rib tumor, Benign	Non-cancerous tumor of rib(s) (e.g., fibrous dysplasia)
2170	Rib tumor, Malignant	Cancerous tumor of rib(s)- primary (e.g., osteosarcoma, chondrosarcoma)
2180	Rib tumor, Metastatic	Cancerous tumor metastasized to rib(s)from a different primary location
2190	Sternal tumor, Benign	Non-cancerous tumor of sternum (e.g., fibrous dysplasia)
2200	Sternal tumor, Malignant	Cancerous tumor of sternum - primary (e.g., osteosarcoma, chondrosarcoma)
2210	Sternal tumor, Metastatic	Cancerous tumor metastasized to sternum from a different primary location
2220	Pectus carinatum	Pectus carinatum represents a spectrum of protrusion abnormalities of the anterior chest wall. Severe deformity may result in dyspnea and decreased endurance. Some patients develop rigidity of the chest wall with decreased lung compliance, progressive emphysema, and increased frequency of respiratory tract infections.
2230	Pectus excavatum	Pectus excavatum is a congenital chest wall deformity in which several ribs and the sternum grow abnormally, producing a concave, or caved-in, appearance in the anterior chest wall. Pectus excavatum is the most common type of congenital chest wall abnormality. It occurs in an estimated 1 in 300-400 births, with male predominance (male-to-female ratio of 3:1). The condition is typically noticed at birth, and more than 90% of cases are diagnosed within the first year of life. Worsening of the chest's appearance and the onset of respiratory symptoms are usually reported during rapid bone growth in the early teenage years.
2240	Thoracic outlet syndrome	Thoracic outlet syndrome (TOS) is caused by compression at the superior thoracic outlet wherein excess pressure is placed on a neurovascular bundle passing between the anterior scalene and middle scalene muscles. It can affect the brachial plexus (nerves that pass into the arm from the neck), the subclavian artery, and - rarely - the vein, which does not normally pass through the scalene hiatus. TOS may occur due to a positional cause - for example, by abnormal compression from the clavicle (collarbone) and shoulder girdle on arm movement. There are also several static

2240		forms, caused by abnormalities, enlargement, or spasm of the various muscles surrounding the arteries, veins, and/or brachial plexus, a fixation of a first rib, or a cervical rib. The most common causes of thoracic outlet syndrome include physical trauma from a car accident, repetitive injuries from a job such as frequent non-ergonomic use of a keyboard, sports-related activities, anatomical defects such as having an extra rib, and pregnancy.
1180	Arrhythmia	Any cardiac rhythm other than normal sinus rhythm.
2440	Arrhythmia, Atrial, Atrial fibrillation	
2450	Arrhythmia, Atrial, Atrial flutter	
2460	Arrhythmia, Atrial, Other	
2050	Arrhythmia, Junctional	Indicate if the patient has the diagnosis of ‘‘Arrhythmia, Junctional’’. “Arrhythmias arising from the atrioventricular junction; may be bradycardia, tachycardia, premature beats, or escape rhythm [1]. [1]. Jacobs JP. (Editor). 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, Volume 18, Supplement S2, pages 1 –530, December 9, 2008, page 379.
2060	Arrhythmia, Ventricular	Indicate if the patient has the diagnosis of ‘‘Arrhythmia, Ventricular’’. “Arrhythmia, Ventricular’’ ROOT Definition = Abnormal rhythm originating from the ventricles [1]. [1]. Jacobs JP. (Editor). 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, Volume 18, Supplement S2, pages 1 –530, December 9, 2008, page 393.
1185	Arrhythmia, Heart block	Atrioventricular block may be congenital or acquired, and may be of varying degree (first, second, or third degree).
1190	Arrhythmia, Heart block, Acquired	Atrioventricular block, when acquired, may be post-surgical, or secondary to myocarditis or other etiologies; the block may be first, second or third degree.
1200	Arrhythmia, Heart block, Congenital	Atrioventricular block, when congenital, may be first, second or third degree block.
1220	Arrhythmia, Pacemaker, Indication for replacement	Indications for pacemaker replacement may include end of generator life, malfunction, or infection.
2530	Short QT syndrome	
2540	Long QT Syndrome (Ward Romano syndrome)	
2550	Wolff-Parkinson-White syndrome (WPW syndrome)	
1230	Atrial Isomerism, Left	In isomerism, both appendages are of like morphology or structure; in left atrial isomerism both the right atrium and left atrium appear to be a left atrium structurally.
1240	Atrial Isomerism, Right	In isomerism, both appendages are of like morphology or

1240		structure; in right atrial isomerism both the right atrium and left atrium appear to be a right atrium structurally.
2090	Dextrocardia	Indicate if the patient has the diagnosis of ‘‘Dextrocardia’’. “Dextrocardia’’ is most usually considered synonymous with a right-sided ventricular mass, whilst ‘‘dextroversion’’ is frequently defined as a configuration where the ventricular apex points to the right. In a patient with the usual atrial arrangement, or situs solitus, dextroversion, therefore, implies a turning to the right of the heart [1]. [1]. Jacobs JP, Anderson RH, Weinberg P, Walters III HL, Tchervenkov CI, Del Duca D, Franklin RCG, Aiello VD, Béland MJ, Colan SD, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Elliott MJ. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. In 2007 Supplement to Cardiology in the Young: Controversies and Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Anderson RH, Jacobs JP, and Wernovsky G, editors. Cardiology in the Young, Volume 17, Supplement 2, pages 1–28, doi: 10.1017/S1047951107001138, September 2007.
2100	Levocardia	Indicate if the patient has the diagnosis of ‘‘Levocardia’’. ‘‘Levocardia’’ usually considered synonymous with a left-sided ventricular mass, whilst ‘‘levoversion’’ is frequently defined as a configuration where the ventricular apex points to the left [1]. [1]. Jacobs JP, Anderson RH, Weinberg P, Walters III HL, Tchervenkov CI, Del Duca D, Franklin RCG, Aiello VD, Béland MJ, Colan SD, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Elliott MJ. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. In 2007 Supplement to Cardiology in the Young: Controversies and Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Anderson RH, Jacobs JP, and Wernovsky G, editors. Cardiology in the Young, Volume 17, Supplement 2, pages 1–28, doi: 10.1017/S1047951107001138, September 2007.
2110	Mesocardia	Indicate if the patient has the diagnosis of ‘‘Mesocardia’’. “Mesocardia’’ is most usually considered synonymous with the ventricular mass occupying the midline [1]. [1]. Jacobs JP, Anderson RH, Weinberg P, Walters III HL, Tchervenkov CI, Del Duca D, Franklin RCG, Aiello VD, Béland MJ, Colan SD, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Elliott MJ. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. In 2007 Supplement to Cardiology in the Young: Controversies and Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Anderson RH, Jacobs JP, and Wernovsky G, editors. Cardiology in the Young, Volume 17, Supplement 2, pages 1–28, doi: 10.1017/S1047951107001138, September 2007.
2120	Situs inversus	Indicate if the patient has the diagnosis of “Situs inversus” of the atrial chambers. The development of morphologically right-sided structures on one side of the body, and morphologically left-sided structures on the other side, is termed lateralization. Normal lateralization, the usual arrangement, is also known as ‘‘situs solitus’’. The

2120		mirror-imaged arrangement is also known as ‘‘situs inversus’’. The term ‘‘visceroatrial situs’’ is often used to refer to the situs of the viscera and atria when their situs is in agreement. The arrangement of the organs themselves, and the arrangement of the atrial chambers, is not always the same. Should such disharmony be encountered, the sidedness of the organs and atrial chambers must be separately specified [1]. [1]. Jacobs JP, Anderson RH, Weinberg P, Walters III HL, Tchervenkov CI, Del Duca D, Franklin RCG, Aiello VD, Béland MJ, Colan SD, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Elliott MJ. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. In 2007 Supplement to Cardiology in the Young: Controversies and Challenges Facing Paediatric Cardiovascular Practitioners and their Patients, Anderson RH, Jacobs JP, and Wernovsky G, editors. Cardiology in the Young, Volume 17, Supplement 2, pages 1–28, doi: 10.1017/S1047951107001138, September 2007.
1250	Aneurysm, Ventricular, Right (including pseudoaneurysm)	An aneurysm of the right ventricle is defined as a localized dilation or enlargement of the right ventricular wall.
1260	Aneurysm, Ventricular, Left (including pseudoaneurysm)	An aneurysm of the left ventricle is defined as a localized dilation or enlargement of the left ventricular wall.
1270	Aneurysm, Pulmonary artery	An aneurysm of the pulmonary artery is defined as a localized dilation or enlargement of the pulmonary artery trunk and its central branches (right and left pulmonary artery).
1280	Aneurysm, Other	A localized dilation or enlargement of a cardiac vessel or chamber not coded in specific fields available for aortic aneurysm, sinus of Valsalva aneurysm, coronary artery aneurysm, right ventricular aneurysm, left ventricular aneurysm, or pulmonary artery aneurysm.
1290	Hypoplastic RV	Small size of the right ventricle. This morphological abnormality usually is an integral part of other congenital cardiac anomalies and, therefore, frequently does not need to be coded separately. It should, however, be coded as secondary to an accompanying congenital cardiac anomaly if the right ventricular hypoplasia is not considered an integral and understood part of the primary congenital cardiac diagnosis. It would rarely be coded as a primary and/or isolated diagnosis.
1300	Hypoplastic LV	Small size of the left ventricle. This morphological abnormality usually is an integral part of other congenital cardiac anomalies and, therefore, frequently does not need to be coded separately. It should, however, be coded as secondary to an accompanying congenital cardiac anomaly if the left ventricular hypoplasia is not considered an integral and understood part of the primary congenital cardiac diagnosis. It would rarely be coded as a primary and/or isolated diagnosis.
2070	Postoperative bleeding	Indicate if the patient has the diagnosis of “Postoperative bleeding”.
1310	Mediastinitis	Inflammation/infection of the mediastinum, the cavity between the lungs which holds the heart, great vessels,

1310		trachea, esophagus, thymus, and connective tissues. In the United States mediastinitis occurs most commonly following chest surgery.
1320	Endocarditis	An infection of the endocardial surface of the heart, which may involve one or more heart valves (native or prosthetic) or septal defects or prosthetic patch material placed at previous surgery.
1325	Rheumatic heart disease	Heart disease, usually valvar (e.g., mitral or aortic), following an infection with group A streptococci
1330	Prosthetic valve failure	Indicate if the patient has the diagnosis of "Prosthetic valve failure". This diagnosis is the primary diagnosis to be entered for patients undergoing replacement of a previously placed valve (not conduit) prosthesis, whatever type (e.g., bioprosthetic, mechanical, etc.). Failure may be due to, among others, patient somatic growth, malfunction of the prosthesis, or calcification or overgrowth of the prosthesis (e.g., pannus formation). Secondary or fundamental diagnosis would relate to the underlying valve disease entity. As an example, a patient undergoing removal or replacement of a prosthetic pulmonary valve previously placed for pulmonary insufficiency after repair of tetralogy of Fallot would have as a primary diagnosis "Prosthetic valve failure", as a secondary diagnosis "Pulmonary insufficiency", and as a fundamental diagnosis "Tetralogy of Fallot".
1340	Myocardial infarction	A myocardial infarction is the development of myocardial necrosis caused by a critical imbalance between the oxygen supply and demand of the myocardium. While a myocardial infarction may be caused by any process that causes this imbalance it most commonly results from plaque rupture with thrombus formation in a coronary vessel, resulting in an acute reduction of blood supply to a portion of the myocardium. Myocardial infarction is a usual accompaniment of anomalous left coronary artery from the pulmonary artery (ALCAPA).
1350	Cardiac tumor	An abnormal growth of tissue in or on the heart, demonstrating partial or complete lack of structural organization, and no functional coordination with normal cardiac tissue. Commonly, a mass is recognized which is distinct from the normal structural components of the heart. A primary cardiac tumor is one that arises directly from tissues of the heart, (e.g., myxoma, fibroelastoma, rhabdomyoma, fibroma, lipoma, pheochromocytoma, teratoma, hemangioma, mesothesioloma, sarcoma). A secondary cardiac tumor is one that arises from tissues distant from the heart, with subsequent spread to the otherwise normal tissues of the heart, (e.g., renal cell tumor with caval extension from the kidney to the level of the heart or tumor with extension from other organs or areas of the body (hepatic, adrenal, uterine, infradiaphragmatic)). N.B., in the nomenclature system developed, cardiac thrombus and cardiac vegetation are categorized as primary cardiac tumors.
1360	Pulmonary AV fistula	An abnormal intrapulmonary connection (fistula) between an artery and vein that occurs in the blood vessels of the

1360		lungs. Pulmonary AV fistulas may be seen in association with congenital heart defects; the associated cardiac defect should be coded as well.
1370	Pulmonary embolism	A pulmonary embolus is a blockage of an artery in the lungs by fat, air, clumped tumor cells, or a blood clot.
1385	Pulmonary vascular obstructive disease	Pulmonary vascular obstructive disease (PVOD) other than those specifically defined elsewhere (Eisenmenger's pulmonary vascular obstructive disease, primary pulmonary hypertension, persistent fetal circulation). The spectrum includes PVOD arising from (1) pulmonary arterial hypertension or (2) pulmonary venous hypertension or (3) portal hypertension, or (4) collage vascular disease, or (5) drug or toxin induced, or (6) diseases of the respiratory system, or (7) chronic thromboembolic disease, among others.
1390	Pulmonary vascular obstructive disease (Eisenmenger's)	"Eisenmenger syndrome" could briefly be described as "Acquired severe pulmonary vascular disease associated with congenital heart disease (Eisenmenger)". Eisenmenger syndrome is an acquired condition. In Eisenmenger-type pulmonary vascular obstructive disease, long-term left-to-right shunting (e.g., through a ventricular or atrial septal defect, patent ductus arteriosus, aortopulmonary window) can lead to chronic pulmonary hypertension with resultant pathological changes in the pulmonary vessels. The vessels become thick-walled, stiff, noncompliant, and may be obstructed. In Eisenmenger syndrome, the long-term left-to-right shunting will reverse and become right to left. Please note that the specific heart defect should be coded as a secondary diagnosis.
1400	Primary pulmonary hypertension	Primary pulmonary hypertension is a rare disease characterized by elevated pulmonary artery hypertension with no apparent cause. Two forms are included in the nomenclature, a sporadic form and a familial form which can be linked to the BMPR-II gene.
1410	Persistent fetal circulation	Persistence of the blood flow pattern seen in fetal life, in which high pulmonary vascular resistance in the lungs results in decreased blood flow to the lungs. Normally, after birth pulmonary pressure falls with a fall in pulmonary vascular resistance and there is increased perfusion of the lungs. Persistent fetal circulation, also known as persistent pulmonary hypertension of the newborn, can be related to lung or diaphragm malformations or lung immaturity.
1420	Meconium aspiration	Aspiration of amniotic fluid stained with meconium before, during, or after birth can lead to pulmonary sequelae including (1) pneumothorax, (2) pneumomediastinum, (3) pneumopericardium, (4) lung infection, and (5) meconium aspiration syndrome (MAS) with persistent pulmonary hypertension.
2250	Kawasaki disease	Kawasaki disease, also known as Kawasaki syndrome, is an acute febrile illness of unknown etiology that primarily affects children younger than 5 years of age. it was first described in Japan in 1967, and the first cases outside of Japan were reported in Hawaii in 1976. It is characterized by fever, rash, swelling of the hands and feet, irritation and redness of the whites of the eyes, swollen lymph glands in

2250		the neck, and irritation and inflammation of the mouth, lips, and throat. Serious complications of Kawasaki disease include coronary artery dilatations and aneurysms, and Kawasaki disease is a leading cause of acquired heart disease in children in the United States. The standard treatment with intravenous immunoglobulin and aspirin substantially decreases the development of coronary artery abnormalities.
1560	Cardiac, Other	Any cardiac diagnosis not specifically delineated in other diagnostic codes.
1570	Thoracic and/or mediastinal, Other	Any thoracic and/or mediastinal disease not specifically delineated in other diagnostic codes.
1580	Peripheral vascular, Other	Any peripheral vascular disease (congenital or acquired) or injury (from trauma or iatrogenic); vessels involved may include, but are not limited to femoral artery, femoral vein, iliac artery, brachial artery, etc.
2260	Complication of cardiovascular catheterization procedure	Unspecified complication of cardiovascular catheterization procedure
2270	Complication of cardiovascular catheterization procedure, Device embolization	Migration or movement of device introduced during a cardiac catheterization procedure to an unintended location
2280	Complication of cardiovascular catheterization procedure, Device malfunction	Malfunction of a device introduced during a cardiac catheterization procedure
2290	Complication of cardiovascular catheterization procedure, Perforation	Perforation or puncture caused by a device introduced during a cardiac catheterization procedure
2300	Complication of interventional radiology procedure	Unspecified complication of interventional radiology procedure
2310	Complication of interventional radiology procedure, Device embolization	Migration or movement of device introduced during an interventional radiology procedure to an unintended location
2320	Complication of interventional radiology procedure, Device malfunction	Malfunction of a device introduced during an interventional radiology procedure
2330	Complication of interventional radiology procedure, Perforation	Perforation or puncture caused by a device introduced during an interventional radiology procedure
2340	Foreign body, Intracardiac foreign body	Presence of a foreign body within the heart
2350	Foreign body, Intravascular foreign body	Presence of a foreign body within an artery or vein
2360	Open sternum with closed skin	Sternotomy edges not re-approximated prior to closure of skin incision
2370	Open sternum with open skin (includes membrane placed to close skin)	Sternotomy and skin incision left open following surgery, covered with a membrane or dressing
2380	Retained sternal wire causing irritation	Surgically placed wire causing soft tissue irritation, pain or swelling (not infected)
2390	Syncope	A transient, self-limited loss of consciousness with an inability to maintain postural tone that is followed by spontaneous recovery. The term syncope excludes seizures,

	2390		coma, shock, or other states of altered consciousness.
	2400	Trauma, Blunt	Injury (ies) sustained from blunt force, caused by motor vehicle accidents, falls, blows or crush injuries
	2410	Trauma, Penetrating	Injury (ies) sustained as a result of sharp force, including cutting or piercing instruments or objects, bites, or firearm injuries from projectiles.
	2560	Cardio-respiratory failure not secondary to known structural heart disease	
	2570	Myocarditis	
	2580	Common AV valve insufficiency	
	2590	Protein-losing enteropathy	
	2600	Plastic bronchitis	
	7000	Normal heart	Normal heart.
	7777	Miscellaneous, Other	Any disease (congenital or acquired) not specifically delineated in other diagnostic codes.
	1590	Status post - Transplant, Heart	
	1610	Status post - Transplant, Heart and lung	
	1600	Status post - Transplant, Lung(s)	
Retired	2040	Arrhythmia, Atrial	Indicate if the patient has the diagnosis of ‘‘Arrhythmia, Atrial’’. ‘‘Arrhythmia, Atrial’’ ROOT Definition = Non-sinus atrial rhythm with or without atrioventricular conduction. [1]. [1]. Jacobs JP. (Editor). 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, Volume 18, Supplement S2, pages 1 –530, December 9, 2008, page 373.

Encounter medical diagnosis

Seq Num: 2560

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUDiagMedical]

Description: Code the medical condition or diagnosis that most directly indicates admission to CICU service. This diagnosis should be the patient's **final** diagnosis, not necessarily what was suspected at the time of CICU admission. For example, a patient admitted with respiratory failure who ultimately was found to have transplant rejection should be coded as "Cardiovasc - Heart transplant rejection." If this is the initial CICU encounter during a hospitalization, and the reason for CICU care is entirely explained by the encounter cardiothoracic diagnosis, you may select "None" for the encounter medical diagnosis. We expect all CICU readmissions to have an encounter medical diagnosis in addition to the encounter cardiothoracic diagnosis (except in the case where a readmission to the CICU is directly from the cardiac OR).

Additional clarifications below between asterisks.

Values

Code	*Text*	
0	None	
14	Cardiovasc - Arrhythmia - Atrial fibrillation	
16	Cardiovasc - Arrhythmia - Atrial flutter	
18	Cardiovasc - Arrhythmia - Atrioventricular block, complete	
22	Cardiovasc - Arrhythmia - Bradycardia	
24	Cardiovasc - Arrhythmia - SVT	SVT separate from atrial flutter or atrial fibrillation, which would include paroxysmal SVT, ectopic atrial tachycardia, and AV node re-entrant tachycardia.
26	Cardiovasc - Arrhythmia - Ventricular tachycardia	
10	Cardiovasc - Cardiac arrest	Patient admitted to the CICU either in active cardiac arrest receiving CPR or resuscitated from cardiac arrest and presenting for post-arrest care. If there is an active underlying condition prompting the cardiac arrest, code this underlying condition as a complication present on arrival.
194	Cardiovasc - Cardiac contusion	Confirmed or suspected cardiac contusion based on history of blunt trauma to chest wall and elevated cardiac enzymes (troponin/CK) or direct inspection of the myocardium by a cardiothoracic surgeon.
2	Cardiovasc - Heart failure, Acute decompensated	Systolic or diastolic cardiac dysfunction that requires at least one of the following therapies: (1) continuous infusion of a vasoactive agent or diuretic agent; (2) respiratory support (non-invasive or invasive positive pressure ventilation); or (3) mechanical circulatory support. If heart failure due to acute myocarditis, use acute myocarditis as medical diagnosis * Does not include intermittent infusion of any agent. *
200	Cardiovasc - Heart failure, Chronic	Chronic or acute on chronic heart failure which was

200	or acute on chronic	managed prior the admission with heart failure medications and/or mechanical circulatory support and requiring the initiation of at least one of the following therapies: 1) new or increased dose of diuretic therapy (IV or enteral), 2) continuous infusion of a new vasoactive agent or increased dose of an existing vasoactive agent, 3) increased respiratory support (HFNC, non-invasive or invasive mechanical ventilation), 4) new mechanical circulatory support.
4	Cardiovasc - Heart transplant rejection	Treatment with anti-rejection medical therapy, or biopsy or explant documented heart transplant rejection (before or after therapy). If patient has acute decompensated heart failure due to rejection, use this diagnosis (not acute decompensated heart failure). * Includes patients admitted to the CICU with other conditions (eg, respiratory failure) who are later determined to be in rejection. *
201	Cardiovasc - Inadequate pulmonary blood flow	Hypoxemia caused by inadequate pulmonary blood flow either confirmed by echo and/or angiogram or suspected clinically after ruling out all other causes. If inadequate pulmonary blood flow is due to pulmonary hypertension or pulmonary embolism, then those should be used as the medical diagnosis.
202	Cardiovasc - Myocarditis, acute	New diagnosis of cardiac dysfunction and/or rhythm disturbance suspected to be secondary to acute myocarditis requiring at least one of the following therapies: (1) continuous infusion of a vasoactive agent; (2) respiratory support (HFNC, CPAP/BiPAP, mechanical ventilation); (3) mechanical circulatory support. * Patients who don't meet this criteria but are diagnosed with myocarditis via MRI can be coded as Other. *
203	Cardiovasc - Myocarditis, acute, infective	Acute myocarditis, as defined above, proven to be infectious by myocardial biopsy, MRI, and/or positive serologies. * Patients who don't meet this criteria but are diagnosed with myocarditis via MRI can be coded as Other. *
6	Cardiovasc - Pacemaker malfunction	Permanent pacemaker malfunction requiring ICU level monitoring and/or therapy.
40	Cardiovasc - Pericardial effusion	Pericardial effusion requiring intensive care level monitoring, medical therapy, and/or drainage
204	Cardiovasc - Pulmonary embolism	Pulmonary embolism is defined as the embolization of a clot or other foreign material to the pulmonary vasculature documented by CT angiogram, nuclear medicine scan, MRI or angiography. This would include patients with superior cavopulmonary anastomosis or Fontan physiology.
30	Cardiovasc - Pulmonary hypertension	The primary reason the patient is admitted to the CICU is for management of pulmonary hypertension. This could include patients newly diagnosed with pulmonary hypertension during this CICU encounter, initiation of pulmonary antihypertensive therapy, or for management of

30		pulmonary hypertension in a patient who is currently being treated with medical (e.g. iNO, sildenafil, bosentan, etc.) therapy.
205	Cardiovasc - Syncope/near syncope	Sudden loss of consciousness without cardiac arrest. If the patient is admitted following a syncopal event and a more definitive diagnosis is identified during the hospitalization, the definitive diagnosis should be used. For example, a patient admitted after a syncopal event who is diagnosed with pulmonary hypertension should carry a medical diagnosis of pulmonary hypertension
206	Cardiovasc - VAD malfunction	Admission for VAD malfunction. Complications related to VAD with specific diagnosis (such as stroke) should be entered with specific diagnosis as the medical diagnosis.
182	GI - Bowel obstruction, unspecified	Gastric outlet, small bowel obstruction, or large bowel obstruction requiring CICU level monitoring/intervention and/or surgical intervention.
184	GI - GI tract hemorrhage, unspecified	Upper or lower GI tract hemorrhage requiring CICU level monitoring and/or transfusion.
207	GI - Hepatic injury	New onset hepatic dysfunction characterized by an ALT > 500.
189	GI - Necrotizing enterocolitis (Bell's criteria)	NEC is defined as Bell's criteria II or III: ileus, pneumatosis, portal vein gas, ascites, and /or pneumoperitoneum AND antibiotics/NPO for > 6 days.
208	GI - PLE	Protein Losing Enteropathy defined as (1) clinical manifestations (peripheral edema, abdominal distention or discomfort, diarrhea, ascites, pleural or pericardial effusion), and (2) serum albumin < 3.5 g/dL and/or serum protein < 6.3 g/dL, or (3) documentation of increased enteric protein loss by fecal alpha 1 anti-trypsin (>27 ml/24 hr without diarrhea and >56 ml/24 hr with diarrhea), spot fecal alpha 1 anti-trypsin concentration (>54 mg/dL) or nuclear scintigraphy.
146	Infectious - Bronchiolitis or other respiratory infection, viral due to RSV	Clinical RSV infection confirmed by PCR or immunoassay. If a patient has respiratory failure or insufficiency due to RSV, use this diagnosis not respiratory failure or respiratory insufficiency.
144	Infectious - Bronchiolitis or other respiratory infection, viral other	Non-RSV viral infection with or without a positive PCR for a known viral pathogen. If a patient has respiratory failure or insufficiency due to a non-RSV viral infection, use this diagnosis, not respiratory failure or respiratory insufficiency. * Please also use this diagnosis to code COVID19 (unless you are coding myocarditis). *
20	Infectious - Endocarditis	Endocarditis as defined by the Duke criteria. This may include infections that began at another institution or at home. These infections may not be adjudicated by the local infection control personnel or person responsible for adjudicating infections because they would not be attributed to the CICU.
162	Infectious - Gastroenteritis	Diarrhea and/or vomiting not due to NEC or other cause (e.g. intestinal obstruction). Does not require positive rotavirus screen.
148	Infectious - Meningitis, bacterial	Fever with lumbar puncture showing positive CSF culture.

154	Infectious - Pneumonia	Clinical diagnosis of pneumonia characterized by fever, increased WBC, and chest X-ray change. If a patient has respiratory failure or insufficiency due to pneumonia, use this diagnosis not respiratory failure or respiratory insufficiency.
140	Infectious - Sepsis	Temperature instability and abnormal WBC (leukopenia or leukocytosis) and hemodynamic instability requiring at least one of the following: (1) volume > 40 cc/kg; (2) new or increased inotropic support; or (3) new or increased mechanical ventilation support. * New or increased mechanical ventilation support refers ONLY to invasive ventilation. *
152	Infectious - Surgical site infection, deep	Deep surgical site infection, as defined by the CDC (requiring incision and drainage). This may include infections that began at another institution or at home. These infections may not be adjudicated by the local infection control personnel or person responsible for adjudicating infections because they would not be attributed to the CICU.
153	Infectious - Surgical site infection, superficial	Superficial surgical site infection, as defined by the CDC. This may include infections that began at another institution or at home. These infections may not be adjudicated by the local infection control personnel or person responsible for adjudicating infections because they would not be attributed to the CICU.
164	Infectious - Urinary tract infection	UTI, as defined by the CDC; select this only if it has been adjudicated by the local infection control personnel or the clinician responsible for adjudicating infections for the purpose of external reporting. This may include infections that began at another institution or at home. These infections may not be adjudicated by the local infection control personnel or person responsible for adjudicating infections because they would not be attributed to the CICU.
172	Neuro - Hypoxic-ischemic brain injury	Hypoxic-ischemic brain injury diagnosed by MRI/CT.
209	Neuro - Intracranial hemorrhage	New or previously unsuspected focus of discrete central nervous system injury consistent with hemorrhage. Patients with hemorrhagic stroke or hemorrhagic conversion of a thromboembolic stroke should be coded as stroke.
160	Neuro - Seizure	
192	Neuro - Skull fracture with intracranial hemorrhage	
170	Neuro - Stroke	Clinical evidence of stroke with thrombotic and/or embolic changes on MRI or CT.
176	Neuro - VP shunt dysfunction	
80	Resp - Chylothorax	
211	Resp - Hemothorax	
212	Resp - Plastic bronchitis	Requires documentation of airway casts at bronchoscopy (on this or a previous admission) or at autopsy.
90	Resp - Pleural effusion	

	110	Resp - Pneumothorax	
	84	Resp - Pulmonary hemorrhage (hemoptysis)	
	82	Resp - Respiratory failure, acute	Respiratory failure requiring institution of invasive mechanical ventilation at the time of or within the first hour of ICU admission. This includes patients intubated for acute respiratory failure at another hospital or in another ward/unit in the current hospital who were admitted to the CICU for management of respiratory failure. This excludes chronically ventilated patients and those who require ongoing respiratory support from a procedure suite (OR, cath lab, etc.).
	120	Resp - Respiratory insufficiency	Respiratory insufficiency requiring initiation of HFNC or any form of non-invasive positive pressure at the time of or within the first hour of CICU admission. This excludes patients who require ongoing respiratory support from a procedure suite (OR, cath lab, etc) or patients requiring respiratory support for procedural sedation.
	196	Other - Drug overdose	Inappropriate pharmacologic therapy (intentional or unintentional) requiring ICU monitoring or intervention.
	210	Other - ICU therapy at home	Patient is receiving ICU level therapy at home (mechanical ventilation, VAD, milrinone, treprostinil, etc) and requires admission for a non-ICU indication but home therapy can only be delivered in the ICU.
	178	Other - Intracardiac tumor	
	180	Other - Oncologic process	
	130	Other - Renal failure, acute	Oliguria with sustained urine output < 0.5 cc/kg/hr for 6 hours and/or a rise in creatinine > 1.5 times upper limits of normal for age.
	190	Other - Rhabdomyolisis	Patient admitted with rhabdomyolysis consisting of CPK elevation or myoglobinuria.
	198	Other - DNR / hospice care	Patients with DNR/DNI order at admission or created within the first hour after admission. This diagnosis is intended to capture patients admitted for comfort care only. * If the DNR/DNI order was put in place more than 1 hour after CICU admission, or if the existing order is cancelled at any time during the CICU admission, do not use this diagnosis. If any attempt was made to provide life sustaining/curative therapy to the patient, then DNR/DNI should NOT be coded as the medical diagnosis. *
	800	Other	
Retired	100	Infectious - Pneumonia	
Retired	150	Infectious - Wound infection	
Retired	50	Cardiovascular dysfunction	Low cardiac output state characterized by some of the following: tachycardia, oliguria, decreased skin perfusion, need for increased inotropic support (10% above baseline at admission), metabolic acidosis, widened Arterial - Venous oxygen saturation, need to open the chest, or need for mechanical support.
Retired	60	Other - Postoperative bleeding	

Retired	70	Cardiovasc - Arrhythmia	
Retired	188	Other - Necrotizing enterocolitis	Evidence of clinical deterioration (apnea, bradycardia, temperature instability, metabolic/lactic acidosis, oliguria, hypotension, biochemical derangement) with associated radiographic (pneumatosis, portal venous free air, fixed/prominent bowel loops, pneumoperitoneum) and gastrointestinal findings (bloody stools, absent bowel sounds, abdominal wall distention/edema/erythema, persistent gastric residuals, surgical intervention).

Medical diagnosis - specify

Seq Num: 2561

Required for case closure: No

Registry field: [CICUEncounter].[CICUDiagMedicalSpec]

Description: If the medical diagnosis is "Other", specify

Source of CICU admission

Seq Num: 2580

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUAdmitSource]

Description: Indicate the patient's location immediately prior to this CICU encounter

Values	*Code*	*Text*
	1	Home
	2	Current hospital
	3	Outside hospital

Specific source of CICU admission

Seq Num: 2600

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUAdmitSourceDesc]

Description: If the patient was hospitalized prior to this CICU encounter, indicate the specific source of transfer to the CICU.

Values	*Code*	*Text*	
	1	Clinic	Current hospital - clinic
	2	ED	Current hospital - ED
	3	Ward-cardiac	Current hospital - Cardiac ward
	4	Ward-non-cardiac	Current hospital - Non-cardiac ward
	5	OR-cardiac	Current hospital - Cardiac OR
	6	OR-general	Current hospital - General OR
	7	Cath lab	Current hospital - Cath lab
	8	PACU/Procedure suite	Current hospital - PACU/Procedure suite
	9	Delivery suite	Current hospital - Delivery suite
	10	NICU service	Current hospital - NICU service. Only select NICU service if the CICU attending was not the primary caregiver in that setting
	11	PICU service	Current hospital - PICU service. Only select PICU service if the CICU attending was not the primary caregiver in that setting
	12	Adult ICU service	Current hospital - Adult ICU service. Only select adult ICU service if the CICU attending was not the primary caregiver in that setting
	21	OSH - ED	Outside hospital - ED
	22	OSH - Floor	Outside hospital - Floor
	23	OSH - NICU / Delivery suite	Outside hospital - NICU or delivery suite
	24	OSH - PICU	Outside hospital - PICU
	25	OSH - CICU	Outside hospital - CICU
	26	OSH - Adult ICU	Outside hospital - Adult ICU

Weight (kg) at CICU admit

Seq Num: 2760

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUWtAdmit]

Description: Indicate the patient's weight in kg closest to the CICU admit date/time or the presumed dry weight as noted in the medical record.

Permanent feeding tube at CICU admit Seq Num: 2805

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUpermTube]

Description: Indicate Yes if patient has a permanent feeding tube (gastrostomy or jejunal gastrostomy tube) at time of CICU admission

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Existing trach at CICU admission Seq Num: 2810

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUTrach]

Description: Indicate Yes if the patient had an actively cannulated tracheostomy at the time of CICU admission.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Advance directive at CICU admission Seq Num: 2806

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUadvDirective]

Description: If patient age is >= 18 years, was there an advance directive in the medical record at the time of CICU admission.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Any non-cardiothoracic surgery during the encounter

Seq Num: 2920

Required for case closure: Yes

Registry field: [CICUEncounter].[NCSurgEnc]

Description: Indicate Yes if the patient had any non-cardiothoracic surgery after CICU admission.
*
If a corrective procedure was done during a bronchoscopy (e.g. balloon of subglottic stenosis, removal of granulation tissue, etc.) it may be captured as non-cardiothoracic surgery. Otherwise, isolated bronchoscopies and endoscopies should NOT be coded here.

If a bronchoscopy is performed as part of a cardiothoracic (STS) surgery, capture it in the 'Therapy - Bronchoscopy' (#6280) field if it occurred during the PC4 encounter.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Non-cardiothoracic surgery date

Seq Num: 2980

Required for case closure: Yes

Registry field: [NCSurg].[NCSurgDt]

Description: Date the non-cardiothoracic surgery was performed.

Non-cardiothoracic surgery type

Seq Num: 3000

Required for case closure: Yes

Registry field: [NCSurg].[NCSurg]

Description: Type of non-cardiothoracic surgery performed.

Values	*Code*	*Text*
	20	ENT - Cervical tracheoplasty
	40	ENT - Cleft lip and palate repair
	210	ENT - Incision and drainage of peritonsilar or retropharyngeal abscess
	30	ENT - Laryngoplasty
	10	ENT - Tracheostomy
	420	ENT - Tympanoplasty
	7000	ENT - Other
	170	General surg - Abdominal laparoscopic procedure
	360	General surg - Cholecystectomy
	260	General surg - Correction of malrotation
	250	General surg - Gastric restrictive procedure
	240	General surg - Gastrostomy or gastrojejunostomy tube placement, open
	230	General surg - Gastrostomy or gastrojejunostomy tube placement, percutaneous
	280	General surg - Hernia repair
	220	General surg - Nissen procedure with or without gastrostomy tube placement
	100	General surg - Pectus excavatum repair
	330	General surg - Proctectomy
	270	General surg - Reduction of volvulus, intussusception, or internal hernia
	200	General surg - Repair of diaphragmatic hernia
	180	General surg - Repair of hiatal hernia
	390	General surg - Repair of omphalocele or gastroschisis
	320	General surg - Repair of perforated

320	colon
310	General surg - Repair of perforated small intestine
190	General surg - Repair of tracheoesophageal fistula
300	General surg - Resection of colon
340	General surg - Resection of liver
290	General surg - Resection of small intestine
160	General surg - Splenectomy
7050	General surg - Other
410	Neurosurg - Craniectomy or craniotomy
50	Neurosurg - Creation or revision of ventriculo-peritoneal or –pleural shunt
60	Neurosurg - Repair of myelomeningocele
7100	Neurosurg - Other
90	Oncologic - Excision of abdominal tumor
70	Oncologic - Excision of benign mass
80	Oncologic - Excision of chest wall tumor
7150	Oncologic - Other
140	Ortho - Fasciotomy
120	Ortho - Open repair of fracture, joint dislocation
130	Ortho - Other open orthopedic procedure
110	Ortho - Spinal fusion, insertion of spinal fixation device, or removal of spinal hardware
7200	Ortho - Other
380	Transplant - Kidney
350	Transplant - Liver
7250	Transplant - Other
400	Urology - Bladder, uterine, or ovarian surgery
370	Urology - Resection of kidney
7300	Urology - Other
150	Vasc - Repair of systemic artery with or without graft
430	Vasc - Tunneled venous catheter placement
440	Vasc - Tunneled venous catheter

440	removal
7350	Vasc - Other
7777	Other non-cardiothoracic surgery

Non-cardiothoracic surgery - specify

Seq Num: 3001

Required for case closure: No

Registry field: [NCSurg].[NCSurgSpec]

Description: Specify the type of non-cardiothoracic surgery

Multiple venues

Seq Num: 2820

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUMultiVenues]

Description: During this encounter, was the patient cared for in more than one ICU venue (i.e., CICU, NICU, PICU, adult ICU) while under CICU care.

Values	*Code*	*Text*
	1	Yes
	0	No

Venue

Seq Num: 2860

Required for case closure: Yes

Registry field: [Venue].[Venue]

Description: Document each ICU venue in which the patient was located while under CICU care. For each venue, you will be asked the admission and discharge dates/times.

Values	*Code*	*Text*
	1	CICU
	2	NICU
	3	PICU
	4	Adult ICU

Venue start date/time

Seq Num: 2880

Required for case closure: Yes

Registry field: [Venue].[VenueStartDtTm]

Description: Indicate the date/time the patient was physically admitted to this venue

Venue end date/time

Seq Num: 2900

Required for case closure: Yes

Registry field: [Venue].[VenueEndDtTm]

Description: Indicate the date/time the patient was physically discharged from this venue

Weight (kg) at CICU discharge

Seq Num: 2780

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUWtDisch]

Description: Indicate the patient's weight in kg closest to the CICU discharge date/time.

CICU disposition

Seq Num: 2620

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUDispo]

Description: Indicate the disposition at CICU discharge date/time

*

Patients discharged to home on hospice care should be coded as 5 - Hospice.

Values	*Code*	*Text*
	1	Home
	2	Current hospital
	3	Outside hospital
	5	Hospice
	4	Deceased

Specific CICU disposition

Seq Num: 2640

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUDispoDesc]

Description: If the patient was transferred to another ward or service, indicate the specific location/service.

Values	*Code*	*Text*	
	1	Ward - Cardiac	
	8	Ward-Cardiac via procedure suite/OR	Patient was sent to OR/procedure suite from ICU with planned admission to cardiac ward thereafter
	2	Ward - Noncardiac	
	3	NICU service	
	4	PICU service	
	5	Adult ICU service	
	6	OSH - CICU	
	7	Rehab unit	

Withdrawal of life-sustaining therapy

Seq Num: 2740

Required for case closure: Yes

Registry field: [CICUEncounter].[CICULSTWithdraw]

Description: Indicate the disposition at CICU discharge date/time

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

CICU critical care end date/time

Seq Num: 2340

Retired in version 2.0

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUEndDtTm]

Description: Date and time the CICU attending physician deems the patient medically ready to leave CICU service.

Initial CICU encounter

Seq Num: 2500

Retired in version 2.0

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUInit]

Description: Select Yes if this is the patient's first CICU encounter during this hospital stay.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Change in cardiothoracic diagnosis from initial encounter

Seq Num: 2520

Retired in version 2.0

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUDiagChange]

Description: If this is not the initial CICU encounter during the hospital stay, indicate whether the primary cardiothoracic anatomy/physiology requiring care in the CICU differs from the initial CICU encounter during this hospitalization. For example, a patient's whose initial CICU encounter was for surgical repair of AVSD would have an initial encounter cardiothoracic diagnosis of "AVC (AVSD), Complete." After discharge from the CICU, the patient develops mitral stenosis requiring readmission to the CICU for respiratory insufficiency. The encounter cardiothoracic diagnosis for this readmission ("Mitral stenosis") is different from his initial encounter. Code this patient as Yes.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

DNR/DNI on File

Seq Num: 2660

Retired in version 2.0

Required for case closure: Yes

Registry field: [CICUEncounter].[DNROnFile]

Description: Indicate whether a DNR/DNI order was on file at any point during this CICU encounter. This includes orders written prior to or during the encounter.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Existing pacemaker/AICD at CICU admission

Seq Num: 2800

Retired in version 2.0

Required for case closure: Yes

Registry field: [CICUEncounter].[CICUDefib]

Description: Indicate Yes if the patient had a permanent implanted pacemaker/AICD at the time of CICU admission.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Respiratory Support

Invasive ventilation

Seq Num: 3040

Required for case closure: Yes

Registry field: [CICUEncounter].[MechVent]

Description: Indicate whether the patient was ever on invasive ventilation during this CICU encounter. If Yes, you will be asked to document each course. Courses that began prior to this encounter should be included if they continued after the CICU service became responsible for care.
*

This only includes patients receiving support through an endotracheal tube or trach. It does not include patients receiving support via a laryngeal mask airway (LMA).

CPAP support delivered via a conventional ventilator for a trached patient should be coded as mechanical ventilation with a mode type of 'Conventional' (#3480).

Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Invasive ventilation began at CICU start

Seq Num: 3210

Required for case closure: Yes

Registry field: [MechanicalVent].[MechVentCICUInit]

Description: Did this course of invasive ventilatory support begin at CICU admission? This includes ventilated patients directly admitted to the CICU from the OR/cath lab if they had NOT been on vent prior to their procedure. If the patient was extubated in the OR/cath lab prior to arrival or was intubated prior to the procedure, please check No.

*

There are two situations where the answer to this question is Yes, and the CICU admit date/time should be used as the intubation date/time:

1) The CICU admission begins immediately post-procedure (surgery or cath). Ventilator support was initiated during the procedure (i.e., the patient was not on support in the immediate pre-op period) and was admitted to the CICU still on vent.

OR

2) A patient is admitted to your hospital on vent support and goes directly to the CICU. This includes patients on home vent support and those transferred from an outside hospital if they were admitted directly to the CICU.

Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values

Code	*Text*
1	Yes
0	No

Ventilation start date/time

Seq Num: 3220

Required for case closure: Yes

Registry field: [MechanicalVent].[IntubDtTm]

Description: For each course of invasive ventilatory support, indicate the date/time it started. If the patient arrived on vent support from the OR or procedure suite and had NOT been on vent prior to their procedure, use the post-procedure CICU arrival time. If this ventilation course began prior to hospital admission, please use the hospital admit date/time. In all other cases, use the actual date/time this course began.

*

If the patient was on invasive ventilation during this hospitalization but prior to CICU admission, please list the actual date and time support began. The only exception to this is for patients who arrive on vent from the OR or procedure suite who were NOT on vent prior to their surgery/procedure. For these patients, use the post-procedure CICU arrival date/time.

If the course of vent began prior to this hospitalization, please enter the hospital admit date/time.

This logic should also be applied to patients who remain on vent at the end of the CICU encounter. If the course ends during this hospitalization, enter the actual end date/time. If this is unknown or the patient remained on support at hospital discharge, use the hospital discharge date/time.

Ventilated for procedure

Seq Num: 3223

Required for case closure: Yes

Registry field: [MechanicalVent].[IntubProc]

Description: Did this course of invasive ventilatory support begin during a procedure (surgery, cath, or radiology)?

If the patient was on vent support prior to the procedure and it continued until the procedure start time, code No.

*

Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Invasive ventilation at CICU discharge

Seq Num: 3240

Required for case closure: Yes

Registry field: [MechanicalVent].[MechVentCICUEnd]

Description: For each course of invasive ventilation, select Yes if the patient remained on invasive ventilator support at the end of the CICU encounter. **This includes patients who expired while intubated.** *

Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values

Code	*Text*
1	Yes
0	No

Vent end date known

Seq Num: 3260

Required for case closure: Yes

Registry field: [MechanicalVent].[ExtubDtKnown]

Description: If the patient remained on ventilator support at the end of the CICU encounter, indicate whether the vent end date is known. If this course of support continued through the end of this hospital admission, answer Yes and use the hospital discharge date/time for the vent end date/time.

Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values

Code	*Text*
1	Yes
0	No

Ventilation end date/time

Seq Num: 3280

Required for case closure: Yes

Registry field: [MechanicalVent].[ExtubDtTm]

Description: For each course of invasive ventilation, indicate the date/time it ended, whether by planned or unplanned extubation. Please note, a planned airway exchange does not signify the end of a course of support. If the patient had been on vent support prior to a procedure but was extubated in the OR/PACU, use the surgery end time. For patients with a tracheostomy, this is the date/time when they are not mechanically ventilated (by any mode, IMV, CPAP, etc.) for the following 24 hours. If the patient remained on a ventilator at hospital discharge, use the hospital discharge date/time. If the patient was on support at CICU discharge and the end date/time is unknown, this will default to noon on the hospital discharge date.

*

A vent course ends when either:

(1) the endotracheal tube is intentionally removed (or, for trached patients, the end of vent support by any mode) for the purpose of allowing the patient to breathe without ventilator support or (2) the endotracheal tube was inadvertently dislodged.

A tube that is removed and immediately replaced, for example to clear a mucus plug, is not an extubation and should not be coded here.

Was the extubation planned

Seq Num: 3300

Required for case closure: Yes

Registry field: [MechanicalVent].[ExtubPlanned]

Description: Did the physician write an extubation order prior to the end of this course of mechanical ventilation?

*

If a patient is extubated and immediately reintubated (for example, to remove a mucus plug), this is not an extubation; the planned/unplanned question is not applicable.

Patients taken off vent because the decision was made to withdraw life-sustaining therapy should be coded as a planned extubation.

If a patient expires while on vent, answer Yes to "Mechanical ventilation at CICU discharge" (# 3240) which will deactivate this planned/unplanned question.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Initial support following invasive vent

Seq Num: 3305

Required for case closure: Yes

Registry field: [MechanicalVent].[ExtubSupport]

Description: Indicate the type of respiratory support provided immediately following discontinuation of mechanical ventilation.

Values	*Code*	*Text*
	1	CPAP or BiPAP
	2	HFNC
	3	Nasal cannula
	4	Other
	5	None
	9	Unk

Initial airway

Seq Num: 3226

Required for case closure: Yes

Registry field: [MechanicalVent].[AirwayInit]

Description: Indicate the type of airway at the beginning of this course.

*

Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values	*Code*	*Text*
	1	Oral
	2	Nasal
	3	Trach
	9	Unk

Final airway

Seq Num: 3229

Required for case closure: Yes

Registry field: [MechanicalVent].[AirwayFinal]

Description: Indicate the type of airway at the end of this course.

*

Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Values	*Code*	*Text*
	1	Oral
	2	Nasal
	3	Trach
	9	Unk

Tracheostomy date/time

Seq Num: 3232

Required for case closure: Yes

Registry field: [MechanicalVent].[AirwayTrachDtTm]

Description: If the patient began this course with an oral or nasal airway but ended with a trach, enter the date/time the tracheostomy was performed
*
Please see the Tracheostomy/Ventilation Scenarios table in the Appendix at the end of this document for coding examples.

Multiple Modes

Seq Num: 3440

Required for case closure: No

Registry field: [MechanicalVent].[MechVentModes]

Description: During this course of mechanical ventilation, was more than one mode of support used? If No, indicate the one mode in the space provided. If Yes, indicate each mode used and its associated start and end date/time.

Values	Code	Text
	1	Yes
	0	No

Mode type

Seq Num: 3480

Required for case closure: No

Registry field: [VentModes].[VentModeType]

Description: Mode of ventilator support

Values	Code	Text	
	1	Conventional (including bi-vent)	
	3	High-frequency	
Retired	2	Bi-Vent	Bi-Vent/APRV is a specific mode on Conventional ventilation. A change between Bi-Vent/APRV and Conventional should be documented as a distinct mode.

Mode start date/time

Seq Num: 3500

Required for case closure: No

Registry field: [VentModes].[VentModeStartDtTm]

Description: Date/time this modality began

Mode end date/time Seq Num: 3520

Required for case closure: No

Registry field: [VentModes].[VentModeEndDtTm]

Description: Date/time this modality began

Positive airway pressure (PAP) Seq Num: 3060

Required for case closure: Yes

Registry field: [CICUEncounter].[PosAirPress]

Description: Was the patient ever on CPAP or BiPAP during this CICU encounter? Courses that began prior to this encounter should be included if they continued after the CICU service became responsible for care. If Yes, document each course that occurred during this encounter. A course is defined as consecutive days during which the patient is on support for any part of that day.
*
For patients who are on CPAP or BiPAP every night and only at night, record this as a single course using the first date on as the start date, and the final date on as the end date. If there is ever a gap of at least one night when the patient is not on that support, start a new course when the support resumes.

This field includes patients receiving respiratory support via a RAM cannula.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

PAP at CICU start Seq Num: 3620

Required for case closure: No

Registry field: [PAPSupport].[PAPCICUStart]

Description: Indicate Yes if the patient was on CPAP or BiPAP at the start of the CICU encounter. This would include patients arriving from the OR/procedural suite and extubated on arrival by anesthesia to CPAP or BiPAP.

Values	*Code*	*Text*
	1	Yes
	0	No

PAP start date Seq Num: 3640

Required for case closure: No

Registry field: [PAPSupport].[PAPStartDt]

Description: Date this course of CPAP or BiPAP began If the patient was on CPAP/BiPAP prior to this encounter, this will default to the CICU start date.

PAP at CICU end Seq Num: 3660

Required for case closure: No

Registry field: [PAPSupport].[PAPCICUEnd]

Description: Did this course of CPAP/BiPAP continue until the end of the CICU encounter? Indicate Yes if the patient was on CPAP or BiPAP for all or part of every calendar day from PAP start date through the end of the CICU encounter.

Values	*Code*	*Text*
	1	Yes
	0	No

PAP end date Seq Num: 3680

Required for case closure: No

Registry field: [PAPSupport].[PAPEndDt]

Description: Indicate the last day during this course that the patient was on CPAP or BiPAP for any part of the calendar day. If the patient was still on CPAP or BiPAP at the end of this CICU encounter, this will default to the CICU discharge date.

High-flow nasal cannula (HFNC) Seq Num: 3070

Required for case closure: Yes

Registry field: [CICUEncounter].[HighFlowNasCan]

Description: Was the patient ever on a high-flow nasal cannula (HFNC) during this CICU encounter? Courses that began prior to this encounter should be included if they continued after the CICU service became responsible for care. If Yes, document each course that occurred during this encounter. A course is defined as consecutive days during which the patient is on support for any part of that day.

*

If a high-flow system is being used, code HFNC support. This delivery system usually involves the use of heated and humidified air which distinguishes it from normal oxygen delivery. The flow rate is not relevant for this definition.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

HFNC at CICU start Seq Num: 3820

Required for case closure: No

Registry field: [HFNCSupport].[HFNCCICUStart]

Description: Indicate Yes if the patient was on HFNC at the start of the CICU encounter. This would include patients arriving from the OR/procedural suite and extubated on arrival by anesthesia to HFNC.

Values	*Code*	*Text*
	1	Yes
	0	No

HFNC start date Seq Num: 3840

Required for case closure: No

Registry field: [HFNCSupport].[HFNCStartDt]

Description: Date this course of HFNC support began. If the patient was on support prior to this encounter, this will default to the CICU start date.

HFNC at CICU end Seq Num: 3860

Required for case closure: No

Registry field: [HFNCSupport].[HFNCCICUEnd]

Description: Did this course of HFNC continue until the end of the CICU encounter? Indicate Yes if the patient was on HFNC for all or part of every calendar day from HFNC start date through the end of the CICU encounter.

Values	*Code*	*Text*
	1	Yes
	0	No

HFNC end date Seq Num: 3900

Required for case closure: No

Registry field: [HFNCSupport].[HFNCEndDt]

Description: Indicate the last day during this course that the patient was on HFNC for any part of the calendar day. If the patient was still on HFNC at the end of this CICU encounter, this will default to the CICU discharge date.

Mechanical ventilation at CICU start

Seq Num: 3206

Retired in version 1.0

Required for case closure: Yes

Registry field: [MechanicalVent].[MechVentCICUStart]

Description: Was the patient on mechanical ventilation at the start of the CICU encounter. This includes patients directly admitted to the ICU from the OR if ventilated. If extubated in the OR prior to arrival, please check No

Values	*Code*	*Text*
Retired	0	No
Retired	1	Yes

Intubation date known

Seq Num: 3207

Retired in version 1.0

Required for case closure: Yes

Registry field: [MechanicalVent].[IntubDtKnown]

Description: If the patient was already ventilated at the start of the CICU encounter, indicate whether the intubation date is known.

Values	*Code*	*Text*
Retired	0	No
Retired	1	Yes

Multiple Airways

Seq Num: 3320

Retired in version 2.0

Required for case closure: No

Registry field: [MechanicalVent].[MultiAirways]

Description: During this course of mechanical ventilation, was more than one airway used? If No, indicate the one airway type in the space provided. If Yes, indicate each type of airway used and its associated start and end date/time.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Airway type Seq Num: 3360

Retired in version 2.0

Required for case closure: No

Registry field: [AirwaySupport].[AirwayType]

Description: Type of airway used

Values	*Code*	*Text*
Retired	1	Oral
Retired	2	Nasal
Retired	3	Trach

Airway cuffed Seq Num: 3380

Retired in version 2.0

Required for case closure: No

Registry field: [AirwaySupport].[AirwayCuffed]

Description: Cuffed or uncuffed airway

Values	*Code*	*Text*
Retired	1	Cuffed
Retired	2	Uncuffed

Airway start date/time Seq Num: 3400

Retired in version 2.0

Required for case closure: No

Registry field: [AirwaySupport].[AirwayStartDtTm]

Description: Date/time this airway began

Airway end date/time Seq Num: 3420

Retired in version 2.0

Required for case closure: No

Registry field: [AirwaySupport].[AirwayEndDtTm]

Description: Date/time this airway ended

Vascular Access

IO access

Seq Num: 3090

Required for case closure: Yes

Registry field: [CICUEncounter].[IOAccess]

Description: Indicate whether intraosseous (IO) access ever used during this CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Venous lines

Seq Num: 3100

Required for case closure: Yes

Registry field: [CICUEncounter].[VenLines]

Description: Were any venous lines used during this CICU encounter (including those placed prior to encounter start)? If Yes, document the type, access site, and start/end dates of each line used during the encounter. Line exchanges over a wire are not considered new lines; lines removed and replaced with a fresh puncture are new lines.

*

This includes sheaths left in place post cath, hemodialysis lines, apharesis lines, and Broviacs placed in a central vein.

It also includes IJ, subclavian and upper extremity lines placed into the Fontan pathway.

For a port-a-cath that is accessed any time during the encounter, code as a tunneled CVL (# 4040) with a cut-down access type (#4020). Use the accessed and de-accessed dates to code the line start (#4121) and line end (#4161) dates.

This field does not include ECMO cannulae.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Venous access type

Seq Num: 4020

Required for case closure: Yes

Registry field: [VenousLine].[VenAccessType]

Description: Access method for this venous line

Values	*Code*	*Text*
	1	Cut-down
	2	Percutaneous

Venous line type

Seq Num: 4040

Required for case closure: Yes

Registry field: [VenousLine].[VenLineType]

Description: Type of venous line

*

Only code PICC here if the line was inserted into a peripheral vessel and threaded into a central vein. If, however, a PICC-type catheter is tunneled directly into a central vein, code it as a tunneled CVL.

We only capture central lines in PC4. Do not code midline CVC's.

Values	*Code*	*Text*	
	1	PICC	
	2	Central venous line (CVL) – percutaneous	
	3	Central venous line (CVL) – tunneled	
	4	Pulmonary artery catheter (PAC)	Pulmonary artery catheter (PAC). Not to include internal jugular lines in patients with cavopulmonary anastomosis.

Venous line site

Seq Num: 4060

Required for case closure: Yes

Registry field: [VenousLine].[VenLineSite]

Description: Anatomic site in which this venous line was placed

Values	*Code*	*Text*
	5	Femoral
	4	Internal jugular (IJ)
	9	Internal jugular (IJ) - Glenn/Fontan
	2	Lower extremity
	6	Subclavian
	10	Subclavian - Glenn/Fontan
	7	Transhepatic
	3	Umbilical
	1	Upper extremity
	11	Upper extremity - Glenn/Fontan
	8	Other

Venous line venue

Seq Num: 4080

Required for case closure: Yes

Registry field: [VenousLine].[VenLineVenue]

Description: Venue in which this venous line was placed

Values	*Code*	*Text*
	1	CICU
	2	Other ICU
	3	OR
	4	Interventional radiology (IR)
	5	Cath lab
	7	Ward
	8	Delivery suite
	6	Outside hospital
	12	Other

Venous line present at CICU start Seq Num: 4100

Required for case closure: Yes

Registry field: [VenousLine].[VenLineCICUStart]

Description: Was this line placed prior to the start of the CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No

Venous line start date Seq Num: 4121

Required for case closure: Yes

Registry field: [VenousLine].[VenLineStartDt]

Description: Date this line was placed. If the line was placed prior to this encounter, this will default to the CICU start date.

Venous line present at CICU end Seq Num: 4140

Required for case closure: Yes

Registry field: [VenousLine].[VenLineCICUEnd]

Description: Was this line still in place at the end of the CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No

Venous line end date Seq Num: 4161

Required for case closure: Yes

Registry field: [VenousLine].[VenLineEndDt]

Description: Date this line was removed. If the line was still in place at the end of this encounter, this will default to the CICU discharge date.

Venous line thrombus requiring treatment

Seq Num: 4180

Required for case closure: Yes

Registry field: [VenousLine].[VenLineThrombus]

Description: Was there ever a thrombus associated with this line that required either (a) the initiation of systemic anticoagulation or (b) an increase in dose or duration of systemic anticoagulation? This should not include tPA of a clotted line. Treatment may begin at any time during/after this CICU encounter.

*

Include patients treated with Lovenox therapy for a thrombus.

For patients already on systemic anticoagulation when a clot was discovered on their line: if the dose and/or duration of therapy changed, capture the thrombus and use the date of therapy change as the treatment start date.

If no changes were made to the patient's systemic anticoagulation regime, do not code the thrombus.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Thrombus treatment start date

Seq Num: 4201

Required for case closure: Yes

Registry field: [VenousLine].[VenLineThrombusDt]

Description: Date treatment initiated for this thrombus

Intracardiac lines

Seq Num: 3130

Required for case closure: Yes

Registry field: [CICUEncounter].[IntraCardLines]

Description: Were any intracardiac lines used during this CICU encounter (including those placed prior to encounter start)? If Yes, document the type, access site, and start/end dates of each intracardiac line used during this encounter.

*

This includes lines placed transthoracically into the Fontan pathway (use 'Right atrium' (#4340), and Broviac lines placed transthoracically directly into the heart.

Broviac lines placed in a central vein should be coded as CVL's (#3100), not intracardiac lines.

Lines placed in the ascending aorta should be coded as arterial lines (#3110) with 'central' site (#4640).

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Intracardiac line type

Seq Num: 4320

Required for case closure: Yes

Registry field: [IntracardLine].[IntraCardLineType]

Description: Type of intracardiac line

Values	*Code*	*Text*
	1	Monitoring / infusions
	2	Broviac

Intracardiac line site

Seq Num: 4340

Required for case closure: Yes

Registry field: [IntracardLine].[IntraCardLineSite]

Description: Anatomic site in which this intracardiac line was placed

*

A PA line migrated to RV should be coded as a PA line.

Values	*Code*	*Text*
	1	Right atrium (RA)
	2	Left atrium (LA) or common atrium
	3	Pulmonary artery
	4	Superior vena cava (SVC)

Intracardiac line venue Seq Num: 4360

Required for case closure: Yes

Registry field: [IntracardLine].[IntraCardLineVenue]

Description: Venue in which this intracardiac line was placed

Values	*Code*	*Text*
	1	CICU
	2	Other ICU
	3	OR
	4	Interventional radiology (IR)
	5	Cath lab
	7	Ward
	8	Delivery suite
	6	Outside hospital
	12	Other

Intracardiac line present at CICU start Seq Num: 4380

Required for case closure: Yes

Registry field: [IntracardLine].[IntraCardLinePresCICUStart]

Description: Was this line placed prior to the start of the CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No

Intracardiac line start date Seq Num: 4401

Required for case closure: Yes

Registry field: [IntracardLine].[IntracardLineStartDt]

Description: Date this line was placed. If the line was placed prior to this encounter, this will default to the CICU start date.

Intracardiac line present at CICU end Seq Num: 4420

Required for case closure: Yes

Registry field: [IntracardLine].[IntraCardLinePresCICUEnd]

Description: Was this line still in place at the end of the CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No

Intracardiac line end date

Seq Num: 4441

Required for case closure: Yes

Registry field: [IntracardLine].[IntracardLineEndDt]

Description: Date this line was removed. If the line was still in place at the end of this encounter, this will default to the CICU discharge date.

Intracardiac line thrombus requiring treatment

Seq Num: 4460

Required for case closure: Yes

Registry field: [IntracardLine].[IntraCardLineThrombus]

Description: Was there ever a thrombus associated with this line that required either (a) the initiation of systemic anticoagulation or (b) an increase in dose or duration of systemic anticoagulation? This should not include tPA of a clotted line. Treatment may begin at any time during/after this CICU encounter.

*

Include patients treated with Lovenox therapy for a thrombus.

For patients already on systemic anticoagulation when a clot was discovered on their line: if the dose and/or duration of therapy changed, capture the thrombus and use the date of therapy change as the treatment start date.

If no changes were made to the patient's systemic anticoagulation regime, do not code the thrombus.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Thrombus treatment start date

Seq Num: 4481

Required for case closure: Yes

Registry field: [IntracardLine].[IntracardLineThrombusDt]

Description: Date treatment initiated for this thrombus

Arterial lines

Seq Num: 3110

Required for case closure: Yes

Registry field: [CICUEncounter].[ArtLines]

Description: Were any arterial lines used during this CICU encounter (including those placed prior to encounter start)? If Yes, document the type, access site, and start/end dates of arterial each line used during this encounter. Line exchanges over a wire are not considered new lines; lines removed and replaced with a fresh puncture are new lines.
*
Lines placed in the ascending aorta should be coded as arterial lines with a 'central' site (# 4640).

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Arterial access type

Seq Num: 4620

Required for case closure: Yes

Registry field: [ArterialLine].[ArtAccessType]

Description: Access method for this arterial line

Values	Code	Text
	1	Cut-down
	2	Percutaneous

Arterial line site

Seq Num: 4640

Required for case closure: Yes

Registry field: [ArterialLine].[ArtLineSite]

Description: Anatomic site in which this arterial line was placed

Values	Code	Text
	1	Peripheral
	4	Central (axillary, femoral)
	5	Umbilical
	3	Internal mammary artery (IMA)
Retired	2	Central (axillary, femoral, umbilical artery)

Arterial line venue Seq Num: 4660

Required for case closure: Yes

Registry field: [ArterialLine].[ArtLineVenue]

Description: Venue in which this arterial line was placed

Values	*Code*	*Text*
	1	CICU
	2	Other ICU
	3	OR
	4	Interventional radiology (IR)
	5	Cath lab
	7	Ward
	8	Delivery suite
	6	Outside hospital
	12	Other

Arterial line present at CICU start Seq Num: 4680

Required for case closure: Yes

Registry field: [ArterialLine].[ArtLineCICUStart]

Description: Was this line placed prior to the start of the CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No

Arterial line start date Seq Num: 4701

Required for case closure: Yes

Registry field: [ArterialLine].[ArtLineStartDt]

Description: Date this line was placed. If the line was placed prior to this encounter, this will default to the CICU start date.

Arterial line present at CICU end Seq Num: 4720

Required for case closure: Yes

Registry field: [ArterialLine].[ArtLineCICUEnd]

Description: Was this line still in place at the end of the CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No

Arterial line end date

Seq Num: 4741

Required for case closure: Yes

Registry field: [ArterialLine].[ArtLineEndDt]

Description: Date this line was removed. If the line was still in place at the end of this encounter, this will default to the CICU discharge date.

Arterial line thrombus requiring treatment

Seq Num: 4760

Required for case closure: Yes

Registry field: [ArterialLine].[ArtLineThrombus]

Description: Was there ever a thrombus associated with this line that required either (a) the initiation of systemic anticoagulation or (b) an increase in dose or duration of systemic anticoagulation? This should not include tPA of a clotted line. Treatment may begin at any time during/after this CICU encounter.
*

Include patients treated with Lovenox therapy for a thrombus.

For patients already on systemic anticoagulation when a clot was discovered on their line: if the dose and/or duration of therapy changed, capture the thrombus and use the date of therapy change as the treatment start date.

If no changes were made to the patient's systemic anticoagulation regime, do not code the thrombus.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Thrombus treatment start date

Seq Num: 4781

Required for case closure: Yes

Registry field: [ArterialLine].[ArtLineThrombusDt]

Description: Date treatment initiated for this thrombus

Venous line start date/time

Seq Num: 4120

Retired in version 2.0

Required for case closure: Yes

Registry field: [VenousLine].[VenLineStartDtTm]

Description: Date/time this line was placed. If the line was placed prior to this encounter, this will default to the CICU start date/time.

Venous line end date/time Seq Num: 4160

Retired in version 2.0

Required for case closure: Yes

Registry field: [VenousLine].[VenLineEndDtTm]

Description: Date/time this line was removed. If the line was still in place at the end of this encounter, this will default to the CICU discharge date/time.

Thrombus treatment start date/time Seq Num: 4200

Retired in version 2.0

Required for case closure: Yes

Registry field: [VenousLine].[VenLineThromTrtDtTm]

Description: Date/time treatment initiated for this thrombus

Intracardiac line start date/time Seq Num: 4400

Retired in version 2.0

Required for case closure: Yes

Registry field: [IntracardLine].[IntraCardLineStartDtTm]

Description: Date/time this line was placed. If the line was placed prior to this encounter, this will default to the CICU start date/time.

Intracardiac line end date/time Seq Num: 4440

Retired in version 2.0

Required for case closure: Yes

Registry field: [IntracardLine].[IntraCardLineEndDtTm]

Description: Date/time this line was removed. If the line was still in place at the end of this encounter, this will default to the CICU discharge date/time.

Thrombus treatment start date/time Seq Num: 4480

Retired in version 2.0

Required for case closure: Yes

Registry field: [IntracardLine].[IntraCardLineThromTrtStartDtTm

Description: Date/time treatment initiated for this thrombus

Arterial line start date/time Seq Num: 4700

Retired in version 2.0

Required for case closure: Yes

Registry field: [ArterialLine].[ArtLineStartDtTm]

Description: Date/time this line was placed. If the line was placed prior to this encounter, this will default to the CICU start date/time.

Arterial line end date/time Seq Num: 4740

Retired in version 2.0

Required for case closure: Yes

Registry field: [ArterialLine].[ArtLineEndDtTm]

Description: Date/time this line was removed. If the line was still in place at the end of this encounter, this will default to the CICU discharge date/time.

Thrombus treatment start date/time Seq Num: 4780

Retired in version 2.0

Required for case closure: Yes

Registry field: [ArterialLine].[ArtLineThromTrtDtTm]

Description: Date/time treatment initiated for this thrombus

Other Devices

Foley Catheter

Seq Num: 3140

Required for case closure: Yes

Registry field: [CICUEncounter].[Foley]

Description: Did the patient have a Foley catheter at any time during the CICU encounter (including those placed prior to encounter start)? If Yes, document each catheter used during the encounter.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Foley at start of CICU encounter

Seq Num: 4920

Required for case closure: Yes

Registry field: [FoleyCath].[FoleyCICUStart]

Description: Was a Foley present at the start of the CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No

Date Foley placed

Seq Num: 4940

Required for case closure: Yes

Registry field: [FoleyCath].[FoleyStartDt]

Description: Date this Foley was placed. If the Foley was placed prior to this encounter, this will default to the CICU start date.

Foley at end of CICU encounter

Seq Num: 4960

Required for case closure: Yes

Registry field: [FoleyCath].[FoleyCICUEnd]

Description: Was this Foley still in place at the end of the CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No

Date Foley removed

Seq Num: 4980

Required for case closure: Yes

Registry field: [FoleyCath].[FoleyEndDt]

Description: Date this Foley was removed. If the Foley was still in place at the end of this encounter, this will default to the CICU discharge date.

Other Therapies

Vasoactive infusions

Seq Num: 5100

Required for case closure: Yes

Registry field: [Therapy].[VasoInfusion]

Description: Was the patient on any vasoactive infusions during this CICU encounter? If Yes, record the start and end date of each course of vasoactive infusions.

A course begins when a patient is on an infusion of any agent.

A course of infusions ends when the patient is off all agents. If any agent is restarted that day or the following calendar day, the course is not considered over.

Do not include infusions of any agents during a procedure that are not continued into the post-procedure CICU stay. For patients on one or more of these agents, but not for the purposes of vasoactive support, code Yes except as noted below.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Vasoactive agents

Seq Num: 5240

Required for case closure: Yes

Registry field: [VasoactiveInfusion].[VasoAgent]

Description: What agent(s) did the patient receive during this CICU encounter

*Isoproterenol (Isuprel) to treat rhythm is NOT considered a vasoactive agent.

* Additional clarifications noted between asterisks.*

Values	Code	Text	
	12	Calcium infusion	Excludes calcium used exclusively to treat hypocalcemia
	2	Dobutamine	
	1	Dopamine	
	4	Epinephrine	
	11	Esmolol	Excludes esmolol used exclusively as an anti-arrhythmic agent, *and esmolol used to bring down a patient's heartrate for imaging *
	14	Fenoldopam	
	16	Isoproterenol	
	17	Levosimendan	
	3	Milrinone	
	15	Nesiritide	
	13	Nicardipine	
	10	Nitroglycerine	
	8	Nitroprusside	
	5	Norepinephrine	
	7	Phentolamine	
	9	Phenylephrine	
	6	Vasopressin	
	88	Other agent	

Other vasoactive agent - specify

Seq Num: 5241

Required for case closure: No

Registry field: [VasoactiveInfusion].[VasoAgentSpec]

Description: Specify the other vasoactive agent used

Was this course active at CICU start? Seq Num: 5254

Required for case closure: Yes

Registry field: [VasoCourse].[VasoEncStart]

Description: Did this course begin prior to or immediately upon CICU admission?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Date vasoactive course began Seq Num: 5255

Required for case closure: Yes

Registry field: [VasoCourse].[VasoStartDt]

Description: Record the date this course of support began in the CICU. If the patient was on any vasoactive infusions at CICU admission, use the CICU admit date.

*

A course of infusions is consecutive days on which the patient is on an infusion of any qualifying agent. When the patient is off all agents, the course ends. The agents themselves may change during the course – we are simply looking for consecutive days on which the patient is on any support.

If all of the patient's infusions are stopped, and then any infusion is restarted that day or the following calendar day, it is a continuation of the same course.

Did this course continue through CICU discharge? Seq Num: 5259

Required for case closure: Yes

Registry field: [VasoCourse].[VasoEncEnd]

Description: Was the patient discharged from the CICU before this course was complete?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Date vasoactive course ended Seq Num: 5260

Required for case closure: Yes

Registry field: [VasoCourse].[VasoEndDt]

Description: Date this course of vasoactive infusions ended. If this course was still active at CICU discharge, this will default to the CICU discharge date.

Sedation/analgesia/neuromuscular block

Seq Num: 5300

Required for case closure: Yes

Registry field: [Therapy].[OTSedation]

Description: Was the patient ever on a sedation, analgesic, or neuromuscular blockade infusions during this CICU encounter? If Yes, list all agents used.

*

If a patient starts on a sedative infusion in preparation for a procedure, and the infusion is discontinued when the procedure is complete, code as No. If the infusion does continue after the procedure, code as Yes.

Do not include a benzodiazepine infusion to treat seizures.

If a dexmedetomidine (Precedex) infusion is used only to treat an arrhythmia, code as No. If used for both an arrhythmia and sedation, code as Yes.

Do not code a PCA pump that is only administering bolus doses. However, if there is a baseline rate infusing, then code as Yes.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Sedation/analgesia/neuromuscular block agent

Seq Num: 5340

Required for case closure: Yes

Registry field: [Sedation].[SedationType]

Description: Sedation, analgesic, or neuromuscular block agent used

*

Do not include a benzodiazepine infusion to treat seizures.

If a dexmedetomidine (Precedex) infusion is used only to treat an arrhythmia, code as No. If used for both an arrhythmia and sedation, code as Yes.

Values

Code	*Text*
1	Benzodiazepines
2	Narcotics
3	Dexmedetomidine
4	Ketamine
5	Barbiturate
6	Propofol
7	Neuromuscular block
8	Epidural anesthesia

Peritoneal drain Seq Num: 5870

Required for case closure: Yes

Registry field: [Therapy].[PDrain]

Description: Was a peritoneal drain present at any time during the CICU encounter? Any device used for peritoneal drainage qualifies.

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Drain present on admit Seq Num: 5872

Required for case closure: Yes

Registry field: [Therapy].[PDrainEncStart]

Description: Was a drain present at CICU admission?

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Drain start date Seq Num: 5874

Required for case closure: Yes

Registry field: [Therapy].[PDrainStartDt]

Description: Earliest date a peritoneal drain was placed. If it was placed prior to this encounter, this will default to the CICU start date

Venue drain placed Seq Num: 5876

Required for case closure: Yes

Registry field: [Therapy].[PDrainVenue]

Description: Indicate the venue in which the earliest drain was placed

Values	Code	Text
	1	CICU
	2	OR
	8	Other
	9	Unk

Drain present on discharge Seq Num: 5878

Required for case closure: Yes

Registry field: [Therapy].[PDrainEncEnd]

Description: Was a drain present at CICU discharge?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Final drain removal date Seq Num: 5880

Required for case closure: Yes

Registry field: [Therapy].[PDrainEndDt]

Description: Date the final peritoneal drain was removed. If it was present at CICU discharge, this will default to the discharge date.

RRT Seq Num: 5900

Required for case closure: Yes

Registry field: [Therapy].[CRRT]

Description: Was the patient ever on renal replacement therapy (RRT) during this CICU encounter. This includes patients who received RRT on ECMO but not those who only received ultrafiltration on ECMO. If a patient had peritoneal drainage only (i.e., no dialysate was used), code this as No.
*

Code intermittent hemodialysis as Yes to RRT and Hemodialysis (#5980).

If peritoneal dialysis catheters are in place for passive drainage only, then do not include them as RRT. Code this patient as having a peritoneal drain (sequence #5870). If, however, dialysate is administered, you would code both a peritoneal drain (sequence #5870) and RRT-peritoneal dialysis (sequence #5900 and #5920).

If at any point the patient has clinical renal failure and RRT continues, you would also answer Yes to 'CRRT for ARF' (sequence #5905) – even if the catheters were initially placed prophylactically.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

CRRT for ARF Seq Num: 5905

Required for case closure: Yes

Registry field: [Therapy].[CRRTarf]

Description: During this CICU encounter, was the patient ever on continuous RRT for treatment of acute renal failure (ARF)?

*

This field is intended to capture all therapeutic (as opposed to prophylactic) RRT. Patients on RRT for chronic or acute renal failure should be coded as Yes to 'CRRT for ARF.'

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

CRRT for ARF date/time Seq Num: 5910

Required for case closure: Yes

Registry field: [Therapy].[CRRTarfDtTm]

Description: Enter the date/time during this CICU encounter when CRRT treatment for acute renal failure began. If treatment for ARF was initiated prior to CICU admission, use the CICU admit date/time.

Peritoneal dialysis Seq Num: 5920

Required for case closure: Yes

Registry field: [Therapy].[CRRTPD]

Description: If the patient required RRT, indicate if he/she was ever on peritoneal dialysis during this CICU encounter. If Yes, also answer Yes to the Peritoneal Drain question above.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Peritoneal dialysis start date Seq Num: 5922

Required for case closure: Yes

Registry field: [Therapy].[CRRTPDstartDt]

Description: Date peritoneal dialysis began in the CICU. If it began prior to CICU admission, use the CICU start date

Peritoneal dialysis end date Seq Num: 5924

Required for case closure: Yes

Registry field: [Therapy].[CRRTPDendDt]

Description: Final date peritoneal dialysis ended in the CICU. If it continued after CICU discharge, use the CICU discharge date

Hemodialysis Seq Num: 5980

Required for case closure: Yes

Registry field: [Therapy].[CRRTHemodial]

Description: If the patient required RRT, indicate if he/she was ever on hemodialysis during this CICU encounter

*

Code intermittent hemodialysis as Yes to Hemodialysis and RRT (#5900).

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Hemodialysis start date Seq Num: 5982

Required for case closure: Yes

Registry field: [Therapy].[CRRTHemodialStartDt]

Description: Date hemodialysis began in the CICU. If it began prior to CICU admission, use the CICU start date

Hemodialysis end date Seq Num: 5984

Required for case closure: Yes

Registry field: [Therapy].[CRRTHemodialEndDt]

Description: Final date hemodialysis ended in the CICU. If it continued after CICU discharge, use the CICU discharge date

CVVH Seq Num: 5940

Required for case closure: Yes

Registry field: [Therapy].[CRRTCVVH]

Description: If the patient required RRT, indicate if he/she was ever on continuous veno-venous hemofiltration (CVVH) during this CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

CVVH start date Seq Num: 5942

Required for case closure: Yes

Registry field: [Therapy].[CRRTCVVHstartDt]

Description: Date CVVH began in the CICU. If it began prior to CICU admission, use the CICU start date

CVVH end date Seq Num: 5944

Required for case closure: Yes

Registry field: [Therapy].[CRRTCVVHendDt]

Description: Final date CVVH ended in the CICU. If it continued after CICU discharge, use the CICU discharge date

Nitric oxide Seq Num: 5600

Required for case closure: Yes

Registry field: [Therapy].[NitricOxide]

Description: Was inhaled nitric oxide ever used during this CICU encounter. If the iNO was used to treat pulmonary hypertension, answer Yes both to this question and the pulmonary hypertension treatment question.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Bronchoscopy Seq Num: 6280

Required for case closure: Yes

Registry field: [Therapy].[OTBronch]

Description: Did the patient undergo bronchoscopy during the CICU encounter? This includes procedures done in the OR as well as the bedside.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Active cooling Seq Num: 5081

Retired in version 1.0

Required for case closure: Yes

Registry field: [Therapy].[OTActiveCool]

Description: Was a cooling blanket used at any time during this CICU encounter

Values	*Code*	*Text*
Retired	0	No
Retired	1	Yes

Enteral feeding volume at CICU end (cc/kg/day) Seq Num: 5839

Retired in version 1.0

Required for case closure: Yes

Registry field: [Therapy].[EntFeedCICUEndVol]

Description: Total volume of enteral feeding, in cc/kg, over the final full 24 hour period of CICU care.

Date first vasoactive infusion began Seq Num: 5120

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[VasoAgentStartDt]

Description: Date the first infusion of any vasoactive agent began during this encounter

Initial vasoactive course active at CICU discharge Seq Num: 5140

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[VasoInfusionCICUEnd]

Description: Was the patient discharged from the CICU before the first course of vasoactive infusions was complete? The course is not considered complete until he/she is off all vasoactive infusions for 24 hours.

Values	Code	Text
Retired	1	Yes
Retired	0	No

Date first course of vasoactive infusions ended Seq Num: 5160

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[VasoAgentEndDt]

Description: First date when a patient treated with vasoactive infusions is no longer on any continuous vasoactive infusion. The course is not complete if any vasoactive infusion is reinitiated within 24 hours. If the patient's initial course is not complete at CICU discharge, this will default to the CICU discharge date.

Vasoactive infusions at CICU discharge Seq Num: 5180

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[VasoInfusionFinalCICUEnd]

Description: Was the patient still on any vasoactive infusions at CICU discharge?

Values	Code	Text
Retired	1	Yes
Retired	0	No

Date final course of vasoactive infusions ended Seq Num: 5200

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[VasoAgentFinalDt]

Description: Final date any vasoactive infusion was used during this encounter. If there was a single course of vasoactive infusions, this will be the same as the date the first course ended. If the patient remained on vasoactive infusions at CICU discharge, this will default to the CICU discharge date.

Apharesis/plasmapharesis during CICU encounter

Seq Num: 5400

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[Apharesis]

Description: Select Yes if apharesis/plasmapharesis were ever used during the CICU encounter. This is defined as the removal, treatment, and return of blood plasma from the circulation, often employed in patients peri-OHT and myocarditis

Values	Code	Text
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Enteral feeding

Seq Num: 5700

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[EntFeed]

Description: Was patient ever on any volume of enteral feedings during this CICU stay, including trophic feedings.

Values	Code	Text
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Date enteral feeding initiated

Seq Num: 5720

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[EntFeedInitDt]

Description: Date enteral feeding was initiated in the CICU

Feeds reinitiated following cardiothoracic surgery

Seq Num: 5740

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[EntFeedReInit]

Description: After enteral feeds were initiated in the CICU, were they subsequently held then reinitiated following cardiothoracic surgery. Select No if the patient was only on enteral feeds postop or feeds were never reinitiated following surgery. Select N/A if the patient did not have cardiothoracic surgery during this CICU encounter.

Values	Code	Text	
Retired	1	Yes	Feeds were held then reinitiated following cardiothoracic surgery
Retired	0	No	Patient had cardiothoracic surgery, but feeds were not held/reinitiated
Retired	8	N/A - No card surg	Patient did not have cardiothoracic surgery during this encounter
Retired	9	Unk	

Date enteral feeding reinitiated

Seq Num: 5760

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[EntFeedReInitDt]

Description: Date enteral feeding was reinitiated following cardiac surgery.

PO ad lib at CICU end

Seq Num: 5780

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[EntFeedAdLib]

Description: For surgical patients age <=30 days at CICU discharge, indicate whether the patient had an order for PO ad lib feeds and was receiving no tube feedings at CICU discharge.

Values	Code	Text
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Caloric density (kCal/oz) at CICU end

Seq Num: 5800

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[EntFeedDens]

Description: For surgical patients age <=30 days at CICU discharge, indicate their caloric density (kCal/oz) at CICU discharge. If the patient was NPO, enter 0.

Final feeds reported in cc/day or kCal/kg/day

Seq Num: 5820

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[EntFeedUnits]

Description: For surgical patients age <=30 days at CICU discharge, select whether their final feeds are reported in cc/day or kCal/kg/day.

Values	*Code*	*Text*
Retired	1	cc/day
Retired	2	kCal/kg/day

Enteral feeding volume at CICU end (cc/day)

Seq Num: 5840

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[EntFeedCCday]

Description: For surgical patients age <=30 days at CICU discharge, document the total volume of enteral feeding, in cc/kg, over the final full 24 hour period of CICU care.

Enteral feeding at CICU end (kCal/kg/day)

Seq Num: 5860

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[EntFeedKcalDay]

Description: For surgical patients age <=30 days at CICU discharge, document the total volume of enteral feeding, in kCal/kg/day, over the final full 24 hour period of CICU care.

CRRT CVAH

Seq Num: 5960

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[CRRTCVAH]

Description: If the patient required CRRT, indicate if he/she was ever on continuous veno-arterial hemofiltration (CVAH) during this CICU encounter

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Other Therapies - Monitoring

EtCO2 Seq Num: 6040

Required for case closure: Yes

Registry field: [Therapy].[MonitorEtCO2]

Description: Was end tidal CO2 (EtCO2) monitoring used at any time during this CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

SVO2- Continuous Seq Num: 6060

Required for case closure: Yes

Registry field: [Therapy].[MonitorSVO2Cont]

Description: Was continuous mixed venous (SvO2) monitoring used at any time during this CICU encounter, excluding such monitoring only done during ECMO.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

NIRS Seq Num: 6200

Required for case closure: Yes

Registry field: [Therapy].[MonitorNIRS]

Description: Was NIRS used at any time during this CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Cerebral Seq Num: 6220

Required for case closure: Yes

Registry field: [Therapy].[NIRSCerebral]

Description: If NIRS was used, was it cerebral

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Somatic Seq Num: 6240

Required for case closure: Yes

Registry field: [Therapy].[NIRSSomatic]

Description: If NIRS was used, was it somatic

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

NIRS - Other Seq Num: 6261

Required for case closure: Yes

Registry field: [Therapy].[NIRSother]

Description: NIRS in any other location

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

CVP Seq Num: 6020

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[MonitorCVP]

Description: Was central venous pressure (CVP) monitoring used at any time during this CICU encounter

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

EEG - Continuous Seq Num: 6080

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[MonitorEEGCont]

Description: Was continuous EEG monitoring used at any time during this CICU encounter

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

BIS Seq Num: 6100

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[MonitorBIS]

Description: Was Bispectral Index (BIS) monitoring used at any time during this CICU encounter

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

LA pressure Seq Num: 6120

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[MonitorLApressure]

Description: Was left atrial or common atrial pressure monitoring used at any time during this CICU encounter

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

PA catheter

Seq Num: 6140

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[MonitorPAcath]

Description: Was pulmonary artery catheter monitoring used at any time during this CICU encounter

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Limb

Seq Num: 6260

Retired in version 2.0

Required for case closure: Yes

Registry field: [Therapy].[NIRSLimb]

Description: If NIRS was used, was it limb

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Medical Events and Complications-Cardiovascular

Cardiac arrest

Seq Num: 6440

Required for case closure: Yes

Registry field: [Complications].[CompCardArrest]

Description: During this CICU encounter, did the patient have a cardiac arrest as defined by AHA GWTG-R, excluding acute respiratory compromise that does not progress to cardiopulmonary arrest. An event is defined as (1) cardiopulmonary arrest requiring chest compressions and/or defibrillation, or (2) acute respiratory compromise requiring emergency assisted ventilation leading to cardiopulmonary arrest requiring chest compressions and/or defibrillation. All events must also elicit a resuscitation response by facility personnel and have a resuscitation record completed. This includes events that occur during a CICU encounter even if they happened outside of the ICU (e.g., imaging suites, procedure suites, etc.) However, cardiac arrest events that begin during a cardiothoracic surgical procedure in any location should be excluded. If Yes, document each arrest that occurred during the CICU encounter.
*

If less than 20 minutes elapsed between the end of one arrest (i.e., return of spontaneous circulation or ECMO initiation) and the beginning of the next arrest, then these are considered part of the same event. Code the arrest date/time (#6450) as the start of the first arrest, and code CPR end date/time (#6480) as the end of the arrest where 20 minutes or more of ROSC was achieved (the second event in this scenario).

If a patient had ventricular tachycardia that ultimately resulted in cardiac arrest, and received ICU-level therapy for the arrhythmia prior to the arrest, then code both events. If there was no ICU-level therapy for the arrhythmia, just code the arrest. In either instance, you would capture VT as the rhythm at CPR onset (#6462).

If patient was admitted to the CICU from home in cardiac arrest upon arrival and into the CICU encounter, code as Yes because it was present upon arrival and treated in the CICU.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Arrest date/time

Seq Num: 6450

Required for case closure: Yes

Registry field: [CompCardArrest].[CardArrestDtTm]

Description: For this arrest, document the date/time it began

ECPR for arrest

Seq Num: 6460

Required for case closure: Yes

Registry field: [CompCardArrest].[CardArrestECPR]

Description: For this arrest, indicate whether ECPR (cannulation during compressions) used during this arrest

*

If a patient had return of spontaneous circulation following CPR, but the decision was made to cannulate onto ECMO, and the patient had a ROSC of less than 20 minutes before the cannulation began, then this is considered ECPR for both the arrest complication and the 'Active CPR at cannulation' (#6730) question in the mechanical circulatory support section.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Rhythm at CPR onset

Seq Num: 6462

Required for case closure: Yes

Registry field: [CompCardArrest].[CardArrestRhythm]

Description: Record the rhythm at the onset of cardiopulmonary resuscitation. Only code JET, SVT, or complete heart block if the patient had a pulse and began receiving CPR for hypotension.

Values	*Code*	*Text*
	1	Complete heart block (CHB)
	2	JET
	3	PEA
	4	Sinus bradycardia / junctional rhythm
	5	SVT
	6	VF
	7	VT
	99	Unk

CPR onset location Seq Num: 6464

Required for case closure: Yes

Registry field: [CompCardArrest].[CardArrestVenue]

Description: Indicate the venue at the onset of cardiopulmonary resuscitation

Values	*Code*	*Text*
	1	CICU
	2	Cath lab
	3	Non-cardiac OR
	4	Procedure suite
	5	Imaging suite
	8	Other
	9	Unk

CPR end time known Seq Num: 6470

Required for case closure: Yes

Registry field: [CompCardArrest].[CardArrestEndKnown]

Description: For this arrest, is the CPR end time known?

Values	*Code*	*Text*
	1	Yes
	0	No

CPR end date/time Seq Num: 6480

Required for case closure: Yes

Registry field: [CompCardArrest].[CardArrestEndDtTm]

Description: If it is known for this arrest, indicate the date/time CPR ended. This is the time when CPR was discontinued for >20 minutes with the return of spontaneous circulation, ECMO initiation, or death.

Cooled to <34 Seq Num: 6490

Required for case closure: Yes

Registry field: [CompCardArrest].[CardArrestHypo]

Description: For this arrest, did the patient have a documented core temperature<34 with stated intent to provide therapeutic

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Cooled to normothermia

Seq Num: 6500

Required for case closure: Yes

Registry field: [CompCardArrest].[CardArrestNormo]

Description: If the patient was not cooled to<34 for this arrest, is there documentation that the patient's temperature was actively controlled with the intent to maintain normothermia.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Pericardial effusion

Seq Num: 6540

Required for case closure: Yes

Registry field: [Complications].[CompPeriEffus]

Description: Did the patient have pericardial effusion during the CICU encounter. Pericardial effusion is defined as abnormal accumulation of fluid in the pericardial space, requiring drainage, by any technique.

*

A pericardial effusion diagnosed and treated while in another unit, and prior to CICU admit, should not be coded as a complication here. That could, however, be captured as a medical diagnosis where appropriate.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Pericardial effusion date/time

Seq Num: 6560

Required for case closure: Yes

Registry field: [Complications].[CompPeriEffusDtTm]

Description: If the patient had pericardial effusion, document the date/time it was first treated during this CICU encounter.

Arrhythmia requiring therapy

Seq Num: 6580

Required for case closure: Yes

Registry field: [Complications].[CompArrhythTherapy]

Description: Did the patient have an arrhythmia requiring ICU-level therapy during the CICU encounter?

An arrhythmia is defined as atrial tachycardia (automatic or re-entrant), ventricular tachycardia (automatic or re-entrant), junctional tachycardia (automatic or re-entrant), complete heart block, second degree heart block or sinus/junctional bradycardia which requires at least one of the following ICU-level therapies: continuous IV medication (excluding electrolyte repletion with the exception of magnesium for torsades), bolus dosing (excluding bolus digoxin), pacing, defibrillation, cardioversion, or cooling.

Premature ventricular beats of any type and PVCs treated with electrolyte replacement should not be included.

This includes therapies while on ECLS/VAD, and arrhythmias clearly documented in the OR for which therapy was initiated in the OR and was ongoing at the time of CICU admission.
*
An arrhythmia that recurs within 24 hours of stopping ICU level intervention and results in re-institution of any ICU level intervention should be coded as a single arrythmia event.

Code NO if the patient:

Changed from oral to IV anti-arrhythmia meds because they were NPO

Is admitted to the CICU with a permanent pacemaker to treat complete heart block, the pacemaker is functioning appropriately, and no arrhythmia is seen in the CICU

Is on a backup pacing mode, or paced to improve cardiac output with underlying sinus rhythm at an appropriate rate

Code YES if the patient:

Had an arrhythmia that began outside of the ICU AND ICU level therapy is ongoing at the time of admission, even if the arrythmia is never documented in the ICU

Already has a defibrillator prior to CICU admission, and the defibrillator delivers an appropriate shock. (The presence of a defibrillator alone does not require coding an arrhythmia complication.)

Has a pacemaker that malfunctions and the patient is in CHB

Is paced for symptomatic bradycardia

These distinctions may be difficult to determine from the medical record, so please discuss with your clinical champion any paced patients without clear documentation of an arrhythmia.

Values

Code	*Text*
1	Yes

0	No
9	Unk

Arrhythmia treatment at CICU start

Seq Num: 6611

Required for case closure: Yes

Registry field: [CompArrhythmia].[ArrhythCICUstart]

Description: Was the patient receiving ICU-level treatment for this arrhythmia at the start of this CICU encounter?

Values

Code	*Text*
1	Yes
0	No
9	Unk

Arrhythmia date/time

Seq Num: 6600

Required for case closure: Yes

Registry field: [CompArrhythmia].[ArrhythStartDtTm]

Description: Document the date and time, to the nearest hour, ICU-level treatment began for the arrhythmia. For patients with multiple cardioversions or defibrillations for the same arrhythmia, the start should be the date/time of the first event. If the patient was receiving ICU-level treatment at the start of the encounter, this will default to the CICU admit date/time.

Arrhythmia type

Seq Num: 6630

Required for case closure: Yes

Registry field: [CompArrhythmia].[ArrhythFoci]

Description: For each arrhythmia, select the type

*

Atrial fibrillation and atrial flutter (treated with ICU-level therapy) should be coded as Atrial Tachycardia/SVT.

Values

Code	*Text*	
1	Atrial tachycardia / SVT	Automatic or re-entry tachy arrhythmia that originates from the atrium. This includes SVT.
2	Ventricular tachycardia	Automatic or re-entry tachy arrhythmia that originates from the ventricle
3	Junctional tachycardia	Automatic or re-entry tachy arrhythmia that originates from the junction
4	Complete heart block	
5	Second degree heart block	
6	Sinus or junctional bradycardia	

Arrhythmia therapy - Drug

Seq Num: 6640

Required for case closure: Yes

Registry field: [CompArrhythmia].[ArrhythTherapyDrug]

Description: Was this arrhythmia treated with ICU-level drug therapy? This includes continuous IV medication (excluding electrolyte repletion with the exception of magnesium for torsades) or bolus dosing (excluding bolus digoxin).

*

Only continuous IV medications and bolus dosing are included (exclusions noted above).

Do not code an arrhythmia event if:

- the only intervention is adenosine and it is used SOLELY as a diagnostic agent

- the patient was switched to IV bolus medication solely due to being made NPO

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Arrhythmia therapy - Electrical Cardioversion/Defibrillation

Seq Num: 6650

Required for case closure: Yes

Registry field: [CompArrhythmia].[ArrhythTherapyCvrsn]

Description: Was this arrhythmia treated with electrical cardioversion (including rapid atrial pacing) or defibrillation?

*

This field includes:

- A shock delivered by an AICD if the shock is delivered in the ICU and is found to be an appropriate shock

-Use of the rapid atrial pacing mode via a temporary pacemaker

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Arrhythmia therapy - Permanent Pacemaker/AICD Seq Num: 6660

Required for case closure: Yes

Registry field: [CompArrhythmia].[ArrhythTherapyPermPace]

Description: Was a permanent pacemaker or AICD used to treat this arrhythmia?

*

An appropriate shock delivered by an AICD is considered ICU level therapy

If the patient is stable without therapy in the ICU and then has a permanent pacemaker placed during the ICU encounter, an arrythmia event should be entered with a start and end date on the date of the pacemaker placement.

If a patient goes to the OR from a non-ICU location and has a pacemaker placed and then is admitted to the ICU, an arrythmia event would only be captured if the pacemaker fails and an ICU level therapy is instituted during the encounter.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Arrhythmia therapy - Temporary Pacemaker Seq Num: 6670

Required for case closure: Yes

Registry field: [CompArrhythmia].[ArrhythTherapyTempPace]

Description: Was a temporary pacemaker used to treat this arrhythmia?

*

Includes transvenous pacing.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Arrhythmia therapy - Cooled <35 Seq Num: 6680

Required for case closure: Yes

Registry field: [CompArrhythmia].[ArrhythTherapyHypo]

Description: For this arrhythmia, did the patient have a documented core temperature<35 with stated intent to provide therapeutic

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Arrhythmia treatment at CICU discharge

Seq Num: 6610

Required for case closure: Yes

Registry field: [CompArrhythmia].[ArrhythCICUEnd]

Description: Was the patient still receiving ICU-level treatment for this arrhythmia at CICU discharge?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Arrhythmia end date

Seq Num: 6621

Required for case closure: Yes

Registry field: [CompArrhythmia].[ArrhythEndDt]

Description: Document the date ICU-level treatment ended for the arrhythmia. For patients with multiple cardioversions or defibrillations for the same arrhythmia the end date should be the date of the last event. If a PPM or AICD was placed for this arrhythmia, use the PPM/AICD placement date as the end date. If the patient was still receiving ICU-level treatment for this arrhythmia at CICU discharge, this will default to the CICU discharge date.

*

The end date is when the ICU-level therapy used to treat the arrhythmia is discontinued and the arrhythmia does not recur. If the patient receives a permanent pacemaker or defibrillator during the encounter and the arrhythmia is not seen again in the CICU (or recurs but is not treated with ICU-level therapy), record the date of pacemaker/AICD placement as the end date.

For patients who are treated with temporary pacing, the end date/time should be when the pacemaker is no longer actively pacing the patient (i.e., attached as a back up/rescue only) or the patient is disconnected from the pacemaker, whichever happens first.

If a patient is transitioned to an oral medication and the arrhythmia doesn't recur, then the end date should be recorded as the final date any ICU-level therapy (e.g., intravenous medication) was stopped.

If the patient is still receiving ICU-level treatment for arrhythmia at CICU discharge (#6610= Yes), set this field to the date of CICU physical end date/time (#2360).

Mechanical circulatory support

Seq Num: 6700

Required for case closure: Yes

Registry field: [Complications].[MechCircSupp]

Description: Did the patient ever require ECMO or VAD support during this CICU encounter? Support that began prior to this encounter should be included if it continued after the CICU service became responsible for care. If Yes, document each course that occurred during this encounter.
*

In a patient supported with a VAD who has an oxygenator added to the circuit (i.e., the assist device is still in place and patient is not fully converted to an ECMO circuit), the VAD event should continue from placement through removal of that device and an ECMO course should also be documented for the duration that the oxygenator is in place.

If a patient's entire course of mechanical support takes place outside of the CICU (example: patient is cannulated in the cath lab, goes to the OR for an intervention, and then comes off support prior to coming to the CICU), a course of mechanical support should not be recorded.

Any decannulation/recannulation where mechanical support is discontinued and then needs to be reinitiated should be documented as separate courses of mechanical support. This is different than ELSO documentation, where recannulation within 12 hours is considered a single course.

These events should all be documented as single courses of mechanical support:
- a cannula change
- a transition between VV and VA ECMO
- a transition from one type of VAD to another
- a patient is bridged with the cannulae in place as a trial off of support but support has to be reinitiated ("failed clamp trial")

Values

Code	*Text*
1	Yes
0	No
9	Unk

Circulatory support type

Seq Num: 6710

Required for case closure: Yes

Registry field: [CompMechSupport].[MechCircSuppType]

Description: Type of mechanical circulatory support for this course

Values

Code	*Text*
1	VAD
2	ECMO

Reason for ECMO

Seq Num: 6720

Required for case closure: Yes

Registry field: [CompMechSupport].[MechCircSuppReason]

Description: If the patient was on ECMO, select the reason it was initiated

Values	Code	Text
	1	LCOS/Cardiac failure/Ventricular dysfunction
	3	Hypoxemia
	4	Hypercarbic respiratory failure
	5	Shunt occlusion
	6	Arrhythmia
	7	Bleeding
	8	Multisystem organ failure
Retired	2	Ventricular dysfunction

Active CPR at cannulation

Seq Num: 6730

Required for case closure: Yes

Registry field: [CompMechSupport].[MechCircSuppCPR]

Description: If the patient was on ECMO, indicate whether the patient was receiving active CPR at the time of cannulation

*

If a patient had return of spontaneous circulation following CPR, but the decision was made to cannulate onto ECMO, and the patient had a ROSC of less than 20 minutes before the cannulation began, then this is considered ECPR for both 'Active CPR at cannulation' (#6730) and the arrest complication (#6460).

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Active CPR within 2 hours of cannulation

Seq Num: 6740

Required for case closure: Yes

Registry field: [CompMechSupport].[MechCircSuppPrevCPR]

Description: If the patient was not receiving CPR at cannulation, indicate whether active CPR was received within 2 hours prior to cannulation

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Initial ECMO cannula site Seq Num: 6742

Required for case closure: Yes

Registry field: [CompMechSupport].[MechCircSuppInitCan]

Description: Indicate the initial cannula site(s)

Values	*Code*	*Text*
	1	Peripheral
	2	Transthoracic
	3	Both
	9	Unk

Final ECMO cannula site Seq Num: 6744

Required for case closure: Yes

Registry field: [CompMechSupport].[MechCircSuppFinalCan]

Description: Indicate the final cannula site(s)

Values	*Code*	*Text*
	1	Peripheral
	2	Transthoracic
	3	Both
	9	Unk

Support present at start of CICU encounter Seq Num: 6750

Required for case closure: Yes

Registry field: [CompMechSupport].[MechCircSuppCICUStart]

Description: Was this course of support initiated prior to the start of the CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No

Date/time of circulatory support initiation

Seq Num: 6760

Required for case closure: Yes

Registry field: [CompMechSupport].[MechCircSuppInitDtTm]

Description: Date/time this course mechanical circulatory support began. If the patient was on support prior to the CICU encounter, this will default to the CICU start date/time. If support was initiated in the OR, use the date/time of postop ICU arrival as the start date/time.

*

The mechanical support start time for patients placed on support in the CICU is the time at which full flows are reached. Consult clinical champion on a case by case basis if full flow time is unclear.

If mechanical support is initiated outside of the CICU prior to the start of a CICU encounter, the start time will be the start of the CICU encounter.

If mechanical support is initiated outside of the CICU but after the start of the CICU encounter (example: a patient is admitted to the CICU and then goes to the cath lab and has ECMO initiated in the cath lab), the mechanical support start time will be the time at which full flows are achieved.

For bedside procedures, use the time full flow is established.

Support present at end of CICU encounter

Seq Num: 6770

Required for case closure: Yes

Registry field: [CompMechSupport].[MechCircSuppCICUEnd]

Description: Was the patient still on this course of support at the end of the CICU encounter? This includes patients who died on support and those who died immediately after support was withdrawn.

Values	*Code*	*Text*
	1	Yes
	0	No

Date/time of circulatory support discontinuation

Seq Num: 6780

Required for case closure: Yes

Registry field: [CompMechSupport].[MechCircSuppDiscDtTm]

Description: Date/time this course mechanical circulatory support ended. If the patient was still on support at the end of the CICU encounter, this will default to the CICU discharge date/time.

*

For a patient who is taken to the OR for discontinuation of mechanical support, use the time the patient leaves the unit as the support end time. If your site's preference is to use the actual time off mechanical support in the OR to be consistent with ELSO data entry, that is acceptable also. Each site should be consistent in their methodology.

If mechanical support is discontinued due to discontinuation of life sustaining measures and the patient does not immediately expire when mechanical support is discontinued, record the actual end time of mechanical support. If the patient expires immediately, record that support was present at time of discharge and use time of death as the end time for mechanical support.

Low cardiac output syndrome (LCOS)

Seq Num: 6851

Required for case closure: Yes

Registry field: [Complications].[CompLCOS2]

Description: Did the patient have any episodes of low cardiac output syndrome (LCOS) during the CICU encounter? LCOS is defined by at least one of the following: 1. VIS >15 at any time 2. Tripling of the VIS during any 48-hour period, including escalation of support following surgery. After tripling, the VIS must be 10 or higher. 3. AVO2 difference >40% by invasive measurement or NIRS 4. LCOS documented in physician note If Yes, document the earliest date any of these criteria were met.
*

Criteria #2:
A patient who is on no vasoactives and then reaches a VIS of 10 or greater in a 48 hour period is considered to meet the tripling criteria.

Do not compare preop and postop values. ONLY compare post operative values to determine if the VIS has tripled.

Criteria #4: the physician note does not have to specifically state 'low cardiac output syndrome.' Synonyms such as “poor oxygen delivery” or “oxygen debt” are sufficient. Please work with your clinical champion to understand what phrases to look for to indicate the patient had LCOS.

Please also note that some sites include LCOS (or one of its synonyms) as part of their templated notes for all patients receiving certain types of ICU-level care. Please work with your clinical champion to determine if this is the case and, if so, how best to distinguish these patients from those with true LCOS.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Initial LCOS date/time

Seq Num: 6861

Required for case closure: Yes

Registry field: [Complications].[CompLCOS2DtTm]

Description: If the patient did have LCOS, record the date/time the patient first met any criteria.

LCOS - VIS >15 Seq Num: 6870

Required for case closure: Yes

Registry field: [Complications].[CompLCOSvIS]

Description: At the initial LCOS date/time, did the patient have a VIS over 15?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

LCOS - VIS tripled Seq Num: 6872

Required for case closure: Yes

Registry field: [Complications].[CompLCOStriple]

Description: At the initial LCOS date/time, had the VIS tripled over a 48-hr period? After tripling, the score must be at least 10. This includes escalation of support following surgery.
*

A patient who is on no vasoactives and then reaches a VIS of 10 or greater in a 48 hour period is considered to meet the tripling criteria.

For this criteria, do not compare preop and postop values. ONLY compare post operative values to determine if the VIS has tripled.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

LCOS - AVO2 >40% Seq Num: 6874

Required for case closure: Yes

Registry field: [Complications].[CompLCOSavo]

Description: At the initial LCOS date/time, was the AVO2 difference >40% by invasive measurement or NIRS?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

LCOS - Physician note

Seq Num: 6876

Required for case closure: Yes

Registry field: [Complications].[CompLCOSnote]

Description: At the initial LCOS date/time, was there documentation of LCOS in a physician note?

*

For this criteria, the physician note does not have to specifically state 'low cardiac output syndrome.' Synonyms such as “poor oxygen delivery” or “oxygen debt” are sufficient. Please work with your clinical champion to understand what phrases to look for to indicate the patient had LCOS.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

LCOS timing

Seq Num: 6881

Required for case closure: Yes

Registry field: [Complications].[CompLCOS2Timing]

Description: If the patient underwent cardiothoracic surgery during or immediately before this encounter, indicate whether this first episode of LCOS occurred preop or postop. If the initial episode was prior to the first cardiothoracic surgery for this encounter, select "Preop". If it was after the first cardiothoracic surgery for this encounter, select "Postop." If the patient did not have cardiothoracic surgery immediately before or during this encounter, select "N/A." Please do not select this answer until the patient has been discharged from the CICU to be sure that no cardiothoracic surgery ever took place.

*

This question refers to the first surgery of type “CPB Cardiovascular” or “No CPB Cardiovascular” during this CICU encounter. If LCOS is initially diagnosed prior to this surgery, it’s preop; if it is after this surgery, it’s postop (even if the patient goes on to have additional surgeries during the encounter.)

Surgeries of type “ECMO”, “VAD” (with or without bypass), “Thoracic”, etc., should not be considered when coding this as pre- or postop.

Values	*Code*	*Text*
	1	Preop
	2	Postop
	3	N/A
	9	Unk

Additional postop LCOS Seq Num: 6901

Required for case closure: Yes

Registry field: [Complications].[CompLCOS2Post]

Description: If the initial LCOS episode recorded was preop, was there a subsequent episode of LCOS during the postoperative period of this CICU encounter?

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Initial postop LCOS date/time Seq Num: 6911

Required for case closure: Yes

Registry field: [Complications].[CompLCOS2PostDtTm]

Description: If the patient did have a subsequent postop LCOS episode, record the date/time the patient first met the criteria in the postoperative period.

Postop LCOS - VIS >15 Seq Num: 6920

Required for case closure: Yes

Registry field: [Complications].[CompLCOSpostVIS]

Description: At the initial postop LCOS date/time, did the patient have a VIS over 15?

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Postop LCOS - VIS tripled Seq Num: 6922

Required for case closure: Yes

Registry field: [Complications].[CompLCOSpostTriple]

Description: At the initial postop LCOS date/time, had the VIS tripled over a 48-hr period? After tripling, the score must be at least 10. This excludes escalation of support following surgery.

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Postop LCOS - AVO2 >40% Seq Num: 6924

Required for case closure: Yes

Registry field: [Complications].[CompLCOSpostAVO]

Description: At the initial postop LCOS date/time, was the AVO2 difference >40% by invasive measurement or NIRS?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Postop LCOS - Physician note Seq Num: 6926

Required for case closure: Yes

Registry field: [Complications].[CompLCOSpostNote]

Description: At the initial postop LCOS date/time, was there documentation of LCOS in a physician note?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

ICU-level treatment for PHTN

Seq Num: 6930

Required for case closure: Yes

Registry field: [Complications].[PHTNtherapy]

Description: During this CICU encounter, was continuous inhaled, continuous IV, or continuous subcutaneous therapy ever given for suspected or defined pulmonary artery hypertension or elevated pulmonary vascular resistance? Examples include inhaled nitric oxide, prostacyclin and remodulin. This does not include nitric oxide given for hypoxemia when there was clearly no pulmonary hypertension, nor does it include enteral therapy.
*

Do not include oral sildenafil here.

Do not code every patient on inhaled nitric oxide (iNO) has having PHTN. Only code patients receiving iNO for pulmonary hypertension / increased pulmonary vascular resistance. For example, patients on prophylactic iNO following stage II surgery and post-transplant patients on iNO for RV afterload reduction should not be coded as having PHTN.

Additionally, PHTN or increased PVR can be presumed by the clinician without other objective data. If the treating clinician believes that the patient has PHTN or elevated PVR and treats the patient with iNO based on that, then code PHTN.

Please note that patients on iNO for any reason should also be coded as Nitric Oxide = Yes (# 5600) in the Therapy section.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

PHTN treatment start date/time

Seq Num: 6940

Required for case closure: Yes

Registry field: [Complications].[PHTNtherDtTm]

Description: Enter the date/time any ICU-level treatment was initiated.

Inhaled NO for PHTN Seq Num: 7000

Required for case closure: Yes

Registry field: [Complications].[PHTNnox]

Description: Was inhaled nitric oxide used for treatment of pulmonary hypertension during this CICU encounter. This does not include nitric oxide given for hypoxemia when there was clearly no pulmonary hypertension.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Prostacyclin for PHTN Seq Num: 7040

Required for case closure: Yes

Registry field: [Complications].[PHTNprost]

Description: Was prostacyclin used for treatment of pulmonary hypertension during this CICU encounter.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Remodulin for PHTN Seq Num: 7060

Required for case closure: Yes

Registry field: [Complications].[PHTNremod]

Description: Was remodulin used for treatment of pulmonary hypertension during this CICU encounter.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Other therapy for PHTN Seq Num: 7065

Required for case closure: Yes

Registry field: [Complications].[PHTNother]

Description: Was another continuous agent used for treatment of PHTN during this CICU encounter?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Other PHTN therapy - specify Seq Num: 7066

Required for case closure: No

Registry field: [Complications].[PHTNotherSpec]

Description: Specify the other continuous agent

PHTN treatment at CICU end Seq Num: 6960

Required for case closure: Yes

Registry field: [Complications].[PHTNCICUend]

Description: Was the patient still receiving ICU-level treatment for PHTN at CICU discharge?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

PHTN treatment end date/time Seq Num: 6980

Required for case closure: Yes

Registry field: [Complications].[PHTNendDtTm]

Description: Date/time all PHTN treatments were discontinued. If the patient was still receiving treatment for PHTN at CICU discharge, this will default to the CICU discharge date/time.

Arrhythmia end date/time Seq Num: 6620

Retired in version 2.0

Required for case closure: Yes

Registry field: [CompArrhythmia].[ArrhythEndDtTm]

Description: Date/time this arrhythmia ended. This is defined as the time all treatments were discontinued. If the patient was still receiving treatment for this arrhythmia at CICU discharge, this will default to the CICU discharge date/time.

Pulmonary vein obstruction Seq Num: 6800

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPulmVeinObstr]

Description: Indicate whether the patient was diagnosed with a pulmonary vein obstruction during the CICU encounter. Pulmonary vein obstruction is defined as clinically significant stenosis or obstruction of pulmonary veins. Typically diagnosed by echocardiography or cardiac catheterization, this may present with or without symptoms. A "clinically significant" event or condition is an event or condition that necessitates a change in treatment.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Pulmonary vein obstruction dx date Seq Num: 6810

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPulmVeinObstrDt]

Description: If the patient had a pulmonary vein obstruction, document the date it was first diagnosed during the CICU encounter.

Systemic vein obstruction Seq Num: 6820

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompSystVeinObstr]

Description: Was the patient diagnosed with a systemic vein obstruction during the CICU encounter? Systemic vein obstruction is defined as clinically significant stenosis or obstruction of any major systemic vein (e.g., superior vena cava, inferior vena cava, femoral veins, internal jugular veins, etc.). A "clinically significant" event or condition is an event or condition that necessitates a change in treatment.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Systemic vein obstruction dx date Seq Num: 6830

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompSystVeinObstrDt]

Description: If the patient had a systemic vein obstruction, document the date it was first diagnosed during the CICU encounter.

Low cardiac output syndrome (LCOS) Seq Num: 6850

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompLCOS]

Description: Did the patient have any episodes of low cardiac output syndrome (LCOS) during the CICU encounter? LCOS is defined by at least one of the following: 1. VIS >15 at any time 2. Addition of a new vasoactive agent (inotrope or pressor or milrinone if note specifies this was started for LCOS, oxygen debt etc.; not Esmolol, Nipride) for patients already on inotropic or vasopressor support 3. New initiation of vasoactive support (inotropes or vasopressors or milrinone; i.e. not Esmolol, Nipride, etc.) after a 24 hour period with no support. (For example, if a patient was weaned off low dose (VIS<15) Dopamine and Milrinone on POD 1, but then developed respiratory failure and required an Epinephrine infusion on POD 4, you would record LCOS on POD 4 and note the date and time. If a patient who is on no continuous vasoactive support has vasoactive agents initiated, they would meet the criteria for LCOS) 4. Widened A-V difference noted by physician 5. LCOS documented in physician note (If an attending physician notes an episode of LCOS, oxygen debt, circulatory failure, the date and time of that event should be

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

LCOS date/time Seq Num: 6860

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompLCOSdtTm]

Description: If the patient did have LCOS, record the date/time the patient first met the criteria.

LCOS timing

Seq Num: 6880

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompLCOStiming]

Description: If the patient underwent cardiothoracic surgery during or immediately before this encounter, indicate whether this first episode of LCOS occurred preop or postop. If the initial episode was prior to the first cardiothoracic surgery for this encounter, select "Preop". If it was after the first cardiothoracic surgery for this encounter, select "Postop." If the patient did not have cardiothoracic surgery immediately before or during this encounter, select "N/A." Please do not select this answer until the patient has been discharged from the CICU to be sure that no cardiothoracic surgery ever took place.

Values	*Code*	*Text*
Retired	1	Preop
Retired	2	Postop
Retired	3	N/A
Retired	9	Unk

Additional postop LCOS

Seq Num: 6900

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompLCOSpostop]

Description: If the initial LCOS episode recorded was preop, was there a subsequent episode of LCOS during the postoperative period of this CICU encounter?

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Postop LCOS date/time

Seq Num: 6910

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompLCOSpostopDtTm]

Description: If the patient did have a subsequent postop LCOS episode, record the date/time the patient first met the criteria in the postoperative period.

Inhaled iloprost for PHTN

Seq Num: 7020

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[PHTNilo]

Description: Was inhaled iloprost used for treatment of pulmonary hypertension during this CICU encounter.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Listed for heart transplant

Seq Num: 7160

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompHeartTxListed]

Description: During the CICU encounter, was the patient placed on the transplant list or transferred to another institution with the expressed intent to list (e.g., the current hospital has no transplant program). If there was an active transplant evaluation during the CICU encounter but the listing occurred at the same hospital after CICU discharge, code Yes.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Date listed for heart transplant

Seq Num: 7180

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompHeartTxListedDt]

Description: If the patient was placed on the heart transplant list, document the date of listing. If the actual listing took place after CICU discharge, use the CICU discharge date.

Medical Events and Complications-Operative/Procedural

Bleeding requiring reoperation

Seq Num: 7200

Required for case closure: Yes

Registry field: [Complications].[CompReopBleed]

Description: Did the patient have any postoperative or postprocedural bleeding requiring re-exploration during the CICU encounter

*

If the patient is taken to the OR by the CT surgeons and re-opened to drain blood in the pericardial and/or pleural space, record as a reoperation for bleeding only (i.e., do not need to also code hemothorax or hemopericardium).

Do not record an event if the bleeding complication was due to a procedure or surgery that was not performed by the CT surgeons.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Date/time first reop for bleeding

Seq Num: 7220

Required for case closure: Yes

Registry field: [Complications].[CompReopBleedDtTm]

Description: Date/time of the first reop for bleeding.

Sternum left open

Seq Num: 7280

Required for case closure: Yes

Registry field: [Complications].[CompSternumOpen]

Description: Was the sternum left open postoperatively, whether planned or unplanned, with the goal of delayed sternotomy closure? Code Yes if the sternum was left open at any time. This includes patients who return from the OR with an open chest as well as those who are re-opened following an initial closure. Record every time the sternum was opened or re-opened during this CICU encounter.

*

This field is designed to capture all patients with an open sternum in the ICU and to calculate the days of open chest in the ICU. All patients whose sternum was left open in the ICU – whether planned or unplanned – must be coded as Yes.

This includes patients who return from the OR with an open chest (#9210) as well as those re-opened following an initial closure.

All patients coded as Yes on #9210 must also be coded as Yes to this complication.

Values	*Code*	*Text*
	1	Yes
	0	No

Date sternum left open

Seq Num: 7288

Required for case closure: Yes

Registry field: [CompSternum].[SternumDt]

Description: Record the date the sternum was left open or re-opened. If this was before the CICU admission, use the CICU admit date.

Venue opened

Seq Num: 7290

Required for case closure: Yes

Registry field: [CompSternum].[SternumVenue]

Description: Record the venue in which the sternum was opened

Values	*Code*	*Text*
	1	OR
	2	CICU
	8	Other
	9	Unk

Closed during this encounter

Seq Num: 7292

Required for case closure: Yes

Registry field: [CompSternum].[SternumClosed]

Description: For this instance of open sternum, was it closed before the end of this CICU encounter?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Date sternum closed

Seq Num: 7294

Required for case closure: Yes

Registry field: [CompSternum].[SternumClosedDt]

Description: Record the date the sternum was closed.

Intraoperative death or intraprocedural death

Seq Num: 7300

Required for case closure: Yes

Registry field: [Complications].[CompIntraopDeath]

Description: Did the patient die in the operating room or procedure room (such as catheterization laboratory or hybrid suite) during an operation or procedure

Values	*Code*	*Text*
	1	Yes
	0	No

Unplanned reoperation or reintervention

Seq Num: 7240

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompReopUnplan]

Description: Did the patient have any additional unplanned cardiac reoperation, unplanned EP, or unplanned interventional cardiac catheterization during the CICU encounter? These include interventions for infection, hemodynamic instability, and residual or recurrent lesion. Delayed sternal closure, ECMO decannulation, VAD decannulation, and removal of Broviac catheter should not be included. Reoperation for bleeding should be documented in the "Bleeding requiring reoperation" field.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Date/time first unplanned reop/reintervention

Seq Num: 7260

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompReopUnplanDtTm]

Description: Date/time of first unplanned reoperation/reintervention (not for bleeding).

Medical Events and Complications-Respiratory

Chylothorax requiring intervention

Seq Num: 7320

Required for case closure: Yes

Registry field: [Complications].[CompChyloIntv]

Description: Was the patient diagnosed with a chylothorax requiring intervention during the CICU encounter? Chylothorax can be determined by clinical status as documented in a note or by laboratory data (fluid with elevated triglyceride, cholesterol and/or lymphocyte count).

For purposes of this definition, a change in diet is considered an intervention.
*

A patient who returned to the CICU from the OR with a chest tube in place, has chylous drainage, but no change in diet should not be coded as chylothorax requiring intervention. There was no intervention directly related to the chylothorax. If a new chest tube had been placed, or the patient's diet was changed, that would be coded as Yes to this field.

A patient who has a chest tube placed in the CICU to treat a pleural effusion, has chylous fluid the next day, and is then started on a low-fat diet, should be coded as chylothorax treated with **both** a chest tube (#7401) and diet change (#7320).

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Chylothorax treatment date

Seq Num: 7341

Required for case closure: Yes

Registry field: [Complications].[CompChyloIntvDt]

Description: Document the date chylothorax treatment was initiated during this CICU encounter

Chest tube for chylothorax

Seq Num: 7360

Required for case closure: Yes

Registry field: [Complications].[CompChyloChestTube]

Description: If the patient was diagnosed with a chylothorax, indicate whether it required placement of a chest tube during this CICU encounter

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Initial chylothorax chest tube date Seq Num: 7381

Required for case closure: Yes

Registry field: [Complications].[CompChyloChestTubeDt]

Description: Document the date the first chest tube was placed to treat the chylothorax

Multiple chest tubes for chylothorax Seq Num: 7382

Required for case closure: Yes

Registry field: [Complications].[CompChyloChestTubeMult]

Description: Were multiple chest tubes placed specifically for chylothorax during this encounter
*
Code Yes both for patients who had multiple tubes placed at the same time, and patients where a new tube was placed a few days after the first one.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Diet change for chylothorax Seq Num: 7383

Required for case closure: Yes

Registry field: [Complications].[CompChyloDiet]

Description: At any time during this encounter, was the chylothorax treated with a change in diet?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

NPO for chylothorax Seq Num: 7384

Required for case closure: Yes

Registry field: [Complications].[CompChyloNPO]

Description: At any time during this encounter, was the patient made NPO specifically due to the chylothorax?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Octreotide for chylothorax Seq Num: 7385

Required for case closure: Yes

Registry field: [Complications].[CompChyloOctreotide]

Description: At any time during this encounter, was the chylothorax treated with an octreotide infusion?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Sildenafil for chylothorax Seq Num: 7386

Required for case closure: Yes

Registry field: [Complications].[CompChyloSildenafil]

Description: At any time during this encounter, was sildenafil used specifically to treat the chylothorax?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Other treatment for chylothorax Seq Num: 7387

Required for case closure: Yes

Registry field: [Complications].[CompChyloOther]

Description: At any time during this encounter, was another therapy used specifically to treat the chylothorax?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Other treatment for chylothorax - specify Seq Num: 7388

Required for case closure: No

Registry field: [Complications].[CompChyloOtherSpec]

Description: Specify the other chylothorax treatment

Pleural effusion/hemothorax requiring chest tube

Seq Num: 7401

Required for case closure: Yes

Registry field: [Complications].[CompEffusionTube]

Description: Was the patient diagnosed with a pleural effusion requiring placement of a chest tube during this CICU encounter? Pleural effusion is defined as an abnormal accumulation of fluid in the pleural space requiring drainage with a chest tube. Pleural effusions include hemothorax (blood accumulation in the pleural space)

*

If the patient is taken to the OR by the CT surgeons and re-opened to drain blood in the pericardial and/or pleural space, record as a reoperation for bleeding only (#7200). You do not need to also code hemothorax or hemopericardium.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Initial pleural effusion/hemothorax chest tube date

Seq Num: 7405

Required for case closure: Yes

Registry field: [Complications].[CompEffusionTubeDt]

Description: Document the date the first chest tube was placed to treat the pleural effusion / hemothorax.

Multiple chest tubes for pleural effusion/hemothorax

Seq Num: 7407

Required for case closure: Yes

Registry field: [Complications].[CompEffusionTubeMult]

Description: Were multiple chest tubes placed specifically for pleural effusion/hemothorax during this encounter?

*

Code Yes for both patients who had multiple tubes placed at the same time, and patients where a new tube was placed a few days after the first one.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Pneumothorax requiring chest tube Seq Num: 7481

Required for case closure: Yes

Registry field: [Complications].[CompPneumothoraxTube]

Description: Was the patient diagnosed with a pneumothorax requiring placement of a chest tube during the CICU encounter? Pneumothorax is defined as a collection of gas in the pleural space resulting in collapse of some or all of the lung on the affected side.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Initial pneumothorax chest tube date Seq Num: 7485

Required for case closure: Yes

Registry field: [Complications].[CompPneumothoraxTubeDt]

Description: Document the date the first chest tube was placed to treat pneumothorax

Multiple chest tubes for pneumothorax Seq Num: 7487

Required for case closure: Yes

Registry field: [Complications].[CompPneumothoraxTubeMult]

Description: Were multiple chest tubes placed specifically for pneumothorax during this encounter?
*
Code Yes for both patients who had multiple tubes placed at the same time, and patients where a new tube was placed a few days after the first one.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Pulmonary embolism Seq Num: 7680

Required for case closure: Yes

Registry field: [Complications].[CompPulmEmbol]

Description: Was the patient diagnosed with a pulmonary embolism during this CICU encounter? Pulmonary embolism is defined as the embolization of a clot or other foreign material to the pulmonary vasculature documented by CT angiogram, nuclear medicine scan, MRI or angiography. A thrombus in a cavopulmonary anastomosis pathway should be coded as a pulmonary embolism.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Pulmonary embolism date Seq Num: 7682

Required for case closure: Yes

Registry field: [Complications].[CompPulmEmbolDt]

Description: Document the date the pulmonary embolism was first diagnosed during this CICU encounter

Chylothorax treatment date/time Seq Num: 7340

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompChyloIntvDtTm]

Description: If the patient was diagnosed with a chylothorax, document the date/time treatment was initiated during this CICU encounter

Chest tube date/time Seq Num: 7380

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompChyloChestTubeDtTm]

Description: If the patient had a chylothorax requiring a chest tube, document the date/time the chest tube was placed

Pleural effusion requiring drainage

Seq Num: 7400

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPleuralEff]

Description: Was the patient diagnosed with a pleural effusion requiring drainage during this CICU encounter? Pleural effusion is defined as an abnormal accumulation of fluid in the pleural space requiring drainage by any technique.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Pleural effusion drainage date/time

Seq Num: 7420

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPleuralEffDrainDtTm]

Description: If the patient was diagnosed with a pleural effusion, document the date/time drainage was initiated during this CICU encounter

Chest tube for pleural effusion

Seq Num: 7440

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPleuralEffChestTube]

Description: If the patient was diagnosed with a pleural effusion, did it require placement of a chest tube during this CICU encounter?

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Chest tube date/time

Seq Num: 7460

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPleuralEffChestTubeDtTm]

Description: If the patient had a pleural effusion requiring a chest tube, document the date/time the chest tube was placed

Pneumothorax requiring intervention Seq Num: 7480

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPneumo]

Description: Was the patient diagnosed with a pneumothorax requiring intervention during the CICU encounter? Pneumothorax is defined as a collection of gas in the pleural space resulting in collapse of some or all of the lung on the affected side, requiring intervention.

Values	Code	Text
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Pneumothorax date/time Seq Num: 7500

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPneumoDtTm]

Description: If the patient was diagnosed with a pneumothorax, document the date/time intervention was initiated during this CICU encounter

Chest tube for pneumothorax Seq Num: 7520

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPneumoChestTube]

Description: If the patient was diagnosed with a pneumothorax, did it require placement of a chest tube during this CICU encounter?

Values	Code	Text
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Chest tube date/time Seq Num: 7540

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPneumoChestTubeDtTm]

Description: If the patient had a pneumothorax requiring a chest tube, document the date/time the chest tube was placed

Hemothorax requiring intervention Seq Num: 7560

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompHemo]

Description: Was the patient diagnosed with a hemothorax requiring intervention during the CICU encounter? Hemothorax is defined as the presence of blood in the pleural space requiring drainage by any technique.

Values	Code	Text
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Hemothorax date/time Seq Num: 7580

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompHemoDtTm]

Description: If the patient was diagnosed with a hemothorax, document the date/time intervention was initiated during this CICU encounter

Chest tube for hemothorax Seq Num: 7600

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompHemoChestTube]

Description: If the patient was diagnosed with a hemothorax, did it require placement of a chest tube during this CICU encounter?

Values	Code	Text
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Chest tube date/time Seq Num: 7620

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompHemoChestTubeDtTm]

Description: If the patient had a hemothorax requiring a chest tube, document the date/time the chest tube was placed

ARDS Seq Num: 7640

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompARDS]

Description: Was the patient diagnosed with acute respiratory distress syndrome (ARDS) during this CICU encounter? ARDS is defined as a clinical syndrome with a variety of etiologies characterized by refractory hypoxemia and bilateral diffuse interstitial infiltrates on chest radiography (CXR), as well as stiff lungs with decreased compliance, increased intrapulmonary shunting, and decreased airway dead space.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

ARDS date/time Seq Num: 7660

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompARDSDtTm]

Description: If the patient was diagnosed with ARDS, document the date/time it was diagnosed during this CICU encounter

Pulmonary embolism date/time Seq Num: 7700

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPulmEmbolDtTm]

Description: If the patient did have a pulmonary embolism, document the date/time it was first diagnosed during this CICU encounter

Medical Events Complications-Infectious

Superficial surgical site infection (SSI)

Seq Num: 7880

Required for case closure: Yes

Registry field: [Complications].[CompSupWoundInf]

Description: Did the patient have a superficial surgical site infection (superficial SSI), as defined by the CDC, during this CICU encounter or within 48 hours of discharge? These procedure-related infections must be adjudicated by local infection control for newly acquired infections in the CICU (i.e., not present on admission). Code clear examples of these infections present on admission that could not be adjudicated to the CICU by local infection control.
*
If a patient is admitted to the CICU with an infection from the floor or an outside hospital, please code these as events on arrival even though your infection control team would not adjudicate these as attributable to the CICU. We will filter such events out on the reporting platform, but if they are entered we can understand how to account for them in other analyses.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Superficial SSI date

Seq Num: 7900

Required for case closure: Yes

Registry field: [Complications].[CompSupWoundInfDt]

Description: Initial date a superficial SSI was diagnosed. If the infection was present on admission, use the CICU admit date.

Deep surgical site infection (SSI)

Seq Num: 7920

Required for case closure: Yes

Registry field: [Complications].[CompWoundInf]

Description: Was the patient diagnosed with a deep incisional infection or organ space infection (mediastinitis), as defined by the CDC, during this CICU encounter or within 48 hours of discharge? These procedure-related infections must be adjudicated by local infection control for newly acquired infections in the CICU (i.e., not present on admission). Code clear examples of these infections present on admission that could not be adjudicated to the CICU by local infection control.

*

If a patient is admitted to the CICU with an infection from the floor or an outside hospital, please code these as events on arrival even though your infection control team would not adjudicate these as attributable to the CICU. We will filter such events out on the reporting platform, but if they are entered we can understand how to account for them in other analyses.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Deep SSI date

Seq Num: 7960

Required for case closure: Yes

Registry field: [CompWoundInfection].[WoundInfDt]

Description: Date this deep SSI was diagnosed during the CICU encounter. If it was present on admission, use the CICU admit date.

Deep SSI organism

Seq Num: 7980

Required for case closure: Yes

Registry field: [CompWoundInfection].[WoundInfOrgism]

Description: Type of organism associated with this deep SSI.

Values	*Code*	*Text*
	1	Gram negative
	2	Gram positive
	3	Mixed
	4	Fungal
	9	Unk

CLABSI

Seq Num: 7760

Required for case closure: Yes

Registry field: [Complications].[CompCABSI]

Description: Did the patient have central line-associated blood stream infection (CLABSI), as defined by the CDC, during this CICU encounter or within 48 hours of discharge? If yes, document every CLABSI that occurred. These device-related infections must be adjudicated by local infection control for newly acquired infections in the CICU (i.e., not present on admission). Code clear examples of these infections present on admission that could not be adjudicated to the CICU by local infection control.

*

If a patient is admitted to the CICU with an infection from the floor or an outside hospital, please code these as events on arrival even though your infection control team would not adjudicate these as attributable to the CICU. We will filter such events out on the reporting platform, but if they are entered we can understand how to account for them in other analyses.

Please follow the rules your local infection control is using for post-transfer CLABSIs. They may say you should only include infections through 1 calendar day following transfer off the unit. On the PC4 reporting platform, we will exclude infections that occur more than 1 calendar day after transfer to ensure that data from all sites are comparable.

We will also remove intracardiac line days from the denominator of the CLABSI/line day metric, since intracardiac lines are not included in the CDC CLABSI definition.

Values

Code	*Text*
1	Yes
0	No
9	Unk

CLABSI date

Seq Num: 7800

Required for case closure: Yes

Registry field: [CompCABSI].[CABSIDt]

Description: Date this CLABSI was diagnosed.

CLABSI organism

Seq Num: 7820

Required for case closure: Yes

Registry field: [CompCABSI].[CABSIOrganism]

Description: Type of organism associated with the CLABSI

Values	*Code*	*Text*
	1	Gram negative
	2	Gram positive
	3	Mixed
	4	Fungal
	9	Unknown

UTI

Seq Num: 8040

Required for case closure: Yes

Registry field: [Complications].[CompUTI]

Description: Was the patient diagnosed with a urinary tract infection (UTI), as defined by the CDC, during the CICU encounter or within 48 hours of CICU discharge? This includes both catheter-associated and non-catheter-associated UTIs. If Yes, document ever UTI that occurred. CAUTIs must be adjudicated by local infection control for newly acquired infections in the CICU (i.e., not present on admission). Code clear examples of these infections present on admission that could not be adjudicated to the CICU by local infection control. Non-CAUTI UTIs need not be adjudicated by local infection control.

*

If a patient is admitted to the CICU with an infection from the floor or an outside hospital, please code these as events on arrival even though your infection control team would not adjudicate these as attributable to the CICU. We will filter such events out on the reporting platform, but if they are entered we can understand how to account for them in other analyses.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

UTI date

Seq Num: 8080

Required for case closure: Yes

Registry field: [CompUTI].[UTIDt]

Description: Date this UTI was diagnosed

UTI organism Seq Num: 8100

Required for case closure: Yes

Registry field: [CompUTI].[UTIOrganism]

Description: Type of organism associated with this UTI

Values	*Code*	*Text*
	1	Gram negative
	2	Gram positive
	3	Mixed
	4	Fungal
	9	Unk

CAUTI Seq Num: 8101

Required for case closure: Yes

Registry field: [CompUTI].[CAUTI]

Description: Was this a catheter-associated UTI as defined by the CDC? These device-related infections must be adjudicated by local infection control for newly acquired infections in the CICU (i.e., not present on admission). Code clear examples of these infections present on admission that could not be adjudicated to the CICU by local infection control.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

VAP

Seq Num: 7721

Required for case closure: Yes

Registry field: [Complications].[CompVAP]

Description: Did the patient have ventilator-associated pneumonia, as defined by the CDC, during this CICU encounter or within 48 hours of discharge? These device-related infections must be adjudicated by local infection control for newly acquired infections in the CICU (i.e., not present on admission). Code clear examples of these infections present on admission that could not be adjudicated to the CICU by local infection control.

*

If a patient is admitted to the CICU with an infection from the floor or an outside hospital, please code these as events on arrival even though your infection control team would not adjudicate these as attributable to the CICU. We will filter such events out on the reporting platform, but if they are entered we can understand how to account for them in other analyses.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

VAP date

Seq Num: 7722

Required for case closure: Yes

Registry field: [Complications].[CompVAPdt]

Description: Record the earliest date VAP was diagnosed during the CICU encounter. If it was present on admission, use the CICU admit date.

Non-VAP pneumonia

Seq Num: 7741

Required for case closure: Yes

Registry field: [Complications].[CompNonVAP]

Description: Did the patient have non-VAP pneumonia as defined by the CDC, during this CICU encounter?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Non-VAP pneumonia date

Seq Num: 7742

Required for case closure: Yes

Registry field: [Complications].[CompNonVAPDt]

Description: Record the date the non-VAP pneumonia was first diagnosed during this CICU encounter. If it was present on admission, use the CICU admit date.

Sepsis Seq Num: 7840

Required for case closure: Yes

Registry field: [Complications].[CompSepsis]

Description: Was the patient diagnosed with sepsis during the CICU encounter? Sepsis is defined as temperature instability and abnormal WBC (leukopenia or leukocytosis) and either (1) initiation or escalation of inotropic support or (2) initiation or escalation of mechanical ventilation. In addition the patient must be treated with antibiotics for > 6 days.
*
Criteria #2, initiation or escalation of mechanical ventilation, refers only to invasive ventilation.

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Sepsis date Seq Num: 7860

Required for case closure: Yes

Registry field: [Complications].[CompSepsisDt]

Description: If the patient had sepsis, document the date it was first diagnosed during the CICU encounter.

Positive culture for sepsis Seq Num: 7862

Required for case closure: Yes

Registry field: [Complications].[CompSepsisCulture]

Description: Did the patient have a positive culture suggesting a pathogenic organism during a sepsis episode? This organism would be presumed as the cause of sepsis.

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Endocarditis Seq Num: 6520

Required for case closure: Yes

Registry field: [Complications].[CompEndocard]

Description: Indicate whether the patient had endocarditis, as defined by the modified Duke criteria, during the CICU encounter.

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Endocarditis dx date Seq Num: 6522

Required for case closure: Yes

Registry field: [Complications].[CompEndocardDt]

Description: Record the date endocarditis was diagnosed during the CICU encounter. If it was present on admission, use the CICU admit date.

Pneumonia Seq Num: 7720

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPneumonia]

Description: Was the patient diagnosed with pneumonia, as defined by the CDC, during the CICU encounter? Pneumonia is defined as a "respiratory disease characterized by inflammation of the lung parenchyma (including alveolar spaces and interstitial tissue), most commonly caused by infection". Pneumonia is diagnosed by appropriate clinical findings (such as fever, leukopenia or leukocytosis, and new onset of purulent sputum) and one or more of the following: positive cultures (of sputum or pulmonary secretions) and/or pulmonary infiltrate on chest x-ray. An endotracheal tube culture may or may not be positive. Patients commonly demonstrate an evolving area of focal lung consolidation accompanied by fever (>38.5). Pneumonia (pneumonitis) may affect an entire lobe (lobar pneumonia), a segment of a lobe (segmental or lobular pneumonia), alveoli contiguous to bronchi (bronchopneumonia), or interstitial tissue (interstitial pneumonia). These distinctions are generally based on x-ray observations. If the infection began during the CICU encounter, mark Yes only if it has been adjudicated by the local infection control personnel. If the institution does not have infection control personnel, the clinician responsible for adjudicating infections for the purpose of external reporting must confirm the presence of the infection. If the infection began prior to CICU admission, code this complication and use the CICU admit date/time as the start date/time. These infections may not be adjudicated by the local infection control personnel or person responsible for adjudicating infections because they would not be attributed to the CICU.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Pneumonia date Seq Num: 7740

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPneumoniaDt]

Description: If the patient did have pneumonia, document the date it was first diagnosed during this CICU encounter

Meningitis

Seq Num: 8000

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompMeningitis]

Description: Was the patient diagnosed with meningitis, as defined by the CDC, during the CICU encounter or within 48 hours of CICU discharge. If the infection began during the CICU encounter, mark Yes only if it has been adjudicated by the local infection control personnel. If the institution does not have infection control personnel, the clinician responsible for adjudicating infections for the purpose of external reporting must confirm the presence of the infection. If the infection began prior to CICU admission, code this complication and use the CICU admit date/time as the start date/time. These infections may not be adjudicated by the local infection control personnel or person responsible for adjudicating infections because they would not be attributed to the CICU.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Meningitis date

Seq Num: 8020

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompMeningitisDt]

Description: If the patient had meningitis during the CICU encounter, document the date it was first diagnosed.

Medical Events and Complications-Neuro

Stroke

Seq Num: 8120

Required for case closure: Yes

Registry field: [Complications].[CompStroke]

Description: Was the patient diagnosed with a stroke during the CICU encounter? A stroke is defined as any confirmed neurological deficit of abrupt onset caused by a disturbance in blood flow to the brain, when the neurologic deficit does not resolve within 24 hours. If the patient is chemically sedated and/or neuromuscularly blocked or is unable to undergo imaging, physician documentation of stroke (e.g., a neurologic consultation) is sufficient for coding this event. Record each new stroke diagnosed during the encounter. The stroke must be in a new territory to be considered a distinct event.

*

Code HIE (hypoxic-ischemic encephalopathy) or other diffuse anoxic brain injury here as Stroke.

If routine brain imaging (e.g., many centers do MRIs on all neonates after bypass) detects a minor abnormality that was not suspected clinically prior to the study and which does not result in any intervention, the finding does not need to be documented as a complication. Adjudicate with the clinical champion as needed.

Please see the Stroke/Intracranial Hemorrhage Scenarios table in the Appendix at the end of this document for coding examples.

Values

Code	*Text*
1	Yes
0	No
9	Unk

Stroke date/time

Seq Num: 8142

Required for case closure: Yes

Registry field: [CompStroke].[StrokeDtTm]

Description: Document the date/time this stroke was diagnosed

*

For patients who exhibit clear clinical symptoms of a stroke, but who do not receive a confirmation via MRI until several days later, use the date/time the symptoms were noted. Code the method of diagnosis (#8144) as both clinical and imaging.

If there are no symptoms, use the imaging date/time.

How was this stroke diagnosed Seq Num: 8144

Required for case closure: Yes

Registry field: [CompStroke].[StrokeDx]

Description: Indicate whether this stroke was diagnosed through clinical findings, imaging, or both.

Values	*Code*	*Text*
	1	Clinical findings
	2	Imaging
	3	Both
	9	Unk

Cranial ultrasound to diagnose stroke Seq Num: 8146

Required for case closure: Yes

Registry field: [CompStroke].[StrokeDxUS]

Description: Was cranial ultrasound used to diagnose this stroke?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

MRI to diagnose stroke Seq Num: 8148

Required for case closure: Yes

Registry field: [CompStroke].[StrokeDxMRI]

Description: Was an MRI used to diagnose this stroke?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

CT to diagnose stroke Seq Num: 8150

Required for case closure: Yes

Registry field: [CompStroke].[StrokeDxCT]

Description: Was a CT scan used to diagnose this stroke?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Primarily hemorrhagic Seq Num: 8152

Required for case closure: Yes

Registry field: [CompStroke].[StrokeHemorrhage]

Description: Was this stroke hemorrhagic at the time of diagnosis?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Hemorrhagic conversion Seq Num: 8154

Required for case closure: Yes

Registry field: [CompStroke].[StrokeConvert]

Description: If this stroke was not primarily hemorrhagic, was there hemorrhagic conversion during this encounter?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Conversion date/time Seq Num: 8156

Required for case closure: Yes

Registry field: [CompStroke].[StrokeConvertDtTm]

Description: Record the date/time of hemorrhagic conversion.

Seizure Seq Num: 8220

Required for case closure: Yes

Registry field: [Complications].[CompSeizure]

Description: Was the patient diagnosed with a seizure during the CICU encounter? A seizure is defined as the clinical and/or electroencephalographic recognition of epileptiform activity.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Seizure date/time

Seq Num: 8240

Required for case closure: Yes

Registry field: [Complications].[CompSeizureDtTm]

Description: If the patient was diagnosed with a seizure, document the date/time of the first occurrence during the CICU encounter.

*

For patients admitted to the CICU with a known seizure disorder, this date/time is still the date/time of the first seizure.

IVH grade II or higher

Seq Num: 8260

Required for case closure: Yes

Registry field: [Complications].[CompIVH]

Description: Did the patient have a new finding of intraventricular hemorrhage (IVH) grade II or higher on cranial ultrasound during the CICU encounter? If using another nomenclature for IVH grading, determine the equivalent grade.

*

If routine brain imaging (e.g., many centers do MRIs on all neonates after bypass) detects a minor abnormality that was not suspected clinically prior to the study and which does not result in any intervention or further testing, the finding does not need to be documented as a complication. Adjudicate with the clinical champion as needed.

Please see the Stroke/Intracranial Hemorrhage Scenarios table in the Appendix at the end of this document for coding examples.

Values

Code	*Text*
1	Yes
0	No
9	Unk

IVH date/time

Seq Num: 8280

Required for case closure: Yes

Registry field: [Complications].[CompIVHDtTm]

Description: If the patient was diagnosed with IVH, document the date/time of the first occurrence during the CICU encounter.

Maximum IVH grade

Seq Num: 8282

Required for case closure: Yes

Registry field: [Complications].[CompIVHmax]

Description: Record the highest IVH grade diagnosed during the CICU encounter.

Values	*Code*	*Text*
	2	II
	3	III
	4	IV
	9	Unk

Max IVH grade date

Seq Num: 8284

Required for case closure: Yes

Registry field: [Complications].[CompIVHmaxDt]

Description: Record the date the highest grade was documented.

Intracranial hemorrhage (non-stroke)

Seq Num: 8302

Required for case closure: Yes

Registry field: [Complications].[CompIntracranial]

Description: Did the patient have an intracranial hemorrhage during the CICU encounter? This includes subdural hemorrhage, subarachnoid hemorrhage, and IVH not captured by the IVH complication. An intracranial hemorrhage is defined as the existence of a neurologic imaging study indicating a new or previously unsuspected focus of discrete central nervous system injury consistent with hemorrhage. Intracranial bleeding found on routine or research imaging studies should not be included. Hemorrhagic strokes, strokes with hemorrhagic conversion, and IVH grade 2 or higher should be coded as "No" and documented in the appropriate complication field.

*

If routine brain imaging (e.g., many centers do MRIs on all neonates after bypass) detects a minor abnormality that was not suspected clinically prior to the study and which does not result in any intervention or further testing, the finding does not need to be documented as a complication. Adjudicate with the clinical champion as needed.

Please see the Stroke/Intracranial Hemorrhage Scenarios table in the Appendix at the end of this document for coding examples.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Intracranial hemorrhage date/time

Seq Num: 8304

Required for case closure: Yes

Registry field: [Complications].[CompIntracranialDtTm]

Description: Document the date/time of the first occurrence of intracranial hemorrhage during the CICU encounter.

Brain death

Seq Num: 8340

Required for case closure: Yes

Registry field: [Complications].[CompBrainDeath]

Description: Was the patient declared brain dead by treating physician during this CICU encounter?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Brain death date/time

Seq Num: 8360

Required for case closure: Yes

Registry field: [Complications].[CompBrainDeathDtTm]

Description: If the patient was declared brain dead, document the date/time of declaration.

Stroke date/time

Seq Num: 8140

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompStrokeDtTm]

Description: If the patient was diagnosed with a stroke, document the date/time of the first occurrence during the CICU encounter.

Cranial ultrasound to diagnose stroke

Seq Num: 8160

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompStrokeDxUS]

Description: If the patient had a stroke, was cranial ultrasound used to diagnose it

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

CT to diagnose stroke

Seq Num: 8180

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompStrokeDxCT]

Description: If the patient had a stroke, was a CT scan used to diagnose it

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

MRI to diagnose stroke

Seq Num: 8200

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompStrokeDxMRI]

Description: If the patient had a stroke, was an MRI used to diagnose it

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Intracranial hemorrhage

Seq Num: 8300

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompHemorrage]

Description: Did the patient have an intracranial hemorrhage during the CICU encounter? Intracranial hemorrhage is defined as a stroke (i.e., any confirmed neurological deficit of abrupt onset caused by a disturbance in blood flow to the brain, when the neurologic deficit does not resolve within 24 hours) plus the existence of a neurologic imaging study indicating a new or previously unsuspected focus of discrete central nervous system injury with an appearance consistent with hemorrhage.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Intracranial hemorrhage date/time Seq Num: 8320

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompHemorrageDtTm]

Description: If the patient had an intracranial hemorrhage, document the date/time of the first occurrence during the CICU encounter.

Paralyzed diaphragm Seq Num: 8380

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompParalyzedDiaphragm]

Description: Was the patient diagnosed with a paralyzed diaphragm during the CICU encounter? A paralyzed diaphragm is defined as the presence of elevated hemi-diaphragm(s) on chest radiograph in conjunction with evidence of weak, immobile, or paradoxical movement assessed by ultrasound or fluoroscopy.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Paralyzed diaphragm dx date Seq Num: 8400

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompParalyzedDiaphragmDt]

Description: If the patient had a paralyzed diaphragm, document the date it was first diagnosed during the CICU encounter.

Vocal cord dysfunction

Seq Num: 8420

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompVocalCordDys]

Description: Was the patient diagnosed with vocal cord dysfunction during the CICU encounter? Vocal cord dysfunction is defined as the presence of poor or no vocal cord movement assessed by endoscopy. Patient may or may not have stridor, hoarse voice or poor cry, in conjunction with endoscopic findings.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Vocal cord dysfunction dx date

Seq Num: 8440

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompVocalCordDysDt]

Description: If the patient had vocal cord dysfunction, document the date/time it was first diagnosed during the CICU encounter.

Medical Events and Complications-Gastrointestinal

Hepatic injury (ALT>500)

Seq Num: 8462

Required for case closure: Yes

Registry field: [Complications].[CompHepaticInj]

Description: Was the patient diagnosed with hepatic injury, defined as an ALT > 500, during the CICU encounter?
*
If a patient did not have a liver panel done, please discuss with your clinical champion whether specific patients should be coded as No or Unknown. In general, if the patient had an uneventful ICU stay and there is no reason to suspect there were any issues with their liver function, you can code this as No.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Hepatic injury date

Seq Num: 8464

Required for case closure: Yes

Registry field: [Complications].[CompHepaticInjDt]

Description: Document the earliest date the patient had an ALT>500.

NEC - Bell's stage II or III

Seq Num: 8501

Required for case closure: Yes

Registry field: [Complications].[CompNECBell]

Description: Did the patient have NEC meeting Bell's criteria for stage II or III during the CICU encounter? If Yes, document each time a new diagnosis of NEC was made during the encounter.
Bell's criteria can be found at the end of this document.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

NEC dx date

Seq Num: 8508

Required for case closure: Yes

Registry field: [CompNECbell].[NECbellDt]

Description: Date this episode of NEC first met Stage II or Stage III criteria.

Surgery for NEC Seq Num: 8510

Required for case closure: Yes

Registry field: [CompNECbell].[NECbellSurg]

Description: Did the patient require surgery for this episode of NEC

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Date of NEC surgery Seq Num: 8512

Required for case closure: Yes

Registry field: [CompNECbell].[NECbellSurgDt]

Description: Date of first surgery for this episode of NEC

Hepatic failure Seq Num: 8460

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompHepaticFail]

Description: Was the patient diagnosed with hepatic failure during the CICU encounter? Hepatic failure is defined as dysfunction of the liver that results in hypoalbuminemia, coagulopathy, and hyperbilirubinemia. Select Yes if the patient develops all 3 of these laboratory abnormalities, or if the patient develops 2 out of these 3 laboratory abnormalities and at least one of the following complications: ascites, cirrhosis, encephalopathy, esophageal varices, and gastrointestinal bleeding.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Hepatic failure date Seq Num: 8480

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompHepaticFailDt]

Description: If the patient had hepatic failure, document the date it was first diagnosed during the CICU encounter.

NEC

Seq Num: 8500

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompNEC]

Description: Did the patient have NEC during the CICU encounter? If Yes, document each time a new diagnosis of NEC was made during the encounter. NEC is defined as an acute reduction in the supply of oxygenated blood to the small intestine or large intestine, typically resulting in acidosis, abdominal distention, pneumatosis, and/or intestinal perforation, that prompts initiation of antibiotics or exploratory laparotomy.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

NEC date

Seq Num: 8540

Retired in version 2.0

Required for case closure: Yes

Registry field: [CompNEC].[NECDt]

Description: Date this episode of NEC was diagnosed

Medical Events and Complications-Dermatologic

Pressure ulcer

Seq Num: 8560

Retired in version 2.0

Required for case closure: Yes

Registry field: [Complications].[CompPressUlcer]

Description: Did the patient have a pressure ulcer during the CICU encounter? A pressure ulcer is defined as a wound that occurs from tissue breakdown as a result of unrelieved pressure with the pressure usually occurring over an underlying bony prominence. Pressure ulcers may be caused by a mechanical device or other factors.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Medical Events and Complications-Other

Pressure ulcer stage III or higher

Seq Num: 8562

Required for case closure: Yes

Registry field: [Complications].[CompUlcer]

Description: Did the patient have a pressure ulcer stage III, stage IV, or unstageable during the CICU encounter? Stage III is full thickness skin loss with subcutaneous fat visible but not bone, tendon or muscle. Stage IV is full thickness tissue loss exposing bone, tendon or muscle. Unstageable is full thickness tissue loss but due to slough or eschar the depth cannot be determined.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Pressure ulcer date

Seq Num: 8564

Required for case closure: Yes

Registry field: [Complications].[CompUlcerDt]

Description: Record the earliest date a pressure ulcer stage III or higher was noted

Max pressure ulcer stage

Seq Num: 8566

Required for case closure: Yes

Registry field: [Complications].[CompUlcerMax]

Description: Record the maximum pressure ulcer stage as per international NPUAP pressure ulcer classification system.

Values	*Code*	*Text*	
	3	III	Full thickness skin loss with subcutaneous fat visible but not bone, tendon, or muscle
	4	IV	Full thickness tissue loss exposing bone, tendon, or muscle
	8	Unstageable	Full thickness tissue loss but, due to slough or eschar, the depth cannot be determined.
	9	Unk	

Hypoglycemia

Seq Num: 8580

Required for case closure: Yes

Registry field: [Complications].[CompHypoglycemia]

Description: Did the patient ever have a blood glucose value of < 40 mg/dL during the CICU encounter? Indicate Yes independent of whether or not patient received treatment.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Narcotic dependence requiring wean

Seq Num: 8600

Required for case closure: Yes

Registry field: [Complications].[CompNarcotic]

Description: Was the patient exposed to narcotic therapy that ultimately warranted transition to a narcotic weaning strategy? This could be exposure during this CICU encounter or those on a wean at admission to treat previous narcotic withdrawal. This does not include patients being treated for neonatal abstinence syndrome.

*

Do not code patients weaning from dexmedetomidine or from barbiturates as “Narcotic dependence requiring wean.” This field only captures patients weaning from narcotics. You may track these other weans if you wish in the 'Other complication' field.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Other complication

Seq Num: 8700

Required for case closure: No

Registry field: [Complications].[CompOther]

Description: Did the patient have any other complication during the CICU encounter?

*

Please use this field to capture a positive COVID19 diagnosis.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Other complication - specify Seq Num: 8701

Required for case closure: No

Registry field: [Complications].[CompOtherSpec]

Description: Specify the other complication(s)

Unplanned return to CICU care Seq Num: 6411

Retired in version 1.0

Required for case closure: Yes

Registry field: [Complications].[CompCICUReturnUnpln]

Description: Did the patient have an unplanned return to CICU service<48 hours after the end of an

Values	Code	Text
Retired	0	No
Retired	1	Yes

Narcotic dependence Seq Num: 6420

Retired in version 2.0

Required for case closure: No

Registry field: [Complications].[CompNarcDep]

Description: During the CICU encounter, was the patient exposed to either a significant amount or duration of narcotic therapy warranting long-term opiate therapy for the prevention of withdrawal

Values	Code	Text
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Encounter type

Cardiac surgery immediately before or during CICU encounter

Seq Num: 8900

Required for case closure: Yes

Registry field: [CICUEncounter].[RiskGroup]

Description: Did the patient have any cardiothoracic surgery of type "CPB Cardiovascular" or "No CPB Cardiovascular" either during or immediately prior to the start of the encounter? Please do not answer this question until (a) the patient has a qualifying surgery or (b) the patient has been discharged from the CICU without any qualifying surgeries.

*

Only surgeries coded as type = “CPB cardiovascular” or “No CPB cardiovascular” are eligible for the Surgical Risk section. If a patient had a VAD placement with CPB, but no qualifying surgeries of type CPB/No-CPB cardiovascular, you would code the encounter as medical.

All of the questions on the subsequent surgical risk tab are related to the first surgery of type CPB cardiovascular or no-CPB cardiovascular during or immediately before this CICU encounter.

If the list of surgeries to choose from is either empty or incomplete, the patient may not have had a qualifying surgery, or the ICU/PACU arrival date/time may be missing from one or more surgeries.

Values

Code	*Text*
1	Yes
0	No

Cardiac surgical encounters

Preop factor

Seq Num: 9000

Required for case closure: Yes

Registry field: [PreopFactor].[PreopFactor]

Description: Indicate the factors that are present preoperatively that may impact the patient's outcome.

Values	Code	Text	
	10	No preoperative factors identified	This patient has no preoperative factors identified.
	200	Cardio-pulmonary resuscitation	Chest compression with medications within 48 hours prior to surgery. Select this factor if chest compression took place during the 48 hours prior to OR Entry Date and Time, or at the time of OR Entry Date and Time.
	210	Preoperative complete AV block	Arrhythmia-Atrioventricular conduction disorder, AV block, Third degree ROOT Definition = Third degree AV block is defined as the absence of AV node conduction. This factor should be selected if it developed before OR Entry Date and Time and was present at the time of OR Entry Date and Time.
	220	Preoperative/Preprocedural mechanical circulatory support (IABP, VAD, ECMO, or CPS)	Code this factor if the patient is supported with mechanical support, of any type (IABP, VAD, ECMO, or CPS), for resuscitation/CPR or support, at the time of OR Entry Date and Time.
	230	Shock, Persistent at time of surgery	Shock ROOT Definition = Shock is defined as "a state of inadequate tissue perfusion". A modern definition according to Simeone states that shock is a "clinical condition characterized by signs and symptoms which arise when the cardiac output is insufficient to fill the arterial tree with blood under sufficient pressure to provide organs and tissues with adequate blood flow." A historic definition according to Blalock in 1940 is that "Shock is a peripheral circulatory failure, resulting from a discrepancy in the size of the vascular bed and the volume of the intravascular fluid". Code this factor if the patient had a metabolic acidosis with pH < 7.2 and/or Lactate > 4 mmol / liter at the time of OR Entry Date and Time.
	240	Shock, Resolved at time of surgery	Shock ROOT Definition = Shock is defined as "a state of inadequate tissue perfusion". A modern definition according to Simeone states that shock is a "clinical condition characterized by signs and symptoms which arise when the cardiac output is insufficient to fill the arterial tree with blood under sufficient pressure to provide organs and tissues with adequate blood flow." A historic definition according to Blalock in 1940 is that "Shock is a peripheral circulatory failure, resulting from a discrepancy in the size of the vascular bed and the volume of the intravascular fluid". Code this factor if the patient had a metabolic acidosis with pH < 7.2 and/or Lactate > 4 mmol / liter at any time after the date and time of admission to the hospital but not at the time of OR Entry Date and Time. This factor should be coded if shock was present at any time after the date and time of admission to the hospital but not at the time of OR Entry Date and Time, including situations where shock was present after admission to the hospital where this operation

240		was performed, and situations where shock was present while the patient was hospitalized at another "transferring facility" that subsequently transferred the patient who ultimately arrived at this hospital in this same hospitalization.
250	Diabetes mellitus, Insulin dependent	Code this factor if the patient has evidence of insulin dependent diabetes mellitus at the time OR Entry Date and Time as manifested by the fact that the patient has the diagnosis of diabetes mellitus that is controlled with insulin.
260	Diabetes mellitus, Non-insulin dependent	Code this factor if the patient has evidence of non-insulin dependent diabetes mellitus at the time OR Entry Date and Time as manifested by the fact that the patient has the diagnosis of diabetes mellitus that is controlled with dietary modification with or without oral medications (oral antihyperglycemic agents).
270	Hypothyroidism	Hypothyroidism refers to decreased levels of triiodothyronine (T3) and thyroxine (T4), and reverse triiodothyronine (reverse T3), with high levels of thyroid-stimulating hormone (TSH). Symptoms of hypothyroidism include bradycardia, pericardial effusions, hypertension and a narrowed pulse pressure and myxedema. Studies have also shown decreases in cardiac output and cardiac contractility, decreased diastolic relaxation and diastolic filling. In those with congestive heart failure (CHF), decreased levels of T3 have been shown to be proportional to New York Heart Association class, poor outcomes, mortality, poor hemodynamics, and hyponatremia. This factor may be coded (1) if the TSH > 20 mU / liter, or (2) if the patient has pituitary failure with hypothyroidism, or (3) if the patient is receiving medication to treat hypothyroidism.
280	Currently taking steroids as treatment for adrenal insufficiency	Code this factor if the patient is taking steroids (as treatment for adrenal insufficiency) at the time of OR Entry Date and Time.
290	Currently taking steroids for any reason other than treatment of adrenal insufficiency	Code this factor if the patient is taking steroids (for any reason other than treatment of adrenal insufficiency) at the time of OR Entry Date and Time.
295	Colostomy present	Code this factor if the patient has a colostomy (involving the large intestine) present at the time of OR Entry Date and Time.
300	Enterostomy of small intestine present	Code this factor if the patient has an enterostomy (involving the small intestine) present at the time of OR Entry Date and Time.
305	Esophagostomy present	Code this factor if the patient has an esophagostomy present at the time of OR Entry Date and Time.
307	Gastrostomy present	Code this factor if the patient has a gastrostomy present at the time of OR Entry Date and Time.
310	Hepatic dysfunction	Hepatic dysfunction is defined as dysfunction of the liver that results in hypoalbuminemia (<2 grams/dL), coagulopathy (PT > 1.5 x upper limits of normal), and hyperbilirubinemia (> 3.0 x upper limits of normal). Select this factor if the patient develops 2 out of these 3 laboratory abnormalities. Code this factor if the patient has evidence of hepatic dysfunction at the time OR Entry Date

310		and Time
320	Necrotizing entero-colitis, Treated medically	Necrotizing enterocolitis (NEC) ROOT Definition = Necrotizing enterocolitis is defined as an acute reduction in the supply of oxygenated blood to the small intestine or large intestine, typically resulting in acidosis, abdominal distention, pneumatosis, and/or intestinal perforation, that prompts initiation of antibiotics or exploratory laparotomy. Select this factor if NEC is present during the same hospitalization as this operation and was managed without surgery to treat the NEC.
330	Necrotizing entero-colitis, Treated surgically	Necrotizing enterocolitis (NEC) ROOT Definition = Necrotizing enterocolitis is defined as an acute reduction in the supply of oxygenated blood to the small intestine or large intestine, typically resulting in acidosis, abdominal distention, pneumatosis, and/or intestinal perforation, that prompts initiation of antibiotics or exploratory laparotomy. Select this factor if NEC is present during the same hospitalization as this operation and was managed with surgery to treat the NEC.
340	Coagulation disorder, Hypercoagulable state	Code this factor if the patient has evidence of a hypercoagulable state at the time OR Entry Date and Time.
350	Coagulation disorder, Hypocoagulable state not secondary to medication (intrinsic hypocoagulable state)	Code this factor if the patient has evidence of a coagulopathy at the time OR Entry Date and Time as manifest by PT/PTT above normal, Thrombocytopenia <100,000,ot Fibrinogen split products positive (>10%) and the coagulopathy is NOT secondary to medications such as Heparin or Warfarin.
360	Coagulation disorder, Hypocoagulable state secondary to medication	Code this factor if the patient has evidence of a coagulopathy at the time OR Entry Date and Time as manifest by PT/PTT above normal, Thrombocytopenia <100,000,ot Fibrinogen split products positive (>10%) and the coagulopathy is secondary to medications such as Heparin or Warfarin.
590	Dyslipidemia	Current or previous diagnosis of dyslipidemia according to National Cholesterol Education Program criteria, defined as any 1 of the following: - Total cholesterol greater than or equal to 200 mg/dL (5.18 mmol/L) - LDL greater than or equal to 130 mg/dL (3.37 mmol/L) - HDL less than or equal to 40 mg/dL (1.04 mmol/L) in males and less than or equal to 50 mg/dL (1.30 mmol/L) in females
370	Endocarditis	This factor should be coded if endocarditis present at any time after the date and time of admission to the hospital and prior to OR Entry Date and Time, including situations where endocarditis was present after admission to the hospital where this operation was performed, and situations where endocarditis was present while the patient was hospitalized at another “transferring facility” that subsequently transferred the patient who ultimately arrived at this hospital in this same hospitalization. Code this factor if endocarditis is diagnosed prior to OR Entry Date and Time, using the Duke Criteria for the Diagnosis of Infective Endocarditis (IE): The definitive diagnosis of infective endocarditis requires one of the following four situations: 1) Histologic and/or microbiologic evidence of infection at

370		surgery or autopsy such as positive valve culture or histology; 2) Two major criteria; 3) One major criterion and three minor criteria; 4) Five minor criteria. The two major criteria are: 1) Blood cultures positive for IE 2) Evidence of endocardial involvement. Blood cultures positive for IE requires: 1) Typical microorganism consistent with IE isolated from 2 separate blood cultures, as noted in number two below (viridans streptococci, Streptococcus bovis, Staphylococcus aureus, or HACEK group [HACEK, Haemophilus species {H. aprophilus and H. paraaphrophilus}, Actinobacillus actinoinycetemcomitans, Cardiobacterium hominis, Eikenella corrodens, and Kingella kingae.]) or (Community-acquired enterococci in the absence of a primary focus); 2) Microorganisms consistent with IE isolated from persistently positive blood cultures defined as: (At least 2 positive cultures of blood samples obtained > 12 hours apart) or (All of 3 or a majority of 4 or more separate cultures of blood, the first and the last sample obtained > 1 hr apart); 3) Single blood culture positive for Coxiella burnetii or an antiphase I IgG antibody titer of >1 :800. Evidence of endocardial involvement requires 1) Positive results of echocardiography for IE defined as: (Oscillating intracardiac mass on the valve or supporting structures in the path of regurgitant jets or on implanted material in the absence of an alternative anatomic explanation) or (Abscess) or (New partial dehiscence of a valvar prosthesis) or 2) New valvar regurgitation (worsening or changing or preexisting murmur not sufficient). The six minor criteria are: 1) Predisposing heart disease or injection drug use (IVDA); 2) Temperature of > 38C; 3) Vascular phenomenon (major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial or conjunctival hemorrhage, Janeway's lesions); 4) Immunologic phenomenon (glomerulonephritis, Osler's nodes, Roth's spots, rheumatoid factor); 5) Microbiologic evidence (a positive blood culture that does not meet a major criterion as noted above) or serologic evidence of active infection with an organism consistent with IE; 6) Echocardiographic findings that are consistent with IE but do not meet a major criterion as noted above. References: 1) Dhawan VK Infectious Endocarditis in Elderly Patients. Clin. Infect. Dis. 2002;34:806-812. 2) Durack DT, Lukes AS, Bright DK. New criteria for diagnosis of infective endocarditis: utilization of specific echocardiographic findings. Duke Endocarditis Service. Am. J. Med. 1994;96:200-209. 3) Li IS, Sexton DJ, Mick N, et al. Proposed modifications to the Duke criteria for the diagnosis of infective endocarditis. Clin. Infect. Dis. 2000;30:633-638. 4) http://gold.aecom.yu.edu/id/almanac/dukeendocarditis.htm, accessed July 5, 2006.
580	Family history of coronary artery disease	Indicate if the patient has/had any direct blood relatives (i.e., parents, siblings, children) who have had any of the following diagnosed at age less than 55 years for male relatives or less than 65 years for female relatives: - Coronary artery disease (i.e., angina, previous CABG or PCI) - MI

580		- Sudden cardiac death without obvious cause
380	Sepsis	Sepsis ROOT Definition = Sepsis is defined as "evidence of serious infection accompanied by a deleterious systemic response". Sepsis may be diagnosed by the presence of a Systemic Inflammatory Response Syndrome (SIRS) resulting from suspected or proven infection. A systemic inflammatory response syndrome (SIRS) is present when at least two of the following criteria are present: hypo- or hyperthermia (>38.5 or <36.0), tachycardia or bradycardia, tachypnea, leukocytosis or leukopenia, and thrombocytopenia. Code this factor if the patient has signs of sepsis within 48 hours of OR Entry Date and Time.
390	Sepsis with positive blood culture	Code this factor if the patient has a positive blood culture within 48 hours of OR Entry Date and Time, combined with the diagnosis of sepsis. Sepsis ROOT Definition = Sepsis is defined as "evidence of serious infection accompanied by a deleterious systemic response". Sepsis may be diagnosed by the presence of a Systemic Inflammatory Response Syndrome (SIRS) resulting from suspected or proven infection. A systemic inflammatory response syndrome (SIRS) is present when at least two of the following criteria are present: hypo- or hyperthermia (>38.5 or <36.0), tachycardia or bradycardia, tachypnea, leukocytosis or leukopenia, and thrombocytopenia. Code this factor if the patient has signs of sepsis and a positive blood culture within 48 hours of OR Entry Date and Time.
400	Preoperative neurological deficit	Code this factor if the patient has any deficit of neurologic function identified by the care team (during the hospitalization of this operation prior to the time of OR Entry Date and Time).
410	Seizure during lifetime	Seizure ROOT Definition = A seizure is defined as the clinical and/or electroencephalographic recognition of epileptiform activity. Select this preoperative factor for any prior seizure during the lifetime of the patient.
420	Seizure within 48 hours prior to surgery	Seizure ROOT Definition = A seizure is defined as the clinical and/or electroencephalographic recognition of epileptiform activity. Select this preoperative factor for any prior seizure during the 48 hours prior to surgery.
430	Stroke, CVA, or Intracranial hemorrhage > Grade 2 during lifetime	Indicate whether the patient had a stroke, CVA, or intracranial hemorrhage > Grade 2 at any time during the patient's lifetime. Stroke ROOT Definition = A stroke is any confirmed neurological deficit of abrupt onset caused by a disturbance in blood flow to the brain, when the neurologic deficit does not resolve within 24 hours. An IVH (Intraventricular hemorrhage) is diagnosed by the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that may extend to include an intraparenchymal component. A Grade 1 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage with a limited germinal matrix involvement. A Grade 2 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that involves an area of up to, but not more than 50% of the

430		ventricular cross-sectional area in sagittal view. A Grade 3 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that involves at least 50% of the ventricular cross-sectional area in sagittal view but not an intraparenchymal component. A Grade 4 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that includes an intraparenchymal component extending beyond the germinal matrix.
440	Stroke, CVA, or Intracranial hemorrhage > Grade 2 within 48 hours prior to surgery	Indicate whether the patient had a stroke, CVA, or intracranial hemorrhage > Grade 2 occurring within the 48 hours prior to surgery. Stroke ROOT Definition = A stroke is any confirmed neurological deficit of abrupt onset caused by a disturbance in blood flow to the brain, when the neurologic deficit does not resolve within 24 hours. An IVH (Intraventricular hemorrhage) is diagnosed by the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that may extend to include an intraparenchymal component. A Grade 1 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage with a limited germinal matrix involvement. A Grade 2 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that involves an area of up to, but not more than 50% of the ventricular cross-sectional area in sagittal view. A Grade 3 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that involves at least 50% of the ventricular cross-sectional area in sagittal view but not an intraparenchymal component. A Grade 4 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that includes an intraparenchymal component extending beyond the germinal matrix.
450	Renal dysfunction	Renal dysfunction is defined as the oliguria with sustained urine output < 0.5 cc/kg/hr for 24 hours and/or a rise in creatinine > 1.5 times upper limits of normal for age, without needing dialysis (including peritoneal dialysis and/or hemodialysis) or hemofiltration.
460	Renal failure requiring dialysis	Renal failure is defined as oliguria with sustained urine output < 0.5 cc/kg/hr for 24 hours and/or a rise in creatinine > 1.5 times upper limits of normal for age, with need for dialysis (including peritoneal dialysis and/or hemodialysis) or hemofiltration.
470	Invasive mechanical ventilation to treat cardiorespiratory failure	This patient was supported with mechanical ventilation to treat cardiorespiratory failure during the hospitalization of this operation and prior to OR Entry Date and Time.
600	Non-invasive mechanical ventilation to treat cardiorespiratory failure	
480	Respiratory Syncytial Virus	Code this factor if the patient is diagnosed with Respiratory Syncytial Virus (RSV) during the hospitalization of this operation prior to the time of OR Entry Date and time and

480		was present at the time of OR Entry Date and Time.
490	Single lung	Code this factor if the patient has only one lung present at the time of OR Entry Date and Time.
500	Tracheostomy present	Code this factor if the patient has a tracheostomy present at the time of OR Entry Date and Time.
510	Asthma	Asthma is the common chronic inflammatory disease of the airways characterized by variable and recurring symptoms, reversible airflow obstruction, and bronchospasm. Symptoms include wheezing, coughing, chest tightness, and shortness of breath. Asthma is clinically classified according to the frequency of symptoms, forced expiratory volume in 1 second (FEV1), and peak expiratory flow rate. Asthma may also be classified as atopic (extrinsic) or non-atopic (intrinsic). It is thought to be caused by a combination of genetic and environmental factors. Treatment of acute symptoms is usually with an inhaled short-acting beta-2 agonist (such as salbutamol). Symptoms can be prevented by avoiding triggers, such as allergens and irritants, and by inhaled corticosteroids.
520	Bronchopulmonary dysplasia (BPD)	Bronchopulmonary dysplasia (BPD) is a chronic lung disorder that is most common among children who were born prematurely, with low birth weights and who received prolonged mechanical ventilation to treat respiratory distress syndrome. BPD is characterized by inflammation and scarring in the lungs. The high pressures of oxygen delivery result in necrotizing bronchiolitis and alveolar septal injury, further compromising oxygenation of blood. Today, with the advent of surfactant therapy and high frequency nasal ventilation and oxygen supplementation, infants with BPD experience much milder injury without necrotizing bronchiolitis or alveolar septal fibrosis. It develops most commonly in the first 4 weeks after birth.
530	ICD (AICD) ([automatic] implantable cardioverter defibrillator) present	An implantable cardioverter-defibrillator (ICD) is a small battery-powered electrical impulse generator that is implanted in patients who are at risk of sudden cardiac death due to ventricular fibrillation and ventricular tachycardia. The device is programmed to detect cardiac arrhythmia and correct it by delivering a jolt of electricity. In current models, the ability to convert tachyarrhythmias has been extended to include both atrial and ventricular arrhythmias. There also exists the ability to perform biventricular pacing for asystole or bradycardia.
540	Pacemaker present	A pacemaker is a medical device that uses electrical impulses, delivered by electrodes contacting the heart muscles, to regulate the beating of the heart. The purpose of a pacemaker is to maintain an adequate heart rate, either because the heart's native pacemaker is not fast enough, or there is a block in the heart's electrical conduction system. Pacemakers are externally programmable and allow the physician to select the optimum pacing modes for individual patients. Some have multiple electrodes stimulating differing positions within the heart to improve synchronization of the upper (atria) and lower (ventricles) chambers of the heart.
570	Tobacco use	Current or previous use of any tobacco product, including

570		cigarettes, cigars, pipes, and chewing tobacco.
610	Transferred from another hospital after undergoing cardiac surgical operation at that hospital during this episode of care	
620	Admitted from home after undergone a cardiac surgical operation within the past 30 days	
777	Other preoperative factors	This patient has other preoperative factor(s) that are not on this list.

Preop PHTN Seq Num: 9005

Required for case closure: Yes

Registry field: [RiskSurg].[RSphtnPre]

Description: Was the patient on treatment for pulmonary hypertension (PHTN) at the time of this surgery? Indicate Yes, if the patient was receiving inhaled, subcutaneous, IV, or oral therapy for PHTN. Exclude patients on therapy for other reasons, such as PLE.
*
Patients on home oxygen therapy for pulmonary hypertension should not be coded as having preop PHTN. The patient must be receiving continuous therapy as described above, and/or enteral meds for PHTN.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Preop PLE Seq Num: 9006

Required for case closure: Yes

Registry field: [RiskSurg].[RSplePre]

Description: Did the patient have a preoperative diagnosis of protein losing enteropathy (PLE)?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Preop chronic lung disease of prematurity Seq Num: 9007

Required for case closure: Yes

Registry field: [RiskSurg].[RScldPre]

Description: Indicate Yes, if patient was born preterm and carries a diagnosis of chronic lung disease or bronchopulmonary dysplasia requiring home oxygen therapy which is persistent at the time of surgery.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Preop arrhythmia Seq Num: 9008

Required for case closure: Yes

Registry field: [RiskSurg].[RSarrhythmiaPre]

Description: Did the patient have a preop arrhythmia? Indicate Yes, if the patient had a known diagnosis of arrhythmia and was receiving temporary pacing or medication (IV or enteral) for arrhythmia at the time of surgery.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Preop creatinine available Seq Num: 9020

Required for case closure: Yes

Registry field: [RiskSurg].[RSCrPreKnown]

Description: Is a preoperative creatinine available within 30 days of this surgery?

Values	*Code*	*Text*
	1	Yes
	0	No

Preop creatinine (mg/dL) Seq Num: 9030

Required for case closure: Yes

Registry field: [RiskSurg].[RSCrPre]

Description: If preop creatinine is available, record the value closest in time to this surgery.

Any ECMO prior to surgery

Seq Num: 9050

Required for case closure: Yes

Registry field: [RiskSurg].[RSECMOpre]

Description: Prior to this surgery, was the patient on ECMO at any time during this hospitalization? This includes ECMO at time of OR entry.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Any CPR prior to surgery

Seq Num: 9150

Required for case closure: Yes

Registry field: [RiskSurg].[RSCPRpre]

Description: Prior to this surgery, did the patient undergo CPR at any time during this hospitalization?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Preop viral respiratory infection

Seq Num: 9170

Required for case closure: Yes

Registry field: [RiskSurg].[RSViralRespPre]

Description: Record Yes if patient had a documented viral respiratory infection at any time during the hospitalization prior to surgery. This could be a clinical diagnosis or confirmed by a PCR test.
*

Code as Yes if a patient has a positive respiratory viral panel at an outside hospital and is directly admitted to the CICU and goes to surgery.

Also code yes if a patient tested positive for RSV at the preop clinic visit prior to admission for surgery, even if the patient is asymptomatic.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Arrest during surgery Seq Num: 9180

Required for case closure: Yes

Registry field: [RiskSurg].[RSarrestIntra]

Description: During this operation, did the patient have a cardiac arrest requiring at least 10 minutes of CPR ?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Left OR with open sternum Seq Num: 9210

Required for case closure: Yes

Registry field: [RiskSurg].[RSOpenChest]

Description: Was the patient admitted to the CICU from the operating room with an open sternum (with or without skin closure).

*

This field is referring to a very specific point in time: Was the patient's sternum open (whether planned or unplanned) when they arrived to the ICU following the first CPB/No CPB Cardiovascular surgery for this ICU encounter (i.e., the surgery selected for the Surgical Risk section.)

If you code "left the OR with an open chest" as Yes, you must answer Yes to the complication 'Sternum left open' (#7280) as well. That field captures patients whose chests were left open at any time in the ICU.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Postop lactate available (first 2 hrs postop) Seq Num: 9260

Required for case closure: Yes

Registry field: [RiskSurg].[RSLactKnown]

Description: Is a lactate available during the first 2 postop hours in the CICU?

*

For patients who required a cardiac reoperation before the 2 hours postop, code this and subsequent surgical risk fields still pertain to the first surgery, and are based on the time period from the initial postop arrival through the return to the operating room.

Values	*Code*	*Text*
	1	Yes
	0	No

Max postop lactate (mmol/L)

Seq Num: 9270

Required for case closure: Yes

Registry field: [RiskSurg].[RSLact]

Description: If it is available, record the highest lactate, in mmol/L, during the first 2 postop hours in the CICU

2 hour postop chest tube output (cc)

Seq Num: 9320

Required for case closure: Yes

Registry field: [RiskSurg].[RSChestOutput]

Description: Record the total output from all chest tubes, in cc, within the first 2 postop hours in the CICU. This should not include output from the operating room or transfer to the CICU.

*

Do not include fluid accumulated in the OR or on transfer from the OR (which may appear as the initial chest tube output on arrival).

Mech vent at 2 hours postop

Seq Num: 9420

Required for case closure: Yes

Registry field: [RiskSurg].[RSintub]

Description: Was the patient mechanically ventilated at 2 hours postop in the CICU?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

FiO2 available

Seq Num: 9440

Required for case closure: Yes

Registry field: [RiskSurg].[RSFIO2known]

Description: If the patient was ventilated, is the FiO2 at 2 hours postop in the CICU available?

Values	*Code*	*Text*
	1	Yes
	0	No

FiO2 Seq Num: 9450

Required for case closure: Yes

Registry field: [RiskSurg].[RSFIO2]

Description: If it is available, record the FiO2 closest to 2 hours postop in the CICU

*

The valid range for the FiO is between 0.00 and 1.00.

Ex: For a patient with postop FiO of 40%, enter 0.40.

Postop mean airway pressure available Seq Num: 9460

Required for case closure: Yes

Registry field: [RiskSurg].[RSAirPressKnown]

Description: If the patient was ventilated, is the mean airway pressure (MAP) available at 2 hours postop in the CICU?

Values	*Code*	*Text*
	1	Yes
	0	No

Postop mean airway pressure Seq Num: 9465

Required for case closure: Yes

Registry field: [RiskSurg].[RSAirPress]

Description: If it is available, record the mean airway pressure closest to 2 hours postop in the CICU

POD0 or POD1 Cr available Seq Num: 9520

Required for case closure: Yes

Registry field: [RiskSurg].[RSCrAdmitAvail]

Description: Were any postop creatinines recorded in the CICU on postop day 0 or postop day 1?

Values	*Code*	*Text*
	1	Yes
	0	No

First postop Cr (mg/dL) Seq Num: 9522

Required for case closure: Yes

Registry field: [RiskSurg].[RSCrAdmitValue]

Description: Record the first postop creatinine in the CICU, in mg/dL. This must be from POD0 or POD1

Any CICU postop Cr (through POD7)

Seq Num: 9524

Required for case closure: Yes

Registry field: [RiskSurg].[RSCr7avail]

Description: Were any postop creatinines recorded in the CICU through postop day 7?

Values	*Code*	*Text*
	1	Yes
	0	No

Max postop Cr (mg/dL)

Seq Num: 9526

Required for case closure: Yes

Registry field: [RiskSurg].[RSCr7value]

Description: Maximum postop creatinine, in mg/dL, recorded in the CICU through POD7.

*

If the patient's maximum postop creatinine was immediately after surgery (#9522), record that value in this question as well.

Max postop Cr date

Seq Num: 9528

Required for case closure: Yes

Registry field: [RiskSurg].[RSCr7date]

Description: Date on which the maximum postop Cr was recorded in the CICU. If this maximum value was recorded more than once, use the earliest date.

*

If the patient's maximum postop creatinine was immediately after surgery (#9522), record that value in this question as well.

This field is intended to capture the maximum postop creatinine through POD7 or ICU discharge, whichever is earlier.

ECMO initiated in OR

Seq Num: 9190

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskSurg].[RSECMOintra]

Description: Was ECMO initiated in the OR during this surgery?

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Postop core temp available Seq Num: 9230

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskSurg].[RSTempKnown]

Description: Is a core temperature (rectal, bladder, esophageal or intracardiac) available during the first 2 postop hours in the CICU?

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Postop core temp (Celsius) Seq Num: 9240

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskSurg].[RSTemp]

Description: If it is available, record the highest core temperature, in Celsius, during the first 2 postop hours in the CICU

Postop systolic BP Seq Num: 9480

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskSurg].[RSSBP]

Description: Find the time during the first 2 postop hours in the CICU where the difference between the systolic and diastolic blood pressure is smallest. Record the systolic BP, in mmHg, at that time.

Postop diastolic BP Seq Num: 9490

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskSurg].[RSDBP]

Description: Find the time during the first 2 postop hours in the CICU where the difference between the systolic and diastolic blood pressure is smallest. Record the diastolic BP, in mmHg, at that time.

Surgical encounters - Neonatal feeding

First surgical encounter

Seq Num: 9322

Required for case closure: Yes

Registry field: [RiskSurg].[RSfirstNeonate]

Description: Is this the patient's first surgical encounter (i.e., the first encounter during which the patient underwent CPB or No CPB Cardiovascular surgery) during this hospitalization?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Any preop enteral feeding

Seq Num: 9324

Required for case closure: Yes

Registry field: [RiskSurg].[RSfeedPreop]

Description: During this hospital admission, did the patient receive any enteral feeds (oral or tube) prior to this surgery? This includes feeding prior to or during this CICU encounter.
*
Anything caloric that passes into the gut counts as enteral feeding, including oral intake, tube feeding, and trophic feeding.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Preop feed in CICU

Seq Num: 9326

Required for case closure: Yes

Registry field: [RiskSurg].[RSfeedPreopCICU]

Description: During this hospitalization, did the patient receive any preop enteral feeds in the CICU?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Preop feed in NICU

Seq Num: 9328

Required for case closure: Yes

Registry field: [RiskSurg].[RSfeedPreopNICU]

Description: During this hospitalization, did the patient receive any preop enteral feeds in the NICU?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Preop feed in another location

Seq Num: 9330

Required for case closure: Yes

Registry field: [RiskSurg].[RSfeedPreopOther]

Description: During this hospitalization, did the patient receive any preop enteral feeds in any other inpatient location?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Any postop enteral feeding in the CICU

Seq Num: 9332

Required for case closure: Yes

Registry field: [RiskSurg].[RSfeedPostop]

Description: During this encounter, did the patient receive any postoperative enteral feeds (oral or tube) prior to CICU discharge?

*

Anything caloric that passes into the gut counts as enteral feeding, including oral intake, tube feeding, and trophic feeding.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Earliest postop feed date

Seq Num: 9334

Required for case closure: Yes

Registry field: [RiskSurg].[RSfeedPostopDt]

Description: What was the earliest date the patient received any postoperative enteral feeds prior to CICU discharge.

Nutrition at CICU discharge

Seq Num: 9336

Required for case closure: Yes

Registry field: [RiskSurg].[RSfeedDisch]

Description: At the time of CICU discharge, how was the child fed?

*

For patients not receiving any feedings, please code as 'Unknown.' (An option for 'None' will be added to version 4.)

Values	*Code*	*Text*
	1	Enteral
	2	TPN
	3	Both
	9	Unk

Inotropic/vasopressor infusion at time of surgery

Seq Num: 9070

Required for case closure: Yes

Registry field: [RiskSurg].[RSVISpre]

Description: Immediately prior to transfer to the operating room, was the patient on an infusion of any of the following inotropes or vasopressors: dopamine, dobutamine, epinephrine, norepinephrine, milrinone, vasopressin?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Dopamine (mcg/kg/min) at surgery

Seq Num: 9080

Required for case closure: Yes

Registry field: [RiskSurg].[RSDopaPre]

Description: Record the dopamine dose (mcg/kg/min) at time of surgery; if the patient was not on dopamine, enter 0.

Dobutamine (mcg/kg/min) at surgery

Seq Num: 9090

Required for case closure: Yes

Registry field: [RiskSurg].[RSDobutPre]

Description: Record the dobutamine dose (mcg/kg/min) at time of surgery; if the patient was not on dobutamine, enter 0

Epinephrine (mcg/kg/min) at surgery

Seq Num: 9100

Required for case closure: Yes

Registry field: [RiskSurg].[RSEpiPre]

Description: Record the epinephrine dose (mcg/kg/min) at time of surgery; if the patient was not on epinephrine, enter 0.

Norepinephrine (mcg/kg/min) at surgery

Seq Num: 9110

Required for case closure: Yes

Registry field: [RiskSurg].[RSNorepiPre]

Description: Record the norepinephrine dose (mcg/kg/min) at time of surgery; if the patient was not on norepinephrine, enter 0.

Milrinone (mcg/kg/min) at surgery

Seq Num: 9120

Required for case closure: Yes

Registry field: [RiskSurg].[RSMilrinPre]

Description: Record the milrinone dose (mcg/kg/min) at time of surgery; if the patient was not on milrinone, enter 0

Vasopressin (units/kg/min) at surgery

Seq Num: 9130

Required for case closure: Yes

Registry field: [RiskSurg].[RSVasopressPre]

Description: Record the vasopressin dose (units/kg/min) at time of surgery; if the patient was not on vasopressin, enter 0. Please note, this must be recorded in units/kg/min.

Inotropic/vasopressor infusion in first 2 postop hrs

Seq Num: 9340

Required for case closure: Yes

Registry field: [RiskSurg].[RSVIS]

Description: Was the patient on an infusion of any of the following within the first 2 postop hours in the CICU: dopamine, dobutamine, epinephrine, norepinephrine, milrinone, vasopressin? If yes, find the time where the vasoactive inotrope score (VIS) is highest and record the doses at that time.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Postop dopamine (mcg/kg/min)

Seq Num: 9350

Required for case closure: Yes

Registry field: [RiskSurg].[RSDopa]

Description: Record the dopamine dose (mcg/kg/min) at the time of maximum VIS; if the patient was not on dopamine, enter 0

*

Looking only at the first 2 postop hours in the CICU, find the time when the vasoactive inotrope score (VIS) is highest and record the dose at that time.

Postop dobutamine (mcg/kg/min) Seq Num: 9360

Required for case closure: Yes

Registry field: [RiskSurg].[RSDobut]

Description: Record the dobutamine dose (mcg/kg/min) at the time of maximum VIS; if the patient was not on dobutamine, enter 0

*

Looking only at the first 2 postop hours in the CICU, find the time when the vasoactive inotrope score (VIS) is highest and record the dose at that time.

Postop epinephrine (mcg/kg/min) Seq Num: 9370

Required for case closure: Yes

Registry field: [RiskSurg].[RSEpi]

Description: Record the epinephrine dose (mcg/kg/min) at the time of maximum VIS; if the patient was not on epinephrine, enter 0

*

Looking only at the first 2 postop hours in the CICU, find the time when the vasoactive inotrope score (VIS) is highest and record the dose at that time.

Postop norepinephrine (mcg/kg/min) Seq Num: 9380

Required for case closure: Yes

Registry field: [RiskSurg].[RSNorepi]

Description: Record the norepinephrine dose (mcg/kg/min) at the time of maximum VIS; if the patient was not on norepinephrine, enter 0

*

Looking only at the first 2 postop hours in the CICU, find the time when the vasoactive inotrope score (VIS) is highest and record the dose at that time.

Postop milrinone (mcg/kg/min) Seq Num: 9390

Required for case closure: Yes

Registry field: [RiskSurg].[RSMilrin]

Description: Record the milrinone dose (mcg/kg/min) at the time of maximum VIS; if the patient was not on milrinone, enter 0

*

Looking only at the first 2 postop hours in the CICU, find the time when the vasoactive inotrope score (VIS) is highest and record the dose at that time.

Postop vasopressin (units/kg/min)

Seq Num: 9400

Required for case closure: Yes

Registry field: [RiskSurg].[RSVasopress]

Description: Record the vasopressin dose (units/kg/min) at the time of maximum VIS; if the patient was not on vasopressin, enter 0.

Please note, this must be recorded in units/kg/min.

*

Looking only at the first 2 postop hours in the CICU, find the time when the vasoactive inotrope score (VIS) is highest and record the dose at that time.

Postop VIS timepoint

Seq Num: 9502

Required for case closure: Yes

Registry field: [RiskSurgVIS].[RSVISpoint]

Description: Record the inotropic/vasopressor support information for each timepoint listed. Only record data for the timepoints prior to CICU discharge.

Values	*Code*	*Text*
	6	6 hrs postop
	12	12 hrs postop
	18	18 hrs postop
	24	24 hrs postop
	30	30 hrs postop
	36	36 hrs postop
	42	42 hrs postop
	48	48 hrs postop
	72	06:00 on POD3
	96	06:00 on POD4
	120	06:00 on POD5
	144	06:00 on POD6
	168	06:00 on POD7

Postop VIS date/time

Seq Num: 9504

Required for case closure: Yes

Registry field: [RiskSurgVIS].[RSVISdttm]

Description: Postop VIS date/time

On support at this postop timepoint Seq Num: 9506

Required for case closure: Yes

Registry field: [RiskSurgVIS].[RSVISsupport]

Description: Was the patient receiving an infusion of any of the 6 listed agents at this timepoint. If the patient is not on any of these infusions at this specific time, answer No.
*
If the patient is back in surgery during one of the VIS timepoints (e.g., 6hrs post-initial op, 12hrs post, etc.), please code 'On support at this timepoint' as No.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Dopamine (mcg/kg/min) at this postop timepoint Seq Num: 9508

Required for case closure: Yes

Registry field: [RiskSurgVIS].[RSVISdopa]

Description: Record the dopamine dose (mcg/kg/min) at this timepoint; if the patient was not on dopamine, enter 0

Dobutamine (mcg/kg/min) at this postop timepoint Seq Num: 9510

Required for case closure: Yes

Registry field: [RiskSurgVIS].[RSVISdobut]

Description: Record the dobutamine dose (mcg/kg/min) at this timepoint; if the patient was not on dobutamine, enter 0

Epinephrine (mcg/kg/min) at this postop timepoint Seq Num: 9512

Required for case closure: Yes

Registry field: [RiskSurgVIS].[RSVISepi]

Description: Record the epinephrine dose (mcg/kg/min) at this timepoint; if the patient was not on epinephrine, enter 0

Norepinephrine (mcg/kg/min) at this postop timepoint Seq Num: 9514

Required for case closure: Yes

Registry field: [RiskSurgVIS].[RSVISnorepi]

Description: Record the norepinephrine dose (mcg/kg/min) at this timepoint; if the patient was not on norepinephrine, enter 0

Milrinone (mcg/kg/min) at this postop timepoint Seq Num: 9516

Required for case closure: Yes

Registry field: [RiskSurgVIS].[RSVISmilrin]

Description: Record the milrinone dose (mcg/kg/min) at this timepoint; if the patient was not on milrinone, enter 0

Vasopressin (units/kg/min) at this postop timepoint Seq Num: 9518

Required for case closure: Yes

Registry field: [RiskSurgVIS].[RSVISvasopress]

Description: Record the vasopressin dose (units/kg/min) at this timepoint; if the patient was not on vasopressin, enter 0. Please note, this must be recorded in units/kg/min.

Non-surgical encounters

High-risk diagnoses on admission

Seq Num: 9700

Required for case closure: Yes

Registry field: [RiskMedDiag].[RiskMedDiag]

Description: Select all conditions that were present at time of CICU admission. If none of them were present, select "None."

* Additional clarifications are below between asterisks. *

Values	Code	Text	
	1	None	
	5	Arrhythmia	Arrhythmia requiring ICU-level therapy * This also applies to arrhythmia present within 1 hour of admission. If an arrythmia occurs that requires ICU-level therapy prior to the CICU admission, whether in an outside hospital, ED, or inpatient unit, it can be captured as a high risk diagnosis on admission even if the therapy did not continue into the ICU, if the arrhythmia was part of the reason for the CICU admission. Both bolus and continuous infusions qualify as ICU-level therapy, with the exception of Digoxin and adenosine used exclusively to diagnose and arrhythmia. Lidocaine is considered ICU-level therapy. *
	2	Cardiomyopathy	
	6	CPR reason for CICU admit	This could be a resolved cardiopulmonary arrest or a patient receiving active compressions. Patients receiving active compressions at the time of admission should also have the arrest recorded in the Complications section. * This is intended to flag patients whose primary reason for CICU admission is an arrest. This is not limited to patients receiving compressions on admission. For example, a teen who arrested playing basketball who is treated by EMTs and then brought to the unit would also be coded with this diagnosis. *
	9	Heart failure, acute decompensated	Systolic or diastolic cardiac dysfunction that requires at least one of the following therapies: 1) continuous infusion of a vasoactive agent or diuretic agent, 2) respiratory support (HFNC, CPAP/BiPAP, or mechanical ventilation), 3) Mechanical circulatory support.
	10	Heart failure, chronic	Pre-admission diagnosis of heart failure requiring medications or VAD support prior to admission.
	11	Heart transplant rejection	Treatment with anti-rejection medical therapy, or biopsy or explant documented heart transplant rejection (before or after therapy). If patient has acute decompensated heart failure due to rejection, use this diagnosis not acute decompensated heart failure.
	3	Myocarditis	New diagnosis of cardiac dysfunction and/or rhythm

	3		disturbance suspected to be secondary to acute myocarditis requiring at least one of the following therapies: (1) continuous infusion of a vasoactive agent; (2) respiratory support (HFNC, CPAP/BiPAP, mechanical ventilation); (3) mechanical circulatory support. * Myocarditis that does not meet the criteria above should NOT be coded here. (You could code the CICU CT diagnosis as myocarditis, and the medical diagnosis could be Other - myocarditis.) *
	7	Pulmonary hypertension	Initiation of pulmonary antihypertensive therapy or management of pulmonary hypertension in a patient currently being treated with medical (e.g., NO, sildenafil, bosentan) therapy.
	4	Systemic AVVR - moderate or worse	Moderate or worse systemic AV valve regurgitation confirmed at or soon after CICU admission
Retired	8	Hospice care and/or DNR	DNR/DNI order at admission or created within 1 hour

BNP available (18hr window)

Seq Num: 9761

Required for case closure: Yes

Registry field: [RiskMed].[RMBNPavail]

Description: Is a BNP or NT-proBNP available from 12 hours pre-CICU admission through 6 hours post-CICU admission?

Values	*Code*	*Text*
	1	Yes
	0	No

BNP type

Seq Num: 9762

Required for case closure: Yes

Registry field: [RiskMed].[RMBNPtype]

Description: Record the BNP type

Values	*Code*	*Text*
	1	BNP
	2	NT-proBNP
	9	Unk

Max BNP (pg/mL)

Seq Num: 9763

Required for case closure: Yes

Registry field: [RiskMed].[RMBNPvalue]

Description: If it is available, record the highest BNP, in pg/mL, from 12 hours pre-CICU admission through 6 hours post-CICU admission.

Creatinine available (18hr window) Seq Num: 9791

Required for case closure: Yes

Registry field: [RiskMed].[RMCrAvail]

Description: Is a creatinine available from 12 hours pre-CICU admission through 6 hours post-CICU admission?

Values	*Code*	*Text*
	1	Yes
	0	No

First creatinine (mg/dL) Seq Num: 9794

Required for case closure: Yes

Registry field: [RiskMed].[RMCrFirst]

Description: If it is available, record the earliest creatinine value, in mg/dL, from 12 hours pre-CICU admission through 6 hours post-CICU admission.

Max creatinine (mg/dL) Seq Num: 9792

Required for case closure: Yes

Registry field: [RiskMed].[RMCrValue]

Description: If it is available, record the highest creatinine value, in mg/dL, from 12 hours pre-CICU admission through 6 hours post-CICU admission.

Creatinine available through CICU day 7 Seq Num: 9796

Required for case closure: Yes

Registry field: [RiskMed].[RMCr7avail]

Description: Were any creatinines recorded in the CICU through day 7?

*

If there are any creatinine values from CICU admission through CICU discharge (for patients in the ICU<7d) or through day 7 (for patients in the ICU 7+ days), you must answer these questions – even if the only creatinine was shortly after ICU admission.

Values	*Code*	*Text*
	1	Yes
	0	No

Max CICU creatinine (mg/dL) Seq Num: 9798

Required for case closure: Yes

Registry field: [RiskMed].[RMCr7value]

Description: Record the highest creatinine value, in mg/dL, through CICU day 7
*
This may be the same value entered in #9792.

Max Cr date Seq Num: 9801

Required for case closure: Yes

Registry field: [RiskMed].[RMCr7Date]

Description: Date on which the maximum Cr was recorded in the CICU. If this maximum value was recorded more than once, use the earliest date.
*
This field is intended to capture the maximum creatinine in the ICU through day 7 or ICU discharge, whichever is earlier.

Hepatic injury (18hr window) Seq Num: 9821

Required for case closure: Yes

Registry field: [RiskMed].[RMHepInj]

Description: Did the patient have transaminitis (AST or ALT >500) from 12 hours pre-CICU admission through 6 hours post-CICU admission?

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Lactate available (4hr window) Seq Num: 9840

Required for case closure: Yes

Registry field: [RiskMed].[RMLactKnown]

Description: Is a lactate available within 2 hours (plus or minus) of CICU admission?

Values	*Code*	*Text*
	1	Yes
	0	No

Max lactate (mmol/L)

Seq Num: 9850

Required for case closure: Yes

Registry field: [RiskMed].[RMLact]

Description: If it is available, record the highest lactate, in mmol/L, within 2 hours (plus or minus) of CICU admission

Pupil reflex on admission

Seq Num: 9901

Required for case closure: Yes

Registry field: [RiskMed].[RMPupilAdmit]

Description: Record the pupil reflex on admission to the CICU

Values	Code	Text
	1	Both reactive
	2	One fixed/one reactive
	3	Both fixed
	9	Unk

Any prior cardiothoracic surgery

Seq Num: 9640

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMPrevSurg]

Description: Prior to this CICU admission, did the patient ever have any cardiothoracic (heart or great vessels) surgical procedures with or without cardiopulmonary bypass (CPB). Also include lung procedures utilizing CPB or tracheal procedures utilizing CPB.

Values	Code	Text
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Current surgical status

Seq Num: 9660

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMStatusPost]

Description: If the patient had prior cardiothoracic surgery, select the category that represents the patient's current status.

Values	Code	Text	
Retired	1	S/P stage I palliation - Norwood	Status post Norwood stage I palliation
Retired	2	S/P stage I palliation - Hybrid	Status post hybrid stage I palliation
Retired	3	S/P stage II palliation	Status post stage II palliation (bi-directional Glenn, hemi-Fontan or Kawashima procedure)
Retired	4	S/P stage III palliation	Status post stage III palliation (fenestrated or non-fenestrated Fontan procedure)
Retired	5	S/P aortopulmonary shunt	Status post aortopulmonary shunt (including MBTS, RVPAS or central shunt) for 1V or 2V palliation
Retired	6	S/P other 1V surgery	Patient with single ventricle anatomy status post other surgery
Retired	7	S/P 2V surgery	Patient with two ventricle anatomy status post other palliative or reparative surgery
Retired	8	S/P thoracic surg (never had cardiac surg)	Patient never had cardiac surgery; status post thoracic surgery, including tracheal reconstruction, with or without CPB.

BNP available

Seq Num: 9760

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMBNPknown]

Description: Is a BNP available within 2 hours (plus or minus) of CICU admission

Values	Code	Text
Retired	1	Yes
Retired	0	No

BNP (pg/mL)

Seq Num: 9770

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMBNP]

Description: If it is available, record the highest BNP, in pg/mL, within 2 hours (plus or minus) of CICU admission

Creatinine available Seq Num: 9790

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMCrKnown]

Description: Is a creatinine available within 2 hours (plus or minus) of CICU admission?

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No

Creatinine (mg/dL) Seq Num: 9800

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMCr]

Description: If it is available, record the highest creatinine value within 2 hours (plus or minus) of CICU admission

Hepatic injury Seq Num: 9820

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMHepFail]

Description: Did the patient have transaminitis (AST or ALT >500) within 2 hours (plus or minus) of CICU admission.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Pupil reflex at 2 hrs Seq Num: 9900

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMPupil]

Description: Record the pupil reflex at 2 hours post CICU admission

Values	*Code*	*Text*
Retired	1	Both reactive
Retired	2	One fixed/one reactive
Retired	3	Both fixed
Retired	9	Unk

Inotropic/vasopressor infusion Seq Num: 9920

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMVIS]

Description: Was the patient on an infusion of any of the following during the first 2 hours of CICU admission: dopamine, dobutamine, epinephrine, norepinephrine, milrinone, vasopressin? If yes, find the time where the vasoactive inotrope score (VIS) is highest and record the doses at that time.

Values	*Code*	*Text*
Retired	1	Yes
Retired	0	No
Retired	9	Unk

Dopamine (mcg/kg/min) Seq Num: 9930

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMDopa]

Description: Record the dopamine dose (mcg/kg/min) at the time of maximum VIS; if the patient was not on dopamine, enter 0.

Dobutamine (mcg/kg/min) Seq Num: 9940

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMDobut]

Description: Record the dobutamine dose (mcg/kg/min) at the time of maximum VIS; if the patient was not on dobutamine, enter 0.

Epinephrine (mcg/kg/min) Seq Num: 9950

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMEpi]

Description: Record the epinephrine dose (mcg/kg/min) at the time of maximum VIS; if the patient was not on epinephrine, enter 0.

Norepinephrine (mcg/kg/min) Seq Num: 9960

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMNorepi]

Description: Record the norepinephrine dose (mcg/kg/min) at the time of maximum VIS; if the patient was not on norepinephrine, enter 0.

Milrinone (mcg/kg/min) Seq Num: 9970

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMMilrin]

Description: Record the milrinone dose (mcg/kg/min) at the time of maximum VIS; if the patient was not on milrinone, enter 0.

Vasopressin (units/kg/min) Seq Num: 9980

Retired in version 2.0

Required for case closure: Yes

Registry field: [RiskMed].[RMVasopress]

Description: Record the vasopressin dose (units/kg/min) at time of maximum VIS; if the patient was not on vasopressin, enter 0. Please note, this must be recorded in units/kg/min.

Non-surgical encounters - VIS

Inotropic/vasopressor infusion on admission

Seq Num: 9921

Required for case closure: Yes

Registry field: [RiskMed].[RMVISadmit]

Description: At the time of CICU admission, was the patient on an infusion of any of the following: dopamine, dobutamine, epinephrine, norepinephrine, milrinone, vasopressin? If yes, record the doses of each of these infusions at the time of CICU admission.

Values	Code	Text
	1	Yes
	0	No
	9	Unk

Dopamine (mcg/kg/min) on admission

Seq Num: 9931

Required for case closure: Yes

Registry field: [RiskMed].[RMDopaAdmit]

Description: Record the dopamine dose (mcg/kg/min) at the time of CICU admission; if the patient was not on dopamine, enter 0.

Dobutamine (mcg/kg/min) on admission

Seq Num: 9941

Required for case closure: Yes

Registry field: [RiskMed].[RMDobutAdmit]

Description: Record the dobutamine dose (mcg/kg/min) at the time of CICU admission; if the patient was not on dobutamine, enter 0.

Epinephrine (mcg/kg/min) on admission

Seq Num: 9951

Required for case closure: Yes

Registry field: [RiskMed].[RMEpiAdmit]

Description: Record the epinephrine dose (mcg/kg/min) at the time of CICU admission; if the patient was not on epinephrine, enter 0.

Norepinephrine (mcg/kg/min) on admission

Seq Num: 9961

Required for case closure: Yes

Registry field: [RiskMed].[RMNorepiAdmit]

Description: Record the norepinephrine dose (mcg/kg/min) at the time of CICU admission; if the patient was not on norepinephrine, enter 0.

Milrinone (mcg/kg/min) on admission Seq Num: 9971

Required for case closure: Yes

Registry field: [RiskMed].[RMMilrinAdmit]

Description: Record the milrinone dose (mcg/kg/min) at the time of CICU admission; if the patient was not on milrinone, enter 0.

Vasopressin (units/kg/min) on admission Seq Num: 9981

Required for case closure: Yes

Registry field: [RiskMed].[RMVasopressAdmit]

Description: Record the vasopressin dose (units/kg/min) at time of CICU admission; if the patient was not on vasopressin, enter 0. Please note, this must be recorded in units/kg/min.

Post-admit VIS timepoint Seq Num: 10002

Required for case closure: Yes

Registry field: [RiskMedVIS].[RMVISpoint]

Description: Record the inotropic/vasopressor support information for each post-admit timepoint listed. Only record data for the timepoints prior to CICU discharge.

Values	*Code*	*Text*
	6	6 hrs post-admit
	12	12 hrs post-admit
	18	18 hrs post-admit
	24	24 hrs post-admit
	30	30 hrs post-admit
	36	36 hrs post-admit
	42	42 hrs post-admit
	48	48 hrs post-admit
	72	06:00 on ICU day 4
	96	06:00 on ICU day 5
	120	06:00 on ICU day 6
	144	06:00 on ICU day 7

Post-admit VIS date/time Seq Num: 10004

Required for case closure: Yes

Registry field: [RiskMedVIS].[RMVISdttm]

Description: Post CICU admission VIS date/time

On support at this post-admit timepoint Seq Num: 10006

Required for case closure: Yes

Registry field: [RiskMedVIS].[RMVISsupport]

Description: Was the patient receiving an infusion of any of the 6 listed agents at this timepoint. If the patient is not on any of these infusions at this specific time, answer No.

Values	*Code*	*Text*
	1	Yes
	0	No
	9	Unk

Dopamine (mcg/kg/min) at this post-admit timepoint Seq Num: 10008

Required for case closure: Yes

Registry field: [RiskMedVIS].[RMVISdopa]

Description: Record the dopamine dose (mcg/kg/min) at this timepoint; if the patient was not on dopamine, enter 0

Dobutamine (mcg/kg/min) at this post-admit timepoint Seq Num: 10010

Required for case closure: Yes

Registry field: [RiskMedVIS].[RMVISdobut]

Description: Record the dobutamine dose (mcg/kg/min) at this timepoint; if the patient was not on dobutamine, enter 0

Epinephrine (mcg/kg/min) at this post-admit timepoint Seq Num: 10012

Required for case closure: Yes

Registry field: [RiskMedVIS].[RMVISepi]

Description: Record the epinephrine dose (mcg/kg/min) at this timepoint; if the patient was not on epinephrine, enter 0

Norepinephrine (mcg/kg/min) at this post-admit timepoint Seq Num: 10014

Required for case closure: Yes

Registry field: [RiskMedVIS].[RMVISnorepi]

Description: Record the norepinephrine dose (mcg/kg/min) at this timepoint; if the patient was not on norepinephrine, enter 0

Milrinone (mcg/kg/min) at this post-admit timepoint Seq Num: 10016

Required for case closure: Yes

Registry field: [RiskMedVIS].[RMVISmilrin]

Description: Record the milrinone dose (mcg/kg/min) at this timepoint; if the patient was not on milrinone, enter 0

Vasopressin (units/kg/min) at this post-admit timepoint Seq Num: 10018

Required for case closure: Yes

Registry field: [RiskMedVIS].[RMVISvasopress]

Description: Record the vasopressin dose (units/kg/min) at this timepoint; if the patient was not on vasopressin, enter 0. Please note, this must be recorded in units/kg/min.

Appendix: Calculating the Vasoactive-Inotropic Score (VIS)

Vasoactive-Inotropic Score (VIS)

To calculate the vasoactive-inotropic score (VIS), multiply the dose of each agent by the factor listed below then sum the results:

Agent	Units	Multiply by
Dopamine	mcg/kg/min	1
Dobutamine	mcg/kg/min	1
Epinephrine	mcg/kg/min	100
Norepinephrine	mcg/kg/min	100
Milrinone	mcg/kg/min	10
Vasopressin*	units/kg/min	10,000

*__PLEASE NOTE:__ Vasopressin must be in units/kg/min. If your MAR records it in any other way (e.g., milliunits/kg/min, units/kg/hour, etc.), you must convert it to units/kg/min.

Example:

A patient is on 5 mcg/kg/min of dopamine, 0.08 mcg/kg/min of epi, and 0.25 mcg/kg/min of milrinone. Her VIS is 15.5

	Units	Multiply by	Dose	Score
Dopamine	mcg/kg/min	1	5	5
Dobutamine	mcg/kg/min	1	0	0
Epinephrine	mcg/kg/min	100	0.08	8
Norepinephrine	mcg/kg/min	100	0	0
Milrinone	mcg/kg/min	10	0.25	2.5
Vasopressin	units/kg/min	10,000	0	0
VIS				**15.5**

The VIS reflects the total support at that point in time. The calculation should only include drugs/doses the patient is receiving concurrently.

Example:

Patient returned to the CICU from the OR on milrinone. Epinephrine was added briefly. When the epinephrine was discontinued, the milrinone dose was increased. The VIS calculations only include the drugs/doses the patient is on concurrently.

			Time 1		Time 2		Time 3	
	Units	Multiply by	Dose	Score	Dose	Score	Dose	Score
Dopamine	mcg/kg/min	1	0	0	5	5	0	0
Dobutamine	mcg/kg/min	1	0	0	0	0	0	0
Epinephrine	mcg/kg/min	100	0	0	0	0	0	0
Norepinephrine	mcg/kg/min	100	0	0	0	0	0	0
Milrinone	mcg/kg/min	10	0.5	5	0.5	5	0.7	7
Vasopressin	units/kg/min	10,000	0	0	0	0	0	0
VIS				**5**		**10**		**7**

Appendix: Modified Duke criteria for endocarditis

The following information is adapted from:

Li JS, Sexton DJ, Mick N, et al. Proposed modifications to the Duke criteria for the diagnosis of infective endocarditis. *Clin Infect Dis.* 2000;30(4):633-638.

Definite infective endocarditis

Pathologic criteria

1) Microorganisms demonstrated by culture or histologic examination of a vegetation, a vegetation that has embolized, or an intracardiac abscess specimen; or
2) Pathologic lesions; vegetation or intracardiac abscess confirmed by histologic examination showing active endocarditis

Clinical criteria*

1) 2 major criteria; or
2) 1 major criterion and 3 minor criteria; or
3) 5 minor criteria

Possible infective endocarditis

1) 1 major criterion and 1 minor criterion; or
2) 3 minor criteria

Rejected

1) Firm alternate diagnosis explaining evidence of infective endocarditis; or
2) Resolution of infective endocarditis syndrome with antibiotic therapy for **<**4 days; or
3) No pathologic evidence of infective endocarditis at surgery or autopsy, with antibiotic therapy for **<**4 days; or
4) Does not meet criteria for possible infective endocarditis, as above

* See following page for definitions of major and minor criteria

Major criteria

- Blood culture positive for IE
  - Typical microorganisms consistent with IE from 2 separate blood cultures:
    - Viridans streptococci, Streptococcus bovis, HACEK group, Staphylococcus aureus; or
    - Community-acquired enterococci, in the absence of a primary focus; or
  - Microorganisms consistent with IE from persistently positive blood cultures, defined as follows:
    - At least 2 positive cultures of blood samples drawn **1**12 h apart; or
    - All of 3 or a majority of **>**4 separate cultures of blood (with first and last sample drawn at least 1 h apart)
  - Single positive blood culture for Coxiella burnetii or antiphase I IgG antibody titer **>**1 : 800

- Evidence of endocardial involvement

- Echocardiogram positive for IE (TEE (transesophageal echocardiography) recommended in patients with prosthetic valves, rated at least "possible IE" by clinical criteria, or complicated IE [paravalvular abscess]; TTE (transthoracic echocardiography) as first test in other patients), defined as follows:
  - Oscillating intracardiac mass on valve or supporting structures, in the path of regurgitant jets, or on implanted material in the absence of an alternative anatomic explanation; or
  - Abscess; or
  - New partial dehiscence of prosthetic valve

- New valvular regurgitation (worsening or changing of pre-existing murmur not sufficient)

Minor criteria

- Predisposition, predisposing heart condition or injection drug use

- Fever, temperature >38 C

- Vascular phenomena, major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial hemorrhage, conjunctival hemorrhages, and Janeway's lesions

- Immunologic phenomena: glomerulonephritis, Osler's nodes, Roth's spots, and rheumatoid factor

- Microbiological evidence: positive blood culture but does not meet a major criterion as noted above[**] or serological evidence of active infection with organism consistent with IE

[**] Excludes single positive cultures for coagulase-negative staphylococci and organisms that do not cause endocarditis.

Appendix: Modified Bell's Staging Criteria for Necrotizing Enterocolitis (NEC)

Modified Bell's Staging Criteria for Necrotizing Enterocolitis (NEC)

Stage	Systemic signs	Intestinal signs	Radiographic signs	Treatment
I: Suspected				
A	Temperature instability, apnea, bradycardia	Elevated pregavage residuals, mild abdominal distension, occult blood in stool	Normal or mild ileus	NPO, antibiotics x 3 days
B	Same as IA	Same as IA, plus gross blood in stool	Same as IA	Same as IA
II: Definite				
A: Mildly ill	Same as IA	Same as I, plus absent bowel sounds, abdominal tenderness	Ileus, pneumatosis intestinalis	NPO, antibiotics x 7 to 10 days
B: Moderately ill	Same as IA, plus mild metabolic acidosis and thrombocytopenia	Same as I, plus absent bowel sounds, definite abdominal tenderness, abdominal cellulitis, right lower quadrant mass	Same as IIA, plus portal vein gas, with or without ascites	NPO, antibiotics x 14 days
III: Advanced				
A: Severely ill, bowel intact	Same as IIB, plus hypotension, bradycardia, respiratory acidosis, metabolic acidosis, disseminated intravascular coagulation, neutropenia	Same as I and II, plus signs of generalized peritonitis, marked tenderness and distension of abdomen	Same as IIB, plus definite ascites	NPO, antibiotics x 14 days, fluid resuscitation, inotropic support, ventilator therapy, paracentesis
B: Severely ill, bowel perforated	Same as IIIA	Same as IIIA	Same as IIB, plus pneumoperitoneum	Same as IIA, plus surgery

Stroke/Intracranial Hemorrhage Scenarios

Scenario 5

A patient is in the unit recovering after stage II palliation with atrial septectomy for HLHS. OnPOD#3, the patient is noted to have rhythmic movement of the right leg. It resolves spontaneously and a cEEG is subsequently placed. Over 48 hours, no seizure activity is noted. An MRI shows a few punctate hemorrhages consistent with exposure to bypass. The neurology consult team recommends initiating therapy with Keppra given the clinical event.

Seq #	Field	Correct Response
	CICU Encounter Fields	
2560	Medical diagnosis	None (surgical dx only)
	Complication/Neuro Fields	
8120	Stroke (yes/no)	No
8142	Diagnosis date/time	--
8144	How diagnosed (clinical findings/imaging/both/unk)	--
	-If imaging = yes:	
8146	• ultrasound (yes/no)	--
8148	• MRI (yes/no)	--
8150	• CT (yes/no)	--
8152	Primarily hemorrhagic (yes/no)	--
8154	- If primarily hemorrhagic = no: Hemorrhagic conversion? (yes/no)	--
8156	› if yes, date/time	--
8220	Seizure (yes/no)	No
8240	- If yes, date/time	--
8260	IVH (≥ grade II)	Yes
8280	- If yes, date/time	Date of initial imaging
8282	- Max grade with date	Grade III, date of initial imaging
8302	Intracranial hemorrhage, non-stroke (yes/no)	No
8304	- first event date/time	--

Appendix: Stroke/Intracranial Hemorrhage Scenarios

Stroke/Intracranial Hemorrhage Scenarios

Scenario 1

Patient is in the CICU on ECMO after stage one palliation for HLHS. The patient is paralyzed and sedated, with cEEG in place. On ECMO day 3, there are HR and BP increases that do not correlate with seizure on cEEG. On day 4, there is left sided electrographic seizure noted and antiepileptic medications are started. An ultrasound is concerning for hypoperfusion in the left hemisphere. The patient is successfully decannulated on ECMO day 6, and an MRI done 2 days later reveals perfusion defect in the distribution of the left MCA without hemorrhage.

Follow up imaging to assess the MCA stroke prior to discharge reveals a small incidental subdural hemorrhage on the left with evolving MCA stroke without hemorrhagic conversion. No intervention was required for the subdural hemorrhage.

Seq #	Field	Correct Response
	CICU Encounter Fields	
2560	Medical diagnosis	None (surgical diagnosis only)
	Complication/Neuro Fields	
8120	Stroke (yes/no)	Yes
8142	Diagnosis date/time	Date/time of cEEG seizure
8144	How diagnosed (clinical findings/imaging/both/unk)	Both (clinical and EEG)
	-If imaging = yes:	
8146	• ultrasound (yes/no)	Yes
8148	• MRI (yes/no)	Yes
8150	• CT (yes/no)	No
8152	Primarily hemorrhagic (yes/no)	No
8154	- If primarily hemorrhagic = no: Hemorrhagic conversion? (yes/no)	No
8156	› if yes, date/time	--
8220	Seizure (yes/no)	Yes
8240	- If yes, date/time	Date/time of cEEG seizure
8260	IVH (≥ grade II)	No
8280	- If yes, date/time	--
8282	- Max grade with date	--
8302	Intracranial hemorrhage, non-stroke (yes/no)	Yes
8304	- If yes, first event date/time	Date/time of study that revealed subdural hemorrhage

Stroke/Intracranial Hemorrhage Scenarios

Scenario 2

A fenestrated Fontan patient presents to an outside hospital with new onset left sided weakness. An MRI done at the outside hospital reveals a right sided intracranial hemorrhage in the right parietal area. The patient is transferred to your CICU where an MRI/MRA reveals a right MCA thrombus. The patient is started on Keppra and no further interventions are planned for the thrombus. On CICU day 3 the patient has a left-sided seizure and is loaded with fosphenytoin.

Seq #	Field	Correct response
	CICU Encounter Fields	
2560	Medical diagnosis	Stroke
	Complication/Neuro Fields	
8120	Stroke (yes/no)	Yes
8142	Diagnosis date/time	CICU admit date/time
8144	How diagnosed (clinical findings/imaging/both/unk)	Both
	-If imaging = yes:	
8146	• ultrasound (yes/no)	No
8148	• MRI (yes/no)	Yes
8150	• CT (yes/no)	No
8152	Primarily hemorrhagic (yes/no)	Yes
8154	- If primarily hemorrhagic = no: Hemorrhagic conversion? (yes/no)	--
8156	› if yes, date/time	--
8220	Seizure (yes/no)	Yes
8240	- If yes, date/time	Date/time of clinical seizure on day 3
8260	IVH (≥ grade II)	No
8280	- If yes, date/time	--
8282	- Max grade with date	--
8302	Intracranial hemorrhage, non-stroke (yes/no)	No
8304	- If yes, first event date/time	--

Stroke/Intracranial Hemorrhage Scenarios

Scenario 3

A 12 year old patient with a prosthetic mitral valve who is on coumadin at baseline is admitted to the CICU after a fall. He presented to the ED with altered mental status but no focal findings and a head CT revealed a small to moderate sized intraparenchymal hemorrhage in the right frontal cortex with minimal midline shift. There is a small amount of blood in right frontal horn of the right lateral ventricle which is ungradable per the radiologist. An echo is concerning for an increased gradient across the prosthetic valve; the patient is transferred to the CICU for monitoring where studies on the valve are reassuring. An MRI/MRA confirms the intraparenchymal hemorrhage but is negative for thrombus. He is transferred to the floor, where coumadin is reinitiated prior to discharge.

Seq #	Field	Correct Response
	CICU Encounter Fields	
2560	Medical diagnosis	Intracranial hemorrhage
	Complication/Neuro Fields	
8120	Stroke (yes/no)	No
8142	Diagnosis date/time	--
8144	How diagnosed (clinical findings/imaging/both/unk)	--
	-If imaging = yes:	
8146	• ultrasound (yes/no)	--
8148	• MRI (yes/no)	--
8150	• CT (yes/no)	--
8152	Primarily hemorrhagic (yes/no)	--
8154	- If primarily hemorrhagic = no: Hemorrhagic conversion? (yes/no)	--
8156	› if yes, date/time	--
8220	Seizure (yes/no)	No
8240	- If yes, date/time	--
8260	IVH (≥ grade II)	No
8280	- If yes, date/time	--
8282	- Max grade with date	--
8302	Intracranial hemorrhage, non-stroke (yes/no)	Yes
8304	- first event date/time	CICU admit date/time

Stroke/Intracranial Hemorrhage Scenarios

Scenario 4

A patient is in the unit recovering after stage II palliation with atrial septectomy for HLHS. OnPOD#3, the patient is noted to have rhythmic movement of the right leg. It resolves spontaneously and a cEEG is subsequently placed. Over 48 hours, no seizure activity is noted. An MRI shows a few punctate hemorrhages consistent with exposure to bypass. The neurology consult team recommends initiating therapy with Keppra given the clinical event.

Seq #	Field	Correct Response
	CICU Encounter Fields	
2560	Medical diagnosis	None (surgical dx only)
	Complication/Neuro Fields	
8120	Stroke (yes/no)	No
8142	Diagnosis date/time	--
8144	How diagnosed (clinical findings/imaging/both/unk)	--
	-If imaging = yes:	
8146	• ultrasound (yes/no)	--
8148	• MRI (yes/no)	--
8150	• CT (yes/no)	--
8152	Primarily hemorrhagic (yes/no)	--
8154	- If primarily hemorrhagic = no: Hemorrhagic conversion? (yes/no)	--
8156	› if yes, date/time	--
8220	Seizure (yes/no)	Yes
8240	- If yes, date/time	Date of clinical even
8260	IVH (≥ grade II)	No
8280	- If yes, date/time	--
8282	- Max grade with date	--
8302	Intracranial hemorrhage, non-stroke (yes/no)	No
8304	- first event date/time	--

Stroke/Intracranial Hemorrhage Scenarios

Scenario 5

A patient is in the unit awaiting stage I palliation for HLHS. Per protocol, the patient undergoes a brain MRI which reveals bilateral grade III IVH. The patient is neurologically intact and hemodynamically stable on low dose prostaglandin and room air, so the decision is made to delay surgery for 5-7 days in order to assess the evolution of the IVH. One week later, a follow up MRI demonstrates no further bleeding and evolution of the previously noted IVH. The patient undergoes stage I palliation and recovers uneventfully.

Seq #	Field	Correct Response
	CICU Encounter Fields	
2560	Medical diagnosis	None (surgical dx only)
	Complication/Neuro Fields	
8120	Stroke (yes/no)	No
8142	Diagnosis date/time	--
8144	How diagnosed (clinical findings/imaging/both/unk)	--
	-If imaging = yes:	
8146	• ultrasound (yes/no)	--
8148	• MRI (yes/no)	--
8150	• CT (yes/no)	--
8152	Primarily hemorrhagic (yes/no)	--
8154	- If primarily hemorrhagic = no: Hemorrhagic conversion? (yes/no)	--
8156	› if yes, date/time	--
8220	Seizure (yes/no)	No
8240	- If yes, date/time	--
8260	IVH (≥ grade II)	Yes
8280	- If yes, date/time	Date of initial imaging
8282	- Max grade with date	Grade III, date of initial imaging
8302	Intracranial hemorrhage, non-stroke (yes/no)	No
8304	- first event date/time	--

Appendix: Tracheostomy/Ventilation Scenarios

Tracheostomy/Ventilation Scenarios

In these tables, rows in italics show coding for a patient who has an interventional cath rather than surgery.

Scenario 1

Patient trached and vented at baseline, comes in from home and has surgical intervention. In the OR, trach is replaced with ETT and patient returns to the CICU ventilated via the ETT. While still being ventilated, ETT is removed and trach recannulated. Patient weans to trach collar and is discharged home.

Seq #	Field	Correct Response
	Hospitalization fields	
1021	Trach at hospital admission	Yes
1022	Home resp support at hospital admission	Yes
	Cardiothoracic Surgery (or Cardiac Catheter) Fields	
1670	Intubated for surgery	Yes
1671	If yes, extubated in OR or upon arrival	No
2272	*Intubated for cath procedure*	*Yes*
2273	*If yes, extubated in cath lab or upon arrival*	*No*
	Respiratory Support Fields	
3040	Invasive ventilation	Yes
3210	Invasive vent began at CICU start	No
3223	Ventilated for procedure	No
3240	Invasive vent at CICU end	No
3260	If yes, end date known	--
3226	Initial airway	ETT
3229	Final airway	trach
3280	Vent end time	Time patient transitioned to trach collar
3232	If initial airway is ETT and final is trach, trach date/time	Date/time of recannulation

Tracheostomy/Ventilation Scenarios

Scenario 2

Patient trached and on heat moisture exchanger (HME) during the day and trach collar at night at baseline, comes in from home and has surgical intervention. Ventilated via the trach during the procedure and returns to the ICU being ventilated via the trach. Patient weaned to trach collar during the day and CPAP at night initially, and subsequently to trach collar around the clock. Discharged home on trach collar only.

Seq #	Field	Correct Response
	Hospitalization fields	
1021	Trach at hospital admission	Yes
1022	Home resp support at hospital admission	No
	Cardiothoracic Surgery (or Cardiac Catheter) Fields	
1670	Intubated for surgery	Yes
1671	If yes, extubated in OR or upon arrival	No
2272	*Intubated for cath procedure*	*Yes*
2273	*If yes, extubated in cath lab or upon arrival*	*No*
	Respiratory Support Fields	
3040	Invasive ventilation	Yes
3210	Invasive vent began at CICU start	Yes
3223	Ventilated for procedure	Yes
3240	Invasive vent at CICU end	No
3260	If yes, end date known	--
3226	Initial airway	trach
3229	Final airway	trach
3280	Vent end time	Time patient transitioned to trach collar
3232	If initial airway is ETT and final is trach, trach date/time	--

Tracheostomy/Ventilation Scenarios

Scenario 3

Patient trached and vented at baseline, admitted to the floor pre-operatively. Went to OR for surgical intervention, and trach is removed and patient is orally intubated for the case. Returns to the CICU with the ETT in place, being mechanically ventilated. Trach recannulated and ETT removed later in the CICU stay. Discharged home from the CICU with trach in place and receiving mechanical ventilation.

Seq #	Field	Correct Response
	Hospitalization fields	
1021	Trach at hospital admission	Yes
1022	Home resp support at hospital admission	Yes
	Cardiothoracic Surgery (or Cardiac Catheter) Fields	
1670	Intubated for surgery	Yes
1671	If yes, extubated in OR or upon arrival	No
2272	*Intubated for cath procedure*	*Yes*
2273	*If yes, extubated in cath lab or upon arrival*	*No*
	Respiratory Support Fields	
3040	Invasive ventilation	Yes
3210	Invasive vent began at CICU start	No (start time=hospital admission)
3223	Ventilated for procedure	No
3240	Invasive vent at CICU end	Yes
3260	If yes, end date known	No
3226	Initial airway	trach
3229	Final airway	trach
3280	Vent end time	CICU discharge date/time
3232	If initial airway is ETT and final is trach, trach date/time	--

Tracheostomy/Ventilation Scenarios

Scenario 4

Patient admitted to the floor, trached and vented at baseline. Goes to OR for surgical intervention and ventilated via the trach throughout the procedure. Returns to the CICU vented via the trach, and then is transferred to the floor still receiving mechanical ventilation via the trach. Discharged home still receiving mechanical ventilation via the trach.

Seq #	Field	Correct Response
	Hospitalization fields	
1021	Trach at hospital admission	Yes
1022	Home resp support at hospital admission	Yes
	Cardiothoracic Surgery (or Cardiac Catheter) Fields	
1670	Intubated for surgery	Yes
1671	If yes, extubated in OR or upon arrival	No
2272	*Intubated for cath procedure*	*Yes*
2273	*If yes, extubated in cath lab or upon arrival*	*No*
	Respiratory Support Fields	
3040	Invasive ventilation	Yes
3210	Invasive vent began at CICU start	No (start time=hospital admission)
3223	Ventilated for procedure	No
3240	Invasive vent at CICU end	Yes
3260	If yes, end date known	No
3226	Initial airway	trach
3229	Final airway	trach
3280	Vent end time	Hospital discharge date
3232	If initial airway is ETT and final is trach, trach date/time	--

Tracheostomy/Ventilation Scenarios

Scenario 5

Patient with trach but not ventilated at baseline, admitted pre-op to the floor. Goes to OR for surgical intervention where trach is removed and ETT placed. Returns to the CICU mechanically ventilated via the ETT. Is later recannulated while still requiring mechanical ventilation and subsequently is weaned to trach collar and discharged home.

Seq #	Field	Correct Response
	Hospitalization fields	
1021	Trach at hospital admission	Yes
1022	Home resp support at hospital admission	No
	Cardiothoracic Surgery (or Cardiac Catheter) Fields	
1670	Intubated for surgery	Yes
1671	If yes, extubated in OR or upon arrival	No
2272	*Intubated for cath procedure*	*Yes*
2273	*If yes, extubated in cath lab or upon arrival*	*No*
	Respiratory Support Fields	
3040	Invasive ventilation	Yes
3210	Invasive vent began at CICU start	Yes
3223	Ventilated for procedure	Yes
3240	Invasive vent at CICU end	No
3260	If yes, end date known	--
3226	Initial airway	ETT
3229	Final airway	trach
3280	Vent end time	Time patient transitioned to trach collar
3232	If initial airway is ETT and final is trach, trach date/time	Date/time of recannulation

Tracheostomy/Ventilation Scenarios

Scenario 6

Patient intubated and on the vent in the NICU. Goes to OR for surgical intervention, returns to the CICU with ETT in place and mechanically ventilated. Fails extubation attempt and is reintubated. Tracheostomy done and patient weaned first to CPAP at night and trach collar during the day, and eventually to around the clock trach collar. Discharged to home.

Seq #	Field	Correct Response
	Hospitalization fields	No
1021	Trach at hospital admission	No
1022	Home resp support at hospital admission	
	Cardiothoracic Surgery (or Cardiac Catheter) Fields	
1670	Intubated for surgery	Yes
1671	If yes, extubated in OR or upon arrival	No
2272	*Intubated for cath procedure*	*Yes*
2273	*If yes, extubated in cath lab or upon arrival*	*No*
	Respiratory Support Fields	
3040	Invasive ventilation	Yes
3210	Invasive vent began at CICU start	No
3223	Ventilated for procedure	No
3240	Invasive vent at CICU end	No
3260	If yes, end date known	--
3226	Initial airway	ETT
3229	Final airway	trach
3280	Vent end time	Time patient transitioned to trach collar
3232	If initial airway is ETT and final is trach, trach date/time	Admit time to CICU after trach