NEW YORK STATE DEPARTMENT OF HEALTH
DIVISION OF QUALITY AND PATIENT SAFETY
CARDIAC SERVICES PROGRAM

Instructions and Data Element Definitions
January 2012

Cardiac Surgery Report, Pediatric
(Under age 18)
Form DOH-2254p

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Revision Highlights and Coding Clarification

New Data Elements

Interventional Cardiologist - A field has been added to collect the NYS Physician License number of interventional cardiologist if the surgical procedure included both surgical and interventional components.

Revised Data Elements

Start time is now collected as time of first skin incision.

Diagnosis #1 is now the Fundamental Diagnosis.
Diagnosis #2 is now the Primary Diagnosis.
Diagnosis #3 - #5 are now Additional Diagnosis #1 - #3.

Procedure #1 is now the Primary Procedure
Procedures #2 - #4 are now Additional Procedures #1 - #3.

The Procedure Code List (Attachment D) and Diagnosis Code List (Attachment E) have been updated. These code sets are compatible with those reported to the STS Congenital Heart Surgery Database v3.0 and are used with permission.

Pulmonary Hypertension has been renamed “Near Systemic PVR.” The definition is unchanged.

Data Element Clarifications

Please see “When to Complete A PedCSRS” form for revised clarifications on what procedures should be reported to PedCSRS.

When to Complete a Pediatric CSRS Form

Complete a Pediatric Cardiac Surgery Reporting System (Pediatric CSRS) form for every patient under the age of 18 at the time of admission undergoing one or more surgical operations **on the heart or great vessels,** with or without extracorporeal circulation.

Complete a Pediatric CSRS form only for procedures that include a surgical intervention on the heart or great vessels. Procedure codes for other types of interventions may be used (as space permits) to indicate non-surgical and/or non-cardiac components of a cardiac surgery. However, non-surgical and non-cardiac procedures are not "form generating". This means that performing one of these procedures by itself, with no cardiac surgical procedure at the same time, is not reportable.

If more than one cardiac surgery occurred during a single hospital stay, complete a separate form for each operation.

Unless otherwise specified, forms should be created for reportable cardiac surgery even if it occurs in a location other than the operating room.

A surgical procedure begins at the time of the FIRST skin incision, unless otherwise stated.

Examples of procedures that are not "form generating" include but are not limited to the following codes found in Attachment D:

- Thoracic and Mediastinal Disease
  - Lung biopsy (1400)
  - Lung procedure, Other (1420)
  - Pectus repair (1430)
  - Tracheal procedure (1440)
- Interventional Cardiology Procedures – All Listed
- Anesthetic Procedures - All Listed
- Pericardial Disease
  - Pericardial drainage procedure (920)
- Thoracic Arteries and Veins
  - PDA closure, Device using transcatheter technique (1340)
- Electrophysiological Procedures
  - Pacemaker implant, Permanent (1450)
  - Pacemaker procedure (1460)
  - Explantation of pacing system (2350)
  - ICD [AICD] implantation (1470)
  - ICD [AICD] procedure (1480)

When to Complete a Pediatric CSRS Form (cont.)

Examples of procedures that are not form generating (continued)

- Mechanical Support
  - ECMO decannulation (2370)
  - IABP insertion (1900)
  - VAD explantation (2390)
- Miscellaneous Procedures
  - Pleural drainage procedure (1810)
  - Pleural procedure, Other (1820)
  - Ligation, Thoracic duct (1830)
  - Decortication (1840)
  - Esophageal procedure (1850)
  - Mediastinal procedure (1860)
  - Bronchoscopy (1870)
  - Diaphragm plication (1880)
  - Diaphragm procedure, Other (1890)
  - VATS – video assisted thoracoscopic surgery (1930)
  - Minimally invasive procedure (1940)
  - Bypass for non-cardiac lesion (1950)
  - Delayed sternal closure (1960)
  - Mediastinal exploration (1970)
  - Sternotomy wound drainage (1980)
  - Thoracotomy, Other (1990)
  - Cardiotomy, Other (2000)
  - Thoracic and/or mediastinal procedure, Other (2020)
  - Peripheral vascular procedure, Other (2030)
  - Miscellaneous procedure, Other (2040)
  - Organ procurement (2050)
  - Other procedure (7777)

PDA closure, Surgical (1330) is form generating only when performed in the operating room on a baby weighing at least 1500 grams. If done at the same time as another cardiac surgical procedure, it should always be reported. This is consistent with the prior PedCSRS instruction to not report an *isolated* PDA on patients less than 1500g or if performed anywhere other than the operating room.

ECMO cannulation (2360): Is form generating only when there is also another PedCSRS reportable procedure ***during the admission***. For these cases, ECMO should be reported regardless of physical location or clinical staff responsible.

Cardiac procedure, Other (2010): Should not be reported for procedures that are not cardiac or that are not surgical. Operative notes will be requested as part of the validation process for cases reported with this procedure code.

Pediatric CSRS Data Reporting Policies

Hospice Policy

Beginning with patients discharged on or after January 1, 2003, any patient that is discharged from the hospital after cardiac surgery or PCI to hospice care (inpatient or home with hospice care) and is still alive 30 days after the discharge from the hospital will be analyzed as a live discharge.

All patients discharged to a hospice or home with hospice care should continue to be reported with Discharge Status – 12: Hospice. If a patient is still alive 30 days after discharge to hospice, whether in hospice or not, appropriate supporting documentation should be sent to Cardiac Services Program. Examples of appropriate documentation include: a dated progress note from the hospice service, evidence of a follow-up doctor's visit 30 days after discharge, evidence of subsequent hospital admission 30 days after initial discharge. It will be the responsibility of the hospital (physician) to send documentation to the Department of Health to support this change. Upon receipt, review, and verification of the documentation, Cardiac Services Program staff will change the discharge status from dead to alive for purposes of analysis. All documentation must be received before the final volume and mortality for a given year of data is confirmed by the hospital.

Reporting Schedule

Pediatric CSRS data is reported quarterly by discharge date. It is due to the Cardiac Services Program two months after the end of the quarter. The 2012 reporting schedule is as follows.

Quarter 1 (1/1/12 – 3/31/12 Discharges) due on or before May 31, 2012
Quarter 2 (4/1/12 – 6/30/12 Discharges) due on or before August 31, 2012
Quarter 3 (7/1/12 – 9/30/12 Discharges) due on or before November 30, 2012
Quarter 4 (10/1/12 – 12/31/12 Discharges) due on or before February 28, 2013

Limited extensions to the above deadlines will be granted on a case by case basis when warranted by extenuating circumstances. They must be requested in writing prior to the required submission date.

Item-by-Item Instructions

PFI Number

Variable Name: PFI

The PFI Number is a Permanent Facility Identifier assigned by the Department of Health. Enter your facility's PFI Number as shown in Attachment A.

Sequence Number

Variable Name: SEQUENCE

If your facility assigns a sequence number to each case on a chronological flow sheet or similar log, enter the sequence number here. The sequence number is not required for the Pediatric Cardiac Surgery Reporting System, but has been included on the form in case your facility finds it useful in identifying and tracking cases.

I. Patient Information

Child's Name

Variable Names: LASTNAME, FIRSTNAME

Enter the child's last name followed by his/her first name.

Medical Record Number

Variable Name: MEDRECNO

Enter the child's medical record number.

Child's Social Security Number

Variable Name: SSNO

Enter the child's social security number.

Patient Information (continued)

Age in Years

Variable Name: AGE

Enter the child's age at admission to the hospital. If the child is less than one year old, enter "0". If the child is admitted on or after his/her 18th birthday, please complete an Adult CSRS form NOT a Pediatric CSRS form.

Date of Birth

Variable Name: DOB

Enter the child's exact date of birth.

Sex

Variable Name: SEX

Check the appropriate box.

Ethnicity

Variable Name: ETHNIC

Check the appropriate box.

Race

Variable Names: RACE, RACESPEC

Select one of the following.

1 - White. A person having origins in any of the original peoples of Europe, the Middle East, or North Africa.

2 - Black or African American. 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."

3 - Native American / American Indian or Alaska Native. 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.

Patient Information (continued)

Race (cont.)

4 - Asian. 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.

5 - Native Hawaiian or Other Pacific Islander. A person having origins in any of the original peoples of Hawaii, Guam, Samoa, or other Pacific Islands.

8 - Other. Report for those responses that are not covered by an above category. Please provide the specific race for any case marked “Other.”

Note: Please note that race should be based on the patient’s racial/ethnic origins, which is not necessarily the same as their country or place of origin.

Multi-racial can be indicated by checking “8-Other” and providing details in the “specify” field.

For White Hispanics, check "White"; for Black Hispanics, check "Black."

Residence Code

Variable Names: RESIDENC, STATE

Enter the county code of the patient’s principal residence, as shown in Attachment B. If the patient lives outside of New York State, use code 99 and print the name of the state or country where the patient resides in the space provided. If you enter a valid NYS County Code then the ‘State or Country” field **should** be left blank.

If the patient is from a foreign country, but is staying in the US during the pre-operative and post-operative time period, you must enter 99 and print the name of the country that the patient is from. Do not enter the residence code of where the patient is staying while in the United States.

Hospital Admission Date

Variable Name: ADMIDATE

Enter the date that the current hospital stay began.

Patient Information (continued)

Primary Payer

Variable Name: PAYER

Enter the primary source of payment for this hospital stay as shown in Appendix C.

Interpretation: Primary Payer and Medicaid

For “Medicaid Pending” code Primary Payer as “11 - Self-Pay” **and** check the box for Medicaid.

Please note the difference between “07 - Other Private Insurance Company” and “19 - Other”. Code 07 refers to a Private Insurance Company (also referred to as “Commercial” insurance) that is not listed elsewhere. Use Code 19 for any other type of insurance that is not given a code of its own (e.g. Corrections).

Code a PPO (Preferred Provider Organization) as Code 06 – HMO/Managed Care.

If you know a patient has Medicare or Medicaid, but do not know if it is Fee for Service or Managed Care, code Fee for Service.

Medicaid

Variable Name: MEDICAID

Check this box if the patient has Medicaid that will provide payment for any portion of this hospital stay. If the patient’s primary payer is Medicaid, check this box in addition to entering “03” or “04” under Primary Payer.

PFI of Transferring Hospital

Variable Name: TRANS_PFI

If the patient was transferred from another acute care facility, enter the PFI of the transferring hospital.

This element only needs to be completed for transfer patients.

A listing of PFIs for cardiac diagnostic centers in New York State (NYS) is provided in Attachment A. If transferred from a Veterans Administration hospital in NYS, enter "8888"; if transferred from outside NYS, enter "9999". For patients transferred from another hospital in NYS, please see http://hospitals.nyhealth.gov for a complete listing of NYS hospitals, including their PFI.

II. Procedural Information

REMINDER: *Complete a separate pediatric cardiac surgery form for each surgery involving the heart or great vessels during the current hospital admission.*

Date of Surgery

Variable Name: SURGDATE

Enter the date on which the cardiac surgical procedure was performed.

Remember to fill out a separate pediatric cardiac surgery form for ***each*** cardiac surgery that occurred during the admission.

Time of First Skin Incision

Variable Names: SURGHOUR, SURGMIN

Enter the time of the first skin incision for this procedure, using military time (e.g. 1:00 am is 01:00, and 1:00 pm is 13:00).

Primary Surgeon Performing Surgery

Variable Name: PHYSNUM

Enter the name and NYS physician license number of the primary or principal surgeon who performed the cardiac surgical procedure(s).

Note: Primary Surgeon name is included on the paper form for abstractor convenience. It is not part of the PedCSRS file structure.

Interventional Cardiologist

Variable Name: CARDNUM

Enter the name and NYS physician license number of the interventional cardiologist participating in the case if this surgical procedure also included an interventional component.

Note: Interventional Cardiologist name is included on the paper form for abstractor convenience. It is not part of the PedCSRS file structure.

II. Procedural Information (continued)

Surgical Priority

Variable Name: PRIORITY

Check the appropriate box.

Elective: All cases not classified as urgent or emergency as defined below.

Urgent: The patient is too ill or unstable to be discharged from the hospital, but is not classified as an emergency as defined below.

This includes patients with ductal-dependent systemic or pulmonary circulation.

Emergency: Patients with cardiac compromise or circulatory compromise of the cardiac organ.

Typical emergency patients include those with obstructed anomalous pulmonary venous return and those with ductal-dependent systemic or pulmonary circulation in whom ductal patency cannot be maintained.

Prior Surgery this Admission

Variable Names: PRIOSURG, PRIODATE

Check the appropriate box to indicate whether the patient underwent any cardiac surgery prior to this one during the current hospital admission.

If “Yes” then the date of the most recent previous cardiac operation **MUST** be entered.

II. Procedural Information (continued)

Fundamental Diagnosis

Variable Names: DIAG1

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.

No "Status - post diagnoses" can be a primary diagnosis or fundamental diagnosis.

Most frequently, the primary diagnosis will also be the fundamental diagnosis. For some operations, however, the fundamental diagnosis and primary diagnosis will be different.

For example, a patient who has a complete AV canal defect and undergoes either palliation or repair of the defect has a primary and fundamental diagnosis of "AVC (AVSD), Complete CAVSD". Subsequently, the child develops mitral insufficiency and is re-hospitalized for mitral valve replacement. The primary diagnosis for the mitral valve replacement operation is "Mitral regurgitation", but the fundamental diagnosis is "AVC (AVSD), Complete CAVSD."

Coding Note: The definition of Fundamental Diagnosis (*DIAG1)* and the Congenital Diagnosis Codes in Attachment E are aligned with STS Congenital Heart Surgery Database v3.0 data element 430.
Society of Thoracic Surgeons, Congenita Heart Surgery Database, Version 3.0 , used with permission.

Primary Diagnosis

Variable Names: DIAG2

Indicate the diagnosis of primary importance at the time of this surgical procedure.

No "Status - post diagnoses" can be a primary diagnosis or fundamental diagnosis.

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.

Coding Note: The definition of Primary Diagnosis (*DIAG1)* and the Congenital Diagnosis Codes in Attachment E are aligned with STS Congenital Heart Surgery Database v3.0 data element 870.
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II. Procedural Information (continued)

Additional Cardiac Diagnosis Codes (#1 - #3)

Variable Names: DIAG3, DIAG4, DIAG5

Report up to three additional diagnoses. Indicate up to three diagnoses noted at the time of the surgical procedure or documented by preoperative studies.

Coding Note: The Congenital Diagnosis Codes in Attachment E are aligned with those used in STS Congenital Heart Surgery Database v3.0 data element 870.
Society of Thoracic Surgeons, Congenita Heart Surgery Database, Version 3.0, used with permission.

Primary Procedure Code

Variable Names: PROC1

Use the codes in Attachment D to report the PRIMARY procedure performed during this surgical procedure.

Coding Note: The definition of Primary Procedure (*PROC1)* and the Procedure Codes in Attachment D are aligned with STS Congenital Heart Surgery Database v3.0 data element 910.
Society of Thoracic Surgeons, Congenita Heart Surgery Database, Version 3.0, used with permission.

Additional Cardiac Procedure Codes (#1 - #3)

Variable Names: PROC2, PROC3, PROC4

Use the procedure codes listed in Attachment D to indicate additional procedure(s) performed during this operation.

Do not repeat the procedure reported as Primary Procedure in these fields.

If there are more than 3 additional procedures, select procedure codes that are both cardiac and surgical in order of significance first. You may use additional spaces for non-surgical interventions that take place during the procedure or portions of the procedure that are not primarily directed at the heart or great vessels only as space permits.

Coding Note: The Procedure Codes in Attachment D are aligned with those used in the STS Congenital Heart Surgery Database v3.0 data element 900.
Society of Thoracic Surgeons, Congenita Heart Surgery Database, Version 3.0, used with permission.

II. Procedural Information (continued)

Mode of Cardiopulmonary (CP) Bypass

Variable Name: LOWFLOW, DEEPHYPO, CIRCARES

Check all that apply. If none apply, leave blank.

Minimally Invasive

Variable Name: MINI_INV

If the cardiac surgical procedure began through an incision other than a complete sternotomy or thoracotomy check "Yes", regardless of whether the case was converted to a standard incision or CP Bypass was used. Otherwise check "No".

Entire Procedure Off Pump

Variable Name: ALL_OFF

Check this box if the cardiac operation was performed entirely without the use of cardiopulmonary bypass.

CABG Information

Variable Names: TOT_COND, ART_COND, DISTAL

If Procedure Code 670 is coded then the following information must be completed.

Total Conduits: List the total number of conduits or grafts performed up to 9. For more than 9, write 9.

Arterial Conduits: List the number of arterial conduits or grafts used up to 9. For more than 9, write 9. The number of arterial conduits **CANNOT** be larger than the total number of conduits

Distal Anastomoses: List the total number of distal anastomoses up to 9. For more than 9, write 9. A distal anastomosis is defined as a hole between a conduit or graft and a coronary touchdown site for the conduit or graft. The number of distal anastomoses could be larger than the total number of conduits, especially in the case of sequential grafts.

III. Pre-Operative Status

Weight at Time of Operation

Variable Names: WGT_UNIT, WEIGHT

Enter the patient's weight at the time of the operation. If less than 10 kilograms, report in grams, if 10 kilograms or more report in kilograms. Check the appropriate box for grams or kilograms.

Gestational Age at Birth in Weeks

Variable Name: GEST_AGE

If the patient is under one year of age at admission, enter the gestational age at birth (in weeks).

If the patient's age at admission was one year or more, this item should be left blank.

Weight at Birth in Grams

Variable Names: BIRTHWGT

If the patient is under one year of age at admission, check the box with the appropriate weight range in grams. If the patient's age at admission was one year or more, this item should be left blank.

Pre-operative Conditions

Check all of the following conditions that existed prior to the start of the procedure, but within the time frame specified.

0. None

Variable Name: NORISK

None of the pre-operative conditions listed below were present prior to surgery.

III. Pre-Operative Status (continued)

1-3. Previous Open Heart Operations

Variable Names: PREVOP_1, PREVOP_2, PREVOP_3

If the patient had an open-heart surgery prior to the current cardiac operation, check the appropriate box to indicate the number of such operations.

Interpretation: For the purposes of this reporting system, minimally invasive procedures are considered open-heart surgery.

"Previous Open Heart Operations" refers to surgeries using CP Bypass and "Previous Closed Heart Operations" refers to those without CP Bypass.

Include any previous surgeries, either from this admission or a previous admission.

If there was a previous surgery this admission, please be sure that the date of the most recent surgery is indicated in the field "Prior Surgery This Admission" on the front of the form.

4-6. Previous Closed Heart Operations

Variable Names: PRECLO_1, PRECLO_2, PRECLO_3

If the patient had a closed heart surgery prior to the current cardiac operation, check the appropriate box to indicate the number of such operations.

Interpretation: "Previous Open Heart Operations" refers to surgeries using CP Bypass and "Previous Closed Heart Operations" refers to those without CP Bypass.

Include any previous surgeries, either from this admission or a previous admission.

If there was a previous surgery this admission, please be sure that the date of the most recent surgery is indicated in the field "Prior Surgery This Admission" on the front of the form.

III. Pre-Operative Status (continued)

7. Pre-op Interventional Cath Procedure

Variable Names: PRE_CATH, INT_DATE

Indicate if the patient has had a pre-operative interventional cardiac catheterization procedure.

If during this admission, enter the date of the most recent procedure in the space provided.

Interpretation: Examples of these procedures include but are not limited to coil embolization of collaterals, balloon valvuloplasty, balloon dilation of coarctation of the aorta, defect closure, pulmonary artery, systemic vein or pulmonary vein. Balloon atrial septostomy would be excluded.

Report this risk factor if the patient underwent a cardiac intervention in-utero (e.g. aortic valve dilation).

11. Severe Cyanosis or Severe Hypoxia

Variable Name: SEV_CYAN

Code if any of the following are present and sustained within 12 hours prior to surgery:

Pulse oximetry saturation <70%
Resting PO2 < 35mmHg
Arterial saturation <75%

Interpretation: The following scenario **would** be coded: Medical record states: "the patient's baseline oxygen saturation is 68% on room air. Central Aorto-Pulmonary Shunt placed for full repair due to cyanosis."

12. Dialysis within 14 Days Prior to Surgery

Variable Name: DIAL_PRE

Code if the patient received either continuous or intermittent hemodialysis or peritoneal dialysis within 14 days prior to surgery. The dialysis does not have to occur in the same hospital stay, it only has to be within 14 days of the procedure.

Note: You may also code this element if the patient had Continuous Renal Replacement Therapy (CRRT), for example PRISMA, within 14 days prior to surgery.

Do not report this risk factor if the patient requires CRRT, for example PRISMA, for fluid management while on ECMO.

III. Pre-Operative Status (continued)

13. Any Ventilator Dependence During the Same Admission or within 14 Days Prior to Surgery

Variable Name: VENT_PRE

Code if the patient was ventilator dependent during the same admission *or* within 14 days prior to surgery.

Interpretation: The following scenario **would** be coded because surgery occurred in the same admission as ventilator dependence even though there was 16 days between ventilator dependence and surgery:

Admitted on 5/15
Ventilator dependent on 6/1
Extubated on 6/10
Surgery on 6/26
Discharged on 6/30

The following scenario **would NOT** be coded because more than 14 days passed between ventilator dependence and surgery:

Admitted on 5/15
Ventilator dependent on 6/1
Extubated on 6/10
Discharged on 6/13
Admitted on 6/20
Surgery on 6/26
Discharged on 6/30

Nasal CPAP is not considered pre-operative ventilator dependence.

14. Inotropic Support Immediately Pre-op within 24 hrs

Variable Name: INOT_PRE

Code if either of the following is present in the patient's medical record:

Dopamine in dosage >5 mcg/kg/minute
Any other agent/dose for inotropic support

15. Positive Blood Cultures within 2 Weeks of Surgery

Variable Name: POS_BLOO
Code if the patient has had positive blood cultures that are documented in the medical record, occurring within 2 weeks prior to surgery.

Interpretation: This can be coded even if the patient had the positive blood cultures within 2 weeks of surgery, was discharged, and was then re-admitted for surgery.

III. Pre-Operative Status (continued)

16. Arterial pH < 7.25, Immediately Pre-op within Hospital Stay

Variable Name: ARTER_PH

Arterial pH is < 7.25 within 12 hours prior to surgery but before the first blood gas taken in the OR.

17. Significant Renal Dysfunction

Variable Name: RENA_DYS

Code if Creatinine levels reach the indicated range for the patient's age:

Preemies and Newborn	Creatinine >1.5 mg /dl
>1 month of age	Creatinine >2.0 mg/dl

18. Trisomy 21

Variable Name: DOWN_SYN

Code for any patients with Trisomy 21 (Down's Syndrome).

III. Pre-Operative Status (continued)

19. Major Extracardiac Anomalies

Variable Name: CARDANOM and ANOM_SPEC

Check this box for any extracardiac anomaly not already captured on the PedCSRS form that is felt to be clinically relevant. Specify the anomaly in the space provided.

Examples include but are not limited to:

Non-Down's Syndrome chromosomal abnormalities	Tracheo-esophageal (TE) fistula
DiGeorge's Syndrome	Choanal Atresia
Cystic Fibrosis	Diaphragmatic hernia
Marfan's Syndrome	Biliary Atresia
Sickle Cell Anemia	Any -ostomy
Blood Dyscrasia	Beecher Muscular Dystrophy
Omphalocele	Tethered Spinal Cord
Hypoplastic lung	Vater Syndrome
	Pierre Robin Syndrome

The following would *not* be accepted as Major Extracardiac Anomalies:

Failure to Thrive	Normothermic
Developmentally Delayed	Cleft lip/palate
Hepatomegaly	Hirschsprung Disease
Preemie	Legally blind
Jaundiced	

Note: As part of the data validation process, you may be asked to provide additional information on the nature, extent, or severity of the "Major Extracardiac Anomaly." Please keep notes on cases with this risk factor to facilitate this validation.

21. Near Systemic Pulmonary Vascular Resistance (PVR)

Variable Name: PULM_HYP

In the case of an unrestrictive ventricular or great vessel communication (e.g. ductus or AP window), the following would constitute evidence of increased PVR (and hence presence of the risk factor):

- bidirectional shunting (meaning at least some R to L shunting) across the defect

OR

- absence of CHF symptoms in patients at least 2 months of age

OR

- evidence of systemic or suprasystemic RV pressure by tricuspid regurgitant jet velocity in the absence of a moderate or large left to right shunt

III. Pre-Operative Status (continued)

22. Ventricular Assist

Variable Name: PREOPVAD

Code if any of the following were used prior to the procedure to maintain vital signs:

Extracorporeal Membrane Oxygenation (ECMO)
Intra-Aortic Balloon Pump (IABP)
Left Ventricular Assist Device (LVAD)
Right Ventricular Assist Device (RVAD)
Bi-Ventricular Assist Device (BIVAD)

24. Pre-existing Neurologic Abnormality

Variable Name: NEUROABN and NEURO_SPEC

Check this box for any pre-existing neurologic abnormality. Specify the abnormality in the space provided.

Pre-existing neurological abnormality includes but is not limited to:

Documented intracranial bleed
Hydrocephalus
Chiari Malformation
Arterial venous malformation
Cerebral vascular accident (CVA)
Seizure disorders

Note: As part of the data validation process, you may be asked to provide additional information on the nature, extent, or severity of the "Pre-existing Neurologic Abnormality." Please keep notes on cases with this risk factor to facilitate this validation.

25. Pneumonia at Time of Surgery

Variable Name: PNEUMONI

As evidenced by:

Chest X-ray with infiltrate and at least **ONE** of the following:

- temperature greater than 101°F (38.5°C)
- white blood count greater than 12,000
- positive blood culture/viral titer.

III. Pre-Operative Status (continued)

26. Prostaglandin Dependence at Time of Surgery

Variable Name: PROSTAGL

At the time of surgery, the child requires prostaglandin to maintain normal respiration

27. Balloon Atrial Septostomy

Variable Name: BALLSEPT

Prior to surgery, but within the same hospital admission, the patient had a balloon atrial septostomy.

28. Any Previous Organ Transplant

Variable Name: ORGN_TRA

The patient has had any organ transplant prior to the current cardiac surgery. This includes, but is not limited to, heart, lung, kidney, and liver transplants. If a heart and/or lung transplant was performed during the operating room visit that generated this form DO NOT code this risk factor.

Interpretation: Also code for bone marrow transplant. Do not code for skin transplant (grafting).

IV. Post-Procedural Events Requiring Intervention

Check all of the listed post-procedural events that occurred following the surgery.

Please Note: *A documented pre-operative condition that persists post-operatively with* ***NO i****ncrease in severity is* ***NOT*** *a post-procedural event.*

0. None

Variable Name: NOEVENTS

Check if none of the post-procedural events listed below occurred following the operation.

1. Cardiac Tamponade

Variable Name: CARDTAMP

Code if cardiac tamponade is present post procedure.

Interpretation: Cardiac Tamponade should be coded if there is post-op chest drainage. Code regardless of where the drainage was performed (operating room, bedside, etc.).

2. Ventricular Fibrillation or CPR

Variable Name: VENT_FIB

Code if the patient experiences V-Fib or requires CPR at any time post-procedure, but before hospital discharge.

3. Bleeding Requiring Reoperation

Variable Name: BLEDREOP

Unplanned reoperation to control bleeding or to evacuate large hematomas in the thorax or pericardium.

Interpretation: This should be coded no matter where the bleeding was controlled (i.e., ICU, OR, bedside).

IV. Post-Procedural Events Requiring Intervention (continued)

4. Deep Sternal Wound Infection

Variable Name: DSW_INF

Drainage of purulent material from the sternotomy or thoracotomy wound.

Report this event only when associated with instability of the sternum.

A sternal wound infection should be reported as a post-procedural event even if it does not become apparent until after the patient is discharged from the hospital.

NOTE: This event is reportable up to one-year post-procedure, regardless of when the patient was discharged.

6. Ventilator Dependency > 10 Days

Variable Name: VENDEP10

The patient is unable to be extubated within 10 days post procedure.

Do not report if the patient had been ventilator dependent within 14 days prior to surgery.

7. Clinical Sepsis with Positive Blood Cultures

Variable Name: SEPSIS

Report if either of the following is present post procedure:

Temperature over 101° F (38.5° C) **and** Increased WBC **and** Positive blood culture

OR

Temperature below 98.6°F (37°C) **and** Decreased WBC **and** Positive blood culture

11. Renal Failure Requiring Dialysis

Variable Name: DIALYSIS

Code if the patient requires either continuous or intermittent hemodialysis or peritoneal dialysis post-procedure. Also code if the patient requires Continuous Renal Replacement Therapy (CRRT), for example PRISMA, post-procedure.

DO NOT code if the patient required dialysis (or CRRT) within 14 days before the procedure. Do not report this major event if the patient requires CRRT, for example PRISMA, for fluid management while on ECMO.

IV. Post-Procedural Events Requiring Intervention (continued)

12. Complete Heart Block at Discharge

Variable Name: COMP_HB

Code if the heart block lasts until the time of discharge with or without permanent pacemaker insertion before discharge.

13. Unplanned Cardiac Reoperation or Interventional Catheterization

Variable Name: UP_REOP

Includes any unplanned cardiac reoperation or interventional catheterization.

The procedure can be done in the operating room, cath lab, or at the bedside.

This would **exclude** a reoperation to control bleeding.

15. New Neurologic Deficit

Variable Name: NEURODEF

New neurologic deficit **present at discharge**.

16. Ventricular Assist

Variable Name: POST_VAD

Code if any of the following were required after the procedure to maintain vital signs:

Extracorporeal Membrane Oxygenation (ECMO)
Intra-Aortic Balloon Pump (IABP)
Left Ventricular Assist Device (LVAD)
Right Ventricular Assist Device (RVAD)
Bi-Ventricular Assist Device (BIVAD)

Do not code if Pre-Operative Status #22 is reported or if VAD/ECMO support was initiated during this procedure (and reported as a procedure code).

V. Discharge Information

Hospital Discharge Date

Variable Name: DISDATE

Enter the date the patient was discharged from the hospital.

If the patient died in the hospital, the hospital discharge date is the date of death.

Discharged Alive To

Variable Name: STATUS, DISWHERE

Check the appropriate box.

If a patient is discharged to Hospice (including Home with Hospice), code the status a "12". NOTE that for purposes of analysis a hospice discharge ("12") is considered an in-hospital mortality, unless the hospital can provide documentation that 30 days after discharge the patient was still alive (even if still in Hospice*).*

Please see the full Hospice policy and reporting requirements under "Pediatric CSRS Data Reporting Policies."

"19 – Other (specify)" should only be checked for a live discharge status not otherwise specified in this section (e.g. AMA).

Any status "19" that is reported without a specific discharge location will be sent back during data validation.

Died in

Variable Name: STATUS, DISWHERE

Check the appropriate box.

If "8 – Elsewhere in Hospital (specify)" is checked, specify where the patient died.

Any status "8" that is reported without an indication of where the patient expired will be sent back during data validation.

30 Day Status

Variable Name: THIRTYDAY

Report the patient's status at 30 days post-procedure using the appropriate code.

Attachment A
PFI Numbers for Cardiac Diagnostic and Surgical Centers

PFI	Facility
ALBANY AREA	
0001	Albany Medical Center Hospital
0135	Champlain Valley Physicians Hospital Medical Center
0829	Ellis Hospital
1005	Glens Falls Hospital
0746	Mary Imogene Bassett Hospital
0755	Rensselaer Regional Heart Institute – St. Mary's
0756	Rensselaer Regional Heart Institute – Samaritan
0818	Saratoga Hospital
0005	St. Peter's Hospital
BUFFALO AREA	
0207	Buffalo General Hospital
0210	Erie County Medical Center
0213	Mercy Hospital of Buffalo
0215	Millard Fillmore Hospital – Gates
0066	Olean General Hospital
0103	Women's Christian Association Hospital
ROCHESTER AREA	
0116	Arnot Ogden Medical Center
0471	Unity Hospital of Rochester
0411	Rochester General Hospital
0413	Strong Memorial Hospital
SYRACUSE AREA	
0977	Cayuga Medical Center at Ithaca
0628	Upstate University Hospital at Community General
0636	Crouse Hospital
0599	Faxton-St. Luke's Healthcare, St. Luke's Division
0598	St. Elizabeth Medical Center
0630	St. Joseph's Hospital Health Center
0058	United Health Services Hospital, Inc.-Wilson Medical Center
0635	University Hospital SUNY Health Science Center (Upstate)

PFI	Facility

NEW ROCHELLE AREA

0989 Benedictine Hospital
0885 Brookhaven Memorial Hospital Miedcal Center
0779 Good Samaritan Hospital of Suffern
0925 Good Samaritan Hospital Medical Center-West Islip
0913 Huntington Hospital
0513 Mercy Medical Center
0528 Nassau University Medical Center
0541 North Shore University Hospital
0686 Orange Regional Medical Center
1072 Sound Shore Medical Center of Westchester
0527 South Nassau Communities Hospital
0924 Southside Hospital
0943 St. Catherine of Siena Medical Center
0563 St. Francis Hospital (aka St. Francis Hospital The Heart Center, Roslyn)
0180 St. Francis Hospital (aka St. Francis Hospital & Health Ctrs, Poughkeepsie)
0694 St. Luke's Cornwall Hospital/Newburgh
0245 University Hospital - Stony Brook
0990 Kingston Hospital
0181 Vassar Brothers Medical Center
1139 Westchester Medical Center
1045 White Plains Hospital Center
0511 Winthrop University Hospital

NY CITY AREA

1438 Bellevue Hospital Center
1439 Beth Israel Medical Center / Petrie Campus
1164 Bronx-Lebanon Hospital Center-Fulton Division
1178 Bronx-Lebanon Hospital Center-Concourse Division
1286 Brookdale Hospital Medical Center
1288 Brooklyn Hospital Center-Downtown
1626 Elmhurst Hospital Center
1294 Coney Island Hospital
1445 Harlem Hospital Center
1300 Interfaith Med Ctr, Jewish Hospital Med Ctr of Brooklyn Division
1165 Jacobi Medical Center
1629 Jamaica Hospital Medical Center
1301 King's County Hospital Center
1450 Lenox Hill Hospital
1302 SUNY Downstate Medical Center @ Long Island College Hospital
1630 Long Island Jewish Medical Center
1304 Lutheran Medical Center
1305 Maimonides Medical Center

PFI	Facility
NY CITY AREA (CONT.)	
3058	Montefiore Medical Center-Jack D. Weiler Hospital of A. Einstein College Division
1169	Montefiore Medical Center-Henry and Lucy Moses Division
1456	Mount Sinai Hospital
1637	NY Hospital Medical Center of Queens
1306	NY Methodist Hospital
1464	NY Presbyterian-Columbia Presbyterian Center
1458	NY Presbyterian-NY Weill Cornell Center
1463	NYU Medical Center
1176	St. Barnabas Hospital
1466	St. Luke's Roosevelt Hospital Center-Roosevelt Hospital Division
1469	St. Luke's Roosevelt Hospital-St. Luke's Hospital Division
1740	Staten Island University Hospital-North
1738	Richmond University Medical Center
1320	University Hospital of Brooklyn
1318	Wyckoff Heights Medical Center
8888	Catheterization Laboratory at a Veterans Administration Hospital in New York. (for use in this reporting system; not an official Permanent Facility Identifier)
9999	Catheterization Laboratory Outside New York State (for use in this reporting system; not an official Permanent Facility Identifier)

A complete listing of NYS hospitals, including their PFI can be found at: http://hospitals.nyhealth.gov/ .

Attachment B
Residence Codes

The county codes shown below are also used in the SPARCS Discharge Data Abstract:

01 Albany	35 Oswego
02 Allegany	36 Otsego
03 Broome	37 Putnam
04 Cattaraugus	38 Rensselaer
05 Cayuga	39 Rockland
06 Chautauqua	40 St. Lawrence
07 Chemung	41 Saratoga
08 Chenango	42 Schenectady
09 Clinton	43 Schoharie
10 Columbia	44 Schuyler
11 Cortland	45 Seneca
12 Delaware	46 Steuben
13 Dutchess	47 Suffolk
14 Erie	48 Sullivan
15 Essex	49 Tioga
16 Franklin	50 Tompkins
17 Fulton	51 Ulster
18 Genesee	52 Warren
19 Greene	53 Washington
20 Hamilton	54 Wayne
21 Herkimer	55 Westchester
22 Jefferson	56 Wyoming
23 Lewis	57 Yates
24 Livingston	58 Bronx
25 Madison	59 Kings
26 Monroe	60 Manhattan
27 Montgomery	61 Queens
28 Nassau	62 Richmond
29 Niagara	
30 Oneida	
31 Onondaga	88 Unknown
32 Ontario	
33 Orange	99 Outside NYS
34 Orleans	

Attachment C
Payer Codes

01 Medicare—Fee For Service
02 Medicare—Managed Care
03 Medicaid—Fee For Service
04 Medicaid—Managed Care
05 Blue Cross
06 HMO/Managed Care
07 Other Private Insurance Company
11 Self Pay
19 Other

Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

SEPTAL DEFECTS

ASD

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

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.

VSD

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.

[1]Society of Thoracic Surgeon, Congenital Heart Surgery Database v3.0, used with permission

Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

SEPTAL DEFECTS (CONTINUED)

VSD (continued)

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.

AV Canal

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

2250 Valvuloplasty converted to valve replacement in the same operation, Common atrioventricular valve

2230 Valve replacement, Common atrioventricular valve

AP Window

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.

[1]Society of Thoracic Surgeon, Congenital Heart Surgery Database v3.0, used with permission

Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

SEPTAL DEFECTS (CONTINUED)

Truncus Arteriosus

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

250 Valve replacement, Truncal valve
Replacement of the truncal valve with a prosthetic valve.

2220 Truncus + Interrupted aortic arch repair (IAA) repair

PULMONARY VENOUS ANOMALIES

Partial Anomalous Pulmonary Venous Connection

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 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)

Total Anomalous Pulmonary Venous Connection

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

[1]Society of Thoracic Surgeon, Congenital Heart Surgery Database v3.0, used with permission

Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

COR TRIATRIATUM

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.

PULMONARY VENOUS STENOSIS

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.

SYSTEMIC VENOUS ANOMALIES

Anomalous Systemic Venous Connection / Obstruction

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.

[1]Society of Thoracic Surgeon, Congenital Heart Surgery Database v3.0, used with permission

Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

RIGHT HEART LESIONS

Tetralogy of Fallot

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 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.

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.

[1]Society of Thoracic Surgeon, Congenital Heart Surgery Database v3.0, used with permission

Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

RIGHT HEART LESIONS (CONTINUED)

Pulmonary Atresia

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.

430 Pulmonary atresia - VSD - MAPCA (pseudotruncus) repair
In the presence of MAPCAs, this code implies implies pulmonary unifocalization (multi- or single-stage), repair of VSD (may be intraventricular tunnel or flat patch VSD closure), and placement of an RV-PA conduit.

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 MAPCA(s)
Occlusion, or closing off, of MAPCAs. This may be done with a transcatheter occluding device, usually a coil, or by surgical techniques.

Tricuspid Valve Disease and Ebstein's Anomaly

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, papillary muscle transplocation 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

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 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.

[1]Society of Thoracic Surgeon, Congenital Heart Surgery Database v3.0, used with permission

Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

Tricuspid Valve Disease and Ebstein's Anomaly (continued)

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 valve prosthesis.

500 Valve surgery, Other, Tricuspid
Other tricuspid valve surgery not specified in procedure codes.

RVOT Obstruction, IVS Pulmonary Stenosis

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 hilar bifurcation)
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.

[1]Society of Thoracic Surgeon, Congenital Heart Surgery Database v3.0, used with permission

Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

RIGHT HEART LESIONS (CONTINUED)

RVOT Obstruction, IVS Pulmonary Stenosis (continued)

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.

Pulmonary Valve Disease

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

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 valve prosthesis.

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.

CONDUIT OPERATIONS

Conduit Operations

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.

1172 Conduit placement, Other
Placement of a conduit from any chamber or vessel to any vessel, valved or valveless, not listed elsewhere.

[1]Society of Thoracic Surgeon, Congenital Heart Surgery Database v3.0, used with permission

Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

CONDUIT OPERATIONS (CONTINUED)

Conduit Stenosis / Insufficiency

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).

LEFT HEART LESIONS

Aortic Valve Disease

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

2310 Valvuloplasty converted to valve replacement in the same operation, Aortic – with Ross procedure

2320 Valvuloplasty converted to valve replacement in the same operation, Aortic – with Ross-Konno procedure

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 mechnical prosthesis in a composite conduit.

[1]Society of Thoracic Surgeon, Congenital Heart Surgery Database v3.0, used with permission

Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

LEFT HEART LESIONS (CONTINUED)

Aortic Valve Disease (continued)

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 Manougian and Nicks aortic annular enlargement procedures.

780 Aortic stenosis, Subvalvar, Repair
Subvalvar aortic stenosis repair by a range of techniques 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

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

LEFT HEART LESIONS (CONTINUED)

Aortic Valve Disease (continued)

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.

Sinus of Valsalva Aneurysm

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.

LV to Aorta Tunnel

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.

Mitral Valve Disease

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

840 Mitral stenosis, Supravalvar mitral ring repair
Supravalvar mitral 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.

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

LEFT HEART LESIONS (CONTINUED)

Hypoplastic Left Heart

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 (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.

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 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”.

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

LEFT HEART LESIONS (CONTINUED)

Hypoplastic Left Heart (continued)

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.

CARDIOMYOPATHY

890 Transplant, Heart

Heart transplantation, any technique, allograft or xenograft.

900 Transplant, Heart and lung

Heart and lung (single or double) transplantation.

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

CARDIOMYOPATHY (CONTINUED)

910 Partial left ventriculectomy (LV volume reduction surgery) (Batista)
Wedge resection of LV muscle, with suturing of cut edges together, to reduce LV volume.

PERICARDIAL DISEASE

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.

SINGLE VENTRICLE

950 Fontan, Atrio-pulmonary connection
Fontan-type procedure with atrio-pulmonary connection.

960 Fontan, Atrio-ventricular connection
Fontan-type procedure with atrio-ventricular connection, either direct or with RA-RV conduit, valved or nonvalved.

970 Fontan, TCPC, Lateral tunnel, Fenestrated
Total cavopulmonary connection using an intraatrial lateral tunnel construction, with fenestration.

980 Fontan, TCPC, Lateral tunnel, Nonfenestrated
Total cavopulmonary connection using an intraatrial lateral tunnel construction, with no fenestration.

1000 Fontan, TCPC, External conduit, Fenestrated
Total cavopulmonary connection using an external conduit to connect the infradiaphragmatic systemic venous return to the pulmonary artery, with fenestration.

1010 Fontan, TCPC, External conduit, Nonfenestrated
Total cavopulmonary connection using an external conduit to connect the infradiaphragmatic systemic venous return to the pulmonary artery, with no fenestration.

1025 Fontan revision or conversion (Re-do Fontan)
Revision of a previous Fontan procedure to a total cavopulmonary connection.

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

SINGLE VENTRICLE (CONTINUED)

1030 Fontan, Other
Other Fontan procedure not specified in procedure codes. May include takedown of a Fontan procedure.

2340 Fontan + Atrioventricular valvuloplasty

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.

TRANSPOSITION OF THE GREAT ARTERIES

Congenitally Corrected TGA

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, VSD closure
Repair of congenitally corrected TGA by VSD closure only.

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 otherwised specified in other listed procedure codes.

Transposition of the Great Arteries

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.

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

Transposition of the Great Arteries (continued)

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 effecting 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 effecting 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 pro 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) 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.

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

TRANSPOSITION OF THE GREAT ARTERIES (CONTINUED)

Transposition of the Great Arteries (continued)

1160 REV (continued)
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)

DORV

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

DOLV

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.

THORACIC ARTERIES AND VEINS

Coarctation of Aorta and Aortic Arch Hypoplasia

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.

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.

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

Coarctation of Aorta and Aortic Arch Hypoplasia (continued

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.

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.

Coronary Artery Anomalies

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, 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.

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

THORACIC ARTERIES AND VEINS (CONTINUED)

Interrupted Arch

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.

Patent Ductus Arteriosus

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.

Vascular Rings and Slings

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.

Aortic Aneurysm

1380 Aortic aneurysm repair
Aortic aneurysm repair by any technique.

Aortic Dissection

1390 Aortic dissection repair
Aortic dissection repair by any technique.

THORACIC AND MEDIASTINAL DISEASE

Lung Disease

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.

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

THORACIC AND MEDIASTINAL DISEASE (CONTINUED)

Pectus Excavatum, Carinatum

1430 Pectus repair
Repair of pectus excavatum or carinatum by any technique.

Tracheal Stenosis

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.

ELECTROPHYSIOLOGICAL

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

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.

INTERVENTIONAL CARDIOLOGY PROCEDURES

2500 Cardiovascular catheterization procedure, Diagnostic

2520 Cardiovascular catheterization procedure, Diagnostic, Angiographic data obtained

2550 Cardiovascular catheterization procedure, Diagnostic, Electrophysiology alteration

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

INTERVENTIONAL CARDIOLOGY PROCEDURES (CONTINUED)

2540 Cardiovascular catheterization procedure, Diagnostic, Hemodynamic alteration

2510 Cardiovascular catheterization procedure, Diagnostic, Hemodynamic data obtained

2530 Cardiovascular catheterization procedure, Diagnostic, Transluminal test occlusion

2410 Cardiovascular catheterization procedure, Therapeutic

2670 Cardiovascular catheterization procedure, Therapeutic, Adjunctive therapy

1540 Cardiovascular catheterization procedure, Therapeutic, Balloon dilation

2590 Cardiovascular catheterization procedure, Therapeutic, Balloon valvotomy

1580 Cardiovascular catheterization procedure, Therapeutic, Coil implantation

1560 Cardiovascular catheterization procedure, Therapeutic, Device implantation

2640 Cardiovascular catheterization procedure, Therapeutic, Perforation (establishing interchamber and/or intervessel communication)

2580 Cardiovascular catheterization procedure, Therapeutic, Septostomy

1550 Cardiovascular catheterization procedure, Therapeutic, Stent insertion

2630 Cardiovascular catheterization procedure, Therapeutic, Stent re-dilation

2650 Cardiovascular catheterization procedure, Therapeutic, Transcatheter Fontan completion

2660 Cardiovascular catheterization procedure, Therapeutic, Transcatheter implantation of valve

2680 Cardiovascular electrophysiological catheterization procedure

2690 Cardiovascular electrophysiological catheterization procedure, Therapeutic ablation

PALLIATIVE PROCEDURES

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 (from aorta or to main pulmonary artery)
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).

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

PALLIATIVE PROCEDURES (CONTINUED)

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

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.

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.

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

PALLIATIVE PROCEDURES (CONTINUED)

2330 Superior cavopulmonary anastomosis(es) (Glenn or HemiFontan) + Atrioventricular valvuloplasty

2130 Superior Cavopulmonary anastomosis(es) + PA reconstruction

1710 Palliation, Other
Any other palliative procedure not specifically listed.

MECHANICAL SUPPORT

2360 ECMO cannulation

2370 ECMO decannulation

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

2380 VAD implantation

ANESTHETIC PROCEDURES

2420 Echocardiography procedure, Sedated transesophageal echocardiogram

2430 Echocardiography procedure, Sedated transthoracic echocardiogram

2435 Non-cardiovascular, Non-thoracic procedure on cardiac patient with cardiac anesthesia

2440 Radiology procedure on cardiac patient, Cardiac Computerized Axial Tomography (CT Scan)

2450 Radiology procedure on cardiac patient, Cardiac Magnetic Resonance Imaging (MRI)

2460 Radiology procedure on cardiac patient, Diagnostic radiology

2470 Radiology procedure on cardiac patient, Non-Cardiac Computerized Tomography (CT) on cardiac patient

2480 Radiology procedure on cardiac patient, Non-cardiac Magnetic Resonance Imaging (MRI) on cardiac patient

2490 Interventional radiology procedure on cardiac patient

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

MISCELLANEOUS PROCEDURES

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.

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Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

MISCELLANEOUS PROCEDURES (CONTINUED)

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.

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.

[1]Society of Thoracic Surgeon, Congenital Heart Surgery Database v3.0, used with permission

Attachment D
Pediatric CSRS Cardiac Procedure Codes[1]

MISCELLANEOUS PROCEDURES (CONTINUED)

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.

[1]Society of Thoracic Surgeon, Congenital Heart Surgery Database v3.0, used with permission

Attachment E
Congenital Cardiac Diagnosis Codes[1]

10 PFO
Small interatrial communication in the region of the foramen ovale characterized by no deficiency of the septum primum and a normal limbus with no deficiency of the septum secundum.

20 ASD, Secundum
An ASD confined to the region of the fossa ovalis; its most common etiology is a deficiency of the septum primum, but deficiency of the limbus or septum secundum may also contribute.

30 ASD, Sinus venosus
Indicate if the patient has the diagnosis of “ASD, Sinus venosus”. An “ASD, Sinus venosus” is defined as a defect with a vena cava or pulmonary vein (or veins) that overrides the atrial septum or the superior interatrial fold (septum secundum) 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 true atrial septum.

40 ASD, Coronary sinus
Deficiency of the wall (sinus septum) separating the left atrium from the coronary sinus, often allowing blood to shunt from the left atrium to the right atrium via the coronary sinus ostium. May or may not be associated with a persistent left superior vena cava.

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) (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.

[1]*Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 (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.)

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 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).

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 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.")

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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; 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 semilunar 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 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.)

[1]*Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 PAVSD 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, VSDMAPCA (pseudotruncus) should be used.

350 Pulmonary atresia, VSD-MAPCA (pseudotruncus)
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 patients 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 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 (valvular 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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.).

[1]*Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Attachment E
Congenital Cardiac Diagnosis Codes[1]

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.]), 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 know 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.

[1]*Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Attachment E
Congenital Cardiac Diagnosis Codes[1]

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.

[1]*Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 neoaorta 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 annuloaortic 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.

[1]*Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Attachment E
Congenital Cardiac Diagnosis Codes[1]

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, myxomatomous 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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), myxomatomous 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 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 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
Single morphologically left ventricle (smooth internal walls, lack chordal attachments of AV valves to the rudimentary septal surface) that receives both atrioventricular valves.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

800 Single ventricle, DIRV
Single morphologically right ventricle (more heavily trabeculated, generally have chordal attachments of AV valve to the septal surfaces) that receives both atrioventricular valves.

810 Single ventricle, Mitral atresia
Single ventricle anomalies with mitral atresia. May also be associated with double outlet right ventricle, congenitally corrected transposition, pulmonary atresia, or pulmonary stenosis.

820 Single ventricle, Tricuspid atresia
Single ventricle anomalies with tricuspid atresia. May also be associated with complete transposition of the great arteries, congenitally corrected transposition of the great arteries, pulmonary atresia, pulmonary stenosis, subaortic stenosis, or ventricular septal defect (small or large).

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.

840 Single ventricle, Heterotaxia syndrome
Visceral heterotaxy syndrome is literally defined as a pattern of anatomic organization of the thoracic and abdominal organs that is neither the expected usual or normal arrangement (so-called situs solitus) nor complete situs inversus (the unusual or mirror-image arrangement of normal). If asymmetry of the thoracic and abdominal viscera is the usual or normal situation, visceral heterotaxy syndrome includes patients with an unusual degree of thoracic and abdominal visceral symmetry. This broad term includes patients with a wide variety of complex cardiac lesions. One way to impose order on this diverse group of cardiac lesions is to stratify them according to the morphology of the atrial appendages. In atrial appendage isomerism, both atrial appendages are similar rather than displaying their usual distinctive morphology. Right or left atrial appendage isomerism exists when both atria have right or left atrial appendage morphologic characteristics, respectively. Right atrial appendage isomerism is frequently associated with bilaterally trilobed lungs (each with short bronchi) and asplenia. Left atrial appendage isomerism frequently is associated with bilaterally bilobed lungs (each with long bronchi) and polysplenia. Many types of anomalies of systemic venous connection are frequently associated with heterotaxy syndrome.

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.

[1]*Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 functionally univentricular heart is defined as a spectrum of cardiac malformations in which entire ventricular mass is functionally univentricular; in other words, whenever only one ventricle is capable, for whatever reason, of supporting either the systemic or the pulmonary circulation.) The consensus of the EACTS and STS Congenital Heart Surgery Database Committees is that the nomenclature proposal for single ventricle hearts would encompass hearts with double inlet atrioventricular connection (both double inlet left ventricle [DILV] and double inlet right ventricle [DIRV]), hearts with absence of one atrioventricular connection (mitral atresia and tricuspid atresia), hearts with a common atrioventricular valve and only one completely well-developed ventricle (unbalanced common atrioventricular canal defect), hearts with only one fully well-developed ventricle and heterotaxia syndrome (single ventricle heterotaxia syndrome), and finally other rare forms of univentricular hearts that do not fit in one of the specified major categories. In the version of the IPCCC derived from the nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and the STS, patients classified in this section of the nomenclature, therefore, include all those who would be coded using the Short List for “Single Ventricle”, specifically: (1) Single ventricle; (2) Single ventricle, DILV; (3) Single ventricle, DIRV; (4) Single ventricle, Heterotaxia syndrome; (5) Single ventricle, Mitral atresia; (6) Single ventricle, Tricuspid atresia; (7) Single ventricle, Unbalanced AV canal. (Despite the recognition that hypoplastic left heart syndrome is a common form of functionally univentricular heart, with a single or dominant ventricle of right ventricular morphology, the EACTS-STS version of the IPCCC includes an entirely separate section for consideration of hypoplastic left heart syndrome. Also, it is recognized that a considerable variety of other structural cardiac malformations, such as pulmonary atresia with intact ventricular septum, biventricular hearts with straddling atrioventricular valves, and some complex forms of double outlet right ventricle (DORV), may at times be best managed in a fashion similar to that which is used to treat other functionally univentricular hearts. Nomenclature for description of those entities, however, is not included in this Single Ventricle section of the EACTS-STS version of the IPCCC.)} [1] [1]. 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.

[1]*Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 ventriculoarterial 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 ventriculoarterial 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.)

[1] *Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Attachment E
Congenital Cardiac Diagnosis Codes[1]

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).

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 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 from aorta (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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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.

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.)}

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 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.

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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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, neoplastm, etc.) which may result in death or lung or heart-lung transplant.

1150 Pectus
Pectus excavatum is a chest wall deformity in which the sternum is depressed. Pectus carinatum is a protrusion of the sternum.

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).

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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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.

1180 Arrhythmia
Any cardiac rhythm other than normal sinus rhythm.

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.

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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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.

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 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 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).

[1]*Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Attachment E
Congenital Cardiac Diagnosis Codes[1]

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 valvular (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

[1]*Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

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 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

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.

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, femorial vein, iliac artery, brachial artery, etc.

7000 Normal heart
Normal heart.

7777 Miscellaneous, Other
Any disease (congenital or acquired) not specifically delineated in other diagnostic codes.

4010 Status post - PFO, Primary closure
Status post - Suture closure of patent foramen ovale (PFO).

4020 Status post - ASD repair, Primary closure
Status post - Suture closure of secundum (most frequently), coronary sinus, sinus venosus or common atrium ASD.

4030 Status post - ASD repair, Patch
Status post - Patch closure (using any type of patch material) of secundum, coronary sinus, or sinus venosus ASD.

4040 Status post - ASD repair, Device
Status post - Closure of any type ASD (including PFO) using a device.

6110 Status post - ASD repair, Patch + PAPVC repair

4050 Status post - ASD, Common atrium (single atrium), Septation
Status post - Septation of common (single) atrium using any type patch material.

4060 Status post - ASD creation/enlargement
Status post - 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.

4070 Status post - ASD partial closure
Status post - Intentional partial closure of any type ASD (partial suture or fenestrated patch closure).

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

4080 Status post - Atrial septal fenestration
Status post - 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.

4085 Status post - Atrial fenestration closure
Status post - Closure of previously created atrial fenestration using any method including device, primary suture, or patch.

4100 Status post - VSD repair, Primary closure
Status post - Suture closure of any type VSD.

4110 Status post - VSD repair, Patch
Status post - Patch closure (using any type of patch material) of any type VSD.

4120 Status post - VSD repair, Device
Status post - Closure of any type VSD using a device.

4130 Status post - VSD, Multiple, Repair
Status post - 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.

4140 Status post - VSD creation/enlargement
Status post - Creation of a ventricular septal defect or enlargement of an existing ventricular septal defect.

4150 Status post - Ventricular septal fenestration
Status post - 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.

4170 Status post - AVC (AVSD) repair, Complete (CAVSD)
Status post - Repair of complete AV canal (AVSD) using one- or two-patch or other technique, with or without mitral valve cleft repair.

4180 Status post - AVC (AVSD) repair, Intermediate (Transitional)
Status post - 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.

4190 Status post - AVC (AVSD) repair, Partial (Incomplete) (PAVSD)
Status post - Repair of partial AV canal defect (primum ASD), any technique, with or without repair of cleft mitral valve.

6300 Status post - Valvuloplasty, Common atrioventricular valve

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

6250 Status post - Valvuloplasty converted to valve replacement in the same operation, Common atrioventricular valve

6230 Status post - Valve replacement, Common atrioventricular valve

4210 Status post - AP window repair
Status post - Repair of AP window using one- or two-patch technique with cardiopulmonary bypass; or, without cardiopulmonary bypass, using transcatheter device or surgical closure.

4220 Status post - Pulmonary artery origin from ascending aorta (hemitruncus) repair
Status post - Repair of pulmonary artery origin from the ascending aorta by direct reimplantation, autogenous flap, or conduit, with or without use of cardiopulmonary bypass.

4230 Status post - Truncus arteriosus repair
Status post - 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).

4240 Status post - Valvuloplasty, Truncal valve
Status post - Truncal valve repair, any type.

6290 Status post - Valvuloplasty converted to valve replacement in the same operation, Truncal valve

4250 Status post - Valve replacement, Truncal valve
Status post - Replacement of the truncal valve with a prosthetic valve.

6220 Status post - Truncus + Interrupted aortic arch repair (IAA) repair

4260 Status post - PAPVC repair
Status post - 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.

4270 Status post - PAPVC, Scimitar, Repair
Status post - In scimitar syndrome, PAPVC repair also 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. 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.

6120 Status post - PAPVC repair, Baffle redirection to left atrium with systemic vein translocation (Warden) (SVC sewn to right atrial appendage)

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

4280 Status post - TAPVC repair
Status post - 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.

6200 Status post - TAPVC repair + Shunt - systemic-to-pulmonary

4290 Status post - Cor triatriatum repair
Status post - 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.

4300 Status post - Pulmonary venous stenosis repair
Status post - Repair of pulmonary venous stenosis, whether congenital or acquired. Repair can be accomplished with a variety of approaches: sutureless, patch venoplasty, stent placement, etc.

4310 Status post - Atrial baffle procedure (non-Mustard, non-Senning)
Status post - 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.

4330 Status post - Anomalous systemic venous connection repair
Status post - 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).

4340 Status post - Systemic venous stenosis repair
Status post - 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.

4350 Status post - TOF repair, No ventriculotomy
Status post - 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 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.

4360 Status post - TOF repair, Ventriculotomy, Nontransanular patch
Status post - 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

4370 Status post - TOF repair, Ventriculotomy, Transanular patch
Status post - 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.

4380 Status post - TOF repair, RV-PA conduit
Status post - 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.

4390 Status post - TOF - AVC (AVSD) repair
Status post - 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.

4400 Status post - TOF - Absent pulmonary valve repair
Status post - 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.

4420 Status post - Pulmonary atresia - VSD (including TOF, PA) repair
Status post - 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.

4430 Status post - Pulmonary atresia - VSD - MAPCA (pseudotruncus) repair
Status post - In the presence of MAPCAs, this code implies pulmonary unifocalization (multi- or single-stage), repair of VSD (may be intraventricular tunnel or flat patch VSD closure), and placement of an RV-PA conduit.

4440 Status post - Unifocalization MAPCA(s)
Status post - 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.

4450 Status post - Occlusion MAPCA(s)
Status post - Occlusion, or closing off, of MAPCAs. This may be done with a transcatheter occluding device, usually a coil, or by surgical techniques.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

4460 Status post - Valvuloplasty, Tricuspid
Status post - Reconstruction of the tricuspid valve may include but not be limited to a wide range of techniques including: leaflet patch extension, artificial chordae placement, papillary muscle transplocation 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, pursestring 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.

6280 Status post - Valvuloplasty converted to valve replacement in the same operation, Tricuspid

4465 Status post - Ebstein's repair
Status post - 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 addressed with surgical ablation procedures. These procedures should be entered as separate procedure codes.

4470 Status post - Valve replacement, Tricuspid (TVR)
Status post - Replacement of the tricuspid valve with a prosthetic valve.

4480 Status post - Valve closure, Tricuspid (exclusion, univentricular approach)
Status post - 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.

4490 Status post - Valve excision, Tricuspid (without replacement)
Status post - Excision of the tricuspid valve without placement of a valve prosthesis.

4500 Status post - Valve surgery, Other, Tricuspid
Status post - Other tricuspid valve surgery not specified in procedure codes.

4510 Status post - RVOT procedure
Status post - 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.

4520 Status post - 1 1/2 ventricular repair
Status post - Partial biventricular repair; includes intracardiac repair with bidirectional cavopulmonary anastomosis to volume unload a small ventricle or poorly functioning ventricle.

4530 Status post - PA, reconstruction (plasty), Main (trunk)
Status post - 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

4540 Status post - PA, reconstruction (plasty), Branch, Central (within the hilar bifurcation)
Status post - 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.

4550 Status post - PA, reconstruction (plasty), Branch, Peripheral (at or beyond the hilar bifurcation)
Status post - 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.

4570 Status post - DCRV repair
Status post - 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.

4590 Status post - Valvuloplasty, Pulmonic
Status post - Valvuloplasty of the pulmonic valve may include a range of techniques including but not limited to: valvotomy with or without bypass, commissurotomy, and valvuloplasty.

6270 Status post - Valvuloplasty converted to valve replacement in the same operation, Pulmonic

4600 Status post - Valve replacement, Pulmonic (PVR)
Status post - 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.

4630 Status post - Valve excision, Pulmonary (without replacement)
Status post - Excision of the pulmonary valve without placement of a valve prosthesis.

4640 Status post - Valve closure, Semilunar
Status post - Closure of a semilunar valve (pulmonic or aortic) by any technique.

4650 Status post - Valve surgery, Other, Pulmonic
Status post - Other pulmonic valve surgery not specified in procedure codes.

4610 Status post - Conduit placement, RV to PA
Status post - Placement of a conduit, any type, from RV to PA.

4620 Status post - Conduit placement, LV to PA
Status post - Placement of a conduit, any type, from LV to PA.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

5774 Status post - Conduit placement, Ventricle to aorta
Status post - Placement of a conduit from the right or left ventricle to the aorta.

5772 Status post - Conduit placement, Other
Status post - Placement of a conduit from any chamber or vessel to any vessel, valved or valveless, not listed elsewhere.

4580 Status post - Conduit reoperation
Status post - 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).

4660 Status post - Valvuloplasty, Aortic
Status post - 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).

6240 Status post - Valvuloplasty converted to valve replacement in the same operation, Aortic

6310 Status post - Valvuloplasty converted to valve replacement in the same operation, Aortic – with Ross procedure

6320 Status post - Valvuloplasty converted to valve replacement in the same operation, Aortic – with Ross-Konno procedure

4670 Status post - Valve replacement, Aortic (AVR)
Status post - 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.

4680 Status post - Valve replacement, Aortic (AVR), Mechanical
Status post - Replacement of the aortic valve with a mechanical prosthetic valve.

4690 Status post - Valve replacement, Aortic (AVR), Bioprosthetic
Status post - Replacement of the aortic valve with a bioprosthetic prosthetic valve.

4700 Status post - Valve replacement, Aortic (AVR), Homograft
Status post - Replacement of the aortic valve with a homograft prosthetic valve.

4715 Status post - Aortic root replacement, Bioprosthetic
Status post - 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

4720 Status post - Aortic root replacement, Mechanical
Status post - Replacement of the aortic root (that portion of the aorta attached to the heart; it gives rise to the coronary arteries) with a mechnical prosthesis in a composite conduit.

4730 Status post - Aortic root replacement, Homograft
Status post - Replacement of the aortic root (that portion of the aorta attached to the heart; it gives rise to the coronary arteries) with a homograft.

4735 Status post - Aortic root replacement, Valve sparing
Status post - 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).

4740 Status post - Ross procedure
Status post - Replacement of the aortic valve with a pulmonary autograft and replacement of the pulmonary valve with a homograft conduit.

4750 Status post - Konno procedure
Status post - 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.

4760 Status post - Ross-Konno procedure
Status post - 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.

4770 Status post - Other annular enlargement procedure
Status post - 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 Manougian and Nicks aortic annular enlargement procedures.

4780 Status post - Aortic stenosis, Subvalvar, Repair
Status post - Subvalvar aortic stenosis repair by a range of techniques 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.

6100 Status post - Aortic stenosis, Subvalvar, Repair, With myectomy for IHSS

4790 Status post - Aortic stenosis, Supravalvar, Repair
Status post - 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

4800 Status post - Valve surgery, Other, Aortic
Status post - Other aortic valve surgery not specified in other procedure codes.

4810 Status post - Sinus of Valsalva, Aneurysm repair
Status post - 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.

4820 Status post - LV to aorta tunnel repair
Status post - 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.

4830 Status post - Valvuloplasty, Mitral
Status post - 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.

6260 Status post - Valvuloplasty converted to valve replacement in the same operation, Mitral

4840 Status post - Mitral stenosis, Supravalvar mitral ring repair
Status post - Supravalvar mitral ring repair.

4850 Status post - Valve replacement, Mitral (MVR)
Status post - Replacement of mitral valve with prosthetic valve, any kind, in suprannular or annular position.

4860 Status post - Valve surgery, Other, Mitral
Status post - Other mitral valve surgery not specified in procedure codes.

4870 Status post - Norwood procedure
Status post - 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 (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

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

4880 Status post - HLHS biventricular repair
Status post - 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.

6160 Status post - Hybrid Approach "Stage 1", Application of RPA & LPA bands
Status post - 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”.

6170 Status post - Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA)
Status post - 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”.

6180 Status post - Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA) + application of RPA & LPA bands
Status post - 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”.

6140 Status post - Hybrid approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Aortic arch repair (Norwood [Stage 1] + Superior Cavopulmonary anastomosis(es) + PA Debanding)
Status post - 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.

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

6150 Status post - Hybrid approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Without aortic arch repair
Status post - 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.

1590 Status post - Transplant, Heart
Status post - Heart transplantation, any technique, allograft or xenograft.

1610 Status post - Transplant, Heart and lung
Status post - Heart and lung (single or double) transplantation.

4910 Status post - Partial left ventriculectomy (LV volume reduction surgery) (Batista)
Status post - Wedge resection of LV muscle, with suturing of cut edges together, to reduce LV volume.

4920 Status post - Pericardial drainage procedure
Status post - 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).

4930 Status post - Pericardiectomy
Status post - Surgical removal of the pericardium.

4940 Status post - Pericardial procedure, Other
Status post - 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.

4950 Status post - Fontan, Atrio-pulmonary connection
Status post - Fontan-type procedure with atrio-pulmonary connection.

4960 Status post - Fontan, Atrio-ventricular connection
Status post - Fontan-type procedure with atrio-ventricular connection, either direct or with RA-RV conduit, valved or nonvalved.

4970 Status post - Fontan, TCPC, Lateral tunnel, Fenestrated
Status post - Total cavopulmonary connection using an intraatrial lateral tunnel construction, with fenestration.

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Congenital Cardiac Diagnosis Codes[1]

4980 Status post - Fontan, TCPC, Lateral tunnel, Nonfenestrated
Status post - Total cavopulmonary connection using an intraatrial lateral tunnel construction, with no fenestration.

5000 Status post - Fontan, TCPC, External conduit, Fenestrated
Status post - Total cavopulmonary connection using an external conduit to connect the infradiaphragmatic systemic venous return to the pulmonary artery, with fenestration.

5010 Status post - Fontan, TCPC, External conduit, Nonfenestrated
Status post - Total cavopulmonary connection using an external conduit to connect the infradiaphragmatic systemic venous return to the pulmonary artery, with no fenestration.

5025 Status post - Fontan revision or conversion (Re-do Fontan)
Status post - Revision of a previous Fontan procedure to a total cavopulmonary connection.

5030 Status post - Fontan, Other
Status post - Other Fontan procedure not specified in procedure codes. May include takedown of a Fontan procedure.

6340 Status post - Fontan + Atrioventricular valvuloplasty

5035 Status post - Ventricular septation
Status post - 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.

5050 Status post - Congenitally corrected TGA repair, Atrial switch and ASO (double switch)
Status post - 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.

5060 Status post - Congenitally corrected TGA repair, Atrial switch and Rastelli
Status post - 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.

5070 Status post - Congenitally corrected TGA repair, VSD closure
Status post - Repair of congenitally corrected TGA by VSD closure only.

5080 Status post - Congenitally corrected TGA repair, VSD closure and LV to PA conduit
Status post - Repair of congenitally corrected TGA by VSD closure and placement of an LV-to-PA conduit.

5090 Status post - Congenitally corrected TGA repair, Other
Status post - Any procedures for correction of CCTGA not otherwised specified in other listed procedure codes.

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Congenital Cardiac Diagnosis Codes[1]

5110 Status post - Arterial switch operation (ASO)
Status post - 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.

5120 Status post - Arterial switch operation (ASO) and VSD repair
Status post - 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.

5123 Status post - Arterial switch procedure + Aortic arch repair
Status post - 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.

5125 Status post - Arterial switch procedure and VSD repair + Aortic arch repair
Status post - 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.

5130 Status post - Senning
Status post - Atrial baffle procedure for rerouting of venous flow in TGA effecting 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.

5140 Status post - Mustard
Status post - Atrial baffle procedure for rerouting of venous flow in TGA effecting 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.

5145 Status post - Atrial baffle procedure, Mustard or Senning revision
Status post - Revision of a previous atrial baffle procedure (either Mustard or Senning), for any reason (e.g., obstruction, baffle leak).

5150 Status post - Rastelli
Status post - 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.

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Congenital Cardiac Diagnosis Codes[1]

5160 Status post - REV
Status post - 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) 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.

6190 Status post - Aortic root translocation over left ventricle (Including Nikaidoh procedure)

6210 Status post - TGA, Other procedures (Kawashima, LV-PA conduit, other)

5180 Status post - DORV, Intraventricular tunnel repair
Status post - Repair of DORV using a tunnel closure of the VSD to the aortic valve. This also includes the posterior straight tunnel repair of Kawashima

5200 Status post - DOLV repair
Status post - 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.

5210 Status post - Coarctation repair, End to end
Status post - Repair of coarctation of aorta by excision of the coarctation segment and end-to-end circumferential anastomosis of the aorta.

5220 Status post - Coarctation repair, End to end, Extended
Status post - 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.

5230 Status post - Coarctation repair, Subclavian flap
Status post - 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.

5240 Status post - Coarctation repair, Patch aortoplasty
Status post - Repair of coarctation of the aorta by incising the coarctation site with placement of a patch sutured in place longitudinally along the aortotomy edge.

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Congenital Cardiac Diagnosis Codes[1]

5250 Status post - Coarctation repair, Interposition graft
Status post - 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.

5260 Status post - Coarctation repair, Other
Status post - 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.

5275 Status post - Coarctation repair + VSD repair
Status post - Coarctation of aorta repair, any technique, and simultaneous VSD repair, any type VSD, any type repair.

5280 Status post - Aortic arch repair
Status post - Aortic arch repair, any technique.

5285 Status post - Aortic arch repair + VSD repair
Status post - Aortic arch repair, any technique, and simultaneous VSD repair, any type VSD, any type repair. This includes repair of IAA with VSD.

5290 Status post - Coronary artery fistula ligation
Status post - 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).

5291 Status post - Anomalous origin of coronary artery from pulmonary artery repair
Status post - Repair of anomalous origin of the coronary artery (any) from the pulmonary artery, by any technique (ligation, translocation with aortic implantation, Takeuchi operation, bypass graft). If additional technique information may be supplied by another procedure code, please list separately (for example, bypass graft).

5300 Status post - Coronary artery bypass
Status post - 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.).

5305 Status post - Anomalous aortic origin of coronary artery from aorta (AAOCA) repair

5310 Status post - Coronary artery procedure, Other
Status post - Any coronary artery procedure not specifically listed.

5320 Status post - Interrupted aortic arch repair
Status post - Repair of interrupted aortic arch (any type) by any technique (direct anastomosis, prosthetic graft, etc). Does not include repair of IAA-VSD.

5330 Status post - PDA closure, Surgical
Status post - Closure of a PDA by any surgical technique (ligation, division, clip) using any approach (i.e., thoracotomy, thoracoscopic, etc).

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Congenital Cardiac Diagnosis Codes[1]

5340 Status post - PDA closure, Device
Status post - Closure of a PDA by device using transcatheter techniques.

5360 Status post - Vascular ring repair
Status post - Repair of vascular ring (any type, except pulmonary artery sling) by any technique.

5365 Status post - Aortopexy
Status post - 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).

5370 Status post - Pulmonary artery sling repair
Status post - Pulmonary artery sling repair by any technique.

5380 Status post - Aortic aneurysm repair
Status post - Aortic aneurysm repair by any technique.

5390 Status post - Aortic dissection repair
Status post - Aortic dissection repair by any technique.

5400 Status post - Lung biopsy
Status post - Lung biopsy, any technique.

1600 Status post - Transplant, Lung(s)
Status post - Lung or lobe transplantation of any type.

5420 Status post - Lung procedure, Other
Status post - 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.

5430 Status post - Pectus repair
Status post - Repair of pectus excavatum or carinatum by any technique.

5440 Status post - Tracheal procedure
Status post - 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.

5450 Status post - Pacemaker implantation, Permanent
Status post - 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).

5460 Status post - Pacemaker procedure
Status post - 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.

6350 Status post - Explantation of pacing system

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Congenital Cardiac Diagnosis Codes[1]

5470 Status post - ICD (AICD) implantation
Status post - Implantation of an (automatic) implantable cardioverter defibrillator system.

5480 Status post - ICD (AICD) ([automatic] implantable cardioverter defibrillator) procedure
Status post - Any revision to a previously placed AICD including revisions to leads, pads, generators, pockets. This may include explantation procedures as well.

5490 Status post - Arrhythmia surgery - atrial, Surgical Ablation
Status post - Surgical ablation (any type) of any atrial arrhythmia.

5500 Status post - Arrhythmia surgery - ventricular, Surgical Ablation
Status post - Surgical ablation (any type) of any ventricular arrhythmia.

6500 Status post - Cardiovascular catheterization procedure, Diagnostic

6520 Status post - Cardiovascular catheterization procedure, Diagnostic, Angiographic data obtained

6550 Status post - Cardiovascular catheterization procedure, Diagnostic, Electrophysiology alteration

6540 Status post - Cardiovascular catheterization procedure, Diagnostic, Hemodynamic alteration

6510 Status post - Cardiovascular catheterization procedure, Diagnostic, Hemodynamic data obtained

6530 Status post - Cardiovascular catheterization procedure, Diagnostic, Transluminal test occlusion

6410 Status post - Cardiovascular catheterization procedure, Therapeutic

6670 Status post - Cardiovascular catheterization procedure, Therapeutic, Adjunctive therapy

6570 Status post - Cardiovascular catheterization procedure, Therapeutic, Balloon dilation

6590 Status post - Cardiovascular catheterization procedure, Therapeutic, Balloon valvotomy

6600 Status post - Cardiovascular catheterization procedure, Therapeutic, Coil implantation

6610 Status post - Cardiovascular catheterization procedure, Therapeutic, Device implantation

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Congenital Cardiac Diagnosis Codes[1]

6640 Status post - Cardiovascular catheterization procedure, Therapeutic, Perforation (establishing interchamber and/or intervessel communication)

6580 Status post - Cardiovascular catheterization procedure, Therapeutic, Septostomy

6620 Status post - Cardiovascular catheterization procedure, Therapeutic, Stent insertion

6630 Status post - Cardiovascular catheterization procedure, Therapeutic, Stent re-dilation

6650 Status post - Cardiovascular catheterization procedure, Therapeutic, Transcatheter Fontan completion

6660 Status post - Cardiovascular catheterization procedure, Therapeutic, Transcatheter implantation of valve

6680 Status post - Cardiovascular electrophysiological catheterization procedure

6690 Status post - Cardiovascular electrophysiological catheterization procedure, Therapeutic ablation

5590 Status post - Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS)
Status post - 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).

5600 Status post - Shunt, Systemic to pulmonary, Central (from aorta or to main pulmonary artery)
Status post - 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).

5610 Status post - Shunt, Systemic to pulmonary, Other
Status post - 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.

5630 Status post - Shunt, Ligation and takedown
Status post - Takedown of any shunt.

6095 Status post - Shunt, Reoperation

5640 Status post - PA banding (PAB)
Status post - Placement of a pulmonary artery band, any type.

5650 Status post - PA debanding
Status post - Debanding of pulmonary artery. Please list separately any pulmonary artery reconstruction required.

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Congenital Cardiac Diagnosis Codes[1]

5660 Status post - Damus-Kaye-Stansel procedure (DKS) (creation of AP anastomosis without arch reconstruction)
Status post - In the Damus-Kaye-Stansel procedure the proximal transected main pulmonary artery is connected by varying techniques to the aorta.

5670 Status post - Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn)
Status post - Superior vena cava to pulmonary artery anastomosis allowing flow to both pulmonary arteries with an end-to-side superior vena-to-pulmonary artery anastomosis.

5680 Status post - Glenn (unidirectional cavopulmonary anastomosis) (unidirectional Glenn)
Status post - Superior vena cava to ipsilateral pulmonary artery anastomosis (i.e., LSVC to LPA, RSVC to RPA).

5690 Status post - Bilateral bidirectional cavopulmonary anastomosis (BBDCPA) (bilateral bidirectional Glenn)
Status post - Bilateral superior vena cava-to-pulmonary artery anastomoses (requires bilateral SVCs).

5700 Status post - HemiFontan
Status post - 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.

6330 Status post - Superior cavopulmonary anastomosis(es) (Glenn or HemiFontan) + Atrioventricular valvuloplasty

6130 Status post - Superior Cavopulmonary anastomosis(es) + PA reconstruction

5710 Status post - Palliation, Other
Status post - Any other palliative procedure not specifically listed.

6360 Status post - ECMO cannulation

6370 Status post - ECMO decannulation

5910 Status post - ECMO procedure
Status post - Any ECMO procedure (cannulation, decannulation, etc.).

5900 Status post - Intraaortic balloon pump (IABP) insertion
Status post - Insertion of intraaortic balloon pump by any technique.

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Congenital Cardiac Diagnosis Codes[1]

5920 Status post - Right/left heart assist device procedure
Status post - Any right, left, or biventricular assist device procedure (placement, removal etc.).

6390 Status post - VAD explantation

6380 Status post - VAD implantation

6420 Status post - Echocardiography procedure, Sedated transesophageal echocardiogram

6430 Status post - Echocardiography procedure, Sedated transthoracic echocardiogram

6435 Status post - Non-cardiovascular, Non-thoracic procedure on cardiac patient with cardiac anesthesia

6440 Status post - Radiology procedure on cardiac patient, Cardiac Computerized Axial Tomography (CT Scan)

6450 Status post - Radiology procedure on cardiac patient, Cardiac Magnetic Resonance Imaging (MRI)

6460 Status post - Radiology procedure on cardiac patient, Diagnostic radiology

6470 Status post - Radiology procedure on cardiac patient, Non-Cardiac Computerized Tomography (CT) on cardiac patient

6480 Status post - Radiology procedure on cardiac patient, Non-cardiac Magnetic Resonance Imaging (MRI) on cardiac patient

6490 Status post - Interventional radiology procedure on cardiac patient

5720 Status post - Aneurysm, Ventricular, Right, Repair
Status post - Repair of right ventricular aneurysm, any technique.

5730 Status post - Aneurysm, Ventricular, Left, Repair
Status post - Repair of left ventricular aneurysm, any technique.

5740 Status post - Aneurysm, Pulmonary artery, Repair
Status post - Repair of pulmonary artery aneurysm, any technique.

5760 Status post - Cardiac tumor resection
Status post - Resection of cardiac tumor, any type.

5780 Status post - Pulmonary AV fistula repair/occlusion
Status post - Repair or occlusion of a pulmonary arteriovenous fistula.

[1]*Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Attachment E
Congenital Cardiac Diagnosis Codes[1]

5790 Status post - Ligation, Pulmonary artery
Status post - Ligation or division of the pulmonary artery. Most often performed as a secondary procedure.

5802 Status post - Pulmonary embolectomy, Acute pulmonary embolus
Status post - Acute pulmonary embolism (clot) removal, through catheter or surgery.

5804 Status post - Pulmonary embolectomy, Chronic pulmonary embolus
Status post - Chronic pulmonary embolism (clot) removal, through catheter or surgery.

5810 Status post - Pleural drainage procedure
Status post - Pleural drainage procedure via thoracocentesis, tube thoracostomy, or open surgical drainage.

5820 Status post - Pleural procedure, Other
Status post - Other pleural procedures not specifically listed; may include pleurodesis (mechanical, talc, antibiotic or other), among others.

5830 Status post - Ligation, Thoracic duct
Status post - Ligation of the thoracic duct; most commonly for persistent chylothorax.

5840 Status post - Decortication
Status post - Decortication of the lung by any technique.

5850 Status post - Esophageal procedure
Status post - Any procedure performed on the esophagus.

5860 Status post - Mediastinal procedure
Status post - Any non-cardiovascular mediastinal procedure not otherwise listed.

5870 Status post - Bronchoscopy
Status post - Bronchoscopy, rigid or flexible, for diagnostic, biopsy, or treatment purposes (laser, stent, dilation, lavage).

5880 Status post - Diaphragm plication
Status post - Plication of the diaphragm; most often for diaphragm paralysis due to phrenic nerve injury.

5890 Status post - Diaphragm procedure, Other
Status post - Any diaphragm procedure not specifically listed.

5930 Status post - VATS (video-assisted thoracoscopic surgery)
Status post - 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).

5940 Status post - Minimally invasive procedure
Status post - 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).

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Attachment E
Congenital Cardiac Diagnosis Codes[1]

5950 Status post - Bypass for noncardiac lesion
Status post - 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).

5960 Status post - Delayed sternal closure
Status post - 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.

5970 Status post - Mediastinal exploration
Status post - Mediastinal exploration, most often for postoperative control of bleeding or tamponade, but may be exploration to assess mediastinal mass, etc.

5980 Status post - Sternotomy wound drainage
Status post - Drainage of the sternotomy wound.

5990 Status post - Thoracotomy, Other
Status post - Any procedure performed through a thoracotomy incision not otherwise listed.

6000 Status post - Cardiotomy, Other
Status post - Any procedure involving an incision in the heart that is not otherwise listed.

6010 Status post - Cardiac procedure, Other
Status post - Any cardiac procedure, bypass or non-bypass, that is not otherwise listed.

6020 Status post - Thoracic and/or mediastinal procedure, Other
Status post - Any thoracic and/or mediastinal procedure not otherwise listed.

6030 Status post - Peripheral vascular procedure, Other
Status post - Any peripheral vascular procedure; may include procedures such as femoral artery repair, iliac artery repair, etc.

6040 Status post - Miscellaneous procedure, Other
Status post - Any miscellaneous procedure not otherwise listed.

6050 Status post - Organ procurement
Status post - Procurement of an organ for transplant (most likely, heart, lungs, or heart and lungs).

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