

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

2012 Discharges
Clarification Document August 2012

**Cardiac Surgery Report, Adult
(Age 18 and Over)**

**Instructions and Data Element
Definitions
Form DOH-2254a**

Note: This document contains several clarifications pertaining to the 2012 CSRS data reporting system. New text is presented as underlined. Any deleted text is noted as ~~strike-through~~.

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Revision Highlights and Coding Clarifications

Deleted Data Elements

The following data elements will no longer be collected effective January 2012.

- Angina Type
- Major Event - Stroke over 24 hours.

New Data Elements

The following data elements have been added to CSRS effective January 2012. Complete data element definitions are located in the main body of this document.

- **Diagnosis Code #3**
- **Interventional Cardiologist, NYS physician license number** (with procedure codes 640-643 and 711 only)
- **Cardiac Presentation on Admission**
- **TIA, Only Cerebrovascular Risk**
- **Diabetes Therapy**

Revised Data Elements

The following revisions are effective January 2012.

Complete data element definitions are located in the main body of this document.

Congenital Diagnosis Codes are now reported when “the patient had a congenital defect repair either in conjunction with, or as the primary surgical procedure.” The diagnosis code list has been replaced.

Induction of Anesthesia (time) is being replaced with “Time of First Skin Incision.”

Pre-Op Beta Blocker is now limited to a 24 hour time frame and the information concerning contraindications has been revised.

Intra-operative Blood Transfusion has a revised definition.

Surgical Priority has a revised definition and an additional response category for “Emergent Salvage.”

Weight definition has been modified to indicate the weight reported should be the one documented closest to the date of the procedure.

CCS Class has been modified to indicate that reporting should be based upon the highest grade of angina or chest pain within the past 2 weeks.

Revision Highlights and Coding Clarifications (continued)

Revised Data Elements (continued)

Creatinine has been modified to indicate the reported creatinine should be the one recorded closest to but before anesthetic management.

Cerebrovascular Disease has a revised definition.

COPD is now called “Chronic Lung Disease” and has a revised definition and expanded response categories.

Diabetes Requiring Medication is now called “Diabetes” and has a revised definition.

Renal Failure, Dialysis has a revised definition.

Emergency Transfer to OR After PCI is now called “Surgery for PCI Complication” and has a revised definition.

Previous PCI This Admission is now called “Previous PCI this Episode of Care” and has a revised definition.

Previous PCI Before This Admission is now called “Previous PCI Before this Episode of Care” and has a revised definition.

Major Event Stroke Intra-Op to 24 Hrs is now called “Major Event - Stroke” and has a revised definition.

Major Event Deep Sternal Wound Infection has a revised definition.

Major Event Bleeding Requiring Reoperation is now collected as either “Acute” or “Late” and has a revised definition.

Major Event Sepsis or Endocarditis is now called “Major Event Sepsis” and has a revised definition.

Major Event GI Bleeding, Perforation or Infarction is now called “Major Event GI Event” and has a revised definition.

Major Event Renal Failure has a revised definition.

Revision Highlights and Coding Clarifications (continued)

New Data Reporting Clarifications

These data reporting clarifications are added effective January 2012.

ECMO should not be reported when it is the only cardiac surgical procedure performed in an admission.

Percutaneous Ventricular Assist Device should not be reported unless it is performed at the same time as a reportable cardiac surgical procedure. If a Percutaneous VAD is placed during a surgical procedure, use procedure code 830.

Aortic wrapping procedures and ligation or excision of left atrial appendage have been added to the list of "Do Not Code" procedures in the "When to Complete an Adult CSRS Form" section.

When to Complete an Adult CSRS Form

Complete an Adult Cardiac Surgery Reporting System (CSRS) form for every patient age 18 or over on admission undergoing one or more operations on the heart or great vessels, with or without extracorporeal circulation.

Unless otherwise specified, forms should be submitted for reportable cardiac surgery no matter where in the hospital the operation is performed. References to the “operating room” in these instructions can be interpreted to mean “the location where the cardiac procedure is occurring.”

If the patient has more than one cardiac surgery during a single hospital stay, complete a separate form for each reportable cardiac surgery.

Transcatheter valve replacement procedures should be reported to CSRS, wherever the procedure may occur. Use Adjunct Valve Information codes (640-643) to indicate a transcatheter valve replacement was performed.

DO NOT CODE:

- Implantation or removal of a pacemaker and its leads or wires
- Removal of an AICD and its leads or wires
- Coronary endarterectomies
- Femoral artery repair or bypass
- Innominate artery bypass
- Aortic subclavian bypass
- Exploration of the atria, aorta, valves, ventricles, or pulmonary artery
- Removal of thymoma
- Thymectomy
- VAD removal
- Intra-cardiac thrombus removal
- Intra-coronary thrombus removal
- Epicardial lead placement
- Ventricular support device (e.g. Heartnet restraint)
- Coronary aneurysm repair (other than CABG)
- Aortic wrapping procedures
- Ligation or excision of left atrial appendage

When to Complete an Adult CSRS Form (continued)

When the following procedures are the ONLY cardiac surgery performed in a hospital admission, code them as a 498 or 998, otherwise, the procedures are NOT CODED.

- Surgical Removal of a Stent
- Aortic Endarterectomy
- Pulmonary Artery Endarterectomy

During quarterly and annual data verification and validation efforts, we will be asking for supporting documentation for cases coded as 398, 498, or 998.

Therefore, we highly recommend that at the time of coding you keep a copy of the operative note as supporting documentation in a place for easy retrieval at a later date.

Code the following procedures only when they are performed at the same time as another reportable cardiac surgery:

- Carotid Endarterectomy (763)
- Implantation of an AICD (764)
- Percutaneous Ventricular Assist Device (use procedure code 830)

Code the following only when performed at the same time as a CABG or valve surgery:

- Percutaneous Coronary Intervention (711)

Code the following procedures only when they are performed in the same admission as a reportable cardiac surgical procedure:

- ECMO (834)

Guidance on Selecting Appropriate Procedure Codes

Repair of Cardiac Laceration Due to Trauma (907): Should be coded for repair of cardiac laceration due to trauma including a procedure to repair an injury to the heart that has resulted from a cardiac diagnostic or interventional procedure or from cardiac surgery.

Radiofrequency or Operative Ablation (770-772): Code 770 (Atrial) or 771 (Ventricle) should be used when lesions are created in the atria or ventricle by an energy source (radiofrequency, microwave, cryothermia, etc.). The lesion then disrupts the abnormal re-entry pathways of electrical signals that can lead to fibrillation.

A 772 (Maze) should be coded when there is a surgical procedure (standard surgical maze procedure) in which full thickness incisions are made in the atria of the heart. Sutures are then used to reapproximate the incised tissue. The resulting lesion disrupts the abnormal re-entry pathways of electrical signals that lead to atrial fibrillation.

All procedures coded 772 will require an operative note to verify coding.

Pericardiectomy (402): Any time the procedure consists of more than a pericardial window (i.e. stripping or partial pericardiectomy) and the procedure is performed on CP bypass it should be coded 402. A pericardial window is a small hole in the pericardium usually done by removing a small amount of the pericardial wall and is usually done for a large or symptomatic collection of pericardial fluid or for diagnosis (biopsy).

Aortic Root Replacement or Repair, With Graft, With Coronary Reimplantation (785): This code only refers to procedures that involve the aortic root repair/replacement and an aortic valve replacement. An Ascending Aorta, with Graft, With Coronary Reimplantation should be coded 780.

Aortic Valve Replacements: Do not code aortic root enlargements when performed with aortic valve replacements.

Valve Debridement: If a valve has had debridement, then a valve repair should be coded.

Bicuspid Aortic Valve: When a bicuspid aortic valve is being operated on for a patient who is not in the childhood era and the operation is required due to acquired valve disease, it should be coded as a standard valve procedure (Code 520-548).

Guidance on Selecting Appropriate Procedure Codes (continued)

Adjunct Valve Information (640-643): Use these codes to indicate a transcatheter valve replacement has been performed by either transfemoral (640), transapical (641), subclavian (642) or direct aortic (643) approach. These procedures should be reported even if they do not occur in the operating room. A valve replacement code must also be reported.

Third Digit for Valve Replacement (510- 608): When reporting valve replacement surgery (codes 510-608), use the third digit to indicate if the valve(s) currently being replaced have been previously intervened upon and if so the reason for the reoperation.

The third digit information is specific to the valve reported. For example, a patient with previous aortic valve replacement who is now having mitral valve replacement (mechanical) would be reported using code 550 because this is not a re-operation on the mitral valve. In the event of multiple valve surgery, the third digit may be different for each valve code reported, i.e. one valve may be a re-op and the other(s) may not.

Codes for re-operation due to failed catheter-based or surgical valve repair and as a complication of a transcatheter valve replacement have also been added. Use code 7 (Complication of Transcatheter Valve Replacement) in the event of an unsuccessful transcatheter valve replacement which requires urgent or emergent surgical valve replacement.

PCI in Same Setting as CABG or Valve Surgery (711): Use this procedure code to indicate percutaneous coronary intervention (PCI) was performed in the same procedure room visit as CABG or valve surgery. This may take place in the OR or some other location such as a hybrid procedure room. This procedure should only be reported if done at the same time as CABG or valve surgery. Data for the PCI must be reported to the Percutaneous Coronary Interventions Reporting System.

Ventricular Assist Device as a Destination Therapy (840): If an LVAD is placed as the final therapy, code 840 in addition to the LVAD. For example, if the patient is not a candidate for a heart transplant, but an LVAD is placed as a long-term treatment option this code would be appropriate.

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, whether in hospice or not, appropriate supporting documentation should be sent to Cardiac Services Program. Examples of appropriate documentation include but are not limited to: 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, and evidence of death 30 days or more after initial discharge.

It will be the responsibility of the hospital (physician) to send documentation to the Department of Health's Cardiac Services Program 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.

Cardiogenic Shock Cases

Beginning with cases discharged January 1, 2006 and continuing for a period of at least two years, cases in pre-procedural Cardiogenic Shock will not be included in the publicly released reports and analyses. This applies only to cases that meet the NYS Cardiac Services Program definition of Cardiogenic Shock (risk factor #13). Data for these cases must still be submitted electronically and will be subject to data verification activities. To ensure that the appropriate cases are identified as "Shock" cases, we will continue to require submission of medical record documentation of any case reported with this risk factor. If appropriate documentation is not provided by your center, the risk factor will be removed from the data and the case will be included in analysis. In addition, we anticipate that there will be increased requirements for medical record documentation for cases coded as "Hemodynamically Unstable" as well. It is strongly suggested that all appropriate staff closely review the definitions and documentation requirements for these two risk factors.

Note: The above policy regarding cases in Shock will be continued for at least another year (2012 discharges).

CSRS Data Reporting Policies (continued)

Physician Assignment

When multiple records exist for the same patient during a hospital admission, and two or more surgeons were reported for those operations, the case will be assigned for analysis to the surgeon performing the first surgery. However, the hospital may submit a letter from the CEO or Medical Director requesting that the case be assigned to the surgeon performing the later surgery.

Reporting Schedule

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 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

Patient Name

Variable Names: LASTNAME, FIRSTNAME

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

Medical Record Number

Variable Name: MEDRECNO

Enter the patient's medical record number.

Social Security Number

Variable Name: SSNO

Enter the patient's Social Security Number as shown in the medical record. If the medical record does not contain the patient's Social Security Number, leave this item blank.

Date of Birth

Variable Name: DOB

Enter the patient's exact date of birth.

I. Patient Information (continued)

Sex

Variable Name: SEX

Check the appropriate box for the patient's sex at birth.

Note: In the absence of any other information, it is reasonable to assume that the sex at birth is the same as at the time of admission.

Ethnicity

Variable Name: ETHNIC

Check the appropriate box.

Note: The term "Hispanic" refers to persons who trace their origin or descent to Mexico, Puerto Rico, Cuba, Central and South America or other Spanish cultures.

Race

Variable Names: RACE, RACESPEC

Choose the appropriate response from the list below.

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.

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. Provide the specific race for any case marked "Other."

I. Patient Information (continued)

Race (continued)

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 NYS, 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 in the US.

Hospital Admission Date

Variable Name: ADMIDATE

Enter the date that the current hospital stay began.

Primary Payer

Variable Name: PAYER

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

Please note that Worker's Compensation, Family Health Plus, and Other Federal Programs are reported as code "19-Other".

Interpretation: Primary Payer and Medicaid: For "Medicaid Pending" code Primary Payer as "11-Self-Pay" and check the box "Medicaid".

For patients in prison, code Primary Payer as "19-Other".

I. Patient Information (continued)

Primary Payer (continued)

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. Code “19” is any other type of insurance that is not given a code of its own (e.g. Corrections).

If the patient has Blue Cross and Medicare, code Medicare if there is no indication of which is primary.

Report a PPO (Preferred Provider Organization) as “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 list of PFIs for cardiac diagnostic centers in 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 PFI.

Note: PFI on the above website is listed without leading 0s. For purposes of cardiac reporting, PFI should always be four (4) numeric characters. For example, PFI “1” should be reported as “0001”.

II. Procedural Information

REMINDER: Fill out a separate CSRS form for each cardiac surgery involving the heart or great vessels during the hospital admission.

Hospital That Performed Diagnostic Cath

Variable Name: CATHPFI

If the cardiac surgery was preceded by a diagnostic catheterization, enter the name and PFI number of the hospital in the spaces provided. If the catheterization was at a cardiac diagnostic center in NYS, enter its PFI Number from Attachment A; if done at a Veterans Administration hospital in NYS, enter "8888"; if done outside NYS, enter "9999". If there was no diagnostic catheterization, leave this item blank.

Do not use this field to report any diagnostic procedure (e.g. CT) other than catheterization.

Note: Diagnostic Catheterization Hospital name is included on the paper form for abstractor convenience. It is not part of the CSRS file structure.

Date of Surgery

Variable Name: SURGDATE

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

Prior Surgery This Admission

Variable Names: PRIOSURG, PRIODATE

Check the appropriate box to indicate whether the patient had any reportable (form generating) cardiac operation prior to the present operation during the same hospital admission.

If "Yes" then the date of the previous cardiac operation **MUST** be entered. This is very important because this date aids in combining multiple procedures that occurred during the same admission in the proper order.

II. Procedural Information (continued)

Cardiac Procedures This OR Visit

Variable Names: PROC1, PROC2, PROC3, PROC4, PROC5

Enter the 3-digit State Cardiac Advisory Committee Code (SCAC) from the procedure code list in Attachment D – Congenital and Acquired Cardiac Procedure Codes.

List up to 5 cardiac procedures performed during this operating room visit.

If there are more than 5, list the 5 most significant.

Note: Please see Attachment D: Congenital and Acquired Cardiac Procedure Codes and “When to Complete an Adult CSRS Form” and “Guidance on Selecting Appropriate Codes” (pg 9-12) for additional coding instructions and scenarios for reporting procedure codes.

Congenital Diagnosis

Variable Names: DIAG1, DIAG2, DIAG3

If the patient had a congenital defect repair either in conjunction with, or as the primary surgical procedure, indicate the three most significant congenital diagnoses.

The diagnosis codes in Attachment E are identical to those used for the Pediatric Cardiac Surgery Reporting System. Inclusion of this information will allow for meaningful evaluation of outcomes for adult congenital cardiac surgery.

Coding Note: Congenital Diagnosis Codes in Attachment E are aligned with STS v2.73 data elements 5310, 5320 and 5330.

Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.

II. Procedural Information (continued)

Primary Physician Performing Operation

Variable Name: PHYSNUM

Enter the name and NYS physician license number of the primary physician who performed the cardiac surgical procedure.

Interpretation: The primary physician should be the one who performed the majority of the cardiac procedure in that surgery.

The following is one of many possible examples: In a single trip to the OR, a radiofrequency ablation is performed by one surgeon and then a CABG by a second surgeon. The primary physician reported on the CSRS form should be the one who performed the CABG. It does not matter that the ablation was performed before the CABG.

If a procedure includes both a cardiac surgeon and a cardiologist (e.g. hybrid revascularization, transcatheter valve replacement) report the cardiac surgeon as the primary physician for these purposes and also report the physician license number for the interventional cardiologist in the “Interventional Cardiologist” field.

Note: Physician name is included on the paper version of the data collection form for abstractor convenience. Physician name is not part of the required CSRS data structure.

Anesthesiologist (1)

Variable Name: ANESNUM1

Enter the name and NYS physician license number of the responsible anesthesiologist at the start of the cardiac surgery.

Note: Anesthesiologist name is included on the paper version of the data collection form for abstractor convenience. Anesthesiologist name is not part of the required CSRS data structure.

Anesthesiologist (2)

Variable Name: ANESNUM2

Enter the name and NYS physician license number of the responsible anesthesiologist at the end of the cardiac surgery.

Note: Anesthesiologist name is included on the paper version of the data collection form for abstractor convenience. Anesthesiologist name is not part of the required CSRS data structure.

II. Procedural Information (continued)

Interventional Cardiologist

Variable Name: CARDNUM

If the procedure is a Transcatheter Valve Implantation (procedure code 640-643) or PCI in same setting as CABG or Valve Surgery (procedure code 711), enter the name and NYS physician license number of the interventional cardiologist participating in the case.

Note: Interventional cardiologist name is included on the paper version of the data collection form for abstractor convenience. Interventional cardiologist name is not part of the required CSRS data structure. NYS physician license number is part of the file upload and must be reported for procedure codes 640-643 or 711. For these procedure codes, if there was no interventional cardiologist participating enter code "000000."

CABG Information

Variable Names: TOT_COND, ART_COND, DISTAL

The following information must be completed for all CABG procedures.

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

Arterial Conduits: List the number of arterial conduits or grafts used up to 9. For more than 9, report 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, report 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.

Minimally Invasive

Variable Name: MINI_INV

If the cardiac surgical procedure began through an incision other than a complete sternotomy or thoracotomy (less than 12 centimeters in length) check "Yes," regardless of whether the case converted to a standard incision or cardiopulmonary bypass was used. Otherwise check "No."

II. Procedural Information (continued)

Converted to Standard Incision

Variable Name: STND_INC

Check this box to indicate that the minimally invasive procedure was modified to a standard incision.

Note: This box should never be checked unless Minimally Invasive is also checked.

Converted from Off Pump to On Pump

Variable Name: CONVERT

Check this box if the procedure began without the use of cardiopulmonary bypass, but prior to the completion of the procedure the patient was placed on pump. This should only be checked if the patient was placed on pump unexpectedly.

Entire Procedure Off Pump

Variable Name: ALL_OFF

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

Internal Mammary Artery (IMA) Grafting

Variable Name: IMA

Enter the appropriate code.

- 0 Never
- 1 This OR Visit
- 2 Prior to this OR Visit

For any patient who has never had a left or right internal mammary artery (IMA) graft, code "0" (Never). If the patient is having an IMA graft during this operation, code "1" (This OR Visit). If at anytime prior to this operating room visit (including this admission) the patient has had an IMA graft, code "2" (Prior to this OR Visit).

If the patient has had an IMA graft anytime prior to this operating room visit and is having one during the operating room visit, code "1".

Ila. Peri-Operative Information

Skin Incision Time

Variable Name: SURG HOUR, SURG MIN

Indicate the time, to the nearest minute (using 24-hour clock), that the skin incision, or its equivalent, was made.

Interpretation: The intent of this field is to capture the time the first skin incision is made regardless of if the first incision is a harvest site incision or a sternal/thoracotomy incision.

Coding Note: *SURG HOUR and SURG MIN* definition is aligned with STS v2.73 data element 2690.

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Skin Closure Time

Variable Name: CLOSE HOUR, CLOSE MIN

Capture the date and time (using 24 hour clock) to the minute, that the skin incision was closed, or its equivalent.

Note: This element refers to the time of the final incision closure prior to leaving the operating room.

If the patient leaves the operating room with an open incision, collect the time that the dressings were applied to the incision.

If the patient expires in the OR prior to skin closure, time of death should be reported in place of skin closure time.

Coding Note: *CLOSE HOUR and CLOSE MIN* definition is aligned with STS v2.73 data element 2700.

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Pre-Induction Blood Pressure

Variable Name: BP_SYS, BP_DIA

Enter the patient's blood pressure just prior to the induction of anesthesia as measured by any means.

Ila. Peri-Operative Information (continued)

Post-Op Temperature

Variable Name: POST_TEMP

Report the patient's post-op temperature in degrees Celsius.

This should be the temperature on arrival at the next level of care after the operating room (e.g. Critical Care, PACU, Recovery, etc).

If a pulmonary artery temperature is available upon arrival at the next level of care, report that value. Otherwise report temperature via other method.

If no post-operative temperature is available (e.g. patient expires prior to arrival at next level of care), report temperature as 00.0.

Temperature Route

Variable Name: TEMP_RT

Report the route of post-operative temperature measurement using the following codes:

- 1 Pulmonary Artery
- 2 Rectal/Bladder
- 3 Nasopharyngeal
- 4 Tympanic
- 8 Other
- 9 Unknown

If Post-op Temperature is reported as "00.0" because none is available (e.g. patient expires prior to arrival at next level of care), report Temperature Route as "9-Unknown".

Hematocrit

Variable Name: CRIT_OR, CRIT_LOW, CRIT_LST, CRIT

Report the patient's hematocrit at the following specified time periods.

- First recorded in the operating room
- Lowest on Cardiopulmonary Bypass - report as "00" or leave blank if entire procedure was "off-pump."
- Last on Cardiopulmonary Bypass - report as "00" or leave blank if entire procedure was "off-pump."
- Post-Op – Value on arrival at next level of care after the operating room (e.g. Critical Care, PACU, Recovery, etc). If no value is available (e.g. patient expires prior to arrival at next level of care) then report as "00" or leave blank.

Ila. Peri-Operative Information (continued)

Hematocrit (continued)

Clarification:

Values from any source are acceptable (e.g. lab, Istat, ABG), however if available from multiple sources for the same time-frame, central lab values are preferred to point of care values.

If blood is drawn for “post-op” lab work just prior to leaving the operating room, that value may be reported for “Post-op, on arrival at next level of care.”

In the event that only one Hematocrit value is recorded for the entire time that the patient is on Cardiopulmonary Bypass, then this value would be reported as both “Lowest” and “Last.”

Pre-Op Beta Blocker Use

Variable Name: PRE_BETA

Use the following codes to indicate pre-op beta blocker use or contraindication.

- 1 Yes - The patient received beta blockers within 24 hours preceding surgery
- 2 Contraindicated - Beta blocker was contraindicated. The contraindication must be documented in the medical record by a physician, nurse practitioner, or physician assistant.
- 3 No - The patient did not receive beta blockers within 24 hours preceding surgery and there is no documented contraindication for beta blockers.

Coding Note: *PRE_BETA* definition is aligned with STS v2.73 data element 1710, however the response values must be mapped. CSRS response 1 = STS response 1; CSRS 2 = STS 3; CSRS 3 = STS 2.

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Ila. Peri-Operative Information (continued)

Extubation at 24 Hours – Report Only for CABG Patients

Variable Name: EXTUBATE

Use the following codes to indicate extubation at 24 hours post-op.

1 Yes - The patient was extubated at 24 hours post-op.

2 Contraindicated - The patient was not extubated at 24 hours post-op due to a contraindication. Contraindications include the following: myocardial dysfunction; valvular heart disease; active systemic illness; respiratory disease; neuropsychiatric disease or problems with communication secondary to language. This would include stroke (new neurological deficit) and neuropsychiatric state (paranoia, confusion, dementia).

3 Neither - The patient was not extubated at 24 hours post-op and there was no contraindication as defined above.

Interpretation: Post-op is defined as starting when the patient leaves the actual procedure room where the cardiac operation occurred.

Post-Op Beta Blocker Use - Report Only for CABG Patients

Variable Name: PO_BETA

1 Yes - The patient received beta-blockers within 24 hours post-op.

2 Contraindicated - The patient did not receive beta-blockers with 24 hours post-op due to a contraindication. Contraindications include the following: allergy, bradycardia (heart rate less than 60 bpm) and not on beta blockers, second or third degree heart block on ECG on arrival or during hospital stay and does not have a pacemaker, systolic blood pressure less than 90 mmHg and not on beta blockers, or other reasons documented by a physician, nurse practitioner, or physician's assistant in the medical chart.

3 Neither- The patient did not receive beta-blockers within 24 hours post-op and there was no contraindication as defined above.

Interpretation: Post-op is defined as starting when the patient leaves the actual procedure room where the cardiac operation occurred.

Ila. Peri-Operative Information (continued)

Intra-Operative Blood Transfusion

Variable Name: TRANSFUS

Indicate if packed red blood cells were transfused intraoperatively. Do not include autologous, cell-saver, pump-residual or chest tube recirculated blood. Intraoperatively is defined as any blood started inside of the OR.

Coding Note: CSRS “TRANSFUS” is a Yes/No variable with a definition such that when STS element 3060 IBdRBCU is ≥ 1 then TRANSFUS should be “checked” (i.e. reported as 1 for text file upload).

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Glucose Control Protocol

Variable Name: GLUCOSE

Check this box if a glucose control protocol was used for this patient.

Interpretation: This element is referring to a post-op glucose control protocol. These may be initiated in the pre or intra-operative period but continued post-op.

Expected documentation would be an order in the patient’s chart indicating use of protocol or evidence that there are standing orders for all patients to be on a protocol.

III. Pre-Op Surgical Risk Factors

Surgical Priority

Variable Name: PRIORITY

Indicate the clinical status of the patient prior to entering the operating room.

- 1 Elective: The patient's cardiac function has been stable in the days or weeks prior to the operation. The procedure could be deferred without increased risk of compromised cardiac outcome.
- 2 Urgent: Procedure required during same hospitalization in order to minimize chance of further clinical deterioration. Examples include but are not limited to: Worsening, sudden chest pain; CHF; acute myocardial infarction; anatomy; IABP; unstable angina with intravenous nitroglycerin or rest angina.
- 3 Emergent: Patients requiring emergency operations will have ongoing, refractory (difficult, complicated, and/or unmanageable) unrelenting cardiac compromise, with or without hemodynamic instability, and not responsive to any form of therapy except cardiac surgery. An emergency operation is one in which there should be no delay in providing operative intervention.
- 4 Emergent Salvage: The patient is undergoing CPR en route to the OR or prior to anesthesia induction or has ongoing ECMO to maintain life.

Coding Note: *PRIORITY* is aligned with STS v2.73 element 2390.

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Height

Variable Name: HEIGHT

Enter the patient's height in centimeters (cm).

Coding Note: HEIGHT definition is consistent with STS v2.73 element 640.

Weight

Variable Name: WEIGHT

Indicate the weight of the patient, in kilograms (kg), closest to the date of the procedure.

Coding Note: *WEIGHT* definition is consistent with STS v2.73 element 630.

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III. Pre-Op Surgical Risk Factors (continued)

Stress Test / Imaging Study Done

Variable Name: STRS_DONE

Use the codes below to indicate if a stress test was performed prior to this procedure but within 6 months.

- 1 Yes
- 2 No
- 9 Unknown

Stress Test / Imaging Study Type

Variable Name: STRS_TYP

Use the codes below to indicate the type of stress test performed

- 1 Standard Exercise Stress Test – without imaging
- 2 Stress Echocardiogram
- 3 Stress Testing with single photon emission computed tomography (SPECT) myocardial perfusion imaging (MPI)
- 4 Stress Testing with cardiac magnetic resonance (CMR)
- 9 Not Done / Unknown

If more than one type of stress test was performed within the past 6 months, report on the most recent test.

Stress Test / Imaging Study Results

Variable Name: STRS_RES

Use the codes below to indicate the stress test results. Definitions and clarification can be found Attachment F: Stress Test Results.

- 1 Negative
- 2 Positive, Low Risk
- 3 Positive, Intermediate Risk
- 4 Positive, High Risk
- 5 Positive, Risk Unavailable
- 6 Indeterminate
- 7 Unavailable
- 9 Not Done/ Unknown

Note: Inclusion of stress test reports in the medical record is encouraged to allow for accurate and complete reporting of these data elements.

III. Pre-Op Surgical Risk Factors (continued)

Ejection Fraction and Measure

Variable Names: EJEC_FRA, MEASURE

Record the pre-operative ejection fraction taken closest to, but before, the start of the cardiac procedure.

If an ejection fraction is unavailable, enter "0" and then enter "9 – Unknown" for the measure.

Indicate how the Ejection Fraction was measured using one of the following:

- 1 LV Angiogram
- 2 Echocardiogram
- 3 Radionuclide Studies
- 4 Transesophageal Echocardiogram (TEE), this includes intra-operative
- 8 Other
- 9 Unknown

Note: Intra-operative direct observation of the heart is NOT an adequate basis for a visual estimate of the ejection fraction.

Interpretation:

Intra-operative TEE is acceptable, if no pre-operative Ejection Fraction is available.

Any ejection fraction that is described as "Normal" in the medical record should be considered 55%.

Any cases with a missing or unusual ejection fraction will be sent back during quarterly and annual data validation to verify accuracy of this data element.

Anginal Classification Within 2 Weeks

Variable Name: CCS_CLAS

Indicate the patient's anginal classification or symptom status within the past 2 weeks prior to surgery. The anginal classification or symptom status is classified as the highest grade of angina or chest pain by the Canadian Cardiovascular Angina Classification System (CCA).

- 1 CCA I Ordinary physical activity does not cause angina; for example walking or climbing stairs, angina occurs with strenuous or rapid or prolonged exertion at work or recreation.
- 2 CCA II Slight limitation of ordinary activity; for example, angina occurs walking or stair climbing after meals, in cold, in wind, under emotional stress or only during the few hours after awakening, walking more than two blocks on the level or climbing more than one flight of ordinary stairs at a normal pace and in normal conditions.

III. Pre-Op Surgical Risk Factors (continued)

Anginal Classification Within 2 Weeks (continued)

- 3 CCA III Marked limitation of ordinary activity; for example, angina occurs walking one or two blocks on the level or climbing one flight of stairs in normal conditions and at a normal pace.
- 4 CCA IV Inability to carry on any physical activity without discomfort - angina syndrome may be present at rest.
- 8 No Symptoms, No Angina The patient has no symptoms, no angina.

Coding Note: CCS_CLAS definition is aligned with STS v2.73 data element 1570. *Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Clarification:

Atypical symptoms (e.g. shortness of breath, upper abdominal pain, left arm pain) may be considered in identifying the CCS class when they are documented as an anginal equivalent or evidence of myocardial ischemia. If these symptoms are not documented as an anginal equivalent, then report response category 8 - No Symptoms, No Angina.

Cardiac Presentation on Admission

Variable Name: CAD_PRE5

Indicate the type of angina present prior to this procedure.

- 1 No Symptoms, No Angina
- 2 Symptoms Unlikely to be Ischemia
Pain, pressure or discomfort in the chest, neck or arms not clearly exertional or not otherwise consistent with pain or discomfort of myocardial ischemic origin. This includes patients with non-cardiac pain (e.g., pulmonary embolism, musculoskeletal, or esophageal discomfort), or cardiac pain not caused by myocardial ischemia (e.g. acute pericarditis).
- 3 Stable Angina
Angina without a change in frequency or pattern for the six weeks prior to this surgical intervention. Angina is controlled by rest and/or oral or transcutaneous medications.
- 4 Unstable Angina
There are three principal presentations of unstable angina:
 - a. Rest angina (occurring at rest and prolonged usually >20 minutes);
 - b. New-onset angina (within the past 2 months, of at least CCS Class III severity); or
 - c. Increasing angina (previously diagnosed angina that has become distinctly more frequent, longer in duration, or increased by 1 or more CCS class to at least CCS III severity).

III. Pre-Op Surgical Risk Factors (continued)

Cardiac Presentation on Admission (continued)

5 Non-ST Elevation MI (Non-STEMI)

Non-ST elevation myocardial infarction as documented in the medical record. Non-STEMIs are characterized by the presence of both criteria:

- a. Cardiac biomarkers (creatinine kinase-myocardial band, Troponin T or I) exceed the upper limit of normal according to the individual hospital's laboratory parameters with a clinical presentation which is consistent or suggestive of ischemia. ECG changes and/or ischemic symptoms may or may not be present.
- b. Absence of ECG changes diagnostic of a STEMI (see #6 STEMI).

6 ST-Elevation MI (STEMI) or equivalent.

The patient presented with a ST elevation myocardial infarction (STEMI) or its equivalent as documented in the medical record.

STEMIs are characterized by the presence of **both criteria**:

- a. ECG evidence of STEMI: New or presumed new ST-segment elevation or new left bundle branch block not documented to be resolved within 20 minutes. ST-segment elevation is defined by new or presumed new sustained ST-segment elevation at the J-point in two contiguous ECG leads with the cut-off points: ≥ 0.2 mV in men or ≥ 0.15 mV in women in leads V2-V3 and/or ≥ 0.1 mV in other leads and lasting greater than or equal to 20 minutes. If no exact ST-elevation measurement is recorded in the medical chart, physician's written documentation of ST-elevation or Q waves is acceptable. If only one ECG is performed, then the assumption that the ST elevation persisted at least the required 20 minutes is acceptable. Left bundle branch block (LBBB) refers to new or presumed new LBBB on the initial ECG.
- b. Cardiac biomarkers (creatinine kinase-myocardial band, Troponin T or I) exceed the upper limit of normal according to the individual hospital's laboratory parameters and a clinical presentation which is consistent or suggestive of ischemia

Note: For purposes of the Registry, ST elevation in the posterior chest leads (V7 through V9), or ST depression that is maximal in V1-3, without ST-segment elevation in other leads, demonstrating posterobasal myocardial infarction, is considered a STEMI equivalent.

Coding Note: *CARD_PRES* definition is aligned with STS v2.73 data element 1610. *Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Clarification:

Report Cardiac Presentation based on the worst status present within 7 days.

Atypical symptoms (e.g. shortness of breath, upper abdominal pain, left arm pain) may be considered in identifying the Cardiac Presentation when they are documented as an anginal equivalent or evidence of myocardial ischemia. If these symptoms are not documented as an anginal equivalent, then report response category 2 - Symptoms Unlikely to be Ischemia.

III. Pre-Op Surgical Risk Factors (continued)

Creatinine

Variable Name: CREATININE

Indicate the creatinine level closest to the date and time of surgery but prior to anesthetic management (induction area or operating room).

Interpretation: For the purposes of this data element, anesthetic management begins when a member of the anesthesiology team initiates care. The administration of IV fluids in the holding area can cause dilution of blood. Do not capture labs drawn after the patient receives fluids in the holding area or O.R.

Acceptable documentation may include that from an out-patient record.

Coding Note: *CREATININE* definition is aligned with STS v2.73 data element 750. *Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

Vessels Diseased

Variable Name: LMT, PROX_LAD, MID_LAD, RCA, LCX

For each diseased vessel, check the appropriate box to indicate the percent diameter stenosis. Include all vessels diseased, even branches.

Interpretation: This section must be completed for all CABG cases. If this information is available for other procedures, please indicate the vessels diseased, otherwise leave blank.

If the diseased segment of the native vessel is bypassed by an open artery or vein graft, do not code as diseased. This vessel is re-vascularized.

Use the ranges listed below when the medical record describes the percent stenosis in the following ways:

MILD	= < 50%
MODERATE	= 50-69%
SEVERE	= > 70%

If a vessel or branch is described as having “Mild” stenosis then the vessel would NOT be coded as diseased, since we only code 50-100% stenosis.

If the medical record reports the range “40-50%” stenosis, then DO NOT CODE as diseased.

If the medical record reports the range “60-70%” stenosis, then code 50-69%.

III. Pre-Op Surgical Risk Factors (continued)

Vessels Diseased (continued)

Proximal LAD is reported by itself. Disease of the Major Diagonal should be reported with Mid/Distal LAD. The Ramus Intermediate should be coded as the Diagonal or Marginal.

Always take the highest stenosis reported for a vessel. If the medical record reports the Proximal RCA with a 70% lesion and the Distal RCA with a 50% you should code the RCA as 70-100%, since the Proximal RCA has a 70% lesion.

If the medical record only has documentation that states the LAD was stenosed then code the Mid LAD and not the Proximal LAD.

Valve Disease

Variable Names: STEN_AOR, STEN_MIT, STEN_TRI, INCO_AOR, INCO_MIT, INCO_TRI

This section is required for valve patients, if the information is available for other patients, please report it.

Enter an assessment of the degree of stenosis or incompetence (acute or chronic) for each valve (Aortic, Mitral, Tricuspid). Both lines should be completed for all valve patients.

Please enter the following values for each valve to indicate the degree of stenosis or incompetence:

- 0 None
- 1 Mild
- 2 Moderate
- 3 Severe

Moderate or Severe Stenosis – Aortic, Mitral, or Tricuspid: Should be demonstrated by appropriate imaging technique, echocardiography, or hemodynamic measurement during cardiac catheterization or operation.

Moderate or Severe Aortic Incompetence: Should be demonstrated by aortography or by pre-op or intraoperative echocardiography.

Moderate or Severe Mitral Incompetence: Should be demonstrated by left ventriculography or by pre-op or intraoperative echocardiography.

Moderate or Severe Tricuspid Incompetence: Should be demonstrated by physical examination or by pre-op or intraoperative echocardiography.

Note: If a patient is not having a valve procedure, but disease (stenosis or incompetence) is indicated, please code.

III. Pre-Op Surgical Risk Factors (continued)

Anti-Anginal Medication Within 2 Weeks

Variable names: MED_BB, MED_CA, MED_NIT, MED_RAN, MED_OTH

Indicate if the patient was taking any of the following agents to treat anginal symptoms within the past two weeks. Check all that apply.

- Beta-Blockers
- Calcium Channel Blockers
- Long Acting Nitrates
- Ranolazine
- Other

Clarification:

Do not report if the patient was given sublingual, IV, or short acting formula of the medications.

Do not report if the patient has been prescribed the medication but is known to be not taking it.

Report if the patient was started on an oral form of the medication after admission but prior to this surgical procedure.

Report if this medication was prescribed for this patient, but you are unsure it has been prescribed specifically to treat anginal symptoms.

Nitro paste and nitro patch are considered Long Acting Nitrates.

“Other” excludes short acting anti-anginal medications such as nitroglycerin sublingual tablets or spray that is used to relieve an acute episode of chest pain.

Other Patient Characteristics

Variable Names: FFR_IVUS, CTO, GRFTFAIL, LIMA_FAIL, LIMA_PAT

Indicate which, if any, of the following characteristics apply to this patient. Check all that apply.

- 50-69% stenosis with significant findings on Fractional Flow Reserve (<0.75) and/or IVUS with significant reduction in cross sectional area.
Note: Significant reduction in cross sectional area by IVUS is defined as 6mm² for the left main and 4mm² for major epicardial vessels other than the left main.

III. Pre-Op Surgical Risk Factors (continued)

Other Patient Characteristics (continued)

- Chronic Total Occlusion (CTO) is the only stenosis – Indicate if patient has a CTO and no other lesion in that vessel or any other vessel. CTO is defined as a vessel with 100% pre-procedure stenosis presumed to be 100% occluded for at least 3 months previous to this procedure.
Note: If timeframe of 3 months is not specified, but lesion is described as “CTO,” this is acceptable.
- Prior CABG with native 3 vessel disease and failure of multiple bypass grafts.
- LIMA was used as a graft but is no longer functional
- LIMA was used as a graft and remains patent to a native coronary artery.

Interpretation: For the items regarding LIMA patency, the graft would be considered “no longer functional” if there is angiographic stenosis of 70% or more or there is evidence of significant flow restriction documented by FFR or by stress test (with echo or nuclear) to localize the ischemia.

0. None

Variable Name: NORISK

Report if none of the pre-operative risk factors listed below are present.

1. Previous CABG - Patent Grafts

Variable Name: PAT_GRAFT

Indicate if, prior to this cardiac surgery, the patient has undergone CABG and currently has one or more patent grafts.

Include any surgeries that occurred prior to this one including those earlier in the current admission.

Note: Check this box if there are any patent grafts, even if there are also occluded grafts. Only check box 1 or box 1a, not both.

If the patient has a history of CABG and a history of other cardiac surgery, you should report both risk factors.

III. Pre-Op Surgical Risk Factors (continued)

1a. Previous CABG – No Patent Grafts

Variable Name: OTH_CABG

Indicate if, prior to this cardiac surgery, the patient has previously undergone CABG and has no patent grafts.

Include any surgeries that occurred prior to this one including those earlier in the current admission.

Note: Check this box only if there are no patent grafts. Only check box 1 or box 1a, not both.

If the patient has a history of CABG and a history of other cardiac surgery, you should report both risk factors.

2a. Previous Valve Surgery

Variable Name: PRE_VALV

Indicate if, prior to this cardiac surgery, the patient has previously undergone surgery or catheter based intervention for valve repair or replacement.

Note: It is acceptable to report this risk factor as well as a risk factor for previous CABG surgery and/or other previous cardiac surgery.

2. Any Other Previous Cardiac Surgery

Variable Name: OTH_SURG

Indicate if prior to this OR visit the patient has had any cardiac surgery other than CABG or valve repair / replacement.

Note: Do not include catheter-based interventions.

If the patient has previously had CABG and/or valve surgery as well as another cardiac surgery, report this risk factor in addition to the appropriate Previous CABG and/or Valve risks.

III. Pre-Op Surgical Risk Factors (continued)

4. - 6. Previous MI (Most Recent)

Variable Names: PREMILT6, PREMI623, PREMIDAY

If the patient had one or more myocardial infarctions before surgery, report the length of time since the most recent MI. Timing should be from the onset of symptoms to the start of the surgery. If the exact time that the symptoms started is not available in the medical record, every effort should be made to create a close estimate based on available documentation.

The diagnosis of Acute Coronary Syndrome (ACS) in the medical record is not sufficient to code risk factors 4 – 6. There must be documentation of a myocardial infarction.

If less than 6 hours, check box “4”.

If 6-23 hours, check box “5”.

If 24 hours or more, enter the number of days in the space provided next to “6”.

If 21 days or more, enter "21".

9. Cerebrovascular Disease

Variable Name: CEREBRO

Indicate whether the patient has cerebrovascular disease, documented by any one of the following:

- CVA (symptoms > 24 hrs after onset, presumed to be from vascular etiology);
- TIA (recovery within 24 hrs);
- Non-invasive carotid test with > 79% diameter occlusion.; or
- Prior carotid surgery or stenting or prior cerebral aneurysm clipping or coil.

Does not include neurological disease processes such as metabolic and/or anoxic ischemic encephalopathy.

Coding Note: *CEREBRO* definition is aligned with STS v2.73 data element 1010. Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.

III. Pre-Op Surgical Risk Factors (continued)

9a. TIA, Only Cerebrovascular Risk

Variable Name: TIA

Indicate whether the patient has a history of a Transient Ischemic Attack (TIA) as the only qualifying feature of "Risk Factor #9 - Cerebrovascular disease." Patient has a history of loss of neurological function that was abrupt in onset but with complete return of function within 24 hours. Patient meets no other elements of the Cerebrovascular disease risk factor.

Interpretation: This element can only be reported if Risk Factor #9 - Cerebrovascular Disease is also reported. TIA should only be reported when the patient meets no other criteria included in the Cerebrovascular Disease definition. For example, if the patient has a history of CVA and TIA, report only #9 - Cerebrovascular Disease.

10. Peripheral Vascular Disease

Variable Name: PERIPH

Angiographic demonstration of at least 50% narrowing in a major aortoiliac or femoral/popliteal vessel, previous surgery for such disease, absent femoral or pedal pulses, or the inability to insert a catheter or intra-aortic balloon due to iliac aneurysm or obstruction of the aortoiliac or femoral arteries. Ankle-Brachial Index < 0.9 is also acceptable documentation.

Examples:

Peripheral Vascular Disease	Code	Do Not Code
1. Tortuosity of the vessel alone		X
2. Tortuosity of the vessel with an inability to insert a catheter	X	
3. Abdominal aortic aneurysm (AAA)	X	
4. Aneurysm in the ascending or descending aorta	X	
5. Absence of femoral pulse on either the right or the left	X	
6. Diminished femoral pulse on either right or left or both		X
7. Claudication		X
8. A negative popliteal pulse alone (1+1- or 1-1+)		X
9. Palpable dorsalis pedis and posterior tibial pulses		X
10. If pulses are non-palpable, but are dopplerable	X	
11. Inability to insert a catheter or IABP in femoral arteries	X	
12. Amputated toes, necrotic toes, gangrene of the foot in the absence of other acceptable criteria		X
13. Renal artery with significant stenosis	X	
14. Subclavian artery with significant stenosis	X	

III. Pre-Op Surgical Risk Factors (continued)

12. Unstable

Variable Name: UNSTABLE

In the immediate pre-operative period, the patient requires pharmacologic or mechanical support to maintain blood pressure or cardiac index.

Interpretation:

Key elements for documentation of Unstable include evidence in the pre-operative period of the following:

1. Hypotension or low cardiac index
and
2. Administration of mechanical or pharmacological support.

For these purposes, the pre-operative period is defined as the period prior to anesthesia taking responsibility for the patient.

- The procedure itself does not constitute support.
- Fluid replacement alone does not constitute support.
- IABP constitutes support only when documented that it was placed for hemodynamics. Pain control, anatomy, or undocumented indication for IABP do not support coding Unstable.

Unstable cannot be coded with SHOCK.

13. Shock

Variable Name: SHOCK

In the immediate pre-operative period, the patient has acute hypotension (*systolic blood pressure < 80 mmHg*) or low cardiac index (*< 2.0 liters/min/m²*), despite pharmacologic or mechanical support.

Interpretation: Key elements for the documentation of Shock include evidence in the immediate pre-operative period of all three of the following elements:

1. Documented acute hypotension (systolic blood pressure < 80 mmHg) or low cardiac index (< 2.0 liters/min/m²), **and**
2. Mechanical or pharmacological support, **and**
3. Persistent acute hypotension (systolic blood pressure < 80 mmHg) or low cardiac index (< 2.0 liters/min/m²) while receiving mechanical or pharmacological support.

For these purposes, the pre-operative period is defined as the period prior to anesthesia taking responsibility for the patient.

III. Pre-Op Surgical Risk Factors (continued)

13. Shock (continued)

- The procedure itself does not constitute mechanical support.
- Fluid replacement alone does not constitute support.

- IABP constitutes support only when documented that it was placed for hemodynamics. Pain control, anatomy, or undocumented indication for IABP do not support coding Shock.

Ongoing resuscitation warrants the coding of Shock.

If the patient has an IABP – the non-augmented BP should be < 80 mmHg to code Shock.

If the patient is Ventricular Assist Device (VAD) dependent then code Shock. The type of VAD (Right, Left, Bi) is not important.

Shock cannot be coded with Unstable.

Clarification: The intent of this data element is to capture patients with pre-operative cardiogenic shock, whose hemodynamics cannot be stabilized with pharmacologic or mechanical support. Patients whose hemodynamics are maintained (SBP \geq 80 or CI \geq 2.0) by pharmacological or mechanical support should be coded as Unstable, not as Shock.

18. Congestive Heart Failure, Current

Variable Name: CHF_CUR

Within 2 weeks prior to the procedure, the patient has a clinical diagnosis of CHF, and symptoms requiring treatment for CHF.

Note: Physician diagnosis of CHF may be based on one of the following:

- Paroxysmal nocturnal dyspnea (PND)
- Dyspnea on exertion (DOE) due to heart failure
- Chest X-Ray showing pulmonary congestion

Documentation must include the presence of a diagnosis of CHF, evidence of symptoms, and treatment for CHF.

III. Pre-Op Surgical Risk Factors (continued)

19. Congestive Heart Failure, Past

Variable Name: CHF_PAST

Between 2 weeks and 6 months prior to the procedure, the patient has a clinical diagnosis/ past medical history of CHF and ongoing treatment for CHF.

Note: Physician diagnosis of CHF may be based on one of the following:

- Paroxysmal nocturnal dyspnea (PND)
- Dyspnea on exertion (DOE) due to heart failure
- Chest X-Ray showing pulmonary congestion

Documentation must include a diagnosis of CHF and evidence of treatment for CHF. Patient's clinical status may be compensated.

It is acceptable to report both Congestive Heart Failure Current and Past.

63. BNP, Three Times Normal

Variable name: BNP3X

Report if prior to surgery but within this admission, the BNP was at least three times the lab's upper limit of normal value.

For transfer patients, BNP from a transferring institution is acceptable.

20. Malignant Ventricular Arrhythmia

Variable Name: MAL_VENT

Recent (within the past 14 days) sustained ventricular tachycardia requiring electrical defibrillation or conversion with intravenous antiarrhythmic agents or ventricular fibrillation requiring electrical defibrillation. Excludes V-Tach or V-Fib occurring within 6 hours of the diagnosis of a myocardial infarction and responding well to treatment.

Interpretation: Sustained arrhythmia is that which continues until something is done to stop it; it does not resolve on its own.

If a patient is experiencing V-Tach or V-Fib that otherwise meets the above criteria, but is within 6 hours of an MI, you may still code this risk factor, IF the arrhythmia is not responding well to treatment. That is, if it continues despite electrical defibrillation or conversion with intravenous anti-arrhythmic agents.

If the patient has an AICD that is documented to have fired then CODE, unless the patient has had an MI within the last 6 hours.

Regular oral medication for a ventricular arrhythmia is NOT sufficient reason to code the risk factor.

III. Pre-Op Surgical Risk Factors (continued)

21. Chronic Lung Disease

Variable name: COPD

Indicate whether the patient has chronic lung disease, and the severity level according to the following classification:

- 1 No
- 2 Mild - FEV1 60% to 75% of predicted, and/or on chronic inhaled or oral bronchodilator therapy.
- 3 Moderate - FEV1 50% to 59% of predicted, and/or on chronic steroid therapy aimed at lung disease.
- 4 Severe - FEV1 <50% predicted, and/or Room Air pO₂ < 60 or Room Air pCO₂ > 50.

Interpretation: The diagnosis of chronic lung disease is not based solely on the fact that a person has or currently is smoking, or is on home oxygen. Diagnostic testing and or pharmacological criteria must be met. Chest x-ray is not included in the data specs for inclusion as chronic lung disease and should not be coded as “Yes.”

A history of chronic inhalation reactive disease (asbestosis, mesothelioma, black lung disease or pneumoconiosis) qualifies as chronic lung disease. Radiation induced pneumonitis or radiation fibrosis also qualifies as chronic lung disease. A history of atelectasis is a transient condition and does not qualify.

Chronic lung disease can include patients with chronic obstructive pulmonary disease, chronic bronchitis, or emphysema. It can also include a patient who is currently being chronically treated with inhaled or oral pharmacological therapy (e.g., beta-adrenergic agonist, anti-inflammatory agent, leukotriene receptor antagonist, or steroid). Patients with asthma or seasonal allergies are not considered to have chronic lung disease.

Coding Note: COPD definition is aligned with STS v2.73 data element 860.
Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.

III. Pre-Op Surgical Risk Factors (continued)

23. Extensive Aortic Atherosclerosis

Variable Name: CALCAORT

Ascending, transverse, and/or descending aortic atherosclerosis marked by either extensive calcification or luminal atheroma such that the intended surgical procedure is altered.

Interpretation: It is necessary to demonstrate that the intended surgical procedure is altered.

Documentation of the advanced aortic pathology by either transesophageal echocardiography, epi aortic echocardiography, intravascular ultrasound, magnetic resonance angiography or other imaging modality performed in the perioperative period should be available either by official report or dictated in the operative notes.

An operative note that dictates a change in the intended surgical procedure (i.e. clamp moved, procedure performed off pump) is acceptable documentation. Changes to the intended surgical procedure may also include documentation that more extensive evaluation/exploration of the aorta, for example epi aortic scanning, was performed.

Calcium in aortic arch on chest X-ray is not enough to code this risk.

24. Diabetes

Variable Name: DIABETES

Indicate whether patient has a history of diabetes diagnosed and/or treated by a physician.

Interpretation: The definition below is informational and data coordinator is not expected to diagnose diabetes.

The American Diabetes Association criteria include documentation of the following:

1. A1c $\geq 6.5\%$; or
2. Fasting plasma glucose ≥ 126 mg/dl (7.0 mmol/l); or
3. Two-hour plasma glucose ≥ 200 mg/dl (11.1 mmol/l) during an oral glucose tolerance test; or
4. In a patient with classic symptoms of hyperglycemia or hyperglycemic crisis, a random plasma glucose ≥ 200 mg/dl (11.1 mmol/l)

It does not include gestational diabetes.

Coding Note: *DIABETES* definition is aligned with STS V2.73 data element 780. Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.

III. Pre-Op Surgical Risk Factors (continued)

24a. Diabetes Therapy

Variable Name: DM_TRT

Indicate the control method the patient presented with on admission. Patients placed on a preprocedure diabetic pathway of insulin drip at admission but were previously controlled by diet or oral method are not coded as insulin treated. Choose the most aggressive therapy used prior to admission.

- 1 No treatment for diabetes
- 2 Diet treatment only
- 3 Oral agent treatment (includes oral agent with/without diet treatment)
- 4 Insulin treatment (includes any combination with insulin)
- 5 Other adjunctive therapy

Report this element for all cases where “Risk Factor #24 - Diabetes” is also reported. If the patient does not qualify for “Risk Factor #24 - Diabetes,” then leave the field blank or enter 0.

Coding Note: *DM_TRT* definition is aligned with STS v2.73 data element 790. *Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

25. Hepatic Failure

Variable Name: HEPATICF

The patient has cirrhosis or other liver disease
and has a bilirubin > 2 mg/dL
and a serum albumin < 3.5 g/dL.

27. Renal Failure, Dialysis

Variable Name: REN_DIAL

Indicate whether the patient is currently undergoing dialysis.

Interpretation: Includes any form of peritoneal or hemodialysis patient is receiving at the time of admission. Also, may include Continuous Veno-Venous Hemofiltration (CVVH, CVVH-D), and Continuous Renal Replacement Therapy (CRRT) as dialysis.

Code “No” for renal dialysis if ultrafiltration is the only documentation found in the record since this is for volume management

Coding Note: *REN_DIAL* definition is aligned with STS v2.73 data element 810 *Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

III. Pre-Op Surgical Risk Factors (continued)

30. Emergency Transfer to OR After Dx Cath

Variable Name: EME_CATH

The patient requires immediate surgery following a diagnostic catheterization.

31. Surgery For PCI Complication

Variable Name: EME_PCI

Indicate if there was a complication during PCI necessitating surgical intervention such as dissection or acute occlusion.

Coding Note: EME_PCI should be reported (file upload value of 1) when STS 1490 POCPCIndSurg = 1.

Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.

32. Previous PCI, This Episode of Care

Variable Name: PCITHIS

Indicate whether there was a previous Percutaneous Cardiac Intervention (PCI) performed within this episode of care. Include those at this facility and at some other acute care facility.

Coding Note: PCITHIS should be reported (file upload value of 1) when STS 1481 POCPCIWhen = 1 or 2.

Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.

33. PCI Before This Episode of Care

Variable Name: PCIBEFO

The patient has had a PCI before this episode of care.

38. Stent Thrombosis

Variable Name: THROMBOS

Formation of a blood clot/thrombus in the stented segment of an artery and/or adjacent area. This usually results in an acute occlusion, chest pain or development of an acute MI. Patient must be currently affected by stent thrombosis as evidenced by AMI, ACS, or clinical angina to code this risk factor.

Interpretation: An occlusion alone, plaque build-up or in-stent restenosis does not constitute coding. There must be documentation noting thrombus. The thrombus needs to be in or around the area that was stented for the risk factor to be coded.

III. Pre-Op Surgical Risk Factors (continued)

39. Any Previous Organ Transplant

Variable Name: ORGAN

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 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 corneal or skin transplant (grafting).

If the patient had a previous organ transplant and that organ was later removed, do not code this risk factor.

40. Heart Transplant Candidate

Variable Name: HT_TRANS

This risk factor should be coded when the patient is an approved heart transplant candidate before the start of the procedure.

Supporting documentation must be included in the patient's medical record showing that the patient was a transplant candidate prior to the start of the procedure. Acceptable documentation includes: notes that a pre-transplant evaluation was performed and patient was accepted, notes from the transplant coordinator that they have discussed this issue with the patient/family, or a note indicating the transplant patient's status based on UNOS urgency criteria.

During quarterly and annual data verification and validation efforts, we will be asking for supporting documentation for cases coded with this risk factor. Therefore, we highly recommend that at the time of coding you keep supporting documentation in a place for easy retrieval at a later date.

62. Active Endocarditis

Variable Name: ENDOCARD

Two or more positive blood cultures without other obvious source with demonstrated valvular vegetations or acute valvular dysfunction caused by infection.

Includes patients who are on antibiotics at the time of surgery.

Excludes patients who have completed antibiotic therapy and have no evidence of residual infection.

IV. Major Events Following Operation

Check to be sure that all of the listed major events occurred during or after the current cardiac surgery. Check at least one box in this section.

Please Note: A documented pre-operative condition that persists post-operatively with no increase in severity is not a major event. This is true even if the pre-operative condition is not part of this reporting system.

Unless otherwise specified, major events are only reported if they occur post-operatively, but before hospital discharge.

0. None

Variable Name: NOCOMPS

Check if none of the major events listed below occurred following the operation.

1. Stroke

Variable Name: STROKE

Indicate whether the patient has a postoperative stroke (i.e., any confirmed neurological deficit of abrupt onset caused by a disturbance in blood supply to the brain) that did not resolve within 24 hours.

Coding Note: *STROKE* definition is aligned with STS v2.73 data element 6030 *Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

2. Q-Wave MI

Variable Name: POSTMI

New Q waves occurring within 48 hours after surgery.

4. Deep Sternal Wound Infection

Variable Name: STERNINF

Indicate whether the patient, within 30 days post-op, had a deep sternal wound infection.

A deep incisional SSI (~~DIP or DIS~~) must meet the following criteria:

Infection occurs within 30 days after the operative procedure

and involves deep soft tissues (e.g., fascial and muscle layers) of the incision

and patient has at least 1 of the following:

- a. purulent drainage from the deep incision but not from the organ/space component of the surgical site

IV. Major Events Following Operation (continued)

4. Deep Sternal Wound Infection (continued)

- b. a deep incision spontaneously dehisces or is deliberately opened by a surgeon and is culture-positive or not cultured when the patient has at least 1 of the following signs or symptoms: fever ($>38^{\circ}\text{C}$), or localized pain or tenderness. A culture-negative finding does not meet this criterion.
- c. an abscess or other evidence of infection involving the deep incision is found on direct examination, during reoperation, or by histopathologic or radiologic examination
- d. diagnosis of a deep incisional SSI by a surgeon or attending physician.

Clarification: Report this element for deep sternal wound infection occurring anytime during the hospitalization or after discharge but within 30 days of the procedure.

Coding Note: *STERNINF* definition is aligned with STS v2.73 data element 5860 *Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

5. Bleeding Requiring Reoperation

Variable Name: BLEDREOP

If ~~a reoperation for bleeding took place~~ the patient was re-explored for mediastinal bleeding with or without tamponade either in the ICU, PACU or returned to the operating room, use the code below to indicate the time frame.

1. Acute (within 24 hours of the end of the case);
2. Late (more than 24 hours after the case ends).

Interpretation: Do not capture reopening of the chest or situations of excessive bleeding that occur prior to the patient leaving the operating room at the time of the primary procedure. Tamponade is a situation which occurs when there is compression or restriction placed on the heart within the chest that creates hemodynamic instability or a hypoperfused state. Do not include medically (non-operatively) treated excessive post-operative bleeding/tamponade events. Include patients that return to an OR suite or equivalent OR environment (i.e., ICU setting) as identified by your institution, that require surgical re-intervention to investigate/correct bleeding with or without tamponade. Include only those interventions that pertain to the mediastinum or thoracic cavity.

Code exactly 24 hours as acute.

Coding Note: *BLEDREOP* definition is aligned with STS v2.73 data element ~~5760~~ 5770

Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.

IV. Major Events Following Operation (continued)

8. Sepsis

Variable Name: SEPSIS

Sepsis is defined as evidence of serious infection accompanied by a deleterious systemic response.

In the time period of the first 48 postoperative or postprocedural hours, the diagnosis of sepsis requires the presence of a Systemic Inflammatory Response Syndrome (SIRS) resulting from a proven infection (such as bacteremia, fungemia or urinary tract infection). A systemic inflammatory response syndrome (SIRS) is present when at least two of the following criteria are present: hypo- or hyperthermia (>38.5 or <36.0), tachycardia or bradycardia, tachypnea, leukocytosis or leukopenia, and thrombocytopenia.

During the first 48 hours, a SIRS may result from the stress associated with surgery and/or cardiopulmonary bypass. Thus, the clinical criteria for sepsis during this time period should be more stringent.

In the time period after the first 48 postoperative or postprocedural hours, sepsis may be diagnosed by the presence of a SIRS resulting from suspected or proven infection.

Coding Note: *SEPSIS* definition is aligned with STS v2.73 data element 6010. *Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

9. G-I Event

Variable Name: GIBLEED

Indicate whether the patient had a postoperative occurrence of any GI event, including but not limited to:

- a. GI bleeding requiring transfusion;
- b. Pancreatitis with abnormal amylase/lipase requiring nasogastric (NG) suction therapy;
- c. Cholecystitis requiring cholecystectomy or drainage;
- d. Mesenteric ischemia requiring exploration;
- e. Hepatic failure;
- f. Prolonged ileus;
- g. *Clostridium difficile*

CLARIFICATION: GI events may require medical management, observational management or surgical intervention to control. DO NOT include events such as prolonged nausea and/or vomiting with no other documented physiological cause. Refer to the specific list included within the definition.

Example # 1: A patient has a placement of a Percutaneous Endoscopic Gastrostomy (PEG). Patients that receive PEG's are generally very sick patients that require long term nutritional support because of multiple postoperative complications and the inability to eat. If a PEG is placed in the stomach, it means that the stomach is working well enough to support the nutritional support that the PEG feedings are providing. Do not code a GI complication in this situation.

Example # 2: A patient experiences a postoperative paralytic ileus that does not increase the length of stay and does not require invasive therapy. Do not code a GI complication.

Example # 3: A patient has elevated liver enzymes postoperatively: A transient rise in the patient's liver enzymes does not represent a GI complication.

Coding Note: *GIBLEED* definition is aligned with STS v2.73 data element 6310. *Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

IV. Major Events Following Operation (continued)

10. Renal Failure

Variable Name: RENAL_FAI

Indicate whether the patient had a new requirement for dialysis postoperatively, which may include hemodialysis, peritoneal dialysis.

Interpretation: May include either hemo or peritoneal dialysis. This includes a onetime need for dialysis as well as implementation of longer term therapy. If the patient was on preoperative peritoneal dialysis and moved to hemodialysis postoperatively, this does not constitute a worsening of the condition and should not be coded as an event.

Continuous Veno-Venous Hemofiltration) (CVVH, CVVH-D) and Continuous Renal Replacement Therapy (CRRT) should be coded here as “Yes.”

Coding Note: *RENAL_FAI* definition is aligned with STS v2.73 data element 6210. *Society of Thoracic Surgeons, Adult Cardiac Surgery Database, Version 2.73, used with permission.*

13. Respiratory Failure

Variable Name: RESP_FAI

Pulmonary insufficiency requiring intubation and ventilation for a period of 72 hours or more, at any time during the post-operative stay. For patients who are placed on and taken off ventilation several times, the total of these episodes should be 72 hours or more.

Interpretation: If the patient is intubated for 72 or more hours after surgery this major event should be coded, even if the patient was intubated prior to the procedure.

The following scenario would be coded:

Patient was extubated 48 hours post-op. Patient was re-intubated sometime the next day. Patient was extubated 32 hours later.

It is not necessary to show that the prolonged ventilatory dependence was due to respiratory failure.

IV. Major Events Following Operation (continued)

14. Unplanned Cardiac Reoperation or Interventional Procedure

Variable Name: UNPLANREOP

Any unplanned cardiac reoperation or percutaneous coronary intervention that is required as a result of the current cardiac surgery. This would exclude a reoperation to control bleeding that occurs within 36 hours of the surgery.

Interpretation: This major event should be reported for any cardiac surgery, not just those reportable in CSRS. Procedures should be directly related to the heart. Examples of reportable surgeries include but are not limited to: CABG, cardiac massage, or cardiac explorations. Some examples of the procedures not reportable are: pacemaker insertion, pericardiocentesis, and pleurocentesis.

If the chest is left open after surgery with a return to the operating room to close, this would not be considered an unplanned cardiac reoperation. If clots need to be removed from an open chest this would not be considered an unplanned cardiac reoperation.

The procedure does not have to be performed in the operating room or cath lab.

This event would not be coded under the following situation: the patient has a reoperation to control bleeding, ~~less than 36 hours after surgery and then goes back greater than 36 hours to once again control bleeding.~~ In this instance coding the major event "5 - Bleeding Requiring Reoperation" is sufficient.

V. Discharge Information

Discharged Alive To

Variable Name: STATUS, DISWHERE

Check the appropriate box.

If a patient is discharged to hospice (including home with hospice), the discharge status should be reported with code “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 on page 13 of the “CSRS Data Reporting Policies”

If the patient came from a prison or correctional facility and is being discharged back to the same setting then “11 – Home” would be coded.

If the patient is discharged to sub-acute rehab that is in a skilled nursing facility then the discharge status would be “14”, if it is unknown where the sub-acute rehab facility is located then the discharge status would be “19”.

If the patient is discharged to an inpatient physical medicine and rehabilitation unit the discharge status should be “15.”

“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.

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.

V. Discharge Information (continued)

30 Day Status

Variable Name: THIRTYDAY

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

VI. Person Completing Report

Name

This space is provided as an aid to the hospital. Enter the name and telephone number of the person completing the report, and the date the report was completed. This field is not required and is not used by the Department of Health. It is provided solely for the use of the individual hospitals.

This field appears only on the hard copy form, it is not part of data entry or file specification for transmission to cardiac services program.

Referring Physician

Variable Name: REF_PHYS

This space is provided as an aid to the hospital. It is intended to allow the name of the referring cardiologist or primary care physician to be entered. For many hospitals this is useful for tracking 30-day status. By entering the name of the referring physician case lists can be generated and sent to the referring physician for follow-up. This field is not required and is not used by the Department of Health. It is provided solely for the use of the individual hospitals.

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 Medical 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

Congenital and Acquired Cardiac Procedure Codes NYSDOH CARDIAC ADVISORY COMMITTEE

100-398 Congenital Heart Disease - Operations With or Without Extracorporeal Circulation

Note: Extracorporeal circulation will be determined from the data element Entire Procedure Off Pump reported under Section II. Procedural Information on the front of the form. Please accurately complete this item for all appropriate cases.

Anomalies of Pulmonary Veins

- 100 Repair of Anomalous Pulmonary Venous Return
- 101 Repair of Pulmonary Vein Stenosis
- 103 Repair of Partial Anomalous Pulmonary Venous Return

Anomalies of Atrial Septum

- 120 ASD Closure
- 121 Creation of ASD
- 122 Repair of Cor Triatriatum
- 123 PFO Closure

Atrioventricular Septal Defect (AVSD)

- 130 Repair of Complete AV Canal
- 131 Repair of Partial AV Canal

Anomalies of Ventricular Septum

- 140 Repair of VSD
- 141 Creation/Enlargement of VSD
- 142 Fenestration of VSD Patch

Anomalies of Atrioventricular Valves

Tricuspid Valve

- 150 Repair (Non-Ebstein's Valve)
Replacement
- 151 Homograft
- 152 Prosthetic
- 153 Tricuspid Valve Closure
- 154 Repair Ebstein's Anomaly

Anomalies of Atrioventricular Valves (continued)

Mitral Valve

- 160 Resect supramitral ring
- 161 Repair (including annuloplasty)
Replacement
- 162 Homograft
- 163 Prosthetic
- 170 Common AV Valve Repair

Anomalies of Ventricular Outflow Tract(s)

Pulmonary Ventricular Outflow Tract

- 180 Pulmonary Valvotomy/Valvectomy
- 181 Resection of subvalvular PS
- 182 Repair of supra-ventricular PS
Pulmonary Valve Replacement
- 190 Homograft
- 191 Prosthetic
- 192 Xenograft

Pulmonary Outflow Conduit

- Valved
- 200 Homograft
- 201 Prosthetic
- 202 Non-Valved
- Transannular Patch
- 210 With Monocusp Valve
- 211 Without Monocusp Valve
- 212 Repair Branch PS

Aortic Ventricular Outflow Tract

- 220 Aortic Valvuloplasty
- 221 Aortic Valvotomy
- 230 Repair Supra-ventricular AS
- 231 Resection of Discrete Subvalvular AS
- 235 Aortoventriculoplasty (Konno Procedure)
Aortic Valve Replacement
- 240 Autograft (Ross Procedure)
- 241 Homograft
- 242 Prosthetic
- 243 Heterograft
- Aortic Root Replacement
- 250 Autograft (Ross Procedure)
- 251 Homograft
- 252 Prosthetic
- 255 LV Apex to Aorta Conduit

Tetralogy of Fallot

- 260 Repair with Pulmonary Valvotomy
- 261 Repair with Transannular Patch
- 262 Repair with Non-valved Conduit
Repair with Valved Conduit
- 263 Homograft
- 264 Prosthetic
- 265 Repair with reduction/plasty of PAs
Repair with pulmonary valve replacement
- 266 Homograft
- 267 Prosthetic

Truncus Arteriosus

- 262 Repair with Non-Valved Conduit
Repair with Valved Conduit
- 263 Homograft
- 264 Prosthetic

Univentricular Heart (Single Ventricle)

- Fontan Operations
- 270 Direct RV-PA Connection
Total Cavopulmonary Connection
- 271 Lateral tunnel – nonfenestrated
- 272 Lateral tunnel – fenestrated
- 273 Extracardiac – nonfenestrated
- 274 Extracardiac – fenestrated
- 275 Septation of Single Ventricle
Hypoplastic Right Ventricle
Valved
- 200 Homograft
- 201 Prosthetic
- 202 Non-Valved
Transannular Patch
- 210 With Monocusp Valve
- 211 Without Monocusp Valve
- Hypoplastic Left Ventricle
- 280 Norwood
- 290 Damus Kaye Stansel (DSK)

Transposition of Great Arteries or Double Outlet RV

- 310 Arterial Switch
- 311 Senning Procedure
- 312 Mustard Procedure
- 313 Intraventricular Repair of DORV

Transposition of Great Arteries or Double Outlet RV (continued)

- Rastelli Procedure
 - RV-PA Conduit
 - Valved
 - 320 Homograft
 - 321 Prosthetic
 - 322 Non-Valved
 - 325 REV operation (Modified Rastelli)
 - LV-PA Conduit
 - Valved
 - 326 Homograft
 - 327 Prosthetic
 - 328 Non-Valved

Great Vessel Anomalies

- 330 PDA Ligation
- 331 Repair Aortopulmonary Window
- 332 Reimplantation of left or right pulmonary artery
- 333 Repair Sinus of Valsalva Aneurysm
- Aortic Repair (Coarctation or Interruption)
 - 340 End to end anastomosis
 - 348 End to side anastomosis
 - 341 Subclavian flap angioplasty
 - 342 Onlay Patch
 - 343 Interposition graft
- 344 Vascular Ring Division
- 345 Repair of PA Sling
- 346 Reimplantation of Innominate Artery
- 347 Aortoplexy

Coronary Artery Anomalies

- Translocation of LCA to Aorta
 - 350 Direct
 - 351 Transpulmonary Tunnel (Takeuchi)
- 352 Coronary Artery Ligation
- 353 Coronary Fistula Ligation

Cardiomyopathies

- 360 Left Ventricular Reconstruction (Batiste Procedure, Surgical Ventricular Restoration)
- 361 Radical Myomectomy

Interval Procedures

- 370 Pulmonary Artery Band
- 375 Unifocalization of Pulmonary Vessels
Shunts
- 381 Central Aortopulmonary Shunt
Blalock Taussig Shunts
- 382 Classical
- 383 Modified
Glenn Shunts
- 384 Unidirectional (Classical)
- 385 Bidirectional
- 386 Bilateral Bidirectional
- 390 Cardiac Arrhythmia Surgery
- 398 Other Operations for Congenital Heart Disease

400-998 Acquired Heart Disease – Operations Performed With or Without Extracorporeal Circulation

- 401 Mitral Valvotomy
- 402 Pericardiectomy (with extracorporeal circulation)
- 403 Stab Wound of Heart or Great Vessel Repair (without extracorporeal
circulation)
- 404 Saccular Aortic Aneurysm

Repair Of Aortic Deceleration Injury

- 420 With Shunt
- 421 Without Shunt

Other

- 498 Other Operation for Acquired Heart Disease (without extracorporeal
circulation)

Valve Repair

- 500 Aortic
- 501 Mitral
- 502 Tricuspid
- 503 Pulmonary

Valve Replacement

- 510-518* Ross Procedure
- 520-528* Aortic Mechanical
- 530-538* Aortic Heterograft
- 540-548* Aortic Homograft

Valve Replacement (continued)

550-558*	Mitral Mechanical
560-568*	Mitral Heterograft
600-608*	Mitral Homograft
570-578*	Tricuspid Mechanical
580-588*	Tricuspid Heterograft
590-598*	Pulmonary

*REOPERATIONS: For Valve Replacement (510-608), use third digit to indicate reason for reoperation, as below. Note, the information below is specific to the valve reported. For example, a patient with previous aortic valve replacement who is now having mitral valve replacement (mechanical) would be reported using code 550 because this is not a re-operation on the mitral valve. In the event of multiple valve surgery, the third digit may be different for each valve code reported, i.e. one valve may be a re-op and the other(s) may not.

Use code 7 – Complication of Transcatheter Valve Replacement in the event of an unsuccessful Transcatheter Valve Replacement which requires surgical valve replacement.

0 Not a Reoperation	5 Disease of Another Valve
1 Periprosthetic Leak	6 Failed Catheter-based Valve Repair
2 Prosthetic Endocarditis	7 Complication of Transcatheter Valve Replacement
3 Prosthetic Malfunction	
4 Failed Surgical Valve Repair	8 Other Reason

Adjunct Valve Information

	Transcatheter Valve Replacement
640	Transfemoral Approach
641	Transapical Approach
642	Subclavian Approach
643	Direct Aortic Approach

Note: Use these codes in conjunction with the valve replacement codes above to indicate if the valve replacement was performed using a transcatheter (transcutaneous) approach. You must also report the appropriate code for valve replacement. Report these procedures no matter where in the hospital they are performed.

Valve Conduits

660	Apical Aortic Conduit
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Note: Record aortic valve and ascending aorta replacement under aneurysms.

Coronary Artery Bypass Grafts

670 Coronary Artery Bypass Graft

Please Note: If you code a 670 then you must complete the CABG Information under the Procedural Information section of the form.

Other Revascularization

710 Transmyocardial Revascularization

711 Percutaneous Coronary Intervention in the same setting as CABG or Valve surgery

715 Growth Factor Installation

Additional Procedures with or without CABG

760 Acquired Ventricular Septal Defect

761 Resection or Plication of LV Aneurysm

762 Ventricular Reconstruction (Batiste Procedure, Surgical Ventricular Restoration)

763 Carotid Endarterectomy (report only if done with another reportable cardiac surgical procedure)

764 Implantation of AICD (report only if done with another reportable cardiac surgical procedure)

Radiofrequency or Operative Ablation

770 Atrial

771 Ventricular

772 Maze Procedure

Aortic Aneurysm Repair/Aortic Root Replacement

780 Ascending Aorta, With Graft, With Coronary Reimplantation

781 Ascending Aorta, Replacement or Repair, Without Coronary Reimplantation

782 Transverse Aorta

783 Descending Thoracic Aorta (Excluding Acute Deceleration Injury)

784 Thoracoabdominal

785 Aortic Root Replacement or Repair, With Graft, With Coronary Reimplantation

Dissecting Aneurysm Surgery

800 Intraluminal Graft

801 Intraluminal Graft with Aortic Valve Suspension

802 Tube Graft with Aortic Valve Suspension

803 Tube Graft with Aortic Valve Replacement

818 Other Dissecting Aneurysm Surgery

Transplant Procedures

- 820 Heart Transplant
- 821 Heart and Lung Transplant
- 822 Lung Transplant
- 830 Left Ventricular Assist Device (LVAD) – Extracorporeal
- 831 Left Ventricular Assist Device (LVAD) – Implantable
- 832 Right Ventricular Assist Device (RVAD)
- 833 Bi-Ventricular Assist Device (BIVAD)
- 834 Extracorporeal Membrane Oxygenation (ECMO)
- 840 Ventricular Assist Device as a Destination Therapy (must also code either 830 or 831)
- 901 Artificial Heart

Other

- 902 Pulmonary Embolectomy
- 903 Stab Wound of Heart or Great Vessel Repair (with extracorporeal circulation)
- 904 Removal of Intracardiac Tumor
- 905 Removal of Intracardiac Catheter (surgical)
- 906 Repair of Aortic Deceleration Injury (With Aortofemoral Bypass)
- 907 Repair of a Cardiac Laceration due to Trauma
- 915 Septal Myomectomy
- 916 Ventricular Myomectomy
- 920 Ventricular Free Wall Rupture
- 998 Other Operation for Acquired Heart Disease (with extracorporeal circulation)

Attachment E

Congenital Cardiac Diagnosis Codes¹

SEPTAL DEFECTS

ASD

- 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.

VSD

- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

SEPTAL DEFECTS (CONTINUED)

AV Canal

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.

AV Window

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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

SEPTAL DEFECTS (CONTINUED)

AV Window (continued)

- 140 AP window (aortopulmonary window) (continued)**
interrupted aortic arch, code "Interrupted aortic arch + AP window (aortopulmonary window)", and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and AP window separately to provide further documentation about the individual interrupted arch and AP window types.)
- 150 Pulmonary artery origin from ascending aorta (hemitruncus)**
One pulmonary artery arises from the ascending aorta and the other pulmonary artery arises from the right ventricle. DOES NOT include origin of the right or left pulmonary artery from the innominate artery or the aortic arch via a patent ductus arteriosus or collateral artery.

Truncus Arteriosus

- 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.}

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Attachment E

Congenital Cardiac Diagnosis Codes¹

PULMONARY VENOUS ANOMALIES

Partial Anomalous Pulmonary Venous Connection

- 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.

Total Anomalous Pulmonary Venous Connection

- 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.

COR TRIARIATUM

- 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 supralvalvar 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 supralvalvar 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

PULMONARY VENOUS STENOSIS

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).

SYSTEMIC VENOUS ANOMALIES

Anomalous Systemic Venous Connection

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.

Systemic Venous Obstruction

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.

RIGHT HEART LESIONS

Tetralogy of Fallot

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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

RIGHT HEART LESIONS (CONTINUED)

Tetralogy of Fallot (continued)

290 TOF (continued)

defect, right aortic arch, left superior vena cava, and coronary artery anomalies must be subspecified as an additional or secondary diagnosis under the primary TOF diagnosis. TOF with absent pulmonary valve or TOF with associated complete atrioventricular canal are NOT to be secondary diagnoses under TOF - they are separate entities and should be coded as such. Controversy surrounds the differentiation between TOF and double outlet right ventricle [DORV]; in the nomenclature used here, DORV is defined as a type of ventriculoarterial connection in which both great vessels arise predominantly from the right ventricle. TOF with pulmonary atresia is to be coded under "Pulmonary atresia-VSD.")

2140 TOF, Pulmonary stenosis

Indicate if the patient has the diagnosis of "TOF, Pulmonary stenosis". Use this diagnosis if the patient has tetralogy of Fallot and pulmonary stenosis. Do not use this diagnosis if the patient has tetralogy of Fallot and pulmonary atresia. Do not use this diagnosis if the patient has tetralogy of Fallot and absent pulmonary valve. Do not use this diagnosis if the patient has tetralogy of Fallot and atrioventricular canal. {Tetralogy of Fallot is defined as a group of malformations with biventricular atrioventricular alignments or connections characterized by anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta. Hearts with tetralogy of Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, and aortic override; 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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

RIGHT HEART LESIONS (CONTINUED)

Tetralogy of Fallot (continued)

310 TOF, Absent pulmonary valve (continued)

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.)

Pulmonary Atresia

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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

RIGHT HEART LESIONS (CONTINUED)

Pulmonary Atresia (continued)

- 350 Pulmonary atresia, VSD-MAPCA (pseudotruncus) (continued)**
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

Tricuspid Valve Disease and Ebstein's Anomaly

- 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)

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Attachment E

Congenital Cardiac Diagnosis Codes¹

RIGHT HEART LESIONS (CONTINUED)

Tricuspid Valve Disease and Ebstein's Anomaly (continued)

- 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.

RVOT Obstruction and/or Pulmonary Stenosis

- 420 Pulmonary stenosis, Valvar**
Pulmonary stenosis, Valvar ranges from critical neonatal pulmonic valve stenosis with hypoplasia of the right ventricle to valvar pulmonary stenosis in the infant, child, or adult, usually better tolerated but potentially associated with infundibular stenosis. Pulmonary branch hypoplasia can be associated. Only 10% of neonates with Pulmonary stenosis, Valvar with intact ventricular septum have RV-to-coronary artery fistula(s). An RV dependent coronary artery circulation is present when coronary artery fistulas (coronary sinusoids) are associated with a proximal coronary artery stenosis; this occurs in only 2% of neonates with Pulmonary stenosis, Valvar with IVS.
- 430 Pulmonary artery stenosis (hypoplasia), Main (trunk)**
Indicate if the patient has the diagnosis of "Pulmonary artery stenosis (hypoplasia), Main (trunk)". "Pulmonary artery stenosis (hypoplasia), Main (trunk)" is defined as a congenital or acquired anomaly with pulmonary trunk (main pulmonary artery) narrowing or hypoplasia. The stenosis or hypoplasia may be isolated or associated with other cardiac lesions. Since the narrowing is distal to the pulmonic valve, it may also be known as supra-valvar 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

RIGHT HEART LESIONS (CONTINUED)

RVOT Obstruction and/or Pulmonary Stenosis (continued)

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.

Pulmonary Valve Disease

510 Pulmonary valve, Other

Other anomalies of the pulmonary valve may be listed here including but not restricted to absent pulmonary valve.

530 Pulmonary insufficiency

Pulmonary valve insufficiency or regurgitation may be due to congenital factors (primary annular dilation, prolapse, leaflet underdevelopment, etc.) or acquired (for example, post cardiac surgery for repair of tetralogy of Fallot, etc.).

540 Pulmonary insufficiency and pulmonary stenosis

Pulmonary valve insufficiency and pulmonary stenosis beyond the neonatal period, in infancy and childhood, may be secondary to leaflet tissue that has become thickened and myxomatous. Retraction of the commissure attachment frequently creates an associated supralvalvar stenosis

SHUNT FAILURE

Shunt Failure

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”.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

CONDUIT FAILURE

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".

LEFT HEART LESIONS

Aortic Valve Disease

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 supralvalvar mitral ring, parachute mitral valve, and coarctation of aorta. Subvalvar aortic stenosis may be categorized into two types: localized subvalvar aortic stenosis, which consists of a fibrous or fibromuscular ridge, and diffuse tunnel subvalvar aortic stenosis, in which circumferential narrowing commences at the annular level and extends downward for 1-3 cm. Idiopathic hypertrophic subaortic stenosis (IHSS) is also known as hypertrophic obstructive cardiomyopathy (HOCM), and is characterized by a primary hypertrophy of the myocardium. The obstructive forms involve different degrees of dynamic subvalvar aortic obstruction from a thickened ventricular wall and anterior motion of the mitral valve. Definitive nomenclature and therapeutic options for IHSS are listed under cardiomyopathy.

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 raphe 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, Supralvalvar

Congenital supralvalvar 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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

LEFT HEART LESIONS (CONTINUED)

Aortic Valve Disease (continued)

570 Aortic stenosis, Supravalvar (continued)

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 neo-aortic reconstruction such as arterial switch, Ross operation, or Norwood procedure; (2) at a suture line from a previous aortotomy or cannulation; and (3) from a narrowed conduit.

590 Aortic valve atresia

Aortic valve atresia will most often be coded under the Hypoplastic left heart syndrome/complex diagnostic codes since it most often occurs as part of a spectrum of cardiac malformations. However, there is a small subset of patients with aortic valve atresia who have a well developed left ventricle and mitral valve and a large VSD (nonrestrictive or restrictive). The diagnostic code "Aortic valve atresia" enables users to report those patients with aortic valve atresia and a well-developed systemic ventricle without recourse to either a hypoplastic left heart syndrome/complex diagnosis or a single ventricle diagnosis.

600 Aortic insufficiency

Congenital aortic regurgitation/insufficiency is rare as an isolated entity. There are rare reports of congenital malformation of the aortic valve that result in aortic insufficiency shortly after birth from an absent or underdeveloped aortic valve cusp. Aortic insufficiency is more commonly seen with other associated cardiac anomalies: (1) in stenotic aortic valves (commonly stenotic congenital bicuspid aortic valves) with some degree of aortic regurgitation due to aortic leaflet abnormality; (2) in association with a VSD (especially in supravalvar or conal type I VSD, more commonly seen in Asian populations); (3) secondary to aortic-left ventricular tunnel; (4) secondary to tethering or retraction of aortic valve leaflets in cases of supravalvar aortic stenosis that may involve the aortic valve; and similarly (5) secondary to encroachment on an aortic cusp by a subaortic membrane; or (6) turbulence caused by a stenotic jet can create progressive aortic regurgitation. Aortic insufficiency may also result from: (1) post-procedure such as closed or open valvotomy or aortic valve repair, VSD closure, balloon valvotomy, or diagnostic catheterization; (2) in the neo-aorta post arterial switch, pulmonary autograft (Ross) procedure, homograft placement, Norwood procedure, or Damus-Kaye-Stansel procedure; (3) as a result of endocarditis secondary to perforated or prolapsed leaflets or annular dehiscence; (4) secondary to 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

LEFT HEART LESIONS (CONTINUED)

Sinus of Valsalva Fistula/Aneurysm

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.

LV to Aorta Tunnel

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.

Mitral Valve Disease

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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

LEFT HEART LESIONS (CONTINUED)

Mitral Valve Disease (continued)

660 Mitral stenosis, Valvar

Valvar mitral stenosis may arise from congenital (annular and / or leaflet) or acquired causes, both surgical (after mitral valve repair or replacement or other cardiac surgery) and non-surgical (post rheumatic heart disease, infective endocarditis, ischemia, myxomatous degeneration, trauma, or cardiomyopathy). Mitral valve annular hypoplasia is distinguished from severe mitral valve hypoplasia and mitral valve atresia, which are typically components of hypoplastic left heart syndrome. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.

670 Mitral stenosis, Subvalvar

Congenital subvalvar mitral stenosis may be due to obstructive pathology of either the chordae tendineae and / or papillary muscles which support the valve leaflets. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.

680 Mitral stenosis, Subvalvar, Parachute

In parachute mitral valve, all chordae are attached to a single papillary muscle originating from the posterior ventricular wall. When the interchordal spaces are partially obliterated valvar stenosis results. This defect also causes valvar insufficiency, most commonly due to a cleft leaflet, a poorly developed anterior leaflet, short chordae, or annular dilatation. This lesion is also part of Shone's anomaly, which consists of the parachute mitral valve, supralvalvar mitral ring, subaortic stenosis, and coarctation of the aorta. When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.

695 Mitral stenosis

Stenotic lesions of the mitral valve not otherwise specified in the diagnosis definitions 650, 660, 670, and 680.

700 Mitral regurgitation and mitral stenosis

Mitral regurgitation and mitral stenosis may arise from congenital or acquired causes or after cardiac surgery. Additional details to aid in coding specific components of the diagnosis are available in the individual mitral stenosis or mitral regurgitation field definitions. When coding multiple mitral valve lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.

710 Mitral regurgitation

Mitral regurgitation may arise from congenital (at the annular, leaflet or subvalvar level) or acquired causes both surgical (after mitral valve repair or replacement, subaortic stenosis repair, atrioventricular canal repair, cardiac transplantation, or other cardiac surgery) and non-surgical (post rheumatic heart disease, infective endocarditis, ischemia (with chordal rupture or papillary muscle infarct), myxomatous degeneration including Barlow's syndrome, trauma, or cardiomyopathy). Congenital lesions at the annular level include annular dilatation or deformation (usually deformation is consequent to associated lesions). At the valve leaflet level, mitral regurgitation may be due to a cleft, hypoplasia or agenesis of leaflet(s), excessive leaflet tissue, or a double orifice valve. At the subvalvar level, mitral regurgitation may be secondary to chordae tendineae anomalies (agenesis, rupture, elongation, or shortening as in funnel valve), or to papillary muscle anomalies (hypoplasia or agenesis, shortening, elongation, single-parachute, or multiple-hammock valve). When coding multiple mitral valvar lesions the predominant defect causing the functional effect (regurgitation, stenosis, or regurgitation and stenosis) should be listed as the primary defect.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

LEFT HEART LESIONS (CONTINUED)

Mitral Valve Disease (continued)

720 Mitral valve, Other

Mitral valve pathology not otherwise coded in diagnosis definitions 650 through 710.

Hypoplastic Left Heart Syndrome

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.

Shone's Syndrome

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) supralvalvar 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", supralvalvar 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.

CARDIOMYOPATHY

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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

CARDIOMYOPATHY (CONTINUED)

- 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.

PERICARDIAL DISEASE

- 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.

SINGLE VENTRICLE

- 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.
- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

SINGLE VENTRICLE (CONTINUED)

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.

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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

SINGLE VENTRICLE (CONTINUED)

- 851 Single Ventricle + Total anomalous pulmonary venous connection (TAPVC) (continued)**
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.

TRANSPOSITION OF THE GREAT ARTERIES

Congenitally Corrected TGA

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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

TRANSPOSITION OF THE GREAT ARTERIES (CONTINUED)

Congenitally Corrected TGA (continued)

872 Congenitally corrected TGA, IVS (continued)

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.)

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*

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Attachment E

Congenital Cardiac Diagnosis Codes¹

TRANSPOSITION OF THE GREAT ARTERIES (CONTINUED)

Congenitally Corrected TGA (continued)

- 878 Congenitally corrected TGA, VSD-LVOTO (continued)**
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.)

Transposition of the Great Arteries

- 880 TGA, IVS**
A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with an intact ventricular septum. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
- 890 TGA, IVS-LVOTO**
A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with an intact ventricular septum and associated left ventricular obstruction. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
- 900 TGA, VSD**
A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with one or more ventricular septal defects. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).
- 910 TGA, VSD-LVOTO**
A malformation of the heart in which there is atrioventricular concordance and ventriculoarterial discordance with one or more ventricular septal defects and left ventricular outflow tract obstruction. There may be d, l, or ambiguous transposition (segmental diagnoses include S,D,D, S,D,L, S,D,A). Also to be included in this diagnostic grouping are those defects with situs inversus, L-loop ventricles and either d or l transposition (segmental diagnosis of I,L,L and I,L,D) and occasionally those defects with ambiguous situs of the atria which behave as physiologically uncorrected transposition and are treated with arterial switch (segmental diagnoses include A,L,L and A,D,D).

DORV

- 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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

DORV (CONTINUED)

- 930 DORV, VSD type (continued)**
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.
- 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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

DORV (CONTINUED)

2030 DORV + AVSD (AV Canal) (continued)

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.

DOLV

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.

THORACIC ARTERIES AND VEINS

Coarctation of Aorta and Aortic Arch Hypoplasia

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.)

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Attachment E

Congenital Cardiac Diagnosis Codes¹

THORACIC ARTERIES AND VEINS (CONTINUED)

Coarctation of Aorta and Aortic Arch Hypoplasia (continued)

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.}

Coronary Artery Anomalies

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.

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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

THORACIC ARTERIES AND VEINS (CONTINUED)

Coronary Artery Anomalies (continued)

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).

Interrupted Arch

1070 Interrupted aortic arch

Indicate if the patient has the diagnosis of "Interrupted aortic arch". Interrupted aortic arch is defined as the loss of luminal continuity between the ascending and descending aorta. In most cases blood flow to the descending thoracic aorta is through a PDA, and there is a large VSD. Arch interruption is further defined by site of interruption. In type A, interruption is distal to the left subclavian artery; in type B interruption is between the left carotid and left subclavian arteries; and in type C interruption occurs between the innominate and left carotid arteries.

2020 Interrupted aortic arch + VSD

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

2000 Interrupted aortic arch + AP window (aortopulmonary window)

Indicate if the patient has the diagnosis of "Interrupted aortic arch + AP window (aortopulmonary window)". In the event of interrupted aortic arch occurring in association with AP window, code "Interrupted aortic arch + AP window (aortopulmonary window)", and then use additional (secondary) diagnostic codes to describe the interrupted aortic arch and the AP window separately to provide further documentation about the individual interrupted aortic arch and AP window types. {Interrupted aortic arch is defined as 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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

THORACIC ARTERIES AND VEINS (CONTINUED)

Interrupted Arch (continued)

2000 Interrupted aortic arch + AP window (aortopulmonary window) (continued)

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.

Patent Ductus Arteriosus

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.)

Vascular Rings and Slings

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.

Aortic Aneurysm

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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

THORACIC ARTERIES AND VEINS (CONTINUED)

Aortic Dissection

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).

THORACIC AND MEDIASTINAL DISEASE

Lung Disease

1130 Lung disease, Benign

Lung disease arising from any etiology (congenital or acquired) which does not result in death or lung or heart-lung transplant; examples might be non-life threatening asthma or emphysema, benign cysts

1140 Lung disease, Malignant

Lung disease arising from any etiology (congenital or acquired, including pulmonary parenchymal disease, pulmonary vascular disease, congenital heart disease, neoplasm, etc.) which may result in death or lung or heart-lung transplant.

Pectus Excavatum, Carinatum

1150 Pectus

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

Tracheal Stenosis

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

Pleural Disease

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 Codes¹

THORACIC AND MEDIASTINAL DISEASE (CONTINUED)

Pleural Disease (continued)

- 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.

Esophageal Disease

- 1490 Esophageal disease, Benign**
Any benign disease of the esophagus
- 1500 Esophageal disease, Malignant**
Any malignant disease of the esophagus

Mediastinal Disease

- 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.

Diaphragmatic Disease

- 1540 Diaphragm paralysis**
Paralysis of diaphragm, unilateral or bilateral
- 1550 Diaphragm disease, Other**
Any disease of the diaphragm other than paralysis.

ELECTROPHYSIOLOGICAL

- 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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

ELECTROPHYSIOLOGICAL (CONTINUED)

- 2050 Arrhythmia, Junctional (continued)**
rhythm [1]. [1]. Jacobs JP. (Editor). 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, Volume 18, Supplement S2, pages 1 – 530, December 9, 2008, page 379.
- 2060 Arrhythmia, Ventricular**
Indicate if the patient has the diagnosis of “Arrhythmia, Ventricular”. “Arrhythmia, Ventricular” ROOT Definition = Abnormal rhythm originating from the ventricles [1]. [1]. Jacobs JP. (Editor). 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, Volume 18, Supplement S2, pages 1 – 530, December 9, 2008, page 393
- 1185 Arrhythmia, Heart block**
Atrioventricular block may be congenital or acquired, and may be of varying degree (first, second, or third degree).
- 1190 Arrhythmia, Heart block, Acquired**
Atrioventricular block, when acquired, may be post-surgical, or secondary to myocarditis or other etiologies; the block may be first, second or third degree.
- 1200 Arrhythmia, Heart block, Congenital**
Atrioventricular block, when congenital, may be first, second or third degree block.
- 1220 Arrhythmia, Pacemaker, Indication for replacement**
Indications for pacemaker replacement may include end of generator life, malfunction, or infection.

MISCELLANEOUS, OTHER

- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

MISCELLANEOUS, OTHER (CONTINUED)

2090 Dextrocardia (continued)

Cardiology in the Young, Volume 17, Supplement 2, pages 1–28, doi: 10.1017/S1047951107001138, September 2007.

2100 Levocardia

Indicate if the patient has the diagnosis of “Levocardia”. “Levocardia” usually considered synonymous with a left-sided ventricular mass, whilst “levoverision” 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

MISCELLANEOUS, OTHER (CONTINUED)

- 1270 Aneurysm, Pulmonary artery**
An aneurysm of the pulmonary artery is defined as a localized dilation or enlargement of the pulmonary artery trunk and its central branches (right and left pulmonary artery).
- 1280 Aneurysm, Other**
A localized dilation or enlargement of a cardiac vessel or chamber not coded in specific fields available for aortic aneurysm, sinus of Valsalva aneurysm, coronary artery aneurysm, right ventricular aneurysm, left ventricular aneurysm, or pulmonary artery aneurysm.
- 1290 Hypoplastic RV**
Small size of the right ventricle. This morphological abnormality usually is an integral part of other congenital cardiac anomalies and, therefore, frequently does not need to be coded separately. It should, however, be coded as secondary to an accompanying congenital cardiac anomaly if the right ventricular hypoplasia is not considered an integral and understood part of the primary congenital cardiac diagnosis. It would rarely be coded as a primary and/or isolated diagnosis.
- 1300 Hypoplastic LV**
Small size of the left ventricle. This morphological abnormality usually is an integral part of other congenital cardiac anomalies and, therefore, frequently does not need to be coded separately. It should, however, be coded as secondary to an accompanying congenital cardiac anomaly if the left ventricular hypoplasia is not considered an integral and understood part of the primary congenital cardiac diagnosis. It would rarely be coded as a primary and/or isolated diagnosis.
- 2070 Postoperative bleeding**
Indicate if the patient has the diagnosis of "Postoperative bleeding"
- 1310 Mediastinitis**
Inflammation/infection of the mediastinum, the cavity between the lungs which holds the heart, great vessels, trachea, esophagus, thymus, and connective tissues. In the United States mediastinitis occurs most commonly following chest surgery.
- 1320 Endocarditis**
An infection of the endocardial surface of the heart, which may involve one or more heart valves (native or prosthetic) or septal defects or prosthetic patch material placed at previous surgery.
- 1325 Rheumatic heart disease**
Heart disease, usually 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".

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Attachment E

Congenital Cardiac Diagnosis Codes¹

MISCELLANEOUS, OTHER (CONTINUED)

1340 Myocardial infarction

A myocardial infarction is the development of myocardial necrosis caused by a critical imbalance between the oxygen supply and demand of the myocardium. While a myocardial infarction may be caused by any process that causes this imbalance it most commonly results from plaque rupture with thrombus formation in a coronary vessel, resulting in an acute reduction of blood supply to a portion of the myocardium. Myocardial infarction is a usual accompaniment of anomalous left coronary artery from the pulmonary artery (ALCAPA).

1350 Cardiac tumor

An abnormal growth of tissue in or on the heart, demonstrating partial or complete lack of structural organization, and no functional coordination with normal cardiac tissue. Commonly, a mass is recognized which is distinct from the normal structural components of the heart. A primary cardiac tumor is one that arises directly from tissues of the heart, (e.g., myxoma, fibroelastoma, rhabdomyoma, fibroma, lipoma, pheochromocytoma, teratoma, hemangioma, mesothelioma, 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) collagen 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

MISCELLANEOUS, OTHER (CONTINUED)

- 1410 Persistent fetal circulation**
Persistence of the blood flow pattern seen in fetal life, in which high pulmonary vascular resistance in the lungs results in decreased blood flow to the lungs. Normally, after birth pulmonary pressure falls with a fall in pulmonary vascular resistance and there is increased perfusion of the lungs. Persistent fetal circulation, also known as persistent pulmonary hypertension of the newborn, can be related to lung or diaphragm malformations or lung immaturity.
- 1420 Meconium aspiration**
Aspiration of amniotic fluid stained with meconium before, during, or after birth can lead to pulmonary sequelae including (1) pneumothorax, (2) pneumomediastinum, (3) pneumopericardium, (4) lung infection, and (5) meconium aspiration syndrome (MAS) with persistent pulmonary hypertension.
- 1560 Cardiac, Other**
Any cardiac diagnosis not specifically delineated in other diagnostic codes.
- 1570 Thoracic and/or mediastinal, Other**
Any thoracic and/or mediastinal disease not specifically delineated in other diagnostic codes.
- 1580 Peripheral vascular, Other**
Any peripheral vascular disease (congenital or acquired) or injury (from trauma or iatrogenic); vessels involved may include, but are not limited to femoral artery, femoral vein, iliac artery, brachial artery, etc.
- 7000 Normal heart**
Normal heart.
- 7777 Miscellaneous, Other**
Any disease (congenital or acquired) not specifically delineated in other diagnostic codes.

STATUS POST SEPTAL DEFECTS

ASD

- 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**

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST SEPTAL DEFECTS (CONTINUED)

ASD (continued)

- 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).
- 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.

VSD

- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST SEPTAL DEFECTS (CONTINUED)

AV Canal

- 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**
- 6250 Status post - Valvuloplasty converted to valve replacement in the same operation, Common atrioventricular valve**
- 6230 Status post - Valve replacement, Common atrioventricular valve**

AP Window

- 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.

Truncus Arteriosus

- 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**

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

PULMONARY VENOUS ANOMALIES

Partial Anomalous Pulmonary Venous Connection

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)

Total Anomalous Pulmonary Venous Connection

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

STATUS POST

COR TRIARIATUM

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.

STATUS POST

PULMONARY VENOUS STENOSIS

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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST SYSTEMIC VENOUS ANOMALIES

Anomalous Systemic Venous Connection

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).

Systemic Venous Obstruction

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.

STATUS POST RIGHT HEART LESIONS

Tetralogy of Fallot

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, Nontransannular 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 transpulmonary 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.

4370 Status post - TOF repair, Ventriculotomy, Transannular 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 transpulmonary 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

RIGHT HEART LESIONS (CONTINUED)

Tetralogy of Fallot (continued)

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.

Pulmonary Atresia

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.

Tricuspid Valve Disease and Ebstein's Anomaly

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 translocation with or without detachment. Annuloplasty techniques that may be done solely or in combination with leaflet, chordae or muscle repair to achieve a competent valve include: eccentric annuloplasty, Kay annular plication, 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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

RIGHT HEART LESIONS (CONTINUED)

Tricuspid Valve Disease and Ebstein's Anomaly (continued)

- 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.

RVOT Obstruction, IVS Pulmonary Stenosis

- 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.
- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

RIGHT HEART LESIONS (CONTINUED)

RVOT Obstruction, IVS Pulmonary Stenosis (continued)

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.

Pulmonary Valve Disease

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.

STATUS POST

CONDUIT OPERATIONS

Conduit Operations

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.

5774 Status post - Conduit placement, Ventricle to aorta

Status post - Placement of a conduit from the right or left ventricle to the aorta.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

CONDUIT OPERATIONS (CONTINUED)

Conduit Operations (continued)

- 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.

Conduit Stenosis/ Insufficiency

- 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).

STATUS POST

LEFT HEART LESIONS

Aortic Valve Disease

- 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 Codes¹

STATUS POST

LEFT HEART LESIONS (CONTINUED)

Aortic Valve Disease (continued)

- 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 mechanical 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 Manouagian 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 myectomy, myotomy, myectomy, 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 Codes¹

STATUS POST

LEFT HEART LESIONS (CONTINUED)

Aortic Valve Disease (continued)

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

Sinus of Valsalva Aneurysm

- 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.

LV to Aorta Tunnel

- 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.

Mitral Valve Disease

- 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, commissurotomy, 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.

Hypoplastic Left Heart

- 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 neo-aortic 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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

LEFT HEART LESIONS (CONTINUED)

Hypoplastic Left Heart (continued)

4870 Status post - Norwood procedure (continued)

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

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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

LEFT HEART LESIONS (CONTINUED)

Hypoplastic Left Heart (continued)

- 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) (continued)**
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.
- 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.

STATUS POST

CARDIOMYOPATHY

- 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.

STATUS POST

PERICARDIAL DISEASE

- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

PERICARDIAL DISEASE (CONTINUED)

- 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.

STATUS POST

SINGLE VENTRICLE

- 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.
- 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.

STATUS POST

TRANSPOSITION OF THE GREAT ARTERIES

Congenitally Corrected TGA

- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

TRANSPOSITION OF THE GREAT ARTERIES (CONTINUED)

Congenitally Corrected TGA (continued)

- 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 otherwise specified in other listed procedure codes.

Transposition of the Great Arteries

- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

TRANSPOSITION OF THE GREAT ARTERIES (CONTINUED)

Transposition of the Great Arteries (continued)

- 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.
- 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)**

STATUS POST

DORV

- 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

STATUS POST

DOLV

- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST THORACIC ARTERIES AND VEINS

Coarctation of Aorta and Aortic Arch Hypoplasia

- 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.
- 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.

Coronary Artery Anomalies

- 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).

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

THORACIC ARTERIES AND VEINS (CONTINUED)

Coronary Artery Anomalies (continued)

- 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.

Interrupted Arch

- 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.

Patent Ductus Arteriosus

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

Vascular Rings and Slings

- 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.

Aortic Aneurysm

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

Aortic Dissection

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

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Congenital Cardiac Diagnosis Codes¹

STATUS POST THORACIC AND MEDIASTINAL DISEASE

Lung Disease

- 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.

Pectus Excavatum, Carinatum

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

Tracheal Stenosis

- 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.

STATUS POST ELECTROPHYSIOLOGICAL

- 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**
- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

ELECTROPHYSIOLOGICAL (CONTINUED)

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

STATUS POST

INTERVENTIONAL CARDIOLOGY PROCEDURES

- 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
- 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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

INTERVENTIONAL CARDIOLOGY PROCEDURES (CONTINUED)

- 6680 Status post - Cardiovascular electrophysiological catheterization procedure**
- 6690 Status post - Cardiovascular electrophysiological catheterization procedure, Therapeutic ablation**

STATUS POST

PALLIATIVE PROCEDURES

- 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.
- 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).

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

PALLIATIVE PROCEDURES (CONTINUED)

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.

STATUS POST

MECHANICAL SUPPORT

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.

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

STATUS POST

ANESTHETIC PROCEDURES

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

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

ANESTHETIC PROCEDURES (CONTINUED)

- 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**

STATUS POST

MISCELLANEOUS PROCEDURES

- 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.
- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

MISCELLANEOUS PROCEDURES (CONTINUED)

- 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).
- 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.

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Attachment E

Congenital Cardiac Diagnosis Codes¹

STATUS POST

MISCELLANEOUS PROCEDURES (CONTINUED)

- 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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Attachment F – Stress Test Results Definition and Clarification

Use the codes and descriptions below to indicate the stress test results based on the type of performed.

Standard Exercise Stress Test

1. Negative: A stress test is negative when the electrocardiogram (ECG) is normal or not suggestive of ischemia. ECGs are not suggestive of ischemia when there is <1 mm of horizontal or downsloping ST-segment depression or elevation for $\geq 60 - 80$ milliseconds after the end of the QRS complex, either during or after exercise.

Positive: A stress test is positive when the electrocardiogram (ECG) suggests ischemia. ECGs suggestive of ischemia can be described as having ≥ 1 mm of horizontal or downsloping ST-segment depression or elevation for $\geq 60-80$ milliseconds after the end of the QRS complex, either during or after exercise. It is also suggestive of ischemia if the patient had symptoms of ischemia (i.e. chest pain), arrhythmias, and/or a fall in blood pressure during or immediately after the procedure. If more than one study was performed with conflicting results and one study suggested coronary artery disease, code positive.

2. Positive, Low Risk: Low-risk treadmill score (score ≥ 5)

3. Positive Intermediate Risk: Intermediate risk treadmill score ($-11 < \text{score} < 5$).

4. Positive, High Risk: High risk treadmill score (score ≤ -11).

5. Positive, Risk Unknown: Positive as above, but risk is unknown.

Stress Echo Imaging Results

1. Negative: The imaging study was normal. There was no change in wall motion during the procedure.

Positive: The imaging study was abnormal. There were changes that reflected wall motion abnormalities during the procedure.

2. Positive Low Risk: (any of the following)

a. Low-risk treadmill score (score ≥ 5).

b. Normal stress echocardiographic wall motion or no change of limiting resting wall motion abnormalities during stress.*

*Although the published data are limited, patients with these findings will probably not be at low risk in the presence of either a high-risk treadmill score or severe resting left ventricular dysfunction (LVEF $< 35\%$).

Stress Echo Imaging Results (continued)

- 3. Positive Intermediate Risk:** (any of the following)
 - a. Mild/moderate resting left ventricular dysfunction (LVEF =35% to 49%)
 - b. Intermediate-risk treadmill score (-11 <score<5).
 - c. Limited stress echocardiographic ischemia with a wall motion abnormality only at higher doses of dobutamine involving less than or equal to two segments
- 4. Positive, High Risk:** (any of the following)
 - a. Severe resting left ventricular dysfunction (LVEF <35%).
 - b. High-risk treadmill score (score <= -11).
 - c. Severe exercise left ventricular dysfunction (exercise LVEF <35%)
 - d. Echocardiographic wall motion abnormality (involving greater than two segments) developing at low dose of dobutamine (<=10 mg/kg/min) or at a low heart rate (<120 beats/min).
 - e. Stress echocardiographic evidence of extensive ischemia.
- 5. Positive, Risk Unknown:** Positive as above, but risk is unknown.

SPECT MPI Imaging Results and Stress Test With CMR :

1. Negative: The results of the imaging study revealed no myocardial perfusion defects.

Positive: The result of the imaging study revealed one or more stress-induced myocardial perfusion defects.

- 2. Positive, Low Risk:** (any of the following)
 - a. Low-risk treadmill score (score >=5).
 - b. Normal or small myocardial perfusion defect at rest or with stress.*

*Although the published data are limited, patients with these findings will probably not be at low risk in the presence of either a high-risk treadmill score or severe resting left ventricular dysfunction (LVEF <35%).

- 3. Positive, Intermediate Risk:** (any of the following)
 - a. Mild/moderate resting left ventricular dysfunction (LVEF=35% to 49%).
 - b. Intermediate-risk treadmill score (-11 < score <5)
 - c. Stress-induced moderate perfusion defect without LV dilation or increased lung intake (thallium-201)
- 4. Positive, High Risk:** (any of the following)
 - a. Severe resting left ventricular dysfunction (LVEF <35%)
 - b. High-risk treadmill score (score <=-11)
 - c. Severe exercise left ventricular dysfunction (exercise LVEF <35%)
 - d. Stress-induced large perfusion defect (particularly if anterior)
 - e. Stress-induced multiple perfusion defects of moderate size
 - f. Large, fixed perfusion defect with LV dilation or increased lung uptake (thallium-201)
 - g. Stress-induced moderate perfusion defect with LV dilation or increased lung uptake (thallium-201)
- 5. Positive, Risk Unknown:** Positive as above, but risk is unknown.

For All Test Types:

6. Indeterminate: The results of the study were indeterminate or uninterpretable. They cannot be considered positive or negative.

7. Unavailable: The results of the study were not available.

9. Not Done / Unknown: No stress test/imaging study was performed within the past 6 months or it is not known if a stress test/imaging study was performed in the past 6 months.