World Database for Pediatric and Congenital Heart Surgery

Appendix A: Surgical Procedure Terms and Definitions

All surgeries are Tier 2 surgeries unless otherwise noted.

Anomalous Systemic Venous Connection

• Anomalous Systemic Venous Connection Repair

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).

Aortic Aneurysm

Aortic aneurysm repair

Aortic aneurysm repair by any technique.

Aortic Dissection

Aortic Dissection repair

Aortic dissection repair by any technique.

Aortic Root Replacement

Aortic Root Replacement, Bioprosthetic

Replacement of the aortic root (that portion of the aorta attached to the heart; it gives rise to the coronary arteries) with a bioprosthesis (e.g., porcine) in a conduit, often composite.

• Aortic Root Replacement, Mechanical

Replacement of the aortic root (that portion of the aorta attached to the heart; it gives rise to the coronary arteries) with a mechanical prosthesis in a composite conduit.

• Aortic Root Replacement, Homograft

Replacement of the aortic root (that portion of the aorta attached to the heart; it gives rise to the coronary arteries) with a homograft

• Aortic Root Replacement, Valve sparing

Replacement of the aortic root (that portion of the aorta attached to the heart; it gives rise to the coronary arteries) without replacing the aortic valve (using a tube graft).

Aortic Valve Disease

Ross Procedure

Replacement of the aortic valve with a pulmonary autograft and replacement of the pulmonary valve with a homograft conduit.

• Konno Procedure (with and without aortic valve replacement)

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.

Ross Konno Procedure

Relief of left ventricular outflow tract obstruction associated with aortic annular hypoplasia, aortic valvar stenosis and/or aortic valvar insufficiency via Konno aortoventriculoplasty using a pulmonary autograft root for the aortic root replacement.

• Repair of Supraaortic Stenosis

• Other aortic annular enlargement procedure

Techniques included under this procedure code include those designed to effect aortic annular enlargement that are not included in other procedure codes. These include the Manouguian and Nicks aortic annular enlargement procedures.

Aortic Valve Repair

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).

Aortic Valve Replacement

• Aortic Valve Replacement, Mechanical

Replacement of the aortic valve with a mechanical prosthetic valve.

• Aortic Valve Replacement, Bioprosthetic

Replacement of the aortic valve with a bioprosthetic prosthetic valve.

• Aortic Valve Replacement, Homograft

Replacement of the aortic valve with a homograft prosthetic valve.

AP Window

Aorto-pulmonary window Repair

Repair of AP window using one- or two-patch technique with cardiopulmonary bypass; or, without cardiopulmonary bypass, using trans catheter device or surgical closure.

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

ASD

• Patent Foramen Ovale, Primary Closure

Suture closure of patent foramen ovale (PFO).

• Atrial Septal Defect Repair, Primary Closure

Suture closure of secundum (most frequently), coronary sinus, sinus venosus or common atrium ASD.

• Atrial Septal Defect Repair, Patch

Patch closure (using any type of patch material) of secundum, coronary sinus, or sinus venosus ASD.

• Atrial Septal Defect Repair, Device

Closure of any type ASD (including PFO) using a device.

• Atrial Septal Defect (ASD) Repair, Partial closure

• Atrial Septal Defect Repair, Patch + Partial anomalous pulmonary venous connection Repair

Patch closure (using any type of patch material) of secundum, coronary sinus, or sinus venosus ASD plus PAPVC repair, any type

Atrial Septal Defect, Common atrium (single atrium), Septation
 Septation of common (single) atrium using any type patch material.

• Atrial Septal Defect creation/enlargement

Creation of an atrial septal defect or enlargement of an existing atrial septal defect using a variety of modalities including balloon septostomy, blade septostomy, or surgical septectomy. Creation may be accomplished with or without use of cardiopulmonary bypass.

Atrial Septal Fenestration

Creation of a fenestration (window) in the septum between the atrial chambers. Usually performed using a hole punch, creating a specifically sized communication in patch material placed on the atrial septum.

• Atrial fenestration closure

Closure of previously created atrial fenestration using any method including device, primary suture, or patch.

AV Canal

• Atrioventricular Septal Repair, Complete (Tier 1)

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

Atrioventricular Septal Repair, Intermediate (Transitional) (Tier 1)

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.

Atrioventricular Septal Repair, Partial (Incomplete) (Tier 1)

Repair of partial AV canal defect (primum ASD), any technique, with or without repair of cleft mitral valve.

• Common atrioventricular valve Repair

Common atrioventricular valve repair, any type

• Common atrioventricular valve Replacement

Replacement of the common atrioventricular valve with a prosthetic valve

Atrioventricular Septal Defect Re-repair (within 90 days)

Cardiomyopathy

- Transplant, Heart
- Transplant, Heart and lung

Coarctaction of Aorta and Aortic arch hypoplasia

Coarctation repair, End to end (Tier 1)

Repair of coarctation of aorta by excision of the coarctation segment and end-to-end circumferential anastomosis of the aorta.

Coarctation repair, End to end, Extended (Tier 1)

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.

Coarctation repair, Subclavian flap (Tier 1)

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.

- Coarctation repair, Patch aortoplasty (Tier 1)
 - Repair of coarctation of the aorta by incising the coarctation site with placement of a patch sutured in place longitudinally along the aortotomy edge.
- Coarctation repair, Interposition graft (Tier 1)
 - 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.
- Coarctation repair, Other (Tier 1)
 - 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.
- Coarctation repair + Ventricular Septal Defect repair
 - Coarctation of aorta repair, any technique, and simultaneous VSD repair, any type VSD, any type repair.
- Aortic arch repair
 - Aortic arch repair, any technique.
- Aortic arch repair + Ventricular Septal Defect repair
 - Aortic arch repair, any technique, and simultaneous VSD repair, any type VSD, any type repair. This includes repair of IAA with VSD.
- Coarctation repair, Extra-anatomic Bypass (Tier 1)
 - Repair of coarctation of the aorta by resection of the coarctation segment and placement of a prosthetic tubular outside the normal anatomic path.
- Coarctation Re-repair (within 90 days)

Conduit Operations

- Conduit Placement, Right Ventricle to Pulmonary Artery (primary or reoperation) Placement of a conduit, any type, from RV to PA.
- Conduit placement, Left Ventricle to Pulmonary Artery

 Placement of according to the Placement of the P
 - Placement of a conduit, any type, from LV to PA.
- Conduit placement, Ventricle to aorta
 - Placement of a conduit from the right or left ventricle to the aorta.

Congenitally Corrected TGA

- Congenitally corrected Transposition of the Great Arteries repair, Atrial switch and ASO (double switch)
 - Repair of congenitally corrected TGA by concomitant atrial switch (Mustard or Senning) and arterial switch operation. VSD closure is usually performed as well; this should be coded separately.
- Congenitally corrected Transposition of the Great Arteries repair, Atrial switch and Rastelli
 - Repair of congenitally corrected TGA by concomitant atrial switch (Mustard or Senning) and VSD closure to the aortic valve with placement of an RV-to-PA conduit.
- Congenitally corrected Transposition of the Great Arteries repair, VSD closure
 Repair of congenitally corrected TGA by VSD closure only

• Congenitally corrected Transposition of the Great Arteries repair, VSD closure and Left ventricular to Pulmonary Artery conduit

Repair of congenitally corrected TGA by VSD closure and placement of an LV-to-PA conduit.

Congenitally corrected Transposition of the Great Arteries repair, Other
Any procedures for correction of CCTGA not otherwise specified in other listed
procedure codes.

Cor Triatriatum

• Cor triatriatum repair

Repair revolves around resecting the anomalous membrane and closing the atrial septal defect

Coronary Artery Anomalies

Coronary artery fistula ligation

Coronary artery fistula repair using any technique. If additional technique information may be supplied by another procedure code, please list separately (e.g., bypass graft).

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

Coronary artery bypass

Coronary artery bypass graft procedure, any technique (with or without CPB, venous or arterial graft, one or more grafts, etc.), for any coronary artery pathology (coronary arterial fistula, aneurysm, coronary bridging, atresia of left main, acquired coronary artery disease, etc.).

• Anomalous aortic origin of coronary artery (AAOCA) repair

Repair of Anomalous coronary either the right coronary artery from the left coronary artery sinus or the left Main coronary artery from the right coronary artery sinus by any technique. These include but not limited to unroofing procedure, translocation of the coronary artery, bypass, ect.

• Coronary artery procedure, Other

Any coronary artery procedure not specifically listed.

DOLV

Double Outlet Left Ventricle repair

Because of the morphologic variability of DOLV, there are many approaches to repair, including: intraventricular tunnel repair directing the VSD to the pulmonary valve, the REV procedure, or the Rastelli procedure. In the case of DOLV use this code for tunnel closure to the pulmonary valve. If the REV or Rastelli procedures are performed, then use those respective codes.

DORV

• Double Outlet Right Ventricle, Intraventricular tunnel repair

Repair of DORV using a tunnel closure of the VSD to the aortic valve. This also includes the posterior straight tunnel repair of Kawashima

Electrophysiological

- Pacemaker implantation, Permanent
 - Implantation of a permanent pacemaker of any type (e.g., single-chamber, dual-chamber, atrial antitachycardia), with any lead configuration or type (atrial, ventricular, atrial and ventricular, transvenous, epicardial, transmural), by any technique (sternotomy, thoracotomy etc.).
- ICD (AICD) implantation
 Implantation of an (automatic) implantable cardioverter defibrillator system.
- Arrhythmia surgery atrial, Surgical Ablation Surgical ablation (any type) of any atrial arrhythmia.
- Arrhythmia surgery ventricular, Surgical Ablation Surgical ablation (any type) of any ventricular arrhythmia.

Hybrid

- Hybrid Approach "Stage 1", Application of RPA & LPA bands
- Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA)
- Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA) + application of RPA & LPA bands
- Hybrid approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Aortic arch repair (Norwood [Stage 1] + Superior Cavopulmonary anastomosis(es) + PA Debanding)
- Hybrid approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Without aortic arch repair Hybrid Approach, Transcardiac balloon dilatation
- Hybrid Approach, Transcardiac balloon dilatation
- Hybrid Approach, Transcardiac transcatheter device placement

Hypoplastic Left Heart and Related malformations

- Norwood procedure (w/mBT shunt) (Tier 1)
- Norwood procedure (Rv-PA Conduit) (Tier 1)
- Conduit insertion right ventricle to pulmonary artery + Intraventricular tunnel left ventricle to neoaorta + arch reconstruction (Rastelli and Norwood type arch reconstruction) (Yasui)
- Norwood procedure Re-repair (within 90 days)
- Hypoplastic Left Heart Syndrome (HLHS) Biventricular Repair (Tier 1)

Interrupted Arch

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

LV to Aorta Tunnel

LV to aorta tunnel repair

Mechanical Support

- Extracorporeal membrane oxygenation Cannulation
 Insertion of cannulas for extracorporeal membrane oxygenation
- Extracorporeal membrane oxygenation Decannulation

Removal of cannulas for extracorporeal membrane oxygenation

- Right Heart Temporary Ventricular Assist Device
- Right Heart Long-Term Ventricular Assist Device
- Left Heart Temporary Ventricular Assist Device
- Left Heart Long-Term Ventricular Assist Device
- Total Artificial Heart

Miscellaneous Procedures

- Aneurysm, Ventricular, Right, Repair
 Repair of right ventricular aneurysm, any technique.
- Aneurysm, Ventricular, Left, Repair Repair of left ventricular aneurysm, any technique.
- Aneurysm, Pulmonary artery, Repair
 Repair of pulmonary artery aneurysm, any technique.
- Cardiac tumor resection

Resection of cardiac tumor, any type.

- Pulmonary AV fistula repair/occlusion
 Repair or occlusion of a pulmonary arteriovenous fistula.
- Ligation, Pulmonary artery
 Ligation or division of the pulmonary artery. Most often performed as a secondary procedure.
- Pulmonary embolectomy, Acute pulmonary embolus
 Acute pulmonary embolism (clot) removal, through catheter or surgery.
- Pulmonary embolectomy, Chronic pulmonary embolus
 Chronic pulmonary embolism (clot) removal, through catheter or surgery.
- Procedures for chylothorax
 Surgical treatment of chylothorax. This may include, but is not limited to: thoracic duct ligation, pleurodesis, pleurectomy, pleuroperitoneal shunt, and external catheter/intermittent drainage.
- Other, specify: ______

Mitral Valve Disease

- Supravalvar mitral ring repair: resection
- Mitral Valve Repair (Left Atrioventricular Valve)

Mitral Valve Replacement (Left Atrioventricular Valve)

- Mitral Valve Replacement, Mechanical
- Mitral Valve Replacement, Bioprosthetic
- Mitral Valve Replacement, Homograft

Palliative Procedures

- Shunt, Ligation and Takedown
- Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS)

 Placement of a tube graft from a branch of the aortic arch to the pulmonary artery with or without bypass, from any approach (thoracotomy, sternotomy).
- Shunt, Systemic to pulmonary, Central (shunt from aorta)

A direct anastomosis or placement of a tube graft from the aorta to the pulmonary artery with or without bypass, from any approach (thoracotomy, sternotomy).

- Shunt, Systemic to pulmonary, Other
 - Placement of any other systemic-to-pulmonary artery shunt, with or without bypass, from any approach (thoracotomy, sternotomy) that is not otherwise coded. Includes classic Blalock-Taussig systemic-to-pulmonary artery shunt.
- Pulmonary Artery banding (PAB)

Placement of a pulmonary artery band, any type.

- Pulmonary Artery debanding
 - Debanding of pulmonary artery. Please list separately any pulmonary artery reconstruction required.
- Damus-Kaye-Stansel procedure (DKS) (creation of Aorto-pulmonary anastomosis without arch reconstruction)
 - In the Damus-Kaye-Stansel procedure the proximal transected main pulmonary artery is connected by varying techniques to the aorta.
- Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn) (Tier 1)
 Superior vena cava to pulmonary artery anastomosis allowing flow to both pulmonary arteries with an end-to- side superior vena-to-pulmonary artery anastomosis.
- Glenn (unidirectional cavopulmonary anastomosis) (unidirectional Glenn) (Tier 1)
 Superior vena cava to ipsilateral pulmonary artery anastomosis (i.e., LSVC to LPA, RSVC to RPA).
- Bilateral bidirectional cavopulmonary anastomosis (BBDCPA) (bilateral (Tier 1) bidirectional Glenn)
 - Bilateral superior vena cava-to-pulmonary artery anastomoses (requires bilateral SVCs).
- Hemi-Fontan (Tier 1)
 - A Hemi-Fontan 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.
- Hepatic vein to azygous vein connection, Direct or with Interposition Graft
- Kawashima operation (superior cavopulmonary connection in setting of interrupted IVC with azygous continuation)
- Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn) Rerepair (within 90 days)

Partial Anomalous Pulmonary Venous Connection (PAPVC)

Partial Anomalous Pulmonary Venous Connection (PAPVC) repair (Tier 1)
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.

- Partial Anomalous Pulmonary Venous Connection (PAPVC), Scimitar, Repair (Tier
 1)
 - 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.
- PAPVC repair, Baffle redirection to left atrium with systemic vein translocation (Warden) (SVC sewn to right atrial appendage) (Tier 1)
 An intracardiac baffle is created to redirect pulmonary venous return to the left atrium and SVC sewn to right atrial appendage)
- Partial Anomalous Pulmonary Venous Connection (PAPVC) Re-repair (within 90 days)

Patent Ductus Arteriosus

- Patent Ductus Arteriosus closure, Surgical
 Closure of a PDA by any surgical technique (ligation, division, clip) using any approach (i.e., thoracotomy, thoracoscopic, etc.).
- Patient Ductus Arteriosus (PDA) closure, device (Option added July 9th, 2019) PDA closure in the catheterization laboratory with an occlusion device.

Pericardial Disease

• Pericardial drainage procedure

Pericardial drainage can include a range of therapies including, but not limited to: pericardiocentesis, pericardiostomy tube placement, pericardial window creation, and open pericardial drainage (pericardiostomy).

- Pericardiectomy
 - Surgical removal of the pericardium.
- Pericardial procedure, Other

Other pericardial procedures that include, but are not limited to pericardial reconstruction for congenital absence of the pericardium, pericardial biopsy, pericardial mass or cyst excision.

Pulmonary Atresia/VSD

• Pulmonary Atresia - VSD (including TOF, PA) repair

For patients with pulmonary atresia with ventricular septal defect without MAPCAs, including those with tetralogy of Fallot with pulmonary atresia, repair may entail either a tetralogy-like repair with transannular patch placement, a VSD closure with placement of an RV-PA conduit, or an intraventricular tunnel VSD closure with transannular patch or RV-PA conduit placement.

 Pulmonary atresia - VSD - MAPCA repair, Complete single stage repair (1 stage that includes pulmonary unifocalization + VSD closure + RV to PA connection [with or without conduit])

One stage repair that includes bilateral pulmonary unifocalizatin + VSD closure + RV to PA connection (with or without conduit).

 Pulmonary atresia - VSD – MAPCA Repair, Status post prior complete unifocalization (includes VSD closure + RV to PA connection [with or without conduit])

VSD closure + RV to PA connection (with or without conduit).

 Pulmonary atresia - VSD – MAPCA repair, Status post prior incomplete unifocalization (includes completion of pulmonary unifocalization + VSD closure + RV to PA connection [with or without conduit])

Completion of pulmonary unifocalization + VSD closure + RV to PA connection (with or without conduit).

- Unifocalization MAPCA(s), Bilateral pulmonary unifocalization Complete unifocalization, all usable MAPCA[s] are incorporated.
- Unifocalization MAPCA(s), Unilateral pulmonary unifocalization
 Unilateral pulmonary unifocalization (one side) usable MAPCA(s) are incorporated.

Pulmonary Valve Disease

- Pulmonary Valve Replacement, Mechanical Replacement of the pulmonic valve with a mechanical valve
- Pulmonary Valve Replacement, Bioprosthetic
 Replacement of the pulmonic valve with a bioprosthetic valve
- Pulmonary Valve Replacement, Homograft
 Replacement of the pulmonic valve with a homograft (allograft)
- Pulmonary Valve Replacement, Other
 Replacement of the pulmonic valve that is not specifically listed. This may include, but is
 not limited to replacement using PTFE (Gore-Tex).
- Pulmonary Valve Repair
 May include a range of techniques including but not limited to: valvotomy with or without bypass, commissurotomy, and valvuloplasty.

Pulmonary Venous Stenosis

Pulmonary venous stenosis repair

Repair involves opening the obstructed vein with a variety of approaches: sutureless, patchvenoplasty, stent placement, etc.

Repair of Subaortic Stenosis

- Membrane Resection
- Myomectomy
- Extended Myomectomy

RVOT Obstruction, IVS Pulmonary Stenosis

- Right ventricular Outflow Tract procedure and/or Transannular patch
 Included in this procedural would be all RVOT procedures not elsewhere specified in the
 nomenclature system. These might be, among others: resection of sub valvar pulmonary
 stenosis (not DCRV type; may be localized fibrous diaphragm or high infundibular
 stenosis), right ventricular patch augmentation, or reduction pulmonary artery
 arterioplasty.
- 1 1/2 ventricular repair

Partial biventricular repair; includes intracardiac repair with bidirectional cavopulmonary anastomosis to volume unload a small ventricle or poorly functioning ventricle.

- Pulmonary Artery, reconstruction, Main
 Reconstruction of the main pulmonary artery trunk commonly using patch material.
- Pulmonary Artery, reconstruction, Central
 Reconstruction of the right or left branch (or both right and left) pulmonary arteries
 (within the hilar bifurcation) commonly using patch material.
- Pulmonary Artery, reconstruction, Peripheral
 Reconstruction of the peripheral right or left branch (of both right and left) pulmonary
 arteries (at or beyond the hilar bifurcation) commonly using patch material.
- Double Chamber Right Ventricle
 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.

Single Ventricle

 Fontan Operation (Complete Cavo-pulmonary anastomosis), Extracardiac Type: Fenestrated (Tier 1)

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

• Fontan Operation (Complete Cavo-pulmonary anastomosis), Extracardiac Type: Non-fenestrated (Tier 1)

The lateral tunnel Fontan is a TCPC type of Fontan Procedure created with anastomosis of SVC and right atrium to the branch pulmonary artery and an intra-atrial baffle to direct IVC flow to pulmonary artery. "The Fontan" is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart. A "TCPC" is a Fontan where both the superior caval vein and the inferior caval vein are connected to the pulmonary circulation through separate connections that are either direct connections or tubular pathways. A fenestration of a Fontan is defined as a communication that is created to allow flow of blood between the systemic and pulmonary venous chambers.

• Fontan Operation (Complete Cavo-pulmonary anastomosis), Lateral Tunnel Type (Tier 1)

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

- Fontan Operation (Complete Cavo-pulmonary anastomosis), Extra/Intra Cardiac Type (Tier 1)
- Fontan Operation (Complete Cavo-pulmonary anastomosis), Internal Conduit Type (Tier 1)
- Fontan Operation (Complete Cavo-pulmonary anastomosis), Other (Tier 1)
- Fontan revision or conversion (Re-do Fontan)

"Fontan revision or conversion (Re-do Fontan)" is defined as an operation where a previously created Fontan circuit is either modified or taken down and changed into a different type of Fontan. "The Fontan" is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart. A "TCPC" is a Fontan where both the superior caval vein and the inferior caval vein are connected to the pulmonary circulation through separate connections that are either direct connections or tubular pathways.

• Fontan, Other (Tier 1)

Fontan procedure not specified in procedure codes. May include takedown of a Fontan procedure. "The Fontan" is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart.

Ventricular septation

Creation of a prosthetic ventricular septum. Surgical procedure used to septate univentricular hearts with two atrioventricular valves. Additional procedures, such as resection of sub pulmonic stenosis, should be listed separately.

• Fontan Re-repair (within 90 days)

Sinus of Valsalva Aneurysm

• Sinus of Valsalva, Aneurysm repair

Systemic Venous Obstruction

Systemic venous stenosis repair

Repair is accomplished (most commonly SVC or IVC) with patch or conduit placement, excision of the stenotic area with primary reanastomosis or direct reimplantation.

Tetralogy of Fallot repair

• Tetralogy of Fallot repair (Tier 1)

This procedure assumes VSD closure and relief of pulmonary stenosis at one or more levels. The repair occurs 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.

- Tetralogy of Fallot repair, Ventriculotomy (Tier 1)
 This procedure assumes VSD closure and relief of pulmonary stenosis at one or more levels. The repair utilizes a ventriculotomy incision, but without placement of a transpulmonary annulus patch.
- Tetralogy of Fallot repair, Transannular patch (Tier 1)

 This procedure assumes VSD closure and relief of pulmonary stenosis at one or more levels, with use of a ventriculotomy incision and placement of a trans-pulmonary annulus patch. The valvar tissue is often removed. 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.

- Tetralogy of Fallot repair, RV-PA Conduit (Tier 1)
 - This procedure assumes VSD closure and relief of Right ventricular outflow tract and pulmonary stenosis at one or more levels, with use of a ventriculotomy incision and placement of a trans-pulmonary annulus patch. Right ventricular to pulmonary artery continuity is created with a homograft, heterograft, or synthetic conduit.
- Tetralogy of Fallot repair / Atrioventricular septal defect repair
 This procedure assumes VSD closure and relief of Right ventricular outflow tract and pulmonary stenosis at one or more levels with repair of associated atrioventricular septal defect.
- Tetralogy of Fallot Absent pulmonary valve repair

 This procedure assumes VSD closure and relief of Right ventricular outflow tract and pulmonary stenosis and with most cases, pulmonary valve replacement (pulmonary or aortic homograft, porcine, other) and a reduction pulmonary artery arterioplasty.
- Tetralogy of Fallot repair, Pulmonary Artery Reconstruction (Tier 1)

 This procedure assumes VSD closure and relief of Right ventricular outflow tract and pulmonary stenosis at one or more levels, with use of a ventriculotomy incision and placement of a trans-pulmonary annulus patch. The pulmonary valve is reconstructed utilizing native valvar tissue or creation of a moncusp synthetic substitute.
- Tetralogy of Fallot repair, Valvotomy (Tier 1)
- Tetralogy of Fallot Re-repair (within 90 days)

Total Anomalous Pulmonary Venous Connection (TAPVC)

- Total Anomalous Pulmonary Venous Connection repair (Tier 1)
 Repair revolves around creating a neo-connection between the pulmonary veins or pulmonary venous confluence to the left atrium
- Total Anomalous Pulmonary Venous Connection Re-repair (within 90 days)

Transposition of the Great Arteries

- Arterial switch operation (ASO) (Tier 1)
 - 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.
- Arterial switch operation (ASO) and VSD repair
 Arterial switch operation is used for repair of transposition of the great arteries (TGA).
 The pulmonary artery and aorta are transected and translocated so that the pulmonary

artery arises from the right ventricle and the aorta from the left ventricle. Coronary artery transfer is also accomplished. The VSD is closed, usually with a patch.

- Arterial switch procedure + Aortic arch repair
 - Concomitant arterial switch operation and repair of the aortic arch in patients with transposition of the great arteries with intact ventricular septum and associated coarctation of the aorta or interrupted aortic arch.
- Arterial switch procedure and VSD repair + Aortic arch repair
 Concomitant arterial switch operation with VSD closure and repair of aortic arch in patients with transposition of the great arteries with VSD and associated coarctation the aorta or interrupted aortic arch.
- Senning

Atrial baffle procedure for rerouting of venous flow in TGA resulting in a "physiological repair". The caval flow is directed behind the baffle to the mitral valve, left ventricle and pulmonary artery while the pulmonary venous flow is directed in front of the baffle to the tricuspid valve, right ventricle, and aorta. The Senning procedure uses atrial wall to construct the baffle.

Mustard

Atrial baffle procedure for rerouting of venous flow in TGA resulting in a "physiological repair". The caval flow is directed behind the baffle to the mitral valve, left ventricle and pulmonary artery while pulmonary venous flow is directed in front of the baffle to the tricuspid valve, right ventricle, and aorta. The Mustard procedure uses patch material to construct the baffle.

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

Rastelli

Most often used for patients with TGA-VSD and significant LVOTO, the Rastelli operation consists of an LV-to-aorta intraventricular baffle closure of the VSD and placement of an RV-to-PA conduit.

Reparation A L Etage Ventriculaire (REV)

The Lecompte (REV) intraventricular repair is designed for patients with abnormalities of ventriculoarterial connection in whom a standard intraventricular tunnel connection 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

- Aortic root translocation over left ventricle (Including Nikaidoh procedure)
- Transposition of the Great Arteries, Other procedures (Kawashima, Left Ventricular to Pulmonary Artery conduit, other)
- Arterial switch Operation (ASO) Re-repair (within 90 days)

Tricuspid Valve Disease and Ebstein's Anomaly

• Ebstein's Repair (Tier 1)

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.

- Tricuspid Valve Replacement (Right Atrioventricular Valve)
 Replacement of the tricuspid valve with a prosthetic valve
- Tricuspid Valve Repair (Right Atrioventricular Valve)

Reconstruction of the tricuspid valve may include but not be limited to a wide range of techniques including: leaflet patch extension, artificial chordae placement, and papillary muscle translocation with or without detachment. Annuloplasty techniques that may be done solely or in combination with leaflet, chordae or muscle repair to achieve a competent valve include eccentric annuloplasty, Kay annular plication, purse-string annuloplasty (including semicircular annuloplasty), sliding annuloplasty, and annuloplasty with ring placement.

Ebstein's Re-repair (within 90 days)

Truncus Arteriosus

Truncus arteriosus repair (Tier 1)

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
 - Truncal valve repair, any type.
- Truncal Valve Replacement

Replacement of the truncal valve with a prosthetic valve.

• Truncus + Interrupted aortic arch (IAA) repair

Truncus arteriosus repair usually includes patch VSD closure and placement of a conduit from RV to PA. In some cases, a conduit is not placed but an RV to PA connection is made by direct association. Also repair of interrupted aortic arch

• Truncus arteriosus Re-repair (within 90 days)

Vascular Rings and Slings

Vascular ring repair

Repair of vascular ring (any type, except pulmonary artery sling) by any technique.

Aortopexy

Surgical fixation of the aorta to another structure (usually the posterior aspect of the sternum) to relieve compression on another vessel or structure (e.g., trachea).

Pulmonary artery sling repair

Pulmonary artery sling repair by any technique.

VSD

• Ventricular Septal Defect repair, Primary closure (Tier 1) Suture closure of any type VSD.

• Ventricular Septal Defect repair, Patch (Tier 1)

Patch closure (using any type of patch material) of any type VSD.

- Ventricular Septal Defect repair, Device (Tier 1)
 - Closure of any type VSD using a device.
- Ventricular Septal Defect, Multiple, Repair (Tier 1)

Closure of more than one VSD using any method or combination of methods.

- Ventricular Septal Defect creation/enlargement (Tier 1)
 - Creation of a ventricular septal defect or enlargement of an existing ventricular septal defect.
- Ventricular Septal patch fenestration

Us pa	reation of a fenestration (window) in the septum between the ventricular chambers. sually performed using a hole punch, creating a specifically sized communication in atch material placed on the ventricular septum. Sentricular Septal Defect Re-repair (within 90 days)
	Page 16 of 16 Updated 07/09/2019