

# 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**

## Coarctation 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 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 neo-aorta + 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** (*Option added March 22, 2017*)

### Mitral Valve Disease

- **Supralvalvar 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 bidirectional Glenn) (Tier 1)**  
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) Re-repair (within 90 days)**

### Partial Anomalous Pulmonary Venous Connection

- **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.).

### 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 unifocalization + 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 trans-pulmonary 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 moncuspid synthetic substitute.
- **Tetralogy of Fallot repair, Valvotomy (Tier 1)**
- **Tetralogy of Fallot Re-repair (within 90 days)**

### Total Anomalous Pulmonary Venous Connection

- **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**  
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.
- **Ventricular Septal Defect Re-repair (within 90 days)**

# World Database for Pediatric and Congenital Heart Surgery

## Appendix A.1: Tier 1 Surgical Procedure Terms and Definitions

### Tier 1 surgeries

#### AV Canal

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

#### Coarctation of Aorta and Aortic arch hypoplasia

- **Coarctation repair, End to end**  
Repair of coarctation of aorta by excision of the coarctation segment and end-to-end circumferential anastomosis of the aorta.
- **Coarctation repair, End to end, Extended**  
Repair of coarctation of the aorta by excision of the coarctation segment and end-to-end anastomosis of the oblique ends of the aorta, creating an extended anastomosis.
- **Coarctation repair, Subclavian flap**  
Repair of coarctation of the aorta by ligating, dividing, and opening the subclavian artery, incising the coarctation site, and folding down the subclavian artery onto the incision in the aorta, suturing the subclavian "flap" in place, creating a roof over the area of the previous coarctation.
- **Coarctation repair, Patch aortoplasty**  
Repair of coarctation of the aorta by incising the coarctation site with placement of a patch sutured in place longitudinally along the aortotomy edge.
- **Coarctation repair, Interposition graft**  
Repair of coarctation of the aorta by resection of the coarctation segment and placement of a prosthetic tubular interposition graft anastomosed circumferentially to the cut ends of the aorta.
- **Coarctation repair, Other**  
Any repair of coarctation not specified in procedure codes. This may include, for example, a combination of two approaches for coarctation repair or extra-anatomic bypass graft, etc.
- **Coarctation repair, Extra-anatomic Bypass**  
Repair of coarctation of the aorta by resection of the coarctation segment and placement of a prosthetic tubular outside the normal anatomic path.

#### Hypoplastic Left Heart and Related malformations

- **Norwood procedure (w/mBT shunt)**
- **Norwood procedure (RV-PA Conduit)**
- **Hypoplastic Left Heart Syndrome (HLHS) Biventricular Repair**



## Palliative Procedures

- **Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn)**  
Superior vena cava to pulmonary artery anastomosis allowing flow to both pulmonary arteries with an end-to-side superior vena-to-pulmonary artery anastomosis.
- **Glenn (unidirectional cavopulmonary anastomosis) (unidirectional Glenn)**  
Superior vena cava to ipsilateral pulmonary artery anastomosis (i.e., LSVC to LPA, RSVC to RPA).
- **Bilateral bidirectional cavopulmonary anastomosis (BBDCPA) (bilateral bidirectional Glenn)**  
Bilateral superior vena cava-to-pulmonary artery anastomoses (requires bilateral SVCs).
- **Hemi-Fontan**  
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.

## Partial Anomalous Pulmonary Venous Connection

- **Partial Anomalous Pulmonary Venous Connection (PAPVC) repair**  
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**  
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)**  
An intracardiac baffle is created to redirect pulmonary venous return to the left atrium and SVC sewn to right atrial appendage)

## Single Ventricle

- **Fontan Operation (Complete Cavo-pulmonary anastomosis), Extracardiac Type: Fenestrated**  
The external conduit Fontan is a TCPC type of Fontan operation created with anastomosis of SVC to the branch pulmonary artery a conduit outside of the heart to connect the infradiaphragmatic systemic venous return to the pulmonary artery. “The Fontan” is defined as an operation or intervention that results in caval flow from both the upper and lower body draining to the pulmonary circulation in a patient with a functionally univentricular heart. A “TCPC” is a Fontan where both the superior caval vein and the inferior caval vein are connected to the pulmonary circulation through

separate connections that are either direct connections or tubular pathways. A fenestration of a Fontan is defined as a communication that is created to allow flow of blood between the systemic and pulmonary venous chambers.

- **Fontan Operation (Complete Cavo-pulmonary anastomosis), Extracardiac Type: Non-fenestrated**

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

- **Fontan Operation (Complete Cavo-pulmonary anastomosis), Lateral Tunnel Type**

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**

- **Fontan Operation (Complete Cavo-pulmonary anastomosis), Internal Conduit Type**

- **Fontan Operation (Complete Cavo-pulmonary anastomosis), Other**

- **Fontan, Other**

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

## **Tetralogy of Fallot repair**

- **Tetralogy of Fallot repair**

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**

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 trans-pulmonary annulus patch.

- **Tetralogy of Fallot repair, Transannular patch**

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**  
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, Pulmonary Artery Reconstruction**  
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 moncuspid synthetic substitute.
- **Tetralogy of Fallot repair, Valvotomy**

### Total Anomalous Pulmonary Venous Connection

- **Total Anomalous Pulmonary Venous Connection repair**  
Repair revolves around creating a neo-connection between the pulmonary veins or pulmonary venous confluence to the left atrium

### Transposition of the Great Arteries

- **Arterial switch operation (ASO)**  
Arterial switch operation is used for repair of transposition of the great arteries (TGA). The pulmonary artery and aorta are transected and translocated so that the pulmonary artery arises from the right ventricle and the aorta from the left ventricle. Coronary artery transfer is also accomplished.

### Tricuspid Valve Disease and Ebstein's Anomaly

- **Ebstein's Repair**  
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.

### Truncus Arteriosus

- **Truncus arteriosus repair**  
Truncus arteriosus repair that most frequently includes patch VSD closure and placement of a conduit from RV to PA. In some cases, a conduit is not placed but an RV to PA connection is made by direct association. Very rarely, there is no VSD to be closed.

### VSD

- **Ventricular Septal Defect repair, Primary closure**  
Suture closure of any type VSD.
- **Ventricular Septal Defect repair, Patch**  
Patch closure (using any type of patch material) of any type VSD.
- **Ventricular Septal Defect repair, Device**  
Closure of any type VSD using a device.

- **Ventricular Septal Defect, Multiple, Repair**  
Closure of more than one VSD using any method or combination of methods.
- **Ventricular Septal Defect creation/enlargement**  
Creation of a ventricular septal defect or enlargement of an existing ventricular septal defect.

# World Database for Pediatric and Congenital Heart Surgery

## Appendix A.2: Tier 2 Surgical Procedure Terms and Definitions

### Tier 2 surgeries

#### 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

- **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**

### Coarctation of Aorta and Aortic arch hypoplasia

- **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 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 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

- **Conduit insertion right ventricle to pulmonary artery + Intraventricular tunnel left ventricle to neo-aorta + arch reconstruction (Rastelli and Norwood type arch reconstruction) (Yasui)**
- **Norwood procedure Re-repair (within 90 days)**

## 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** (*Option added March 22, 2017*)

### Mitral Valve Disease

- **Supralvalvar 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.
- **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) Re-repair (within 90 days)**

### **Partial Anomalous Pulmonary Venous Connection**

- **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.).

### **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 unifocalization + 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 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.
- **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 / 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 Re-repair (within 90 days)**

## Total Anomalous Pulmonary Venous Connection

- **Total Anomalous Pulmonary Venous Connection Re-repair (within 90 days)**

## Transposition of the Great Arteries

- **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

- **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

- **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 patch fenestration**  
Creation of a fenestration (window) in the septum between the ventricular chambers. Usually performed using a hole punch, creating a specifically sized communication in patch material placed on the ventricular septum.
- **Ventricular Septal Defect Re-repair (within 90 days)**



## World Database for Pediatric and Congenital Heart Surgery

### Appendix B: Diagnosis (International Paediatric and Congenital Cardiac Codes (IPCCC) and definitions)

#### Anomalous Systemic Venous Connection Systemic venous anomaly

- **Systemic venous anomaly**  
A congenital cardiovascular malformation in which there is an abnormality of the mediastinal systemic veins including but not limited to: caval veins, coronary sinus, hepatic veins connecting to the heart, brachiocephalic veins, and/or azygos veins.

#### Aortic Aneurysm

- **Aortic aneurysm (including pseudoaneurysm)**  
A congenital cardiovascular malformation in which the luminal diameter of the aorta between its sinotubular junction and the origin of its first branch is dilated (above the upper limit of normal adjusted for body size).

#### Aortic dissection

- **Aortic dissection**

#### Aortic Valve Disease

- **Aortic stenosis, Subvalvar**  
A congenital cardiovascular malformation associated with narrowing within the outflow tract supporting the aortic valve.
- **Aortic stenosis, Valvar**  
A congenital cardiovascular malformation of the aortic valve in which there is narrowing or stricture (obstruction to flow).
- **Aortic stenosis, Supravalvar**  
A congenital cardiovascular malformation with narrowing of the aorta at the level of the sinotubular junction which may extend into the ascending aorta.
- **Aortic valve atresia**  
A congenital cardiovascular malformation in which there is no orifice of the aortic valve.
- **Aortic insufficiency**  
Congenital cardiovascular malformation of the aortic valve allowing backward flow into the ventricle
- **Aortic insufficiency and aortic stenosis**
- **Aortic valve, Other**

#### AP Window

- **Aorto-pulmonary window (aortopulmonary window)**  
A congenital cardiovascular malformation in which there is side-to-side continuity of the lumens of the ascending aorta and pulmonary trunk in association with separate aortic and pulmonary valves or their atretic remnants.
- **Pulmonary artery origin from ascending aorta (hemitruncus)**  
A congenital cardiovascular malformation in which one branch pulmonary artery arises from the ascending aorta and the other branch pulmonary artery arises from the pulmonary trunk (main pulmonary artery).

## ASD

- **Patent oval foramen (patent foramen ovale)**  
A small interatrial communication (or potential communication) confined to the region of the oval fossa (fossa ovalis) characterized by no deficiency of the primary atrial septum (septum primum) and a normal limbus with no deficiency of the septum secundum (superior interatrial fold).
- **Atrial Septal Defect, Secundum**  
A congenital cardiac malformation in which there is an interatrial communication confined to the region of the oval fossa (fossa ovalis), most commonly due to a deficiency of the primary atrial septum (septum primum) but deficiency of the septum secundum (superior interatrial fold) may also contribute.
- **Atrial Septal Defect, Venosus**  
A congenital cardiovascular malformation in which there is a caval vein (vena cava) and/or pulmonary vein (or veins) that overrides the atrial septum or the septum secundum (superior interatrial fold) producing an interatrial or anomalous veno-atrial communication.
- **Atrial Septal Defect, Coronary Sinus**  
A congenital cardiovascular malformation in which there is a communication between the left atrium and the coronary sinus allowing interatrial communication through the coronary sinus ostium.
- **Atrial Septal Defect, Common Atrium (single Atrium)**  
A congenital cardiovascular malformation in which there is near-complete absence of the interatrial septum.

## AV Canal

- **Atrioventricular Canal Defect, Intermediate (transitional)**  
A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal defect) with a single atrioventricular valve annulus and distinct left and right atrioventricular valvar orifices, an interatrial communication immediately above the atrioventricular valve, and a restrictive interventricular communication (interventricular pressure gradient) immediately below the atrioventricular valve.
- **Atrioventricular Canal Defect, Partial (incomplete) (PAVSD) (ASD, primum)**  
A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal defect) with an interatrial communication just above the atrioventricular valve, no interventricular communication just below the atrioventricular valve, separate right and left atrioventricular valvar orifices, and varying degrees of malformation of the left-sided component of the common atrioventricular valve. The bridging leaflets of the common atrioventricular valve are bound down to the crest of the scooped-out ventricular septum so that the potential for shunting through the atrioventricular septal defect is possible only at the atrial level and not at the ventricular level.
- **Complete Atrioventricular Canal Defect**  
A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal defect) with an interatrial communication just above the atrioventricular valve, an interventricular communication just below the atrioventricular valve, and varying degrees of malformation of the left ventricular component of the common atrioventricular valve. There is unrestrictive interventricular communication (no interventricular pressure gradient) and the bridging leaflets usually float to varying extent within the atrioventricular septal defect.

## Cardiomyopathy

- **Cardiomyopathy (including dilated, restrictive, and hypertrophic)**
- **Cardiomyopathy, End-stage congenital heart disease**

## Coarctation of Aorta and Aortic arch hypoplasia

- **Coarctation of aorta**  
A congenital cardiovascular malformation in which there is a discrete luminal narrowing of the junction between the aortic arch and the descending aorta.
- **Aortic arch hypoplasia**  
A congenital cardiovascular malformation in which there is diffuse luminal narrowing of the aortic arch (below the lower limit of normal adjusted for body size).
- **Ventricular Septal Defect + Aortic arch hypoplasia**
- **Ventricular Septal Defect + Coarctation of aorta**

## Conduit Failure

- **Conduit Failure**

## Congenitally Corrected TGA

- **Congenitally corrected Transposition of the Great Arteries, Intact Ventricular Septum**
- **Congenitally corrected Transposition of the Great Arteries**  
A congenital cardiovascular malformation in which the morphologically right atrium connects to the morphologically left ventricle, the morphologically left atrium connects to the morphologically right ventricle, the morphologically right ventricle connects to the aorta, and the morphologically left ventricle connects to the pulmonary trunk.
- **Congenitally Corrected Transposition of the Great Arteries, Intact Ventricular Septum-Left Ventricular Outflow Tract Obstruction**
- **Congenitally Corrected Transposition of the Great Arteries, Ventricular Septal Defect**
- **Congenitally Corrected Transposition of the Great Arteries, Ventricular Septal Defect-Left Ventricular Outflow Tract Obstruction**

## Cor triatriatum

- **Cor triatriatum**  
A congenital cardiac malformation in which there is a partition that divides the left atrium into a posterior chamber that receives some or all of the pulmonary veins and an anterior chamber that communicates with the left atrial appendage and atrioventricular junction (usually the mitral valve).

## Coronary Artery Anomalies

- **Coronary Artery Anomaly, Aneurysm**  
A congenital cardiovascular malformation in which there is one or more localized dilation(s) of a coronary vessel. It is usually defined as an increase in luminal diameter that exceeds 1.5 times the luminal diameter of the adjacent normal coronary arteries.
- **Coronary Artery Anomaly, Anomalous aortic origin of coronary artery (AAOCA)**  
A congenital cardiovascular malformation in which the origin and/or course of a coronary artery is abnormal.
- **Coronary Artery Anomaly, Anomalous pulmonary origin (includes ALCAPA)**

A congenital cardiovascular malformation in which the left coronary artery originates from the pulmonary trunk or one of its branches.

- **Coronary Artery Anomaly, Fistula**

A congenital cardiovascular malformation in which a coronary artery communicates, through an anomalous channel, with a cardiac chamber or with any segment of the pulmonary circulation.

- **Coronary artery anomaly, Other**

## DOLV

- **Double Outlet Left Ventricle**

A congenital cardiovascular malformation in which both great arteries arise entirely or predominantly from the morphologically left ventricle.

## DORV

- **Double Outlet Right Ventricle**

A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections, a subaortic or doubly-committed (with absence or deficiency of the conal septum) ventricular septal defect, and unobstructed pulmonary outflow tract.

- **Double Outlet Right Ventricle, Atrioventricular Septal Defect**

- **Double Outlet Right Ventricle, Intact Ventricular Septum (IVS)**

A congenital cardiovascular malformation that is a variant of double outlet right ventricle that is associated with an intact ventricular septum.

- **Double Outlet Right Ventricle, Remote VSD (Uncommitted)**

A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections that is associated with ventricular septal defect that is remote from the ventricular outflow tracts and usually within the inlet or muscular septum.

- **Double Outlet Right Ventricle, Tetralogy of Fallot type**

A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections, a subaortic or doubly-committed (with absence or deficiency of the conal septum) ventricular septal defect, and pulmonary outflow tract obstruction.

- **Double Outlet Right Ventricle, Transposition of Great Arteries Type**

A congenital cardiovascular malformation that is a variant of double outlet right ventricle with concordant atrioventricular connections that is associated with a subpulmonary ventricular septal defect (includes Taussig-Bing heart).

## Electrophysiological

- **Arrhythmia**
- **Arrhythmia, atrial**
- **Arrhythmia, heart block**
- **Arrhythmia, ventricular**

## Hypoplastic left heart syndrome

- **Hypoplastic left heart syndrome (HLHS)**

A spectrum of congenital cardiovascular malformations with normally aligned great arteries without a common atrioventricular junction with significant hypoplasia of the left ventricle and including atresia, stenosis, or hypoplasia of the aortic or mitral valve, or both valves, and hypoplasia of the ascending aorta and aortic arch.

\*When Hypoplastic Left Heart Syndrome is selected, the following questions will be asked to further characterize:

Aortic Valve Atresia: Select yes, no, or unknown.

Aortic Valve Stenosis: Select yes, no, or unknown.

Aortic Valve Hypoplasia: Select yes, no, or unknown.

Mitral Valve Atresia: Select yes, no, or unknown.

Mitral Valve Stenosis: Select yes, no, or unknown.

Mitral Valve Hypoplasia: Select yes, no, or unknown.

Ventricular Septal Defect: Select yes, no, or unknown.

Left Ventricle size: Select normal, small, or unknown.

\*Hypoplastic Right Ventricle (HRV) is not a diagnosis option. We have elected to identify the anatomic details, which give rise to a small right ventricle rather than using the term HRV.

### Interrupted Arch

- **Interrupted aortic arch**  
A congenital cardiovascular malformation in which there is an absence of luminal continuity between the ascending and descending aorta.
- **Interrupted aortic arch + Aorto-Pulmonary window**
- **Interrupted aortic arch + Ventricular Septal Defect**

### LV to Aorta Tunnel

- **Left Ventricular to aorta tunnel**  
A congenital cardiovascular malformation in which there is a paravalvar communication between the aorta and a ventricle.

### Miscellaneous, Other

- **Atrial Isomerism, Left**  
A congenital cardiovascular malformation that is a variant of a heterotaxy syndrome in which some paired structures on opposite sides of the left-right axis of the body are symmetrical mirror images of each other, and have the morphology of the normal left-sided structures.
- **Atrial Isomerism, Right**  
A congenital cardiovascular malformation that is a variant of heterotaxy syndrome in which some paired structures on opposite sides of the left-right axis of the body are symmetrical mirror images of each other, and have the morphology of the normal right-sided structures.
- **Dextrocardia**  
A congenital cardiovascular malformation in which the heart is predominantly in right hemithorax.
- **Levocardia**  
A congenital cardiovascular finding in which the heart is predominantly in left hemithorax.
- **Mesocardia**  
A congenital cardiovascular malformation in which the heart is central or midline within the thorax.
- **Aneurysm, Pulmonary artery**  
A congenital cardiovascular malformation in which there is an enlargement of the luminal diameter of the pulmonary trunk (main pulmonary artery) and/or branch pulmonary arteries (above the upper limit of normal adjusted for body size).

- **Prosthetic valve failure**
- **Cardiac tumor**
- **Pulmonary vascular obstructive disease (Eisenmenger's)**
- **Prosthetic valve Endocarditis**
- **Active Endocarditis**
- **Rheumatic Heart Disease**
- **Situs inversus**  
A congenital cardiac malformation in which the atrial morphologies and positions are the mirror image of normal.
- **Aneurysm, Other**
- **Aneurysm, Ventricular, left (including pseudoaneurysm)**  
A congenital cardiac malformation in which there is an outpouching of the left ventricular wall.
- **Aneurysm, Ventricular, Right (including pseudoaneurysm)**
- **Other, specify** (*Option added March 22, 2017*)

### Mitral Valve Disease

- **Mitral stenosis (Annular Hypoplasia)**  
A congenital cardiac malformation of the mitral valve in which there is annular hypoplasia (incomplete development or underdevelopment so that it is abnormally small [below the lower limit of normal adjusted for body size]). Hypoplasia may or may not be associated with stenosis.
- **Mitral stenosis, Subvalvar**  
A congenital cardiac malformation in which the mitral chords, chordal attachments, or papillary muscles are abnormal.
- **Mitral stenosis, Subvalvar, Parachute**  
A congenital cardiac malformation in which the chords of the mitral valve attach to a single or to closely adjacent papillary muscles.
- **Mitral stenosis, Supravalvar mitral ring**  
A congenital cardiac malformation in which a ridge of tissue is attached or integral to the atrial side of the mitral valvar leaflet(s)
- **Mitral stenosis, Valvar**  
A congenital cardiac malformation of the mitral valve in which there is narrowing or stricture of the valvar orifice (obstruction to flow).
- **Mitral regurgitation**  
A congenital cardiac finding in which there is backward flow through the mitral valve.
- **Mitral regurgitation and mitral stenosis**
- **Mitral valve, Other**

### Partial anomalous pulmonary venous connection

- **Partial anomalous pulmonary venous connection (PAPVC)**  
A congenital cardiovascular malformation in which one or more (but not all) of the pulmonary veins connect anomalously to the right atrium or to one or more of its venous tributaries and the remaining pulmonary veins connect to the left atrium.
- **Partial anomalous pulmonary venous connection (PAPVC), scimitar**  
A congenital cardiovascular malformation with partial anomalous pulmonary venous connection in which some of the pulmonary veins (usually the right pulmonary veins) connect anomalously to the inferior caval vein (inferior vena cava) or to the right atrium at the insertion of the inferior vena cava.

## Patent ductus arteriosus

- **Patent ductus arteriosus**  
A congenital cardiovascular malformation of the arterial duct (ductus arteriosus) or its fibrous remnant (ligamentum arteriosum).

## Pulmonary atresia

- **Pulmonary atresia**  
A congenital cardiovascular malformation where the pulmonary trunk (main pulmonary artery) is not present or has luminal occlusion, excluding common arterial trunk.
- **Pulmonary atresia, Intact Ventricular Septum**  
A congenital cardiovascular malformation in which there are normally aligned great arteries, no opening between the morphologically right ventricle and the pulmonary trunk, and no ventricular level communication.
- **Pulmonary atresia, VSD (Including TOF, PA)**  
A congenital cardiovascular malformation that is a variant of tetralogy of Fallot in which there is no direct communication between the right ventricle and the pulmonary arterial tree.
- **Pulmonary atresia, Ventricular Septal Defect-Multiple aorto-pulmonary collateral artery**  
A congenital cardiovascular malformation that is a variant of tetralogy of Fallot in which there is no direct communication between the right ventricle and the pulmonary arterial tree and there are collateral blood vessels between the systemic and pulmonary arteries.
- **Pulmonary atresia, MAPCA(s) (major aortopulmonary collateral[s]) (without PA-VSD)**  
A congenital cardiovascular malformation in which the blood supply to the lungs is derived completely or in part from collateral vessels that arise from the aorta or its branches.

## Pulmonary Valve Disease

- **Pulmonary insufficiency**  
Congenital cardiovascular malformation of the pulmonary valve allowing backward flow into the ventricle
- **Pulmonary valve, Other**
- **Pulmonary insufficiency and pulmonary stenosis**

## Pulmonary venous stenosis

- **Pulmonary venous stenosis**  
A congenital cardiovascular malformation with a pathologic narrowing of one or more pulmonary veins including diffuse hypoplasia, long segment focal/tubular stenosis and/or discrete stenosis.

## RVOT Obstruction and/or Pulmonary Stenosis

- **Pulmonary stenosis, Valvar**  
A congenital cardiovascular malformation of the pulmonary valve in which there is narrowing or stricture (obstruction to flow)
- **Pulmonary stenosis, Subvalvar**  
A congenital cardiovascular malformation associated with narrowing within the outflow tract supporting the pulmonary valve.
- **Pulmonary artery stenosis (hypoplasia), Main (trunk) (Supravalvar Stenosis)**

A congenital cardiovascular malformation associated with narrowing at the level of the pulmonary sinotubular junction.

- **Pulmonary artery stenosis, Branch, Central (within the hilar bifurcation)**  
A congenital cardiovascular malformation of a pulmonary artery, proximal to its first branch, in which there is luminal narrowing (below the lower limit of normal adjusted for body size).
- **Pulmonary artery stenosis, Branch, Peripheral (at or beyond the hilar bifurcation)**  
A congenital cardiovascular malformation of a pulmonary artery, distal to its first branch, in which there is luminal narrowing (below the lower limit of normal adjusted for body size).
- **Pulmonary artery, Discontinuous**  
A congenital cardiovascular malformation in which there is absence of luminal continuity between the right and left branch pulmonary arteries.
- **Double Chamber Right Ventricle**  
A congenital cardiac malformation in which the right ventricle is divided into two chambers, one inferior including the inlet and trabecular portions of the right ventricle and one superior including the trabecular portion and infundibulum.

### Shone's syndrome

- **Shone's syndrome**  
A congenital cardiovascular malformation in which more than one of the following lesions are present: (1) supralvalvar or intravalvar mitral ring, (2) mitral subvalvar stenosis, (3) a parachute deformity of the mitral valve, (4) subaortic stenosis, (5) valvar aortic stenosis, and (6) aortic coarctation.

### Shunt Failure

- **Shunt Failure**

### Single Ventricle

- **Single ventricle, Double Inlet left ventricle**  
A congenital cardiovascular malformation with a univentricular atrioventricular connection wherein both atria connect to a morphologically left ventricle either via two separate atrioventricular valves or a common atrioventricular valve, such that all or nearly all of the total atrioventricular junctional (annular) area is committed to the left ventricular chamber.
- **Single ventricle, Double Inlet Right Ventricle**  
A congenital cardiovascular malformation with a univentricular atrioventricular connection wherein both atria connect to a morphologically right ventricle either via two separate atrioventricular valves or a common atrioventricular valve, such that all or nearly all of the total atrioventricular junctional (annular) area is committed to the right ventricular chamber.
- **Single ventricle, Mitral atresia**  
A congenital cardiovascular malformation with absence of the mitral valvar annulus (connection/junction) or an imperforate mitral valve.
- **Single ventricle, Unbalanced Atrio-ventricular canal Defect**  
A congenital cardiac malformation in which the common atrioventricular valve is primarily related to one ventricle, usually but not always associated with hypoplasia of the other ventricle.
- **Single ventricle, Heterotaxia syndrome**
- **Single ventricle, Other**



- **Single ventricle + Total anomalous pulmonary venous connection (TAPVC)**
- **Single ventricle, Tricuspid atresia**  
A congenital cardiovascular malformation with absence of the tricuspid valvar annulus (connection/junction) or an imperforate tricuspid valve.

### Sinus of Valsalva Fistula/Aneurysm

- **Sinus of Valsalva aneurysm**  
A congenital cardiovascular malformation in which there is dilation of a single aortic sinus of Valsalva.

### Systemic venous obstruction

- **Systemic venous obstruction**  
A congenital cardiovascular malformation in which there is an abnormality of the mediastinal systemic veins including but not limited to: caval veins, coronary sinus, hepatic veins connecting to the heart, brachiocephalic veins, and/or azygos veins.

### Tetralogy of Fallot

- **Tetralogy of Fallot**  
A group of congenital cardiac 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. Tetralogy of Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, aortic override, and most often right ventricular hypertrophy.
- **Tetralogy of Fallot, Pulmonary stenosis**
- **Tetralogy of Fallot, complete Atrio-ventricular septal Defect**  
A congenital cardiac malformation with both an atrioventricular septal defect (atrioventricular canal defect) and tetralogy of Fallot.
- **Tetralogy of Fallot, Absent pulmonary valve**  
A congenital cardiovascular malformation that is a variant of tetralogy of Fallot in which the ventriculo-arterial junction of the right ventricle with the pulmonary trunk features an atypical valve with rudimentary leaflets (cusps) which do not coapt.

### Total anomalous pulmonary venous connection

- **Total anomalous pulmonary venous connection (TAPVC), Type 1 (supracardiac)**  
A congenital cardiovascular malformation with total anomalous pulmonary venous connection to the superior caval vein (superior vena cava) or one of its venous tributaries.
- **Total anomalous pulmonary venous connection (TAPVC), Type 2 (cardiac)**  
A congenital cardiovascular malformation with total anomalous pulmonary venous connection to the right atrium directly or to the coronary sinus or to both.
- **Total anomalous pulmonary venous connection (TAPVC), Type 3 (infracardiac)**  
A congenital cardiovascular malformation with infradiaphragmatic total anomalous pulmonary venous connection.
- **Total anomalous pulmonary venous connection (TAPVC), Type 4 (mixed)**  
A congenital cardiovascular malformation with total anomalous pulmonary venous connection at two or more levels (supracardiac, cardiac, or infracardiac).

### Transposition of the Great Arteries

- **Transposition of the Great Arteries, Intact Ventricular Septum-Left Ventricular Outflow Tract Obstruction**
- **Transposition of the Great Arteries, Ventricular Septal Defect**  
A congenital cardiovascular malformation in which the morphologically right atrium connects to the morphologically right ventricle, the morphologically left atrium connects to the morphologically left ventricle, the morphologically right ventricle connects to the aorta, the morphologically left ventricle connects to the pulmonary trunk, and one or more ventricular septal defects are present.
- **Transposition of the Great Arteries, Intact Ventricular Septum**  
A congenital cardiovascular malformation in which the morphologically right atrium connects to the morphologically right ventricle, the morphologically left atrium connects to the morphologically left ventricle, the morphologically right ventricle connects to the aorta, the morphologically left ventricle connects to the pulmonary trunk, and a ventricular septal defect is not present.
- **Transposition of the Great Arteries, Ventricular Septal Defect-Left Ventricular Outflow Tract Obstruction**  
A congenital cardiovascular malformation in which the morphologically right atrium connects to the morphologically right ventricle, the morphologically left atrium connects to the morphologically left ventricle, the morphologically right ventricle connects to the aorta, the morphologically left ventricle connects to the pulmonary trunk, one or more ventricular septal defects are present, and left ventricular outflow tract obstruction is present.

### Tricuspid Valve Disease and Ebstein's Anomaly

- **Ebstein's anomaly**  
A congenital cardiac malformation of the tricuspid valve and right ventricle that is characterized by downward (apical) displacement of the functional annulus, usually involving the septal and inferior (posterior) leaflets.
- **Tricuspid regurgitation, non-Ebstein's related**  
A congenital cardiac finding in which there is backward flow through the tricuspid valve.
- **Tricuspid regurgitation and tricuspid stenosis**
- **Tricuspid stenosis**  
A congenital cardiovascular malformation of the tricuspid valve in which there is narrowing or stricture (obstruction to flow)
- **Tricuspid valve, Other**

### Truncus arteriosus

- **Truncus arteriosus**  
A congenital cardiovascular malformation in which a single arterial trunk arises from the heart, giving origin sequentially to the coronary arteries, one or more pulmonary arteries, and the systemic arterial circulation.
- **Truncus arteriosus + Interrupted aortic arch**  
A congenital cardiovascular malformation in which a common arterial trunk is associated with an interrupted aortic arch.
- **Truncal valve insufficiency**  
A congenital cardiovascular malformation in which there is backward flow through the truncal valve.

### Vascular rings and Slings

- **Vascular Ring**

A congenital cardiovascular malformation in which one or more of the following encircle the trachea and esophagus: the aorta and its major branches, the pulmonary trunk and its major branches, and the arterial duct (ductus arteriosus) or their vascular remnant(s).

- **Pulmonary Artery Slings**

A congenital cardiovascular malformation in which the left pulmonary artery originates from the right pulmonary artery and passes between the trachea and esophagus.

## VSD

- **Ventricular Septal Defect, Type 1 (Subarterial) (Supracristal) (Conal septal defect) (Infundibular)**

A congenital cardiac malformation in which there is a ventricular septal defect that opens to the outlet of the right ventricle.

- **Ventricular Septal Defect, Type 2 (Perimembranous) (Paramembranous) (Conoventricular)**

A congenital cardiovascular malformation in which there is a ventricular septal defect that 1) occupies the space that is usually closed by the interventricular part of the membranous septum, 2) is adjacent to the area of fibrous continuity between the leaflets of an atrioventricular valve and an arterial valve, and 3) is located at the center of the base of the ventricular mass.

- **Ventricular Septal Defect, Type 3 (Inlet) (AV canal type)**

A congenital cardiac malformation in which there is a ventricular septal defect that opens into the inlet component of the right ventricle.

- **Ventricular Septal Defect, Type 4 (Muscular)**

A congenital cardiac malformation in which there is a ventricular septal defect with exclusively muscular borders that opens to the outlet of the right ventricle, and in which the muscular outlet septum is aligned with the apical part of the muscular septum.

- **Ventricular Septal Defect, Type: Gerbode type (LV-RA communication)**

- **Ventricular Septal Defect, Multiple**

A congenital cardiac malformation in which there are multiple ventricular septal defects.

# World Database for Pediatric and Congenital Heart Surgery

## Appendix C: Birth Country

AFG	Afghanistan
ALA	Åland Islands
ALB	Albania
DZA	Algeria
ASM	American Samoa
AND	Andorra
AGO	Angola
AIA	Anguilla
ATG	Antigua and Barbuda
ARG	Argentina
ARM	Armenia
ABW	Aruba
AUS	Australia
AUT	Austria)
AZE	Azerbaijan
BHS	Bahamas
BHR	Bahrain
BGD	Bangladesh
BRB	Barbados
BLR	Belarus
BEL	Belgium
BLZ	Belize
BEN	Benin
BMU	Bermuda
BTN	Bhutan
BOL	Bolivia, Plurinational State of
BES	Bonaire, Saint Eustatius and Saba
BIH	Bosnia and Herzegovina
BWA	Botswana
BRA	Brazil)
VGB	British Virgin Islands
BRN	Brunei Darussalam
BGR	Bulgaria
BFA	Burkina Faso
BDI	Burundi
KHM	Cambodia
CMR	Cameroon
CAN	Canada
CPV	Cape Verde
CYM	Cayman Islands
CAF	Central African Republic
TCD	Chad
CHL	Chile
CHN	China
COL	Colombia
COM	Comoros
COG	Congo
COK	Cook Islands

CRI	Costa Rica
CIV	Côte d'Ivoire
HRV	Croatia
CUB	Cuba
CUW	Curaçao
CYP	Cyprus
CZE	Czech Republic
PRK	Democratic People's Republic of Korea
COD	Democratic Republic of the Congo
DNK	Denmark
DJI	Djibouti
DMA	Dominica
DOM	Dominican Republic
ECU	Ecuador
EGY	Egypt
SLV	El Salvador
GNQ	Equatorial Guinea
ERI	Eritrea
EST	Estonia
ETH	Ethiopia
FRO	Faeroe Islands
FLK	Falkland Islands, Malvinas
FJI	Fiji
FIN	Finland
FRA	France
GUF	French Guiana
PYF	French Polynesia
GAB	Gabon
GMB	Gambia
GEO	Georgia
DEU	Germany
GHA	Ghana
GIB	Gibraltar
GRC	Greece
GRL	Greenland
GRD	Grenada
GLP	Guadeloupe
GUM	Guam
GTM	Guatemala
GGY	Guernsey
GIN	Guinea
GNB	Guinea-Bissau
GUY	Guyana
HTI	Haiti
VAT	Holy See
HND	Honduras
HKG	China, Hong Kong Special
HUN	Hungary
ISL	Iceland
IND	India
IDN	Indonesia

IRN	Iran, Islamic Republic of
IRQ	Iraq
IRL	Ireland
IMN	Isle of Man
ISR	Israel
ITA	Italy
JAM	Jamaica
JPN	Japan
JEY	Jersey
JOR	Jordan
KAZ	Kazakhstan
KEN	Kenya
KIR	Kiribati
KWT	Kuwait
KGZ	Kyrgyzstan
LAO	Lao, People's Democratic Republic
LVA	Latvia
LBN	Lebanon
LSO	Lesotho
LBR	Liberia
LBY	Libya
LIE	Liechtenstein
LTU	Lithuania
LUX	Luxembourg
MAC	China, Macao Special Administrative Region
MDG	Madagascar
MWI	Malawi
MYS	Malaysia
MDV	Maldives
MLI	Mali
MLT	Malta
MHL	Marshall Islands
MTQ	Martinique
MRT	Mauritania
MUS	Mauritius
MYT	Mayotte
MEX	Mexico
FSM	Micronesia, Federated States of
MCO	Monaco
MNG	Mongolia
MNE	Montenegro
MSR	Montserrat
MAR	Morocco
MOZ	Mozambique
MMR	Myanmar
NAM	Namibia
NRU	Nauru
NPL	Nepal
NLD	Netherlands
NCL	New Caledonia
NZL	New Zealand

NIC	Nicaragua
NER	Niger
NGA	Nigeria
NIU	Niue
NFK	Norfolk Island
MNP	Northern Mariana Islands
NOR	Norway
PSE	Occupied Palestinian Territory
OMN	Oman
PAK	Pakistan
PLW	Palau
PAN	Panama
PNG	Papua New Guinea
PRY	Paraguay
PER	Peru
PHL	Philippines
PCN	Pitcairn
POL	Poland
PRT	Portugal
PRI	Puerto Rico
QAT	Qatar
KOR	Republic of Korea
MDA	Republic of Moldova
REU	Réunion
ROU	Romania
RUS	Russian Federation
RWA	Rwanda
SHN	Saint Helena
KNA	Saint Kitts and Nevis
LCA	Saint Lucia
SPM	Saint Pierre and Miquelon
VCT	Saint Vincent and the Grenadines
BLM	Saint-Barthélemy
MAF	Saint-Martin, French Part
WSM	Samoa
SMR	San Marino
STP	Sao Tome and Principe
SAU	Saudi Arabia
SEN	Senegal
SRB	Serbia
SYC	Seychelles
SLE	Sierra Leone
SGP	Singapore
SXM	Sint Maarten, Dutch Part
SVK	Slovakia
SVN	Slovenia
SLB	Solomon Islands
SOM	Somalia
ZAF	South Africa
SSD	South Sudan
ESP	Spain

LKA Sri Lanka  
SDN Sudan  
SUR Suriname  
SJM Svalbard and Jan Mayen Islands  
SWZ Swaziland  
SWE Sweden  
CHE Switzerland  
SYR Syrian Arab Republic  
TJK Tajikistan  
THA Thailand  
MKD The Former Yugoslav Republic of Macedonia  
TLS Timor-Leste  
TGO Togo  
TKL Tokelau  
TON Tonga  
TTO Trinidad and Tobago  
TUN Tunisia  
TUR Turkey  
TKM Turkmenistan  
TCA Turks and Caicos Islands  
TUV Tuvalu  
UGA Uganda  
UKR Ukraine  
ARE United Arab Emirates  
GBR United Kingdom of  
TZA United Republic of Tanzania  
USA United States of America  
VIR United States Virgin Islands  
URY Uruguay  
UZB Uzbekistan  
VUT Vanuatu  
VEN Venezuela, Bolivarian Republic of  
VNM Vietnam  
WLF Wallis and Futuna Islands  
ESH Western Sahara  
YEM Yemen  
ZMB Zambia  
ZWE Zimbabwe  
OTH Other, specify  
Unknown



## World Database for Pediatric and Congenital Heart Surgery

### Appendix D: Non Congenital Cardiac Anatomic Abnormality Terms and Definitions

#### Major Abnormality of Craniofacial (Head)

Any abnormality of the head including: **Choanal Atresia**, a congenital anomaly in which a bony or membranous occlusion blocks the passageway between the nose and pharynx; **Cleft lip**, a congenital anomaly consisting of one or more clefts in the upper lip that results from failure of the maxillary and median nasal process to close during embryonic development; **Cleft Palate**, a congenital fissure in the roof of the mouth, resulting from incomplete fusion of the palate during embryonic development. It may involve only the uvula or extend through the entire palate

#### Major Abnormalities of the Abdominal Wall.

Any abnormality of the abdominal wall including: **Congenital Diaphragmatic Hernia (CDH)**, A developmental defect of the diaphragm that allows abdominal viscera to herniate into the chest; **Gastroschisis**, a congenital defect characterized by a defect in the abdominal wall through which intestines protrude; **Omphalocele**, a defect in the medial anterior abdominal wall through which intraabdominal contents are extruded, usually at the base of the umbilical cord

#### Major Abnormality of the Biliary System

Biliary atresia is characterized by absence or discontinuity of the extrahepatic biliary system, resulting in obstruction to bile flow.

#### Major Abnormality of the Brain

Any abnormality of the brain including: **Hydrocephalus**, excessive CSF accumulation in the brain creating potentially harmful pressures; **Macrocephaly**, defined as a head circumference which is greater than 2 standard deviations larger than the average for age and sex; **Microcephaly**, defined as smaller than normal circumference of the head because the cerebral cortex has not developed properly or has stopped growing.

#### Major Abnormality of the Diaphragm

A developmental defect of the diaphragm that allows abdominal viscera to herniate into the chest.

#### Major Abnormality of the Gastrointestinal System

Any abnormality of the gastrointestinal system including: **Duodenal atresia and stenosis**, an absence or narrowing of the duodenum; **Jujenal atresia or stenosis**, an absence or narrowing of the middle section of the small bowel; **Ileal atresia or stenosis**, and absence or narrowing of a portion of the ileum; **Intestinal malrotation**, an abnormal placement and fixation of intestine; **Hirschsprung's disease**, A disorder of the enteric nervous system characterized by an absence of ganglion cells in the distal colon resulting in functional obstruction; **Colonic Atresia or stenosis**, an absence or narrowing of the large bowel extending to the rectum; **Rectal atresia or stenosis**, absence or narrowing of a portion of the rectum; **Imperforate anus**, a specific type atresia or the anal canal with or without a fistulous opening to an ectopic location on the perineum, within the urinary system, or into the vaginal vestibule

**Q:** If a patient has a condition that was corrected, for example, "imperforate anus", should this still be recorded?

**A:** Yes.

### **Major Abnormality of the Kidney, Ureter, or Bladder**

A major abnormality of the kidney(s), ureter(s) or bladder

### **Major Abnormality of the Larynx-Trachea- or Bronchus**

Any abnormality including: **Laryngomalacia**, abnormal laxity of the laryngeal support cartilage resulting in excessive inward collapse and collapse of the lumen with inspiration during spontaneous ventilation. Characterized by inspiratory stridor; **Congenital Tracheal Stenosis**, a primary tracheal narrowing at any level between the larynx and carina with significantly smaller than expected luminal diameter (not secondary to trauma or prolonged intubation). Frequently related to complete cartilagenous tracheal rings; **Trachesophageal Fistula (TEF)**, the presence of any type of patent communication below the larynx connecting the tracheo-bronchial tree to the esophagus. May be associated with other anomalies, including VATER, VACTERL and tracheal clefts. Typically, congenital, but may occur due to trauma or pressure necrosis; **Bronchomalacia**, A deficiency in the cartilaginous wall of the bronchus that may lead to atelectasis or obstructive emphysema.

### **Major Abnormality of the Lung**

Any abnormality of the lung including; **Congenital Lobar Emphysema (CLE)**, a developmental anomaly of the lower respiratory tract characterized by isolated hyperinflation of a lobe in the absence of extrinsic bronchial obstruction; **Cystic Congenital Adenomatous Malformation(CAM)**, a spectrum of cystic and solid lesions of the lung that result from abnormal embryogenesis and typically present with symptoms of respiratory distress in newborns and infants; **Cystic Fibrosis**, is an autosomal recessive genetic disorder affecting most critically the lungs, and also the pancreas, liver, and intestine. It is characterized by abnormal transport of chloride and sodium across an epithelium, leading to thick, viscous secretions; **Pulmonary Lymphangiectasia**, a rare developmental disorder involving the lung characterized by pulmonary subpleural, interlobar, perivascular and peribronchial lymphatic dilatation. PL presents at birth with severe respiratory distress, tachypnea and cyanosis, with a very high mortality rate at or within a few hours.

### **Major Abnormality of the Spine and Spinal cord**

Any abnormality of the spine or spinal cord including: **Myelomeningocele**, a defect in which a portion of the spinal cord protrudes through a gap in the vertebral column, frequently accompanied by hydrocephalus and mental retardation; **Spina Bifida**, characterized by defective closure of the vertebral canal with herniation of the spinal cord and or meninges; **Spinal Scoliosis**, a lateral curve in the spine, usually combined with rotation of the vertebra

**Q:** I have a patient who was adopted at age of 2 years old, and this information is unknown. What should I enter?

**A:** If not known to be present, then nothing would be recorded.

## World Database for Pediatric and Congenital Heart Surgery

### Appendix E: Chromosomal Abnormalities: Terms and Definitions

11p15.5

11q

12p1.21

12p12.1

12q24

15q21.1

1q42.1

20p12

#### **22q11 deletion**

Deletions or mutations involving the long arm of chromosome 22 (critical region 22q11.2) are associated with the DiGeorge sequence, velocardiofacial syndrome, conotruncal face anomaly syndrome, CATCH 22, and some isolated conotruncal malformations.

2p21

3p22

#### **45XOTurner syndrome**

(45XO) is a chromosomal deletion abnormality, which occurs in 1:5000 live female births. Although common in first trimester, most 45XO conceptuses are spontaneously aborted. Affected individuals are missing one X chromosome. The major features include short stature, primary amenorrhea due to ovarian dysgenesis, webbed neck, congenital lymphedema, and cubitus valgus. Cardiovascular abnormalities occur in 20-40% of cases, the most common of which is coarctation of the aorta (70%). Additional defects include commissural aortic valve, aortic stenosis, a spectrum of left-sided obstructive defects and/or hypoplastic defects; hypoplastic left heart syndrome, aortic dilation, dissection, and rupture

#### **47XXY**

Klinefelter, or 47XXY syndrome, is a sporadic chromosomal abnormality in which males have at least two X chromosomes and at least one Y chromosome. Incidence is 1:500 males or 1:1000 births. Klinefelter syndrome occurs usually in association with advanced maternal age at conception. It is the most common sex chromosome disorder and the most common cause of hypogonadism and infertility. Cardiovascular abnormalities in more than 50% of cases include mitral valve prolapse, varicose veins and deep venous thrombosis.

4p

4p16

**5p**

**6p12**

**7q11**

**7q11.23**

**7q32**

**7q34**

**8q12**

**TGFBR1 oor 2**

### **Trisomy 08**

Trisomy 8, or Warkany syndrome, is a chromosomal abnormality, which is a frequent cause of first trimester spontaneous abortions. Complete Trisomy 8 is usually an early lethal disorder. Incidence is 1:25,000-50,000 births. Affected individuals have an extra (or third) copy of chromosome 8. Cardiovascular abnormalities include septal defects and great vessel anomalies.

### **Trisomy 09**

Trisomy 9 or Rethore syndrome is a rare chromosomal abnormality, which is a frequent cause of first trimester spontaneous abortions. Incidence is 1:100,000 births. Affected individuals have an extra (or third) copy of chromosome 9. Most affected individuals die during infancy or early childhood. Cardiovascular abnormalities occur in 75% of cases and include VSD, ASD, PDA, valve defects, DORV, persistent left SVC, and endocardial fibroelastosis.

### **Trisomy 13**

Patau or Bartholin-Patau syndrome, or Trisomy 13, is a chromosomal abnormality. Incidence is 1:5000-10,000 births. Sporadic cases occur usually in association with advanced maternal age at conception. Affected individuals have an extra (or third) copy of chromosome 13. More than 90% of individuals with Trisomy 13 die within their first days or weeks of life. Only 5-10% survive beyond 1 year of age. Cardiovascular abnormalities in 80% of cases include VSD, PDA, ASD; dextrocardia in more than 50% of cases; and anomalous pulmonary venous connection, overriding aorta, pulmonary stenosis, hypoplastic aorta, mitral valve atresia, aortic valve atresia, and bicuspid aortic valve in fewer than 50% of cases.

### **Trisomy 18**

Edwards syndrome, or Trisomy 18, is a chromosomal abnormality. Incidence is 1:3000-5000 births. Sporadic cases of Edwards syndrome occur usually in association with advanced maternal age at conception. Affected individuals have an extra (or third) copy of chromosome 18. Approximately 50% of infants with Trisomy 18 die within the first week of life, approximately 40% die within the first month of life, only 5-10% survive beyond the first year. Cardiovascular abnormalities in more than 50% of cases include VSD, ASD and PDA; bicuspid aortic and/or pulmonary valves, nodularity of valve leaflets, pulmonic stenosis, coarctation of the aorta in 10-50% of cases; and anomalous coronary artery, TGA, TOF, dextrocardia and aberrant subclavian artery in less than 10% of cases.

**Trisomy 21**

Down syndrome, or Trisomy 21, is the most frequent chromosomal abnormality. Incidence is 1:600-1000 live births. Sporadic cases of Down syndrome occur in strong association with advanced maternal age at conception. Affected individuals have an extra (or third) copy of chromosome 21. Cardiovascular abnormalities in 40-50% of cases, in decreasing order of frequency, include AVSD, VSD, TOF and PDA. Left- sided obstructive defects, such as coarctation and aortic valve stenosis, are rare.

# World Database for Pediatric and Congenital Heart Surgery

## Appendix F: Pre-Operative Risk Factors

### Cardio-pulmonary resuscitation

Chest compression took place during the 48 hours prior to OR Entry Date and Time, or at the time of OR Entry Date and Time.

### Coagulation Disorder

Evidence of a PT/PTT above normal. Thrombocytopenia <100,000, or Fibrinogen split products positive (>10%) and the coagulopathy is NOT secondary to medications such as Heparin or Warfarin.

### Diabetes mellitus

Evidence of insulin dependent diabetes mellitus as manifested by the fact that the patient has the diagnosis of diabetes mellitus that is controlled with insulin or that is controlled with dietary modification with or without oral medications (oral antihyperglycemic agents).

### Previous History of Endocarditis

Use the Duke Criteria for the Diagnosis of Infective Endocarditis (IE): The definitive diagnosis of infective endocarditis requires one of the following four situations: 1) Histologic and/or microbiologic evidence of infection at surgery or autopsy such as positive valve culture or histology; 2) Two major criteria; 3) One major criterion and three minor criteria; 4) Five minor criteria. The two major criteria are: 1) Blood cultures positive for IE 2) Evidence of endocardial involvement. Blood cultures positive for IE requires: 1) Typical microorganism consistent with IE isolated from 2 separate blood cultures, as noted in number two below (viridans streptococci, Streptococcus bovis, Staphylococcus aureus, or HACEK group [HACEK, Haemophilus species {H. aphrophilus and Hparaaphrophilus}, Actinobacillusactinoinycetemcomitans, Cardiobacterium hominis, Eikenella corrodens, and Kingella kingae.]) or (Community-acquired enterococci in the absence of a primary focus); 2) Microorganisms consistent with IE isolated from persistently positive blood cultures defined as: (At least 2 positive cultures of blood samples obtained > 12 hours apart) or (All of 3 or a majority of 4 or more separate cultures of blood, the first and the last sample obtained > 1 hr apart); 3) Single blood culture positive for Coxiella burnetii or an antiphase I IgG antibody titer of >1 :800. Evidence of endocardial involvement requires 1) Positive results of echocardiography for IE defined as: (Oscillating intracardiac mass on the valve or supporting structures in the path of regurgitant jets or on implanted material in the absence of an alternative anatomic explanation) or (Abscess) or (New partial dehiscence of a valvar prosthesis) or 2) New valvar regurgitation (worsening or changing or preexisting murmur not sufficient). The six minor criteria are: 1) Predisposing heart disease or injection drug use (IVDA); 2) Temperature of > 38C; 3) Vascular phenomenon (major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial or conjunctival hemorrhage, Janeway's lesions); 4) Immunologic phenomenon (glomerulonephritis, Osler's nodes, Roth's spots, rheumatoid factor); 5) Microbiologic evidence (a positive blood culture that does not meet a major criterion as noted above) or serologic evidence of active infection with an organism consistent with IE; 6) Echocardiographic findings that are consistent with IE but do not meet a major criterion as noted above.

## **Endocrine Abnormalities**

Hypothyroidism refers to decreased levels of triiodothyronine (T3) and thyroxine (T4), and reverse triiodothyronine (reverse T3), with high levels of thyroid-stimulating hormone (TSH). Symptoms of hypothyroidism include bradycardia, pericardial effusions, hypertension and a narrowed pulse pressure and myxedema. Studies have also shown decreases in cardiac output and cardiac contractility, decreased diastolic relaxation and diastolic filling. In those with congestive heart failure (CHF), decreased levels of T3 have been shown to be proportional to New York Heart Association class, poor outcomes, mortality, poor hemodynamics, and hyponatremia. This factor may be coded (1) if the TSH > 20 mU / liter, or (2) if the patient has pituitary failure with hypothyroidism, or (3) if the patient is receiving medication to treat hypothyroidism.

## **Failure to Thrive**

Failure to thrive in childhood is a state of undernourishment due to inadequate caloric intake, inadequate caloric absorption, or excessive caloric expenditure.

## **Hepatic dysfunction**

Hepatic dysfunction is defined as dysfunction of the liver that results in hypoalbuminemia (<2 grams/dL), coagulopathy (PT > 1.5 x upper limits of normal), and hyperbilirubinemia (> 3.0 x upper limits of normal). Select this factor if the patient develops 2 out of these 3 laboratory abnormalities.

## **Mechanical ventilation to treat cardiorespiratory failure**

Supported with mechanical ventilation to treat cardiorespiratory failure during the hospitalization.

**Q:** Should this be selected if a patient was intubated at an outside hospital prior to coming to our hospital for surgery?

**A:** If the patient was intubated to treat respiratory failure, then you should indicate “yes” even if the patient was intubated at an outside hospital.

## **Necrotizing entero-colitis**

Necrotizing enterocolitis is defined as an acute reduction in the supply of oxygenated blood to the small intestine or large intestine, typically resulting in acidosis, abdominal distention, pneumatosis, and/or intestinal perforation, that prompts initiation of antibiotics or exploratory laparotomy.

## **Neurological deficit**

Indicate whether the patient had a stroke, CVA, or intracranial hemorrhage > Grade 2 at any time during the patient’s lifetime. A stroke is any confirmed neurological deficit of abrupt onset caused by a disturbance in blood flow to the brain, when the neurologic deficit does not resolve within 24 hours. An IVH (Intraventricular hemorrhage) is diagnosed by the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that may extend to include an intraparenchymal component. A Grade 1 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage with a limited germinal matrix involvement. A Grade 2 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that involves an area of up to, but not more than 50% of the ventricular cross-sectional area in sagittal view. A Grade 3 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that involves at least 50% of the

ventricular cross-sectional area in sagittal view but not an intraparenchymal component. A Grade 4 IVH requires the existence of a neurologic imaging study indicating a new or previously unsuspected collection of intraventricular hemorrhage that includes an intraparenchymal component extending beyond the germinal matrix.

### **Pacemaker present**

A pacemaker is a medical device that uses electrical impulses, delivered by electrodes contacting the heart muscles, to regulate the beating of the heart. The purpose of a pacemaker is to maintain an adequate heart rate, either because the heart's native pacemaker is not fast enough, or there is a block in the heart's electrical conduction system.

### **Preoperative complete AV block**

The absence of AV node conduction.

### **Preoperative/Preprocedural mechanical circulatory support (IABP, VAD, ECMO, or CPS)**

Patient is supported with mechanical support, of any type (IABP, VAD, ECMO, or CPS), for resuscitation/CPR or support,

### **Renal dysfunction**

Renal dysfunction is defined as the oliguria with sustained urine output  $< 0.5$  cc/kg/hr for 24 hours and/or a rise in creatinine  $> 1.5$  times upper limits of normal for age, without needing dialysis (including peritoneal dialysis and/or hemodialysis) or hemofiltration.

### **Renal failure requiring dialysis**

Renal failure is defined as oliguria with sustained urine output  $< 0.5$  cc/kg/hr for 24 hours and/or a rise in creatinine  $> 1.5$  times upper limits of normal for age, with need for dialysis (including peritoneal dialysis and/or hemodialysis) or hemofiltration.

### **Respiratory Failure not requiring ventilation**

Respiratory failure results from inadequate gas exchange by the respiratory system, meaning that the arterial oxygen, carbon dioxide or both cannot be kept at normal levels. A drop in the oxygen carried in blood is known as hypoxemia; a rise in arterial carbon dioxide levels is called hypercapnia.

### **Seizure**

A seizure is defined as the clinical and/or electroencephalographic recognition of epileptiform activity.

### **Sepsis**

Sepsis is defined as "evidence of serious infection accompanied by a deleterious systemic response". Sepsis may be diagnosed by the presence of a Systemic Inflammatory Response Syndrome (SIRS) resulting from suspected or proven infection. A systemic inflammatory response syndrome (SIRS) is present when at least two of the following criteria are present: hypo- or hyperthermia ( $>38.5$  or  $<36.0$ ), tachycardia or bradycardia, tachypnea, leukocytosis or leukopenia, and thrombocytopenia.



### **Shock, Persistent at time of surgery**

Shock is a "clinical condition characterized by signs and symptoms which arise when the cardiac output is insufficient to fill the arterial tree with blood under sufficient pressure to provide organs and tissues with adequate blood flow."

### **Shock, Resolved at time of surgery**

Shock is a "clinical condition characterized by signs and symptoms which arise when the cardiac output is insufficient to fill the arterial tree with blood under sufficient pressure to provide organs and tissues with adequate blood flow." This factor should be coded if shock was present at any time after the date and time of admission to the hospital but not at the time of OR Entry Date and Time, including situations where shock was present after admission to the hospital where this operation was performed, and situations where shock was present while the patient was hospitalized at another "transferring facility" that subsequently transferred the patient who ultimately arrived at this hospital in this same hospitalization.

### **Tracheostomy**

The patient has a tracheostomy present

### **None**

No pre-operative risk factors present

## **World Database for Pediatric and Congenital Heart Surgery**

### **Appendix G: Syndromes Terms and Definitions**

#### **Alagille Syndrome (intrahepatic biliary duct agenesis)**

Alagille-Watson syndrome, is an autosomal dominant condition [mapped to 20p12 & 1p13-p11] of intrahepatic biliary duct agenesis or arteriohepatic dysplasia. Incidence is 1:70,000 births. The 20-year predicted life expectancy is 75% for all patients, 80% for those not requiring a liver transplant, and 60% for those requiring a liver transplant. Typical manifestations include intrahepatic cholestasis, distinctive facies, anterior chamber abnormalities of the eye, and butterfly hemiverterbrae. The most common cardiovascular abnormality is peripheral pulmonary artery stenosis. Additional defects include ASD, VSD, coarctation of the aorta and TOF.

#### **Apert Syndrome**

Also known as Apert-Crouzon disease or Vogt cephalodactyly, is an autosomal dominant condition [mapped to 10q26] of acrocephalosyndactyly. Incidence is 1:65,000-88,000 births; it occurs in strong association with advanced paternal age at conception. Apert syndrome is similar to Crouzon and Pfeiffer syndromes. Cardiovascular abnormalities include pulmonic stenosis, VSD, overriding aorta, and endocardial fibroelastosis.

#### **Brugada Syndrome**

Also known as sudden unexplained nocturnal death syndrome (SUNDS), is an autosomal dominant condition [mapped to 3p21, 3p22.3, 12p13.3 & 10p12], occurring in 1:2000 births. Brugada syndrome is associated with the risk of sudden cardiac death. Mean age of sudden death is approximately 40 years. Symptoms include right bundle branch block and ST segment elevation on ECG, idiopathic ventricular fibrillation, and cardiac arrest. Brugada syndrome, in its typical form is sinus rhythm with anterior raised ST segment in V1 and V2 due to a genetic ion-channel defect involving a sodium-channel defect isolated to SCN5A gene. Brugada syndrome is a type of "Channelopathy". A ventricular tachycardia due to a genetic ion-channel defect is also known as a "Channelopathy" or "Ion channelopathy". This diagnosis is most commonly Long QT syndrome, but also includes Brugada syndrome, Jervell and Lange-Nielsen syndrome, Romano-Ward syndrome, Andersen syndrome, etc.

#### **Cardiofaciocutaneous Syndrome**

Cardiofaciocutaneous syndrome (CFC) is a sporadic condition [mapped to 7q34] affecting the heart, face, skin and hair. Incidence is 1:333,000-500,000 births. CFC is similar to Noonan and Costello syndromes. Cardiovascular abnormalities include pulmonary valve stenosis, ASD and hypertrophic cardiomyopathy.

#### **Carpenter Syndrome**

An autosomal recessive condition [mapped to 6p11] of acrocephalopolysyndactyly, type II. Incidence is 1:1,000,000 births. Cardiovascular abnormalities in 50% of cases include ASD, VSD, pulmonic stenosis, TOF, TGA and PDA.

#### **Cat-eye Syndrome**

Also known as Schmid-Fraccaro syndrome, is an autosomal dominant condition [mapped to 22q11], associated with coloboma of the iris. Incidence is 1:50,000-150,000 births. The classic pattern of malformations includes mild mental deficiency, hypertelorism, down-slanting palpebral fissures, iris, coloboma, pre-auricular pits or tags, and anal and renal malformations. Cardiovascular abnormalities in 40% of cases include TAPVC, ASD, VSD, persistent left superior vena cava, TOF, interruption of the inferior vena cava, and tricuspid atresia.

### **Charge Association**

Also known as Hall-Hittner syndrome, is an autosomal dominant condition [mapped to 8q12.1 & 7q21.11]; some sporadic cases have been reported. Incidence is 1:8500-10,000 births. CHARGE syndrome is a nonrandom association of congenital anomalies which may include Coloboma, Heart defects, Atresia choanae, Retarded growth and development and/or central nervous system anomalies, Genital anomalies and/or hypogonadism and Ear anomalies and/or deafness. Diagnosis is made if 4/6 major (or 3 major & 3 minor) defects are present. Heart defects are present in 75% to 80% of cases. Of those with heart defects, most have conotruncal anomalies (TOF, DORV, truncus arteriosus) and aortic arch anomalies (vascular ring, aberrant subclavian artery, IAA, coarctation of the aorta, right aortic arch, aortic valve stenosis). Other cardiovascular abnormalities include PDA, AVSD, VSD, and ASD.

### **Cornelia de Lange Syndrome**

Also known as de Lange or Brachmann-de Lange syndrome, is an autosomal dominant condition [mapped to 5p13.1, Xp11.22-11.21 & 10q25]; some X-linked and sporadic cases have been reported. Incidence is 1:10,000-30,000 births. Cardiovascular abnormalities in 25% of cases most commonly include VSD and ASD.

### **Costello Syndrome**

An autosomal dominant condition [mapped to 12p12.1 & 11p15.5]; some sporadic cases have been reported. Incidence is 1:1,000,000 births. Cardiovascular abnormalities include ASD, VSD, pulmonic stenosis, mitral valve prolapse, hypertrophic cardiomyopathy and arrhythmias.

### **Cri-du-chat Syndrome**

Also known as LeJeunesyndrome, is a chromosome deletion syndrome [mapped to 5p15.2]. Incidence is 1:20,000-50,000 births. Cri-du-chat refers to the distinctive cry of children with this disorder, caused by abnormal larynx development. Cardiovascular abnormalities in 30% of cases most commonly include VSD and ASD. Rare defects include TOF and AVSD.

### **Deletion 10p Syndrome**

Deletions on the short arm of chromosome 10 are associated with septal defects, particularly ASDs, and DiGeorge/velocardiofacial 2 syndrome.

### **Deletion 8p Syndrome**

Deletions on the short arm of chromosome 8 are associated with ASD, AVSC, conotruncal abnormalities, pulmonic valve stenosis and Tetralogy of Fallot.

### **DiGeorge Syndrome**

Also known as Shprintzen, Takao, velocardiofacial, or conotruncal anomaly face syndrome, is an autosomal dominant condition [mapped to 22q11.2]. Incidence is 1:4000 births. Cardiovascular anomalies are seen in association with hypoplasia or aplasia of the thymus and parathyroid gland, which are derivatives of pharyngeal pouches III and IV, and which can result in abnormalities of the immune system and calcium metabolism respectively. Cardiovascular abnormalities include conotruncal or outflow tract defects of the heart, such as tetralogy of Fallot, truncus arteriosus, and interrupted aortic arch, particularly type B IAA. Additional defects include VSD, right aortic arch, aberrant right subclavian artery, and PDA.

### **Down Syndrome**

Also known as Trisomy 21, is the most frequent chromosomal abnormality. Incidence is 1:600-1000 live births. Sporadic cases of Down syndrome occur in strong association with advanced

maternal age at conception. Affected individuals have an extra (or third) copy of chromosome 21. Cardiovascular abnormalities in 40-50% of cases, in decreasing order of frequency, include AVSD, VSD, TOF and PDA. Left-sided obstructive defects, such as coarctation and aortic valve stenosis, are rare.

### **Edwards Syndrome**

Also known as Trisomy 18, is a chromosomal abnormality. Incidence is 1:3000-5000 births. Sporadic cases of Edwards syndrome occur usually in association with advanced maternal age at conception. Affected individuals have an extra (or third) copy of chromosome 18. Approximately 50% of infants with Trisomy 18 die within the first week of life, approximately 40% die within the first month of life, only 5-10% survive beyond the first year. Cardiovascular abnormalities in more than 50% of cases include VSD, ASD and PDA; bicuspid aortic and/or pulmonary valves, nodularity of valve leaflets, pulmonic stenosis, coarctation of the aorta in 10-50% of cases; and anomalous coronary artery, TGA, TOF, dextrocardia and aberrant subclavian artery in less than 10% of cases.

### **Ehlers-Danlos Syndrome**

Is a group of inherited disorders marked by extremely loose joints, hyperelastic skin that bruises easily, and easily damaged blood vessels. A variety of gene mutations involve collagen of the skin, bone, blood vessels, and internal organs. The abnormal collagen leads to the symptom which can include rupture of internal organs or abnormal heart valves.

### **Ellis-van Creveld Syndrome**

Also known as chondroectodermal dysplasia, is an autosomal recessive condition [mapped to 4p16] of skeletal dysplasia. Incidence is 1:60,000- 200,000 births. Major features include short stature of prenatal onset (short limbs), hypoplastic nails and dental anomalies, postaxial polydactyly, narrow thorax, and cardiac defects. Cardiovascular abnormalities in more than 50% of cases most commonly include ASD or common atrium. Additional defects include PDA, persistent left superior vena cava, hypoplastic left heart defects, coarctation of the aorta, TAPVC, and TGA.

### **Fetal Alcohol Syndrome**

Indicate whether the patient has a history of Fetal alcohol syndrome (FAS). Fetal alcohol syndrome (FAS) is a condition that results from prenatal alcohol exposure. FAS is a group of problems that can include mental retardation, birth defects, abnormal facial features, growth problems, problems with the central nervous system, trouble remembering and/or learning, vision or hearing problems, and behavior problems. Mothers who consume large quantities of alcohol during pregnancy may have babies who are born with Fetal Alcohol Syndrome (or FAS). A diagnosis of FAS is based on three factors: 1) prenatal and postnatal growth retardation; 2) central nervous system abnormalities, and, 3) abnormalities of the face.

### **Fetal Drug Exposure**

Indicate whether the patient has a history of Fetal drug exposure. Fetal drug exposure can lead to numerous problems including low birth weight, prematurity, small for Gestational Age (SGA), failure to Thrive (FTT), neurobehavioral symptoms, infectious diseases, and Sudden Infant Death Syndrome (SIDS).

### **Fetal Rubella Syndrome (Congenital Rubella Syndrome)**

Indicate whether the patient has a history of maternal rubella virus infection during first trimester of pregnancy. Fetal rubella syndrome is associated with PDA, peripheral pulmonary stenosis, fibromuscular and intimal proliferation of medium and large arteries, VSD and ASD.

### **Goldenhar Syndrome**

Also known as hemifacialmicrosomia, oculoauriculovertebral dysplasia or spectrum, and facioauriculovertebralsequence, is an autosomal dominant condition [mapped to 14q32]. Incidence is 1:3000-5000 births. Cardiovascular abnormalities include VSD, PDA, TOF and coarctation

### **Heterotaxy Syndrome**

Synonymous with 'visceral heterotaxy' is defined as an abnormality where the internal thoraco-abdominal organs demonstrate abnormal arrangement across the left-right axis of the body. By convention, heterotaxy does not include patients with either the expected usual or normal arrangement of the internal organs along the left-right axis, also known as '**situs solitus**', nor patients with complete mirror-imaged arrangement of the internal organs along the left-right axis also known as '**situs inversus**'.

### **Heterotaxy Syndrome, Asplenia**

Is defined as a subset of heterotaxy with components of bilateral right-sidedness, usually associated with absence of the spleen.

### **Heterotaxy Syndrome, Polysplenia**

Is defined as a subset of Heterotaxy with components of bilateral left-sidedness, usually associated with multiple spleens.

### **Holt-Oram Syndrome**

Also known as heart hand, syndrome is an autosomal dominant condition [mapped to 12q24.1]. Incidence is 1:100,000 births. Holt-Oram syndrome was first described in 1960 by Holt and Oram who noted the association of radial anomalies with atrial septal defects. Cardiovascular abnormalities in 75% of cases most commonly include ASD. Additional defects include first degree AV block, bradycardia, fibrillation, AVSD, VSD, HLHS and PDA.

### **Jacobsen Syndrome**

Jacobsen syndrome is a chromosome deletion syndrome [mapped to 11q23]. Incidence is 1:100,000 births. Associated cardiovascular abnormalities include VSD and ASD.

### **Kabuki Syndrome**

Kabuki, or Niikawa-Kuroki, syndrome is an autosomal dominant condition. Incidence is 1:32,000 births. Affected individuals have a facial appearance similar to Japanese Kabuki theatre actors. Cardiovascular abnormalities in 50% of cases include ASD, VSD, coarctation of the aorta, bicuspid aortic valve, mitral valve prolapse, TOF, single ventricle with common atrium, DORV, TGA, and pulmonic, aortic and mitral valve stenosis.

### **Kartagener Syndrome**

Kartagener syndrome, also known as Siewert syndrome or primary ciliary dyskinesia, is an autosomal recessive condition [mapped to 9p21-p13]. Incidence is 1:30,000 births. Features include situs inversus and asplenia. Cardiovascular abnormalities include dextrocardia.

### **Klinefelter Syndrome (XXY)**

Klinefelter, or 47XXY syndrome, is a sporadic chromosomal abnormality in which males have at least two X chromosomes and at least one Y chromosome. Incidence is 1:500 males or 1:1000 births. Klinefelter syndrome occurs usually in association with advanced maternal age at conception. It is the most common sex chromosome disorder and the most common cause of

hypogonadism and infertility. Cardiovascular abnormalities in more than 50% of cases include mitral valve prolapse, varicose veins and deep venous thrombosis.

### **Leopard Syndrome**

LEOPARD is an acronym for multiple Lentigines, Electrocardiographic conduction abnormalities, Ocular hypertelorism, Pulmonic stenosis, Abnormal genitalia, Retardation of growth, and sensorineural Deafness. LEOPARD syndrome is an autosomal dominant condition [mapped to 12q24.1 & 3p25]. Cardiovascular abnormalities include pulmonic stenosis in 40% of cases, and hypertrophic cardiomyopathy in 20% of cases. Additional **defects include** subaortic stenosis, complete heart block, bundle branch block, prolonged P-R and QRS, and abnormal P waves.

### **Loeys-Dietz Syndrome**

Loeys-Dietz syndrome is an autosomal dominant condition [mapped to 3p22 & 9q22]. Cardiovascular abnormalities include aortic and arterial aneurysms/dissections with tortuosity of the arteries, PDA, ASD, bicuspid aortic and pulmonic valves, and mitral valve prolapse.

### **Long QT Syndrome (Ward Romano Syndrome)**

Long QT syndrome (LQTS), or Ward Romano syndrome, is an autosomal dominant condition [LQTS1 mapped to 11p15.5]. There are at least 11 known mutations for LQTS which have been mapped to at least 7 chromosomes. Long QT syndrome is characterized by a prolonged QT interval on EKG and is associated with the risk of sudden cardiac death. Incidence is 1:2000 births. Symptoms include ventricular arrhythmias, recurrent syncope, ventricular fibrillation, torsade de pointes, and cardiac arrest. Long QT syndrome (Ward Romano syndrome) is a ventricular tachycardia occurring in the setting of prolonged Q-T interval. Long QT syndrome is a type of "Channelopathy". A ventricular tachycardia due to a genetic ion-channel defect is also known as a "Channelopathy" or "Ion channelopathy". This diagnosis is most commonly Long QT syndrome, but also includes Brugada syndrome, Jervell and Lange-Nielsen syndrome, Romano-Ward syndrome, Andersen syndrome, etc.

### **Marfans Syndrome**

The most common connective tissue disorder, and is associated with the risk of sudden cardiac death. Cardiovascular abnormalities include aortic root dilation, aortic dissection and rupture, aortic regurgitation, ascending aortic aneurysm, mitral valve prolapse, mitral regurgitation, tricuspid valve prolapse, premature calcification of the mitral annulus, pulmonary artery dilatation and CHF.

### **Mucopolysaccharidosis Syndrome**

A group of disorders, including Hurlers syndrome, Hurler-Scheie syndrome, Hunter syndrome, and Scheie syndrome of lysosomal storage. Cardiovascular abnormalities include valve disease, coronary artery narrowing, and ventricular hypertrophy. Survival varies depending on the particular genetic configuration.

### **Noonan Syndrome**

Noonan syndrome is an autosomal dominant condition [mapped to 12q24.1]. Incidence is 1:1000-2500 births. Major features include short stature, seen in about half, mental retardation (usually mild), characteristic facial features, a shield chest deformity, cubitus valgus, and a short webbed neck. Cardiovascular abnormalities occur in at least 50% of cases and include pulmonary valve stenosis (75%) secondary to a dysplastic pulmonary valve with thickened valve leaflets, ASD (30%) usually associated with pulmonary stenosis, PDA (10%), VSD (10%), and

hypertrophic cardiomyopathy (10-20%) that can involve both ventricles. Rare lesions include TOF, coarctation of the aorta, subaortic stenosis, and Ebstein malformation.

### **Patau Syndrome (Trisomy 13)**

This is a disorder with an incidence of 1:5000-10,000 births. Sporadic cases occur usually in association with advanced maternal age at conception. Affected individuals have an extra (or third) copy of chromosome 13. More than 90% of individuals with Trisomy 13 die within their first days or weeks of life. Only 5-10% survive beyond 1 year of age. Cardiovascular abnormalities in 80% of cases include VSD, PDA, ASD; dextrocardia in more than 50% of cases; and anomalous pulmonary venous connection, overriding aorta, pulmonary stenosis, hypoplastic aorta, mitral valve atresia, aortic valve atresia, and bicuspid aortic valve in fewer than 50% of cases.

### **Pierre Robin Syndrome**

Characterized by an unusually small mandible (micrognathia), posterior displacement or retraction of the tongue (glossoptosis), and upper airway obstruction. Incomplete closure of the roof of the mouth (cleft palate) is present in the majority of patients, and is commonly U-shaped.

### **Prune Belly Syndrome**

Characterized by three main features: Anterior abdominal wall musculature ("stomach muscles") deficient or absent, urinary tract anomalies (such as a very large bladder) and bilateral cryptorchidism (two undescended testicles.) The incidence of prune belly syndrome is about 1 in 40,000 births; 95% of cases occur in males. It is thought that prune belly syndrome is a multisystem disease complex that derives from a primary defect in mesodermal development at about 8 weeks' gestation. The major prognostic factor is the degree of dilation of the urinary tract; 20% of patients are stillborn, 30% die of renal failure or urosepsis within the first two years of life, and the remaining 50% have varying degrees of urinary pathology.

### **Rethore Syndrome (Trisomy 9)**

A rare chromosomal abnormality, which is a frequent cause of first trimester spontaneous abortions. Incidence is 1:100,000 births. Affected individuals have an extra (or third) copy of chromosome 9. Most affected individuals die during infancy or early childhood. Cardiovascular abnormalities occur in 75% of cases and include VSD, ASD, PDA, valve defects, DORV, persistent left SVC, and endocardial fibroelastosis.

### **Rubinstein-Taybi Syndrome**

An autosomal dominant condition [mapped to 16p13.3 & 22q13]. Incidence is 1:100,000-125,000 births. Cardiovascular abnormalities occur in 30% of cases and include ASD, VSD and PDA.

### **Short QT Syndrome**

Short QT syndrome appears to be an autosomal dominant condition [mapped to 7q35-q36, 11p15.5, 17q23.1-q24.2]. Short QT syndrome is characterized by a shortened QT interval on EKG and is associated with the risk of sudden cardiac death. Symptoms include episodes of syncope, palpitations, paroxysmal atrial fibrillation, ventricular fibrillation, and cardiac arrest. Short QT syndrome is a ventricular tachycardia occurring in the setting of short Q-T interval. Short QT syndrome is a type of "Channelopathy". A ventricular tachycardia due to a genetic ion-channel defect is also known as a "Channelopathy" or "Ion channelopathy". This diagnosis is most commonly Long QT syndrome, but also includes Brugada syndrome, Jervell and Lange-Nielsen syndrome, Romano-Ward syndrome, Andersen syndrome, etc.

### **Sickle Cell Disease**

An autosomal recessive genetic blood disorder with over dominance, characterized by red blood cells that assume an abnormal, rigid, sickle shape. Sickling decreases the cells' flexibility and results in a risk of various complications. The sickling occurs because of a mutation in the hemoglobin gene. Sickle-cell disease occurs more commonly in people (or their descendants) from parts of tropical and sub-tropical regions where malaria is or was common.

### **Sickle Cell Trait**

A condition in which a person has one abnormal allele of the hemoglobin beta gene (is heterozygous), but does not display the severe symptoms of sickle cell disease that occur in a person who has two copies of that allele (is homozygous). Those who are heterozygous for the sickle cell allele produce both normal and abnormal hemoglobin (the two alleles are co-dominant). Sickle cell disease is a blood disorder in which the body produces an abnormal type of the oxygen-carrying substance hemoglobin in the red blood cells. Sickling and sickle cell disease also confer some resistance to malaria parasitization of red blood cells, so that individuals with sickle-cell trait (heterozygotes) have a selective advantage in some environments.

### **Situs Inversus**

Defined as an abnormality where the internal thoraco-abdominal organs demonstrate mirror-imaged atrial arrangement across the left-right axis of the body.

### **Smith-Lemli-Opitz Syndrome**

An autosomal recessive condition [mapped to 11q12-q13]. Incidence is 1:20,000-40,000 births. Cardiovascular abnormalities include VSD, ASD, coarctation of the aorta, and PDA.

### **Turner Syndrome (45XO)**

A chromosomal deletion abnormality, which occurs in 1:5000 live female births. Although common in first trimester, most 45XO conceptuses are spontaneously aborted. Affected individuals are missing one X chromosome. The major features include short stature, primary amenorrhea due to ovarian dysgenesis, webbed neck, congenital lymphedema, and cubitus valgus. Cardiovascular abnormalities occur in 20-40% of cases, the most common of which is coarctation of the aorta (70%). Additional defects include bicommissural aortic valve, aortic stenosis, a spectrum of left-sided obstructive defects and/or hypoplastic defects, hypoplastic left heart syndrome; aortic dilation, dissection, and rupture.

### **VACTERL Syndrome (VACTER/VATER/VATERR Syndrome)**

A nonrandom association of defects, including Vertebral anomalies, Anal atresia, Cardiovascular anomalies, Tracheoesophageal fistula, Esophageal atresia, Renal and/or Radial anomalies, and Limb anomalies. Diagnosis is made if 3/7 defects are present. Incidence is 1:6000 births. Cardiovascular malformations include VSD, TOF, TGA and PDA.

### **Von Willebrand disease (vWD)**

The most common hereditary coagulation abnormality described in humans, although it can also be acquired as a result of other medical conditions. It arises from a qualitative or quantitative deficiency of von Willebrand factor (vWF), a multimeric protein that is required for platelet adhesion. There are three forms of vWD: inherited, acquired and pseudo or platelet type. There are three types of hereditary vWD: vWD Type I, vWD Type II and vWD III. Within the three inherited types of vWD there are various subtypes. Platelet type vWD is also an inherited condition. vWD Type I is the most common type of the disorder and those that have it are



typically asymptomatic or may experience mild symptoms such as nosebleeds although there may be severe symptoms in some cases. There are various factors that affect the presentation and severity of symptoms of vWD such as blood type.

### **Williams Syndrome (Williams-Beurn Syndrome) (7q11/7q11.23)**

An autosomal dominant condition [mapped to 7q11.23]. Incidence is 1:7500 births. Williams syndrome was initially described by Williams and colleagues in four unrelated children with mental retardation, an unusual facial appearance, and supravalvar stenosis. Cardiovascular abnormalities occur in at least 50% of cases and include supravalvar aortic stenosis, bicuspid aortic valve, mitral valve prolapse, mitral regurgitation, coronary artery stenosis, pulmonary valve stenosis, ASD, VSD and peripheral pulmonary artery stenosis. Supravalvar aortic stenosis is the most frequent single defect, but any of the systemic or pulmonary arteries can be affected.

### **Wolff-Parkinson-White Syndrome (WPW Syndrome)**

A condition in which there is an extra electrical pathway or circuit in the heart. Incidence is 1:500. WPW is one of the most common causes of tachycardia in infants and children. Symptoms may include chest pain, syncope, palpitations, shortness of breath, and can lead to episodes of tachycardia with heart rate >230 bpm. The "Wolff-Parkinson-White Syndrome (WPW syndrome)" is an "accessory connection-mediated tachycardia". An "accessory connection-mediated tachycardia" is a tachycardia secondary to accessory connection(s) or pathway(s). Typical accessory pathways are extra nodal pathways that connect the myocardium of the atrium and the ventricle across the AV groove and are classified by location, type of conduction (decremental versus nondecremental), and whether they are capable of anterograde (manifest, demonstrating pre-excitation on standard ECG) or retrograde (concealed) conduction, or both. The "Wolff- Parkinson-White Syndrome (WPW syndrome)" diagnosis is reserved for patients who have both pre- excitation on ECG (manifest conduction) and tachyarrhythmias.

### **Wolf-Hirschhorn Syndrome**

A chromosome deletion syndrome [mapped to 4p16.3]. Incidence is 1:96,000 births. Affected individuals have a 35% risk of mortality prior to age 2. Cardiovascular abnormalities include ASD and VSD.

### **Other Syndrome**

Select if patient has a syndrome other than those listed above.

**Q:** Does a syndrome like Tourette's belong in the syndrome field, the pre-operative factor field (as a neurological deficit), or in both?

**A:** Enter as Other Syndrome.

# World Database for Pediatric and Congenital Heart Surgery

## Appendix H: Complications

### **Arrhythmia requiring drug therapy**

A condition in which the heart beats with an irregular or abnormal rhythm requiring drug therapy.

### **Arrhythmia requiring electrical cardioversion or defibrillation**

A condition in which the heart beats with an irregular or abnormal rhythm requiring electrical cardioversion or defibrillation

### **Arrhythmia requiring Permanent pacemaker**

Implantation and utilization of a permanent pacemaker for treatment of any arrhythmia including heart block (atrioventricular [AV] heart block).

### **Bleeding, requiring reoperation**

Postoperative/postprocedural bleeding requiring reoperation

### **Cardiac dysfunction resulting in low cardiac output**

Low cardiac output state characterized by some of the following: tachycardia, oliguria, decreased skin perfusion, need for increased inotropic support (10% above baseline at admission), metabolic acidosis, widened Arterial - Venous oxygen saturation, need to open the chest, or need for mechanical support. If the cardiac dysfunction is of a severity that results in inotrope dependence, mechanical circulatory support, or listing for cardiac transplantation, please also code as "Cardiac failure (severe cardiac dysfunction)". A patient will be considered to have "inotrope dependence" if they cannot be weaned from inotropic support (10% above baseline at admission) after any period of 48 consecutive hours that occurs after the time of OR Exit Date and Time, and either (1) within 30 days after surgery in or out of the hospital, and (2) after 30 days during the same hospitalization subsequent to the operation. If patient meets criteria for severe cardiac dysfunction, only code "severe".

### **Cardiac failure (severe cardiac dysfunction)**

Low cardiac output state characterized by some of the following: tachycardia, oliguria, decreased skin perfusion, need for increased inotropic support (10% above baseline at admission), metabolic acidosis, widened Arterial - Venous oxygen saturation, need to open the chest, or need for mechanical support. This complication should be selected if the cardiac dysfunction is of a severity that results in inotrope dependence, mechanical circulatory support, or listing for cardiac transplantation. A patient will be considered to have "inotrope dependence" if they cannot be weaned from inotropic support (10% above baseline at admission) after any period of 48 consecutive hours that occurs after the time of OR Exit Date and Time and either (1) within 30 days after surgery in or out of the hospital, and (2) after 30 days during the same hospitalization subsequent to the operation. If patient meets criteria for severe cardiac dysfunction, only code "severe".

### **Chylothorax or pleural effusion, requiring drainage**

Presence of lymphatic fluid in the pleural space, commonly secondary to leakage from the thoracic duct or one of its main tributaries. Thoracocentesis is the gold standard for diagnosis and generally reveals a predominance of lymphocytes and/or a triglyceride level greater than 110 mg/dL

### **Endocarditis-postprocedural infective endocarditis**

Infective endocarditis in the setting of a heart which has been altered by surgery or intervention. Duke Criteria for the Diagnosis of Infective Endocarditis (IE): The definitive diagnosis of infective endocarditis requires one of the following four situations: 1) Histologic and/or microbiologic evidence of infection at surgery or autopsy such as positive valve culture or histology; 2) Two major criteria; 3) One major criterion and three minor criteria; 4) Five minor criteria. The two major criteria are: 1) Blood cultures positive for IE 2) Evidence of endocardial involvement. Blood cultures positive for IE requires: 1) Typical microorganism consistent with IE isolated from 2 separate blood cultures, as noted in number two below (viridans streptococci, Streptococcus bovis, Staphylococcus aureus, or HACEK group [HACEK, Haemophilus species {H. arophilus and H. paraaerophilus}, Actinobacillus actinoinyctemcomitans, Cardiobacterium hominis, Eikenella corrodens, and Kingella kingae.]) or (Community-acquired enterococci in the absence of a primary focus); 2) Microorganisms consistent with IE isolated from persistently positive blood cultures defined as: (At least 2 positive cultures of blood samples obtained > 12 hours apart) or (All of 3 or a majority of 4 or more separate cultures of blood, the first and the last sample obtained > 1 hr apart); 3) Single blood culture positive for Coxiella burnetii or an antiphase I IgG antibody titer of >1 :800. Evidence of endocardial involvement requires 1) Positive results of echocardiography for IE defined as: (Oscillating intracardiac mass on the valve or supporting structures in the path of regurgitant jets or on implanted material in the absence of an alternative anatomic explanation) or (Abscess) or (New partial dehiscence of a valvular prosthesis) or 2) New valvular regurgitation (worsening or changing or preexisting murmur not sufficient). The six minor criteria are: 1) Predisposing heart disease or injection drug use (IVDA); 2) Temperature of > 38C; 3) Vascular phenomenon (major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial or conjunctival hemorrhage, Janeway's lesions); 4) Immunologic phenomenon (glomerulonephritis, Osler's nodes, Roth's spots, rheumatoid factor); 5) Microbiologic evidence (a positive blood culture that does not meet a major criterion as noted above) or serologic evidence of active infection with an organism consistent with IE; 6) Echocardiographic findings that are consistent with IE but do not meet a major criterion as noted above.

### **Intraventricular hemorrhage (IVH) > grade 2**

An intraventricular hemorrhage involves bleeding into the brain's ventricular system, where the cerebrospinal fluid is produced and circulates towards the subarachnoid space. Grade I - bleeding occurs just in the germinal matrix; Grade II - bleeding also occurs inside the ventricles, but they are not enlarged; Grade III - ventricles are enlarged by the accumulated blood; Grade IV - bleeding extends into the brain tissue around the ventricles

### **Mechanical circulatory support (IABP, VAD, ECMO, or CPS)**

Utilization of postoperative/postprocedural mechanical support, of any type (IABP, VAD, ECMO, or CPS), for resuscitation/CPR or support, during the postoperative/postprocedural time period. Code this complication if it occurs (1) within 30 days after surgery or intervention regardless of the date of hospital discharge, or (2) after 30 days during the same hospitalization

### **Multi-System Organ Failure (MSOF) = Multi-Organ Dysfunction Syndrome (MODS)**

Multi-System Organ Failure (MSOF) is a condition where more than one organ system has failed (for example, respiratory failure requiring mechanical ventilation combined with renal failure requiring dialysis). (MODS). Only code this complication if the patient has failure of two or more than two organs. Do not code MSOF if only failing organs are the heart and lungs.

**Neurological deficit diagnosed in the operating room, persisting at discharge or 91 days if patient is still in hospital.**

Newly recognized and/or newly acquired (diagnosed in the operating room) deficit of neurologic function leading to inpatient referral, therapy, or intervention not otherwise practiced for a similar unaffected inpatient, with a persisting neurologic deficit present at hospital discharge or 91 days if patient is still in hospital.

**Neurological deficit diagnosed in the operating room, not present at discharge**

Newly recognized and/or newly acquired (in the operating room) deficit of neurologic function leading to inpatient referral, therapy, or intervention not otherwise practiced for a similar unaffected inpatient, with no persisting neurologic deficit present at hospital discharge or 91 days if patient is still in hospital.

**Neurological deficit that occurred after the operating room visit, persisting at discharge**

Newly recognized and/or newly acquired (diagnosed after the operating room visit) deficit of neurologic function leading to inpatient referral, therapy, or intervention not otherwise practiced for a similar unaffected inpatient, with a persisting neurologic deficit present at hospital discharge or 91 days if patient is still in hospital.

**Neurological deficit that occurred after the operating room visit, not present at discharge**

Newly recognized and/or newly acquired (diagnosed after the operating room visit) deficit of neurologic function leading to inpatient referral, therapy, or intervention not otherwise practiced for a similar unaffected inpatient, with no persisting neurologic deficit present at hospital discharge or 91 days if patient is still in hospital.

**Paralyzed diaphragm (possible phrenic nerve injury), requiring surgical plication**

Presence of elevated hemi-diaphragm(s) on chest radiograph in conjunction with evidence of weak, immobile, or paradoxical movement assessed by ultrasound or fluoroscopy.

**Pericardial Effusion, requiring drainage**

Abnormal accumulation of fluid in the pericardial space, Requiring drainage, By any technique.

**Peripheral nerve injury persisting at discharge or 91 days if patient is still in hospital**

Newly acquired or newly recognized deficit of unilateral or bilateral peripheral nerve function indicated by physical exam findings, imaging studies, or both.

**Peripheral nerve injury not present at discharge or 91 days if patient is still in hospital**

Newly acquired or newly recognized deficit of unilateral or bilateral peripheral nerve function indicated by physical exam findings, imaging studies, or both.

**Postoperative/Postprocedural respiratory insufficiency requiring mechanical ventilatory support > 7 days**

Respiratory insufficiency requiring mechanical ventilatory support from surgery or procedure to greater than 7 days postoperatively/postprocedurally. In other words, the inability of the patient to exchange oxygen and carbon dioxide in sufficient quantities to avoid unacceptable hypercarbia, hypoxemia, or both, without mechanical ventilatory support for greater than 7 days during the postoperative or postprocedural period. The patient therefore does utilize mechanical ventilatory support for greater than 7 days during the postoperative or post procedural period.

**Postoperative/Postprocedural respiratory insufficiency requiring reintubation**

Reintubation required after initial extubation. In other words, the need to reinstitute postoperative or postprocedural mechanical ventilation after a planned extubation and prior to discharge, or after a planned extubation and after discharge but within 30 days of surgery. The

intent of this field is to capture Postoperative/Postprocedural respiratory insufficiency requiring reintubation. It is not intended to capture situations where a patient may undergo elective intubations for other additional operations or procedures (including percutaneous endoscopic gastrostomy [PEG], tube insertions, catheter placement, cardiac catheterizations, etc.). However, these elective intubations and extubations are included and counted when determining "Final Extubation Date and Time".

### **Pulmonary vein obstruction**

Clinically significant stenosis or obstruction of pulmonary veins. Typically diagnosed by echocardiography or cardiac catheterization, this may present with or without symptoms. A "clinically significant" event or condition is an event or condition that necessitates a change in treatment

### **Renal failure - acute renal failure, Acute renal failure requiring dialysis at the time of hospital discharge or 91 days if patient is still in hospital**

Acute renal failure is defined as new onset oliguria with sustained urine output < 0.5 cc/kg/hr for 24 hours and/or a rise in creatinine > 1.5 times upper limits of normal for age (or twice the most recent preoperative/preprocedural values if these are available), with eventual need for dialysis (including peritoneal dialysis and/or hemodialysis) or hemofiltration. Code this complication if the patient requires dialysis at the time of hospital discharge or death in the hospital.

### **Renal failure - acute renal failure, Acute renal failure requiring temporary dialysis with the need for dialysis not present at hospital discharge or 91 days if patient is still in hospital**

Acute renal failure is defined as new onset oliguria with sustained urine output < 0.5 cc/kg/hr for 24 hours and/or a rise in creatinine > 1.5 times upper limits of normal for age (or twice the most recent preoperative/preprocedural values if these are available), with eventual need for dialysis (including peritoneal dialysis and/or hemodialysis) or hemofiltration. Code this complication if the patient does not require dialysis at the time of hospital discharge or death in the hospital.

### **Renal failure - acute renal failure, Acute renal failure requiring temporary hemofiltration with the need for dialysis not present at hospital discharge or 91 days if patient is still in hospital**

Acute renal failure is defined as new onset oliguria with sustained urine output < 0.5 cc/kg/hr for 24 hours and/or a rise in creatinine > 1.5 times upper limits of normal for age (or twice the most recent preoperative/preprocedural values if these are available), with eventual need for dialysis (including peritoneal dialysis and/or hemodialysis) or hemofiltration. Code this complication if the patient does not require dialysis at the time of hospital discharge or death in the hospital. (This complication should be chosen only if the hemofiltration was associated with acute renal failure.)

### **Respiratory failure, requiring tracheostomy**

Failure to wean from mechanical ventilation necessitating the creation of a surgical airway

### **Seizure**

A seizure is defined as the clinical and/or electroencephalographic recognition of epileptiform activity.

### **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). 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. 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. 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.

### **Spinal cord injury, Neurological deficit persisting at discharge**

*(Option added October 11, 2017)*

Newly acquired or newly recognized deficit of spinal cord function indicated by physical exam findings, imaging studies, or both.

### **Stroke: Ischemic**

A stroke is any confirmed neurological deficit of abrupt onset caused by a disturbance in blood flow to the brain, when the neurologic deficit does not resolve within 24 hours.

### **Subdural Bleed**

Bleeding between the dura mater, and the brain. Usually resulting from tears in bridging veins which cross the subdural space, subdural hemorrhages may cause an increase in intracranial pressure (ICP), which can cause compression of and damage to delicate brain tissue. Subdural hematomas are often life-threatening when acute. Chronic subdural hematomas, however, have a better prognosis if properly managed

### **Systemic vein obstruction**

Clinically significant stenosis or obstruction of any major systemic vein (e.g., superior vena cava, inferior vena cava, femoral veins, internal jugular veins, etc.). A "clinically significant" event or condition is an event or condition that necessitates a change in treatment

### **Unplanned cardiac reoperation during the postoperative or postprocedural time period, exclusive of reoperation for bleeding**

Any additional unplanned cardiac operation occurring (1) within 30 days after surgery or intervention in or out of the hospital, or (2) after 30 days during the same hospitalization subsequent to the operation or intervention. A cardiac operation is defined as any operation that is of the operation type of "CPB" or "No CPB Cardiovascular". The following operations will always be coded as "Planned Reoperation": (1) Delayed Sternal Closure, (2) ECMO Decannulation, (3) VAD Decannulation, (4) Removal of Broviac catheter. The following operations will always be coded as "Unplanned Reoperation": (1) Mediastinal exploration for infection, (2) Mediastinal exploration for hemodynamic instability, (3) Emergent mediastinal exploration for initiation of ECMO or VAD, (4) Reoperation for residual or recurrent lesion. Mediastinal exploration for bleeding is always coded separately as "Bleeding, Requiring reoperation".

### **Vocal cord dysfunction (possible recurrent laryngeal nerve injury)**

Presence of poor or no vocal cord movement assessed by endoscopy. Patient may or may not have stridor, hoarse voice or poor cry, in conjunction with endoscopic findings.

### **Wound dehiscence-Median Sternotomy**

*(Option added October 11, 2017)*

Wound dehiscence (sterile) is defined as separation of the layers of a surgical wound. This separation is deep to include the sternotomy. Wound dehiscence due to wound infections should be listed as Wound infection-Mediastinitis.

### **Wound infection-Mediastinitis**

The diagnosis of Mediastinitis must meet one of the following criteria: Criterion 1: Patient has organisms cultured from mediastinal tissue or fluid that is obtained during a surgical operation or by needle aspiration. Criterion 2: Patient has evidence of mediastinitis by histopathologic examination or visual evidence of mediastinitis seen during a surgical operation. Criterion 3: Patient has at least ONE of the following numbered signs or symptoms with no other recognized cause: 1) fever, 2) chest pain, or 3) sternal instability AND at least one of the following numbered features: 1) purulent mediastinal drainage, 2) organisms cultured from mediastinal blood, drainage or tissue, or 3) widening of the cardio-mediastinal silhouette. Criterion 4: Patient  $\leq$  1 year of age has at least one of the following numbered signs or symptoms with no other recognized cause: 1) fever, 2) hypothermia, 3) apnea, 4) bradycardia, or 5) sternal instability AND at least one of the following numbered features: 1) purulent mediastinal discharge, 2) organisms cultured from mediastinal blood, drainage or tissue, or 3) widening of the cardio-mediastinal silhouette. Infections of the sternum (sternal osteomyelitis) should be classified as mediastinitis.

### **Wound infection-Superficial wound infection**

A superficial wound infection must meet the following numbered criteria: 1) The infection involves only the skin and the subcutaneous tissue of the incision and 2) The patient has at least ONE of the following lettered features: A) purulent drainage from the superficial portion of the incision, B) organisms isolated from an aseptically obtained culture of fluid or tissue from the superficial portion of the incision, C) at least ONE of the following numbered signs or symptoms: [1] pain or tenderness, [2] localized swelling, redness, or heat, and [3] the superficial portion of the incision is deliberately opened by a surgeon, unless the incision is culture negative, or D) a diagnosis of superficial wound infection by the surgeon or by the attending physician.

