

# 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.