

## 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**
- **Cardiac Trauma** (*Option added July 2, 2019*)
- **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 July 9, 2019*)

### **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).

### **Pericardial Disease** *(Added April 24, 2018)*

- **Pericardial Disease (non specific)**

### **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) supravalvar 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 Sling**  
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.