

SELECTED TERMS USED IN ADULT CONGENITAL HEART DISEASE

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A

aberrant innominate artery

A rare abnormality associated with right aortic arch comprising a sequence of arteries arising from the aortic arch—right carotid artery, right subclavian artery, and then (left) innominate artery—with the last passing behind the esophagus. This is in contrast to the general rule that the first arch artery gives rise to the carotid artery contralateral to the side of the aortic arch (ie, right carotid artery in left aortic arch and left carotid artery in right aortic arch). *Syn:* retroesophageal innominate artery.

aberrant subclavian artery

Right subclavian artery arising from the aorta distal to the left subclavian artery. Left aortic arch with (retroesophageal) aberrant right subclavian artery is the most common aortic arch anomaly. It was first described in 1735 by Hunauld and occurs in 0.5% of the general population. *Syn:* lusorian artery. *See also* [vascular ring](#).

absent pulmonary valve syndrome

Absent pulmonary valvular tissue, resulting in pulmonary regurgitation. This rare anomaly uncommonly may be isolated, or it may be associated with ventricular septal defect, obstructed pulmonary valve annulus, and massive dilation and distortion of the pulmonary arteries. Absent pulmonary valve may occur in association with other simple or complex congenital heart lesions, for instance as a variant of tetralogy of Fallot.

ACHD

Adult congenital heart disease.

ACHD eLearning Center (www.achdlearningcenter.org)

The ACHD e-Learning Center is jointly developed and maintained by the Cincinnati Children's Hospital Heart Institute and the International Society for Adult Congenital Heart Disease (www.isachd.org). High quality educational resources at basic and advanced levels are offered for: (1) healthcare professionals caring for adult patients with congenital heart defects; (2) cardiology trainees and cardiologists interested in learning more about adult patients with congenital heart defects; (3) ACHD teachers and training program directors.

Alagille syndrome

See [arteriohepatic dysplasia](#).

ALCAPA

Anomalous left coronary artery arising from the pulmonary artery. *See* [Bland-White-Garland syndrome](#).

ambiguus

With reference to cardiac situs, neither right nor left sided (indeterminate). Latin spelling is generally used for situs ambiguus. *Syn:* ambiguous sidedness. *See also* [situs](#).

Amplatzer device

A group of self-centering devices delivered percutaneously by catheter for closure of abnormal intracardiac and vascular connections such as secundum atrial septal defect, patent foramen ovale or patent ductus arteriosus.

Anderson-Fabry disease

See [Fabry disease](#)

aneurysm of sinus of Valsalva

See [sinus of Valsalva/aneurysm](#).

anomalous pulmonary venous connection

Pulmonary venous connection to the right side of the heart, which may be total or partial.

- **partial anomalous pulmonary venous connection (PAPVC)**

One or more, but not all, the pulmonary veins connect to the right atrium directly or via a vena cava. This anomaly is frequently associated with sinus venosus defect. *See also* [Scimitar syndrome](#), [sinus venosus defect](#).

- **total anomalous pulmonary venous connection (TAPVC)**

All pulmonary veins connect to the right side of the heart, either directly or via venous tributaries. The connection may be supradiaphragmatic, usually via a vertical vein to the innominate vein or the superior vena cava. The connection may also be infradiaphragmatic via a descending vein to the portal vein, the inferior vena cava, or one of its tributaries. Pulmonary venous obstruction is common in supradiaphragmatic connection and almost universal in infradiaphragmatic connection.

aortic arch anomalies

Abnormalities of the aortic arch and its branching. Left or right aortic arch is defined by the mainstem bronchus that is crossed by the descending thoracic aorta and does not refer to the side of the midline on which the aorta descends. In left aortic arch (normal anatomic arrangement), the descending thoracic aorta crosses over the left mainstem bronchus; the innominate artery branching into the right carotid and right subclavian artery arises first, the left carotid artery second, and the left subclavian artery third. Usually, the first aortic arch vessel gives rise to the carotid artery that is opposite to the side of the aortic arch (ie, the right carotid artery in left aortic arch and the left carotid artery in right aortic

arch). The most important anomalies are described as follows:

- **abnormal left aortic arch:** Left aortic arch with minor branching anomalies; left aortic arch with aberrant right subclavian artery.
- **cervical aortic arch:** The arch extends above the level of the clavicle.
- **double aortic arch:** Both right and left aortic arches are present; that is, the ascending aorta splits into two limbs encircling the trachea and esophagus, and the two limbs join to form a single descending aorta. There are several forms, such as widely open right and left arches or hypoplasia/atresia of one arch (usually the left). This anomaly is commonly associated with patent ductus arteriosus. Double aortic arch creates a vascular ring around the trachea and the esophagus. *See also* [vascular ring](#).
- **interrupted aortic arch:** Complete discontinuation between the ascending and descending thoracic aorta.
 - **type A interrupted aortic arch:** Interruption distal to the subclavian artery that is ipsilateral to the second carotid artery.
 - **type B interrupted aortic arch:** Interruption between the second carotid artery and the ipsilateral subclavian artery.
 - **type C interrupted aortic arch:** Interruption between carotid arteries.
- **isolation of contralateral arch vessels:** An aortic arch vessel arises from the pulmonary artery via the ductus arteriosus without connection to the aorta; this anomaly is very uncommon. Isolation of the left subclavian artery is the most common form.
- **persistent fifth aortic arch:** Double-lumen aortic arch with both lumina on the same side of the trachea. Degree of luminal patency varies from full patency of both lumina to complete atresia of one of them.
- **right aortic arch:** The descending thoracic aorta crosses the right mainstem bronchus. Right aortic arch is often associated with tetralogy of Fallot, pulmonary atresia, truncus arteriosus, and other conotruncal anomalies. Types of right aortic arch branching include:
 - mirror image branching (left innominate artery, right carotid artery, right subclavian artery);
 - aberrant left subclavian artery with a normal caliber; sequence of branching: left carotid artery, right carotid artery, right subclavian artery, left subclavian artery;
 - retroesophageal diverticulum of Kommerell; *See* [diverticulum of Kommerell](#);
 - right aortic arch with left descending aorta (i.e., retroesophageal segment of right aortic arch); the descending aortic arch crosses the midline toward the left by a retroesophageal route.

aortic override

See [tetralogy of Fallot](#).

aortic valve-sparing aortic root replacement

See [David operation](#); [Yacoub procedure](#).

aorto–left ventricular defect (tunnel)

Vascular connection between the aorta and the left ventricle resulting in left ventricular volume overload due to regurgitation from the aorta via the tunnel to the left ventricle.

aortopulmonary collateral

Abnormal arterial vessel arising from the aorta, providing blood supply to the lungs. This vessel may be single or multiple and small or large (*see also* [MAPCA](#)) and may be associated with tetralogy of Fallot, pulmonary atresia, or other complex cyanotic congenital heart disease.

aortopulmonary septal defect

See [aortopulmonary window](#).

aortopulmonary window

A congenital connection between the ascending aorta and the main pulmonary artery, which may be contiguous with the semilunar valves or, less often, separated from them. It simulates the physiology of a large patent ductus arteriosus but requires a more demanding repair. *Syn:* aortopulmonary septal defect.

APSACHD

Asia-Pacific Society for Adult Congenital Heart Disease (<http://www.apsachd.org>)

arterial switch operation (ASO)

See [Jatene procedure](#).

arteriohepatic dysplasia

An autosomal dominant multisystem syndrome consisting of intrahepatic cholestasis, characteristic facies, butterfly-like vertebral anomalies, and varying degrees of peripheral pulmonary artery stenosis or diffuse hypoplasia of the pulmonary artery and its branches. It is associated with microdeletion in chromosome 20p. *Syn:* Alagille syndrome.

ASO

arterial switch operation. *See* [Jatene procedure](#).

asplenia syndrome

See [isomerism/right isomerism](#).

atresia, atretic

Imperforate; used with reference to an orifice, valve, or vessel.

atrial maze procedure

An intervention for atrial fibrillation directed toward restoring normal rhythm by interrupting conduction of the abnormal atrial impulses. It was originally (and is still) performed by creating surgical incisions in the atrium that are then re-sewn, thus creating electrical barriers that disrupt reentrant circuits. Similar electrical barriers often can be created in the electrophysiology laboratory without the need for thoracotomy. The originally described procedure, involving incisions in both atria, is called the Cox maze procedure (Cox JL, Schuessler RB, D'Agostino HJ Jr, et al. The surgical treatment of atrial fibrillation: III. Development of a definitive surgical procedure. *J Thorac Cardiovasc Surg*. 1991;101:569-583).

atrial septal defect (ASD)

An interatrial communication, classified according to its location relative to the oval fossa (fossa ovalis).

- **coronary sinus “ASD” (properly termed coronary sinus defect):** Inferior and anterior to the oval fossa at the anticipated site of the orifice of the coronary sinus. May be part of a

complex anomaly including absence of the coronary sinus and a persistent left superior vena cava. *See also* [Raghib complex](#).

- **ostium primum ASD:** Part of the spectrum of atrioventricular septal defect (AVSD). Located anterior and inferior to the oval fossa such that there is no atrial septal tissue between the lower edge of the defect and the atrioventricular valves. The atrioventricular valves are located on the same plane. An ostium primum ASD is almost always associated with a “cleft” in the left atrioventricular valve (“anterior mitral leaflet”). This cleft is actually the separation between the left-sided portions of the primitive anterosuperior and postero-inferior bridging leaflets. *See also* [atrioventricular septal defect \(AVSD\)](#).
- **ostium secundum ASD:** Located within the true interatrial septum at the level of the oval fossa. The defect is actually a defect in the embryologic septum primum that otherwise constitutes the floor of the oval fossa.
- **“sinus venosus ASD” (so-called)** *See* [sinus venosus defect](#).

atrial switch procedure

A procedure to redirect venous return to the contralateral ventricle. When used in complete transposition of the great arteries (Mustard or Senning procedure), this accomplishes physiologic correction of the circulation while leaving the right ventricle to support the systemic circulation. It is also used in congenitally corrected transposition of the great arteries in combination with an arterial switch operation (Jatene procedure). *See also* [double-switch procedure](#).

atrioventricular concordance

See [concordant atrioventricular connections](#).

atrioventricular discordance

See [discordant atrioventricular connections](#).

atrioventricular septal defect (AVSD)

A group of anomalies resulting from a deficiency of the atrioventricular (AV) septum that have in common (1) a common AV junction with a common fibrous ring and a unique, five-leaflet, AV valve; (2) unwedging of the aorta from its usual position deeply wedged between the mitral and tricuspid valves; (3) a narrowed subaortic outflow tract; (4) disproportion between the inlet and outlet portions of the ventricular septum. The valves should be referred to as *left AV valve* and *right AV valve* or as *common AV valve*. There is no true mitral or tricuspid valve associated with an AVSD. Echocardiographic recognition is aided by the observation that left and right AV valves are located in the same anatomic plane. Included in this group of conditions are anomalies previously known as (and often still described as) ostium primum ASD (partial AVSD), “cleft” in the adjoining leaflet of the left AV valve and/or right AV valve, inlet VSD, and complete AVSD (“complete AV canal defect”). An older, obsolete term describing such a defect is *endocardial cushion defect*. *See also* [endocardial cushion defect](#).

atrioventricular septum

The atrioventricular septum separates the right atrium from the left ventricle. Its superior border is the septal attachment of the anterior mitral leaflet, and its inferior border the attachment of the septal leaflet of the tricuspid valve, which is apically offset from the anterior mitral leaflet. The atrioventricular septum has two parts: an inferior muscular portion which is at the apex of the muscular interventricular septum and a superior fibrous portion. *See also* [Gerbode defect](#).

atrioventricular valve (AV valve)

A valve guarding the inlet to a ventricle. AV valves correspond with their respective ventricles: the tricuspid valve is always associated with the right ventricle, and the mitral valve is associated with the left ventricle. However, in the setting of an atrioventricular septal defect, there is neither a true mitral nor a true tricuspid valve. Rather, in severe forms there is a single atrioventricular orifice, guarded by a five-leaflet AV valve. The left AV valve comprises the left lateral leaflet and the left portions of the superior (anterior) and inferior (posterior) bridging leaflets, while the right AV valve comprises the right inferior leaflet, the right anterosuperior leaflet, and the right portions of the superior and inferior bridging leaflets.

- **cleft AV valve:** A defect due to incomplete fusion of the superior and inferior bridging leaflets that conjoin to form the left AV valve in AVSD. A cleft may also be seen in the septal tricuspid leaflet. A similar-appearing, but morphogenetically distinct, entity may involve the anterior or rarely the posterior leaflet of the mitral valve in otherwise normal hearts.
- **common AV valve:** Describes a five-leaflet AV valve in complete AVSD that is related to both ventricles.
- **overriding AV valve:** Describes an AV valve that empties into both ventricles. It overrides the interventricular septum above a ventricular septal defect.
- **straddling AV valve:** Describes an AV valve with anomalous insertion of tendinous cords or papillary muscles into the contralateral ventricle (an associated ventricular septal defect is obligatory).

autograft

Tissue or organ transplanted to a new site within the same individual.

AV septal defect (AVSD)

See [atrioventricular septal defect \(AVSD\)](#).

AV valve

See [atrioventricular valve](#).

azygos continuation of the inferior vena cava

An anomaly of systemic venous connections in which the inferior vena cava (IVC) is interrupted or absent prior to its passage through the liver. IVC flow reaches the right atrium through an enlarged azygos vein connecting the IVC to the superior vena cava. Usually, only hepatic venous flow reaches the right atrium from below. *See also* [isomerism](#).

B

Baffles operation

Diversion of the right pulmonary venous drainage to the right atrium and the inferior vena cava (IVC) drainage to the left atrium by using an allograft aortic tube to connect the IVC and the left atrium (Baffles TG. A new method for surgical correction of transposition of the aorta and pulmonary artery. *Surg Gynecol Obstet*. 1956;102:227-233). This obsolete operation provided partial physiologic correction in patients with complete transposition of the great arteries and was originally described by Lillehei and Varco in 1953 (Lillehei CW, Varco RL. Certain physiologic, pathologic, and surgical features of complete transposition of great vessels. *Surgery*. 1953;34:376-400).

baffle

A structure created surgically to divert blood flow. For instance, in atrial switch operations for complete transposition of the

great arteries, an intraatrial baffle is constructed to divert systemic venous return across the mitral valve thence to left ventricle and pulmonary artery and to divert pulmonary venous return across the tricuspid valve thence to right ventricle and aorta. *See also* [Mustard procedure](#), [Senning procedure](#).

balanced

As in “balanced circulation”—for example, in the setting of ventricular septal defect and pulmonary stenosis. The pulmonary stenosis is such that there is neither excessive pulmonary blood flow (which might lead to pulmonary hypertension) nor inadequate pulmonary blood flow (which might lead to marked cyanosis). *See also* [ventricular imbalance](#).

balloon atrial septostomy

See [Rashkind procedure](#).

Bentall procedure

Replacement of the ascending aorta and aortic valve with a composite graft/valve device and re-implantation of the coronary ostia into the sides of the conduit (Bentall H, DeBono A. A technique for complete replacement of the ascending aorta. *Thorax* 1968;23:338-339).

- **exclusion technique:** The native aorta is resected and replaced by the prosthetic graft.
- **inclusion technique:** The walls of the native aorta are wrapped around the graft so that the prosthetic material is “included.”

bicuspid aortic valve

An anomaly in which the aortic valve is composed of only two cusps instead of the usual three. There is often a raphe or aborted commissure dividing the larger cusp anatomically but not functionally. This anomaly is seen in approximately 2% of the general population, with male predominance (3-4:1). Associated aortic abnormality is common, including ascending aortopathy and coarctation of the aorta; in coarctation, approximately 75% of patients have an associated bicuspid aortic valve.

bidirectional cavopulmonary anastomosis (BCPC)

See [Glenn anastomosis/bidirectional Glenn cavopulmonary anastomosis](#).

Björk modification

See [Fontan procedure/RA-RV Fontan](#).

Blalock-Hanlon atrial septectomy

A palliative procedure to improve arterial oxygen saturation in patients with complete transposition of the great arteries. A surgical atrial septectomy is accomplished through a right lateral thoracotomy, excising the posterior aspect of the interatrial septum to provide mixing of systemic and pulmonary venous return at the atrial level (Blalock A, Hanlon CR. Surgical treatment of complete transposition of aorta and pulmonary artery. *Surg Gynecol Obstet*. 1950;90:1-15.)

Blalock-Taussig-Thomas anastomosis

A palliative operation (sometimes called Blalock-Taussig-Thomas shunt, BT shunt, BTT Shunt, Blalock-Taussig anastomosis) for the purpose of increasing pulmonary blood flow and hence systemic oxygen saturation. It involves creating an anastomosis between a subclavian artery and the ipsilateral pulmonary artery either directly with an end-to-side anastomosis (classic) or using an interposition tube graft (modified). (Blalock A, Taussig HB. The surgical treatment of malformations of the

heart in which there is pulmonary stenosis or pulmonary atresia. *J Am Med Assoc*. 1945;128:189-202; Brogan TV, Alfieri GM. Has the time come to rename the Blalock-Taussig shunt? *Pediatr Crit Care Med*. 2003;4(4):450-453).

Bland-White-Garland syndrome

The left main coronary artery arises from the main pulmonary artery (Bland EF, White PD, Garland J. Congenital anomalies of the coronary arteries: report of an unusual case associated with cardiac hypertrophy. *Am Heart J*. 1933;8:787-801). *Syn:* ALCAPA, anomalous left coronary artery from the pulmonary artery.

bridging leaflets

The superior and the inferior bridging leaflets of the AV valve are two leaflets uniquely found in association with AVSD. They “bridge,” or pass across, the interventricular septum. When the central part of the bridging leaflet tissue runs within the interventricular septum, the AV valve is functionally separated into left and right components; when the bridging leaflets do not run within the interventricular septum, but pass over its crest, a common AV valve guarding a common AV orifice (with an obligatory ventricular septal defect) is the result.

Brock procedure

An obsolete palliative operation to increase pulmonary blood flow and reduce right-to-left shunting in tetralogy of Fallot. It involved resection of part of the right ventricular infundibulum using a punch or biopsy-like instrument introduced through the right ventricle so as to reduce right ventricular outflow tract obstruction, without closure of the ventricular septal defect. The operation was performed without cardiopulmonary bypass (Brock RC. Pulmonary valvotomy for the relief of congenital pulmonary stenosis: report of three cases. *Br Med J*. 1948;1:1121-1126).

bulboventricular foramen

An embryologic term describing the connection between the left-sided inflow segments (primitive atrium and presumptive left ventricle) and the right-sided outflow segments (presumptive right ventricle and conotruncus) in the primitive heart tube. *Syn:* primary foramen, primary ventricular foramen, primary interventricular foramen.

C

Canadian Adult Congenital Heart (CACH) Network

A cooperative nationwide association of Canadian cardiologists, cardiac surgeons, and others, many of whom are situated in regional referral centers for adult congenital heart disease, dedicated to improving the care of patients with ACHD (<http://cachnet.ca>).

cardiac position

Position of the heart in the chest with regard to its location and the orientation of its apex.

- **cardiac location:** Location of the heart in the chest. Cardiac location is affected by many factors, including underlying cardiac malformation, abnormalities of mediastinal and thoracic structures, tumors, kyphoscoliosis, and abnormalities of the diaphragm:
 - levoposition: to the left
 - mesoposition: central
 - dextroposition: to the right
- **cardiac orientation:** The orientation of the apex of the heart relative to the base. The base to apex axis of the heart is defined by the alignment of the ventricles and is independent

of cardiac situs (sidedness). This axis is best described by echocardiography using the subcostal four-chamber views:

- levocardia: apex directed to the left
- mesocardia: apex oriented inferiorly
- dextrocardia: apex directed to the right

cardiac sidedness

See [situs](#).

cardiopulmonary study

A rest and stress study of cardiopulmonary physiology, including at least the following elements: resting pulmonary function, stress study to assess maximum workload, maximum oxygen uptake (VO_2 max), anaerobic threshold (AT), ventilatory efficiency (VE/VCO_2), and oxygen saturation with effort.

CardioSEAL device

A proprietary device delivered percutaneously by catheter for closure of a secundum atrial septal defect or patent foramen ovale.

Carpentier classification of Ebstein anomaly

A classification of severity of Ebstein anomaly from Type A to Type D according to increasing severity of the disease. (Carpentier A, Chauvaud S, Macé L, et al. A new reconstructive operation for Ebstein anomaly of the tricuspid valve. *J Thorac Cardiovasc Surg*. 1988;96:92-101.)

Carpentier-Chauvaud monocusp repair

Technique for tricuspid valve repair in Ebstein anomaly involving mobilization of the anterior tricuspid valve leaflet with reattachment to the anterior annulus, a tricuspid annuloplasty and plication of the atrialized RV. (Carpentier A, Chauvaud S, Macé L, et al. A new reconstructive operation for Ebstein anomaly of the tricuspid valve. *J Thorac Cardiovasc Surg*. 1988;96:92-101.)

CATCH-22

See [microdeletion 22q11.2 syndrome](#).

cat's eye syndrome

A syndrome due to a tandem duplication of chromosome 22q or an isodicentric chromosome 22, such that the critical region 22pter-22q11 is duplicated. Phenotypic features include mental deficiency, anal and renal malformations, hypertelorism, and others. Total anomalous pulmonary venous return is the most common congenital cardiac lesion, occurring in up to 40% of patients.

cavopulmonary connection

Surgically created connection between a vena cava and the pulmonary artery intended to deliver systemic venous blood to the pulmonary circulation. See also [Glenn anastomosis](#).

- **bidirectional cavopulmonary connection:** A cavopulmonary connection between the superior vena cava and the pulmonary arteries in which the pulmonary arteries are confluent, allowing caval blood to be delivered to both lungs. *Syn:* bidirectional Glenn shunt
- **total cavopulmonary connection (TCPC):** See [Fontan procedure/total cavopulmonary connection](#).

Celermajer index

An echocardiographic score for assessing the anatomic severity of Ebstein anomaly based on measurements of atrial and ventricular areas on an apical 4-chamber view. This score, originally described in neonates, involves calculating the ratio of the

combined area of the right atrium and atrialized right ventricle to that of the functional right ventricle and left atrium and left ventricle [RA area + area of the atrialized RV]/[area of functional RV + LA area + LV area]. (Celermajer DS, Cullen S, Sullivan ID, Spiegelhalter DJ, Wyse RK, Deanfield JE. *J Am Coll Cardiol*. 1992;19:1041-1046.)

Char syndrome

A genetic condition with autosomal dominant inheritance, caused by a mutation in TFAP2B gene and characterized by three major features: distinctive facial appearance, a congenital heart defect (usually a patent ductus arteriosus), and deformities of the fifth digits. The facial phenotype includes flattened cheekbones, a flat nasal bridge, ptosis, downslanting palpebral fissures, triangular-shaped mouth, and prominent, thick lips.

CHARGE association

Anomaly characterized by the presence of coloboma or choanal atresia and three of the following defects: congenital heart disease, nervous system anomaly or intellectual impairment, genital abnormalities, and ear abnormality or deafness. If coloboma and choanal atresia are both present, only two of the additional (minor) abnormalities are needed for diagnosis. Congenital heart defects seen in the CHARGE association are tetralogy of Fallot with or without other cardiac defects, atrioventricular septal defect, double-outlet right ventricle, double-inlet left ventricle, transposition of the great arteries, interrupted aortic arch, and others.

Chiari network

Fenestrated remnant of the right valve of the sinus venosus resulting from incomplete regression of this structure during embryogenesis. It was first described in 1897 (Chiari H. Ueber Netzbildungen im rechten Vorhof. *Beitr Pathol Anat*. 1897;22:1-10). The prevalence is 2% in autopsy and echocardiographic studies. It presents as coarse right atrial reticula connected to the eustachian and thebesian valves and attached to the crista terminalis. It may be associated with a patent foramen ovale and interatrial septal aneurysm. See also [sinus venosus](#).

cleft AV valve

See [atrioventricular valve](#); See also [atrial septal defect/ostium primum ASD](#).

coarctation of the aorta

A stenosis of the proximal descending aorta varying in anatomy, physiology, and clinical presentation. It may present as discrete or long-segment stenosis, is frequently associated with hypoplasia of the aortic arch and bicuspid aortic valve, and may be part of a [Shone complex](#).

common (as in AV valve, atrium, ventricle, etc.)

Implies bilateral structures with absent septation. Contrasts to "single," which implies absence of corresponding contralateral structure. See also [single](#).

common arterial trunk

See [truncus arteriosus](#).

common atrium

Large atrium characterized by a nonrestrictive communication between the bilateral atria due to absence of most of the atrial septum. Frequently associated with complex congenital heart disease (eg, isomerism, AVSD). See also [single](#) (atrium).

complete transposition of the great arteries (TGA)

An anomaly in which the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle. The right ventricle supports the systemic circulation. *Syn:* classic transposition; atrioventricular concordance with ventriculoarterial discordance; the terms *d*-transposition and *d*-TGA, although not true synonyms, are often used to refer to complete transposition of the great arteries.

concordant atrioventricular connections

Appropriate connection of morphologic right atrium to morphologic right ventricle and of morphologic left atrium to morphologic left ventricle. *Syn:* atrioventricular concordance.

concordant ventriculoarterial connections

Appropriate origin of pulmonary trunk from morphologic right ventricle and of aorta from morphologic left ventricle. *Syn:* ventriculoarterial concordance.

conduit

A structure that connects nonadjacent parts of the cardiovascular system, allowing blood to flow between them. It is often fashioned from prosthetic material and may include a valve.

cone reconstruction

A technique for tricuspid valve repair in Ebstein anomaly involving mobilization of the anterior and posterior tricuspid valve leaflets from their anomalous attachments in the right ventricle, rotating the complex clockwise to be sutured to the septal border of the anterior leaflet, thus creating a cone the vertex of which remains fixed at the right ventricular apex and the base of which is sutured to the true tricuspid valve annulus. The septal leaflet is incorporated into the cone wall if possible and the atrial septal defect is closed. (da Silva JP, Baumgratz JF, da Fonseca L, et al. The cone reconstruction of the tricuspid valve in Ebstein anomaly: the operation: early and midterm results. *J Thorac Cardiovasc Surg.* 2007;133:215-223.)

congenital coronary arteriovenous fistula (CCAVF)

A direct communication between a coronary artery and cardiac chamber, great artery, or vena cava, bypassing the coronary capillary network.

congenital heart disease

Anomalies of the heart originating in fetal life. Their expression may, however, be delayed beyond the neonatal period and may change with time as further postnatal physiologic and anatomic changes occur.

congenital heart block

Heart block developing during fetal life and present at birth. Can be isolated in an otherwise structurally normal heart or can be associated with congenital heart defects, particularly defects associated with left atrial isomerism. When congenital heart block occurs in a normal heart, it is not infrequently associated with maternal systemic lupus erythematosus with anti-Rh_o and anti-La autoantibodies.

congenital pericardial defect

A defect in the pericardium due to defective formation of the pleuropericardial membrane of the septum transversum. The spectrum of pericardial deficiency is wide. It may be partial or total. Its clinical diagnosis is difficult. Left-sided defects are more

common. Total absence of the pericardium may be associated with other defects such as bronchogenic cyst, pulmonary sequestration, hypoplastic lung, and other congenital heart diseases.

congenitally corrected transposition of the great arteries

An anomaly in which the atrioventricular connection is discordant such that the right atrium connects to the left ventricle and the left atrium connects to the right ventricle, and the ventriculoarterial connection is discordant such that the aorta arises from the right ventricle and the pulmonary artery from the left ventricle. There are usually associated anomalies, the most common being ventricular septal defect, left ventricular (pulmonary) outflow tract obstruction, and anomalies of the tricuspid valve (Ebstein-like malformation); any combination of these anomalies can coexist. Other associated anomalies (eg, aortic arch anomalies) are rare. The right ventricle supports the systemic circulation. *Syn:* CCTGA, atrioventricular discordance with ventriculoarterial discordance, double discordance. The terms l-transposition and l-TGA, although not true synonyms, are often and inappropriately used to refer to congenitally corrected transposition of the great arteries.

connection

Anatomic link between two structures (eg, venoatrial, atrioventricular, ventriculoarterial). An abnormal connection implies abnormal anatomic attachment of the structures. *Connection* and *drainage* are not interchangeable terms. *See also* [drainage](#).

conotruncal abnormality

Neural crest cell migration is crucial for conotruncal septation and the development of both the pulmonary and aortic outflow tracts. If neural crest cell migration fails, conotruncal abnormalities occur. Conotruncal anomalies include tetralogy of Fallot, truncus arteriosus, interrupted aortic arch, pulmonary atresia with ventricular septal defect, absent pulmonary valve, and *d*-malposition of the great arteries with double-outlet right ventricle, single ventricle, or tricuspid atresia. Abnormal neural crest migration may also be associated with complex clinical entities, such as 22q11.2 microdeletion syndrome.

conus

See [infundibulum](#).

CorMatrix patch

Prosthetic patch material used mainly for valve repairs.

cor triatriatum dexter

Abnormal septation of the right atrium due to failure of regression of the right valve of the sinus venosus. This yields a smooth-walled posteromedial "sinus" chamber (arising embryologically from the sinus venosus) that receives the venae cavae and (usually) the coronary sinus and a trabeculated anterolateral "atrial" chamber (arising embryologically from the primitive right atrium) that includes the right atrial appendage and is related to the tricuspid valve. Usually there is free communication between these two compartments, but variable obstruction to systemic venous flow from the "sinus" chamber to the "atrial" chamber may occur and may be associated with underdevelopment of downstream right-sided heart structures (eg, hypoplastic tricuspid valve, tricuspid atresia, pulmonary stenosis, or pulmonary atresia). A patent foramen ovale or an atrial septal defect is often present in relation to the posteromedial sinus chamber. *See also* [sinus venosus](#).

cor triatriatum sinister

Abnormal septation of the left atrium by a membrane that divides the left atrium into an accessory pulmonary venous chamber and a true left atrial chamber contiguous with the mitral valve. The pulmonary veins enter the accessory chamber, and the left atrial appendage is associated with the true left atrium. The connection between the accessory chamber and the true left atrium varies in size and may produce pulmonary venous obstruction. *Cor triatriatum* otherwise unmodified implies cor triatriatum sinister. Cor triatriatum sinister is to be differentiated from supraventricular mitral ring. See also [supraventricular mitral ring](#).

coronary sinus defect

See [atrial septal defect \(ASD\)/coronary sinus "ASD."](#)

Cox maze procedure

See [atrial maze procedure](#).

crisscross heart

A rotational abnormality of the ventricular mass around its long axis resulting in relationships of the ventricular chambers not anticipated from the given atrioventricular connections. If the rotated ventricles are in a markedly superoinferior relationship, the heart may also be described as a superoinferior or upstairs-downstairs heart. There may be ventriculoarterial concordance or discordance. *Syn:* crisscross atrioventricular connection.

crista supraventricularis

A saddle-shaped muscular crest in the right ventricular outflow tract intervening between the tricuspid valve and the pulmonary valve, consisting of septal and parietal components, which demarcates the junction between the outlet septum and the pulmonary infundibulum. Occasionally, but less accurately, this structure is termed *crista ventricularis*.

crista terminalis

A vestigial remnant of the right valve of the sinus venosus located at the junction of the trabeculated right atrial appendage and the smooth-walled "sinus" component of the right atrium that receives the inferior vena cava, the superior vena cava, and the coronary sinus. This is a feature of normal right atrial internal anatomy. *Syn:* terminal crest.

crista ventricularis

See [crista supraventricularis](#).

cyanosis

A bluish discoloration due to the presence of an increased quantity of desaturated hemoglobin in tissues. In congenital heart disease, cyanosis is generally due to right-to-left shunting through congenital cardiac defects, bypassing the pulmonary alveoli, or due to acquired intrapulmonary shunts (central cyanosis). Cyanosis can also occur as a result of increased peripheral extraction due, for instance, to critically reduced cutaneous flow (peripheral cyanosis).

D**d-transposition**

See [Complete transposition of the great arteries](#).

d-TGA

See [Complete transposition of the great arteries](#).

Dacron

A synthetic material often used to fashion conduits and other prosthetic devices for the surgical palliation or repair of congenital heart disease.

Damus-Kaye-Stansel (DKS) operation

The original operation involves anastomosis of the proximal end of the transected main pulmonary artery in an end-to-side fashion to the ascending aorta to provide blood flow from the systemic ventricle to the aorta; coronary arteries are not translocated and are perfused in a retrograde fashion. (Damus PS. Correspondence. *Ann Thorac Surg*. 1975;20:724-725; Kaye MP. Anatomic correction of transposition of the great arteries. *Mayo Clin Proc*. 1975;50:638-640; and Stansel HC Jr. A new operation for *d*-loop transposition of the great vessels. *Ann Thorac Surg*. 1975;19:565-567.) Originally, the procedure was introduced for biventricular repair in patients with transposition of the great arteries, and later applied for patients with abnormal ventriculoarterial connections who were not suitable for an arterial switch operation (e.g., transposition of the great arteries and unsuitable coronary patterns, double-outlet right ventricle with severe subaortic stenosis). The principle of the DKS operation has also been applied in patients with single ventricle physiology with transposed great arteries (e.g., double inlet left ventricle with transposition of the great arteries, tricuspid atresia with transposition of the great arteries) to address risk of systemic ventricular outflow tract obstruction after the Fontan palliation, as the ventricular septal defect/bulboventricular foramen may become restrictive, and the DKS anastomosis prevents consequent subaortic obstruction.

Danielson technique

A technique for tricuspid valve repair in Ebstein anomaly involving repositioning of the tricuspid valve to the true annulus by plication of the free wall of the atrialized RV, posterior tricuspid annuloplasty, and right reduction atrioplasty (Danielson GK, Maloney JD, Devloo REA. Surgical repair of Ebstein's anomaly. *Mayo Clin Proc*. 1979;54:185).

David operation

A form of valve-sparing aortic root replacement (VSRR) for the management of aortic root and ascending aortic aneurysm. In the David I procedure, described as the "re-implantation" procedure, a tailored synthetic tube graft is fixed to the left ventricular outflow tract at the subannular level, and the native aortic valve is re-implanted inside the fabric graft, thus stabilizing the size of the aortic annulus (David TE, Feindel CM. An aortic valve sparing operation for patients with aortic incompetence and aneurysm of the ascending aorta. *J Thorac Cardiovasc Surg*. 1992;103:617-621). See also [Yacoub procedure](#).

DCRV

See [double-chambered right ventricle](#).

De Vega annuloplasty

A surgical method for management of tricuspid regurgitation that involves decreasing the size of the tricuspid valve annulus by placing a circumferential suture around the tricuspid valve, with due care to avoid the atrioventricular node.

dextrocardia

Cardiac apex directed to the right relative to the cardiac base. See [cardiac position](#).

dextroposition

Rightward shift of the heart. *See* [cardiac position](#).

dextroversion

An older term for dextrocardia. *See* [cardiac position](#).

differential hypoxemia, differential cyanosis

A difference in the degree of hypoxemia or cyanosis in different extremities as a result of the site of a right-to-left shunt. The most common situation involves greater hypoxemia and cyanosis in the feet, and sometimes the left hand, compared with the right hand and head, in a patient with an Eisenmenger patent ductus arteriosus.

DiGeorge syndrome

An autosomal dominant syndrome now known to be part of microdeletion 22q11.2 or CATCH-22 syndrome. As originally described, it consisted of infantile hypocalcemia, immunodeficiency due to thymic hypoplasia, and a conotruncal cardiac abnormality. *See also* [microdeletion 22q11.2 syndrome](#).

DILV, double-inlet left ventricle

See [double inlet ventricle](#). *See also* [univentricular connection](#), [Holmes heart](#).

DIRV, double-inlet right ventricle

See [double inlet ventricle](#). *See also* [univentricular connection](#).

discordant atrioventricular connections

Anomalous connection of atria and ventricles such that the morphologic right atrium connects via a mitral valve to a morphologic left ventricle, while the morphologic left atrium connects via a tricuspid valve to a morphologic right ventricle. *Syn:* atrioventricular discordance.

discordant ventriculoarterial connections

Anomalous connection of the great arteries and ventricles such that the pulmonary trunk arises from the left ventricle and the aorta arises from the right ventricle. *Syn:* ventriculoarterial discordance.

diverticulum of Kommerell

Enlarged origin of an aberrant left subclavian artery associated with right aortic arch. Its diameter may be equal to that of the descending aorta and tapers to the left subclavian diameter. It is found at the origin of the aberrant left subclavian artery, the fourth branch off the right aortic arch.

double aortic arch

See [aortic arch anomalies](#).

double-chambered right ventricle

Separation of the right ventricle into a higher-pressure inflow chamber and a lower-pressure infundibular chamber; the separation usually being produced by hypertrophy of the septomarginal trabeculation. (Other mechanisms are possible; obstruction always occurs proximal to the infundibulum, and is often progressive.) When a ventricular septal defect is present, it usually communicates with the higher pressure right ventricular inflow chamber.

double discordance

See [congenitally corrected transposition of the great arteries](#).

double-inlet ventricle

The morphologic arrangement in which more than 50% of both atria are connected to one dominant ventricular chamber. *See also* [univentricular connection](#).

double-orifice mitral valve

The mitral valve orifice is partially or completely divided into two parts by a fibrous bridge of tissue. Both orifices enter the left ventricle. Mitral regurgitation and/or mitral stenosis may be present. Aortic coarctation and atrioventricular septal defect may be associated.

double-outlet left ventricle (DOLV)

Both the pulmonary artery and the aorta arise predominantly from the morphologic left ventricle. DOLV is rare and much less frequent than double-outlet right ventricle (DORV).

double-outlet right ventricle (DORV)

Both great arteries arise predominantly from the morphologic right ventricle; there is usually no fibrous continuity between the semilunar and the AV valves; a ventricular septal defect (VSD) is present. When the VSD is in the subaortic position without right ventricular (RV) outflow tract obstruction, the physiology simulates a simple VSD. With RV outflow tract obstruction, the physiology simulates tetralogy of Fallot. When the VSD is in the subpulmonary position (Taussig-Bing anomaly), the physiology simulates complete transposition of the great arteries with VSD. *See also* [Taussig-Bing anomaly](#).

double-switch procedure

An operation used in patients with congenitally corrected transposition of the great arteries (CCTGA) and also in patients who have had a previous Mustard or Senning atrial switch operation for complete transposition of the great arteries. It leads to anatomic correction of the ventricle to great artery relationships such that the left ventricle supports the systemic circulation. It includes an arterial switch procedure (*see* [Jatene procedure](#)) in all cases, as well as an [atrial switch procedure](#) (Mustard or Senning) in the case of CCTGA, or reversal of the previous Mustard or Senning procedure in the case of previously operated complete transposition of the great arteries. When used in relation to revision of a prior Mustard or Senning operation, it is more accurately termed a *switch-reversal* or *switch-conversion*.

doubly committed VSD

See [ventricular septal defect](#).

Down syndrome

The most common malformation caused by trisomy 21. Most patients (95%) have complete trisomy of chromosome 21; some have translocation or mosaic forms. The phenotype is diagnostic (short stature, characteristic facial appearance, intellectual impairment, brachydactyly, atlantoaxial instability, thyroid and white blood cell disorders). Congenital heart defects are frequent, with atrioventricular septal defect, ventricular septal defect, patent ductus arteriosus, and tetralogy of Fallot being the most common. Mitral valve prolapse and aortic regurgitation may be present. Patients with Down syndrome are prone to earlier and more severe pulmonary vascular disease than might otherwise be expected as a consequence of the lesions identified.

drainage

A physiologic term describing the direction of blood flow. Anomalous drainage can occur in the absence of anomalous

connection (eg, common atrium with normal connection of the pulmonary veins). *See also* [connection](#).

dural ectasia

Expansion of the dural sac in the lumbosacral area, seen on computed tomography or magnetic resonance imaging. It has been one of the criteria used to confirm the diagnosis of Marfan syndrome (Pyeritz RE, Fishman EK, Bernhardt BA, Siegelman SS. Dural ectasia is a common feature of the Marfan syndrome. *Am J Hum Genet*. 1988;43:726-732; Fattori R, et al. Importance of dural ectasia in phenotypic assessment of Marfan's syndrome. *Lancet*. 1999;354:910-913). It has assumed lesser importance in diagnosis in the 2010 iteration of the "Ghent criteria."

E

Ebstein anomaly

An anomaly of the tricuspid valve and other cardiac structures in which the basal attachments of tricuspid valve leaflets, septal > posterior > anterior, are displaced apically within the right ventricle. Apical displacement of the septal tricuspid leaflet of more than 8 mm/m² is diagnostic (the extent of apical displacement should be indexed to body surface area). Abnormal structure of all three leaflets is seen, with the anterior leaflet typically large with abnormal attachments to the right ventricular wall due to incomplete delamination. The pathologic and clinical spectrum is broad and includes not only valve abnormalities but also myocardial structural changes in both ventricles. Tricuspid regurgitation is common, tricuspid stenosis occurs occasionally, and right-to-left shunting through a patent foramen ovale or atrial septal defect is a regular but not invariable concomitant. Other congenital lesions are often associated, such as ventricular septal defect, pulmonary stenosis, and/or accessory conduction pathways. (Ebstein W. Über einen sehr seltenen Fall von Insuffizienz der Valvula tricuspidalis, bedingt durch eine angeborene hochgradige Missbildung derselben. *Arch Anat Physiol*. 1866;7:238-254.)

Ehlers-Danlos syndrome (EDS)

A group of heritable disorders of connective tissue (specifically, abnormalities of collagen). Hyperextensibility of the joints and hyperelasticity and fragility of the skin are common to all forms; patients bruise easily. Some forms of EDS are associated with postural orthostatic tachycardia syndrome (POTS) and dysautonomia.

- **Classic EDS, formerly known as EDS types I and II:** Demonstrates autosomal dominant inheritance. The cardiovascular abnormalities are generally mild, consisting of mitral and tricuspid valve prolapse. Dilation of major arteries, including the aorta, may occur. Aortic rupture is rare, described in what was formerly classified as EDS Type I, but not in type II.
- **Hypermobility EDS, formerly known as EDS Type III:** Is a condition with a broad spectrum of manifestations, for which a specific molecular genetic cause has not been defined. Cardiovascular abnormalities are very mild or nonexistent.
- **Vascular EDS, formerly known as EDS type IV:** Is an autosomal dominant condition, frequently appearing de novo. This is the "arterial" form, associated with aortic dilation and rupture of medium and large arteries spontaneously or after trauma. Rupture of internal organs may also occur. Vascular EDS is due to an abnormality of type III procollagen and comprises about 10% of cases of EDS.
- There are several other rare types of EDS.

Eisenmenger syndrome

An extreme form of pulmonary vascular obstructive disease arising as a consequence of preexisting systemic to pulmonary shunting in which pulmonary vascular resistance increases such that pulmonary pressures are at or near systemic levels and there is reversed (right to left) or bidirectional shunting at great artery, ventricular, and/or atrial levels. *See also* [Heath-Edwards classification](#).

Ellis-van Creveld syndrome

An autosomal recessive syndrome in which the most common cardiac lesions are common atrium and partial atrioventricular septal defect (including primum atrial septal defect).

endocardial cushion defect

See [atrioventricular septal defect](#). The term *endocardial cushion defect* has fallen into disuse because it infers an outdated concept of the morphogenesis of the atrioventricular septum.

endocardial fibroelastosis

Fibrosis of the endocardium and adjacent myocardium that may be present at birth or develop later in life and often causes diastolic ventricular dysfunction.

erythrocytosis

Increased red blood cell concentration secondary to chronic tissue hypoxia, as seen in cyanotic congenital heart disease and in chronic pulmonary disease. It results from a hypoxia-induced physiologic response, resulting in increased erythropoietin levels, and affects only the red cell line. It is also called secondary erythrocytosis. The term *polycythemia* is inaccurate in this context because other blood cell lines are not affected. *See also* [polycythemia vera](#). Erythrocytosis may cause hyperviscosity symptoms. *See also* [hyperviscosity](#).

eustachian valve

A remnant of the right valve of the sinus venosus guarding the entrance to the right atrium from the inferior vena cava.

extracardiac Fontan

See [Fontan procedure](#).

F

Fabry disease

An X-linked lysosomal storage disease with frequent involvement of the myocardium, presenting as a phenocopy of sarcomeric hypertrophic cardiomyopathy.

fenestration

An opening or "window" (usually small) between two structures, which may be spontaneous, traumatic, or created interventionally or surgically. The usual purpose of a therapeutic fenestration is to act as a pressure relief mechanism, for example after Fontan palliation or repair of Ebstein anomaly, in which atrial fenestration reduces pressure in the systemic atrium and increases cardiac output at the expense of an increase in cyanosis.

fibrillin

A large glycoprotein closely involved with collagen in the structure of connective tissue. Mutations in the fibrillin gene on chromosome 15 are responsible for the manifestations of Marfan syndrome. *See also* [Marfan syndrome](#).

fluorescence in situ hybridization (FISH)

An early cytogenetic technique to detect and to localize the presence or absence of specific nucleic acids sequences (DNA sequences) on a chromosome by the use of fluorescent probes that bind to those parts of the chromosome with a high degree of sequence similarity (eg, in diagnosis of microdeletion 22q11.2).

Fontan procedure (operation)

A palliative operation for patients with a univentricular circulation involving diversion of the systemic venous return to the pulmonary artery, usually without the interposition of a subpulmonary ventricle. There are many variations, all directed toward normalization of systemic oxygen saturation and elimination of volume overload of the functioning ventricle.

- **atriopulmonary Fontan:** An early modification of the original Fontan operation in which a nonvalved connection is created between the right atrium and the pulmonary artery by a variety of techniques. *Syn:* Fontan-Kreutzer procedure, Kreutzer procedure. *See also* [Kreutzer procedure](#).
- **Björk modification Fontan:** *See* [Fontan procedure/RA-RV Fontan](#).
- **classic Fontan:** Originally a valved conduit between the right atrium and the pulmonary artery (Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax*. 1971;26:240-248). Subsequently modified to a direct (nonvalved) anastomosis between the right atrium and the pulmonary artery, as described by Kreutzer.
- **extracardiac Fontan:** *See* [Fontan procedure/total cavopulmonary connection \(TCPC\)](#).
- **fenestrated Fontan:** Surgical creation of a defect in the atrial patch or baffle to provide a pressure-relief mechanism, allowing some right-to-left shunting, thus reducing pressure in the systemic venous circuit and increasing systemic blood flow. Increase in systemic hypoxemia is an undesired but inevitable consequence. *See also* [fenestration](#).
- **lateral tunnel Fontan:** Inferior vena caval flow is directed by a baffle within the right atrium into the lower portion of the divided superior vena cava or the right atrial appendage, which is connected to the pulmonary artery. The upper part of the superior vena cava is connected to the superior aspect of the pulmonary artery constituting a bidirectional cavopulmonary (Glenn) anastomosis. The majority of the right atrium is excluded from the systemic venous circuit.
- **RA-RV Fontan:** A conduit (often valved) is placed between the right atrium and the hypoplastic right ventricle. Also known as the Björk modification (Björk VO, Olin CL, Bjarke BB, Thorén CA. Right atrial-right ventricular anastomosis for correction of tricuspid atresia. *J Thorac Cardiovasc Surg*. 1979;77:452-458).
- **total cavopulmonary connection (TCPC):** Inferior vena caval blood is directed to the pulmonary artery via an extracardiac conduit. The superior vena cava is anastomosed to the pulmonary artery as in the bidirectional superior cavopulmonary (Glenn) anastomosis.

fossa ovalis

An oval depression in the lower part of the right atrial surface of the interatrial septum. It is a vestige of the foramen ovale, and its floor corresponds to the septum primum of the fetal heart. *Syn:* oval fossa.

Friedreich ataxia

An autosomal recessive inherited neurodegenerative disease, typically presenting in children and young adults, and

frequently associated with cardiomyopathy presenting as left ventricular hypertrophy.

G**Gallavardin phenomenon**

A clinical sign found in patients with aortic stenosis, in which the musical high frequency component of an aortic stenosis murmur is heard at the cardiac apex, where it may be misinterpreted as mitral regurgitation (in spite of its difference in timing). The musical component is dissociated from the harsh noisy component best heard at the upper right sternal border radiating to the neck.

Gerbode defect

An unusual variant of atrioventricular septal defect in which the defect is in the superior portion of the atrioventricular septum above the insertion of the septal leaflet of the tricuspid valve, resulting in a direct connection and shunt between the left ventricle and the right atrium. *See also* [atrioventricular septum](#).

Ghent criteria

A set of criteria for the diagnosis of Marfan syndrome. The 1996 version of the Ghent nosology required the involvement of three organ systems (two systems must have “major” involvement) or of two organ systems (one major) and a positive family history (DePaepe A, Devereux RB, Dietz HC, Hennekam RC, Pyeritz RE. Revised diagnostic criteria for the Marfan syndrome. *Am J Med Genet*. 1996;62:417-426). The Ghent criteria were revised in 2010, placing more emphasis on genetic testing. Apart from genetic testing with documentation of pathogenic fibrillin gene (FBN1) mutations, diagnosis of Marfan syndrome is based on typical eye involvement, cardiovascular involvement (ie, aortic root dilatation), and typical skeletal features (standardized as *systemic score*). (Loeys BL, Dietz HC, Braverman AC, et al. The revised Ghent nosology for the Marfan syndrome. *J Med Genet*. 2010;47(7):476-485.)

Glenn anastomosis

Syn: Glenn operation (often called “Glenn shunt”), superior cavopulmonary anastomosis. A palliative operation for the purpose of increasing pulmonary blood flow, and hence systemic oxygen saturation, in which a direct anastomosis is created between the superior vena cava (SVC) and a pulmonary artery. This procedure does not cause systemic (subaortic) ventricular volume overload.

- **bidirectional Glenn:** End-to-side anastomosis of the divided SVC to the undivided pulmonary artery (Haller JA Jr, Adkins JC, Worthington M, Rauenhorst J. Experimental studies in permanent bypass of the right heart. *Surgery*. 1966;59:1128-1132; Azzolina G, Eufrate S, Pensa P. Tricuspid atresia: experience in surgical management with a modified cavopulmonary anastomosis. *Thorax*. 1972;27:111-115; Hopkins RA, Armstrong BE, Serwer GA, Peterson RJ, Oldham Jr HN. Physiologic rationale for a bidirectional cavopulmonary shunt: a versatile complement to the Fontan principle. *J Thorac Cardiovasc Surg*. 1985;90:391-398). *Syn:* bidirectional superior cavo-pulmonary anastomosis.
- **classic Glenn:** Anastomosis of the SVC to the distal end of the divided right pulmonary artery with division/ligation of the SVC below the anastomosis. Acquired pulmonary arteriovenous malformations with associated systemic arterial desaturation are a common long-term complication. (Glenn WW. Circulatory bypass of the right side of the heart. IV.

Shunt between superior vena cava and distal right pulmonary artery: report of clinical application. *N Engl J Med.* 1958;259:117-120.)

goose-neck deformity

Describes the angiographic appearance of the distorted, elongated, and narrowed left ventricular outflow tract, and ascending aorta during diastole in patients with AV septal defect.

Gore-Tex

A synthetic material often used to fashion conduits and other prosthetic devices for the surgical palliation or repair of congenital heart disease.

GUCH

Grown-up congenital heart disease. A term originated by Dr. Jane Somerville, a famous cardiologist, one of the founders of the discipline of ACHD/GUCH. *Syn:* adult congenital heart disease (ACHD).

GUCH Working Group 22

Working Group for Grown-Up Congenital Heart Disease of the European Society of Cardiology (Working Group 22) <http://www.esccardio.org/Working-groups/Working-Group-on-Grown-Up-Congenital-Heart-Disease>

H

hammock effect

A form of malfunction of a percutaneously implanted pulmonary valve prosthesis (described for the Melody valve) caused by insufficient alignment of the stent to the conduit in which it has been implanted. Usually causes prosthetic pulmonary valve stenosis.

hammock mitral valve

A rare congenital mitral valve anomaly characterized by fibrous continuity between subvalvar mitral apparatus and mitral valve leaflets, typically causing mitral regurgitation. It is also known as an anomalous mitral arcade. *See also* [mitral arcade](#).

Heath-Edwards classification

A legacy histopathologic classification useful in assessing the potential for reversibility of pulmonary vascular disease (Heath D, Edwards JE. The pathology of hypertensive pulmonary vascular disease: a description of six grades of structural changes in the pulmonary arteries with special reference to congenital cardiac septal defects. *Circulation.* 1958;18:533-547).

- **Grade I:** Hypertrophy of the media of small muscular arteries and arterioles
- **Grade II:** Intimal cellular proliferation in addition to medial hypertrophy
- **Grade III:** Advanced medial thickening with hypertrophy and hyperplasia including progressive intimal proliferation and concentric fibrosis. This results in obliteration of arterioles and small arteries.
- **Grade IV:** “Plexiform lesions” of the muscular pulmonary arteries and arterioles with a plexiform network of capillary-like channels within a dilated segment
- **Grade V:** Complex plexiform, angiomatous and cavernous lesions, and hyalinization of intimal fibrosis
- **Grade VI:** Necrotizing arteritis

hemi-Fontan

A modification of the bidirectional cavopulmonary anastomosis utilizing the right atrium, believed to improve flow dynamics from the superior vena cava into the pulmonary arteries and also to simplify the additional surgery required to “complete” a Fontan procedure at a later time. A side-to-side anastomosis is created between the SVC-RA junction and the PA; venous flow from SVC into RA is occluded by a homograft dam. *See also* [Glenn anastomosis/bidirectional Glenn](#).

hemi-truncus

An old term describing an anomalous pulmonary artery branch to one lung arising from the ascending aorta in the presence of a main pulmonary artery arising normally from the right ventricle and supplying the other lung. The preferred terminology is aberrant pulmonary artery arising from the aorta, because it is morphogenetically distinct from “truncus arteriosus.” Individual aortic and pulmonary valves are present and there is no truncal valve. *See also* [truncus arteriosus](#).

hepatic factor

A yet to be defined humoral factor contained in hepatic venous blood that is essential for normal development and function of pulmonary blood vessels. Patients in whom hepatic venous blood is absent from pulmonary perfusion (eg, classic Glenn shunt, Kawashima procedure) are at high risk of developing pulmonary arteriovenous fistulae that may be reverted by directing hepatic venous blood through the lungs.

heterograft

Transplanted tissue or organ from a different species.

heterotaxy

Abnormal arrangement (Gk. *taxo*) of viscera that differs from the arrangement seen in either situs solitus or situs inversus. It is often described as “visceral heterotaxy.”

heterotopic

Located in an anatomically abnormal site, often in reference to transplantation of an organ.

HLHS

See [hypoplastic left heart syndrome](#).

Holmes heart

In 1824, Andrew F. Holmes, later the first Dean of the Medical Faculty of McGill University, published autopsy findings of a 21-year-old man who had died with chronic cyanosis and congestive heart failure. This was the first documented case of single ventricle. There was absence of the sinus (body or inflow tract) of the morphologically right ventricle (RV), hence a single (unpaired) morphologically left ventricle (LV), double-inlet LV, infundibular outlet chamber (IOC), and normally related great arteries, with the pulmonary artery arising from the IOC and the aorta from the single LV. In view of its rarity, William Osler urged Maude Abbott to republish this case, which she did in 1901, catalyzing her career. Maude Abbott then proceeded to become the world authority on congenital heart disease until her death in 1940 (Dobell AR, van Praagh R. The Holmes heart: historic associations and pathologic anatomy. *Am Heart J.* 1996;132:437-445).

Holt-Oram syndrome

Autosomal dominant syndrome consisting of radial abnormalities of the forearm and hand associated with secundum atrial septal defect (most common), ventricular septal defect, or, rarely, other cardiac malformations (Holt M, Oram S. Familial heart disease with skeletal manifestations. *Br Heart J*. 1960;22:236-242). The gene for this syndrome is on chromosome 12q2 (Basson CT, Bachinsky DR, Lin RC, et al. Mutations in human TBX5 cause limb and cardiac malformation in Holt-Oram syndrome. *Nat Genet*. 1997;15:30-35).

homograft

Transplanted tissue or organ from another individual of the same species.

Hunter syndrome

A genetic syndrome due to a deficiency of the enzyme iduronate sulfate (mucopolysaccharidase) with X-linked recessive inheritance. Clinical spectrum is wide. Patients present with skeletal changes, intellectual impairment, arterial hypertension, and involvement of atrioventricular and semilunar valves resulting in valvular regurgitation.

Hurler syndrome

A genetic syndrome due to a deficiency of the enzyme α -L-iduronidase (mucopolysaccharidase) with autosomal recessive inheritance. Phenotype presents with a wide spectrum including severe skeletal abnormalities, corneal clouding, hepatosplenomegaly, intellectual impairment, and mitral valve stenosis.

hyperviscosity

An excessive increase in viscosity of blood as may occur secondary to erythrocytosis in patients with cyanotic congenital heart disease. Hyperviscosity symptoms include headache; impaired alertness, depressed mentation, or a sense of distance; visual disturbances (blurred vision, double vision, amaurosis fugax); paresthesias of fingers, toes, or lips; tinnitus; fatigue, lassitude; myalgias (including chest, abdominal muscles); and muscle weakness. (Perloff JK, Rosove MH, Child JS, Wright GB. Adults with cyanotic congenital heart disease: hematologic management. *Ann Intern Med*. 1988;109:406-413.) Restless legs or a sensation of cold legs may reflect hyperviscosity (observation of Dr. E. Oechslin). Because the symptoms are nonspecific, their relation to hyperviscosity is supported if they are alleviated by phlebotomy. Iron deficiency and dehydration worsen hyperviscosity and must be avoided—or treated if present.

hypoplastic left heart syndrome (HLHS)

A heterogeneous syndrome with a wide variety and severity of manifestations involving hypoplasia, stenosis, or atresia at different levels of the left heart including the aorta, aortic valve, left ventricular outflow tract, left ventricular body, mitral valve, and left atrium. See also [Norwood procedure](#), [Sano modification](#).

I**ILAA**

Isomerism of the left atrial appendage. See also [heterotaxy](#), [isomerism](#).

IRAA

Isomerism of the right atrial appendage. See also [heterotaxy](#), [isomerism](#).

Ilbawi procedure (operation)

An operation for congenitally corrected transposition of the great arteries with ventricular septal defect (VSD) and pulmonary stenosis in which continuity is established between the left ventricle and the aorta via the VSD using a baffle within the right ventricle. The right ventricle is connected to the pulmonary artery using a valved conduit. An atrial switch procedure is done. The left ventricle then supports the systemic circulation (Ilbawi MN, DeLeon SY, Backer CL. An alternative approach to the surgical management of physiologically corrected transposition with ventricular septal defect and pulmonary stenosis or atresia. *J Thorac Cardiovasc Surg*. 1990;100:410-415).

infracristal

Located below the crista supraventricularis in the right ventricular outflow tract. See also [crista supraventricularis](#).

infundibular, infundibulum

(Pertaining to) a ventricular–great arterial connecting segment that normally is subpulmonary but can be subaortic and may be bilateral or absent. Bilateral infundibulum may be seen in patients with transposition of the great arteries with ventricular septal defect (VSD) and pulmonary stenosis (PS), double-outlet right ventricle with VSD and PS, and anatomically corrected malposition. *Syn*: conus.

inlet VSD

See [ventricular septal defect](#).

interrupted aortic arch

See [aortic arch anomaly](#).

interrupted inferior vena cava

The inferior vena cava is interrupted or absent below the hepatic veins with ongoing systemic venous drainage via the azygos vein to the superior vena cava. The hepatic veins enter the right atrium directly. This anomaly is frequently associated with complex congenital heart disease, particularly left isomerism.

intraatrial reentrant tachycardia (IART)

A term describing macroreentrant atrial arrhythmias, mainly atypical atrial flutter, commonly seen in CHD, in which reentrant circuits around (surgical) scars are often responsible for the arrhythmia. The atrial rate tends to be slower than that seen in typical atrial flutter.

ISACHD

International Society for Adult Congenital Heart Disease (formerly ISACCD, International Society for Adult Congenital Cardiac Disease; <http://www.isachd.org>).

isolation of arch vessels

See [aortic arch anomalies](#).

isomerism

Paired, mirror-image sets of normally single or nonidentical organ systems (atria, lungs, and viscera), often associated with other abnormalities.

- **left isomerism:** *Syn*: polysplenia syndrome. A congenital syndrome consisting of paired, morphologically left structures: multiple bilateral spleens, bilateral left bronchi, bilateral bilobed (left) lungs, midline liver, two morphologic left atria, multiple anomalies of systemic venous

connections, and other complex cardiac and noncardiac malformations.

- **right isomerism:** *Syn:* asplenia syndrome, Ivemark syndrome. Congenital syndrome consisting of paired morphologically right structures: absence of spleen, bilateral right bronchi, bilateral trilobed (right) lungs, two morphologic right atria, multiple anomalies of pulmonary venous connections, and other complex cardiac and noncardiac anomalies.

J

Jacobsen syndrome

A genetic disorder, usually a spontaneous mutation, characterized by the deletion of several genes on chromosome 11q23. Affected individuals present with developmental disorder affecting speech and motor skills and cognitive impairment. Many patients have attention deficit hyperactivity disorder (ADHD), distinctive facial features, and bleeding disorders. Congenital heart defects include mainly left-sided obstructive lesions. An affected individual can inherit the deletion from an unaffected parent with a balanced translocation.

Jatene procedure (operation)

Syn: arterial switch operation (ASO). An operation for complete transposition of the great arteries, involving removal of the aorta from its attachment to the right ventricle, removal of the pulmonary artery from the left ventricle, and reattachment of the great arteries to the contralateral ventricles, with re-implantation of the coronary arteries into the neo-aorta. As a consequence, the left ventricle supports the systemic circulation (Jatene AD, Fontes VF, Souza LC, Paulista PP, Neto CA, Sousa JE. Anatomic correction of transposition of the great vessels. *J Thorac Cardiovasc Surg.* 1976;72:364-370). See also [LeCompte maneuver](#).

juxtaposition of atrial appendages

A rare anomaly seen in patients with transposition of the great arteries and other complex congenital heart defects in which the atrial appendages are situated side by side. The right atrial appendage passes immediately behind the transposed main pulmonary artery in patients with leftward juxtaposition of atrial appendages. In the presence of juxtaposition of the atrial appendages, cannulation for heart-lung-bypass, an atrial switch operation or an atriopulmonary Fontan operation (no longer performed) is more challenging.

K

Kartagener syndrome

Autosomal recessive syndrome consisting of situs inversus totalis, dextrocardia, and a defect of ciliary motility called primary ciliary dyskinesia (PCD) leading to sinusitis, bronchiectasis, and sperm immobility. PCD can occur in absence of situs inversus, in which case it is not called Kartagener syndrome (Kartagener M. Zur Pathogenese der Bronchiektasien: Bronchiektasien bei Situs viscerum inversus. *Beitr Klink Tuberkul.* 1933;28:231-234; Kartagener M, Stucki P. Bronchiectasis with situs inversus. *Arch Pediatr.* 1962;79:193-196; Miller RD, Divertie MB. Kartagener's syndrome. *Chest.* 1972;62:130-136).

Kawasaki disease

An acute febrile, systemic disease characterized by vascular inflammation of small and medium-large arteries that affects especially small children and imitates in its initial manifestations infectious diseases such as measles or scarlet fever. Cardiac

complications are common. In particular, the coronary arteries can be affected leading to coronary artery aneurysms and late coronary stenoses that may persist into adult life (Kawasaki T, Kosaki F, Okawa S, Shigematsu I, Yanagawa H. A new infantile acute febrile mucocutaneous lymph node syndrome [MLNS] prevailing in Japan. *Pediatrics.* 1974;54:271-276).

Kawashima procedure

A modification of the Fontan operation used in patients with inferior vena caval interruption, typically found in patients with left atrial isomerism. In such patients, drainage of systemic venous blood from the lower body occurs via azygos or hemiazygos veins to the superior vena cava (left, right, or bilateral). As a consequence, a superior bidirectional cavopulmonary anastomosis diverts most of the systemic venous blood into the lungs. In the setting of bilateral superior vena cava, bilateral bidirectional cavopulmonary anastomoses are constructed. In patients with left atrial isomerism, hepatic veins typically connect directly to the atrium; thus hepatic venous blood is not diverted to the lungs, predisposing to the formation of pulmonary arteriovenous malformations (see also [hepatic factor](#)). To reduce the likelihood of the latter complication, most patients require completion of the Fontan circulation by diversion of the hepatic venous blood to the pulmonary artery via an extracardiac conduit. (Kawashima Y, Kitamura S, Matsuda H, Shimazaki Y, Nakano S, Hirose H. Total cavopulmonary shunt operation in complex cardiac anomalies. A new operation. *J Thorac Cardiovasc Surg.* 1984;87[1]:74-81.) Contrast to Kawashima repair. See also [Kawashima repair](#).

Kawashima repair

An operation for repair of double-outlet right ventricle with subpulmonary ventricular septal defect (Taussig-Bing anomaly), consisting of an intraventricular diversion using a pericardial patch to direct left ventricular output through the ventricular septal defect to the aorta. This procedure is to be distinguished from a different Kawashima procedure, which is a modified bidirectional cavopulmonary anastomosis performed in patients with interrupted inferior vena cava and azygos continuation. See also [Kawashima procedure](#).

Kay plication

A surgical method for management of tricuspid regurgitation that involves plication of the posterior leaflet, thus converting the tricuspid valve into a functionally bicuspid valve (Kay GL, Morita S, Mendez M, Zubiato P, Kay JH. Tricuspid regurgitation associated with mitral valve disease: repair and replacement. *Ann Thorac Surg.* 1989;48:S93-S95).

Konno procedure (operation)

Repair of tunnel-like subvalvular left ventricular outflow tract obstruction (LVOTO) by aortoventriculoplasty. The operation involves enlargement of the left ventricular outflow tract by inserting a patch in the ventricular septum, as well as aortic valve replacement and enlargement of the aortic annulus and ascending aorta (Konno S, Imai Y, Iida Y, Nakajima M, Tatsuno K. A new method for prosthetic valve replacement in congenital aortic stenosis associated with hypoplasia of the aortic valve ring. *J Thorac Cardiovasc Surg.* 1975;70:909-917). Modification of the original technique with preservation of the aortic valve is described as the "modified Konno procedure." In severe forms of LVOTO not amenable to a Konno procedure, a prosthetic valve-containing conduit may be inserted between the left ventricular apex and descending aorta (DiDonato RM, Danielson GK, McGoon DC, Driscoll DJ, Julsrud PR, Edwards WD. Left

ventricular–aortic conduits in paediatric patients. *J Thorac Cardiovasc Surg.* 1984;88:82-91; Frommelt PC, Rocchini AP, Bove EL. Natural history of apical left ventricular to aortic conduits in paediatric patients. *Circulation.* 1991;84[suppl III]:213-218).

Kreutzer procedure

An operation performed for the management of tricuspid atresia, in which, in contrast to the classic Fontan procedure, an atriopulmonary anastomosis is performed without interposition of a valve between the inferior vena cava and the right atrium. Kreutzer used a homograft for the RAA-PA connection in his first patient; in subsequent early patients he performed a direct atriopulmonary anastomosis by removing the patient's own pulmonary valve and PA from the outflow tract of the right ventricle and attaching it to the RAA. However, the concept of a valve in the RA-PA connection was quickly abandoned by Kreutzer as well as by Fontan. (Kreutzer G, Galíndez E, Bono H, De Palma C, Laura JP. An operation for the correction of tricuspid atresia. *J Thorac Cardiovasc Surg.* 1973;66:613-621). See also [Fontan procedure/atriopulmonary Fontan](#).

• **Fontan-Kreutzer procedure:** Direct atriopulmonary connection for single ventricle circulation. See also [Fontan procedure/atriopulmonary Fontan](#).

L

LeCompte maneuver

The pulmonary artery is brought anterior to the aorta during an arterial switch procedure in patients with complete transposition of the great arteries. See also [Jatene procedure](#).

LEOPARD syndrome

An autosomal dominant condition including lentigines, electrocardiographic abnormalities, ocular hypertelorism, pulmonary stenosis, abnormal genitalia, retardation of growth, and deafness. Rarely, cardiomyopathy, or complex congenital heart disease may be present.

levocardia

Leftward-oriented cardiac apex (normal). See [cardiac position](#).

levoposition

Leftward shift of the heart. See [cardiac position](#).

ligamentum arteriosum

A normal fibrous structure that is the residuum of the ductus arteriosus after its spontaneous closure.

Loeys-Dietz syndrome

An autosomal dominant aneurysm syndrome characterized by the triad of arterial tortuosity and aneurysms, hypertelorism, and bifid uvula or cleft palate (Loeys BL, Chen J, Neptune ER. A syndrome of altered cardiovascular, craniofacial, neurocognitive and skeletal development caused by mutations in *TGFBR1*. *Nat Genet.* 2005;37:275-281). It is caused by mutations in the transforming growth factor- β receptor gene (Loeys BL, Schwarze U, Holm T. Aneurysm syndromes caused by mutations in the TGF-beta receptor. *N Engl J Med.* 2006;355:788-798).

long QT syndrome

Abnormal prolongation of QT duration imparting risk of torsades de pointes, syncope, and sudden cardiac death. It may be congenital or acquired (from medications such as antiarrhythmics, antihistamines, some antibiotics; electrolyte disturbances

such as hypocalcemia, hypomagnesemia, hypokalemia; hypothyroidism; and other factors). Normal QT interval is inversely related to heart rate.

looping

Bending of the primitive heart tube (normally to the right, dextro, *d-*), which determines the atrioventricular relationship.

- ***d*-loop:** Morphologic right ventricle lies to the right of the morphologic left ventricle (normal rightward bend).
- ***l*-loop:** Morphologic right ventricle lies to the left of the morphologic left ventricle (leftward bend).

Lutembacher syndrome

Atrial septal defect associated with mitral valve stenosis. The mitral valve stenosis is usually acquired (rheumatic).

LVOTO

Left ventricular outflow tract obstruction.

M

maladie de Roger

Eponymous designation for a small restrictive ventricular septal defect (VSD) that is not associated with significant left ventricular volume overload or raised pulmonary artery pressure. There is a loud VSD murmur due to the high-velocity turbulent left-to-right shunt across the defect.

malposition

An abnormality of cardiac position. See [cardiac position](#).

MAPCA

Major aortopulmonary collateral artery. A large abnormal arterial vessel arising from the aorta that connects to a pulmonary artery (usually in the pulmonary hilum) and provides blood supply to a portion of the lungs. It is found in complex pulmonary atresia and other complex congenital heart disorders associated with a severe reduction or absence of antegrade pulmonary blood flow from the ventricle(s).

Marfan syndrome

A connective tissue disorder with autosomal dominant inheritance caused by a defect in the fibrillin gene on chromosome 15. The phenotypic expression varies. Patients may have tall stature, abnormal body proportions, ocular abnormalities, dural ectasia, and protrusio acetabulae, and present with skeletal and cardiovascular abnormalities. Mitral valve prolapse with mitral regurgitation, ascending aortic dilation or aneurysm with subsequent aortic regurgitation, and aortic dissection are the most common cardiovascular abnormalities. See also [Ghent criteria](#).

McGoon ratio

Angiographic index to determine if the branch pulmonary arteries are large enough to permit surgical repair in patients with tetralogy of Fallot/pulmonary atresia. The McGoon ratio is the ratio of the combined diameter of the right and left pulmonary artery at the prebranching point divided by the diameter of the aorta at the level of the diaphragm. A ratio greater than 2 is normal. A ratio greater than 1.2 is associated with an acceptable postoperative right ventricular pressure; a ratio less than 0.8 is deemed inadequate for surgical repair. This ratio tends to overestimate the adequacy of the size of pulmonary arteries because the diameter of the descending thoracic aorta at the level of diaphragm is frequently smaller

in patients with pulmonary atresia/tetralogy of Fallot. *See also* [Nakata index](#).

Melbourne shunt

An operation involving the creation of an end-to-side anastomosis between a hypoplastic pulmonary artery and the ascending aorta to increase pulmonary blood flow and to promote growth of the central pulmonary artery (eg, in a patient with pulmonary atresia, ventricular septal defect, and major systemic-to-pulmonary collateral arteries with a diminutive central pulmonary artery). The Melbourne shunt was developed by Dr. Roger Mee's group from Melbourne, Australia. (Watterson KG, Wilkinson JL, Karl TR, Mee RB. Very small pulmonary arteries: central end-to-side shunt. *Ann Thorac Surg*. 1991;52:1132-1137.)

mesocardia

Cardiac apex directed to midchest. *See* [cardiac position](#).

mesoposition

Shift of the heart toward the midline. *See* [cardiac position](#).

Metras modification

An operative technique to address transposition of the great arteries with ventricular septal defect (VSD) and left ventricular outflow tract obstruction. In the Metras modification, in contrast to the Rastelli operation, the right ventricle-pulmonary artery connection is maintained with a segment of tubular aortic autograft that allows for growth of the RV-PA conduit, reducing or delaying the need for reoperation. (Metras D, et al. Modified Rastelli using an autograft: a new concept for correction of transposition of the great arteries with ventricular septal defect and left ventricular outflow tract obstruction [with an extension to other congenital heart defects]. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann*. 2000;3:117-124). *See also* [Rastelli procedure](#).

microdeletion 22q11.2 syndrome

Syndrome due to a microdeletion at chromosome 22q11.2 resulting in a wide clinical spectrum. Also called CATCH-22 syndrome, diGeorge syndrome. CATCH stands for cardiac defect, abnormal facies, thymic hypoplasia, cleft palate, and hypocalcemia. Cardiac defects include conotruncal defects such as interrupted aortic arch, tetralogy of Fallot, truncus arteriosus, and double-outlet right ventricle. *See also* [diGeorge syndrome](#), [velocardiofacial syndrome](#).

mitral arcade

Chordae of the mitral valve are shortened or absent, and the thickened mitral valve leaflets insert directly into the papillary muscle ("hammock valve"). Mitral valve excursion is limited and results in mitral stenosis. *See also* [hammock mitral valve](#).

mitral valve prolapse

Systolic billowing of one or both mitral valve leaflets into the left atrium superior to the annular plane with or without associated mitral regurgitation. Echocardiographic definition: dislocation greater than 2 mm into the left atrium of at least one of the mitral valve leaflets during systole and a thickening of 5 mm or more of the prolapsing valve leaflet during diastole.

moderator band

A prominent muscular structure traversing the right ventricle from the base of the anterior papillary muscle to the septum near the apex.

Moenchkeberg sling

A sling of conduction tissue in the setting of right atrial isomerism with dual AV-nodes. The presence of a Moenchkeberg sling predisposes to reentrant arrhythmias.

muscular VSD

See [ventricular septal defect](#).

Mustard procedure (operation)

An operation for complete transposition of the great arteries in which venous return is directed to the contralateral ventricle by means of an atrial baffle made from autologous pericardial tissue or (rarely) synthetic material, after resection of most of the atrial septum. As a consequence the right ventricle supports the systemic circulation. This is a type of "atrial switch" operation. *See also* [Senning procedure](#), [atrial switch procedure](#); [double-switch procedure](#). (Mustard WT. Successful two-stage correction of transposition of the great vessels. *Surgery*. 1964;55:469-472).

N

Nakata index

Angiographic index to determine if the branch pulmonary arteries are large enough to permit surgical repair in patients with tetralogy of Fallot/complex pulmonary atresia. The Nakata index is the sum of the combined cross-sectional area of the branch pulmonary arteries indexed to body surface area (normal value: $330 \pm 30 \text{ mm}^2/\text{m}^2$; a Nakata index $< 150 \text{ mm}^2/\text{m}^2$ is considered diminutive [Nakata S, Imai Y, Takanashi Y. A new method for the quantitative standardization of cross-sectional areas of the pulmonary arteries in congenital heart diseases with decreased pulmonary blood flow. *J Thorac Cardiovasc Surg*. 1984;88:610-619]). *See also* [McGoan ratio](#).

national referral center

See [supraregional referral center \(SRRC\)](#).

neoaortic valve/neopulmonary valve

In patients undergoing an arterial switch operation (ASO, Jatene procedure), the pulmonary root and aortic root are detached from their native sites and reattached to the opposite valve; thus the pulmonary valve becomes the neo-aortic valve, and the aortic valve becomes the neopulmonary valve. *See* [Jatene procedure](#).

Nikaidoh procedure

Surgical repair of double-outlet right ventricle or transposition of the great arteries with pulmonary stenosis, consisting of aortic translocation with reconstruction of the right ventricular outflow tract. The aortic root, with valve and coronary arteries attached, is removed from the right ventricular outflow tract and translocated posteriorly to lie primarily over the left ventricle. The ventricular septal defect is repaired with a patch that is attached to the aortic root at its superior margin. The pulmonary artery is attached to the right ventriculotomy with another pericardial patch.

nonrestrictive VSD

See [ventricular septal defect](#).

Noonan syndrome

An autosomal dominant syndrome phenotypically somewhat similar to Turner syndrome, with a normal chromosomal complement, due in some instances to an abnormality in chromosome 12q. It is associated with congenital cardiac anomalies,

especially dysplastic pulmonic valve stenosis, pulmonary artery stenosis, atrial septal defect, tetralogy of Fallot, and hypertrophic cardiomyopathy. Congenital lymphedema is a common associated anomaly that may be unrecognized (Noonan JA, Ehmke DA. Associated noncardiac malformations in children with congenital heart disease. *Midwest Soc Pediatr Res.* 1963;63:468).

Norwood procedure

The first operation in a multistage strategy for management of hypoplastic left heart syndrome. In stage 1 (Norwood), a systemic to pulmonary arterial shunt (generally a modified Blalock-Taussig-Thomas anastomosis) is created to maintain pulmonary blood flow and the main pulmonary artery is disconnected from the lungs and anastomosed to a reconstructed aorta to provide systemic blood flow. By this strategy, the right ventricle becomes the subaortic ventricle supporting the systemic circulation. This procedure is followed by a second stage some months later when a bidirectional cavopulmonary anastomosis is created to increase pulmonary blood flow and decrease volume loading of the subaortic right ventricle. The Blalock-Taussig-Thomas shunt is closed. The third stage is the completion of a Fontan-type operation. *See also* Fontan procedure, Sano modification.

Norwood-Rastelli procedure

An operation for transposed great arteries associated with a ventricular septal defect (VSD), left ventricular outflow tract obstruction, and hypoplastic ascending aorta and arch, in which the proximal main pulmonary artery is anastomosed to and augments the ascending aorta and arch (Norwood), while the left ventricle is baffled to the pulmonary, now neo-aortic, valve via the VSD and a right ventricle (RV) to distal pulmonary artery (PA) conduit is constructed to restore RV-PA continuity (Rastelli).

O

orthotopic

Located in an anatomically normal recipient site, often in reference to transplantation of an organ.

ostium primum ASD

See atrial septal defect.

outlet VSD

See ventricular septal defect.

oval fossa

See fossa ovalis.

over-and-under ventricles

See superior-inferior heart.

overriding valve

An atrioventricular valve that empties into both ventricles, or a semilunar valve that originates from both ventricles.

P

PAIVS, PA-IVS

Pulmonary atresia with intact ventricular septum.

palliation, palliative operation

A procedure carried out for the purpose of relieving symptoms or ameliorating some of the adverse effects of an anomaly that

does not address the fundamental anatomic or physiologic disturbance. Contrasts to “repair” or “reparative operation.”

PAPVC

Partial anomalous pulmonary venous connection. *See* anomalous pulmonary venous connection.

PAPVD

Partial anomalous pulmonary venous drainage. *See* anomalous pulmonary venous connection. *See also* drainage.

parachute mitral valve

A mitral valve abnormality in which all chordae tendineae of the mitral valve, which may be shortened and thickened, insert into a single, abnormal, papillary muscle, usually causing mitral stenosis. The parachute mitral valve may be part of the Shone complex. *See also* Shone complex.

partial AV septal defect

See atrioventricular septal defect.

patent ductus arteriosus (PDA)

An arterial duct that fails to undergo normal closure in the early postnatal period. *Syn:* persistently patent ductus arteriosus, persistent arterial duct.

patent foramen ovale (PFO)

Failure of anatomic fusion of the valve of the foramen ovale with the limbus of the fossa ovalis that normally occurs when left atrial pressure exceeds right atrial pressure after birth. There is no structural deficiency of tissue of the atrial septum. The foramen is functionally closed as long as left atrial pressure exceeds right atrial pressure, but can reopen if right atrial pressure rises. In pathologic studies, patent foramen ovale is found in up to 35% of the adult population. The lower and variable prevalence reported in clinical series depends on the techniques used to find it. Patent foramen ovale is not classified as an atrial septal defect or as a heart condition; it is a frequently found normal variant. *Syn:* probe-patent foramen ovale (PFO).

pectus carinatum

Malformation of the chest wall with anterior protrusion of the sternum. One of the classic skeletal features in patients with Marfan syndrome. Often asymmetric.

pectus excavatum

Malformation of the chest wall consisting of posterior indentation of the sternum. One of the classic skeletal features in patients with Marfan syndrome, though seen commonly in other conditions or as an isolated finding.

pentalogy of Fallot

Tetralogy of Fallot with an atrial septal defect or patent foramen ovale. *See* tetralogy of Fallot.

perimembranous ventricular septal defect

See ventricular septal defect.

persistent left superior vena cava (LSVC)

Persistence of the left anterior cardinal vein (which normally obliterates during embryogenesis) results in persistent LSVC. LSVC connects via the coronary sinus to the right atrium in more than 90% of patients; rarely, it may directly connect to the

left atrium in association with other congenital heart defects (eg, isomerism). Its prevalence is up to 0.5% in the general population and higher in patients with congenital heart disease.

PFO

See [patent foramen ovale](#).

phlebotomy

A palliative procedure involving withdrawal of whole blood (usually in up to 500-mL increments) that may be offered to patients with cyanotic congenital heart disease and secondary erythrocytosis who are experiencing hyperviscosity symptoms. Concomitant volume replacement is usually indicated.

pink tetralogy of Fallot

See [tetralogy of Fallot](#).

plastic bronchitis

A rare complication following Fontan surgery characterized by the development of bronchial casts consisting of fibrinous acellular infiltrate that obstruct the tracheobronchial tree. These casts may be life-threatening because of resulting airway obstruction and asphyxiation. Plastic bronchitis is associated with high central venous pressures and multiple abnormal lymphatic collateral vessels within lung parenchyma.

polycythemia vera

A neoplastic transformation of all blood cell lines (erythrocyte, leukocyte, and platelet) associated with increased numbers of cells in the peripheral blood. Contrast to secondary erythrocytosis, as seen in cyanotic heart disease. See also [erythrocytosis](#).

polysplenia syndrome

See [isomerism/left isomerism](#).

Potts anastomosis

A palliative operation for the purpose of increasing pulmonary blood flow and, hence, systemic oxygen saturation. The procedure involves creating a small communication between a pulmonary artery and the ipsilateral descending thoracic aorta. It is often complicated by the development of pulmonary vascular obstructive disease if too large or by acquired stenosis or atresia of the pulmonary artery if distortion occurs. It is sometimes called “Potts shunt” (Potts WJ, Smith S, Gibson S. Anastomosis of the aorta to pulmonary artery in certain types of congenital heart disease. *J Am Med Assoc*. 1946;132:627-631).

PPH

Primary pulmonary hypertension, an obsolete term, now reclassified as idiopathic pulmonary arterial hypertension. See [pulmonary arterial hypertension](#).

probe-patent foramen ovale

See [patent foramen ovale](#).

protein-losing enteropathy (PLE)

A complication that may be seen after the Fontan operation (as well as in other conditions) in which protein is lost via the gut, resulting in ascites, peripheral edema, pleural and pericardial effusions, and chronic diarrhea. It is of unknown cause, although exacerbated by high systemic venous pressure. If serum protein and albumin levels are low, increased α 1-antitrypsin in the stool supports the diagnosis.

protrusio acetabulae

Abnormal medial displacement of the head of the femur within the acetabulum. This is a radiologic finding useful in the diagnosis of Marfan syndrome. See also [Ghent criteria](#).

pseudotruncus arteriosus

An old term used to describe pulmonary atresia with a ventricular septal defect and pulmonary blood flow provided by systemic to pulmonary collaterals (MAPCAs). This anatomic arrangement had previously been called “truncus arteriosus type IV” but is morphogenetically a separate lesion from truncus arteriosus. In pseudotruncus, the single vessel arising from the ventricles is an aorta with an aortic valve, not a truncus with a truncal valve, and pulmonary blood flow derives from aortopulmonary collateral arteries, not from anomalously connected true pulmonary arteries. *Syn*: tetralogy of Fallot with pulmonary atresia and MAPCAs.

pulmonary arterial hypertension (PAH)

Abnormally elevated mean pulmonary arterial pressure ≥ 25 mm Hg at rest as assessed by right heart catheterization. The clinical significance of a mean pulmonary arterial pressure between 21 and 24 mm Hg is not known, and the term borderline pulmonary hypertension is not used anymore. Hemodynamic definitions include precapillary pulmonary hypertension (pulmonary arterial wedge pressure [PAWP] ≤ 15 mm Hg) and postcapillary pulmonary hypertension (PAWP > 15 mm Hg). (Galie N, et al. ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Heart J*. 2016;37:67-119; and Hoeper MM, et al. *J Am Coll Cardiol*. 2013;62:D42-50).

pulmonary arteriovenous malformation (PAVM)

Defect of the pulmonary circulation consisting of direct connection between arterioles and venules without intervening capillaries. These defects are believed to arise during fetal development or very soon after birth and may enlarge under certain circumstances (eg, in a lung supplied by a classic Glenn anastomosis). When large enough, they may cause central cyanosis.

pulmonary artery band

Surgically created stenosis of the main pulmonary artery performed as a palliative procedure to protect the lungs against high blood flow and pressure when definitive correction of an underlying anomaly is not immediately possible/advisable (eg, in the setting of a nonrestrictive ventricular septal defect).

pulmonary artery sling

Anomalous origin of the left pulmonary artery from the right pulmonary artery such that it loops around the trachea. It may be associated with complete cartilaginous rings in the distal trachea and tracheal stenosis. It may occur as an isolated entity or in association with other congenital heart defects.

pulmonary atresia

An imperforate pulmonary valve. When associated with a ventricular septal defect (PA + VSD; variant of tetralogy of Fallot), pulmonary blood flow arises from aortopulmonary collateral vessels and systemic venous return exits the right side of the heart via the VSD. When associated with intact interventricular septum, (PA + IVS; PAIVS) pulmonary artery blood supply is via a patent ductus arteriosus and the systemic venous return exits the right side of the heart via an obligatory atrial septal defect.

Q**quadricuspid aortic valve**

A congenital anomaly in which the aortic valve is composed of four cusps. Aortic regurgitation is frequent; severe aortic stenosis may occur.

R**Raghib complex**

A rare congenital anomaly that consists of persistent left superior vena cava connecting to the left atrium, atresia of the ostium of the coronary sinus or absence of the coronary sinus, and an atrial septal defect. Raghib complex may be associated with cor triatriatum sinister. *See also* [cor triatriatum sinister](#), [persistent left superior vena cava](#). (Raghib G, Ruttenberg HD, Anderson RC, et al. Termination of left superior vena cava in left atrium, atrial septal defect, and absence of coronary sinus: a developmental complex. *Circulation*. 1965;31:906-918.)

Rashkind procedure

A balloon atrial septostomy performed as a palliative procedure to allow increased mixing of pulmonary and systemic venous return in infants born with complete transposition of the great arteries (*d*-TGA; Rashkind WJ, Miller WW. Creation of an atrial septal defect without thoracotomy: a palliative approach to complete transposition of the great arteries. *J Am Med Assoc*. 1966;196:991-992).

Rastelli procedure (operation)

An operation for repair of complete transposition of the great arteries in association with a large ventricular septal defect (VSD) and pulmonary stenosis in which a communication is established between the left ventricle (LV) and the aorta via the VSD using a baffle within the right ventricle (RV). The RV is connected to the pulmonary artery using a valved conduit, and the LV-pulmonary artery (PA) connection is obliterated. As a consequence, the LV supports the systemic circulation (Rastelli GC, McGoon DC, Wallace RB. Anatomic correction of transposition of the great arteries with ventricular septal defect and subpulmonary stenosis. *J Thorac Cardiovasc Surg*. 1969;58:545-552).

regional referral center (RRC)

A center for the care of adult patients with congenital heart disease incorporating, at a minimum, cardiology staff with special skills, training, and experience in the management of such patients and highly skilled echocardiographers.

restrictive right ventricular physiology

Physiologic behavior of the ventricles of some patients (eg, after repair of tetralogy of Fallot or of pulmonary atresia with intact ventricular septum). It may be defined by echocardiography as antegrade pulmonary artery flow in late diastole (a wave) through all phases of respiration. The pulsed recordings are obtained with the sample volume at the midpoint between the pulmonary valve cusps or remnants and the pulmonary artery bifurcation (Redington AN, et al. Antegrade diastolic pulmonary artery flow as a marker of right ventricular restriction after complete repair of pulmonary atresia with intact ventricular septum and critical pulmonary valve stenosis. *Cardiol Young*. 1992;2:382-386).

restrictive VSD

See [ventricular septal defect](#).

REV procedure (réparation à l'étage ventriculaire)

A surgical technique used to treat transposition of the great arteries (TGA) with ventricular septal defect (VSD) and pulmonary stenosis and malpositions similar to TGA, such as double-outlet right ventricle. Resection of a posteriorly deviated conal septum enlarges the VSD and facilitates the construction of a wide and straight left ventricle to aorta tunnel. Transection of the great arteries and the LeCompte maneuver permit direct implantation of the pulmonary artery on the right ventriculotomy.

right aortic arch

See [aortic arch anomalies](#).

right ventricle (RV)-dependent coronary circulation

Describes the coronary circulation in some patients with pulmonary atresia with intact ventricular septum, wherein fistulous communications develop between the RV and the coronary arteries in association with the hypertensive RV that preclude biventricular repair because RV decompression may lead to coronary artery steal, ischemia, and/or sudden death. (Freedom RM, Anderson RH, Perrin D. The significance of ventriculo-coronary arterial connections in the setting of pulmonary atresia with an intact ventricular septum. *Cardiol Young*. 2005;15:447-468.)

right ventricular dysplasia

See [Uhl anomaly](#).

Ross procedure; Ross operation

A method of aortic valve replacement involving autograft transplantation of the pulmonary valve, annulus, and trunk into the aortic position, with re-implantation of the coronary ostia into the neo-aorta. The right ventricular outflow tract is reconstructed with a homograft conduit (or a bioprosthetic valve and conduit). (Ross DN. Replacement of aortic valve with a pulmonary autograft. *Lancet*. 1967;2:956-958; Ross D. Pulmonary valve autotransplantation [the Ross operation]. *J Cardiac Surg*. 1988;3:313-319).

rubella syndrome

A wide spectrum of malformations including cataracts, retinopathy, deafness, bone lesions, intellectual impairment and congenital heart disease caused by maternal rubella infection early in pregnancy. The associated congenital heart lesions include pulmonary artery stenosis, patent ductus arteriosus, tetralogy of Fallot, and ventricular septal defect.

RV infundibulum

A normal connecting segment between the body of the right ventricle and the pulmonary artery. *Syn:* RV conus. *See also* [infundibulum](#).

RVOTO

Right ventricular outflow tract obstruction.

S**sail sound**

An auscultatory finding heard in early systole in some patients with Ebstein anomaly. The first heart sound includes mitral valve closure as its first component, with a delayed tricuspid component. The abnormally large tricuspid anterior leaflet snapping like a sail catching the wind causes this delayed closure sound. The sail sound is not an ejection click, although it may simulate one.

Sano modification

A modification of the Norwood procedure for hypoplastic left heart syndrome, in which a right ventricular–pulmonary artery (RV-PA) conduit is constructed to maintain pulmonary blood flow instead of a modified Blalock-Taussig-Thomas anastomosis. (Sano S, Ishino K, Kawada M. Right ventricle-pulmonary artery shunt in first-stage palliation of hypoplastic left heart syndrome. *J Thorac Cardiovasc Surg.* 2003;126(2):504-509.) See also [Norwood procedure](#).

SAS

See [subaortic stenosis](#).

scimitar syndrome

A constellation of anomalies including infradiaphragmatic total or partial anomalous pulmonary venous connection of the right lung to the inferior vena cava, often associated with hypoplasia of the right lung and right pulmonary artery. The lower portion of the right lung tends to receive its arterial supply from the abdominal aorta. The name of the syndrome derives from the appearance on the posteroanterior chest radiograph of the shadow formed by the anomalous pulmonary venous connection that resembles a Turkish sword, or scimitar.

secondary erythrocytosis

See [erythrocytosis](#); See also [polycythemia vera](#).

segmental analysis

Syn: sequential segmental analysis. A systematic approach of describing congenital heart defects, based on the identification and arrangement of the atrial segment, the ventricular segment, the arterial segment, the connection between these three segments, and then the description the associated intracardiac defects.

Senning procedure (operation)

An operation for complete transposition of the great arteries in which venous return is directed to the contralateral ventricle by means of an atrial baffle fashioned in situ by using the right atrial wall and interatrial septum. As a consequence, the right ventricle supports the systemic circulation. This is a type of “atrial switch” operation (Senning A. Surgical correction of transposition of the great vessels. *Surgery.* 1959;45:966-980). See also [Mustard procedure](#), [atrial switch procedure](#), [double-switch procedure](#), [Ilbawi procedure](#).

septomarginal trabeculation

Prominent muscular structure on the septal surface of the right ventricle consisting of a body and two limbs. The body extends distally towards the apex of the right ventricle. Proximally, the septomarginal trabeculation splits into two limbs at the base of the right ventricle: the posteroinferior or postero-caudal limb, which gives rise to the papillary muscle; and the anterosuperior or anterocephalad limb, which extends to the pulmonary valve. The supraventricular crest inserts between the two limbs of the septomarginal trabeculation. The ventricular septal defect in tetralogy of Fallot is between the limbs of the septomarginal trabeculation. *Syn:* septal band.

septoparietal trabeculation

Prominent muscular structure that extends from the anterior surface of the septomarginal trabeculation to the lateral (parietal) wall of the right ventricle.

Shone complex (syndrome)

An association of multiple levels of left ventricular inflow and outflow obstruction: subvalvular and valvular left ventricular outflow tract obstruction, coarctation of the aorta, and mitral stenosis (parachute mitral valve and supramitral ring). (Shone JD, Sellers RD, Anderson RC, Adams P Jr, Lillehei CW, Edwards JE. The developmental complex of “parachute mitral valve,” supraventricular ring of left atrium, subaortic stenosis and coarctation of aorta. *Am J Cardiol.* 1963;11:714-725.)

Shprintzen syndrome

See [velocardiofacial syndrome](#), [CATCH-22](#), [microdeletion 22q11.2 syndrome](#).

shunt

Movement of blood through a congenitally abnormal or surgically created connection between two circuits, at the level of the atria, ventricles, or great arteries. *Shunt* is a physiologic term relating to flow, in contrast to *connection*, which is an anatomic term.

Silver-Russell syndrome

Growth disorder characterized by slow growth in utero and after birth. Affected patients typically have a low birth weight and show failure to thrive. Silver-Russell syndrome may be associated with cor triatriatum sinister.

single (as in atrium, ventricle, etc.)

Implies absence of the corresponding contralateral structure. Contrasts to “common,” which implies bilateral structures with absent septation. See also [common](#).

sinus of Valsalva

An anatomic dilation of the ascending aorta that occurs just above the aortic valve (*Syn:* aortic sinus, sinus of Morgagni, Petit’s sinus). There are generally three aortic sinuses: the left, which gives rise to the left coronary artery; the right, which gives rise to the right coronary artery; and the posterior, or noncoronary sinus.

- **aneurysm of sinus of Valsalva:** Localized dilation of a sinus of Valsalva due to a separation between the aortic media and the annulus fibrosus, associated with a deficiency of elastic tissue and abnormal development of the bulbus cordis. Congenital sinus of Valsalva aneurysm typically involves only one sinus, whereas acquired sinus of Valsalva aneurysms (eg, atherosclerosis, trauma, syphilis) usually involve multiple sinuses.

sinus venosus

An embryologic structure, the anatomic precursor of the inferior vena cava, superior vena cava, coronary sinus, and part of the definitive right atrium, which is located external to the primitive right atrium in the early embryologic period (3 to 4 weeks’ gestation). The sinus portion of the right atrium receives the inferior vena cava, superior vena cava, and coronary sinus. The right and left valves of the sinus venosus separate the sinus venosus from the primitive right atrium, which is the embryologic precursor of the trabeculated or muscular portion of the right atrium, including the right atrial appendage, and in turn communicates with the tricuspid valve. The left valve of the sinus venosus joins the interatrial septum, retrogresses, and is absorbed. The right valve of the sinus venosus enlarges and functions to deflect oxygenated fetal blood coming from the placenta via the inferior vena cava across the foramen ovale. Through partial resorption of the right valve of the sinus venosus, remnants form the

eustachian valve related to the inferior vena cava, the thebesian valve related to the coronary sinus, and the crista terminalis. *Chiari network* describes right atrial reticula that are an extensively fenestrated remnant of the right valve of the sinus venosus. See also [cor triatriatum dexter](#), [eustachian valve](#), [thebesian valve](#), [sinus venosus defect](#), [Chiari network](#).

sinus venosus defect

A communication located posterosuperior to the oval fossa (called superior sinus venosus defect) or, rarely, posteroinferior to the oval fossa (called inferior sinus venosus defect). It is commonly associated with partial anomalous pulmonary venous connection (most often right pulmonary veins, especially the right upper pulmonary vein in association with a superior sinus venosus defect). The defect is functionally identical to an atrial septal defect but properly named a sinus venosus defect because it occurs due to abnormal development of the sinus venosus in relation to the systemic and pulmonary veins and is not a defect in the interatrial septum. See also [atrial septal defect](#).

situs

Sidedness. The position of the morphologic right atrium determines the sidedness and is independent of the direction of the cardiac apex or the position of the ventricles or the great arteries.

- **situs ambiguus:** Indeterminate sidedness (in the setting of atrial isomerism).
- **situs inversus:** Mirror-image sidedness (ie, opposite of normal); left-sided morphologic right atrium.
- **situs inversus totalis:** Total mirror-image sidedness; the position of all lateralized organs is inverted.
- **situs solitus:** Normal sidedness; right-sided morphologic right atrium.

Smith-Lemli-Opitz (SLO) syndrome

The SLO syndrome is an inherited disorder with a reduced enzyme activity of DHCR7 leading to severe deficiency of cholesterol. The syndrome is associated with congenital heart disease, most commonly tetralogy of Fallot, atrial septal defect, ventricular septal defect, and patent ductus arteriosus (Smith DW, Lemli L, Opitz JM. A newly recognized syndrome of multiple congenital anomalies. *J Pediatr*. 1964;64:210-221).

Starnes procedure

A strategy of fenestrated right ventricular exclusion and systemic to pulmonary shunt applied in severe Ebstein anomaly and also in some cases of [PAIVS](#) in order to facilitate univentricular repair (Starnes VA, Pitlick PT, Bernstein D, Griffin ML, Choy M, Shumway NE. Ebstein's anomaly appearing in the neonate: a new surgical approach. *J Thorac Cardiovasc Surg*. 1991;101:1082-1087).

stent

Intravascular (intraluminal) prosthesis that scaffolds a vessel for the purpose of maintaining patency.

Sterling-Edwards procedure

A legacy palliative operation for transposition of the great arteries in which the atrial septum is resected, repositioned, and sutured to the left of the right pulmonary veins to produce drainage to the right atrium. The procedure produces left-to-right shunt of oxygenated blood directly into the systemic atrium and ventricle and offloads the pulmonary circulation in patients with complete transposition of the great arteries

and high pulmonary flow (Edwards WS, Bargeron Jr LM, Lyons C. Reposition of right pulmonary veins in transposition of the great vessels. *J Am Med Assoc*. 1964;188:522-523; Edwards WS, Bargeron LM. More effective palliation of the transposition of the great vessels. *J Thorac Cardiovasc Surg*. 1965;19:790-795).

straddling atrioventricular valve

See [atrioventricular valve](#).

subaortic stenosis (SAS)

A group of defects leading to subvalvar left ventricular outflow tract obstruction. The lesion can be discrete, membranous, or tubular, and may occur in isolation or be associated with other defects.

subaortic ventricle

The ventricle that relates most directly to the aorta.

subpulmonary ventricle

The ventricle that relates most directly to the pulmonary artery.

superoinferior heart

A term applied to a heart in which the ventricles are in a markedly superoinferior relationship due to abnormal displacement of the ventricular mass along the horizontal plane of its long axis. This anomaly often coexists with crisscross atrioventricular relationships. See also [crisscross heart](#). *Syn:* over-and-under ventricles; upstairs-downstairs heart.

supracristal

Located above the crista supraventricularis in the right ventricular outflow tract, hence contiguous with the origin of the great arteries. See [crista supraventricularis](#).

supraregional referral center (SRRC)

A "full service" center for providing optimal care of adult patients with congenital heart disease comprising specialized resources, the availability of cardiology specialists with specific training and experience in ACHD, the availability of other cardiology subspecialists, and other medical and paramedical personnel with special training or experience in the problems of congenital heart disease and offering opportunities for training, research, and education in the field. *Syn:* national referral center.

supravalvar aortic stenosis (SVAS)

A rare form of aortic stenosis, characterized by supravalvar narrowing of the proximal ascending aorta. The narrowing is typically hourglass-shaped but can be more diffuse and tubular, and it may involve the entire aortic root, including the coronary ostiae. SVAS is often associated with the Williams-Beuren syndrome (see [Williams syndrome](#)). It can also occur as a familial form, as a sporadic disease, or in conjunction with other congenital cardiac defects involving left ventricular outflow tract obstruction.

supravalvular mitral ring

An anomaly found in the left atrium that may produce congenital mitral stenosis. Supravalvular mitral ring is to be differentiated from [cor triatriatum sinister](#). A supravalvular mitral ring separates the left atrial appendage from the mitral valve, whereas in [cor triatriatum sinister](#) the left atrial appendage is always in the same chamber as the mitral valve. See also [Shone complex](#), [cor triatriatum](#).

SVAS

See [supravalvar aortic stenosis](#).

switch conversion of transposition

An operation performed in patients who had previously had a Mustard or Senning procedure for complete transposition of the great arteries to allow the left ventricle to assume the function of the systemic (subaortic) ventricle. The first stage may involve pulmonary artery banding to induce subpulmonary left ventricular hypertrophy. The second stage involves an arterial switch operation, removal of the Mustard/Senning atrial baffle, and reconstruction of an atrial septum. See also [double-switch operation](#).

systemic AV valve

The atrioventricular valve guarding the inlet to the systemic (subaortic) ventricle.

systemic-to-pulmonary artery shunt

An overarching term describing palliative surgical shunts from the aorta and its branches to the pulmonary artery for the purpose of increasing pulmonary blood flow. See also [Blalock-Taussig-Thomas shunt](#), [Waterston shunt](#), [Potts shunt](#), central shunt.

T**Takeuchi procedure (Takeuchi intrapulmonary tunnel)**

A surgical procedure for the management of anomalous left coronary artery arising from the pulmonary artery (ALCAPA) that involves creating an aortopulmonary window and an intrapulmonary-artery tunnel to connect the ostium of the anomalous left coronary artery to the ascending aorta. (Takeuchi S, Imamura H, Katsumoto K. New surgical method for repair of anomalous left coronary artery from pulmonary artery. *J Thorac Cardiovasc Surg*. 1979;78:7-11.) See also [Bland-White-Garland syndrome \(ALCAPA\)](#).

TAPVC

Total anomalous pulmonary venous connection. See [anomalous pulmonary venous connection](#).

TAPVD

Total anomalous pulmonary venous drainage. This term is sometimes used to refer to the entity properly called total anomalous pulmonary venous connection. See [anomalous pulmonary venous connection](#). See also [connection](#), [drainage](#).

Taussig-Bing anomaly

A form of double-outlet right ventricle in which the great arteries arise side by side with the aorta to the right of the pulmonary artery and the ventricular septal defect (VSD) in a subpulmonary position. Because the left ventricle empties across the VSD preferentially into the pulmonary artery, the physiology simulates complete transposition of the great arteries with a VSD.

TCPC

Total cavopulmonary connection. See [Fontan procedure/total cavopulmonary connection](#).

tetralogy of Fallot

A congenital anomaly, the primary pathophysiologic components of which are obstruction to right ventricular outflow at the

infundibular level and a large nonrestrictive ventricular septal defect (VSD). The other two components of the “tetralogy” are an overriding aorta and concentric right ventricular hypertrophy. Valvular right ventricular outflow tract obstruction (RVOTO; pulmonic stenosis) and distal pulmonary artery stenosis are often present. The essential morphogenetic anomaly is malalignment of the infundibular (outlet) septum such that it fails to unite with the trabecular septum (hence the VSD), owing to anterior deviation (hence the RVOTO). Lillehei and colleagues first described the repair in 1955 (Lillehei CW, Cohen M, Warden HE. Direct vision intracardiac surgical correction of the tetralogy of Fallot, pentalogy of Fallot, and pulmonary atresia defects: reports of first ten cases. *Ann Surg*. 1955;142:418-445).

- **pentalogy of Fallot:** Tetralogy of Fallot with an associated atrial septal defect or patent foramen ovale.
- **pink tetralogy of Fallot:** Tetralogy of Fallot presenting with increased pulmonary blood flow and minimal cyanosis because of a lesser degree of RVOTO. *Syn:* acyanotic Fallot.
- **tetralogy of Fallot with pulmonary atresia:** Tetralogy of Fallot with no anterograde flow from RV to PA. Pulmonary blood supply is by MAPCAs. A form of complex pulmonary atresia. Sometimes called *pseudotruncus arteriosus*. See also [MAPCA](#), [pseudotruncus arteriosus](#).

thebesian valve

A remnant of the right valve of the sinus venosus guarding the opening of the coronary sinus.

thebesian veins

Minute veins within the heart wall that drain directly into the cavity of the heart. *Syn:* thebesian vessels.

total anomalous pulmonary venous connection (drainage, return)

See [anomalous pulmonary venous connection/total anomalous pulmonary venous connection](#).

total cavopulmonary connection (TCPC)

See [Fontan procedure/total cavopulmonary connection](#).

trabecular VSD

See [ventricular septal defect](#).

transannular

Crossing the annulus. In connection with the right ventricular outflow tract in tetralogy of Fallot, the term refers to the pulmonary valve annulus, which often must be enlarged by a transannular patch, with consequent obligatory pulmonary insufficiency. Transannular patching was first described in 1959 (Kirklin JW, Ellis Jr FH, McGoon DC, Dushane JW, Swan HJ. Surgical treatment for tetralogy of Fallot by open intracardiac repair. *J Thorac Surg*. 1959;37:22-51).

transfer

The physical relocation of patients and their health records from one institution or practitioner to another. Transfer refers to an event and is to be differentiated from “transition,” which describes a process. See also [transition](#).

transition

The purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented health care systems. Transition is a

process and is to be differentiated from the event of “transfer” (Rosen DS, Blum RW, Britto M Sawyer SM, Siegel DM, Society for Adolescent Medicine. Transition to adult health care for adolescents and young adults with chronic conditions: a position paper of the Society for Adolescent Medicine. *J Adolesc Health*. 2003;33:309-311). See also [transfer](#).

transposition of the great arteries (TGA)

Syn: complete transposition of the great arteries. See [discordant ventriculoarterial connections](#); See also [congenitally corrected transposition of the great arteries](#).

- **complex transposition of the great arteries**: Discordant connection of the great arteries and ventricles such that the pulmonary trunk arises from the left ventricle and the aorta arises from the right ventricle, with associated abnormalities, most commonly a ventricular septal defect.
- **simple transposition of the great arteries**: Discordant connection of the great arteries and ventricles such that the pulmonary trunk arises from the left ventricle and the aorta arises from the right ventricle, without any additional associated abnormality.

tricuspid atresia

A congenital anomaly in which there is no physiologic or gross morphologic connection between the right atrium and right ventricle. An internal connection allows mixing of systemic and pulmonary venous return at the atrial level. There is a variable degree of hypoplasia of the right ventricle. The left ventricle and mitral valve are normal.

truncus arteriosus

A single artery (truncus, trunk) arises from the base of the heart because of failure of proximal septation into the aorta and the pulmonary artery. Both pulmonary and systemic arteries, as well as the coronary arteries, arise from the common trunk. Truncus arteriosus is divided into two types, depending on whether there is a ventricular septal defect or an intact ventricular septum. *Syn*: common arterial trunk.

Turner syndrome

A clinical syndrome resulting from sex chromosome abnormalities affecting females. The syndrome is due to a 45 XO karyotype in about 50% of cases, with 45XO/45XX mosaicism and other X-chromosome abnormalities comprising the remainder. There is a characteristic but variable phenotype and association with congenital cardiac anomalies, especially post-ductal coarctation of the aorta and other left-sided obstructive lesions, as well as partial anomalous pulmonary venous connection without atrial septal defect. The phenotype varies and is somewhat similar to that of Noonan syndrome. See also [Noonan syndrome](#).

U

Uhl anomaly

Congenital malformation consisting of hypoplasia or aplasia of the right ventricular myocardium, presenting with marked enlargement of both the right ventricle and right atrium and consequent tricuspid regurgitation. Arrhythmogenic right ventricular dysplasia/cardiomyopathy, in which there is progressive replacement of right ventricular myocardium by adipose and fibrous tissue, associated with ventricular arrhythmias, may be one end of a spectrum and Uhl anomaly the other.

unbalanced atrioventricular canal

See [ventricular imbalance](#).

unguarded tricuspid orifice

A congenital abnormality of the tricuspid valve consisting of partial or complete agenesis of valvular tissue leading to congenital tricuspid regurgitation.

unicuspid aortic valve

A congenital anomaly in which the aortic valve is comprised of only a single cusp

- **acommissural unicuspid aortic valve** is characterized by a single cusp, a stenotic central orifice, and rudimentary commissures that do not divide the valve, generally presenting in infancy with severe stenosis.
- **unicommissural unicuspid aortic valve** is characterized by a single cusp with a single commissural attachment to the aortic wall and an elongated orifice.

unifocalization

A surgical technique that creates a common trunk for major aortopulmonary collateral arteries (MAPCAs), often utilizing a Blalock-Taussig-Thomas shunt or conduit, as part of the surgical management of complex pulmonary atresia.

univentricular connection

Both atria are connected to only one ventricle. The connection is univentricular, but the heart is usually biventricular. *Syn*: double-inlet ventricle.

unroofed coronary sinus

An anomaly in which there is a deficiency in the normal separation of the coronary sinus from the left atrium as the coronary sinus passes behind the left atrium in the atrioventricular groove, such that the coronary sinus drains to the left atrium, thus an atrial level left to right shunt. A form of absence of the coronary sinus.

upstairs-downstairs heart

See [superoinferior heart](#).

V

VACTERL association

Describes a spectrum of defects including vertebral abnormalities, anal atresia, tracheoesophageal fistula, radial dysplasia, renal abnormalities, and congenital heart defects (atrial and ventricular septal defect, tetralogy of Fallot, truncus arteriosus, aortic coarctation, patent ductus arteriosus, etc.).

Valsalva, sinus of

See [sinus of Valsalva](#).

valve-sparing aortic root replacement (VSRR)

See [David operation](#); [Yacoub procedure](#).

vascular ring

A wide spectrum of aortic arch anomalies, including double aortic arch and other vascular structures that surround the trachea and the esophagus resulting in their compression. The vascular structures may or may not be patent. Vascular rings may be isolated (in 1% to 2% of patients with congenital heart disease) or associated with other congenital heart malformations, such as tetralogy of Fallot. See [aortic arch anomalies](#).

velocardiofacial syndrome

Syndrome of cleft palate, abnormal facies (square nasal root, long nose with narrow alar base, long face with malar hypoplasia, long philtrum, thickened helix, low-set ears), velopharyngeal incompetence, and congenital cardiac defects (conotruncal anomalies, isolated ventricular septal defect, tetralogy of Fallot). It is due to microdeletion at chromosome 22q11.2. *Syn:* Shprintzen syndrome, microdeletion 22q11.2 syndrome. *See also* [CATCH-22](#), [microdeletion 22q11.2 syndrome](#).

venous (or subpulmonary) AV valve

The atrioventricular valve guarding the inlet to the subpulmonary or “venous” ventricle.

ventricle repair

- **1-ventricle repair:** *See* [Fontan procedure](#).
- **1.5-ventricle repair:** A term used to describe operations for cyanotic congenital heart disease as well as some cases of Ebstein anomaly, performed when the subpulmonary ventricle is insufficiently developed to accept the entire systemic venous return. A bidirectional cavopulmonary connection is constructed to divert superior vena cava flow directly to the lungs, while inferior vena cava flow is directed to the lungs via the functioning but small subpulmonary ventricle.
- **2-ventricle repair:** A term used to describe operations for cyanotic congenital heart disease with a common ventricle in which functioning systemic (subaortic) and subpulmonary ventricles are created by means of surgical septation of the common ventricle.

ventricular imbalance

In the setting of atrioventricular septal defect, ventricular imbalance refers to relative hypoplasia of one or the other of the ventricles in association with small size of the ipsilateral component of the atrioventricular annulus.

ventricular septal defect (VSD)

A defect in the ventricular septum, such that there is direct communication between the two ventricles. The anatomic classification system for VSDs proposed by the Society for Thoracic Surgery—Congenital Heart Surgery Database Committee in association with the European Association for Cardiothoracic Surgery is presented, followed by additional definitions for specific types of VSD:

- **doubly committed (juxta-arterial) VSD:** A defect in the outlet septum such that there is fibrous continuity between the aortic and pulmonary valves, with the VSD situated directly beneath both semilunar valves. *See* type 1 VSD.
- **inlet VSD:** A defect in the lightly trabeculated inlet portion of the muscular interventricular septum, typically seen as part of an atrioventricular septal defect. *See* type 3 VSD.
- **muscular VSD:** A defect entirely surrounded by muscular interventricular septum. *See* type 4 VSD.
- **nonrestrictive VSD:** A ventricular septal defect of such a size that there is no significant pressure gradient between the ventricles. Hence the pulmonary artery is exposed to systemic pressure unless there is right ventricular outflow tract obstruction.
- **outlet VSD:** A defect in the nontrabeculated outlet portion of the muscular interventricular septum, hence above the crista supraventricularis. *Syn:* supracristal VSD. Sometimes also described as subpulmonary, subarterial, or doubly committed subarterial VSD. *See* type 1 VSD.

- **perimembranous VSD:** A VSD located in the membranous portion of the interventricular septum with variable extension into the contiguous portions of the inlet, trabecular, or outlet portions of the muscular septum but not involving the atrioventricular septum. *Syn:* membranous VSD; infracristal VSD, type 2 VSD.
- **restrictive VSD:** A VSD of sufficiently small size that there is a pressure gradient between the ventricles, such that the pulmonary ventricle (hence pulmonary vasculature) is protected from the systemic pressure of the contralateral ventricle.
- **Swiss cheese VSD:** Multiple muscular VSDs. *See* type 4 VSD.
- **trabecular VSD:** A defect in the heavily trabeculated central or trabecular portion of the muscular interventricular septum. May be multiple. *See* type 4 VSD.
- **type 1 VSD:** Located in the outlet portion of the muscular septum. It is also termed a conal, subpulmonary, infundibular, or supracristal defect. Also in this category is the doubly committed juxta-arterial VSD.
- **type 2 VSD:** Confluent with the membranous septum. These defects usually extend into one of the three components of the muscular septum. *Syn:* perimembranous or membranous VSD.
- **type 3 VSD:** Located in the inlet portion of the muscular septum inferior to the atrioventricular valves. *Syn:* inlet VSD or atrioventricular canal type VSD.
- **type 4 VSD:** Located in the trabecular portion of the muscular septum completely surrounded by muscle. *Syn:* trabecular VSD, muscular VSD. Location may be midmuscular, apical, posterior, or anterior.

ventriculoarterial concordance

See [concordant ventriculoarterial connections](#).

ventriculoarterial discordance

See [discordant ventriculoarterial connections](#).

ventriculo-infundibular fold

Muscle interposed between the leaflets of an atrioventricular and of a subarterial valve, thus separating the inlet and outlet portions of a ventricle.

W**Warden procedure**

A surgical procedure for correction of sinus venosus defect with partial anomalous pulmonary venous connection to the superior caval vein, involving transposing the superior caval connection to the right atrial appendage and closing the sinus venosus defect incorporating the stump of the SVC to baffle the anomalous veins to the LA (Warden HE, Gustafson RA, Tarnay TJ, Neal WA. An alternative method for repair of partial anomalous pulmonary venous connection to the superior vena cava. *Ann Thorac Surg*. 1984;38:601-605).

Waterston anastomosis

Syn: Waterston shunt, Waterston-Cooley shunt. A palliative operation for the purpose of increasing pulmonary blood flow, and hence systemic oxygen saturation, which involves creating a small communication between the right pulmonary artery and the ascending aorta. It is often complicated by the development of pulmonary vascular obstructive disease, if too large. Not uncommonly it causes distortion of the pulmonary artery

(Waterston DJ. Treatment of Fallo's tetralogy in children under one year of age. *Rozhl Chir.* 1962;41:181-183).

Williams syndrome

Syn: Williams-Beuren syndrome. An autosomal dominant syndrome, often arising de novo, associated with an abnormality of elastin, infantile hypercalcemia, mild cognitive impairment and the so-called cocktail personality, and congenital heart disease, especially supravalvular aortic stenosis and multiple peripheral pulmonary stenoses (Williams JC, Barratt-Boyes BG, Lowe JB. Supravalvular aortic stenosis. *Circulation.* 1961;24:1311-1318; Beuren AJ, Apitz J, Harmjan D. Supravalvular aortic stenosis in association with mental retardation and certain facial features. *Circulation.* 1962;26:1235-1240).

Wolff-Parkinson-White (WPW) syndrome

Accessory lateral atrioventricular conduction pathway causing characteristic electrocardiographic changes and atrial (and sometimes ventricular) arrhythmias. WPW syndrome may be isolated or associated with congenital heart defects. It is found in up to 25% of patients with Ebstein anomaly, who typically have more than one accessory pathway.

Wood unit

A nonstandard unit for expressing pulmonary vascular resistance (mm Hg/L), named after Paul Wood, the famous British cardiologist. One Wood unit is equivalent to 80 dyn/s/cm⁵.

X

xenograft

Tissue or organ used for transplant derived from another species. *Syn:* heterograft.

Y

Yacoub procedure

A form of valve-sparing aortic root replacement described as the "remodeling" procedure, in which an aortic tube graft is sutured onto the aortic annulus above the insertion line of the aortic cusps, thus leaving the annulus mobile (but unsupported) and allowing billowing of the graft. This contrasts to the David I valve-sparing aortic root replacement, described as the "re-implantation" procedure, in which the graft is fixed to the left ventricular outflow tract at the subannular level and the valve is re-implanted inside the fabric graft, thus fixing the size of the aortic annulus permanently. The Yacoub procedure has also been called the David II procedure (Sarsam MA, Yacoub M.

Remodeling of the aortic valve annulus. *J Thorac Cardiovasc Surg.* 1993;105:435-438). See also [David operation](#).

Yasui operation

A complex operation for patients with interrupted aortic arch, hypoplastic ascending aorta and ventricular septal defect, combining elements of the Norwood operation for hypoplastic left heart syndrome and of the Rastelli operation. Components of the Yasui operation include (1) reconstruction of the aortic arch; (2) Damus-Kaye-Stansel anastomosis to reconstruct the hypoplastic ascending aorta; (3) diversion of the left ventricular blood to the pulmonary valve with a patch; (4) connection of the right ventricle to the pulmonary artery with a valved conduit. (Yasui H, Kado E, Nakano, et al. Primary repair of interrupted aortic arch and severe aortic stenosis in neonates. *Thorac Cardiovasc Surg.* 1987;93:539-545.)

Z

Z score, Z value

The number of standard deviations a measurement departs from mean normal for a given body surface area. It is a way of expressing a physiologic variable in a form corrected for age and body size that is important in pediatrics. (Rimoldi HJA, et al. A note on the concept of normality and abnormality in quantitation of pathologic findings in congenital heart disease. *Pediatr Clin North Am.* 1963;10:589-591; Daubeney PEF, et al. Relationship of the dimension of cardiac structures to body size: an echocardiographic study in infants and children. *Cardiol Young.* 1999;9:402-410.)

Acknowledgments

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