Further Reading


Further Reading

Bibliography

*Diagnosis and Management of Adult Congenital Heart Disease* by Michael A. Gatzoulis, Gary D. Webb, Piers Daubeney. Churchill Livingstone; 2003
This practical resource provides essential guidance on the anatomic issues, clinical presentation, diagnosis and clinical management of adults with congenital heart disease. Each consistently structured, disease-oriented chapter discusses incidence, genetics, morphology, presentation, investigation and imaging, treatment and intervention. A wealth of illustrations, including line drawings, EKGs, radiographs and echocardiograms clearly depict the clinical manifestations of congenital defects.

Clinical reference for cardiologists. Perloff and Child’s book provides excellent clinical information on the special needs and concerns faced in caring for adults with congenital heart disease. First textbook of its kind in the field.

*Congenital Heart Disease Adult* by Welton M. Gersony, Marlon, S. Rosenbaum, Myron L. Weisfeldt. McGraw-Hill Professional; 2001
This guide to the broad spectrum of congenital heart defects helps to optimize adult patient care.

*Congenital Heart Disease in Adults: A Practical Guide* by Andrew Redington, Darryl Shore, Paul Oldershaw. W B Saunders; 1997
Concise text for cardiologists and family practitioners on the special aspects of managing congenital heart disease in adults. Takes the approach that these adults can’t be treated as large children with congenital disease.

*Cardiac Surgery* by Nicholas Kououchkos, Eugene Blackstone, Donald Doty, Frank Hanley, Robert Karp. W B Saunders; 3rd edition 2003
Essential textbook in both adult and pediatric cardiac surgery, updated and revised in a new, third edition. It thoroughly covers the full range of new and classic surgical procedures and presents the up-to-date clinical evidence practitioners need to make effective management decisions.
Bibliography

*Moss and Adams’ Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult* by Hugh D. Allen, Howard P. Gutgesell, Edward B. Clark, David J. Driscoll. Lippincott Williams & Wilkins; 6th edition 2000

Updated throughout, the sixth edition of Moss and Adams continues to be the primary cardiology text for those who care for infants, children, adolescents, young adults, and fetuses with heart disease. A comprehensive text covering basic science theory through clinical practice of cardiovascular disease in the young, this edition includes an expanded special section on young adults and a greatly expanded genetics section.


A correlation of the clinical and pathological features of congenital heart disease, including anatomy, imaging and pathology. This work covers all aspects of structural defects of the heart and major vessels arising during the development of the fetus, as well as conditions seen in neonates and young children. It provides a comprehensive review of incidence and actuality of various conditions, their characteristic features as defined by a range of investigative techniques, and detailed discussion of underlying pathology.


A comprehensive and exhaustive reference of fundamental and clinical aspects of heart disease in infancy and childhood. The contributors are well-known experts in the field and the editors are a world-class group who have published extensively in the field. Provides an up-to-date and authoritative account of pediatric cardiovascular fields covering embryology, morphology, pathophysiology, specific clinical conditions, treatments and the psychosocial aspects of caring for patients with heart disease.

*Cardiac Arrhythmias after Surgery for Congenital Heart Disease* by S. Balaji, P.C. Gillette, C.L. Case. Hodder Arnold; 2001

This comprehensive text discusses all aspects of atrial and ventricular cardiac arrhythmias in patients undergoing cardiac surgery for congenital heart problems. This area is one of growing interest, as an increasing number of individuals with heart defects live longer due to improved therapy, and are now facing new problems. The numbers and types of problems being tackled by invasive ablation techniques have been growing dramatically since the mid-1980s, and this publication aims to tie together the aspects of these conditions. Appropriate management of these patients critically depends on knowledge of the type of surgery, the types of arrhythmias these patients are prone to, and the therapeutic modalities that can be used to treat them.


The essential *Textbook of Cardiovascular Medicine*, bringing cutting-edge advances in the field. Ninety-eight world authorities synthesize everything from the newest findings in molecular biology and genetics to the latest imaging modalities, interventional procedures and medications. The two adult congenital heart editors Drs Therrien and Webb in this latest edition encompass all of today’s essential knowledge in the field.
Task Force on the Management of Cardiovascular Diseases During Pregnancy of the European Society of Cardiology

Canadian Cardiovascular Society Consensus Conference 2001 update

Task Force on the Management of Grown Up Congenital Heart Disease of the European Society of Cardiology
*Eur Heart J* 2003, 24, 1035–84.

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The purpose of this glossary is to help guide those reading and researching in the area of adult congenital heart disease. It is meant to be a living document, constantly under revision, improvement, correction, as you, its users, find ways to ease the path for those who follow. To this end, if you cannot find a term you think should be here, or if you disagree with a definition, or see a way to improve it, drop us an e-mail before you move on. We promise to consider all feedback carefully, and to make additions and revisions on the website (http://www.achd-library.com/) often. We hope you find the glossary helpful.

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aberrant innominate artery
A rare abnormality associated with right aortic arch wherein the sequence of arteries arising from the aortic arch is: right carotid artery, right subclavian artery, then (left) innominate artery. The latter passes 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 (i.e. right carotid artery in left aortic arch and left carotid artery in right aortic arch). syn. retro-esophageal innominate artery.

aberrant subclavian artery
The right subclavian artery arises 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, first described in 1735 by Hunauld, and occurring in 0.5% of the general population.

absent pulmonary valve syndrome
Pulmonary valvular tissue is absent, 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 dilatation and distortion of the pulmonary arteries. Absent pulmonary valve may also occur in association with other simple or complex congenital heart lesions.

ACHD
Adult congenital heart disease.

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). see situs.

Amplatzer® device
A self-centering device delivered percutaneously by catheter for closure of an atrial septal defect, a patent foramen ovale or a patent ductus arteriosus.

anomalous pulmonary venous connection
Pulmonary venous return to the right heart, which may be total or partial.

• 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 (SVC). The connection may also be infradiaphragmatic via a descending vein to the portal vein, the inferior vena cava (IVC) or one of its tributaries. Pulmonary venous obstruction is common in supradiaphragmatic connection, and almost universal in infradiaphragmatic connection.

- 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 atrial septal defect. see also scimitar syndrome.

aortic arch anomalies
Abnormalities of the aortic arch and its branching. Note that 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 (i.e. the right carotid artery in left aortic arch and the left carotid artery in right aortic arch). The most important anomalies are:

- abnormal left aortic arch
  - left aortic arch with minor branching anomalies;
  - left aortic arch with retroesophageal right subclavian artery.
- right aortic arch. In right aortic arch the descending thoracic aorta crosses the right mainstem bronchus. It is often associated with tetralogy of Fallot, pulmonary atresia, truncus arteriosus and other cono-truncal anomalies.

Types of right aortic arch branching include:
- mirror image branching (left innominate artery, right carotid artery, right subclavian artery);
- retroesophageal left (aberrant) subclavian artery with a normal calibre.
  Sequence of branching: left carotid artery, right carotid artery, right subclavian artery, then 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;
- 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.
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- **cervical aortic arch.** The arch is located above the level of the clavicle.
- **double aortic arch.** Both right and left aortic arches are present, i.e. the ascending aorta splits into two limbs encircling the trachea and esophagus. 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.
- **persistent 5th aortic arch.** Double-lumen aortic arch with both lumina on the same side of the trachea. Degree of lumen patency varies from full patency of both lumina to complete atresia of one of them. Seen in some patients with coarctation of the aorta or interruption of the aortic arch.
- **interrupted aortic arch.** Complete discontinuation between the ascending and descending thoracic aorta.
  - Type A: interruption distal to the subclavian artery that is ipsilateral to the second carotid artery.
  - Type B: interruption between second carotid artery and ipsilateral subclavian artery.
  - Interruption between carotid arteries.

**aortic-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.

**aortic override**
*see* tetralogy of Fallot.

**aortic valve-sparing ascending aortic replacement**
*see* David operation.

**aorto-pulmonary collateral**
Abnormal arterial vessel arising from the aorta, providing blood supply to the pulmonary arteries. May be single or multiple, and small or large (see also MAPCA). May be associated with tetralogy of Fallot, pulmonary atresia or other complex cyanotic congenital heart disease.

**aorto-pulmonary septal defect**
*see* aorto-pulmonary window.

**aorto-pulmonary window**
A congenital connection between the ascending aorta and main pulmonary artery, which may be contiguous with the semi-lunar valves, or, less often, separated from them. Simulates the physiology of a large PDA, but requires a more demanding repair. *syn.* aorto-pulmonary septal defect.
arterial switch operation  
*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 stenoses or diffuse hypoplasia of the pulmonary artery and its branches. Associated with microdeletion in chromosome 20p. *syn.* Alagille syndrome.

asplenia syndrome  
*see* isomerism/right isomerism.

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

atrial septal defect (ASD)  
an inter-atrial communication, classified according to its location relative to the oval fossa (fossa ovalis):

- coronary sinus ASD. Inferior and anterior location 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.
- 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 that are located on the same plane; almost always associated with a “cleft” in the “anterior mitral leaflet”. This cleft is actually the separation between the leftsided portions of the primitive antero-superior and postero-inferior bridging leaflets. *see also* AVSD.
- ostium secundum ASD. Located at the level of the oval fossa.
- sinus venosus ASD. *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 (either the Mustard or the Senning procedure) this accomplishes physiologic correction of the circulation, while leaving the right ventricle to support the systemic circulation. In patients with l-transposition of the great arteries and in patients who have had a previous Mustard or Senning procedure, it is used as part of a “double switch procedure” which results in anatomic correction of the circulation, with the left ventricle supporting the systemic circulation. *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 septum which have in common: 1) a common atrioventricular junction with a common fibrous ring, and a unique, 5-leaflet, atrioventricular 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. 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” anterior mitral and/or septal tricuspid valve leaflet, 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 left ventricular inlet from the right atrium. It has two parts: a muscular portion which exists because the attachment of the septal leaflet of the tricuspid valve is more towards the apex of the ventricle than the corresponding attachment of the mitral valve, and a fibrous portion superior to the attachment of the septal leaflet of the tricuspid valve. This latter portion separates the right atrium from the sub-aortic left ventricular outflow tract. 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 always associated with the right ventricle, and the mitral valve 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 5-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 antero-superior leaflet, and the right portions of the superior and inferior bridging leaflets.

• cleft AV valve. A defect often involving the left AV valve in AVSD formed by the conjunction of the superior and inferior bridging leaflets. A cleft may also be seen in the septal tricuspid leaflet. A similar but morphogenetically distinct entity may involve the anterior or rarely posterior leaflet of the mitral valve in otherwise normal hearts.
• common AV valve. Describes a 5-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 VSD.
• straddling AV valve. Describes an AV valve with anomalous insertion of tendinous cords or papillary muscles into the contralateral ventricle (VSD required).

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 wherein the inferior vena cava (IVC) is interrupted distal to its passage through the liver, and 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.

Baffes operation
Anastomosis of the right pulmonary veins to the right atrium (RA) and the IVC to the left atrium (LA) by using an allograft aortic tube to connect the IVC and the LA. (Baffes TG. A new method for surgical correction of transposition of the aorta and pulmonary artery. Surg Gynecol Obstet 1956, 102, 227–233). This operation provided partial physiologic correction in patients with complete TGA. Lillehei and Varco originally described such a procedure 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 surgically created to divert blood flow. For instance, in atrial switch operations for complete transposition of the great vessels, an intra-atrial baffle is constructed to divert systemic venous return across the mitral valve, thence to the left ventricle and pulmonary artery, and pulmonary venous return across the tricuspid valve, thence to the right ventricle and aorta. see also Mustard procedure. see also Senning procedure.

balanced
As in “balanced circulation”, e.g. in the setting of VSD 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.

**Bentall procedure**
Replacement of the ascending aorta and the aortic valve with a composite graft valve device and reimplantation 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 wherein the aortic valve is comprised 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 2% of the general population and in 75% of patients with aortic coarctation.

**bidirectional cavopulmonary anastomosis**
see Glenn shunt/bidirectional Glenn.

**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, first described in 1950. 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 shunt**
A palliative operation for the purpose of increasing pulmonary blood flow, 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 (classical) 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. *Journal of the American Medical Association* 1945, 128, 189–202.)
Bland-White-Garland Syndrome
The left main coronary artery arises from the main pulmonary artery. The first report describing clinical and pathologic features was published in 1933. (Bland EF, White PD, Garland J. Congenital anomalies of the coronary arteries: report of an unusual case associated with cardiac hypertrophy. *American Heart Journal* 1933, 8, 787; 801) syn. ALCAPA.

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 VSD) is the result.

Brock procedure
A 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 ventricle (RV) infundibulum using a punch or biopsy-like instrument introduced through the right ventricle so as to reduce RV outflow tract obstruction, without VSD closure. The operation was performed without cardiopulmonary bypass. (Brock RC. Pulmonary valvotomy for the relief of congenital pulmonary stenosis: report of three cases. *British Medical Journal* 1948, 1, 1121–1126.)

bulbo-ventricular foramen
*syn.* primary foramen, primary ventricular foramen, primary interventricular foramen. An embryological 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.

CACH (Canadian Adult Congenital Heart) Network
A co-operative 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 ACHD patients. For more information, visit http://www.cachnet.org.

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:
  - levoposition—to the left;
Cardiac location is affected by many factors including underlying cardiac malformation, abnormalities of mediastinal and thoracic structures, tumors, kyphoscoliosis, abnormalities of the diaphragm.

- cardiac orientation—the base to apex orientation of the heart:
  - levocardia—apex directed to the left of the midline;
  - mesocardia—apex oriented inferiorly in the midline;
  - dextrocardia—apex directed to the right of the midline.

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 apical and subcostal 4-chamber views.

- 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 (MVO₂), anaerobic threshold (AT), and oxygen saturation with effort.

Cardio-Seal® device
A device delivered percutaneously by catheter for closure of an ASD or PFO.

CATCH 22
Syndrome due to microdeletion at chromosome 22q11 resulting in a wide clinical spectrum. 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.

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→q11 is duplicated. Phenotypic features include mental deficiency, anal and renal malformations, hypertelorism and others. Total anomalous pulmonary venous return is the commonest congenital cardiac lesion (in up to 40% of patients).

CHARGE association
This anomaly is characterized by the presence of coloboma or choanal atresia and three of the following defects: congenital heart disease, nervous system anomaly or mental retardation, genital abnormalities, 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 and 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 echocardiography studies. It presents with coarse right atrial reticula connected to the Eustachian and Thebesian valves and attached to the crista terminalis. It may be associated with patent foramen ovale and interatrial septal aneurysm.

cleft AV valve
*see* atrioventricular valve; *see also* atrial septal defect. *see also* ostium primum ASD.

coarctation of the aorta
A stenosis of the proximal descending aorta varying in anatomy, physiology and clinical presentation. It may present with 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 with “single”, which implies absence of corresponding contralateral structure. *see also* single.

common atrium
Large atrium characterized by a nonrestrictive communication between the bilateral atria due to the absence of most of the atrial septum. Frequently associated with complex congenital heart disease (isomerism, atrioventricular septal defect, etc.). *see also* single (atrium).

common arterial trunk
*see* truncus arteriosus.

complete transposition of the great arteries
*syn.* classic transposition; d-transposition; d-TGA; atrioventricular concordance with ventriculo-arterial discordance. An anomaly wherein the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle. The right ventricle supports the systemic circulation.
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 ventriculo-arterial connections
Appropriate origin of pulmonary trunk from morphologic right ventricle and of aorta from morphologic left ventricle. syn. ventriculo-arterial concordance.

conduit
A structure that connects non-adjacent parts of the cardiovascular system, allowing blood to flow between them. Often fashioned from prosthetic material. May include a valve.

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 (CHD)
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.

congenitally corrected transposition of the great arteries
syn. cc-TGA; l-transposition; l-TGA; atrioventricular discordance with ventriculo-arterial discordance; double discordance. An anomaly wherein the aorta arises from the right ventricle and the pulmonary artery from the left ventricle, and, in addition, the atrioventricular connection is discordant such that the right atrium connects to the left ventricle and the left atrium connects to the right ventricle. There are usually associated anomalies, the most common being ventricular septal defect, pulmonic stenosis, and/or a hypoplastic ventricle. The right ventricle supports the systemic circulation.

congenital pericardial defect
A defect in the pericardium due to defective formation of the pleuro-pericardial 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.

connection
Anatomic link between two structures (e.g. veno-atrial, atrioventricular, ventriculo-arterial).
cono-truncal abnormality
Neural crest cell migration is crucial for cono-truncal septation and the development of both the pulmonary and aortic outflow tracts. If neural crest cell migration fails, cono-truncal abnormalities occur. The most common cono-truncal anomalies are truncus arteriosus and interrupted aortic arch. Other defects may include tetralogy of Fallot, pulmonary atresia with ventricular septal defect, absent pulmonary valve or 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 CATCH 22.

conus
*see* infundibulum.

cor triatriatum sinister
A membrane divides the left atrium into an accessory pulmonary venous chamber and a left atrial chamber contiguous with the mitral valve. The pulmonary veins enter the accessory chamber. The connection between the accessory chamber and the true left atrium varies in size and may produce pulmonary venous obstruction.

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 (embryologic origin of the sinus venosus) that receives the venae cavae and (usually) the coronary sinus, and a trabeculated anterolateral “atrial” chamber (embryologic origin of 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 heart structures (e.g. hypoplastic tricuspid valve, tricuspid atresia, pulmonary stenosis or pulmonary atresia). A patent foramen ovale or an atrial septal defect are often present in relation to the posteromedial chamber.

When there is more extensive 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, which are extensively fenestrated remnants of the right sinus venosus valve. *see* sinus venosus.

criss-cross heart
*syn.* criss-cross atrioventricular connection. 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 supero-inferior relationship, the heart may
also be described as a supero-inferior or upstairs-downstairs heart. There may be ventriculo-arterial concordance or discordance.

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 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 atrial component receiving the inferior vena cava, the superior vena cava, and the coronary sinus. A feature of 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 due to increased peripheral extraction due, for instance, to critically reduced cutaneous flow (peripheral cyanosis).

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 operation
A procedure reserved for patients with abnormal ventriculo-arterial connections who are not suitable for an arterial switch operation (e.g. TGA and non-suitable coronary patterns, DORV with severe subaortic stenosis, systemic ventricular outflow tract obstruction in hearts with a univentricular AV connection). The 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. The aortic orifice and a VSD (if present) are closed with a patch. A conduit between the right ventricle and the distal pulmonary artery provides venous blood to the lungs. The procedure was described in 1975. (Damus PS. Correspondence. Annals of Thoracic Surgery 1975, 20, 724–725.) (Kaye MP. Anatomic correction of transposition of the great arteries. Mayo Clinic Proceedings 1975, 50, 638–640.)

**David operation**

A surgical procedure for ascending aortic aneurysm, involving replacement of the ascending aorta with a synthetic tube and remodeling of the aortic root so the preserved aortic valve is no longer regurgitant (David TE, Feindel CM. An aortic valve sparing operation for patients with aortic incompetence and aneurysm of the ascending aorta. *Journal of Thoracic and Cardiovascular Surgery* 1992, 103, 617–621.)

**dextrocardia**

Cardiac apex directed to the right of the midline. *see* cardiac position.

**dextroposition**

Rightward shift of the heart. *see* cardiac position.

**dextroversion**

An old term for dextrocardia. *see* cardiac position.

**differential hypoxemia; differential cyanosis**

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

**DiGeorge syndrome**

An autosomal dominant syndrome now known to be part of “CATCH 22”. As originally described, it consisted of infantile hypocalcemia, immunodeficiency due to thymic hypoplasia, and a cono-truncal cardiac abnormality. *see also* CATCH 22.

**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.

**discordant ventriculo-arterial 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.

**diverticulum of Kommerell**

Enlarged origin of the 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 anomaly.

**double-chambered RV**
Separation of the right ventricle (RV) into a higher-pressure infl ow chamber, and a lower pressure infundibular chamber, the separation usually being produced by hypertrophy of the “septomarginal band”. When a VSD is present, it usually communicates with the high pressure RV infl ow chamber.

**double discordance**
*see* congenitally corrected transposition of the great arteries.

**double inlet left ventricle (DILV)**
*see* 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 atrio-ventricular septal defect are commonly associated defects.

**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 is present. When the VSD is in the subaortic position without 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 (the 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 l-transposition of the great arteries (l-TGA; congenitally corrected transposition of the great arteries; cc-TGA) and also in patients who have had a prior Mustard or Senning atrial switch operation for complete transposition of the great arteries (d-TGA). 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 operation) in all cases, as well as an atrial switch procedure (Mustard or Senning) in the case of l-TGA, or reversal of the previously done Mustard or Senning procedure in the case of d-TGA.

doubly-committed VSD
see ventricular septal defect.

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

dural ectasia
Expansion of the dural sac in the lumbo-sacral area, seen on CT or MRI. It is one of the criteria used to confirm the diagnosis of Marfan syndrome. (Peryitz RE, et al. Dural ectasia is a common feature of the Marfan syndrome American Journal of Human Genetics 1988, 43, 726–732.) (Fattori R, et al. Importance of dural ectasia in phenotypic assessment of Marfan’s syndrome. Lancet 1999, 354, 910–913.)

Ebstein anomaly
An anomaly of the tricuspid valve in which the basal attachments of both the septal and the posterior valve leaflets are displaced apically within the right ventricle. Apical displacement of the septal tricuspid leaflet of >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. 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 VSD, pulmonary stenosis, and/or accessory conduction pathways.

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.
• Ehlers-Danlos types I, II and III, which demonstrate autosomal dominant inheritance, are the commonest forms, each representing about 30% of cases. The cardiovascular abnormalities are generally mild, consisting of mitral and tricuspid valve prolapse. Dilatation of major arteries, including the aorta, may occur. Aortic rupture is seen rarely in type I, but not in types II and III.

• Ehlers-Danlos syndrome type IV is also autosomal dominant, but frequently appears de novo. This is the “arterial” form, presenting with aortic dilatation and rupture of medium and large arteries spontaneously or after trauma. It is due to an abnormality of type III procollagen, and comprises about 10% of cases of Ehlers-Danlos syndrome.

• There are 6 other rare types of Ehlers-Danlos syndrome.

Eisenmenger syndrome
An extreme form of pulmonary vascular obstructive disease arising as a consequence of pre-existing systemic to pulmonary shunt, wherein pulmonary vascular resistance rises such that pulmonary pressures are at or near systemic levels and there is reversed (right-to-left) or bidirectional shunting at great vessel, ventricular, and/or atrial levels. see also Heath-Edwards classification. see also pulmonary hypertension.

Ellis-van Creveld syndrome
An autosomal recessive syndrome in which common atrium, primum ASD and partial AV septal defect are the most common cardiac lesions.

endocardial cushion defect
see atrioventricular septal defect. The term endocardial cushion defect has fallen into disuse because it implies an outdated concept of the morphogenesis of the atrioventricular septum.

erthrocytosis
Increase in red blood cell concentration secondary to chronic tissue hypoxia, as seen in cyanotic CHD 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, since 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 of the inferior vena cava to the right atrium.

extracardiac Fontan
see Fontan procedure.
fenestration
An opening, or “window” (usually small) between two structures, which may be spontaneous, traumatic, or created surgically.

fibrillin
Fibrillin is a large glycoprotein, closely involved with collagen in the structure of connective tissue. Mutations in the fibrillin gene on chromosome 15 are responsible for all manifestations of Marfan syndrome. see also Marfan syndrome.

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 leading to normalization of systemic oxygen saturation and elimination of volume overload of the functioning ventricle.

- 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 changed to a direct anastomosis between right atrium (RA) and pulmonary artery (PA).
- extracardiac Fontan. Inferior vena cava (IVC) blood is directed to the pulmonary artery via an extracardiac conduit. The superior vena cava (SVC) is anastomosed to the PA as in the bidirectional Glenn shunt.
- fenestrated Fontan. Surgical creation of an ASD in the atrial patch or baffle to provide an escape valve, allowing right-to-left shunting to reduce pressure in the systemic venous circuit, at the expense of systemic hypoxemia.
- lateral tunnel. see total cavopulmonary connection (TCPC).
- RA-RV Fontan. Conduit (often valved) between right atrium (RA) and right ventricle (RV). Also known as the Björk modification. (Björk VO, et al. Right atrial-ventricular anastomosis for correction of tricuspid atresia. Journal of Thoracic and Cardiovascular Surgery 1979, 77, 452–458.)
- total cavopulmonary connection (TCPC). IVC flow is directed by a baffle within the RA into the lower portion of the divided SVC or the right atrial appendage, which is connected to the pulmonary artery. The upper part of the SVC is connected to the superior aspect of the pulmonary artery as in the bidirectional Glenn procedure. The majority of the RA is excluded from the systemic venous circuit. syn. lateral tunnel Fontan.

Gerbode defect
An unusual variant of atroioventricular septal defect, wherein the defect is in the superior portion of the atroioventricular septum above the insertion of the septal leaflet of the tricuspid valve, resulting in a direct communication and shunt between the left ventricle and the right atrium. see also atroioventricular septum.
Ghent criteria
A set of criteria for the diagnosis of Marfan syndrome, requiring involvement of three organ systems (one system must have “major” involvement), or two organ systems and a positive family history. (DePaepe A, Deitz HC, Devereux RB, et al. Revised diagnostic criteria for the Marfan syndrome. American Journal of Medical Genetics 1996, 62, 417–426)

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

- classic Glenn. Anastomosis of the SVC to the distal end of the divided right PA with division/ligation of the SVC below the anastomosis. Acquired pulmonary arterio-venous 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. New England Journal of Medicine 1958, 259, 117–120.)


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GUCH

Heath-Edwards classification

- 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**
The first part of a “staged Fontan”, sometimes chosen to reduce the morbidity and mortality that might be associated with performing the complete Fontan at one operation. The hemi-Fontan includes a bidirectional cavopulmonary anastomosis and obliteration of central shunts. The second step to complete the Fontan procedure may be performed at a later time.

**hemi-truncus**
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.

**heterograft**
Transplanted tissue or organ from a different species.

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

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

**Holt-Oram syndrome**

**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, mental retardation, arterial hypertension and involvement of atrioventricular and semilunar valves resulting in valve regurgitation.

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

eytive 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); paresthesiae of fingers, toes or lips; tinnitus; fatigue, lassitude; myalgias (including chest, abdominal muscles), and muscle weakness. (Perloff JK, et al. Adults with cyanotic congenital heart disease: hematologic management. *Annals of Internal Medicine* 1988, 109, 406–413.) Restless legs or a sensation of cold legs may reflect hyperviscosity (observation of Dr E. Oechslin). As the symptoms are non-specific, 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
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.

Ilbawi procedure (operation)
An operation for congenitally corrected transposition of the great arteries with VSD and pulmonary stenosis, wherein 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. An atrial switch procedure is done. The LV then supports the systemic circulation. (Ilbawi MN, et al. An alternative approach to the surgical management of physiologically corrected transposition with ventricular septal defect and pulmonary stenosis or atresia. *Journal of Thoracic and Cardiovascular Surgery* 1990, 100, 410–415.)
infracristal
Located below the crista supraventricularis in the right ventricular outflow tract. see crista supraventricularis.

infundibular, infundibulum
(Pertaining to) a ventricular-great arterial connecting segment. Normally subpulmonary, but can be sub-aortic, and may be bilateral or absent. Bilateral infundibulum may be seen in patients with TGA/VSD/pulmonary stenosis (PS), DORV with VSD/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 below the hepatic veins with subsequent 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.

ISACCD
International Society for Adult Congenital Cardiac Disease. For information link through http://www.isaccd.org

isolation of arch vessels
see aortic arch anomalies.

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

• right isomerism. syn. asplenia syndrome. Congenital syndrome consisting of paired morphologically right structures: absence of spleen, bilateral right bronchi, bilateral tri-lobed (right) lungs, two morphologic right atria, and multiple anomalies of systemic and pulmonary venous connections and other complex cardiac and non-cardiac anomalies.

• 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, and complex congenital heart disease and other associated non-cardiac malformations.
Jatene procedure (operation)
syn. arterial switch procedure. An operation used in complete transposition of the great arteries, involving removal of the aorta from its attachment to the right ventricle, and of the pulmonary artery from the left ventricle, and the reattachment of the great arteries to the contralateral ventricles, with reimplantation of the coronary arteries into the neo-aorta. As a consequence, the left ventricle supports the systemic circulation. (Jatene AD, et al. Anatomic correction of transposition of the great vessels. *Journal of Thoracic and Cardiovascular Surgery* 1976, 72, 364–370.) see also Lecompte manoeuvre.

juxtaposition of atrial appendages
A rare anomaly seen in patients with transposition of the great arteries and other complex congenital heart defects (dextrocardia, tricuspid atresia, etc.), wherein 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.

Kartagener syndrome

Kommerell
see diverticulum of Kommerell.

Konno procedure (operation)
Lecompte manoeuvre
The pulmonary artery is brought anterior to the aorta during an arterial switch procedure in patients with d-transposition of the great arteries. see also Jatene procedure.

LEOPARD syndrome
This autosomal dominant condition includes Lentigines, EKG 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.

long-QT syndrome
Abnormal prolongation of QT-duration with subsequent risk for torsade de pointes, syncope and sudden cardiac death. It may be congenital or acquired (medications such as antiarrhythmics, antihistamines, some antibiotics; electrolyte disturbances such as hypocalcemia, hypomagnesemia, hypokalemia; hypothyroidism; and other factors.). QT-interval must be adjusted to heart rate.

looping
Bending of the primitive heart tube (normally to the right, dextro, d-) that 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.
maladie de Roger
Eponymous designation for a small restrictive ventricular septal defect that is not associated with significant left ventricular volume overload or elevated 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 aorto-pulmonary collateral artery. A large abnormal arterial vessel arising from the aorta, connecting to a pulmonary artery (usually in the pulmonary hilum) and providing blood supply to the lungs. Found in complex pulmonary atresia and other complex CHD associated with 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, protrusio acetabulae, and present with skeletal and cardiovascular abnormalities. Mitral valve prolapse with mitral regurgitation, ascending aortic dilatation/aneurysm with subsequent aortic regurgitation, and aortic dissection are the most common cardiovascular abnormalities. see also Ghent criteria.

mesocardia
Cardiac apex directed to mid-chest. see cardiac position.

mesoposition
Shift of the heart toward the midline. see cardiac position.

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.

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

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. 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.)

national referral center
see supraregional referral center (SRRC).

nonrestrictive VSD
see ventricular septal defect.

Noonan syndrome
An autosomal dominant syndrome phenotypically somewhat similar to Turner syndrome, with a normal chromosomal complement, due to an abnormality in chromosome 12q. It is associated with congenital cardiac anomalies, especially dysplastic pulmonic valve stenosis, pulmonary artery stenosis, ASD, tetralogy of Fallot, or hypertrophic cardiomyopathy. Congenital lymphedema is a common associated anomaly that may be unrecognized. (Noonan JA, Ehmke DA. Associated non-cardiac malformations in children with congenital heart disease. Midwest Society for Pediatric Research 1963, 63, 468.)

Norwood procedure
A multistage operation for hypoplastic left heart syndrome. A systemic to pulmonary arterial shunt is created, followed by a staged Fontan-type operation (usually via a hemi-Fontan procedure) resulting in single ventricle physiology. The morphologic right ventricle supports the systemic circulation.

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.

over-and-under ventricles
see supero-inferior heart.
overriding valve
An AV valve that empties into both ventricles or a semilunar valve that originates from both ventricles.

palliation, palliative operation
A procedure carried out for the purpose of relieving symptoms or ameliorating some of the adverse effects of an anomaly, which does not address the fundamental anatomic/physiologic disturbance. Contrasts with “repair” or “reparative operation”.

PAPVC
Partial anomalous pulmonary venous connection. see anomalous pulmonary venous connection.

parachute mitral valve
A mitral valve abnormality in which all chordae tendineae of the mitral valve, which may be shortened and thickened, insert in 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)
A ductus 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. Patent foramen ovale is found in up to 35% of the adult population in pathological studies. The lower and variable prevalence reported in clinical series depends on the techniques used to find it. syn: probe-patent foramen ovale, PFO.

pentology of Fallot
Tetralogy of Fallot with, in addition, an ASD or PFO. see tetralogy of Fallot.

perimembranous VSD
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 left superior vena cava. LSVC drains via the coronary sinus to the right atrium in more than 90% of patients. Rarely, it may directly drain to the left atrium in association with other congenital heart defects (e.g. 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) which may be offered to patients with cyanotic CHD and secondary erythrocytosis who are experiencing hyperviscosity symptoms. Concomitant volume replacement is usually indicated.

**pink tetralogy of Fallot**
*see* tetralogy of Fallot.

**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 with secondary erythrocytosis as seen in cyanotic heart disease. *see also* erythrocytosis.

**polysplenia syndrome**
*see* isomerism/left isomerism.

**Potts shunt**
A palliative operation for the purpose of increasing pulmonary blood flow, hence systemic oxygen saturation. The procedure involves creating a small communication between a pulmonary artery and the ipsilateral descending thoracic aorta. Often complicated by the development of pulmonary vascular obstructive disease if too large, or acquired stenosis or atresia of the pulmonary artery if distortion occurs. (Potts WJ, *et al.* Anastomosis of aorta to pulmonary artery: certain types of congenital heart disease. *Journal of the American Medical Association* 1946, 132, 627–631.)

**PPH**
Primary pulmonary hypertension. *see* pulmonary hypertension.

**probe-patent foramen ovale**
*see* patent foramen ovale.
protein-losing enteropathy (PLE)
A complication seen following the Fontan operation in which protein is lost via the gut, resulting in ascites, peripheral edema, pleural and pericardial effusions. It is of unknown cause, though exacerbated by high systemic venous pressure. If serum protein and albumin are low, increased alpha-1 antitrypsin in the stool supports the diagnosis of PLE.

protrusio acetabulae
Abnormal displacement of the head of the femur within the acetabulum. A radiological finding useful in the diagnosis of Marfan syndrome.

pseudotruncus arteriosus
Pulmonary atresia with a VSD, biventricular aorta, and pulmonary blood flow provided by systemic to pulmonary collaterals. This anatomic arrangement had previously been called “truncus arteriosus type IV” but is morphogenetically a different 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 aorto-pulmonary collateral arteries, not from anomalously connected true pulmonary arteries.

pulmonary artery banding
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 the underlying anomaly is not immediately advisable, e.g. in the setting of a non-restrictive VSD.

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 VSD (variant of tetralogy of Fallot), pulmonary blood flow arises from aorto-pulmonary collaterals, and systemic venous return exits the right heart via the VSD. When associated with intact interventricular septum, pulmonary artery blood supply is via a patent ductus arteriosus, and systemic venous return exits the right heart via an obligatory ASD.

pulmonary hypertension
Raised pulmonary arterial pressure. A common method to define the severity of pulmonary hypertension is the pulmonary/aortic systolic pressure ratio:
Rashkind procedure
A balloon atrial septostomy performed as a palliative procedure in children with d-TGA. (Rashkind WJ, Miller WW. Creation of an atrial septal defect without thoracotomy: a palliative approach to complete transposition of the great arteries. *Journal of the American Medical Association* 1966, **196**, 991–992.)

Rastelli procedure (operation)
An operation for repair of complete transposition of the great arteries in association with a large VSD and pulmonic stenosis, wherein 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-PA connection is obliterated. As a consequence, the left ventricle supports the systemic circulation. (Rastelli GC, *et al.* Anatomic correction of transposition of the great arteries with ventricular septal defect and subpulmonary stenosis. *Journal of Thoracic and Cardiovascular Surgery* 1969, **58**, 545–552.)

regional referral center (RRC)
A center for the care of adult patients with CHD, 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, e.g. after repair of tetralogy of Fallot. 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. *Cardiology in the Young* 1992, **2**, 382–386.)

restrictive VSD
*see* ventricular septal defect.

<table>
<thead>
<tr>
<th>Severity</th>
<th>Ratio</th>
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</thead>
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<tr>
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<td>0.3, &lt;0.6</td>
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<tr>
<td>moderate</td>
<td>0.6, &lt;0.9</td>
</tr>
<tr>
<td>severe</td>
<td>0.9 (Eisenmenger syndrome)</td>
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see aortic arch anomalies.

right ventricular dysplasia  
see Uhl anomaly.

Ross procedure  

rubella syndrome  
A wide spectrum of malformations caused by rubella infection early in pregnancy, including cataracts, retinopathy, deafness, congenital heart disease, bone lesions, mental retardation, etc. The spectrum of congenital heart lesions is wide and includes pulmonary artery stenosis, patent ductus arteriosus, tetralogy of Fallot, and ventricular septal defect.

Right ventricle (RV) infundibulum  
A normal connecting segment between the body of the RV and the pulmonary artery. *syn.* RV conus. *see also* infundibulum.

RVOTO  
Right ventricular outflow tract obstruction.

sail sound  
An auscultatory finding in some patients with Ebstein anomaly. The S₁ 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. The sail sound is not an ejection click, although it may simulate one.

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 (PA). 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 PA chest x-ray of the shadow formed by the anomalous pulmonary venous connection, which resembles a Turkish sword, or scimitar.

secondary erythrocytosis  
*see* erythrocytosis. *see also* polycythemia vera.
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 right atrial wall and interatrial septum. As a consequence, the right ventricle supports the systemic circulation. A type of “atrial switch” operation. see also Mustard procedure, atrial switch operation, double switch operation. (Senning A. Surgical correction of transposition of the great vessels. Surgery 1959, 45, 966–980.)

Shone complex (syndrome)
An association of multiple levels of left ventricular inflow and outflow obstruction (subvalvar and valvar LVOTO, coarctation of the aorta and mitral stenosis [parachute mitral valve and supramitral ring]). (Shone JD et al. The developmental complex of “parachute mitral valve”, supravalvular ring of left atrium, subaortic stenosis and coarctation of aorta. American Journal of Cardiology 1963, 11, 714–725.)

Shprintzen syndrome
see velo-cardio-facial syndrome. see CATCH 22.

shunt
Movement of blood through a congenitally abnormal or surgically created connection and communication between two circuits, at the level of the atria, ventricles, or great vessels. “Shunt” is a physiologic term, in contrast to “connection” which is an anatomic term.

single (as in atrium, ventricle, etc.)
Implies absence of the corresponding contralateral structure. Contrasts with “common”, which implies bilateral structures with absent septation. see also common.

sinus venosus
An embryologic structure, the anatomic precursor of the inferior vena cava, superior vena cava and 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, the embryologic precursor of the trabeculated or muscular portion of the right atrium, and includes the right atrial appendage, which 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 the oxygenated fetal blood coming from the placenta and via the inferior vena cava across the foramen ovale. see also cor triatriatum dexter, sinus venosus defect.
sinus venosus defect
A communication located postero-superior (or rarely postero-inferior) to the oval fossa, commonly associated with partial anomalous pulmonary venous connection (most often right pulmonary veins, especially the right upper pulmonary vein in association with a postero-superior defect), which 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 pulmonary veins and is not a defect in the interatrial septum. see also atrial septal defect

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

- situs ambiguous. Indeterminate sidedness (in the setting of atrial isomerism).
- situs inversus. Mirror-image sidedness, i.e. opposite of normal. Left-sided morphologic right atrium.
- situs inversus totalis. Total mirror-image sidedness. The position of all laterlized organs is inverted.
- situs solitus. Normal sidedness. Right-sided morphologic right atrium.

stent
Intravascular (intraluminal) prosthesis to scaffold a vessel following transluminal balloon dilatation, for the purpose of maintaining patency.

Sterling Edwards procedure

straddling AV valve
see atroventricular valve.

subpulmonary ventricle
The ventricle that relates most directly to the pulmonary artery.
supero-inferior heart
A term applied to a heart the ventricles of which are in a markedly supero-inferior relationship due to abnormal displacement of the ventricular mass along the horizontal plane of its long axis. Often coexists with criss-cross atrioventricular relationships. see also criss-cross 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 CHD comprising specialized resources, the availability of cardiology specialists with specific training and experience in ACHD, the availability of other cardiology sub-specialists and other medical and paramedical personnel with special training/experience in the problems of congenital heart disease, and offering opportunities for training, research and education in the field. syn. national referral center.

supravalvar mitral ring
An anomaly found in the left atrium that produces congenital mitral stenosis. see also cor triatriatum. see also Shone complex.

switch-conversion of transposition
An operation performed in patients with congenitally corrected transposition of the great arteries, or 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 ventricle. The first stage may involve pulmonary artery banding to induce pulmonary left ventricular hypertrophy. The second stage involves an arterial switch operation in both groups and a Mustard or Senning operation in patients with congenitally corrected transposition, or removal of the Mustard/Senning atrial baffles and reconstruction of an atrial septum in patients with complete TGA. see also double switch operation.

systemic AV valve
The atrioventricular valve guarding the inlet to the systemic ventricle.

TAPVC
Total anomalous pulmonary venous connection. see anomalous pulmonary venous connection.

TAPVD
Total anomalous pulmonary venous drainage. A term sometimes used to refer to the entity properly called total anomalous pulmonary venous connection. see anomalous pulmonary venous connection.
**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 in a subpulmonary position. Since the left ventricle empties across the VSD preferentially into the pulmonary artery, the physiology simulates complete transposition of the great arteries with a VSD.

**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 VSD. The other two components of the “tetralogy” are an overriding aorta and concentric right ventricular hypertrophy. Valvar 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) due to anterior deviation (hence the RVOTO). Lillehei first described the repair in 1955. (Lillehei CW, et al. Direct vision intracardiac surgical correction of the tetralogy of Fallot, pentalogy of Fallot, and pulmonary atresia defects; reports of first ten cases. *Annals of Surgery* 1955, 142, 418–445.)

- pentalogy of Fallot. Tetralogy of Fallot with an associated ASD or PFO.
- 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.

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

**total anomalous pulmonary venous connection (drainage, return)**
*see* anomalous pulmonary venous connection/total anomalous pulmonary venous connection.

**trabecular VSD**
*see* ventricular septal defect

**transannular**
Crossing the annulus. In connection with the RVOT 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, et al. Surgical treatment for tetralogy of Fallot by open intracardiac repair. *Journal of Thoracic Surgery* 1959, 37, 22–51.)

**transposition of the great arteries (TGA)**
*see* discordant ventriculo-arterial connections and see below
• simple TGA. 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 associated abnormality.

• 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.

tricuspid atresia
A congenital anomaly in which there is no physiologic or gross morphologic connection between the right atrium and right ventricle, and there is an inter-atrial connection allowing 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) arises from the base of the heart because of failure of proximal division into the aorta and the pulmonary artery. Thus, 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 VSD or an intact ventricular septum. syn. common arterial trunk.

Turner syndrome
A clinical syndrome due to the 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 drainage without ASD. The female phenotype varies with the age of presentation, and is somewhat similar to that of Noonan syndrome.

Uhl anomaly
Congenital malformation consisting of nearly total absence of the right ventricular myocardium, presenting with marked enlargement of both the right ventricle and right atrium and subsequent tricuspid regurgitation. Arrhythmogenic right ventricular cardiomyopathy may be one end of a spectrum and Uhl anomaly the other.

unbalanced AV canal
see ventricular imbalance.

unifocalization
A surgical technique that creates a common trunk for multiple direct aortopulmonary collateral arteries, 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.

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 (LA) in the AV groove, such that the coronary sinus drains into the LA. A form of absence of the coronary sinus.

upstairs-downstairs heart
see supero-inferior heart.

VACTERL association
Describes a spectrum of defects including vertebral abnormalities, anal atresia, tracheo-esophageal 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.).

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 CHD) or associated with other CHD malformations, such as tetralogy of Fallot. see aortic arch anomalies.

velo-cardio-facial 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 (cono-truncal anomalies, isolated VSD, tetralogy of Fallot). Due to microdeletion at chromosome 22q11. syn. Shprintzen syndrome. see also CATCH 22.

venous (or pulmonary) AV valve
The AV valve guarding the inlet to the venous, or pulmonary, ventricle.

ventricle repair
• 1-ventricle repair. see Fontan operation
• 1.5-ventricle repair (one and one-half ventricle repair). A term used to describe operations for cyanotic congenital heart disease performed when the pulmonary 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 pulmonary ventricle.
• 2-ventricle repair. A term used to describe operations for cyanotic congenital heart disease with common ventricle wherein functioning systemic and pulmonary 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.

- doubly-committed 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.
- inlet VSD. A defect in the lightly trabeculated inlet portion of the muscular interventricular septum, typically seen as part of an atrioventricular septal defect.
- muscular VSD. A defect entirely surrounded by muscular interventricular septum.
- 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 RVOTO.
- outlet VSD. A defect in the non-trabeculated 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.
- 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.
- restrictive VSD. A ventricular septal defect of small enough 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.
- trabecular VSD. A defect in the heavily trabeculated central or trabecular portion of the muscular interventricular septum. May be multiple.

**ventriculo-arterial concordance**
*see* concordant ventriculo-arterial connections.

**ventriculo-arterial discordance**
*see* discordant ventriculo-arterial connections.
**Waterston shunt**
A palliative operation for the purpose of increasing pulmonary blood flow, hence systemic oxygen saturation, which involves creating a small communication between the main pulmonary artery and the ascending aorta. Often complicated by the development of pulmonary vascular obstructive disease if too large. Not uncommonly caused distortion of the pulmonary artery. (Waterston DJ. Treatment of Fallot’s tetralogy in children under one year of age. *Rozhl Chir* 1962, 41, 181–183.)

**Williams syndrome**

**Wolff-Parkinson-White (WPW) syndrome**
Accessory lateral atrioventricular conduction pathway causing characteristic EKG 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. Typically, they have more than one accessory pathway.

**Wood unit**
A non-standard unit for expressing pulmonary vascular resistance (mmHg/L), named after Paul Wood, the famous British cardiologist. One Wood unit is equivalent to 80 dyn.cm.sec⁻⁵.

**xenograft**
Tissue or organ used for transplant, derived from another species. *syn.* Heterograft.

**Z-score, Z-value**
A way of expressing a physiologic variable in a form corrected for age and body size. Important in pediatrics. This is the number of standard deviations a measurement departs from mean normal. (Rimoldi HJA, *et al.* A note on the concept of normality and abnormality in quantitation of pathologic findings in congenital heart disease. *Pediatric Clinics of North America* 1963, 10, 589–591.) (Daubeney PEF, *et al.* Relationship of the dimension of cardiac structures to body size: an echocardiographic study in normal infants and children. *Cardiology in the Young* 1999, 9, 402–410.)
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