4 Basic Nomenclature in Adult Congenital Heart Disease

The development of multidetector computed tomography has resulted in marked advancements in the diagnosis of congenital heart disease (CHD) which has renewed interest in the classifications and definitions used to describe the anatomy of the heart and great vessels. The sequential, segmental approach to analyzing CHD patient images was introduced nearly three decades ago and is used worldwide [1–5]. Thus, a thorough understanding of both the nomenclature and anatomy applied in this approach is essential to the proper and error-free interpretation of CHD CTA images [6]. The definitions described below are based on the segmental, sequential approach to classifying CHD which is described in Chap. 5.

4.1 Cardiac Orientation

Cardiac orientation or position refers to the relationship or axis of the base to the apex of the heart and may help to predict the presence of CHD. Early in fetal development, normal embryogenesis [situs solitus (normal atrial positions) and D-bulboventricular loop (normal ventricular positions)], results in the apex of the heart being situated in the right hemithorax. At the beginning of the second month, the apex of the heart migrates to the left hemithorax (normal adult position). In situs inversus with an L-loop (atria are reversed and ventricles are reversed), the opposite occurs (the apex of the heart migrates from the left hemithorax to the right hemithorax). Regardless of atrial situs, all D-loops should complete their development with the heart in the left chest (levocardia). On the contrary, all L-loops should end development with the heart in the right chest (dextrocardia). If the cardiac apex fails to shift, it may result in situs solitus with dextrocardia which is termed dextroversion or situs inversus with levocardia called levoversion. Thus, dextrocardia may occur from a D-loop that fails to shift leftward or an L-loop that completes its rightward shift. Mesocardia (midline heart) occurs when the ventricular apex does not complete its shift. Mesocardia may be associated with concordant (D-loop) or discordant (L-loop) ventricles as well as with heterotaxy syndromes.

Of note, positioning of the heart in the right chest with a left-sided cardiac apex can occur when the contents of the left chest force the heart to the right or when the volume of the right lung is reduced (for instance, due to pulmonary hypoplasia or collapse). This type of positioning is more appropriately termed dextroposition since the axis of the heart is usually normal.

To summarize, three different cardiac situs abnormalities are possible: levocardia, dextrocardia, and mesocardia (Fig. 4.1). Levocardia is defined as a normal cardiac position in the left chest with the cardiac base-to-apex axis pointing from upper right to lower left (normal cardiac base-to-apex axis points left to right). Dextrocardia refers to a heart located in the right chest with the base-to-apex axis pointing from the upper left to the lower right. Mesocardia refers to a heart that is usually in the midline with the base-to-apex axis directly from superior to inferior. Situs solitus with dextrocardia is termed dextroversion and situs inversus with levocardia is called levoversion.

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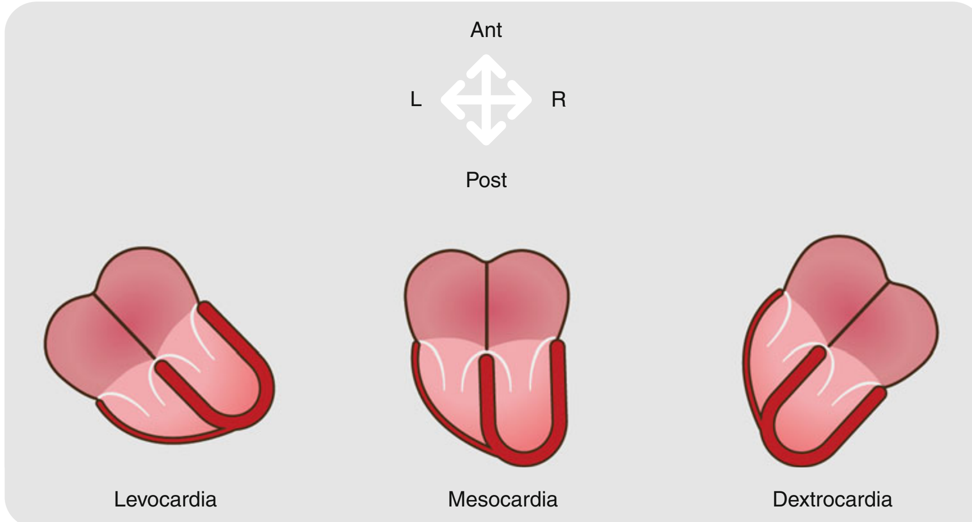


Fig. 4.1 A depiction of the three possible cardiac situs abnormalities

4.2 Nomenclature for Thoracic and Visceral Situs

Situs or sidedness refers to the position or arrangement of structures or organs that are not bilaterally symmetric. There are three possible arrangements: normal also known as solitus, inversus (mirror image of normal), and ambiguous (not clearly solitus or inversus). In the ambiguous situation, features of situs solitus and situs inversus are present in the same person. Here the thoracic and abdominal organs cannot be lateralized and have neither the normal nor mirror image arrangement.

4.2.1 Situs Definitions

Thoracic Situs: The situs of the left and right lung is independent of the cardiac or abdominal situs but instead is identified by the anatomy of the respective bronchi, especially the relationship between the bronchi and pulmonary arteries [7]. The right main bronchus takes a more vertical course and branches at an earlier point than the more horizontally oriented left bronchus. See Fig. 4.2.

Abdominal Situs: Refers to the sidedness of the main visceral organs such as the liver, stomach, and spleen.

Atriovisceral Situs: Atrial situs is determined by the position of the morphologic right and left atria. Atrial development is simultaneous to venous return formation, and thus, the atrial locations are fixed early in development by the entering veins. The atrial situs is independently determined from and unaffected by the shape of the bulboventricular loop or the final locations of the ventricles. In turn, the atrial situs corresponds to the visceral and thoracic situs and abnormalities in atrial position usually correspond with parallel abnormalities in the visceral and thoracic situs.

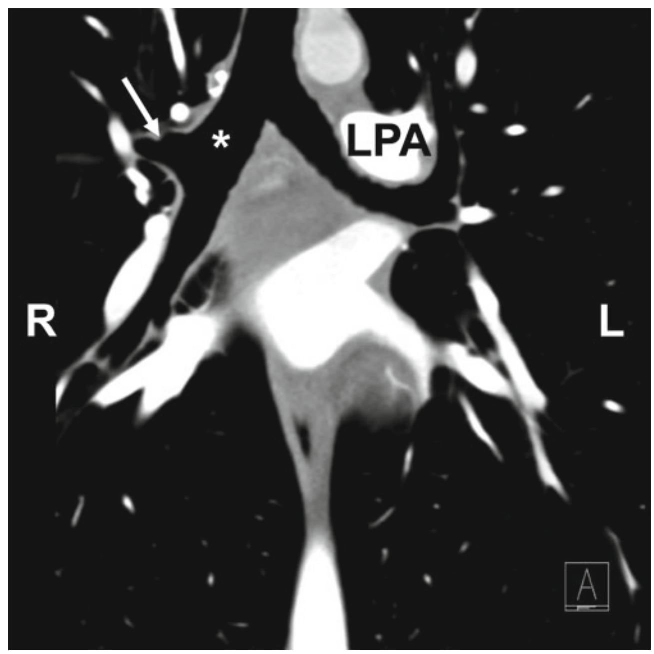


Fig. 4.2 A coronal CT image demonstrating the normal central bronchial anatomy which can be used to help differentiate the morphologic right lung from the morphologic left lung. The right main stem bronchus (*asterisk*) is more vertically oriented with an early branch point (*white arrow*), *LPA* left pulmonary artery

Three designations are used in characterizing atrial situs: situs solitus, situs inversus, or situs ambiguous. Situs solitus (normal) and situs inversus are present when there is identifiable lateralization of the atrial chambers, meaning that atrial arrangement is congruent with thoracoabdominal organ arrangement [8, 9]. Situs solitus is present when the right atrium and liver are on the right side and the left atrium, stomach, and spleen are on the left side. The right lung is trilobed and the left lung is bilobed each with their usual

Table 4.1 The characteristics of left and right isomerism

Right isomerism	Left isomerism
Bilateral morphologic right atria	Bilateral morphologic left atria
Asplenia	Bilateral bilobed lungs
Bilateral trilobed lungs	Interrupted inferior vena cava
Symmetric liver	Partial anomalous pulmonary venous return
Total anomalous pulmonary venous return	

bronchial pattern. The right pulmonary artery lies anterior to the right bronchus (eparterial position), and the left bronchus lies behind the left pulmonary artery (hyparterial position). Situs inversus is a mirror image of situs solitus.

It should be noted that atrial situs most often follows the thoracic and visceral situs. That is, atrial situs inversus is most often concomitant with thoracic and visceral situs inversus.

Ambiguous situs indicates that assignment or position of the right and left atria cannot be determined. Ambiguous situs may also be termed heterotaxy. Atrial situs ambiguity usually occurs concomitantly with visceral heterotaxia or situs ambiguity where the visceral organs are abnormally positioned across the left–right axis of the body. The term "isomerism" has been used to describe the combination of atrial situs ambiguity (heterotaxy) and visceral heterotaxy [8, 9]. Two forms of isomerism are recognized: right isomerism and left isomerism. Right isomerism is when there are bilateral morphologic right atria and is associated with asplenia and left isomerism (bilateral morphologic left atria) is associated with polysplenia. Right isomerism is also often associated with bilateral trilobed lungs (two right lungs, a symmetric liver across the midline, and total anomalous pulmonary venous return). Left isomerism is usually associated with bilateral bilobed lungs (two left lungs), interrupted inferior vena cava, and partial anomalous pulmonary venous return.

Note that the aorta and great veins generally have specific orientations depending on the situs and type of isomerism discussed in detail in Chap. 5. See Fig. 5.3.

See Table 4.1 for a characterization of isomerization.

Figure 4.3 illustrates the thoracic, atrial, and visceral situs seen with situs solitus, situs inversus, and situs ambiguous (right and left isomerism).

Atrial situs and ventricular looping are independent. Thus, any version of atrial situs may accompany a D-looped (correct ventricular sidedness) or L-looped (incorrect ventricular sidedness) heart. Figure 4.3 demonstrates this concept.

4.2.2 Definitions for the Atrial Chambers

Anatomically, atrial chamber differentiation is usually based on the distinctive features of the atrial appendages. Each appendage has intrinsic morphologic features. Typically, the right atrial appendage is a triangular, broad-based structure, while the left atrial appendage is smaller, thinner, and finger-like. The right atrium may be identified by locating the crista terminalis, a muscular band that runs from the entrance of the superior vena cava to that of the inferior vena cava. This structure separates the trabeculated right atrial appendage from the remainder of the right atrium. The left atrium lacks the crista terminalis. The thicker superior limbus of the atrial septum around the fossa ovalis is associated with the right atrium. The flap of the valve at the oval fossa is located in the left atrium. Additionally, the identification of inferior vena cava and the coronary sinus entering the right atrium is used as a criterion for identification.

The superior vena cava (SVC) is not an acceptable criterion to identify the morphologic right atrium since an anomalous SVC is not uncommon. Similarly, pulmonary vein anomalies are also commonly seen, and as such, identifying pulmonary venous connections is not a stern rule for identifying the morphologic left atrium.

Table 4.2 identifies the distinctive features differentiating the right from the left atrium.

4.2.3 Definitions for Cardiac Ventricles

The ventricular cardiac chambers are identified by their associated respective inlet valves. That is, the left ventricle is associated with the mitral (bileaflet) inlet valve (arises more superior than the tricuspid valve), and the right ventricle is associated with the tricuspid (trileaflet) inlet valve which arises below the membranous septum. Ventricles are composed of three parts: the inlet, outlet, and trabecular portions. The inlet contains the atrioventricular valves and the subvalvular apparatuses. The ventricular outlet portions are cephalad and lead to the great arteries. The trabecular portion extends from the papillary muscles to the ventricular apices.

There are several characteristics which are specific for the right ventricle and are useful in distinguishing the right from the left ventricle. These are the (a) infundibulum, (b) tricuspid valve apparatus, (c) apical trabeculations, and (d) moderator band. The infundibulum is a term that indicates the right ventricular outlet, which is a smooth muscular structure (also known as the conus or muscular conus). The left ventricular outlet is partially fibrous due to the aortic-mitral fibrous continuity.

The tricuspid valve apparatus has three leaflets and three papillary muscles; the papillary muscles are attached to both the interventricular septum and the free wall of the right ventricle. The mitral valve apparatus consists of an annulus and two leaflets, which connect to two papillary muscles via cord-like tendons called chordae tendineae. The papillary muscles insert only on the free lateral wall of the left ventricle

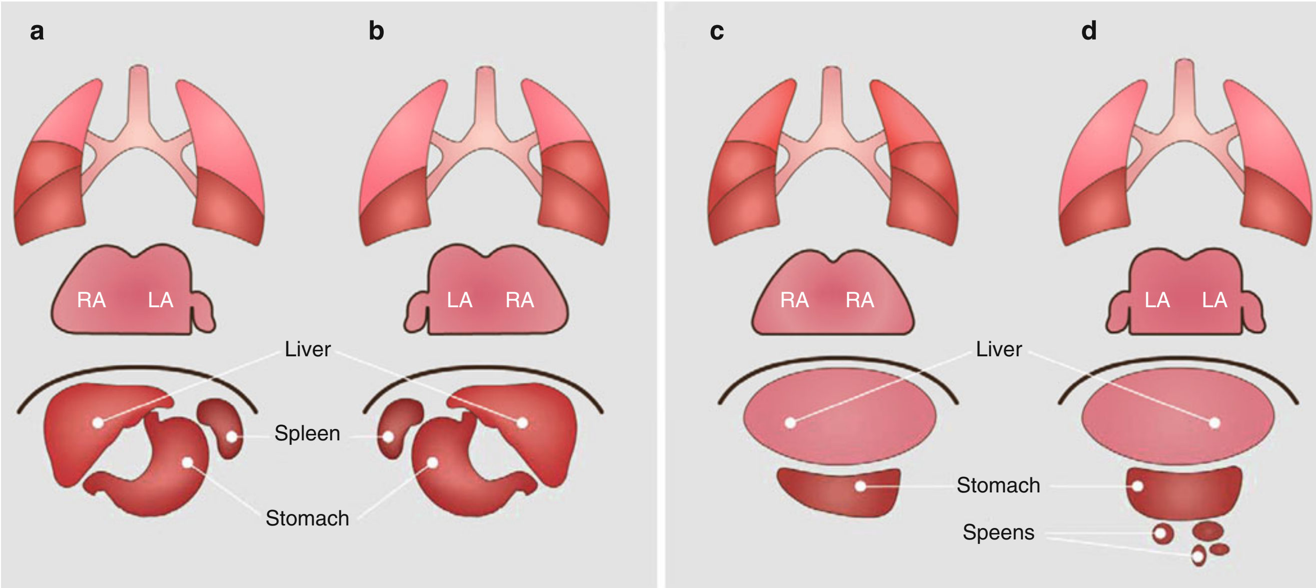


Fig. 4.3 The correspondence of atrial, thoracic, and abdominal organs arrangement in situs solitus (**a**), situs inversus (**b**), right isomerism (**c**), and left isomerism (**d**). Note in panel (a) the trilobed right lung is rightward, the bilobed left lung is leftward, and the atria, liver, spleen, and stomach are on their correct sides. In panel (b) the lungs, atria, and visceral organs are reversed, mirror image. Panel (c) illustrates bilateral right lungs, bilateral morphologic right atria, and heterotaxy of the visceral organs. Panel (d) shows bilateral left lungs, bilateral morphologic left atria, and visceral heterotaxy with polysplenia

Table 4.2 The characteristics that differentiate the right from the left atria

Atrial differentiation	
Right atrium	Left atrium
Triangular broad appendage	Narrow based, thinner, and finger-like appendage
Contains crista terminalis	No crista terminalis
Thick superior limbus around fossa ovalis	Contains fossa ovalis flap
Inferior vena cava and coronary sinus connection	

Table 4.3 The characteristics that differentiate the right from the left ventricle

Ventricular differentiation	
Right ventricle	Left ventricle
Presence of the infundibulum	Partially fibrous ventricular outlet
Associated with the tricuspid valve and tricuspid valve apparatus	Associated with the mitral valve and mitral valve apparatus
Course trabeculations	Fine trabeculations
Presence of the moderator band	Absence of moderator band

(not on the septum). Again, the tricuspid valve is identified by its caudal location below the membranous septum.

The characteristics of the internal trabeculae of the ventricles also help to differentiate between the right and left ventricles. The right ventricular trabeculations are coarse; the left ventricular trabeculations are thinner, delicate structures. The moderator band is another distinguishing feature of the right ventricle. It extends from the septum to the free lateral wall of the right ventricle and contains part of the right bundle branch of the cardiac conduction system, which plays a role in the electrophysiologic conduction of the right ventricle's free wall.

Table 4.3 identifies the distinguishing characteristics of the right and left ventricles.

As discussed in Chap. 1, the ventricles may form according to a D-loop (normal) or L-loop (reversed) pattern. Because cardiac looping is independent of visceral situs development, both D-loop ventricles and L-loop ventricles may be concordant or discordant relative to visceroatrial situs. Concordant signifies a D-loop with situs solitus or L-loop with situs inversus. Discordant means D-loop with situs inversus or L-loop with situs solitus. In the case of ambiguous situs, concordance or discordance cannot be determined.

4.2.4 Definitions for Great Arteries

The great vessels are typically described by the terms solitus, inversus, dextro, and levo. Solitus refers to the normal anatomically relationship between the great vessels. Inversus refers to the mirror image anatomic relationship. Dextro describes great vessels on the right side of the body and levo indicates the great vessels are on the left.

The great arteries are most easily identified by their branches and not by their relationship to the ventriculoarterial valves. The left aortic arch (normal) typically gives rise to a brachiocephalic artery, left common carotid artery, and left subclavian artery. The pulmonary artery typically bifurcates into left and right pulmonary arteries. The coronary arteries predominantly arise from the sinuses of the aorta. The location of the conus can help to identify the pulmonary artery. Typically, the muscular conus is subpulmonic in location.

A common arterial trunk is defined as a vessel connected to the ventricle (or ventricles) via a common ventriculoarterial valve. The common trunk supplies the coronary, systemic, and pulmonary circulations directly. A solitary arterial trunk is defined as a vessel arising from ventricle or ventricles that does not give rise to intrapericardial pulmonary arteries. In this anomaly, the blood supply to the lung usually comes from collateral vessels originating from either the ascending or descending thoracic aorta.

4.3 Nomenclature to Describe Connecting Segments

A segment is the term used to describe a part or section of the cardiovascular system. A connection is the term that describes the junction between two cardiovascular segments. Again, note that atrial situs, great artery orientation, and ventricular looping do not specify atrioventricular connections. All of these variables are independent of each other.

4.3.1 Nomenclature for Atrioventricular Connections

There are five types of atrioventricular connection: concordant (normal), discordant, ambiguous, double-inlet, and absent connection. Concordant refers to a normal connection between segments. The right atrium connects to the morphologic right ventricle and the left atrium connects to the morphologic left ventricle. Discordant refers to the opposite of the normal connection. The right atrium connects to the morphologic left ventricle and the left atrium drains into the morphologic right ventricle. Ambiguous connection refers to connections in which half the atrioventricular junction is connected concordantly and the other half is discordantly connected. That is, there may be bilateral morphologic right or left atria. For example, one of the atria may correctly connect with its concordant ventricle and the opposite-sided morphologically identical atrium discordantly connects with the opposite ventricle. Concordant, discordant, and ambiguous connections occur in a biventricular heart. See Fig. 4.4.

In functionally univentricular hearts, there are three possible inlets: double inlet, single inlet, or common inlet. Double inlet refers to a functional univentricle connected to two separate atria with two separate atrioventicular valves. Single inlet refers to two separate atria with only one of the atria connected to the functional univentricle via one atrioventricular valve. The other atria connection is atretic. Single inlet may be either a right single inlet where the left-sided atrioventricular connection is atretic or left single inlet where the right-sided atrioventricular connection is atretic. A common inlet refers to when both atria are connected to a functional univentricle via one atrioventricular valve.

Figure 4.5 depicts an artist's rendition of the possible univentricular inlets.

4.3.2 Nomenclature for Atrioventricular Valvular Connections

Straddling is a feature of the chordae tendineae of an atrioventricular valve and describes chordae that cross a ventricular septal defect and have their attachments in the opposite ventricle. Overriding is a feature of the valve annulus which describes an annulus that crosses a ventricular septal defect and thus lies "over" more than one ventricle. Straddling and overriding often coexist. Figure 4.6 depicts the concepts of straddling and overriding.

4.3.3 Nomenclature for Ventriculoarterial Connections

There are four potential types of ventriculoarterial connection: concordant, discordant, double-outlet right ventricle, and double-outlet left ventricle. Similar to the atrioventricular connection, concordant refers to a normal connection between segments. The pulmonary artery arises from the right ventricle and the aorta from the left ventricle. Discordant refers to the opposite type of connection. The aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle, which is seen in transposition of the great vessels. Double-outlet right ventricle means both great vessels originate from the right ventricle, and double-outlet left ventricle means the great vessels arise from the left ventricle. In both settings, only one valve, either the aortic or pulmonic valve, can be identified. Figure 4.7 depicts the possible ventriculoarterial connections.

Ventriculoarterial overriding refers to a situation when more than half of the area of the outlet overrides the ventricular septum. It is commonly seen in tetralogy of Fallot, where the aorta overrides the ventricular septum.

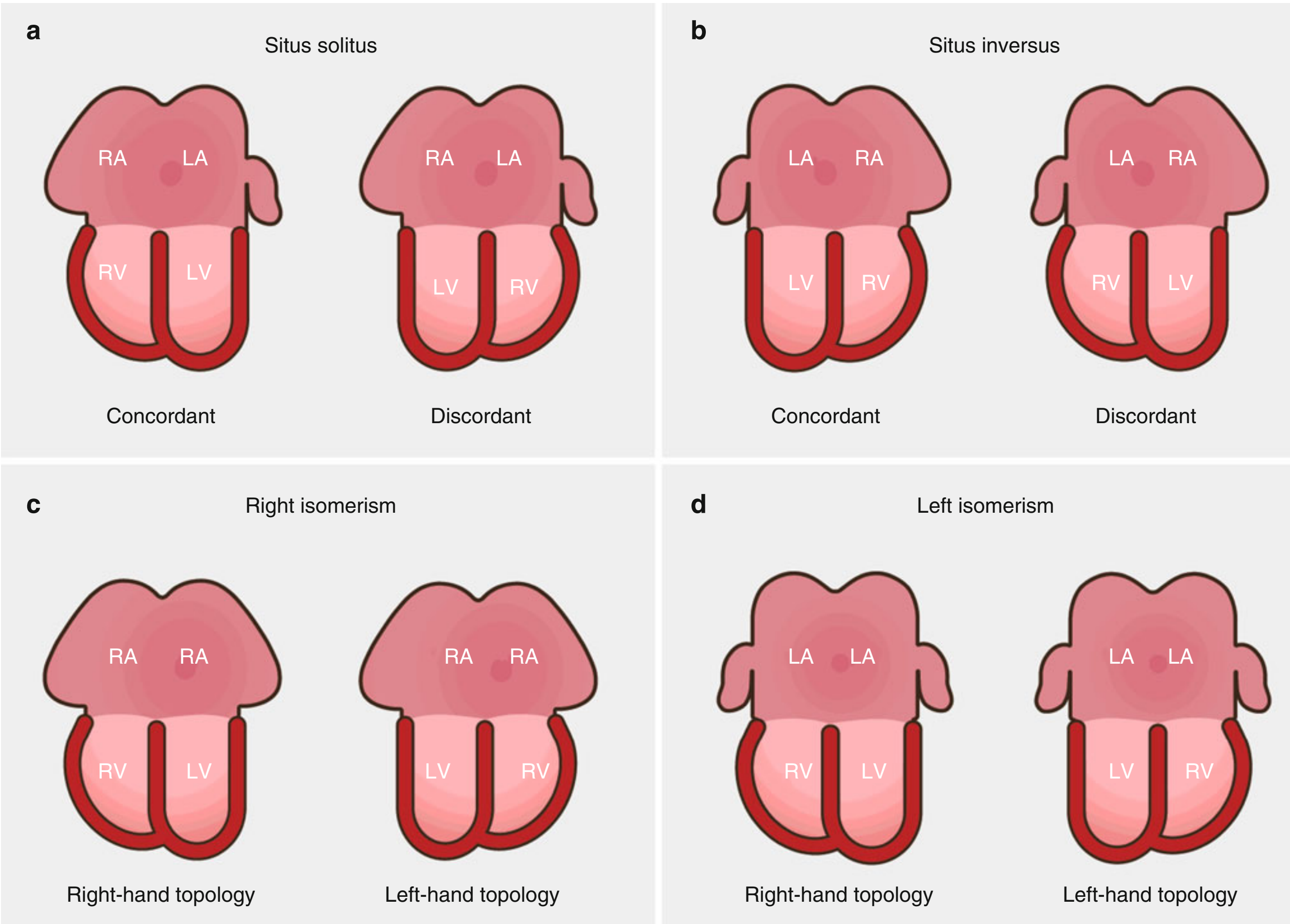
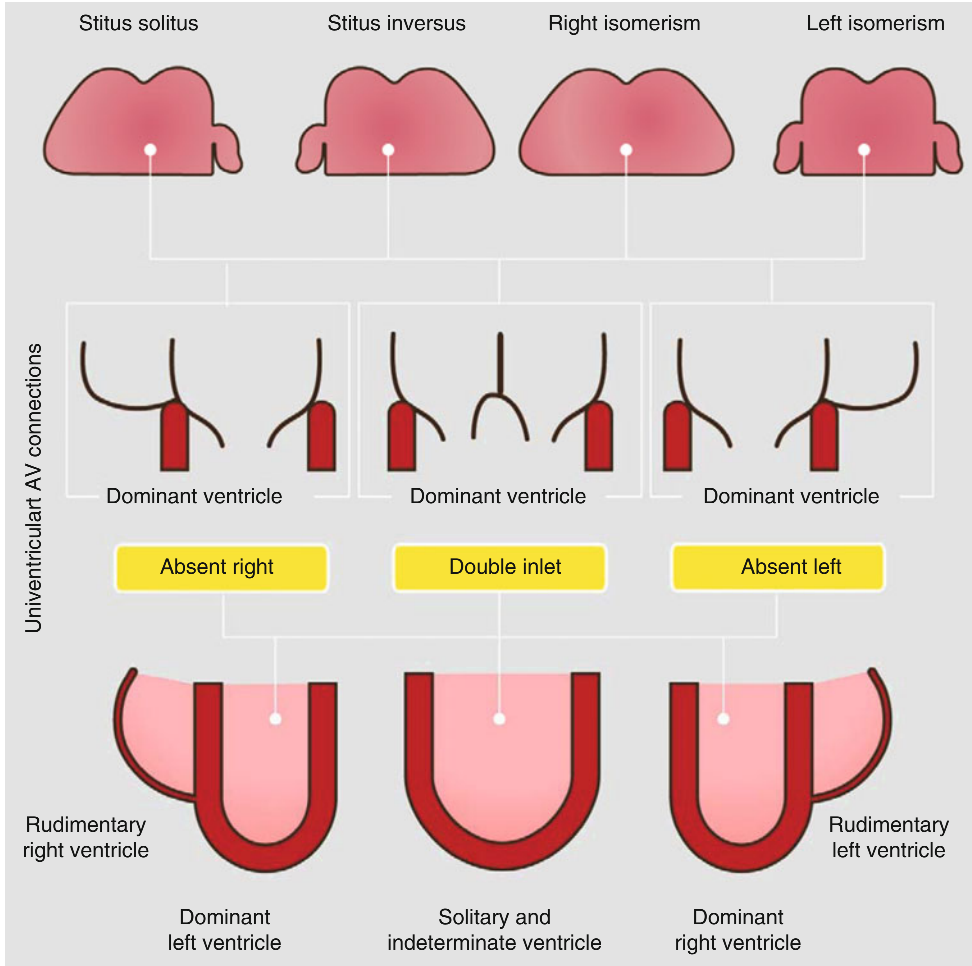


Fig. 4.4 The variants of biventricular atrioventricular connections in situs solitus (**a**), situs inversus (**b**), right isomerism (**c**), and left isomerism (**d**) demonstrating the independence of situs and looping. In panels (a) and (b), concordant demonstrates normal ventricular sidedness (D-looping), while discordant demonstrates opposite ventricular sidedness (L-looping). In panels (c) and (d), right-handed topology refers to D-looping and left-handed topology refers to L-looping

Fig. 4.5 The variants of univentricular atrioventricular (*AV*) connections in situs solitus, situs inversus, and right and left isomerism



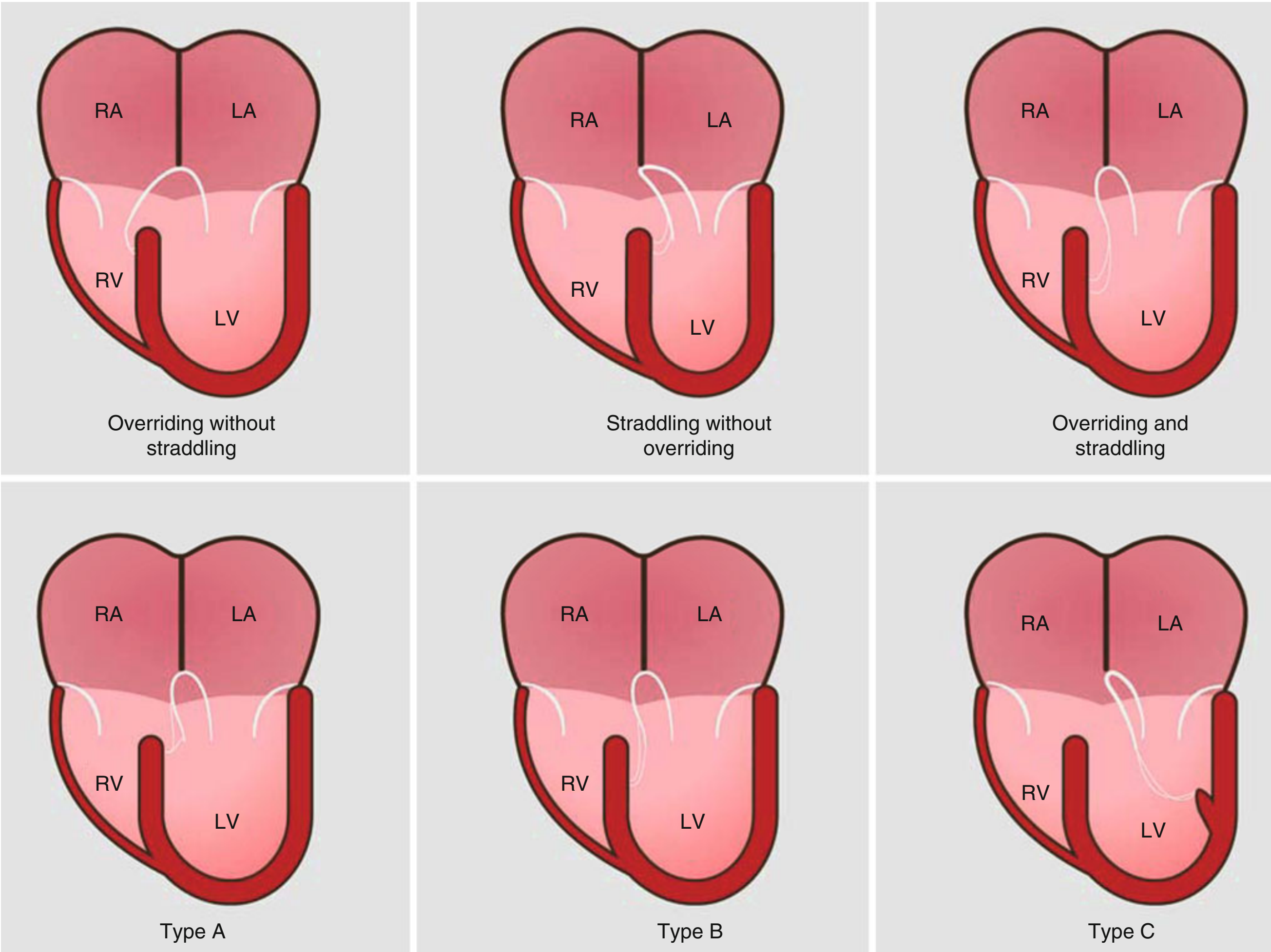


Fig. 4.6 An artist's depiction of the concepts of straddling and overriding

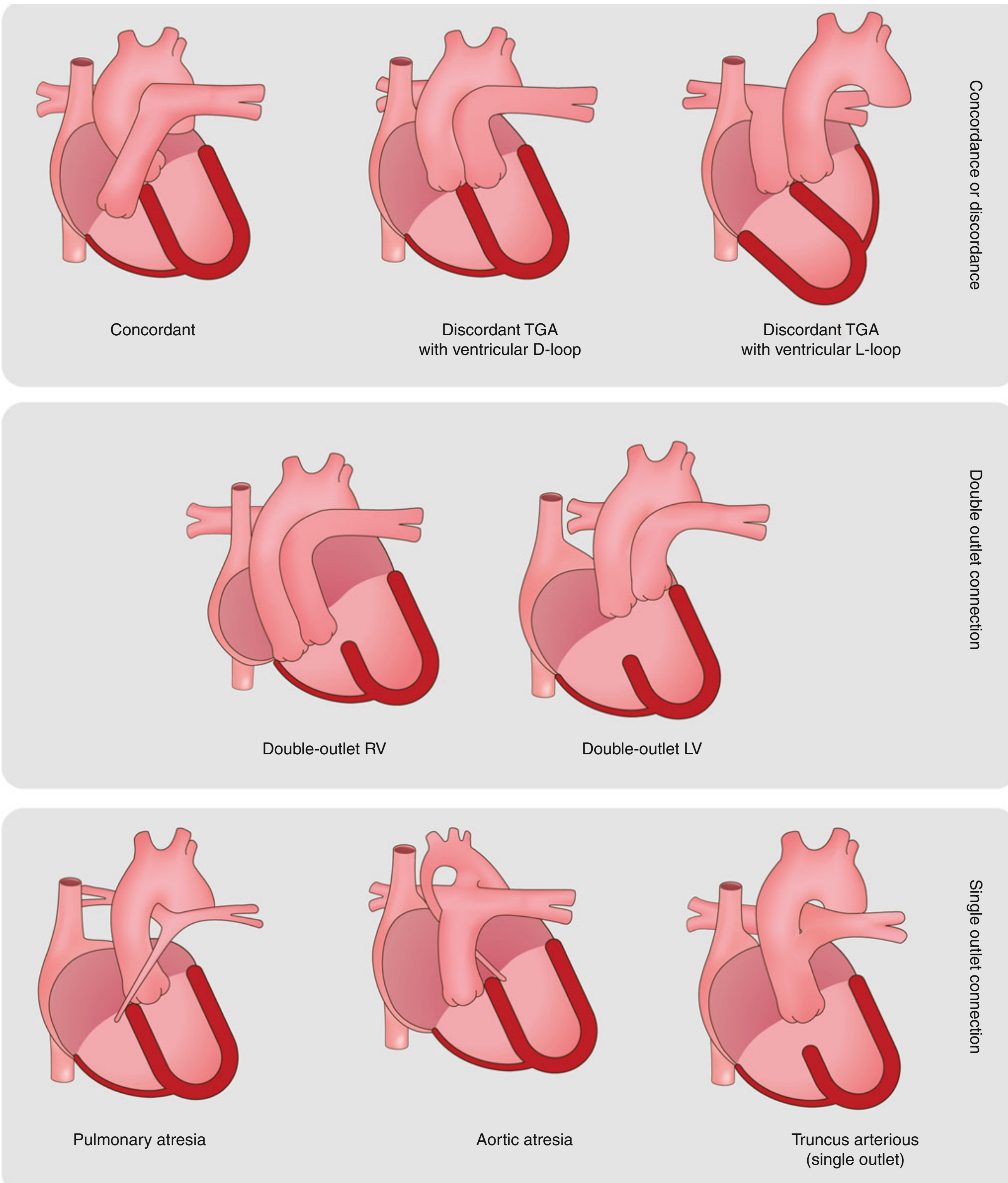


Fig. 4.7 A depiction of the possible ventriculoarterial connections

4.4 Van Praagh Notation System

The nomenclature system used in the segmental, sequential approach developed by Van Praagh also includes a series of three letters, separated by commas, within parentheses which are used for conveying anatomic positioning. The first initial denotes the visceroarterial situs (S: solitus, I: inversus). The second initial marks the ventricular looping pattern (D: D-looping, L: L-looping). The third initial conveys the great artery positioning (S: solitus (normal)), I: inversus (with no transposition), D-TGA (D-transposition of the great arteries), L-TGA (L-transposition of the great arteries), D-MGA (D-malposition of the great arteries), L-MGA (L-malposition of the great arteries), A-TGA (ambiguous transposition of the great arteries), and finally A-MGA (ambiguous malposition of the great arteries) (Fig. 4.8). Thus, (S,D,S) would indicate the normal anatomy (solitus, D-looping, solitus), whereas (I, L, I) would indicate atrial situs inversus, ventricular L-looping, and inversus of the great vessels and (S,L,L-TGA) indicates atrial situs solitus, ventricular L-looping, and L-TGA.

Figure 4.9 depicts the various types of human hearts emphasizing the Van Praagh notation system [10].

Note that TGA is only one form of malposition. Malposition of the great arteries may also include double-outlet right ventricle, double-outlet left ventricle, and anatomically corrected malposition.

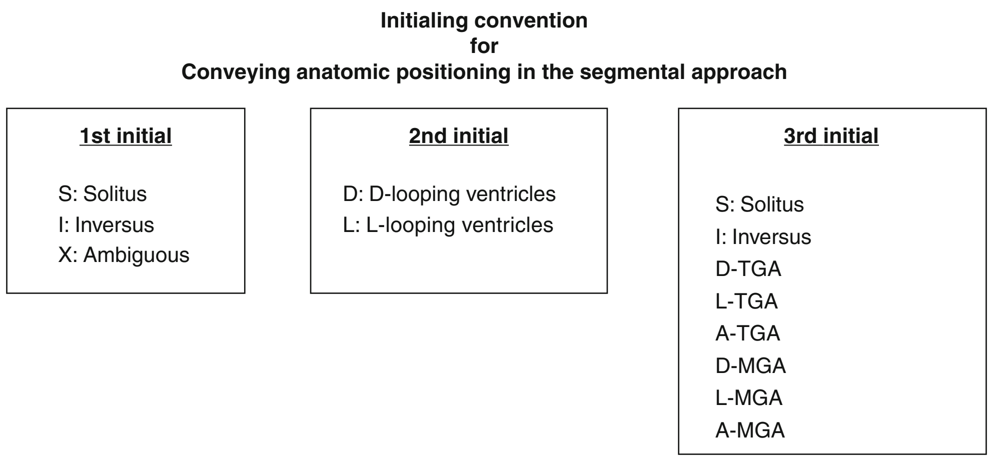


Fig. 4.8 The initialing system used in the labeling of congenital heart disease patterns. The first initial is the atrial situs (*S* for solitus, *I* for inversus, *X* for ambiguous). The second initial involves the ventricular looping pattern (*D* for D-loop, *L* for L-loop) and the third letter is the spatial orientation of the great vessels (*S* for solitus, *I* for inversus, *D-TGA* for D-transposition of the great vessels, *L-TGA* for L-transposition of the great vessels, *A-TGA* for ambiguous transposition of the great vessels, *D-MGA* for D-malposition of the great vessels, *L-MGA* for L-malposition of the great vessels, *A-MGA* for ambiguous transposition of the great vessels)

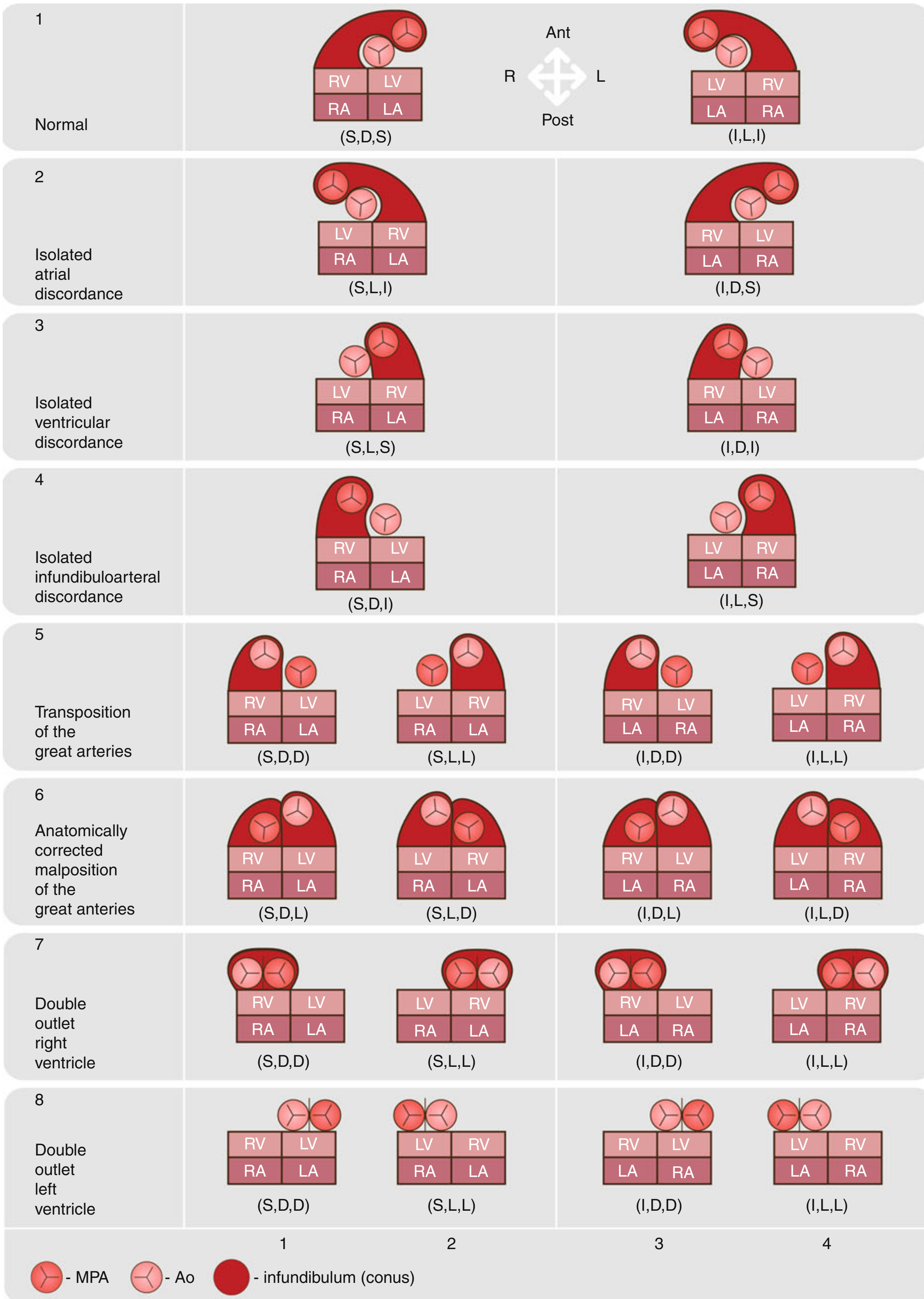


Fig. 4.9 Depicts the various types of human hearts emphasizing the Van Praagh notation system (Reproduced with the kind permission of Ronald B. Foran, MD, Pediatric Cardiology Associates, Rockford Il)

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