Sequential Segmental approach for Imaging of congenital heart disease by MSCT

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Contents

Introduction

Congenital heart diseases are still a major cardiac problem in the pediatric age group. Echocardiography remains a first-line non-invasive imaging tool for establishing the diagnosis and follow-up in most patients (**Khatri S et al., 2008).**

Diagnostic imaging is crucial in the evaluation of thoracic congenital anomalies in pediatric patients. Although clinical assessment may provide insight into the possible diagnosis, imaging is usually necessary to confirm the diagnosis, treatment planning and postoperative evaluation (**Goo Hyun 2011)**.

Imaging of CHD is performed for evaluation of anatomy and function prior to surgery/intervention and following the procedure for evaluation of complications as well as assessment for re-interventions. Several imaging modalities, each with advantages and disadvantages, are deployed in the evaluation of CHD. **Echocardiography** is the most commonly used modality due to its wide availability, portability, low cost, and ability to accurately evaluate cardiac anatomy and function. Limitations include operator dependence, limited acoustic window in large patients or COPD/chest wall deformities, limited field-of-view for evaluating extra cardiac vascular structures (aorta, peripheral pulmonary arteries, veins, cavopulmonary shunts) and limited quantification of right/single ventricular function **(Orwat S, et al., 2013).**

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 (**(Lapierre C et al., 2010).**

Computed tomography (CT) has several advantages in the evaluation of CHD, including high spatial resolution, good temporal resolution, and isotropic multi-planar reconstruction. The latest scanners are capable of scanning the entire heart within a single heartbeat at high temporal resolution, thus obviating the need for sedation/general anesthesia that was required with the older scanners **(Kelly Han B et al., 2015).**

Due to the above mentioned advantages, CT is a generally accepted robust modality in the evaluation of CHD, both in adults and children, before as well as after surgery/intervention. Prior to surgery/intervention, CT provides a comprehensive mapping of the anatomy of these complex anomalies especially that of extracardiac structures such as aorta, systemic veins, pulmonary veins, and pulmonary arteries, beyond what is possible with echocardiography and hence is a good complement to echocardiography. Ventricular volumes and functions can be quantified with high accuracy, particularly for the right ventricle and single ventricle. Evaluation of ventricular and Valvular function with CT is useful in patients with contraindications to MRI or those patients who have devices/implants which produce artifacts in MRI. The accuracy is comparable to MRI **(Kelly Han B et al., 2015).**

This review article describes the sequential steps required for accurate and complete analysis of complex congenital heart disease during a review of any imaging study. By following these steps, the reader of cardiac computed tomographic angiography studies, will more likely end with an accurate interpretation.

Chapter 1

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 five decades ago and is used worldwide (**Lapierre C et al., 2010).**

The definitions described below are based on the segmental, sequential approach to classifying CHD which is described in the next Chapter.

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) (**Van Praagh R. 1977).**

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 (**Van Praagh R**. **1977)**.

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 **(Van Praagh R**.**1977).**

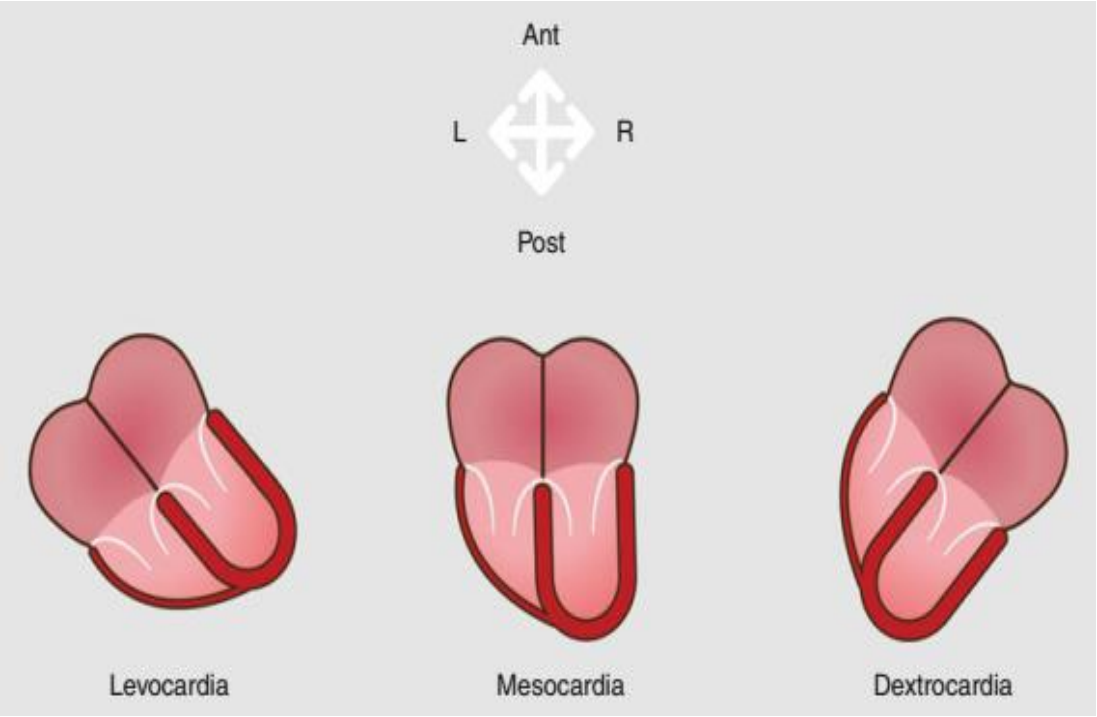


Fig. 1 a depiction of the three possible cardiac situs abnormalities

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 (**Jacobs JP et al., 2007).**

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 .The right main bronchus takes a more vertical course and branches at an earlier point than the more horizontally oriented left bronchus (**Van Mierop et al., 1970).**

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. 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. Three designations are used in characterizing atrial situs: situs Solitus, situs inversus, or situs ambiguous (**Jacobs JP et al., 2007)**.

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 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) (**Van Mierop et al., 1970).**

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 heterotaxy 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 (**Jacobs JP et al., 2007)**.

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. See Table 1 for a characterization of isomerization. Figure 2 illustrates the thoracic, atrial, and visceral situs seen with situs Solitus, situs inversus, and situs ambiguous (right and left isomerism). **(Moller JH et al., 1967).**

Table 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	

Quoted from Mazur et al., 2013

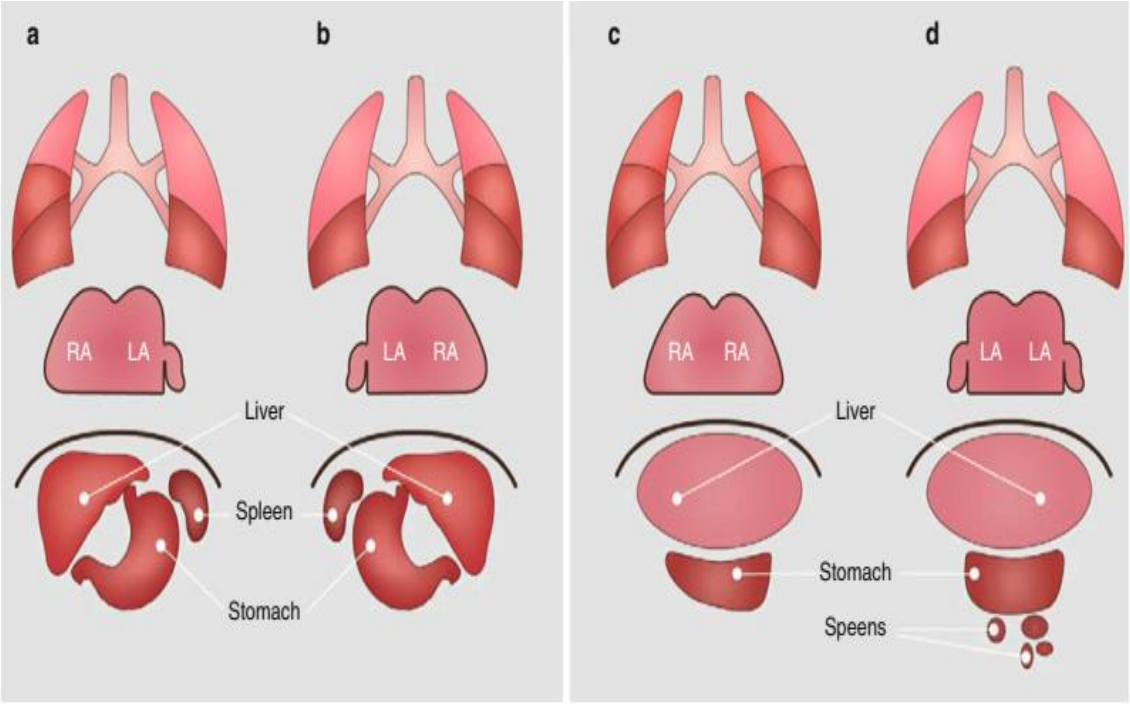


Fig. 2 The correspondence of atrial, thoracic, and abdominal organs arrangement in situs solitus (**a**), situs inversus (**b**), right isomerism (**c**), and left isomerism (**d**).

Definitions for the Atrial Chambers

The atria are often considered ‘‘viscera’’, and usually follow corresponding thoracovisceral connections The right atrium or systemic atrium is a developmental derivative of the major horn of the sinus venosus which receives blood from the superior, inferior vena cava and ostium of the coronary sinus. It is generally located on the same side as the liver, resulting in concordance of the visceroatrial situs. However, this situation is variable in patients with situs ambiguous .The main body of the right atrium is composed of a smooth segment (closer to the atrial septum) and a more trabeculated portion, separated by the terminal crest or crista terminalis. **(Edwards WD., 2008).**

A helpful method of identifying each atrium is the appearance of its corresponding appendage. The morphological right atrial appendage is more anterior than the left, has a pyramidal configuration and a broad based connection to the rest of the right atrium. In comparison, the left atrial appendage is more posterior, has a tubular beaked appearance and exhibits a narrower connection to the left atrium. Moreover, while the right atrium is smoother than the left atrium, the right atrial appendage tends to be less trabeculated than the left atrial appendage. The left atrium usually receives the pulmonary veins (the most notable exception being anomalous pulmonary venous return) and, except in extreme circumstances, does not receive systemic venous blood. Normally, there are two right pulmonary veins (occasionally three) and two left pulmonary veins (occasionally one). In addition, the left atrium has a more cephalad location within the mediastinum than the right atrium. The left atrial wall is thicker than the right atrial wall as seen in **table 2** (**Jacobs JP et al., 2007).**

Table 2: The Characteristics that differentiate the right from left atria

	Right atrium	Left atrium
Myocardial features	Crista terminalis, tinea sagittalis, extension of pectinate muscles toward AV valve	Pectinate muscles confined to appendage*
Appendage	Broad-based, triangular, anterior	Long and narrow (finger-like), posterior
Septum	Septum secundum (limbus of the fossa ovale)	Septum primum (valve of the foramen ovale)
Veins	Receives the major horn of the sinus venosus: IVC†, SVC‡, CS§.	Normally receives all pulmonary veins**

Quoted from Mazur et al., 2013

Definitions of Cardiac chambers

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 (**Lapierre et al., 2010).**

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. (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 3** identifies the distinguishing characteristics of the right and left ventricles **(Anderson RH et al., 1984).**

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 (**Lapierre C et al., 2010).**

Table 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

Quoted from Mazur et al., 2013

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 (**Praagh V 2010).**

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 (**Praagh V 2010).**

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. (**Lapierre C et al., 2010).**

A- 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. 3. (**Lapierre C et al., 2010).**

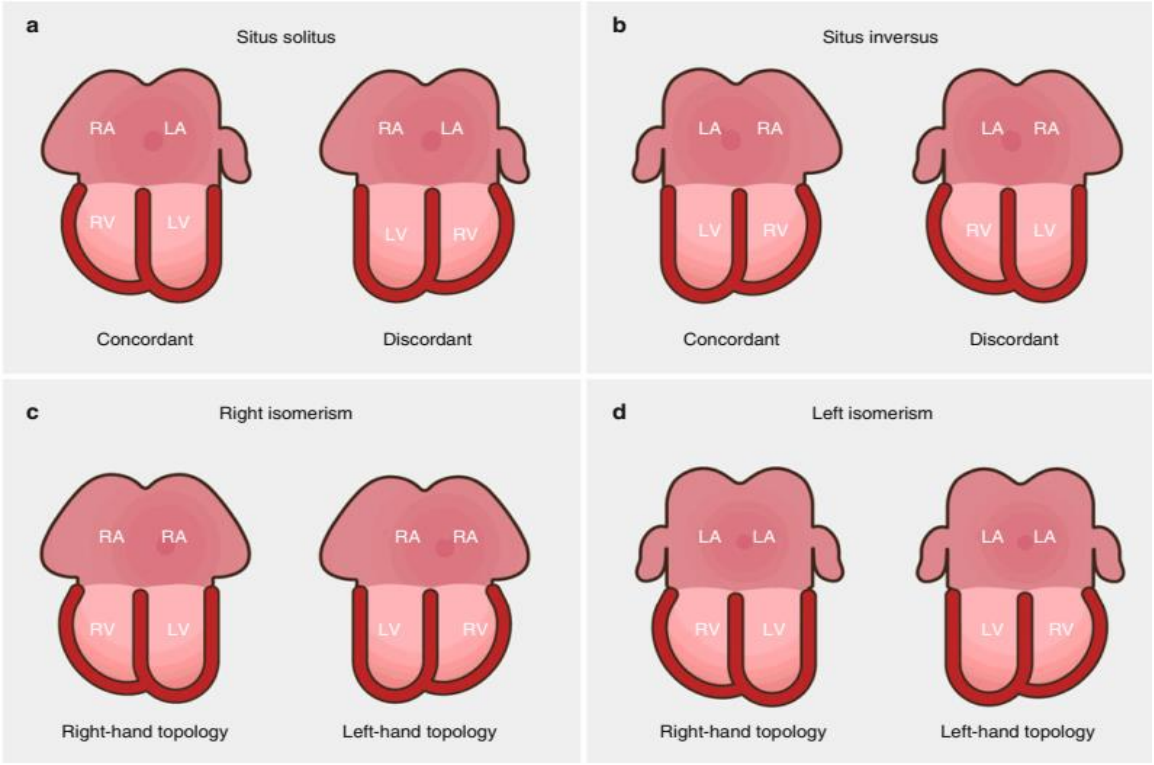


Fig. 3 The variants of biventricular atrioventricular connections in situs solitus (**a**), situs inversus (**b**), right isomerism (**c**), and left isomerism (**d**) demonstrating the independence of situs and looping

In 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 atrioventricular

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 atrioventicular valve (**Praagh V 2010).**Figure 4

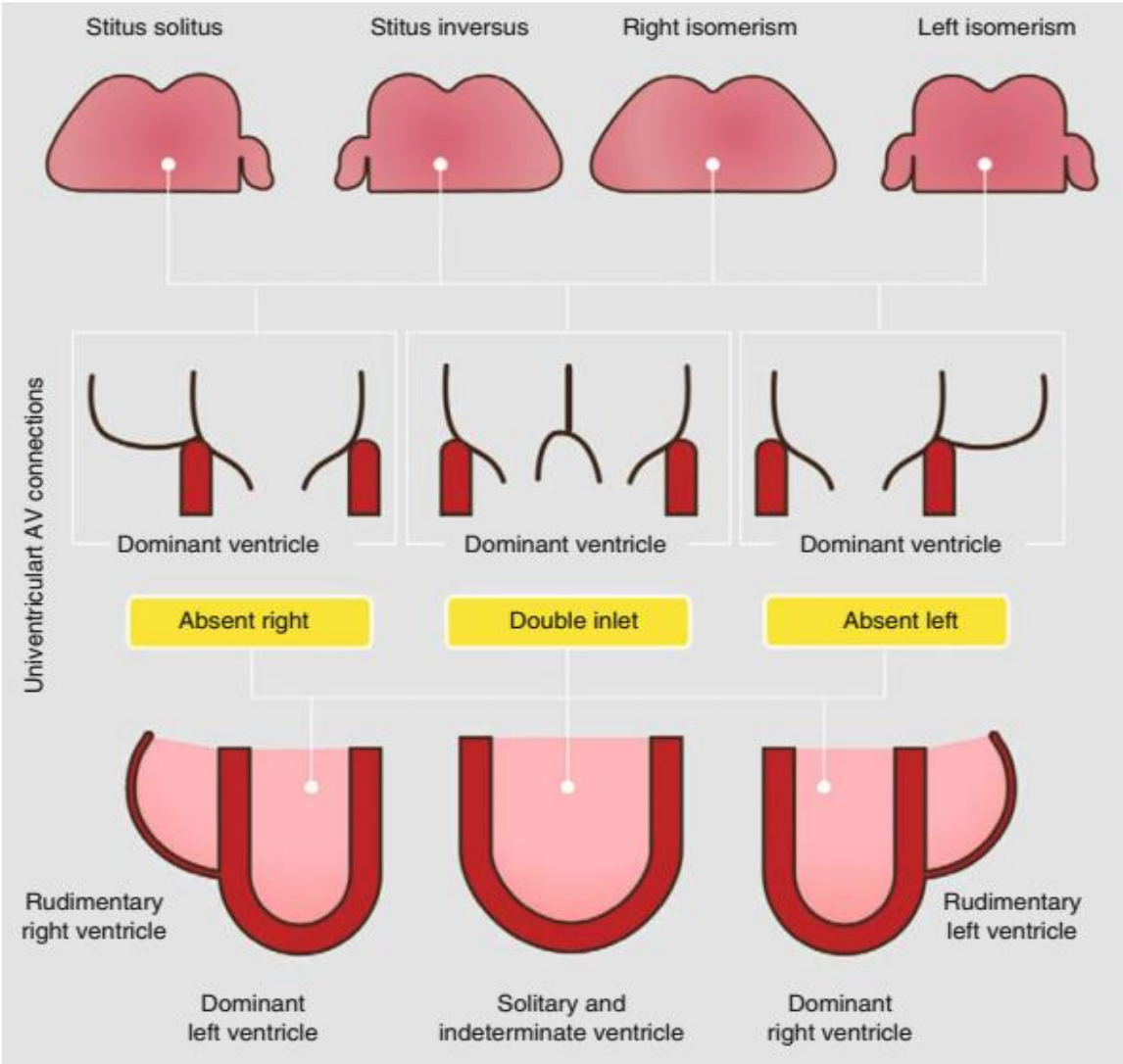


Figure 4 : The variations of univentricular atrioventricular (*AV*) connections in situs Solitus, situs inversus, and right and left isomerism

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. (**Lapierre C et al., 2010).**

B - 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 5** 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 **(Van Praagh R 1977)**.

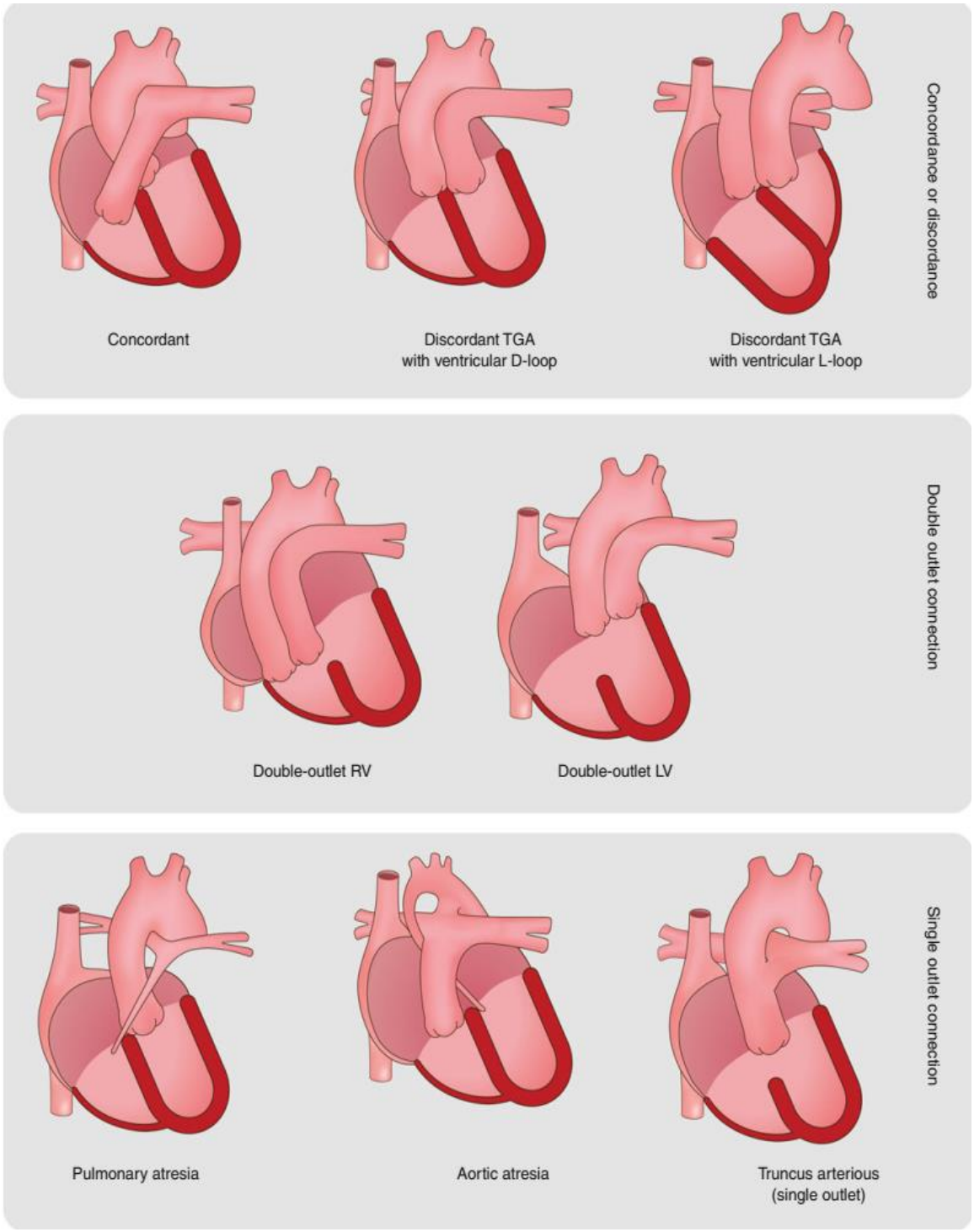


Fig. 5: A depiction of the possible ventriculoarterial connections

Chapter 2

Segmental, Sequential Approach in Congenital Heart Disease

Morphologically, malformations affecting the heart can be categorized into two broad categories. The first category is the malformations affecting the heart in which the cardiac chambers and the great arteries are normally related and connected. Typical examples are septal defects and valvular stenosis. The second category includes more complex malformations that are characterized by an abnormal relationship between the components of a segment or segments and abnormal connections between cardiac segments. Examples include various forms of so-called single ventricles or univentricular hearts, complete and corrected transposition of the great arteries, and double-outlet ventricles. These complex malformations require a systematic analysis in a step-by-step fashion that is called the sequential segmental approach **(Jacobs JP et al., 2007).**

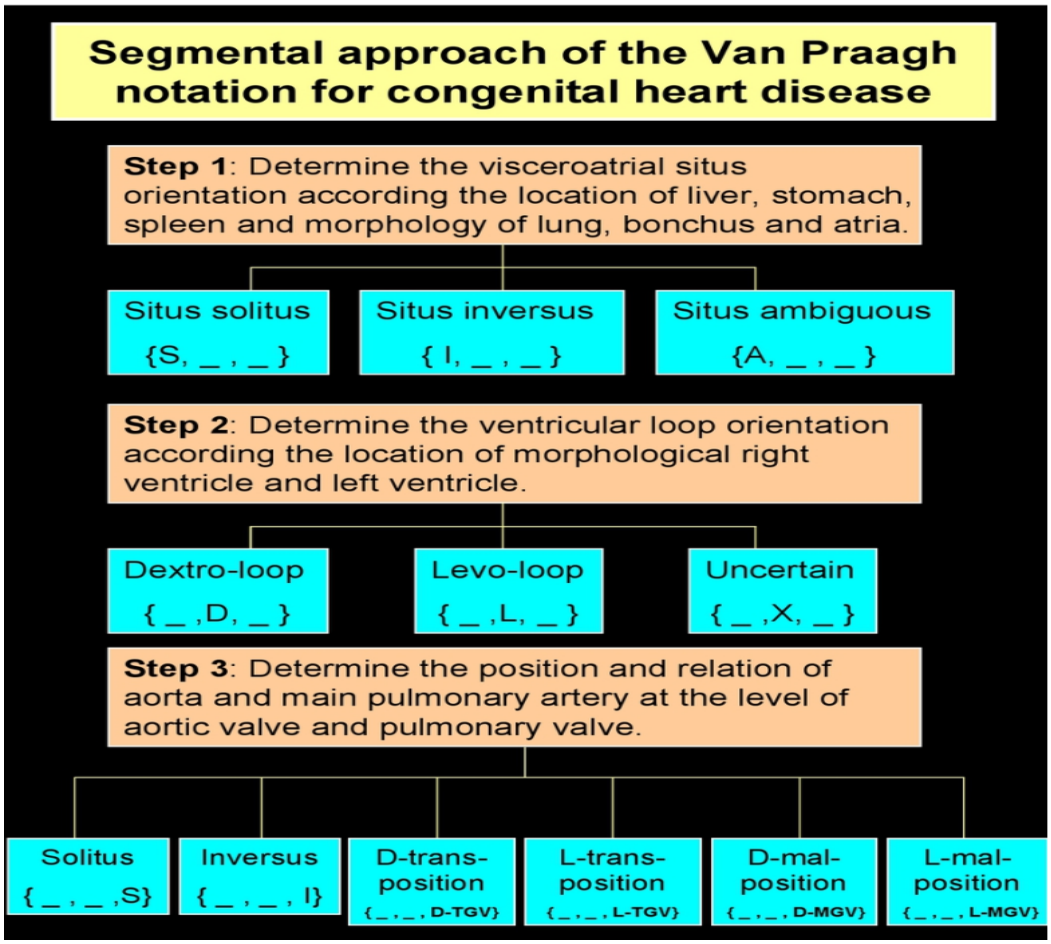
The concept of sequential segmental approach was first introduced by **Van Praagh et al in 1960.** Since then, there have been discussions and debates regarding the system of segmental analysis and its terminology. The most intense and serious debates have been between Richard Van Praagh of Boston, Massachusetts, and **Robert H. Anderson** of London, UK. Unfortunately, the debates have polarized the pediatric cardiology group into two schools, Van Praaghnians and Andersonians **(Lapierre C et al., 2010).**

Van Praagh first conceptualized the segmental classification of cardiac anatomy. Their description was limited to relationships of three main cardiac segments, namely, the atrial chambers, the ventricles, and the arterial trunks.

Later in the **1970s, Anderson *et al*.** highlighted the importance of morphology of the connecting segments, atrioventricular (AV) and ventriculoarterial (VA) junctions, in defining cardiac malformations (**Anderson RH et al., 1974).**

It is also well known that the assessment of each cardiac segment should be strictly based on its morphologic characteristics and not on its location, orientation, and connection with other segments. This assumes greater significance in the setting of CHD, where the variation in the orientation and connection of various cardiac segments is common (**Anderson RH et al., 1974)**.

The Van Praagh segmental approach



This flowchart demonstrates the three-step approach of the Van Praagh notation for complex congenital heart disease. Notice the major concepts of each step and each condition mentioned in every step.

What's is the sequential segmental analysis?

As discussed in Chapter 1, the heart consists of three morphologically and functionally distinct segments: the atria, the ventricles, and the arterial trunks **Figure 6.** They are joined together by two connecting units, the atrioventricular junction and the ventriculoarterial junction, both of which are usually guarded by the valves. Each component of each segment of the heart is characterized by its own morphologic characteristics. The components of each segment can be related in various ways, and the components of a segment can be connected to the components of the next segment in various ways. Therefore, there are three facets in the make-up of the heart: the ***morphologies,*** the ***connections,*** and the ***relations.*** Sequential segmental analysis is the systematic approach to the diagnosis of congenital heart disease in which the three facets of the make-up in the heart are analyzed in a segment-by-segment fashion **(Lapierre C et al., 2010).**

The major steps of sequential segmental approach include (Fig. 7)

1. **Determination of the visceral situs and cardiac position**
2. **Morphologic identification of the cardiac chambers and great arteries**
3. **Analysis of the connections and the relations**
    a. **Determination of the atrial situs**
    b. **Evaluation of the atrioventricular connections and ventricular relationship**
    c. **Evaluation of the ventriculoarterial connections and great arterial relationship**
4. **Evaluation of the associated anomalies at each segment**

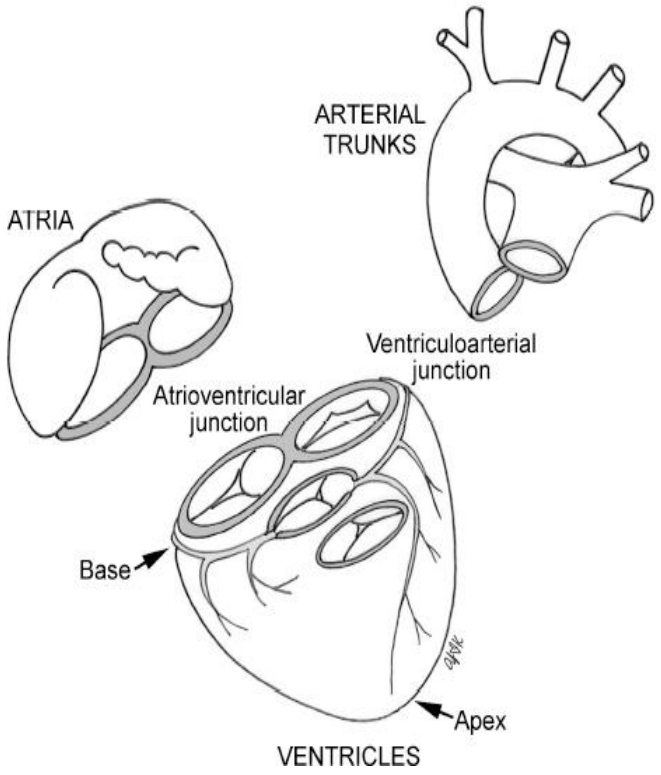


Fig. 6 Basic cardiac segments and intervening junctions

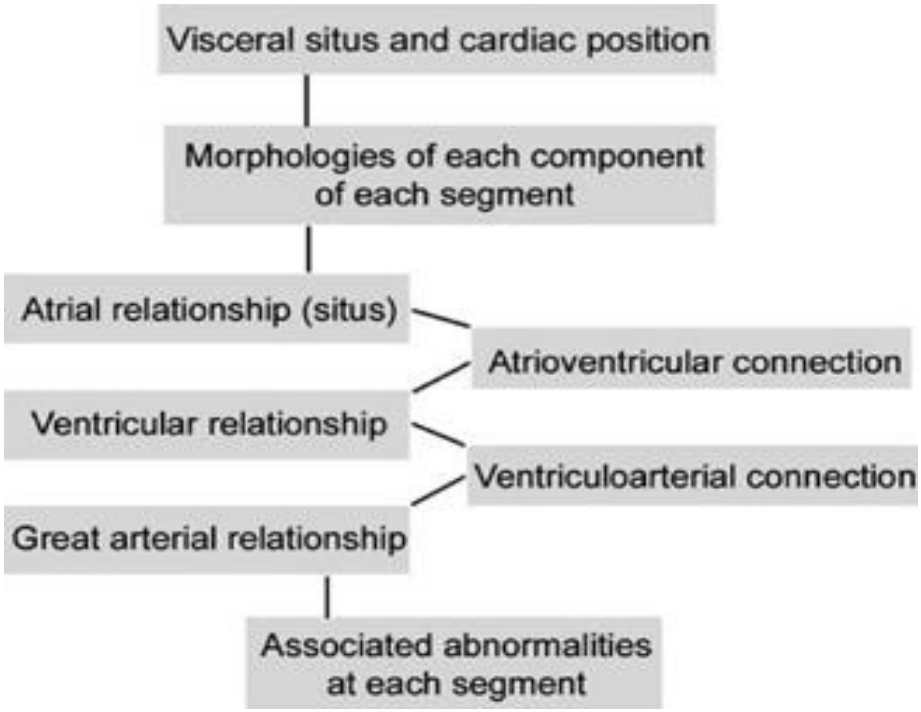


Fig. 7 Steps of the sequential segmental approach

Step 1: determine the visceroatrial situ and cardiac position

A- Cardiac position

Three different cardiac situs abnormalities are possible: levocardia, dextrocardia, and mesocardia**.** 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.

B- Determination of visceroatrial situs

The visceroatrial situs is determined by the relationship of bilateral artria and adjacent thoraco-abdominal visceral organs. It would be much easier to observe the location of abdominal visceral organs at first. In the normal anatomical configuration (situs Solitus, designated as {S, _, _ }, the largest lobe of the liver is on the right side and the spleen and stomach are on the left. If the largest lobe of the liver is on the left side and the spleen and stomach are on the right side, situs inversus (designated as **{I, _, _})** (**Figure 7)** should be considered **(Stanger P et al., 1977).**

Second, determine the thoraco-abdominal situs via broncho-pulmonary anatomy since pulmonary sideness usually coordinate with atria sideness. The main bronchus of morphological left lung locates below the left pulmonary artery **(hyparterial position)** and its upper lobe bronchus originates from a

more distal site compared to the right side. In contrast, the main bronchus of morphological right lung locates above the right pulmonary artery **(eparterial position)** and its upper lobe bronchus has a more proximal takeoff compared to left side **(Figure 8).** The number of pulmonary lobe also helps: the morphological right lung has three lobes while the morphological left lung has two lobes. (**Van Mierop et al., 1970)**.

The number of pulmonary lobes also helps establish sidedness; the right lung has three lobes, whereas the left lung has two. Even if the morphologic right lung is located on the left side, it generally maintains its trilobed character and right-sided bronchial anatomy. However, in the presence of right or left isomerism, duplication of the morphologic right or left bronchial tree renders pulmonary correlation unhelpful for determining the sidedness of the morphologic right atrium **(Van Mierop et al., 1970)**.

Third, determine the location of the morphological right atrium and left atrium. Identify the morphological right atrium according to image features of right atrial appendage **(Figure 9):** blunt, trapezoidal shape with a broad connection to the rest of the atrium. On the contrary, left atrial appendage **(Figure 10),** with a tubular, fingerlike shape, is usually narrower than the right and has a narrow connection to the rest of the atria. Sometimes the right and left atrial appendages are not well determined according to the radiologic image, we can observe the drainage chamber of systemic venous return (including inferior vena cava, superior vena cava and coronary sinus) instead. Due to the venoatrial concordance, the drainage chamber of systemic venous return is usually the morphological right atrium. **(Lapierre C et al., 2010).**

At last, determine the visceroatrial situs according to the morphological features just mentioned. If the sideness of thoraco-visceral structures (atria, bronchus, pulmonary lobes, liver, stomach and spleen) is as usual location, assign the letter S for situs solitus, {S, _, _}. If the sideness of thoraco-visceral structures is all reversed, assign the letter I for situs inversus,{I, _, **_}(Figure 11, 12)**. If the orientation of thoraco -visceral structures does not fit into either of condition, assign the letter A for situs ambiguous,{A, _, _}. Situs ambiguous is synonymous with Heterotaxy syndrome. Famous examples of heterotaxy syndrome are asplenic syndrome **(Figure 13 , 14)** and polysplenic syndrome **(Figure 15)** which demonstrates bilateral right-sideness (right isomerism) and bilateral left-sideness (left isomerism) of the thoraco-visceral structures respectively **(Stanger P et al., 1977)**.

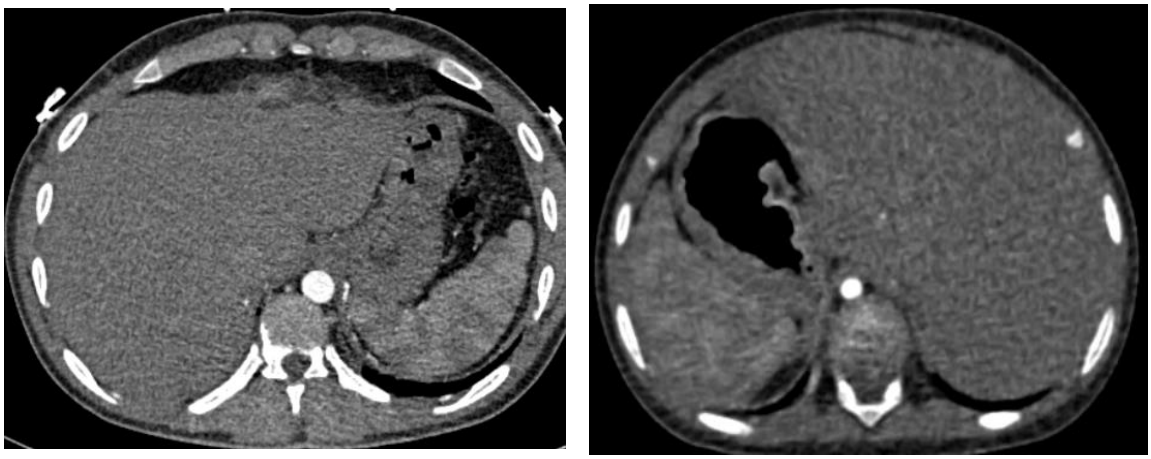


Fig. 7: A- CT axial MIP image shows the largest lobe of liver at the right side and stomach and spleen at the left side, which is regarded as **situs Solitus {S, _, _}.**

B- CT axial MIP image shows the largest lobe of liver at the left side and stomach and spleen at the right side, which is regarded as **situs inversus, {I, _, _}.**

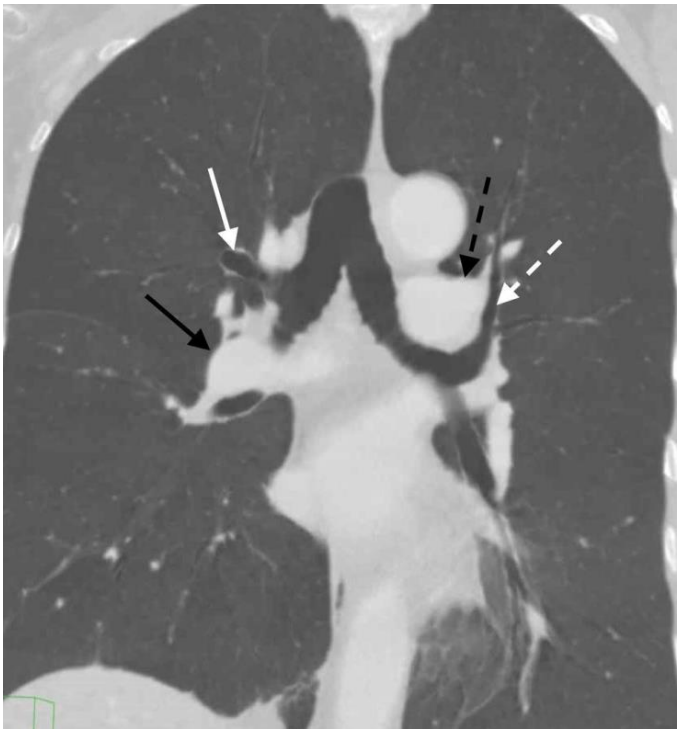


Fig. 8: CT coronal MIP images shows Normal pulmonary sideness of broncho -pulmonary anatomy,{S,_,_}: (1) Hyparterial position of left main bronchus and eparterial position of right main bronchus(black arrow: right main pulmonary artery; black dashed arrow: left main pulmonary artery). (2) Proximal take-off of the right upper lobe bronchus (white arrow) and distal take-off of the left upper lobe bronchus (white dashed arrow)

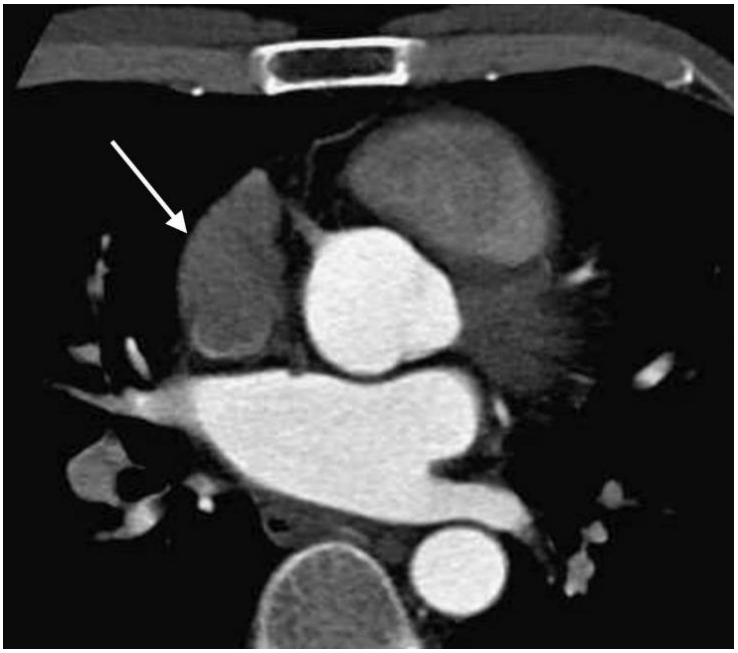


Fig. 9: CT axial image shows typical appearance of right atrial appendage (white arrow): blunt, trapezoidal shape with a broad connection to the rest of the atrium.

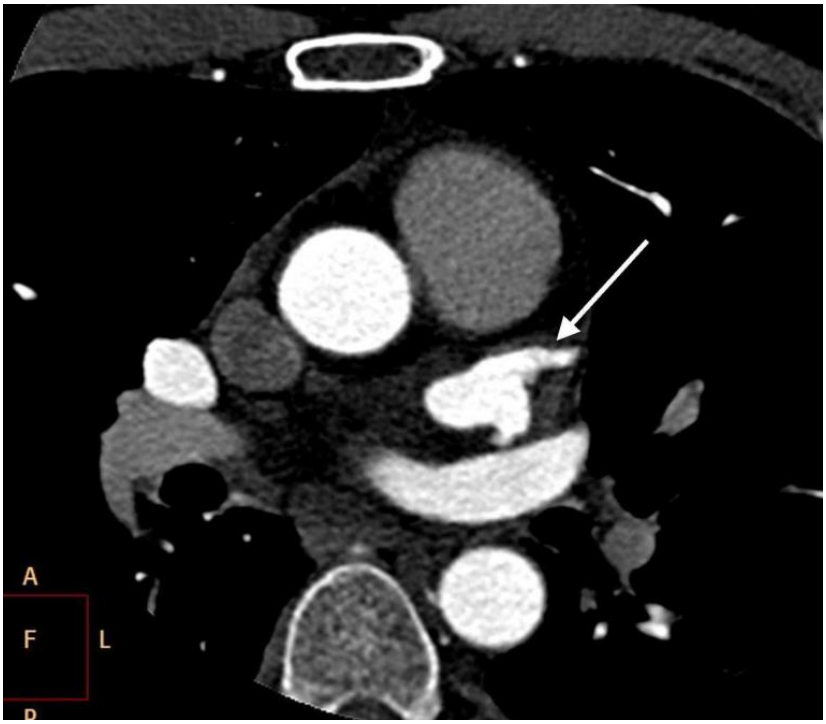


Fig. 10: CT axial images shows typical appearance of left atrial appendage (white arrow): tubular, fingerlike shape, narrower than right atrial appendage

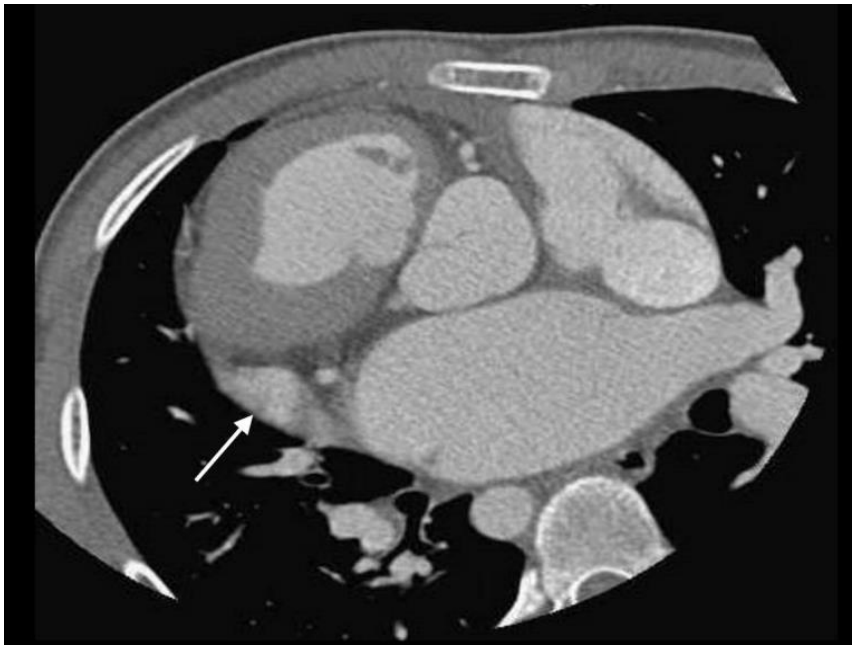


Fig. 11: CT axial image shows the morphological left appendage (white arrrow), which is tubular, narrow and finger-like shape, on the right side. Situs inversus is considered and is designated as {I, _, _ }

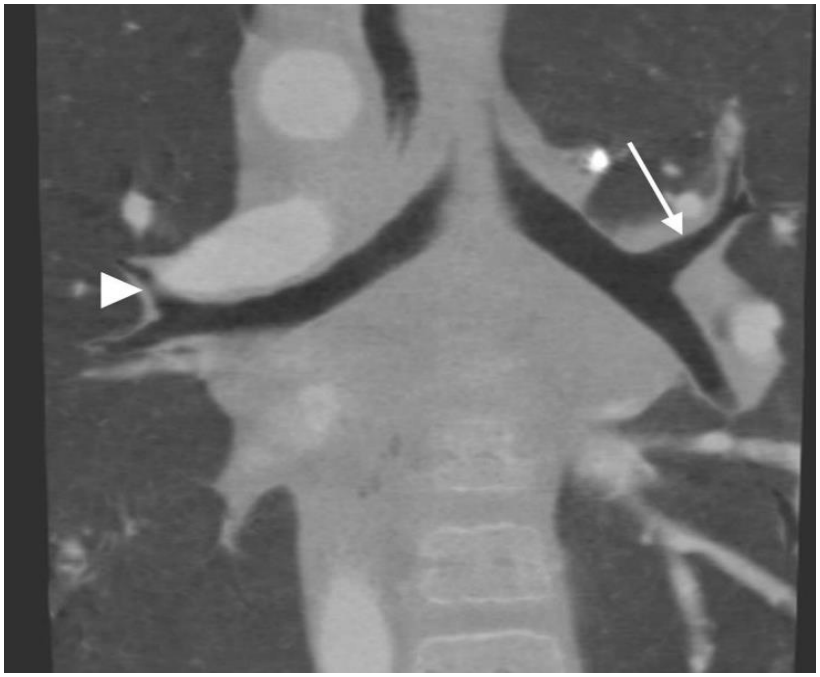


Fig. 12: CT coronal images (same case as Figure 5) has hyparterial location of right main bronchus with distal take-off of the right upper lobe bronchus (arrowhead). On the other hand, the left upper lobe bronchus takes off from a relative proximal location (white arrow). These findings also confirm situs inversus.

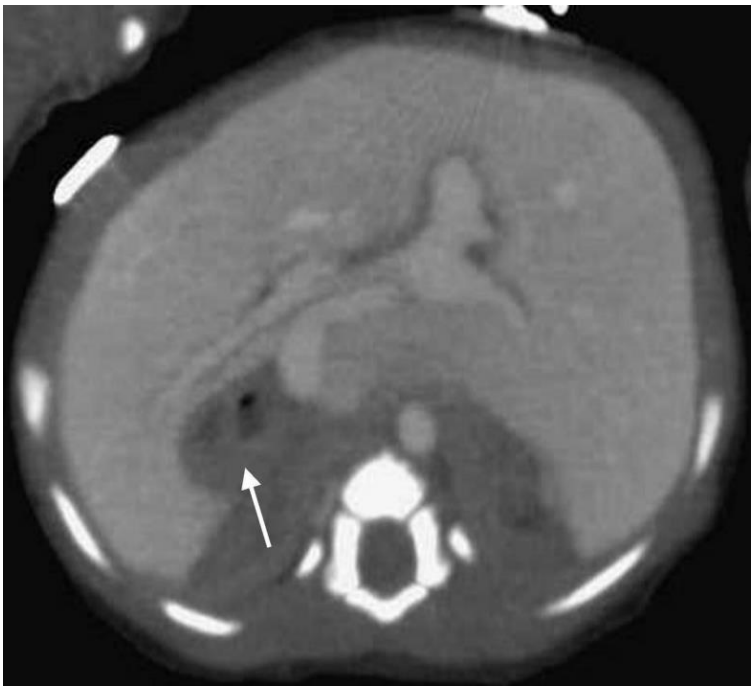


Fig. 13: CT axial image of 1 month old girl with huge transverse liver and right side stomach (white arrow). No definite spleen can be found in the abdomen. Situs ambiguous is suspected and {A,_,_} is assigned

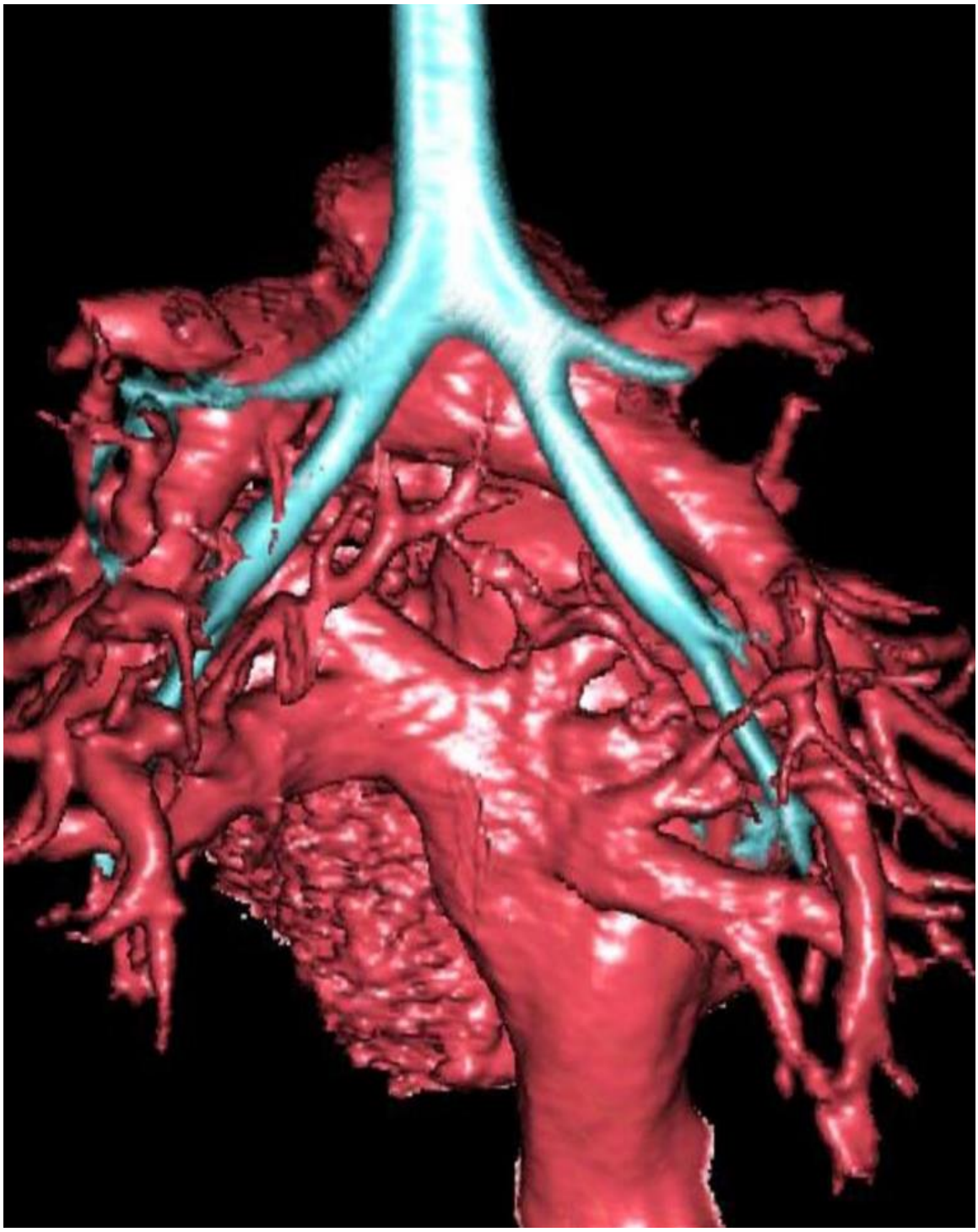


Fig. 14: 3D reconstruction of MDCT of 1 month old girl (same case as figure 13) has eparterial location of bilateral main bronchi with proximal take-off of the bilateral upper lobe bronchi. Bilateral right sideness of broncho-pulmonary anatomy is confirmed, and splenic syndrome is considered.

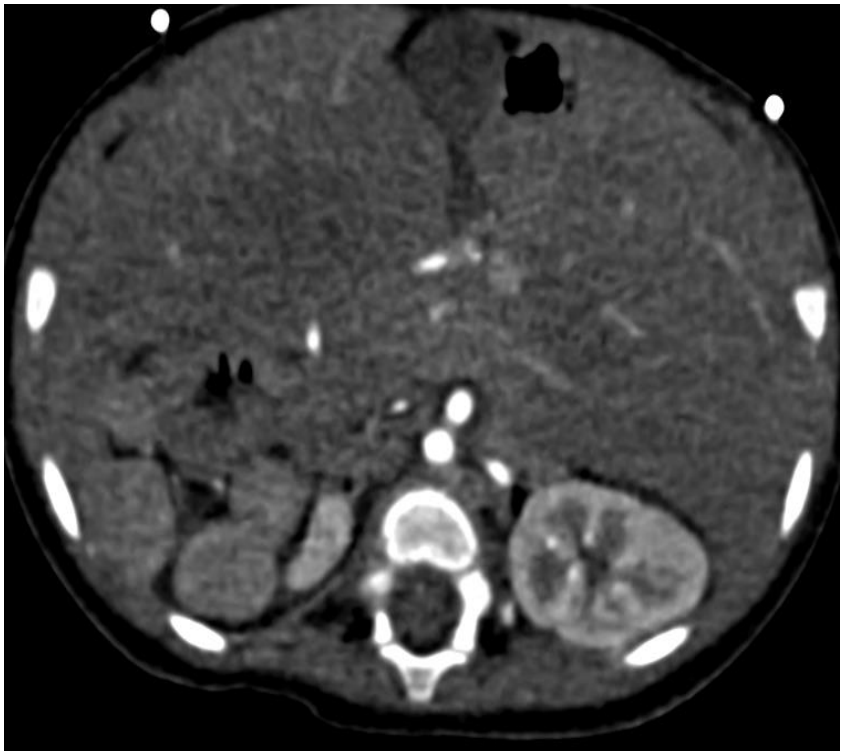


Fig. 15 : CT axial image of upper abdomen shows abdominal situs ambiguous (LT. Isomerism with midline liver and polysplenia)

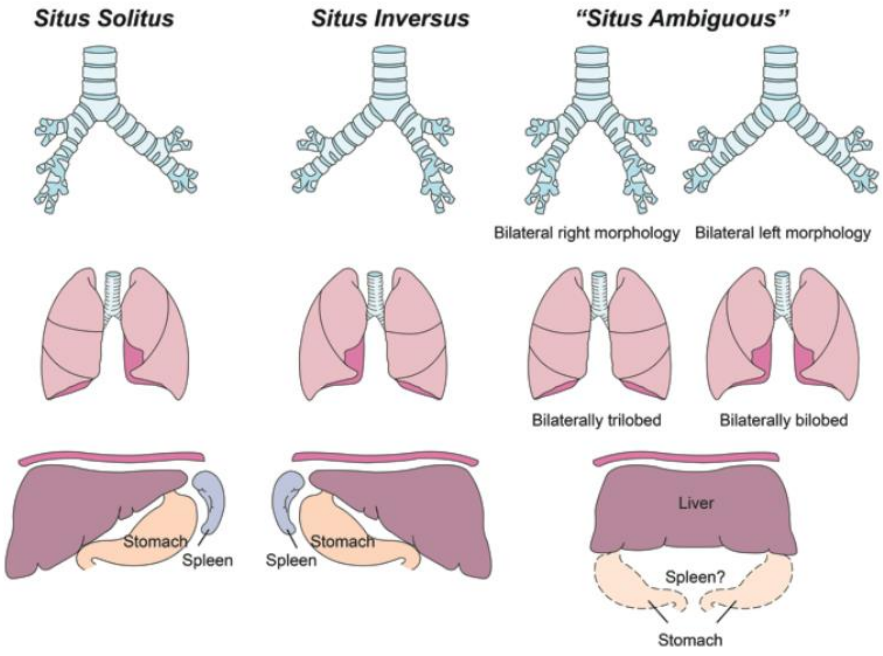


Fig. 16 The correspondence of thoracic, and abdominal organs arrangement in situs solitus , situs inversus , Situs Ambiguous

Step 2: Determine the ventricular loop orientation. {_, X, _}

Developmental Anatomy

During early development of embryo, the primitive heart is a tubular structure. This primitive tubular structure is composed of truncus arteriosus, bulbus cordis, primitive left ventricle and primitive atria in a cranio-caudal direction **(Figure 17)**. These segmental structures will develop into great vessel, right ventricle, left ventricle and atria in the future, respectively (Van **Praagh R & Vlad P. 1978)** .

At about gestational age of 4th week, this cardiac tube begins to elongate and fold on itself to right side or left side **(Figure 17)**. In normal condition, the cardiac tube bends toward right side, making the future right ventricle right side to the future left ventricle and forming a **D-loop** orientation. In contrast, if the cardiac tube folds toward left side, positioning the truncus arteriosus left side to the future left ventricle, an **L-loop** orientation develops. This folding process is important in the embryologic development and determines the position and relationship of major structures of cardiac tube (**Van Praagh R 1980).**

Identification of Right and Left Ventricles

To describe the orientation of ventricular loop, we identify the morphological right and left ventricles first. There are several distinguishing features of morphological right and left ventricles. **In morphological right ventricle**, the trabeculae are coarse and the papillary muscles attach to both free wall and interventricular septum **(Figure 18).** It also contains the famous "moderator band" **(Figure 18),** a trabecular structure extending across the right ventricular apex from the anterior papillary muscle to the interventricular septum. On the

other hand, the trabeculae of morphological left ventricle are thin and fine, with smooth superior septal surface **(Figure 19).** The papillary muscles in morphological left ventricle attached to the free wall only. (**Van Praagh R et al., 1981)**

After identifying the morphological right and left ventricles, we can determine the ventricular loop orientation. If the morphological right ventricle is located right side to the morphological left ventricle, assign the letter **D for D-loop** orientation, **{_, D, _}.** If the morphological right ventricle is located leftward of the morphological left ventricle, it indicates a **L-loop** orientation and the letter L can be denoted **(Figure 20)**. In complex cases, such as single ventricle, superior-inferior ventricles or failure to determine morphological right and left ventricles, use the loop rule or Hand rule. With the **loop rule**, we predict the ventricular loop orientation based on the position and relation of the great vessels. In the presence of a right-sided aortic valve, the morphologic right ventricle is located rightward of the morphologic left ventricle **(D-loop)** In the presence of a left-sided aortic valve, the morphologic right ventricle is located leftward of the morphologic left ventricle (**L-loop) (Figure 21) (Männer J. 2009).**

The hand rule reflects the principle of chirality: just as the right hand and left hand are mirror opposites, so too are the d-loop and the l-loop. To use the hand rule, first identify the right ventricle. Next, imagine approaching the heart from the anterior direction and placing your hand inside the right ventricle with your palm against the interventricular septum, the thumb in the right ventricular inflow tract (AV valve), and the fingers in the outflow tract. If you can accomplish this imaginary exercise with your right hand, the ventricular loop has a rightward orientation (d loop). If you can do it with your

left hand but not your right, the ventricular loop has a leftward orientation (l-loop). In other words, if the exercise is performed in the presence of a normal ventricular configuration (ie, a d-loop), the right hand will always fit into the right ventricle, whereas the left hand will always fit into the left ventricle. If the left hand fits into the right ventricle and the right hand fits into the left ventricle while the thumb is positioned in the right ventricular inflow tract and the fingers in the outflow tract, then the ventricular loop is reversed (ie, an l-loop). **(Van Praagh R 1985).**

At multidetector CT, the hand rule is best applied by using either a three-dimensional hollow view rotated so that the right ventricular side of the septum is visible or, alternatively, a vertical long axis image obtained with oblique multiplanar reconstruction at an angle that allows visualization of the outflow tract. If multiplanar reconstruction is used, the interventricular septum should come into view as the radiologist scrolls forward through the image sections. **(Van Praagh R 1985)**.

The "loop" rule is based on the observation that the position and relation of the great vessels are predictive of the ventricular loop orientation. In the presence of a right-sided aortic valve, the right ventricle is located rightward of the left ventricle, signifying the presence of a d-loop; and in the presence of a left-sided aortic valve, the right ventricle is located leftward of the left ventricle, signifying an l-loop. If all attempts fail and the direction of ventricular folding cannot be determined by using either the hand rule or the loop rule, assign an ***X*** *for* unknown loop orientation (denoted as **"{_, X, _}**"). Such instances should be rare. (**Van Praagh R 1985)**.

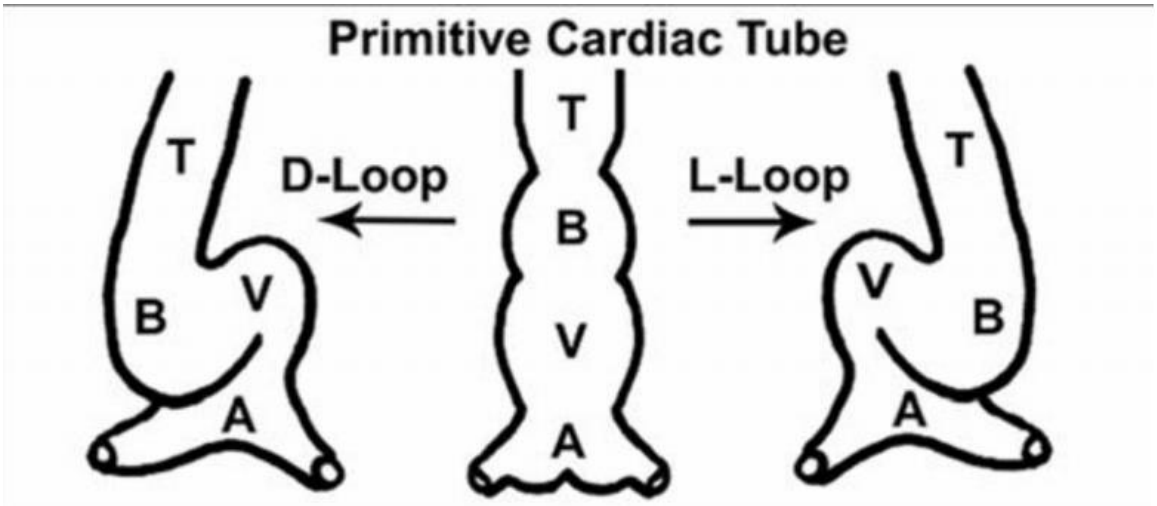


Fig. 17: Diagram illustrates bending of primitive cardiac tube. Cardiac tube is demonstrated from anterior view. Cardiac tube is composed of atrium (A), ventricle (V), bulbus cordis (B) and truncus arteriosus (T). Cardiac tube normally bends to right side, forming a D-bulboventricular loop. Rarely, cardiac tube may bend leftward, forming a L - bulboventricular loop.

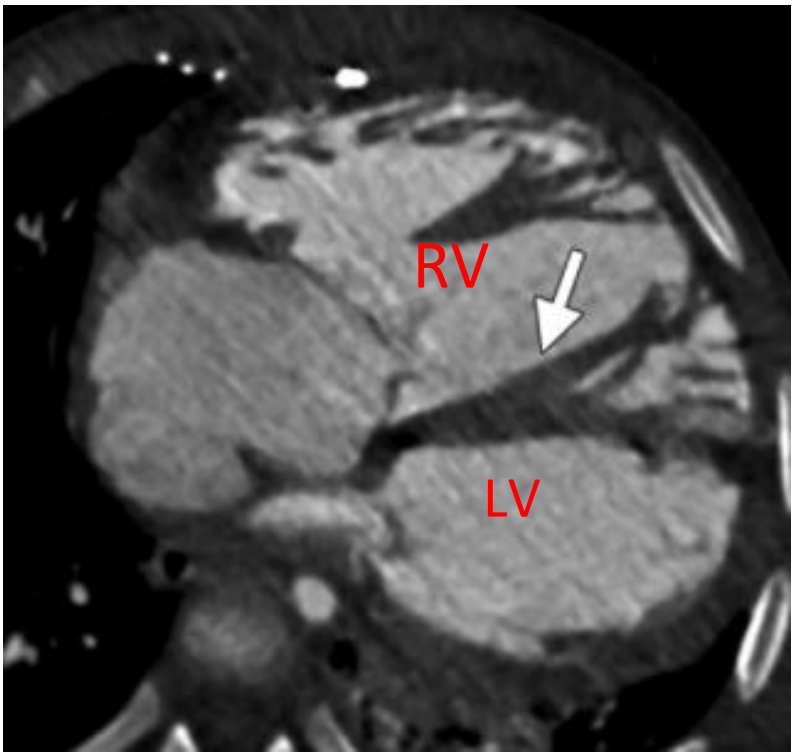


Fig. 18: CT axial image shows in morphological right ventricle, the trabeculae are coarse and the papillary muscles attach to both free wall and interventricular septum. It contains "moderator band"(white arrow), a thick trabecular structure extending from anterior papillary muscle to interventricular septum **(D- Loop)**

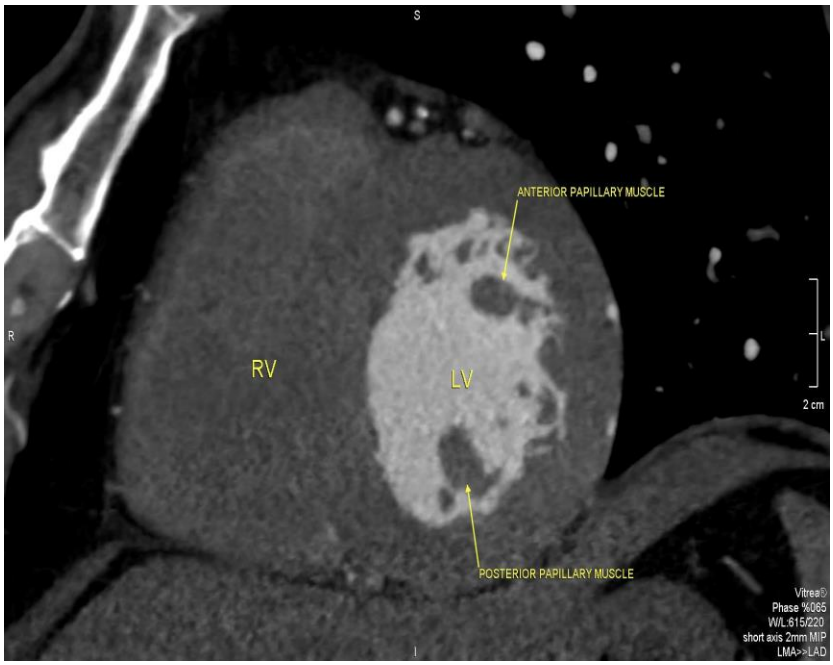


Fig. 19: CT oblique image shows the morphological left ventricle has fine and thin trabeculae with smooth septal surface and two papillary muscles attached to the free wall

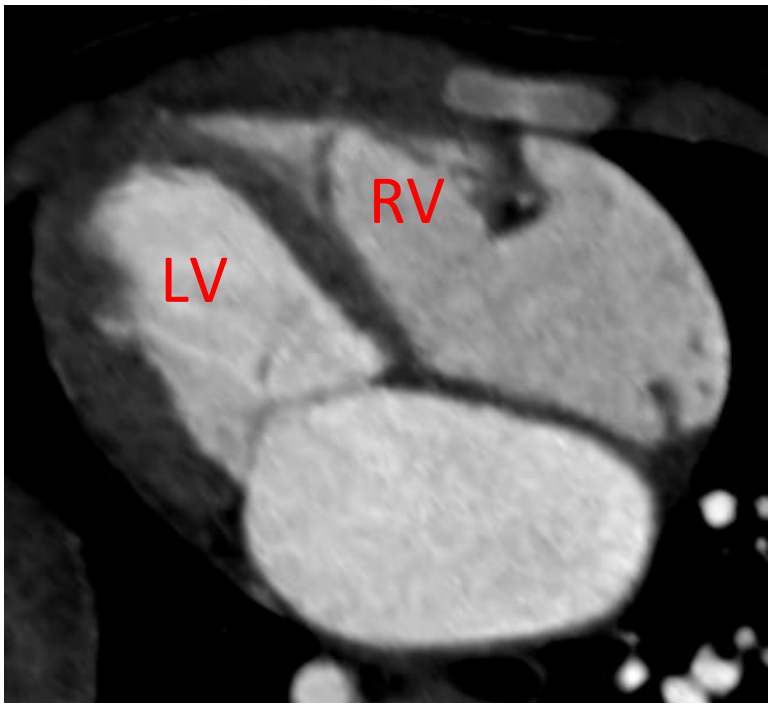


Fig. 20 : CT axial image shows the morphological right ventricle located leftward of the morphologic left ventricle a L-loop ventricular orientation as the loop rule predicts, assigned as {_,L,_}. Notice thick moderator band extending from ventricular wall to septum.

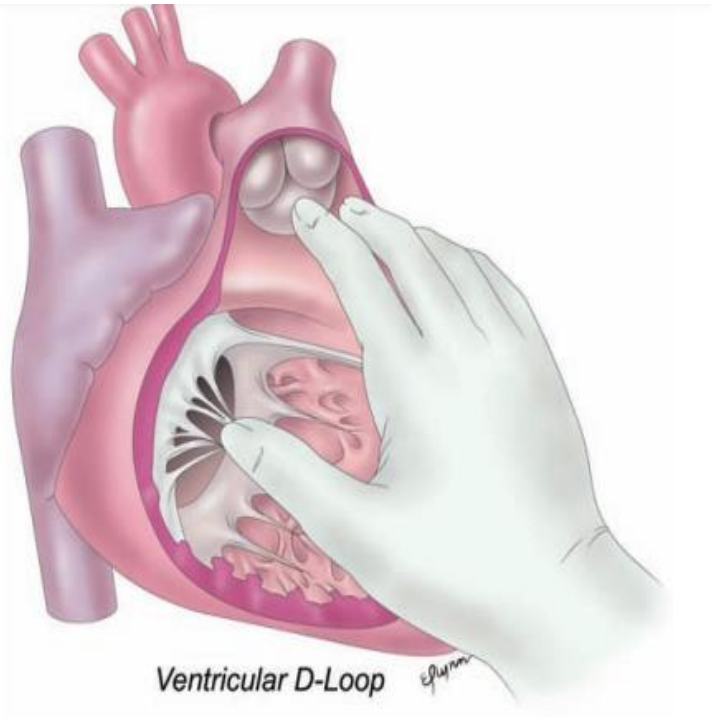


Figure 20 : Hand rule

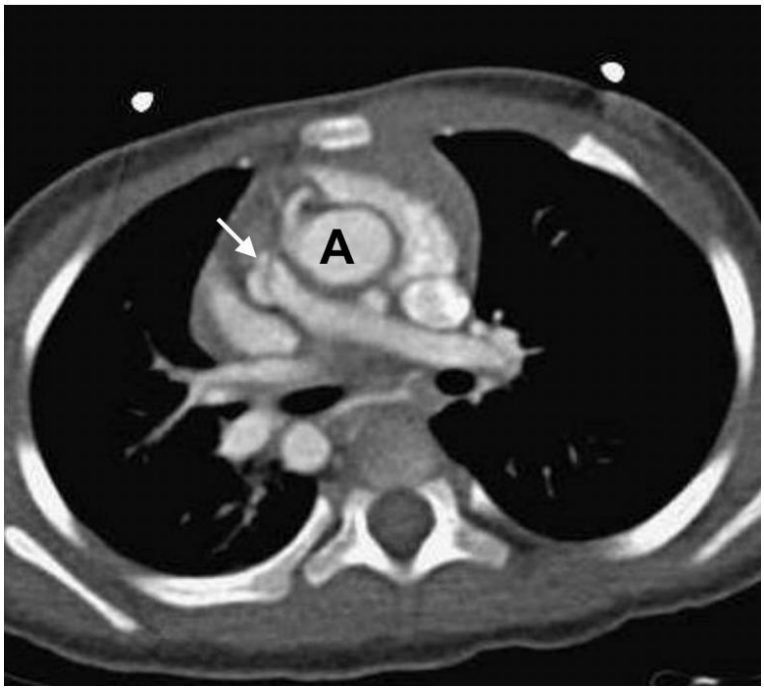


Fig. 21: CT axial image of the great vessels shows (A) Aorta leftward to the main pulmonary trunk (white arrow). According to the loop rule, she would have **L-loop** ventricular orientation.

Step 3: determine the relation and anomaly of great vessels origin. {_, _, X}

There are a group of congenital heart disease with anomaly developing at the level of junction of great vessels and ventricles, known as conotruncal anomalies, such as LTGA, D-TGA, DORV, TOF…etc. Normally we observe the connection between great vessel origin and ventricles for diagnosis of these malformations. However, in 1980, Van Praagh and Layton et al. proposed the relationship of conotruncal anomalies and the positions of great vessel origin In their concept, the orientation of great vessel origin can be demonstrated and categorized in a six-point diagram (depicted as Figure 22). Observe the level of aortic valve and pulmonic valve representing origin of great vessels. Look for the aortic root and main pulmonary trunk (MPA) instead if the aortic and pulmonic valves are difficult to identify due to previous operation (**Van Praagh R, et al**., **1980).**

The great vessels are identified by their branch vessels. The aorta gives rise to at least one coronary artery and most of the systemic circulation. The main pulmonary trunk feeds at least one pulmonary artery. In the normal arrangement, the aorta is posterior and to the right of the pulmonary artery (**Van Praagh R, et al**., **1980).**

In the normal orientation of great vessels origin, the aortic root is posterior and rightward of the MPA. We assign the letter "S" to denote this normal configuration, which is situs Solitus of great vessels: **{_, _, S}** (Figure 23). If the aortic root is posterior and but leftward of the MPA, the letter "I" is assigned to represent situs inversus of great vessels: **{_, _, I}** (Figure 24). When malformation develops at connection of great vessels and ventricles, it

may be either transposition or malposition of the great vessels. Transposition of great vessels means the aorta originates from morphological right ventricle and MPA originates from morphological left ventricle. Malposition of the great vessel means both great vessels originate from the same ventricle or are overriding (**Van Praagh R, et al**., **1980).**

In case of transposition, the aorta is usually but not always anterior to the MPA. If the aorta is anterior and right to the MPA, the anomaly is regarded as dextro-transposition of the great vessels, which is recorded as **{_, _, D-TGV} (Figure 25).** If the aorta is anterior and left to the MPA, the anomaly is described as levo- transposition of the great vessels or congenitally corrected transposition of great vessels, which is recorded as **{_, _, L-TGV} (Figure 26).** If the aorta is not anterior or posterior to the MPA, the condition is usually malposition of the great vessels. If the aorta is right side to the MPA, it is described as D malposition and recorded as **{_, _, D-MGV}** (Figure 27 & 28). If the aorta is left side to the MPA, it is described as L-malposition and recorded as **{_, _, L-MGV}** (Figure 29 & 30) (**Van Praagh R, et al**., **1980).**

Additionally, since the situs solitus, dextroposition and dextro-malposition of great vessels result from rightward folding of the primitive cardiac tube (the aorta is rightward of the MPA), these conditions usually concur with D type ventricular loop. On the other hand, situs inversus, levoposition and levo-malposition of great vessels (the aorta is leftward of MPA) often accompany with L type ventricular loop due to leftward folding of the primitive cardiac tube. These phenomena verify the **"loop rule"** mentioned before **(Männer J. 2009)**.

There are four possible types of conal (infundibular) anatomy that must be potentially identified: sub pulmonary conus (normal), sub aortic conus,

bilateral conus, and bilaterally absent conus (Fig. 31). Sub aortic conus is seen in cases of D- or L-transposition. Bilateral conus is found in cases of double-outlet right ventricle and bilaterally absent conus is noted in cases of double-outlet left ventricle (**Lapierre C et al., 2010).**

Double outlet ventricle

There is a bewildering heterogeneity in this group of heart malformations that results in ventricular arterial misalignment. The right, as well as, the left ventricle can exhibit a ‘‘double outlet’’ type of architecture, in which more than one and a half portions of both great arteries arise from one ventricular cavity. There is generally a ventricular septal defect. The anatomic hallmark in double outlet right ventricle is discontinuity of the mitral and aortic valves. There may be associated pulmonic stenosis, aortic stenosis, transposition of the great vessels and aortic Coarctation or interruption of the aortic arch **(Männer J. 2009)**.

Criss-cross heart

Criss-cross heart describes a series of cardiac malformations resulting in crossed atrioventricular connections. This is a consequence of ventricular twisting along its long axis while the base of the heart remains fixed. This gives the ventricles a superoinferior relationship with crossing of their inflow streams. This condition is frequently associated with large ventricular septal defects and L-looping of the ventricles and may exhibit straddling AV valve chordae (**Anderson RH & Shirali G 2009).**

Truncus arteriosus

Truncus arteriosus (persistent truncus arteriosus) is a congenital condition in which a single vessel or trunk supplies the coronary, systemic and pulmonary circulations. Embryologically, it represents a failure of spiral division of the truncus arteriosus into an aorta and pulmonary artery. This common trunk overrides both ventricles and, as a result of deficiency or absence of the conal or infundibular septum, there is typically an associated ventricular septal defect. Occasionally, it is known as a solitary arterial trunk. A single semilunar valve is the rule, and it is usually in fibrous continuity with the mitral valve (**Tynan M et al., 1979).**

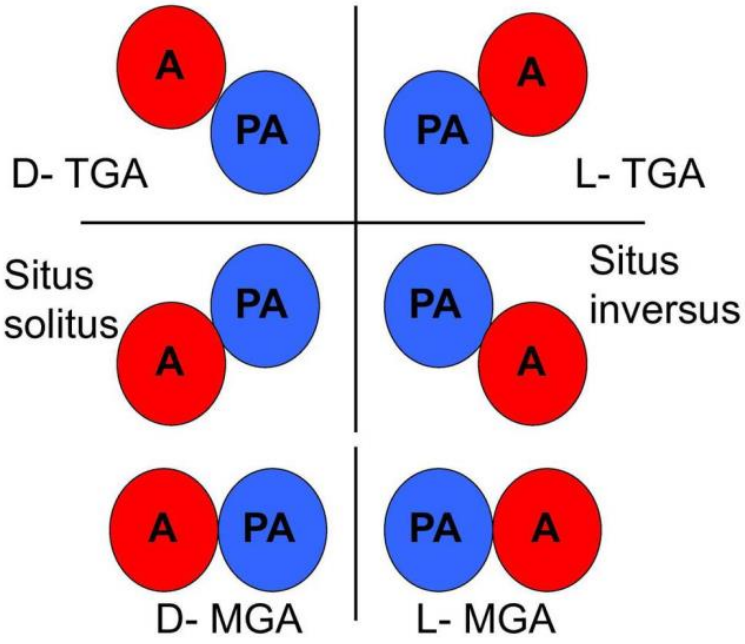


Fig. 22: This simple diagram demonstrates the relationship of great vessel origins on cross sectional image in all six condition, including: situs solitus, situs inversus, D-TGA, L-TGA, D-MGA, L-MGA. A=aorta, PA= pulmonary trunk, TGA= transposition of the great arteries, MGA= malposition of the great arteries.

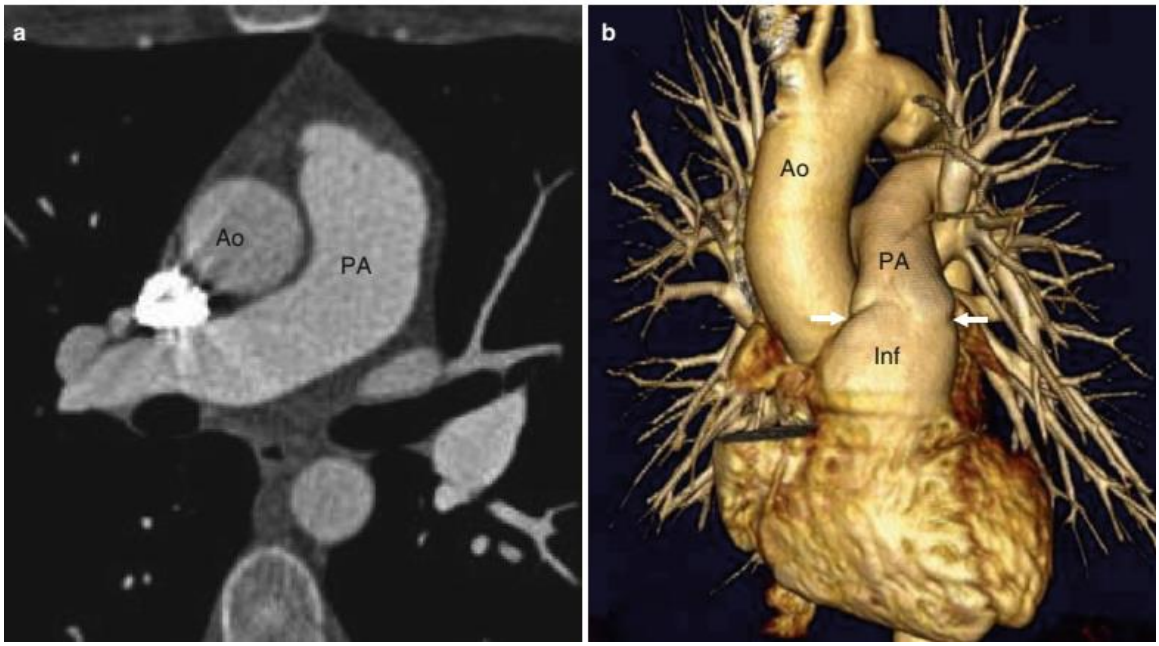


Fig. 23: Panel **(a)** is an axial, maximum-intensity projection, CT image demonstrating the normal spatial orientation of the great vessels.
The PA is to the left of the aorta. Panel **(b)** is a 3-dimensional volume rendered CT image showing the aorta posterior and to the right of the pulmonary artery. Which is designated as situs Solitus of great vessels **"{_, _, S}".**

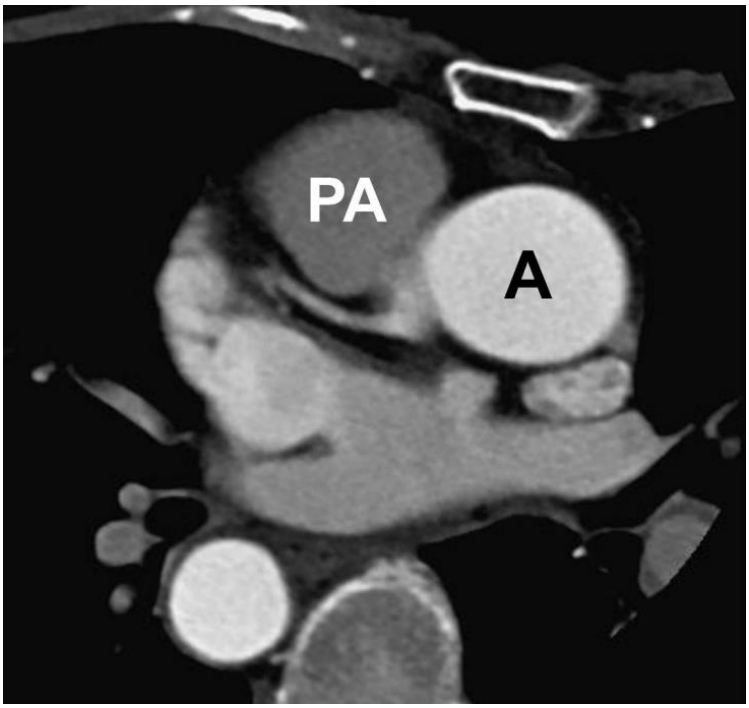


Fig. 24: CT axial image shows Inversion of the great vessels: the aortic root(A) is located posterior and rightward to the main pulmonary trunk(PA), at the level of the valves, which is designated as situs inversus of great vessels "{_,_,I}".

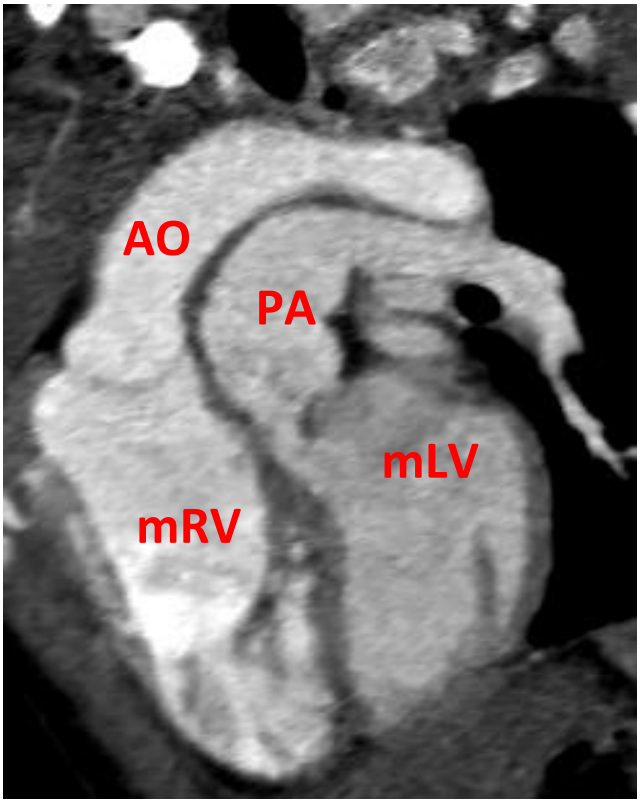


Fig. 25: CT image shows D-TGA has aorta arising from morphological right ventricle (m RV) and The aortic root locates anterior and rightward of the pulmonary trunk (PA), which is designated as **"{_,_,D-TGA}".**

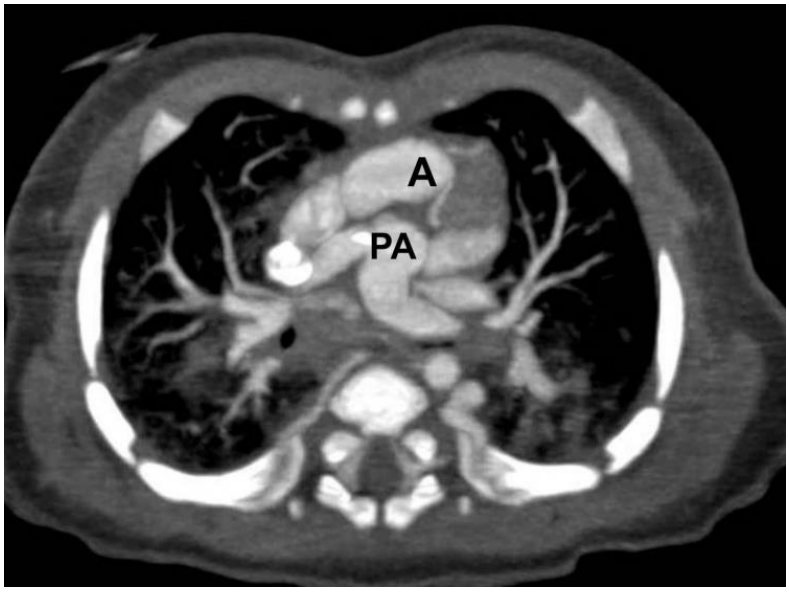


Fig. 26: CT image shows L-TGA has aortic root (A) locates anterior and leftward to the main pulmonary trunk (PA), this is assigned as **"{_, _, L-TGA}".**

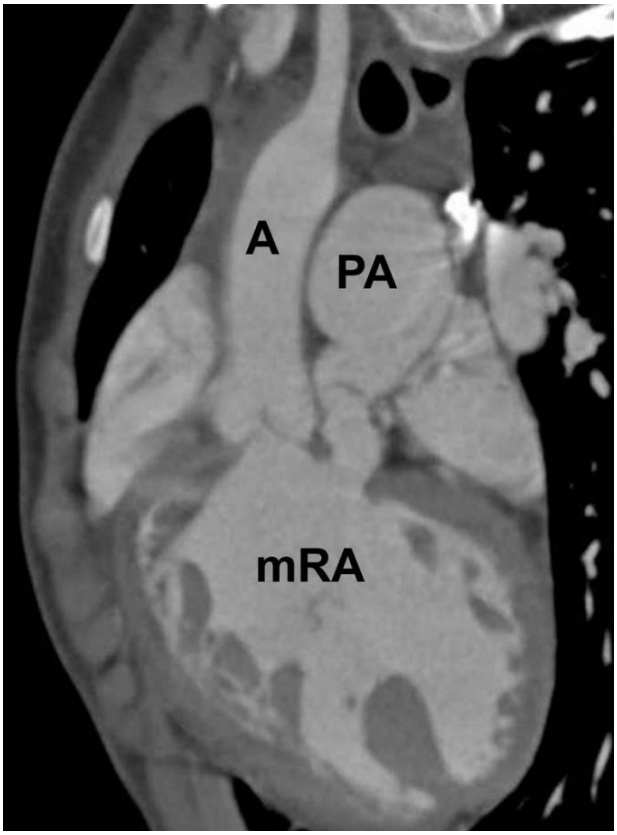


Fig. 27: CT coronal image in 6 year old girl with double outlet right ventricle, has aortic root (A) and main pulmonary trunk (PA) arising in a parallel plane from morphological right ventricle (mRV), which has thick and coarse trabeculae

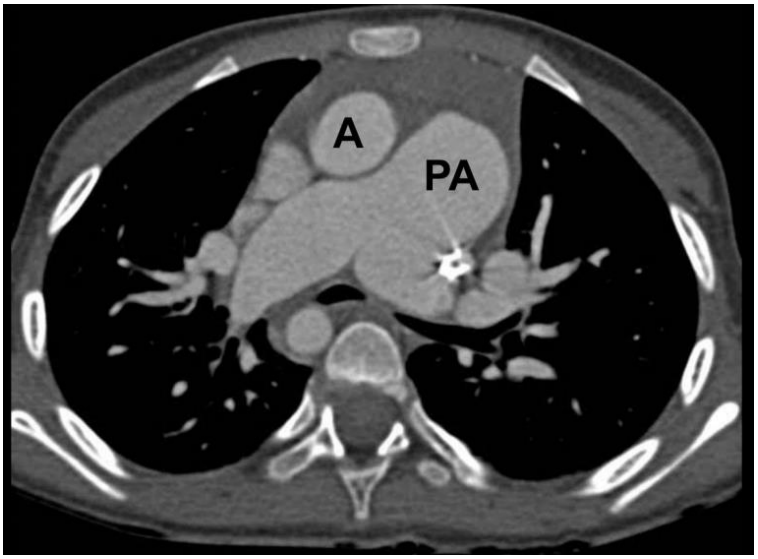


Fig. 28: CT axial image of 6 year old girl with double outlet right ventricle has aortic root(A) and main pulmonary trunk (PA) arising in a parallel plane with aorta at right side and main pulmonary trunk at left side, which is designated as **{_, _, D-MGA}.**

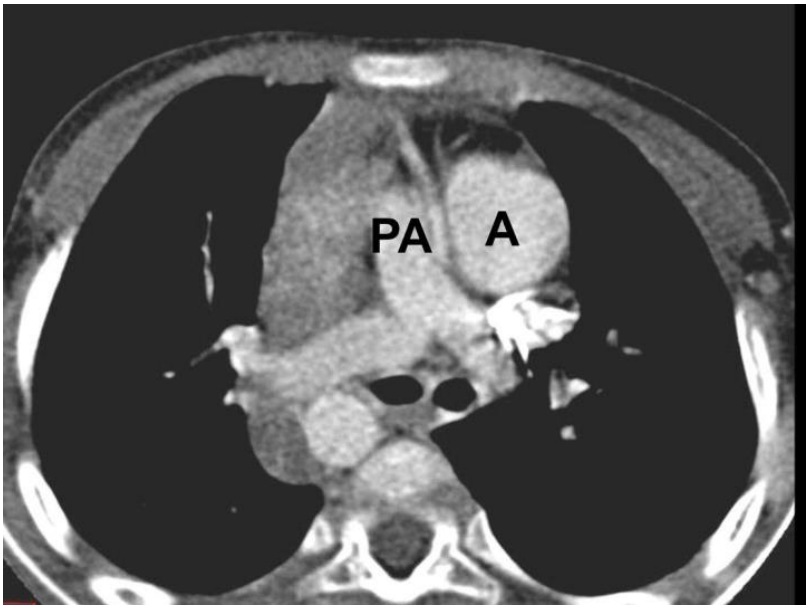


Fig. 29: CT axial image of 10 year old boy with double outlet left ventricle has aortic root (A) and main pulmonary trunk (PA) arising in a parallel plane with aorta at left side and main pulmonary trunk at right side, which is designated as **{_, _, L-MGA}.**

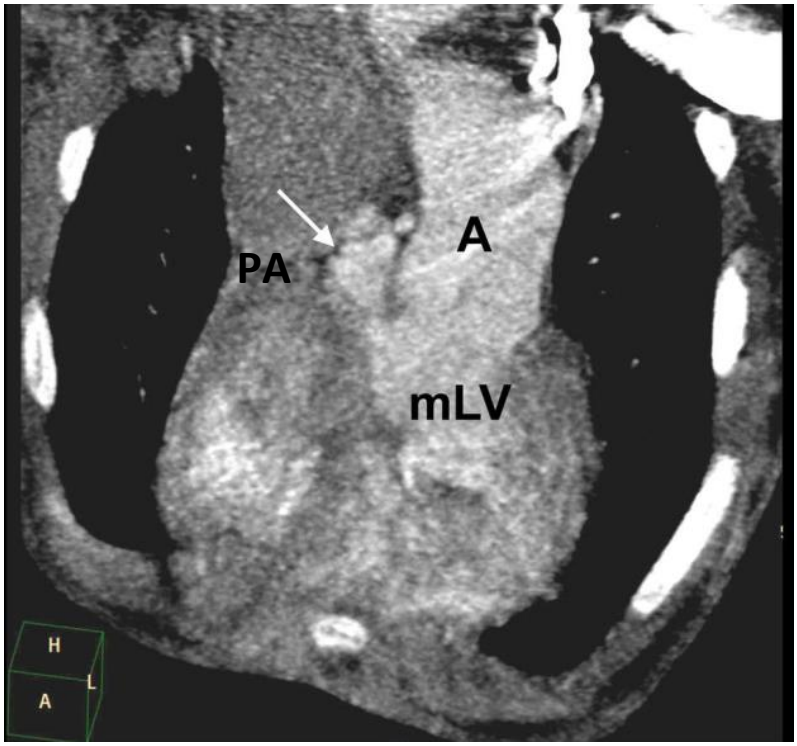


Fig. 30: CT coronal image of 6 year old girl with double outlet left ventricle(same case as figure 23) has aortic root (A) and main pulmonary trunk (PA) arising in a parallel plane and both from morphological left ventricle (mLV). Which is designated as **{_, _, L-MGA}.**

Assessment of Connecting Segments

Specification of Atriovisceral situs, orientation of the ventricular loop, and spatial orientation of the great vessels do not specify the atrioventricular and ventriculoarterial connections. The atrioventricular and ventriculoarterial connections are independently determined Embryologically and, thus, need to be separately evaluated in terms of biventricular versus univentricular connections **(**step 4**).(Van Praagh R , 1984) .**

Atrioventricular valve

Morphologically, the AV valve represents the ventricular chamber and it is one of the features used to identify a ventricle as right or left. In hearts with concordant AV connections, the tricuspid valve guarding the right AV junction has three leaflets and is positioned distally (apical offsetting) compared to the left-sided bi-leaflet mitral valve . Unlike the mitral valve, the tensor apparatus of the tricuspid valve connects to the ventricular septum. These findings are useful in echocardiographic identification of tricuspid and mitral valves. In the setting of atrioventricular septal defect (AVSD), the AV valve is common with no apical offsetting of the left and the right components of the valve (**Anderson RH, et al., 2010)**

Since the valve is common and does not possess characteristics of a normal mitral or tricuspid valve, it is better to use the term left and right AV valves, instead of tricuspid and mitral valves. The apical offsetting is also absent in cases with inlet type of ventricular septal defect (VSD) and both AV valves are at the same level (**Anderson RH, et al., 2010)**

Ventriculoarterial Connections

Step 4 begins with an assessment of the junction between the ventricles and great arteries. Besides permanent truncus arteriosus, four types of ventriculoarterial connection may develop: **concordant** connection (the pulmonary artery arises from the right ventricle, and the aorta arises from the left ventricle); discordant connection, which is synonymous with transposition of the great vessels (the pulmonary artery arises from the left ventricle, and the aorta arises from the right ventricle); double outlet right ventricle (the great vessels arise from the right ventricle); and double outlet left ventricle (the great vessels arise from the left ventricle)**(Van Praagh R , 1984)**. **Figure 32**

Infundibulum

The infundibulum is the connecting segment between the ventricles and the arterial trunks. In normal hearts, there is a complete subpulmonary conus with muscular separation between the pulmonary and the right-sided tricuspid valves, whereas the subaortic conus is absent, allowing fibrous continuity between the left and non-coronary cusps of the aortic valve and the base of the anterior mitral leaflet . In some hearts, the aortic valve is separated from the mitral valve when it is labeled as aorta–mitral discontinuity. In morphological terms, this indicates subaortic conus. Any arrangement other than isolated subpulmonary conus, i.e., bilateral conus, subaortic conus with absent subpulmonary conus, and bilaterally absent conus, is abnormal (**Capuani A et al., 1995).**

A subpulmonary conus is typically absent in the setting of transposition of great arteries (TGA), which in turn results in continuity between the pulmonary valve and the mitral valve, although this is not an essential morphological feature to define TGA. Similarly, bilateral conus is commonly associated with a double-outlet RV but is not necessary for the diagnosis. Thus, although the infundibulum provides an important clue about cardiac anatomy, it is not the defining feature of either ventricle or VA connection, and therefore, the morphology of the infundibulum should not be used to define the ventricle or VA connection(**Capuani A et al., 1995).**.

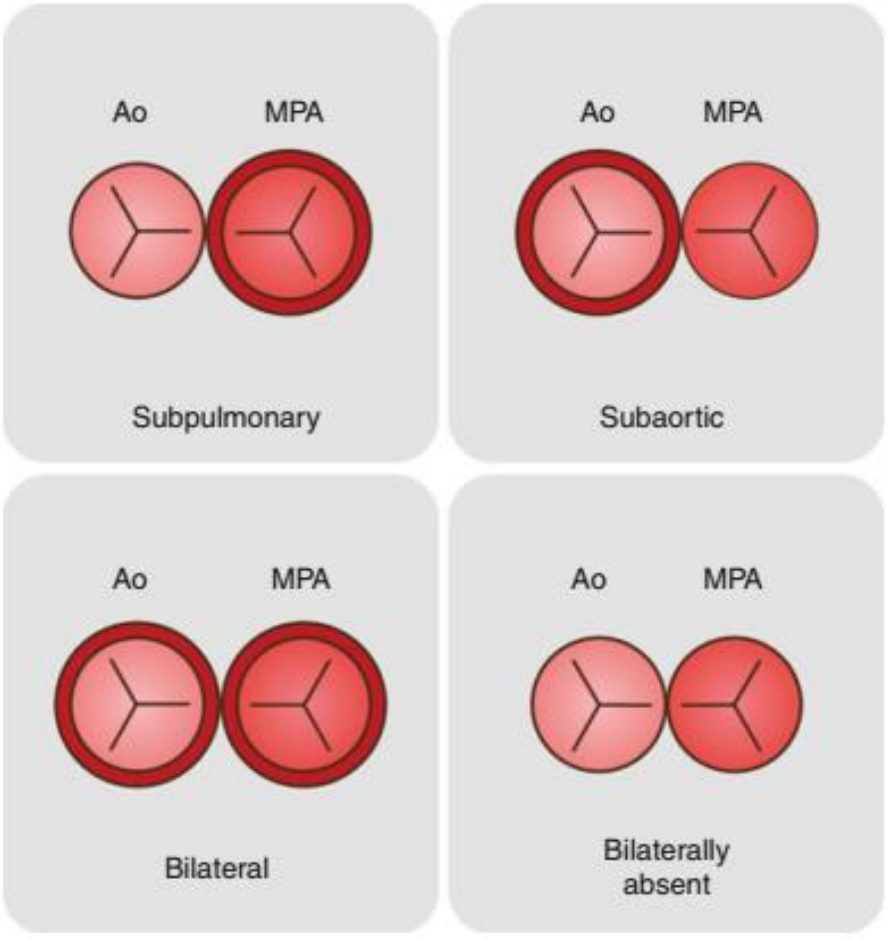


Fig. 31 : Normal conus anatomy: sub pulmonary (normal), sub aortic, bilateral, and bilaterally absent. Sub aortic conus is common in transposition, whereas bilateral conus is seen in cases of double-outlet right ventricle and bilaterally absent conus is noted in double-outlet left ventricle. In this picture, the conus is represented by the *dark, red ring*

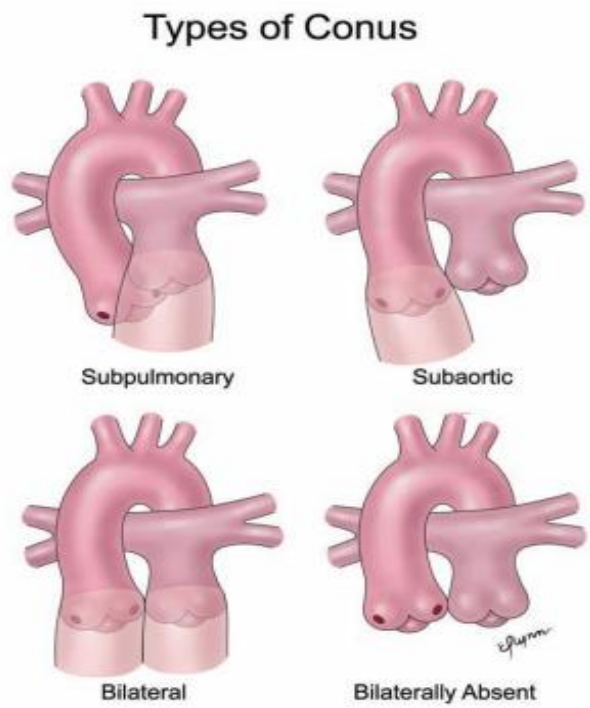


Figure 31 : Types of Conus

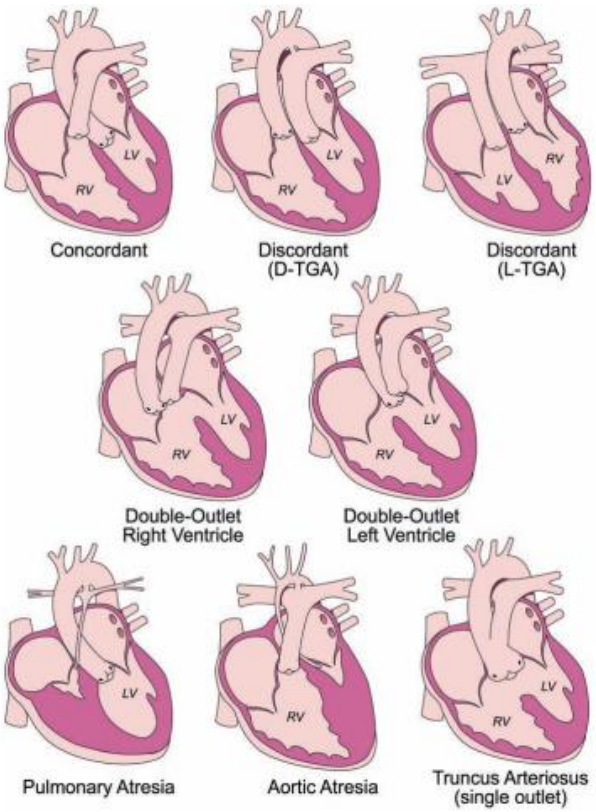
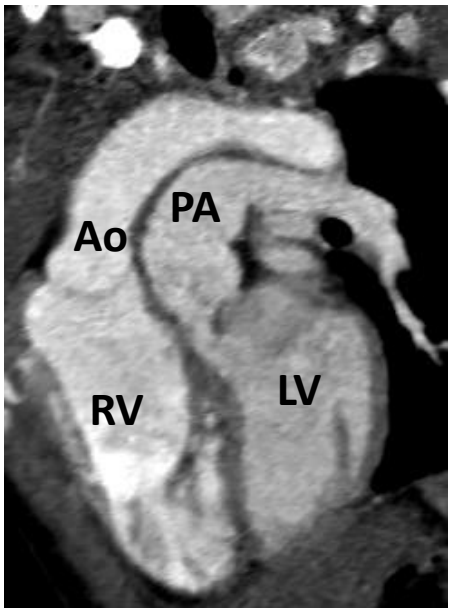
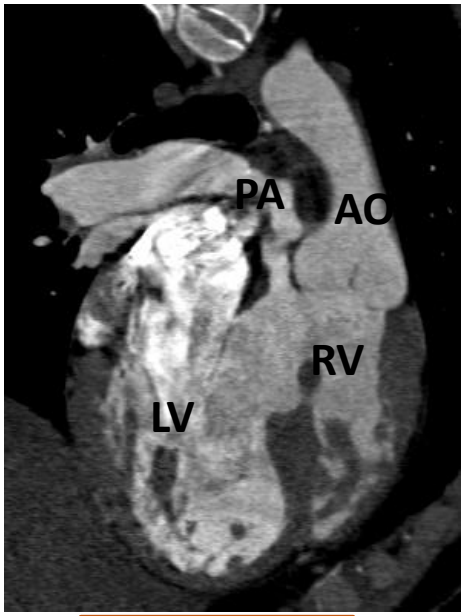


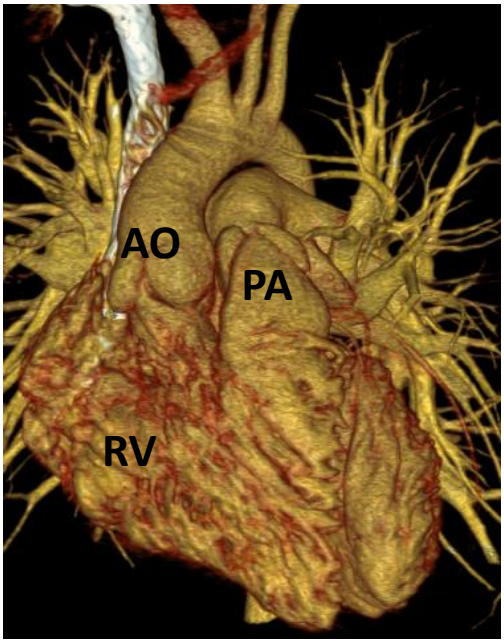
Figure 32 : shows types of Ventriculoarterial Connections



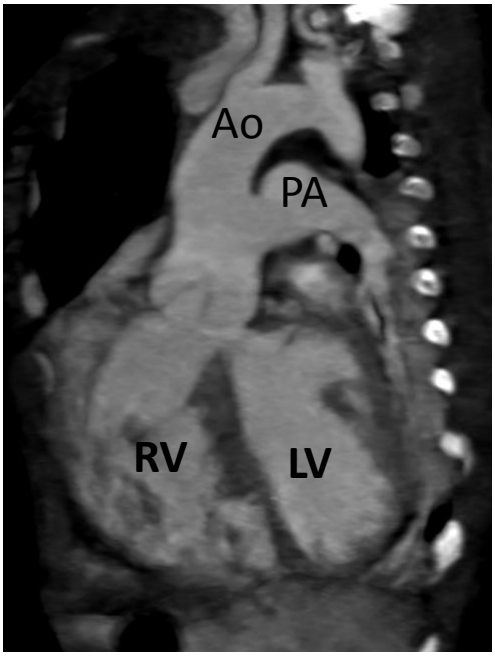
D-TGA



L-TGA



DORV



Truncus arteriosus

Figure 32 : shows types of Ventriculoarterial Connections

Atrioventricular Connections

Following the ventriculoarterial connection analysis, **step 4** also includes an evaluation of the atrioventricular connections. Biventricular atrioventricular connections refer to an arrangement where each atrium is connected to its own ventricle. Univentricular atrioventricular connections describe connections where only one ventricle is connected to the atrial mass. According to the approach developed by **Anderson and colleagues**, there are five types of atrioventricular connection: concordant (normal), discordant, ambiguous, double inlet, and absent right or left connection. With a normal or concordant connection, the right atrium drains into the right ventricle, and the left atrium drains into the left ventricle. With a discordant connection, the right atrium drains into the left ventricle, and the left atrium drains into the right ventricle. Transposition of the great vessels frequently occurs in association with discordant atrioventricular connection (**Anderson RH et al., 1984).**

In cases of heterotaxy, the connection is described as *ambiguous*. *Concordant*, *discordant*, and *ambiguous* may be used to describe the connections when two ventricles are present, whereas *double inlet* and *absent right* (or *left*) *connection* are used for a univentricular heart. For more precise description, the anatomy and the position of the atrioventricular valve annuli also may be described. **(Lapierre C et al., 2010).**

Connection-wise, as highlighted earlier, the AV valves are usually committed fully to one of the ventricles, although, in the setting of single-ventricle physiology, one of the valves may be atresia, e.g., tricuspid atresia and mitral atresia. In some cases, mostly in the presence of a VSD, the AV valve can be connected to both the ventricles. In this regard, the term overriding is used if the Valvular annulus overrides the ventricular septum. The degree of override

greater than 50% assigns the valve to the ventricle, receiving a greater share of the annulus. The AV valve is termed as straddling when the tensor apparatus is supported by the other ventricle, in addition to the ventricle with the dominant connection (**Milo S et al., 1979).**

The identification of straddling and overriding of AV valve is important as it is often associated with hypoplasia of the ipsilateral ventricle precluding biventricular surgical repair. Rarely, the AV connection can have both atriums connected to one ventricle (double-inlet ventricle) or one atrium connecting to both the ventricles (double-outlet atrium) creating single-ventricle physiology (**Smallhorn JF et al., 1981)**

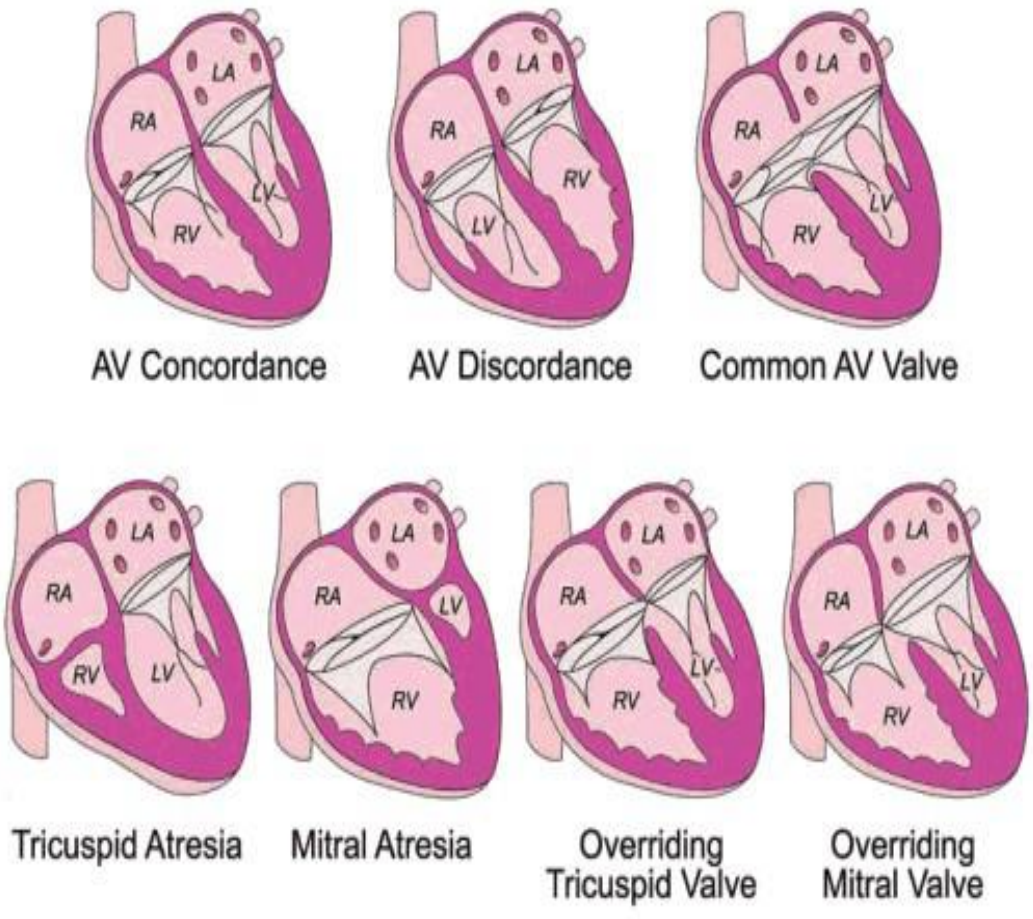
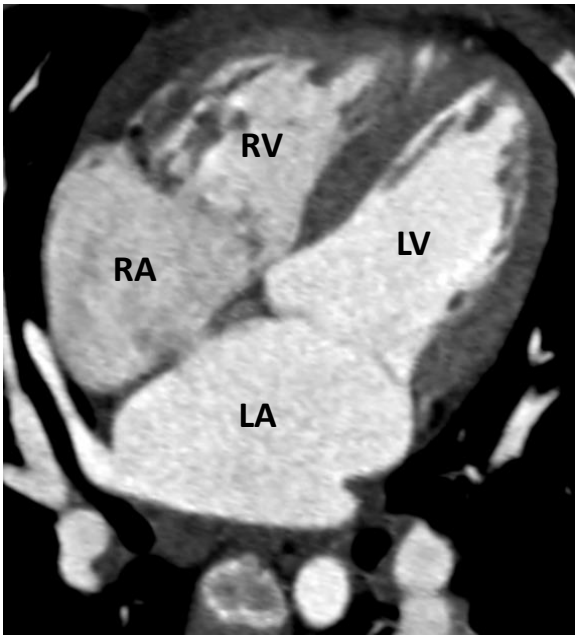
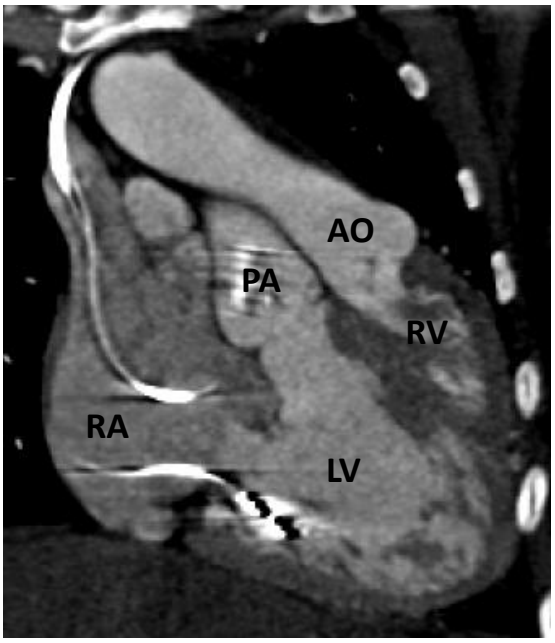


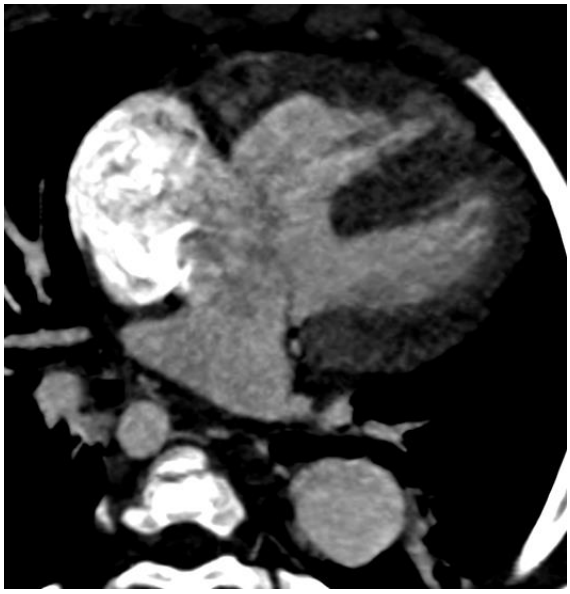
Figure 33 : types of atrioventricular connections



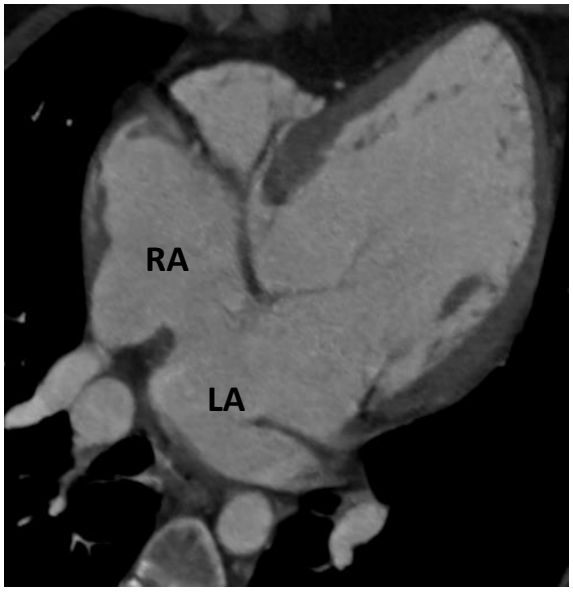
Concordant



Discordant; cc-TGA



CAVC



Tricuspid atresia

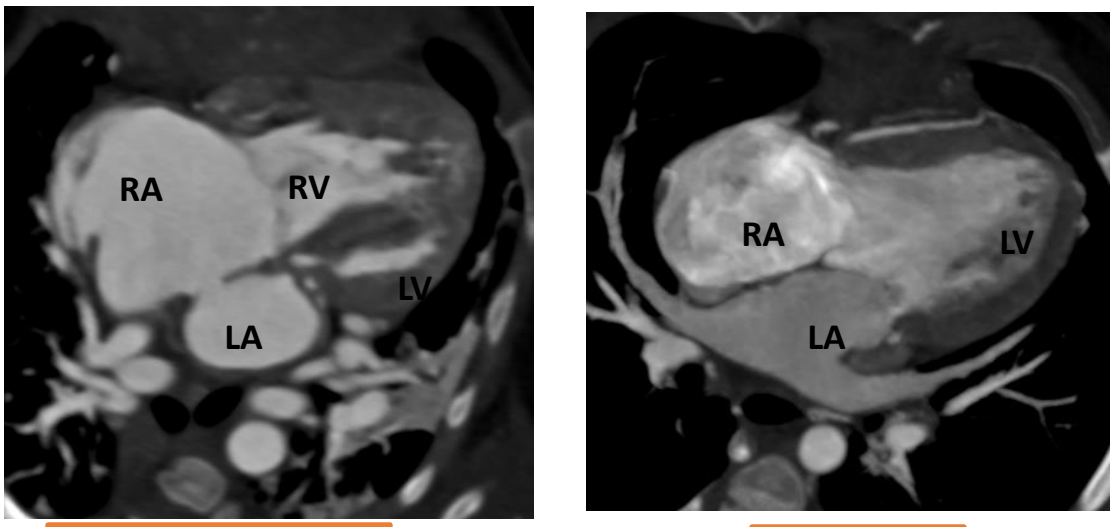


Figure 33 : types of atrioventricular connections

Assessment of Associated Malformations

Associated malformations need to be assessed in a segmental fashion at three levels: the heart, great vessels, and coronary arteries. At the level of the heart, the presence, size, and location of atrial and ventricular septal defects, the size of the cardiac chambers, and the presence and severity of ventricular out flow obstructions need to be documented. Great vessel anatomy needs to be reviewed for the presence of stenotic or hypoplastic segments as well as the presence of a patent ductus arteriosus. Next, an evaluation of systemic and pulmonary venous return should be performed. Coronary anatomy and the presence of coronary anomalies need to be noted as well, and coronary artery disease needs to be described according to published guidelines. Finally, the presence and any complications of palliative shunts, closure devices, and stents need to be reported. **(Van Praagh R, 1984).**

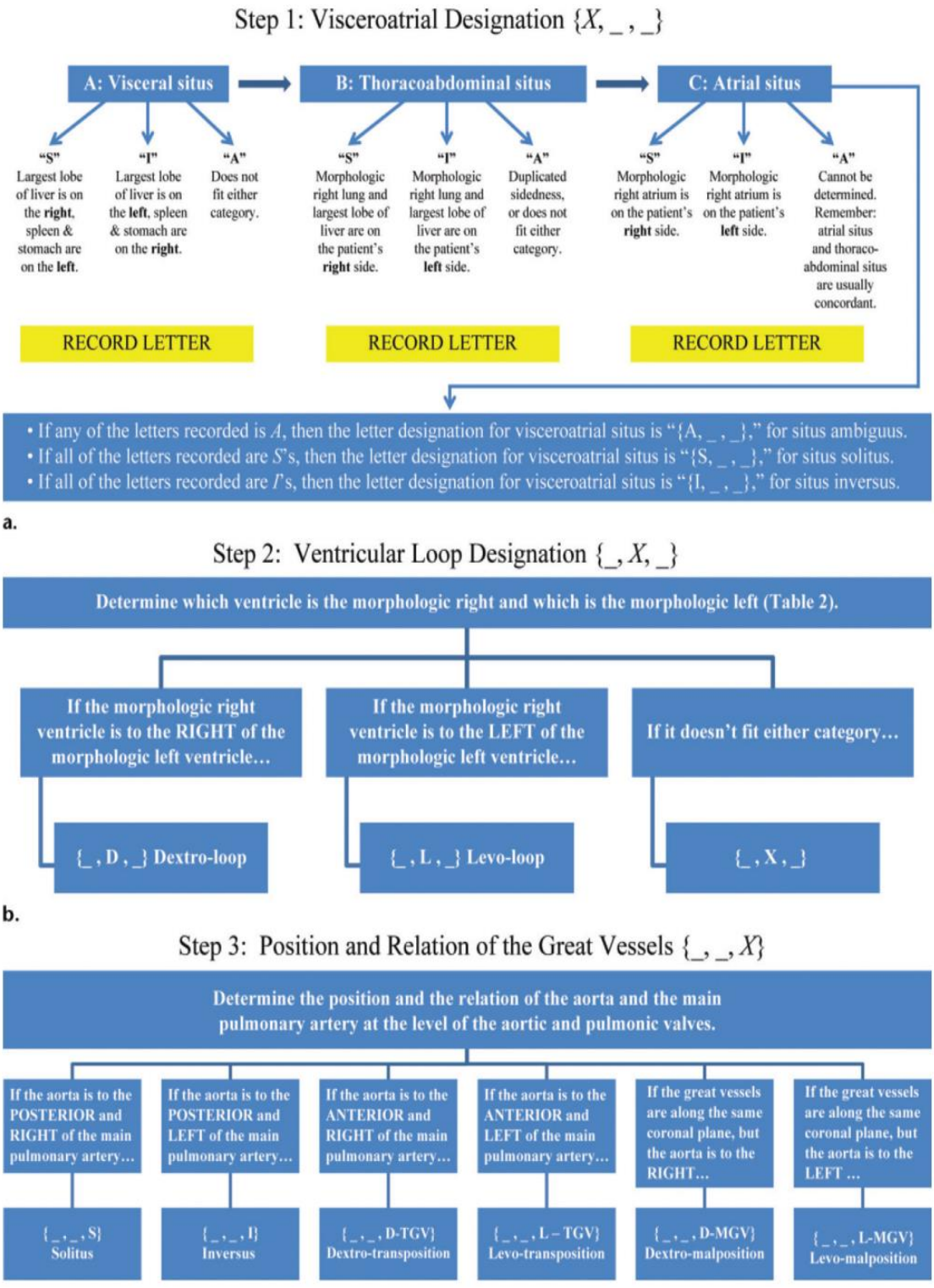
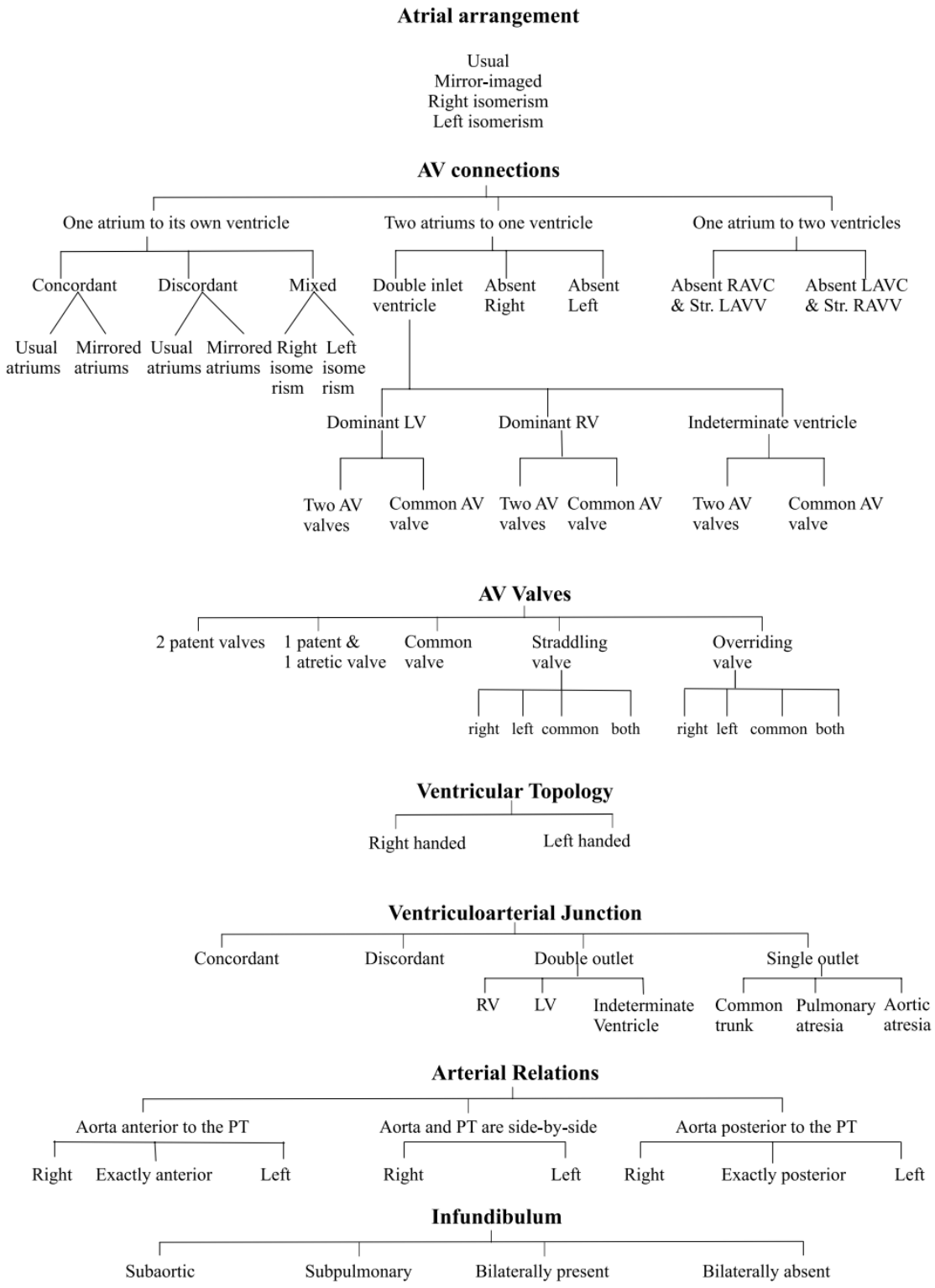


Figure 35 : Flowcharts show the three steps in reporting the **Van Praagh** segmental classification of congenital heart disease: describing the visceroatrial situs {*X*, _, _} **(a)**, describing the ventricular loop orientation {_, *X*, _} **(b)**, and describing the position and relation of the great vessels {_, _, *X*}



Flow diagram: Variable features in sequential segmental analysis

Refrences

Anderson RH, Becker AE, Freedom RM, et al. Sequential segmental analysis of congenital heart disease. Pediatr Cardiol 1984;5(4):281–287.

Anderson RH, Shirali G (2009) Sequential segmental analysis. Ann Pediatr Cardiol 2(1):24–35.

Anderson RH, Shinebourne EA, Gerlis LM. Criss-cross atrioventricular relationships producing paradoxical atrioventricular concordance or discordance. Their significance to nomenclature of congenital heart disease. Circulation 1974;50:176-80.

Anderson RH, Wessels A, Vettukattil JJ. Morphology and morphogenesis of atrioventricular septal defect with common atrioventricular junction. World J Pediatr Congenit Heart Surg 2010;1:59-67.

Asferg C, Usinger L, Kristensen TS, et al. Accuracy of multi-slice computed tomography for measurement of left ventricular ejection fraction compared with cardiac magnetic resonance imaging and two-dimensional transthoracic echocardiography: a systematic review and meta-analysis. Eur J Radiol. 2012;81:e757–62

Capuani A, Uemura H, Ho SY, Anderson RH. Anatomic spectrum of abnormal ventriculoarterial connections: Surgical implications. Ann Thorac Surg 1995;59:352-60.

De la Cruz MV, Espino-Vela J, Attie F et al (1967) An embryological theory for the ventricular inversions and their classification. Am Heart J 73:777–793

Edwards WD (2008) Classification and terminology of cardiovascular anomalies. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF (eds) Moss and Adams' heart disease in infants children and adolescents, 7th edn. Lippincott Williams & Wilkins, Philadelphia, pp 34–56

Goo Hyun Woo. Cardiac MDCT in children: ct technology overview and interpretation. Radiol Clin North Am 2011;49:997–1010.

Jacobs JP, Anderson RH, Weinberg PM, et al. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. Cardiol Young. 2007;17 suppl 2:1–28

Kelly Han B, Rigsby CK, Hlavacek A, et al. Computed tomography imaging in patients with congenital heart disease, Part I: Rationale and Utility, An Expert Consensus Document of the Society of Cardiovascular Computed Tomography (SCCT). J Cardiovasc Comput Tomogr. 2015;9:475–92.

Khatri S, Varma SK, Khatri P, et al. 64-slice multidetector-row computed tomographic angiography for evaluating congenital heart disease. Pediatr. Cardiol. 2008;29():755–62.

Lapierre C, Dery J, Guerin R, Viremouneix L, Dubois J, Garel L. Segmental approach to imaging of congenital heart disease. Radiographics. 2010;30:397–411.

Linda C, Pamela T, Elliot K. Cardiac CT angiography beyond the coronary arteries: what radiologists need to know and why they need to know it. AJR2014;203:W583–95

Männer J. The anatomy of cardiac looping: a step towards the understanding of the morphogenesis of several forms of congenital cardiac malformations Clin Anat 2009;22(1):21–35.

Milo S, Ho SY, Macartney FJ, Wilkinson JL, Becker AE, Wenink AC, *et al*. Straddling and overriding atrioventricular valves: Morphology and classification. Am J Cardiol 1979;44: 1122-34.

Moller JH, Nakib A, Anderson RC, Edwards JE. Congenital cardiac disease associated with polysplenia: a developmental complex of bilateral "left sidedness". Circulation. 1967;36(5):789–99.

Orwat S, Diller GP, Baumgartner H. Imaging of congenital heart disease in adults: choice of modalities. Eur Heart J Cardiovasc Imaging.2013:15(1):6-17

Praagh V. Normally and abnormally related great arteries. What have we learned. World J Pediatr Congenit Heart Surg. 2010;1(3):364–85.

Smallhorn JF, Tommasini G, Macartney FJ. Detection and assessment of straddling and overriding atrioventricular valves by two dimensional echocardiography. Br Heart J 1981;46:254-62.

Stanger P, Rudolph AM, Edwards JE. Cardiac malpositions: an overview based on study of sixty five necropsy specimens. Circulation 1977;56(2):159–172.

Tynan M, Becker A, Macartney F et al (1979) Nomenclature and classification of congenital heart disease. Br Heart J 41:544–553

Van Mierop LH, Eisen S, Schiebler GL. The radiographic appearance of the tracheobronchial tree as an indicator of visceral situs. Am J Cardiol. 1970;26(4):432–5

Van Praagh R. The segmental approach to diagnosis in congenital heart disease. In: Bergsma D, ed. Birth defects original article series, vol 8, no. 5. National Foundation—March of Dimes. Baltimore: Williams & Wilkins, 1972; 4–23.

Van Praagh R. The story of anatomically corrected malposition of the great arteries [editorial]. Chest 1976;69(1):2–4

Van Praagh R. Terminology of congenital heart disease: glossary and commentary. Circulation 1977;56 (2):139–143

Van Praagh R. The importance of segmental situs in the diagnosis of congenital heart disease. Semin Roentgenol 1985;20(3):254–271

Van Praagh R, David I, Gordon D, Wright GB, Van Praagh S. Ventricular diagnosis and designation. In: Godman MJ, ed. World congress, London, 1980. Paediatric cardiology, vol 4. Edinburgh, Scotland: Churchill Livingstone, 1981; 153-168.

Van Praagh R, Layton WM, Van Praagh S. The morphogenesis of normal and abnormal relationships between the great arteries and the ventricles: pathologic and experimental data. In: Van Praagh R, Takao A, eds. Etiology and morphogenesis of congenital heart disease. Mount Kisco, NY: Futurama, 1980; 271-31