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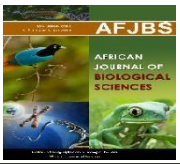

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Detection Of Tricuspid Valve Lesions in Functional Single Ventricle

Nada Abozaid Elsayed Abozaid, Elsayed Hamed Zidan, Samar Mohammed Shehata, Rabab Mohammed Abdelhay

Radiodiagnosis Department, Faculty of Medicine, Zagazig University, Egypt

Email:nada.abozayed@medicine.zu.edu.eg, nodaabozaid@gmail.com

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Abstract:Background: Tricuspid valve lesions are commonly encountered in patients with functional single ventricle, a complex congenital heart defect where only one ventricle is able to effectively pump blood to the body, In cases of FSV with tricuspid valve lesion a Univentricular connection is present: when the atria are connected predominantly with one ventricular chamber due to absence of one AV valve (tricuspid valve) or the presence of two valves or a common valve predominately draining in one ventricle of left ventricular morphology (double inlet connection). In both conditions, the left ventricle is of good size and the right ventricle is hypoplastic, missing the inlet portion, and the two ventricular cavities are separated by an interventricular septum. Echocardiography as well as cardiac catheterization are common cardiac imaging modalities. Both modalities have drawbacks; Echocardiography includes operator dependence, limited acoustic shadows, a small field of view, and poor evaluation of pulmonary veins. Cardiac catheterization includes the overlapping of great vessels, difficulty in demonstrating systemic and pulmonary vessels at the same time, catheter-related complications, high doses of iodinated contrast material and ionizing radiation. Cardiac computed tomography (CT) can overcome these limitations. Multidetector computed tomography (MDCT) has recently expanded as advanced imaging modality for structural heart disease imaging.

Keywords:*Tricuspid valve lesions, functional single ventricle*

Introduction

Tricuspid valve lesions are commonly encountered in patients with functional single ventricle, a complex congenital heart defect where only one ventricle is able to effectively pump blood to the body **(1)**. The tricuspid valve, which controls blood flow from the right atrium to the right ventricle, can become dysfunctional in these patients **(2).**

Defects in the atrial septum, which separates the right and left atria, can also contribute to tricuspid valve dysfunction in single ventricle patients. The position and size of septal defects within the oval fossa can

impact the anatomy and function of the tricuspid valve, making it challenging to achieve an effective repair. Furthermore, the presence of a thick Eustachian valve or displacement of the oval fossa can further complicate the anatomy and limit options for transcatheter closure of septal defects **(3).**

In cases of FSV with tricuspid valve lesion a *Univentricular* connection is present: when the atria are connected predominantly with one ventricular chamber due to absence of one AV valve (tricuspid valve) or the presence of two valves or a common valve predominately draining in one ventricle of left ventricular morphology (double inlet connection). In both conditions, the left ventricle is of good size and the right ventricle is hypoplastic, missing the inlet portion, and the two ventricular cavities are separated by an interventricular septum (2).

These lesions include tricuspid atresia occur in about 1 per 10,000 live births. Ebstein anomaly occurs in about 0.5 per 10,000 live births with gender predilection. However, with the maternal use of lithium, the Ebstein anomaly can increase nearly seven-fold. Double inlet left ventricle occurs in up to 0.01 per 10,000 live births. Atrioventricular canal defect occurs in 0.03 to 0.04 per 10,000 live births **(4).**

Echocardiography as well as cardiac catheterization are common cardiac imaging modalities. Both modalities have drawbacks; Echocardiography includes operator dependence, limited acoustic shadows, a small field of view, and poor evaluation of pulmonary veins. Cardiac catheterization includes the overlapping of great vessels, difficulty in demonstrating systemic and pulmonary vessels at the same time, catheter-related complications, high doses of iodinated contrast material and ionizing radiation. Cardiac computed tomography (CT) can overcome these limitations **(5).**

Multidetector computed tomography (MDCT) has recently expanded as advanced imaging modality for structural heart disease imaging **(6)**. The 3-dimensional reconstruction of cardiac anatomy is cornerstone in diagnosis of congenital heart diseases which is limited in other imaging modalities **(7).**

We aimed to Accurately delineate the tricuspid valve lesions associated with functional single ventricle.

Embryology of the heart

The heart is the first organ to form and become functional, emphasizing the importance of transport of material to and from the developing infant. It originates about day 18 or 19 from the mesoderm and begins beating and pumping blood about day 21 or 22. It forms from the cardiogenic region near the head and is visible as a prominent heart bulge on the surface of the embryo. Originally, it consists of a pair of strands called cardiogenic cords that quickly form a hollow lumen and are referred to as endocardial tubes. These then fuse into a single heart tube and differentiate into the truncus arteriosus, bulbus cordis, primitive ventricle, primitive atrium, and sinus venosus, starting about day 22. **(8)**

The heart tube continues to elongate and begins looping at around day 23 of development. The bulbus cordis moves ventrally, caudally, and to the right (forward, down and right), and the caudal portion – the primitive ventricle – moves dorsally, cranially and to the left (backwards, up and left). This process produces a shape that is much closer to the fully developed heart. **(9)**

Disturbances will lead to abnormal looping that can vary from random, anterior to leftward looping resulting in: abnormal atrial situs (situs inversus or isomerism), dextrocardia, and ventricular inversion. It is important to realize that only the atria of the heart may present with isomerism, whereas the ventricles never show heterotaxia patterns.**(10)**

The five regions of the primitive heart tube develop into recognizable structures in a fully developed heart: **(fig.1).**

1. The **truncus arteriosus** will eventually divide and give rise to the ascending aorta and pulmonary trunk.
2. The **bulbus cordis** develops into the trabeculated part of right ventricle and smooth outflow tracts.
3. The **primitive ventricle** forms the left ventricle; however, the primitive ventricle is separated from the next expansion (i.e., the bulbus cordis) by the bulbo-ventricular sulcus; the latter segment will form much of the right ventricle.

4. The **primitive atrium** becomes the anterior portions of both the right and left atria, and the two auricles.
5. The **sinus venosus** is originally formed of two horns, right and left. The right horn grows larger than the left horn. As the heart develops, the sinus venosus gets integrated into the wall of the right atrium as posterior smooth portion of the right atrium (sinus venarum). The rest of the internal surface of the right atrium and auricle has a thicker, trabeculated appearance; originate from the primordial atrium. The transition from the smooth to the rough internal surface of the right atrium is demarcated on the inside of the atrium by a ridge called the crista terminalis, and on the outside by a groove called the sulcus terminalis. The smaller left horn develops into the coronary sinus. **(11)**

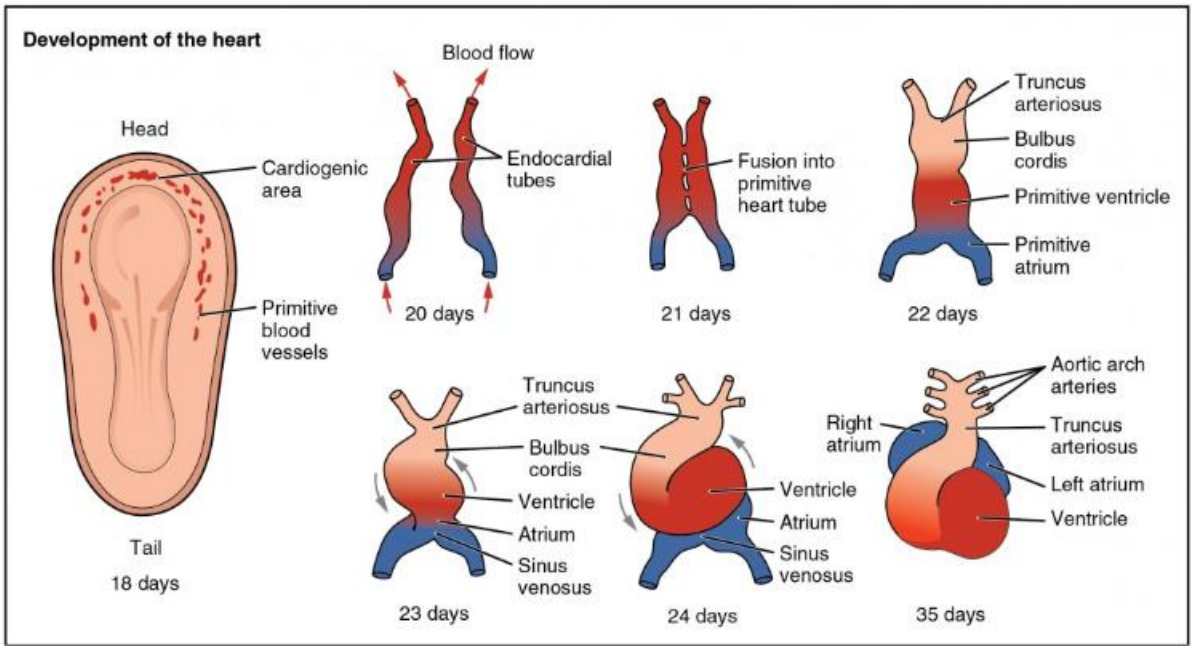

Figure 1: *heart tube formation and looping process.* ***(J. Gordon Betts, 2013)***

The Primordial Heart Septation:

Partitioning of the AV canal, primordial atrium, ventricle, and outflow tract begins during the middle of the fourth week. Partitioning is essentially completed by the end of the 8th week.

1. **Septation of the AV canal:**

At the end of the fourth week, two mesenchymal cushions, the atrioventricular endocardial cushions, appear at the superior and inferior borders of the atrioventricular canal. As well as the two lateral atrioventricular cushions appear on the right and left borders of the canal. The superior and inferior cushions, in the meantime, project further into the lumen and fuse, resulting in a complete division of the canal into right and left atrioventricular orifices by the end of the fifth week **(Fig. 2)**. **(12)**

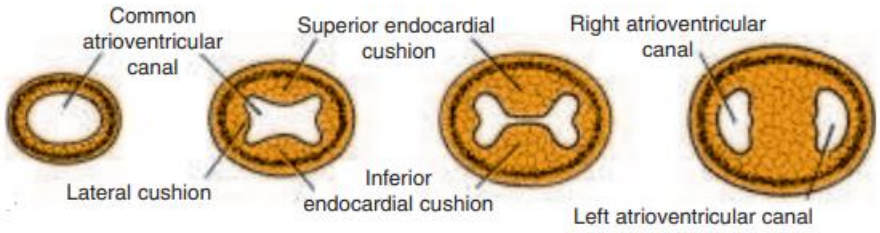

Figure 2 *formation of septum in atrioventricular canal.* ***(Morphologia, 2019)***

2. **Septation of the primordial ventricle:**

Minor trabeculations appear during early development of the primordial ventricle. Following growth of the ventricles further trabeculations appear and grow as larger, muscular structures. Some researchers believe that as the trabeculations grow they coalesce resulting in the formation of the ventricular septum. However, the more commonly described theory of septation begins with the appearance of a primordial muscular interventricular (IV) ridge developing in the floor of the ventricle near the apex. As either side of the ventricle grows and dilates, their medial walls fuse forming the prominent IV septum. The foramen located between the cranial portion of the IV septum and the endocardial cushions: the IV foramen, closes by the end of the seventh week as the bulbar ridges (formed by active proliferation of neural crest cells and continuous in truncus arteriosus) fuse with the endocardial cushions, forming the membranous IVS.

The mature heart at 9 weeks' gestation shows the following structures: (13)

1. A venous pole that is now connected to systemic veins from the upper body (superior vena cava), lower body (inferior vena cava), liver (hepatic veins), and coronary circulation (coronary sinus), all of which pass into the right atrium.
2. A separate left atrium, connected to the venous system separately by ingrowth of pulmonary veins from the lungs.
3. An atrioventricular canal that begins as a single unrestricted opening and septates into two atrioventricular valves. The tricuspid valve opens from right atrium into right ventricle, and the mitral valve opens from left atrium to left ventricle.
4. A primitive ventricular segment that has grown in cell number and by cell hypertrophy into two distinct and separate chambers, a right and left ventricle. By processes known as compaction and trabeculation, the working myocardium of each ventricle has become highly organized and adapted to the unique requirement of a pulmonary (right) or systemic (left) ventricle.
5. An arterial pole that has separated in a spiral fashion to create two separate outflow tracts of the heart. The right ventricle retains the proximal part of the original bulbus cordis connection as the conus; thus, its structure differs from that of the left ventricle.

Anatomy of the heart:

The heart has the shape of a pyramid, with its apex pointing towards the left nipple while its base forms the posterior surface of the heart. Other surfaces are the anterior, inferior (or diaphragmatic), and two pulmonary surfaces facing the lungs. Its primary role is to receive the blood from the body, pump it to the lungs to be oxygenated, and receive it once more to pump it again to the rest of the human body tissues. The right side of the heart (which consists of the right atrium and the right ventricle) receives the desaturated blood, while the left side (consisting of the left atrium and left ventricle) receives the oxygenated blood. **(14)**

The pericardium is a thick membrane that covers the heart. It consists of two layers: the fibrous pericardium and the serous pericardium. The serous pericardium is thin and covers the heart, also called the epicardium. The fibrous pericardium is much thicker. Together they form the pericardial cavity, a thin sac hosting a small amount of fluid. **(15)**

The heart has four chambers: the left and right atriums and the left and right ventricles. They form a shallow groove at the line of their junction, which form the atrioventricular groove. The atrioventricular groove hosts major coronary arteries while they travel along to the line of attachment of atrioventricular valves. The right and left ventricles are separated by a septum, which corresponds to the interventricular grooves that travel from the posterior to the anterior surface of the heart.

- **CT anatomy of the Ventricles:**

1. **Right ventricle:**
   - The right ventricle is shaped differently to the left ventricle: the left ventricle is cylindrical in shape and the cavity of the right ventricle is effectively wrapped around it. The right ventricle also has a thinner wall which is more trabecularized, especially towards the apex.
   - It consists of an inlet portion that receives blood from the right atrium through the tricuspid valve and a muscular outlet portion (infundibulum) that pumps the blood to the pulmonary artery. **(14)**

- The **moderator band** is another distinguishing feature of the right ventricle. It runs from the septum to the lateral wall of the right ventricle **(Fig. 2)**
- **Crista supraventricularis** is a muscular ridge within the right ventricle of the heart. It is located between the tricuspid and pulmonic valves, at the junction of the right ventricular anterior (free) wall and the interventricular septum **(Fig. 3)**. **(16)**

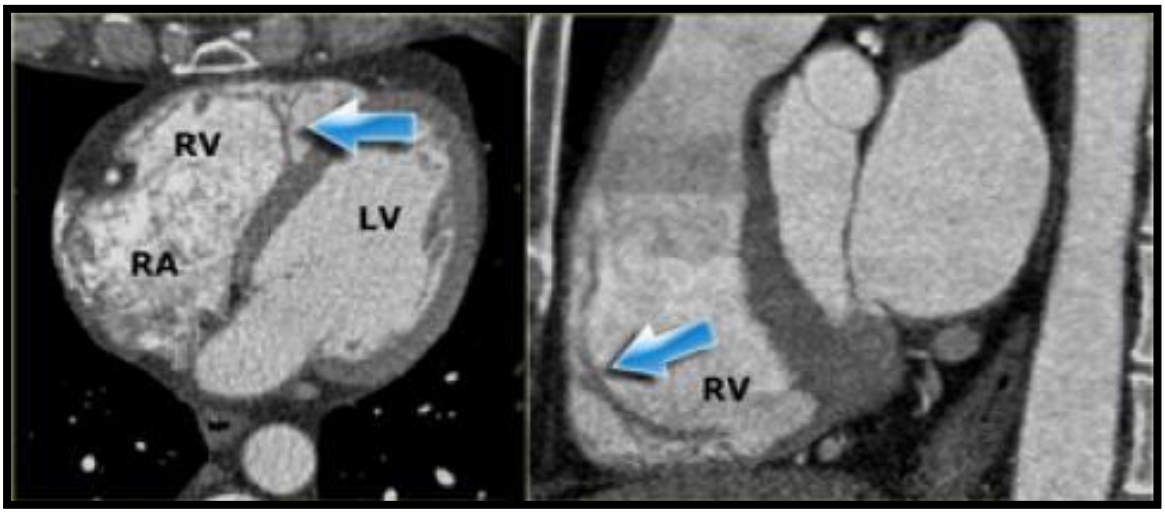

Figure 3: *Axial (left) and sagittal oblique (right) reconstructions showing the right ventricle. The blue arrows indicate the moderator band. RA=right atrium, RV=right ventricle, LV=left ventricle.*

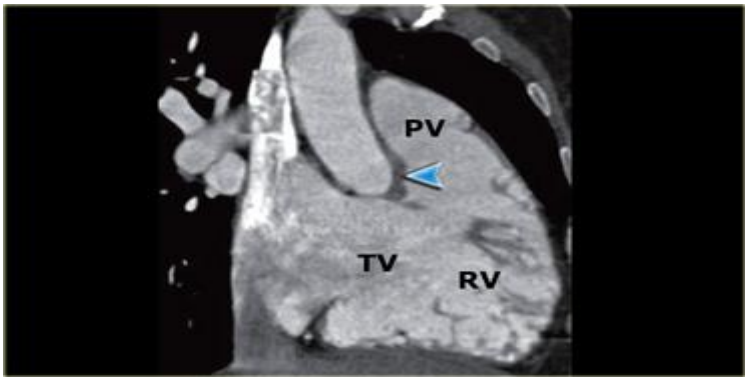

Figure 4: *Reconstruction showing the tricuspid (TV) and pulmonary (PV) valves as well as the cavity of the right ventricle (RV). The blue arrow indicates the crista supraventricularis.*

Left ventricle:

- The left ventricle is made of thick muscle walls because a lot of power is needed to push blood to the arterial system of the body.
- It is conical in shape, and it is longer than the right ventricle in length.
- It also occupies part of the anterior (sternocostal), inferior (diaphragmatic), and left wall of heart.
- The left ventricle forms the apex of the heart, and it receives blood from the left atrium through the mitral valve and pumps it to the body through the aortic valve **(Fig. 4)**.
- It consists of an inlet portion (ostium venosum), an outlet portion (ostium arteriosum), and an apical portion. The apical portion is conical and consists of fine trabeculations. **(14)**
- The left ventricle has a uniform thickness, varying end-diastolically from 0.6 to 1.0 cm. **(17)**

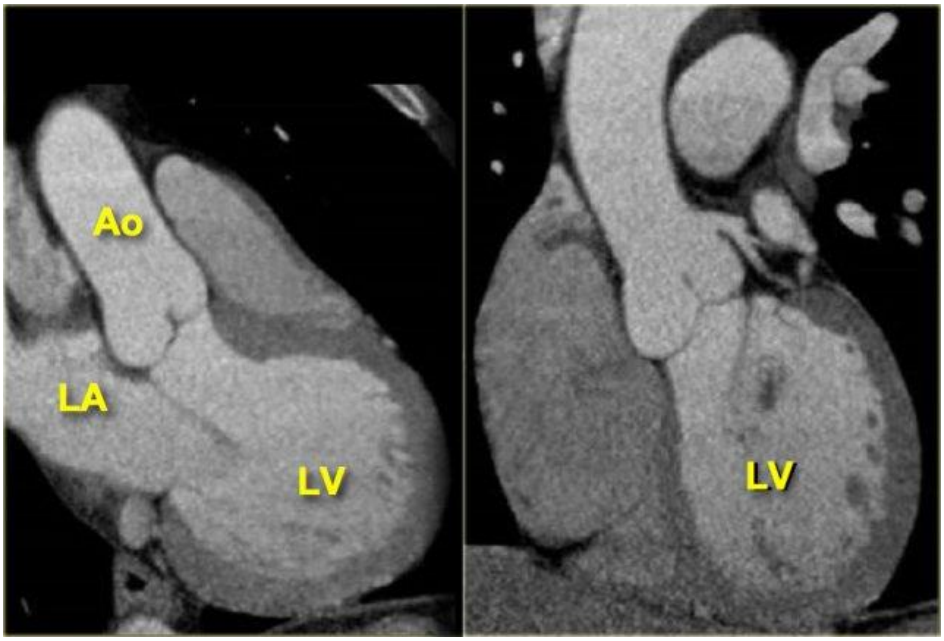

Figure 5: *3-chamber view (middle) and coronal reconstruction (right) of the heart illustrating the relationships between the left atrium, ventricle, and aortic root. LA=left atrium, Ao=aorta, LV=left ventricle.* ***(Willems et al,2009).***

2. **There are several characteristics which are specific for the right ventricle and are useful in distinguishing the right from the left ventricle: (18)**
    a. The infundibulum:
        i. 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 supraventricular crest is a prominent muscular structure that forms the posterior wall of the RV outlet and separates the inlet from the outlet components of RV. The tricuspid and pulmonary valves are widely separated by this muscular wall. **(19)**
        ii. The left ventricular outlet is partially fibrous due to the aortic-mitral fibrous continuity. The aortic and mitral valves are adjacent to each other and connected by a fibrous band.
    b. Tricuspid valve apparatus:
        i. 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 tricuspid valve is identified by its caudal location below the membranous septum.
        ii. 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).
    c. Apical trabeculations:
        i. 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.
        ii. The left ventricular trabeculations are thinner, delicate structures.
    d. Moderator band: 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 1: *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.
Thinner walls.	Thick walls.

3. Segmentation of the left ventricle:

All methods of cardiac imaging, from ventriculography to scintigraphy, subdivide the left ventricle into a series of slices or segments. The resolution of the segmental approach is a function of the number of segments; thus, it can range from 20% (in the 5-segment model) to 5% (in the 20-segment model). However, increasing the number of segments, and thus reducing their size, leads to an unacceptable complication in the analysis with a greater need for approximation and interpolation. A reasonable trade-off between accuracy and feasibility is represented by the 16-segment model proposed by the American Heart Association, recently modified to include the 17th segment, i.e., the true apex. **(20)**

17-Segment Model (AHA)

Left Ventricle Segmentation Procedure

The muscle and cavity of the left ventricle can be divided into a variable number of segments. Based on autopsy data the AHA recommends a division into 17 segments for the regional analysis of left ventricular function or myocardial perfusion: **(Fig. 5) (21)**

- The left ventricle is divided into equal thirds perpendicular to the long axis of the heart. This generates three circular sections of the left ventricle named basal, mid-cavity, and apical. Only slices containing myocardium in all 360° are included.
- The basal part is divided into six segments of 60° each. The segment nomenclature along the circumference is: basal anterior, basal anteroseptal, basal infero-septal, basal inferior, basal inferolateral, and basal anterolateral. The attachment of the right ventricular wall to the left ventricle can be used to identify the septum.
- Similarly, the mid-cavity part is divided into six 60° segments called mid anterior, mid anteroseptal, mid infero-septal, mid inferior, mid inferolateral, and mid anterolateral.
- Only four segments of 90° each are used for the apex because of the myocardial tapering. The segment names are apical anterior, apical septal, apical inferior, and apical lateral.
- The apical cap represents the true muscle at the extreme tip of the ventricle where there is no longer cavity present. This segment is called the apex.

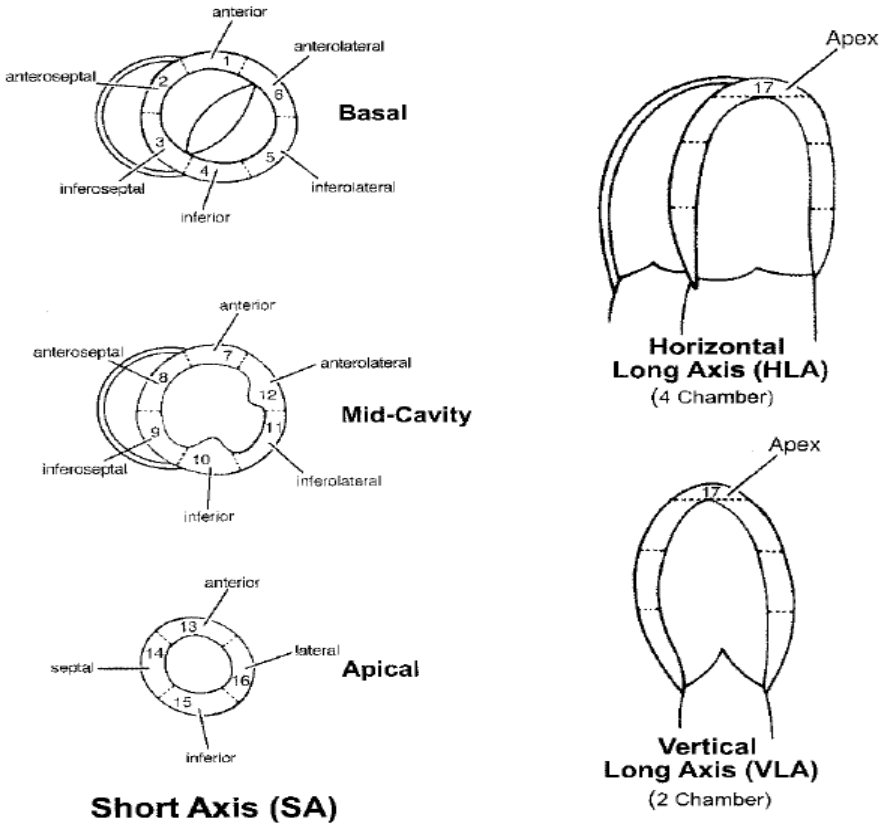

Figure 6:*Diagram of vertical long-axis (VLA, approximating the 2-chamber view), horizontal long-axis (HLA, approximating the 4-chamber view), and short-axis (SA) planes showing the name, location, and anatomic landmarks for selection of the basal (tips of the mitral valve leaflets), mid-cavity (papillary muscles), and apical (beyond papillary muscles but before cavity ends) short-axis slices for the recommended 17-segment system.* ***(Cerqueira, Weissman et al. 2002).***

Polar Plots

If functional values have been obtained in the 17 cardiac segments by some quantification method, they can be arranged as a polar plot with the:

- apex in the center,
- the four apical segments as a first ring,
- the six mid-cavity segments as the second ring,
- and the six apical segments as the outermost ring.

Such an arrangement makes it easy to compare the outcome in different conditions (eg. rest/stress) or between patients. The arrangement together with numbers identifying the cardiac segments is illustrated below **(Fig. 6)**.

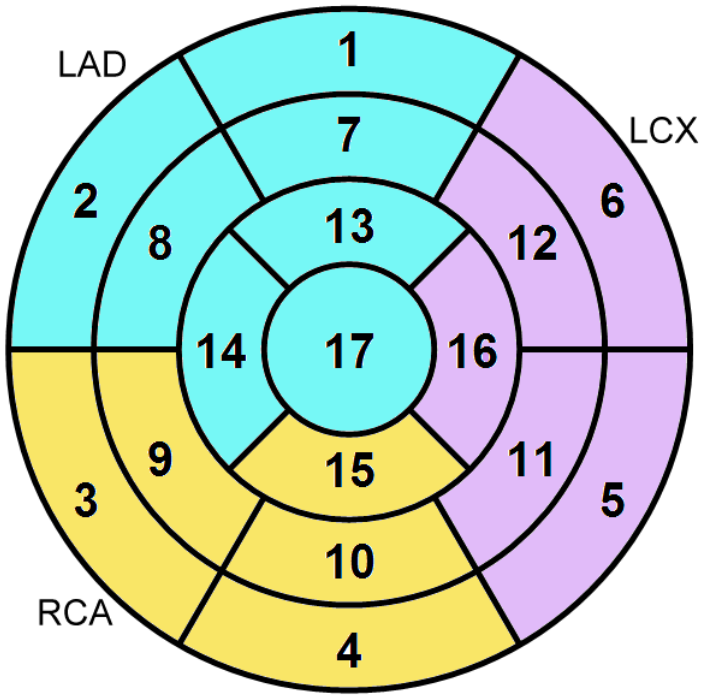

Figure 7: *Diagram displaying a circumferential polar plot, of the 17 myocardial segments.*

Table 2: *The17 segments of left ventricle:*

	Basal Segments		**Mid-cavity Segments**		**Apical Segments**
1.	basal anterior	7.	mid anterior	13.	apical anterior
2.	basal anteroseptal	8.	mid anteroseptal	14.	apical septal
3.	basal infero-septal	9.	mid infero-septal	15.	apical inferior
4.	basal inferior	10.	mid inferior	16.	apical lateral
5.	basal inferolateral	11.	mid inferolateral	17.	apex
6.	basal anterolateral	12.	mid anterolateral		

The relative contribution of the basal, mid-cavity, and apical segments are 35% (6/17), 35% (6/17), and 30% (5/17), respectively.

Coronary Artery Territories

The AHA guidelines emphasize that there is a "tremendous variability in the coronary artery blood supply to myocardial segments". The greatest variability occurs at the apical cap, which can be supplied by any of the three arteries. With the recognition of the anatomic variability the individual segments may be assigned to specific coronary artery territories as follows: **(Fig.7)**. **(21)**

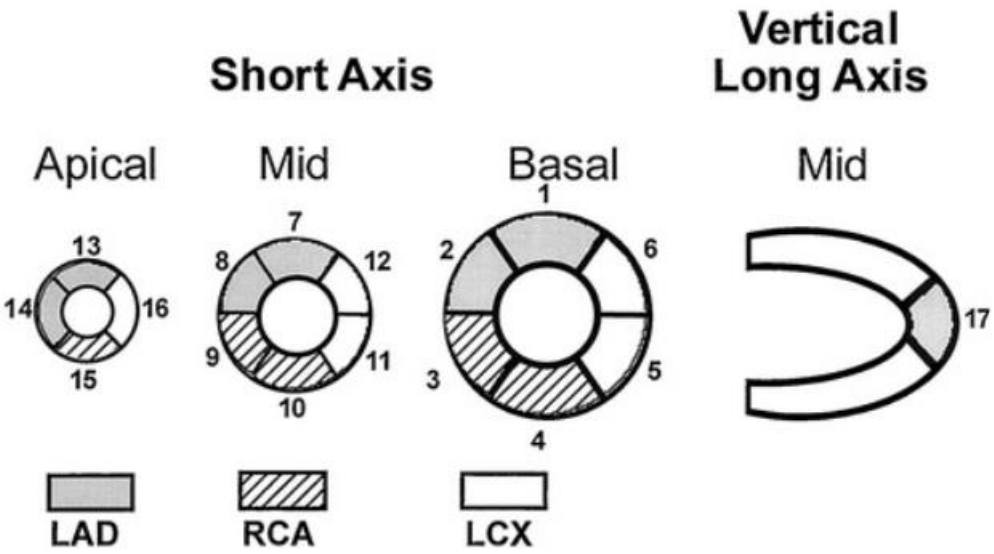

Figure 8: *Assignment of the 17 myocardial segments to the territories of the left anterior descending (LAD), right coronary artery (RCA), and the left circumflex coronary artery (LCX).* ***(Cerqueira, Weissman et al. 2002).***

The 17-segment model meets the basic requirements of any reasonable segmentation: it is simple enough to be employed in practice; it has an anatomical basis; there is good correspondence with the distribution of coronary arteries. There are at least two good reasons to accept the 17-segment system, updating the 16-segment system. First, the 16-segment system did not include a true apical myocardial segment devoid of cavity – with the development of echocardiographic contrast agents for the assessment of myocardial perfusion, the myocardial apex segment or apical cap beyond the left ventricular cavity becomes pertinent, and a 17-segment model may become more appropriate for both the assessment of wall motion and myocardial perfusion with echocardiography. The second – and more important – reason to adopt the 17-segment model is that it proposes a standardized myocardial segmentation and nomenclature for tomographic imaging of the heart that is shared by all imaging modalities: nuclear cardiology, cardiovascular magnetic resonance, cardiac computed tomography, positron emission tomography, and coronary angiography. **(20)**

- **Cardiac valves:**

They are specialized flap or cusp-like structures, located on each end of the two ventricles that maintain the directed one-way flow of the blood inside the heart. Normal valves have three flaps, except the mitral valve, which has two flaps. The mitral and tricuspid atrio-ventricular (AV) valves separate the atria from the ventricles, whereas the aortic and pulmonary semilunar (SL) valves separate the ventricles from the great arteries. **(22)**

1. Tricuspid valve:

- The tricuspid valve is located posteriorly and to the right of the aortic valve.
- The tricuspid valve complex consists of the tricuspid annulus, tricuspid valve leaflets, chordae tendineae, and papillary muscles.
- There are three tricuspid valve leaflets: the anterior, posterior, and septal leaflets.
- The tricuspid valve leaflets attach to papillary muscles (three papillary muscles: anterior, posterior, and septal papillary muscles) via the chordae tendineae.
- During ventricular diastole, the tricuspid valve leaflets open to allow blood flow from the right atrial chamber into the right ventricular chamber. During ventricular diastole, the tricuspid valve leaflets close to prevent regurgitated blood flow into the right atrial chamber **(Fig. 8)**. **(14)**

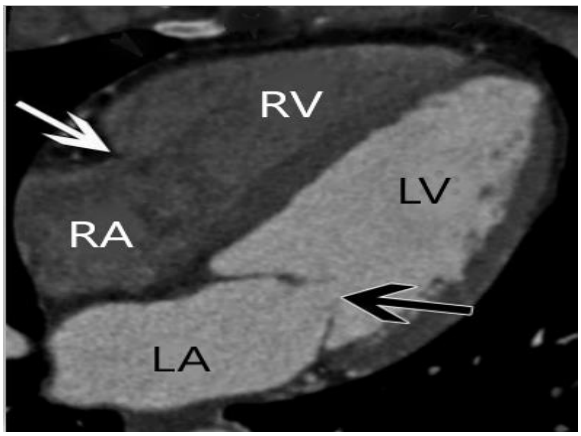

Figure 9: *Horizontal long-axis MPR image shows the left ventricle (LV), right ventricle (RV), left atrium (LA), right atrium (RA), mitral valve (MV: black arrow), tricuspid valve (white arrow).* ***(O'Brien et al, 2007) (23).***

Functional Single Ventricle (FSV):

A congenital heart disease is defined as a structural abnormality of the heart and (or) great vessels that is present at birth **(24)**. Congenital heart disease is the most common congenital anomaly, occurring in almost 1% of live births. **(25)**

Functional single ventricle, or functionally univentricular heart, refers to a group of patients with congenital cardiac malformations with one ventricle being small, incomplete, underdeveloped and/or functionally inadequate. **(26)**

Pathology of Functional single ventricle:

Normally the vena caval blood is emptied into the right atrium (RA), and from there is passed on to the RV, and pulmonary artery (PA) and is transported into the lungs for oxygenation. From there, it is returned to the LA, LV, and aorta and is passed on to the body for delivery of oxygen and nutrients. The resultant systemic arterial saturation is ≥96%. The blood is returned back to the vena cavae, and the cycle is repeated. **(Fig. 9A).**

By contrast, in babies with single ventricle, the systemic (SBF) and pulmonary (PBF) blood flow returns mix with each other in the single functioning ventricle with consequent lower systemic oxygen saturations. The single ventricle then provides both the SBF and PBF. This admixture in the single ventricle causes systemic arterial oxygen desaturation. The O2 saturations vary between 75% and 85%, depending upon the quantity of PBF and pulmonary to systemic flow ratio (Qp:Qs). **(Fig. 9).(1)**

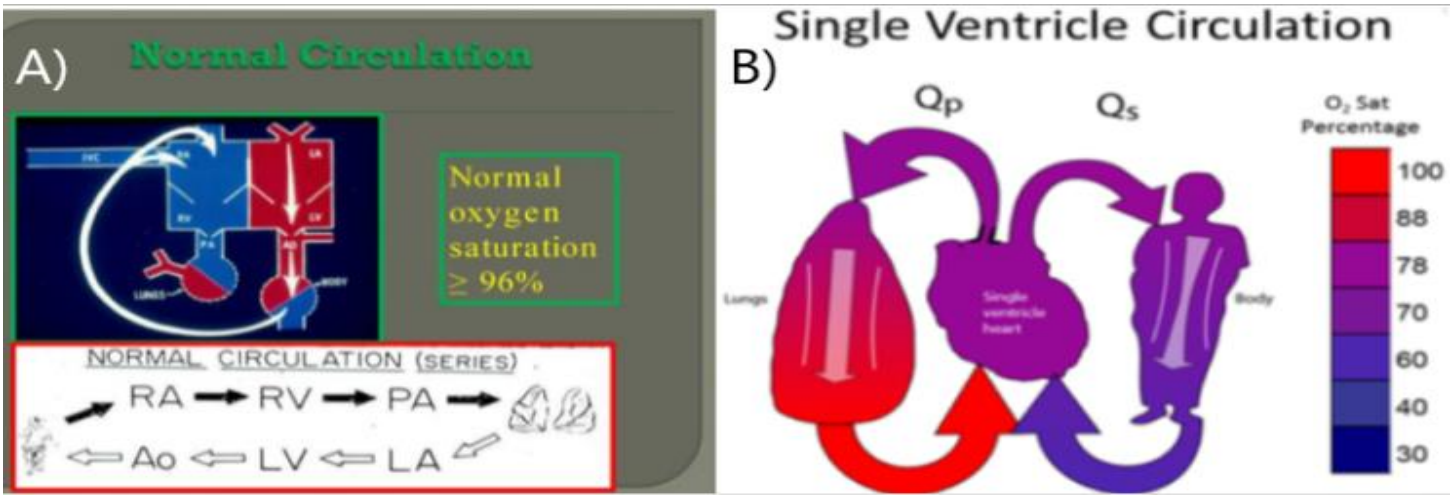

Figure 9: Diagram showing: A) normal circulation, B) Single ventricle circulation. (Rao 2021).

Image findings of tricuspid valve lesions in functional single ventricle:

1. Hypoplastic right heart syndrome (HRHS):

HRHS is not a commonly used generic term like hypoplastic left heart syndrome. Hypoplastic right heart syndrome (HRHS) is characterized by under-development of the tricuspid and/or pulmonary valves and of the right ventricle (RV) and pulmonary artery with right to left shunting through inter-atrial communication. The extreme forms are associated with tricuspid and/or pulmonary atresia. There may be other multiple associated congenital cardiac defects. The more common variety i.e. tricuspid atresia. **(27).**

A case of 2-weak-old female child presented with cyanosis, lethargy, difficult breathing, and poor feeding. Clinical examination revealed abnormal heart murmurs and decreased O_2 saturation. **(Figure 10).**

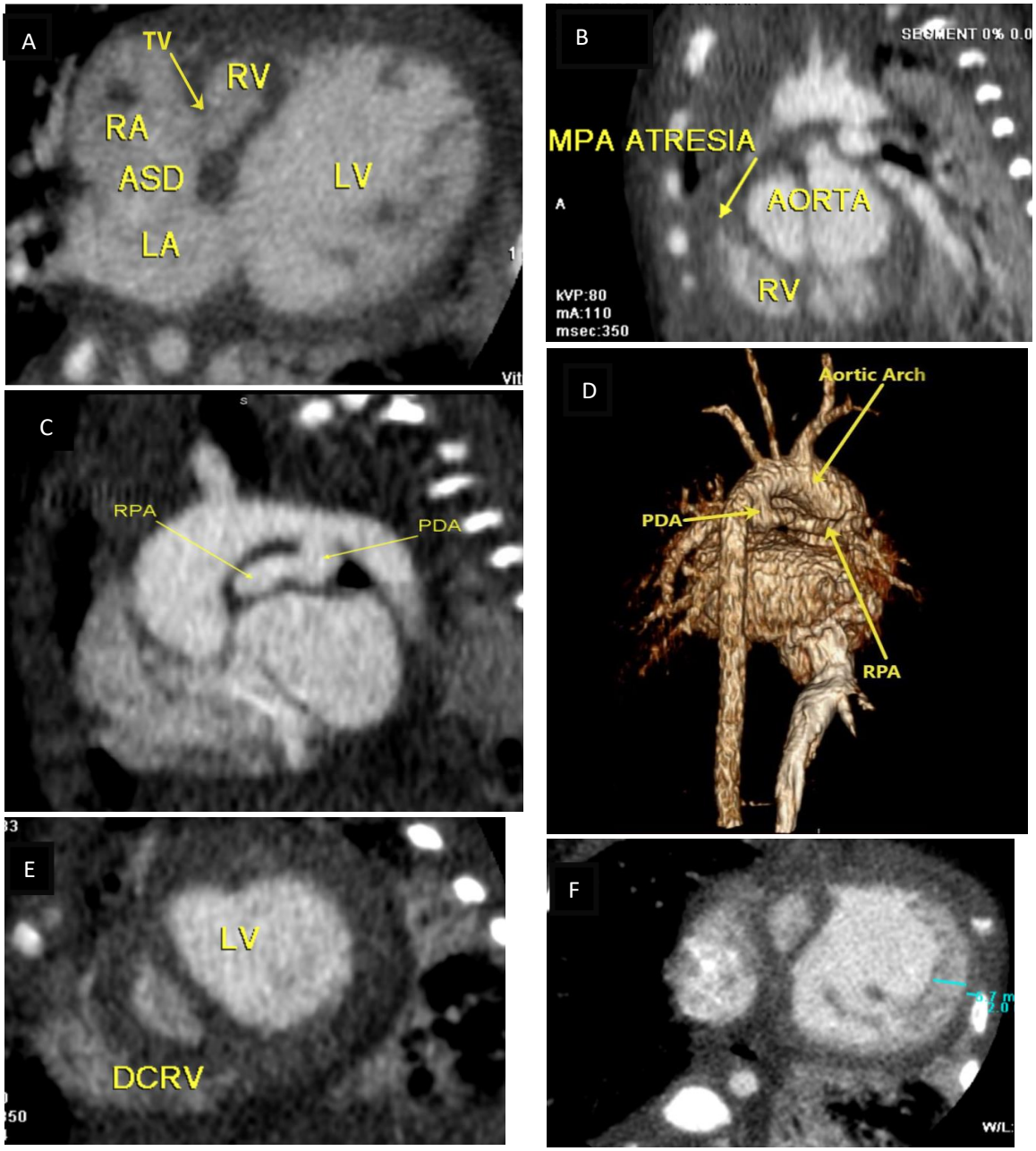

Figure 10: Hypoplastic right heart syndrome with functional single ventricle of left ventricular morphology

A. **Axial oblique MIP image** of 4-chamber view shows enlarged right atrium (RA) with wide ASD, tricuspid valve (TV) stenosis, hypo-plastic right ventricle (RV) and hypertrophied left ventricle (LV).

B. **Sagittal MIP image** show atretic main pulmonary artery (MPA atresia) arising from hypoplastic right ventricle (RV).

C. **Sagittal Oblique MIP image** shows straight PDA supplying right pulmonary artery (RPA).

D. **3D volume rendering (VR) image (Posterior view)** shows normal branching of the left sided aortic arch, PDA arising from inferior surface of aortic arch to confluent right and left pulmonary arteries.

E. **Short axis view** shows double chambered hypoplastic right ventricle (DCRV).

F. **Short axis view** shows left ventricular non -compaction with non-compaction/compaction (NC/C) ration =2.85 at diastolic phase.

2. Tricuspid atresia (TA):

CT has proven to be an excellent imaging modality to determine the anatomic features of tricuspid atresia. The classic finding is fatty tissue between the right atrium and the right ventricle in the expected location of the tricuspid valve, a small right ventricle, right atrial and left ventricular enlargement. Other features include atrial and/or ventricular septal defects. **(Fig. 11). (28)**

- **Etiology:** Tricuspid atresiaoccurs due to the disruption of the normal development of the atrioventricular valves from the endocardial cushion. **(29)**
- **Classification:** TA is classified based on the associate abnormalities, and it was proposed by Rao: **(30)**

•**Type I:** Normally related great arteries.
•**Type II:** D-transposition of the great arteries (TGA).
•**Type III:** Other than type II malposition of the great arteries.
•**Type IV:** Persistent truncus arteriosus.

- Each type can be further subdivided into three types according to the pulmonary arteries' status:

•**Subtype a:** Pulmonary atresia.
•**Subtype b:** Pulmonary stenosis or pulmonary hypoplasia.
•**Subtype c:** Normal anatomy of the pulmonary arteries.

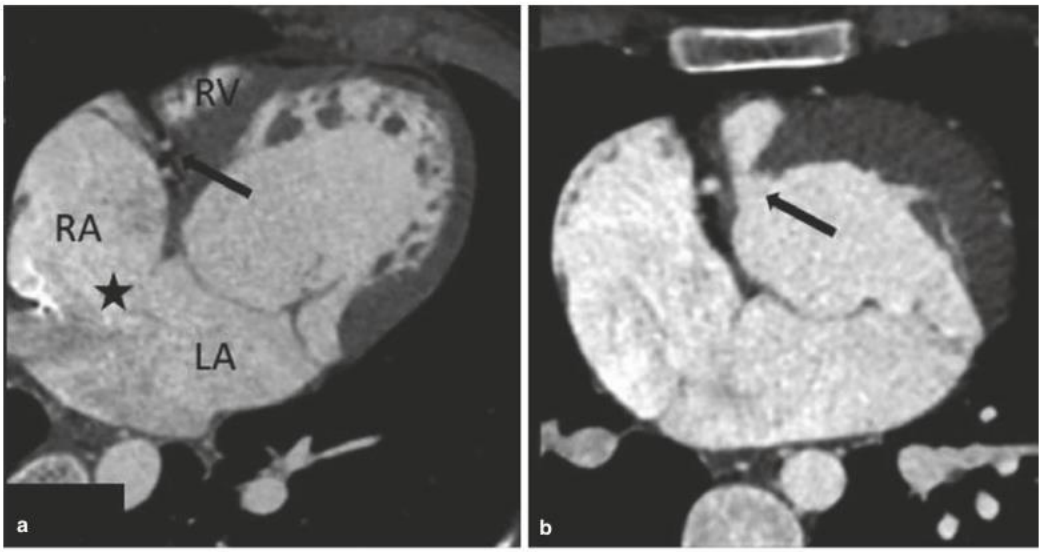

Figure 11: *Cardiac CT axial images shows: (a) ASD (black star) and presence of fat in AV right groove (Black arrow). (b) Restrictive VSD (black arrow) and hypoplastic right ventricle.* ***(Nagulakonda and Malhi 2023).***

3. Ebstein's Anomaly of the Tricuspid Valve:

Ebstein anomaly is easily detected by CT. Imaging findings include a large right atrium proper, a dilated atrialized right ventricle, a small true right ventricle along with apical displacement of the septal leaflet of the tricuspid valve, and a "sail-like" anterior tricuspid valve leaflet deformity. The atrialized ventricular wall may be thinner than the distal functional right ventricle, and the heart may rotate posteriorly into the left hemithorax with bulging of the ventricles posteriorly. An atrial septal defect is an associated finding. **(18)(Fig. 12)**

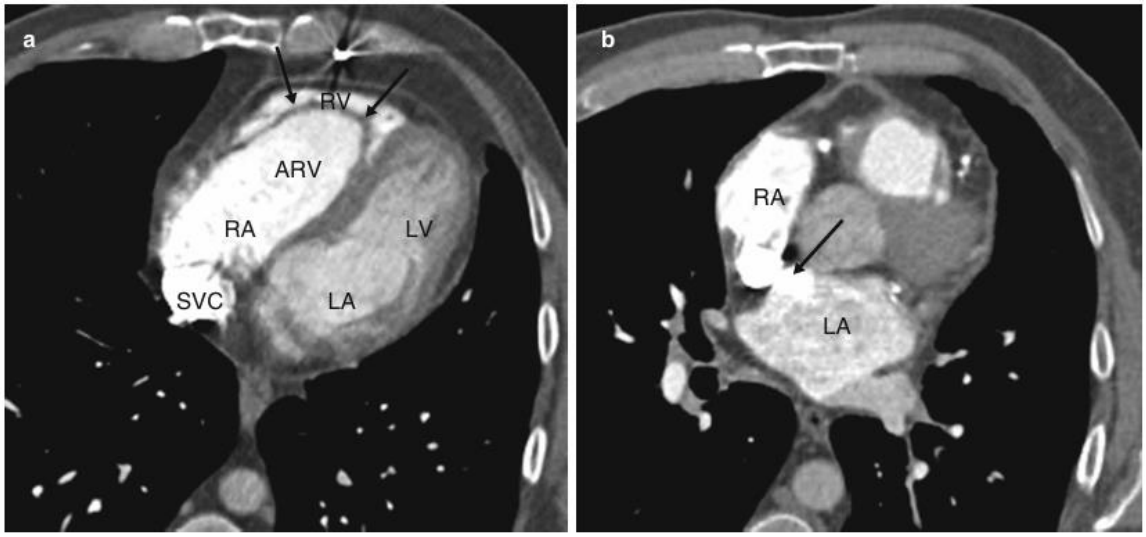

Figure 10: *Ebstein anomaly, (a) is an axial tomogram showing marked apical displacement of the septal tricuspid leaflet (arrows) resulting in atrialization of the right ventricle (ARV). (b) is an axial cut at a more superior level demonstrating an atrial septal defect LA with contrast-enhanced blood (arrow) flowing into the left atrium (LA) from the right atrium (RA). SVC superior vena cava, RV rightventricleproper.* ***(Mazur, Siegel et al. 2013).***

4. Unbalanced atrio-ventricular defect:

Cases of common AV canal in which there is marked discrepancy in the amount of attachment of the common valve to the two ventricles are designated as unbalanced AV canal. Frequently the ventricle with fewer attachments is also hypoplastic which could be of right or left ventricular morphology, but this is not necessarily the case. In fact, even when both ventricles are normal in size, if the canal is significantly unbalanced (i.e. minimal attachment of the valve to one ventricle) this will still be a functional single ventricle, since the ventricle receiving few attachments cannot function independently because the inflow cannot be enlarged **(Fig. 13)**. **(31).**

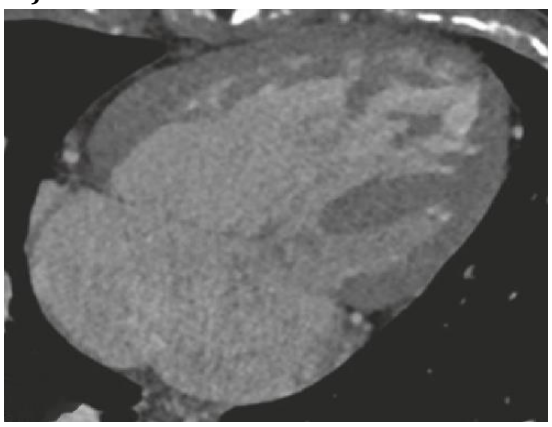

Figure 11: *Axial CT images show presence of unbalanced AVSD with RV larger than LV. (Right dominant).* ***(Nagulakonda and Malhi 2023).***

5. **Double inlet ventricle:** is the condition in which both atria are predominantly connected to one ventricle. The mode of atrioventricular connection can be through two atrioventricular valves or a common valve **(Fig. 14).** In the most common form, both atria connect to a dominant left ventricle in the presence of an anteriorly situated rudimentary right ventricle. However, the dominant ventricle may also be a right ventricle with a posterior rudimentary left ventricle or a solitary indeterminate ventricle. **(32)**

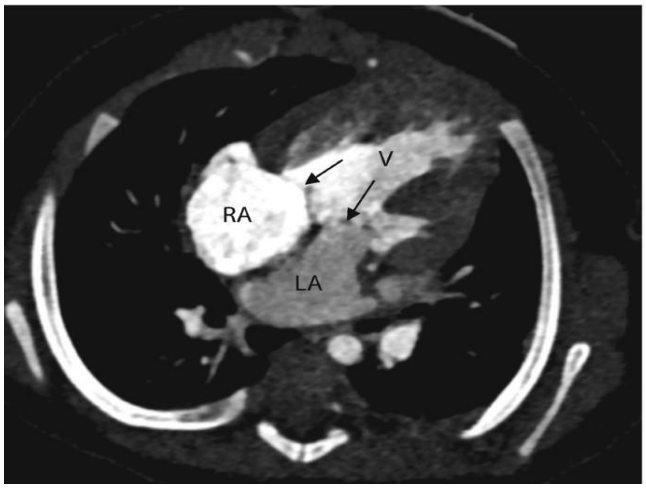

Figure 14: *Cardiac CT axial view showing double-inlet ventricle with both atrioventricular valves (arrows) communicating with common ventricle. RA right atrium, LA left atrium, V common ventricle.* ***(Kharouf and Adebo 2021).***

a. Double inlet left ventricle (DILV): is the majority of double inlet ventricle cases: **(33)**

- Both AV valves empty into a large ventricle of LV morphology, and there is generally a smaller RV outflow chamber located anteriorly. In most cases of DILV, it is very difficult to determine which AV valve is tricuspid, and which is mitral.
- **There are three general classes of DILV in the series of single ventricles analyzed by Van Praagh et al:**
  - i. L-transposed great arteries; the most frequent (38%).
  - ii. D-transposed great arteries; 2nd most common (25%): the aorta arising from a rightward, RV outflow chamber.
  - iii. Normally related great arteries; the least common (15%). Also known as Holmes' heart.

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