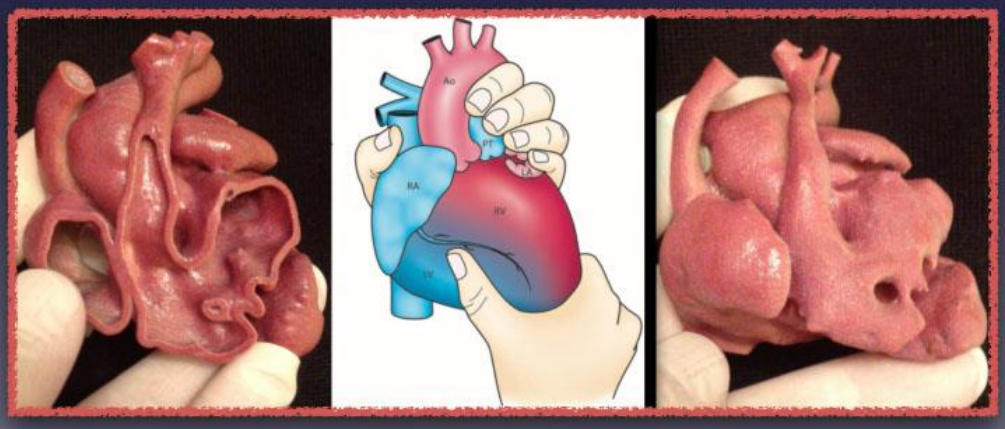
Most Peculiar Hearts in Your Hands

Criss-cross, superoinferior, twisted, topsy-turvy, etc. What do they all mean?



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List of Abbreviations

Ao, ascending aorta
AV, aortic valve
CS, crista supraventricularis
DORV, double outlet right ventricle
LA, left atrium
LAA, left atrial appendage
LBA, left brachiocephalic artery
LCCA, left common carotid artery
LSA, left subclavian artery
LV, left ventricle
MV, mitral valve
OS, outlet septum
PDA, patent ductus arteriosus
PT, pulmonary arterial trunk
PV, pulmonary valve
RA, right atrium
RAA, right atrial appendage
RBA, right brachiocephalic artery
RCCA, right common carotid artery
RSA, right subclavian artery
RV, right ventricle
TV, tricuspid valve
VIF, ventriculoinfundibular fold
VSD, ventricular septal defect

CHAPTER 1. OVERVIEW

INTRODUCTION AND HISTORIC BACKGROUND

In most hearts, the atria connect to the ventricles through the atrioventricular valves in a parallel fashion, which allows visualization of the four chambers of the heart in a horizontal long axis plane called a 'four-chamber view'. The parallel nature of the atrioventricular connection is present in most hearts regardless of the type and mode of the given atrioventricular connection including double inlet right or left ventricle and the hearts with a common atrioventricular valve (**Figures 1-1 and 1-2, upper panels**). In addition, the great arterial trunks usually show predictable spatial relationship for the given segmental connections. When the segmental connections are normal, the aorta and the pulmonary arterial trunk spiral each other with the aortic valve positioned rightward and posterior. The hearts with transposition of the great arteries typically show parallel great arteries. In most cases of complete transposition, the aorta is positioned rightward and anterior. In most cases of congenitally corrected transposition, the aorta is positioned leftward and anterior. Because of these classic spatial relationships of the great arteries in the majority of the hearts in situs solitus with transposition, complete and congenitally corrected transpositions are also called 'D-transposition' and 'L-transposition', respectively.

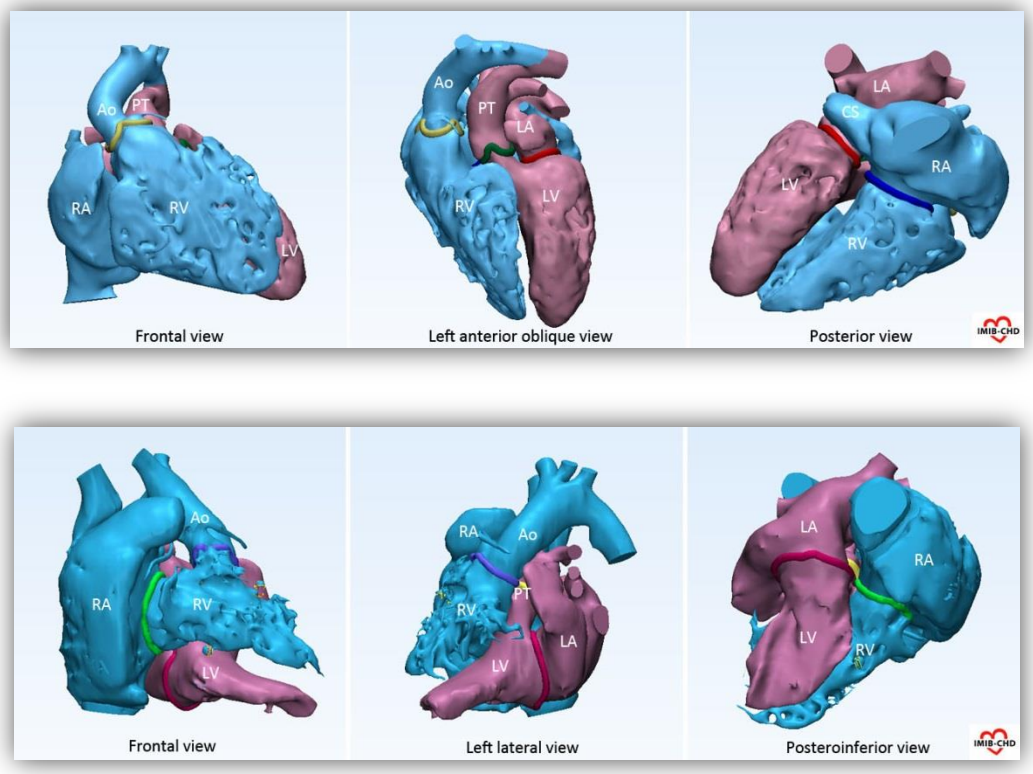

Figure 1-1. Volume rendered MR images of the cardiac cavities showing a classic form of complete transposition (upper panels) and complete transposition with twisted atrioventricular connection (lower panels). Note that the atrioventricular valves are arranged in parallel in the classic case, while they are not parallel in the atypical case with a twisted connection.

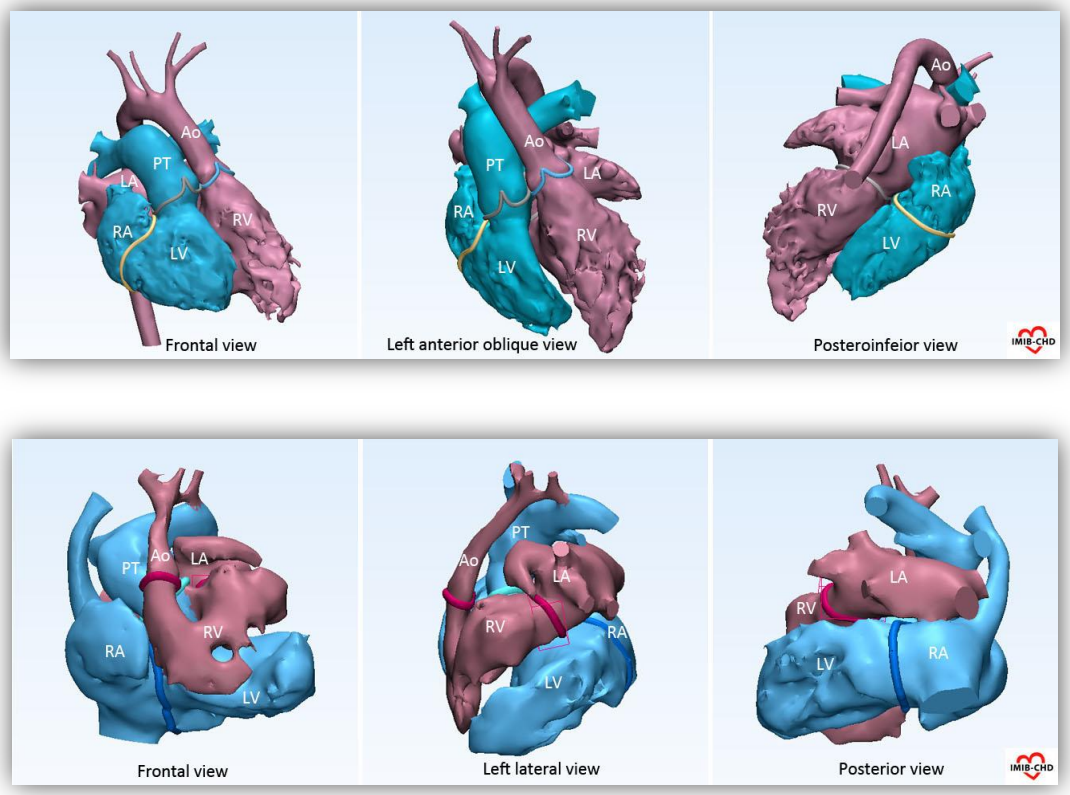

Figure 1-2. Volume rendered MR images of the cardiac cavities showing a classic form of congenitally corrected transposition (upper panels) and congenitally corrected transposition with twisted atrioventricular connection (lower panels). Note that the atrioventricular valves are arranged in parallel in the classic case, while they are not parallel in the atypical case with a twisted connection.

Rarely, the spatial relationships of the cardiac chambers and great arteries are not as expected for the given segmental connections (**Figures 1-1 and 1-2, lower panels**). As these hearts show unusual external and internal appearances, they often cause diagnostic dilemmas and difficulties in describing their complex anatomic features. These hearts have been described using various descriptive terms as listed in Table 1-1 [1-11]. It is known that the first description of the hearts with unusual spatial relationship of the cardiac chambers for the given pathology was given by Lev and Rowlatt in 1961[1]. Certainly, the concept and terminology regarding such rare pathologic entities have evolved with accumulation of case examples. Among many terms, 'criss-cross heart' [4, 5] and 'superoinferior ventricles' [8, 9] have been most commonly opted. Although both terms are excellent in highlighting a striking feature of the classic forms, there have been debates on whether one term is any better than the other and whether the terms could be used interchangeably.

Table 1-1. Evolution of terminology.

Year	Authors	Remarks
1961	Lev M and Rowlatt UF [1]	Mixed levocardia
1974	Kinsley RH, et al [2]	Ventricular rotation
1974	Anderson RH, et al [3, 4]	Criss-cross atrioventricular relationship or criss-cross heart
1976	Ando M, et al [5]	Crossing atrioventricular valves
1976	Guthaner DG, et al [6]	Uncorrected transposition with horizontal ventricular septum
1977	Momma K, et al [7]	Upstairs-downstairs alignment of the ventricles
1977	Van Praagh R, et al [8, 9]	Superoinferior ventricles
1991	Seo JW, et al [10, 11]	Twisted atrioventricular connection

The most commonly used terms 'criss-cross heart' and 'superoinferior ventricles' describe the hearts with the same or similar pathology from the different points of view and perspectives. The term 'criss-cross heart' describes the directions of the two blood streams from the atria to the ventricular apices or the axes of the atrioventricular valve openings, while the term 'superoinferior ventricles' obviously describes the spatial relationship between the two ventricles. Although Richard van Praagh claimed that criss-crossing appearance is an illusion [12], the majority of the hearts described as criss-cross heart did show real anatomical crossing of the connections between the atria and underlying ventricles as shown in the lower panels of **Figures 1-1** and **1-2**. In the majority of the hearts of this kind, both features coexist. The hearts described as 'criss-cross hearts' almost invariably show the inlet of one ventricle located superior to the inlet of the other ventricle. The right ventricular inlet is almost always superior to the left ventricular inlet. However, the positions of the apices of the ventricles vary, showing more superoinferior relationship of the apices in some and more side-by-side relationship in others [13]. The majority of the hearts described as 'superoinferior ventricles' also show crossed blood streams across the atrioventricular junction. However, there are hearts with their ventricles superoinferiorly related but without a significant degree of crossing of the atrioventricular blood streams [4]. As there are such exceptional cases showing only one apparent feature, the terms, 'criss-cross heart' and 'superoinferior ventricles' are not completely interchangeable. Therefore, both terms fail to accommodate the whole spectrum of similar malformations because of too distinctive and restrictive nature of the adjectives "criss-cross" and "superoinferior". In fact, minor forms of a kind needed to be described as "partial criss-cross heart" when the relationship of the inlets is not altered enough to be recognized as "criss-cross" [4, 13].

In 1992, Seo et al published the paper entitled "*Further morphological observations on hearts with twisted atrioventricular connections (criss-cross hearts*)" where they emphasized the varying degrees of twisting in the spectrum of pathology [11]. They found that the adjective "twisted' introduced by Peter Brandt in 1980 [10] best explains the common morphologic features and spectral nature of the pathology.

Freedom, et al described two cases with the superoinferiorly related ventricles with the great arteries exiting from the diaphragmatic aspect of the ventricular mass in their 1984 seminal textbook "*Angiography of Congenital Heart Disease*". Because of the bizarre posterior-inferior displacement of the arterial pole, the brachiocephalic arteries were markedly elongated. Freedom, et al called this peculiar heart a 'topsy-turvy heart" [14].

THEORETICAL PATHOGENETIC MECHANISMS

With growing experience, we find that there are three theoretical mechanisms that can produce unexpected ventricular relationships for the given segmental connections. They are *twisting, tilting* and *organoaxial rotation* [15].

Twisting: Uncommonly, the cardiac chambers and arterial trunks show unusual spatial relationships for the given atrioventricular and ventriculoarterial connections as if the heart were twisted clockwise or counterclockwise around its long axis by the right hand placed on the cardiac apex, with the left hand holding the posterior part of the heart in a fixed position (**Figure 1-3**) [11, 16]. Most of the hearts described as criss-cross heart or superoinferior ventricles show varying degrees of twisting of the heart around the long axis. Criss-cross heart is an extreme form of this category in which the atrioventricular connection axes appear to cross each other at a right angle. A minor degree of twisting is seen in cases where the ventricles are superoinferiorly related without apparent crossing of the atrioventricular connections. In the majority of the cases, twisting is in a direction to place the right ventricle above the left ventricle. Only a few cases have been reported in which the left ventricle is superior to the right ventricle [17-19]. The common characteristics of the twisted hearts are the loss of normal parallel orientation of the opening axes of the two atrioventricular valves, a curved or angled configuration of the atrial and ventricular septa [11, 16, 20, 21]. In addition, the great arterial trunks show unusual spatial relationship for the given atrioventricular and ventriculoarterial connection. The twisted hearts with complete transposition of the great arteries usually show the ascending aorta on the left anterior aspect of the pulmonary arterial trunk. On the contrary, the twisted hearts with congenitally corrected transposition typically show the ascending aorta on the right anterior aspect of the pulmonary arterial trunk. Therefore, the whole heart including the atrioventricular opening axes and the great arteries appears twisted.

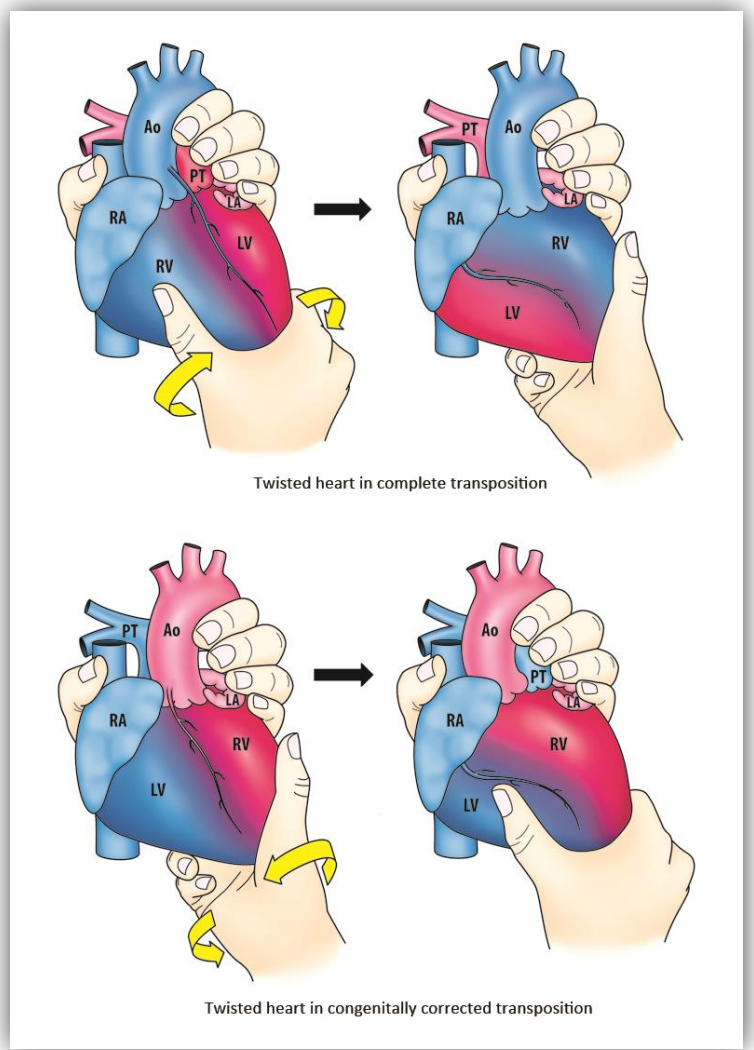

Figure 1-3. ***Twisting***. Cartoons showing a pathogenetic mechanism for so-called criss-cross hearts and most hearts with superoinferior ventricles in the presence of complete transposition (upper panel) and congenitally corrected transposition (lower panel). The heart appears twisted around the long axis of the heart by the observer's right hand, clockwise in the heart with complete transposition and counterclockwise in the heart with congenitally corrected transposition. Twisting occurs in a direction to displace the right ventricular inlet superiorly above the left ventricular inlet with the inlet part of the ventricular septum oriented horizontally. Note that the ascending aorta is displaced to the other side of the pulmonary trunk, leftward in complete transposition and rightward in congenitally corrected transposition.

Tilting: Rarely, the ventricles are related superoinferiorly with the two atrioventricular opening axes being kept parallel as if the ventricles are tilted by lifting or displacing the ventricular apex upward (**Figures 1-4 and 1-5**) [4.22]. The atria and atrial septum are oriented in a usual fashion, while the

ventricular septum is tilted to a relatively horizontal plane. Tilting occurs almost exclusively in hearts with a discordant atrioventricular connection. Tilting is more commonly in the direction that places the right ventricle superior to the left ventricle [22]. However, tilting may occur toward the right side in which situation, the left ventricle is superior to the right ventricle. The latter relationship is typically seen in congenitally corrected transposition occurring in situs solitus and dextrocardia.

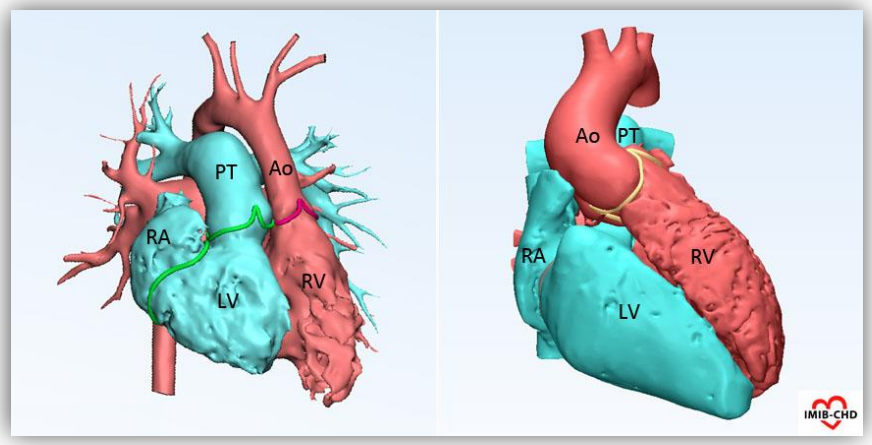

Figure 1-4. Volume rendered images in two cases of congenitally corrected transposition. Left-hand panel shows the ventricles related side-by-side. Right-hand panel shows the ventricles obliquely positioned with the right ventricle positioned to the left and superior relative to the left ventricle. In extreme examples, the right ventricle may be positioned superior to the left ventricle with the atrioventricular valve axes remaining aligned in parallel.

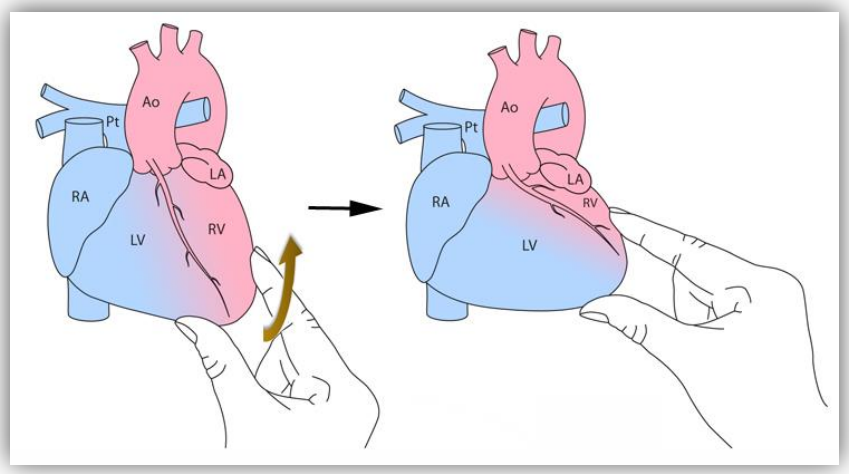

Figure 1-5. ***Tilting***. Cartoons showing a pathogenetic mechanism for superoinferior ventricular relationship in congenitally corrected transposition. The ventricular apex is lifted leftward or rightward without rotation. As a result, the ventricles are related superoinferiorly while the atrioventricular opening axes remain parallel.

Organoaxial rotation: Topsy-turvy hearts are characterized by the cardiac chambers showing superoinferior relationship and the great arterial trunks exiting the heart very low close to the diaphragm as if the entire heart were rotated around its long axis by the two hands of the observer [14]. As the entire heart is rotated, the parallel axes of the atrioventricular connections are maintained (**Figure 1-6**). The great arterial trunks are displaced in the direction of rotation so that they are located far down in the thorax near the diaphragm. A few years ago, we experienced a fetus with a peculiar pathology that was very similar to what Freedom et al described (**Figure 1-7**) [23, 24]. Although the fetus showed the four chambers of the heart oriented in a coronal plane and had a disproportionately large ductus arteriosus, there was no intracardiac pathology. The arterial valves were displaced inferiorly, resulting in unusually low position of the aortic and ductal arches and marked elongation of the head and neck branches of the aortic arch. Most importantly, the trachea and left main bronchus was elongated and compressed by the low lying aortic and ductal arches and the left pulmonary artery. Erek et al subsequently reported three cases showing almost identical morphological findings [25].

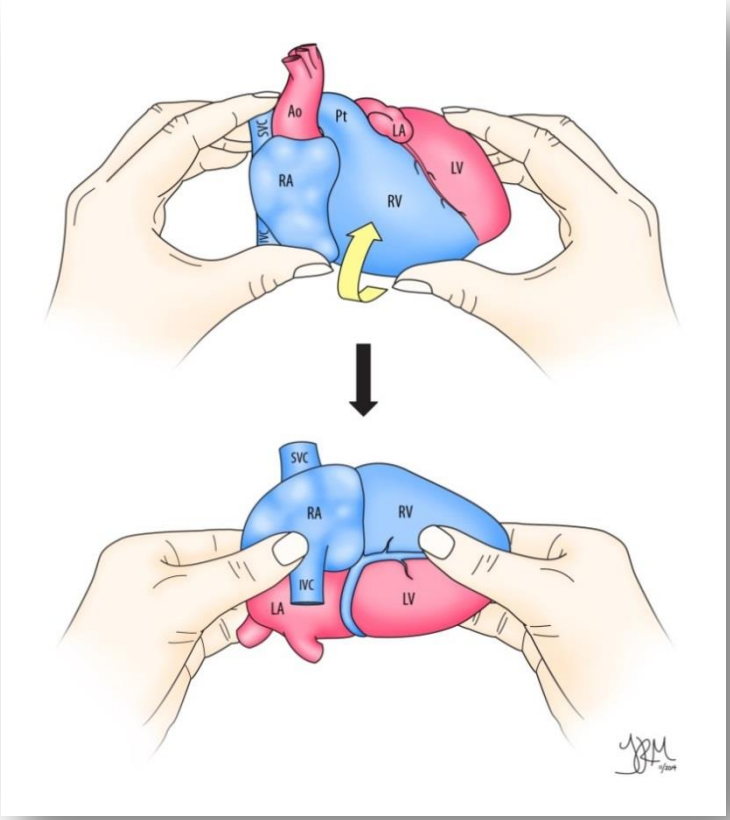

Figure 1-6. ***Organoaxial rotation***. Cartoons showing a pathogenetic mechanism for the so-called topsy-turvy heart in the presence of normal segmental connections. The heart is rotated backward around its long axis by the observer's two hands. The right atrium and right ventricle are displaced upward on top of the left atrium and left ventricle, respectively. As a result, the arterial roots arise from the lower aspects of the displaced right and left ventricles.

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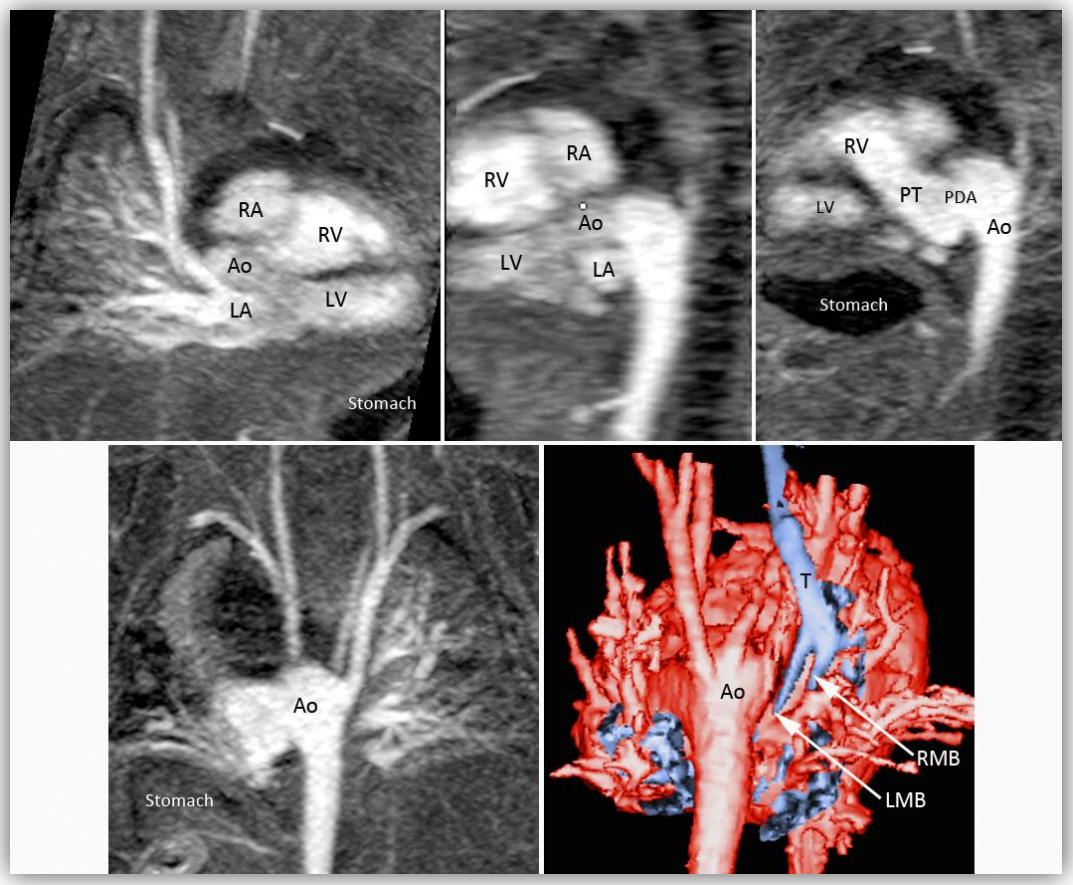


Figure 1-7. Postnatal MR angiograms of a newborn with a fetal diagnosis of topsy-turvy heart. Frontal view (left upper panel) shows the four cardiac chambers arranged in a coronal plane with the left ventricular outflow tract and the aortic root (Ao) in the center. Lateral views (upper middle and right panels) show that the aorta arises from the inferiorly located left ventricle and the pulmonary arterial trunk from the superiorly located right ventricle. There is a large patent ductus arteriosus (PDA). Posterior views of MR angiogram (left lower panel) and CT angiogram (right lower panel) show very low position of the aortic arch and superoinferior elongation of the head and neck branches. The left main bronchus (LMB) is elongated and compressed by the low-lying aortic arch. RMB, right main bronchus; T, trachea.

Most hearts typically exhibit the features that can be explained predominantly by one of these three mechanisms. However, a small number of cases may display mixed features that can be explained by combination of two or three of the above mechanisms.

On the other hand, the ventricles may also appear superoinferiorly located without any obvious signs of twisting, tilting or rotation. Cap-like double-horned right ventricle may mimic the appearance of

superoinferior ventricles [26-29]. In these rare cases, the right ventricular apex is severely hypoplastic or absent, and the right ventricle appears to consist of two horns; the inlet and outlet. Because of the absence of its apex, the right ventricle is largely located superior to the left ventricle and the atrioventricular connection may appear criss-crossing. This characteristic feature is elegantly illustrated in the 3D magnetic resonance angiograms in the case reported recently by Lopez et al as criss-cross heart in otherwise normal heart in situs inversus [29]. We believe this case an example of double-horned right ventricle.

VENTRICULAR CHIRALITY OR TOPOLOGY

When there is an abnormal ventricular relationship for the given atrioventricular connection as in twisted and topsy-turvy hearts, the classic right-left relationship between the ventricles is broken and may even appear inverted. To define the ventricular relationship in such abnormal situations, van Praagh, et al introduced a method called 'ventricular chirality' in which the observer places the palmar surface of his or her hand on the septum of the right ventricle, the wrist in the apex, the thumb in the right ventricular inlet and the fingers in the right ventricular outlet (**Figure 1-8**) [8, 9]. If the right ventricular aspect of the septum accepts the palmar surface of the observer's right hand, there is a normal ventricular chirality or D-loop ventricular relationship. When the right ventricular aspect of the septum accepts the palmar surface of the observer's left hand, there is an inverted ventricular chirality or L-loop ventricular relationship. Anderson, et al adopted this concept in their segmental approach but described D-loop and L-loop ventricular relationships as right-hand and left-hand patterns of 'ventricular topology', respectively [4].

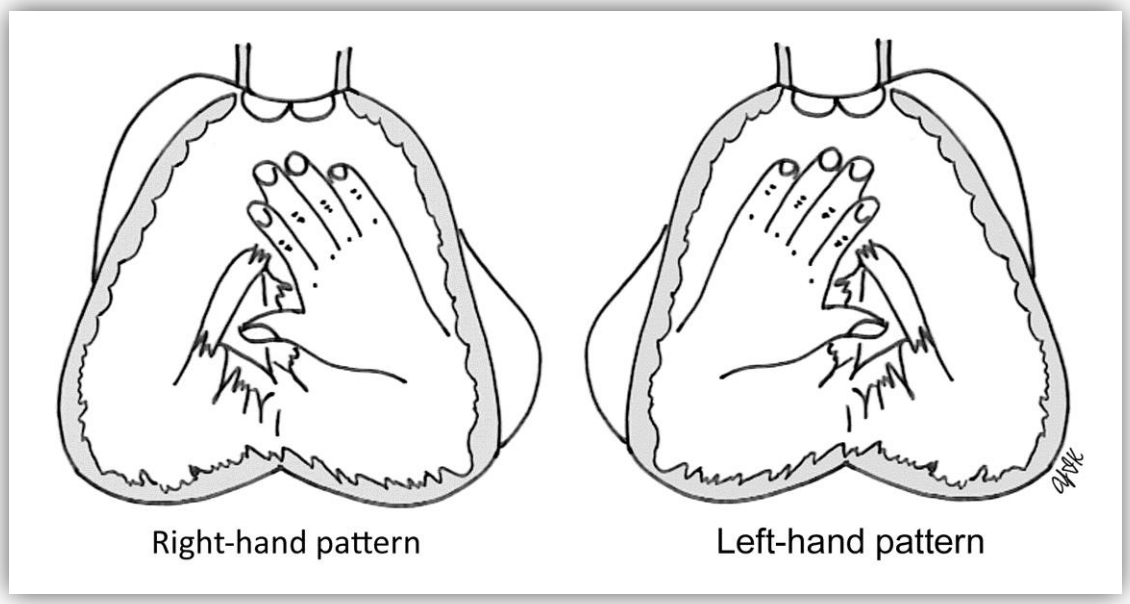


Figure 1-8. Cartoons showing the concept of ventricular chirality or topology. The palmar surface of the observer's hand is placed on the right ventricular septal surface with the wrist on the apex, the thumb in the inlet and the fingers in the outlet. In the presence of right-hand topology or D-loop ventricles, the right ventricular septum accepts the palm of the observer's right hand. In the presence of left-hand topology or L-loop, the right ventricular septum accepts the palm of the observer's left hand.

Regardless of the spatial relationship between the ventricles, the pattern of atrioventricular connection is in accordance with the ventricular topology or chirality in the majority of cases. When there is concordant atrioventricular connection in atrial situs solitus, the ventricular topology or chirality is almost always a right-hand pattern or d-loop. When there is discordant atrioventricular connection in atrial situs solitus, the ventricular topology or chirality is almost always a left-hand pattern. Extremely rarely, the ventricular topology or chirality is not in accordance with the given atrioventricular connection, for which Anderson et al described as 'disharmony' between the atrioventricular connection and segmental topology [30-34].

Although the concept of ventricular 'chirality' or 'topology' has been introduced to facilitate identification of the ventricular relationship or 'loop' and is applicable in the assessment of the pathological specimens, it is difficult to apply this concept during image interpretation especially when the ventricular relationship is unusual for the given pathology. It is indeed a pity that the concept is hardly applicable in those rare situations where the concept is intended to be applied for and needed. Furthermore, the arterial trunk or trunks often arise from the top of the free wall of the right ventricle, not allowing the observer's fingers to be placed in the outlet of the right ventricle (**Figure 1-9**). Despite the fact that the ventricular 'chirality' or 'topology' concept is needed only in the interpretation of the extremely rare cases, it has been advised to apply the concept to segmental approach to any form of congenital heart disease. Although it might appear logical, its application in the assessment and description of most usual cases of congenital heart disease is not only cumbersome and time consuming but also liable for mistakes. Furthermore, chirality or topology hardly has any clinical or surgical significance.

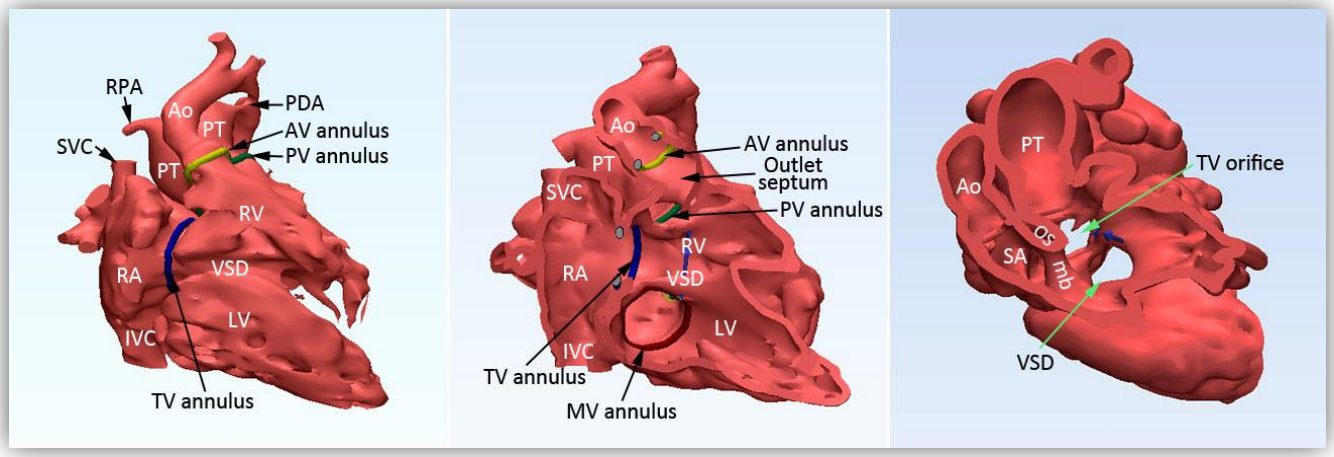


Figure 1-9. Volume rendered images of a twisted heart with double outlet right ventricle. Cast model (left-hand panel) and endocardial surface model of the interior of the heart in coronal cut (middle panel) and the interior of the right ventricle in a cut through the ventricular septum (right-hand panel) show the ventricular septum in a horizontal plane between the superior right ventricle and the inferior left ventricle. Both great arterial trunks arise from the top of the superior right ventricle. The observer's hand can be placed on the ventricular septum. However, the fingers cannot be placed through the arterial outlets.

GREAT ARTERIAL RELATIONSHIPAND SO-CALLED "LOOP RULE"

In the majority of the abnormal hearts as well as the normal hearts, the ascending aorta and its valve are on the side of the morphologically right ventricle. Van Praagh called this interesting tendency a "loop rule", highlighting that the "D-loop" ventricles (the right ventricle to the right side of the left ventricle) go with the "D-positioned aorta" (aorta on the right relative to the pulmonary arterial trunk), and the "L-loop ventricles" go with the "L-positioned aorta" (i.e., the segmental set being {S,D,S}. {S,D,D}, {S.L.L}, etc) (**Figure 1-1 and 2, upper panels**) [35, 36]. This strong tendency is typically broken when there is twisted atrioventricular connection [37-41] (**Figure 1-1 and 2, lower panels**). When there is twisted concordant atrioventricular connection, the aorta is typically displaced to the left and anterior or directly anterior to the pulmonary arterial trunk (i.e., {S,D,L} or {S,D,A}) (**Figure 1-1, lower panel**). It is in contrast to the usual complete transposition, double outlet right ventricle and normal heart in which the aorta is located to the right side of the pulmonary arterial trunk (i.e., {S,D,D} or {S,D,S}) (**Figure 1-1, upper panel**). When there is twisted discordant atrioventricular connection, the aorta is typically displaced to the right and anterior or directly anterior to the pulmonary arterial trunk (i.e., {S,L,D} or {S,L,A}) (**Figure 1-2, lower panel**). It is in contrast to classic congenitally corrected transposition in which the aorta is located to the left and anterior to the pulmonary arterial trunk (i.e., {S,L,L}) (**Figure 1-2, upper panel**). Other malformation that commonly breaks the "loop rule" is anatomically corrected malposition of the great arteries in which the aorta and pulmonary arterial trunk arise from the appropriate ventricle but the great arteries are malposed due to the presence of subaortic infundibulum or conus [42-44]. Although broken "loop rule" can be found any form of congenital heart disease, it may be a valuable clue to the diagnosis of twisted heart and anatomically corrected malposition of the great arteries.

In most of the hearts with the ventricles tilted, the usual great arterial relationship for the given pathology is largely maintained [4]. In topsy-turvy hearts, the right and left relationship of the great arterial trunks appears to be maintained, although the great arterial roots are displaced inferiorly [23-25].

ASSOCIATED ABNORMALTIES

Among three types of the hearts showing unusual or unexpected spatial relationships of the cardiac chambers for the given segmental combinations, twisted or tilted hearts are almost always associated with intracardiac defects, while topsy-turvy hearts are usually seen without major defects.

Twisted heart : Twisted hearts are most commonly seen in the setting of situs solitus and concordant atrioventricular connection and less commonly in the setting of situs solitus and discordant atrioventricular connection [37-60]. Twisted hearts are also seen in individuals with situs inversus and heterotaxy syndromes [54, 57, 58]. Majority of the twisted hearts show transposition or double outlet right ventricle, while concordant ventriculoarterial connection is rarely seen. Because of the curved or twisted configuration of the ventricular septum, the distinction between transposition and double outlet ventricle is difficult. Depending on the part of the ventricular septum the observer references, the arterial valve in question may appear to arise from one or the other ventricle. As twisting implies malalignment between the atrial and ventricular septal planes, a VSD is almost always present. Very rarely, twisted heart occurs with intact ventricular septum [3, 65-67]. The VSD typically involves the inlet part of the ventricular septum with variable extension toward the outlet and trabecular parts as observed from the right ventricle. It may also involve predominantly the

outlet part of the septum as shown in **Cases 6** and **8**. Although the VSD may appear a perimembranous type, its relationship with the remnant membranous septum is difficult to clearly define. Because of the malalignment of the atrial and ventricular septa, the one or both atrioventricular valves often show annular overriding with or without actual straddling of the tension apparatuses across the ventricular septum [40, 39, 59, 68]. The right ventricle is often hypoplastic with its inlet dimension significantly compromised especially when there is straddling or overriding atrioventricular valve or valves. The right ventricular outflow tract is often narrowed due to deviation of the outlet septum. In the presence of transposition, the right ventricular outflow tract obstruction is associated with tubular hypoplasia of the aortic arch and or coarctation of the aorta. Subpulmonary stenosis or pulmonary valvar stenosis or atresia is common. Right or left juxtaposition of the atrial appendages is not uncommonly seen especially in extremely complicated cases [32, 69, 70]. The atrioventricular connection may also appear twisted in double inlet right or left ventricles [4, 62, 63]. It has also reported that the heart appears twisted in the presence of tricuspid atresia [64]. The conduction tissue disposition including the location of the atrioventricular node and the course of the His bundle and its branches is considered abnormal in the majority of the cases of twisted heart [22, 46, 68]. However, it has not been described in a large number of cases because of the rarity and complexity of twisted hearts. The origins and distribution of the coronary arteries are also expected to be abnormal but the information is scarce [37, 71]. The origins and proximal courses of the coronary arteries should be carefully assessed when intracardiac repair is planned especially when an arterial switch operation or placement of a ventriculoarterial conduit is considered.

Topsy-turvy heart: In contrast to the twisted hearts, most reported cases of topsy-turvy heart are not associated with intracardiac defects. The cases we experienced and the recently reported cases have a great similarity showing a large aortopulmonary connection through a large and short patent ductus arteriosus or an unusual form of aortopulmonary window without intracardiac defects [23-25]. A coauthor of this article, Whal Lee and his colleague have recently experienced the most severe and peculiar form in which there was a large VSD, double outlet from the inferiorly located right ventricle, a small patent ductus arteriosus and right juxtaposition of the atrial appendages (**Case 10**). Most importantly, it should be reminded that the low lying position of the great arterial trunks in the thorax is associated with elongation and compression of the trachea and one or both bronchi.

SURGICAL MANAGEMENT

Although biventricular repair is preferred, the associated abnormalities such as significant right ventricular hypoplasia and straddling atrioventricular valve(s) often preclude this option [2, 48, 72-77]. Fang et al reviewed 150 patients reported as criss-cross heart or superoinferior ventricles between 1977 and 2008 [59]. Among 150 patients, interventional or surgical procedures were described in 37 patients. Biventricular repair with arterial switch, double switch or Rastelli procedure was performed in 34% of 37 patients, while 38% were staged towards Fontan-type operations. In the earlier era, biventricular repair might have been discouraged because of inadequate demonstration of the intracardiac anatomy at conventional angiograms or 2D echocardiograms. With modern 3D imaging technology with CT or MRI and 3D printing technology as introduced in this article, increasing number of cases would turn out to be eligible to biventricular repair.

REFERENCES

1. Lev M, Rowlatt UF. The pathologic anatomy of mixed levocardia. A review of thirteen cases of atrial or ventricular inversion with or without corrected transposition. Am J Cardiol 1961;8:216-263.
2. Kinsley RH, McGoon DC, Danielson GK. Corrected transposition of the great arteries. Associated ventricular rotation. Circulation 1974;49:574-578.
3. Anderson RH, Shinebourne EA, Gerlis LM. Criss-cross atrioventricular relationships producing paradoxical atrioventricular concordance or discordance. Circulation 1974;50:176-180.
4. Anderson RH. A question of definition. Criss-cross hearts revisted. Ped Cardiol 1982;3:305-313.
5. Ando M, Takao A, Nihmura I, Mori K: Crossing atrioventricular valves. A clinical study of 8 cases. Circulation 1976;53, 54 (suppl II):11-90.
6. Guthaner DG, Higgins CB, Silverman JF, Hayden WG, Wexler L. An unusual form of the transposition complex. Uncorrected levo-transposition with horizontal ventricular septum: report of two cases. Circulation 1976;53:190-195.
7. Momma K, Takao A, Mimori 5, et al. Corrected transposition of the great arteries with upstairs downstairs alignment of the ventricles. Heart 1976; 8:86-93. [Japanese]
8. Van Praagh R, Weinberg PM, Van Praagh S. Malposition of the heart. In: Moss AJ, Adams FH, Emmanouilides GC, editors. Heart disease in infants, children and adolescents. Baltimore: Williams & Wilkins; 1977. pp. 394–417.
9. Van Praagh S, LaCorte M, Fellows KE, Bossina K, Bush HJ, Keck EW, Weinberg PM, Van Praagh R. Supero-inferior ventricles: anatomic and angiographic findings in ten post-mortem cases. In: Van Praagh R, Takao A, editors. Etiology and morphogenesis of congenital heart disease. Mount Kisco (NY): Futura Publishing Co.; 1980. p. 317–378.
10. Brandt PWT. Cineangiocardiography atrioventricular and ventriculoarterial connections. In: Pediatric cardiology. Vol 4. Edinburgh, Scotland: Churchill Livingstone, 1980; 189-210.
11. Seo JW, Yoo SY, Ho SY, et al. Further morphological observations on hearts with twisted atrioventricular connections (criss-cross hearts). Cardiovasc Pathol 1992;1: 211-217.
12. Van Praagh R. Editorial comment. Progress in the understanding of congenital heart disease of congenital heart disease. Double-outlet right ventricle {S,D,L}, definition of ventriculocarterial discordance, definition of transposition of the great arteries, and the illusion of crisscross AV relations. Texas Heart Institute J 1988;15:183-186.
13. Thilenius OG[1], Bharati S, Lev M, Karp RB, Arcilla RA. Horizontal ventricular septum with dextroversion: hearts with and without aortic atresia. Pediatr Cardiol 1987;8:1871-93.
14. Freedom RM, Culham JG, Moes CF. Angiocardiography of Congenital Heart Disease. New York, NY: Macmillan Ed; 1984:629-642.
15. Anderson RH, Yoo SJ. Abnormal positions and relationship of the heart. In: Anderson RH, Baker EJ, Penny D, Redington AN, Rigby ML, eds. Pediatric Cardiology. 3rd ed. Philadelphia, Churchill Livingstone: Elsevier; 2010:991-1001.
16. Yoo SJ, Seo JW, Lim TH, Park IS, Hong CY, Song MG, Kim SH, Choe KO, Cho BK, Lee HJ. Hearts with twisted atrioventricular connections: findings at MR imaging. Radiology 1993;188:109-113.
17. Porras D, Kratz C, Loukas M, van Doesburg NH, Davignon A, Van Praagh R. Superoinferior ventricles with superior left ventricle and inferior right ventricle: a newly recognized form of congenital heart disease. Pediatr Cardiol 2003;24:604.
18. Yang G, Wang Q, He J, Wu M. Superior left ventricle in combination with inferior right ventricle presenting with balanced hemodynamics and mild symptoms in a later adolescent. Tex Heart Inst J 2010;37:445-448.
19. Angelini P. Left ventricle on top versus right ventricle on top in superoinferior ventricles: what are we talking about? Tex Heart Inst J 2010;37:442-444.

20. Robinson JP, Kumpeng V, MaCartney JF. Cross sectional echocardiographic and angiocardiographic correlation in crisscross hearts. Br Heart J 1985;54:61–67.
21. Yang YL, Wang XF, Cheng TO, Xie MX, Lü Q, He L, Lu XF, Wang J, Li L, Anderson RH. Echocardiographic characteristics of the criss-cross heart. Int J Cardiol 2010;140:133-137.
22. Wilkinson JL, Anderson RH. Anatomy of discordant atrioventricular connections. Wold J Pediatr Cong Heart Surg 2011;2:43-53.
23. Jaeggi E, Chitayat D, Golding F, Kim P, Yoo SJ. Prenatal diagnosis of topsy-turvy heart. Cardiol Young 2008;18:337-342.
24. Herrera P, Caldarone CA, Forte V, et al.; and Airway Reconstruction Team. Topsy-Turvy heart with associated congenital tracheobronchial stenosis and airway compression requiring surgical reconstruction. Ann Thorac Surg 2008;86:282-283.
25. Erek E, Guzeltas A, Ozturk NY, Kiyan G, Karakoc F, Akalin F, Odemis E, Arsan S. Topsy-turvy heart: a very rare congenital rotational heart disease with tracheobronchial anomalies. World J Pediatr Congenit Heart Surg 2013;4:308-311.
26. Ciavarella JM, McGoon DC, Hagler DJ, Fulton RE. Caplike double-horned double-outlet right ventricle: report of two cases. J Thorac Cardiovasc Surg 1979;77:536-542.
27. Beitzke A, Anderson RH, Wilkinson JL, Shinebourne EA. Two-chambered right ventricle simulating two-chambered left ventricle. Br Heart J 1979;42:22-26.
28. Muster AJ, Mavroudis C, Backer CL, Berdusis K, Alboliras ET, Ilbawi MN. Double-horned or caplike right ventricle: diagnosis and operative treatment. Ann Thorac Surg 1996;61:823-828.
29. Lopez AJ, Angelini P, Lufschanowski R. Successful ablation of atrioventricular node reentry tachycardia in a patient with crisscross heart and situs inversus levocardia. J Interv Card Electrophysiol 2006;17:133-137.
30. Anderson RH, Smith A, Wilkinson JL. Disharmony between atrioventricular connections and segmental combinations: unusual variants of "crisscross" hearts. J Am Coll Cardiol 1987;10:1274-1277.
31. Anderson RH, Ho SY. Editorial note: Segmental interconexions versus topological congruency in complex congenital malformations. Int J Cardiol 1989;25:229-233.
32. Coto EO, Wilkinson JL, Dickinson DF, Rufilanchas JJ, Márquez J. Gross distortion of atrioventricular and ventriculo-arterial relations associated with left juxtaposition of atrial appendages. Bizarre form of atrioventricular criss-cross. Br Heart J 1979;41:486-492.
33. Van Praagh R. When concordant or discordant atrioventricular alignments predict the ventricular situs wrongly. I. Solitus atria, concordant alignments, and L-loop ventricles. II. Solitus atria, discordant alignments, and D-loop ventricles. J Am Coll Cardiol 1987;10:1278-1279.
34. Geva T, Sanders SP, Ayres NA, O'Laughlin MP, Parness IA. Two-dimensional echocardiographic anatomy of atrioventricular alignment discordance with situs concordance. Am Heart J 1993;125:459-464.
35. Van Praagh R. The segmental approach to diagnosis in congenital heart disease. In Birth Defects, Vol VIII. Williams & Wilkins, Baltimore. 1972, pp 4-23.
36. Van Praagh R. Terminology of congenital heart disease. Glossary and commentary. Circulation 1977;56:139-143.
37. Houyel L, Van Praagh R, Lacour-Gayet F, Serraf A, Petit J, Bruniaux J, Planché C. Transposition of the great arteries [S,D,L]. Pathologic anatomy, diagnosis, and surgical management of a newly recognized complex. J Thorac Cardiovasc Surg 1995;110:613-624.
38. Marino B, Sanders S, Pasquini L, et al. Two-dimensional echocardiographic anatomy in criss-cross heart. Am J Cardiol 1986;58:325–333
39. Héry E, Jimenez M, Didier D, van Doesburg NH, Guérin R, Fouron JC, Davignon A. Echocardiographic and angiographic findings in superior-inferior cardiac ventricles. Am J Cardiol 1989;63:1385-1389.
40. Geva T, Van Praagh S, Sanders SP, Mayer JE Jr, Van Praagh R. Straddling mitral valve with hypoplastic right ventricle, crisscross atrioventricular relations, double outlet right ventricle and dextrocardia: morphologic, diagnostic and surgical considerations. J Am Coll Cardiol 1991;17:1603-1612.
41. Fraisse A, del Nido PJ, Gaudart J, Geva T. Echocardiographic characteristics and outcome of straddling mitral valve. J Am Coll Cardio 2001;38:819-826.

42. Freedom RM and Harrington DP. Anatomically corrected malposition of the great arteries: Report of 2 cases, one with congenital asplenia, frequent association with juxtaposition of atrial appendages. Br Heart J 1974; 36: 207-212.
43. Van Praagh R, Durnin RE, Jockin H, Wagner HR, Korns M, Garabedian H, Ando M, Calder AL. Anatomically corrected malposition of the great arteries (S, D, L). Circulation 1975;51:20-31.
44. Anderson RH, Becker AE, Losekoot TG, Gerlis LM. Anatomically corrected malposition of great arteries. Br Heart J 1975; 37: 993-1013.
45. Anderson KR, Lie JT, Sieg K, Hagler DJ, Ritter DG, Davis GD. A criss-cross heart. Detailed anatomic description and discussion of morphogenesis. Mayo Clin Proc 1977;52:569-575.
46. Symons JC, Shinebourne EA, Joseph MC, Lincoln C, Ho Y, Anderson RH. Criss-cross heart with congenitally corrected transposition: report of a case with d-transposed aorta and ventricular preexcitation. Europ J Cardiol 1977;5:493-505.
47. Freedom RM, Culham G, Rowe RD. The criss-cross and superoinferior ventricular heart: an angiographic study. Am J Cardiol 1978;42:620-628.
48. Sato K, Ohara S, Tsukaguchi I, Yasui K, Nakada T, Tmai M, Kobayashi Y, Kozuka T. A criss-cross heart with concordant atrioventriculo-arterial connections. Report of a case. Circulation 1978;57:396-400.
49. Attie F, Muñoz-Castellanos L, Oveseyevitz J, Flores-Delgado I, Testelli MR, Buendia A, Kuri J, Molina B. Crossed atrioventricular connections. Am Heart J 1980;99:163-172.
50. Tadavarthy SM, Formanek A, Castaneda-Zuniga W, Moller JH, Edwards JE, Amplatz K. The three types of criss-cross heart: a simple rotational anomaly. Br J Radiol 1981;54:736-743.
51. Van Mill G, Moulaert A, Harinck E, et al. Subcostal two-dimensional echocardiographic recognition of a criss-cross heart with discordant ventriculo-arterial connection. Pediatr Cardiol 1982;3:319–23.
52. Schneeweiss A, Shem-Tov A, Blieden LC, Deutsch V, Neufeld HN. Criss-cross heart--a case with horizontal septum, complete transposition, pulmonary atresia and ventricular septal defect. Pediatr Cardiol 1982;3:325-328.
53. Marino B, Chiariello L, Bosman C, Marsico F, Calabro R, Reale A, Marion B. Criss-cross heart with discordant atrioventricular connections. Pediatr Cardiol 1982;3:315-318.
54. Yamagishi M, Imai Y, Kurosawa H, Takanashi Y, Soejima K, Nagase Y, Shinoka T, Nakazawa M, Ando M, Takao A. Superoinferior ventricular heart with situs inversus, levo-loop and dextro-malposition (I,L,D), and double-outlet right ventricle: a case report. J Thorac Cardiovasc Surg 1986;91:633-637.
55. Carminati M, Valsecchi O, Borghi A, Balduzzi A, Bande A, Crupi G, Ferrazzi P, Invernizzi G. Cross-sectional echocardiographic study of criss-cross hearts and superoinferior ventricles. Am J Cardiol 1987;59:114-118.
56. Han HS[1], Seo JW, Choi JY. Echocardiographic evaluation of hearts with twisted atrioventricular connections (criss-cross heart). Heart Vessels 1994;9:322-326.
57. Ozkutlu S, Ayabakan C, Demircin M, Yilmaz M. A case of superoinferior ventricular heart with situs ambiguus, dextroventricular loop, and levo transposition of the arteries: prenatal and postnatal echocardiographic diagnosis. Pediatr Cardiol 2003;24:498-502.
58. Zhu M, Zhong Y. Magnetic resonance evaluation of criss-cross heart. Pediatr Cardiol 2008;29:359-365.
59. Fang F, Li ZA, Yang Y, Zheng CH, Lam YY. Deciphering the mysteries of crisscross heart by transthoracic echocardiography. Echocardiography 2011;28:104-108.
60. De Oliveira ÍM, Aiello VD, Mindêllo MM, Martins Yde O, Pinto VC Jr. Criss-cross heart: report of two cases, anatomic and surgical description and literature review. Rev Bras Cir Cardiovasc 2013;28:93-102.
61. Kim TH, Yoo SJ, Ho SY, et al. Twisted atrioventricular connections in double inlet right ventricle: evaluation by magnetic resonance imaging. Cardiol Young 2000;10:567–573.
62. Abdullah M, Yoo SJ, Hornberger L. Fetal echocardiographic features of twisted atrioventricular connections. Cardiol Young 2000;10:409–412.
63. Ngeh N[1], Api O, Iasci A, Ho SY, Carvalho JS. Criss-cross heart: report of three cases with double-inlet ventricles diagnosed in utero. Ultrasound Obstet Gynecol 2008;31:461-465.

64. Ayabakan C, Binnetoğlu K, Sarısoy Ö, Tokel K. Crisscross heart with tricuspid atresia diagnosed in utero. Congenit Heart Dis 2013;8:E153-156
65. Santos MA, Simões LC. Paroxysmal supraventricular tachycardia in supero-inferior ventricles with intact ventricular septum. Int J Cardiol 1987;14:232-235.
66. Fontes VF[1], de Souza JA, Pontes Jùnior SC. Criss-cross heart with intact ventricular septum. Int J Cardiol 1990;26:382-385.
67. Alday LE, Juaneda E. Superoinferior ventricles with criss-cross atrioventricular connections and intact ventricular septum. Pediatr Cardiol 1993;14:238-241.
68. Sieg K, Hagler DJ, Ritter DG, McGoon DC, Maloney JD, Seward JB, Davis GD. Straddling right atrioventricular valve in criss-cross atrioventricular relationship. Mayo Clin Proc 1977;52:361-368.
69. Wagner HR, Alday LE, Vlad P. Juxtaposition of the atrial appendages: a report of six necropsied cases. Circulation 1970;42:157-163.
70. Seo JW, Choe GY, Chi JG. An unusual ventricular loop associated with right juxtaposition of the atrial appendages. Int J Cardiol 1989;25:219-228
71. Angelini P, Lopez A, Lufschanowski R, Nemeth MA, Flamm SD. Coronary arteries in crisscross heart. Tex Heart Inst J 2003;30:208–213.
72. Waldhausen JA, Williams SP, Whitman V. Horizontal interventricular septum in congenital heart disease: surgical considerations. Ann Thorac Surg 1977;23:271-275.
73. Danielson GK, Tabry IF, Ritter DG, Fulton RE. Surgical repair of criss-cross heart with straddling atrioventricular valve. Thorac Cardiovasc Surg 1979;77:847-851.
74. Nagatsu M[1], Harada Y, Takeuchi T, Goto H, Ota Y. Can concordant criss-cross heart be ameliorated by hemodynamic changes? Ann Thorac Surg 1995;60:699-701.
75. Kashiwagi J, Imai Y, Aoki M, Shin'oka T, Hagino I, Nakazawa M. An arterial switch operation for a concordant crisscross heart with the complete transposition of the great arteries. J Thorac Cardiovasc Surg 2002;124:176-178.
76. Nakada I, Nakamura T, Matsumoto H, Sezaki T. Successful repair of criss-cross heart using modified Fontan operation. Chest 1983;83:569-570.
77. Podzolkov VP, Ivanitsky AV, Makhachev OA, Alekian BG, Chiaureli MR, Ragimov FR. Fontan-type operation for correcting complex congenital defects in a criss-cross heart. Pediatr Cardiol 1990;11:105-110.

Chapter 2. Case series 1-12

Case 1. Twisted heart with complete transposition of the great arteries

Case 2. Twisted heart with atrioventricular concordant connection and double outlet right ventricle

Case 3. Mildly twisted atrioventricular connection to superoinferiorly related ventricles and double outlet right ventricle

Case 4. Twisted heart with congenitally corrected transposition of the great arteries

Case 5. Twisted heart with congenitally corrected transposition of the great arteries

Case 6. Twisted heart with atrioventricular discordant connection, double outlet right ventricle with a subpulmonary VSD, and partial anomalous pulmonary venous connection

Case 7. Twisted heart with aorta as a single arterial trunk from the right ventricle and pulmonary atresia in situs solitus and dextrocardia

Case 8. Superoinferiorly related ventricles with double outlet right ventricle in situs solitus and dextrocardia

Case 9. Twisted heart with the left ventricle superiorly located in situs solitus, mesocardia and tetralogy of Fallot

Case 10. So-called topsy-turvy heart with double outlet right ventricle

Case 11. Congenitally corrected transposition of the great arteries with superoinferior ventricles and parallel atrioventricular valves

Case 12. Classic double inlet left ventricle and transposition of the great arteries

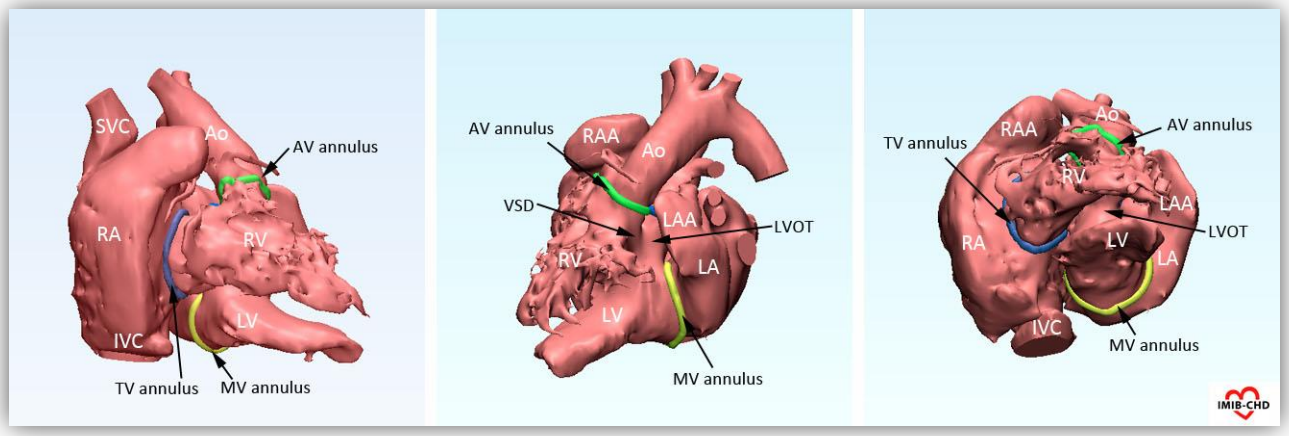
CASE 1. Twisted heart with complete transposition of the great arteries

- Source images: ECG-gated contrast-enhanced CT angiograms.

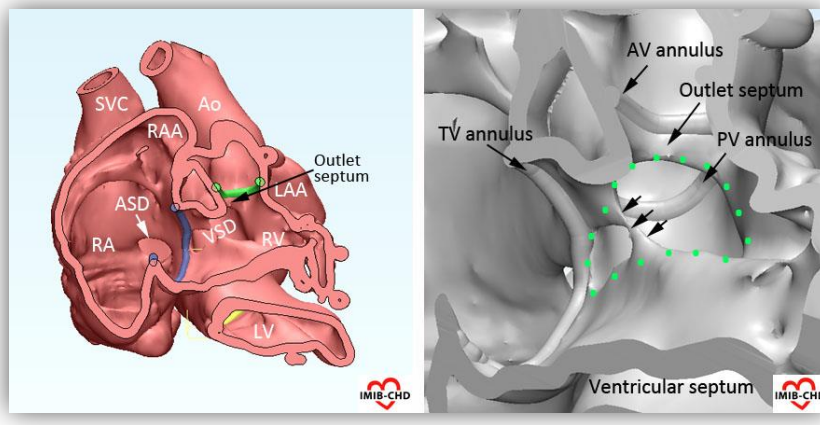
Summary:

- Situs solitus / Levocardia / Left aortic arch
- Concordant atrioventricular connection / Discordant ventriculoarterial connection
- Superoinferior relationship of the ventricles with the right ventricle on top of the left ventricle
- Aorta located anteriorly and slightly leftward relative to the pulmonary arterial trunk
- Bilateral subaortic and subpulmonary infundibulum. The subpulmonary infundibulum is short.
- Hypoplastic outlet septum, mildly malaligned anteriorly relative to the rest of the ventricular septum. Mild overriding of the pulmonary valve across the VSD
- Confluent inlet and outlet VSD at a short distance from the semilunar valves
- Tight pulmonary artery banding located close to the pulmonary arterial bifurcation

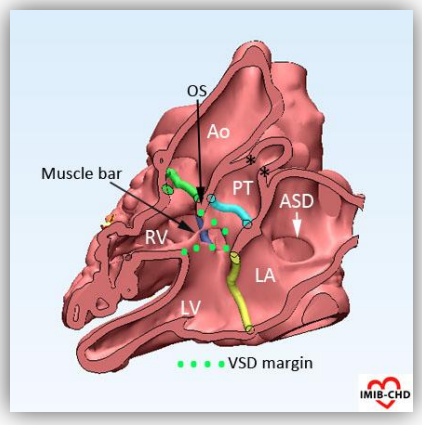
Models (4 pieces):



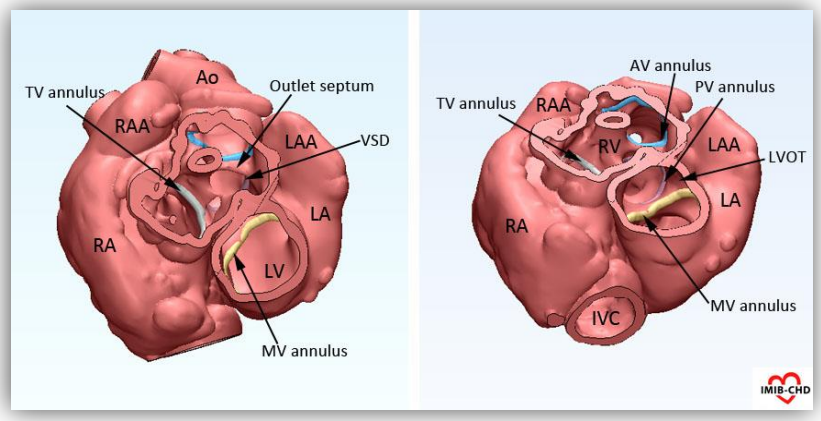
Model 1A. Volume rendered images of the cardiac chambers viewed from anterior (left panel), left posterior (middle panel) and postero-inferior (right panel) aspects.



Model 1B. Volume rendered image of the interior of the ventricles and right atrium viewed from front (left panel), and the close up view of the right ventricular aspect of the VSD (marked with green dots). Arrows in right panel indicate a muscle bar traversing the VSD.



Model 1C. Volume rendered image of the ventricles and the left atrium viewed from behind. Black asterisks indicate pulmonary artery banding.



Model 1D. Volume rendered images of the base of the ventricles viewed from the apex at different angles. The apical two thirds of the ventricles were removed for visualization of the basal plane of the ventricles

Findings:

- There is atrial situs solitus. The systemic and pulmonary venous connections are normal.
- The right and left ventricles are supero-inferiorly related with the ventricular septum horizontally oriented (**1A-1C**).
- The right atrium connects to the superiorly-located right ventricle. The left atrium connects to the inferiorly-located left ventricle (**1A-1C**).
- The right atrium is elongated with the tricuspid valve superiorly located at a distance from the inferior vena cava (**1A, 1B**). The opening axes of the tricuspid and mitral valves are not parallel (**1A-1D**). The tricuspid valve is oriented in a sagittal plane to open directly leftward. The mitral valve is oriented obliquely to open leftward, forward and slightly downward.
- The aorta arises from the right ventricle (**1A-1D**). The aortic valve is supported by a mildly narrow muscular infundibulum. The pulmonary arterial trunk arises from the left ventricle (**1A, 1C, 1D**). The pulmonary valve is supported by a short but wide muscular infundibulum. The left ventricular outflow tract is posterior and inferior to the right ventricular outflow tract.
- The VSD primarily involves the inlet septum and extends toward the outlet part (**1B-1D**). As its posterior rim abuts the tricuspid valve annulus, it is considered a perimembranous type. It is crossed by a thin muscle bar (arrows **1B**, right panel).
- There is mild overriding of the pulmonary valve across the ventricular septal crest due to mild anterior malalignment of the hypoplastic outlet septum relative to the rest of the ventricular septum (**1C, 1D**).
- The right ventricular side of the ventricular septum accepts the palmar surface of the observer's right hand with the thumb in the inlet, the wrist on the apex and the fingers in the outlet (**1B**). Therefore, there is a right hand pattern or chirality of the ventricular topology.
- There is severe stenosis of the distal part of the pulmonary arterial trunk due to tight surgical pulmonary artery banding (**1C**).
- The great arteries take parallel courses with the aorta located anteriorly and slightly leftward.
- A single coronary artery arises from the right anterior facing sinus (Sinus 2) (**1A**).
- There is a secundum type atrial septal defect.

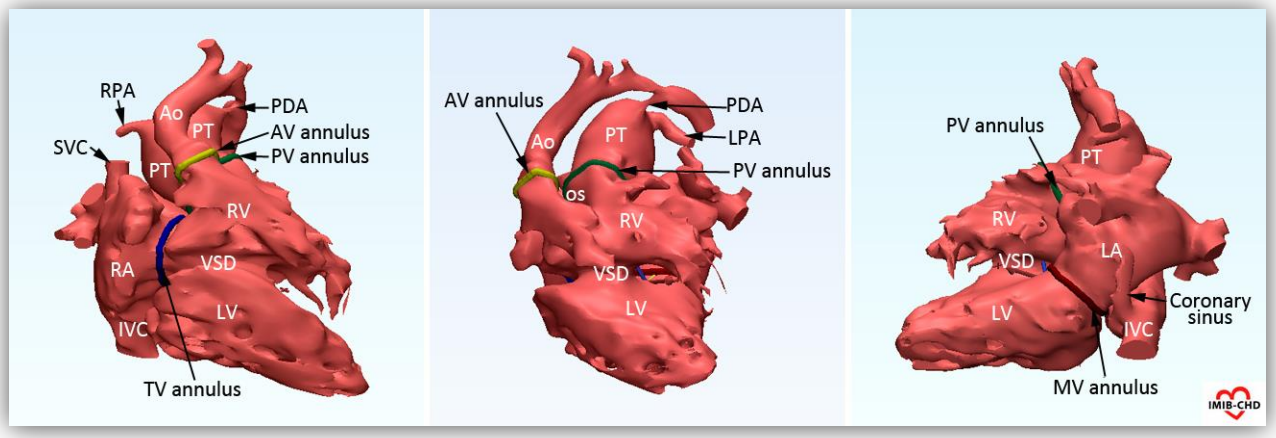
CASE 2. Twisted heart with concordant atrioventricular connection and double outlet right ventricle

- Source images: Non-ECG-gated contrast-enhanced MR angiograms.

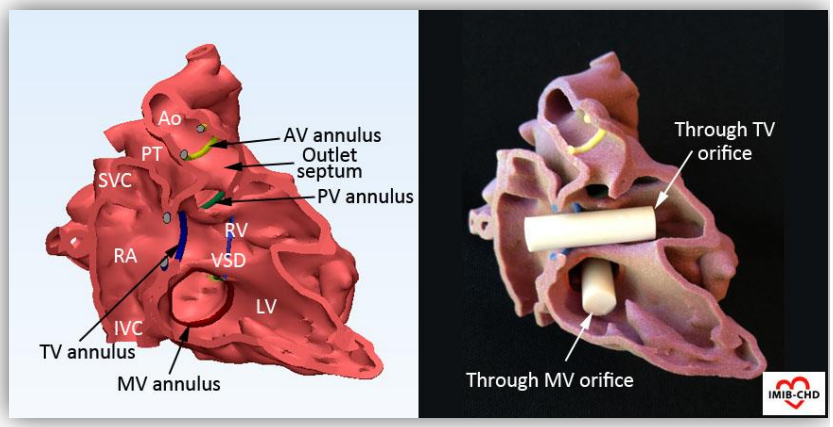
Summary:

- Situs solitus / Levocardia / Left aortic arch
- Concordant atrioventricular connection / Double outlet right ventricle
- Superoinferior relationship of the ventricles with the right ventricle above and behind the left ventricle.
- Aorta located directly anterior to the pulmonary arterial trunk
- Bilateral infundibulum with a long subaortic and a short subpulmonary infundibulum
- Subaortic narrowing due to deviated outlet septum
- A large inlet VSD remote from the semilunar valves. Mild overriding of the tricuspid annulus across the VSD
- Tubular hypoplasia of the aortic arch and coarctation of the aorta with small patent ductus arteriosus

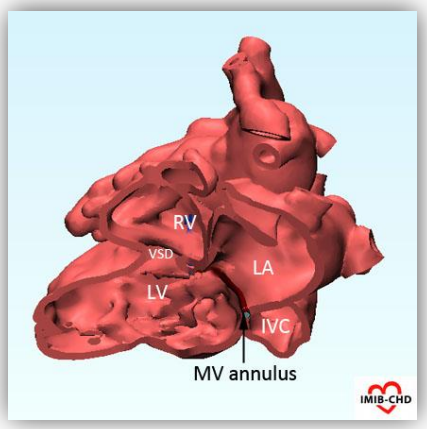
Models (5 pieces):



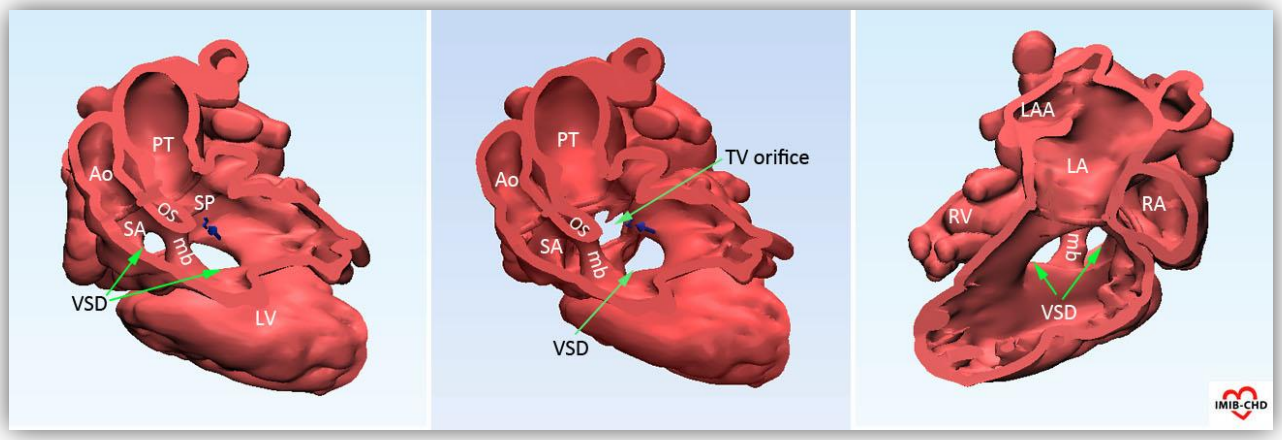
Model 2A. Volume rendered images of the cardiac chambers viewed from anterior (left panel), left anterior and superior (middle panel), and posterior (right panel) aspects.



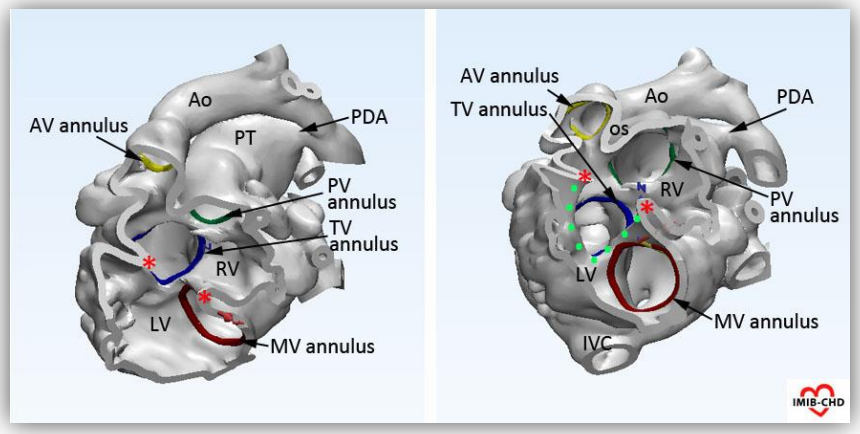
Model 2B. Volume rendered image and photograph of the model showing the interior of the ventricles and right atrium viewed from the front. White bars are passed through the tricuspid and mitral valve openings of the model.



Model 2C. Volume rendered image of the ventricles and left atrium viewed from behind.



Model 2D. Volume rendered images of the right ventricular aspect (left and middle panels) and the left ventricular aspect (right panel) of the ventricular septum. The free walls of the right and left ventricles are removed. os, outlet septum; mb, muscle bundle; SA, subaortic outflow tract; SP, subpulmonary outflow tract.



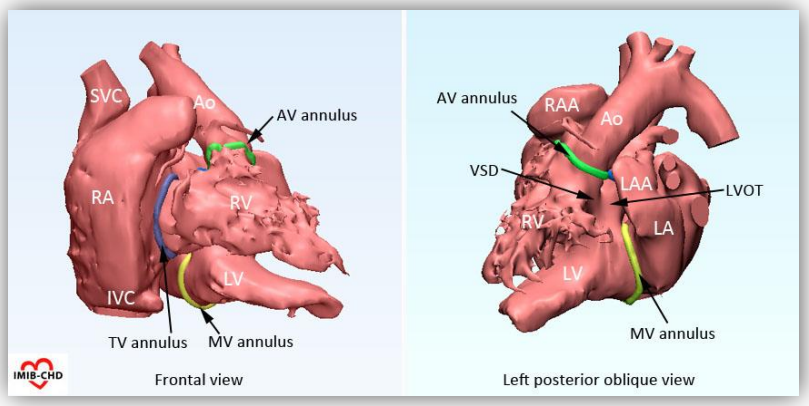
Model 2E. Volume rendered images of the interior of the base of the ventricles as viewed from the apex at different angles. Red asterisks indicate the cut-edge of the VSD. Green dots on right panel indicate VSD margin viewed from the left ventricle. os, outlet septum.

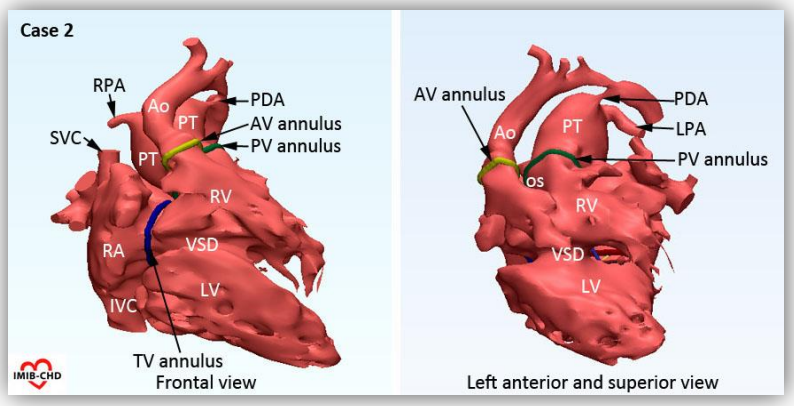
Findings:

- There is atrial situs solitus. The systemic and pulmonary venous connections are normal.
- The right and left ventricles are supero-inferiorly related with the major part of the right ventricle above and behind the left ventricle (**2A-2C, 2E**).
- The right atrium connects to the superiorly-located right ventricle. The left atrium connects to the inferiorly-located left ventricle. The horizontally oriented ventricular septum is slanted backward because of a high degree of twisting (**2A, 2B, 2E**). A large part of the left ventricle is spatially related to the lower part of the right atrium.
- The right atrium is elongated with the tricuspid valve superiorly located at a distance from the inferior vena cava (**2A, 2B**). The opening axes of the tricuspid and mitral valves are not parallel. The tricuspid valve is oriented in a sagittal plane to open directly leftward (**2B, 2E**). The mitral valve is oriented obliquely to open leftward, forward and downward (**2B, 2C, 2E**).
- Both the aorta and pulmonary arterial trunk arise from the right ventricle through muscular infundibulum (**2A-2C**). The outlet septum is an exclusively a right ventricular structure separating the subaortic and subpulmonary outflow tracts. The outlet septum encroaches on the subaortic outflow tract (**2B, 2D, 2E**). There is a muscle bundle arising from the lower margin of the outlet septum and attaching to the anterior free wall of the right ventricle (**2B, 2D, 2E**).

- The VSD is seen along the lower part of the tricuspid valve annulus and the medial superior part of the mitral valve annulus, suggesting that the defect is a perimembranous type involving predominantly the inlet part (**2B-2E**). It is remote from the semilunar valves.
- The inferior part of the annulus of the tricuspid valve overrides the ventricular septum to be related to the left ventricular inlet (**2B, 2E**).
- The right ventricular side of the ventricular septum accepts the palmar surface of the observer's right hand with the thumb in the inlet and the wrist on the apex (**2B**). It is considered a right-hand pattern of ventricular topology. However, the fingers of the right hand cannot be placed in the outlet because of its unusual superior position in the right ventricle directly opposite to the horizontal ventricular septum.
- The subaortic outflow tract is narrow and the aortic valve annulus is small. The aortic arch shows severe tubular hypoplasia. A narrow patent ductus arteriosus connects the pulmonary arterial trunk to the descending aorta. There is a shallow posterior shelf at the junction between the aortic isthmus and descending aorta (**2A**).
- The great arteries take a parallel course with the aorta directly anterior to the pulmonary arterial trunk (**2A, 2D**).
- The pulmonary arterial trunk is aneurysmally dilated and the branch pulmonary arteries are hypoplastic.

Comparison with Case 1:





There is a higher degree of clockwise twisting of the ventricles in Case 2 than in Case 1. The tricuspid annulus is higher in Case 2 than in Case 1 (left panels). In Case 1, the ventricles are largely supero-inferiorly related and the ventricular septum is in a horizontal plane. In Case 2, the major part of the right ventricle is positioned posteriorly to the major part of the left ventricle with the ventricular septum slanted backward. In Case 1, the pulmonary arterial trunk arises mostly from the left ventricle (right panel). In Case 2, the pulmonary arterial trunk is obviously arising from the right ventricle (right panel).

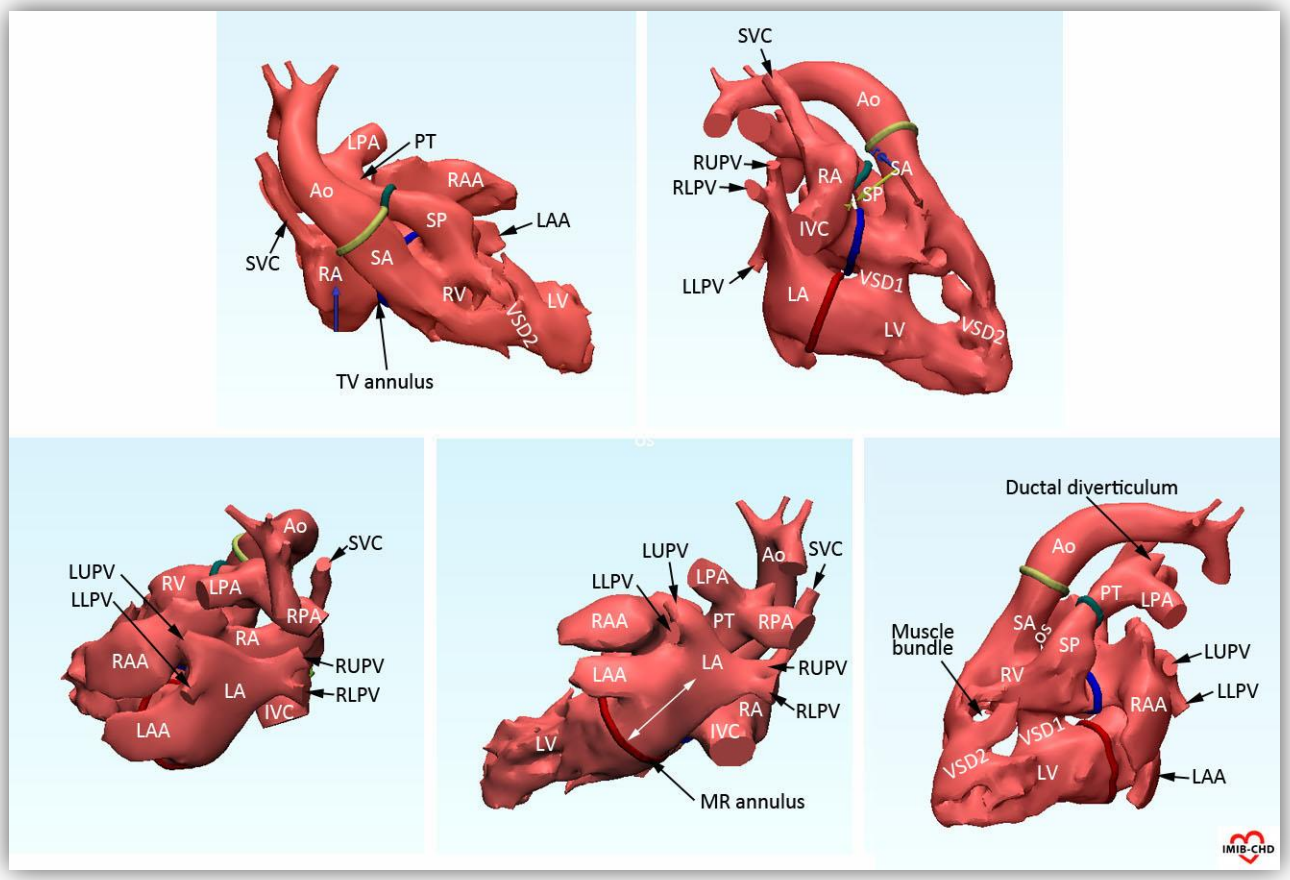
CASE 3. Mildly twisted atrioventricular connection to superoinferiorly related ventricles with double outlet right ventricle.

- Source images: Non-ECG-gated contrast-enhanced MR angiograms.

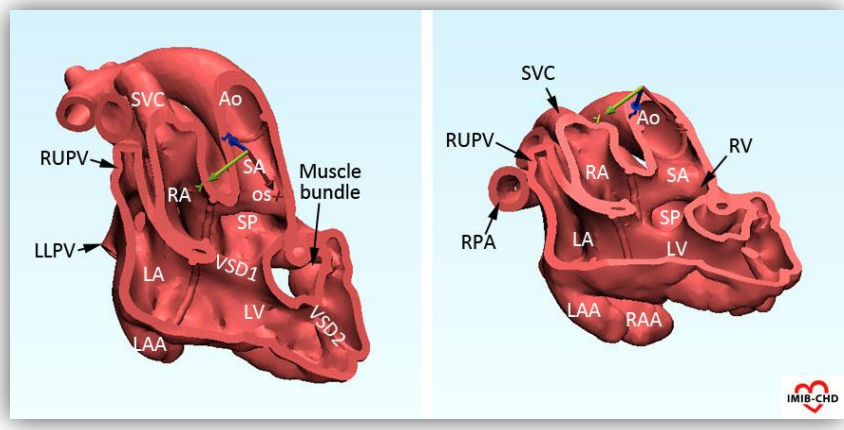
Summary:

- Situs solitus / Levocardia / Right aortic arch with mirror-image branching
- Concordant atrioventricular connection / Double outlet right ventricle
- Right ventricle positioned superior and anterior to the left ventricle
- Double outlet right ventricle with subaortic and subpulmonary infundibulum
- Aorta on the right and anterior to pulmonary arterial trunk
- VSD involving mostly the inlet part of the right ventricular aspect of the septum. An additional apical muscular VSD
- Left juxtaposition of the atrial appendages

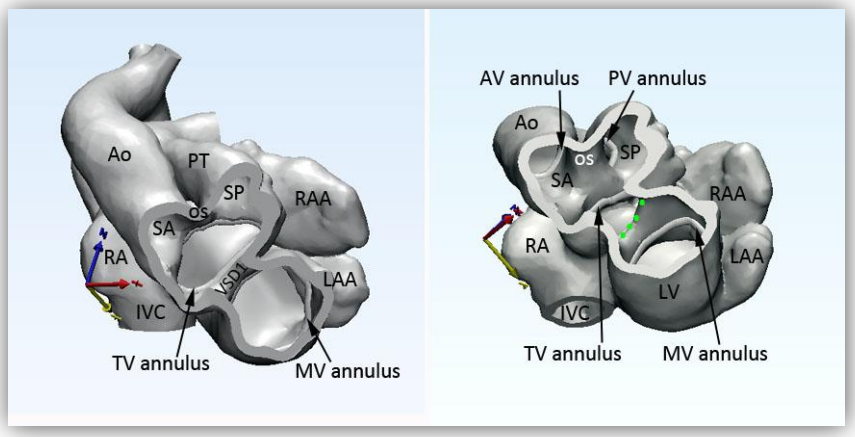
Models (3 pieces):



Model 3A. Volume rendered images of the cardiac chambers viewed from anterior (upper left panel), right anterior and inferior (upper right panel), right posterior and superior (lower left panel), posterior (lower middle panel), and left posterior and inferior (lower right panel) aspects. Cardiac valve annuli are marked with color. Double headed white arrow indicates the long vestibular part of the left atrium between the pulmonary venous confluence and the mitral valve. os, outlet septum; SA, subaortic outflow tract; SP, subpulmonary outflow tract.



Model 3B. Volume rendered images of the interior of the ventricles and atria viewed from front at different angles. os, outlet septum; SA, subaortic outflow tract; SP, subpulmonary outflow tract.



Model 3C. Volume rendered images of the interior of the base of the ventricles viewed from the apex at different angles. os, outlet septum; SA, subaortic outflow tract; SP, subpulmonary outflow tract.

Findings:

- There is atrial situs solitus. The systemic and pulmonary venous connections are normal.
- The right atrium has an elongated tubular configuration above the left atrium. The right atrial appendage is displaced to the left and the two appendages are juxtaposed superoinferiorly (**3A**, lower left panel). The left atrium is elongated supero-inferiorly with a large vesitibular part (double headed white arrow in **3A**, lower middle panel) between the pulmonary venous confluence and the mitral valve.
- The right ventricle is hypoplastic. It is located anterior and superior to the left ventricle (**3A**).
- The right atrium connects to the right ventricle. The left atrium connects to the left ventricle (**3A-3C**).
- There is mild clockwise rotation of the cardiac chambers around the long axis of the heart as seen from the apex. The right atrium and right ventricle are located superior and anterior to the left atrium and left ventricle (**3A, 3B**). There is only minimal twisting of the atrioventricular connection with the opening axes of the tricuspid and mitral valves aligned almost in parallel (**3B, 3C**)..
- Both aorta and pulmonary arterial trunk arise from the right ventricle through muscular infundibulum (**3A-3C**). The outlet septum is exclusively a right ventricular structure separating the subaortic and subpulmonary outflow tracts.
- There are two VSDs. One involves the basal part and is related to the inlets of the right and left ventricles. The other involves the apical part. Both VSDs are remote from the semilunar valves (**3A-3C**).
- The right ventricular side of the ventricular septum accepts the palmar surface of the observer's right hand with the thumb in the inlet and the wrist on the apex. It is considered a right-hand pattern of ventricular topology. However, the fingers of the right hand cannot be placed in the outlet because of its unusual position occupying the top of the right ventricle.
- The great arteries take a parallel course with the aorta located anterior and slightly rightward relative to the pulmonary arterial trunk (**3A, 3C**).

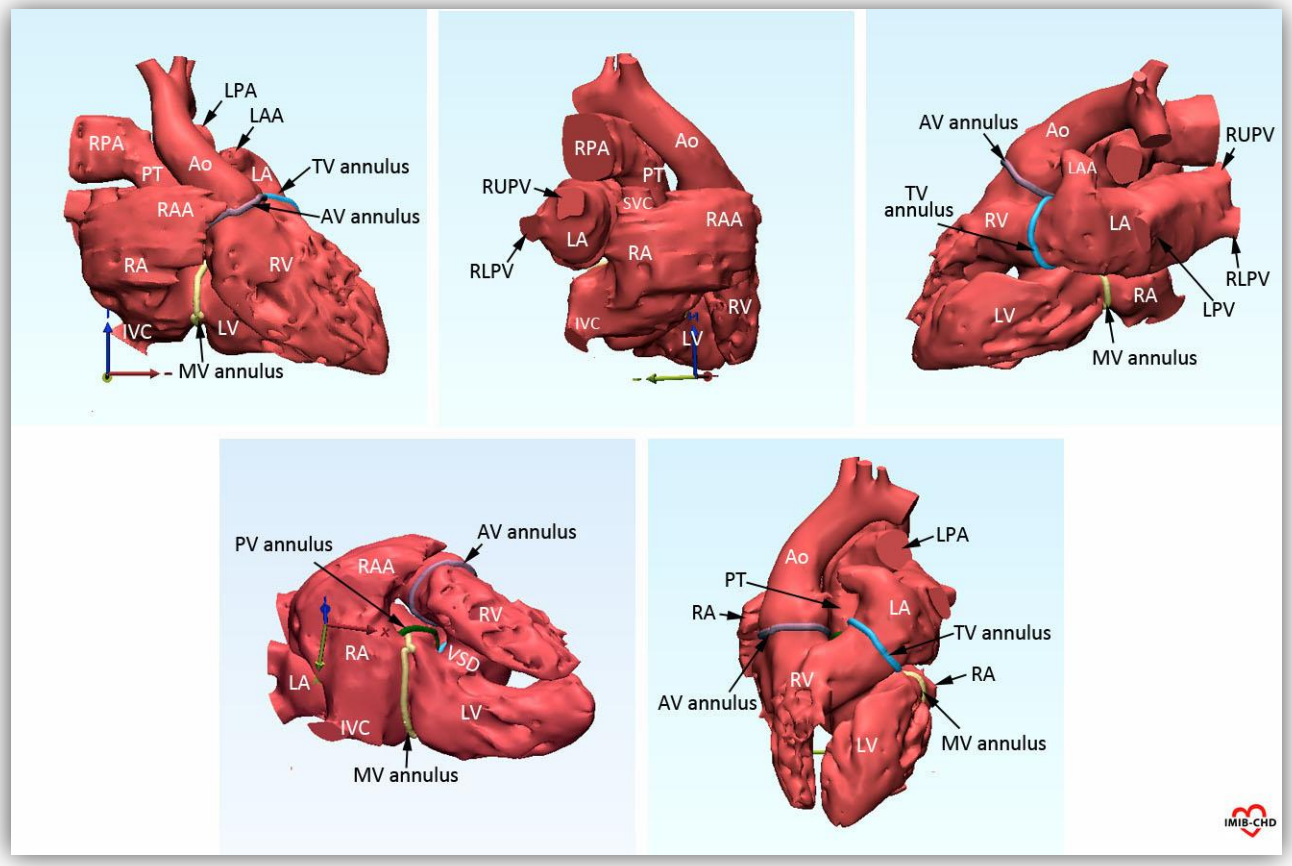
CASE 4. Twisted heart with congenitally corrected transposition of the great arteries

- Source images: ECG-gated contrast-enhanced CT angiograms.

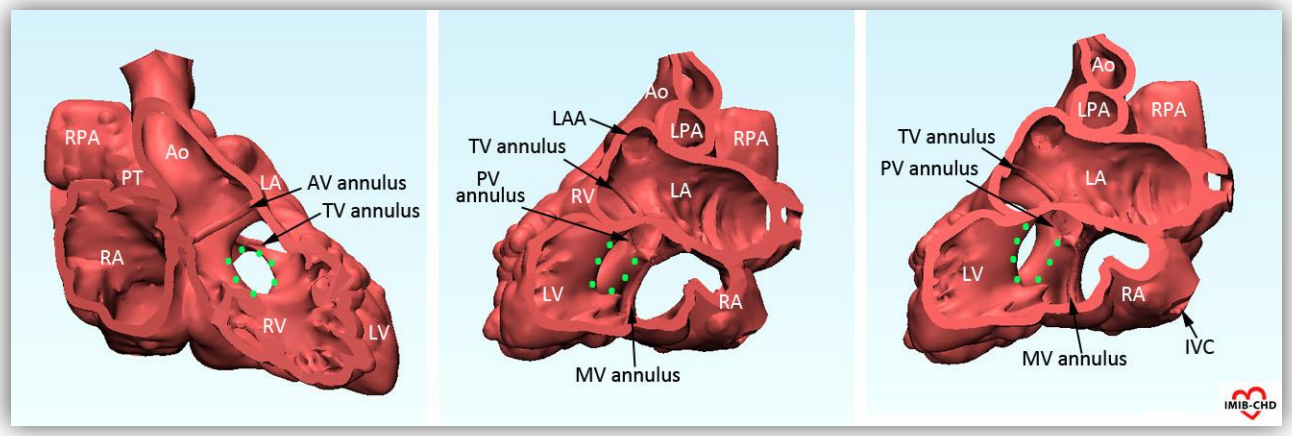
Summary:

- Situs solitus / Levocardia / Left aortic arch
- Discordant atrioventricular connection / Discordant ventriculoarterial connection
- Inverted anteroposterior relationship of the ventricles for the given atrioventricular connection with the right ventricle anterior and the left ventricle posterior
- Aorta located anteriorly and slightly leftward relative to the pulmonary arterial trunk
- Aorta arising from the right ventricle through a muscular infundibulum and pulmonary arterial trunk arising from the left ventricle without muscular infundibulum
- Confluent inlet and outlet VSD with its upper margin close to the semilunar valves
- Dilated right pulmonary artery

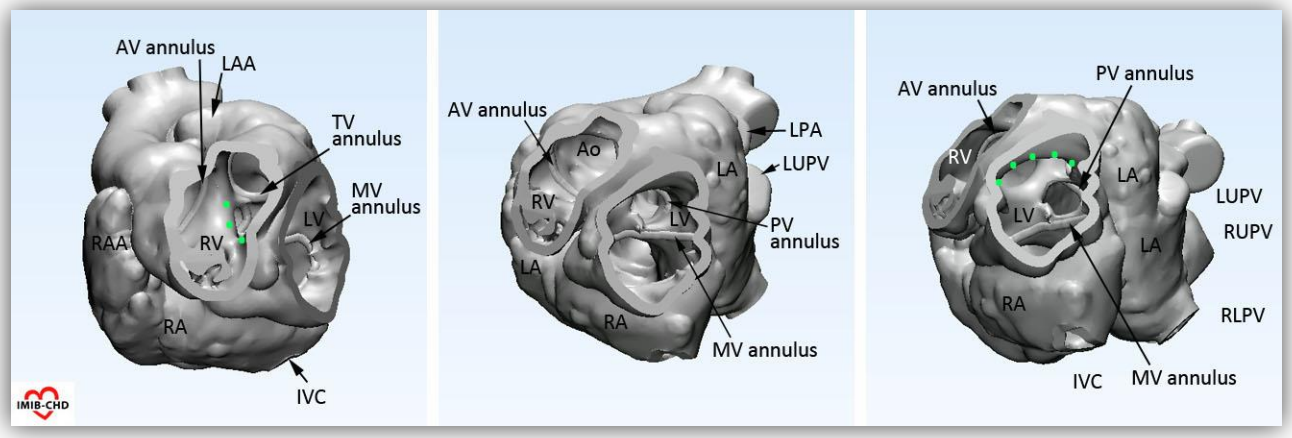
Models (3 pieces):



Model 4A. Volume rendered images of the cardiac chambers viewed from anterior (upper left panel), right lateral (upper middle panel), left posterior (upper right panel), inferior (lower left panel) and left lateral superior (lower right panel) aspects.



Model 4B. Volume rendered images of the ventricular septum viewed from the right ventricle (left panel) and from the left ventricle (middle and left panels). The VSD margin is demarcated by green dots.



Model 4C. Volume rendered images of the base of the ventricles viewed from the apex at different angles. The VSD margin is demarcated by green dots.

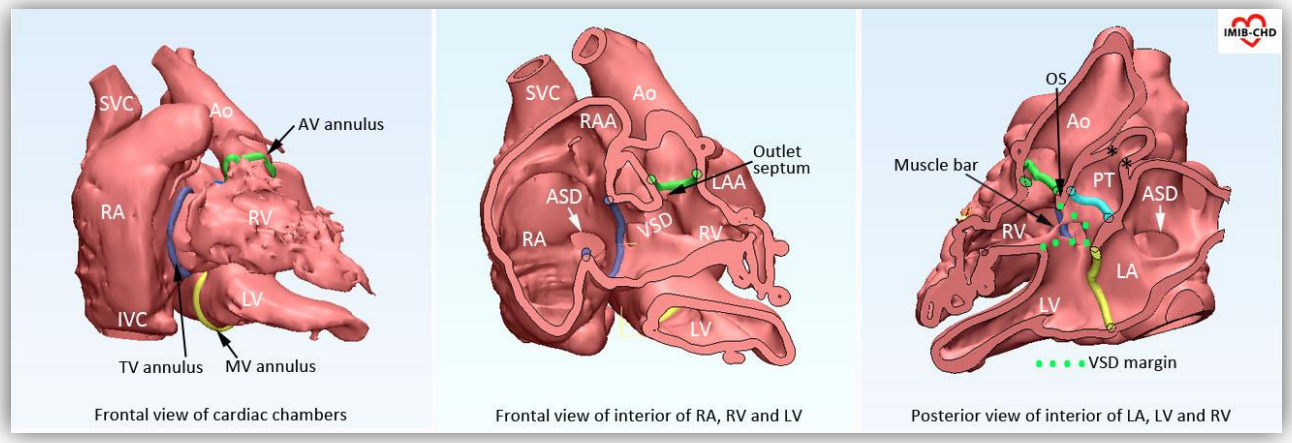
Findings:

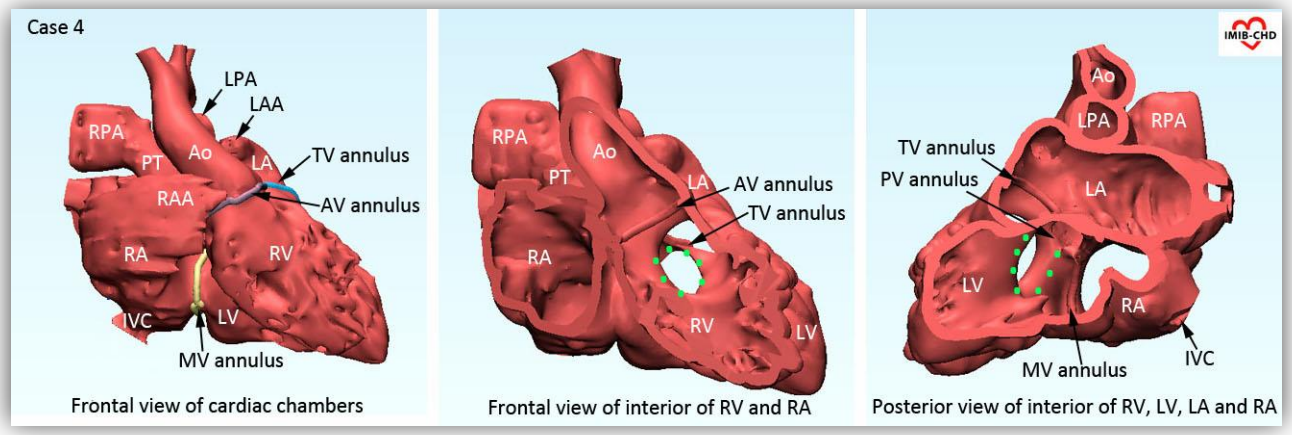
- There is atrial situs solitus. The systemic and pulmonary venous connections are normal.
- The right and left atria are normally related but are shaped unusually. The lower part of the right atrium is elongated anteroposteriorly to harbor its postero-inferiorly located atrioventricular valve (**4A-4C**). The left atrium is elongated from right to left with a long vestibular part between the pulmonary venous confluence and the tricuspid valve (**4A-4C**).
- The right and left ventricles are anteroposteriorly related with vertically oriented ventricular spetum (**4A**).
- The right atrium connects to the posteriorly-located left ventricle (**4A-4C**). The left atrium connects to the anteriorly-located right ventricle (**4A-4C**).
- The opening axes of the tricuspid and mitral valves are not parallel (**4A-4C**). The mitral valve is oriented in a sagittal plane to open directly leftward. The tricuspid valve is oriented obliquely to open forward, downward and slightly leftward.
- The aorta arises from the right ventricle through a muscular infundibulum (**4A-4C**). The pulmonary arterial trunk arises from the left ventricle (**4B, 4C**). The mitral valve is in direct contact with the

pulmonary valve. As a consequence, the pulmonary valve is not supported by muscular infundibulum. The ventricular outflow tracts are parallel to each other. It is noteworthy that the left ventricular outflow tract is posterior and slightly rightward relative to the right ventricular inlet (**4A**).

- The VSD involves the basal part of the ventricular septum (**4B, 4C**). As its posterior rim abuts the tricuspid valve annulus, it is considered a perimembranous type. The upper margin to the VSD is close to both semilunar valves (**4A-4C**).
- As the VSD involves the curved part of the ventricular septum, the pulmonary valve may appear to mildly override the ventricular septum (**4B, 4C**).
- The right ventricular side of the ventricular septum accepts the palmar surface of the observer's left hand with the thumb in the inlet, the wrist on the apex and the fingers in the outlet (**4B**). Therefore, there is a left hand pattern or chirality of the ventricular topology.
- The great arteries take a parallel course with the aorta located anteriorly and slightly leftward.
- The pulmonary valve annulus is smaller than the aortic valve annulus.
- The right pulmonary artery is dilated.

Comparison with Case 1:





In both cases, the ventricles show similar spatial relationship. However, there is a concordant atrioventricular connection in Case 1 and discordant atrioventricular connection in Case 4. In Case 1, the right atrium has its atrioventricular valve (tricuspid valve) positioned high at a distance from the inferior vena caval orifice. In Case 4, the right atrium has its atrioventricular valve (mitral valve) positioned low in close proximity to the inferior vena caval orifice.

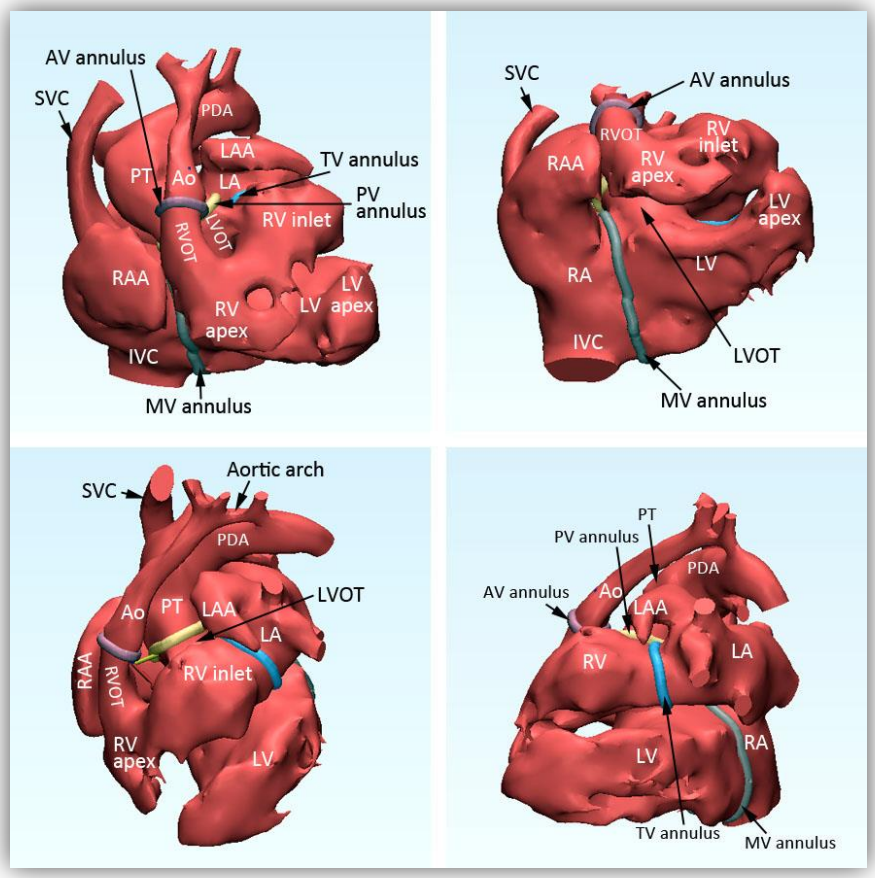
CASE 5. Twisted heart with congenitally corrected transposition of the great arteries

- Source images: Non-ECG-gated contrast-enhanced MR angiograms.

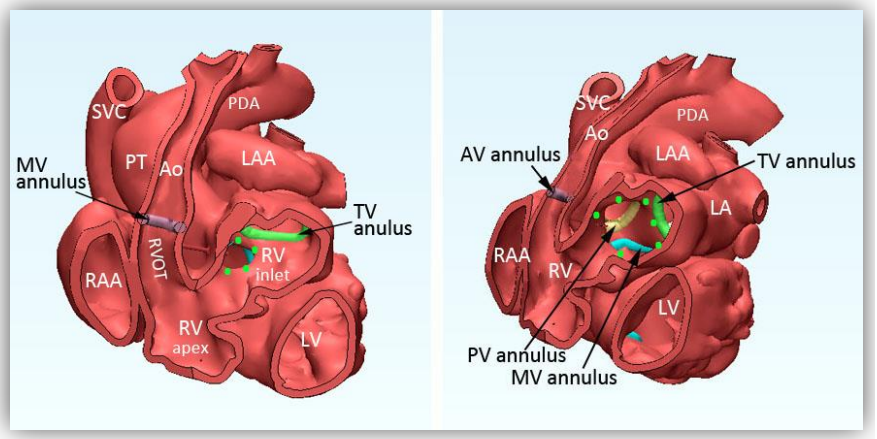
Summary:

- Situs solitus / Levocardia / Left aortic arch
- Discordant atrioventricular connection / Discordant ventriculoarterial connection
- Right ventricle, positioned superior and anterior to the Left ventricle
- Right and left ventricular apices pointing the opposite directions
- Non-parallel, twisted atrioventricular connection axes
- Aortic valve located directly anterior to the pulmonary valve
- Well-developed subaortic infundibulum. Very short subpulmonary infundibulum
- Long and narrow muscular subaortic outflow tract.
- Diffusely small aorta with severe tubular hypoplasia of the aortic arch and coarctation of the aorta. A large patent ductus arteriosus
- A large VSD between the inlet of the right ventricle and the outlet of the left ventricle.

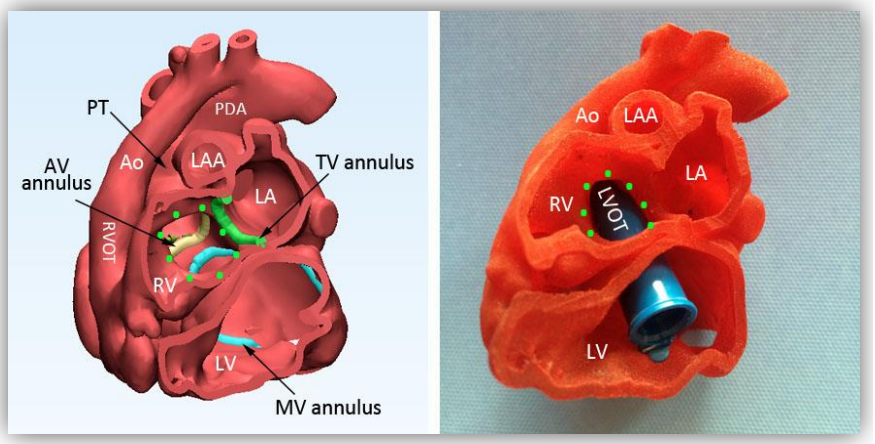
Models (5 pieces):



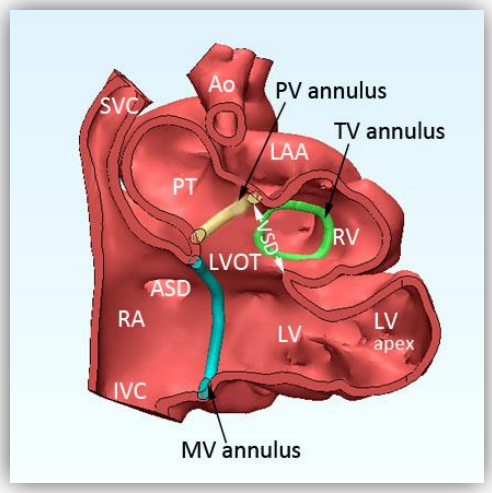
Model 5A. Volume rendered images of the cardiac chambers viewed from anterior (upper left panel), anterior and inferior (upper right panel), left anterior (lower left panel), and left posterior (lower right panel) aspects. Cardiac valve annuli are marked with color.



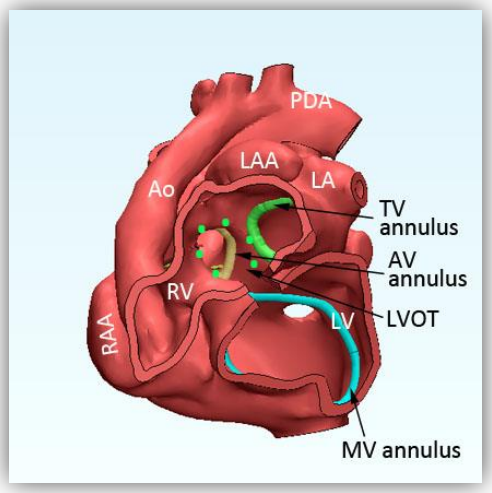
Model 5B. Volume rendered images showing the interior of the right ventricle viewed from the front at different angles. VSD is marked with green dots.



Model 5C. Volume rendered image (left panel) showing the interior of the right and left ventricles viewed from behind. Corresponding photograph of the model with a plastic bar passed through the left ventricular outflow tract (LVOT). The VSD margin is marked with green dots.



Model 5D. Volume rendered image of the interior of the left ventricle.

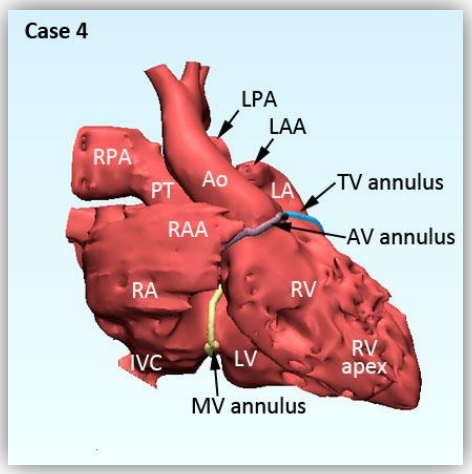


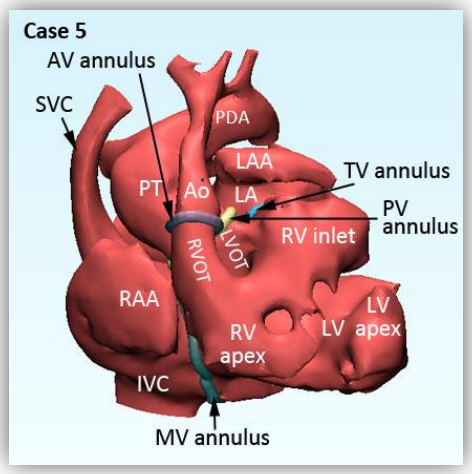
Model 5E. Volume rendered image of the base of the ventricles. The VSD margin is marked with green dots.

Findings:

- There is atrial situs solitus. The systemic and pulmonary venous connections are normal.
- There is a superoinferior relationship of the major parts of the atria with the left atrium superior to the right atrium (**5A**, lower right panel). There is a secundum type defect (**5D**). The atrial septum is oriented in a slanted coronal plane.
- The right ventricle is superior and anterior to the left ventricle (**5A-5D**).
- The right atrium connects to the inferiorly located left ventricle. In contrast to the high position of the tricuspid valve within the right atrium in Cases 1 and 2, the mitral valve is located inferiorly and posteriorly to connect the right atrium to the inferiorly located left ventricle (**5A, 5C**).
- The opening axes of the tricuspid and mitral valves are not parallel. The mitral valve is oriented in a sagittal plane to open directly leftward (**5C, 5E**). The tricuspid valve is oriented obliquely to open forward and slightly leftward (**5B-5E**).
- The right ventricular apex points to the right, anterior and inferior, while the left ventricular apex points leftward and anterior (**5A, 5B, 5D**).
- There is a large VSD between the right ventricular inlet and the left ventricular outlet (**5B-5E**).
- The right ventricle is moderately hypoplastic.
- The right ventricular side of the ventricular septum accepts the palmar surface of the observer's left hand with the thumb in the inlet, the wrist on the apex and the fingers in the outlet. Therefore, there is a so-called left-hand pattern or chirality of the ventricular topology (**5B, 5C**).
- The aorta arises from the right ventricle through a long infundibulum. The pulmonary arterial trunk arises from the superior aspect of the left ventricle. The left ventricular outlet consists of a short muscular infundibulum with close proximity of the pulmonary and mitral valve annuli. The right ventricular inlet wraps around the left ventricular outlet (**5B, 5C**).
- The subaortic outflow tract is narrow and the aortic valve is small.
- The aortic valve is located directly anterior to the pulmonary valve. The aorta courses posteriorly, while the pulmonary trunk courses from the left to the right.
- The aortic arch shows severe tubular hypoplasia. A large patent ductus arteriosus connects the pulmonary arterial trunk to the descending aorta distal to the left subclavian arterial origin. There is focal coarctation due to a posterior shelf above the insertion of the ductus arteriosus to the aorta.

Comparison with Case 4:





Both cases have twisted atrioventricular discordant connection, Case 5 showing a higher degree of twisting than Case 4. Note that the right ventricular apex points leftward in Case 4, while it points rightward in Case 5. In Case 5, the right and left ventricular apices point in opposite directions.

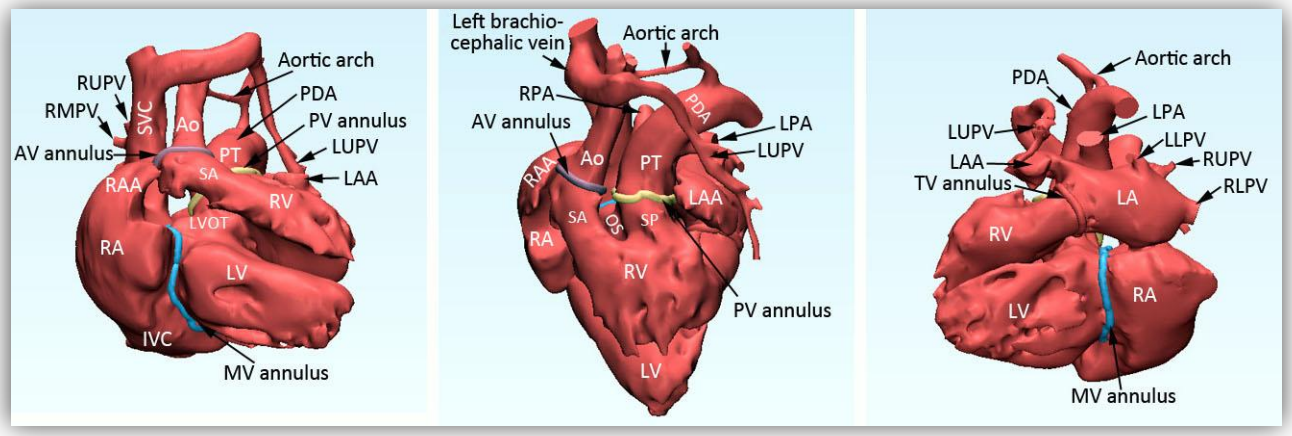
CASE 6. Mildly twisted heart with atrioventricular discordant connection, double outlet right ventricle with a subpulmonary VSD, and partial anomalous pulmonary venous connection

- Source images: ECG-gated contrast-enhanced CT angiograms.

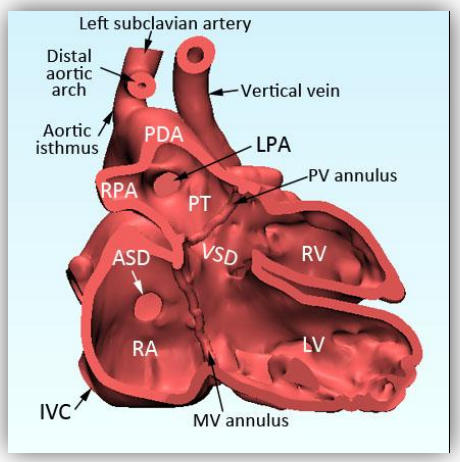
Summary:

- Situs solitus / Levocardia / Atrioventricular discordant connection / Double outlet right ventricle
- Superoinferior relationship of the ventricles with mildly twisted atrioventricular connection axes
- Aorta located anteriorly and rightward relative to the pulmonary arterial trunk.
- Bilateral infundibulum with a long subaortic and short subpulmonary infundibulum
- A unrestrictive VSD extending toward the outlet and committed to pulmonary outflow tract
- Severe tubular hypoplasia of the aortic arch
- Anomalous connection of the left upper pulmonary vein to the left brachiocephalic vein, anomalous connection of the right upper and middle pulmonary veins to the superior vena cava

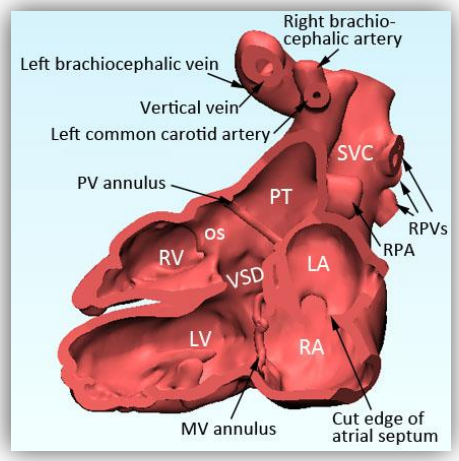
Models (5 pieces):



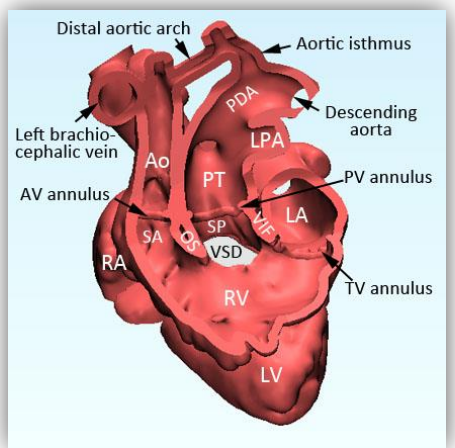
Model 6A. Volume rendered images of the cardiac chambers viewed from anterior (left panel), anterior and superior (middle panel), and posterior and inferior (right panel) aspects. OS, outlet septum; SA, subaortic infundibulum; SP, subpulmonary infundibulum.



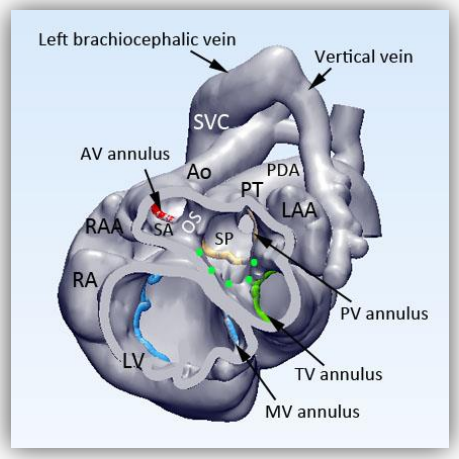
Model 6B. Volume rendered image of the interior of the ventricles and right atrium in coronal plane viewed from front.



Model 6C. Volume rendered image of the interior of the atria and ventricles in coronal plane viewed from behind. OS, outlet septum.



Model 6D. Volume rendered image of the ventricular septum viewed from the right ventricular aspect. OS, outlet septum; SA, subaortic infundibulum; SP, subpulmonary infundibulum; VIF, ventriculoinfundibular fold.



Model 6E. Volume rendered image of the base of the ventricles viewed from the apex. VSD margin is marked by green dots. OS, outlet septum; SA, subaortic infundibulum; SP, subpulmonary infundibulum.

Findings:

- There is atrial situs solitus. The atrial septum shows a curved configuration mostly in an oblique vertical plane (**6B**). The systemic venous connection is normal.
- The left upper pulmonary vein connects to the left brachiocephalic vein through a vertical vein. The right upper and middle pulmonary veins connect directly to the superior vena cava. Both lower pulmonary veins connect to the left atrium (**6A**).
- The right ventricle is superior to the left ventricle with the ventricular septum in a mostly horizontal plane. The atrial and ventricular septa are significantly malaligned (**6B, 6C**).
- The right atrium connects to the inferiorly located left ventricle (**6A-6C**). The left atrium connects to the superiorly located right ventricle (**6A** right panel and **6D**).
- The mitral valve is positioned antero-inferiorly relative to the tricuspid valve (**6A, 6E**) and low in the right atrium (**6B, 6C, 6E**). The tricuspid valve annulus is small. The opening axes of the tricuspid and mitral valves are not parallel (**6A** right panel). The mitral valve is oriented in a sagittal plane and opens left ward and mildly upward (**6A** left and right panels, **6B, 6C**). The tricuspid valve is oriented obliquely and opens forward, leftward and slightly downward (**6A** right panel, **6D**).
- Both the aorta and pulmonary arterial trunk arise from the right ventricle (**6A, 6D**). The right ventricular outflow tract is divided into the right anterior subaortic outflow tract (SA) and left posterior subpulmonary outflow tract (SP) by the outlet septum (OS). The outlet septum is exclusively a right ventricular structure, appearing deviated forward and rightward relative to the rest of the septum (**6D**). The subaortic outflow tract is a long narrow muscular infundibulum (**6A, 6D**). The subpulmonary outflow tract is a short unobstructed muscular infundibulum with the ventriculoinfundibular fold (VIF) intervening between the tricuspid and pulmonary valve annuli (**6A-6D**).
- The right ventricular side of the ventricular septum accepts the palmar surface of the observer's left hand with the thumb in the inlet, the wrist on the apex and the fingers in the outlet. Therefore, there is a so-called left hand pattern or chirality of the ventricular topology (**6D**).
- A large VSD involves the posterior basal part of the horizontally oriented ventricular septum (**6A-6E**). The VSD extends rightward and is located exclusively below the subpulmonary outflow tract. The VSD is the only outlet of the left ventricle (**6B, 6C**).
- The great arterial trunks are in parallel. There is severe tubular hypoplasia of the aortic arch (**6A, 6D**). A large patent ductus arteriosus connects the pulmonary arterial trunk to the descending aorta, forming a posterior shelf.

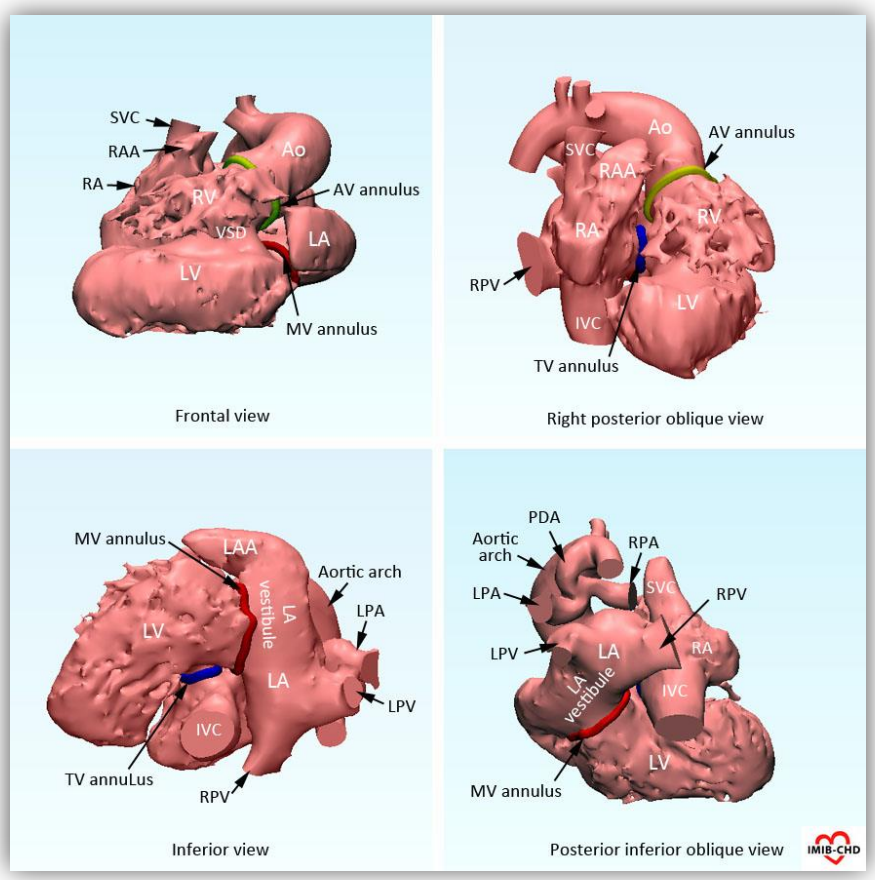
CASE 7. Twisted heart with aorta as a single arterial trunk from the right ventricle and pulmonary atresia in situs solitus and dextrocardia

- Source images: ECG-gated contrast-enhanced CT angiograms.

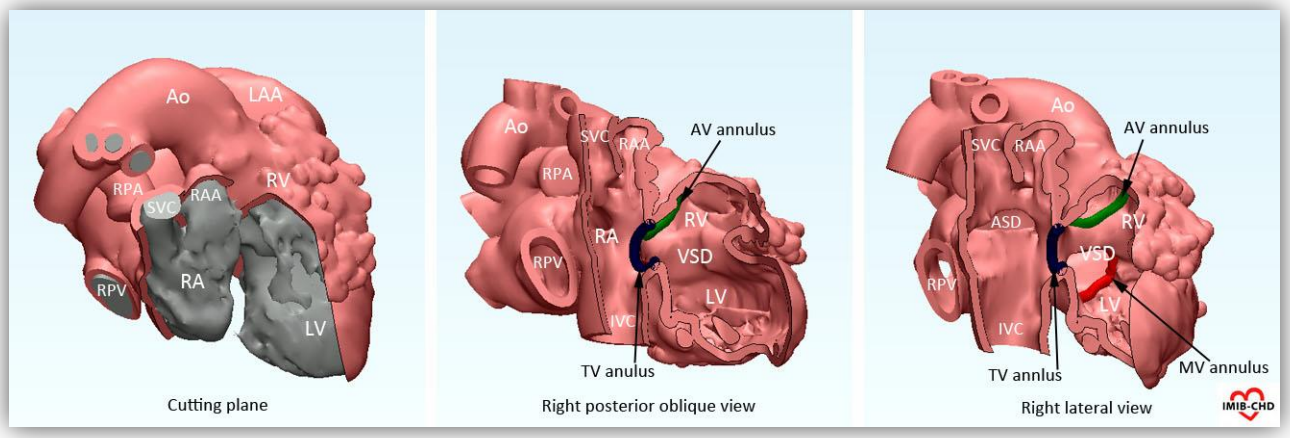
Summary:

- Situs solitus / Dextrocardia / Left aortic arch
- Concordant atrioventricular connection / aorta as the single arterial trunk arising from right ventricle
- Superoinferior relationship of the ventricles with the hypoplastic right ventricle on top of the left ventricle
- Non-parallel, twisted atrioventricular connection axes: tricuspid valve opening directly forward and mitral valve opening rightward and forward
- A large VSD between the inlets of the ventricles.
- Small tricuspid valve annulus overriding the ventricular septum through the VSD.
- Aorta arising from right ventricle through well-developed subaortic infundibulum
- Confluent pulmonary arteries supplied by a large left-side patent ductus arteriosus. No main pulmonary arterial segment.

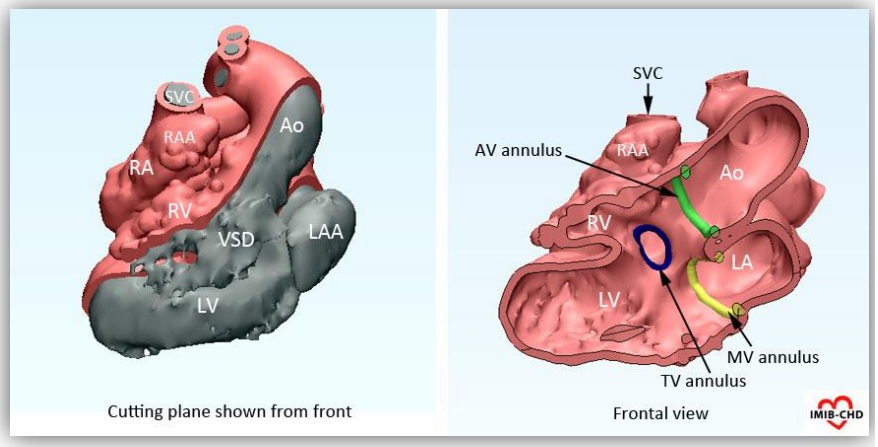
Models (5 pieces):



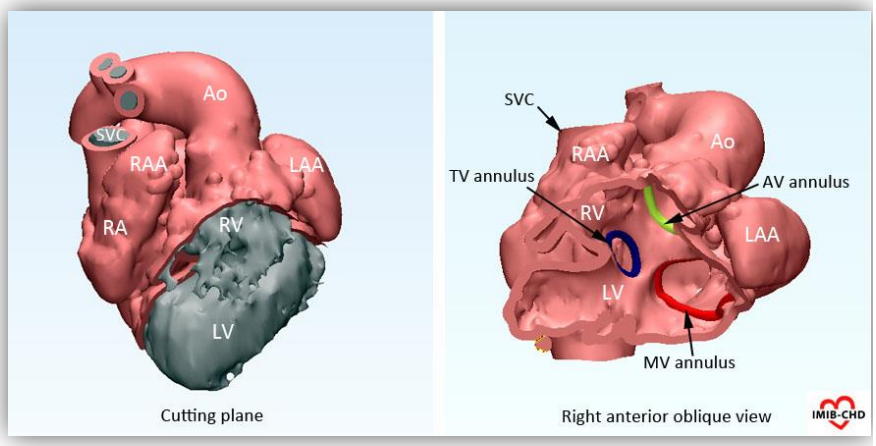
Model 7A. Volume rendered images of the cardiac chambers with the cardiac vale annuli marked with color.



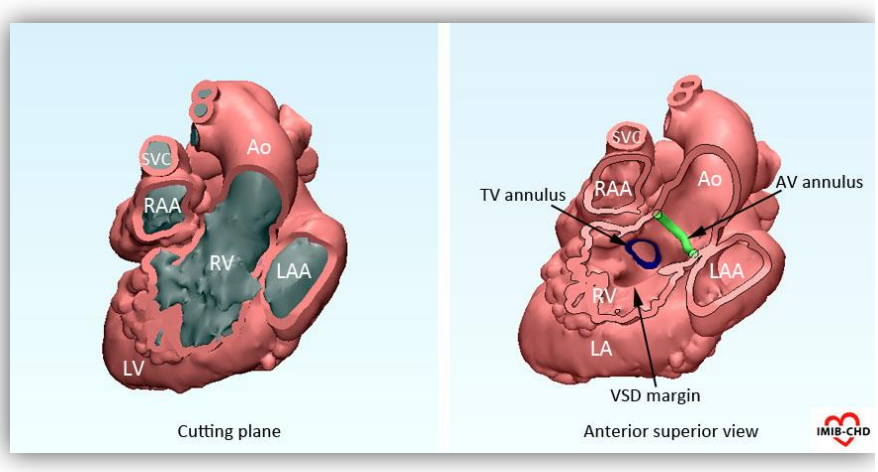
Model 7B. Volume rendered images showing the interior of the right atrium, right ventricle and left ventricle seen through a curved cut plane as shown in left panel figure.



Model 7C. Volume rendered images showing the interior of the left atrium, left ventricle and right ventricle seen through a curved cut plane as shown in left panel figure.



Model 7D. Volume rendered images showing the interior of the base of the ventricles seen through a curved cut plane as shown in left panel figure.



Model 7E. Volume rendered images showing the interior of the right ventricle seen after removal of the free wall of the right ventricle along the plane as shown in left panel figure.

Findings:

- There is atrial situs solitus and dextrocardia. The systemic and pulmonary venous connections are normal.
- The right ventricle is superior to the left ventricle with the ventricular septum in a horizontal plane (**7A-7E**).
- The right atrium connects to the superiorly located right ventricle through a small tricuspid valve (**7A, 7B**). The left atrium connects to the inferiorly located left ventricle (**7A, 7C, 7D**).
- The opening axes of the tricuspid and mitral valves are not parallel. The tricuspid valve opens forward and slightly leftward (**7A, 7B**). The mitral valve opens forward, rightward and downward (**7A, 7C**).
- The right ventricular apex is pointing rightward and slightly forward, while the left ventricular apex is pointing rightward and slightly posterior (**7A**).
- There is a large inlet VSD (**7B-7E**). There is overriding of the tricuspid valve annulus.
- The right ventricle is moderately hypoplastic.
- The right ventricular side of the ventricular septum accepts the palmar surface of the observer's right hand with the thumb in the inlet, the wrist on the right-sided apex and the fingers in the outlet. Therefore, there is a so-called right-hand pattern or chirality of the ventricular topology (**7E**).
- The aorta arises from the right ventricle through a long infundibulum (**7B, 7E**). The aortic arch is left sided and unobstructed.
- The pulmonary arterial trunk is not formed. The confluent pulmonary arteries are supplied by a tortuous left-sided ductus arteriosus.

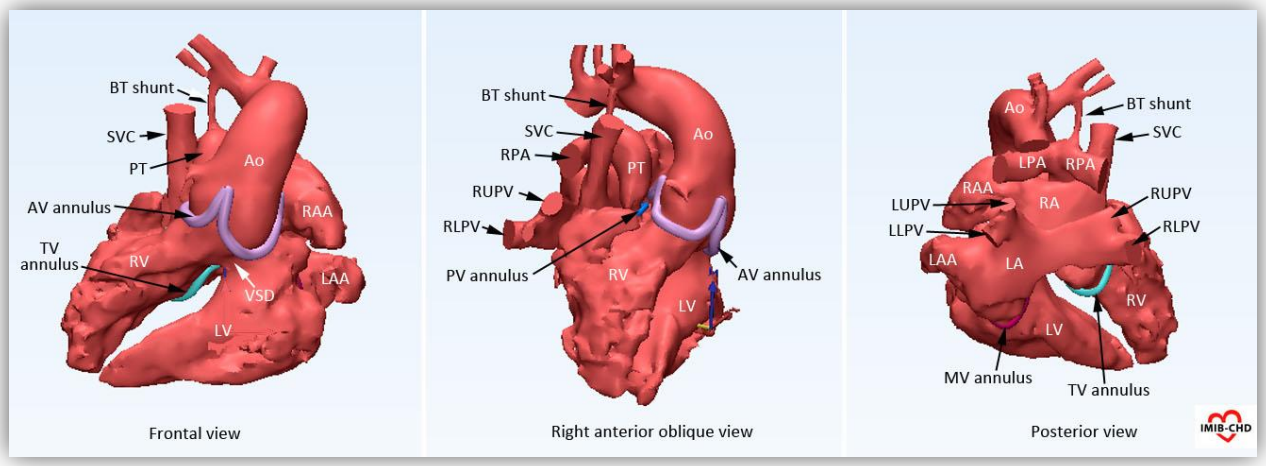
CASE 8. Superoinferiorly related ventricles with double outlet right ventricle in situs solitus and dextrocardia

- Source images: Non-ECG-gated contrast-enhanced MR angiograms.

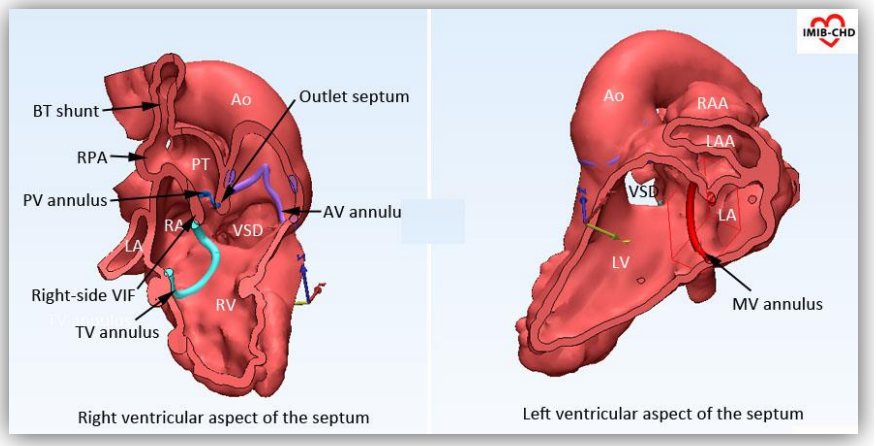
Summary:

- Situs solitus / Dextrocardia / Left aortic arch
- Major part of the right atrium positioned superior to the left atrium with juxtaposition of the atrial appendages on the left side
- Concordant atrioventricular connection / Double outlet right ventricle
- Superoinferior relationship of the ventricles with the right ventricle superior and posterior to the left ventricle
- Parallel atrioventricular connection axes
- Both aorta and pulmonary arterial trunk arising from the right ventricle through a longer subaortic infundibulum and a shorter and severely narrowed subpulmonary infundibulum
- Ascending aorta positioned leftward and anterior to the pulmonary arterial trunk
- A large outlet VSD closer to the aortic valve

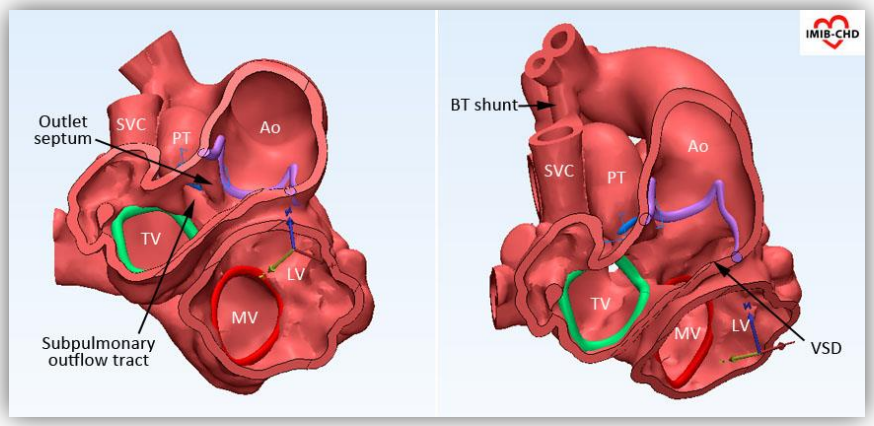
Models (3 pieces):



Model 8A. Volume rendered images of cardiac chambers with the cardiac valves marked with color. BT, Blalock-Taussig.



Model 8B. Volume rendered images showing the interior of the right and left ventricles after removal of the free wall of the atria and ventricles.



Model 8C. Volume rendered images showing the interior of the base of the ventricles viewed from the apex.

Findings:

- There is atrial situs solitus and dextrocardia. The systemic and pulmonary venous connections are normal.
- The right atrium is located to the right, anterior and superior relative to the left atrium. There is left juxtaposition of the atrial appendages with the right atrial appendage displaced to the left side and positioned above the left atrial appendage (**8A**).
- The right ventricle is superior and posterior to the left ventricle with the ventricular septum in an oblique plane between the coronal and horizontal planes (**8A**).
- There is a concordant atrioventricular connection (**8A and 8B**). Despite the superoinferior relationship between the ventricles, the opening axes of the tricuspid and mitral valves are parallel and the apices of the ventricles are pointing the same direction, rightward and forward.
- There is a large outlet VSD (**8B and 8C**). The VSD is immediately below the aortic valve, while it is at some distance from the pulmonary valve.
- The right ventricular side of the ventricular septum accepts the palmar surface of the observer's right hand with the thumb in the inlet, the wrist on the right-sided apex and the fingers in the outlet. Therefore, there is a so-called right-hand pattern or chirality of the ventricular topology (**8B**).
- Both the aorta and pulmonary trunk arise from the right ventricle through muscular infundibuli (**8A-C**). The subaortic infundibulum is longer than the subpulmonary infundibulum. The subpulmonary infundibulum or outflow tract is narrow between the right-sided ventriculoinfundibular fold (VIF) and the outlet septum.
- The ascending aorta is anterior to and slightly to the left of the small pulmonary arterial trunk (**8A-C**). There is a modified Blalock-Taussig (BT) shunt between the right subclavian artery and the right pulmonary artery.

CASE 9. Twisted heart with the left ventricle superiorly located in situs solitus, mesocardia and tetralogy of Fallot

- Source images: ECG-gated contrast-enhanced CT angiograms.

Summary:

- Situs solitus / Mesocardia / Left aortic arch
- Protruding heart underneath the deficient anterior chest wall (**Figure1**)
- Right atrial vestibular part elongated in a right-left direction with the tricuspid valve displaced far leftward. Elongated left atrial vestibular part in anteroposterior direction with the mitral valve displaced superiorly.
- Large secundum atrial septal defect
- Concordant atrioventricular connection with non-parallel atrioventricular connection axes.
- Mitral valve positioned superior to the tricuspid valve. Opening axis of the tricuspid valve directed forward and leftward. Opening axis of the mitral valve directed forward and slightly upward.
- Left ventricle located anterior and superior to the right ventricle. Right ventricle wrapping around the left ventricle from below and behind.
- Concordant ventriculoarterial connection with an exaggerated intertwined relationship between the right and left ventricular outflow tracts and arterial trunks.
- A large confluent perimembranous VSD in close proximity to the aortic valve.
- Aortic valve located posteriorly and inferiorly relative to the pulmonary valve
- Elongated pulmonary trunk.

Anterior chest wall:

The sternum as well as the muscle layer of the lower part of the midline anterior chest wall is deficient. The heart protrudes forward through the defect underneath the intact skin and is seen as a pulsating mass. Only the first segment of the sternum is present but is not normally formed. The sternum is bifid with its two lateral parts not fused.

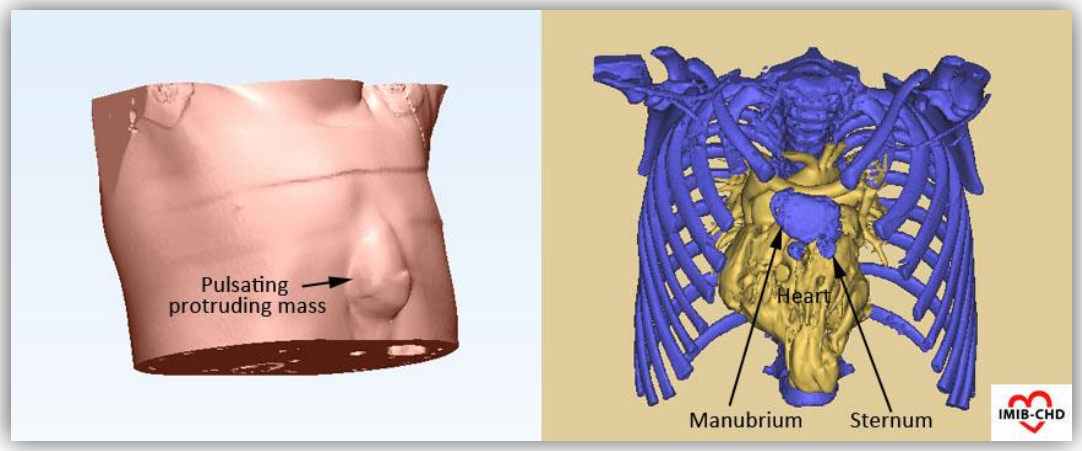
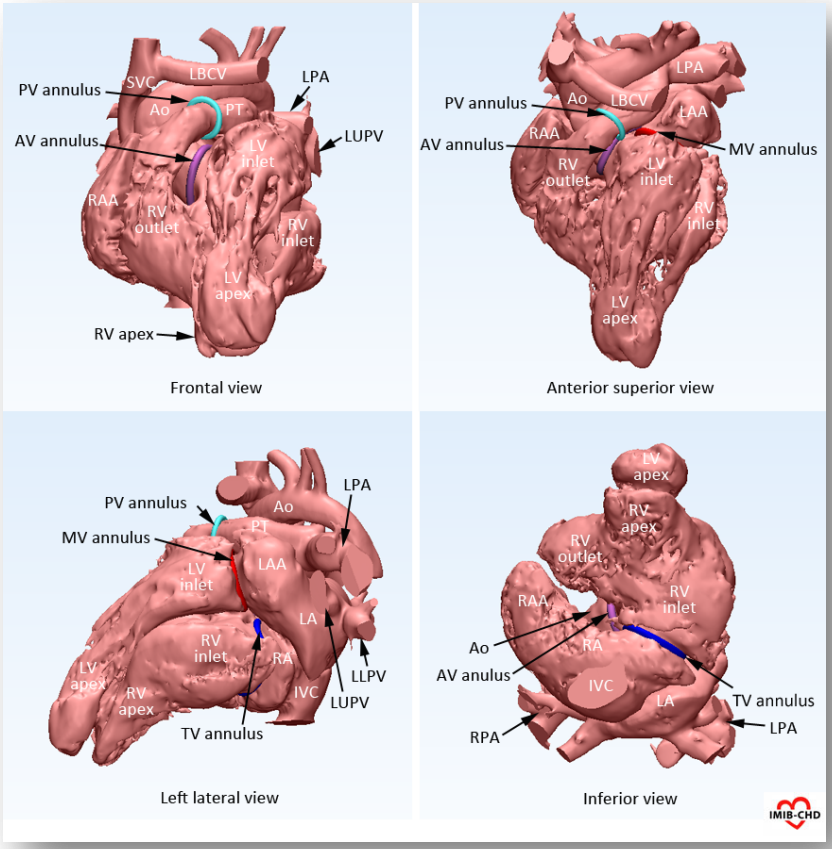
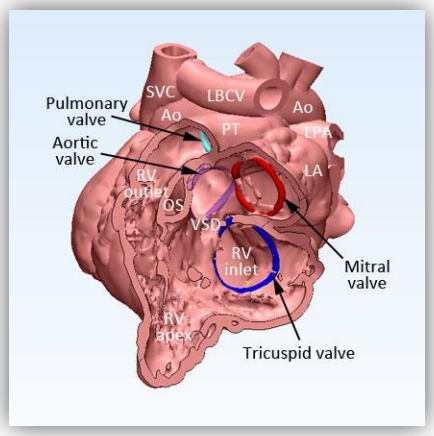


Figure 1. Volume rendered images of the skin surface of the chest wall (left panel) and bony thorax.

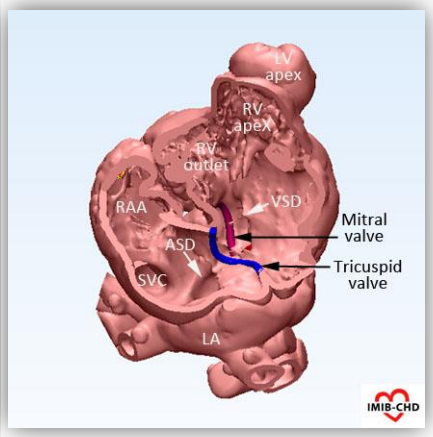
Models (4 pieces):



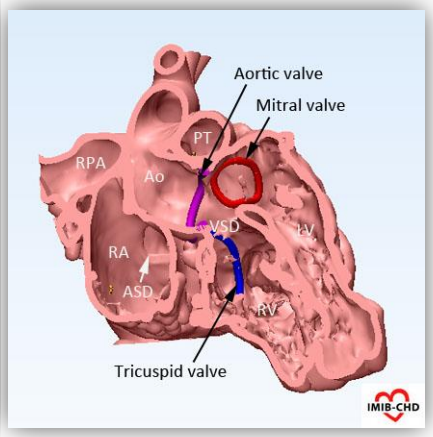
Model 9A. Volume rendered images of the cardiac chambers with the cardiac vale annuli marked with color.



Model 9B. Volume rendered image showing the interior of the right ventricle and base of the heart.



Model 9C. Volume rendered image showing the interior of the right atrium and right ventricle viewed from below.



Model 9D. Volume rendered image showing the interior of the left ventricle and ventricular septum viewed from the front.

Findings:

- The atrial situs is solitus. There is mesocardia with the apex of the ventricles protruding forward through the midline defect in the anterior chest wall. The systemic and pulmonary venous connections are normal.
- The right and left atria are grossly normally related (**9A**). However, the tricuspid valve annulus is displaced leftward, backward and downward, while the mitral valve annulus is displaced upward and forward (**9B- 9D**). As a consequence, the mitral valve annulus is above the tricuspid valve annulus. The opening axes of the tricuspid and mitral valves are not parallel. The opening axis of the tricuspid valve is directed forward and leftward. Opening axis of the mitral valve is directed forward and slightly upward. The left atrial outlet is elongated to form a tubular vestibulum underneath its appendage (**9A left lower panel**).
- The atrioventricular connection is concordant (**9A, 9B and 9D**). The ventricles are unusually oriented with the left ventricle located anteriorly and superiorly. The right ventricle wraps around the left ventricle from behind (**9A**).
- The right ventricular inlet with its tricuspid valve is displaced leftward, while the right ventricular outlet is displaced rightward (**9A**, **9B**). The right and left relationship between the inlet and outlet of the right ventricle is reversed.
- The ventriculoarterial connection is concordant (**9A-D**). The right and left ventricular outflow tracts demonstrate an exaggerated spiraling or intertwined relationship. The left ventricular outflow tract shows an acutely angulated in appearance and the right ventricular outflow tract is elongated (**9A**). The right ventricular aspect of the ventricular septum accepts the observer's right hand with the palm facing forward, the thumb in the tricuspid valve orifice, the fingers in the outlet and the wrist at the apex. Therefore, there is a right-hand pattern of ventricular topology **(9A-B**).
- The subpulmonary outflow tract shows mild narrowing due to deviation of the outlet septum (OS) (**9A, 9B**).
- The spiral relationship of the arterial trunks is kept with the pulmonary valve directly anterior and superior to the aortic valve (**9A**).
- There is a perimembranous VSD in close proximity to the aortic valve. The aortic valve overrides the ventricular septum.

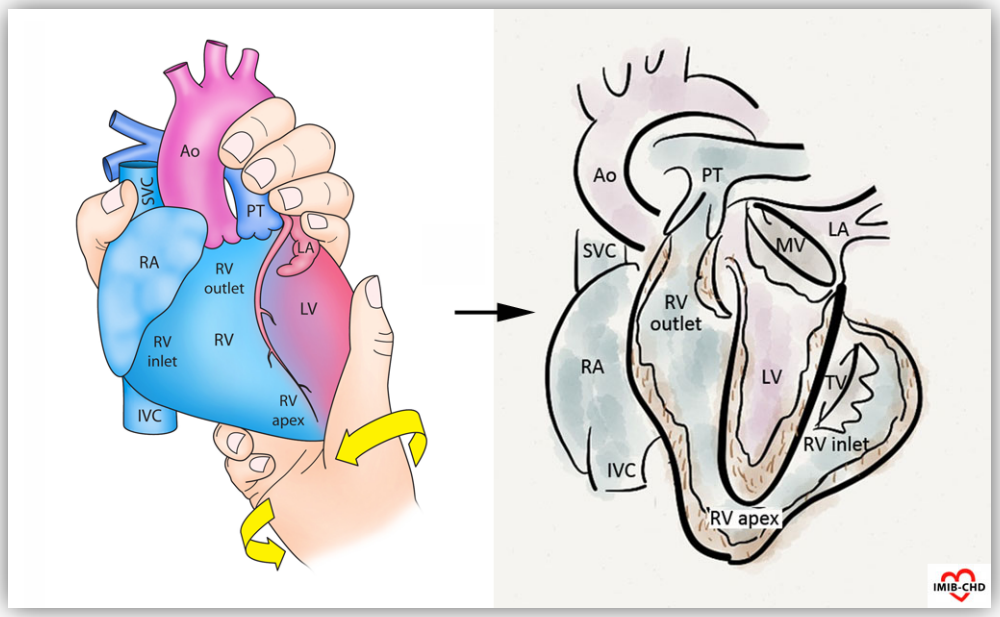


Figure 2. Hypothetical pathogenesis of the twisted atrioventricular connection in this case. The usual heart with tetralogy of Fallot is twisted counter-clockwise by the observer's right hand with the left hand holding the posterior aspect of the heart. The left ventricle is moved forward in front and above the right ventricle.

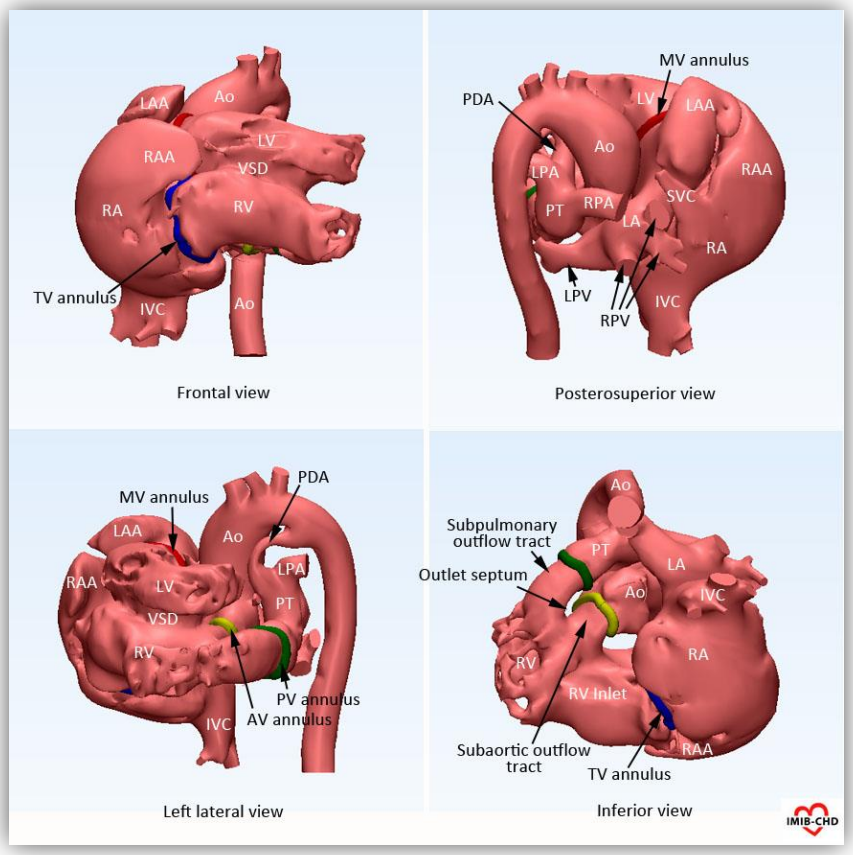
CASE 10. So-called topsy-turvy heart with double outlet right ventricle

- Source images: ECG-gated contrast-enhanced CT angiograms.

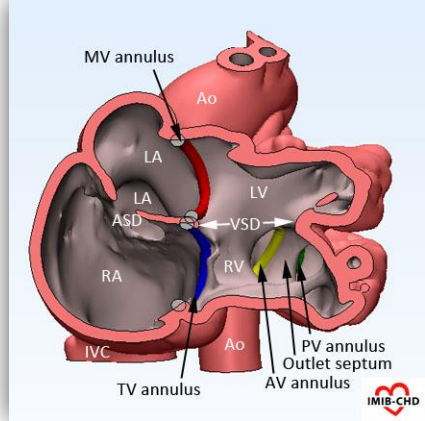
Summary:

- Situs solitus / Levocardia / Left aortic arch
- Unusual superoinferior relationship of the cardiac chambers with the left atrium and left ventricle above and the major part of the right atrium and right ventricle below.
- Juxtaposition of the atrial appendages on the right side
- Large secundum atrial septal defect
- Concordant atrioventricular connection with parallel atrioventricular connection axes
- Right ventricular outflow tracts arising from the far posterior aspect of the right ventricle.
- Double outlet right ventricle through a muscular infundibulum that is evenly divided into the subaortic and subpulmonary outflow tract
- A large confluent VSD that is closer to the subaortic outflow tract
- Aortic valve located anterior and slightly to the right of the pulmonary valve
- Long ascending aorta and pulmonary arterial trunk taking acutely angulated proximal courses.
- Confluent pulmonary arteries with a tortuous patent ductus arteriosus.

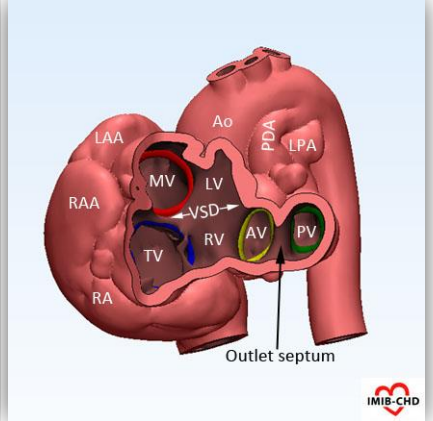
Models (4 pieces):



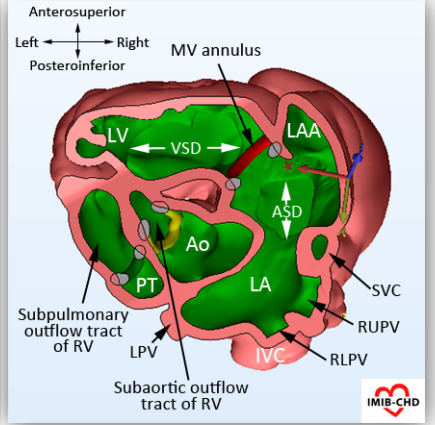
Model 10A. Volume rendered images of the cardiac chambers with the cardiac valve annuli marked with color.



Model 10B. Volume rendered image showing the interior of the four chambers of the heart.



Model 10C. Volume rendered image showing the interior of the base of the ventricles viewed from the apex.



Model 10D. Volume rendered image showing the interior of the left atrium and left ventricle viewed from behind and above.

Findings:

- There is atrial situs solitus and levocardia. The systemic and pulmonary venous connections are normal.
- The right atrium is located to the right, anterior and inferior relative to the left atrium. There is right juxtaposition of the atrial appendages with the left atrial appendage being displaced to the right side and sitting above the right atrial appendage (**10A**). There is a large secundum type atrial septal defect.
- The four-chambers of the heart are superoinferiorly related with the left atrium and ventricle located on top of the right atrium and ventricle (**10A and 10B**). The atrial and ventricular septa are horizontally oriented. Note that the left atrial vestibular part is markedly elongated between the pulmonary venous confluence and the mitral valve (**10D**).
- There is a concordant atrioventricular connection (**10A and 10B**). Despite a superoinferior relationship between the ventricles, the opening axes of the tricuspid and mitral valves are parallel and the apices of the ventricles are pointing the same leftward direction.
- The right ventricular side of the ventricular septum accepts the palmar surface of the observer's left hand with the thumb in the inlet, the wrist on the left-sided apex and the fingers in the outlet. Therefore, there is a so-called left-hand pattern or chirality of the ventricular topology (**10B and 10C**).
- The right ventricular outlet is a completely muscular infundibulum projecting backward from the inferiorly located right ventricle (**10A right upper panel**). It is divided equally into the right-sided subaortic and left-sided subpulmonary outflow tract by the outlet septum.
- There is a large confluent VSD (**10B-C**). The VSD is closer to the aortic valve.
- The ascending aorta is superoinferiorly elongated as it arises from the inferiorly located right ventricle. It takes an acutely angled course in its proximal part (**10A**). The main pulmonary artery is also long and takes an angulated course.
- A long and tortuous patent ductus arteriosus connects the descending aorta to the top of the pulmonary arterial trunk (**10A left lower panel**).

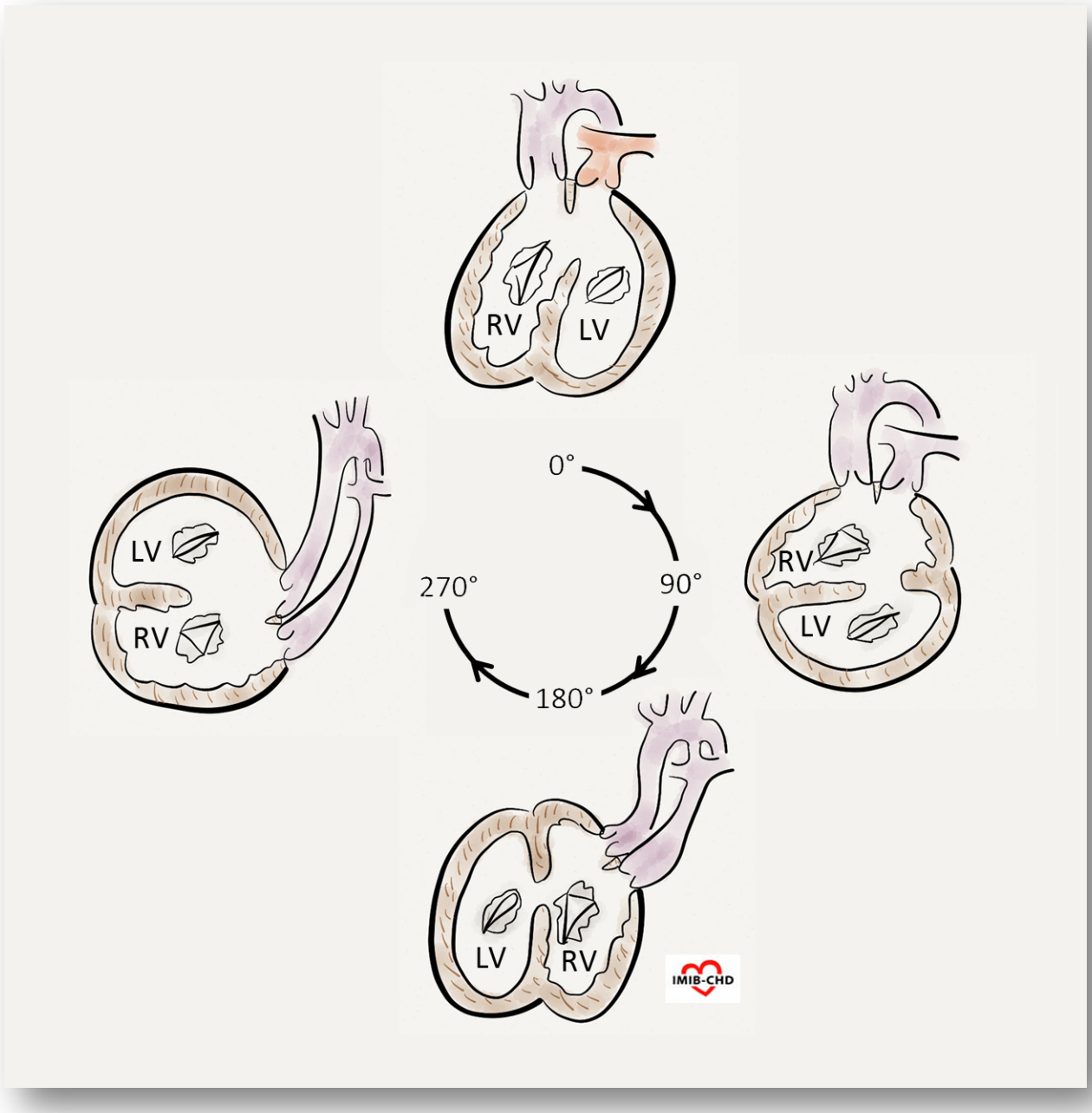


Figure. Hypothetical pathogenesis of topsy-turvy ventricular relationship in this case. The usual orientation of cardiac chambers and great arterial trunks is shown on the top. With 90 degrees of organoaxial rotation of the cardiac chambers as seen from the apex of the ventricles, the atria and ventricles show a superoinferior relationship with the right atrium and right ventricle located above the left atrium and left ventricle. Further rotation to 180 degrees results in the left atrium and left ventricle lying in front of the right atrium and right ventricle. With 270 degrees of rotation, the left atrium and left ventricle are placed on top of the right atrium and right ventricle, respectively. As both arterial trunks leave the inferiorly located right ventricle, they are markedly elongated.

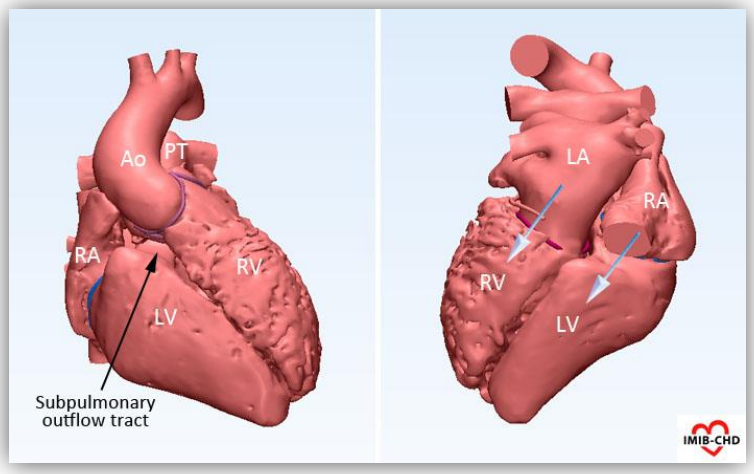
CASE 11. Congenitally corrected transposition of the great arteries with superoinferior ventricles and parallel atrioventricular valves

- Source images: ECG-gated contrast-enhanced CT angiograms.

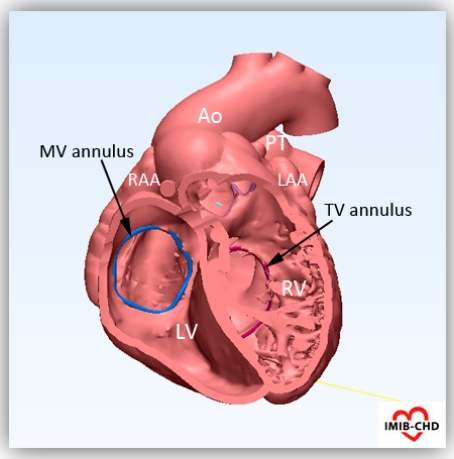
Summary:

- Situs solitus / Levocardia / Left aortic arch
- Atrioventricular discordant connection with parallel axes of mitral and tricuspid valve openings
- Superoinferior relationship of the ventricles with the right ventricle located leftward and superior relative to the left ventricle
- Small permembranous ventricular septal defect
- Long subaortic and short subpulmonary infundibulum
- Ascending aorta anterior and slightly left of the pulmonary arterial trunk

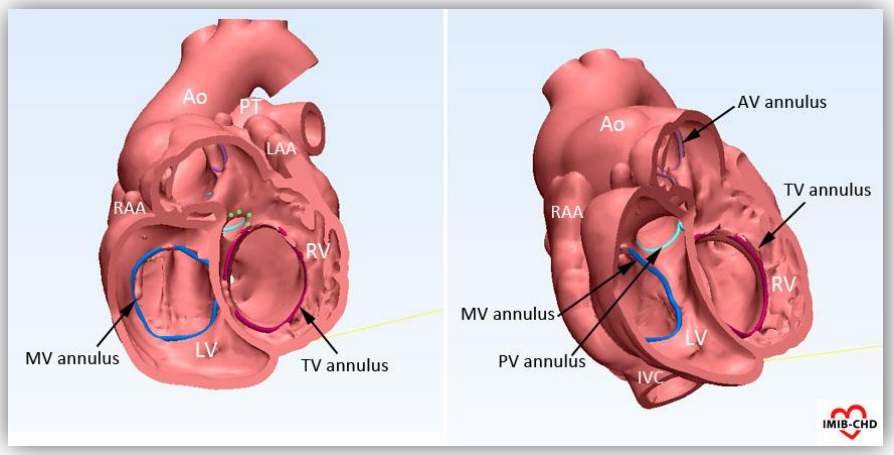
Models (3pieces):



Model 11A. Volume rendered images of the cardiac chambers with the cardiac valve annuli marked with color.



Model 11B. Volume rendered image showing the interior of the base of the ventricles viewed from the apex.



Model 11C. Volume rendered image of the interior of the heart after removal of the anterosuperior walls of the ventricles. VSD is marked with green dots.

Findings:

- There is atrial situs solitus and levocardia. The systemic and pulmonary venous connections are normal.
- The atrial relationship is normal (**11A**).
- The right ventricle is located leftward and superior relative to the left ventricle.
- The right atrium connects to the morphologically left ventricle on the right, and the left atrium connects to the morphologically right ventricle on the left. The atrioventricular connection axes are completely parallel to each other.
- There is a small ventricular septal defect (**11B**). It abuts on the top of tricuspid valve, characterizing the defect as a perimembranous type.
- The pulmonary arterial trunk arises from the left ventricle through a short muscular infundibulum (**11B**). Note that the pulmonary valve annulus (marked in green color) is not in direct contact with the mitral valve annulus (marked in blue) (**11B and 11C**). The aorta arises from the right ventricle through a long muscular infundibulum (**11A-C**).
- The aortic valve is located anterior and slightly leftward relative to the pulmonary valve (**11A-C**).
- The ascending aorta takes an oblique course on the far left upper aspect of the heart to form a left aortic arch (**12A**).

Discussion:

When congenitally corrected transposition occurs in situs solitus and levocardia, the ventricles tends to be arranged superoinferiorly as if the heart is tilted upward by a hand placed in the apex. The extreme example of such relationship is seen in classic double inlet left ventricle in which the right ventricle is located superiorly and leftward as shown in **Case 12**.

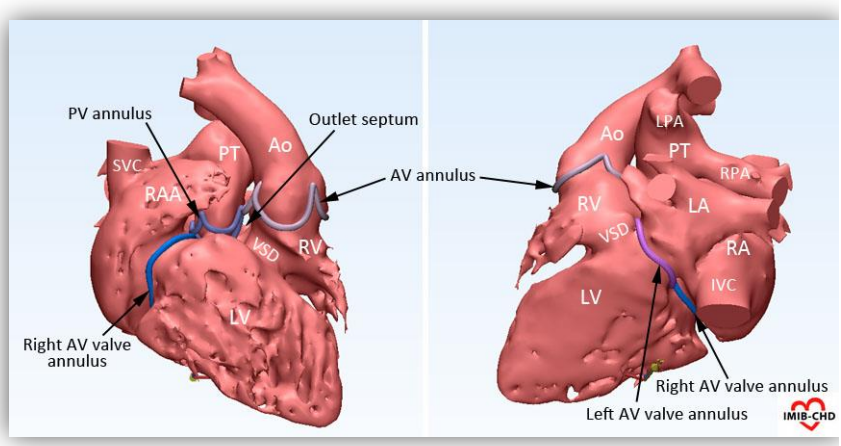
CASE 12. Classic double inlet left ventricle and transposition of the great arteries

- Source images: ECG-gated contrast-enhanced CT angiograms.

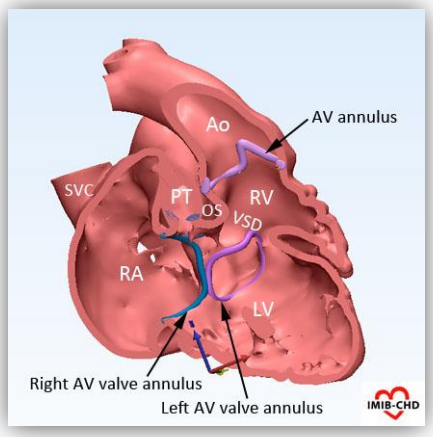
Summary:

- Situs solitus / Levocardia / Left aortic arch
- Both right and left atria connecting to the main chamber of left ventricular morphology (double inlet left ventricle). Parallel open axes of the right and left atrioventricular valves.
- Hypoplastic right ventricle located superior, left ward and slightly posterior to the left ventricle
- A large ventricular septal defect.
- Pulmonary arterial trunk arising from the left ventricle without a muscular infundibulum. Aorta arising from the right ventricle through a muscular infundibulum
- Subpulmonary outflow stenosis and small pulmonary valve annulus

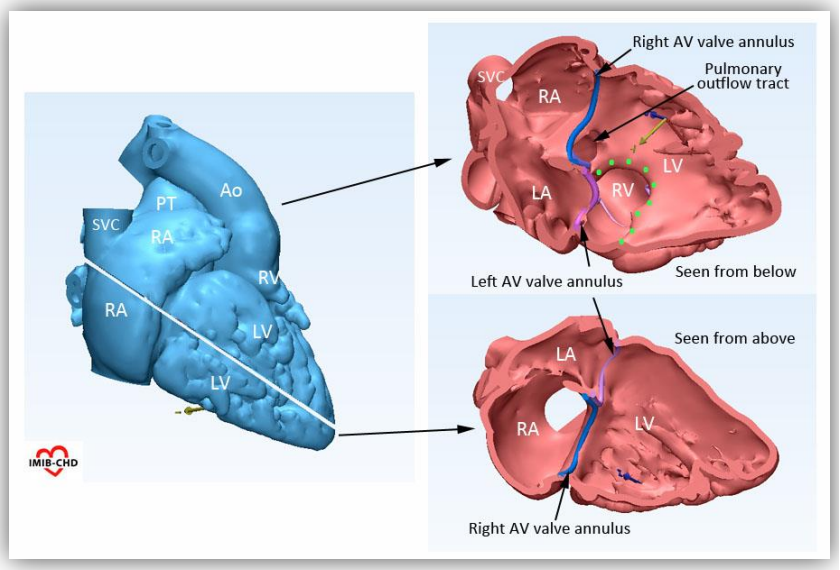
Models (3pieces):



Model 12A. Volume rendered images of the cardiac chambers with the cardiac valve annuli marked with color.



Model 12B. Volume rendered endocardial surface image of the heart with the anterior wall of the right atrium and ventricles removed.



Model 12C. Volume rendered endocardial surface images of the heart. The heart was divided into two parts through the plane shown on the left panel. The cutting plane is through both atrioventricular valves and the apex of the left ventricle. The margin of the VSD is marked with green dots.

Findings:

- There is atrial situs solitus and levocardia. The systemic and pulmonary venous connections are normal.
- The atrial relationship is normal (**12A and 12C**).
- There is a large secundum atrial septal defect.
- The right ventricle is small and located superior, leftward and slightly posterior to the left ventricle. The spatial relationship between the ventricles is similar to that of the typical form of congenitally corrected transposition in situs solitus and levocardia as shown in **Case 11**, although the right ventricle is small in this case.
- Both right and left atria are connected to the main chamber of left ventricular morphology through the separate atrioventricular valves that have parallel opening axes. The right ventricle does not have any direct connection to the atria.
- There is a large muscular ventricular septal defect (**12A-C**). This is the only inflow to the right ventricle.
- As there is no atrioventricular valve in the right ventricle, the ventricular topology cannot be defined using the observer's hands. Considering that the case is an extreme form of straddling left atrioventricular or tricuspid valve in the presence of congenitally corrected transposition skeleton, it can safely be regarded that there is a so-called left-hand pattern or chirality of the ventricular topology.
- The left ventricle gives rise to the pulmonary arterial trunk. There is fibrous continuity or direct contact without intervening muscle (ventriculoinfundibular fold) between the right-side atrioventricular valve and the pulmonary valve (**12B**). The outlet septum (**OS** in **12B**) is deviated rightward and backward, encroaching of subpulmonary out flow tract.
- The right ventricular outlet is a completely muscular infundibulum giving rise to the aorta (**12A and 12B**).
- The aortic valve is located anterior and leftward relative to the pulmonary valve (**12A and 12B**).
- The ascending aorta takes an oblique course on the far left upper aspect of the heart to form a left aortic arch (**12A**).

Discussion:

Double inlet left ventricle is characterized by the small morphologically right ventricle positioned superiorly above the left ventricle. The right ventricle can be on the left, right or directly anterior to the left ventricle. **Case 12** is the most common form of double inlet left ventricle in the presence of situs solitus and levocardia, showing the right ventricle on the left and giving rise to the left-sided aorta. The ventricular and great arterial relationships are similar to those seen in classical congenitally corrected transposition in situs solitus and levocardia as shown **Case 11**. Between these two uncommon but well recognized entities, there is a spectrum of rare abnormalities that show varying degrees of straddling / overriding of the tricuspid valve.