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.

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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 suprinferior 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 suprinferiorly 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](image-url) **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.
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 RELATIONSHIP AND 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 ABNORMALITIES

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 inversus 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 discordant 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 venticule. As twisting implies malignment 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**


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