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# Anatomy and echocardiography of discordant atrioventricular connections

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THE MOST APPROPRIATE WAY OF DESCRIBING THE congenital cardiac malformations unified because the atrial chambers are joined across the atrioventricular junctions to morphologically inappropriate ventricles has long been contentious. In the past, the lesions have been described in such arcane terms as mixed levocardia,[1] while "ventricular inversion" still retains it currency in some circles. As we will show in this review, the abnormal arrangements at the atrioventricular junctions can be found with various patterns, but most frequently the patients also have the arterial trunks arising from morphologically inappropriate ventricles. This combination is best described as congenitally corrected transposition, and will form the focus of our review. It is salutary to note that, when von Rokitansky gave the first description of this combination,[2] one of his illustrations was ideally suited to aid the understanding of modern-day echocardiographers (Fig. 1). We hope to emulate von Rokitansky in our own review.

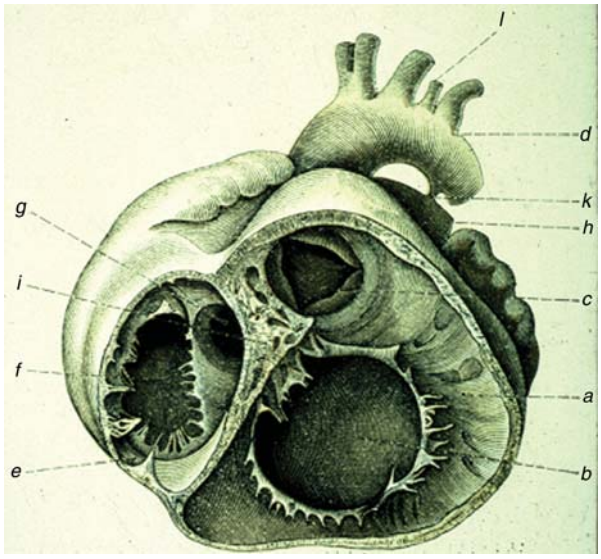


**Figure 1.**
*This lithograph from the atlas of Rokitansky[2] shows the anatomic arrangement of congenitally corrected transposition as seen in a short axis cut across the ventricular mass. To the left hand, the mitral and pulmonary valves (f,g) overlap within the roof of the morphologically left ventricle, which is right sided. The aortic and tricuspid valves (c,b) are separated by the supraventricular crest in the roof of the left-sided morphologically right ventricle.*

## Anatomic arrangement

The unifying feature of the hearts to be discussed in our review is that the atrial chambers, which can be usually arranged or mirror-imaged, are joined across the atrioventricular junctions to morphologically inappropriate ventricles. In the majority of patients with this arrangement, well described as discordant atrioventricular connections, the ventricles themselves are also joined across the ventriculo-arterial junctions to morphologically inappropriate arterial trunks, in other words they also possess discordant ventriculo-arterial connections (Fig. 2).

In this respect, several aspects of this description warrant further comment. First, the combination of discordant atrioventricular and ventriculo-arterial

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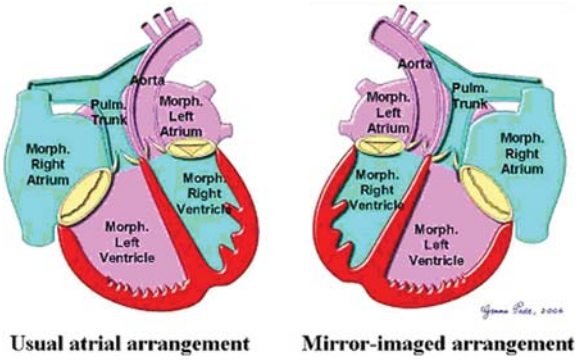


**Figure 2.**
*The cartoon shows the segmental combinations producing the arrangement well described as congenitally corrected transposition.*

connections can only exist when the atrial chambers themselves are usually arranged or mirror-imaged. Comparable arrangements of the ventricular mass can be found in the setting of visceral heterotaxy, when the atrial appendages are isomeric, but the connections across the atrioventricular junctions themselves cannot be discordant when both appendages have the same morphology. Second, the connections are discordant across both atrioventricular junctions. Patients with similar arrangements of the ventricular mass can also be found when both atriums join to the same ventricle, or else only one atrium is joined to the ventricular mass. It is not correct to describe these "close cousins" as having discordant atrioventricular connections. The arrangement discussed in this review can only exist when both atrioventricular connections are discordant. Third, many investigators consider discordant atrioventricular connections to be synonymous with atrioventricular discordance. Indeed, when some of us first described the arrangement in our own version of segmental analysis,[3] we used the term atrioventricular discordance. We now know that this was incorrect, and probably contributed to some of the confusions regarding the difference between segmental analysis[4] as opposed to the sequential format, which was subsequently popularized in Europe.[5] When Van Praagh et al. first introduced the segmental approach,[6] they used the term "atrioventricular discordance" to describe the combination of usual atrial arrangement, which they labeled solitus, or "S", with left hand ventricular topology, or l-loop, which they labeled "L" in their short hand notation. They then pointed out that the combination of atrial mirror-imagery, which they labeled "I", and right hand ventricular topology, which they shortened to "D", also produced atrioventricular discordance. They also emphasized, however, that these combinations of {S,L,*} or {I,D,*}, could be found with double inlet ventricle, or tricuspid atresia, or various other combinations. In their approach as initially used, therefore, "atrioventricular discordance" did not refer specifically to the arrangement in which atrial chambers were joined to morphologically inappropriate ventricles. When the Europeans first promulgated their sequential modifications, this nuance escaped their attention, and we are unsure whether they would still describe patients with so-called "single left ventricle {S,L,L}" as atrioventricular discordance. Be that as it may, any potential confusion can be avoided if the term "discordant atrioventricular connections" is used to describe the abnormal junctional pattern, since this emphasizes the plurality of the junctions, along with the fact that it specifically describes the abnormal connections. It should also be noted that, in the approach to nomenclature now recommended by Van Praagh,[7] it is argued that the atrial chambers do not connect directly to the ventricles, but that the atrioventricular canal interposes between the two segments. This is not the case with the sequential segmental approach[8] to be followed subsequently in this review, since the atrial chambers and ventricles are considered to join one another across the atrioventricular junctions, and hence their cavities are truly in connection one with the other.

The discordant connections, which cancel one another out so as to produce the congenital correction of the circulatory patterns, are then well demonstrated by sections taken in the four chamber plane. These show that the atrial chamber with the broad-based appendage, receiving the systemic venous tributaries, is connected across one of the atrioventricular junctions with the ventricle possessing fine apical trabeculations, while the atrium with the tubular appendage, which receives the pulmonary venous return, is connected across the other atrioventricular junction to the ventricle possessing coarse apical trabeculations (Fig. 3 – left hand panel). The atrioventricular valve within the finely trabeculated ventricle is a morphologically mitral valve, lacking any direct cordal attachments to the ventricular septum. In most instances, the hinge of its atrioventricular valve is situated more basally that that of the other atrioventricular valve. The atrioventricular valve entering the coarsely trabeculated ventricle does possess septal attachments, and its hinge is positioned more towards the ventricular apex, making it a morphologically tricuspid valve. Sections taken more superiorly through the ventricular mass show that the rapidly bifurcating arterial trunk arises from the finely trabeculated ventricle, almost always with fibrous continuity between its valve and the morphologically mitral valve (Fig. 3 – middle panel). The most superior sections show that the arterial trunk supplying the coronary and brachiocephalic arteries arises from the coarsely trabeculated ventricle (Fig. 3 – right hand panel).

It is rare, however, to find the arrangement as shown in Figure 3, in which both the atrial and ventricular

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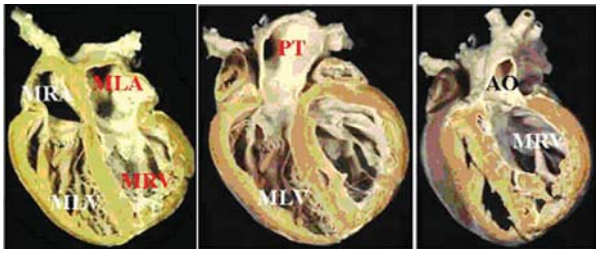


**Figure 3.**
*These sections through a heart from a patient with congenitally corrected transposition in the setting of usual atrial arrangement show the discordant connections across both the atrioventricular and ventriculo-arterial junctions, the two sets of discordant connections cancelling each other out so as congenitally to correct the patterns of circulation. The left hand panel is through the atrioventricular junctions (MRA, MLA – morphologically right and left atriums), the middle panel shows the pulmonary trunk (PT) arising from the morphologically left ventricle (MLV), while the right hand panel shows the aorta (AO) arising from the morphologically right ventricle (MRV).*

septal structures were intact. Patients with this arrangement can survive to ripe old ages without the abnormal junctional arrangements coming to attention. Much more frequently, the patterns of circulation are disturbed by the presence of associated anomalies, or else because of the development of abnormalities of atrioventricular conduction. Of the associated malformations, three are sufficiently frequent to be considered part and parcel of congenitally corrected transposition.[9] These are a ventricular septal defect, obstruction of the outflow tract from the morphologically left ventricle, and abnormalities of the morphologically tricuspid valve.[10] These lesions can show just as much variability as expected in patients with concordant atrioventricular connections, and the same approach is taken to categorization and diagnosis. Any other associated malformation, however, can also co-exist in the setting of congenitally corrected transposition. Variations can also be found in the relationships of the chambers, and in the structure of the ventricular outflow tracts, but these are not constant. Thus, whilst in the majority of patients the aorta will be found anterior and leftward relative to the pulmonary trunk, this is not a constant finding. It is a mistake, therefore, to consider "l-transposition" as synonymous with congenitally corrected transposition. From the surgical stance, it is important to note that there is an abnormal location of the atrioventricular node in most patients having congenitally corrected transposition,[11] albeit that the bundle can arise from a regular atrioventricular node in the presence of better alignment between the atrial and ventricular septal structures.[12] The arrangement of the coronary arteries can be described in the same fashion as that adopted for physiologically uncorrected transposition. This information, of course, is essential for the surgeon contemplating a double switch procedure. As we will show, all of these features are now readily demonstrable using echocardiographic techniques.

## Echocardiographic approach to diagnosis

As with all congenital cardiac malformations, the echocardiographic approach to patients with discordant atrioventricular connections requires a methodical sequential segmental approach. Such a strategy is all the more important in initial diagnosis of those with discordant atrioventricular connections, since these children may be asymptomatic and otherwise elude clinical detection. Recognition of discordant atrioventricular connections requires a complete understanding of the anatomic features of the left and right atriums, the mitral and tricuspid valves, and the morphologically left and right ventricles.

As in the normal heart, the morphologically left ventricle is characterized by fine apical trabeculations, and lack of cordal attachments of its atrioventricular valve to the ventricular septum. The morphologically right ventricle has coarse trabeculations, along with direct attachment of the tension apparatus of its atrioventricular valve to the ventricular septum (Fig. 3 – left hand panel). In addition, the moderator band is a characteristic feature of the morphologically right ventricle. Identification of this structure can be particularly helpful in fetal echocardiography, where characterization of the difference between the apical trabeculations can be more challenging. In the setting of discordant atrioventricular and ventriculo-arterial connections, attention to the origins of the great arteries demonstrates that the leaflets of the pulmonary valve, arising from the morphologically left ventricle, are almost always in fibrous continuity with the leaflets of the morphologically mitral valve (Fig. 3 – middle panel). The leaflets of the aortic valve, in contrast, arising from the morphologically right ventricle, have no continuity with the leaflets of the morphologically tricuspid valve, but rather are supported by the infundibulum of the right ventricle, described by some morphologists as a "conus" (Fig. 3 – right hand panel).

These features which distinguish between the morphologically right and left ventricles can be obtained in multiple imaging planes. Apical images often permit identification of the nature of the trabeculations, including the presence of the moderator band, and the attachments of the atrioventricular valves to the ventricular septum (Fig. 4).

Subcostal transverse and sagittal imaging planes are particularly helpful for defining the relationship of the atrioventricular and the arterial valves. In particular, these sections show well the fibrous continuity between the leaflets of the mitral and pulmonary valves

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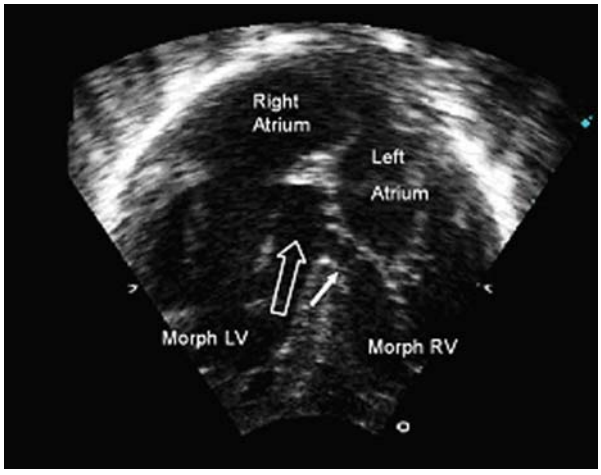


**Figure 4.**
*This apical cut from a patient with congenitally corrected transposition shows well the discordant atrioventricular connections. Note the lack of direct attachments of the right-sided morphologically mitral valve to the ventricular septum (large open arrow), as opposed to the direct septal attachments of the left-sided morphologically tricuspid valve (small white arrow).*

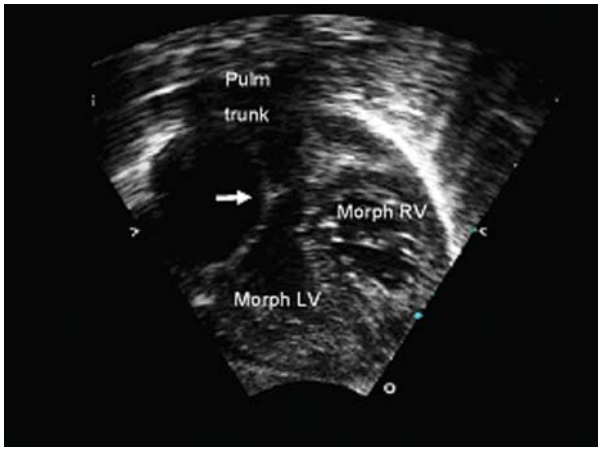


**Figure 5.**
*This subcostal transverse image of the outflow tract from the morphologically left ventricle (Morph LV) in a patient with congenitally corrected transposition shows well the fibrous continuity between the leaflets of the morphologically mitral and pulmonary valves (white arrow).*

in the morphologically left ventricle (Fig. 5). This feature can also be well seen in parasternal long axis planes, which also show the discontinuity between the leaflets of the tricuspid and aortic valves within the morphologically right ventricle (Fig. 6).

As already discussed, a number of important associated lesions occur in the setting of discordant atrioventricular connections. These lesions include ventricular septal defects, sub-pulmonary obstruction, and anomalies of the morphologically tricuspid valve, such as Ebstein's malformation. A complete delineation of such

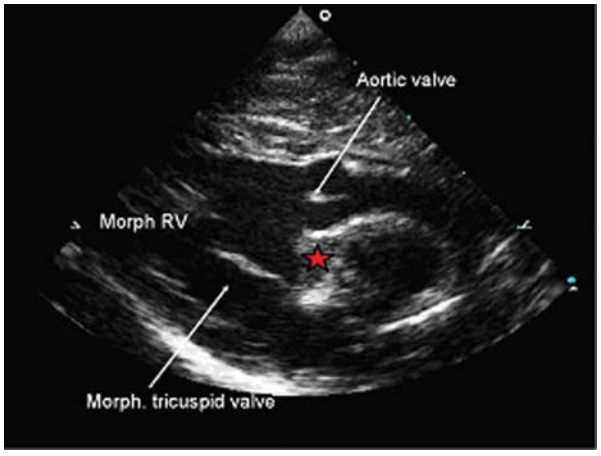


**Figure 6.**
*This parasternal long axis image shows well the discontinuity between the leaflets of the tricuspid valve and those of the aortic valve in the morphologically right ventricle (Morph RV), the aortic valve being supported by a complete muscular infundibulum, or conus (red star).*

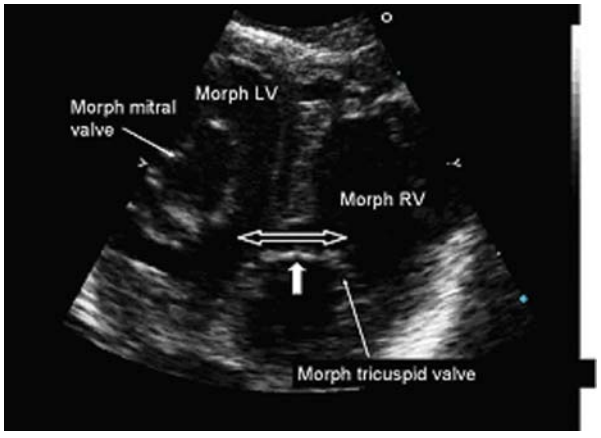


**Figure 7.**
*This parasternal short axis image is from a patient with congenitally corrected transposition and a perimembranous ventricular septal defect.*

lesions by echocardiography is particularly important, as the surgical approaches, and timing of surgical interventions, will be determined by these associated lesions.

A ventricular septal defect is the commonest associated malformation, and is seen in the majority of patients presenting for treatment. The ventricular septal defect is typically perimembranous. It can be imaged in multiple echocardiographic imaging planes. The subcostal transverse plane shows well the crucial relationship in the postero-inferior margin of the defect between the septal leaflet of the morphologically tricuspid valve and the leaflets of the pulmonary valve, this being the phenotypic feature of the perimembranous defect (Fig. 7). Doppler, and colour Doppler, interrogation can help plan medical and surgical management. The patient with a large and unrestrictive

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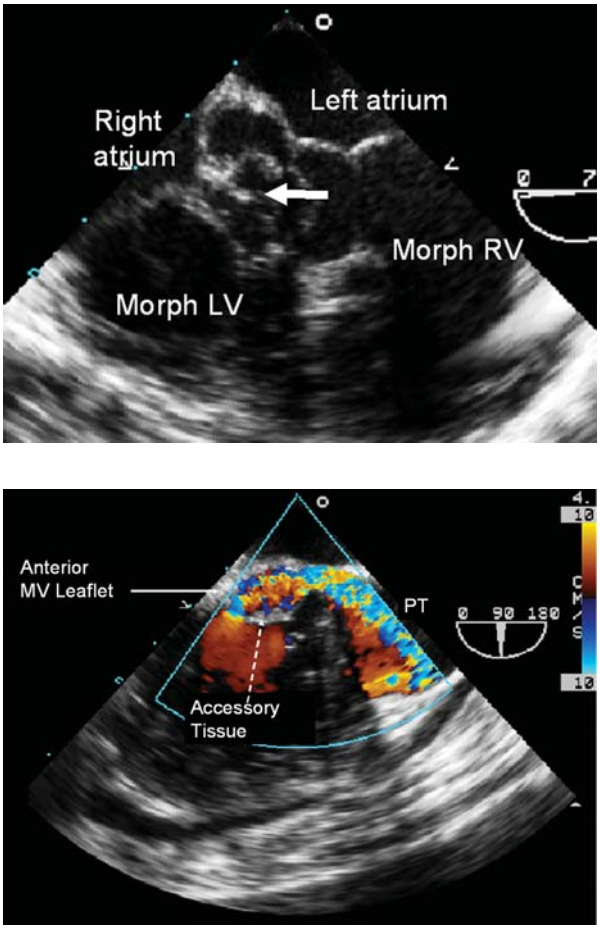


**Figure 8.**
*This transoesophageal transverse image (upper panel) demonstrates accessory tissue arising from the membranous septum that obstructs the subpulmonary outflow tract in this patient with congenitally corrected transposition. The Doppler colour interrogation confirms the obstructive nature of the lesions (lower panel).*

defect can be managed with complete surgical repair, or by banding the pulmonary trunk in those institutions adopting the strategy of combined atrial and arterial switch procedures. Conversely, patients with restrictive defects of moderate size may not require any intervention in infancy, regardless of the surgical strategy.

The greatest concern in those patients having a ventricular septal defect is the co-existing presence of obstruction within the outflow tract of the morphologically left ventricle. Such obstruction can exist in isolation, but typically is seen along with deficient ventricular septation. The mechanisms of obstruction include fibrous tissue originating from the membranous septum (Fig. 8). Alternatively, the muscular outlet septum can be deviated so as to impinge on the left ventricular outflow tract. It is usually possible to identify the mechanism of sub-pulmonary stenosis using parasternal long axis, as well as a sub-costal sagittal, planes of imaging. Transoesophageal imaging, along with Doppler interrogation, is particularly helpful (Fig. 8).

It is also possible that the obstruction can be caused by abnormalities of the pulmonary valve, such as annular hypoplasia, or thickening of the valvar leaflets. When present, obstruction of the pulmonary outflow tract can complicate the strategy of combined atrial and arterial switch procedures, as such patients are at risk for significant sub-arterial or arterial obstruction post-operatively. Obstruction of the sub-pulmonary outflow tract, nonetheless, can protect the pulmonary arterial capillary bed. Doppler interrogation permits a reasonable estimate to be made of pulmonary arterial pressures. In many children, an understanding of this process allows the paediatric cardiologist to recommend deferring surgical intervention to an older age.

According to autopsy studies, around nine-tenths of all patients with discordant atrioventricular connections have significant structural anomalies of the morphologically tricuspid valve.[10] The most common anomaly is Ebstein's malformation. A detailed description of Ebstein's malformation as it involves the normally positioned morphologically tricuspid valve is reported elsewhere in this supplement. Briefly, the essence of the lesion is that the effective valvar orifice is formed within the cavity of the morphologically right ventricle, at the junction of the atrialised inlet and functional ventricular components. The severity of the lesion varies considerably. In the setting of discordant atrioventricular connections, it should be appreciated that significant regurgitation of the morphologically tricuspid valve can occur with minimal, or even in the absence of, structural abnormalities of the valve or its tension apparatus.[13] In those patients in whom the pressure within the morphologically right ventricle is high relative to that in the left ventricle, as might be seen when the ventricular septum is intact, there can be significant tricuspid regurgitation. This regurgitation is due to the bowing of the ventricular septum away from the tricuspid valve, which in turn distorts the normal relationship between the septal leaflet of the tricuspid valve and the supporting tension apparatus (Fig. 9).

Recognition of this process is particularly important, since surgical measures to increase the pressure in the morphologically left ventricle, such as banding of the pulmonary trunk, may result in improvement of complete resolution of the tricuspid valvar regurgitation. Echocardiography readily permits identification of non-coaptation of the leaflets of the morphologically tricuspid valve in the absence of associated structural anomalies.

The origin of the coronary arteries in discordant atrioventricular connections can be reliably demonstrated by echocardiography, the parasternal short-axis plane being particularly valuable for this task.

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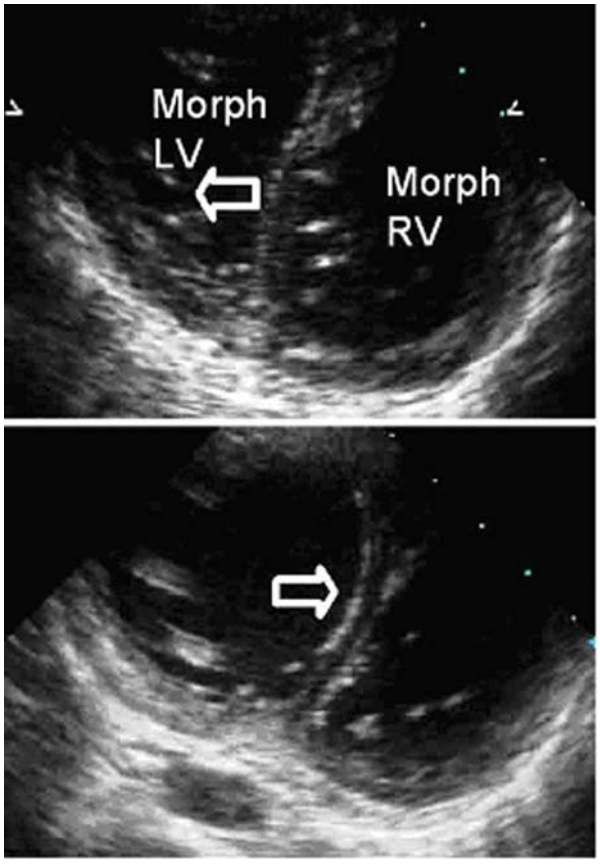


**Figure 9.**
*The parasternal short axis image in the upper panel, from a patient with discordant atrioventricular connections and intact ventricular septal structures, shows the bowing of the ventricular septum towards the morphologically left ventricle (arrow). Subsequent to banding of the pulmonary trunk, the bowing of the septum was reversed (lower panel), concomitant with "training" of the morphologically left ventricle in anticipation of a double switch procedure.*

Historically, abnormalities of coronary arterial origins were most of academic interest. With the increased use of the double-switch approach for surgical repair, delineation of these features has become of utmost clinical importance. In particular, the identification of an intramural course of a coronary artery may have great bearing on the surgical management.

One of the challenges for the echocardiographer when dealing with patients with discordant atrioventricular connections is characterization of the right ventricular volume and contractility. Many studies have shown that, in such patients, impaired function of the systemic morphologically right ventricle is common, and increases with time. Conventional M-mode techniques are poorly suited to assess the volume or contractility of the morphologically right ventricle. A number of Doppler techniques, therefore, have been found to be helpful in assessing right ventricular health. More exciting have been the advances in magnetic resonance imaging and three-dimensional echocardiography to characterize the morphologically right ventricle. In addition, with the recent enthusiasm for the double switch approach for surgical correction, the echocardiographer has come to play an increasingly important role in determining the appropriate timing of surgery.[14] As already discussed, in those patients with intact ventricular septal structures who have undergone banding of the pulmonary trunk, echocardiography can be particularly helpful in determining when the morphologically left ventricle is sufficiently "trained" to support the systemic circulation.

## Conclusions

Discordant atrioventricular connections describe the specific arrangement in which the atrial chambers are joined to morphologically inappropriate ventricles. In order to identify such discordant atrioventricular connections, it is necessary to define first the characteristics of the left and right atriums, and then the left and right ventricles. In the majority of patients with discordant atrioventricular connections, the ventriculo-arterial connections are also discordant, the double discordance then underscoring congenital correction of the circulations. Other ventriculo-arterial connections, however, must be anticipated to co-exist with discordant atrioventricular connections, and should be identified by the echocardiographer. It is also the duty of the echocardiographer to identify any, and all, of a number of important associated lesions that exist in the patients with discordant atrioventricular connections, concentrating in particular on the integrity of the ventricular septum, the subpulmonary outflow tract, and the structure of the morphologically tricuspid valve. Given the surgical strategies now available to treat the patient with discordant atrioventricular connections, the echocardiographer must incorporate the anatomic features and haemodynamic assessment so as to determine the most appropriate management.

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