Cardiol Young 2006; 16 (Suppl. 1): 35–42

ISSN 1047-9511
doi: 10.1017/S1047951105002301

Echo-morphological correlates concerning the functionally univentricular heart in the setting of isomeric atrial appendages

William T. Mahle,[1] Norman H. Silverman,[2] Gerald R. Marx,[3] Robert H. Anderson[4]

[1]Children's Healthcare of Atlanta and the Emory University School of Medicine, Atlanta, Georgia, [2]Lucille Packard Children's Hospital, Stanford University School of Medicine, Palo Alto, California, [3]Children's Hospital, Harvard University School of Medicine, Boston, Massachusetts, United States of America; [4]Cardiac Unit, Institute of Child Health, University College, London, United Kingdom

Keywords: Asplenia; polysplenia; visceral heterotaxy; double inlet ventricle

IT HAS LONG BEEN KNOWN THAT THE MOST COMPLEX combinations of cardiac malformations are those found in the setting of the so-called "splenic syndromes".[1] Many aspects of these syndromes have been controversial over recent years, not least the presence or absence of features of isomerism within the heart.[2,3] Recent experience with genetic manipulation of mice, nonetheless, has now shown that it is possible to generate unequivocal evidence of cardiac isomerism, particularly in those animals which show features of right isomerism when the genes responsible for morphologically leftness are knocked out.[4] Furthermore, when the crucial philosophical principle known as the "morphological method"[5] is applied to the hearts of patients known to have visceral heterotaxy, it is equally clear that patients falling within these groups, when judged on the extent of the pectinate muscles relative to the atrioventricular junctions, exhibit isomerism of either the morphologically right or left atrial appendages.[3] The morphological method[5] states that one variable feature in any structure should not be used as the criterion of another feature that is itself variable. Application of this principle to identification of the atrial chambers shows that venous connections, known to be markedly variable in the setting of visceral heterotaxy,[1,3,6] should not be used to arbitrate atrial arrangement, or "situs". Instead, those wishing to determine atrial arrangement should use the most constant feature of the atrial chambers to distinguish morphologically rightness from leftness, and ideally establish whether this chosen feature is independent of changes that might be induced by altered haemodynamics. Fortunately, the extent of the pectinate muscles within the atrial appendages as judged relative to the atrioventricular junctions has been shown to serve this purpose, and has also been shown to distinguish between morphologically right and left atrial appendages in the setting of visceral heterotaxy.[3] On this basis, it is possible to stratify patients with visceral heterotaxy into those with isomerism of either the morphologically right or left atrial appendages.[3] For the most part, these distinctions correlate with the presence or absence of the spleen, or multiple spleens, respectively. But this is not always the case.[7] In situations where there is disharmony between the arrangements of the organs of the various bodily systems, nonetheless, this does not mean that the arrangement is "ambiguous". Rather, it means that the observer should take the necessary time to describe the patterns in each system, pointing out the discrepancies, if any, between the arrangement of the atrial appendages within the heart and the patterns of formation of the thoracic and abdominal organs, particularly the spleen. When considering the heart in this fashion, it will then become apparent that patients with functionally univentricular hearts will be encountered most frequently in the setting of isomerism of the morphologically right atrial appendages, and that these patients potentially have a poor prognosis.[8] Patients with isomerism of the morphologically left appendages can also be found, less frequently, with functionally univentricular hearts, albeit that the finding of isomerism in this latter setting is less likely to

Correspondence to: William T. Mahle MD, Children's Healthcare of Atlanta, Emory University School of Medicine, 1405 Clifton Road, NE, Atlanta, GA 30322-1062, USA. Tel: +1 404 315 2672; Fax: +1 404 325 6021; E-mail: mahlew@kidsheart.com

impact on clinical management. In this review, we will discuss all the implications of the finding of patients having a functionally univentricular heart in the setting of isomeric atrial appendages.

Bodily arrangement

It is now well established that there are four basic patterns of arrangement of the organs. In the usual pattern, also known as "situs solitus" (Fig. 1: left-hand panel), then the muscles, nerves and vascular system of the trunks and limbs show evidence of symmetry, while the bodily organs are lateralized, with the lungs and bronchial tree showing obvious differences on the right and left sides, and within the abdomen the liver being predominantly right sided, while the stomach and spleen are left sided. Within the heart itself, the atrial appendage containing pectinate muscles that extend all round the atrioventricular junction is right sided, while the tubular appendage with pectinate muscles confined within it is left sided. In a very small proportion of the population, all of these features, including the arrangement of the atrial appendages, are mirror imaged, an arrangement usually called "situs inversus", although the essence of the difference is mirror imagery rather than "upside-downness" (Fig. 1: right-hand panel). In a significantly larger proportion of the population when compared to those having mirror imagery, but small in comparison to those having the usual arrangement, there is symmetry not only of the parietal structures, but also the lungs and bronchial tree, and the atrial appendages. In these settings, the abdominal organs are jumbled up, producing the situation known as visceral heterotaxy (Fig. 2). When analysed in terms of the thoracic organs or atrial appendages, the morphologic patterns can be distinguished as showing evidence of right isomerism (Fig. 2: left-hand panel) or left isomerism (Fig. 2: right-hand panel). Diagnosis of the bodily arrangement, and the arrangement of the atrial appendages, nonetheless, gives no information concerning the venoatrial connections, nor the arrangement of the rest of the heart. It is the task of the diagnostician seeking to define the cardiac malformation in the setting of visceral heterotaxy, therefore, first to establish the presence of right as opposed to left isomerism, and then to demonstrate the abnormalities present at the atrioventricular and ventriculoarterial junctions, as well as determining the structure of the ventricular mass.

Anatomic distinction of right versus left isomerism within the heart

As emphasized, it is only the appendages within the heart that show the anatomic features of isomerism. Thus, in the patient with right isomerism, the pectinate muscles extend round both atrioventricular junctions, meeting at the crux of the heart. In very many patients showing such isomerism of the right atrial appendages, there is a common atrioventricular junction rather than separate right and left junctions, and the extent of the pectinate muscles is then clear cut in autopsied specimens (Fig. 3). The situation is less obvious for the clinician, although as we will discuss, knowledge of the morphologic patterns

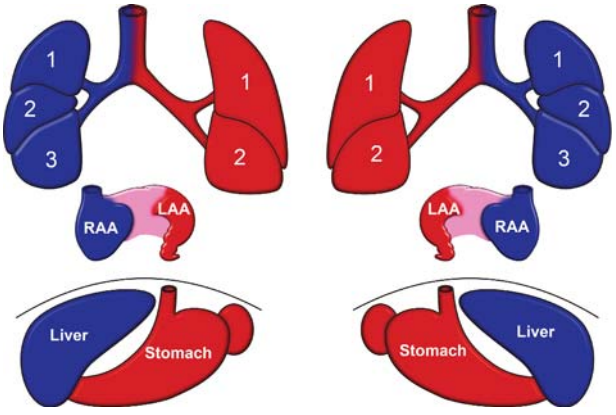

Figure 1.
The Figures show the usual (left-hand panel) and mirror-imaged (right-hand panel) patterns of arrangement of the thoracic and abdominal organs. As can be seen, there are morphologically different features on the right and left sides of the body. The features characteristic of the morphologically right side are coloured blue, while those characteristic for the morphologically left side are coloured red. Within the heart, it is the appendages that are different. RAA, LAA: right and left atrial appendages.

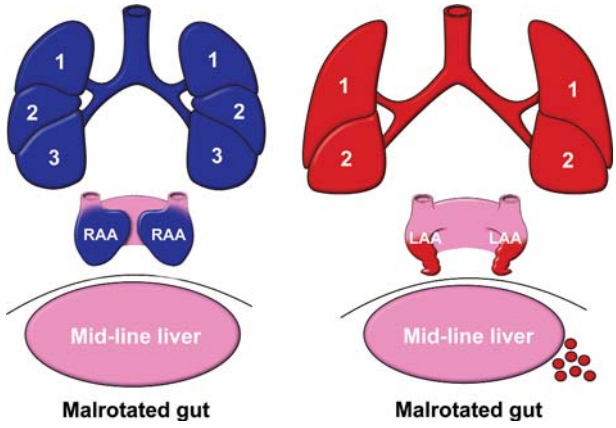

Figure 2.
These Figures show the isomeric bodily arrangements using the same colour codes as for Figure 1. Right isomerism (left-hand panel) is usually, but not always, associated with absence of the spleen, while left isomerism (right-hand panel) is typically found with multiple spleens, albeit that exceptions can occur. In these cases with disharmony between the systems of organs, it is necessary to describe each system separately. RAA, LAA: right and left atrial appendages.

https://doi.org/10.1017/S1047951105002301 Published online by Cambridge University Press

should now make it easier for the echocardiographer to diagnose as right or left isomerism. An important clue to the presence of right isomerism, nonetheless, is found in the universal absence of the coronary sinus. Failure to find a venous channel within the left-sided atrioventricular junction, therefore, is strong evidence for the existence of either isomerism or mirror-imaged arrangement, with right isomerism being the most likely diagnosis. When there is left isomerism, then both of the appendages are typically narrow and tube-like, having a narrow junction with the bodies of the right- and left-sided atriums. This feature, however, is less reliable than finding the pectinate muscles confined within the tubular appendages, both atrial vestibules being smooth when traced to the cardiac crux (Fig. 4). The finding of smooth vestibules on both sides has yet to be proven as being of diagnostic value to the echocardiographer, a possibility we will discuss below.

The functionally univentricular heart in the setting of isomeric atrial appendages

As pointed out recently by Freedom et al.,[8] the combination of a functionally univentricular heart in the setting of isomeric right atrial appendages continues to constitute one of the most egregious forms of congenital cardiac disease. Indeed, functionally univentricular hearts are much more frequent in the setting of right as opposed to left isomerism, and serve as one of the distinguishing features of the two types of isomerism. Any form of functionally univentricular heart can be found with right isomerism, but double inlet ventricle is by far the commonest atrioventricular connection, and typically through a common atrioventricular valve (Fig. 5). The dominant ventricle can be of left, right or indeterminate morphology. When considering the overall number of patients with right isomerism having double inlet ventricle, then the majority will prove to have a dominant left ventricle. When considering the overall cohorts of patients with double inlet right ventricle, or double inlet to a solitary and indeterminate ventricle, in contrast, a significant proportion will be found to have isomeric atrial appendages, typically with the double inlet atrioventricular connection guarded by a common atrioventricular valve. When the common valve opens to a dominant right ventricle, then the left ventricle is incomplete and hypoplastic. It can be difficult, nonetheless, to make the distinction between biventricular atrioventricular connections through a common

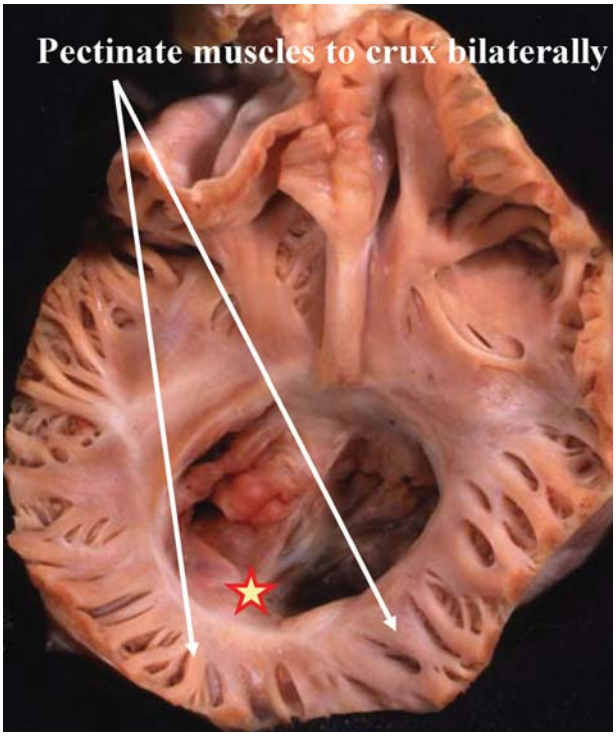

Figure 3.
This photograph shows isomerism of the right atrial appendages in the setting of a heart with a common atrioventricular junction. The pectinate muscles extend to the crux of the heart (star) on both sides.

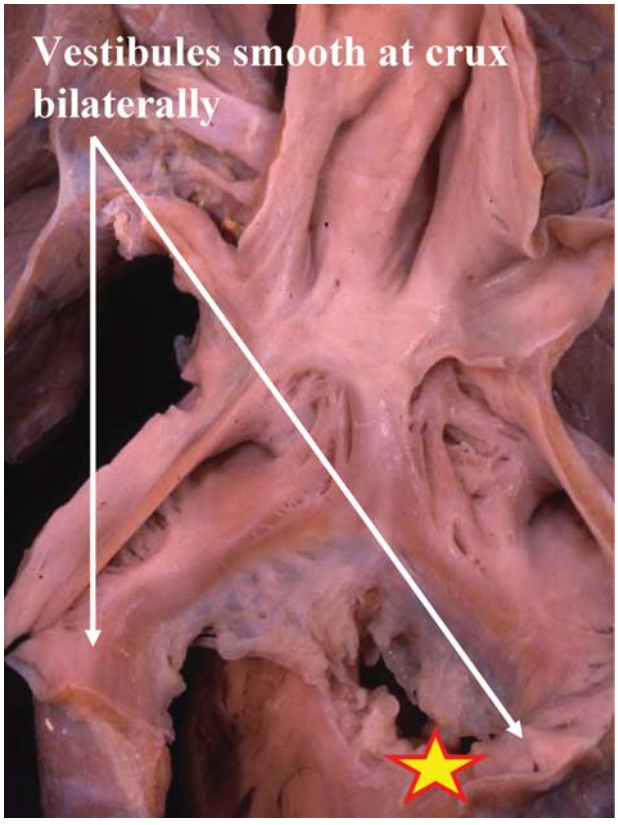

Figure 4.
In this specimen, again with a common atrioventricular junction, it is the morphologically left appendages that are arranged in isomeric fashion. Both vestibules are smooth at the crux, albeit that there is some spillage of the pectinate muscles out of the mouths of the appendages.

https://doi.org/10.1017/S1047951105002301 Published online by Cambridge University Press

valve and double inlet. It is the task of the echocardiographer in this setting, therefore, to determine the proportion of the common junction committed to the dominant as opposed to the incomplete ventricle, and to correlate this with the size of the hypoplastic left ventricle. It is also the task of the echocardiographer to determine the competency of the common valve, and to establish the precise venoatrial connections, and the arrangements at the ventriculo-arterial junctions. When both appendages are of right morphology, then the pulmonary venous connections must be anatomically abnormal even if they return to the atrial chambers. In half of patients with isomeric right appendages, the pulmonary veins will drain to an extracardiac site. The other half are just as important, since the pulmonary veins typically join together in a narrow midline confluence that drains to the atrial roof (Fig. 5). The pulmonary veins can drain anomalously to one or other atrial chamber with a morphologically right appendage, but this is unusual. Also important is to note the universal absence of the coronary sinus. Due to this, the coronary veins drain directly into the atrial chambers, often crossing the atrioventricular junctions that are extending for some distance within the atrial musculature before terminating, frequently adjacent to a venous orifice. These features should also now be potentially recognizable by the echocardiographer. It may be difficult for the echocardiographer to diagnose double inlet to a solitary and indeterminate ventricle. This remains a diagnosis of exclusion for the clinician, being made when it is impossible in life to find the incomplete second ventricular chamber. At the ventriculo-arterial junctions, the connections are typically discordant or double outlet from the right or solitary ventricle. Most typically, each arterial valve is supported by its own infundibulum. In the setting of right isomerism, the echocardiographer should anticipate subpulmonary obstruction, or more likely pulmonary atresia. It is also possible, nonetheless, to find subaortic obstruction or atresia.

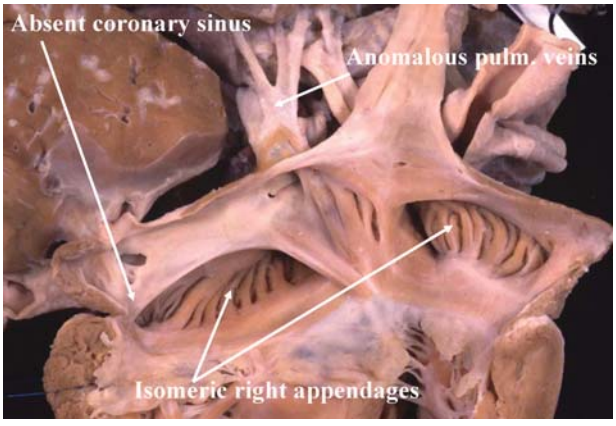

Figure 5.
In this specimen, with double inlet through a common valve to a dominant left ventricle, the isomerism of the right atrial appendages is readily recognized. The pulmonary veins drain together through a midline confluence. Note the absence of a coronary sinus.

Obstruction in the aortic pathways is more likely to be associated with isomeric left rather than right atrial appendages. In the setting of the functionally univentricular heart, this combination will typically be found with hypoplasia of the left ventricle, and with mitral atresia or stenosis with aortic atresia. When encountered, the presence of the isomeric atrial appendages is unlikely to create additional difficulties, but the investigator needs to be aware of the likely co-existence of interruption of the inferior caval vein, with continuation of the venous pathways from the abdomen through the azygos system of veins. When both appendages are of left morphology, there is also an abnormal disposition of the sinus node, with obvious correlations for interpretation of the electrocardiogram. Any form of functionally univentricular heart, nonetheless, must be anticipated to co-exist in the patient with isomeric left atrial appendages, so the key to diagnosis is recognition of the presence of isomerism, and full sequential analysis of the atrioventricular and ventriculo-arterial junctions.

In the setting of isomerism, the investigator should always remember the importance of splenic morphology, and take note of the fact that, while usually concordant with the arrangement of the atrial appendages, this is not always the case.[7]

Echocardiographic interrogation of the patient with isomeric atrial appendages and a functionally univentricular heart

As the surgical management of the malformations found in patients with heterotaxy has improved, so it has become necessary to establish accurately the atrial arrangement of each individual patient. To cite but one example, the association of abnormalities of atrioventricular conduction with isomerism of the left atrial appendages makes accurate identification valuable to the clinician. It is still the case, nonetheless, that it is difficult for the echocardiographer precisely to identify the nature of the appendages, so still it is inferential findings that give the clue to the likelihood of an isomeric arrangement.

Abdominal arrangement and the systemic veins

The echocardiographer begins the interrogation by displaying the abdominal great vessels in a transverse projection at the level of the tenth thoracic vertebra. The transducer can then be tracked cephalad to trace the inferior caval vein and hepatic veins until

https://doi.org/10.1017/S1047951105002301 Published online by Cambridge University Press

they connect with the heart. Though an alternative approach might include a sagittal view of the abdomen, such an approach may fail to identify some details, such as the inferior caval vein crossing the midline. In addition, defining the position of the aorta relative to the midline of the spine is often simpler using transverse scans. The inferior caval vein lies within the liver, and can sit either to the right or the left of the spine. Should the inferior caval vein be interrupted, as is frequently the case with left isomerism, then no structure will be found at the anticipated location. Instead, an azygos vein will be found posterior to both the liver and the aorta (Fig. 6). Scanning sagitally and parasternally will then demonstrate the drainage of the azygos vein, which is to one or other of the superior caval veins before reaching the atriums.

The hepatic venous connections are best identified using both transverse and parasagittal views. Complete identification of the drainage of the hepatic veins is crucial when a patient has a functionally univentricular heart in the setting of isomerism, since the hepatic veins must be incorporated into any proposed total cavopulmonary connection. Failure to include all hepatic veins in the systemic venous system can result in significant right-to-left shunting after surgery.

The atriums and their appendages

Although still difficult, using modern day cross-sectional echocardiography, it should be possible to display reliably the morphology of the atrial appendages, based upon their distinctive shape and the nature of their junctions with the rest of the atrial chamber.[9] An atrial appendage is morphologically right if it has a broad junction with the rest of the atrial chamber. An appendage is morphologically left if it displays a narrow junction with the rest of the atrial chamber, and if the orifice has the same or smaller width as the proximal portion of the appendage. Obviously the echocardiographer will need to identify two broad-based appendages so as to diagnose right isomerism, and two tubular and narrow appendages to make the diagnosis of left isomerism, although there is always the danger that mistakes will be made if it is not possible, with certainty, to identify the extent of the pectinate muscles. Both transverse and sagittal image planes can help identify these features, while interrogation from the suprasternal transverse plane also permits differentiation of the appendages. As discussed above, in the past the relationship of the aorta and the inferior caval vein and the hepatic veins as seen in subcostal planes was used so as to make the diagnosis of isomerism by inference.[10] Unfortunately, such algorithms as proposed by Huhta et al.,[10] do not always agree with direct assessment of the morphology of the appendages. Nowadays, we have also recognized that identification of the coronary sinus in subcostal transverse images is an invaluable guide to the presence of isomerism. The coronary sinus is always absent in patients with right isomerism, and in many patients with left isomerism. So, if this structure is not found in the left-sided atrioventricular junction, this is strong evidence for the presence of either an isomeric arrangement, or of mirror-imaged arrangement.

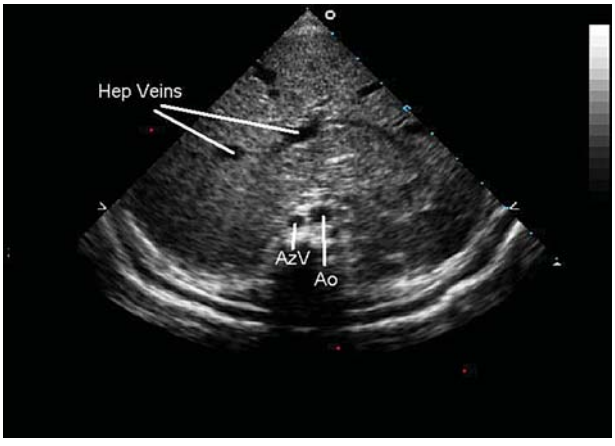

Figure 6.
This subcostal transverse scan demonstrates the typical situation seen with interruption of the inferior caval vein in the setting of isomerism of the left atrial appendages. The dilated azygos vein (AzV), located posterior to the aorta (Ao) and adjacent with the veterbras, carries the venous return from the abdomen to the heart. Hepatic veins (Hep veins) are also shown.

Pulmonary veins

Given that, in patients with right isomerism, the pulmonary veins frequently drain to an extracardiac site, or to a narrow confluence in the midline that drains to the atrial roof, sufficient attention must be given fully to define the pulmonary venous anatomy. Subcostal transverse scanning, or interrogation from the apical window, are usually satisfactory when seeking to identify the midline confluence between the pulmonary veins and the atrial roof that is characteristic of right isomerism (Fig. 7). To define totally anomalous subdiaphragmatic drainage of pulmonary veins, the echocardiographer should begin by displaying the abdominal great vessels in a transverse view. It is often possible to visualize a descending vertical vein in cross-section in this fashion. The aorta is the circular structure adjacent to the vertebral column. If there is an enlarged azygos vein, this structure is also typically adjacent to the vertebral column. Descending vertical veins are usually circular structures. The descending vertical vein can be traced to its termination, almost always a junction with the portal venous system. Often the easiest way to identify an ascending vertical vein

https://doi.org/10.1017/S1047951105002301 Published online by Cambridge University Press

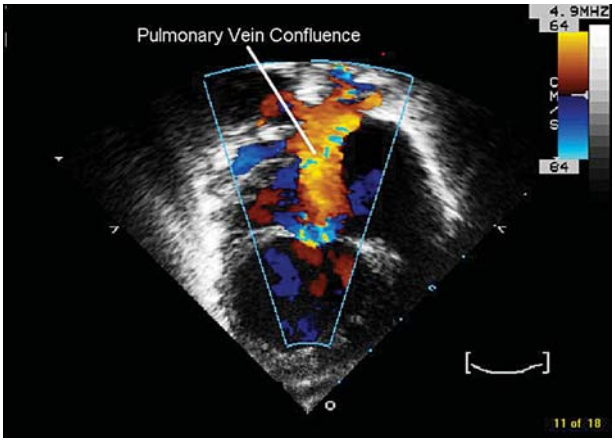

Figure 7.
This apical image demonstrates the midline confluence of the pulmonary veins that drains to the roof of the atrium in a patient with right isomerism.

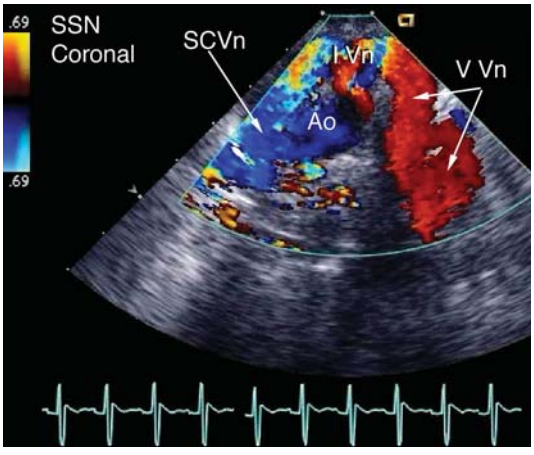

Figure 8.
This image obtained from the suprasternal notch in a patient with right isomerism shows the presence of a vertical vein (VVn), draining superiorly (red flow) into the brachiocephalic vein (I Vn), and the blue flow in the superior caval vein (SCVn), as these vessels course over the transverse aorta (Ao).

is to survey the entire superior aspect of the pulmonary venous confluence between the pulmonary hilums. The ascending vertical vein should be tracked until it connects to the systemic venous system (Fig. 8). A sagittal view can also be helpful, particularly if there is a short connecting vein between the confluence and the right superior caval vein. It is the mixed type of totally anomalous pulmonary venous connection that is most difficult to identify, owing to the absence of an easily discernible confluence. In order to avoid overlooking this variant, it is essential that the echocardiographer identifies all four pulmonary veins.

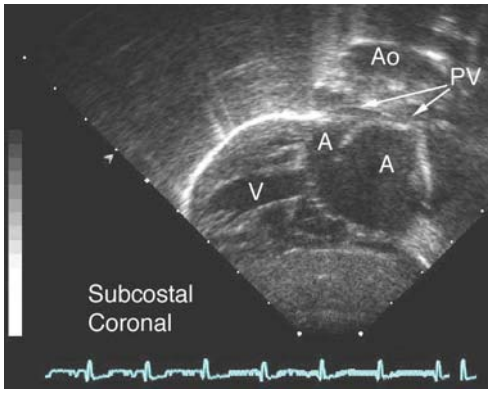

Figure 9.
In this subcostal transverse image from a patient with right isomerism and a right-sided heart, the two atriums (A) are separated by a remnant of atrial septal strand. The area below this is part of the atrioventricular septal defect. A common atrioventricular valve, guarding a common atrioventricular junction, is connected mostly to the larger left ventricle (V), but in part to the inferior incomplete right ventricle. The pulmonary venous confluence (PV) is seen immediately above the roof of the atrium. Ao: aorta.

Atrioventricular connections

Though a variety of atrioventricular connections can be identified in patients with isomeric atrial appendages, it is double inlet through a common valve that is found most frequently in the setting of the functionally univentricular heart. When seen in this setting, the left ventricle is usually dominant, although a dominant right ventricle is not uncommon. The relationship of the common atrioventricular valve to the respective ventricles, and delineation of the subvalvar apparatus, is best achieved in subcostal transverse and sagittal planes (Fig. 9). From the standpoint of echocardiographic imaging, it is important to determine if the heart is indeed functionally univentricular. While considerable attention has been given to specific measures of left ventricular hypoplasia in normal atrioventricular connections, much less has been reported regarding assessment of ventricular adequacy in the setting of a common atrioventricular junction. In examining subjects with small left ventricles and common atrioventricular junction, Cohen et al.,[11] assessed the commitment of the common valve to the respective ventricles from a subcostal long-axial oblique plane.[11] They suggested that, when less than one-third of the common valvar orifice was committed to the left ventricle, the left ventricle would not support systemic circulation, and hence the heart would be functionally univentricular. Earlier studies had suggested that presence of a single papillary muscle precluded the use of the left ventricle as the systemic ventricle.[9] Subsequent studies have shown that the architecture of the papillary muscles

https://doi.org/10.1017/S1047951105002301 Published online by Cambridge University Press

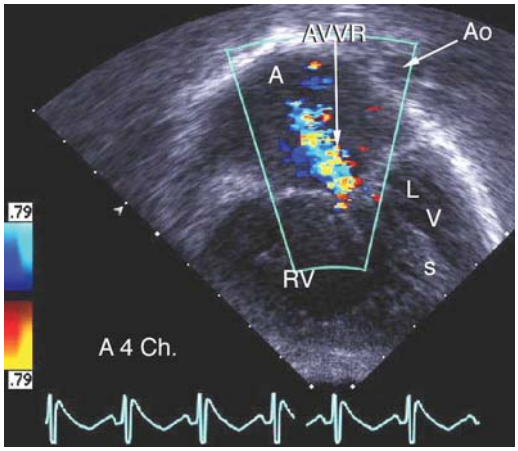

Figure 10.
This patient with isomerism of the right atrial appendages and a common atrioventricular valve has gross regurgitation across the valve (AVVR). The colour echocardiogram in systole in the same patient shows the moderate central regurgitation. A: atrium; LV: left ventricle; RV: right ventricle; S: septum; Ao: aorta; A4Ch: apical four chamber.

is an imperfect measure of left ventricular hypoplasia.[11,12] More recent data suggest that an effective commitment of less than one-third of the common valve to the right ventricle would similarly characterize a functionally univentricular heart.[13] Less commonly, it is possible to find double inlet through two valves to the dominant ventricle, almost always of left ventricular morphology. In this variant, subcostal and apical imaging defines the size and relationship of the two atrioventricular valves. Insufficiency of atrioventricular valves is best assessed using colour Doppler in apical or parasternal long-axis imaging planes (Fig. 10).

Ventriculo-arterial connections

These are markedly variable, though double outlet from the right ventricle is most commonly seen (Fig. 11). Discordant ventriculo-arterial pattern is also prevalent. The subcostal sagittal scan is usually ideal for defining the connections and relationships of the great arterial trunks, as well as the presence of obstruction or stenosis in the outflow tracts. In those with double outlet right ventricle, it is frequent to find bilateral infundibulums, although this should no longer be considered a defining feature of the connection. Whenever there is subaortic obstruction, however, the echocardiographer should also look for the presence of aortic coarctation. Careful delineation of the size of the subaortic area, and the diameter of the aortic valve, as well as the excursion of the leaflets, are important. While some patients can be managed by repair of the coarctation and limitation of flow of blood to the lungs by banding the pulmonary trunk,

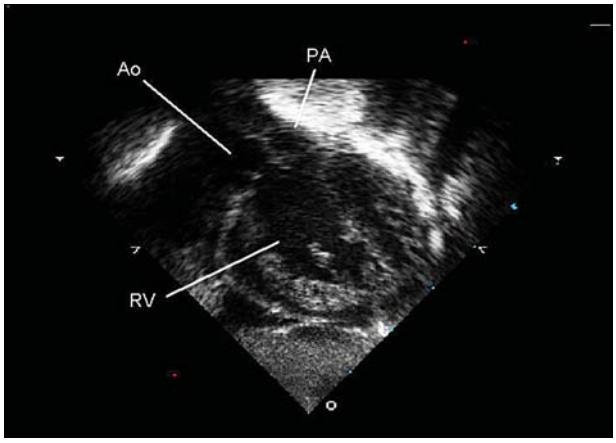

Figure 11.
This subcostal transverse scan demonstrates double outlet right ventricle (RV) with side-by-side great vessels, the aorta (Ao) being to the right of the pulmonary trunk (PA). There is mild subpulmonary stenosis.

those with more severe aortic or subaortic obstruction may require a Damus–Kaye–Stansel procedure to ensure adequate systemic blood flow. Finding significant reversal of flow subsequent to colour or pulse Doppler interrogation of the transverse aortic arch imaged from a suprasternal plane would support the latter approach. Pulmonary stenosis or atresia can also be visualized in various planes. Subcostal sagittal imaging is usually ideal to delineate the subpulmonary region, whereas parasternal long- and short-axis planes provide better visualization of the pulmonary valve itself, as well as the right and left pulmonary arteries.

Conclusion

In patients with isomeric atrial appendages, a detailed segmental approach to non-invasive imaging is of great importance. Though common associations occur within the spectrum of right and left isomerism, a detailed approach, with particular attention to morphology of the atrial appendages, is needed fully to describe and treat in appropriate fashion the patients with these complex cardiac lesions.

Acknowledgement

Professor Anderson is supported by grants from the British Heart Foundation together with the Joseph Levy Foundation. Research at the Institute of Child Health and Great Ormond Street Hospital for Children NHS Trust benefits from R&D funding received from the NHS Executive.

References

1. Ivemark BI. Implications of agenesis of the spleen on the pathogenesis of conotruncus anomalies in childhood; an analysis of the

heart malformations in the splenic agenesis syndrome, with fourteen new cases. Acta Paediatr 1955; 44: 7–110.
2. Van Praagh R, Van Praagh S. Atrial isomerism in the heterotaxy syndromes with asplenia, or polysplenia, or normally formed spleen: an erroneous concept. Am J Cardiol 1990; 66: 1504–1506.
3. Uemura H, Ho SY, Devine WA, Kilpatrick LL, Anderson RH. Atrial appendages and venoatrial connections in hearts from patients with visceral heterotaxy. Ann Thorac Surg 1995; 60: 561–569.
4. Bamforth SD, Braganca J, Farthing CR, et al. Cited2 controls left-right patterning and heart development through a Nodal-Pitx2c pathway. Nat Genet 2004; 36: 1189–1196.
5. Van Praagh R, Leidenfrost RD, Lee SK, Marx G, Wright GB, Van Praagh S. The morphologic method applied to the problem of "single" right ventricle. Am J Cardiol 1982; 50: 929–932.
6. Moller JH, Nakib A, Anderson RC, Edwards JE. Congenital cardiac disease associated with polysplenia. A developmental complex of bilateral "left-sidedness". Circulation 1967; 36: 789–799.
7. Uemura H, Ho SY, Devine WA, Anderson RH. Analysis of visceral heterotaxy according to splenic status, appendage morphology, or both. Am J Cardiol 1995; 76: 846–849.
8. Freedom RM. The asplenia syndrome: a review of significant extracardiac structural abnormalities in 29 necropsied patients. J Pediatr 1972; 81: 1130–1133.
9. Chin AJ, Fogel MA. Noninvasive Imaging of Congenital Heart Disease: Before and After Surgical Reconstruction. Futura, Armonk, NY, 1994.
10. Huhta JC, Smallhorn JF, Macartney FJ. Two dimensional echocardiographic diagnosis of situs. Br Heart J 1982; 48: 97–108.
11. Cohen MS, Jacobs ML, Weinberg PM, Rychik J. Morphometric analysis of unbalanced common atrioventricular canal using two-dimensional echocardiography. J Am Coll Cardiol 1996; 28: 1017–1023.
12. van Son JA, Phoon CK, Silverman NH, Haas GS. Predicting feasibility of biventricular repair of right-dominant unbalanced atrioventricular canal. Ann Thorac Surg 1997; 63: 1657–1663.
13. De Oliveira NC, Sittiwangkul R, McCrindle BW, et al. Biventricular repair in children with atrioventricular septal defects and a small right ventricle: anatomic and surgical considerations. J Thorac Cardiovasc Surg 2005; 130: 250–257.

https://doi.org/10.1017/S1047951105002301 Published online by Cambridge University Press