

REVIEW

EDUCATIONAL SERIES IN CONGENITAL HEART DISEASE: The sequential segmental approach to assessment

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Abstract

Sequential segmental analysis allows clear description of the cardiac structure in a logical fashion without assumptions and confusing nomenclature. Each segment is analysed, and then the connections described followed by any associated anomalies. For the echocardiographer there are several key features of the cardiac structures to help differentiate and accurately describe them.

Key Words

- ▶ echocardiography
- ▶ congenital heart defects

Introduction

Echocardiography has an essential role in the assessment of cardiac morphology in children and adults with suspected or confirmed congenital heart disease. Whilst knowledge and understanding of cardiac morphology is a fundamental part of training for paediatric cardiologists and specialist congenital heart echocardiographers, it is less familiar territory for adult cardiologists and echocardiographers. Nevertheless, with the majority of children with congenital heart disease surviving into adulthood, an understanding of these conditions is increasingly important, and highly rewarding, for the adult cardiology department – patients with these conditions may present to their local hospital under a variety of different circumstances. The *de novo* diagnosis of complex cardiac morphological abnormalities in adults is uncommon; however, adults with known congenital heart disease increasingly present to general cardiologists – as an initial presentation, for geographical or logistic reasons, during acute/concurrent illnesses or loss of regular follow-up. Knowledge of cardiac morphology in patients is frequently incomplete or incorrect, even those who have undergone detailed investigation or treatment in childhood. The knowledge and ability to perform a

‘congenital heart echocardiogram’, with a link to a local or regional expert team if appropriate, is invaluable in this situation.

For adult echocardiographers, the systematic approach to performing a detailed study is well described (BSE minimum dataset). However, this approach is designed to detect acquired disease in (the vast majority of) patients who have a normal arrangement of cardiac structures. It is not well suited to the approach required in congenital heart disease where there is a spectrum of disease affecting the location and arrangement (connections) of cardiac chambers and valves. Undertaking echocardiography in patients with new, undocumented, untreated or surgically palliated congenital heart disease requires an approach that allows for the assessment of cardiac structure in a systematic manner, without any assumptions, e.g:

- is the right atrium on the patient's right side?
- is there a right ventricle?
- does the right atrium connect to the right ventricle?
- does the right ventricle connect to the pulmonary artery or the aorta, or both, or neither?

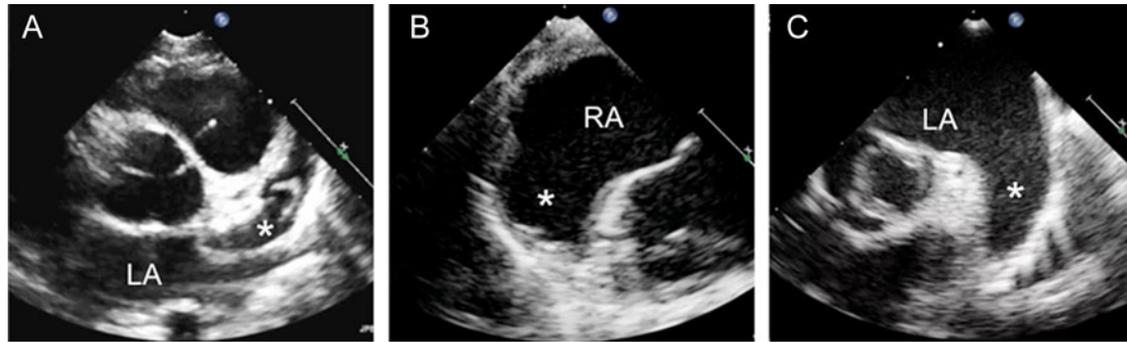


Figure 1

Atrial appendages. (A) Parasternal short axis in an older child of the left atrial appendage. (B) Transoesophageal view of right atrial appendage: short and stubby with pectinate muscles. (C) Transoesophageal view of left atrial appendage: long and thin. LA, left atrium; RA, right atrium.

The approach that we used to assess these questions in congenital heart disease is the sequential segmental analysis method.

The sequential segmental analysis method for describing cardiac morphology was developed by Anderson *et al.* in the early 1980s (1) as a clear way of describing the malformed heart. Instead of using eponymous names which did not tell the clinician anything ('Holmes Heart', 'Taussig-Bing'), this approach allows anyone to read the diagnosis and immediately envision the anatomy. The premise is describing the cardiac segments and their connections in a logical manner, and by identifying salient features of each segment. For the echocardiographer this creates a template for the examination and subsequent report.

Cardiac position

Cardiac position is defined by both where it is (left chest, midline, right chest) and also by where the apex points (apex to the right, apex to midline, apex to the left).

This avoids confusing terms such as 'dextrocardia', which could mean a heart pulled to the right such as in Scimitar syndrome (anomalous pulmonary venous return associated with hypoplasia of the right lung) or totally rotated on its axis (frequently referred to as 'situs inversus').

Atrial arrangement

The first step in sequential analysis is to identify the atrial arrangement. It is important to note that the atriums are not defined by which venous drainage they receive nor by the side of the body on which the atrium lies. For example, pulmonary veins can drain into the left atrium (normal) or right atrium (anomalous).

The gold standard for naming of the atriums, the morphology of the atrial appendages is challenging on echocardiography, especially in older children and adults or if there is juxtaposition of the atrial appendages, in which the atrial appendages lie alongside each other,

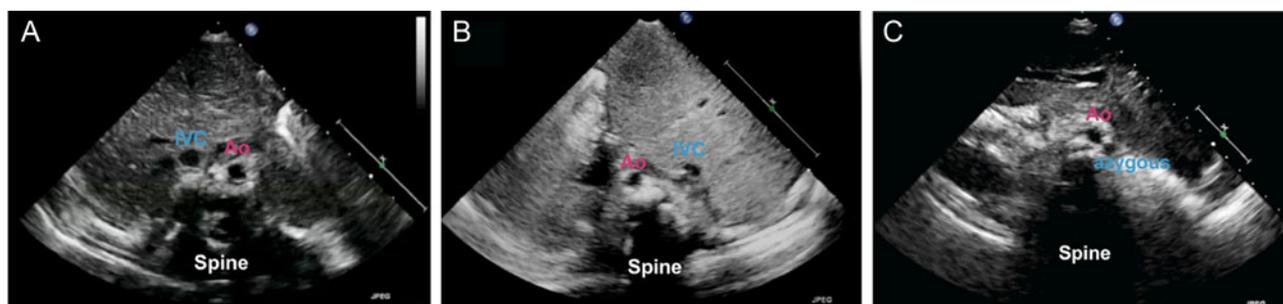


Figure 2

Atrial arrangement. (A) Normal atrial arrangement. (B) Mirror image atrial arrangement. (C) Laterality disturbance (left atrial isomerism). Ao, aorta; IVC, inferior caval vein.

Table 1 Features of laterality disturbances.

Right atrial isomerism	Left atrial isomerism
Bilateral right bronchi	Bilateral left bronchi
Midline liver	Midline liver
Asplenia	Polysplenia (often functionally asplenic)
Malrotation	Malrotation
Two sinoatrial nodes	Absent sinoatrial node (complete heart block can occur, but more usually an ectopic atrial rhythm)
By definition total anomalous pulmonary venous connection	Interrupted IVC with azygous continuation

to the left or to the right of the great arteries. The parasternal short axis view is useful in transthoracic echocardiography (in children and adults if there are good acoustic windows Fig. 1A), but if they cannot be seen then atrial appendages can be seen better on transoesophageal echocardiography. The right atrial appendage is short and stubby (Fig. 1B), whereas the left atrial appendage is long and thin (Fig. 1C).

In clinical practice, if visualisation of appendage morphology is not possible, identification of atrial arrangement is based on defining the relative position of the inferior caval vein (IVC) and the aorta, coupled with defining the drainage of the IVC to the right atrium and the pulmonary veins to the left atrium. Therefore, in paediatric echocardiography, the initial subcostal

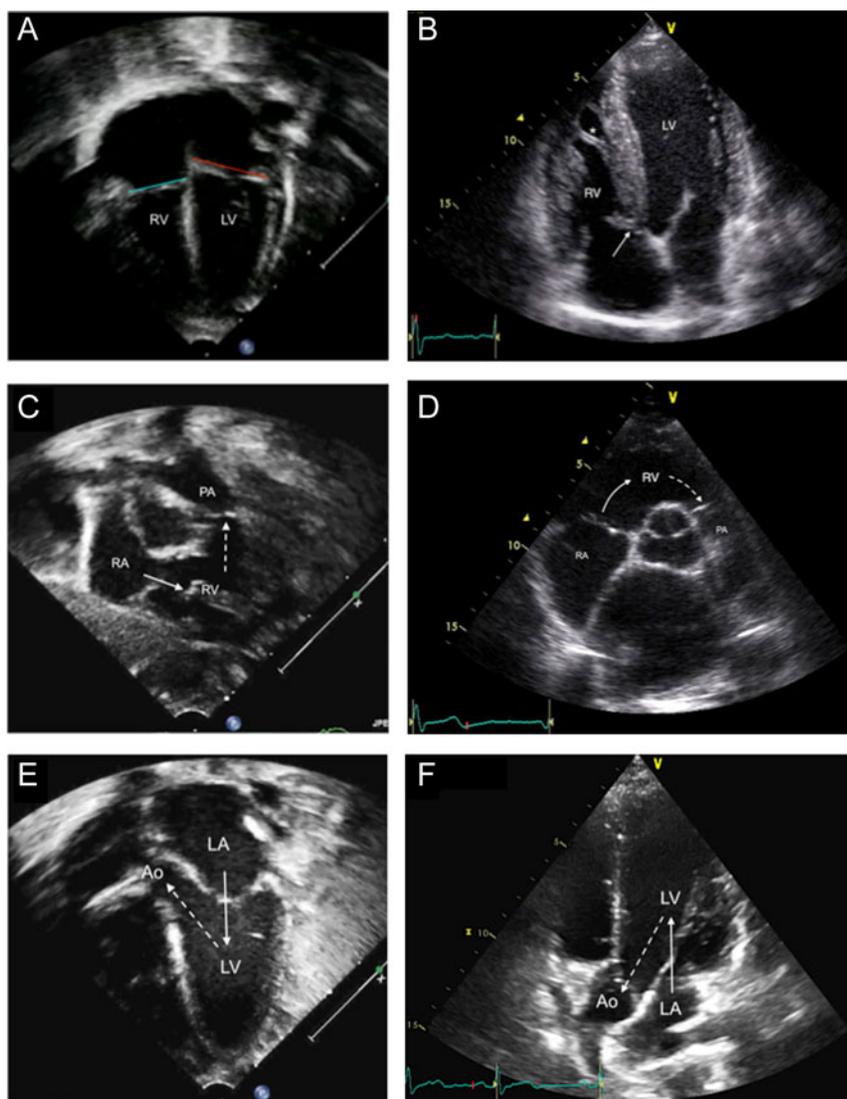


Figure 3

Ventricular morphology. (A) Normal off-setting of the atrioventricular valves (paediatric orientation) indicated by the red and blue lines. (B) Normal off-setting of the atrioventricular valves (adult orientation). The RV is more trabeculated than the LV and there is a distinct moderator band (*). The most reliable sign used to identify the right ventricle is the apical offset of insertion point of the right-sided atrioventricular valve (arrow) – this is therefore the tricuspid valve and right ventricle. (C) Right ventricle with its three portions in paediatric orientation and (D) adult orientation: inflow (solid arrow) and outflow (dashed arrow), separated by the body of the RV. (E) LV with steeper outflow angle and continuity between inflow and outflow in paediatric orientation and (F) adult orientation. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle.

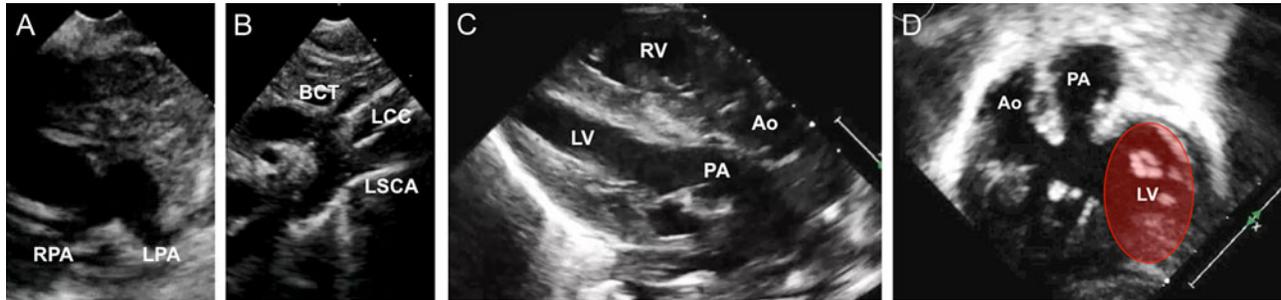


Figure 4

Great vessels. (A) Pulmonary artery bifurcation. (B) Aortic arch with head and neck vessels. (C) Parasternal long axis view in transposed great arteries showing parallel great vessels. (D) Subcostal view showing the aorta and pulmonary arteries both arising from the right ventricle. Ao, aorta; BCT, brachiocephalic trunk; LCC, left common carotid; LPA, left pulmonary artery; LSCA, left subclavian artery; LV, left ventricle; PA, pulmonary artery; RPA, right pulmonary artery; RV, right ventricle.

transverse image allows demonstration of the position of the aorta and IVC in relation to the spine. The aorta is pulsatile and round whereas the IVC is flatter and non-pulsatile.

In normal atrial arrangement, the aorta is to the left of the spine and the IVC anterior and to the right (Fig. 2A). In mirror image atrial arrangement this is reversed, with the aorta to the right of the spine and IVC anterior and to the left (Fig. 2B). As a rule, the IVC does not join the left atrium (this has been described but is exceedingly rare); therefore, unless it is 'interrupted' (see below), the IVC can be followed up towards the right atrium. In laterality disturbances (failure of the normal left–right arrangements of the heart and other organs), there can be a midline liver with abnormal venous drainage (Table 1). In left atrial isomerism an interrupted IVC with azygous continuation may be seen. This is seen as a vessel behind the aorta with no IVC seen (Fig. 2C).

Once identified, their position can also be defined, for example, the pulmonary veins drain to the right-sided morphological left atrium and the superior caval vein to the left-sided morphological left atrium. This allows the reader to understand position as well as morphology.

Identifying segments: the ventricles

After identifying the atriums, the ventricles must be assessed (Fig. 3A, B, C, D, E and F). The left ventricle tends to have a more elliptical shape and has a smooth septal surface, fine trabeculations and with no attachments of

the papillary muscles to the septum. The right ventricle has a more complex geometry with coarser trabeculations and valvar support apparatus that connects to the septum. A moderator band of muscle is seen in the right ventricle and it is described as having three sections: the inflow, the apex and the outflow. In the normal heart, the tricuspid valve inserts more towards the apex, well seen in the apical four-chamber view (Fig. 3A and B). If reverse off-setting of the valves is seen, there must be a suspicion of discordant atrioventricular connections (i.e. the right atrium connecting to the LEFT ventricle and the left atrium connecting to the RIGHT ventricle). As well as the off-setting, the valvar morphology itself can be of use. The bileaflet mitral valve is a feature of the left ventricle and the tricuspid valve a feature of the right.

The normal right ventricle has inlet, apical and outlet portions and so there is no continuity between the tricuspid valve and the outlet valve (Fig. 3C and D). The normal left ventricle has a more acute angle between the inflow and outflow and continuity between the mitral and outflow valve (Fig. 3E and F). These hold true in the structurally normal heart but are not of use in the congenitally abnormal heart and so are not generally used to define the ventricles.

In adults, apical off-setting of the tricuspid valve (TV) and identification of separate inlet, apical and outlet portions, are the most reliable methods of differentiating the RV from the LV as well as looking at the atrioventricular valve morphology. Prominent trabeculations and large muscle bundles within the LV are fairly common and chordal attachments can be difficult to identify; therefore, these are less reliable factors.

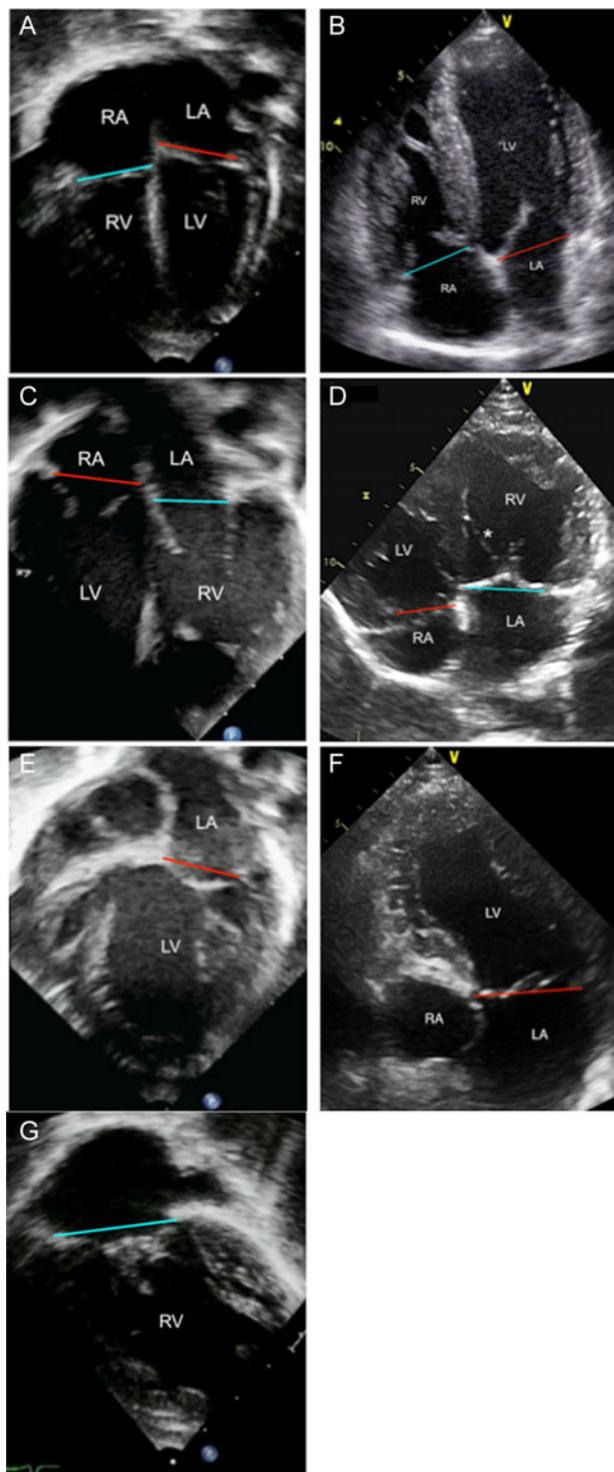


Figure 5
Atrioventricular connections. (A) Concordant atrioventricular connections, paediatric orientation. (B) Concordant atrioventricular connections, adult orientation. (C) Discordant atrioventricular connections in paediatric orientation and (D) adult orientation; note the direct chordal attachment to the septum (*) which helps identify the tricuspid valve (E and F) Absent right atrioventricular connection. (G) Absent left atrioventricular connection.

Identifying segments: the great vessels

The great vessels are usually the easiest to identify – the pulmonary artery bifurcates to supply each lung and, in a neonate, the arterial duct can be seen inserting into it (Fig. 4A). The aorta gives rise to the coronary arteries as well as the head and neck vessels (Fig. 4B). Sweeps are best used to demonstrate which vessels arise from which ventricles. In normal, concordant ventriculoarterial connections, the great vessels have a criss-cross arrangement. This is easily appreciated in the short axis views where the aorta is seen in cross section whilst the pulmonary artery is seen along its length. In discordant ventriculoarterial connections (i.e. various forms of ‘transposition of the great arteries’), the great vessels runs in parallel (a ‘shotgun’ arrangement); the parasternal long axis view shows parallel great vessels (Fig. 4C). In paediatric patients’ subcostal views can be useful in seeing two vessels arising from the right ventricle in double outlet right ventricle (Fig. 4D).

Types of atrioventricular connection

Once each segment has been defined, we can then assess the connections. Atrioventricular connections can be concordant (e.g. right atrium to right ventricle), discordant (e.g. right atrium to left ventricle) or absent (e.g. absent right atrioventricular connection). In some hearts, there may be a double inlet connection (e.g. both atriums open to a single ventricle) and very rarely the connection may be ambiguous (e.g. the ventricular morphology cannot be defined and so concordance/discordance cannot be ascertained) (Fig. 5).

Types of ventriculoarterial connection

Again, connections can be concordant (e.g. right ventricle to pulmonary artery), discordant (e.g. right ventricle to aorta, Fig. 4C) or absent (e.g. pulmonary atresia or aortic atresia). Double outlet connection describes both vessels arising from the same ventricle (e.g. double outlet right ventricle where both the aorta and pulmonary artery arise from the right ventricle, Fig. 4D). This should then be further clarified by describing the relation of the great vessels to each other (e.g. aorta anterior and rightward). There may be a solitary trunk that gives rise to the coronary arteries, head and neck vessels and pulmonary arteries (i.e. common arterial trunk).



Figure 6

Subcostal views in adults. (A) Subcostal view in an adult with a 'laterality disturbance' (atrial isomerism). The liver is in the midline and abnormal systemic veins (*) draining into a large, single atrium. (B) In the same patient, the apex of the heart (arrow) points to the patient's right. Conventional left parasternal and apical views in this patient were completely unhelpful! (C) Subcostal view in a different patient with the apex pointing to the right (arrow). The ventricle gives rise to two, parallel great arteries – a double outlet ventricle. Ao, aorta; PA, pulmonary artery.

Practical notes for the adult echocardiographer

Since the vast majority of congenital heart disease presents in childhood, paediatric echocardiographers are familiar with the nomenclature, spectrum of abnormalities and the approach to echocardiography referred to in this article. Accordingly, the echocardiography study is organised in a very different manner to the conventional adult echocardiogram: it is standard for the study to start in the subcostal view, allowing for initial identification of the IVC, aorta and therefore atrial arrangement. The atrial and ventricular connections are often well seen in the apical views. The approach of the echocardiographer is segment by segment, utilising views sweeping from one structure to another.

By contrast, the vast majority of echocardiograms performed in general adult cardiology are for acquired heart disease; disorders affecting the arrangement and connections of cardiac structures are extremely rare. On the other hand, the survival of patients with complex congenital heart disease (with or without surgical palliation) into adulthood is increasingly common and presentation to their local hospital is becoming a more frequent occurrence. There is, therefore, an increasing need for echocardiography for such patients in all hospital settings.

The conventional adult echocardiogram starts in the parasternal long axis view – least helpful in establishing the fundamentals of the segmental assessment. For this reason, if faced with a situation in which complex congenital heart disease is suspected – for instance where the *orientation* of structures seen in the parasternal long axis view appears highly abnormal or when there is a history of complex congenital heart disease – adapting the 'paediatric' sequential segmental approach can be invaluable. **Figures 6** and **7** illustrate some of these principles in adult patients:

- (i) Starting from the subcostal view to establish the position of the IVC and arrangement of the atriums. Even in adults, the subcostal views are often the most valuable in establishing the orientation of the heart and obtaining an initial understanding of the sequence and connections of cardiac segments (**Fig. 6A, B and C**).
- (ii) Using this information to move on to the apical views to assess the atrioventricular connections and assessment of the ventricle(s).
- (iii) In addition to standard parasternal views, including a short axis 'sweep' and off axis views (**Fig. 7A, B and C**) to identify and confirm specific segmental features and connections.
- (iv) Ensuring that suprasternal views are included.

In these studies, it is crucial to think about each segment/structure in turn and use intermediate/off axis views to complete the comprehensive assessment outlined above.

Associated abnormalities: a systematic approach

Once the cardiac segments have been identified and the connections have been described, then associated anomalies can be assessed and described using the same systematic approach:

Atrial segment:

- *Is the atrial arrangement normal, mirror image or is there a laterality disturbance?*
- Systemic venous drainage – does the SVC (and IVC) drain normally into the RA? Is the appearance of the coronary sinus normal or enlarged, suggesting a persistent left SVC?



Figure 7

Parasternal, demonstrating normal anatomy in an adult patient. (A) Short axis views at the level of the aortic valve leaflets, the long, thin left atrial appendage is dilated and clearly visible (*). LA, left atrium. (B) An upward angulation, above the level of the aortic valve leaflets, demonstrates the normal relationship between the great arteries: the aorta (Ao) is seen in cross section and the pulmonary artery (PA) is seen along its length – the normal ‘criss-cross’ arrangement. (C) By contrast to Fig. 7B, this off axis parasternal long axis view shows parallel great arteries indicating *discordant* ventriculoarterial connections. The aorta (Ao) is seen anteriorly, and the pulmonary artery (PA), with its branching pattern (arrows), is posterior. In this case, there is a prominent membrane in the left ventricular outflow tract (LV) causing sub-pulmonary stenosis. RV, right ventricle.

- Pulmonary venous drainage – is there evidence of pulmonary veins draining into the RA or systemic veins (this can be difficult to assess by transthoracic echocardiography in adults)?
- Size and appearance of atria.
- Presence, location and size of atrial septal defects.

Atrioventricular connection:

- *Is this concordant, discordant, double, absent?*
- Appearance and function of atrioventricular valves (e.g. mitral valve, tricuspid valve, common A–V valve).

Ventricular segment:

- Morphology, size and function of the ventricles.
- Presence, location and size of ventricular septal defects.

Ventriculoarterial connection:

- *Is this concordant, discordant, double, absent?*
- Appearance of the outflow tracts, presence and severity of obstruction – this may be subvalvar, valvar, supravalvar or a combination.
- Appearance and function of ventriculoarterial valves (e.g. aortic valve, pulmonary valve, common ‘truncal’ valve).

Great arteries:

- Size, position and branching pattern of great arteries.
- Presence, position and size of arterial duct.
- Presence and severity of obstruction (e.g. branch pulmonary stenosis, coarctation of the aorta).
- Origins of the coronary arteries.

Template for congenital heart disease report

Similarly, the structure of the congenital heart disease echocardiogram report differs from the standard approach for the adult echocardiographer. A ‘normal’ report would include:

- Usual atrial arrangement, atrioventricular and ventriculoarterial concordance.
- Pulsatile abdominal aorta.
- IVC/SVC to right atrium.
- Four pulmonary veins to left atrium.
- Competent mitral and tricuspid valves with no stenosis.
- Good biventricular systolic function.
- No left or right ventricular outflow tract obstruction.
- Competent aortic and pulmonary valves.
- Usual coronary artery origins.
- Unobstructed branch pulmonary arteries.
- Unobstructed left-sided aortic arch with a normal branching pattern.
- No ASD/VSD/PDA.

Summary

This method provides a template for the echocardiographer’s study and report in congenital heart disease. Each segment is identified by its characteristic features and then each connection is described along with any associated abnormalities.

Declaration of interest

The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of this review.

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