Editorial

Sequential segmental analysis in complex fetal cardiac abnormalities: a logical approach to diagnosis

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Introduction

Cardiac abnormalities are common. A wide spectrum of lesions can be encountered in the child and fetus. Complex abnormalities are over-represented in prenatal series as these may be more readily recognized during routine anatomical surveys. While the diagnosis of ‘simple’ defects such as pulmonary stenosis or an isolated ventricular septal defect in an otherwise normally connected heart is usually considered straightforward, the description and understanding of the infinite variety of ‘complex’ cardiac abnormalities may be perceived as a difficult task. It need not be so. Sequential segmental analysis offers a step-by-step approach to describing the cardiac anatomy in different malformations and leads to a thorough appreciation of its pathophysiology. This approach was developed and disseminated in the late 1970s and early 1980s1–3 and is based on the concept that all heart malformations can be readily analyzed with reference to three basic segments4–6.

‘Complex’ congenital heart anomalies become simple when approached in a logical fashion. Knowledge of cardiac embryogenesis is unnecessary to describe the anatomy of the heart after 8 weeks’ gestation. All hearts, normal or malformed, are made up of three segments: atria, ventricles and great arteries (Figure 1). In sequential segmental analysis, the cardiac segments (i.e. the morphologically right and left atria, the morphologically right and left ventricles and the great arteries) are identified separately based on their anatomical features, not their spatial orientation. The atria are separated from the ventricles at the level of the fibrous tissue plane of the valves at the atrioventricular (AV) junction. The ventriculo-arterial junction is marked by the attachment of the arterial valves to the ventricular mass. Atrial situs is established first by determining the position of each atrium in relation to each other within the chest and the AV and ventriculo-arterial connections ascertained.

The questions asked are which atrium (atria) connects to which ventricle(s) and which ventricle connects to which great artery (or arteries). These connections form the basic template of the heart. A distinction is made between ‘connections’ and ‘relationships’. The spatial relationships (i.e. what is to the right or left, or anterior or posterior) are described separately from the cardiac
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Figure 2 Steps for determining fetal laterality. (1) Obtain a transverse section through the fetal abdomen and locate the spine (sp) as shown in (a). The right and left sides of the fetus are not yet established. Only two possibilities exist. They are dependent on fetal lie, which is determined by the position of the spine and head. (2) Establish the position of the fetal head in relation to the abdominal cross-sectional view (i.e. head above or below the level of the abdominal scanning plane). (3) In the example shown, with the fetal head located below the plane of the abdomen, the left side is lowermost (b). One can then ascertain that the stomach is on the left. If, however, the fetal head is above the plane of the abdomen it follows that the left side is uppermost (c). In this case the stomach will be to the right side of the fetus. ant, anterior; post, posterior.

connections. In postnatal life various diagnostic tools can be used to help define the sequential analysis of the heart in addition to cross-sectional echocardiography, such as the electrocardiogram, chest radiograph, angiography and magnetic resonance imaging. Antenatal diagnosis, however, is based primarily on ultrasound features. Thus, fetal echocardiography is the technique used.

The nomenclature used in sequential segmental analysis has been well established in pediatric cardiology practice for decades and increasingly so in fetal cardiology. Any nomenclature, however, has to be based on clearly defined but ultimately descriptive terms. The aim of this editorial is therefore to clarify the various concepts necessary for full understanding of the sequential segmental approach to the (fetal) heart so that the descriptive terms can be used in a consistent manner.

Fetal laterality

Contrary to the practice of pediatric cardiology, sequential segmental analysis in the fetus starts with ascertaining of fetal laterality, which is based on fetal orientation in the maternal abdomen. Obstetricians and obstetric sonographers are in general used to sorting out the right and left side of the fetus as, unlike a child, a fetus can be any way up and cannot be asked to lie still in a predetermined position.

Various approaches to determine fetal laterality have been proposed but ultimately this is based on establishing fetal lie. The observer’s three-dimensional visualization of the fetus within the maternal abdomen promptly allows appreciation of the fetus’ right and left sides. A worked-out example is presented in Figure 2.

Atrial arrangement or situs

Analysis of the atrial segment considers atrial arrangement or situs. Situs is the location or position that an organ occupies in a bilateral system of symmetry. Determining atrial arrangement means describing the position of each atrium in relation to each other, irrespective of where the heart is within the chest. It starts with distinguishing the right atrium from the left atrium. The morphologically right atrium is characterized by a broad-based triangular appendage, whereas the left atrial appendage has a narrow base and is tubular and hooked (Figure 3).

The shape of the atrial appendages is the most constant feature of the atrium but it has not been systematically

Figure 3 Cross-sectional view obtained through the fetal chest. Right and left side of the fetus were established first, as labeled. Note the distinct appearance of the right and left atrial appendages (asterisks, RAA and LAA, respectively), which are correctly located on the right and left sides of the fetus, respectively. ant, anterior; lt, left; post, posterior; rt, right.

Figure 4 Anatomically, atrial situs is determined by the arrangement of the atrial appendages as illustrated in the diagram.
imaged in the fetus or child. In current pediatric cardiology practice, atrial situs is most commonly recognized from the arrangement of the abdominal great vessels with respect to the spine, at the level of the diaphragm.

The three types of situs are (Figure 4): (1) solitus or usual atrial arrangement, (2) inversus or mirror-image arrangement and (3) ambiguous: isomerism of right or left atrial appendages.

In usual atrial arrangement (situs solitus), the inferior vena cava lies to the right of the spine and more anterior than the left-sided aorta (Figure 5a). This arrangement only infers that the morphologically right atrium is to the right of the morphologically left atrium and no assumption should be made regarding the position of the heart within the chest. Additionally, the stomach lies to the left and the liver and portal vein to the right. The opposite applies to situs inversus where a mirror-image arrangement of the normal pattern applies (Figure 5b). In right or left isomerism (situs ambiguous) there are two morphologically right or two morphologically left atrial appendages (i.e. the two appendages have similar morphology). In right isomerism the abdominal great vessels are both positioned on the same side of the spine (either right or left) with inferior caval vein anterior and the aorta posterior (Figure 5c). In left isomerism the inferior caval vein is usually interrupted and the aygos vein can be seen as a venous structure posterior to the aorta. The aorta tends to have a more central location with respect to the spine and the aygos vein may be located to either the right or left side (Figure 5d). Generally, right isomerism is associated with bilateral right-sidedness (and absence of left-sided structures such as the spleen, asplenia syndromes). Left isomerism is associated with polysplenia. Malposition of organs such as the stomach, liver, portal vein and heart are often encountered in situs ambiguous and may alert the sonographer to the possibility of a cardiac abnormality.

Inferring atrial situs from the arrangement of the abdominal vessels often suffices as the first step in sequential segmental analysis as this arrangement is usually similar to the arrangement of the lungs (thoracic situs) and atria (atrial situs)\(^9,12\).

**Atrioventricular (AV) junction**

To describe the AV junction, it is necessary first to determine how the atria are connected to the ventricular

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**Figure 5** In ultrasound practice, atrial arrangement is usually inferred from the relative position of the abdominal vessels in relation to the spine, at the level of the diaphragm (approximately T10). The options for atrial situs are: (a) situs solitus: aorta (Ao) to the left, inferior vena cava (IVC) to the right; (b) situs inversus: Ao to the right, IVC to the left; (c) right isomerism: Ao and IVC to the same side. In this example, both are to the right side; (d) left isomerism: Ao is a central vessel, IVC not imaged (interrupted). The venous structure (aygos, Az) is posterior to the Ao (in this example, the Az is to the right). ant, anterior; lt, left; post, posterior; rt, right.

**Figure 6** Diagram illustrating the options for biventricular atrioventricular connections. LV, left ventricle; RV, right ventricle.
mass. An AV connection exists when the cavity of an atrial chamber is in actual or potential continuity with a ventricular cavity. The area of continuity is the AV junction where atrial myocardium is connected to ventricular myocardium at the parietal AV fibrous plane. When each atrium is connected to its own ventricle, a biventricular AV connection exists. When each atrium does not connect to a separate chamber in the ventricular mass, a univentricular AV connection exists.

The possibilities for AV connection are: (1) biventricular AV connection (Figure 6) (subdivided into concordant, discordant or ambiguous) or (2) univentricular AV connection (Figure 7) (subdivided into double-inlet ventricle, absent right AV connection or absent left AV connection).

**Biventricular AV connection**

In concordant AV connection, the right atrium connects to the morphologically right ventricle and the left atrium to the morphologically left ventricle whereas in discordant connection, the right atrium connects to the morphologically left ventricle and the left atrium.

![Diagram](image)

**Figure 7** Diagram illustrating the options for univentricular atrioventricular connections. AV, atrioventricular; LV, left ventricle; RV, right ventricle.

![Cross-sectional views](image)

**Figure 8** Cross-sectional views of the fetal chest at the level of the four-chamber view in two fetuses with usual atrial arrangement (as labeled) and a biventricular atrioventricular (AV) connection. In (a) the connection is concordant as normal offsetting of the AV valves can be seen (asterisk) thus identifying the right atrium (RA) to be on the right side. In (b) the connection is discordant as the presence of reverse offsetting (asterisk) identifies the right ventricle (RV) to be on the left. ant, anterior; LA, left atrium; lt, left; LV, left ventricle; post, posterior; rt, right.

to the morphologically right ventricle. Concordant and discordant connections can only exist in hearts with lateralized atria (Figure 8). In hearts with atrial isomerism a biventricular connection is termed ambiguous. In right isomerism one right atrium connects to the right ventricle and the other to the left ventricle, whereas in left isomerism one left atrium connects to the right ventricle and the other to the left ventricle. In these cases it is therefore necessary to state which ventricle is to the right or left of each other.

The morphologically right and left ventricles in hearts with a biventricular connection have relatively distinct anatomical features that allow each to be recognized on ultrasound. Irrespective of their spatial orientation, apical trabeculations of the morphologically right ventricle are coarser than those of the left ventricle and there is typically a muscle bar in the right ventricle, the moderator band. In contrast, the morphologically left ventricle is characterized by smooth walls with fine apical trabeculations. Another feature of the right ventricle is that its AV valve (the tricuspid valve) has chordal attachments to the interventricular septum, whereas the AV valve connected to the left ventricle (mitral valve) does not. In addition, on echocardiography, the tricuspid valve has a slightly lower insertion on the interventricular septum (in relation to the cardiac apex) compared to the mitral valve (normal offsetting of the valves) (Figure 8).

Univentricular AV connection

In this category, one or two atrial chambers connect to one ventricle (called the ‘main’ or ‘dominant’ ventricle). When both atria, irrespective of their arrangement, connect to the same ventricle, the term used is double-inlet ventricle (Figure 9a). The other types of univentricular atrioventricular connection occur when one of the atria has no connection – actual or potential – with the underlying ventricular mass and sulcus tissue separates the atrial from ventricular myocardium. In these instances only one atrium connects to the ventricular mass. Thus, there may be either absent right or absent left AV connection (Figure 9b, 9c). The term ‘univentricular’ applies to the type of AV connection and not to the heart itself as there are usually two chambers in the ventricular mass, the second chamber lacking an inlet portion and being called rudimentary typically being smaller than the main chamber (Figure 9b).

If the dominant ventricle is of left ventricular morphology, the rudimentary right ventricle occupies an antero-superior position and may be right- or left-sided or directly anterior. Conversely, a rudimentary left ventricle is postero-inferior and either right- or left-sided. Rarely, there is only one ventricular chamber, which is of indeterminate morphology.

Mode of AV connection

In addition to the type or category of AV connection, the morphology of the AV valves (mode of AV connection) also requires description. The possibilities are: (1) two perforate valves, (2) one imperforate and one perforate, (3) common valve and (4) straddling and overriding valves.

In straddling, the valvar tensor apparatus, the chordae tendineae are attached to both sides of the interventricular septum. In contrast, overriding is used to describe the arrangement in which the valve orifice lies above both ventricles often above a ventricular septal defect. Straddling valves are usually associated with ventricular septal defect as are the majority of, but not all, overriding valves. A 50% rule defines to which ventricle an atrium is connected depending on the degree of overriding. This therefore influences the category of AV connection to which the heart is assigned.

Figure 9 Cross-sectional views of the fetal chest at the level of the four-chamber view in three fetuses with usual atrial arrangement (as labeled) and a univentricular atrioventricular (AV) connection. In (a) the connection is double-inlet as the two atria are connected to one main chamber (left ventricle, LV) via two AV valves (asterisks). In (b) there is absence of the right AV connection (asterisk). A small rudimentary right ventricle (rud rv) can be seen in addition to the dominant LV. In (c), there is absence of the left AV connection (asterisk). ant, anterior; LA, left atrium; lt, left; post, posterior; RA, right atrium; rt, right.
**Ventriculo-arterial junction**

Segmental analysis next involves determining the ventriculo-arterial connection and describing which great artery connects to which ventricle. To describe ventriculo-arterial connections, the echocardiographic features used to distinguish aorta, pulmonary trunk and common arterial trunk have to be known. The aorta is the great artery that gives branches to the head, neck and arm arteries and coronary arteries, although the latter are not routinely imaged in the fetus. The pulmonary trunk usually bifurcates into left and right pulmonary arteries, and in the fetus it continues as the arterial duct, which connects to the descending aorta. A common arterial trunk is recognizable as a single great artery arising from the ventricles that gives rise to coronary arteries, pulmonary arteries and head and neck vessels.

There are four types of ventriculo-arterial connection (Figure 10): (1) concordant, (2) discordant, (3) double-outlet and (4) single-outlet.

Concordant ventriculo-arterial connection means the pulmonary artery is connected to the morphologically right ventricle and the aorta to the morphologically left ventricle (Figure 10a, 10b). In discordant ventriculo-arterial connections the pulmonary trunk connects with the morphologically left ventricle and the aorta connects with the morphologically right ventricle (Figure 10c). Double-outlet is when more than 50% of each great artery is connected to the same ventricular chamber (Figure 10d). Double-outlet can occur from a morphologically right, a morphologically left or from a ventricle of indeterminate morphology. Single-outlet of the heart applies to when only one arterial trunk connects with the ventricular mass. The single outlet may be via a common arterial trunk (persistent truncus arteriosus) (Figure 10e) or the solitary arterial trunk may be associated with either pulmonary or aortic atresia where the atretic trunk cannot be traced with certainty to a particular ventricle. The ventricles to which one or both great arteries are attached may be a normal right or left ventricle in a biventricular connection or a dominant or rudimentary ventricle in a univentricular AV connection.

Arterial valves can override but cannot straddle because, unlike AV valves, they do not have a tensor apparatus. The degree of overriding influences the category into which the ventriculo-arterial connection is assigned.

**Associated abnormalities**

Having established the basic arrangement of the heart, each segment is analyzed for abnormalities. In the presence of a normally connected heart, the associated lesions will constitute the major (or minor) cardiac defect. In the presence of abnormalities of situs and/or cardiac connections, identification of associated lesions is important as their presence may alter the pathophysiology otherwise expected as a consequence of the abnormal cardiac connections.

**Atrial**

Systemic and pulmonary venous connections can be imaged in the fetus. The arrangement of the abdominal veins has already been identified in determining atrial situs. The superior vena cava is seen in the three-vessel view and with normal drainage can be seen entering the morphologically right atrium. Bilateral superior caval veins can also be identified. The left superior vena cava typically enters the coronary sinus. Normally, four pulmonary veins enter the left atrium. When present, anomalous pulmonary venous connections, partial or total, can be identified in the fetus.

**AV and ventriculo-arterial junction**

Seen in a four-chamber view, the morphology of the AV valves is identified and any inlet ventricular septal defect seen whereas outlet defects are seen when angling the transducer anteriorly from the four-chamber view. Ventricular morphology and size are also ascertained. Valve stenosis if present can be documented, as can abnormalities of the great arteries such as interruption of the aortic arch. Coarctation of the aorta is normally inferred from ventricular or great vessel disproportion, but can be imaged directly in some fetuses.

**Cardiac position**

The location of the heart within the thorax is independent of the chamber connections. The heart may be within

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**Figure 10** Transverse oblique views of the fetal chest demonstrating the various types of ventriculo-arterial connections. In (a-b) the connection is concordant (left ventricle (LV) to aorta (Ao) and right ventricle (RV) to the pulmonary artery (PA)). In (c) the connection is discordant (transposition, Ao from RV and PA from LV). In (d) both great arteries arise from the same ventricle (double outlet RV). An example of a single outlet heart is shown in (e); the common arterial trunk is shown to give rise to both the Ao and PA (asterisk). ant, anterior; post, posterior; T, common arterial trunk.
the right chest (dextrocardia), within the left chest (levocardia) or centrally placed (mesocardia). Similarly, the apex of the heart may point to the left, right or not be identifiable. In situs solitus the heart will almost always be mainly in the left chest with its apex pointing to the left. In situs inversus with mirror image the heart is usually mainly in the right chest with its apex pointing to the right, but there will be occasions when the heart is left-sided. It should be emphasized that description of cardiac position gives no definitive information concerning the internal segmental arrangement of a given malpositioned heart.

We hope to have provided a practical way of describing the heart in the fetus based on identifying the different cardiac segments. With practice, ‘complex’ malformations can then also be described in a simple descriptive fashion (Figure 11).

REFERENCES