



# Sequential Segmental Analysis of the Heart: A Malformation Screening Technique

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## Abstract

The diagnosis of fetal cardiac malformations has a reputation for being complex and difficult. It is not necessary to have knowledge of cardiac embryogenesis to describe the malformed heart. But it is essential to identify the normal cardiac chambers and great arteries from their morphological characteristics. Fetal cardiac abnormalities can be easily understood if approached in a simple and straightforward fashion. Sequential segmental analysis and the use of simple descriptive terms allows description of the most complex cardiac anomalies. In this review, we highlight features of the heart that can be identified by obstetricians who scan the fetal heart.

**Key words:** sequential segmental analysis, fetal heart, malformation

## Özet

### Kalbin Ardışık Segmental Analizi: Malformasyon Tarama Tekniği

Fetal kardiyak anomalilerin tanısının karmaşık ve zor olduğu düşünülür. Malforme kalbin tanımının yapılabilmesi için kardiyak embriyogenez hakkında bilgi sahibi olunması gerekli değildir. Fakat normal kardiyak boşlukların ve büyük arterlerin morfolojik özellikleri arasında ayırım yapmak yeterlidir. Yaklaşım basit ve karmaşıklığa yol açmadan yalın olursa fetal kardiyak anomaliler kolaylıkla tanınabilir. Ardışık segmental analiz ve temel tanımlayıcı terimlerin kullanımı birçok kompleks kardiyak anomalilerin tanımlanmasına olanak verir. Bu derlemede fetal kardiyak ultrasonu yapan obstetrisyenlerin bilmesi gereken temel özellikler vurgulanmıştır.

**Anahtar sözcükler:** ardışık segmental analiz, fetal kalp, malformasyon

## Introduction

The diagnosis of fetal cardiac malformations has a reputation for being complex and difficult. The notion that knowledge of cardiac embryogenesis is necessary to describe anomalies is unwarranted. In this article, we show how fetal cardiac abnormalities can be easily understood if approached in a simple and straightforward fashion.

All hearts can be analysed irrespective of how complex the anomaly, on the basis of three segments; the atria, the ventricles and the great arteries with simple descriptive words being used to describe how the different segments connect to another, their spatial relationships and any associated anomalies. This comprehensive system for describing the heart was first applied by Shinebourne et al in 1976 and its use in practice is called Sequential Segmental Analysis of the Heart (1,2). The basic template is to determine the atrial arrangement and describe the atrioventricular and ventriculoarterial connections. For

prenatal screening, cross sectional echocardiography is used to image the heart.

## Atrial Arrangement

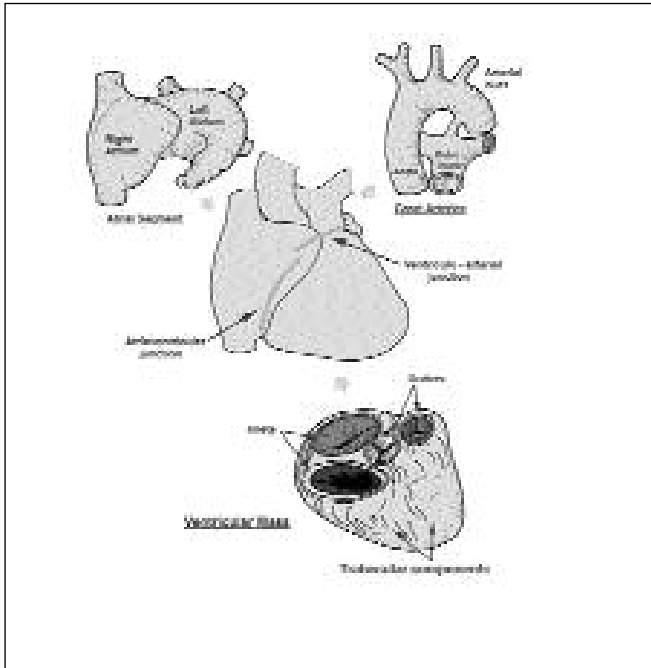
The first step in segmental analysis is to determine the arrangement of the atria. The morphologically right atrium is characterised by a broad based triangular appendage, whereas the left atrial appendage has a narrow base and is tubular and hooked. Analysis of the atrial segment considers atrial arrangement or situs. Situs is the location or position that an organ occupies in a bilateral system of symmetry. The three types of situs are:

- Solitus (usual arrangement).
  - Inversus (mirror image).
  - Ambiguus (isomerism of right or left atrial appendages).
- Although the shape of the atrial appendage is the most constant feature of the atrium, it is not always visualised. In practice, atrial situs can be most commonly recognized from the arrangement of the abdominal great vessels.

In usual atrial arrangement (situs solitus), the aorta lies to the left of the spine and the inferior vena cava to the right, the opposite being the case in situs inversus (3).

In right or left isomerism (situs ambiguus), there are two morphologically right or two morphologically left atrial

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**Figure 1.** Diagram showing the three segments of the heart and the two junctions, atrioventricular and ventriculo-arterial, that are analysed in the sequential segmental approach.

appendages. The abdominal great vessels are on the same side of the spine with the inferior caval vein anterior and the aorta posterior, in right isomerism. In left isomerism, the inferior caval vein is interrupted and the venous structure (azygous vein) is posterior to the aorta. Generally, right isomerism associated with bilateral right sidedness (and absence of left sided structures such as the spleen, asplenia syndromes) (4). Left isomerism is associated with polysplenia (5).

### Atrioventricular Junction

To describe the atrioventricular junction, it is first necessary to determine how the atria are connected to the ventricular mass. When each atrium is connected to its own ventricle, a biventricular atrioventricular connection exists.

The possibilities are:

- Concordant
  - Right Atrium to Right Ventricle, Left Atrium to Left Ventricle
- Discordant
  - Right Atrium to Left Ventricle, Left Atrium to Right Ventricle
- Ambiguous
  - Right Atrium to Right Ventricle, Right Atrium to Left Ventricle or Left Atrium to Right Ventricle, Left Atrium to Left Ventricle

When each atrium does not connect to a separate chamber in the ventricular mass, a univentricular atrioventricular connection exists.

The possibilities are:

- Double inlet ventricle
- Absent right atrioventricular connection
- Absent left atrioventricular connection

When both atria, irrespective of their arrangement, connect to the same ventricle, the term used is double inlet ventricle. There are usually two chambers in the ventricular mass, the second chamber being rudimentary. The other types of univentricular atrioventricular connection occur when one of the atria has no connection with the underlying ventricular mass. Thus, there may be either absent right or left atrioventricular connection.

The morphologically right and left ventricles have a constellation of anatomic features which allow each to be recognized by a sonographer. Irrespective of their spatial orientation, apical trabeculations of the morphologically right ventricle are more coarse than that 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 the tricuspid valve has chordal attachments to the interventricular septum, whereas the normal mitral valve does not. In addition, on echocardiography, the tricuspid valve has a slightly lower insertion on the interventricular septum compared to the mitral valve.

In addition to the type or category of atrioventricular connection, the morphology of the atrioventricular valves (mode of atrioventricular connection ) also requires description.

The possibilities are:

- Two perforate valves
- Common valve
- One imperforate and one perforate valve
- Straddling and/or overriding valves

In straddling, the valvar tensor (chordae tendineae) apparatus is connected on both sides of the interventricular septum. In contrast, overriding is used to describe the arrangement in which the valve orifice lies above both ventricles usually above a ventricular septal defect (6). Straddling valves are usually associated with an inlet ventricular septal defect as are the majority, but not all overriding valves.

### Ventriculo-arterial Junction

Segmental analysis next involves determining the ventriculo-arterial junction, describing the connections, spatial relations and infundibular morphology. Separately, connections specifically state the fashion in which two structures are connected one to the other irrespective of their relations. Relations describe the position of the one structure to the other structure in terms of right/left and anterior/posterior coordinates (7). Connections are paramount in describing the architecture of a given heart. The possibilities are shown in Table 1. To describe ventriculo-arterial connections, distinction must be made between aorta, pulmonary trunk and common arterial trunk. On scanning the aorta is the great artery that continues into the aortic arch with its branches to the head, neck and arm arteries whereas the pulmonary trunk

**Table 1.** Ventriculo-arterial connections

Connections		Chambers and arteries connected
Concordant ventriculo-arterial connections		Aorta from left ventricle or its rudiment. Pulmonary trunk from right ventricle or rudiment.
Discordant ventriculo-arterial connections		Pulmonary trunk from left ventricle or rudiment. Aorta from right ventricle or rudiment.
Double outlet connection		More than half of both arteries from left, right, or indeterminate ventricle.
Single outlet heart	Truncus arteriosus	Single arterial trunk giving rise to coronary arteries, head and neck vessels and pulmonary artery(s).
Single outlet heart	Single aortic trunk	Aorta from ventricular chambers; atretic pulmonary trunk with no connection to ventricular chamber.
Single outlet heart	Single pulmonary trunk	Pulmonary trunk from ventricular chambers; atretic aorta with no connection to ventricular chamber.

usually bifurcates into the left and right pulmonary arteries. The arterial duct connects the pulmonary bifurcation to the underside of the aortic arch. The common arterial trunk is recognizable as a single great artery that, when traced, continues into the pulmonary arteries as well as the aortic arch. There are four types of ventriculo-arterial connection:

1. Concordant ventriculo-arterial connection in which the aorta is connected to a morphologically left ventricle and pulmonary trunk to a morphologically right ventricle.
2. Discordant ventriculo-arterial connections in which the pulmonary trunk connects with the morphologically left ventricle while the aorta connects with the morphologically right ventricle.
3. Double outlet in which more than 50% of each great artery is connected to the same ventricular chamber. Double outlet can occur from a morphologically right, a morphologically left or from a ventricle of indeterminate morphology.
4. Single outlet of the heart in which only one arterial trunk connects with the ventricular mass. The single outlet may be via a common arterial trunk (persistent truncus arteriosus), a solitary arterial trunk or be associated with pulmonary or aortic atresia where the atretic trunk cannot be traced with certainty to a particular ventricle.

Arterial valves can override, but cannot straddle because, unlike atrioventricular valves, they do not have a tensor apparatus.

### Cardiac Position

The location of the heart within the thorax is independent of the chamber connections. The heart may be within 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 is no reason why the heart should not be 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.

### Comment

In this review, we highlight features of the heart that should become familiar to obstetricians who scan the fetal heart. Sequential segmental analysis imposes a systematic approach to simple and complex cardiac anomalies. It is not complete until all intracardiac and vascular anomalies have been listed. With practice, the commoner anomalies such as ventricular septal defect and atrioventricular septal defect can be detected by obstetricians. Using sequential segmental analysis should simplify diagnosis and avoid omissions.

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