Utility of 4 Chamber view in detecting cardiac anomalies

Poster No.: C-1505
Congress: ECR 2014
Type: Educational Exhibit
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Keywords: Congenital, Intrauterine diagnosis, Ultrasound, Foetal imaging
DOI: 10.1594/ecr2014/C-1505

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Learning objectives

Cardiovascular anomalies are frequently encountered during antenatal evaluations. The aim of the study is to be able to identify the diverse anomalies of the foetal heart chambers using the standard 4 chamber view on Ultrasound examination in order to be able to aid in the further management of the pregnancy and to reduce the postnatal morbidity and mortality.

Background

Congenital cardiac anomalies are estimated to occur in approximately 8 out of 1000 live births. The 4 chamber view during the evaluation of the foetal heart forms an integral part of an anomaly scan between the age of 18 - 22 weeks (calculated from the last menstrual period).

The four-chamber view is obtained by a transverse projection through the fetal thorax above the level of the diaphragm, either apical (parallel to the interventricular septum) or subcostal (perpendicular to the interventricular septum).

This view shows the two atria and ventricles along with atrioventricular (AV) valves (mitral and tricuspid) and septa (interventricular and interatrial).

Cardiac anatomy is typically evaluated using a sequential segmental approach, which depends on morphologic identification of the atria, ventricles, and great arteries, not on their spatial relationship. The morphologic right atrium (RA) has a triangular appendage, whereas the morphologic left atrium (LA) has a hook-shaped appendage.

The tricuspid valve opens into the RV and the mitral valve opens into the LV, with the septal leaflet of tricuspid valve inserting more apically than the mitral valve.

The importance of detection of anomalies on four chamber view are as follows:

1. Allows time for a second look using other modalities like foetal echocardiography for more precise evaluation
2. Helps to reduce postnatal morbidity and mortality
3. Allows prenatal counselling
4. Allows time planned for optimum antenatal care and planning optimal delivery techniques
5. Allows time to relocate a patient to a centre equipped to handle a complex case
6. Helps to characterize according to specific syndromes and look for associated extracardiac anomalies
ISUOG Practice Guidelines (updated): Sonographic screening examination of the fetal heart

Situs and general aspects
- Fetal laterality (identify right and left sides of fetus)
- Stomach and heart on left
- Heart occupies a third of thoracic area
- Majority of heart in left chest
- Cardiac axis (apex) points to left by 45°±20°
- Four chambers present
- Regular cardiac rhythm
- No pericardial effusion

Atrial chambers
- Two atria, approximately equal in size
- Foramen ovale flap in left atrium
- Atrial septum primum present (near to crux)
- Pulmonary veins entering left atrium

Ventricular chambers
- Two ventricles, approximately equal in size
- No ventricular wall hypertrophy
- Moderator band at right ventricular apex
- Ventricular septum intact (apex to crux)

Atrioventricular junction and valves
- Intact cardiac crux
- Two atrioventricular valves open and move freely
- Differential offsetting: tricuspid valve leaflet inserts on ventricular septum closer to cardiac apex than does mitral valve

*Ultrasound Obstet Gynecol* 2013; 41: 348–359

**Fig. 1:** 4 CHAMBER VIEW GUIDELINES
Findings and procedure details

AXIS DEVIATION

FIGURE 2

The cardiac axis is calculated from a line drawn from the posterior spine to the anterior sternum (spinosternal line). The ventricular septum typically intersects this line at 40-45

SITUS ANOMALIES

FIGURE 3

<table>
<thead>
<tr>
<th>SITUS</th>
<th>RIGHT SIDE</th>
<th>LEFT SIDE</th>
</tr>
</thead>
<tbody>
<tr>
<td>INVERSUS</td>
<td>• Morphologic left atrium</td>
<td>• Morphologic right atrium</td>
</tr>
<tr>
<td></td>
<td>• Stomach</td>
<td>• Major hepatic lobe</td>
</tr>
<tr>
<td></td>
<td>• Descending aorta</td>
<td>• Inferior vena cava</td>
</tr>
<tr>
<td></td>
<td>• Bilobed lung</td>
<td>• Trilobed lung</td>
</tr>
<tr>
<td></td>
<td>• Long hyparterial bronchus</td>
<td>• Short epiarterial bronchus</td>
</tr>
<tr>
<td>AMBIGUOUS</td>
<td>Variable</td>
<td>Variable</td>
</tr>
</tbody>
</table>

FOETAL CARDIAC POSITION

FIGURE 4, 5, 6, 7, 8

Foetal cardiac position independent of the axis

- Dextrocardia - heart located in the right chest
  - Dextroposition - heart in right chest with apex medial or to the left commonly due to extrinsic factors
  - Dextroversion - heart in right chest with axis pointing to right, found in situs inversus & situs ambiguous. Commonly associated with CHD.

- Mesocardia - central position of the heart

- Levocardia - left sided position of heart
• Levocardia, normal position of heart - term used with visceral situs abnormality
• Levoposition - heart displaced further towards left

SINGLE VENTRICLE

FIGURE 9, 10

Single ventricle is characterized by a single or two AV valves opening into a single ventricle.

• It accounts for 2% of congenital heart defects and is caused by failure of development of the interventricular septum.
• The single ventricle may be the morphologic LV (85%) or the RV.
• The ventricle may not have an outflow tract.
• It may be associated with VSD, ASD, common atrium, pulmonary stenosis, and cardiosplenic syndromes.
• Ultrasound shows a single ventricle without an interventricular septum. Differential diagnosis includes large VSD or hypoplastic RV or LV

HYPOPLASTIC LEFT HEART

FIGURE 11, 12

• Hypoplastic left heart syndrome is characterized by hypoplastic left-sided cardiac structures, including the LV, mitral valve, aortic valve, and aorta.
• It accounts for 2-4% of congenital cardiac defects and is seen in 0.16-0.25 per 1000 live births.
• It is more common in boys and is caused by decreased flow in and out of the LV during development (e.g., mitral or aortic stenosis or atresia).
• Blood flow to the systemic circulation (coronary arteries, brain, liver, and kidneys) in these patients is dependent on flow through the ductus arteriosus.
• On ultrasound, the LV is small (LV:RV ratio < 1) in size the ventricular septum makes an angle of 90° with the spinosternal line, and the aortic outflow is smaller than the pulmonary outflow tract.
• Mitral and aortic valves are hypoplastic or atretic. A single area of flow is seen at the AV level and bidirectional flow at the proximal aorta because of distal aortic coarctation

MITRAL REGURGITATION

FIGURE 13

Rarely occurs isolated. It is commonly associated with mitral stenosis
Mitral valve prolapse: It is a cause for mitral regurgitation caused due to myxomatous proliferation which can lead to myxomatous degeneration of the loose spongiosa and fragmentation of the collagen fibrils.

**PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM**

**FIGURE 14, 15**

Characterized by

- Complete obstruction to the right ventricular outflow tract
- Hypoplastic right ventricle
- Thickened right ventricular wall
- Intact ventricular septum

**TRICUSPID VALVE DYSPLASIA**

**FIGURE 16, 17**

Characterized by

- Thickened dysplastic tricuspid valve leaflets
- Normal insertion of tricuspid valve leaflets on the annulus
- Varying degrees of tricuspid valve regurgitation
- Dilatation of the right atrium

**TRICUSPID REGURGITATION**

Causes

- Trivial
- Heart defects with dysplastic TV
- Heart diseases with right ventricular outflow obstruction
- Heart defects with 'facultative' TV regurgitation
- Volume overload
- Impaired myocardial contractility

**TRICUSPID ATRESIA**

**FIGURE 18, 19, 20**

A muscular ridge separates the right atrium from the rudimentary ventricle and only a single AV valve can be demonstrated which is morphologically the mitral valve.

**TRICUSPID ATRESIA WITH VENTRICULAR SEPTAL DEFECT**
Characterized by

- Absence of right atrioventricular connection
- Diminutive right ventricle
- Widely patent foramen ovale
- VSD
- Right ventricular obstruction

VENTRICULAR SEPTAL DEFECTS

FIGURE 21, 22

<table>
<thead>
<tr>
<th>TYPES</th>
<th>ALSO CALLED</th>
<th>LOCATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perimembranous</td>
<td>Infracristal conoventricular</td>
<td>In the inflow tract beneath the aortic valve &amp; under the subventricular crest</td>
</tr>
<tr>
<td>Outlet</td>
<td>Supracristal , subpulmonic , subarterial</td>
<td>Under the pulmonary valve and above the supraventricular crest</td>
</tr>
<tr>
<td>Muscular</td>
<td>Trabecular</td>
<td>Can be apical , midmuscular or multiple</td>
</tr>
<tr>
<td>Inlet</td>
<td>Posterior aterioventricular septum</td>
<td>Posterior to the septal leaflet of the tricuspid valve</td>
</tr>
</tbody>
</table>

TYPES OF VENTRICULAR SEPTAL DEFECTS

ATRIAL SEPTAL DEFECTS

FIGURE 23

- Atrial septal defect (ASD) is characterized by defect in a portion of the atrial septum.
- It is the fifth most common congenital heart disease, seen in 1 of 1500 live births, and is caused by abnormal tissue resorption and deposition during development of the atrial septum.
- According to its location, it is classified as ostium secundum (midatrial septum), ostium primum (lower atrial septum), sinus venosus (outside the atrial septum in the wall separating the SVC or IVC from the LA), and coronary sinus defect, which can be partial or complete.
- ASD may be difficult to visualize in a fetus because of the presence of foramen ovale. However, with high-resolution ultrasound, the septum primum is seen in the four-chamber view as a circular or linear structure with a loose pocket configuration, and the septum secundum is seen as a thick stationary structure with the foramen ovale opening into it.
• Normal foramen ovale measures almost the same as the aortic root, with the difference being 1 mm or less
• A secundum defect is seen as a larger defect in the central portion of the atrial septum or a deficient foramen flap. A primum defect is seen in the lower part of the atrial septum

ATRIOVENTRICULAR SEPTAL DEFECT

(FIGURE 24, 25, 26)

• AV septal defect (AV canal defect or endocardial cushion defect) is caused by failure of fusion of the endocardial cushion, resulting in defects of the atrial ostium primum, the ventricular inlet septum common AV valve, and the biventricular AV connections.
• AV septal defect accounts for 2-7% of congenital heart defects and is seen in 0.19-0.56 per 1000 live births.
• It is associated with trisomy 21 syndrome, left atrial isomerism, hypoplastic left heart, pulmonary stenosis, coarctation, tetralogy, complete heart block, and extracardiac anomalies.
• There are two types: the complete type (97% of cases), with common valvular orifice, and the incomplete type, with separate right and left valve orifices.
• The valve of common AV junction has five leaflets, which are separate in the complete type, but two leaflets are connected by narrow tissue in the incomplete type.
• It is associated with a cleft in the anterior mitral leaflet. Free regurgitation is seen across the common AV valve.
• Direct shunting may be seen from the LV into the RA. In severe forms, all four chambers communicate, causing left-to-right and right-to-left shunt.
• Ultrasound shows a defect in the endocardial cushion, with an inlet VSD and primum ASD associated with a single abnormal AV valve that has a T-shaped arrangement.
• Color Doppler shows open flow across the defect and abnormal AV valve.

EBSTEIN ANOMALY

FIGURE 27, 28

• Ebstein anomaly is characterized by displacement and attachment of one or more tricuspid leaflets (usually septal or posterior leaflets) toward the apex of the RV.
• The RV is divided into an "atrialized" portion above the leaflets and a muscular portion below the leaflets. It accounts for less than 1% of congenital heart defects, occurs at a rate of 7% in the fetal population, and occurs in 1 per 20,000 live births.
• It is associated with maternal lithium use, chromosomal abnormalities, ASD, patent foramen ovale, and pulmonary stenosis or atresia.
• Ultrasound shows apical displacement of the tricuspid valve into the RV, tethered leaflets, reduction in the size of the functional RV (increase in the size of the RV (including the atrialized portion), and tricuspid regurgitation.
• Cardiomegaly, hydrops, and tachyarrhythmias may be seen. Intrauterine mortality is as high as 85%.
• Differential diagnosis includes Uhl anomaly, tricuspid valve dysplasia, and idiopathic RA enlargement

TETRALOGY OF FALLOT

FIGURE 29

Tetralogy of Fallot is characterized by narrowing of the RVOT, VSD, overriding aorta, and right ventricular hypertrophy. It accounts for 5-10% of congenital cardiac defects and is seen in 0.24-0.56 per 1000 live births. It is caused by anterior displacement of the conotruncus, resulting in unequal division of conus into a small anterior RV portion and large posterior LV portion. The incomplete closure of the septum results in aortic overriding. It is associated with chromosomal and extracardiac abnormalities.

On ultrasound, the aorta is seen straddling a large membranous VSD. Depending on the size of the PA, it may not be easily seen and the normal crossing of aorta and pulmonary arteries is not seen.

The aorta may be dilated, and the pulmonary valve is stenosed or atretic with a dilated PA.

Because of the presence of normal fetal shunts, RV hypertrophy is not seen in the fetus.

CARDIAC MASSES

FIGURE 30, 31

Cardiac neoplasms are uncommon in the fetus, with most of them being primary rather than metastatic.

Approximately 50% of masses are intracavitary, with inflow or outflow obstruction; 10% of these masses are malignant. Rhabdomyoma (60%), teratoma (25%), fibroma (12%), hemangioma, and hamartoma are the common masses. Myxoma, oncocytic cardiomyopathy, lymphangioma, metastasis, epithelial cysts, mesothelioma of AV node, and valvular blood cysts are rare.

Rhabdomyoma is a sessile, smooth, lobulated, and round tumor that is homogeneously echogenic. It is more commonly multiple and typically located in the ventricular septum, atrial, or ventricular free walls. Rhabdomyomas usually do not cause hemodynamic
compromise, grow because of maternal hormones, and spontaneously regress after birth. Multiple rhabdomyomas are associated with tuberous sclerosis in 100% and solitary tumors in 50% of cases. Rhabdomyosarcoma, however, appears clustered, irregular, or fragmented these cases.

**Fibroma** is less common and is seen as a smooth homogeneous mass that blends with the myocardium, often indistinguishable from rhabdomyosarcoma. It is commonly seen in the interventricular septum and less commonly in the ventricular free walls. It is heterogeneous if there is cystic degeneration or calcification. It may be associated with Beckwith-Wiedemann syndrome or Gorlin syndrome.

**CARDIOMYOPATHY**

**FIGURE 32, 33**

- Cardiomyopathies account for 8-11% of fetal cardiovascular abnormalities with one third of fetuses dying in utero
- Cardiomyopathies can be broadly classified as dilated, hypertrophic, and restrictive types. Intrinsic causes of primary cardiomyopathy are single-gene disorders ( Noonan syndrome, familial cardiomyopathy, and metabolic abnormalities), mitochondrial and storage disorders, chromosomal abnormalities, and #-thalassemia.
- Extrinsic causes are intrauterine infections, maternal diseases (autoantibodies and diabetes), and twin-twin transfusion syndrome.
- Dilated cardiomyopathy is the end result of various cardiac disease processes and is characterized by the dilation of cardiac chambers and the reduction of systolic function.
- In hypertrophic cardiomyopathy, the LV-RV myocardial thickness is increased without an underlying structural abnormality. It has been associated with maternal diabetes and often regresses during the first 6 months of life. Ventricular hypertrophy can also be seen because of increased afterload.
- Decreased LV compliance results in cardiac and respiratory distress. Restrictive cardiomyopathy is characterized typically by normal ventricular size and systolic function, but abnormal diastolic function and elevated filling pressure. Secondary cardiomyopathies have better prognosis than do idiopathic or familial cases.

**RHYTHM ABNORMALITIES**

**FIGURE 34**

Fetal arrhythmias are a rare but serious condition occurring in an estimated 1-2% of pregnancies. In about 10% of those cases, morbidity or even mortality occurs.
Depending on the type of arrhythmia, hydrops fetalis, neurological sequelae and fetal demise can be expected

Persistent fetal arrhythmias are associated with structural cardiac defects in about 15% of cases.

The risk of intrauterine death, which varies greatly and depends on the underlying form of arrhythmia, is estimated to lie anywhere from 9% to 50%

Arrhythmias can be grouped into

Irregular rhythm: irregularities caused by premature atrial contractions. The average fetal heart rate is normal or near normal in most cases.

Tachycardias: rhythms exceeding physiologic variation, more than 180 beats per minute (bpm), can be sustained or intermittent. It can be further divided into

- Supraventricular tachycardia involving the conductive system
- Atrial tachycardia: Atrial Flutter, Atrial Fibrillation, Atrial ectopic tachycardia
- Ventricular tachycardia

Bradycardia: rhythms with less than 100 bpm. Types include

- Sinus bradycardia
- Atrioventricular block
- Complete heart block

PERICARDIAL EFFUSION

Common associations

- Hydrops foetalis
- Foetal arrhythmias
- Fetal cardiac masses
- Trisomy 21

Images for this section:
Fig. 2: AXIS DEVIATION
<table>
<thead>
<tr>
<th></th>
<th>Vertex</th>
<th>Breech</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anticlock Wise</td>
<td>![Vertex Image]</td>
<td>![Breech Image]</td>
</tr>
<tr>
<td>Clock Wise</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Fig. 3:** APPEARANCE OF SITUS INVERSUS IN VETEX AND BREECH PRESENTATION
Fig. 4: DEXTROVERSION
**Fig. 5:** DEXTROPOSITION WITH THE HEART PUSHED TO THE RIGHT
Fig. 6: LEVOCARDIA
Fig. 7: LEVOPOSITION
Fig. 8: MESOCARDIA
Fig. 9: COR TRILOCULARE - BIATRIXUM (double inlet ventricle)
Fig. 10: UNIVENTRICULAR HEART
Fig. 11: HYPOPLASTIC LEFT HEART With dysplastic mitral valve, atretic aortic valve and hypoplastic aorta
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Fig. 16: LINE DIAGRAM OF TRICUSPID VALVE DYSPLASIA
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Fig. 19: TRICUSPID ATRESIA
Fig. 20: TRICUSPID ATRESIA WITH VENTRICULAR SEPTAL DEFECT
Fig. 21: LINE DIAGRAM OF TYPES OF VENTRICULAR SEPTAL DEFECTS
Fig. 22: VENTRICULAR SEPTAL DEFECT
**Fig. 23:** LINE DIAGRAM OF ATRIAL SEPTAL DEFECT
Fig. 24: LINE DIAGRAM OF ATRIOVENTRICULAR SEPTAL DEFECTS
Fig. 25: ATRIOVENTRICULAR SEPTAL DEFECTS
Diastole (AV valve opened)  
Two dimensional: gap in the centre of heart  
**Color:** single outflow channel with mixture of blood in both atria & ventricles

Systole (AV valve closed)  
Two dimensional: common AV valve with both leaflets at one level, showing a linear line  
Two dimensional: AV length ratio > 0.6 (long atrium)  
**Color:** AV valve regurgitation

**Fig. 26:** DIAGNOSIS OF ATRIOVENTRICULAR SEPTAL DEFECTS ON 4 CHAMBER VIEW
Fig. 27: LINE DIAGRAM OF EBSTEIN ANOMALY
Fig. 28: EBSTEIN ANOMALY
Fig. 29: A 4 CHAMBER VIEW FOR TETRALOGY OF FALLOT
Fig. 30: MULTIPLE RHABDOMYOMAS
Fig. 31: FIBROMA
Fig. 32: DILATED CARDIOMYOPATHY
Fig. 33: FIBROELASTOSIS
Fig. 34: BRADYCARDIA IN A CASE OF FIBROELASTOSIS
Fig. 35: PERICARDIAL EFFUSION
Conclusion

Routine fetal cardiac ultrasound using four-chamber enables the detection and characterization of most of the cardiac anomalies. However vast number of cardiovascular anomalies are not limited to the chambers. Infact they the chamber anomalies form a part of the spectrum. It is essential to evaluate vascular and outflow anomalies as well in each scan. A further comprehensive evaluation can be performed with fetal echocardiography, particularly in high-risk pregnancies and extracardiac anomalies. Doppler imaging is used in the evaluation of vascular and valvular lesions. MRI is a complementary tool, especially when fetal cardiac structures are visualized suboptimally with ultrasound.

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