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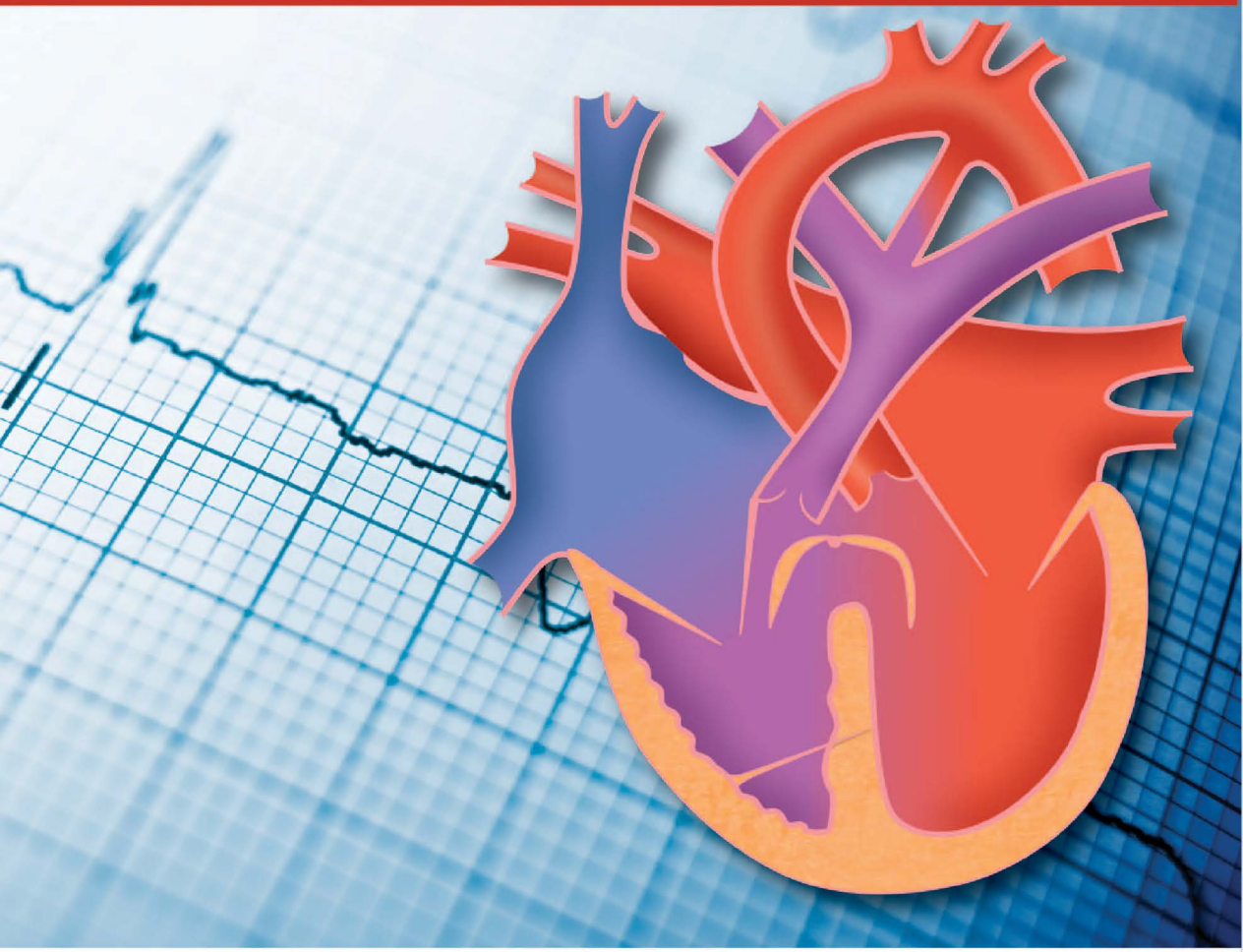
FETAL HEART

Screening, Diagnosis and Intervention



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What is Different in Fetal Heart Examination and Echocardiography?

Olus Api

Worldwide, 1.35 million infants are born with congenital heart disease (CHD) each year, with a worldwide occurrence of 7 per 1,000 live births.[1] Among the CHDs, at least 50% of cardiac malformations are minor and have little long-term consequences for the developing child. The remaining 50% are major or critical CHDs. Most critical CHDs typically require early neonatal intervention due to closure of the ductus arteriosus in ductus-dependent lesions or inadequacy of the foramen ovale for oxygenation or cardiac output. It is well known that 25% of deaths due to CHD occur before the diagnosis of CHD in the first week of life.[2] Therefore, prenatal detection of critical CHDs may help to enhance perinatal counseling and delivery planning. Additionally, several reports have shown that an improved outcome is associated with prenatal diagnosis for specific types of major CHD.[3–5] Furthermore, many major cardiac malformations are associated with chromosome abnormalities, genetic syndromes or are part of a multiple malformation disorder.[6]

It is well known that 25% of deaths due to CHD occur before the diagnosis of CHD in the first week of life.[7] The concept of prenatal screening for CHD was introduced in the United Kingdom in 1986.[8] Following this introduction, the French study recommended incorporation of the four-chamber view of the heart into the routine obstetric scan performed at 18–22 weeks of gestation.[9] Consequently, a concentrated screening program was set up in 10 obstetric centers in the south-east Thames region in 1988, which showed that some forms of major CHDs might be detected by examining the four-chamber view of the heart during routine obstetric scan.[10]

Although the four-chamber view examination has been shown to be an effective method of detecting some of the critical CHDs in utero, this will not be able to detect all major forms of CHD such as double-outlet right ventricle (DORV), tetralogy of Fallot (TOF), transposition of the great arteries (TGA) and truncus arteriosus.[10–12] It was shown that adding ventricular outflow tract views to the four-chamber view of the heart at routine fetal anomaly scans at >18 weeks is the most effective technique to detect CHD prenatally.[13] Similarly, it has also been shown by that including examination of the arterial

connections of the heart would improve the detection rate.[14,15] Although the previous formal guidelines recommended including the outflow tract views to the four-chamber view when technically feasible, the current guidelines emphasize that the cardiac screening examination of the fetus should include both the four-chamber view and outflow tract views.[16–18] Additional cross-sectional views which include the three-vessel (3V) view and the three-vessel and trachea (3VT) view may show different aspects of the great vessels and surrounding structures. The 3V view which has been proposed by Yoo et al. includes the transverse view of the fetal upper mediastinum.[19] It can be easily obtained by moving the transducer cephalad from the four-chamber plane. However, the aortic arch and trachea were not included in this view. Later on, Yagel et al. demonstrated the clinical applicability of the 3VT view, a transverse view of the upper mediastinum slightly cranial to the original 3V view, to evaluate the anatomy of the major vessels in the mediastinum.[20] The 3VT view demonstrates the main pulmonary trunk in direct communication with the ductus arteriosus, a transverse section of the aortic arch and the superior vena cava (SVC) from left to right and a cross-section of the trachea is visualized posterior to and between the great arteries and SVC. Yagel et al. suggested the integration of all these views in a sequential segmental approach to the diagnosis of CHD.[21] The five axial views for the fetal heart examination include the transverse section of the upper abdomen to demonstrate the relation of aorta and inferior vena cava (IVC), four-chamber view of the heart, left ventricular outflow tract view, right ventricular outflow tract view along with 3V view and the 3VT view.

THE TECHNIQUE OF FETAL HEART EXAMINATION

Basic cardiac structures that should be evaluated for fetal heart examination are as follows:

1. *General inspection*:
   - Atrial and abdominal situs
   - Fetal laterality
   - Heart occupies a third of thoracic area
   - Cardiac apex points to left by 45° ± 20°
   - Four chambers present
   - Regular cardiac rhythm
   - No pericardial effusion
2. *Evaluation of atria (mandatory)*:
   - Two atria, approximately equal in size
   - Foramen ovale flap movement from right atrium to left atrium
   - Atrial septum primum present (near to crux)
   - At least two pulmonary veins entering left atrium
3. *Evaluation of ventricles (mandatory)*:
   - Two ventricles, approximately equal in size

- No ventricular wall hypertrophy
- Right ventricle with the moderator band is situated at the fetal right side and oval shaped left ventricle at the fetal left side
- Ventricular septum intact (apex to crux)

4. *Evaluation of atrioventricular junction and valves (mandatory)*:
   - Intact cardiac crux
   - Two atrioventricular valves open and move freely
   - Tricuspid valve leaflet more apical insertion on the ventricular septum compared to mitral valve
5. *Evaluation of left ventricular outflow tract view (mandatory)*:
   - Continuity between the interventricular septum and the anterior wall of aorta
   - Aortic valve should not be thickened and move freely
6. *Evaluation of right ventricular outflow tract view (mandatory)*:
   - Pulmonary artery (PA) should be crossing over the aorta
   - The bifurcation of the PA into both pulmonary branches should be seen
   - The pulmonary valve should not be thickened and move freely
7. *Evaluation of 3VT view (mandatory)*:
   - The main pulmonary trunk in direct communication with the ductus arteriosus, a transverse section of the aortic arch and the SVC should be aligned from left to right, anterior to posterior and from larger to smaller size
   - Both the aortic arch and the ductal arch should be located to the left of the trachea, in a "V"-shaped configuration
   - Any abnormality in vessel size, alignment, arrangement and number should be noted

We recommend the fetal heart examination to maximize the detection of heart anomalies during a second-trimester scan in all fetuses and this examination should be an integral part of routine prenatal care. The fetal heart examination should be used in the evaluation of all fetuses carrying low risk for CHD. This kind of approach will help also to identify fetuses at risk for genetic syndromes and provide useful information for patient counseling and obstetric management. The fetal heart examination should optimally be performed between 18 and 22 weeks' gestational age as recommended by many formal guidelines.[14–18] The examination should include a transverse sweep with cephalad movement of the transducer from the fetal abdomen (at the level of the standard abdominal circumference) through the four-chamber view and toward the upper mediastinum to visualize the left and right ventricular outflow tract views together with 3V and 3VT views. Real-time gray-scale sonography is the required modality for the fetal heart examination. Fetal heart examination is usually obtained when the cardiac apex is directed toward the anterior maternal wall. Maternal obesity, abdominal scars and

prone fetal position can make the fetal heart examination difficult due to acoustic shadowing.[16] It may be necessary to examine the patient at a different time if the heart is poorly visualized. Optimization of the images such as image magnification, frame rate, focus and frequency selection, gain intensity, is almost always recommended on a minimum basis. System settings should emphasize a high frame rate, with increased contrast and high resolution.

THE TECHNIQUE OF FETAL ECHOCARDIOGRAPHY

Suspected heart anomalies on the fetal heart examination will require more comprehensive evaluation using fetal echocardiography. One of the main goals for a fetal echocardiography is to confirm the presence or absence of cardiac disease. If this scan is abnormal, the examiner should characterize these abnormalities, make a differential diagnosis of the most probable defects, and identify fetuses that will require immediate medical or surgical intervention following delivery.[22] Real-time gray-scale sonography, complementary spectral and color Doppler ultrasound are important components of the fetal echocardiography.[22,23] Continuous-wave Doppler sonography may be sometimes necessary to quantify very high velocity flow across stenotic or incompetent valves such as in critical aortic stenosis. Occasionally, M-mode echocardiography, three-dimensional (3D) and four-dimensional (4D) ultrasonography and advanced techniques may be required to evaluate fetal cardiac function using measurements of ventricular shortening fraction, stroke volume, cardiac output, PR intervals, left and right ventricular Tei indices.[22–24]

Indications for fetal echocardiography are often based on a variety of parental and fetal risk factors for CHD.[23,25,26] However, most cases are not associated with any maternal or fetal risk factors. Common indications for fetal echocardiography include but are not limited to the following:[22,23]

1. *Maternal indications*:
   - Family (maternal, paternal, sibling) history of CHD
   - Metabolic disease (e.g. diabetes mellitus and phenylketonuria)
   - Teratogen exposure (e.g. retinoids and lithium).
   - Autoimmune antibodies, anti-Ro (SSA)/anti-La (SSB)
   - In vitro fertilization
2. *Fetal indications*:
   - Abnormal fetal heart examination (abnormal cardiac axis, abnormal cardiac position, abnormal four-chamber view, abnormal outflow tract views, abnormal 3V and 3VT views)
   - Abnormal heart rate or rhythm
   - Fetal chromosomal anomaly
   - Fetal extracardiac anomaly
   - Fetal hydrops

- Increased nuchal translucency
- Abnormal ductus venosus waveform
- Monochorionic twins

Among these indications, abnormal fetal heart examination carries the highest risk for the fetus for having a CHD. Therefore, the fetal heart examination should be offered to all second-trimester fetuses without any known risk factor for having a CHD.

Fetal echocardiography is commonly performed between 18 and 22 weeks' gestational age. Some forms of CHD may even be recognized during earlier stages of pregnancy. Fetal nuchal translucency measurement screening programs and the developing technology in ultrasound have created a new population of at-risk pregnancies that will be referred for early fetal echocardiography performed between the 12th and 16th week of pregnancy. Early detailed assessment of the fetal heart requires a high level of expertise in early anomaly scanning and fetal echocardiography. The use of transvaginal ultrasound and complementary Doppler ultrasound are the modalities which help to improve the detection rate for early fetal echocardiography. However, there still remains some limitations of fetal echocardiography in the first trimester and follow-up at second-trimester echocardiography may be required in most of the cases.

Fetal echocardiography is usually obtained when the cardiac apex is directed toward the anterior maternal wall—similar to routine fetal heart examination. Optimization of the images may again be needed such as image magnification, frame rate, focus, frequency selection, gain intensity, compound and harmonic imaging and Doppler-adjustments such as color-box scale, pulse-repetition frequency scale, wall motion filtering may be required to be established in every individual examination.

As a minimum, fetal echocardiography involves a thorough examination of the fetal heart by sequential segmental analysis.[27] The examiner should confirm anatomical relationships and functional flow characteristics through a systematic analysis of cardiac axis and situs, ventricular morphology, pericardial effusions, venous-atrial, atrioventricular and ventriculoarterial connections of the heart, size and relationships of the left and right ventricular outflow tracts, ductal and aortic arches, interventricular septum, atrial septum, atrial chamber size and foramen ovale and atrioventricular and semilunar valves flow across each heart connection, as seen with Doppler flow mapping.[22,23,27] These anatomical structures are usually evaluated using both transverse and sagittal views. Two-dimensional (2D) measurements of cardiac chambers or vessels are may also be required in order to interpret findings when compared against expected values. *Z*-scores may be used to improve the interpretation of cardiac measurements.[22,28]

Basic cardiac structures that should be evaluated for fetal echocardiography are as follows:

1. *Evaluation of four-chamber view (as described for fetal heart examination)*:
   - Evaluation of color Doppler sonography of systemic veins (including SVC and IVC and ductus venosus) (mandatory)
   - Evaluation of color Doppler sonography of pulmonary veins (mandatory)
   - Evaluation of color Doppler sonography of foramen ovale (mandatory)
   - Evaluation of color Doppler sonography of atrioventricular valves (mandatory)
   - 2D measurements of mitral valve and tricuspid valve (optional)
2. Evaluation of color Doppler sonography of atrial and ventricular septa
3. *Evaluation of the left ventricular outflow tract (as described for fetal heart examination)*:
   - Evaluation of color Doppler sonography of aortic valve (mandatory)
   - 2D measurements of aortic valve (mandatory)
4. *Evaluation of the right ventricular outflow tract (as described for fetal heart examination)*:
   - Evaluation of color Doppler sonography of PA valve (mandatory)
   - 2D measurements of PA valve (mandatory)
5. Evaluation of the 3VT view (as described for fetal heart examination)
6. Evaluation of short-axis views ventricles, aorta and PA (mandatory)
7. *Evaluation of aortic arch view (sagittal view optional if satisfactory 3VT views are obtained)*:
   - Evaluation of color Doppler sonography of aortic arch
8. *Evaluation of ductal arch view (sagittal view optional if satisfactory 3VT views are obtained)*:
   - Evaluation of color Doppler sonography of ductal arch
9. Evaluation of SVC and IVC views (mandatory).

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FETAL HEART

Screening, Diagnosis and Intervention

Salient Features

- Congenital anomalies are the greatest contributor to the infant death and congenital heart diseases (CHDs) account for up to 50% of these deaths
- Congenital heart diseases with the highest incidence of extracardiac abnormalities (>25%) included heterotaxy, single left ventricle and tricuspid atresia, hypoplastic left heart syndrome, and tetralogy of Fallot (TOF)
- Congenital heart disease is the most common type of birth defect, accounting for one-third of all major congenital anomalies
- For the clinicians and families, the major or critical cardiac defects carry the utmost importance since they may be either lethal or may require intervention in infancy of long-term follow-up
- About 1.35 million infants are born with CHD each year, with a worldwide occurrence of 7 per 1,000 live births
- Prenatal detection of critical CHDs may help to enhance perinatal counseling and delivery planning
- The fetal heart examination should be used in the evaluation of all fetuses carrying low risk for CHD
- The fetal heart examination to maximize the detection of heart anomalies during a second-trimester scan in all fetuses and this examination should be an integral part of routine prenatal care.

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