

ISUOG consensus statement: what constitutes a fetal echocardiogram?

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KEYWORDS: fetal echocardiography; guidelines; heart; pregnancy

Introduction

In 2006 the International Society of Ultrasound in Obstetrics and Gynecology (ISUOG) published practice guidelines for the sonographic screening of congenital heart disease (CHD) during the second trimester of pregnancy¹. This document described two levels for screening low-risk fetuses for heart anomalies. Firstly, a 'basic' scan should be performed by analyzing a four-chamber view of the fetal heart. Secondly, an 'extended-basic' scan further examines the size and relationships of both arterial outflow tracts. The term 'fetal echocardiogram' was also mentioned as a more detailed sonographic evaluation to be performed by specialists in the prenatal diagnosis of CHD. Although common indications and imaging techniques were briefly discussed, it was thought important to further explain how this advanced diagnostic procedure differs from the basic and extended-basic cardiac screening examinations.

A Fetal Echocardiography Task Force was subsequently asked to develop a standard description of 'what constitutes a fetal echocardiogram'. Our original goal was to develop guidelines for a detailed examination of the fetal heart that were based on the literature and a consensus opinion of an expert panel. However, we soon realized that this project was quite different from developing minimum practice guidelines for fetal cardiac screening. There are several imaging modalities that can be used to evaluate fetal heart anomalies, ranging from M-mode techniques and color Doppler sonography to the use of four-dimensional (4D) ultrasonography with spatiotemporal image correlation (STIC). Consequently, our original efforts to develop a minimum practice guideline for fetal echocardiography evolved into a consensus statement that covers the following topics:

1) guidance about timing of and indications for fetal echocardiography;

2) a definition of what constitutes a fetal echocardiogram;
3) a description of imaging modalities for fetal echocardiography;
4) the importance of multidisciplinary collaboration; and
5) documentation of diagnostic findings and conclusions.

One of the main goals for a fetal echocardiogram is to confirm the presence or absence of cardiac disease. If this scan is abnormal, the examiner should characterize these abnormalities, develop an accurate differential diagnosis of the most probable defects, and specifically identify fetuses that will require immediate medical or surgical attention after birth. Special emphasis should be directed toward fetuses that are likely to have ductal-dependent lesions^{2–4}.

When should a fetal echocardiogram be performed?

Fetal echocardiography can be performed at any time during the second trimester when cardiac anatomical details can be satisfactorily visualized. For example, a mother who is at a slightly increased risk for CHD – e.g. 2% against a background rate of 0.8% – should be electively scheduled for a detailed cardiac scan at some time between 18 and 22 weeks' menstrual age. These include mothers with a family history of CHD, maternal diabetes or exposure to teratogenic drugs, and fetuses that have had an increased nuchal translucency thickness measurement. Fetal cardiac abnormalities may occur in association with extracardiac anomalies and therefore a detailed cardiac scan may be indicated when such anomalies are detected^{5–7}. A mother who is particularly anxious because of a family history, perhaps loss of a previous child for example, or where the nuchal translucency measurement is ≥ 3.5 mm, may be offered a scan at or before 14 weeks' gestation, with a follow-up scan at 20–22 weeks⁸. If a fetus is suspected of having CHD at any scan, it should be seen as soon as possible, regardless of menstrual age.

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Others have suggested using a fetal echocardiogram as a component of the genetic sonogram, to evaluate fetuses at risk for chromosome abnormalities^{9,10}.

Who should be referred for fetal echocardiography?

The detection rate of structural heart anomalies will be higher with fetal echocardiography than with screening examinations. Women with recognized increased risks for fetal cardiac anomalies should be offered a more detailed scan. Risk factors for fetal cardiac anomalies can be categorized as fetal or maternal in origin. Some of the more common indications for fetal echocardiography are summarized in Table 1. Acquired cardiac lesions that become apparent later in life, even those of genetic origin such as Marfan's syndrome and hypertrophic subaortic stenosis, are not generally detectable by prenatal ultrasonography.

What constitutes a fetal echocardiogram during the second trimester of pregnancy?

Many experienced healthcare professionals, whether they are midwives, sonographers, obstetricians, perinatologists or radiologists, can evaluate the fetal heart with a high degree of diagnostic accuracy during an obstetric ultrasound evaluation. The echocardiogram can be individualized, depending on the nature of the suspected cardiac lesion. As a minimum, it involves a thorough examination of the four-chamber view, both arterial outflow tracts, three vessels and trachea view, and an assessment of pulmonary venous return¹¹⁻¹⁴. The examiner should confirm anatomical relationships and functional flow characteristics through a systematic analysis of the following areas:

- 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
- atrioventricular and semilunar valves
- flow across each heart connection, as seen with Doppler flow mapping.

These anatomical features are usually evaluated using transverse views, although sagittal scanning planes are also used as necessary. However, the specific views are less important as long as the relevant cardiac structures are being satisfactorily visualized. Color Doppler ultrasonography is an important component of the fetal echocardiogram. Although spectral Doppler ultrasonography is not essential, it can be used to further characterize the nature and severity of suspected flow disturbances^{15,16}. Continuous-wave Doppler sonography is sometimes necessary to quantify very high velocity flow across stenotic or incompetent valves. Occasionally, advanced techniques may be required to evaluate fetal cardiac function using measurements of ventricular ejection fraction, stroke volume, cardiac output, mechanical PR intervals, Tei indices, and ventricular strain parameters¹⁷⁻²⁰. Volume sonography allows a supplemental approach for analyzing complex cardiac lesions and may also provide important benefits for telemedicine, educational and research applications²¹⁻²⁴.

Although two-dimensional (2D) measurements of cardiac chambers or vessels are not always required for fetal echocardiography, they may help to interpret findings when compared against expected values. Quantitative measurements can be used to objectively interpret the severity of some cardiac lesions and as a basis for comparison over time. Measurements can be especially helpful where several different examiners are obtaining cardiac biometry throughout pregnancy. Standardized techniques must be used to maintain appropriate quality control. Some investigators have proposed the use of Z-scores to improve the interpretation of cardiac measurements²⁵⁻²⁷.

What type of imaging techniques are used for fetal echocardiography?

Suspected anomalies can be evaluated using several imaging modalities, although the examination can be individualized for specific cardiac anomalies (Table 2). Real-time gray-scale sonography and complementary Doppler ultrasound techniques (e.g. spectral and color) are often applied. M-mode echocardiography and Doppler ultrasonography are also important tools for the analysis of fetal cardiac dysrhythmias. Additional results, from three-dimensional (3D) and 4D ultrasonography,

Table 1 Common indications for fetal echocardiography

| <i>Maternal indications</i> | <i>Fetal indications</i> |
|--|---|
| First-degree relative of proband (mother or father) with CHD | Increased nuchal translucency thickness |
| Prior child with CHD born to mother and/or father | Abnormal ductus venosus waveform |
| Pre-existing metabolic disease | Abnormal fetal cardiac screening exam |
| Type 1 diabetes | Major extracardiac abnormality |
| Phenylketonuria | Abnormal fetal karyotype |
| Infections | Hydrops |
| Parvovirus B19 | Effusion |
| Rubella | Fetal cardiac dysrhythmias |
| Coxsackie | Persistent bradycardia or tachycardia |
| Autoimmune antibodies | |
| Anti-Ro (SSA) | |
| Anti-La (SSB) | |
| Teratogen exposure | |
| Retinoids | |
| Phenytoin | |
| Carbamazepine | |
| Lithium carbonate | |
| Valproic acid | |
| Paroxetine | |

CHD, congenital heart disease.

may further increase diagnostic confidence for the accurate characterization of some complex lesions.

The examiner must be completely familiar with the potential impact of technical factors for acquiring the best diagnostic heart images within the constraints of acceptable thermal and mechanical safety indices. Ultrasound signal gain, image magnification, compound imaging, and harmonic imaging should be optimized. Other important factors should also be considered, including menstrual age, maternal body habitus, fetal movement and position, and ultrasound transducer frequency. It may be necessary to wait until an optimal fetal position is obtained or even to reschedule the patient to be scanned on another day.

An accurate prenatal diagnosis is extremely important for healthcare professionals who will be counseling parents about the nature, severity, clinical management and prognosis of their unborn child. Some aspects of how to obtain information concerning the entire fetus may be well beyond the scope of a pediatric cardiologist working in isolation from the obstetric imaging specialist. The interpretation of some cardiac abnormalities can be challenging, and minor differences can substantially alter the surgical approach and clinical prognosis. For example, abnormal mitral valve attachments can preclude the switch operation for transposition of the great arteries, a remote or small VSD in a double-outlet right ventricle can only be treated by a one ventricle repair, or a restrictive atrial septum can change the immediate postnatal management and prognosis in fetuses with hypoplastic left heart syndrome. It should also be recognized that some cardiac lesions will evolve over time and may not be apparent until later in pregnancy²⁸.

Any cardiac abnormality must be interpreted in the context of the entire fetus and there should be a careful search for possible associations with extracardiac abnormalities. The clinical significance of fetal cardiac

disease can be quite different from what is found in a pediatric population. For example, prenatal recognition of trisomy 18 in a fetus with an inlet ventricular septal defect may dramatically alter management of the pregnancy or child after delivery. The rarity of individuals with both cardiac and extracardiac diagnostic skills supports the need for close teamwork to provide the most precise fetal prognosis. Only an accurate differential diagnosis will lead to appropriate prenatal counseling. For these reasons, we recommend multidisciplinary counseling for parents who expect to deliver an infant with heart disease. This may include collaboration between specialists in pediatric cardiology, maternal fetal medicine, fetal imaging, neonatology, genetics, cardiothoracic surgery, social work and psychology.

How should fetal echocardiogram results be documented?

There is currently no universal legal requirement to document, record and archive a fetal echocardiogram. Both stored images and a report to the managing/referring clinician are highly recommended and are mandatory in some jurisdictions. However, we suggest that those providing fetal echocardiography should have the facilities for recording still and moving images of the heart, and these should be available for future reference. Another alternative is to store volume data sets using 3D and 4D ultrasonography.

Acknowledgments

This consensus statement was developed under the auspices of the ISUOG Clinical Standards Committee. Chair: Dr W. Lee, Division of Fetal Imaging, William Beaumont Hospital, Royal Oak, Michigan, USA.

Appreciation is particularly extended to specialty consultants on the Fetal Echocardiography Task Force

Table 2 General recommendations for fetal echocardiography

| <i>Echocardiographic component</i> | <i>Recommendation</i> |
|---|--------------------------------|
| Structure | |
| Laterality, situs, cardiac connections, other anomalies | Mandatory |
| Biometry | |
| Normal heart | Optional |
| Abnormal heart | Desirable depending on anomaly |
| Cardiac rhythm (M-mode or spectral Doppler) | |
| Normal rhythm on two-dimensional ultrasonography | Optional |
| Abnormal rhythm on two-dimensional ultrasonography | Mandatory |
| Cardiac function (e.g. M-mode or Doppler) | |
| Normal function on two-dimensional ultrasonography | Optional |
| Abnormal function on two-dimensional ultrasonography | Desirable depending on anomaly |
| Color Doppler ultrasonography | Mandatory |
| Power Doppler ultrasonography | Optional |
| Spectral Doppler ultrasonography | |
| Normal cardiac anatomy | Optional |
| Abnormal cardiac anatomy | Desirable depending on anomaly |
| Continuous-wave Doppler ultrasonography | Optional |
| Tissue Doppler ultrasonography | Optional |
| Three- and four-dimensional ultrasonography | Optional |

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