THE INTERNATIONAL SOCIETY OF ULTRASOUND in Obstetrics & Gynecology

GUIDELINES

Cardiac screening examination of the fetus: guidelines for performing the 'basic' and 'extended basic' cardiac scan

INTRODUCTION

Congenital heart disease (CHD) is a leading cause of infant mortality, with an estimated incidence of about 4–13 per 1000 live births[1–3]. Between 1950 and 1994, 42% of infant deaths reported to the World Health Organization were attributable to cardiac defects[4]. Structural cardiac anomalies were also among the most frequently missed abnormalities by prenatal ultrasonography[5,6]. Prenatal detection of CHD may improve the pregnancy outcome of fetuses with specific types of cardiac lesions[7–11].

Prenatal detection rates have varied widely for CHD[12]. Some of this variation can be attributed to examiner experience, maternal obesity, transducer frequency, abdominal scars, gestational age, amniotic fluid volume, and fetal position[13,14]. Continuous training of health-care professionals based on feedback, a low threshold for echocardiography referrals and convenient access to fetal heart specialists are particularly important factors that can improve the effectiveness of a screening program[3,15]. As one example, the major cardiac anomaly detection rate doubled after implementing a two-year training program at a medical facility in Northern England[16].

The 'basic' and 'extended basic' cardiac ultrasound examinations are designed to maximize the detection of heart anomalies during a second-trimester scan[17]. These guidelines can be used for evaluating low-risk fetuses that are examined as a part of routine prenatal care[18–20]. This approach helps to identify fetuses at risk for genetic syndromes and provides useful information for patient counseling, obstetrical management and multidisciplinary care. Suspected heart anomalies will require more comprehensive evaluation using fetal echocardiography.

GENERAL CONSIDERATIONS

Gestational age

The fetal cardiac examination is optimally performed between 18 and 22 weeks' menstrual age. Some anomalies may be identified during the late first and early second trimesters of pregnancy, especially when increased nuchal translucency is identified[21–26]. Some countries, however, do not offer a medical insurance system for financial reimbursement of earlier scans at a time when more subtle cardiac defects may be undetectable or not present. Subsequent screening at 20–22 weeks' gestation is less likely to require an additional scan for completion of this evaluation, although many patients would prefer knowing about major defects at an earlier stage of pregnancy[27]. Many anatomic structures can still be satisfactorily visualized beyond 22 weeks, especially if the fetus is not prone.

Despite the well-documented utility of a four-chamber view, one should be aware of potential diagnostic pitfalls that can prevent timely detection of CHD[28–30]. Detection rates can be optimized by performing a thorough examination of the heart, recognizing that the four-chamber view is much more than a simple count of cardiac chambers, understanding that some lesions are not discovered until later pregnancy, and being aware that specific types of abnormalities (e.g. transposition of the great arteries or aortic coarctation) may not be evident from this scanning plane alone.

Technical factors

Ultrasound transducer

Higher-frequency probes will improve the likelihood of detecting subtle defects at the expense of reduced acoustic penetration. The highest possible transducer frequency should be used for all examinations, recognizing the trade-off between penetration and resolution. Harmonic imaging may provide improved images especially for patients with increased maternal abdominal wall thickness during the third trimester of pregnancy.[31]

Imaging parameters

Gray scale is still the basis of a reliable fetal cardiac scan. System settings should emphasize a high frame rate with increased contrast resolution. Low frame persistence, a single acoustic focal zone, and a relatively narrow image field should also be used for this purpose.

Zoom and cine-loop

Images should be magnified until the heart fills at least a third to one half of the display screen. If available, a

cine-loop feature can be used to assist the evaluation of ventricular septal defects and heart valve leaflets throughout the cardiac cycle.

BASIC CARDIAC EXAMINATION

The basic cardiac screening examination relies on a four-chamber view of the fetal heart[32,33]. This view should not be mistaken for a simple chamber count because it involves a careful evaluation of specific criteria (Figure 1). Major elements for a basic examination of the fetal heart are shown in Table 1. A normal heart is usually no larger than one-third the area of the chest. Some views may reveal a small hypoechogenic rim around the fetal heart that can be mistaken for a pericardial effusion. An isolated finding of this type usually represents a normal variation[34,35].

Cardiac rate and regular rhythm should be confirmed. The normal rate ranges from 120 to 160 beats per minute. Mild bradycardia is transiently observed in normal second-trimester fetuses. Fixed bradycardia, especially heart rates that remain below 110 beats per minute, requires timely evaluation for possible heart block. Repetitive heart rate decelerations during the third trimester can be caused by fetal distress. Occasional skipped beats are typically not associated with an increased risk of structural fetal heart disease. However, this finding may occur with clinically significant cardiac rate or rhythm disturbances as an indication for fetal echocardiography[36]. Mild tachycardia (> 160 beats per minute) can occur as a normal variant during fetal movement. Persistent tachycardia, however, should be further evaluated for possible fetal distress or more serious tachydysrhythmias.

The heart is normally deviated about $45 \pm 20°$ (2 standard deviations (SD)) toward the left side of the fetus (Figure 2)[37]. Careful attention should be given to cardiac axis and position because they can be

Table 1 Basic cardiac screening examination. Adapted with permission from: Lee W. American Institute of Ultrasound in Medicine. Performance of the basic fetal cardiac ultrasound examination. *J Ultrasound Med* 1998; **17**: 601–607

General	Normal cardiac situs, axis and position
	Heart occupies a third of thoracic area
	Majority of heart in left chest
	Four cardiac chambers present
	No pericardial effusion or hypertrophy
Atria	Atria approximately equal in size
	Foramen ovale flap in left atrium
	Atrial septum primum present
Ventricles	Ventricles about equal in size
	No cardiac wall hypertrophy
	Moderator band at right ventricular apex
	Ventricular septum intact (apex to crux)
Atrioventricular valves	Both atrioventricular valves open and move freely
	Tricuspid valve leaflet inserts on ventricular septum closer to the cardiac apex than to the mitral valve

easily evaluated even if the four-chamber view is not satisfactorily visualized[38]. Situs abnormalities should be suspected when the fetal heart and/or stomach is/are not found on the left side as well. Abnormal axis increases the risk of a cardiac malformation, especially involving the outflow tracts. This finding may be associated with a chromosomal anomaly. Some hearts are abnormally displaced from their usual position in the anterior left central chest. Abnormal cardiac position can be caused by a diaphragmatic hernia or space-occupying lesion, such as cystic adenomatoid malformation. Position abnormalities can also be secondary to fetal lung hypoplasia or agenesis.

Both atrial chambers normally appear similar in size and the foramen ovale flap should open into the left
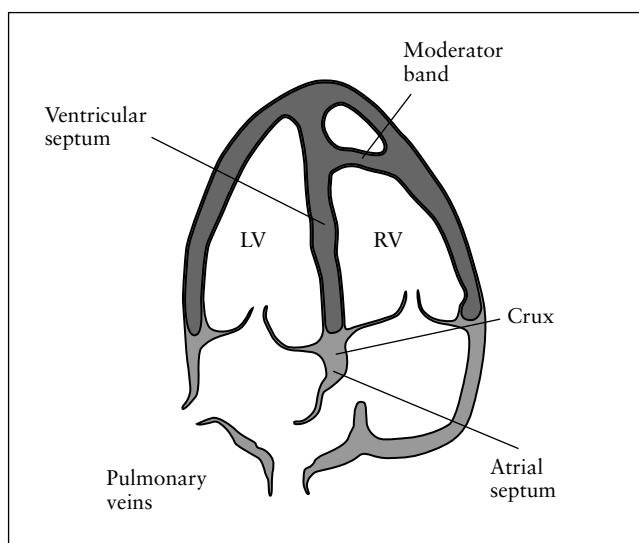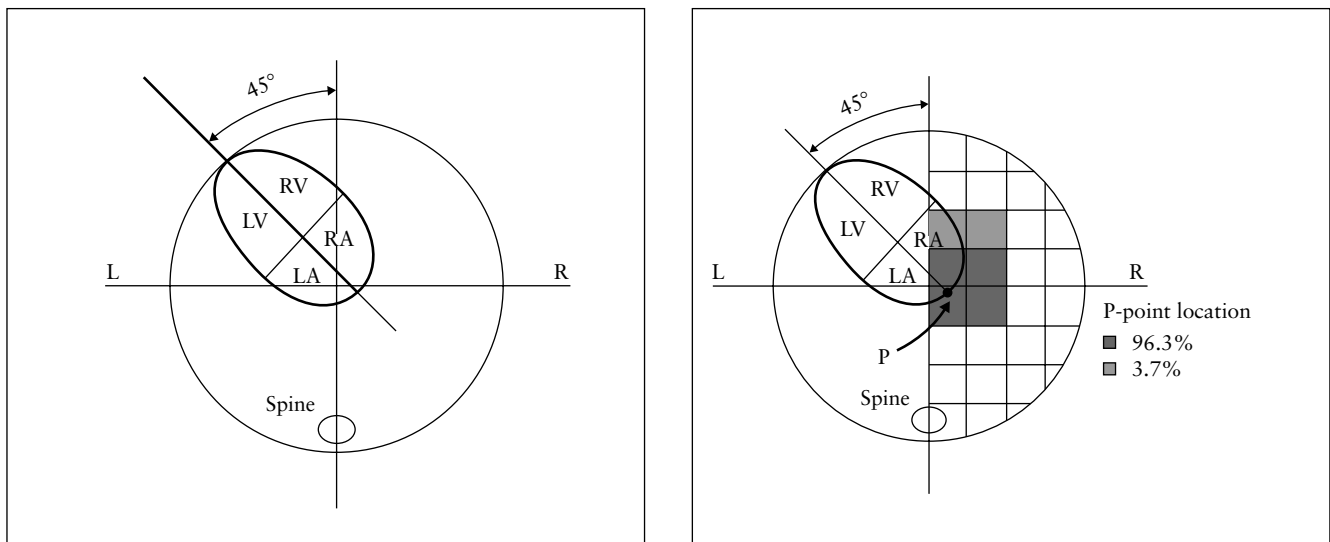

Figure 1 Four-chamber view of the fetal heart. Key components of a normal four-chamber view include an intact interventricular septum and atrial septum primum. There is no disproportion between the left (LV) and right (RV) ventricles. A moderator band helps to identify the morphologic right ventricle. Note how the 'offset' atrioventricular septal valve leaflets insert into the crux. Reproduced with permission from: Lee W. American Institute of Ultrasound in Medicine. Performance of the basic fetal cardiac ultrasound examination. *J Ultrasound Med* 1998; **17**: 601–607.

Figure 2 Fetal cardiac axis and position. The cardiac axis can be measured from a four-chamber view of the fetal heart. A line through the interventricular axis is extended to the posterior border of the heart to produce point P, the location of which can be used to define fetal cardiac position. LA, left atrium; LV, left ventricle; RA, right atium; RV, right ventricle. Adapted with permission from: Comstock CH. Normal fetal heart axis and position. *Obstet Gynecol* 1987; **70**: 255–259.

atrium. Pulmonary veins can often be seen entering the left atrium. However, their identification should not be considered a mandatory part of a basic cardiac screening examination. The lower rim of atrial septal tissue, called the septum primum, should be present. A moderator band helps to identify the morphologic right ventricle. Both ventricles should also appear similar in size without evidence for thickened walls. Although mild ventricular disproportion can occur as a normal variant, hypoplastic left heart syndrome and aortic coarctation are important causes of this disparity[39,40].

The ventricular septum should be carefully examined for cardiac wall defects from the apex to the crux. Septal wall defects may be difficult to detect when the transducer's angle of insonation is directly parallel to the ventricular wall. Under these circumstances, a defect may be falsely suspected because of acoustic 'drop-out' artifact. Small septal defects (1–2 mm) can be very difficult to confirm if the ultrasound imaging system fails to provide a sufficient degree of lateral resolution, especially if fetal size and position are unfavorable.

Two distinct atrioventricular valves (right-sided, tricuspid and left-sided, mitral) should be seen to open separately and freely. The septal leaflet of the tricuspid valve is inserted to the septum closer to the apex when compared to the mitral valve (i.e. normal offset). Abnormal alignment of the atrioventricular valves can be a key sonographic finding for cardiac anomalies such as atrioventricular septal defect.

EXTENDED BASIC CARDIAC EXAMINATION

If technically feasible, routine views of the outflow tracts should be attempted as part of an 'extended basic' cardiac screening examination. Evaluation of outflow tracts can increase the detection rates for

major cardiac malformations above those achievable by the four-chamber view alone[41,42]. Additional views to the basic cardiac examination are more likely to identify conotruncal anomalies such as tetralogy of Fallot, transposition of the great arteries, double outlet right ventricle, and truncus arteriosus.

An extended basic examination minimally requires that normal great vessels are approximately equal in size and that they cross each other at right angles from their origins as they exit from their respective ventricular chambers. Failure to confirm these findings in a well-visualized study warrants further evaluation.

Sonographic technique

The outflow tracts are usually obtained by angling the transducer toward the fetal head from a four-chamber view when the interventricular septum is tangential to the ultrasound beam (Figure 3). Another method for evaluating the outflow tracts has also been described for the fetus when the interventricular septum is perpendicular to the ultrasound beam[43]. This approach requires a four-chamber view of the heart where the probe is rotated until the left ventricular outflow tract is seen. Once this view is obtained, the transducer is rocked cephalad until the pulmonary arterial outflow tract is observed in a plane that is perpendicular to the aorta.

Yoo *et al.* have also described a 'three-vessel view' to evaluate the pulmonary artery, ascending aorta, and superior vena cava in relation to their relative sizes and relationships (Figure 4)[44,45]. Others have used this view to emphasize vascular relationships to the fetal trachea as well[46,47].

Left ventricular outflow tract

The left ventricular outflow tract (LVOT) view confirms the presence of a great vessel originating from the left
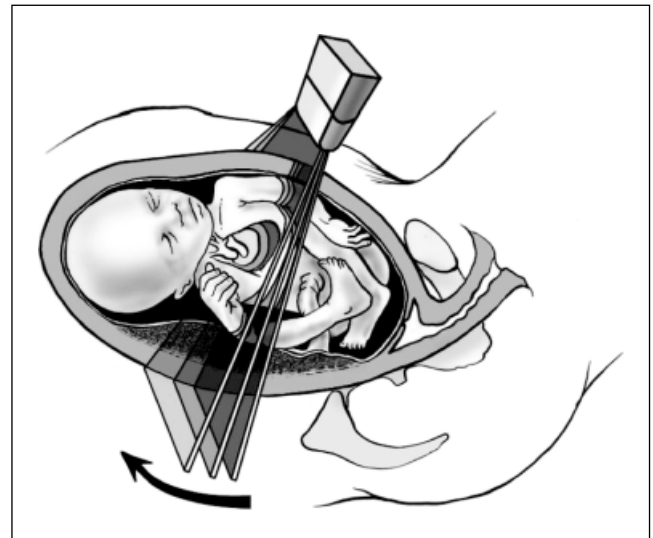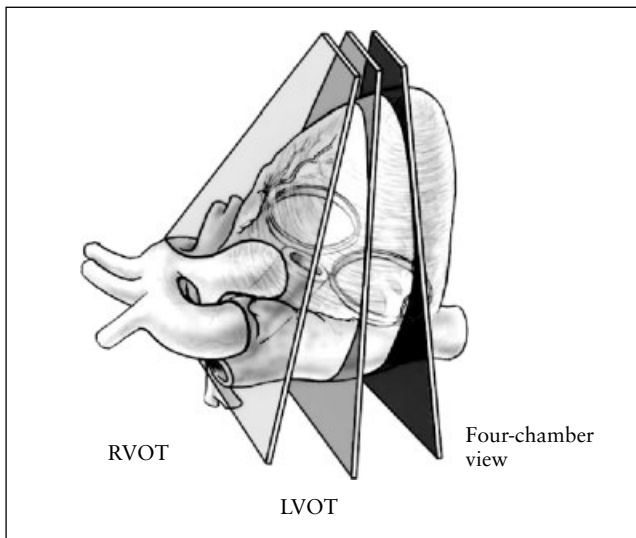
Figure 3 Fetal heart scanning technique. The four-chamber view of the heart is obtained from an axial scanning plane across the fetal thorax. Corresponding views of the left (LVOT) and right (RVOT) ventricular outflow tracts are found by angling the transducer toward the fetal head. Reproduced with permission from: Lee W. American Institute of Ultrasound in Medicine. Performance of the basic fetal cardiac ultrasound examination. *J Ultrasound Med* 1998; **17**: 601–607.

Figure 4 Three-vessel view of the fetal heart. This view demonstrates the relationship of the pulmonary artery (PA), aorta (Ao) and superior vena cava (SVC) in the upper mediastinum. Note the alignment as well as the relative sizes of the three vessels. The pulmonary artery has the largest diameter and is the most anterior vessel while the superior vena cava is the smallest and the most posterior. ant, anterior; Lt, left; post, posterior; Rt, right. Image courtesy of Dr J. S. Carvalho.

ventricle (Figure 5). Continuity should be documented between the anterior aortic wall and ventricular septum. The aortic valve moves freely and should not be thickened. When the LVOT is truly the aorta, it should even be possible to trace the vessel into its arch, from which three arteries originate into the neck. However, identification of these aortic arch vessels should not be considered as a routine part of the extended basic cardiac examination. The LVOT view may help to identify ventricular septal defects and conotruncal abnormalities that are not seen during the basic cardiac examination alone.

Right ventricular outflow tract

A view of the right ventricular outflow tract (RVOT) documents the presence of a great vessel from a morphologic right ventricle with a moderator band (Figure 6). The pulmonary artery normally arises from the right ventricle and courses toward the left of the more posterior ascending aorta. It is usually slightly larger than the aortic root during fetal life and crosses the ascending aorta at about a 70° angle just above its origin.

The pulmonary arterial valves move freely and should not be thickened. The RVOT can be confirmed as a pulmonary artery only if its distal end appears bifurcated, although this division cannot always be seen owing to fetal position. The distal pulmonary artery normally divides toward the left side into a ductus arteriosus that continues into the descending aorta. The right side branches into the right pulmonary artery.

A large obstetrical ultrasound survey of over 18 000 fetuses examined the standardized practice of incorporating a basic cardiac examination into the routine 30 minutes[48]. When technically feasible, an extended basic evaluation of the outflow tracts was also attempted. Of the studies that included an adequate four-chamber view, most of them (93%) were associated with satisfactory evaluation of the outflow tracts. Non-visualization rates were: left ventricular outflow tract, 4.2%; right ventricular outflow tract, 1.6%; both outflow tracts, 1.3%.

FETAL ECHOCARDIOGRAM

A fetal echocardiogram should be performed if recognized risk factors raise the likelihood of congenital heart disease beyond what would be expected for a low-risk screening population. Unfortunately, a high proportion of prenatally detectable cases of congenital heart disease
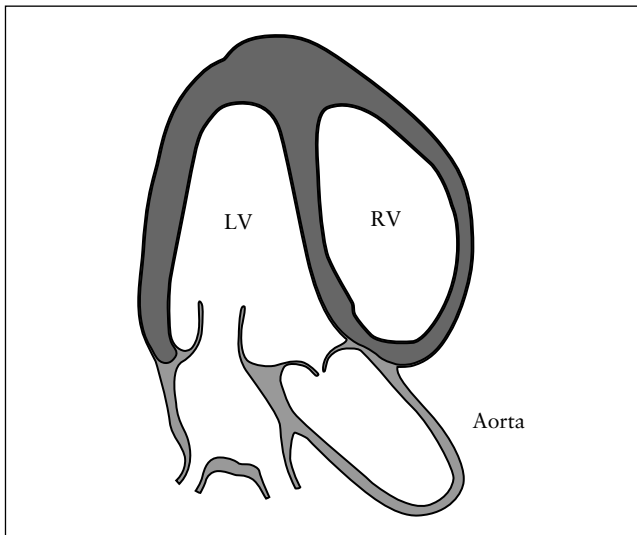
Figure 5 Left ventricular outflow tract (LVOT). This view demonstrates a great artery that exits the left ventricle. The aortic valve leaflets should be freely moving and not thickened. LV, left ventricle; RV, right ventricle. Reproduced with permission from: Lee W. American Institute of Ultrasound in Medicine. Performance of the basic fetal cardiac ultrasound examination. *J Ultrasound Med* 1998; **17**: 601–607.
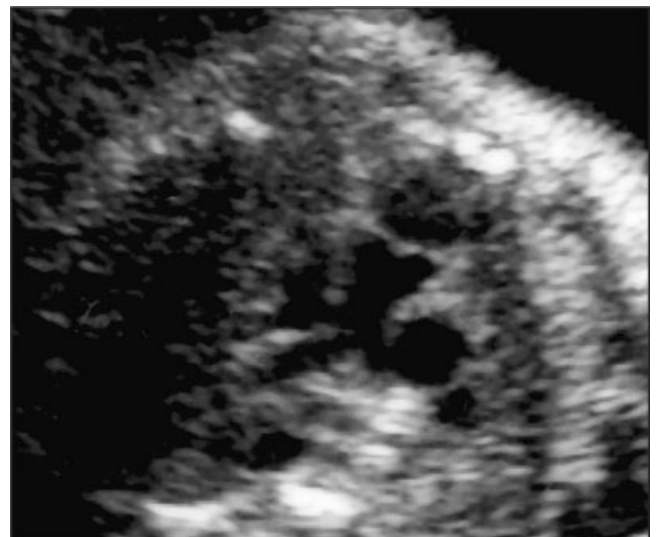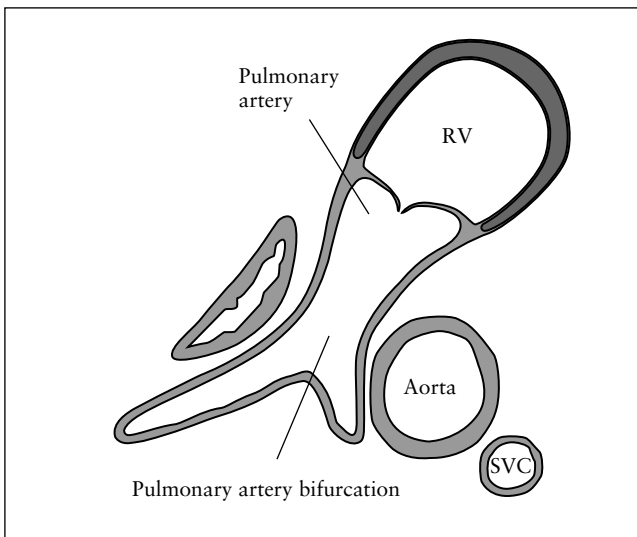

Figure 6 Right ventricular outflow tract (RVOT). This view emphasizes that a great vessel can be seen exiting the morphologic right ventricle (RV). The bifurcation is not always clearly seen in this manner. Note that the RVOT exits the ventricle at about 70° to the aortic outflow tract. Occasionally, the right superior vena cava (SVC) will be seen as the most posterior vessel. Adapted with permission from: Lee W. American Institute of Ultrasound in Medicine. Performance of the basic fetal cardiac ultrasound examination. *J Ultrasound Med* 1998; **17**: 601–607.

occurs in patients without any risk factors or extra-cardiac anomalies[49]. Specific details of this specialized procedure are not within the scope of this article. Healthcare practitioners, however, should be familiar with some of the reasons why patients could be referred for this comprehensive evaluation (Table 2)[50]. As an example, increased nuchal translucency of greater than 3.5 mm at 11–14 weeks' gestation, is an indication for a detailed cardiac evaluation even if this measurement subsequently falls into the normal range later in pregnancy[51–54].

Fetal echocardiography should be performed by specialists who are familiar with the prenatal diagnosis of congenital heart disease. In addition to information provided by the basic screening examination, a detailed analysis of cardiac structure and function may further characterize visceroatrial situs, systemic and pulmonary venous connections, foramen ovale mechanism, atrioventricular connections, ventriculoarterial connections, great vessel relationships and sagittal views of the aortic and ductal arches.

Advanced sonographic techniques can be used to study the heart. For example, Doppler ultrasonography can measure blood flow velocity or identify abnormal flow patterns across valves and within heart chambers. M-mode echocardiography also offers an important method for analyzing cardiac dysrhythmias, suspected ventricular dysfunction, and abnormal wall thickness.

Table 2 Common indications for fetal echocardiography

Maternal indications

Family history	First-degree relative of proband
Pre-existing metabolic disease	Diabetes
	Phenylketonuria
Maternal infections	Parvovirus B19
	Rubella
	Coxsackie
Cardiac teratogen exposure	Retinoids
	Phenytoin
	Carbamazepine
	Lithium carbonate
	Valproic acid
Maternal antibodies	Anti-Ro (SSA)
	Anti-La (SSB)

Fetal indications

Suspected fetal heart anomaly	
Abnormal fetal karyotype	
Major extracardiac anomaly	
Abnormal nuchal translucency	≥ 3.5 mm before 14 weeks' gestation
Fetal cardiac rate or rhythm disturbances	Persistent bradycardia
	Persistent tachycardia
	Persistent irregular heart rhythm

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