

Recommendations and guidelines for perinatal practice

Perinatal echocardiography: protocols for evaluating the fetal and neonatal heart*

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Abstract

This *Protocol for Evaluating the Fetal and Neonatal Heart* details the indications, views, and measurements to be obtained for both (1) the basic screening examination of the fetal heart (a necessary component of all complete fetal anatomy evaluations) and the specialty study called (2) echocardiography as applied to either the fetus or neonate, using 2D and Doppler ultrasound. While the purpose of the screening study is to detect or exclude the possibility of a cardiac abnormality, echocardiography attempts to diagnose the specific anatomic and physiologic disruption. Also emphasized is the value of a collaborative team approach to management of the fetus and its parents when a cardiovascular anomaly is present, in an effort to achieve a smoother transition from fetus to neonate across the continuum of perinatal care.

Keywords: Arteriosus; fetal echocardiography; neonatal echocardiography; protocol.

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Introduction

The World Association of Perinatal Medicine (WAPM) seeks to establish a set of guidelines and protocols for the performance of fetal and neonatal echocardiography (echo). This set of protocols is constructed upon the foundation of established evidence-based echo standards and guidelines published by other international organizations. These guidelines are comprehensive and we have attempted to include all aspects of evaluation of the cardiovascular system that may be performed. The primary purpose of these protocols is to offer the ultrasound practitioner methodologies that will allow the complete study of the anatomy and physiology of the fetal or neonatal heart and aid in the proliferation of knowledge and information that will lead to an appropriate diagnosis and possible treatment plan.

Fetal echocardiography is the ultrasonic evaluation of the human fetal cardiovascular system. The study is difficult and requires both experience and a fundamental knowledge of both normal and abnormal fetal cardiac anatomy and physiology. The ultrasonic study of the fetal heart occurs in two separate settings: (1) as a preliminary evaluation during the routine anatomic fetal survey, and (2) as a targeted comprehensive anatomic and physiologic study of the entire cardiovascular system. Both of these settings are screening tools for describing structural or functional congenital heart disease and heart failure, although targeted fetal echocardiography may also be considered diagnostic.

A) Basic fetal heart screening provides a set of information that leads to confidence in stating that the heart is within normal limits for specific parameters recorded as still images and observed during real-time ultrasound. There should be at least five transverse images of the heart and vasculature in the view and three sagittal images:

1. Abdominal cross-sectional view: the stomach should be seen on the left side of the fetus and just below the heart and diaphragm. The abdominal aorta should be on the left of the spine and the inferior vena cava on the right of the spine anterior to the level of the aorta.
2. Transthoracic four chamber view: the atria should be approximately equal in size. The flap of the foramen ovale (septum primum) should move into the left atrial chamber. The ventricles should be approximately equal in size with the tricuspid valve attaching to the

- septum slightly apical relative to the attachment of the mitral valve. The right ventricular apex should contain the moderator band. Both ventricular chambers should reach the apex of the heart and should squeeze equally.
3. Transthoracic four-chamber sweep to the Left Ventricular (LV) Outflow Tract (also known as the five-chamber view): sweeping toward the fetal neck, the aortic valve and ascending aorta are seen arising centrally from the four-chamber view with the ascending aorta aiming toward the right shoulder. Pulmonary veins should be seen in this view attached to the posterior wall of the left atrium.
 4. Transthoracic four-chamber sweep to the Right Ventricular (RV) Outflow Tract View: continuing the sweep toward the fetal neck, the pulmonic valve and the main pulmonary artery are seen arising at a 90° angle to the left of the ascending aorta and aiming toward the right shoulder. The bifurcation of the pulmonary artery and ductus arteriosus completes this view.
 5. The three-vessel view: continuing the sweep toward the fetal neck with a medial twist of the transducer, the short axis views of the main pulmonary artery, the ascending aorta and the superior vena cava are seen with their relationships to the trachea and esophagus. The main pulmonary artery diameter is slightly greater than the aorta which is slightly greater than the superior vena cava.
 6. Sagittal sweep view showing the superior vena cava, right atrium and inferior vena cava: both the superior vena cava (SVC) and the inferior vena cava (IVC) drain from a posterior position to a medial connection to the right atrium. The hepatic veins descend from an anterior position to the right atrial (RA) junction with the IVC.
 7. Sagittal sweep view showing the centrally arising aortic arch: twisting the transducer slightly to the fetal right shoulder reveals the aortic arch with the brachiocephalic vessels aiming to the fetal neck.
 8. Sagittal sweep view showing the ductal arch: straightening out the transducer and sweeping to the left of the spine reveals the anterior, superiorly arising pulmonary artery to ductus arteriosus to descending aorta portions of the ductal arch. The branch pulmonary arteries and the ductus arteriosus are seen aiming toward the fetal abdomen.
2. Size: in the transthoracic four-chamber view, the heart should occupy approximately 1/3 of the chest and seen such that “3 hearts” could fit into the thorax.
 3. Function: in the transthoracic four-chamber view, both ventricles should squeeze vigorously and equally toward the ventricular septum, such that the heart does not appear to “rock” within the chest.
 4. Rhythm: while observing the heart, there should be a relatively steady rate within the range of 120–180 beats per minute without early beats, blocked beats, or periods of bradycardia or tachycardia.
 5. Proportion: pairs of cardiac chamber structures should be seen as approximately the same size, right to left, including the atria, ventricles, atrio-ventricular (AV) valves, semilunar valves, great vessels and the SVC and IVC.

Any deviation from the expected normal images should elicit an offer to perform a comprehensive fetal echocardiogram.

B) Comprehensive fetal echocardiography or a targeted cardiovascular study is performed by a specialist (sonographer or physician) with advanced training, and builds upon the findings of the basic fetal heart screening by incorporating a segmental analysis to establish specific relationships between the vessels, valves and chambers and to evaluate the cause of any disproportion or abnormality in position, size, function or rhythm. These guidelines suggest what may be performed during a targeted cardiovascular study. The fetal echo uses real-time 2-dimensional ultrasound, m-mode measurements and various Doppler modalities to measure and provide information about structure, function and physiology of the entire cardiovascular system of the fetus. Color Doppler is used extensively to identify both direction of flow across vessels and valves and to identify any abnormal flow direction, absence of flow or turbulent flow. Routine indications for fetal echo include: abnormal cardiac views on a screening anatomy scan, family history of congenital heart disease, maternal diabetes or lupus, exposure to teratogens or infection, fetal arrhythmia, fetal size significantly less than dates, and increased nuchal translucency. The highest yield of congenital heart disease or congestive heart failure comes from studies performed for suspected cardiac structural abnormalities or abnormal fluid collections or hydrops fetalis. Any suggested diagnosis of fetal congenital heart disease requires post-natal confirmation. In targeted fetal cardiac studies, the basic tenets of screening are enhanced by specific measurements:

Taken as a whole, these images show normalcy in the following fashion:

1. Position: the axis of the fetal heart, made up of the interventricular and interatrial septa, should be seen in coming from the center of the chest into the left hemi thorax at a 45° angle to the sternum to spine axis in a transthoracic four-chamber view.
1. Position: cardiac structures: In an abdominal to cephalic transverse sweep through the fetal body, certain relationships should be maintained:

- a. The IVC should be to the right of the spine and the abdominal aorta should be to the left of the spine.
 - b. The intraabdominal umbilical vein should be medial and lead to the absolutely central ductus venosus found just to the right of the fundus of the stomach and immediately below the diaphragm.
 - c. Above the diaphragm, the RA should be seen to the right of center, the flap of the foramen ovale immediately above the ductus venosus and the LA seen left of center within the thorax.
 - d. As the sweep continues cephalad to reveal the four-chamber view, the insertion of the tricuspid valve on the ventricular septum should sit slightly apical relative to the mitral valve insertion on the ventricular septum.
 - e. The RV should be the anterior ventricular chamber demarcated by a moderator band near its apex.
 - f. The LV should be wedge shaped and occupy the ventricular apex within the left thorax.
 - g. The aorta should appear to arise centrally from the LV while being attached to the mitral valve posteriorly and to the IVS anteriorly. The ascending aorta aims toward the right fetal shoulder.
 - h. The main pulmonary artery should arise superiorly above the RV and to the left of the aorta, aiming toward the ductus arteriosus and to the left fetal shoulder.
2. Size: Cardio-thoracic ratio:
- a. The simple ratio of circumferences drawn as ellipses around the pericardium of the heart and around the thorax at the outer edge of the ribs and spine in the transthoracic four-chamber view should be approximately 50% (CC/TC ratio). Any measurement >55% before 34 weeks' gestation suggests cardiomegaly.
 - b. Cardiothoracic Area Ratio (CTAR) is performed as above in the four-chamber view but with the areas within the ellipses with the normal range from 0.25 to 0.35.
 - c. The measurement of diameters of the heart and chest at the level of the diaphragm in the coronal view (similar to the AP chest X-ray) should be approximately 0.50.
3. Size: Vessels and chambers: all cardiac structures can be measured either from a frozen 2D image or by m-mode technique. Size also refers to relative hypoplasia. A small tricuspid valve implies hypoplastic right heart that can result in pulmonary valve atresia later in gestation. Similarly, a small aortic valve may result in a coarctation of the aorta and a relatively hypoplastic left ventricle later in gestation.
4. Function: an m-mode set of measurements can be made with the beam set perpendicular to the lower half of the interventricular septum (IVS) such that the RV and LV have approximately equal sizes during diastole without imaging the AV valves. The function can be measured as a shortening fraction >30% (diastole – systole/diastole). The diastolic dimensions of the IVS, LV wall and RV wall measurements should measure approximately the same. Both the RV wall and IVS should move toward the LV wall in systole. An approximation of good LV function can be made when the systolic LV chamber dimension measurement is at least 1/3 less than the diastolic LV chamber dimension measurement. Another method of evaluation is the myocardial performance index (Tei Index) that uses the Doppler inflow and outflow patterns across in the left ventricular outflow tract to determine global function (MV closure to opening time – LV ejection time/LV ejection time).
 5. Rhythm abnormalities: both Doppler recordings of the arteries and proximal veins and m-mode images through an atrium and a ventricle or semilunar valve should be used to establish the atrio-ventricular conduction relationship. Timing of the AV conduction including the mechanical AV interval can be made from imaging the Doppler flow in the LVOT showing mitral inflow and LVOT outflow.
 6. Proportion: an abnormal measurement of any valve suggests structural congenital heart disease. Relative disproportion with cardiomegaly in an otherwise normally structured heart suggests impending heart failure which can lead to hydrops fetalis. Blood flow should be equal across both the AV valves and the semilunar valves by Doppler imaging. Abnormal flow patterns suggest either volume overload or downstream obstruction.
 7. Cardiomegaly: an increased cardio to thoracic ratio in what otherwise appears to be a normally structured heart suggests a volume load increase on the fetal heart. This may occur with extracardiac shunts from arterio-venous malformation (AVM) and/or fistulae detected as choriohemangioma, cord AV fistula, intraabdominal umbilical vein varix AVM, sacrococcygeal and neck teratomas, hepatic or vein of Galen AVM. Cardiomegaly with increased velocities across the AV and semilunar valves and ductus arteriosus can suggest fetal anemia. Persistent bradycardia or complete heart block may cause cardiomegaly and ventricular hypertrophy.
 8. Peripheral Doppler measurements:
 - a. Umbilical artery: high impedance flow patterns with a pulsatility index >1.75 or absent or reversed flow (adjusted for gestational age) during end diastole suggests increased afterload on the heart, most likely from placental insufficiency or partial placental abruption.

- b. Middle cerebral artery: increased diastolic flow with a pulsatility index < 1.50 suggests redistribution of cardiac output away from other organs to preserve cerebral perfusion. This in turn may lead to cardiomegaly from increased volume and then to dilation of the coronary arteries suggesting relative hypoxia. Increased diastolic MCA flow can also be seen with cerebral arteriovenous malformations. The MCA maximal velocity should be evaluated in line with the flow pattern ascertained by color or power Doppler. Any MCA flow > 55 cm/s at < 30 weeks' gestation suggests anemia, and the values should be interpreted according to gestational age.
 - c. Ductus venosus: an exaggerated A wave or reversal across the baseline in the flow pattern suggests either right heart obstruction or congestive heart failure, and, in IUGR fetuses, hypoxia or acidemia. In studies performed before 18 weeks gestation, an exaggerated A wave may be a marker for fetal aneuploidy.
 - d. Ductus arteriosus: the peak velocity in systole in the ductus arteriosus is the highest in the fetal cardiovascular system. There should be little to no flow during diastole. Velocities higher than 150 cm/s with increased diastolic flow indicate ductal constriction. Tricuspid valve regurgitation is associated with ductal constriction and occlusion and may be associated with non-steroidal anti-inflammatory medications.
9. Use of peripheral color Doppler: any time the heart appears large with relatively normal structures, the placenta, cord and fetal organs should be scanned with color pulsed or power Doppler to rule out AV malformations. These can be identified in the placenta as choriohemangiomas, and as AVMs in the umbilical cord, at the cord insertions sites, in the fetal liver, lungs, neck or head. Any external or internal teratoma should be considered a potential site for AV malformations.

The best gestational age period for performing fetal echocardiography is 18–22 weeks. Useful information can be identified earlier, even in the last third of the first trimester by transvaginal imaging. Congenital heart disease and findings of congestive heart failure should be monitored by repeat echocardiograms with counseling for delivery timing, location, and post-natal management. Poorly controlled diabetes mellitus warrants at least limited scheduled follow-up cardiac evaluation during the third trimester and into the newborn period to look for evidence of diabetic hypertrophic cardiomyopathy. When fetal echocardiography demonstrates structural or function abnormalities, parental counseling should begin immediately. This includes offering karyotype evaluation with additional study seeking 22q11 – micro-deletions. A

collaborative counseling team may include the obstetrician, maternal-fetal medicine specialist, neonatologist, pediatric cardiologist and cardiac surgeon, geneticist, social worker, and other specialists as indicated including clergy. They can provide pertinent information about the diagnosis, management options, and prognosis. This allows the parents to make autonomous informed decisions about the future of the pregnancy and also permit for ongoing planning for monitoring the fetus as well as the timing and location of delivery. Communication between the team members and the parents is essential to optimize the parents' understanding as well as the outcome of the pregnancy.

C) Neonatal echocardiography is performed for a variety of reasons, including neonatal evaluation of heart murmurs, persistent cyanosis, cardiomegaly on chest X-ray, tachypnea, an increased oxygen requirement, arrhythmia, and for known structural or functional congenital heart disease seen *in utero*. This evaluation uses the same modalities as fetal cardiovascular ultrasound, but because the newborn lungs are now filled with air, the imaging windows are limited and the study is restricted to proximal abdominal and thoracic vessels and cardiac structures. By far, the most common reason for neonatal echo is to evaluate a premature infant for the presence of a persistent patent ductus arteriosus either before or after medical treatment.

A standard neonatal echo protocol involves sonographic sweeps through the various cardiovascular structures to define spatial and functional parameters from specific acoustic windows including sub-xyphoid, apical, mid parasternal long- and short-axis, high parasternal, and suprasternal views. An m-mode set of measurements for LV function and RV size is performed from the mid parasternal view, using shortening fraction $> 28\%$ as normal. Pulsed Doppler is used across each valve with continuous wave Doppler used for velocities > 2 ms. While color Doppler is used extensively to identify normal and abnormal flow patterns, atrial septal defects, ventricular septal defects and abnormal or persistent connections between the great arteries, the jet streaming of abnormal color Doppler flow patterns allows correct alignment of the directional Doppler beam for estimation of pressures and gradients.

Sub-xyphoid sweeps

In newborns, all the basic questions of relative position, size, function and structure can be answered from sub-xyphoid or sub-costal imaging sweeps. Rhythm questions can be better answered by electrocardiography. Function can be globally assessed from below or measured from the parasternal images. These sweeps should be performed and then repeated with color Doppler:

1. Frontal sweep: with the image inverted on the screen, the frontal sweep begins with the transducer aimed

at the spine to show the position of the abdominal aorta and IVC. The transducer is then slowly swept upward through the heart and toward the anterior chest. This sweep identifies the relationships of the systemic and pulmonary veins, the AV valves, the morphologic positions and structure of the ventricles and outflow tracts and then the great vessels.

2. Hepato-left clavicular sweep: this sweep identifies the atrial septum and foramen ovale and continues through the ventricular septum to identify defects and functional wall movement.
3. Sagittal sweep: this sweep identifies other portions of the atrial and ventricular septa and the anterior-posterior relationships of the great vessels.
4. Right anterior oblique sweep: this sweep identifies the lower portion of the atrial septum, the aortic valve in cross section and the long axis of the RVOT to pulmonary artery anatomy.

Apical sweeps

In the newborn, most structural defects identified in the sub-xiphoid views can be confirmed and are sometimes more easily defined in the four-chamber views. The image remains inverted on the screen.

1. Four-chamber views: transducer sweep is horizontal. These views allow evaluation of the competency of all 4 valves by color Doppler and pulsed or continuous wave recordings. The integrity of the lower muscular and apical portions of the ventricular septum and the inflow portion of the posterior and anterior muscular septum may be best seen in these views.
2. Two-chamber views: transducer sweep is vertical. This view, identifying the left atrium and ventricle, best shows regional left ventricular wall motion and performance.

Para-sternal sweeps

LV function is best measured with m-mode techniques from these sweeps. They can also be used to better identify wayward jets of AV valve regurgitation. They may be the best views for muscular and sub aortic and sub pulmonary ventricular septal defects.

1. Long axis sweep: this sweep begins with the long axis of the left ventricle showing the apex to ascending aorta and mitral valve apparatus and is angled across to the tricuspid valve and then to the right ventricular outflow tract and back to the LVOT.
2. Short axis sweep: used for M-mode measurements, this sweep shows the regional wall motion function of all areas of the right and left ventricular free walls and the interventricular septum, the relative areas of hypertrophy and the relative positional anatomy of the papillary muscles. In normal situations the IVS

should always approximate the LV free wall during systole.

High parasternal sweeps

These views show the great vessel relationships, the aortic arch, the pulmonary artery bifurcation and the ductus arteriosus.

1. Transverse sweep: this sweep begins with the short axis of the ascending aorta and pulmonary bifurcation, and with downward deflection, the entry of the pulmonary veins into the left atrium. With upward deflection, the ascending aorta is followed cephalad to the branching of the first brachiocephalic vessel which indicates right or left aortic arch.
2. Sagittal sweep: this sweep shows the aortic arch and the patent ductus arteriosus. Color Doppler may be necessary to show the direction of flow in the PDA. Directional Doppler velocity in the PDA can determine pulmonary artery pressure in systole and diastole relative to aortic pressure.

Suprasternal sweeps

These rotational sweeping views may be used in term infants with a neck cushion elevation, but may be difficult to obtain in a premature infant because of transducer size. They are used to show the side of the aortic arch, the pulmonary artery branching and pulmonary venous connections.

Neonatal echocardiography is typically unscheduled and may be either urgent or emergent, unless it is requested to follow-up a prenatally-diagnosed cardiovascular abnormality. Diagnosis of a ductal dependent lesion should lead to immediate transfer to a center capable of neonatal cardiothoracic surgery and management after medical treatment to maintain ductal patency has begun. Evaluation of the cyanotic newborn should differentiate pulmonary versus cardiac pathology. "Routine" evaluation of the premature newborn should be performed to rule out congenital heart disease, evaluate the patency of the ductus arteriosus and evaluate the pulmonary arterial pressure. Counseling of the parents becomes the responsibility of the neonatologist who acts as the primary manager of the newborn until transfer to a pediatric cardiology/cardiothoracic unit or neonatal intensive care unit. A collaborative counseling team may include the neonatologist, pediatric cardiologist, cardiac surgeon, geneticist, social worker and other specialists as indicated. They can provide pertinent information about the diagnosis, management options, and prognosis. Communication between the team members and the parents is essential to optimize the parents' understanding and the plans for the management of their newborn.

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