AIUM Practice Parameter for the Performance of Fetal Echocardiography

Introduction

The American Institute of Ultrasound in Medicine (AIUM) is a multidisciplinary association dedicated to advancing the safe and effective use of ultrasound in medicine through professional and public education, research, development of clinical practice parameters, and accreditation of practices performing ultrasound examinations.

The *AIUM Practice Parameter for the Performance of Fetal Echocardiography* was developed (or revised) by the AIUM in collaboration with other organizations whose members use ultrasound for performing this examination(s) (see "Acknowledgments"). Recommendations for personnel requirements, the request for the examination, documentation, quality assurance, and safety may vary among the organizations and may be addressed by each separately.

This Practice Parameter is intended to provide the medical ultrasound community with recommendations for the performance and recording of high-quality ultrasound examinations. The parameter reflects what the AIUM considers the appropriate criteria for this type of ultrasound examination but is not intended to establish a legal standard of care. Examinations performed in this specialty area are expected to follow the parameter with recognition that deviations may occur depending on the clinical situation.

Congenital heart disease (CHD) is a leading cause of infant morbidity and mortality from birth defects, with an estimated incidence of 6 per 1000 live births for moderate to severe forms.[1,2] Accurate prenatal diagnosis offers potential clinical benefits with regard to infant outcomes, especially in those cases that are likely to require prostaglandin infusion to maintain patency of the ductus arteriosus.[3–6] Fetal echocardiography is broadly defined as a detailed ultrasound evaluation that is used to identify and characterize heart anomalies before delivery. This specialized diagnostic procedure is an extension of fetal cardiac screening parameters that have been previously described for the 4-chamber view and outflow tracts.[7] It should be performed only for a valid medical reason, and the lowest possible ultrasonic exposure settings should be used to gain the necessary diagnostic information. Although it is not possible to detect every abnormality, adherence to this parameter will maximize the probability of detecting and correctly diagnosing most cases of clinically significant CHD.

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This specialized diagnostic examination is an extension of the standard ultrasound fetal assessment described in the *AIUM-ACR-ACOG-SMFM-SRU Practice Parameter for the Performance of Standard Diagnostic Obstetric Ultrasound Examinations* and the American College of Obstetricians and Gynecologists Practice Bulletin No. 175: Ultrasound in Pregnancy.

Qualifications and Responsibilities of Personnel

Physicians interpreting or performing this type of ultrasound examination should meet the specified AIUM Training Guidelines in accordance with AIUM accreditation policies. Sonographers performing the ultrasound examination should be appropriately credentialed in the specialty area in accordance with AIUM accreditation policies. Physicians not personally performing the examination must provide supervision, as defined by the Centers for Medicare and Medicaid Services Code of Federal Regulations 42 CFR §410.32.

Request for the Examination

The written or electronic request for an ultrasound examination must originate from a physician or other appropriately licensed health care provider or under the provider's direction. The clinical information provided should allow for the performance and interpretation of the appropriate ultrasound examination and should be consistent with relevant legal and local health care facility requirements.

Indications

Clinical indications for fetal echocardiography are often based on a variety of parental and fetal risk factors for CHD.[1] However, most CHD cases are not associated with known fetal and/or maternal risk factors but, rather, are often suspected at the time of an anatomic ultrasound survey. For fetuses suspected of having an abnormal fetal heart at the time of a basic or detailed anatomic ultrasound examination, referral for fetal echocardiography is indicated, as the risk of significant disease is high. For pregnancies at low risk for CHD, cardiac screening ultrasound is primarily used to examine the fetal heart as a part of a standard second-trimester obstetric ultrasound examination. When risk is elevated above that of the general population, referral for fetal echocardiography may be indicated depending on the local resources, clinical settings, examiner availability, and results of a fetal cardiac screening evaluation.[7,8]

Although precise estimates of risk are outside the scope of this document, the following is a list of common fetal and maternal conditions associated with an increased risk of CHD[1]:

Fetal Factors

Fetal echocardiography is indicated if there is:

- Suspected cardiac structural anomaly
- Suspected abnormality in cardiac function
- Hydrops fetalis
- Persistent fetal tachycardia (heart rate > 180 beats per minute)
- Persistent fetal bradycardia (heart rate < 120 beats per minute) or a suspected heart block
- Frequent episodes or a persistently irregular cardiac rhythm
- Major fetal extracardiac anomaly
- Nuchal translucency of 3.5 mm or greater or at or above the 99th percentile for gestational age[9,10]
- Chromosomal abnormality by invasive genetic testing or with cell-free fetal DNA screening
- Monochorionic twinning

Fetal echocardiography may be considered if there is:

- Systemic venous anomaly (eg, a persistent right umbilical vein, left superior vena cava, or absent ductus venosus)[11,12]
- Greater-than-normal nuchal translucency measurement between 3.0 and 3.4 mm

Maternal or Familial Disease or Maternal Environmental Exposure

Fetal echocardiography is indicated if there is:

- Pregestational diabetes regardless of the hemoglobin A_{1C} level[13]
- Gestational diabetes diagnosed in the first or early second trimester

- In vitro fertilization, including intracytoplasmic sperm injection[14,15]
- Phenylketonuria (unknown status or a periconceptional phenylalanine level > 10 mg/dL)[16]
- Autoimmune disease with anti-Sjogren syndrome–related antigen A antibodies and with a prior affected fetus
- First-degree relative of a fetus with CHD (parents, siblings, or prior pregnancy)
- First- or second-degree relative with disease of Mendelian inheritance and a history of childhood cardiac manifestations
- Retinoid exposure
- First-trimester rubella infection

Fetal echocardiography may be considered if there is:

- Selected teratogen exposure (eg, paroxetine, carbamazepine, or lithium)
- Antihypertensive medication limited to angiotensin-converting enzyme inhibitors[17]
- Autoimmune disease with Sjogren syndrome–related antigen A positivity and without a prior affected fetus
- Second-degree relative of a fetus with CHD

Other Considerations

Limited data exist to support the utility of fetal echocardiography for the following isolated conditions, given minimal risk to the fetus and potential difficulty in implementing fetal echocardiography as routine in some clinical settings. A detailed fetal anatomic ultrasound examination (*Current Procedural Terminology* code 76811), which includes an evaluation of the fetal heart, may be appropriate instead, with fetal echocardiography performed only if an abnormality is suspected[8]:

- Obesity (body mass index ≥30 kg/m^2)[18,19]
- Selective serotonin reuptake inhibitor antidepressant exposure other than paroxetine
- Noncardiac "soft marker" for aneuploidy in the absence of karyotype information[20]
- Abnormal maternal serum analytes (eg, α-fetoprotein level)[21]
- Isolated single umbilical artery

Earlier studies may have previously suggested an increased risk of fetal heart disease for certain conditions or exposures that have not been borne out in larger follow-up studies. Fetal echocardiography in these cases is only indicated if the results of a detailed fetal ultrasound examination (*Current Procedural Terminology* code 76811) are abnormal.[8] These conditions include:

- Gestational diabetes diagnosed after the second trimester
- Warfarin exposure
- Alcohol exposure[22,23]
- Echogenic intracardiac focus
- Maternal fever or viral infection with seroconversion only[24]
- Isolated CHD in a relative further removed from second degree to the fetus

Specifications of the Examination

The following section describes required and optional elements for fetal echocardiography.

Technical Considerations

Fetal echocardiography is commonly performed between 18 and 22 weeks' gestational age, although some cardiac structures may be better visualized before or after this period. Various forms of CHD may also be recognized at early stages of pregnancy, including during the nuchal translucency examination.[25] *Optimal views are typically obtained when the cardiac apex is up (±45°) toward the transducer. However, evaluations of the atrial and ventricular septa and wall thickness are improved when the ultrasound beam is tangential or perpendicular to these structures.* Technical limitations (eg, maternal obesity, fetal position, and advanced gestation) may impede a detailed evaluation of cardiac anatomy due to poor penetration and posterior acoustic shadowing, especially during the third trimester.

Optimizing transducer placement on the maternal abdomen, applying adequate transducer pressure, and changing the maternal position are techniques that may improve fetal positioning and image quality. System settings should be adjusted with an emphasis on maintaining high frame rates (eg, using a narrow field of view, small imaging depth, single acoustic focus, and narrow color Doppler ultrasound region of interest box) with application of acceptable acoustic

output levels under the ALARA (as low as reasonably achievable) principle. The degree of image magnification should be adjusted so that the heart fills about one-third of the imaging sector display. In some cases, it may be necessary to reexamine the patient at a different time during gestation if the heart is poorly visualized due to technical factors.

Cardiac Imaging Guidelines: Basic Approach

The fetal echocardiogram is a detailed evaluation of cardiac structure and function. This assessment involves a sequential segmental analysis of 4 basic areas that include the situs, atria, ventricles, and great arteries and their connections.[26–28] This analysis includes an initial assessment of the fetal right/left orientation, followed by an assessment of the following segments and their relationships:

- Visceral/abdominal situs:
  - Position of the stomach, portal vein, descending aorta, and inferior vena cava in the axial view of the abdomen
  - Cardiac apex position and cardiac axis in the axial view of the chest
- Atria:
  - Situs
  - Systemic and pulmonary venous connections
  - Systemic venous anatomy, including normal/abnormal variations (eg, ductus venosus)
  - Pulmonary venous anatomy, noting normal connection of at least one right and one left pulmonary vein
  - Atrial anatomy (including the septum, foramen ovale, and septum primum)
- Ventricles:
  - Position
  - Atrioventricular connections (including offsetting of the mitral and tricuspid valves)
  - Right and left ventricular anatomy (including the septum)
  - Relative and absolute sizes
  - Systolic function
  - Pericardium
- Great arteries (aorta, main and branch pulmonary arteries, and ductus arteriosus):
  - Ventricular connections
  - Vessel size, patency, and flow (both velocity and direction)
  - Relative and absolute sizes of the aortic isthmus and ductus arteriosus
  - Pulmonary artery bifurcation
  - Position of the transverse aortic arch and ductus arteriosus relative to the trachea

  The following connections should be also evaluated as part of a segmental analysis:
- Atrioventricular junction: anatomy, size, and function (stenosis or regurgitation) of atrioventricular (eg, mitral and tricuspid or common atrioventricular) valves
- Ventriculoarterial junction: anatomy, size, and function (stenosis or regurgitation) of semilunar (eg, aortic and pulmonary or truncal) valves, including assessments of both the subpulmonary and subaortic regions

Grayscale Imaging (Required)

Key scanning planes can provide useful diagnostic information about the fetal heart (Figures 1–3).[29–32] The evaluation should include the following anatomic regions, including the upper abdomen for situs, cardiac chambers, valves, vessels, and pericardium:

- Four-chamber view, including pulmonary veins
- Left ventricular outflow tract
- Right ventricular outflow tract
- Branch pulmonary artery bifurcation
- Three-vessel view (including a view with pulmonary artery bifurcation and a more superior view with the ductal arch)
- Short-axis views ("low" for ventricles and "high" for outflow tracts)
- Long-axis view (if clinically relevant)
- Aortic arch
- Ductal arch
- Superior and inferior venae cavae

Color Doppler Ultrasound (Required)

Color Doppler ultrasound should be used to evaluate the following structures for potential flow disturbances[33–35]:

- Systemic veins (including superior and inferior venae cavae and ductus venosus)
- Pulmonary veins (at least two: one right vein and one left vein)
- Atrial septum and foramen ovale
- Atrioventricular valves

Figure 1. Representative scan planes for fetal echocardiography include an evaluation of the 4-chamber view (1), left and right arterial outflow tracts (2 and 3, respectively), two variants of the 3-vessel view, one demonstrating the main pulmonary artery bifurcation (4) with another more superior plane that demonstrates the ductal arch (5), and the 3-vessel and trachea view (6). Not all views may be seen from a single cephalic transducer sweep without some minor adjustments in the position and orientation of the transducer due to anatomic variations and the fetal lie. Asc Ao indicates ascending aorta; DAo, descending aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle; and Tr, trachea.

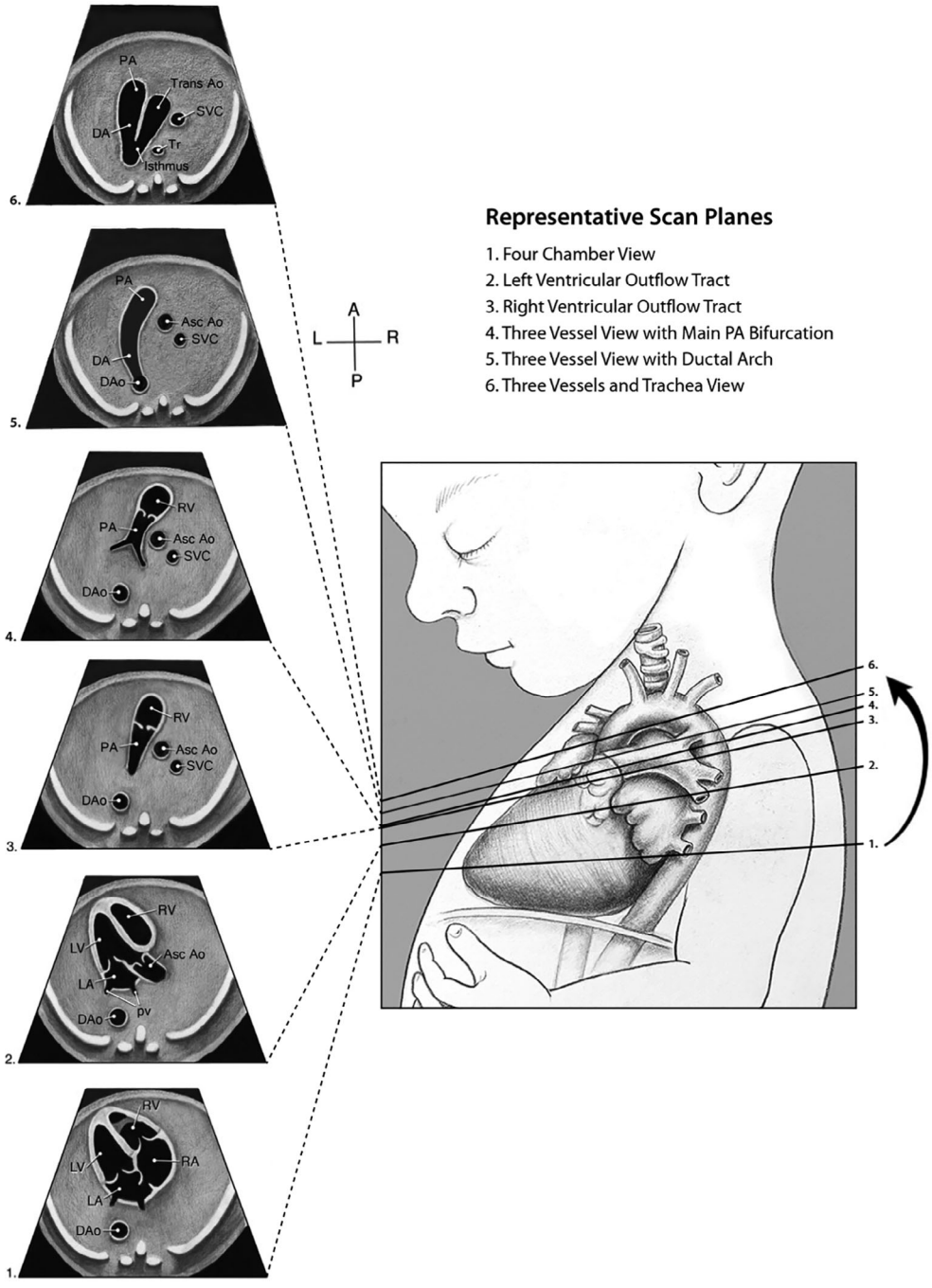


Figure 2. Sagittal views of the superior and inferior venae cavae (1), aortic arch (2), and ductal arch (3). The scan angle between the ductal arch and thoracic aorta ranges between 10° and 19° during pregnancy,[53] as illustrated by the 4-chamber view diagram (lower right). Ao indicates descending aorta; Ao Root, aortic root; DA, ductus arteriosus; IVC, inferior vena cava; LA, left atrium; LV, left ventricle; PV, pulmonary valve; RA, right atrium; RPA, right pulmonary artery; RV, right ventricle; and SVC, superior vena cava.

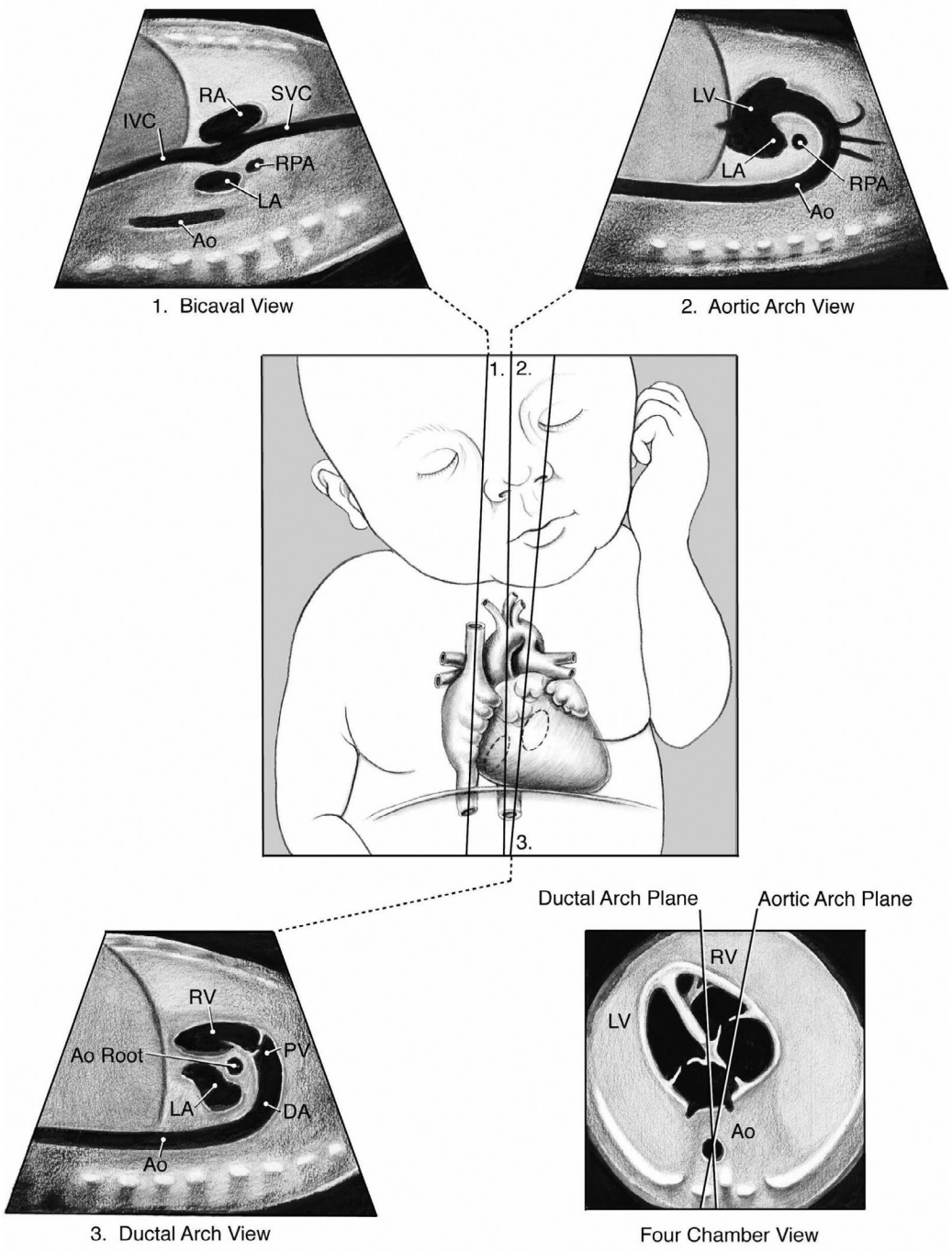
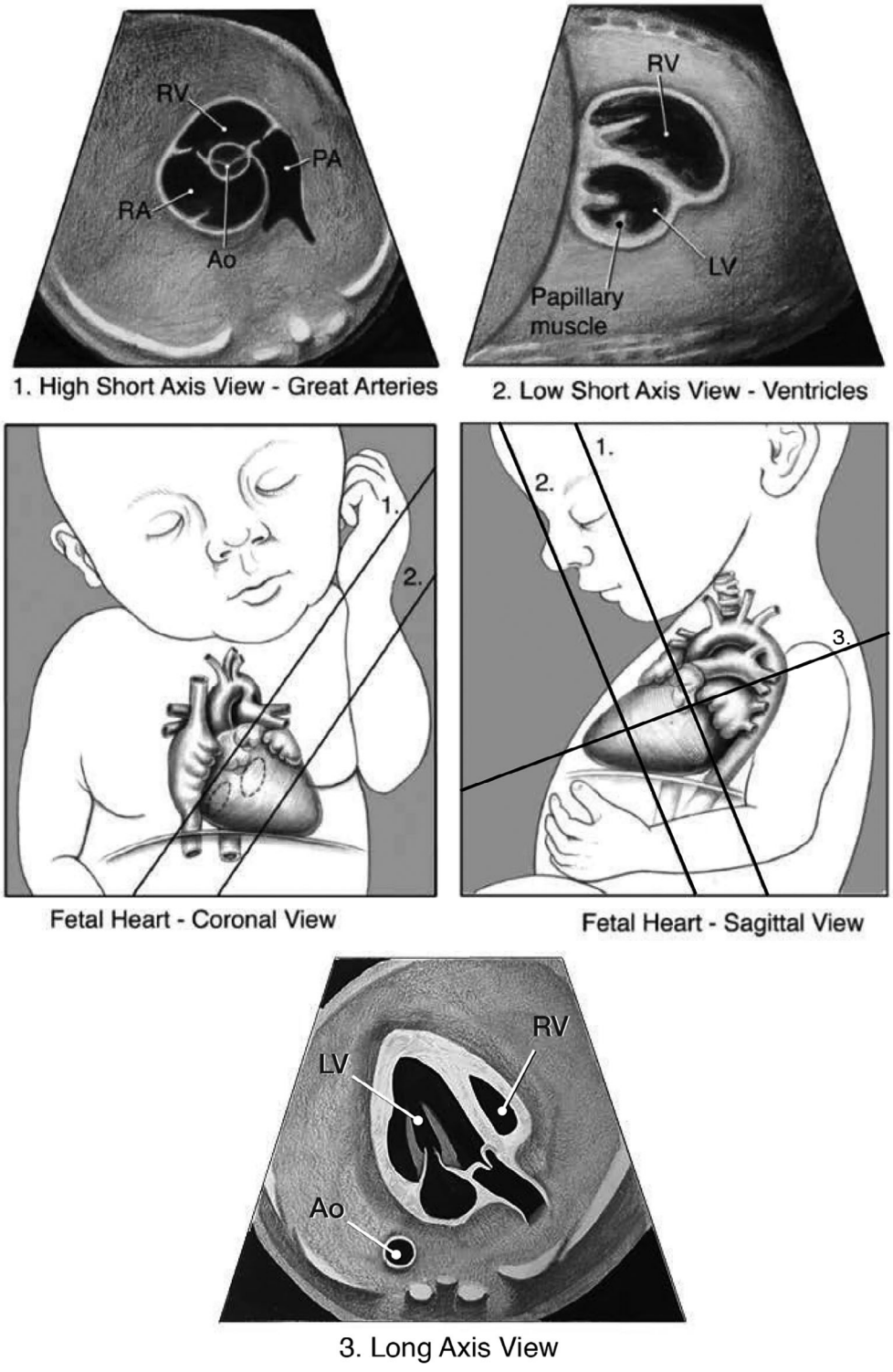


Figure 3. High short-axis view (1), low short-axis view (2), and long-axis view (3) of the fetal heart. Ao indicates aortic valve; LV, left ventricle; PA, pulmonary artery; RA, right atrium; and RV, right ventricle.



- Ventricular septum
- Semilunar valves
- Ductal arch
- Aortic arch

***Pulsed Doppler Ultrasound* (Required)**
Pulsed-wave Doppler ultrasound should be used to evaluate the following:

- Right and left atrioventricular valves
- Right and left semilunar valves
- Pulmonary veins (at least two: one right vein and one left vein)
- Ductus venosus
- Suspected structural or flow abnormality on color Doppler imaging

Pulsed-wave Doppler ultrasound may also be clinically relevant for evaluating the ductus arteriosus, systemic veins (eg, superior vena cava, inferior vena cava, and hepatic veins), aortic arch at the isthmus, branch pulmonary arteries, middle cerebral artery, and umbilical artery or vein.

***Heart Rate and Rhythm Assessment* (Required)**
Documentation of the heart rate and rhythm should be made by cardiac cycle length measurements obtained by the Doppler technique or M-mode interrogation. A normal fetal heart rate at midgestation is 120 to 180 beats per minute. If bradycardia or tachycardia is documented, or if the rhythm is noted to be irregular, a detailed assessment of atrial and ventricular contractions should be performed.

***Cardiac Biometry* (Required)**
Normal ranges for fetal cardiac measurements have been published as percentiles and z scores that are based on gestational age or fetal biometry.[36–41] Individual measurements should be determined from 2-dimensional (2D) images and include the following parameters:

- Aortic and pulmonary valve annulus in systole (absolute size with comparison of left- to right-sided valves)
- Tricuspid and mitral valve annulus in diastole (absolute size with comparison of left- to right-sided valves)

Additional fetal cardiac biometry can also be performed for suspected structural and functional cardiac anomalies, including but not limited to:

- Right and left ventricular lengths
- Aortic arch and isthmus diameter measurements from the sagittal arch view or 3-vessel and trachea view with comparison of the aortic isthmus to ductus arteriosus
- Main pulmonary artery and ductus arteriosus measurements
- End-diastolic ventricular diameter just inferior to the atrioventricular valve leaflets in the short- or long-axis view
- Thickness of the ventricular free walls and interventricular septum in diastole just inferior to the atrioventricular valves
- Cardiothoracic ratio
- Additional measurements if clinically relevant, including:
  - Systolic ventricular dimensions (short or long axis views)
  - Transverse atrial dimensions
  - Branch pulmonary artery diameters

***Cardiac Function Assessment* (If Clinically Relevant)**
Right and left heart function should be qualitatively assessed. Signs of cardiomegaly, atrioventricular valve regurgitation, and hydrops fetalis are key circulatory findings that can indicate fetal cardiac dysfunction and should be noted if present. If compromised function is suspected, a quantitative assessment of heart function may be performed using several measures, including but not limited to fractional shortening,[42,43] ventricular strain,[44,45] and the myocardial performance index.[46]

***Complementary Imaging Strategies* (If Clinically Relevant)**
Other adjunctive imaging modalities, such as 3- and 4-dimensional ultrasound, have been used to evaluate anatomic defects and to quantify fetal hemodynamic parameters, such as cardiac output.[47] Adjunctive Doppler modalities include tissue and continuous wave Doppler ultrasound.[48–50] Additional fetal cardiac functional assessment modalities such as tricuspid annular plane systolic excursion[51] and the sphericity index[52] have also been reported, although their role in clinical care should be considered investigational at this time.

Specific Documentation of Heart Views

In addition to still-frame acquisition and storage documenting the grayscale, color, and pulsed Doppler views, the following motion video clips should be obtained for routine documentation. If there are suspected structural or functional cardiac anomalies, additional motion video clips should be considered. Required clips include:

- Axial sweep from the stomach to the upper mediastinum, to include the 4-chamber view, arterial outflow tracts, as well as the 3-vessel and trachea view
- Four-chamber view: 2D and color Doppler ultrasound
- Left ventricular outflow tract view: 2D and color Doppler ultrasound
- Right ventricular outflow tract view: 2D and color Doppler ultrasound
- Three-vessel and trachea view: 2D and color Doppler ultrasound
- Sagittal view of the aortic and ductal arches: 2D and color Doppler ultrasound

Documentation

Accurate and complete documentation is essential for high-quality patient care. Written reports and ultrasound images/video clips that contain diagnostic information should be obtained and archived, with recommendations for follow-up studies if clinically applicable, in accordance with the *AIUM Practice Parameter for Documentation of an Ultrasound Examination*.

A complete evaluation can only be accomplished if acquisition of analog recordings or digital motion video clips, in conjunction with still images, is used as a standard part of every fetal echocardiogram.

Equipment Specifications

An ultrasound examination of the fetal heart should be conducted using an ultrasound system equipped with the ability to obtain M-mode, pulsed Doppler, and power/color Doppler images. Sector, curvilinear, and endovaginal transducers are used for this purpose. Use of 3- and 4-dimensional technology and continuous wave Doppler ultrasound is optional if clinically relevant. The transducer should be adjusted to operate at the highest clinically appropriate frequency, using acoustic power settings that follow the ALARA principle.

A trade-off exists between image resolution and beam penetration. With modern equipment, fetal imaging studies performed from the anterior abdominal wall can usually use frequencies that vary between 1 and 9 MHz, depending on the body habitus of the patient. Furthermore, acoustic shadowing and the maternal body habitus may limit the ability of higher-frequency transducers from providing greater anatomic detail for the fetal heart. Endovaginal scans should be performed using frequencies of 5 MHz or higher.

Quality and Safety

Policies and procedures related to quality assurance and improvement, safety, infection control, and equipment performance monitoring should be developed and implemented in accordance with the AIUM Standards and Guidelines for the Accreditation of Ultrasound Practices.

ALARA Principle

The potential benefits and risks of each examination should be considered. The ALARA principle should be observed for factors that affect the acoustic output and by considering the transducer dwell time and total scanning time. Further details on ALARA may be found in the current AIUM publication *Medical Ultrasound Safety*.

Fetal Safety

Diagnostic ultrasound studies of the fetus are generally considered safe during pregnancy (Conclusions Regarding Epidemiology for Obstetric Ultrasound).

Diagnostic ultrasound should be performed only when there is a valid medical indication (Prudent Use in Pregnancy). The lowest possible ultrasonic exposure setting should be used to gain the necessary diagnostic information under the ALARA principle.

The output display standard, an on-screen real-time display of acoustic output, should be visible and monitored for the thermal index (TI) and mechanical index (MI). The dwell time should be kept to a minimum. A TI for soft tissue (TIs) should be used before

10 weeks' gestation, and a TI for bone (TIb) should be used at or after 10 weeks' gestation when bone ossification is evident (*Recommended Maximum Scanning Times for Displayed Thermal Index (TI) Values*).

Doppler ultrasound may be used to answer specific clinical questions. Spectral pulsed Doppler ultrasound is associated with higher energy output and should be used judiciously as part of an evaluation for anomalies. The promotion, selling, or leasing of ultrasound equipment for making "keepsake fetal videos" is considered by the US Food and Drug Administration to be an unapproved use of a medical device. Use of a diagnostic ultrasound system for keepsake fetal imaging, without a physician's order, may be in violation of state laws or regulations.

Infection Control

Transducer preparation, cleaning, and disinfection should follow manufacturer recommendations and be consistent with the AIUM Guidelines for Cleaning and Preparing External- and Internal-Use Ultrasound Transducers Between Patients, Safe Handling, and Use of Ultrasound Coupling Gel.

Equipment Performance Monitoring

Monitoring protocols for equipment performance should be developed and implemented in accordance with the *AIUM Standards and Guidelines for the Accreditation of Ultrasound Practices*.

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