GUIDELINES AND STANDARDS

Guidelines and Recommendations for Performance of the Fetal Echocardiogram: An Update from the American Society of Echocardiography

Check for updates

Anita J. Moon-Grady, MD (Co-Chair), Mary T. Donofrio, MD (Co-Chair), Sarah Gelehrter, MD, Lisa Hornberger, MD, Joe Kreeger, ACS, RCCS, RDCS, Wesley Lee, MD, Erik Michelfelder, MD, Shaine A. Morris, MD, Shabnam Peyvandi, MD, Nelangi M. Pinto, MD, Jay Pruetz, MD, Neeta Sethi, MD, John Simpson, MD, Shubhika Srivastava, MD, and Zhiyun Tian, MD, *San Francisco and Los Angeles, California; Washington, District of Columbia; Ann Arbor, Michigan; Edmonton, Alberta, Canada; Atlanta, Georgia; Houston, Texas; Salt Lake City, Utah; Durham, North Carolina; London, United Kingdom; Wilmington, Delaware; and Philadelphia, Pennsylvania*

Keywords: Prenatal diagnosis, Congenital heart disease, Fetal arrhythmia, Hydrops, Cardiomyopathy

This document is endorsed by the Fetal Heart Society, the Society of Pediatric Echocardiography, and the American Institute of Ultrasound in Medicine.

TABLE OF CONTENTS

From the University of California, San Francisco, San Francisco, California (A.J.M.-G., S.P.); Children's National Hospital, Washington, District of Columbia (M.T.D.); the University of Michigan, Ann Arbor, Michigan (S.G.); the University of Alberta, Edmonton, Alberta, Canada (L.H.); Children's Healthcare of Atlanta, Atlanta, Georgia (J.K.); Baylor College of Medicine, Houston, Texas (W.L., S.A.M.); Emory University/Children's Healthcare of Atlanta, Atlanta, Georgia (E.M.); Texas Children's Hospital, Houston, Texas (S.A.M.); the University of Utah, Salt Lake City, Utah (N.M.P.); Children's Hospital of Los Angeles, Los Angeles, California (J.P.); Duke University, Durham, North Carolina (N.S.); Evelina London Children's Hospital, London, United Kingdom (J.S.); Dupont/Nemours Children's Hospital, Wilmington, Delaware (S.S.); and Children's Hospital of Philadelphia, Philadelphia, Pennsylvania (Z.T.).

Attention ASE Members:

Login at www.ASELearningHub.org to earn continuing medical education credit through an online activity related to this article. Certificates are available for immediate access upon successful completion of the activity and post-work. This activity is free for ASE Members, and $40 for nonmembers.

Abbreviations
2D = Two-dimensional
3VT = Three-vessel-and-trachea view
3VV = Three-vessel view
AHA = American Heart Association
AIUM = American Institute of Ultrasound in Medicine
APVS = Absent pulmonary valve syndrome
ASE = American Society of Echocardiography
AV = Atrioventricular
CHD = Congenital heart disease
CW = Continuous-wave
d-TGA = D-transposition of the great arteries
DTI = Doppler tissue imaging
HLHS = Hypoplastic left heart syndrome
IVC = Inferior vena cava
l-TGA = L-transposition of the great arteries
LV = Left ventricular
MI = Mechanical index
PW = Pulsed-wave
RV = Right ventricular
SV = Single ventricle
SVC = Superior vena cava
TI = Thermal index
TOF = Tetralogy of Fallot
TV = Tricuspid valve
TVD = Tricuspid valve disease
VSD = Ventricular septal defect
VTI = Velocity-time integral

I. INTRODUCTION

In 2004 the American Society of Echocardiography (ASE) guidelines and standards for the performance of fetal echocardiography were published.[1] The writing group, commissioned by the Pediatric Council of the ASE, provided a review of the medical literature as well as a summary of expert consensus to create a document that was much needed and served the fetal and pediatric echocardiography community for almost two decades. However, the field continues to advance rapidly, and since the time of that publication, a great deal of new information has become available.

Over the past 5 years, several professional groups, including the American Heart Association (AHA),[2] the International Society of Ultrasound in Obstetrics and Gynecology,[3] and the American Institute of Ultrasound in Medicine (AIUM),[4] have published updated guidelines and standards documents on fetal cardiac evaluation. Since the publication of the ASE's document in 2004, much of what has been learned is due to an intense desire for multidisciplinary collaboration, pooling multicenter data and expertise from subspecialists including pediatric cardiologists, radiologists, obstetricians, maternal fetal medicine specialists, and sonography practitioners, all important stakeholders in the care of fetal patients with cardiovascular disease. Ongoing research and collaboration have supported efforts toward a better understanding of fetal physiology and disease processes and progression. This has led to significant improvements in fetal imaging and in clinical practice and outcomes for patients.

Given that the future of fetal cardiology includes multidisciplinary cooperation,[5] there is new information available that should be summarized and relayed in a revised ASE statement to maintain the best practice for fetal echocardiography and fetal and perinatal cardiovascular care across disciplines. The purpose of this document is to provide updated recommendations for the performance and interpretation of fetal echocardiography in the detection, classification, risk assessment, and perinatal care planning of the pregnancy in which the fetus has cardiovascular disease. This document replaces the 2004 ASE guideline mentioned earlier.

II. INDICATIONS

Indications for performance of fetal echocardiography (Table 1), the gold standard test to diagnose congenital heart disease (CHD) in the fetus, have evolved over the years. The discussion in this section hinges on the quality and sensitivity of prenatal ultrasound screening for cardiac abnormalities, the basic or detailed obstetric anatomic ultrasound performed in the second trimester of pregnancy. As is the case with most screening tests, a basic or detailed anatomy ultrasound to detect a congenital anomaly is offered to a population of healthy individuals with a low probability of disease. However, in contrast to other medical screening tests, there is tremendous regional variability in the performance of the obstetric anatomic ultrasound as a screening tool to detect CHD. Thus, decisions about indications for referral for fetal echocardiography are challenging at the population and health policy level. In discussion of disease detection within a population-based health care delivery system, it is useful to understand that recommendation for referral for definitive testing depends on both the rate of disease in the population being subjected to screening and on the performance of the screening test.

A true assessment of individual baseline risk for CHD on the basis of factors in the pregnant person and/or fetus is difficult. In the United States population-based estimates of even the overall *incidence* of fetal CHD are not clear because of lack of data on termination and fetal demise and lack of standardized recording of data in the prenatal period. Thus, most of the population-based estimates of CHD are based on birth records and are prevalence estimates, ranging from 0.3% to 1.2%.[13] In most situations, referral for fetal echocardiography is recommended when the likelihood of fetal CHD is greater than about 3 times the background population risk, or >1%. Patients with a likelihood of disease lower than the background level after normal results on screening ultrasound would not need additional testing with fetal echocardiography, but the postscreening likelihood depends on both the pretest probability of fetal CHD and the sensitivity and specificity of the screening method used (Figure 1). *If screening suggests a cardiac abnormality, there is a high likelihood that the results of fetal echocardiography will be abnormal, and therefore fetal echocardiography must be performed to confirm and refine the diagnosis.* Where to set the threshold for referral for fetal echocardiography despite normal screening results, though, depends upon how well the screening ultrasound performs in the particular community and the tolerance for missed diagnoses. Over the past decade, standard obstetric cardiac screening guidelines have expanded from a four-chamber view alone to include imaging of both outflow tracts[3] and the three-vessel view (3VV) and three-vessel-and-trachea view (3VT) *when technically feasible.*[15] Obtaining additional views increases the sensitivity of obstetric cardiac screening,[16] but local practice and experience influence the degree of improvement. The presence of certain risk factors (maternal, fetal and/or familial) may increase the pretest probability of CHD in offspring enough so that fetal echocardiography is recommended regardless of normal findings on obstetric ultrasound. Practice patterns may ultimately differ on the basis of regional assessments of said sensitivity and specificity of detecting CHD with obstetric ultrasound in a particular region or community as well as considerations of cost-effectiveness and resource allocation as

discussed below. Additional important factors may include the health care system, insurance challenges, and available resources that can exacerbate existing health care disparities and access to care, as well as the skill of screening operators and requirements for and access to continuing education and skill assessments. In situations in which fetal echocardiography is a limited resource, or there is increased risk associated with patient travel to perform fetal echocardiography, a triage system that considers local screening detection rates such as that described in the ASE coronavirus disease 2019 guidelines[17] can be used. If available, telemedicine use should be considered.

Costs related to fetal echocardiography on the basis of different indications are also a critical consideration for both the patient and the health care system. Costs and cost-effectiveness related to prenatal screening for CHD vary significantly depending on the strategy used. Published cost-effectiveness analyses in fetal CHD have focused primarily on screening strategies for high-risk conditions such as

Table 1 Potential indications for fetal echocardiography

	ASE 2023 recommendation	AIUM 2020[4]	AHA 2014[2]*
Maternal factors (absolute risk)[†]			
Pre–gestational diabetes (3%-5%)	Is indicated	Is indicated	I (indicated)
Gestational diabetes diagnosed after second trimester (<1%)	Not indicated	Not indicated	III (no benefit)
Phenylketonuria (12%-14%)	Is indicated	Is indicated	I (indicated)
Autoimmune disease: SSA/SSB positive (1%-5%)[‡]	Is indicated	Is indicated	IIa (probably indicated)
In vitro fertilization (1.1%-3.3%)	May be considered[§]	Is indicated	IIa (Probably indicated)
Maternal infection: rubella (3%-4%)	Is indicated	Is indicated	I (indicated)
Family history of CHD: first-degree relative (3%-20%)[¶]	Is indicated	Is indicated	I (indicated)
Family history of CHD: second-degree or more distant relative (<2%)[‖]	Not indicated	May be indicated	IIb (may be indicated)
Obesity (BMI > 30 kg/m^2) (1-2%)	Not indicated	Not indicated	—
Retinoids (8%-20%)	Is indicated	Is indicated	I (indicated)
ACE inhibitors (3%)	May be considered[§]	May be indicated	IIa (probably indicated)
Paroxetine (3%)	May be considered[§]	May be indicated	IIb (may be indicated)
Other selective serotonin reuptake inhibitors (1%-2%)[6,7]	Not indicated	Not indicated	III (no benefit)
Anticonvulsants (1%-2%)	Not indicated	May be indicated	IIb (may be indicated)
Lithium (1%-2%)	Not indicated	May be indicated	IIb (may be indicated)
Warfarin (<1%)[8]	Not indicated	Not indicated	III (no benefit)
Fetal factors identified during screening (absolute risk)			
Fetal hydrops (15%-20%)[9]	Is indicated	Is indicated	I (indicated)
Extracardiac anomaly (20%-45%)[10,11]	Is indicated	Is indicated	I (indicated)
Chromosomal abnormalities (10%-90%)	Is indicated	Is indicated	I (indicated)
Monochorionic twinning (2%-10%)	Is indicated	Is indicated	I (indicated)
Nuchal translucency 3.0-3.4 mm (~3%)	May be considered[§]	May be indicated	IIa (probably indicated)
Nuchal translucency ≥3.5 mm (6%-60%)	Is indicated	Is indicated	I (indicated)
Single umbilical artery in isolation (1.2%-1.8%)[12]	Not indicated	Not indicated	IIb (may be indicated)

ACE, Angiotensin-converting enzyme; *BMI*, Body mass index.

Fetal echocardiography is indicated in the setting of abnormal results on screening ultrasound of the heart regardless of additional risk factors; below are recommended indications for fetal echocardiography on the basis of a priori risk from previously published guidelines and the current document. In the "ASE 2023" (present document) recommendations, a classification of "not indicated" assumes a normal cardiac screening result at second-trimester obstetric anatomy scan. Reference data reviewed in Donofrio et al.[2] unless otherwise noted.

*Using "classification of recommendations"[2]: I = procedure should be performed; IIa = it is reasonable to perform procedure; IIb = procedure may be considered; III = harm/no benefit.

[†]"Absolute risk": baseline CHD risk assessment independent of ultrasound findings.

[‡]Prior affected child with complete heart block, risk increases to 11% to 19%.

[§]Decision to refer for fetal echocardiography despite negative results on screening ultrasound should be based on the sensitivity and specificity of CHD detection in local community screening practices.

[¶]Can be up to 50% with genetic disorders with Mendelian inheritance.

[‖]Not indicated unless a genetic disorder with Mendelian inheritance.
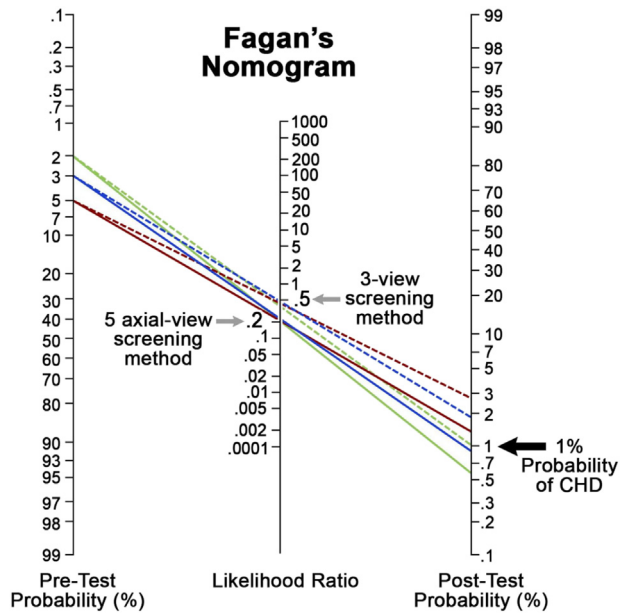
Figure 1 Pre- and posttest probability of fetal CHD following normal results on screening ultrasound. The likelihood of fetal CHD being ≤1% (background risk) following screening ultrasound depends on the pretest probability of fetal CHD and the sensitivity and specificity of the screening method used. Using a three-view screening method *(dotted lines)*, pretest probability of fetal CHD needs to be ≤2% for postscreening ultrasound probability of fetal CHD to drop to 1% *(dotted green line)*. Using a five-axial-view screening method *(solid lines)*, pretest probability of fetal CHD needs to be ≤3% for postscreening ultrasound probability of fetal CHD to drop to 1% *(solid blue line)*. With a pretest probability of fetal CHD of 5%, neither method results in a postscreening ultrasound probability of fetal CHD of <1% *(red lines)*. Negative likelihood ratio calculations are based on sensitivity and specificity reported by Bak *et al.*[14]

pregestational diabetes and in vitro fertilization. Some conclude that a well-executed obstetric scan is cost effective compared with referring these higher risk pregnant patients directly for fetal echocardiography. However, this conclusion depends heavily on the quality of the screening ultrasound and its interpretation, and the cost-benefit ratio of screening is related to the sensitivity, specificity, and posttest probability as described earlier.[18,19] Conversely, when considering the low-risk population, multiple studies have shown that a high-quality screening ultrasound performed in general obstetrical practice is most cost effective, while the use of universal fetal echocardiography is not. Inclusion of additional views, as shown in a recent analysis of a five-view screening approach,[14] is also cost effective by increasing sensitivity for very little additional cost. The optimal strategy for screening and referral thus depends on society's willingness to pay for increased effectiveness. CHD is more common and lethal than trisomy 21,[13] for which prenatal screening programs have been found to have an average cost of $27,000 to $78,000 per defect detected.[20,21] In comparison, costs per CHD detected average only $38,000 to $47,000 using current second-trimester ultrasound screening,[22] although this results in only an estimated 50% to 70% detection rate. Understanding the trade-off between costs and effectiveness in CHD screening in the low- and high-risk populations and improving obstetric screening may provide further evidence for performing fetal echocardiography, consideration of alternative strategies, and/or

focusing efforts for effective implementation in practice. A summary of key studies in this area is provided in Table 2.

With this information as a background, maternal and fetal risk factors with accompanying absolute risk for CHD in the fetus are presented in Table 1; also included are consensus recommendations for fetal echocardiography referral as well as a comparison with published recommendations from other organizations, including the AIUM[4] and the AHA.[2] Again, fetal echocardiography is indicated if the results of screening ultrasound of the heart are abnormal, regardless of additional risk factors, as the risk for true CHD approaches 40% to 70% in these cases.[25]

Key Points

- Fetal echocardiography is indicated in the setting of abnormal results on screening ultrasound of the heart regardless of the presence or absence of additional risk factors.
- There is wide variability in the performance of midgestation obstetric ultrasound. It is thus reasonable to continue the current strategy of incorporating both maternal and fetal risk factors in the decision to refer for fetal echocardiography even when the basic or detailed obstetric anatomic scan appears normal.
- In situations in which fetal echocardiography is a limited resource or increased risk is involved with obtaining testing, a triage system that considers local screening detection rates and the option of use of telemedicine may be considered.

III. THE FETAL ECHOCARDIOGRAPHIC EXAMINATION

Specific training requirements and maintenance of competency are outside the scope of this document. However, guidelines have been developed by the American College of Cardiology in conjunction with the AHA and ASE and are endorsed by our group.[1] It is recommended by the AHA[2] and endorsed by our group that "only well-trained or experienced pediatric cardiologists, maternal-fetal medicine specialists, obstetricians, or radiologists who have acquired the appropriate knowledge base and skills should supervise and perform fetal echocardiograms" and that ongoing quality improvement efforts be documented.

A. Timing of Examination, Equipment, and Image Storage

Timing. Optimal timing for performance of comprehensive transabdominal fetal echocardiography is 18 to 22 weeks' gestation. Advances in ultrasound technology have allowed fetal heart evaluation to be performed as early as 12 to 14 weeks' gestation.[26] Such early scans are particularly suited for fetuses at high risk for cardiac abnormality. Early scans should be repeated later in the second trimester, even if the initial imaging results are normal.[27]

Serial assessment of the abnormal heart is usually done at 2- to 8-week intervals (depending on lesion and clinical concerns) until about 34 to 36 weeks' gestation. Assessment of the fetal heart nearer to term is targeted to situations in which findings may influence immediate postnatal management, such as assessment of atrial restriction in d-transposition of the great arteries (d-TGA) or hypoplastic left heart syndrome (HLHS). If the results of midtrimester fetal echocardiography are normal, subsequent assessment is generally unnecessary, however is recommended at up to 2- to 4-week intervals when there is a concern for development of a progressive cardiac lesion such as cardiomyopathy, left heart obstruction, or significant valve stenosis or regurgitation. Precise follow-up interval timing should be decided upon by the fetal cardiology and obstetric teams caring for the patient, as progression may affect in utero care, delivery timing, and postnatal management.

Table 2 Summary of key cost-effectiveness studies related to screening for CHD in standard-risk and certain high- or intermediate-risk conditions

Paper	Population	Strategies compared	Effectiveness measure	Conclusions	Considerations
Odibo et al. (2006)[18]	Pre–gestational diabetes	1. No FE 2. Selective FE after abnormal scan 3. FE for Hgb A$_{1c}$ > 7 4. Universal FE	QALYs	Selective FE abnormal scan most cost effective	Model assumed detailed OB anatomy scan sensitivity 61% and specificity 99% Termination probability 50% Did perform sensitivity analyses
Chung et al. (2021)[23]	In vitro fertilization	1. Selective FE after abnormal scan 2. FE for ICSI only 3. FE for all IVF	QALYs CHD detected	ICER for (2) and (3) $2 million to $5 million per QALY Only selective FE after abnormal scan was cost effective	Same assumptions of OB ultrasound above Sensitivity of FE assumed 88%
NHS (2008)[24]	Standard-risk pregnancies	1. Standard screen with four-chamber 2. Screen with four-chamber + outflow views	QALYs	33% more CHD detected ~$24,000 per QALY for outflow	Assumed 36% of detected with d-TGA
Pinto et al. (2014)[22]	Standard-risk pregnancies	1. Standard screen with four-chamber 2. Screen with four-chamber + outflow views 3. Tiered with NT 4. Four-chamber + outflow performed by MFM 5. Universal FE Also considered referral strategy	CHD detected	Four-chamber + outflow with referral to MFM most cost effective For an additional $580 per CHD detected, four-chamber + outflow with referral to cardiology increased CHD detection 13%	Assumed OB scan sensitivity 67% MFM screening 77% MFM FE 80% Cardiology FE 94%
Bak et al. (2020)[14]	Standard-risk pregnancies	1. Three cardiac views 2. Five views 3. Six views 4. Five axial views	QALYs	ICER for five axial views compared with three views was ~35,000 per QALY	Model assumed costs per extra view $10 Sensitivity Three views: 44% Five views: 55% Six views: 67% Five axial views: 66% FE: 94%

FE, Fetal echocardiography; *ICER*, incremental cost-effectiveness ratio; *Hgb A$_{1c}$*, glycated hemoglobin; *ICSI*, intracytoplasmic sperm injection; *IVF*, in vitro fertilization; *MFM*, maternal-fetal medicine specialist; *NT*, nuchal translucency screen at 10-14 weeks; *OB*, obstetric; *QALY*, quality-adjusted life year.

Equipment. Ultrasound systems used for fetal echocardiography should have high spatial and temporal resolution given the small size of a rapidly beating fetal heart. Transabdominal scanning is the standard except in very early gestation. Curvilinear probes are the most frequently used type of transducer given the wide near-field view and near parallel ultrasound beams; these transducers are specifically designed for transabdominal scanning and preferred for the fetal cardiac examination, although linear and sector transducers may be used. The transducer frequency range should be 2 to 7 MHz for late second- and third-trimester scans, while a transducer with a frequency range of 5 to 12 MHz may be useful for late first-trimester and early second-trimester scans. Technological capabilities should include two-dimensional (2D;

B-mode), M-mode, color flow and pulsed-wave (PW) Doppler ultrasound as minimum requirements. Continuous-wave (CW) Doppler ultrasound may be used to characterize high flow velocities if available. Settings such as frequency, harmonics, sector width, and depth should be adjustable to maximize frame rate and lateral resolution at the necessary depth.

Storage. The following is adapted from the AIUM practice parameter for documentation of an ultrasound examination[28] and Intersocietal Accreditation Commission standards and guidelines for pediatric echocardiography accreditation (published June 1, 2017, revised April 27, 2018; https://intersocietal.org/wp-content/uploads/2021/10/IAC PediatricEchocardiographyStandards2017.pdf).

Fetal echocardiographic studies should include both still and moving images, accompanied by a structured report. Required motion clips include but are not limited to the following:

- Axial sweep from the stomach to the upper mediastinum, to include the four-chamber view, arterial outflow tracts, as well as the great vessel and arch views
- Four-chamber view: 2D and color Doppler ultrasound
- Left ventricular (LV) outflow tract view: 2D and color Doppler ultrasound
- Right ventricular (RV) outflow tract view: 2D and color Doppler ultrasound
- 3VV and 3VT views: 2D and color Doppler ultrasound
- Sagittal view of the aortic and ductal arches: 2D and color Doppler ultrasound

Images should be stored in a secure digital archive so that they can be retrieved and reviewed as necessary. Recommendations for content of stored images have been provided by the AIUM.[28] Compliance with local legal requirements for duration of storage is important. The following minimum information should be contained in the metadata and be readable/displayable during review of the stored images (for analog records, identifiers should be present on the image):

- Patient's name and other identifying information
- Facility's identifying information
- Date and time of the ultrasound examination
- Output display standard (thermal index [TI] and mechanical index [MI])
- Label of the anatomic location and laterality, when appropriate
- Image orientation when appropriate

B. Safety

Infection Control. Fetal echocardiographers should adhere to all current institutional and national recommendations with respect to infection control, as it pertains to equipment, pregnant people, and health care professionals. Specific recommendations can be tailored according to circumstances such as the coronavirus disease 2019 pandemic.[29]

Acoustic Bioeffects. To date, there have been no confirmed harmful effects attributable to ultrasound examination of the fetal heart.[30] However, the fetal echocardiographic examination uses a variety of ultrasound modalities, including 2D imaging, PW and CW Doppler, color flow Doppler, and Doppler tissue imaging (DTI), all of which emit energy. Hence, fetal echocardiographers need to be aware that there are theoretical safety considerations in the application of ultrasound to the developing fetus.

The effects of diagnostic ultrasound can be divided into thermal (related to an increase of temperature in the region of insonation) and mechanical (e.g., cavitation from expansion and contraction or collapse of bubbles because of acoustic pressure from the ultrasound beam) types. In view of these potential bioeffects, ultrasound systems have output display standards—TI and MI—to provide guidance to examiners who scan fetuses.

Individuals performing fetal echocardiography should be aware of the TI and MI and should limit power output and time of exposure to no more than that which is necessary to complete the examination. All fetal echocardiographic studies should be guided by the principle of "as low as reasonably achievable," which influences ultrasound modalities and duration of examination.[31] Different ultrasound systems,

even from the same manufacturer, may differ with respect to MI and TI.[32]

C. Examination Technique

In preparation for the examination, the patient is typically placed in a supine position with a soft pillow under the head and with a slight incline of the upper body to improve comfort. The ultrasound system should be positioned to minimize tension and repetitive strain on the operator's scanning arm and upper body. If maternal body habitus and fetal lie and motion limit the examination, the patient may be offered reexamination at another time and the limitations should be made clear in the report.

After establishing the position of the fetus and the right/left and anterior/posterior orientation, an initial survey of the fetus is used to estimate the approximate gestational age and to establish abdominal situs and cardiac position. The presence or absence of fluid in the pericardial, pleural, and peritoneal spaces should be noted. Fetal weight can be estimated at the time of fetal echocardiography, at the discretion of the clinical care team.

D. Image Optimization

Several methods are useful in optimizing image quality and visualization of fetal cardiac anatomy during an examination. Applying adequate transducer pressure and changing maternal position are techniques that may improve fetal positioning and image quality, particularly in obese patients with excessive abdominal panniculus. In such patients, it may be beneficial to scan below the level of the panniculus or through the umbilicus. System settings should be adjusted to maintain high frame rates (e.g., narrowing the sector or field of view, using an appropriate imaging depth, and narrowing the color Doppler region of interest).

Because fetal heart rates normally range from 120 to 160 beats/min, maintaining scanning frame rates >30 to 40 Hz is recommended to optimize temporal and spatial resolution. Additionally, even higher frame rates of 80 to 100 Hz (i.e., about 40 frames/cardiac cycle) may be needed to view important events occurring at heart rates >140 beats/min or to acquire cardiac functional information from speckle-tracking analysis.[33] Image magnification should be set so that the heart takes up approximately one-third of the imaging display and the imaging focal zone should be appropriately set to the level of the desired region of interest to achieve the best possible lateral resolution. The cardiac septa should be imaged in orthogonal projections with the septal planes perpendicular to the imaging plane. Color and spectral Doppler interrogation of intracardiac shunts, as well as valvular and vascular hemodynamics, must be performed at angles coaxial to blood flow to optimize accuracy. Power Doppler ultrasound can be particularly helpful in providing more detail of blood flow associated with small vascular structures and cardiac septal defects. If, despite optimization, imaging remains nondiagnostic, the patient may be offered reexamination at another time and the limitations should be made clear in the report.

E. Standard Views and Imaging Planes

Multiple scanning positions and sweeps are necessary to adequately image the fetal heart. Current societal guidelines advocate for a combination of comprehensive axial plane evaluation with sagittal and
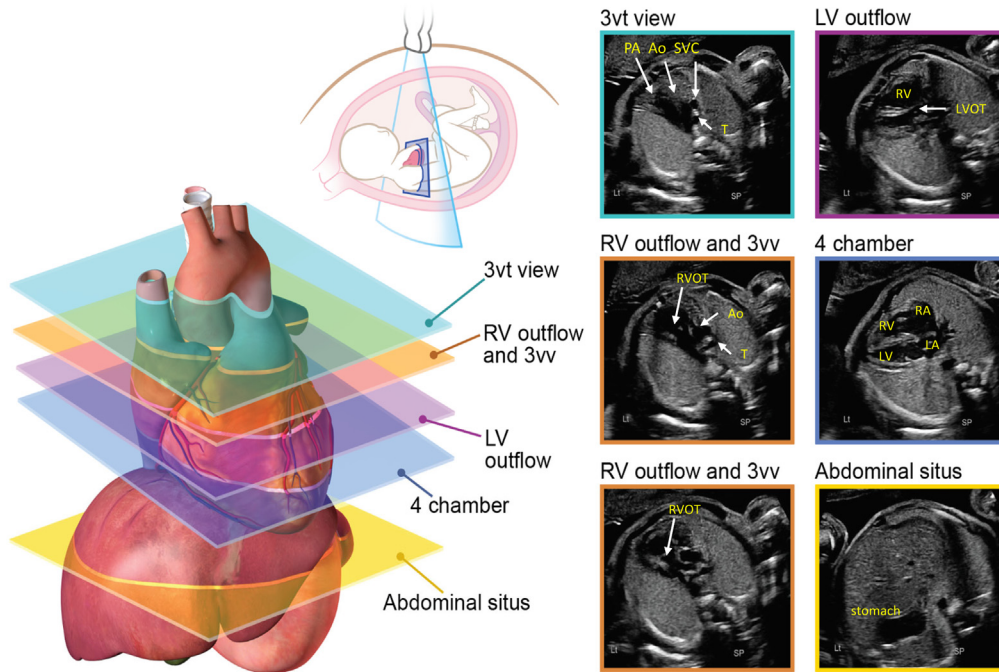
Figure 2 Axial planes suggested for screening the fetal heart at the time of the obstetric anatomic survey and as an initial series obtained during fetal echocardiography. Note that the images depict a fetus in cephalic presentation; breech presentation will result in mirror-image reversal from that shown here. *Ao*, Aorta; *DA*, ductus arteriosus; *LV*, left ventricle; *LVOT*, left ventricular outflow tract; *PA*, pulmonary artery; *RV*, right ventricle; *RVOT*, right ventricular outflow tract; *T*, trachea. Redrawn and adapted with permission from Yagel et al.[34] See also Video 1 (available at www.onlinejase.com).

parasagittal views of targeted structures.[4] Figures 2 and 3 demonstrate the anatomic correlates to the tomographic imaging planes that should be used to evaluate cardiac anatomy and function. A detailed list of structural elements for a complete fetal echocardiographic examination is presented in Table 3, and illustrative examples are shown in Figures 4-9.
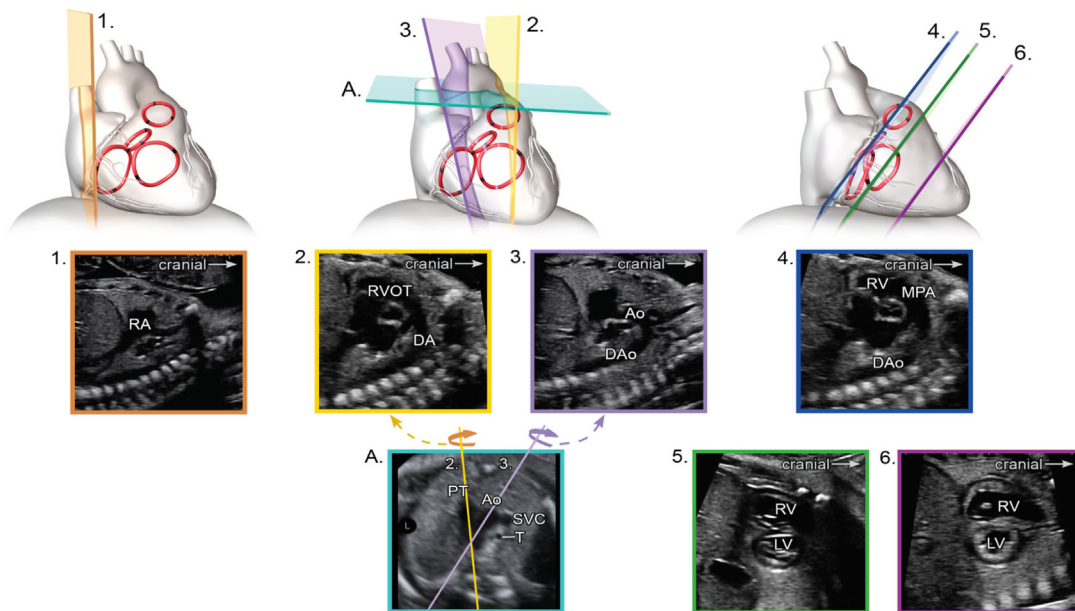
Figure 3 Sagittal and parasagittal planes for fetal echocardiographic evaluation. *Ao*, Aorta; *DA*, ductus arteriosus; *DAo*, descending aorta; *LA*, left atrium; *LV*, left ventricle; *MPA*, main pulmonary artery; *RA*, right atrium; *RV*, right ventricle; *RVOT*, right ventricular outflow tract. See also Video 2 (available at www.onlinejase.com) for sagittal sweep and Video 3 (available at www.onlinejase.com) for arch imaging technique demonstration.

Table 3 Recommended essential and optional components of fetal echocardiographic 2D imaging using 2D and color Doppler

| Component	2D	Color Doppler	Other
Cardiac size	**Required**	NA	**Required**: measurement of cardiothoracic area ratio (normal <0.34)
Cardiac axis (apex left, right, anterior)	**Required**	May be useful if imaging is challenging	Optional: measurement of axis (normal 35°-65°)
Cardiac position in thorax (dextroposition, levoposition)	**Required**	NA	
Visceral and atrial situs determination	**Required**	Optional	
Systemic venous anatomy/connections	**Required**	**Required**	
Pulmonary venous anatomy/connections	Required	**Required**	At least one from each side
Qualitative atrial size and atrial septal morphology	**Required**	**Required**	
AV connections	**Required**	**Required**	
Tricuspid and mitral valve morphology and size (with comparison of right and left)	**Required**	May be useful if imaging is challenging	**Required**: tricuspid and mitral valve annulus measurement
Ventricular morphology, looping determination, size (with comparison of right and left)	**Required**	NA	Optional: maximum length and width measurement
Ventricular septal morphology with exclusion/localization of defect if present	**Required**	**Required**	Optional: wall thickness measurement
Ventricular-arterial connections	**Required**	**Required**	
Pulmonary and aortic valve morphology and size (with comparison of right and left)	**Required**	**Required**	**Required**: aortic and pulmonary valve dimension measurement
Great artery relationship and size (with comparison of right and left)	**Required**	May be useful if imaging is challenging	Optional: ascending aorta and main pulmonary artery measurement
Aortic and ductal arch morphology and size (with comparison of right and left)	**Required**	**Required**	Optional: ductal and aortic isthmus measurement
Aortic/ductal relationship relative to the trachea	**Required**	**Required**	Optional: color Doppler for aberrant subclavian
Proximal right and left branch pulmonary arteries (bifurcation)	**Required**	**Required**	Optional: Measurement of branch diameters
Assessment for pericardial or pleural effusions	**Required**	NA	

NA, Not applicable.
Boldface denotes required elements.
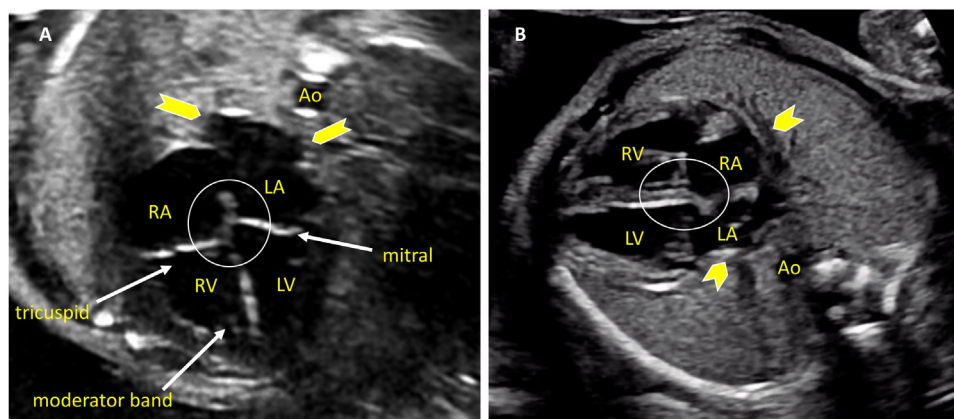
Figure 4 Four-chamber axial images in orthogonal planes. The fetal heart should be examined from at least two orthogonal planes, depending on the structure size, thickness, and orientation to the insonating beam. In **(A)**, the crux of the heart *(circle)* and the tricuspid and mitral valves are well seen, but the atrial and ventricular septa are best viewed as in **(B)**, where the foramen ovale flap is more clearly seen *(oval)*. Note that other structures are visible in addition to the "four chambers" and include pulmonary veins *(arrowheads)*, the descending aorta (Ao) and moderator. band. *LA*, Left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle. See also Video 4 (available at www.onlinejase.com).
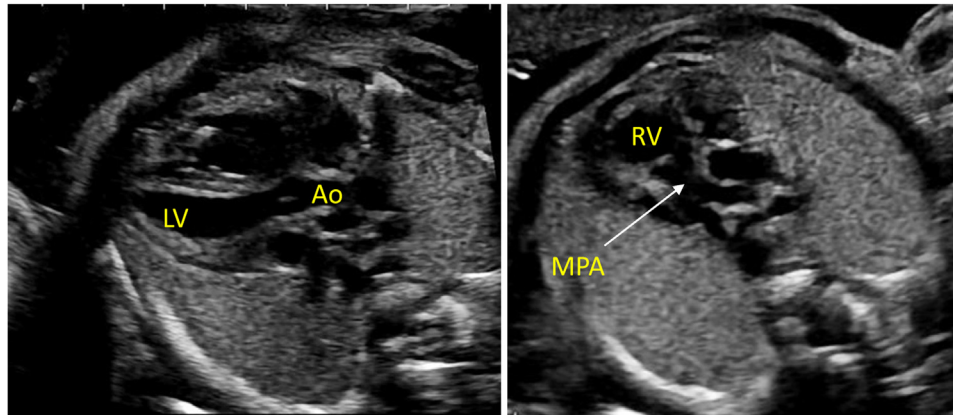
**Figure 5** Detailed imaging of the left *(left)* and right *(right)* ventricular outflow tracts. The ventricular septum is intact from the left ventricle (LV) to the ascending aorta (Ao) in the left ventricular outflow tract or "long-axis" view, and the RV outflow, pulmonary valve, and branch pulmonary artery bifurcation distal to the main pulmonary artery are detailed in this view. The aortic and pulmonary valves are seen in the closed position as thin, nondysplastic structures. See also Video 5 (available at www.onlinejase.com).

i. **Abdominal situs:** Careful attention to fetal lie and orientation of the transducer (notch left or notch right) and display orientation is necessary when assigning abdominal situs. A transverse view of the upper fetal abdomen (Figure 2, *bottom right*) showing the stomach, liver, descending aorta, and systemic venous structures is evaluated to assign situs (usually labeled). This view typically shows a left-sided stomach and a right-sided main hepatic lobe and gallbladder. As the scan plane is moved cephalad toward the thorax and four-chamber view, the hepatic veins and their connections can be visualized.

ii. **Four-chamber view:** The plane of insonation just cephalad to the fetal diaphragm reveals the "four-chamber" view of the heart, including both atria and ventricles (Figure 2, *right middle*, and Figure 4). In addition to cardiac position and the sizes of the atria and ventricles, atrial and ventricular septal anatomy, atrioventricular (AV) valve morphology, and pulmonary venous connections can be discerned. Color Doppler should be used to assess AV valve regurgitation, direction of flow at the foramen ovale, integrity of the ventricular septum, and pulmonary venous connections.
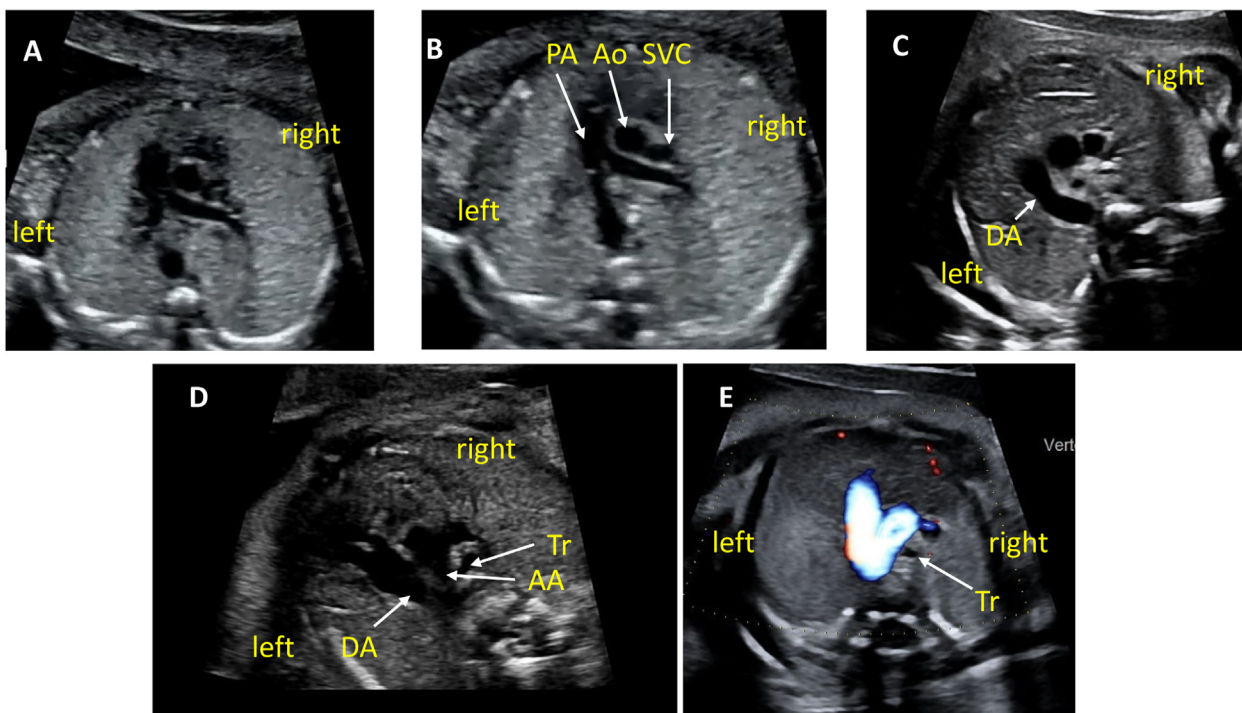
Figure 6 Three-vessel view and 3VT. Very slight movement of the transducer when imaging the upper mediastinum in the axial plane can produce these views (see also Video 6 available at www.onlinejase.com). **(A)** to **(C)** are all acceptable versions of the 3VV, with **(A)** showing more of the RV outflow tract (RVOT) and pulmonary valve, **(B)** demonstrating the main pulmonary artery and branch pulmonary arteries, and **(C)** imaging the ductal arch and no longer in a plane that shows the pulmonary artery branches. **(D)** and **(E)** are slightly more cranial and a bit oblique to show both the ductal and aortic arches and their normal relation to the trachea. When imaging conditions are less optimal or in early gestation, addition of color or power Doppler may make imaging the 3VT view easier **(E)**. *AA*, Aortic arch; *Ao*, aorta; *DA*, ductal arch; *PA*, pulmonary artery; *Tr*, trachea.
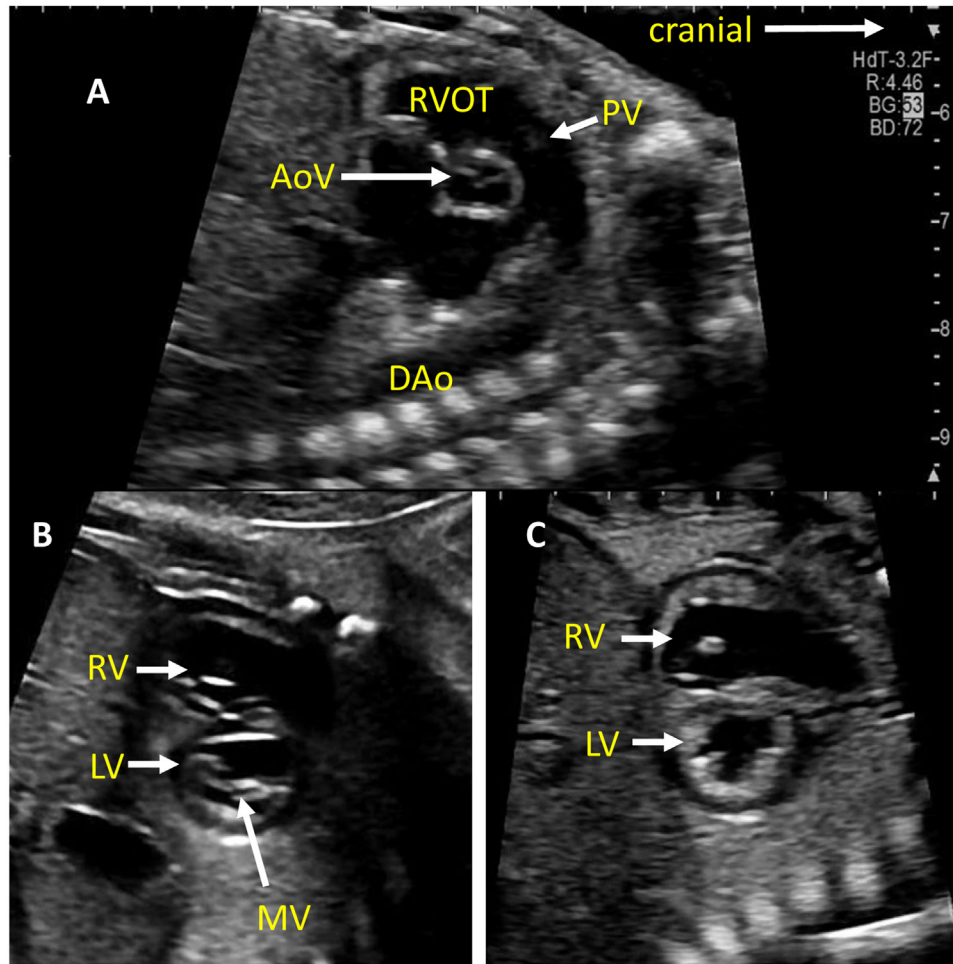
Figure 7 Short-axis views of the fetal heart. **(A)** shows the high short axis at the base, showing aortic valve (AoV) and pulmonary valve (PV) oriented orthogonally to each other, the TV in its normal position, the IVC entering the right atrium, and the more posterior left atrium. **(B)** and **(C)** are obtained by sweeping toward the apex of the heart slightly. In **(B)** the mitral valve (MV) is clearly seen, with two appropriately spaced papillary muscles and a normal anterior leaflet. In **(C)**, the bodies of both ventricles are clearly seen and measurements of the ventricular walls and septum can be made. Cine imaging at this level allows assessment of function and calculation of LV fractional shortening.

iii. **LV outflow tract:** This view is performed by moving the insonation plane slightly cephalad from the four-chamber view (Figure 2, *right upper,* and Figure 5). In the normal heart, the subaortic area, aortic valve, supravalvular region, and ascending aorta can be visualized. Color Doppler should be used to evaluate for the presence of aortic valvular regurgitation or outflow obstruction.

iv. **RV outflow tract:** As the sweep continues cephalad from the LV outflow tract, the RV outflow tract comes into view. In a normal heart, this view
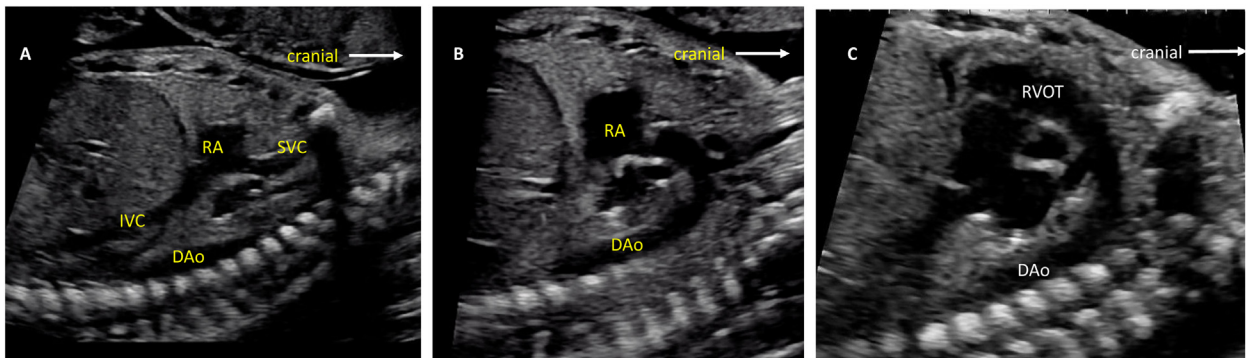
Figure 8 Sagittal **(A, C)** and parasagittal **(B)** images of the bicaval view and the aortic and ductal arches. Note the flattened arch characteristic of the ductus arteriosus **(C)** and the higher arc of the aorta **(B)** with head and upper extremity vessels visible. *DAo,* Descending aorta; *RA,* right atrium; *RVOT,* RV outflow tract. See also Videos 7-9 (available at www.onlinejase.com).
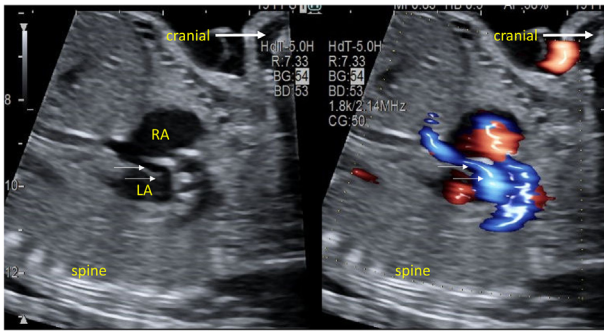
Figure 9 Sagittal caval view in 2D and with color Doppler shows the right-to-left *(blue)* atrial flow across the foramen ovale *(arrows)*. *LA*, Left atrium; *RA*, right atrium.

demonstrates the subpulmonary area, the pulmonary valve, and the proximal main pulmonary artery and may include the pulmonary artery bifurcation (Figures 2 and 6A). With color and PW Doppler, pulmonary valve stenosis or regurgitation can be detected.

v. **3VV:** Cephalad from the RV outflow tract, the 3VV is obtained (Figure 2, *left lower and middle*, and Figures 6A-C). In this plane in a normal heart from left to right, the main pulmonary artery, ascending aorta in cross-section, and superior vena cava (SVC) in cross-section are evident. One or both branch pulmonary arteries may also be included. Abnormalities in vessel number, size, alignment, and arrangements should be noted.

vi. **3VT:** Cephalad from the 3VV, the 3VT is obtained. In the normal heart, the aortic and ductal arches can be seen in a long axis of the transverse segments, both located to the left of the trachea and converging to form the proximal descending thoracic aorta (Figure 2, *top left*, and Figure 6, *bottom panels*). Color Doppler should be used to demonstrate patency and direction of flow in both arches.

vii. **Bicaval view:** The bicaval view is obtained by sagittal imaging of the fetal chest and abdomen (Figure 3, *panel 1*, and Figure 8A). This view will confirm whether the inferior vena cava (IVC) is intact rather than interrupted, and whether the SVC is on the same side as the IVC. This view is also used to assess atrial septal anatomy and patency of and flow direction across the foramen ovale using color Doppler (Figure 9).

viii. **Long-axis view of the aortic arch:** Parasagittal imaging of the fetal thorax (Figure 3, *panel 3*, and Figure 8B) is necessary for complete visualization of the aortic arch. The arch should be assessed to determine continuity between ascending and descending aorta and for hypoplasia of the specific segments (ascending, transverse, isthmus). Color Doppler should be used to assess flow. Normal flow should be antegrade, toward the descending aorta, with minimal forward flow during diastole.

ix. **Long-axis view of the ductal arch:** The ductal arch is usually obtained from a direct sagittal view of the fetal thorax, moving the plane of insonation just left of the midline (Figure 3, *panel 2*, and Figure 8C). The ductus arteriosus should be assessed for restriction at both aortic and main pulmonary artery insertions by noting velocity of flow in systole and diastole using color and/or spectral Doppler. The direction of flow should also be noted.

x. **Short-axis views:** Imaging the short-axis plane of the heart is useful for assessing the ventricular septum, cardiac function, and the anatomy of the AV valves and ventriculoarterial relationships. The high short-axis view at the base of the heart (Figure 3, *panel 4*, and Figure 7A), demonstrates the aortic valve en face, surrounded by the RV outflow tract and pulmonary artery wrapping anteriorly around it. The branch pulmonary artery bifurcation and the tricuspid valve (TV) are usually well seen and the membranous and outlet ventricular septa are evident. TV regurgitation and integrity of the membranous ventricular septum can also be assessed from this view using color Doppler.

The low short-axis view toward the ventricular apex (Figure 3, *panels 5 and 6*, and Figures 7B and C) is used to assess ventricular

function and to evaluate for muscular ventricular septal defects (VSDs) with the application of color Doppler. With slight tilting of the scan plane toward the cardiac base, the morphology of the AV valves can be assessed (Figure 7B).

F. Cardiac Biometry and Measurement Data

Cardiac structures that should be measured for all fetal studies include the aortic and pulmonary valve annuli in systole and the mitral and TV annuli in diastole (Table 4). Fetal heart rate and rhythm should be documented for all studies. Additional cardiac and vessel measurements should be made in specific structural defects (Table 4).

Typically, indexing of measurements involves transformation to a Z score, or the number of SDs from the mean the observed value represents.[35] Several Z score equations for measurements adjusting for gestational age have been published, though alternative formulas exist using biparietal diameter and femur length[36] (Table 5).
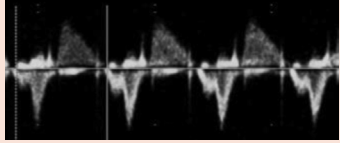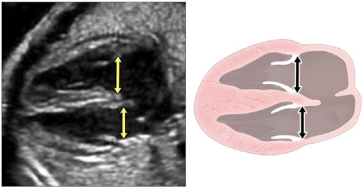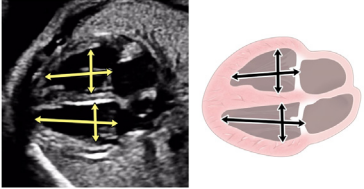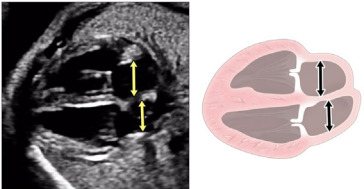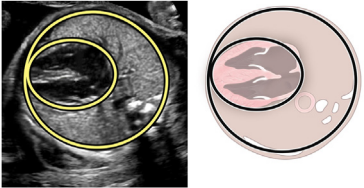
G. Doppler Ultrasonography

Doppler interrogation is an integral component of a complete fetal echocardiographic examination. Detailed assessment of cardiac structures, which will capture major cardiac pathology, should be the initial focus of this examination. The complementary use of Doppler ultrasonography provides additional information regarding normal and abnormal blood flow, as well as cardiac function, rhythm, and conduction that cannot be provided through grayscale imaging alone.

Color and Power Doppler Ultrasound. The value of color Doppler in assessing normal and abnormal fetal cardiac anatomy and blood flows has been recognized for more than three decades.[42] Use of this imaging modality begins with ensuring the Nyquist limit is set to demonstrate flow velocities of interest and to exclude valve stenosis and abnormal flow. Color gain and persistence must also be adjusted during the examination to ensure the color fills the chamber or blood vessel without obscuring the grayscale tissue morphology.

The power Doppler modality analyzes the amplitude of Doppler signals instead of frequency shifts[43]; other modalities such as directional power Doppler combine amplitudes and frequency shifts thus permitting bidirectional imaging of flow.[44] Use of power Doppler as an adjunct to color Doppler can further delineate and define smaller vascular structures and low flow velocities such as in pulmonary and systemic veins.
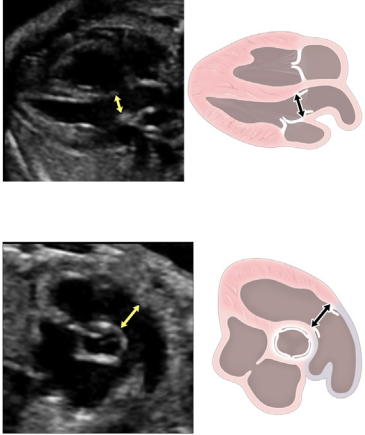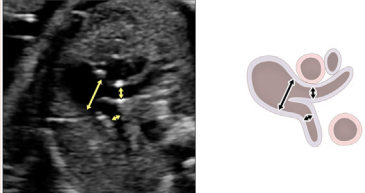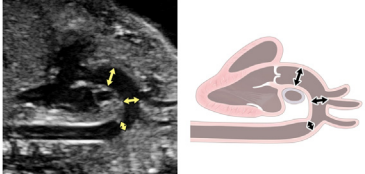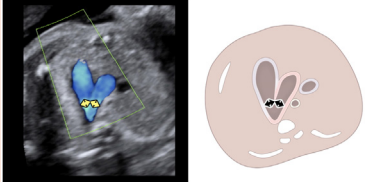
PW Doppler Ultrasound. PW Doppler provides additional data regarding blood flow profiles and direction. Accurate velocity assessment requires the line of interrogation to be coaxial to flow (or at an angle $<20°$). Minimizing the sample volume gate reduces risks for unwanted simultaneous sampling of multiple sites in the fetal heart. Normal values for ventricular inflows and outflows, as well as foramen ovale, pulmonary veins, and IVC flows have been published.[45-49] PW Doppler should be used routinely to assess, at a minimum, flow patterns in ventricular inflows, outflows, arches, pulmonary veins, the ductus venosus, and umbilical vessels (Table 6, Figure 10). Evaluation of umbilical artery

Table 4 Recommended cardiac biometrics

Measurement	Potential uses (not all inclusive)	Example
Heart rate and rhythm	**Required** as part of complete echocardiography	
AV valve (tricuspid and mitral) annulus in early diastole	**Required** as part of complete echocardiography	
RV and LV length in diastole (from four-chamber view)	Right or left heart obstruction	
Transverse atrial dimensions	AV valve regurgitation	
Cardiothoracic ratio	**Required** as part of complete echocardiography Prognostic in TTTS, anemia, SCT, CPAM, and CHAOS	
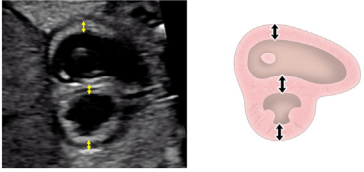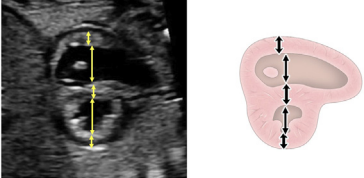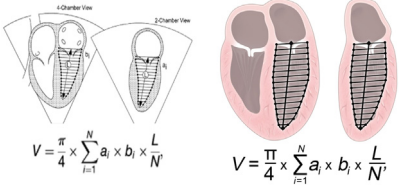
(Continued)

Table 4 (*Continued*)

Measurement	Potential uses (not all inclusive)	Example
Semilunar valve (aortic and pulmonary annulus) measured at end-diastole/ early systole open or closed	**Required** as part of complete echocardiography	
Main pulmonary artery diameter proximal to bifurcation in systole Branch pulmonary diameter(s) in systole	Right or left heart obstruction Lung pathology including diaphragmatic hernia, pulmonary hypoplasia	
Ascending aorta in systole Transverse aortic arch (from long axis of the aortic arch and/or 3VT), with comparison with the ductal arch) in systole	Left heart hypoplasia, bicuspid aortic valve Coarctation, arch hypoplasia	
Aortic isthmus (from long axis of the aortic arch and/or 3VT) with comparison of the aortic isthmus to distal ductus arteriosus in systole	Coarctation, arch hypoplasia	

(*Continued*)

Table 4 (Continued)

Measurement	Potential uses (not all inclusive)	Example
Thickness of the ventricular free walls and interventricular septum in diastole (from low short axis of the ventricles)	TTTS, maternal pregestational diabetes	
RV and LV dimension in diastole (from low short axis of the ventricles), with additional measurements in systole if function concerns	Right or left heart obstruction; allows calculation of shortening fraction	
RV and LV volumes	LV function assessment, calculation of ejection fraction by bullet (also known as 5/6 area × length) or other method	$$V = \frac{\pi}{4} \times \sum_{i=1}^{N} a_i \times b_i \times \frac{L}{N} \qquad V = \frac{\pi}{4} \times \sum_{i=1}^{N} a_i \times b_i \times \frac{L}{N}$$
Calculated combined cardiac output, indexed to estimated fetal weight	High-output lesions, tamponade physiology (e.g., chest masses, hydrothorax)	Output = cross-sectional area $[\pi(\text{diameter}/2)^2] \times$ VTI $\times$ HR pulmonary + aortic = combined

CHAOS, Congenital high-airway obstruction syndrome; *CPAM*, congenital pulmonary airway malformation; *HR*, heart rate; *SCT*, sacrococcygeal teratoma; *TTTS*, twin-twin transfusion syndrome; *VTI*, velocity-time integral.

Measurements for all fetal studies, for comparison with published or internal normal values, include heart rate and rhythm, aortic and pulmonary valves in systole, and mitral and TV annuli in diastole. Optional measurements, especially if abnormalities are noted, include but are not limited to those listed in the table. Boldface denotes required elements.

(free-floating loop of cord) and middle cerebral artery velocities, including measurement of pulsatility and/or resistance indices, can provide additional information regarding the health of the placenta and changes in cerebral resistance, a common phenomenon in certain CHD.[50]

CW Doppler Ultrasound. CW Doppler is not needed for imaging the normal fetal heart but may be necessary to evaluate cardiac and extracardiac pathologies that contribute to abnormal flows and ventricular pressures. As with PW Doppler, alignment of the Doppler cursor coaxial to flow is critical for accurate velocity assessment and gradient calculation. In evaluating valve stenosis, CW Doppler gradient calculation provides insight into the severity of the obstruction. Additional data can be acquired through estimating right or LV pressures with interrogation of the corresponding AV valve regurgitant jet.[51]

H. Complete Fetal Cardiac Assessment Using Segmental Analysis

A segmental analysis of the fetal heart is proposed in Table 7, incorporating all required views and Doppler information. On occasion not all standard imaging planes may be obtainable, although all the cardiovascular structures must be adequately visualized in some manner for the examination to be considered complete, unless the structure is indeed not present. The order in which the images are acquired may be varied.

Table 5 Published Z score equations

Study	Year	GA range	n	Type	Models	Independent variable	Strengths	Weaknesses
Schneider et al.[37]	2005	15-39	130	Z score	Linear regression	Femur length, biparietal diameter, menstrual age	17 measures studied	Small sample size
Lee et al.[36]	2010	20-40	2,735	Z score	Linear regression	Femur length, biparietal diameter, menstrual age	SD formula published, large sample size	Only aortic and pulmonary valves, ventricular short-axis dimensions, and cardiac circumference studied
Colan[38]	2016	16-40	104	Z score	Linear regression	GA	In many ultrasound systems, 17 measures studied	Methods not published, small sample size
Krishnan et al.[39]	2016	12-39	296-414	Z score	Polynomial regression	Femur length, biparietal diameter, GA	13 measures studied	Does not perform well at extremes of GA and measurement
Gu et al.[40]	2018	17-39	6,343	q score	Quantile regression	GA	Better account for nonlinear changes over time, large sample	Clinicians less familiar, hard to calculate quickly
Vigneswaran et al.[41]	2018	13-36	7,945	Z score	Polynomial regression	GA	Large sample size	Only aortic and pulmonary valves, arterial duct, distal transverse arch reported

GA, Gestational age based on first trimester dating.
Currently available websites and calculators: Schneider, McElhinney (subset of Colan scores), Krishnan, and Vigneswaran: http://fetal.parameterz.com/app; Lee: obsono.org/; Colan: https://zscore.chboston.org/.

Table 6 Recommended PW Doppler components of a complete fetal echocardiographic examination (required and optional)

Component	Required/optional
Umbilical vein	**Required***
Ductus venosus	**Required**
Hepatic vein(s)	Optional
SVC and IVC	Optional
Pulmonary veins (at least one each right and left)	**Required**
TV and mitral (inflow) valve	**Required**
Aortic and pulmonary (semilunar, outflow) valves	**Required**
Great arteries and arches (transverse aortic, isthmus)	Optional
Ductus arteriosus (distal, at aortic insertion)	**Required***
Branch pulmonary arteries	Optional
Umbilical artery	**Required***
Middle cerebral artery	Optional
LV inflow-outflow (IVRT)	Optional
Tricuspid/mitral lateral annulus tissue Doppler	Optional

IVRT, Isovolumic relaxation time.
Boldface denotes required elements.
*This represents a consensus-driven change from prior documents.

I. Alternative Ultrasound Modalities

Other ultrasound technologies may be used to image fetal cardiovascular structure and physiology. Spatiotemporal imaging correlation captures a static or dynamic three-dimensional volume data set using a specially designed ultrasound transducer and analysis software.[53] Real-time three-dimensional echocardiographic imaging with this method can be used to enhance detection of anatomic defects and quantify hemodynamics such as ventricular function and cardiac output,[54] although spatiotemporal imaging correlation has not been validated for clinical use. DTI and myocardial strain imaging have been extensively investigated in the postnatal population and may provide a more thorough evaluation of fetal myocardial mechanics.[55] On-cart or offline analysis of cardiac function with 2D strain (speckle-tracking) can be used to detect subclinical cardiac dysfunction.[56] How these modalities can best be applied during pregnancy requires further investigation.

J. Fetal Echocardiography in Early Gestation (<15 Weeks)

The diagnosis of CHD at <15 weeks was first reported in the early 1990s, shortly after the introduction of endovaginal transducers into obstetric practice.[57] Early diagnosis has many advantages including more time for decision-making and for exclusion of associated pathologies, option of earlier pregnancy termination, and insight into CHD evolution from earlier gestations.[58] To date, a broad spectrum of structural and myocardial diseases and arrhythmias have
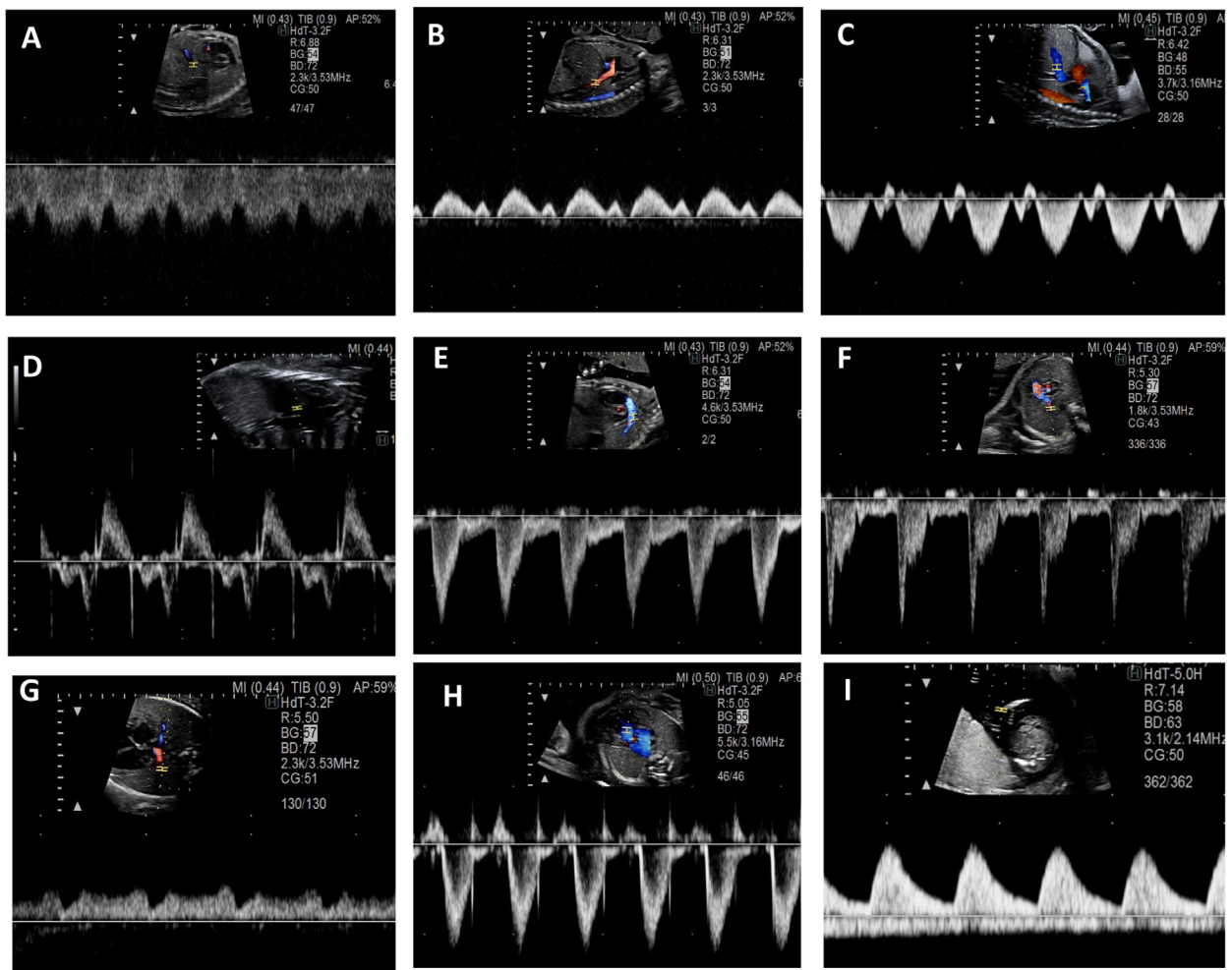
Figure 10 PW Doppler. This modality should be used routinely to assess, at a minimum, flow patterns in ventricular inflows, outflows, arches, pulmonary and systemic veins, the ductus venosus, and umbilical vessels. Representative waveforms include. **(A)** Ductus venosus, **(B)** IVC, **(C)** hepatic vein, **(D)** LV inflow-outflow, **(E)** ductus arteriosus, **(F)** branch pulmonary artery, **(G)** pulmonary vein, **(H)** pulmonary valve, and **(I)** umbilical artery and vein.

Table 7 Segmental analysis of the fetal heart with recommended and optional components of fetal echocardiography

To demonstrate/establish	Recommended view	Additional views
Fetal position in the uterus and anatomic overview Fetal number and position in the uterus Stomach position and abdominal situs Cardiac position Umbilical cord	A single **axial sweep** from abdomen to head (or head to abdomen) Umbilical cord in short axis or fetal bladder with color Doppler to demonstrate number of umbilical vessels **PW UA**, **UV**, **ductus venosus** $\pm$ hepatic vein, $\pm$ IVC	Cordes method for determining sidedness[52]
Systemic venous connections	**3VV sagittal "bicaval" view**	Axial sweep of abdomen from cord insertion to diaphragms, 2D and color
Pulmonary venous connections	**Four-chamber view**: 2D Four-chamber with color (low Nyquist), PW Doppler (two veins, one left and one right)	Additional pulmonary veins from right and left lungs
Atrial anatomy and AV relationship AV valve morphology and function Ventricular morphology and function	**Four-chamber view** 2D, color Doppler AV valves and atrial/ventricular septum, PW tricuspid and mitral inflow, LV inflow-outflow, DTI lateral tricuspid annulus **Ventricular short axis**: 2D, color	Orthogonal plane, four-chamber 2D, color atrial septum and ventricular septum, PW foramen ovale flow **Measure** tricuspid and mitral valves in diastole
Ventriculoarterial relationships, aortic and pulmonary valves	**Long-axis of LV**, 2D color Sweep cranial to **RVOT**, 2D and color **High short-axis base** 2D, color, PW main pulmonary artery Apical **four-chamber** sweep, PW LVOT	**Measure** aortic and pulmonary valves in systole
Aortic arch	**Sagittal arch view**, 2D and color, PW **3VV**, start at pulmonary artery bifurcation and sweep to 3VT with 2D and color	Measure ductal dimension and aortic isthmus on 2D in sagittal or 3VT
Ductal arch	**Sagittal ductal arch**, 2D and color PW ductus arteriosus	Short-axis base, rotate to show ductus, 2D and color
Branch pulmonary arteries	**3VV**, show bifurcation view	Short axis base, 2D Four-chamber, angle cranial, measure branch diameters PW as each branch PA enters lung (hilum)

LVOT, LV outflow tract; *PA*, pulmonary artery; *RVOT*, RV outflow tract; *UA*, umbilical artery; *UV*, umbilical vein.
Boldface denotes required elements.

Table 8 Early (<15 weeks) fetal echocardiography: recommendations, unique elements, strengths, and limitations

Element	Recommendations and challenges	Strengths, limitations, comments
Timing	>10 wk for EV imaging >12 wk for transabdominal imaging (ideal 13-15 wk)	Depends on time of referral, comfort level of sonographer, and availability of EV imaging
Indications	Highest yield indications (see Table 1)	Increased NT: 3-3.5 mm (95th percentile) 3% risk, ≥3.5 mm (99th percentile) 6% risk, 6 mm 24% risk, and >8.5 mm >60% risk Holosystolic TR, DV A-wave reversal
Imaging	Observe ALARA principle[61,62]	TI and MI at ≤1.0 TIs <10 wk and TIb ≥10 wk Limited use of Doppler modalities Narrow sector and zoom
	High-frequency transducer (>8 MHz)	Small cardiac structures
	Should be as close to the fetal heart as possible	At <12 wk <4 cm from fetal heart suggested[63]
	EV imaging	May only be necessary before 12 wk[63]
Cardiac examination expectations[63]	After 11 wk four-chamber ~100%	Symmetric four chambers Cardiac axis changes from 8-14 wk
	At 11 wk >90% great arteries and arches	Symmetric great arteries and arches expected
	Use of color flow mapping when 2D is limited	Assists with confirming presence and patency of four chambers, outflows, and arches
	Pulmonary and systemic vein assessment limited	Most challenging aspect of the examination at early gestations
	Situs determination	May be easier to perform transabdominally, not via EV
Cardiac pathology	General sensitivity and specificity good	78.6% and 98.9%, respectively[57]
	Major structural heart disease similar in appearance to later gestation	Most can be detected at 10-14 wk Subtle defects including valve lesions due to suboptimal resolution difficult
	Major limitations due to resolution (potentially can be overcome with technology advances) and timing	If unclear or suspicious, suggest return within 1-2 wk given exponential growth of the heart at this stage
Counseling and reporting	Integrate role of genetic screening and testing, limitations of examination	Potential for missed diagnoses and progressive lesions
Additional considerations	Higher rates of pregnancy termination[64]	Some disease, especially with associated aneuploidy, may be more severe than later gestation due to natural history resulting in pregnancy loss
	Although detailed, considered a screening examination	Reassessment at 18-20 wk recommended

ALARA, As low as reasonably achievable; *DV*, ductus venosus; *EV*, endovaginal; *MI*, mechanical index; *NT*, nuchal translucency; *TI*, thermal index; *TIb*, thermal index for bone; *TIs*, thermal index for soft tissue.
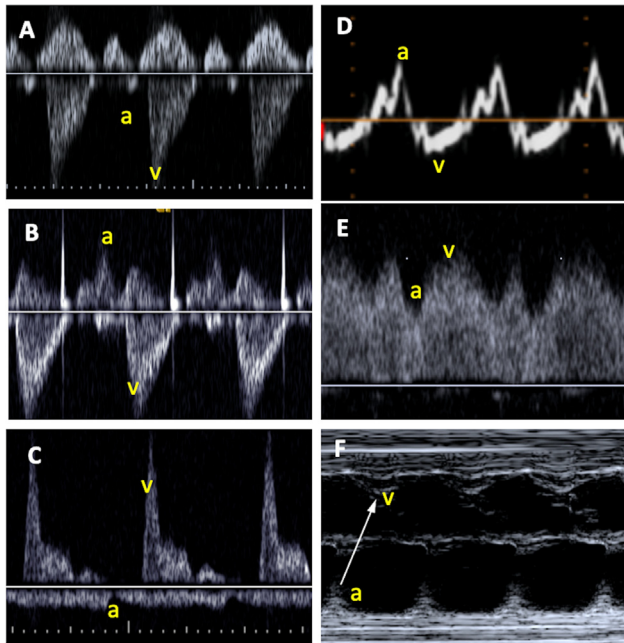
Figure 11 Ultrasound tools for evaluation of fetal heart rate, rhythm, and AV conduction. **(A)** SVC–ascending aorta (Ao) Doppler. **(B)** LV inflow-outflow Doppler, obtained with a wide range gate simultaneously sampling mitral inflow at the anterior leaflet and aortic outflow just below the aortic valve. **(C)** Pulmonary artery–pulmonary vein simultaneous sampling. **(D)** Tissue Doppler at the lateral tricuspid annulus. **(E)** Ductus venosus Doppler. **(F)** M-mode with cursor placed through the atrial wall and the free wall of the right or left ventricle. The fetal rate and rhythm are normal in all the above tracings. *a*, Atrial contraction; *v*, ventricular contraction.

been diagnosed at <15 weeks. The sensitivity and specificity of early fetal echocardiography have been reported as 78.6% and 98.9%, respectively,[57] and in experienced hands, sensitivity of 89% and specificity of 100% are possible for a basic cardiac diagnosis.[59,60]
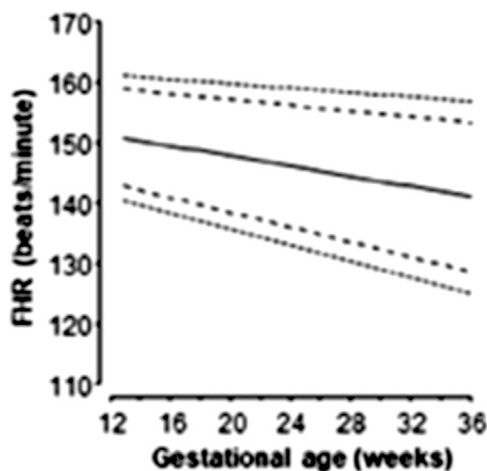
Figure 12 Fetal heart rate (FHR) measured by echocardiography according to gestational age. The first, fifth, 50th, 95th, and 99th percentiles are marked. Adapted with permission from Zidere *et al.*[65]

Table 8 provides an overview of the approach to early fetal echocardiography, including its strengths and limitations, and important considerations in comparison with fetal echocardiography in the second and third trimesters.

Key Points

- Fetal echocardiography must be performed using appropriate equipment and in observance of the principle of "as low as reasonably achievable."
- Existing guidelines for storage and documentation must be followed.
- Two-dimensional, spectral Doppler, and color Doppler echocardiographic modalities should be used, and imaging settings optimized specifically for the fetal heart examination.
- Standard views and imaging planes should be used.
- Minimum standards for 2D, spectral Doppler, and color Doppler static and dynamic image capture including fetal position, cardiac structure and function, and cardiac biometry must be met.
- Fetal echocardiography can be performed earlier than 18 weeks and should be considered for patients presenting in the late first trimester with suspicion for cardiac anomaly, depending on local resources and expertise.
- There is insufficient evidence to recommend early (<16 weeks) fetal echocardiography for low-risk pregnancies, but it has been shown to be feasible and may be used for high-risk pregnancies or those with known abnormal screening findings.

IV. GUIDANCE FOR DISEASE-SPECIFIC ANATOMIC, PHYSIOLOGIC, AND FUNCTIONAL FETAL ECHOCARDIOGRAPHIC EVALUATION

A. Rhythm Disturbances and Tools for Assessment

Echocardiographic assessment of fetal cardiac rhythm uses 2D, M-mode, PW Doppler, and tissue Doppler modalities (Figure 11). Evaluation of the type and mechanism of arrhythmia is essential to plan treatment and to prognosticate. The normal fetal heart rate varies
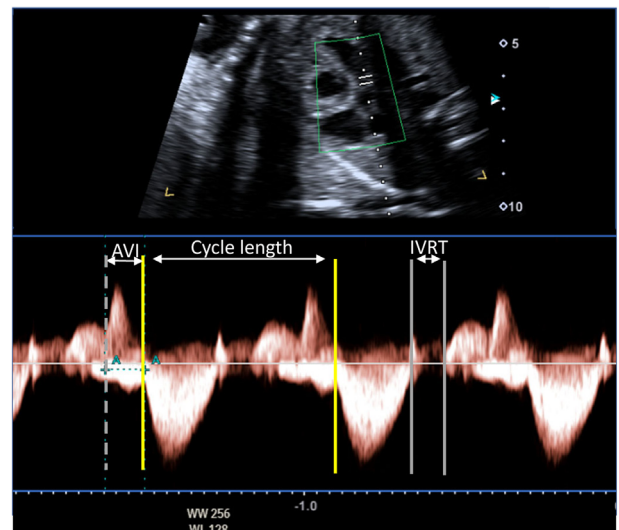
Figure 13 Doppler interrogation of a fetus with suspected long-QT syndrome. The Doppler cursor is placed between mitral valve inflow (above baseline) and aortic flow (below baseline). The cycle length is 560 msec (heart rate 110 beats/min). The AV interval (AVI) is normal and there is AV concordance, excluding AV block. The time between aortic valve closure and onset of mitral valve flow (isovolumic relaxation time [IVRT]) is 100 msec. The IVRT expressed as a percentage of the cycle length is 17%, which is well above the normal range for gestation, consistent with a diagnosis of long-QT syndrome.

Table 9 Recommendations for evaluation of disturbances of fetal heart rate and rhythm

Disturbance	Diagnosis	Evaluation	Notes
Irregular fetal heart rhythms with a normal overall rate	Atrial ectopic beats (>95%) Ventricular ectopic beats much less frequent (<5%)	Any available tool: a-a and V-V time intervals will demonstrate resetting of atrial rhythm in PACs (with an "incomplete compensatory pause") while PVCs do not change the atrial a-a interval	Follow-up heart rate monitoring until resolved
Tachycardia	Sinus tachycardia	M-mode or arterial/venous Doppler; long VA tachycardia with a regular rhythm, typically <200 beats/min	
	SVT	M-mode or arterial/venous Doppler; 1:1 ratio between the atrial and ventricular contractions (Figure 14); most commonly orthodromic reciprocating (pathway mediated), a short VA tachycardia	Assessment of mitral valve/aortic flow is not recommended because of fusion of inflow patterns so that the timing of atrial contraction cannot be measured
	Ectopic atrial tachycardia or PJRT	M-mode or arterial/venous Doppler; tachycardia with a 1:1 A/V ratio but of "long VA" type, may have variable V-V intervals	Echocardiographic determination of mechanism is important for selection of arrhythmia treatment
	Atrial flutter	M-mode, 2D, Doppler; extremely rapid atrial rate with a characteristic "sawtooth" pattern on M-mode (Figure 14); atrial rate > 400 beats/min, and associated with a variable degree of AV block (2:1, 3:1 most common)	The atria are frequently dilated with associated AV regurgitation and impaired ventricular function
	Ventricular tachycardia	M-mode or arterial/venous Doppler; tachycardia with A/V dissociation, regular atrial rate with (usually) slower ventricular rate, may have variable V-V intervals	
Bradycardia	Sinus bradycardia	Any modality demonstrating atrial and ventricular contraction simultaneously; most sinus bradycardias have a ventricular rate >90-100 beats/min; echocardiographic findings in a fetus with sinus bradycardia due to long-QT syndrome are shown in Figure 13, with the use of Doppler to measure IVRT in this context	DTI at the lateral tricuspid annulus may be useful
	Complete heart block	Any modality demonstrating atrial and ventricular contraction simultaneously; in complete heart block the atrial and ventricular rates are completely dissociated (Figure15), and ventricular rates are typically in the range of 50-80 beats/min	
	Multiple blocked atrial ectopic beats, atrial bigeminy and blocked atrial bigeminy vs second- or third-degree AV block	Multiple blocked atrial ectopic beats can lead to a subnormal or irregular ventricular rate, which must be distinguished from second- or third-degree AV block; the echocardiographic approach to ectopic beats is shown in Figures 17 and 18 and uses any modality available	Confirm with ultrasound, as external fetal monitoring cannot distinguish from bradycardia because of fetal distress (M-mode may be better than Doppler methods when sinus-ectopic intervals are long)

IVRT, Isovolumic relaxation time; *PAC,* premature atrial contraction; *PJRT,* permanent junctional reciprocating tachycardia; *PVC,* premature ventricular contraction; *SVT,* supraventricular tachycardia; *VA,* ventriculoarterial.
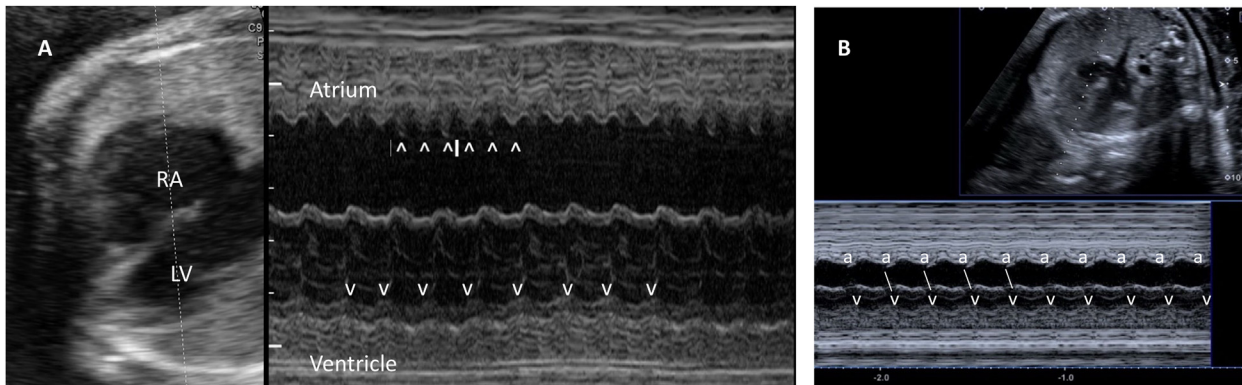
Figure 14 Fetal tachyarrhythmia diagnosis using M-mode. **(A)** Atrial flutter. The atrial contractions demonstrate the typical "sawtooth" pattern of atrial flutter *(open arrow [^])*. Alternate flutter waves are conducted to the ventricles (V) giving 2:1 block. The atrial rate was 474 beats/min. **(B)** Reentrant (pathway-mediated) supraventricular tachycardia. The atrial (a) and ventricular (v) rates are equal (240 beats/min), and the rhythm is regular.

with gestational age and normal reference ranges have been published (Figure 12).[65,66] At any gestational age, a sustained heart rate of >160 to 180 beats/min or <120 beats/min is abnormal.

M-mode echocardiography to assess fetal rhythm takes advantage of high temporal resolution to investigate precise timing of atrial and ventricular myocardial contraction, usually assessed simultaneously. Doppler echocardiography of simultaneous mitral valve inflow/aortic outflow, pulmonary vein/branch pulmonary artery, or SVC/aortic flow can be used to time atrial and ventricular contraction. PW tissue Doppler can be used to measure atrial and ventricular myocardial velocities and time intervals by interrogation of the lateral mitral or TV annulus. Doppler techniques of rhythm assessment permit quantification of fetal heart rate, the relationship between atrial and ventricular contraction, and assessment of conduction time intervals such as the mechanical PR interval (AV interval). In pregnant people known to carry anti-Ro/La antibodies, measurement of the fetal mechanical PR interval can be performed to evaluate for AV conduction abnormalities, although there is not yet clear evidence of efficacy of routine surveillance in decreasing the incidence of higher degrees of

AV block.[67] The measurement of the Doppler-derived isovolumic time intervals (Figure 13) may aid in the diagnosis of fetal congenital long-QT syndrome[68] or in distinguishing challenging rhythms such as blocked atrial bigeminy versus 2:1 AV block.[69]

Common arrhythmias and their diagnostic evaluation using echocardiography are presented in Table 9 and Figures 14-17. If echocardiographic findings are equivocal, fetal magnetocardiography and electrocardiography can be used to directly assess the fetal cardiac electrical signal, but they currently have limitations to widespread clinical use. Fetal magnetocardiography can precisely characterize fetal heart rhythm, as well as conduction and repolarization properties.[70]

Assessment of Hemodynamic Compromise Due to Arrhythmia. Sustained fetal arrhythmias may lead to fetal congestive heart failure and nonimmune fetal hydrops, increasing the risk for fetal demise.[71,72] Careful assessment of fetal cardiovascular status and well-being is critical to guiding treatment.

Fetal cardiovascular status is determined by measuring the cardiothoracic area ratio (Figure 18), noting the presence of AV valve regurgitation and/or ventricular dysfunction, and noting findings suggestive of hydrops. Doppler waveforms, particularly increases in ductus venosus and umbilical venous pulsatility, may be difficult to interpret if abnormal in the setting of fetal atrial ectopic beats, tachycardia, or complete heart block. An abnormal fetal heart rhythm disrupts the normal sequence of electrical events in the cardiac cycle, such that A-wave flow reversal in the ductus venosus or umbilical venous notching may be reflective of the fetal arrhythmia rather than reflective of impaired cardiac function or fetal compromise.
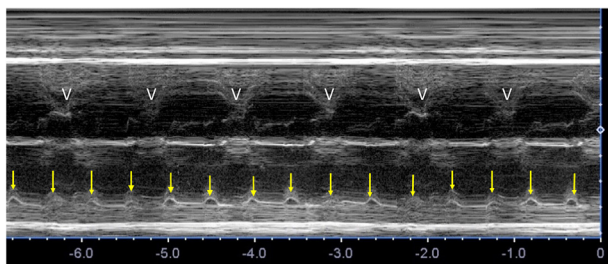
Figure 15 M-mode tracing of complete heart block. The atrial rate *(arrows)* is normal at 120 beats/min. The ventricular rate (V) is much slower at 55 beats/min, with no relationship between atrial and ventricular contractions (AV dissociation). In this example the heart structure was normal, and heart block was due to maternal anti-Ro antibodies.

Key Points

- Fetal heart rate and rhythm must be documented as part of a fetal echocardiographic examination.
- If the heart rhythm is irregular, or if bradycardia or tachycardia are observed, the mechanism should be documented using M-mode, PW Doppler, or tissue Doppler echocardiography or a combination of these modalities.
- Fetal echocardiographic measurement of time intervals in specific situations (maternal anti-Ro/La positivity, suspected or confirmed genetic long-QT syndrome) may be useful.
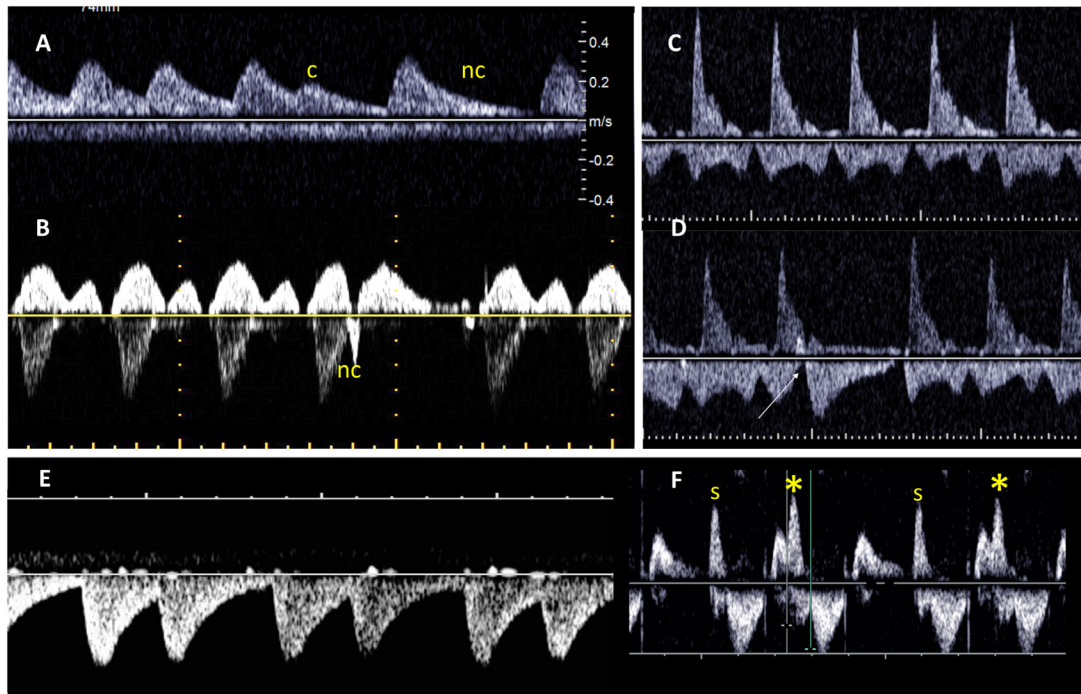
Figure 16 Irregular rhythm due to conducted and nonconducted premature atrial contractions (PACs). **(A)** Umbilical artery/vein Doppler demonstrating conducted (c) and nonconducted (nc) PACs in the same patient. If the AV conduction is normal, a premature atrial event is followed by a related premature ventricular event. If, however, the PAC is premature enough to fail to conduct across the refractory AV node, there is no ventricular activation. **(B)** Doppler recordings from the SVC and aorta in a fetus with PACs. The tracing shows normal venous flow above baseline with a single nonconducted premature atrial contraction (nc) resulting in temporary flow reversal at the end of systole; flow reverses due to atrial contraction against a closed TV. There is a long pause due to lack of conduction as the impulse is blocked in the AV node. **(C, D)** Pulmonary venous inflow–branch pulmonary artery outflow. Pulmonary artery flow above baseline and venous flow below baseline sampled simultaneously due to their proximity in the hilum, demonstrating normal rhythm **(C)** and a single nonconducted PAC, denoted by *arrow* **(D)**. **(E, F)** Frequent PACs. Umbilical artery Doppler **(E)** showing an irregular rhythm with an average rate of about 120 beats/min. Using LV inflow-outflow Doppler, the reason for the irregularity is clearly demonstrated to be frequent conducted premature atrial contractions *(asterisk)* alternating with normal sinus beats (s) in a pattern of bigeminy.
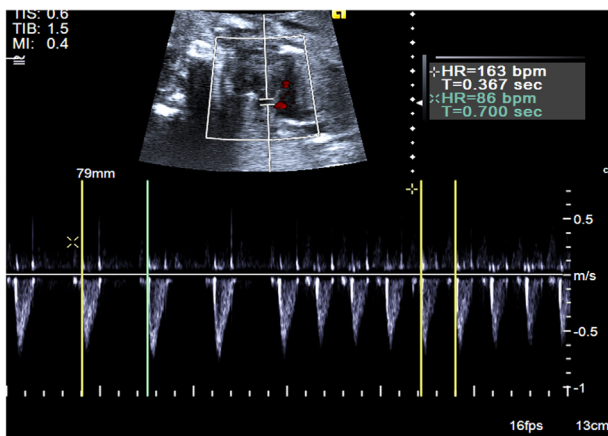
Figure 17 Fetal heart rate alternating between frequent PACs in a pattern of blocked atrial bigeminy and normal sinus rhythm, not to be mistaken for second-degree AV block. Because of the incomplete compensatory pause after the extrasystolic atrial beat, the previous V-V interval is shortened, therefore the apparent ventricular rate (86 beats/min) is more than half of the rate seen in the following period of sinus rhythm (163 beats/min), distinguishing the slower rhythm from intermittent second-degree AV block and too fast for typical third-degree block.
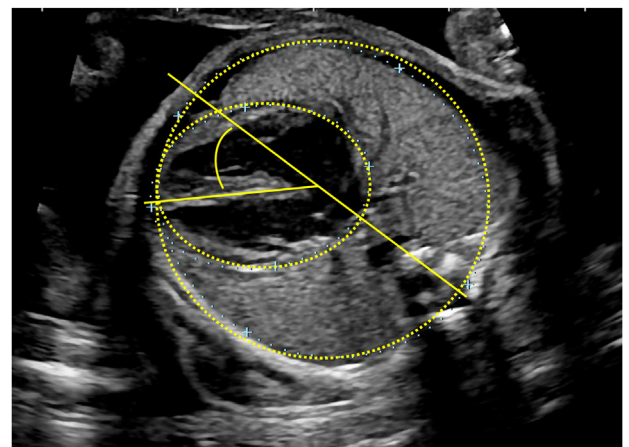
Figure 18 Standard biometric measurements from an axial image at the level of the four-chamber view. The cardiothoracic ratio is calculated from measurement of the inside circumference (or area) of the chest and the epicardial surface of the heart, as shown in *yellow dotted lines*. The cardiac angle is determined by bisecting the chest; with a line from the sternum to the spine and measuring the angle at which the ventricular septum would intersect the line *(yellow solid lines)*.

Table 10 Recommendations for components of a comprehensive fetal cardiac functional assessment

	Function index	Modality
Systolic function	• **LV and RV qualitative assessment or** measured shortening fraction • TAPSE, MAPSE • Myocardial performance (Tei) index (global function)	• 2D, M-mode • M-mode • PW or tissue Doppler
Diastolic function	• **AV valve inflow profile** (E/A velocity, E/A ratio, presence of monophasic inflow pattern) • DFTc • Myocardial performance index (MPI, Tei; global function) • **IVC or hepatic venous flow** (ratio of A-wave VTI/forward flow VTI) • **Ductus venosus flow (presence/absence of A-wave reversal**, pulsatility index**)** • **Umbilical venous flow (presence/absence of venous pulsation)**	• PW Doppler • PW or tissue Doppler • PW Doppler • PW Doppler • PW Doppler • PW Doppler
Valve function	• **Presence/absence of AV valve regurgitation**	• Color flow Doppler
Cardiac output	• Cardiothoracic ratio • Combined cardiac output, indexed to EFW	• 2D • 2D and PW Doppler
Cardiovascular profile score (see Table 13)	• Composite score of overall cardiac function	• 2D, M-mode, PW Doppler

DFTc, Diastolic filling time corrected for cardiac cycle length; *EFW*, estimated fetal weight; *MAPSE*, mitral annular plane systolic excursion; *TAPSE*, tricuspid annular plane systolic excursion.

Components in boldface type should be considered mandatory in any fetus suspected of having cardiovascular dysfunction; other components may be considered depending on the specific cardiac condition (see Table 12).

B. Assessment of Fetal Cardiac Functional Abnormalities

Two-dimensional and Doppler echocardiography are important tools in the assessment of fetal cardiac function. Various methods can be used to assess cardiovascular function (Table 10) in the fetus with cardiac and noncardiac conditions at risk for cardiac dysfunction and fetal compromise (Table 11). Identifying fetal cardiac dysfunction

Table 11 Types of hemodynamic derangement caused by fetal conditions

Hemodynamic derangement	Fetal condition	Common abnormalities detected by echocardiography
High–cardiac output lesions[73-75]	• Arteriovenous malformation • Agenesis of ductus venosus • Fetal anemia • TRAP • TTTS • Vascular masses/tumors	• Elevated CCOi • Cardiomegaly (CTAR) • AV valve regurgitation • Abnormal or normal vein Doppler • LV/RV systolic dysfunction • Hydrops
Increased preload (volume overload)[76,77]	• AVSD with severe AV valve regurgitation • Complete AV block • TOF/absent pulmonary valve	• Elevated MPI • Systolic dysfunction • Abnormal venous Doppler • Cardiomegaly (CTAR) • Hydrops
Increased afterload lesions[78,79]	• Bilateral semilunar valve stenosis including truncal valve stenosis • Constriction of the ductus arteriosus • TTTS	• Elevated MPI • AV valve regurgitation • Diastolic dysfunction • Abnormal venous Doppler • Hydrops
Cardiac compression (decreased preload)[75,80-82]	• CDH • CHAOS • CPAM	• Low CCOi • Abnormal tricuspid inflow and/or venous Doppler (CHAOS, CPAM) • Hydrops • Left heart compression, reduced LV stroke volume (CDH)

AVSD, Atrioventricular septal defect; *CCOi*, combined cardiac output index; *CDH*, congenital diaphragmatic hernia, *CHAOS*, congenital high-airway obstruction; *CPAM*, congenital pulmonary airway malformation; *CTAR*, cardiothoracic area ratio; *MPI*, myocardial performance index; *TRAP*, twin-reversed arterial perfusion; *TTTS*, twin-twin transfusion syndrome.
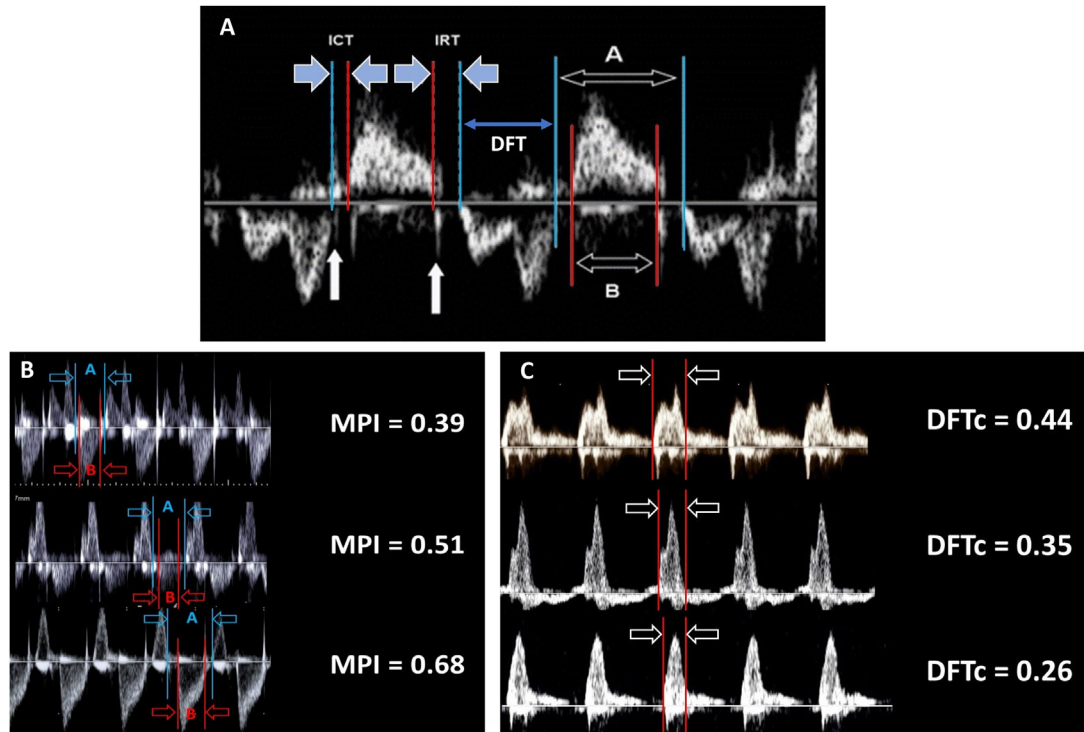
Figure 19 Myocardial performance index (MPI) and diastolic filling time (DFT), normal and abnormal. **(A)** PW Doppler tracing of LV inflow-outflow demonstrating calculation of the Doppler myocardial performance index. The MPI is expressed as the total time in iso-volumic relaxation time (IRT) and isovolumic contraction time (ICT), divided by the ejection time (interval B) or (interval A − interval B)/ interval B. Note the valve clicks *(white arrows)* used to mark the beginning and end of IRT and ICT. DFT from the onset of the E wave to the end of the A wave is also illustrated. **(B)** Changes in myocardial performance index in fetal disease states. A normal MPI is seen in the top panel. The middle and bottom panels are examples of worsening MPI in the recipient twin in a monochorionic/diamniotic twin gestation with twin-twin transfusion syndrome. **(C)** Diastolic filling time, corrected for heart rate (DFTc). The DFTc is calculated as the total duration of the inflow Doppler spectral tracing, divided by the cardiac cycle length. As diastolic function worsens the normal DFTc progressively shortens and inflow becomes monophasic.

is important for determining the need and timing of potential intervention for conditions that can be treated prenatally. Characterization of fetal cardiac dysfunction can provide clinicians with the means to better select patients for intervention, predict outcome, and to perform follow-up assessment after fetal interventional procedures. Assessment of cardiac function is also important in determining delivery planning and timing even in cases in which fetal intervention may not be an option.
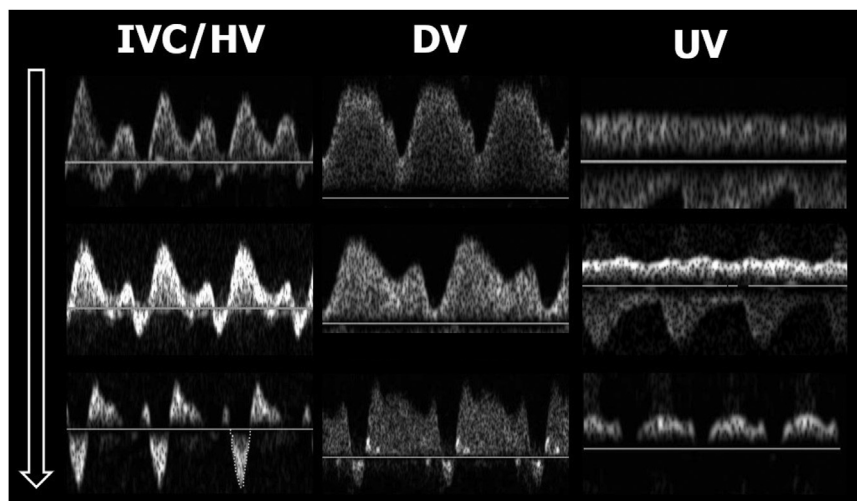
Figure 20 Venous Doppler changes with increasing central venous pressure (CVP). As CVP rises *(open white arrow)*, representative changes in the IVC, hepatic veins (HV), ductus venosus (DV), and umbilical vein (UV) are noted.

Table 12 Cardiovascular profile score*

Category	Subscale points		
	2	1	0
Hydrops	None	Ascites or pleural or pericardial effusion	Skin edema
Heart size (CA/TA ratio)	0.20-0.35	0.35-0.50	>0.50 or <0.20
Cardiac function	Normal biphasic AV inflow, RV and LV SF > 0.28	Holosystolic TR or RV/LV SF < 0.28	Holosystolic MR or TR dP/dt < 400 or monophasic AV inflow
Venous Doppler	Nonpulsatile UV and normal DV	Nonpulsatile UV and negative A wave in DV	Pulsatile UV
Arterial Doppler	Forward diastolic flow in UA	Absent end-diastolic flow in UA	Retrograde end-diastolic flow in UA

CA, Cardiac area; *DV*, ductus venosus; *MR*, mitral regurgitation; *SF*, shortening fraction; *TA*, thoracic area; *TR*, tricuspid regurgitation; *UA*, umbilical artery; *UV*, umbilical vein.
Adapted with permission from Mäkikallio *et al.*[88]
*Sum of points in all five categories = score; "10" best, "0" worst.

i. Imaging Tools for Assessing Fetal Cardiac Function. The components of a comprehensive cardiac functional assessment are listed in Table 10. On initial imaging of the fetus with suspected cardiac dysfunction, it is important to assess for the presence of fetal hydrops and/or its component findings. Overall cardiac size, as determined by cardiothoracic area ratio, may be a sign of altered hemodynamics in the fetus.[83] Two-dimensional and/or M-mode echocardiographic assessment of cardiac chamber size and ventricular function should be performed. AV valve Doppler flow patterns[65] can indicate diastolic dysfunction and AV valve regurgitation imaged with color Doppler or PW Doppler may indicate systolic dysfunction. The Doppler myocardial performance index assesses global ventricular function by incorporating both systolic and diastolic time intervals (Figure 19). Venous Doppler flow patterns will display increased retrograde blood flow velocity during atrial contraction as fetal heart function worsens (Figure 20). The magnitude of this A-wave reversal is also influenced by the size of the vessels, the net umbilical blood flow through the vessels, and the adrenergic state of the fetus. Venous Doppler A-wave reversal is unreliable in rhythms other than sinus rhythm.

PW Doppler ultrasound can be used to quantify RV and LV stroke volumes and outputs for additional insight into the human fetal circulation.[84,85] Stroke volume is calculated with the semilunar valve annulus diameter and Doppler velocity-time integral (VTI) using the following formula:

Stroke volume = [(0.785 × annular diameter^2) × VTI].

Given that the fetal circulation is in parallel, the combined cardiac output from both ventricles is often used. Normal combined cardiac output index in the fetus is 425 mL/min/kg and ranges from 225 to 625 mL/min/kg.[85] Normative data for gestational age have been published[85] and have been used to predict outcomes in high-output states.[73]

DTI. Although there is no evidence to suggest a benefit in the routine application of DTI during fetal imaging, it may be useful in the evaluation of fetal cardiac function and rhythm. Normative data for DTI e′, a′, and s′ velocities, isovolumic relaxation and contraction time intervals, and RV and LV Tei indices and E/e′ ratio have been published.[86,87]

Cardiovascular Profile Score. The cardiovascular profile score is a global measure of cardiovascular status and incorporates multiple fetal cardiovascular findings, expressing them as a cumulative score with the best being 10 and the worst being 0 (Table 12). The score has been used to describe the degree of fetal cardiovascular compromise for several disease states[89] including twin-twin transfusion syndrome,[90] fetal high–cardiac output states,[73,74] fetal growth restriction,[88] and nonimmune fetal hydrops.[75,83] This measure may be useful in describing and tracking longitudinal changes in the degree of cardiac compromise.

ii. Specific Types of Cardiovascular Perturbation in the Fetus. When considering fetal abnormalities that can produce cardiovascular compromise, it is useful to consider which type of hemodynamic derangement is primarily involved, and how the fetal heart adapts to these changes. Although all fetuses at risk for cardiac dysfunction should undergo complete assessment of systolic and diastolic function, an understanding of the hemodynamic derangements specific to a particular condition allows clinicians to "target" which parameter of fetal cardiac function may be most useful in assessing the degree of hemodynamic derangement. Although it is beyond the scope of this statement to fully discuss specific echocardiographic findings in each fetal condition, understanding and documenting the findings that are prognostic in each condition is important, as reviewed in Table 11.

iii. Additional Imaging: Extracardiac Doppler and Adjunct/ Advanced Techniques. Complete cardiovascular assessment in the compromised fetus should include PW Doppler evaluation of both umbilical and middle cerebral artery flow patterns, although this can be accomplished either during obstetrical scanning or during fetal echocardiography. In the fetus with cardiovascular compromise, it has been shown that redistribution of cardiac output—so-called brain sparing—will occur as a compensatory response, with demonstrable decreases in cerebral resistance. This phenomenon has been demonstrated in fetal growth restriction,[76] high–cardiac output states,[91] and twin-twin transfusion syndrome.[78] These changes may precede changes in venous flow pattern, and thus may be an important early sign of fetal cardiac compromise. Measurement of the peak systolic velocity in the middle cerebral artery should also be performed and compared with gestational age normal values in the setting of cardiac dysfunction, to exclude fetal anemia.[92]

Other advanced techniques, such as DTI and myocardial strain, have been described. The utility of these techniques in evaluating function and predicting outcomes is less clear and use is largely limited to research applications.

Key Points

- Qualitative assessment of cardiac systolic and diastolic function must be part of a fetal echocardiographic examination. This includes 2D imaging, color and PW Doppler of AV valve inflow and regurgitation (if present), and ductus venosus PW Doppler.
- Quantitative measures should be performed when cardiac function or output is suspected to be abnormal, either on the basis of extracardiac diagnosis or if the fetus is compromised (i.e., effusions, abnormal 2D appearance, abnormal venous Doppler patterns); serial measurement may also be indicated.
- A cardiovascular profile score may be useful in describing and documenting longitudinal changes in the degree of cardiac compromise.

C. Structural Heart Disease

i. Single Ventricles and Assessment for Ductal Dependence.

Single ventricle (SV) CHD is often detected on routine ultrasound screening because of the presence of an abnormality in the four-chamber view; most patients are diagnosed before birth.[93] The most common form of SV CHD is HLHS, with an incidence of 0.26 per 1,000 live births.[13] Other forms of SV CHD include hypoplastic right heart syndrome (0.22 per 1,000 live births) and univentricular AV connection, which describes a group of cardiac malformations in which the AV connection is completely or predominantly to a single ventricular chamber and includes double-inlet left ventricle, single inlet (with atresia of the left or right AV valve), and common inlet (AV septal defect with a common ventricle). SV forms of CHD are some of the most complex forms of CHD, all of which require a series of palliative operations ultimately culminating in total cavopulmonary connection ("Fontan") circulation. In this section, we review common imaging approaches to all forms of SV CHD necessary for the counseling of families about diagnosis and prognosis as well as what imaging information is needed for delivery room and neonatal management.

General Imaging Approach. As with fetal echocardiography in structurally normal hearts, visceroatrial situs, systemic venous connections, AV junction, morphology and function of the SV, and fetal heart rate and rhythm should be assessed. A segmental approach to fetal echocardiographic imaging of SV CHD is recommended to identify important associated findings.

Venous Connections. Systemic and pulmonary venous connections and atrial situs/morphology should be evaluated, as abnormalities can have implications for counseling regarding outcomes and surgical approach for SV palliation. For systemic veins, evaluation of the IVC for intrahepatic interruption (with the hepatic vein connections directly to the atrium) and the presence of bilateral SVCs should be performed. Pulmonary venous connections should be assessed given that anomalous drainage significantly influences surgical approach and outcomes.[94] For HLHS, pulmonary venous Doppler and its forward/reverse VTI ratio is used to detect left atrial hypertension Figure 21. Some centers use maternal hyperoxygenation testing to assess pulmonary vascular reactivity in the third trimester in the fetus with HLHS to improve the specificity and sensitivity of predicting need for urgent intervention. Studies have shown that lack of pulmonary vasoreactivity (<10% decline in pulsatility index of the pulmonary artery)[95] is associated with neonatal hemodynamic instability and the need for an urgent intervention to open the atrial septum after birth.

AV Junctions. SV CHD may have hypoplastic or atretic tricuspid or mitral valves, or a common AV valve that is unbalanced (primarily committed to one ventricle; Figure 22). AV valve size should be measured (see Table 13) and the degree of valvular regurgitation assessed by color Doppler. The presence of significant AV valvular regurgitation can affect both fetal well-being and postnatal outcome of SV surgical palliation.[99]

Ventriculoarterial Junction. SV lesions can have an array of ventriculoarterial connections and variations in obstruction to either pulmonary or systemic blood flow. It is therefore imperative in the fetus with SV CHD that the outflow(s) be thoroughly imaged, including orientation (normal, transposed, malposed), patency, size, and direction of blood flow. The diameter of the ascending aorta should be measured in patients with aortic stenosis or atresia. Color Doppler imaging of the arches in the 3VV, 3VT, and sagittal planes is key as retrograde flow in the aortic or ductal arch is perhaps the single most powerful indicator of need for prostaglandin infusion after birth.[100] In fetuses with aortic or pulmonary valve hypoplasia, valve stenosis can progress through gestation, and serial hemodynamic assessment of the valves and great arteries should be obtained (Tables 13 and 14).

Rhythm, Function, and Morphology. In clinical practice, function is assessed qualitatively, although quantitative measures including fractional shortening, tricuspid and mitral annular plane excursion, and sphericity index have been reported.[104] Myocardial imaging techniques such as strain and strain rate may provide a more quantitative picture of ventricular function but are not currently used routinely in clinical practice.[105] Bradyarrhythmias (e.g., complete heart block or sinus node dysfunction) are associated with left atrial isomerism and discordant AV connections. This combination carries a poor prognosis and high risk for mortality in utero.[106]

Key Points

- A complete fetal echocardiographic examination must be performed for suspected SV before a reasonable prediction of prognosis can be made.
- Demonstration of the direction of flow and any restriction at the atrial septum, ductus arteriosus (if present), and aortic isthmus (if present) is required.
- Imaging of the systemic and pulmonary venous anatomy should be performed for counseling and planning postnatal management.
- AV valve diameter or area should be measured and the degree of AV valve regurgitation assessed by color Doppler.
- For fetuses with HLHS (and variants with a lack of egress from the left atrium), the pulmonary venous forward/reverse VTI ratio should be measured to determine risk for postnatal urgent intervention at every fetal echocardiographic evaluation, with the last measurement near term.
- For CHD with HLHS (and variants with a lack of egress from the left atrium) maternal hyperoxygenation testing in the late third trimester may be considered in select cases to assess pulmonary vascular reactivity.

ii. Complex AV Connections.

Lesions affecting the AV junction may be isolated (although often are associated with aneuploidy, including trisomy 21) or may be exceedingly complex and found in combination with multiple associated defects.[107] The region of the AV junction is referred to as the "crux" of the heart and is the anatomic connection point of the atrial chamber(s) to the ventricles. Normally, the left-sided mitral valve is a bileaflet valve with chordal attachments to two separate papillary muscles on the free wall of the left ventricle. The right-sided TV is slightly more apically displaced with three leaflets and chordal attachments distributed throughout the body of the right ventricle, including the ventricular septal surface. Although the four-chamber view allows optimal visualization of the cardiac crux

Table 13 Guidance for disease-specific anatomic, physiologic, and functional evaluation for commonly encountered fetal CHD lesions

Category	Suspected when					
SV and ventricular disproportion	Axial chest view shows clear lack of two symmetric ventricular chambers					
Lesion	Key observations	Common venous variations	Specific AV valve issues	Outflow imaging	Additional measurements	Other key points
Unbalanced AVSD (right or left dominant)	Presence of AVSD features with a single AV valve, ± primum atrial septal defect, ± inlet VSD	Course and laterality of SVC and IVC, primitive azygous systems, separate hepatic connections; TAPVR	Common valve regurgitation; may be progressive En face imaging of the AV junction required	Define ventriculoarterial connections Outflow tract of the hypoplastic ventricle may be stenotic or atretic; may need to assess serially	Measure right and left AV valve diameter or area (Figure 23) Cohen index AVVI RV/LV inflow angle[96]	For assessment of balanced AV valve see below ("Common AV valve") Consider right or left atrial isomerism/ heterotaxy
Tricuspid atresia	Absent AV connection, small right ventricle, VSD	Bilateral SVC Benign A-wave reversal in DV	Mitral valve prolapse, dilation, or regurgitation	Define ventriculoarterial connections, may be transposed Outflow tract from the morphologic right ventricle may have subvalvular or valvular stenosis; Assess blood flow in arches for possible ductal dependent	Can measure VSD dimensions in two orthogonal planes	VSD obstruction can be progressive, compromising blood flow to the outflow tract off the right ventricle (leading to pulmonary stenosis, coarctation)
HLHS	Small left ventricle on four-chamber view with mitral and aortic stenosis or atresia	Bilateral SVC	TR with RV dysfunction may increase the risk for fetal death and postnatal morbidity/ mortality	Assess aortic valve patency, size of the ascending aorta, and transverse aortic arch Color Doppler interrogation of pulmonary valve Direction of flow in the distal transverse aortic arch	Measure the ascending aorta diameter, which can be predictive of postnatal outcomes after the Norwood procedure[97]	Evaluate for atrial septal restriction - pulmonary vein Doppler VTI forward/ reverse ratio < 3 predicts inadequate left atrial egress and increased likeliness for neonatal emergent atrial septoplasty[98]
Double-inlet left ventricle	Both AV valves open into the morphologic LV, four-chamber view shows "large" VSD or absent ventricular septum	Bilateral SVC	Assess size of each AV valve (one valve may be stenotic or atretic), as well as degree of valvular regurgitation AV valve may straddle into outlet chamber	Define AV and ventriculoarterial connections (may have D- or L-looped ventricles, DORV) Assess patency of outflow tract connected to the morphologic right ventricle for subvalvular or valvular obstruction		At risk for complete heart block with L-looped ventricles If left AV valve is hypoplastic or atretic, assess adequacy of atrial septal defect

Complex AV junction abnormality	Axial chest four-chamber view does not demonstrate two clearly separate AV valves with offset at crux					
Lesion	Key observations	Common venous variations	Specific AV valve issues	Outflow imaging	Additional measurements	Other key points
Common AV valve, AVSD	Crux lacks offset or AV valves appear at same level, large atrial or VSDs	Bilateral SVC, interrupted IVC, TAPVR	Morphology, balance, regurgitation; papillary muscle configuration	Normally related or transposed, subvalvular obstruction or valve hypoplasia, reverse flow in ductal or aortic arch	Cohen index AVVI RV/LV inflow angle[96]	Additional VSDs may be missed if not specifically looked for
Crisscross hearts	Two separate AV valves but not visible in same plane Confusion as to single vs two ventricles (foreshortening of right ventricle or left ventricle in axial plane)	Visceroatrial situs abnormalities bilateral SVC	TR or MR, straddle, override	Carefully define the connections, can be DORV/d-TGA, l-TGA when L-looping is present, outflow tract obstructions	AV valve annular dimensions	Postnatal physiology difficult to predict if imaging is not specifically directed toward defining connections and not relationships
Straddling valves	Four-chamber view shows VSD or overriding aorta	Bilateral SVC	Size, regurgitation, degree of valve straddle and chordal distribution	D- vs L- looped ventricles and risk for heart block, subvalvular and valvular obstruction, malposed aorta		If left AV valve is hypoplastic, at risk for left atrial hypertension
Mitral stenosis: parachute, hypoplasia	RV > LV asymmetry	LSVC Aortic stenosis, coarctation	Morphology, papillary muscles; Z score measurement	Consider DORV if VSD present and normal ascending aorta size and flow	Foramen ovale dimension; Pulmonary vein forward/reverse ratio	Foramen ovale flow direction critical in determining adequacy of mitral valve

Outflow tract lesions with normal 4-ch	Axis abnormal (<30° or >60°) LV outflow tract view abnormal or not obtainable Abnormal 3VV					
Lesion	Key observations	Common venous variations	Specific AV valve issues	Outflow imaging	Additional measurements	Other key points
TOF	Abnormal axis or LV outflow tract, with VSD and overriding vessel; abnormal 3VV	Bilateral SVC, retroaortic innominate vein Right aortic arch Vascular rings Aortopulmonary window	Rare mitral or tricuspid abnormalities and association with AVSD	3VV: smaller caliber pulmonary artery Tracheal view: presence of duct with direction of flow, absence of ductus arteriosus Continuity of branch pulmonary arteries	Branch pulmonary arteries, serial PV annulus, infundibular measurements Major aortopulmonary collateral vessels	Ductal direction at 28 wk predicts need for neonatal intervention Ao/PV valve ratio < 0.6 and PV Z score < −5 with absence of ductus arteriosus predicts need for neonatal intervention[119]
Truncus arteriosus (common arterial trunk)		Bilateral SVC Right aortic arch, vascular ring Retro-aortic innominate vein	TR, MR	Aortic vs pulmonary dominant, one vs two arches vs aortic interruption (type) PA origins	Truncal velocity, regurgitation Proximal branch PA dimensions	Right aortic arch associated with >50% risk for del22q11
DORV		Bilateral SVC	Size, regurgitation, mitral valve straddle into right ventricle TV straddling through inlet VSD AVSD AV valve: imaging best defined in short-axis or sagittal imaging	Normally related or transposed, subvalvular obstruction or valve hypoplasia, reverse flow in ductal or aortic arch, AP window, branch pulmonary artery patency and continuity	Relationship of VSD to great vessels Identifying LV to aortic pathway: size of VSD and any AV valve attachments that could preclude biventricular repair	Ventricular looping, relationship and hypoplasia
d-TGA	Abnormal 3VV, LV outflow view abnormal, parallel outflows	None	d-TGA with VSD ± pulmonary stenosis can have straddling and abnormal MV: cleft MV	Aortic and pulmonary valves should be normal size; aortic hypoplasia associated with TV hypoplasia and pulmonary valve may be bicuspid	Foramen ovale size (best defined in sagittal view) Septal excursion index[143]	Common for aortic valve to be larger than normal

(Continued)

I-TGA	None	Abnormal crux on four-chamber view; TV displacement, TR common and affect prognosis	Pulmonary or subpulmonary obstruction assessment Arch hypoplasia	AV intervals VSD presence and size	May have abnormal axis, meso- or dextroposition Serial monitoring for heart block

Progressive semilunar valve obstruction	**3VV abnormal; four-chamber asymmetry**				
Lesion	Key observations	Common venous variations	Specific AV valve issues	Outflow imaging	Additional measurements	Other key points
Aortic stenosis	Thickened valve visible in systole on LV outflow tract view, ± dilated ascending aorta on 3VV, LV hypertrophy or dilation/dysfunction on four-chamber view	Pulmonary vein Doppler with reversal in atrial systole	Mitral valve abnormalities include hypoplasia, arcade MR velocity can be used to assess LV systolic pressure and function	Aortic arch, flow reversal in transverse arch predicts severe obstruction; Doppler of aortic valve may not reflect severity with severe LV dysfunction	Aortic arch and isthmus (risk for coarctation) Mitral inflow duration MR peak velocity (if present)	Direction of foramen ovale flow must be documented Assess for elevated LA pressure due to atrial septal restriction
Pulmonary stenosis	Abnormal four-chamber view, with right atrial enlargement, RV < LV size, RV hypertrophy with decreased function; may appear normal but with color flow reversal in the ductus/main PA in 3VV	Increased A-wave reversal in systemic veins is common	Tricuspid annulus dimension, inflow duration by PW TR by color > mild associated with low risk for sinusoids	Ductal anatomy Left PA stenosis due to abnormal ductal insertion, outflow anatomy (infundibular stenosis/atresia) Ductal flow reversal suggests ductal dependent	Branch PA dimensions TR peak velocity (if present)	Assessment for coronary-cameral fistulae "sinusoids" in free wall and septum with low-velocity color Doppler
Ebstein anomaly and TV dysplasia	Right atrial enlargement and cardiomegaly, dysplastic or tethered leaflets displacement of septal ± posterior leaflets of TV To-and-fro flow through "annulus" at the level of the AV groove	Increased A-wave reversal in IVC, and ductus venosus and UV pulsations	Tricuspid annulus dimension predicts progression Tricuspid regurgitation peak velocity also predictive	Pulmonary stenosis or atresia, functional atresia; PR Hypoplasia of branch PAs Direction of ductal shunt Rare associations with coarctation	Serial measurement of: TV annulus, TR peak velocity, pulmonary flow, PR, evaluation of LV function and UA and MCA Doppler	

Lesion	Key observations	Common venous variations	Specific AV valve issues	Outflow imaging	Additional measurements	Other key points
APVS	Association with TOF or in isolation Hypoplastic PV annulus with diminutive leaflets and to-and-fro color Doppler across PV Main and branch PA dilation RA and RV dilation	May see A-wave reversal in IVC and ductus venosus with increased RA pressure	Assess for TR and PW and CW Doppler of TR velocity to assess RV systolic pressure/function	Ductus arteriosus is typically absent but presence should be assessed as affects prognosis	Serial measurement of branch PA size, biventricular function, CTR UA and MCA Doppler	

Arch abnormalities

3VV or 3VT abnormal

Lesion	Key observations	Common venous variations	Specific AV valve issues	Outflow imaging	Additional measurements	Other key points
Coarctation	RV > LV discrepancy, ascending Ao smaller than SVC, small transverse and descending/isthmus	LSVC to coronary sinus	Mitral anatomy, atrial septal flow direction	Aortic valve stenosis, bicuspid aortic valve	Aortic isthmus	False positive and false negative rates are substantial
Vascular rings	3VT has "U" shape around esophagus and trachea, instead of "V" to left of trachea	None	None	Exclude aberrant subclavian arteries, double aortic arch	As needed	Increased risk for tracheal anomalies, tracheal rings
Ductal aneurysm	Late gestation, abnormal 3VV, 3VT, or sagittal arch images with pulsatile, tortuous appearance		None	None	Aortic isthmus Diameter of arterial duct at its widest	Generally normal variant, no clinical correlate in newborn

Ao, Aorta; *AVSD*, atrioventricular septal defect; *AVVI*, AV valve index; *CTR*, cardiothoracic ratio; *DORV*, double-outlet right ventricle; *DV*, ductus venosus; *LA*, left atrial; *LSVC*, left SVC; *MCA*, middle cerebral artery; *PA*, pulmonary artery; *PV*, pulmonary valve; *RA*, right atrial; *TAPVR*, total anomalous pulmonary venous return.

This general information is not comprehensive and does not replace lesion-specific literature regarding fetal cardiac anatomy and physiology in CHD.

Table 14 Mechanisms and patterns of progression of fetal CHD

| Development/ progression	Heart disease associated with evolution	Evolution/progression
AV and semilunar valve/outflow obstruction	• Aortic stenosis • Pulmonary stenosis with intact ventricular septum • Truncus arteriosus with truncal valve stenosis • Mitral or tricuspid stenosis • TOF • Other conotruncal pathology • Coarctation • Ebstein/TV dysplasia (second-degree pulmonary obstruction/atresia)	• Increased severity of structural heart disease • Development of ventricular dysfunction for outflow obstruction in particular • Progressive ventricular dilation (not expected with AV valve stenosis) • Progressive ventricular hypoplasia • Progressive great artery dilation or hypoplasia for semilunar obstruction • Progressive AV valve regurgitation with outflow obstruction
AV and semilunar valve regurgitation	• Ebstein anomaly/TV dysplasia • APVS • Semilunar (aortic or pulmonary) • Truncus arteriosus with truncal valve regurgitation • Severe semilunar valve obstruction with AV regurgitation	• Potential for evolving heart failure, hydrops and/ or IUFD • Pulmonary regurgitation in the presence of a patent ductus can cause circular shunt and placental steal
Ventricular, valve, great artery, branch PA, arch hypoplasia	• Severe semilunar valve obstruction • Conotruncal pathology with AV valve obstruction • Coarctation	• Increased severity of structural heart disease
Foramen ovale restriction	• d-TGA • HLHS • Critical left heart obstruction • Isolated (can occur in absence of other CHD)	• Development of pulmonary congestion, nutmeg lung • Hypoxemia after birth, left atrial hypertension (HLHS), need for urgent BAS • In isolation, can give appearance of smaller left-sided structures but after delivery no consequence
Ductal constriction or aneurysm formation	• Constriction in isolation, with cyclooxygenase inhibitors/NSAIDs, associated with certain foods/herbal teas • Constriction rarely occurs in presence of CHD • Ductal aneurysm occurs at 35-40 wk in 1%-2% of all fetuses	• Isolated ductal constriction can result in development of right heart failure and hydrops and after birth PPHN[101] • Ductal constriction can be lethal in critical CHD including TGA at higher risk • Aneurysm largely close spontaneously, low risk for rupture or compression and association with connective tissue disorders
Tumor regression/ progression	• Rhabdomyoma, teratoma, others	• Depends on the tumor nature and its impact on cardiac filling, function, and rhythm (e.g., rhabdomyomas evolve in the midtrimester but regress after 32 wk)
Chamber dilation	• Valve regurgitation • High–cardiac output states • Ventricular dysfunction	• Increased severity of disease, compression of the contralateral chamber which could lead to high filling pressures and poor output • Atrial or ventricular arrhythmias • Risk for pulmonary hypoplasia
Ventricular dysfunction	• Primary myocardial disease • Structural CHD with volume load • Bilateral outflow obstruction • Arrhythmias	• Increased severity of heart disease including evolution of heart failure, hydrops and risk for IUFD
Heart failure	• Ventricular dysfunction inadequate redistribution of the cardiac preload • Significant volume or pressure overload • Arrhythmia • Cardiac compression	• More severe heart disease, fetal hydrops and IUFD

(Continued)

Table 14 (*Continued*)

Development/ progression	Heart disease associated with evolution	Evolution/progression
Arrhythmias in fetal CHD	• Higher risk lesions include Ebstein/TV dysplasia • Any with atrial or ventricular dilation • Tumor • Cardiomyopathies	• More severe if associated with structural CHD or myocardial disease
Reduction in size of VSD	• Usually with simple isolated ventricular or muscular VSDs but can occur in other CHD	• Decreased severity or resolution of heart disease[102,103]

BAS, Balloon atrial septostomy; *IUFD*, intrauterine fetal demise; *PA*, pulmonary artery; *PPHN*, persistent pulmonary hypertension of the newborn; *TGA*, transposition of the great arteries.
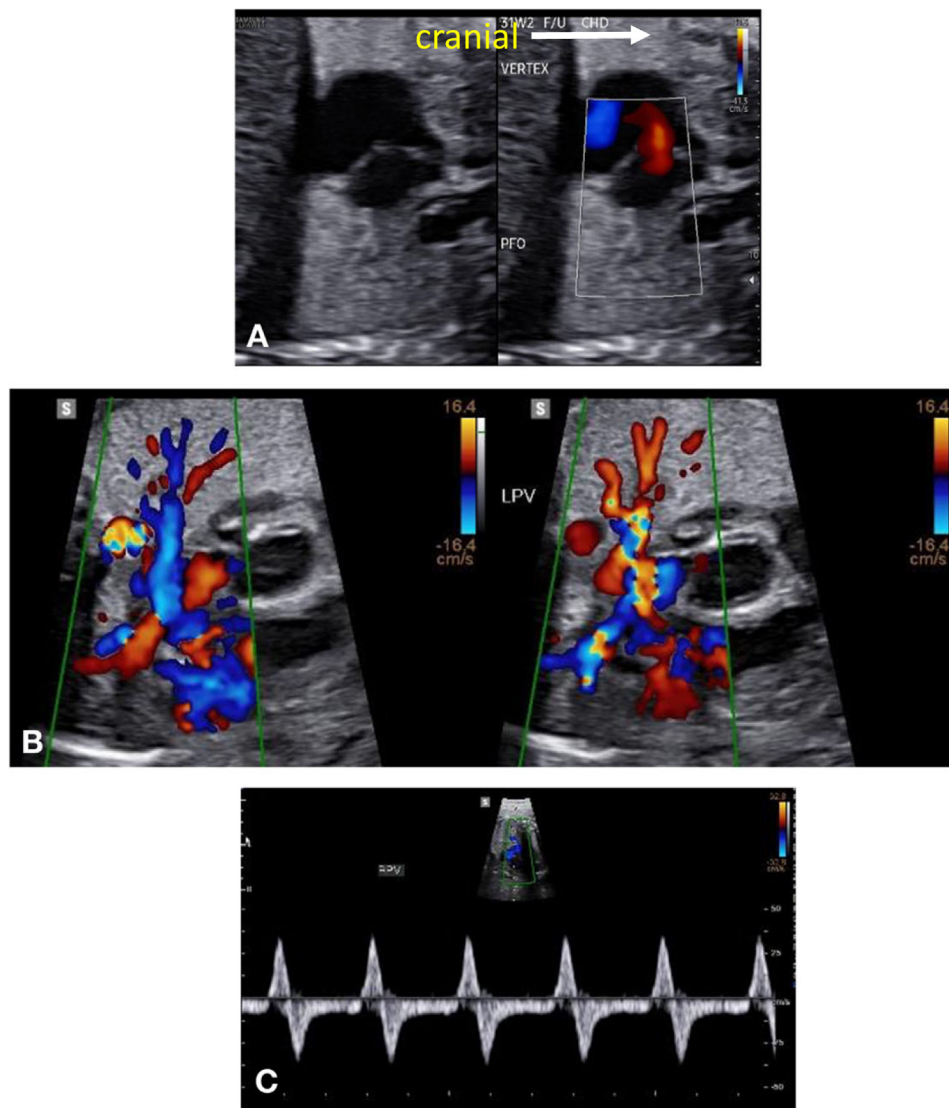
Figure 21 Complete fetal echocardiographic assessment of the degree of atrial septal restriction in a fetus with left heart obstruction. **(A)** Left-to-right flow across the interatrial septum is shown. **(B)** Pulmonary vein enlargement and to-and-fro flow are demonstrated. **(C)** PW Doppler showing abnormal forward/reverse flow pattern consistent with moderately elevated left atrial pressure.
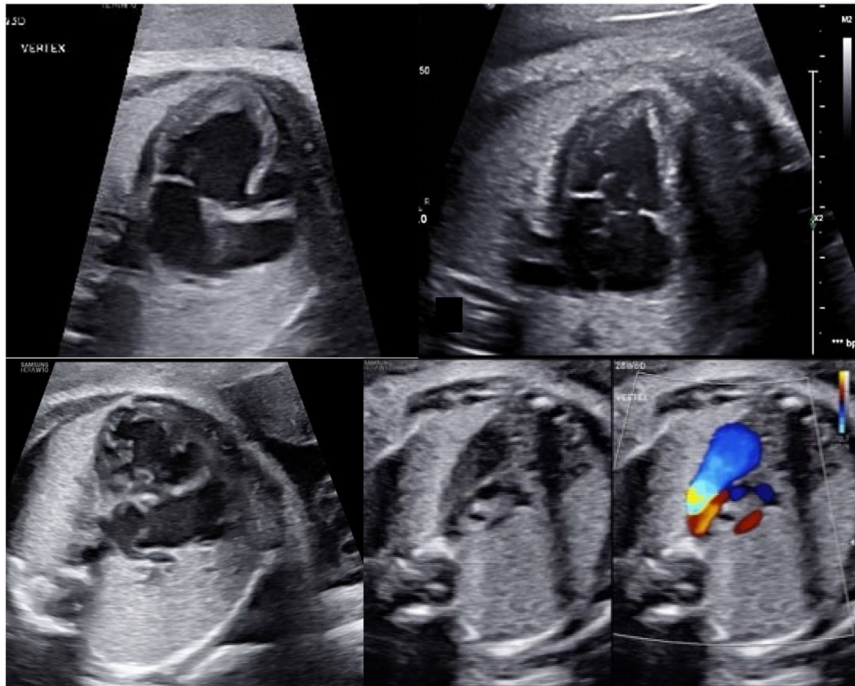
Figure 22 Examples of SV heart lesions on four-chamber view and 3VT with and without color Doppler. See also Video 10 (available at www.onlinejase.com).

and AV valve morphology as well as the presence of VSDs and outflow tract abnormalities, additional imaging is needed when an abnormality is suggested. CHD involving abnormal or malformed AV valves have a high incidence of ventricular disproportion, associated defects in the ventricular septum, and maldevelopment of the outflows and great arteries that may result in the need for SV palliative surgical approach.

General Imaging Approach. In the presence of AV valve abnormalities, the evaluation of overall fetal cardiac structures, including ventricular size and function, as well as AV balance over the ventricles, is essential. Following initial situs determination, the four-chamber view is the optimal starting point for evaluating these defects. In this view, care should be taken to align the AV septum in a more perpendicular plane to clearly define the AV valve morphology and leaflet insertion points, as well as chordal distribution and insertion. Ventricular looping and alignment are determined. The AV valve annular dimensions should be measured in early diastole at the valve hinge points. As outflow tract and great artery abnormalities are common in this group of anomalies, it is imperative that cephalad sweeps are obtained to define both the outflow tract and great artery anatomy, size, and patency. A sagittal view of the fetal thorax best facilitates short-axis imaging of the AV valve morphology, chordal attachments, and papillary muscle arrangement and morphology (Figure 23). Abnormal AV valve inflow or regurgitation should be assessed with color Doppler. PW Doppler evaluation of inflow patterns is essential. However because the fetal circulation allows redistribution of flow such that stenotic valve severity may be underestimated by peak velocity, annulus size in right AV valve disease and hemodynamic effects such as left-to-right shunting at the atrial level in left AV valve obstruction should be documented.

AV Septal Defects. A common AV connection or AV septal defect is visualized in the four-chamber view, noting the abnormal crux of the heart in which the septal leaflet of the right-sided AV valve is at the same level as the left-sided AV valve anterior leaflet when closed (Figures 23 and 24). This finding should prompt assessment of the left AV valve en face for cleft or common orifice, the atrial septum for primum defects, and inlet ventricular septum. If by 2D or color flow imaging the valve is predominantly committed to either of the ventricles with or without ventricular size disproportion, the area of the common AV valve apportioned over each ventricle can be measured from a parasagittal view in diastole (Figure 23), and an AV valve index can be calculated as left/right valve area[108] or the RV/LV inflow angle can be measured from an apical four-chamber view (Figure 24). The outflow tracts should be assessed to determine the presence of obstruction from the conal septum, subvalvular fibrous tissue, or AV valve or accessory tissue.

Congenital Mitral Valve Abnormalities. Mitral valve abnormalities may result in decreased flow through the left heart and reversal of flow at the foramen ovale. Two-dimensional imaging and color Doppler interrogation of the mitral valve and atrial septum should be performed, as well as PW Doppler of the pulmonary veins to assess for increased left atrial pressure. Isolated mitral valve dysplasia and mitral valve "arcade"[109,110] are abnormalities that cause significant mitral regurgitation, although any degree of mitral valve regurgitation is abnormal. Associated LV outflow obstructive lesions should be considered when mitral regurgitation is present.[111]

Straddling/Overriding AV Valves and Crisscross Heart. "Straddling AV valve" is a term applied when the chordal apparatus passes through a VSD and attaches to the wall or a papillary
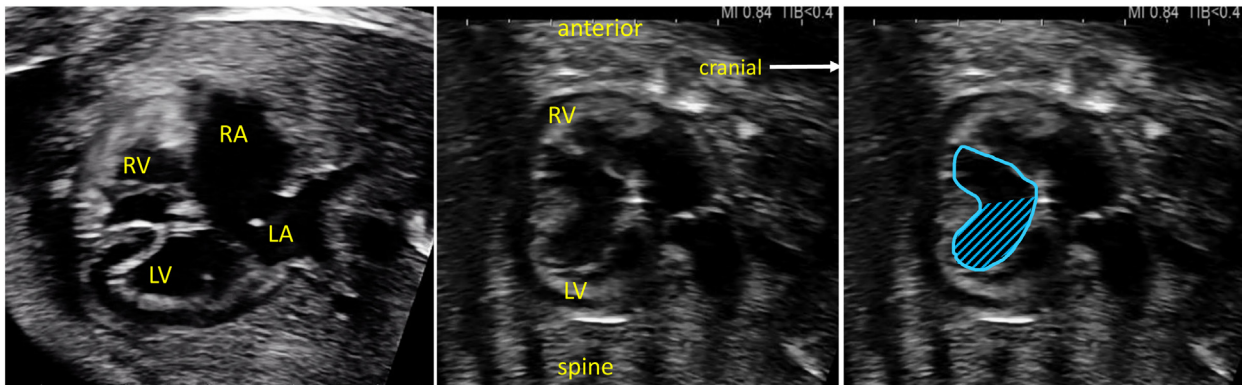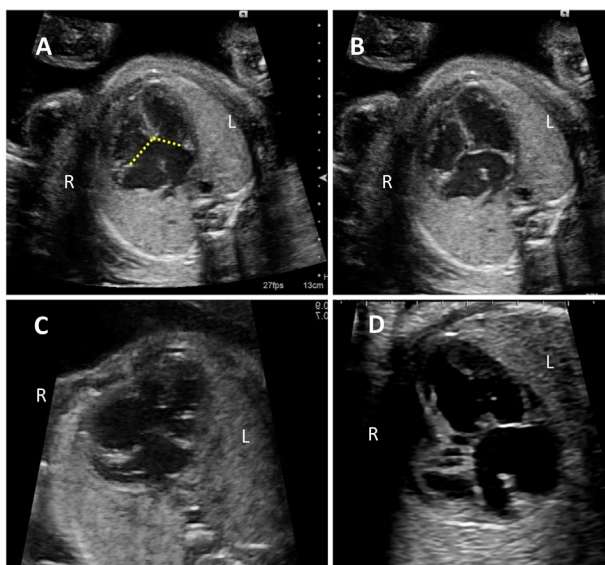
Figure 23 Quantification of degree of imbalance in AV septal defect. The area of the common AV valve apportioned over each ventricle can be measured from a parasagittal view in diastole and an AV valve index (AVVI) can be calculated. Depending on the equation used, a ratio of about 2:1 or higher suggests need for SV palliation. *LA*, Left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle.

muscle structure of the opposite ventricle. An overriding AV valve is characterized by a valve annulus that is anatomically positioned over the interventricular septum and opens into both ventricles.[112] In criss-cross AV relations or "twisted" AV connections, the apparatus of each of the two AV valves cross over each other, usually such that the atrium on the right side opens into the left-sided ventricle, and the atrium on the left side opens into the right-sided ventricle. Additionally, the ventricular looping may be such that there is AV concordance or discordance.[113] In the setting of AV discordance, there is almost always a VSD with a high incidence of AV valve straddling through the defect. Careful imaging of the valves including dimensions, function, and attachments using a combination of 2D, color Doppler, and PW Doppler echocardiography from multiple views (Figure 25) should accompany the diagnosis. In the presence of either straddling or overriding AV valves, repair by surgical closure

Figure 24 Balanced **(A, B)** and unbalanced right-dominant **(C)** and left-dominant **(D)** AV septal defect appearance in four-chamber axial views. Degree of imbalance can be quantified by measurement of the valve angle **(A,** *dotted line***)** with a smaller angle being associated with increased imbalance.

of the VSD may not be possible and may necessitate a palliative SV surgical approach.

Key Points

- In the presence of AV valve abnormalities, all fetal cardiac structures should be evaluated, including assessment of ventricular size and function as well as balance of the AV valve.
- In the fetus with suspected abnormality of the AV junction, nonstandard planes and en face imaging should be used to perform a detailed evaluation of the anatomy of the valves, chordae, and their insertions.
- A complete Doppler evaluation of the fetal heart including assessment of the AV valve(s) for stenosis, regurgitation, and direction of flow at the atrial septum and ductus arteriosus must be performed at each examination.
- Additional lesions that might influence surgical approach must be delineated to allow determination of ultimate surgical palliation strategy (SV palliation vs biventricular repair).

iii. "Look-Alike" Outflow Tract Lesions. Lesions involving the outflow tracts are identified on obstetric anatomic ultrasound screening using the outflow tract, 3VV, and 3VT views. Often with outflow tract anomalies there are minimal abnormalities noted in the four-chamber view. Although many of these defects may appear similar on ultrasound screening, complete fetal echocardiographic evaluation provides detailed information, which then influences perinatal planning, interventions needed at birth, and long-term management and prognosis depending on precise physiologic and anatomic diagnosis.

General Imaging Approach. CHD involving the outflow tracts are often associated with other anomalies, including abnormalities of the systemic or pulmonary venous connections, AV valve, atrial or ventricular septa, branch pulmonary arteries, and aortic arch. As with other CHD, visceroatrial situs, systemic venous connections, the AV junction, ventricular morphology and function, and heart rate and rhythm should be assessed carefully. Imaging views and sweeps from the four-chamber view to the outflow tract, 3VV, and 3VT views will detail the segmental anatomy, including AV and ventriculoarterial connections, and the relationship of the great arteries. The short-axis and coronal views can be used to help localize any defects in the ventricular septum and determine the relationship of the great arteries to each other and to the ventricles.

Lesions with a VSD and Overriding Great Vessel. The differential diagnosis for an outflow tract view that includes a VSD and an overriding great vessel includes tetralogy of Fallot (TOF) and
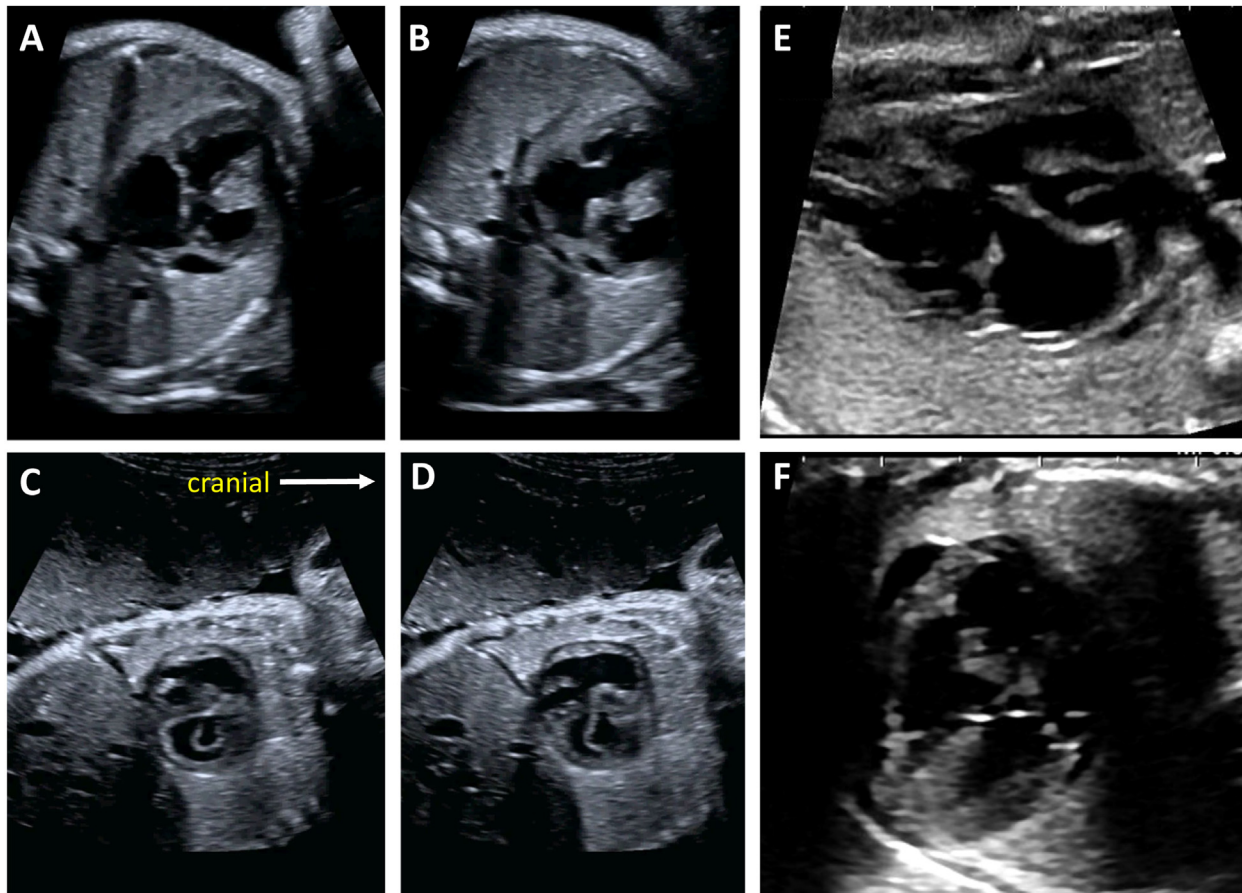
Figure 25 Straddling left AV valve that complicates or precludes biventricular repair strategy. **(A-D)** d-TGA with VSD. Fetus with atypical mitral valve cleft and straddling of the mitral valve chordae through an outlet VSD. **(A)** and **(B)** (and Video 11 available at www.onlinejase.com) show the axial plane with abnormal opening of the mitral valve toward the septum, while the valve anatomy is better seen in **(C)** and **(D)** (and Video 12 available at www.onlinejase.com) where a parasagittal plane demonstrates en face views of the mitral valve in two different diastolic frames. In practice, this anatomy is better demonstrated using video clips and live scanning. **(E, F)** Double-outlet right ventricle with subpulmonary VSD and straddling mitral valve, making a neonatal Damus-Kaye-Stansel procedure more feasible than an arterial switch operation.

pulmonary atresia with VSD, double-outlet right ventricle, and common arterial trunk (Figure 26). TOF is characterized by an overriding aorta and varying severity of subpulmonary and pulmonary valve obstruction, ranging from mild pulmonary stenosis to pulmonary atresia or dilation in the presence of absent pulmonary valve syndrome (APVS). TOF may also be seen in association with AV septal defect.[114] The branch pulmonary arteries must be identified at their origins, as discontinuity, segmental stenosis, and major aortopulmonary collateral vessels can coexist with an apparently patent outflow tract and/or ductus arteriosus and have implications for management and long-term outcome[115-119] (Table 13). Imaging for associated arch anomalies and thymus allow tailored counseling regarding risk of 22q11 deletion.[120] In TOF with absent pulmonary valve, ventricular dysfunction predicts fetal and perinatal morbidity and mortality.[121]

In double-outlet right ventricle, the VSD can be subaortic, subpulmonary, doubly committed, or remote, and therefore it is necessary to determine the orientation and relative sizes of the aorta and pulmonary artery as they exit the ventricle, as well as their relationship to the VSD. If the VSD is subpulmonary, an anterior aorta may be obstructed in the subaortic region, necessitating careful aortic arch imaging with 2D and color Doppler as coarctation often accompanies this

situation. If the VSD is subaortic, the anatomy and postnatal physiology may be like that of TOF.

Truncus arteriosus, also called common arterial trunk, is characterized by a single arterial trunk that gives rise directly to the coronary arteries, aorta, and one or both pulmonary arteries. Truncal valve function, especially significant regurgitation, can affect in utero survival.[77] A fair amount of anatomic variability exists with dominant commitment to the pulmonary circulation (branch pulmonary arteries from the common trunk, smaller aortic partition with or without arch interruption) or to the aortic circulation (dominant ascending aorta with branch pulmonary arteries arising together or separately from the common trunk).[122] Although the ductus arteriosus is often absent, one may be present in all forms of common arterial trunk, with the more pulmonary-dominant variants and truncus with interrupted aortic arch entirely dependent on ductus arteriosus patency for perfusion of the lower body. These fetuses are dependent on prostaglandin E_1 initiation at birth.

Lesions with LV Outflow Giving Rise to a Posterior Pulmonary Artery. Both d-TGA and l-transposition of the great arteries (l-TGA) demonstrate a similar posterior pulmonary artery arising from the LV outflow tract (Figure 26). A diagnosis of d-TGA should prompt
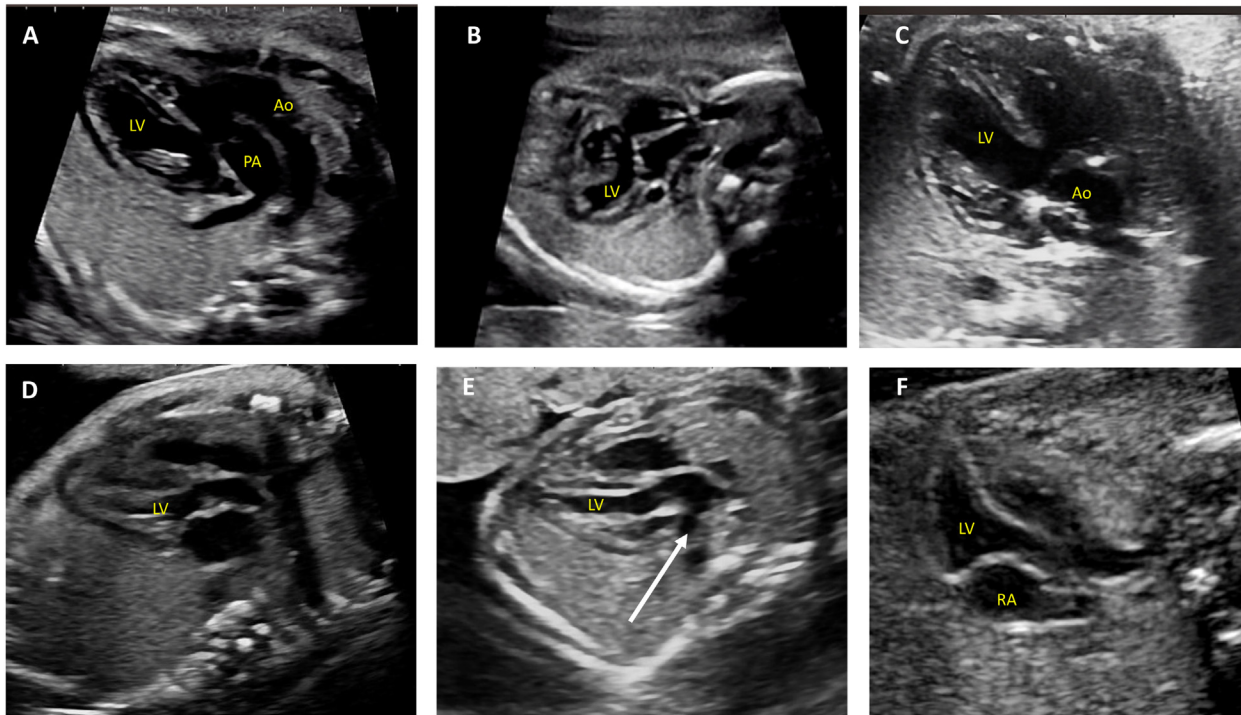
Figure 26 Abnormalities seen on evaluation of the left ventricular outflow tract (LVOT) that may not be visible in the four-chamber view. **(A)** shows a posterior vessel that gives rise to a pulmonary artery, with a subpulmonary VSD and an anteriorly positioned aorta (Taussig-Bing anomaly). Image **(B)** is a fetus with a common arterial trunk, **(C)** is a fetus with TOF, **(D)** shows d-TGA with parallel outflow tracts. **(E)** demonstrates a vessel arising from the LVOT branches early *(arrow)* and thus is a pulmonary artery, making this also a diagnosis of d-TGA. **(F)** l-TGA, showing that the right atrium (with SVC visualized) is connected to the left ventricle and the LVOT gives rise to the posterior pulmonary artery. *Ao*, Aorta; *LV*, left ventricle; *PA*, pulmonary artery; *RA*, right atrium. See also Videos 13-16 (available at www.onlinejase.com).

late-gestation imaging of the interatrial septum as well as delivery in a setting that permits enlargement of the interatrial septal defect if shunting is inadequate. l-TGA may be identified in the four-chamber view by visualization of an abnormal crux and the moderator band in the apex of the left-sided right ventricle. The combination of AV and ventriculoarterial discordance results in physiologically corrected flow. The incidence of VSD in l-TGA may be 60% to 70%, although the type and location of VSD is variable.[123] TV dysplasia and Ebstein malformation can be associated with l-TGA, as is aortic coarctation. Rhythm abnormalities and congenital heart block can be noted during fetal life and affect survival; therefore, serial evaluation is indicated.[124]

Key Points

- Suspected outflow tract abnormalities should be evaluated on fetal echocardiography before discussing prognosis, associated abnormalities, and additional testing (including genetic testing) with the patient.
- For suspected outflow tract abnormalities, complete evaluation includes imaging the branch pulmonary arteries at their origins. Discontinuity, segmental stenosis, and major aortopulmonary collateral vessels can coexist with an apparently patent outflow tract and/or ductus arteriosus; direction of ductal flow by Doppler must be documented.
- Aortic arch laterality must be documented at least once during gestation, and an attempt made to delineate branching pattern especially if the aortic and ductal arches are on opposite sides of the trachea.
- The aortic arch is at risk in any lesion with subaortic obstruction or subpulmonary VSD; the arch should be imaged by 2D and Doppler echocardiography in the axial and sagittal planes to evaluate patency and dimensions.
- Any fetus with d-TGA should have late-gestation assessment of foramen ovale patency.
- In pregnancies involving fetal diagnosis of l-TGA, serial echocardiographic evaluation of the fetus should be offered, to identify progressive AV valve regurgitation and rhythm abnormalities, including complete heart block.

iv. Progressive Lesions, Obstructive and Regurgitant. What appears initially to be mild fetal heart disease can evolve to more severe structural and functional disease (Tables 13 and 14). Lesions with the greatest potential for progression in utero include semilunar valve obstruction with an intact ventricular septum and lesions associated with significant valvular regurgitation. Fetal echocardiographic evaluation of such lesions requires comprehensive definition of segmental and valve-specific anatomy and of biventricular and AV valve function, and, given their progressive nature, serial assessments are essential. With progression, these lesions can result in cardiovascular compromise; therefore, routine Doppler interrogation of systemic venous, ductus venosus, umbilical arterial and venous, and middle cerebral arterial flows is essential.

Semilunar Valve Obstruction with an Intact Ventricular Septum. Fetal aortic and pulmonary valve stenoses span a broad spectrum of severity. More severe obstruction, particularly early in gestation, may lead to progressive ventricular hypoplasia[125,126] or result in fetal hydrops if there is compromised filling of the contralateral ventricle.[127] Evaluation of semilunar valve morphology, size, patency, and competency, including the use of color, PW and CW Doppler, is important in making the diagnosis and determining lesion severity (Figure 27). Growth of the great artery downstream from the obstruction may be affected, resulting in dilation or hypoplasia in longer-standing obstruction, necessitating repeat measurements. Retrograde flow in the ipsilateral arch heralds critical semilunar valve obstruction at any gestational age[100] and should
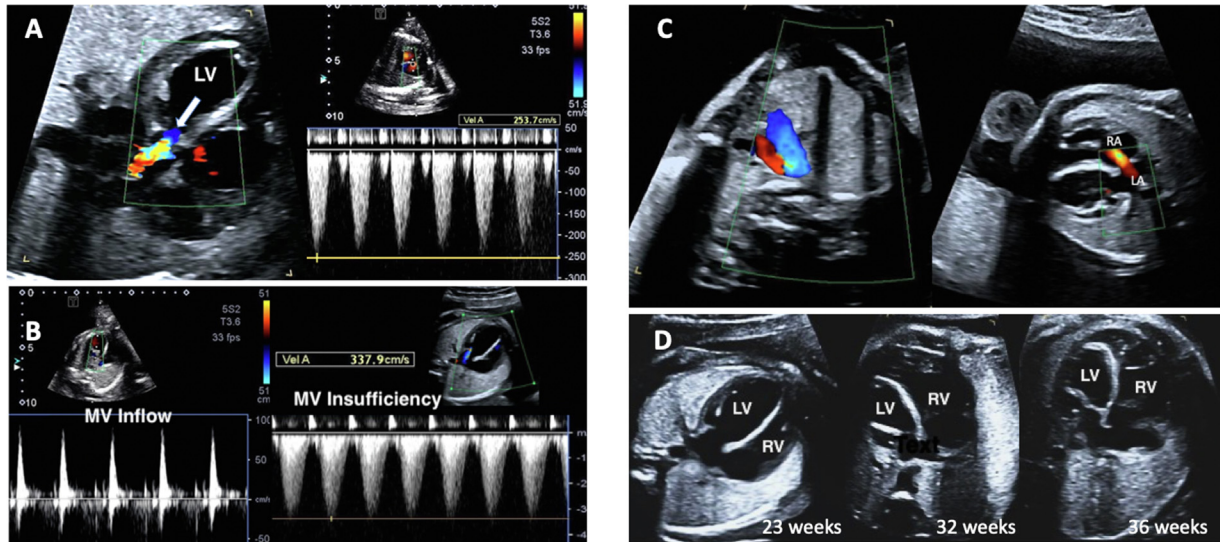
Figure 27 Progression of aortic stenosis. Severe aortic stenosis in a 23-week fetus with a dilated and hypokinetic left ventricle (LV) with endocardial fibroelastosis. **(A)** By color Doppler, aliasing beginning at the aortic valve *(arrow)* is demonstrated *(left)* with a gradient by spectral Doppler of 25 mm Hg *(right)*. **(B)** Mitral valve (MV) inflow is uniphasic and of short duration *(left)*, and the jet of mitral regurgitation suggests a high LV systolic pressure (46 mm Hg pressure gradient plus the left atrial pressure; *right panel*). **(C)** The presence of retrograde aortic arch flow *(red, left)* and left-to-right atrial flow *(red, right)* are additional findings that predict progressive left heart hypoplasia evolving through gestation due to reduced flow through the left heart. **(D)** The LV becomes progressively more globular and reduced in length, and is no longer apex forming by 36 weeks. *RV*, Right ventricle.

be looked for repeatedly through gestation if not present at initial evaluation (Figure 28). The severity of the stenosis, the likelihood of progressive valvular hypoplasia, and candidacy for prenatal intervention may be determined by assessing ventricular size (end-diastolic diameter and length), identifying the apex-forming ventricle as well as the presence of hypertrophy and/or endocardial fibroelastosis,[128] and assessing both systolic and diastolic function of the ipsilateral ventricle.[128-131] CW Doppler of mitral and TV regurgitation provides key hemodynamic insight into the severity of aortic and pulmonary stenosis, respectively, as well as ventricular function, with higher velocities in more severe disease suggesting preserved ventricular function. Conversely, low regurgitant jet velocities could suggest less severe obstruction, severe myocardial dysfunction, or indicate that the primary lesion is myocardial or related to severe AV valve regurgitation (see below) and not due to outflow obstruction. Assessment of the contralateral ventricle and AV valve function is critical in predicting potential for cardiovascular compromise.

In addition to common features, there are unique aspects to fetal aortic and pulmonary valve obstruction. In aortic stenosis the size of and flow through the foramen ovale should be assessed. High left atrial and LV filling pressures in more severe obstruction results in left-to-right atrial shunting and may lead to atrial restriction with left atrial hypertension, ultimately affecting pulmonary vein Doppler flow patterns.[132] In critical pulmonary outflow obstruction, particularly in valvular atresia or near atresia with a small right ventricle, coronary artery fistulae may be present and are suspected in the presence of to-and-fro flow in the RV free wall and septum.[133]

Key Points

- Serial evaluation of the heart should be performed if valve stenosis is suspected, including assessment of ventricular size and function, as well as the presence of endocardial fibroelastosis.
- Direction of flow in the ductal and aortic arches must be documented at every examination; abnormal flow direction heralds critical postnatal obstruction.
- Doppler pressure gradients may be measured and reported but are not necessarily indicative of stenosis severity.
- Direction of flow at the atrial septum must be documented at every examination.
- Evaluation of biventricular function is recommended in aortic and pulmonary valve stenosis, including but not limited to Doppler venous and ventricular inflow patterns, time intervals, presence of valve regurgitation, fractional area change, fractional shortening, and cardiothoracic ratio.

Severe Atrioventricular and Semilunar Valve Regurgitation. Significant AV or semilunar valvular regurgitation is associated with increasing volume load, manifested by progressive cardiomegaly, and is not well tolerated by the fetal circulation. Volume load and consequent biventricular dysfunction can result in an inability to redistribute the cardiac preload, leading to hemodynamic instability, increased filling pressures, and hydrops.[121,128] Fetal TV disease (TVD) and APVS are the most common lesions with such pathophysiology, although common AV valve regurgitation (discussed in previous sections) may also be progressive and can affect prognosis, especially in patients with left or right atrial isomerism (heterotaxy).

General Imaging Approach. The first clue to the presence of TVD, as observed in Ebstein anomaly and TV dysplasia, is significant right atrial dilation with or without RV dilation. Evaluation of TV anatomy in four-chamber and short-axis views elucidates the underlying
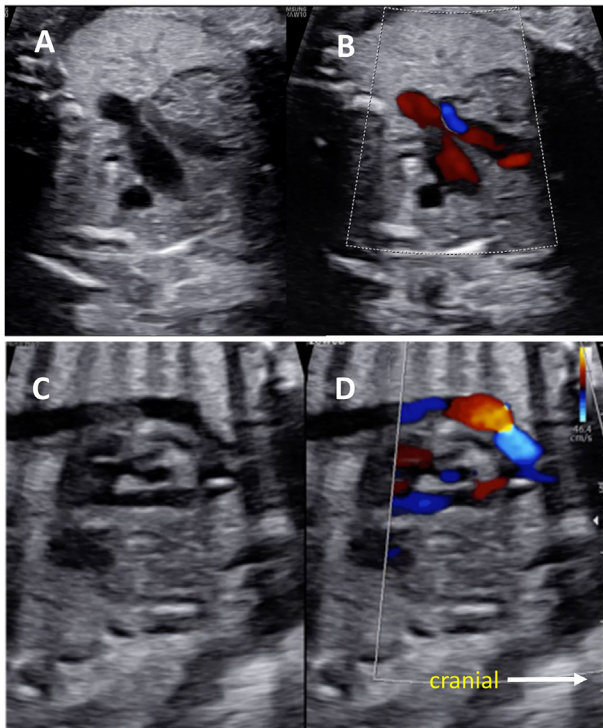
Figure 28 Retrograde flow in the arch ipsilateral to an obstructed semilunar valve. *(Upper panel)* Severe RV outflow tract obstruction, in which the 3VT appears somewhat normal but color Doppler shows retrograde flow in the ductus arteriosus *(blue)*. See also Video 17 (available at www.onlinejase.com). *(Lower panel)* Sagittal view of the aortic arch appears normal on the 2D image, but color Doppler imaging reveals retrograde *(blue)* systolic flow in the transverse arch.

diagnosis (Table 13). TV regurgitation severity can be assessed by measuring the vena contracta width relative to the valve annulus. Doppler interrogation (PW and CW) of the insufficient jet velocity is critical in excluding primary pulmonary obstruction (high velocities) and in determining the right ventricle's ability to generate a systolic pressure. Assessment of pulmonary valve flow and patency is essential as this lesion can be associated with anatomic or pseudo-pulmonary atresia, the latter where the pulmonary valve does not open as the RV pressure cannot exceed the systemic pressure (Figure 29). Structural or functional pulmonary atresia in severe TVD is always associated with retrograde ductal flow, which can develop at any point in gestation, therefore flow direction in the ductus arteriosus must be documented serially through gestation. Pulmonary valve and main pulmonary artery hypoplasia are common due to reduced RV outflow. Branch pulmonary arteries should be imaged and measured as arterial hypoplasia correlates with associated lung hypoplasia.[129] LV morphology (eccentricity, presence or absence of noncompaction), size, and function should be assessed as LV dysfunction in TVD can lead to cardiovascular compromise.[128,130] Lack of augmented radial displacement, reduced circumferential strain, and the presence of LV dyssynchrony are also key features in severe TVD that likely reflect suboptimal ventricular-ventricular interaction,[128] although these modalities are still considered to be investigational. Cardiac rhythm should be assessed to exclude atrial ectopy

and supraventricular tachyarrhythmias, which are common.[131] Finally, it is important to assess for other structural defects including aortic coarctation and VSD.

APVS is most often associated with TOF with a subaortic VSD but may also occur with an intact ventricular septum. Given the implications for surgical management as well as associated 22q11.2 deletion, differentiating between the two is critical. Progressive RV dysfunction is predictive of perinatal mortality.[121] In addition to assessing the branch pulmonary artery dimensions and cardiothoracic ratio, evaluation of cardiac position is important as alteration in APVS suggests the presence of airway obstruction with hyperinflation of the lung ipsilateral to the obstructed airway.[121,134] A ductus arteriosus is rarely present but may be found in association with discontinuous pulmonary arteries or in fetuses without a VSD. Left heart anatomy and LV function should be assessed as LV dysfunction has also been shown to independently predict fetal demise.[121] Assessment of the aortic arch position in TOF or APVS is important as it may contribute to airway compression when the branch pulmonary artery ipsilateral to the arch is aneurysmal.

Key Points

- Serial evaluation of valvular regurgitation should be performed, as it may worsen with advancing gestation.
- Progressive cardiomegaly is a common feature of fetal heart disease associated with significant AV and semilunar valve regurgitation.
- In severe Ebstein anomaly or severe TV dysplasia, PW and/or CW Doppler interrogation of the tricuspid regurgitation jet should be performed as a measure of RV function because it can be used to predict clinical outcome.
- Assessing cardiac position in TOF with APVS should be performed, given that an abnormal axis due to segmental lung fluid trapping may be associated with significant airway obstruction after birth and can be used to predict outcome.

V. Isolated Arch Abnormalities. With the introduction of the 3VV and 3VT views into obstetric anatomic ultrasound protocols over the past 10 years, there has been an increase in referrals for suspected great artery and arch abnormalities despite an otherwise normal appearing four-chamber and outflow tract evaluation.[135] In particular, the 3VT view has a characteristic appearance with two similarly sized arches forming a "V" to the left of the trachea (Figures 6D and E). Size discrepancy, abnormal color flow direction, "U" shape, and separation of the SVC and trachea with one or two intervening arches will alert the examiner to abnormalities including coarctation, ductal aneurysm, right aortic arch, and vascular rings, including double aortic arch and right aortic arch with left ductus arteriosus with or without aberrant subclavian artery (Figure 30).

General Imaging Approach. Assuming intracardiac abnormalities have been excluded, imaging is straightforward. A combination of axial, sagittal, and parasagittal imaging with 2D and color Doppler to follow the course of both the ductal and aortic arches with attention to the origins of the head and upper extremity vessels will lead to a correct diagnosis. The use of color and power Doppler in the 3VT view is helpful in detecting an aberrant subclavian artery, retrograde flow in vessels, or antegrade flow in the smaller of two arches in double aortic arch. A pulmonary artery sling can also be recognized in this view; although a sling is not a vascular ring, these patients may have airway abnormalities, including tracheal rings, and delivery should occur in an appropriate setting to allow assisted ventilation if needed.

Key Points

- Aortic and ductal arch laterality (sidedness) should be documented as part of every fetal echocardiographic examination.
- Vascular anatomy should be assessed in the 3VT view given that vascular rings, slings, and right aortic arch are readily detectable by fetal echocardiography.
- Attempts should be made to diagnose aberrant subclavian artery origin, especially if an aortic arch laterality abnormality is suspected.

D. Imaging-Based Risk Assessment and Delivery Planning for the Pregnancy Affected by Fetal Heart Disease

Fetal echocardiographic diagnosis is central to the care of fetuses with CHD, or cardiac rhythm or function disorders as it allows the opportunity for complete prenatal counseling of the family, arrangement of psychosocial supports, provision of comprehensive perinatal care, and coordination of delivery planning. Once a diagnosis of CHD is made, the fetal cardiologist must work with the obstetric care provider(s) to determine the implications for the fetus and pregnant person during the remainder of gestation and the need for follow-up visits as well as the anticipated neonatal level of care necessary.[136] Depending on the type and severity of fetal heart disease it may be prudent to have serial fetal echocardiographic follow-up to assess changes in fetal status and guide anticipated postnatal management.[137,138] Tables 14 and 15 detail the various situations in which serial fetal echocardiographic imaging is recommended because of the risk for CHD progression. There are certain forms of
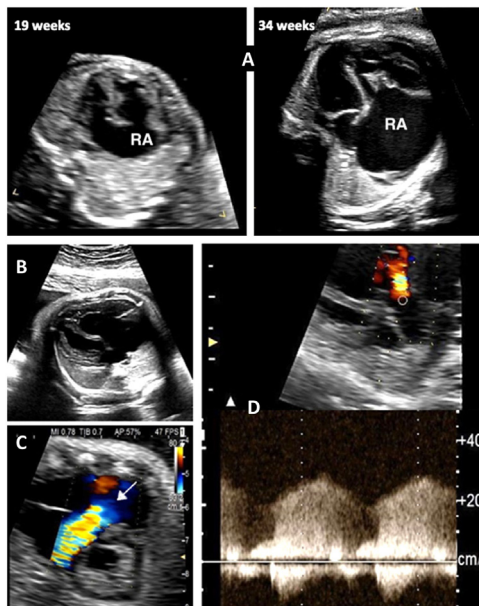
Figure 29 Fetal Ebstein anomaly and TV dysplasia. **(A)** In this fetus severe tricuspid regurgitation is associated with progressive cardiomegaly, especially right atrial (RA) dilation, as demonstrated in these serial images at 19 and then 34 weeks, and can be associated with the evolution of heart failure/hydrops. **(B-D)** Images from a fetus with severe Ebstein anomaly demonstrating severe cardiomegaly in the four-chamber view. **(C)** A broad jet of tricuspid regurgitation *(arrow)* from the sagittal view with the regurgitant jet originating well below the TV annulus and even toward the RV outflow tract due to septal leaflet displacement. There was also continuous pulmonary regurgitation by color and CW Doppler. **(D)** The latter finding suggests the right ventricle is unable to generate sufficient pressure to open the pulmonary valve.
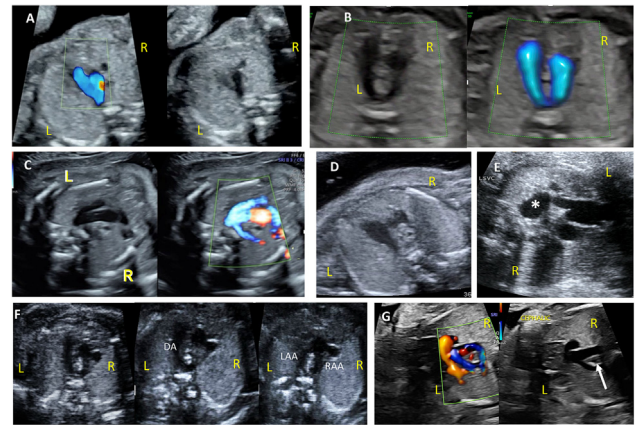
Figure 30 Isolated arch abnormalities. **(A)** Normal heart at 13 weeks, showing a 2D image *(right)* and the added value of power Doppler *(left)* to visualize the "V" to the left of the SVC and trachea. **(B)** Right aortic arch with left ductus arteriosus. **(C)** Isolated aberrant right subclavian artery seen coursing behind the esophagus, more clearly with color Doppler (see also Video 18 available at www.onlinejase.com). **(D)** Isolated coarctation was suspected when transverse aorta was noted to be smaller than the ductal arch (see also Video 19 available at www.onlinejase.com). **(E)** Ductal aneurysm *(asterisk)* in the third trimester. **(F)** Double aortic arch with three "arches" visualized in three slightly different planes (see also Video 20 available at www.onlinejase.com): ductal arch (DA), left aortic arch (LAA), and right aortic arch (RAA). The right aortic arch is dominant. **(G)** Another example of double aortic arch with color Doppler showing the two aortic arches encircling the trachea and esophagus *(arrow)* and the flow in the innominate vein in red as it courses anterior to both arches toward the SVC on the right.

critical CHD that require cardiac intervention immediately after birth to decrease the risk for neonatal death and/or severe complications. To better communicate the risk level to the care team and to coordinate appropriate, prompt postnatal care for these patients it is recommended to use image-based risk stratification systems paired with specific level of care plans for prenatally diagnosed CHD.[2] Although making a detailed prenatal diagnosis is key, it is the coordinated perinatal plan of care that has the greatest potential to influence outcomes for these patients. There remain gaps in the ability of fetal echocardiography to predict the need for immediate postnatal intervention for such lesions as d-TGA with restrictive atrial septum.[139,140] The adjunctive use of maternal hyperoxia testing to simulate postnatal physiology has been studied, however data are incomplete thus far. Fetal magnetic resonance imaging can be done to measure lung volumes or fluid trapping in TOF with absent pulmonary valve, or to assess for evidence of lymphangiectasis in HLHS with a restrictive or intact atrial septum or obstructed total anomalous pulmonary venous return. This may be useful in prognosticating outcome and planning.[141,142]

Key Points

- Maternal, fetal, and neonatal care providers must work together to determine the implications for the fetus and pregnant person during the remainder of pregnancy and the perinatal/neonatal period once a diagnosis of fetal CHD is made.
- Serial fetal echocardiographic follow-up to assess changes in fetal status and guide anticipated postnatal management is indicated for specific cardiac diagnoses at risk for postnatal hemodynamic compromise.
- Risk stratification on the basis of fetal echocardiography may be used to facilitate communication and standardize clinical approach to perinatal management of the pregnant person and fetus.

Table 15 Recommendations regarding imaging-based risk assessment and follow-up imaging for specific CHD diagnoses

Indication for follow-up fetal echocardiography	Suggested follow-up (discretion of cardiology and obstetrics providers)	Key fetal echocardiographic imaging findings to assess	Example diagnosis
Fetal CHD at risk for CHF	Every 2-4 wk	Comprehensive cardiac function assessment*	HLHS with MR, tetralogy with absent pulmonary valve, Ebstein anomaly, AVSD with AV valve regurgitation, cardiac tumors
Fetal CHD at risk for arrhythmia	Every 2-4 wk	Rate and rhythm documentation	Long-QT syndrome, l-TGA, LAI
Fetal extracardiac anomaly at risk for hydrops	Every 2-4 wk	Comprehensive cardiac function assessment*	CPAM, SCT, TTTS, arteriovenous malformation TRAP, anemia
Fetus at risk for postnatal cyanosis	Every 4-8 wk + late third trimester	Atrial septal excursion and foramen opening, ductal shunting pattern, consider maternal hyperoxygenation testing	d-TGA/IVS, d-TGA VSD, DORV with subpulmonary VSD, TOF
Fetus at risk for postnatal pulmonary venous obstruction	Every 4-8 wk + late third trimester	Doppler forward/reverse VTI ratio; atrial septal position and thickness, vertical vein Doppler peak velocity and waveform, consider maternal hyperoxygenation testing	HLHS/RAS, obstructed TAPVR, cor triatriatum
Fetal tachycardia	Every 2 weeks unless well controlled	Assess heart rate and rhythm, heart size, hydrops, CHF	Atrial flutter, SVT
Fetus at risk for postnatal respiratory compromise	Every 4-8 weeks + late third trimester	Mediastinal shift, cardiothoracic area ratio, RV and LV function Consider fetal MRI to assess lungs	Tetralogy with absent pulmonary valve, Ebstein anomaly, HLHS/RAS
At risk for fetal AV block (l-TGA, LAI, other)	Every 2-4 weeks	Assess fetal AV interval, fetal heart rate (atrial and ventricular), hydrops, extracardiac Doppler	First-degree AV block; second- and third-degree AV block
Post-FCI for fetal AS	Third trimester, every 4 weeks	Assess aortic outflow, arch flow direction, atrial septal flow velocity and direction, left ventricle size and function, mitral regurgitation, endocardial echo brightness	Severe AS with evolving HLHS
Post-FCI for HLHS/RAS	Third trimester, every 4 weeks	Assess patency of atrial communication, pulmonary vein Doppler forward/reverse VTI	HLHS/RAS

AS, Aortic stenosis; *AVSD*, atrioventricular septal defect; *CPAM*, congenital pulmonary airway malformation; *CHF*, congestive heart failure; *DORV*, double-outlet right ventricle; *FCI*, fetal cardiac intervention; *LAI*, left atrial isomerism; *MR*, mitral regurgitation; *RAS*, restrictive interatrial septum; *SCT*, sacrococcygeal teratoma; *SVT*, supraventricular tachycardia; *TAPVR*, total anomalous pulmonary venous return; *TTTS*, twin-twin transfusion syndrome; *TRAP*, twin reversed-arterial perfusion; *VTI*, velocity-time integral.
*See Section "Assessment of Fetal Cardiac Functional Abnormalities" for details.

V. SUMMARY AND CONCLUSIONS

Fetal echocardiography has evolved over the past four decades to become a highly sensitive and specific noninvasive tool for detection, classification, and risk assessment of fetal cardiovascular disease. Standards for imaging, reporting, and communication of test results have also evolved. The current document contains evidence- and consensus-based recommendations for performance of fetal echocardiography in both normal and abnormal fetal hearts, and for detailed evaluation of structural, functional, and rhythm-related abnormalities. Although this document is far from comprehensive, the writing group has presented here general guidelines for what constitutes a complete fetal echocardiographic examination in the presence of anomalies that can be used as a guide for both learners and experienced practitioners.

NOTICE AND DISCLAIMER

other parties in reliance on this information. Nor does your use of this information constitute the offering of medical advice by ASE or create any physician-patient relationship between ASE and your patients or anyone else.

ACKNOWLEDGMENT

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.echo.2023.04.014.

REFERENCES

1. Rychik J, Ayres N, Cuneo B, et al. American Society of Echocardiography guidelines and standards for performance of the fetal echocardiogram. J Am Soc Echocardiogr 2004;17:803-10.
2. Donofrio MT, Moon-Grady AJ, Hornberger LK, et al. Diagnosis and treatment of fetal cardiac disease: a scientific statement from the American Heart Association. Circulation 2014;129:2183-242.
3. International Society of Ultrasound in Obstetrics and Gynecology, Carvalho JS, Allan LD, Chaoui R, et al. ISUOG Practice Guidelines (updated): sonographic screening examination of the fetal heart. Ultrasound Obstet Gynecol 2013;41:348-59.
4. AIUM practice parameter for the performance of fetal echocardiography. J Ultrasound Med 2020;39:E5-16.
5. Pinto NM, Morris SA, Moon-Grady AJ, et al. Prenatal cardiac care: goals, priorities & gaps in knowledge in fetal cardiovascular disease: perspectives of the Fetal Heart Society. Prog Pediatr Cardiol 2020;59:101312.
6. Wang S, Yang L, Wang L, et al. Selective serotonin reuptake inhibitors (SSRIs) and the risk of congenital heart defects: a meta-analysis of prospective cohort studies. J Am Heart Assoc 2015;4:e001681.
7. Melov SJ, Shetty PS, Pasupathy D, et al. Selective serotonin reuptake inhibitor or serotonin-norepinephrine reuptake inhibitors and epidemiological characteristics associated with prenatal diagnosis of congenital heart disease. Prenat Diagn 2021;41:35-42.
8. Schaefer C, Hannemann D, Meister R, et al. Vitamin K antagonists and pregnancy outcome. A multi-centre prospective study. Thromb Haemost 2006;95:949-57.
9. Society for Maternal-Fetal Medicine (SMFM), Norton ME, Chauhan SP, Dashe JS. Society for maternal-fetal medicine (SMFM) clinical guideline #7: nonimmune hydrops fetalis. Am J Obstet Gynecol 2015;212:127-39.
10. Barris DM, Brailovschi Y, Shah A, et al. The role of fetal echocardiogram after detection of extracardiac anomalies in utero (fetal echocardiogram for extracardiac malformations). Prenat Diagn 2021;41:1134-9.
11. Ghimire LV, Chou FS, Moon-Grady AJ. Risk of congenital heart disease is increased among newborns with non-cardiac congenital anomalies: survey-based analysis using 2016 Kids' Inpatient Database. Ultrasound Obstet Gynecol 2021;57:841-2.
12. Tai C, Conner T, Pevyandi S, et al. Prevalence of congenital heart disease in an isolated single umbilical artery is low at a tertiary referral center. J Ultrasound Med 2021;40:1729-30.
13. Hoffman JIE, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol 2002;39:1890-900.
14. Bak GS, Shaffer BL, Madriago E, et al. Detection of fetal cardiac anomalies: cost-effectiveness of increased number of cardiac views. Ultrasound Obstet Gynecol 2020;55:758-67.
15. AIUM practice parameter for the performance of detailed second- and third-trimester diagnostic obstetric ultrasound examinations. J Ultrasound Med 2019;38:3093-100.
16. Li Y, Hua Y, Fang J, et al. Performance of different scan protocols of fetal echocardiography in the diagnosis of fetal congenital heart disease: a systematic review and meta-analysis. PLoS One 2013;8:e65484.
17. Barker PCA, Lewin MB, Donofrio MT, et al. Specific considerations for pediatric, fetal, and congenital heart disease patients and echocardiography service providers during the 2019 Novel Coronavirus outbreak: Council on Pediatric and Congenital Heart Disease. J Am Soc Echocardiogr 2020;33:658-65.
18. Odibo AO, Coassolo KM, Stamilio DM, et al. Should all pregnant diabetic women undergo a fetal echocardiography? A cost-effectiveness analysis comparing four screening strategies. Prenat Diagn 2006 Jan;26:39-44.
19. Bernard LS, Ramos GA, Fines V, et al. Reducing the cost of detection of congenital heart disease in fetuses of women with pregestational diabetes mellitus. Ultrasound Obstet Gynecol 2009;33:676-82.
20. Gekas J, Gagné G, Bujold E, et al. Comparison of different strategies in prenatal screening for Down's syndrome: cost effectiveness analysis of computer simulation. BMJ 2009;338:b134.
21. Cunningham GC, Tompkinison DG. Cost and effectiveness of the California triple marker prenatal screening program. Genet Med 1999;1:199-206.
22. Pinto NM, Nelson R, Puchalski M, et al. Cost-effectiveness of prenatal screening strategies for congenital heart disease. Ultrasound Obstet Gynecol 2014;44:50-7.
23. Chung EH, Lim SL, Havrilesky LJ, et al. Cost-effectiveness of prenatal screening methods for congenital heart defects in pregnancies conceived by in-vitro fertilization. Ultrasound Obstet Gynecol 2021;57:979-86.
24. National Collaborating Centre for Women's and Children's Health (UK). Antenatal Care: Routine care for the healthy pregnant woman. London: RCOG Press; 2008.
25. Simpson LL. Indications for fetal echocardiography from a tertiary-care obstetric sonography practice. J Clin Ultrasound 2004;32:123-8.
26. Huggon IC, Ghi T, Cook AC, et al. Fetal cardiac abnormalities identified prior to 14 weeks' gestation. Ultrasound Obstet Gynecol 2002;20:22-9.
27. Zidere V, Bellsham-Revell H, Persico N, et al. Comparison of echocardiographic findings in fetuses at less than 15 weeks' gestation with later cardiac evaluation. Ultrasound Obstet Gynecol 2013;42:679-86.
28. AIUM practice parameter for documentation of an ultrasound examination. J Ultrasound Med 2020;39:E1-4.
29. Abramowicz JS, Basseal JM, Brezinka C, et al. ISUOG Safety Committee Position Statement on use of personal protective equipment and hazard mitigation in relation to SARS-CoV-2 for practitioners undertaking obstetric and gynecological ultrasound. Ultrasound Obstet Gynecol 2020;55:886-91.
30. Torloni MR, Vedmedovska N, Merialdi M, et al. Safety of ultrasonography in pregnancy: WHO systematic review of the literature and meta-analysis. Ultrasound Obstet Gynecol 2009 May;33:599-608.
31. American Institute of Ultrasound in Medicine. Prudent use and safety of diagnostic ultrasound in pregnancy; 2020. Available at: https://www.aium.org/officialStatements/79. Accessed November 19, 2022.
32. Nemescu D, Berescu A, Onofriescu M, et al. Safety indices during fetal echocardiography at the time of first-trimester scan are machine dependent. PLoS One 2015;10:e0127570.
33. Semmler J, Day TG, Georgiopoulos G, et al. Fetal speckle-tracking: impact of angle of insonation and frame rate on global longitudinal strain. J Am Soc Echocardiogr 2020;33:1141-6.e2.
34. Yagel S, Cohen SM, Achiron R. Examination of the fetal heart by five short-axis views: a proposed screening method for comprehensive cardiac evaluation. Ultrasound Obstet Gynecol 2001;17:367-9.

35. DeVore GR. Computing the Z Score and centiles for cross-sectional analysis: a practical approach. J Ultrasound Med 2017;36:459-73.

36. Lee W, Riggs T, Amula V, et al. Fetal echocardiography: z-score reference ranges for a large patient population. Ultrasound Obstet Gynecol 2010; 35:28-34.

37. Schneider C, McCrindle BW, Carvalho JS, et al. Development of Z-scores for fetal cardiac dimensions from echocardiography. Ultrasound Obstet Gynecol 2005;26:599-605.

38. McElhinney DB, Marshall AC, Wilkins-Haug LE, et al. Predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for aortic stenosis with evolving hypoplastic left heart syndrome. Circulation 2009;120:1482-90.

39. Krishnan A, Pike JI, McCarter R, et al. Predictive models for normal fetal cardiac structures. J Am Soc Echocardiogr 2016;29:1197-206.

40. Gu X, Zhu H, Zhang Y, et al. Quantile score: a new reference system for quantitative fetal echocardiography based on a large multicenter study. J Am Soc Echocardiogr 2019;32:296-302.e5.

41. Vigneswaran TV, Akolekar R, Syngelaki A, et al. Reference ranges for the size of the fetal cardiac outflow tracts from 13 to 36 weeks gestation: a single-center study of over 7000 cases. Circ Cardiovasc Imaging 2018; 11:e007575.

42. Chiba Y, Kanzaki T, Kobayashi H, et al. Evaluation of fetal structural heart disease using color flow mapping. Ultrasound Med Biol 1990;16:221-9.

43. Abuhamad A, Chaoui R. A Practical Guide to Fetal Echocardiography: Normal and Abnormal Hearts. Fourth Edition. Philadelphia: Lippincott-Williams & Wilkins; 2016.

44. Heling KS, Chaoui R, Bollmann R. Advanced dynamic flow – a new method of vascular imaging in prenatal medicine. A pilot study of its applicability. Ultraschall Med 2004;25:280-4.

45. Huisman TW, Stewart PA, Wladimiroff JW, et al. Flow velocity waveforms in the ductus venosus, umbilical vein and inferior vena cava in normal human fetuses at 12-15 weeks of gestation. Ultrasound Med Biol 1993;19:441-5.

46. Carceller-Blanchard AM, Fouron JC. Determinants of the Doppler flow velocity profile through the mitral valve of the human fetus. Br Heart J 1993;70:457-60.

47. Paladini D, Palmieri S, Celentano E, et al. Pulmonary venous blood flow in the human fetus. Ultrasound Obstet Gynecol 1997;10:27-31.

48. Mielke G, Benda N. Blood flow velocity waveforms of the fetal pulmonary artery and the ductus arteriosus: reference ranges from 13 weeks to term. Ultrasound Obstet Gynecol 2000;15:213-8.

49. Fouron JC. The unrecognized physiological and clinical significance of the fetal aortic isthmus. Ultrasound Obstet Gynecol 2003;22:441-7.

50. Bhide A, Acharya G, Baschat A, et al. ISUOG Practice Guidelines (updated): use of Doppler velocimetry in obstetrics. Ultrasound Obstet Gynecol 2021;58:331-9.

51. Yamamoto Y, Hornberger LK. Progression of outflow tract obstruction in the fetus. Early Hum Dev 2012;88:279-85.

52. Cordes TM, O'Leary PW, Seward JB, et al. Distinguishing right from left: a standardized technique for fetal echocardiography. J Am Soc Echocardiogr 1994;7:47-53.

53. Gonçalves LF, Lee W, Chaiworapongsa T, et al. Four-dimensional ultrasonography of the fetal heart with spatiotemporal image correlation. Am J Obstet Gynecol 2003;189:1792-802.

54. Wang Y, Fan M, Siddiqui FA, et al. Strategies for accurate diagnosis of fetal aortic arch anomalies: benefits of three-dimensional sonography with spatiotemporal image correlation and a novel algorithm for volume analysis. J Am Soc Echocardiogr 2018;31:1238-51.

55. Patey O, Carvalho JS, Thilaganathan B. Intervendor discordance of fetal and neonatal myocardial tissue Doppler and speckle-tracking measurements. J Am Soc Echocardiogr 2019;32:1339-49.e23.

56. DeVore GR, Polanco B, Satou G, et al. Two-dimensional speckle tracking of the fetal heart: a practical step-by-step approach for the fetal sonologist. J Ultrasound Med 2016;35:1765-81.

57. Clur SAB, Bilardo CM. Early detection of fetal cardiac abnormalities: how effective is it and how should we manage these patients? Prenat Diagn 2014;34:1235-45.

58. McBrien A, Hornberger LK. Early fetal echocardiography. Birth Defects Res 2019;111:370-9.

59. Wiechec M, Knafel A, Nocun A. Prenatal detection of congenital heart defects at the 11- to 13-week scan using a simple color Doppler protocol including the 4-chamber and 3-vessel and trachea views. J Ultrasound Med 2015;34:585-94.

60. Quarello E, Lafouge A, Fries N, et al. Basic heart examination: feasibility study of first-trimester systematic simplified fetal echocardiography. Ultrasound Obstet Gynecol 2017;49:224-30.

61. American Institute of Ultrasound in Medicine. Medical Ultrasound Safety. Laurel, Maryland; 2020. Available at: http://aium.s3.amazonaws.com/resourceLibrary/mus4.pdf. Accessed November 17, 2022.

62. Salvesen K, Lees C, Abramowicz J, et al. ISUOG statement on the safe use of Doppler in the 11 to 13 +6-week fetal ultrasound examination. Ultrasound Obstet Gynecol 2011;37:628.

63. Hutchinson D, McBrien A, Howley L, et al. First-trimester fetal echocardiography: identification of cardiac structures for screening from 6 to 13 Weeks' gestational age. J Am Soc Echocardiogr 2017;30:763-72.

64. Carvalho JS, Moscoso G, Tekay A, et al. Clinical impact of first and early second trimester fetal echocardiography on high risk pregnancies. Heart 2004;90:921-6.

65. Zidere V, Vigneswaran TV, Syngelaki A, et al. Reference ranges for pulsed-wave Doppler of the fetal cardiac inflow and outflow tracts from 13 to 36 weeks' gestation. J Am Soc Echocardiogr 2021;34:1007-16.e10.

66. Mitchell JL, Cuneo BF, Etheridge SP, et al. Fetal heart rate predictors of long QT syndrome. Circulation 2012;126:2688-95.

67. Friedman DM, Kim MY, Copel JA, et al. Utility of cardiac monitoring in fetuses at risk for congenital heart block: the PR Interval and Dexamethasone Evaluation (PRIDE) prospective study. Circulation 2008;117:485-93.

68. Clur SAB, Vink AS, Etheridge SP, et al. Left ventricular isovolumetric relaxation time is prolonged in fetal Long-QT Syndrome. Circ Arrhythm Electrophysiol 2018;11:e005797.

69. Sonesson SE, Eliasson H, Conner P, et al. Doppler echocardiographic isovolumetric time intervals in diagnosis of fetal blocked atrial bigeminy and 2:1 atrioventricular block. Ultrasound Obstet Gynecol 2014;44: 171-5.

70. Cuneo BF, Strasburger JF, Yu S, et al. In utero diagnosis of long QT syndrome by magnetocardiography. Circulation 2013;128:2183-91.

71. Jaeggi ET, Carvalho JS, De Groot E, et al. Comparison of transplacental treatment of fetal supraventricular tachyarrhythmias with digoxin, flecainide, and sotalol: results of a nonrandomized multicenter study. Circulation 2011;124:1747-54.

72. Krapp M, Kohl T, Simpson JM, et al. Review of diagnosis, treatment, and outcome of fetal atrial flutter compared with supraventricular tachycardia. Heart 2003;89:913-7.

73. Byrne FA, Lee H, Kipps AK, et al. Echocardiographic risk stratification of fetuses with sacrococcygeal teratoma and twin-reversed arterial perfusion. Fetal Diagn Ther 2011;30:280-8.

74. Statile CJ, Cnota JF, Gomien S, et al. Estimated cardiac output and cardiovascular profile score in fetuses with high cardiac output lesions. Ultrasound Obstet Gynecol 2013;41:54-8.

75. Derderian SC, Jeanty C, Fleck SR, et al. The many faces of hydrops. J Pediatr Surg 2015;50:50-4; discussion 54.

76. Flood K, Unterscheider J, Daly S, et al. The role of brain sparing in the prediction of adverse outcomes in intrauterine growth restriction: results of the multicenter PORTO Study. Am J Obstet Gynecol 2014;211: 288.e1-5.

77. Duke C, Sharland GK, Jones AM, et al. Echocardiographic features and outcome of truncus arteriosus diagnosed during fetal life. Am J Cardiol 2001;88:1379-84.

78. Votava-Smith JK, Habli M, Cnota JF, et al. Diastolic dysfunction and cerebrovascular redistribution precede overt recipient twin cardiomyopathy in early-stage twin-twin transfusion syndrome. J Am Soc Echocardiogr 2015;28:533-40.

79. Moon-Grady AJ, Rand L, Gallardo S, et al. Diastolic cardiac pathology and clinical twin-twin transfusion syndrome in monochorionic/diamniotic twins. Am J Obstet Gynecol 2011;205:279.e1-11.

80. Mahle WT, Rychik J, Tian ZY, et al. Echocardiographic evaluation of the fetus with congenital cystic adenomatoid malformation. Ultrasound Obstet Gynecol 2000;16:620-4.

81. Vogel M, McElhinney DB, Marcus E, et al. Significance and outcome of left heart hypoplasia in fetal congenital diaphragmatic hernia. Ultrasound Obstet Gynecol 2010;35:310-7.

82. Byrne FA, Keller RL, Meadows J, et al. Severe left diaphragmatic hernia limits size of fetal left heart more than does right diaphragmatic hernia. Ultrasound Obstet Gynecol 2015;46:688-94.

83. Hofstaetter C, Hansmann M, Eik-Nes SH, et al. A cardiovascular profile score in the surveillance of fetal hydrops. J Matern Fetal Neonatal Med 2006;19:407-13.

84. Rasanen J, Wood DC, Weiner S, et al. Role of the pulmonary circulation in the distribution of human fetal cardiac output during the second half of pregnancy. Circulation 1996;94:1068-73.

85. Mielke G, Benda N. Cardiac output and central distribution of blood flow in the human fetus. Circulation 2001;103:1662-8.

86. Harada K, Tsuda A, Orino T, et al. Tissue Doppler imaging in the normal fetus. Int J Cardiol 1999;71:227-34.

87. Chan LY, Fok WY, Wong JT, et al. Reference charts of gestation-specific tissue Doppler imaging indices of systolic and diastolic functions in the normal fetal heart. Am Heart J 2005;150:750-5.

88. Mäkikallio K, Räsänen J, Mäkikallio T, et al. Human fetal cardiovascular profile score and neonatal outcome in intrauterine growth restriction. Ultrasound Obstet Gynecol 2008;31:48-54.

89. Huhta JC. Fetal congestive heart failure. Semin Fetal Neonatal Med 2005;10:542-52.

90. Shah AD, Border WL, Crombleholme TM, et al. Initial fetal cardiovascular profile score predicts recipient twin outcome in twin-twin transfusion syndrome. J Am Soc Echocardiogr 2008;21:1105-8.

91. Peyvandi S, Feldstein VA, Hirose S, et al. Twin-reversed arterial perfusion sequence associated with decreased fetal cerebral vascular impedance. Ultrasound Obstet Gynecol 2015;45:447-51.

92. Mari G, Adrignolo A, Abuhamad AZ, et al. Diagnosis of fetal anemia with Doppler ultrasound in the pregnancy complicated by maternal blood group immunization. Ultrasound Obstet Gynecol 1995;5: 400-5.

93. Krishnan A, Jacobs MB, Morris SA, et al. Impact of socioeconomic status, race and ethnicity, and geography on prenatal detection of hypoplastic left heart syndrome and transposition of the great arteries. Circulation 2021;143:2049-60.

94. Hancock HS, Romano JC, Armstrong A, et al. Single ventricle and total anomalous pulmonary venous connection: implications of prenatal diagnosis. World J Pediatr Congenit Heart Surg 2018;9:434-9.

95. Szwast A, Tian Z, McCann M, et al. Vasoreactive response to maternal hyperoxygenation in the fetus with hypoplastic left heart syndrome. Circ Cardiovasc Imaging 2010;3:172-8.

96. Cohen MS, Jegatheeswaran A, Baffa JM, et al. Echocardiographic features defining right dominant unbalanced atrioventricular septal defect: a multi-institutional Congenital Heart Surgeons' Society study. Circ Cardiovasc Imaging 2013;6:508-13.

97. Carvajal HG, Canter MW, Abarbanell AM, et al. Does ascending aorta size affect Norwood outcomes in hypoplastic left heart with aortic atresia? Ann Thorac Surg 2020;110:1651-8.

98. Divanović A, Hor K, Cnota J, et al. Prediction and perinatal management of severely restrictive atrial septum in fetuses with critical left heart obstruction: clinical experience using pulmonary venous Doppler analysis. J Thorac Cardiovasc Surg 2011;141:988-94.

99. Shamszad P, Gospin TA, Hong BJ, et al. Impact of preoperative risk factors on outcomes after Norwood palliation for hypoplastic left heart syndrome. J Thorac Cardiovasc Surg 2014;147:897-901.

100. Berning RA, Silverman NH, Villegas M, et al. Reversed shunting across the ductus arteriosus or atrial septum in utero heralds severe congenital heart disease. J Am Coll Cardiol 1996;27:481-6.

101. Dyamenahalli U, Smallhorn JF, Geva T, et al. Isolated ductus arteriosus aneurysm in the fetus and infant: a multi-institutional experience. J Am Coll Cardiol 2000;36:262-9.

102. Paladini D, Palmieri S, Lamberti A, et al. Characterization and natural history of ventricular septal defects in the fetus. Ultrasound Obstet Gynecol 2000;16:118-22.

103. Axt-Fliedner R, Schwarze A, Smrcek J, et al. Isolated ventricular septal defects detected by color Doppler imaging: evolution during fetal and first year of postnatal life. Ultrasound Obstet Gynecol 2006;27:266-73.

104. Peixoto AB, Bravo-Valenzuela NJ, Martins WP, et al. Reference ranges for the fetal mitral, tricuspid, and interventricular septum annular plane systolic excursions (mitral annular plane systolic excursion, tricuspid annular plane systolic excursion, and septum annular plane systolic excursion) between 20 and 36 + 6 weeks of gestation. J Perinat Med 2020;48: 601-8.

105. Crispi F, Sepulveda-Swatson E, Cruz-Lemini M, et al. Feasibility and reproducibility of a standard protocol for 2D speckle tracking and tissue Doppler-based strain and strain rate analysis of the fetal heart. Fetal Diagn Ther 2012;32:96-108.

106. Friedberg MK, Ursell PC, Silverman NH. Isomerism of the left atrial appendage associated with ventricular noncompaction. Am J Cardiol 2005;96:985-90.

107. Huggon IC, Cook AC, Smeeton NC, et al. Atrioventricular septal defects diagnosed in fetal life: associated cardiac and extra-cardiac abnormalities and outcome. J Am Coll Cardiol 2000;36:593-601.

108. Szwast AL, Marino BS, Rychik J, et al. Usefulness of left ventricular inflow index to predict successful biventricular repair in right-dominant unbalanced atrioventricular canal. Am J Cardiol 2011;107:103-9.

109. Losada E, Moon-Grady AJ, Strohsnitter WC, et al. Anomalous mitral arcade in twin-twin transfusion syndrome. Circulation 2010;122: 1456-63.

110. Layman TE, Edwards JE. Anomalous mitral arcade. A type of congenital mitral insufficiency. Circulation 1967;35:389-95.

111. Pluchinotta FR, Teot LA, Sanders SP. Left ventricular aneurysm, aortic stenosis, and arcade mitral valve: coexistence of three rare congenital heart defects. Pediatr Cardiol 2012;33:1206-9.

112. Bevilacqua M, Sanders SP, Van Praagh S, et al. Double-inlet single left ventricle: echocardiographic anatomy with emphasis on the morphology of the atrioventricular valves and ventricular septal defect. J Am Coll Cardiol 1991;18:559-68.

113. Yang YL, Wang XF, Cheng TO, et al. Echocardiographic characteristics of the criss-cross heart. Int J Cardiol 2010;140:133-7.

114. Apitz C, Webb GD, Redington AN. Tetralogy of Fallot. Lancet 2009;374: 1462-71.

115. Rodenbarger A, Thorsson T, Stiver C, et al. Third trimester predictors of interventional timing and accuracy of fetal anticipatory guidance in tetralogy of Fallot: a multi-center study. Prenat Diagn 2020;40:870-7.

116. Arya B, Levasseur SM, Woldu K, et al. Fetal echocardiographic measurements and the need for neonatal surgical intervention in Tetralogy of Fallot. Pediatr Cardiol 2014;35:810-6.

117. Arunamata A, Balasubramanian S, Punn R, et al. Impact of fetal somatic growth on pulmonary valve annulus Z-Scores during gestation and through birth in patients with Tetralogy of Fallot. Pediatr Cardiol 2018;39:1181-7.

118. Kwon EN, Parness IA, Srivastava S, et al. Subpulmonary stenosis assessed in midtrimester fetuses with tetralogy of Fallot: a novel method for predicting postnatal clinical outcome. Pediatr Cardiol 2013;34: 1314-20.

119. Friedman K, Balasubramanian S, Tworetzky W. Midgestation fetal pulmonary annulus size is predictive of outcome in tetralogy of Fallot. Congenit Heart Dis 2014;9:187-93.

120. Goldmuntz E. 22q11.2 deletion syndrome and congenital heart disease. Am J Med Genet C Semin Med Genet 2020;184:64-72.

121. Chelliah A, Moon-Grady AJ, Peyvandi S, et al. Contemporary outcomes in tetralogy of Fallot with absent pulmonary valve after fetal diagnosis. J Am Heart Assoc 2021;10:e019713.

122. Russell HM, Jacobs ML, Anderson RH, et al. A simplified categorization for common arterial trunk. J Thorac Cardiovasc Surg 2011;141:645-53.

123. Krummholz A, Gottschalk I, Geipel A, et al. Prenatal diagnosis, associated findings and postnatal outcome in fetuses with congenitally corrected transposition of the great arteries. Arch Gynecol Obstet 2021;303:1469-81.

124. Vorisek CN, Enzensberger C, Willomeit S, et al. Prenatal diagnosis and outcome of congenital corrected transposition of the great arteries - a multicenter report of 69 cases. Ultraschall Med 2021;42:291-6.

125. Hornberger LK, Sanders SP, Rein AJ, et al. Left heart obstructive lesions and left ventricular growth in the midtrimester fetus. A longitudinal study. Circulation 1995;92:1531-8.

126. Gardiner HM, Kovacevic A, Tulzer G, et al. Natural history of 107 cases of fetal aortic stenosis from a European multicenter retrospective study. Ultrasound Obstet Gynecol 2016;48:373-81.

127. Vogel M, McElhinney DB, Wilkins-Haug LE, et al. Aortic stenosis and severe mitral regurgitation in the fetus resulting in giant left atrium and hydrops: pathophysiology, outcomes, and preliminary experience with pre-natal cardiac intervention. J Am Coll Cardiol 2011;57: 348-55.

128. Eckersley LG, Howley LW, van der Velde ME, et al. Quantitative assessment of left ventricular dysfunction in fetal Ebstein's anomaly and tricuspid valve dysplasia. J Am Soc Echocardiogr 2019;32:1598-607.

129. Nathan AT, Marino BS, Dominguez T, et al. Tricuspid valve dysplasia with severe tricuspid regurgitation: fetal pulmonary artery size predicts lung viability in the presence of small lung volumes. Fetal Diagn Ther 2010;27:101-5.

130. Selamet Tierney ES, McElhinney DB, Freud LR, et al. Assessment of progressive pathophysiology after early prenatal diagnosis of the Ebstein anomaly or tricuspid valve dysplasia. Am J Cardiol 2017; 119:106-11.

131. Hornberger LK, Sahn DJ, Kleinman CS, et al. Tricuspid valve disease with significant tricuspid insufficiency in the fetus: diagnosis and outcome. J Am Coll Cardiol 1991;17:167-73.

132. Taketazu M, Barrea C, Smallhorn JF, et al. Intrauterine pulmonary venous flow and restrictive foramen ovale in fetal hypoplastic left heart syndrome. J Am Coll Cardiol 2004;43:1902-7.

133. Maeno YV, Boutin C, Hornberger LK, et al. Prenatal diagnosis of right ventricular outflow tract obstruction with intact ventricular septum, and detection of ventriculocoronary connections. Heart 1999;81:661-8.

134. Sun HY, Boe J, Rubesova E, et al. Fetal MRI correlates with postnatal CT angiogram assessment of pulmonary anatomy in tetralogy of Fallot with absent pulmonary valve. Congenit Heart Dis 2014;9:E105-9.

135. Jain S, Kleiner B, Moon-Grady A, et al. Prenatal diagnosis of vascular rings. J Ultrasound Med 2010;29:287-94.

136. American Academy of Pediatrics Committee on Fetus And Newborn. Levels of neonatal care. Pediatrics 2012;130:587-97.

137. Donofrio MT, Skurow-Todd K, Berger JT, et al. Risk-stratified postnatal care of newborns with congenital heart disease determined by fetal echocardiography. J Am Soc Echocardiogr 2015;28:1339-49.

138. Sanapo L, Pruetz JD, Słodki M, et al. Fetal echocardiography for planning perinatal and delivery room care of neonates with congenital heart disease. Echocardiography 2017;34:1804-21.

139. Patey O, Carvalho JS, Thilaganathan B. Urgent neonatal balloon atrial septostomy in simple transposition of the great arteries: predictive value of fetal cardiac parameters. Ultrasound Obstet Gynecol 2021;57:756-68.

140. Punn R, Silverman NH. Fetal predictors of urgent balloon atrial septostomy in neonates with complete transposition. J Am Soc Echocardiogr 2011;24:425-30.

141. Chelliah A, Berger JT, Blask A, et al. Clinical utility of fetal magnetic resonance imaging in tetralogy of Fallot with absent pulmonary valve. Circulation 2013;127:757-9.

142. Barrera CA, Johnson AM, Rychik J, et al. Prognostic value of the nutmeg lung pattern/lymphangiectasia on fetal magnetic resonance imaging. Pediatr Radiol 2021;51:1809-17.

143. Mekjarasnapha M, Traisrisilp K, Luewan S, et al. Reference ranges for fetal septum primum excursion from 14 to 40 weeks' gestation. J Ultrasound Med 2013;32:1729-34.