THE WHO, WHAT, WHEN, WHY, AND HOW'S from ASE's New Fetal Echocardiography Guideline

This is a companion article for the recently released updated "Guidelines and Recommendations for Performance of the Fetal Echocardiogram: An Update from the American Society of Echocardiography." It provides a summary of key clinically relevant aspects of the guidelines and updates from the previous guidelines. However, practitioners should note that the tables for these guidelines are particularly useful in summarizing important points for use in practice and are referenced throughout this article.

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WHY an update?

It has been almost two decades since ASE released its last guidelines on fetal echocardiography in 2004. Since then, advances in the field and increased collaboration have improved imaging and increased our understanding of fetal cardiac physiology and disease progression. The time had come for pediatric cardiologists who focus on fetal imaging to provide an updated statement on current best practices on fetal cardiac imaging and care to complement recent guidelines published by the AHA, AIUM, and ISUOG.

WHO needs a fetal echocardiogram?

One of the biggest questions that is debated in the literature is who needs a fetal echocardiogram. If fetal echocardiography was not a limited and costly endeavor, we would easily say fetal echocardiograms all around. Instead, pregnancies that are at low risk for fetal cardiac disease are screened during an obstetric anatomy scan. There is increasing debate about where to draw the line regarding who warrants referral for a fetal echocardiogram because of a condition that increases the baseline risk for cardiac disease and much literature published in this area since the last guidelines. These debates center around the need for additional testing when a screening anatomy scan is normal. However, there is huge variability in how well screening performs in different settings and prenatal detection of cardiac disease remains much lower than desired. Thus, these guidelines continue to recommend that certain fetal and maternal conditions should reach that convey an estimated risk of >3% of cardiac disease in their fetus based on available evidence still warrant fetal echocardiograms even in the setting of a normal screen (Table 1 in

Guidelines and Recommendations for Performance of the Fetal Echocardiogram: An Update from the American Society of Echocardiography) but goes into further detail by specific condition and compares these recommendations with AIUM and AHA guidelines. Again, these indications and referrals for fetal echocardiography must be considered within the local context and the sensitivity and specificity of the screening anatomy scan in the clinical practice of the provider as well as issues of access. Finally, the primary reason for referral which remains the strongest indication is of course an abnormal cardiac screening.

WHEN should you perform a fetal echocardiogram?

The best window for a transabdominal fetal echo is 18-22 weeks gestation which is similar to the timing of the anatomy scan. While we are increasingly pushing the envelope of how early we can image the heart when there is a concern or higher risk of fetal cardiac disease, first-trimester exams have a lower sensitivity and (78.6% and 98.9%) should be repeated in the second trimester. Serial fetal echos are necessary for abnormalities of the heart up until 34-36 weeks gestation; how many and how often depends on the condition and discretion of the care team.

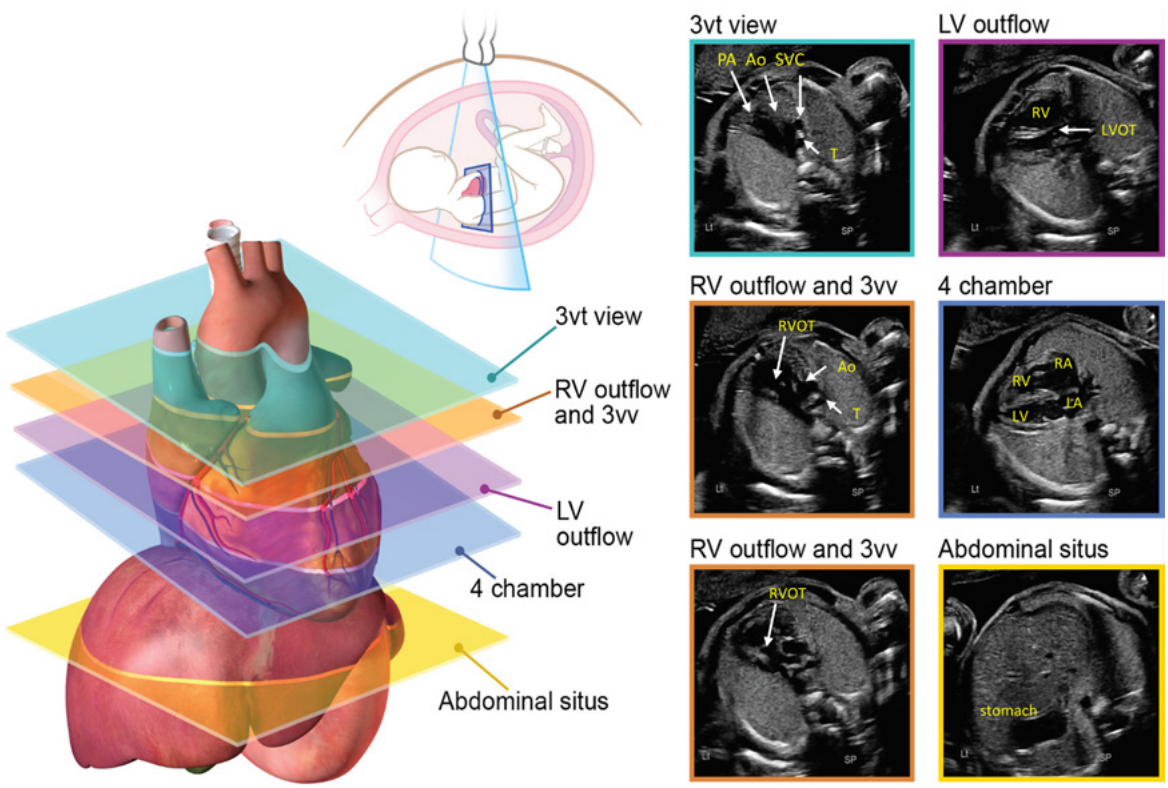

FIGURE 1 (from Figure 2 in Guidelines and Recommendations for Performance of the Fetal Echocardiogram: An Update from the American Society of Echocardiography). Axial planes suggested for screening the fetal heart at the time of the obstetric anatomic survey and as an initial series obtained during a fetal echocardiogram. Note that the images depict a fetus in cephalic presentation; breech presentation will result in mirror-image reversal from that shown here. 3VT, three vessels and trachea view; 3VV, three vessel view; Ao, aorta; DA, ductus arteriosus; LV, left ventricle; LVOT, left ventricular outflow tract; PA, pulmonary artery; RV, right ventricle; RVOT, right ventricular outflow tract; SVC, superior vena cava; T, trachea. Redrawn and adapted from Yagel 2001[113].

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WHAT equipment, settings, and images?

While there is no evidence of harmful effects of ultrasound on a fetus, the principle of As Low As Reasonably Achievable minimizes power output and duration of exam to decrease exposure. The size and frequency of motion of a beating fetal heart require a high spatial and temporal resolution in ultrasound imaging platforms. Frame rates should be above 40 Hz and higher for visualizing finer structures and performing advanced techniques such as strain. In addition to typical image optimization techniques in pediatric echocardiograms, fetal requires consideration of maternal and fetal position with flexible and comprehensive scanning to obtain the best imaging windows. Consideration of depth of field is particularly important when scanning through the maternal abdomen and the fetal cardiac image should be magnified to occupy at least 1/3 of the imaging screen. Multiple scanning planes and sweeps are required to visualize and interrogate all the structures that are recommended for inclusion in a fetal echocardiogram (**Figures 1 and 2**). These views include abdominal situs, three-vessel, and three-vessel trachea sweeps in addition to views discussed in the previous guidelines. The structures and use of modalities such as M-mode, color Doppler and color are detailed in the guidelines and do not significantly differ from the previous document except for the addition of newer tools such as power Doppler to visualize smaller vascular structures and those with low velocity flow.

The many HOWS...

HOW to assess rate and rhythm?

Rhythm/rate abnormalities are a frequent referral indication for fetal echocardiogram but of course, should be assessed in all fetal echocardiograms. References for normal values of heart rate by gestational age are published and deviations from these or typical rhythm warrant further investigation. While rate is relatively straightforward to assess, determining rhythm and specific atrioventricular conduction relies on assessing the relationship between mechanical atrio and ventricular contraction and may include assessments of timing. This relationship can be analyzed using several complementary echocardiogram modalities including M-mode of the atria and ventricles and simultaneous Doppler interrogation of mitral/aortic or tricuspid/pulmonary inflow and outflow, aortic and SVC, and/or pulmonary artery and pulmonary vein. Tissue Doppler, color M-mode, and isovolumic time intervals can also help deduce challenging rhythms. Finally, rhythm abnormalities require careful assessment for hemodynamic compromise covered further below.

HOW to assess function?

While often taken for granted in a "normal" study, accurate assessment of cardiac function with a combination of qualitative and quantitative imaging methods is a critical part of the fetal echocardiogram. A more detailed assessment of function and signs of hemodynamic compromise is essential for conditions with primary cardiac dysfunction, tumors, rhythm disturbances, a subset of structural heart disease, and fetal conditions or extracardiac pathologies that can impact cardiac output or resistance. These evaluations can be critical to determining eligibility and timing of interventions and or delivery. In general assessment, the function should include 2D imaging of systolic and diastolic function, Doppler and color Doppler of the atrioventricular valves, umbilical vessels, and the ductus venosus, and quantification of the cardiac size. Further quantitative assessments of function such as

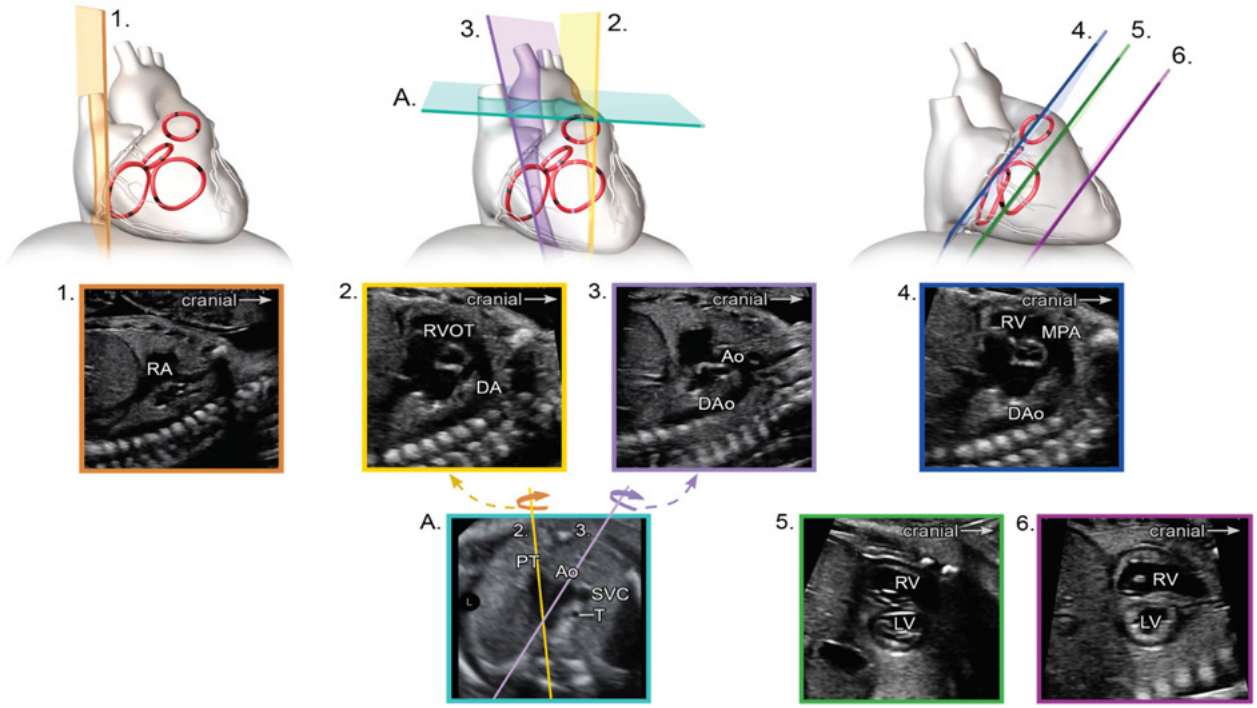

FIGURE 2 (from Figure 3 in Guidelines and Recommendations for Performance of the Fetal Echocardiogram: An Update from the American Society of Echocardiography). Sagittal and parasagittal planes for fetal echocardiogram evaluation. Ao, aorta; DA, ductus arteriosus; DAo, descending aorta; LA, left atrium; LV, left ventricle; MPA, main pulmonary artery; RA, right atrium; RV, right ventricle; RVOT, right ventricular outflow tract; SVC, superior vena cava.

the myocardial performance index and cardiac outputs are warranted when there is concern for the potential for or existing cardiac compromise. The cardiovascular profile score can be used in such cases. Tissue Doppler and strain are on the horizon for potential use as well.

HOW to assess structural abnormalities?

A significant portion of these guidelines are devoted to imaging structural heart defects in the fetus. The updated document expands significantly on key considerations and additional imaging to guide management and counseling for structural abnormalities. This is divided into major sections for single ventricle heart disease, complex atrioventricular connections, "look-alike" outflow tract anomalies, progressive lesions (such as stenosis or valve regurgitation), and isolated arch abnormalities. The common themes across the general approach to imaging among all these lesions is that detailed, comprehensive, and serial assessment

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of the fetal heart is important to appropriate counseling, prognostication, delivery, and intervention planning. This includes imaging at a minimum, determinants of ventricular adequacy (size and function), ductal dependency of either systemic or pulmonary blood, semilunar and atrioventricular valve competence, and ventriculoarterial relationships. However, many important aspects require further in-depth imaging depending on the type of anatomic defect observe. Key imaging pearls that allow such assessment for each anatomic subset are presented in the guidelines, while specific detailed imaging recommendations by type of defect are presented in a comprehensive table (Table 13 from Guidelines and Recommendations for Performance of the Fetal Echocardiogram: An Update from ASE, partially reproduced here) to guide the practitioner. As an example, the detailed evaluation of single ventricle fetal heart disease would include complete standard planes and Doppler examination plus assessment of:

- Morphology of dominant ventricle
- Ductal dependency of either systemic or pulmonary blood flow as determined by direction of ductal flow, outflow tract obstruction, and size of downstream vessels (pulmonary arteries and aortic arch and isthmus)
- Restriction of flow at the atrial septum, pulmonary venous Doppler VTI
- Atrioventricular valve regurgitation and ventricular function
- Systemic or pulmonary venous abnormalities that could impact outcome or repair

The WHY?

Diagnosis and ongoing assessment of fetuses with cardiac disease by fetal echocardiography allows for multidisciplinary comprehensive care, counseling, and support. What we learn from our fetal cardiac imaging, allows us to optimize the perinatal and delivery plan for the fetus and family. In certain cases, with evolving disease, fetal cardiac imaging is critical to determining the timing and need for delivery and/or prenatal or postnatal intervention. Existing risk stratification systems in the literature can facilitate coordination and communication of care plans. Evolving knowledge around additional techniques such as maternal hyperoxia, imaging modalities such as fetal MRI, and advanced echo techniques may provide additional prognostic information. This knowledge will further our efforts to provide accurate prognoses and timely care, which is of course the why which drives those of us who practice in this field.

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Category	Suspected when:					
Single ventricle 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 ventricular septal defect	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 ventriculo-arterial 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 [137]	For assessment of balanced AV valve see below ("Common atrioventricular valve") Consider right or left atrial isomerism/ heterotaxy
Tricuspid atresia	Absent AV connection, small RV, VSD	Bilateral SVC Benign A-wave reversal in DV	Mitral valve prolapse, dilation, or regurgitation	Define ventriculo-arterial connections, may be transposed Outflow tract from the morphologic RV 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 RV (leading to pulmonary stenosis, coarctation)
Hypoplastic left heart syndrome	Small LV on 4-chamber view with mitral and aortic stenosis or atresia	Bilateral SVC	TR with RV dysfunction may increase the risk of a 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 [138]	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 [139]
Double inlet left ventricle	Both AV valves open into the morphologic LV, 4-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 atrioventricular and ventriculo-arterial connections (may have D- or L-looped ventricles, DORV) Assess patency of out-flow tract connected to the morphologic RV 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

Excerpt from Table 13 in Guidelines and Recommendations for Performance of the Fetal Echocardiogram:
An Update from the American Society of Echocardiography: Guidance for disease-specific anatomic, physiologic, and functional evaluation for commonly encountered fetal congenital heart disease (CHD) lesions. This general information is not comprehensive and does not replace lesion-specific literature regarding fetal cardiac anatomy and physiology in congenital heart disease