Fetal Cardiac US: Techniques and Normal Anatomy Correlated with Adult CT and MR Imaging[1]

Neel Patel, MD
Evan Narasimhan, MD
Anne Kennedy, MB, BCh

Abbreviations: ASD = atrial septal defect, AV = atrioventricular, CHD = congenital heart disease, CPT = *Current Procedural Terminology*, IVC = inferior vena cava, LVOT = left ventricular outflow tract, PA = pulmonary artery, RVOT = right ventricular outflow tract, SVC = superior vena cava, 3VT = three-vessel trachea view, 3VV = three-vessel view, VSD = ventricular septal defect

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[1]From the Department of Radiology and Imaging Sciences, University of Utah Hospital and Clinics, 30 N 1099 E, Room 1A71, Salt Lake City, Utah 84132. Presented as an education exhibit at the 2015 RSNA Annual Meeting. Received May 2, 2016; revision requested August 5 and received September 1; accepted October 12. For this journal-based SA-CME activity, the authors, editor, and reviewers have disclosed no relevant relationships. **Address correspondence to** A.K. (e-mail: *Anne.kennedy@hsc.utah.edu*).

SA-CME LEARNING OBJECTIVES

After completing this journal-based SA-CME activity, participants will be able to:

- Discuss the importance of prenatal diagnosis of congenital heart disease.
- Perform a systematic approach to evaluation of the four-chamber view of the fetal heart.
- Describe the anatomy of the outflow tract views in comparison with CT and MR imaging.

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Congenital heart disease (CHD) is an important cause of childhood mortality. Despite the widespread use of ultrasonography (US) as a screening tool, the prenatal detection rate is suboptimal. Improvement of the initial screening examination, which is performed in low-risk populations and often interpreted by community radiologists, targets a point in the screening process that is likely to have the largest population effect. If the goal of community-based screenings is to detect cases that may be abnormal and refer those to specialized centers for complete assessment, it is logical to use a checklist to confirm normal anatomy. This article presents a stepwise process to evaluate fetal cardiac anatomy using comparison with computed tomography (CT) and magnetic resonance (MR) images, which are more familiar to radiologists in busy general practices. In addition, this article presents a checklist for assessment of the four-chamber view and demonstrates the expected normal appearance of the outflow tract views as well as the additional views required for complex obstetric US. These additional views include the aortic arch and bicaval views, three-vessel view (3VV), and three-vessel trachea view (3TV). CHD may be isolated, but it may indicate aneuploidy or a syndrome that, if present, determines the prognosis. In isolated CHD, the prognosis is determined by the exact nature of the abnormalities. In particular, duct-dependent disease if undiagnosed results in circulatory collapse in the infant once the ductus closes. If the heart does not look normal, the patient should be referred for detailed evaluation. Timely diagnosis of significant CHD allows for development of a personalized pregnancy management plan.

Introduction

Cardiac defects are the most common congenital abnormality; they occur in five to nine per 1000 births. Fifty percent of childhood deaths attributed to congenital malformations are due to congenital heart disease (CHD), and of liveborn infants with CHD 18% die within their first year (1).

Obstetric ultrasonography (US) is now considered a routine part of prenatal care; the optimal timing for a single scan is at 18–20 weeks gestation (2). The goal of the midtrimester scan is to confirm

TEACHING POINTS

- Detection of all cardiac anatomy is easier on cine clips than on static images. In particular, chamber wall contraction (ie, ventricular squeeze) pulmonary vein drainage, and AV valve assessment require cine clips.
- The LVOT is formed by the aortic root and trunk; it arises in the center of the heart, runs cephalad, forms a tight turn, and descends in the posterior mediastinum. The head and neck vessels arise from the apex of the curve.
- The RVOT is formed by the pulmonary conus and main PA. These wrap around the root of the aorta. As soon as the RVOT exits the pericardium, it branches; the ductus arteriosus then runs posteriorly toward the spine, and the right PA continues to wrap around the aorta.
- The 3VV is another way to look at the outflow tracts. It is obtained by sweeping toward the fetal head from the axial four-chamber view. Look for the size of the three vessels seen from right to left; normally the SVC is smaller than the aorta, which is smaller than the PA. The ductus arteriosus should be directed posteriorly toward the spine to unite with the descending aorta.
- The 3VT is a variation of the 3VV that includes views of the trachea and esophagus. It is obtained by sweeping superior and toward the left from the 3VV. It shows the confluence of the ductal and aortic arches, which come together in a V shape with the *V* open to the anterior chest wall and separated from the sternum by the thymus. The limbs of the *V* (the ductal and aortic arches) should be similar in size and show flow in the same direction. The ductal limb becomes slightly larger than the aortic limb in late pregnancy.

gestational age, evaluate anatomy, and clarify the relationship of the placenta to the cervix. Detection of anomalies varies with expertise and, in general, detection of CHD has not been good, with prenatal detection rates of only 30%–50% reported in developed countries (3,4). Medical society guidelines for performance of obstetric US now mandate evaluation of the four-chamber view of the heart and the cardiac outflow tracts in all second- and third-trimester obstetric scans. This is because the detection of CHD has improved from 55%–65% with just a four-chamber evaluation to 80%–84% if outflow tract assessment is included (5,6).

Prenatal detection of CHD should result in a pregnancy management plan. The options for pregnancy management include continuation of the pregnancy with intent to treat the infant, continuation of pregnancy with planned comfort care and no intervention at birth, or termination. In each case, it is crucial to plan the delivery location and method, including when and whether to induce labor, deliver by cesarean section, or use prostaglandin infusion at birth. In some cases, infants will need emergency intervention and thus every attempt is made to have a controlled delivery in a unit with the necessary facilities to treat critically ill newborns and access to pediatric interventional cardiology and cardiothoracic surgery services (7).

Prenatal diagnosis improves the outcome for fetuses with complex CHD. Tworetzky et al (8) reviewed a cohort of 88 patients with hypoplastic left heart syndrome; 33 patients were prenatally diagnosed, 55 were diagnosed postnatally. The prenatal diagnosis group had less acidosis, tricuspid regurgitation, and ventricular dysfunction and less need for preoperative inotrope. Furthermore, the postoperative mortality in the prenatal diagnosis group was 0% compared with 34% for the postnatal diagnosis group (8). Prenatal detection not only improves outcome, it decreases the cost of care. Jegatheeswaran et al (9) showed that infants with prenatal diagnosis of CHD were 16.5 times less likely to require emergency transport after birth. Prenatal transfer costs were $390 per fetus versus $5140 per infant transfer (9). Planned maternal transport to facilities with appropriate resources to manage children with complex CHD allows for a controlled delivery. The infant is immediately placed in the care of experts and is thus in the best physiologic condition for intervention (8,10).

What Are the Obstacles to Prenatal Detection?

Prenatal diagnosis of CHD is possible; it has been shown to improve outcomes for pregnancies that continue and it decreases costs, but the prenatal detection rate is still not as good as it could be. What are the obstacles that prevent effective community-based cardiac screenings? There are socioeconomic factors (eg, equipment types, personnel training and experience, and cost of follow-up studies), technical challenges, (eg, maternal habitus, late gestational age, multiple pregnancy), and a widely held perception that the assessment of the fetal heart is difficult. Pinto et al (1) identified multiple points in the screening process that were amenable to improvement. They identified improvement of the initial screening examination performed in low-risk populations as the one likely to have the largest population effect (1). As in any sonographic study, the technical parameters should be optimized. Views of the heart should be magnified so that the cardiac structures occupy most of the field of view. The use of several angles of insonation is important to avoid shadowing from adjacent structures, and the use of the dedicated fetal echo setting on the machine is helpful to improve contrast and resolution of small rapidly moving structures. Color Doppler flow imaging is helpful to confirm flow across a ventricular septal defect (VSD) and across the valves. Also, the use of two-dimensional cine clips has been shown to improve the ability to clear cardiac structures

Table 1: Checklist for Four-Chamber View

Situs
Heart position
Cardiac axis
Heart size
Is the "squeeze" OK?
Chamber identification and symmetry
Septum appearance
Atrioventricular (AV) valve offset
Foramen ovale flap
Area behind the heart
Rate and rhythm

as normal when compared with assessment with static images alone (11–13).

If the goal of community-based screenings is to detect cases that may be abnormal and refer them to specialized centers for complete assessment, use of a checklist to confirm normal anatomy makes sense. A systematic approach will allow determination of normal versus abnormal; those thought to be abnormal can be referred for more detailed evaluation. Table 1 shows an example of such a checklist for the four-chamber view.

Basic versus Complex Scan

In the United States, the images obtained in the basic fetal cardiac examination billed under the *Current Procedural Terminology* (CPT) code 76805 should include the four-chamber view, the left ventricular outflow tract (LVOT) view, and the right ventricular outflow tract (RVOT) view. These are also the recommended views in the American Institute of Ultrasound in Medicine and International Society of Ultrasound in Obstetrics and Gynecology guidelines (5,6).

Additional cardiothoracic views should be obtained when performing a complex obstetric US examination (CPT code 78611). These views include the aortic arch view, bicaval view, three-vessel view (3VV), three-vessel trachea view (3VT), and illustration of diaphragmatic integrity (14). These will be illustrated in detail later in the article.

Normal Anatomy: Basic Fetal Cardiac Scan

The standard four-chamber view of the fetal heart is an axial image through the chest, similar to a chest computed tomography (CT) scan or axial images in cardiac magnetic resonance (MR) imaging studies. The main difference is that the fetal lungs are not aerated; thus, the heart is in a true axial plane in the fetus but in a more oblique orientation postnatally. The outflow tract views

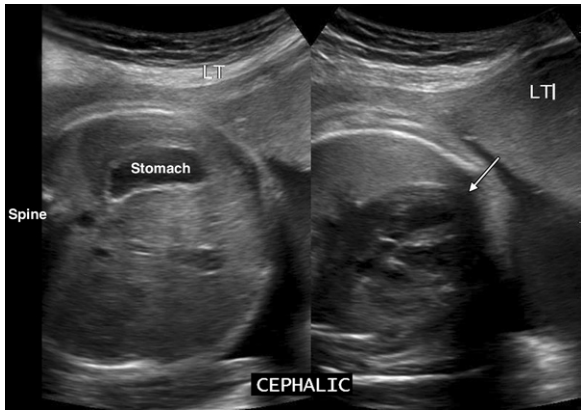


Figure 1. Axial US images of situs solitus in a third-trimester fetus. The images show the fetal abdomen (left) and chest (right) and were obtained using the split-screen feature on the US machine. The fetus is in cephalic presentation with the spine to the maternal right. Thus, the fetal left side is closest to the transducer, which is placed on the maternal abdominal wall. These images help confirm situs solitus with the heart and the cardiac apex (arrow) both on the fetal left side.

are obtained in nonaxial scan planes, but again the anatomy is the same as that seen countless times a day on chest imaging studies seen by community radiologists. Detection of all cardiac anatomy is easier on cine clips than on static images. In particular, chamber wall contraction (ie, ventricular squeeze), pulmonary vein drainage, and atrioventricular (AV) valve assessment require cine clips.

Situs

To determine situs, check the orientation of the fetal head and spine.

If the fetus is in cephalic presentation with its spine to the maternal left, then the fetal right side is "up" (ie, closest to the maternal abdominal wall) so the cardiac apex should point "down" and the stomach should be "down" or left as well. In our practice, we obtain a video clip of the four-chamber view extended into the upper abdomen, which shows the stomach and cardiac apex on the same side. A split-screen static image can be used for documentation as well (Fig 1).

Situs solitus is used to describe the normal arrangement with the cardiac apex and stomach on the left. Situs inversus implies right-left inversion with the cardiac apex and stomach on the right and the liver on the left. The term *situs ambiguous* is used to describe anything other than normal or complete situs inversus. Situs ambiguous is associated with heterotaxy syndromes that are in turn associated with complex CHD (15).

Heart Position

The normal heart is situated in the midline, apex directed left. A line that bisects the chest

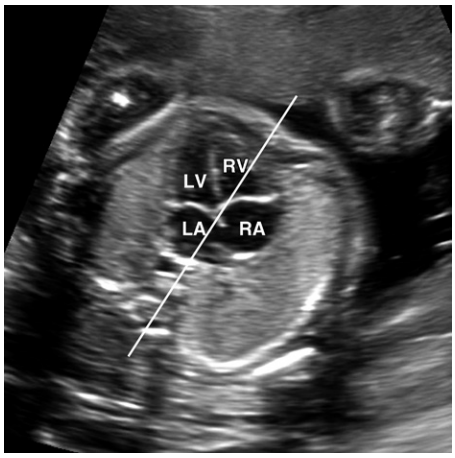


Figure 2. Axial US image of the heart of a fetus at 19 weeks gestation. A line drawn to bisect the chest from spine to sternum through the fetal heart at the level of the four-chamber view should pass through the left atrium *(LA)* and right ventricle *(RV)*. Large chest masses cause cardiac displacement. Subtle changes in position and axis may be the first indication of CHD. *LV* = left ventricle, *RA* = right atrium.

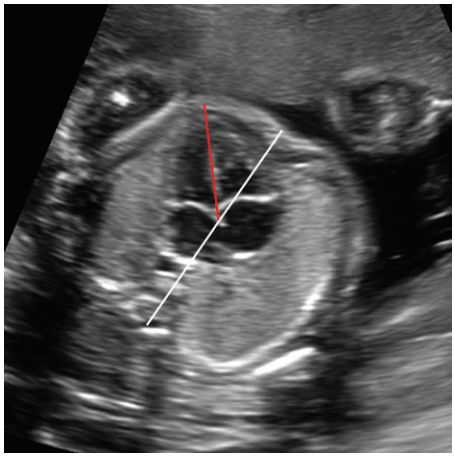


Figure 3. Axial US image of the cardiac axis in a fetus at 19 weeks gestation. Image through the fetal heart at the level of the four-chamber view shows a white line bisecting the fetal chest from spine to sternum. A red line is drawn along the plane of the interventricular septum. The cardiac axis is the angle between the red line and the white line. An abnormal cardiac axis is associated with conotruncal heart disease. True axial orientation is confirmed by the C shape of the rib. If multiple ribs are visible, the scan plane is oblique; nonaxial images may simulate disease states.

from spine to sternum should pass through the left atrium and right ventricle. Subtle cardiac displacement may be apparent only when checking this line (Fig 2).

Cardiac Axis

The axis is measured between the line that bisects the chest and a line along the axis of the ventricular septum. In the second and third trimesters, the axis normally ranges from 30° to 45° (Fig 3). In the first trimester, the range is higher from 34.5° to 56.8° (mean ± standard deviation = 47.6° ± 5.6) (16). An abnormal cardiac axis in the first trimester at the time of nuchal translucency measurement is an effective tool for detection of CHD. In a study by Sinkovskaya et al (17), it performed significantly better than other sonographic signs, including enlarged nuchal translucency, tricuspid regurgitation, or reversed A-wave in the ductus venosus (reversed flow during atrial contraction) used alone or in combination for detection of CHD (17).

Size

The heart should occupy approximately one-third of the chest. Heart area to chest area ratio should be ~33%, and heart circumference to chest circumference ratio should be ~50%. These measurements can be easily obtained with the same calipers as those used in measurements of the abdominal circumferences. The thoracic circumference measurement includes the skin (18).

Squeeze

The term *squeeze* refers to the ventricular contraction. This can be assessed only on cine clips. Right ventricle and left ventricle wall thickness and chamber contractility should be comparable. Left ventricular outflow obstruction (eg, critical aortic stenosis) may cause left ventricular dilatation and poor contractility, whereas right ventricular outflow obstruction (eg, pulmonary atresia) may cause right ventricular hypertrophy (19). The septal hypertrophy in diabetes mellitus (DM) cardiomyopathy may be severe enough to reduce the ventricles to slit-like chambers. (20). Nomograms are available for heart size, ventricular diameter, and wall thickness (21).

Chamber Identification and Symmetry

The left ventricle has a smooth interior contour and no septal valve attachment. The right ventricle has a trabeculated interior. The moderator band (also known as the septomarginal trabecula) is an important landmark for identification of the right ventricle as it traverses the cavity near the ventricular apex to connect the interventricular septum to the anterior papillary muscle (Fig 4). The septal leaflet of the tricuspid valve attaches to the ventricular septum (Fig 4). With normal embryologic looping, the right ventricle is the anterior ventricle (ie, closest to the chest wall).

At the time of the midtrimester scan, the ventricles should be symmetric in size; but in the normal fetal heart, physiologic enlargement of the right atrium (the only cardiac chamber that receives the entire cardiac output) and a simultaneous increase

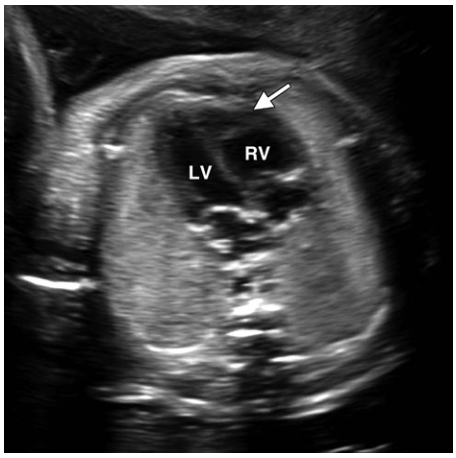


Figure 4. US image of ventricles in a fetus at 28 weeks gestation. The right ventricle *(RV)* is identified by the presence of the moderator band (arrow). It should be the anterior ventricle. The left ventricle *(LV)* is smooth-walled.

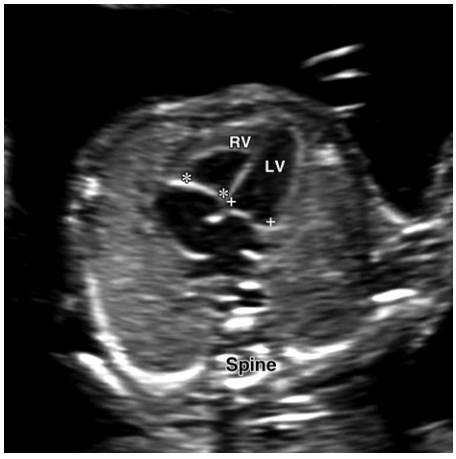
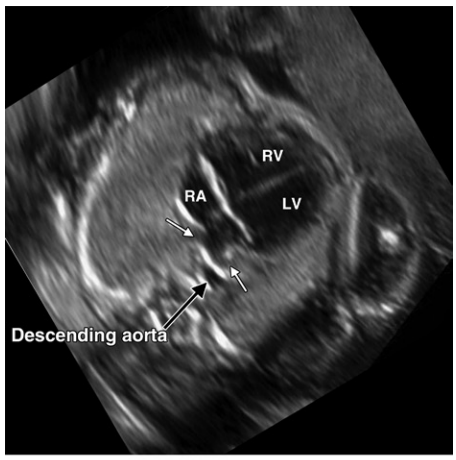
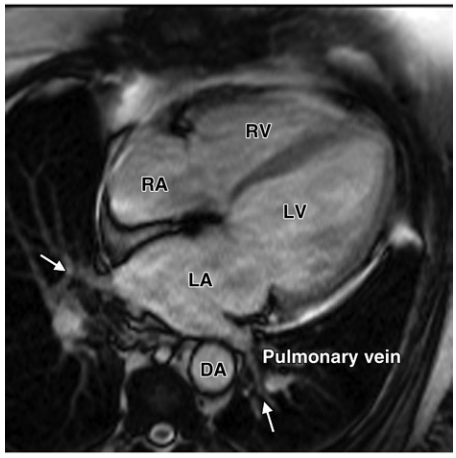


Figure 5. US image of the chamber symmetry of a fetus at 20 weeks gestation. The ventricles should be approximately equal in size and both should be apex-forming. Ventricular diameters can be measured as shown; ∗ shows level of the tricuspid valve and measures the right ventricle *(RV)*; + shows the level of the mitral valve and measures the left ventricle *(LV)*. The atria were symmetric in real time. This still image was obtained to measure the ventricular chambers.



a.



b.

Figure 6. Identification of the atria in a fetus at 21 weeks gestation. *LV* = left ventricle, *RV* = right ventricle. **(a)** Axial four-chamber US image shows the left atrium, which is the most posterior chamber of the heart. It is identified by the pulmonary veins (white arrows), which drain into it; these are visible on the four-chamber view. The descending aorta (black arrow) descends to the left of midline. The right atrium *(RA)* drains the SVC and IVC, which are seen on the bicaval view, not the four-chamber view. **(b)** Axial cardiac MR image shows the same anatomy with the pulmonary veins (arrows) entering the left atrium *(LA)* and the descending aorta *(DA)* touching the LA wall.

in the right ventricle can be observed later in pregnancy. The width of the right ventricle can be 1.3 times that of the left ventricle by term (22–24).

The width of the ventricles is measured at the level of the AV valves (Fig 5). The AV valves belong to the ventricle, so the tricuspid valve opens into the right ventricle, the mitral into the left ventricle. Both ventricles should be apex-forming.

The right atrium receives the systemic veins, superior vena cava (SVC), and inferior vena cava (IVC). The SVC and IVC are not visible on the four-chamber view, but the left atrium is identified by its drainage of the pulmonary veins, which are visible on the four-chamber view (Fig 6). The atria should be similar in size. With normal fetal circulation, the foramen ovale flap moves from the right atrium into the left atrium as the oxygenated blood from the ductus venosus (which enters the right atrium via the IVC) streams across the foramen ovale to reach the left heart (Fig 7).

Septum Appearance

The ventricular septum is about twice the length of the atrial septum (Fig 8). The ventricular septum thins from muscular to membranous; this normal

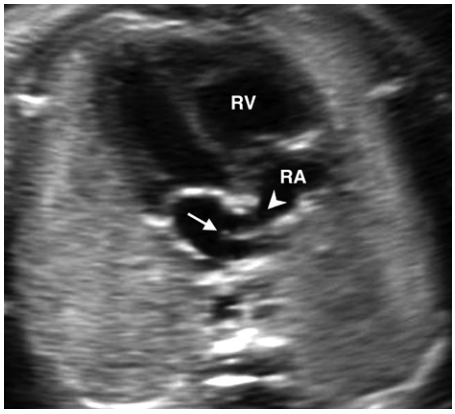


Figure 7. Axial US image of the foramen ovale in a fetus at 19 weeks gestation. Four-chamber view shows the foramen ovale as a well-demarcated defect in the atrial septum (arrowhead). In real time, the foramen ovale flap (arrow) can be seen to move with atrial contraction. The flap may be quite redundant and bulge into the lumen of the left atrium; this is described as a foramen ovale aneurysm. It is of no particular significance. *RA* = right atrium, *RV* = right ventricle.

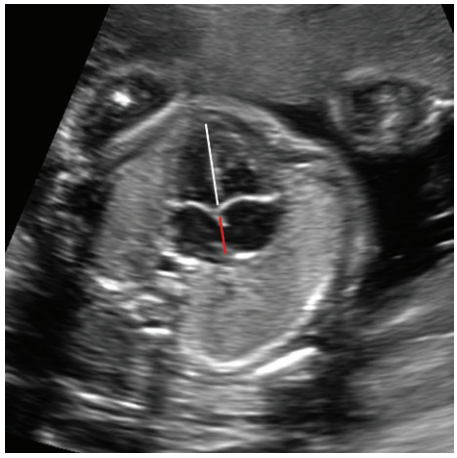


Figure 8. Axial US image (four-chamber view) shows relative sizes of the atrial and ventricular septa in a fetus at 19 weeks gestation. The ventricular septum (white line) is approximately twice the length of the atrial septum (red line). If these lines appear more equal in length, check carefully for a common atrioventricular valve in atrioventricular septal defect.

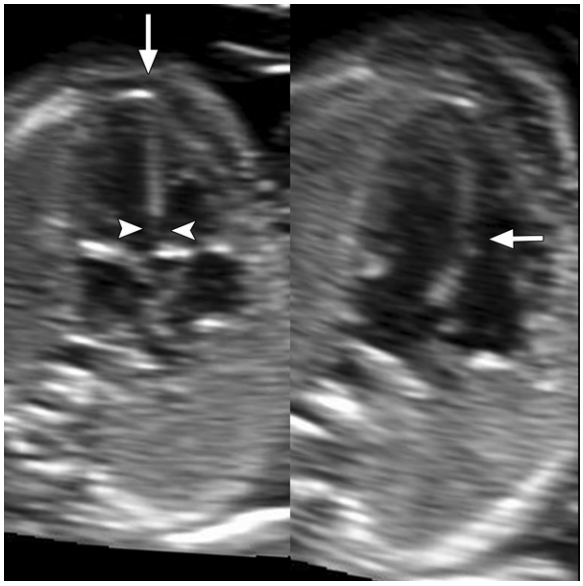


Figure 9. US image of the ventricular septum. Composite images in the same case viewed from different angles of insonation. In the left image, the septum (arrow) is viewed with the beam parallel to its long axis; the normal transition from muscular to membranous septum results in signal dropout (arrowheads), which can be mistaken for a VSD. In the right image, the angle of insonation is changed so that the beam is perpendicular to the septum (arrow) and shows that it is intact. Color Doppler imaging may also be used to look for flow across any possible VSD.

change in thickness can be misinterpreted as a VSD if the beam traverses from the apex toward the crux of the heart (Fig 9). Use additional scan planes or color Doppler flow imaging to exclude a VSD at this level, and check the angle of the aorta to the left ventricle and the continuity of the septum and anterior aortic wall on the LVOT view.

The atrial septum is thin; the foramen ovale is the normal opening in it that allows passage of oxygenated blood from the right heart to the left. A fetal atrial septal defect (ASD) will be of the primum type, located at the crux of the heart, and unlike the foramen ovale, a septum primum ASD is not covered by a flap. ASD is an extremely difficult diagnosis to make in the fetus.

AV Valve Offset

The AV valves belong to the relevant ventricle. The tricuspid valve is part of the right ventricle; it is more apically placed, located a millimeter closer to the apex of the heart than the mitral valve, and has a septal leaflet attached to the interventricular septum (25). The mitral valve is part of the left ventricle and does not have a septal leaflet. Together with the atrial and ventricular septa, the AV valves form the crux of the heart (Fig 10).

Area Behind the Heart

The descending aorta should be left of midline, touching the left atrium wall. It should be the only discretely identifiable tubular structure behind the heart at the 18–20-week scan (Fig 11). If there is increased space behind the left atrium or another tubular structure there, think about the esophagus, just as it is seen in the azygoesophageal recess on a chest CT image. Fetal swallowing changes the shape and size of the esophagus; therefore, a period of real-time evaluation should allow confident recognition of the esophagus. Persistent vascular structures posterior to the heart may be seen with anomalous pulmonary venous return or with azygos continuation of the IVC (26). Azygos

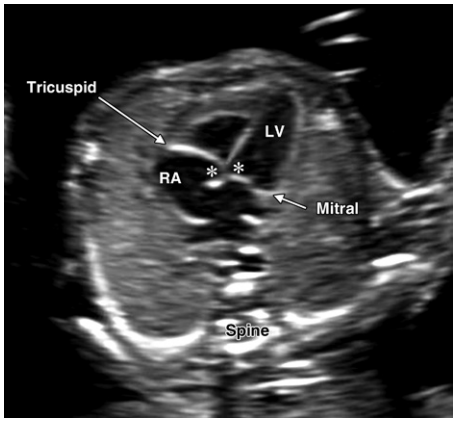


Figure 10. Axial US image of AV valve offset in a fetus at 19 weeks gestation. Four-chamber view shows the normal relationship of the AV valves. The mitral valve belongs to the left ventricle *(LV)*; the tricuspid valve between the right atrium *(RA)* and right ventricle is more apically placed. The septum and AV valves together create the crux of the heart.

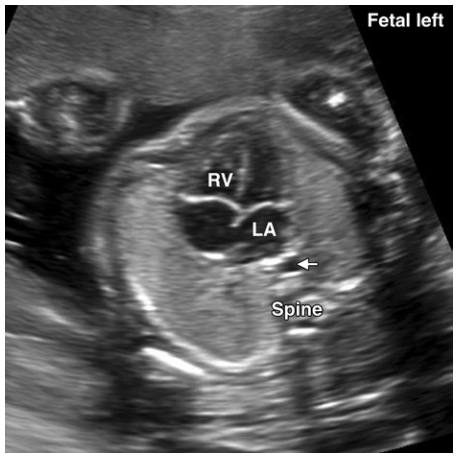


Figure 11. Axial four-chamber US view of the area behind the heart in a fetus at 19 weeks gestation. This view shows a single vessel seen in circular cross section (arrow), left of midline, behind the heart, and closely applied to the left atrial *(LA)* wall. This is the descending aorta. *RV* = right ventricle.

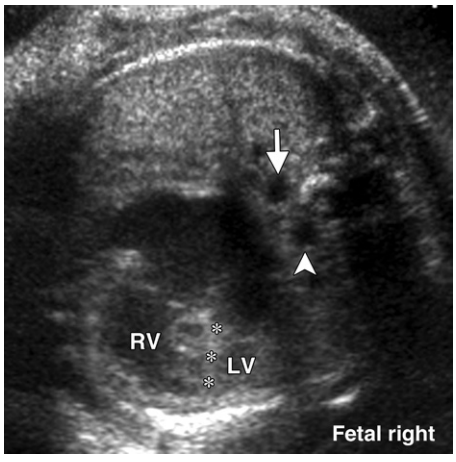


Figure 12. Axial US image shows azygos continuation of the IVC in a fetus at 27 weeks gestation. Four-chamber view in a fetus with situs ambiguous shows the cardiac apex directed to the fetal right and a right-dominant, unbalanced atrioventricular septal defect. ∗ = the septum between the larger right ventricle *(RV)* and smaller left ventricle *(LV)*. There are two similar-sized vessels behind the heart: the aorta (arrow) and the enlarged azygos vein (arrowhead), which is the continuation of an interrupted IVC.

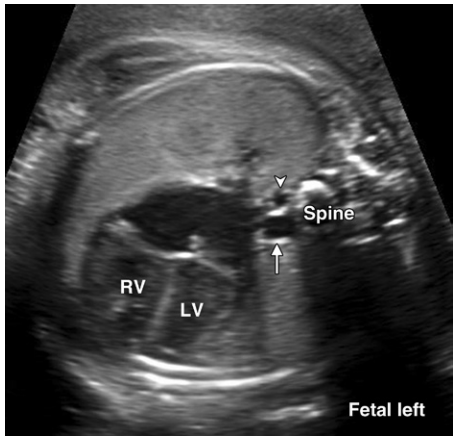


Figure 13. Normal azygos vein in a fetus at 35 weeks gestation. Axial US four-chamber view obtained during a growth follow-up scan shows the normal aorta (arrow). The smaller vessel beside it is the normal azygos vein (arrowhead) In this case, the hepatic veins and IVC were normal, and formal fetal echocardiography helped to confirm normal cardiac anatomy. With modern high-resolution equipment and slender maternal habitus, it is not uncommon to see a normal azygos vein in the late third trimester. This case does not need to be referred to fetal echocardiography. *LV* = left ventricle, *RV* = right ventricle.

continuation of the IVC is associated with situs ambiguous and/or heterotaxy syndromes and severe CHD; this manifests as two vessels of similar size behind the heart (Fig 12). With modern equipment, it is possible to see the normal azygos vein beside the aorta in the third trimester, but it is smaller than the aorta, and the intrahepatic IVC will be present (27) (Fig 13).

Rate and Rhythm

Cardiac rate and rhythm can be documented using M-mode or pulsed Doppler imaging. For M-mode imaging, the beam is directed through one atrium and one ventricle to evaluate atrioventricular conduction (Fig 14). To use pulsed Doppler imaging, the sample volume is placed near the LVOT, adjacent to where the mitral and aortic valves are in fibrous continuity. Atrial contraction results in flow toward the ventricle, while ventricular contraction leads to flow away from the ventricle into the aorta. Thus, the Doppler tracing will show the atrial rate to one

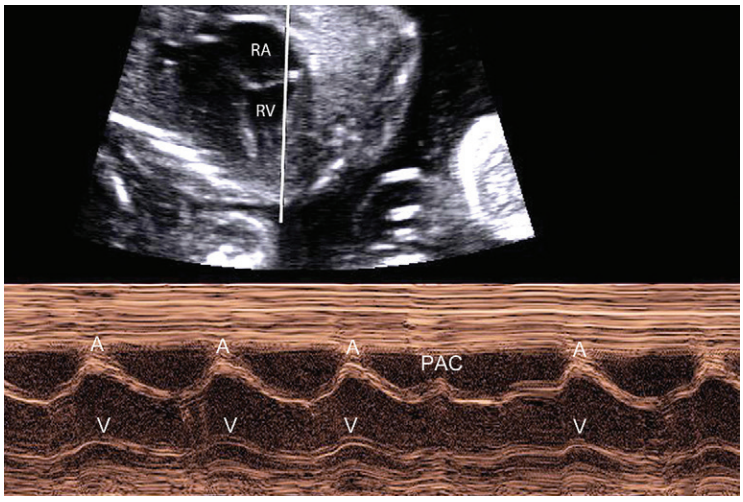


Figure 14. M-mode image of heart rate and rhythm. The M-mode cursor is placed so that it goes through an atrium and a ventricle. The first motion encountered is the atrial contraction *(A)*; a premature atrial contraction *(PAC)* is seen. The ventricle *(V)* normally contracts 1:1 in time with atrial contraction. When there is a blocked PAC, there is a delayed ventricular contraction as seen here. This results in an irregular fetal heart rate at auscultation. Cursor placement through both atria or through both ventricles provides information on the atrial or ventricular rate but not atrioventricular conduction. *RA* = right atrium, *RV* = right ventricle.

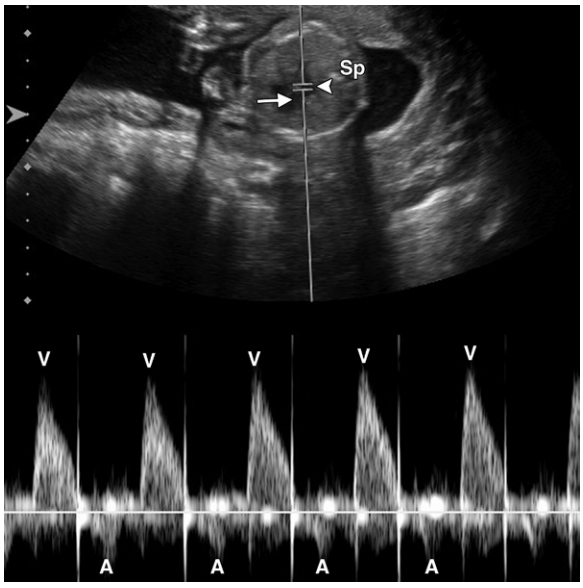


Figure 15. Pulsed Doppler image of heart rate and rhythm. The Doppler sample volume is placed between the left atrium (arrowhead) and left ventricle (arrow). In this example, ventricular contraction *(V)* results in flow out of the ventricle toward the transducer, thus, above the baseline. With atrial contraction *(A)*, the blood moves from the atrium to the ventricle and away from the transducer, shown as below the baseline. This allows assessment of atrial and ventricular rates and atrioventricular conduction.

side of baseline and the ventricular rate on the other (Fig 15).

Outflow Tract Views

The LVOT is formed by the aortic root and trunk; it arises in the center of the heart, runs cephalad, forms a tight turn, and descends in the posterior mediastinum. The head and neck vessels arise from the apex of the curve. Check that there is an angle between the ventricular septum and the ascending aorta; if absent, look for a VSD (Fig 16). Make sure that the vessel exiting the left ventricle does not branch as it exits the pericardium; that is the branch pattern of the pulmonary artery (PA). In transposition of the great arteries, there is ventriculoarterial discordance with the aorta arising from the right ventricle and the PA arising from the left ventricle.

The RVOT is formed by the pulmonary conus and main PA. These wrap around the root of the aorta. As soon as the RVOT exits the pericardium, it branches; the ductus arteriosus then runs posteriorly toward the spine, and the right PA continues to wrap around the aorta. The left PA is present but not visible in this plane (Fig 17).

The crisscross sign is used to describe the real-time visualization of the outflow tracts crossing each other as they exit the heart; they are never parallel at the level of the aortic and pulmonary valves. The normal crisscross sign is to be differentiated from the extremely rare pathologic entity

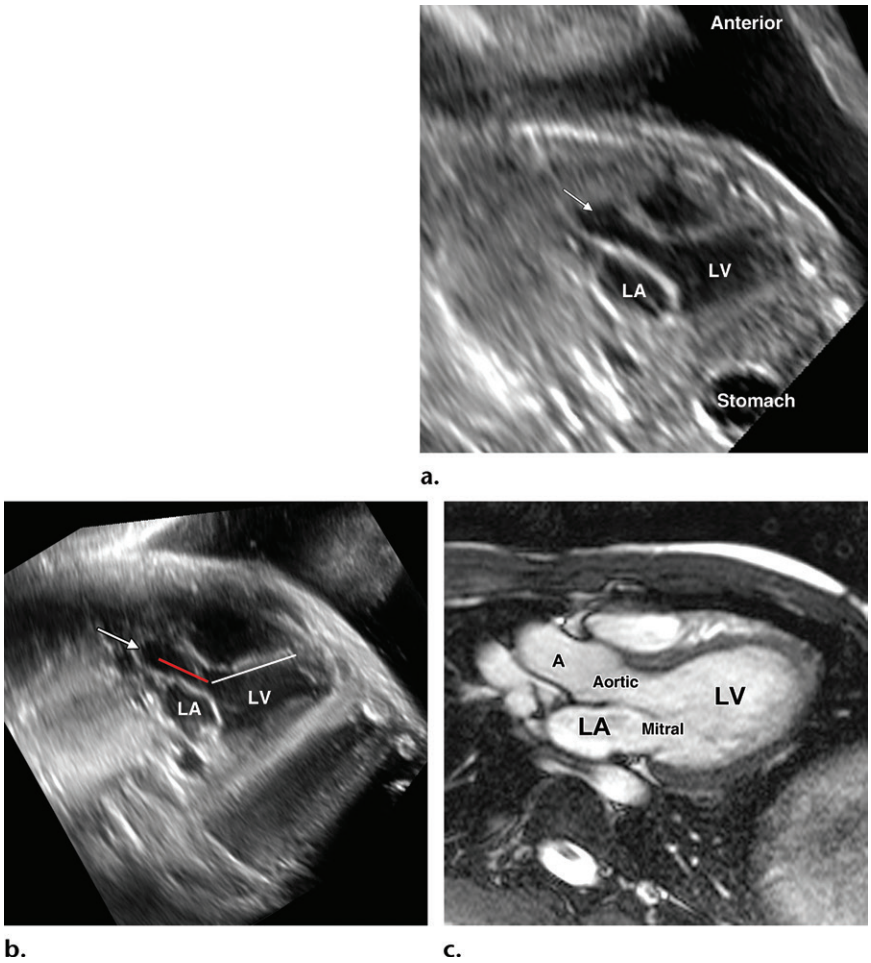


Figure 16. Images of LVOT in a fetus at 20 weeks gestation **(a, b)** and an adult cardiac patient **(c)**. **(a)** Long-axis US view shows the vessel exiting the smooth-walled ventricular chamber. The left atrium *(LA)* and left ventricle *(LV)* are separated by the mitral valve, and the aorta (arrow) does not branch. Both the anterior and posterior walls of the aorta form distinct echogenic lines; any signal dropout is suspicious for a VSD. In real time, this view shows the fibrous continuity of the mitral and aortic valves. **(b)** In the same orientation, the ventricular wall (white line) creates an angle with the long axis of the aorta (red line). This angle is lost with the gooseneck aortic deformity seen when the aorta is "sprung" anteriorly in the setting of an atrioventricular septal defect. **(c)** LVOT view from adult cardiac MR imaging shows the same anatomy in a format that may be more familiar to radiologists. The mitral and aortic valve leaflets are visible. *A* = aorta, *LA* = left atrium, *LV* = left ventricle.

of crisscross heart, in which the atria connect with the contralateral ventricles and the ventricular chambers are arranged in a superoinferior fashion. On a cine clip at this level, the aortic and pulmonary valve leaflets should "come and go" throughout the cardiac cycles if they are normal in thickness and mobility. The outflows are perpendicular to each other; if one is seen in circular cross section, the other should be seen in the long axis as a tube (Fig 18). Remember that the SVC and ascending aorta are close to each other, but the plane of the LVOT view does not include the SVC. If two parallel arteries are seen exiting the heart, the most likely diagnoses are transposition of the great arteries or double-outlet right ventricle. Table 2 shows checklist items for the outflow tract views.

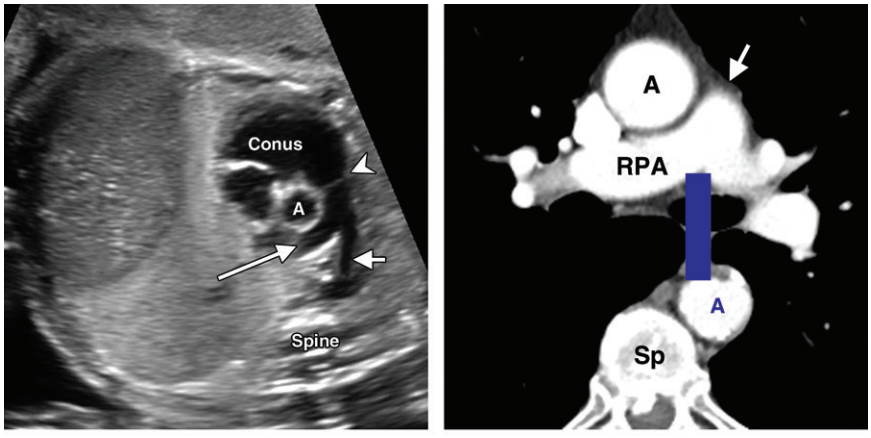
Normal Anatomy: Complex Fetal Cardiac Scan

Additional cardiothoracic views required for performance of the complex obstetric US examination (CPT code 78611) include the aortic arch view, the bicaval view, 3VV, 3VT, and illustration of diaphragmatic integrity.

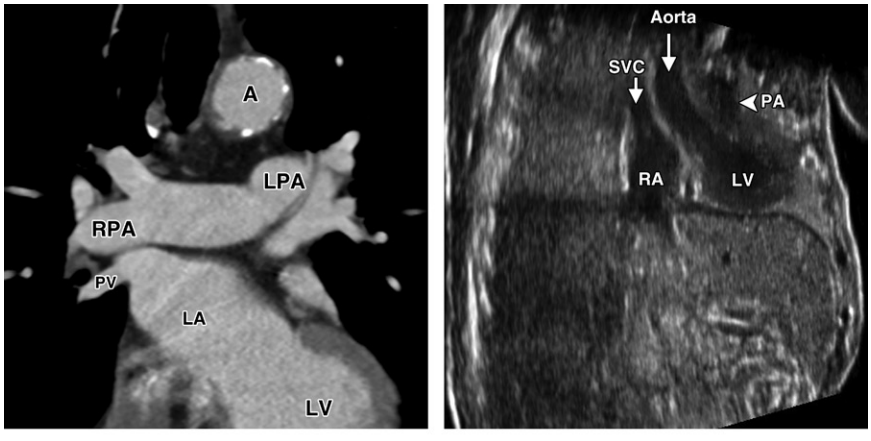
Aortic Arch View

This is an oblique sagittal view similar to a left anterior oblique angiogram or the sagittal arch view obtained in CT arteriography (Fig 19). The isthmus, after the takeoff of the left subclavian artery, is the narrowest part of the arch. Unfortunately, this is also the commonest site of coarctation, which is a difficult diagnosis to make in the fetus.

Figure 17. RVOT images. **(a)** Long-axis view shows the other great vessel exiting the heart. Note that the aorta *(A)* is now seen in circular cross section because it runs 90° to the main PA. The pulmonary valve (arrowhead) is at the origin of the main PA from the pulmonary conus. The main PA divides early such that in this scan plane the ductus arteriosus (short arrow) runs toward the spine and the right pulmonary artery (long arrow) wraps around the aortic root. **(b)** Adult contrast-enhanced chest CT image shows the same anatomy in a format that may be more familiar to radiologists. The aortic root (black *A*) is "hugged" by the right pulmonary artery *(RPA)*, which arises from the main PA (arrow). The ductus arteriosus (blue rectangle) connects it to the descending aorta (blue *A*) in fetal life. *Sp* = spine.



a. b.



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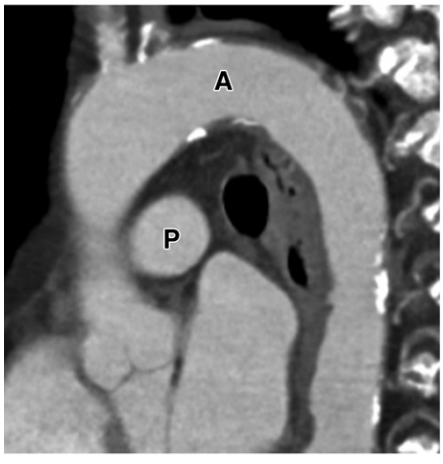

c.

Figure 18. Outflow tract orientation. **(a)** Coronal enhanced CT image shows the aorta *(A)* in circular cross section but the PA in the long axis as the right and left PAs *(RPA, LPA)* enter the lungs. The pulmonary vein *(PV)* enters the left atrium *(LA)*. *LV* = left ventricle. **(b)** Coronal oblique fetal US through the fetal chest is not a standard cardiac view, but it shows the crisscross orientation of the great arteries. The aorta arises from the left ventricle *(LV)* and ascends parallel to the SVC as it descends to drain into the right atrium *(RA)*. The PA is seen in oval cross section as it curves around the aortic root. **(c)** Sagittal enhanced CT image shows the aortic arch *(A)* in the long axis and the PA *(P)* in circular cross section because the great vessels are at 90° to each other.

Table 2: Checklist for Outflow Tract Views

LVOT
Left atrium to left ventricle to aortic root
Smooth ventricular contour
Septal continuity with anterior wall of aorta
Angle of aorta to septum
Gives rise to head and neck vessels from apex of arch
RVOT
Wraps around root of the aorta, which is central in the heart
Bifurcates into ductus arteriosus and right PA as it exits pericardium
3VV
SVC < aorta < PA
PA continues toward spine as ductus arteriosus
3VT
Aorta and PA converge to form a V shape to left of trachea
Direction of flow same in aorta and PA
No vessels to left of PA

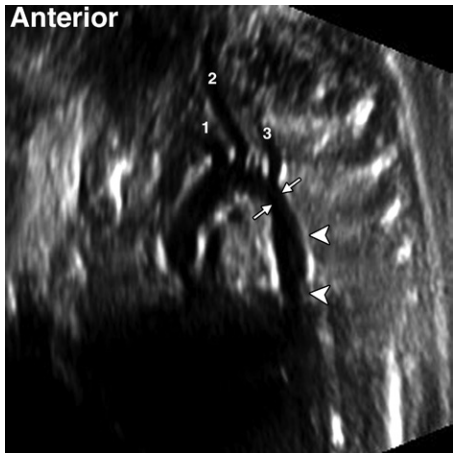

Figure 19. Left parasagittal US image of the aortic arch in a fetus at 30 weeks gestation. The apical curve is small in radius; as a result, this view is often called the candy-cane view. The head and neck vessels arise from the apex of the arch (*1* = innominate artery, *2* = left common carotid artery, *3* = left subclavian artery). The arrows indicate the aortic isthmus, which is the most common site for coarctation to develop. After uniting with the ductus arteriosus, the vessel continues toward the abdomen as the descending aorta (arrowheads). This is the same anatomy as seen on Figure 18c.

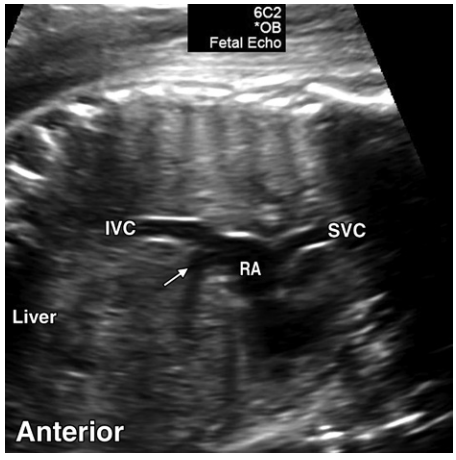

Figure 20. Bicaval view in a fetus at 19 weeks gestation. Right parasagittal US image shows the SVC and IVC entering the right atrium *(RA)*. The hepatic veins (arrow) are visible within the fetal liver. Be careful not to confuse hepatic veins with the IVC if there is a question of azygos continuation of the IVC. Hepatic veins are smaller and travel in an oblique course through the liver.

Bicaval View

The bicaval view is a parasagittal view showing the SVC and IVC entering the right atrium. The SVC and IVC should be similar in size, and it is important to follow the IVC into the liver for some distance to make sure that it is not interrupted as may be seen in azygos continuation of the IVC, which is associated with heterotaxy syndromes. The right hepatic vein is frequently visible in the liver on this view as well (Fig 20).

Three-Vessel View

The 3VV is another way to look at the outflow tracts. It is obtained by sweeping toward the fetal head from the axial four-chamber view. Look for the size of the three vessels seen from right to left; normally the SVC is smaller than the aorta, which is smaller than the PA. The ductus arteriosus should be directed posteriorly toward the spine to unite with the descending aorta (28,29) (Fig 21).

Three-Vessel Trachea View

The 3VT is a variation of the 3VV that includes views of the trachea and esophagus. It is obtained by sweeping superior and toward the left from the 3VV. It shows the confluence of the ductal and aortic arches, which come together in a V shape with the *V* open to the anterior chest wall and separated from the sternum by the thymus. The limbs of the *V* (the ductal and aortic arches) should be similar in size and show flow in the same direction. The ductal limb becomes slightly larger than the aortic limb in late pregnancy. The vertex points left of the trachea, anterior to the spine. There should be no vessels left of the PA. A fourth vessel may be seen in this view in the setting of a persistent left SVC or in total anomalous pulmonary venous return (TAPVR) superior drainage via a vertical vein (30).

Diaphragmatic Integrity

Document that both sides of the diaphragm are continuous anterior to posterior on parasagit-

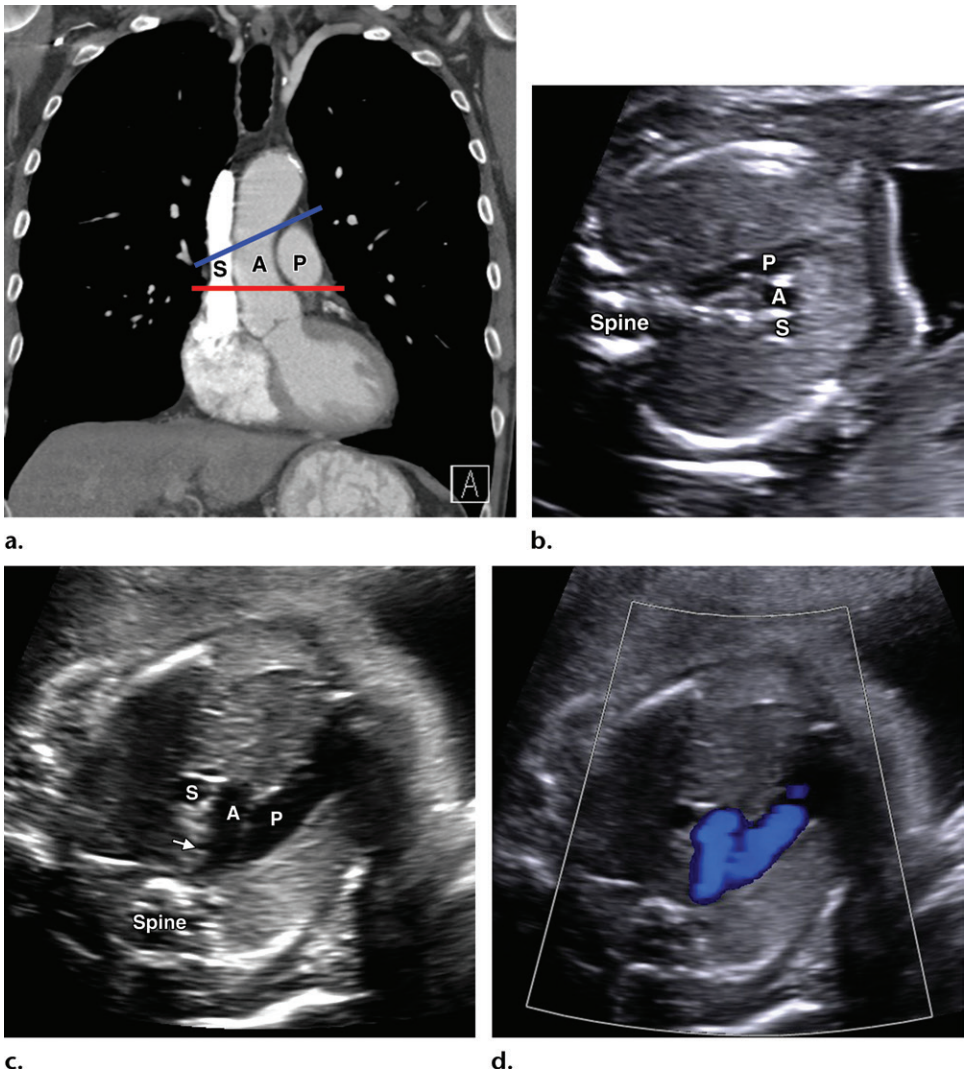

Figure 21. 3VV and 3VT. *A* = aorta, *P* = PA, *S* = SVC. **(a)** Coronal enhanced CT image shows the approximate scan planes for the 3VV (red line) and 3VT (blue line). **(b)** Axial fetal US image shows a normal 3VV with the SVC, the aorta, and the PA as it continues posteriorly into the ductal arch. **(c)** Axial oblique US image shows a normal 3VT with the SVC and the fluid-filled trachea (arrow). The aortic and ductal (PA to ductus arteriosus) arches come together in a V shape to the left of the trachea. Note that the PA is slightly larger than the aorta. **(d)** Color Doppler image at the same level shows that the direction of flow is the same in both great vessels. With duct-dependent CHD, one of the great vessels fills retrogradely from the ductus. If this occurs, the direction of flow (ie, color) in the limbs of the *V* will differ.

tal views. This is also best demonstrated on cine clips, which can be obtained at the same time as the longitudinal views of the fetal spine.

Conclusion

CHD may be isolated but it may indicate aneuploidy or a syndrome. When present, aneuploidy and other anomalies determine the prognosis. When isolated, the prognosis is determined by the exact nature of the abnormalities. A simple VSD will likely resolve with no intervention, and mild forms of CHD may just require serial monitoring of the child over time. However, hypoplastic left heart syndrome requires a series of surgical procedures at best and, in some cases, can only be managed with cardiac transplantation. Duct-dependent CHD means that there is obstruction of either the right- or left-sided circulation with retrograde filling of one great vessel from the other via the ductus arteriosus. At birth, the normal transition from fetal to postnatal circulation results in closure of the ductus venosus and ductus arteriosus. Once the ductus arteriosus closes, there is no option for retrograde filling; the pulmonary and systemic circulations are separated with shutdown of whichever outflow is obstructed. The result is circulatory collapse in the infant.

In any fetal US examination, possible CHD needs to be taken seriously. At the community level, the sonologist's function is to identify those cases that require additional evaluation. Use of the systematic approach outlined in this article

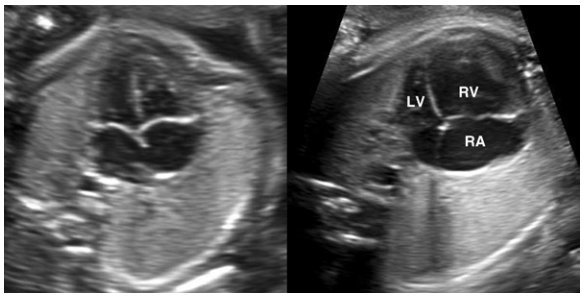

Figure 22. Application of the checklist to assess the four-chamber view. Left image shows a normal four-chamber view. In the right image, the position is rotated, the axis is increased, and neither the atria nor the ventricles are symmetric in size. This four-chamber view is abnormal. The patient needs referral for formal fetal echocardiography. In this case, the final diagnosis was a severe coarctation of the aorta. *LV* = left ventricle, *RA* = right atrium, *RV* = right ventricle.

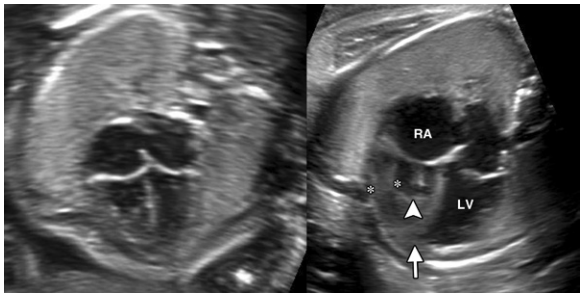

Figure 24. Application of the checklist to assess the four-chamber view. Left image shows a normal four-chamber view. In the right image, there is loss of ventricular symmetry; the left ventricular *(LV)* apex (arrow) wraps around that of the right ventricle (arrowhead). The right ventricular wall (*) is markedly thickened in comparison with that of the LV. This four-chamber view is abnormal. The patient needs referral for formal fetal echocardiography. In this case, the final diagnosis was pulmonary atresia with an intact ventricular septum. *RA* = right atrium.

should allow more confident determination of normal versus abnormal (Figs 22–25). If the heart does not look normal, refer the patient for expert evaluation. Once an abnormality is confirmed, a personalized pregnancy management plan can be developed depending on the nature of the lesion and the desires of the family.

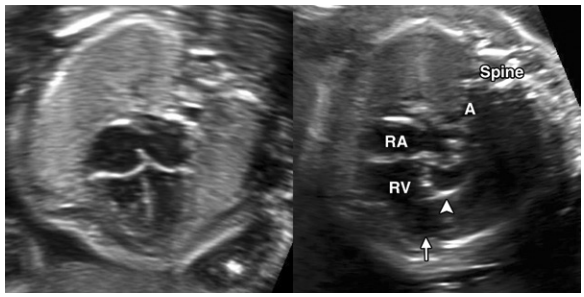

Figure 23. Application of the checklist to assess the four-chamber view. Left image shows a normal four-chamber view. In the right image, there is loss of ventricular and atrial symmetry; the left atrium and left ventricle are smaller than the right atrium *(RA)* and right ventricle *(RV)*. The left ventricle is not apex-forming (arrowhead = left ventricular apex, arrow = right ventricular apex). Despite the small left heart chambers, the aorta *(A)* is normal and fills via retrograde flow from the ductus arteriosus. This four-chamber view is abnormal, and the patient needs referral for formal fetal echocardiography. In this case, the final diagnosis was hypoplastic left heart syndrome.

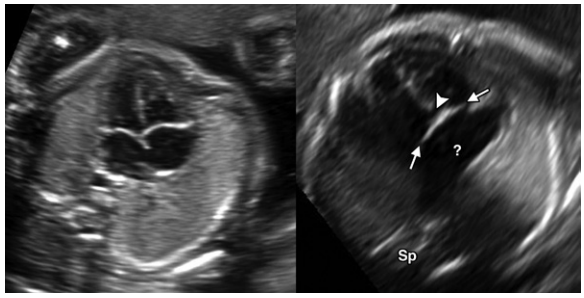

Figure 25. Application of the checklist to assess the four-chamber view. Left image shows a normal four-chamber view. In the right image, the axis is abnormal, and the "crux" of the heart is absent. Instead of the normal offset AV valves, there is a single low-slung valve (arrows). Note the VSD (arrowhead) and the lack of the atrial septum and foramen ovale *(?)*. The four-chamber view is abnormal, and the patient needs referral for formal fetal echocardiography. In this case, the final diagnosis was a balanced atrioventricular septal defect. The abnormal cardiac axis was detected at the time of nuchal translucency screening. Cell-free fetal deoxyribonucleic acid (DNA) testing indicated increased risk for trisomy 21. The family opted for termination because of the heart defect, which was diagnosed at 17 weeks gestation.

References

1. Pinto NM, Keenan HT, Minich LL, Puchalski MD, Heywood M, Botto LD. Barriers to prenatal detection of congenital heart disease: a population-based study. Ultrasound Obstet Gynecol 2012;40(4):418–425.
2. Reddy UM, Abuhamad AZ, Levine D, Saade GR; Fetal Imaging Workshop Invited Participants. Fetal imaging: executive summary of a joint Eunice Kennedy Shriver National Institute of Child Health and Human Development, Society for Maternal-Fetal Medicine, American Institute of Ultrasound in Medicine, American College of Obstetricians and Gynecologists, American College of Radiology, Society for Pediatric Radiology, and Society of Radiologists in Ultrasound fetal imaging workshop. J Ultrasound Med 2014;33(5):745–757.
3. Khoo NS, Van Essen P, Richardson M, Robertson T. Effectiveness of prenatal diagnosis of congenital heart defects in South Australia: a population analysis 1999-2003. Aust N Z J Obstet Gynaecol 2008;48(6):559–563.
4. Stoll C, Garne E, Clementi M; EUROSCAN Study Group. Evaluation of prenatal diagnosis of associated congenital heart diseases by fetal ultrasonographic examination in Europe. Prenat Diagn 2001;21(4):243–252.
5. Carvalho JS, Allan LD, Chaoui R, et al; International Society of Ultrasound in Obstetrics and Gynecology. ISUOG practice guidelines (updated): sonographic screening examination of the fetal heart. Ultrasound Obstet Gynecol 2013;41(3):348–359.
6. American Institute of Ultrasound in Medicine. AIUM practice guideline for the performance of obstetric ultrasound examinations. J Ultrasound Med 2013;32(6):1083–1101.
7. Pruetz JD, Carroll C, Trento LU, et al. Outcomes of critical congenital heart disease requiring emergent neonatal cardiac intervention. Prenat Diagn 2014;34(12):1127–1132.
8. Tworetzky W, McElhinney DB, Reddy VM, Brook MM, Hanley FL, Silverman NH. Improved surgical outcome after

fetal diagnosis of hypoplastic left heart syndrome. Circulation 2001;103(9):1269–1273.
9. Jegatheeswaran A, Oliveira C, Batsos C, et al. Costs of prenatal detection of congenital heart disease. Am J Cardiol 2011;108(12):1808–1814.
10. Levey A, Glickstein JS, Kleinman CS, et al. The impact of prenatal diagnosis of complex congenital heart disease on neonatal outcomes. Pediatr Cardiol 2010;31(5):587–597.
11. Poole PS, Chung R, Lacoursiere Y, et al. Two-dimensional sonographic cine imaging improves confidence in the initial evaluation of the fetal heart. J Ultrasound Med 2013;32(6):963–971.
12. Sklansky M. Current guidelines for fetal echocardiography: time to raise the bar. J Ultrasound Med 2011;30(2):284–286; author reply 286.
13. Chaoui R. The four-chamber view: four reasons why it seems to fail in screening for cardiac abnormalities and suggestions to improve detection rate. Ultrasound Obstet Gynecol 2003;22(1):3–10.
14. Wax J, Minkoff H, Johnson A, et al. Consensus report on the detailed fetal anatomic ultrasound examination: indications, components, and qualifications. J Ultrasound Med 2014;33(2):189–195.
15. Applegate KE, Goske MJ, Pierce G, Murphy D. Situs revisited: imaging of the heterotaxy syndrome. RadioGraphics 1999;19(4):837–852; discussion 853–854.
16. Sinkovskaya E, Horton S, Berkley EM, Cooper JK, Indika S, Abuhamad A. Defining the fetal cardiac axis between 11 + 0 and 14 + 6 weeks of gestation: experience with 100 consecutive pregnancies. Ultrasound Obstet Gynecol 2010; 36(6):676–681.
17. Sinkovskaya ES, Chaoui R, Karl K, Andreeva E, Zhuchenko L, Abuhamad AZ. Fetal cardiac axis and congenital heart defects in early gestation. Obstet Gynecol 2015;125(2):453–460.
18. Awadh AM, Prefumo F, Bland JM, Carvalho JS. Assessment of the intraobserver variability in the measurement of fetal cardiothoracic ratio using ellipse and diameter methods. Ultrasound Obstet Gynecol 2006;28(1):53–56.
19. Todros T, Paladini D, Chiappa E, et al. Pulmonary stenosis and atresia with intact ventricular septum during prenatal life. Ultrasound Obstet Gynecol 2003;21(3):228–233.
20. Pauliks LB. The effect of pregestational diabetes on fetal heart function. Expert Rev Cardiovasc Ther 2015;13(1):67–74.
21. Chanthasenanont A, Somprasit C, Pongrojpaw D. Nomograms of the fetal heart between 16 and 39 weeks of gestation. J Med Assoc Thai 2008;91(12):1774–1778.
22. Sharland GK, Allan LD. Normal fetal cardiac measurements derived by cross-sectional echocardiography. Ultrasound Obstet Gynecol 1992;2(3):175–181.
23. Carrilho MC, Barros FS, Araujo Júnior E. Increasing isolated right heart chambers in second trimester ultrasound: always a warning sign? Rev Bras Cir Cardiovasc 2014; 29(2):IV–V.
24. Quartermain MD, Cohen MS, Dominguez TE, Tian Z, Donaghue DD, Rychik J. Left ventricle to right ventricle size discrepancy in the fetus: the presence of critical congenital heart disease can be reliably predicted. J Am Soc Echocardiogr 2009;22(11):1296–1301.
25. Lapierre C, Rypens F, Grignon A, Dubois J, Déry J, Garel L. Prenatal ultrasound screening of congenital heart disease in the general population: general concepts, guidelines, differential diagnoses. Ultrasound Q 2013;29(2):111–124.
26. Ganesan S, Brook MM, Silverman NH, Moon-Grady AJ. Prenatal findings in total anomalous pulmonary venous return: a diagnostic road map starts with obstetric screening views. J Ultrasound Med 2014;33(7):1193–1207.
27. Berg C, Georgiadis M, Geipel A, Gembruch U. The area behind the heart in the four-chamber view and the quest for congenital heart defects. Ultrasound Obstet Gynecol 2007;30(5):721–727.
28. Yoo SJ, Lee YH, Kim ES, et al. Three-vessel view of the fetal upper mediastinum: an easy means of detecting abnormalities of the ventricular outflow tracts and great arteries during obstetric screening. Ultrasound Obstet Gynecol 1997;9(3):173–182.
29. Yoo SJ, Lee YH, Cho KS. Abnormal three-vessel view on sonography: a clue to the diagnosis of congenital heart disease in the fetus. AJR Am J Roentgenol 1999;172(3):825–830.
30. Jeanty P, Chaoui R, Tihonenko I, Grochal F. A review of findings in fetal cardiac section drawings. III. The 3-vessel-trachea view and variants. J Ultrasound Med 2008;27(1):109–117.