

# Prenatal Diagnosis of Congenital Cardiac Anomalies: A Practical Approach Using Two Basic Views<sup>1</sup>

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## ONLINE-ONLY CME

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## LEARNING OBJECTIVES

After reading this article and taking the test, the reader will be able to:

- Describe the technique used to obtain the four-chamber view of the fetal heart.
- Describe the technique used to obtain the base view of the fetal heart.
- Discuss the advantage of adding the base view to a routine fetal US examination.

## SUPPLEMENTAL MATERIAL

Movie clips to supplement this article are available online at [radiographics.rsna.org/cgi/content/full/22/5/1125/DC1](http://radiographics.rsna.org/cgi/content/full/22/5/1125/DC1).

Structural cardiac anomalies are estimated to occur in 8 of every 1,000 live births. Cardiovascular anomalies are frequently associated with other congenital anomalies because the heart is among the last organs to develop completely in the embryo. The guidelines for routine prenatal evaluation of both the American College of Radiology and the American Institute of Ultrasound in Medicine require evaluation of the fetal heart. The ultrasonographic (US) view that is most commonly used is the four-chamber view, which allows assessment of abnormalities involving the atria and the ventricles. However, this view is not adequate for demonstrating the outflow tracts of the aorta and pulmonary artery. A base view that demonstrates the crossing of the great vessels can be obtained just superior to the four-chamber view. Addition of the base view to routine US evaluation with the four-chamber view increases not only the sensitivity for detection of cardiac anomalies but also the accuracy of diagnosis.

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**Abbreviation:** VSD = ventricular septal defect

**Index terms:** Fetus, cardiovascular system, 856.8751 • Fetus, US, 856.1298 • Heart, abnormalities, 50.143, 50.16, 50.1715 • Heart, US, 50.1298 Tetralogy of Fallot, 50.145 • Ventricular septal defect, 515.142

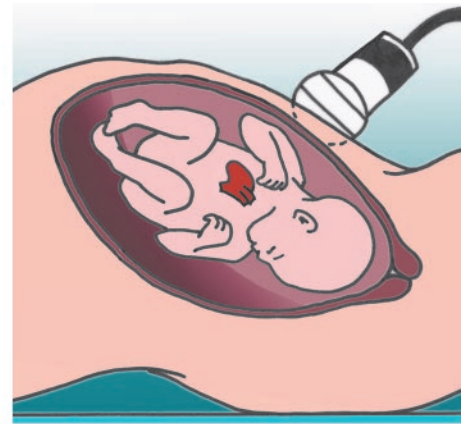
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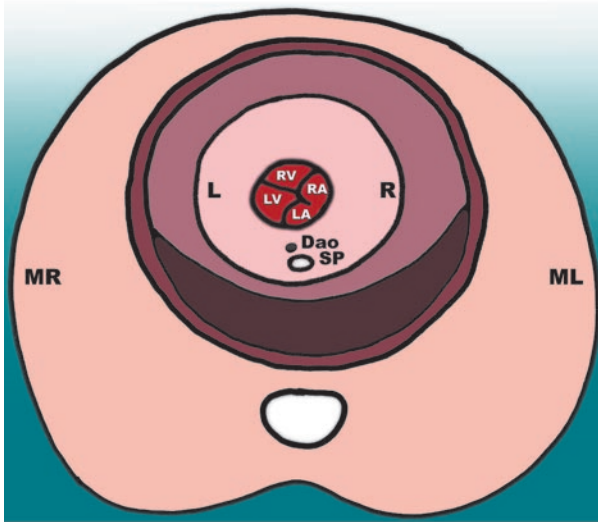
See the commentary by McGahan following this article.

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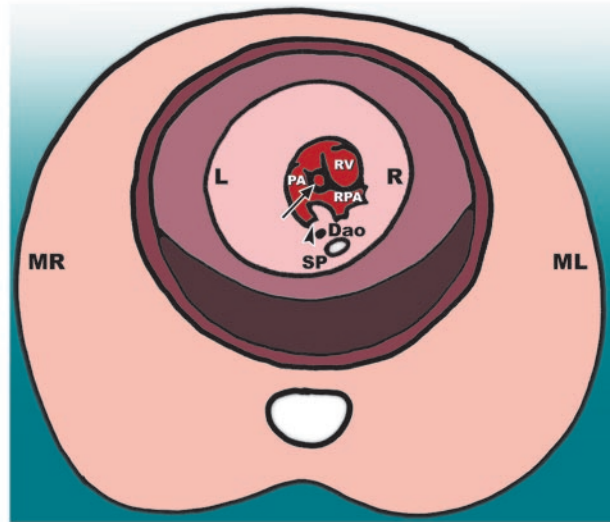
**Figure 1.** Determination of fetal orientation with a cephalic presentation. Throughout the figure legends, *L* = fetal left, *LA* = left atrium, *LV* = left ventricle, *R* = fetal right, *RA* = right atrium, *RV* = right ventricle. **(a)** Sagittal drawing shows the fetal head near the maternal cervix and the fetal rump near the uterine fundus. **(b)** Drawing of a four-chamber view of the fetal heart with the fetus in a supine position. *Dao* = descending aorta, *ML* = maternal left, *MR* = maternal right, *SP* = spine. **(c)** Drawing of a base view of the fetal heart with the fetus in a supine position shows the main pulmonary artery (*PA*) crossing from right to left over the ascending aorta (arrow). Arrowhead = ductus arteriosus, *Dao* = descending aorta, *ML* = maternal left, *MR* = maternal right, *RPA* = right pulmonary artery, *SP* = spine (*Fig 1 continues*).



a.



b.



c.

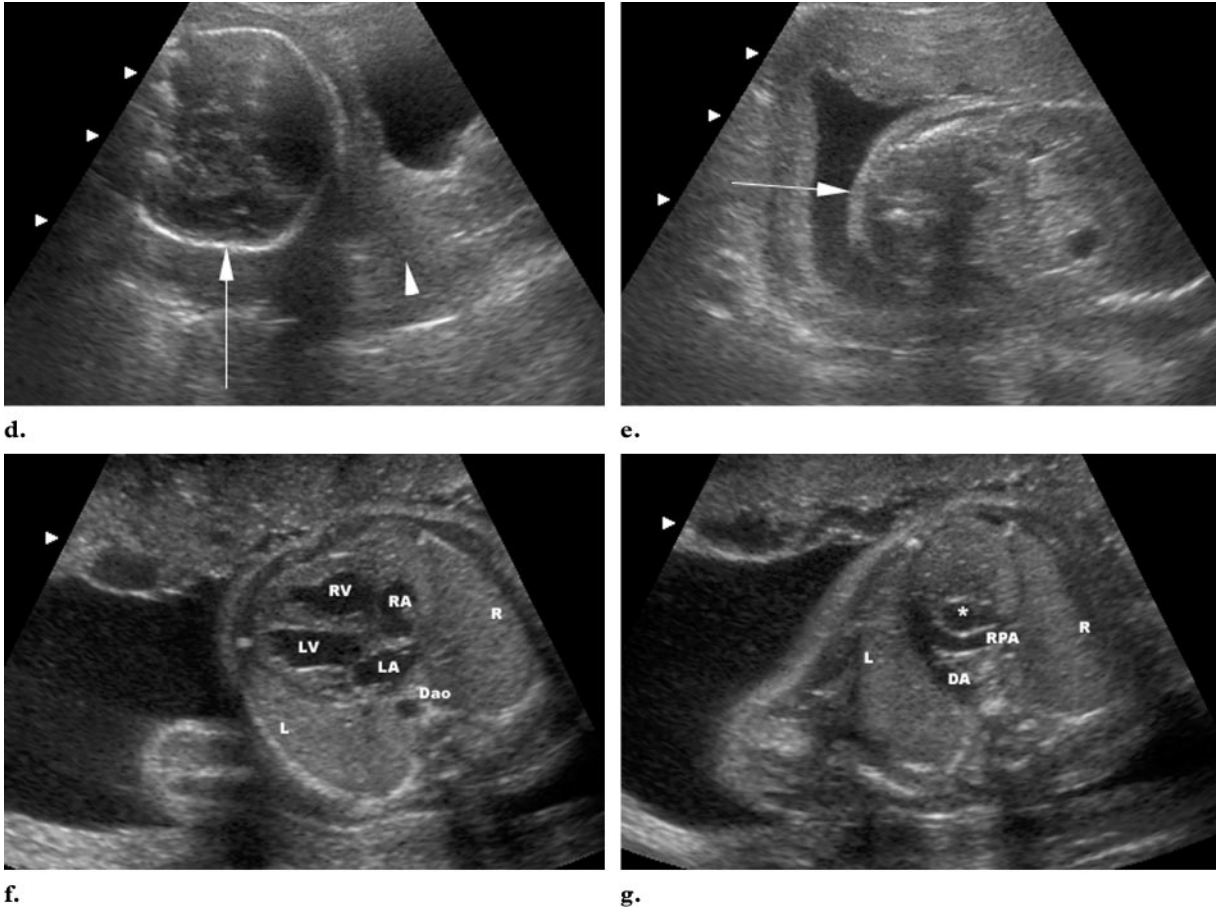
### Introduction

Structural cardiac anomalies are estimated to occur in 8 of 1,000 live births (1–4). Cardiovascular anomalies are frequently associated with other congenital anomalies because the heart begins to develop the 3rd week after conception and continues to develop until the end of the 8th week (5). Since the heart is basically developing during the entire period of organogenesis, it is susceptible to the development of anomalies. Since most cardiac abnormalities are found in patients without associated risk factors (1), evaluation of the fetal heart is an important component of a routine obstetric ultrasonographic (US) examination. The guidelines for routine prenatal evaluation of both the American College of Radiology and the

American Institute of Ultrasound in Medicine require evaluation of the fetal heart. The view most commonly obtained is the four-chamber view, which allows assessment of abnormalities involving the atria and the ventricles. However, the four-chamber view does not demonstrate the outflow tracts of the aorta and pulmonary artery. Therefore, abnormalities involving the great vessels can be missed.

A base view that demonstrates the crossing of the great vessels can be obtained just superior to the four-chamber view. In the base view, the diameters of both the ascending aorta and the main pulmonary artery can be measured. Also, the bifurcation of the main pulmonary artery into the right pulmonary artery and the ductus arteriosus can be visualized. The addition of this second view is valuable in the determination of anomalies involving the great vessels at the base of the heart.

**Figure 1** (continued). (d) Sagittal transabdominal US image shows the fetal head (arrow) near the maternal cervix (arrowhead). (e) Sagittal transabdominal US image of the uterine fundus shows the fetal rump (arrow). (f) Transabdominal US image (four-chamber view) of a supine fetus. *Dao* = descending aorta. (g) Transabdominal US image (base view) of a supine fetus shows the right pulmonary artery (*RPA*) crossing over the ascending aorta (\*). *DA* = descending aorta.



Obtaining both views of the heart is a relatively simple approach that allows inclusive assessment of the major cardiac anomalies. Incorporating this approach as part of the routine makes for a shorter learning curve than when it is used only on occasion.

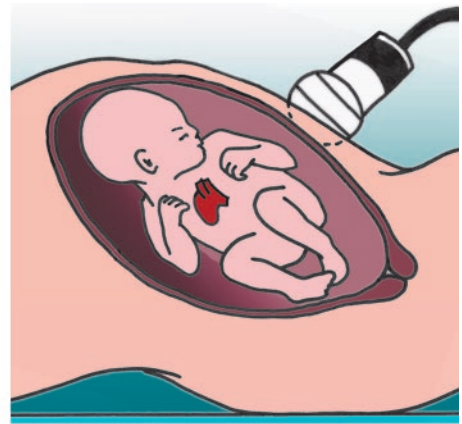
Before any abnormalities can be described, the proper technique of obtaining both the four-chamber view and the base view of the heart needs to be discussed. To emphasize the importance of each technique, abnormalities are presented in the following order: those that can be diagnosed primarily with the four-chamber view and those that can be diagnosed primarily with the base view. Of those that can best be diagnosed with the four-chamber view, the following are described: hypoplastic left heart syndrome, endocardial cushion defect, and ventricular septal defect. Heart defects that may be missed on the

four-chamber view but detected on the base view include persistent truncus arteriosus, complete transposition of the great arteries, double-outlet right ventricle, and tetralogy of Fallot.

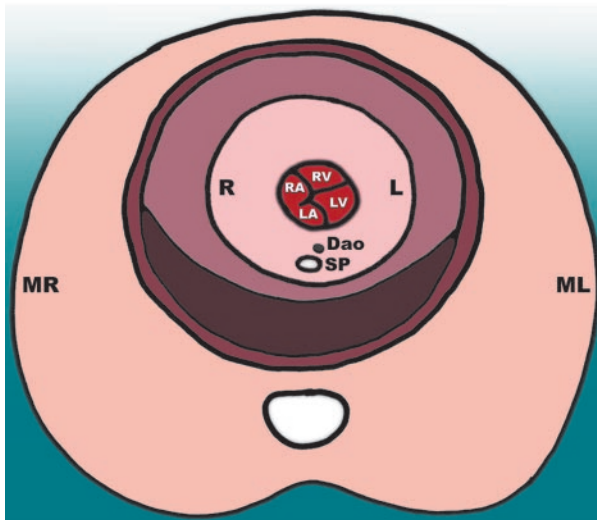
### Technique

In evaluating the fetal heart, the presentation and lie of the fetus should first be documented by obtaining longitudinal images of the cervix and uterine fundus. Following this, transverse images are obtained to determine the orientation of the fetal right side and left side. With a fetus in the supine or prone position, the spine becomes the point of reference in determining fetal orientation. If the fetus is lying on one side, note is made as to whether the right or left side is dependent (Figs 1, 2). The usual scanning sequence begins with the

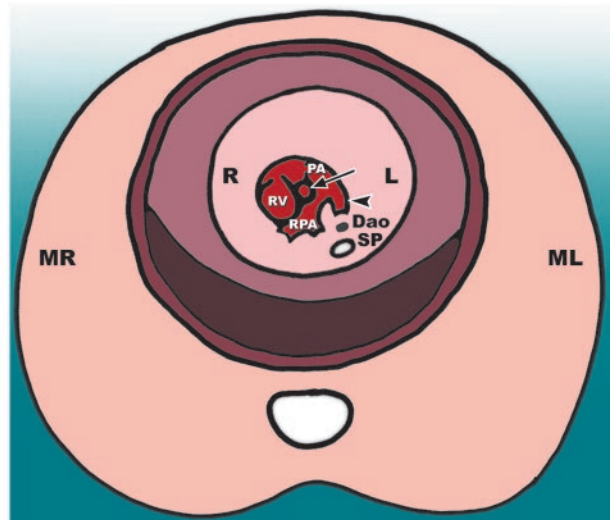
**Figure 2.** Determination of fetal orientation with a breech presentation. **(a)** Sagittal drawing shows the fetal rump near the maternal cervix and the fetal head near the uterine fundus. **(b)** Drawing of a four-chamber view of the fetal heart with the fetus in a supine position. *Dao* = descending aorta, *ML* = maternal left, *MR* = maternal right, *SP* = spine. **(c)** Drawing of a base view of the fetal heart with the fetus in a supine position shows normal crossing of the pulmonary artery (*PA*) over the ascending aorta (arrow). Arrowhead = ductus arteriosus, *Dao* = descending aorta, *ML* = maternal left, *MR* = maternal right, *RPA* = right pulmonary artery, *SP* = spine (*Fig 2 continues*).



a.



b.

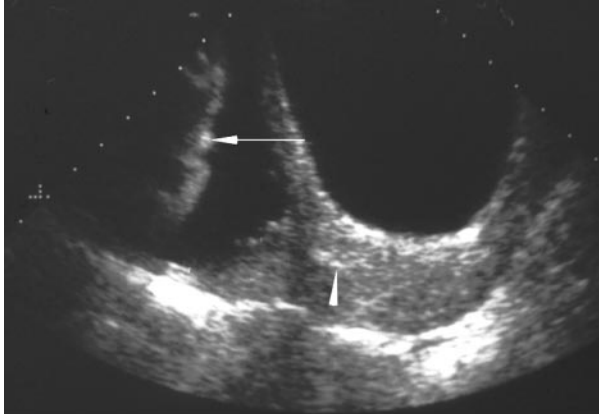


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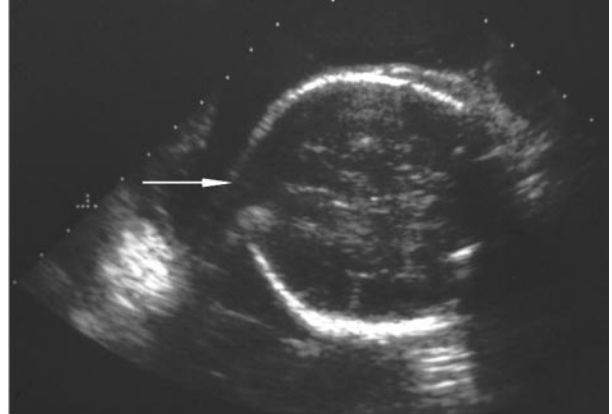
long axis of the thoracic spine; the transducer is rotated 90° until a four-chamber view of the heart is seen. Once the four-chamber view is visualized, the base view of the heart can be obtained by an-

gling the transducer slightly cephalad or minimally sliding the transducer superiorly. It may be difficult to obtain high-quality images of the heart in a fetus with congenital heart disease. This is because other organs may be involved in a spectrum of anomalies.

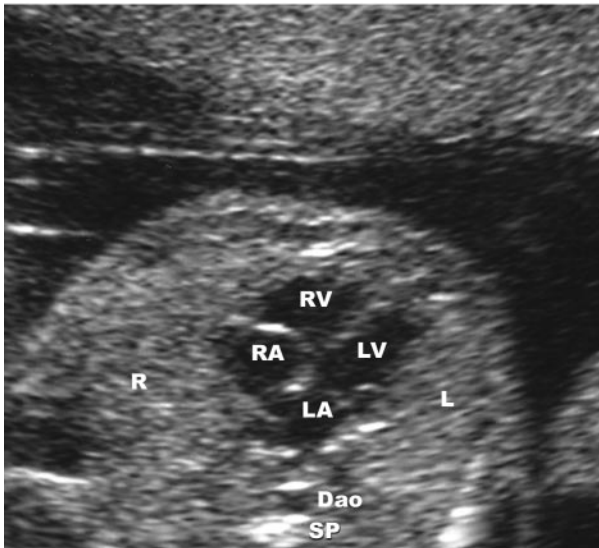
**Figure 2 (continued).** (d) Sagittal transabdominal US image shows the fetal rump (arrow) near the maternal cervix (arrowhead). (e) Sagittal transabdominal US image of the uterine fundus shows the fetal head (arrow). (f) Transabdominal US image (four-chamber view) of a supine fetus. *Dao* = descending aorta, *SP* = spine. (g) Transabdominal US image (base view) of a supine fetus shows the right pulmonary artery (*RPA*) crossing over the ascending aorta (\*). *DA* = ductus arteriosus, *PA* = main pulmonary artery.



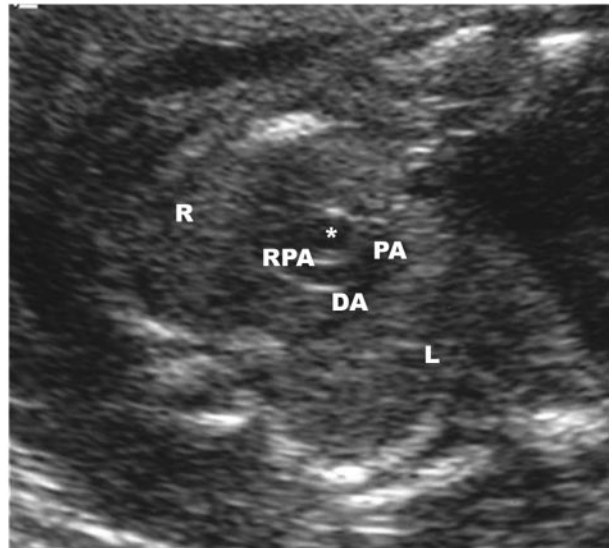
d.



e.



f.

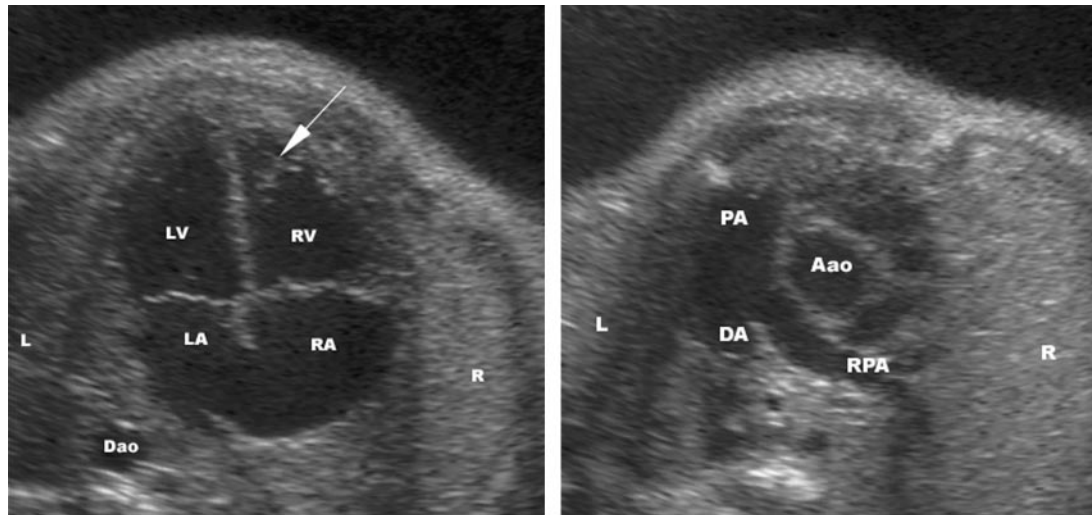


g.

**Normal Views**

The four chambers of the heart are delineated by the ventricular septum, the atrial septum, the tricuspid valve, and the mitral valve. The axis of the ventricular septum is directed to the left and forms an angle of 45°–50° with the midsagittal

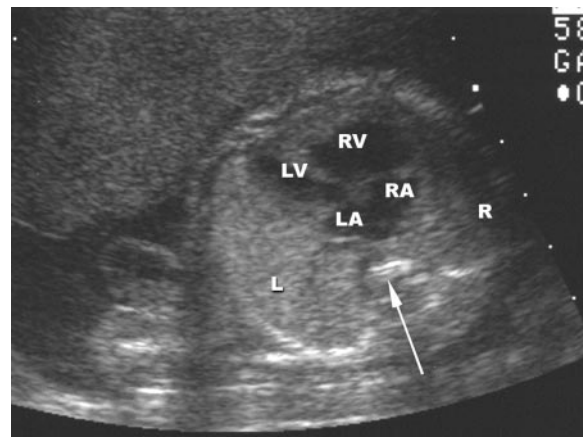
plane. The cardiac circumference is approximately one-third to one-half of the thoracic circumference throughout gestation. The foramen ovale interrupts the interatrial septum, with the flap of the foramen always opening toward the left



**Figure 3.** Normal views. **(a)** Transabdominal US image (four-chamber view) shows the moderator band (arrow) in the right ventricle, the left ventricle, the right atrium, the left atrium, and the descending aorta (*Dao*), which is anterior to the echogenic spine. **(b)** Transabdominal US image (base view) shows the main pulmonary artery (*PA*), the right pulmonary artery (*RPA*) draping over the ascending aorta (*Aao*), and the ductus arteriosus (*DA*), which connects to the descending aorta.

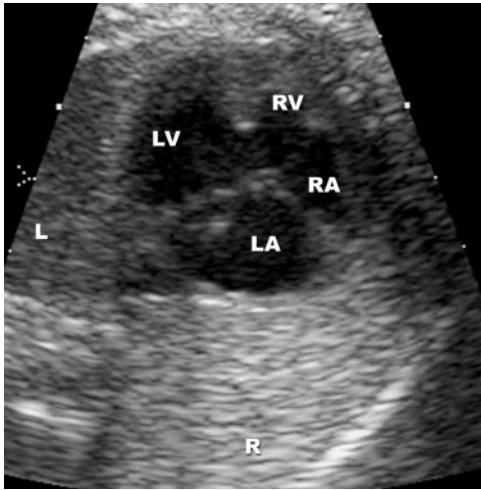
atrium. The right ventricle is located behind the sternum and is characterized by the presence of the moderator band (3,6). Normally, both ventricles are approximately the same size. The left ventricle is posterior and to the left of the right ventricle, and the mitral valve insertion is slightly cephalad to the insertion of the tricuspid valve (3,6). These features help distinguish the right ventricle from the left ventricle (Fig 3a).

The base view of the heart demonstrates the normal crossing of the great vessels just above the four-chamber view. The pulmonary artery arises from the right ventricle, proceeds across the midline, and passes anterior to the ascending aorta. As the pulmonary artery bifurcates, the right pulmonary artery loops around the circular ascending aorta (see Movie 1 at [radiographics.rsna.org/cgi/content/full/22/5/1125/DC1](http://radiographics.rsna.org/cgi/content/full/22/5/1125/DC1)). At this plane of bifurcation, the left pulmonary artery is not imaged. Instead, the ductus arteriosus forms the other limb of the bifurcation, connecting with the de-



**Figure 4.** Hypoplastic left heart syndrome in a fetus with a cephalic presentation. Transabdominal US image (four-chamber view) shows that the left ventricle is small relative to the right ventricle and the left atrium is small relative to the right atrium. Arrow = spine.

scending aorta (Fig 3b). The pitfalls associated with this view lie primarily in the inability to obtain an optimum plane of scanning or failure to maintain the proper orientation as the fetus



**Figure 5.** Endocardial cushion defect in a fetus with a cephalic presentation. Transabdominal US image (four-chamber view) shows absence of the interventricular and interatrial septa, thus producing connections between the ventricles and between the atria.

changes position. False-negative and false-positive diagnoses arise when both the aorta and the pulmonary artery are not seen in the same image. Assumptions should not be made as to the presence or normalcy of the vessel not imaged unless it is directly viewed. Also, constant reorientation of the transducer should be achieved by checking the position of the fetal head when a change in fetal position is suspected.

### Hypoplastic Left Heart Syndrome

Hypoplastic left heart syndrome is a spectrum of heart malformations that consists of a small left ventricle, which is associated with aortic atresia and an atretic or hypoplastic mitral valve. The prevalence of hypoplastic left heart syndrome ranges from 0.16 to 0.25 per 1,000 live births, which represents 2%–4% of congenital heart defects (4,7,8). The early developmental changes that lead to hypoplastic left heart syndrome are

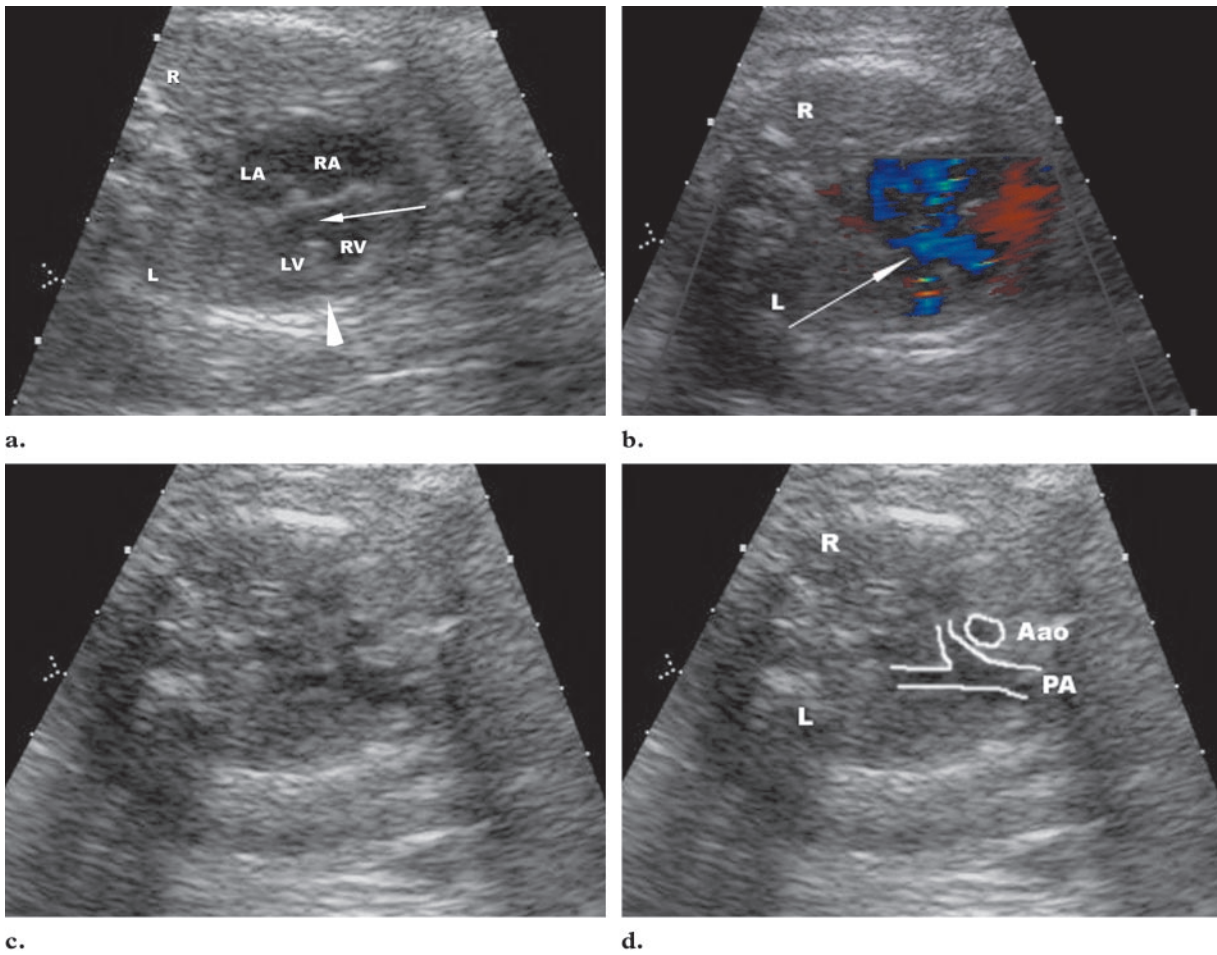
unknown. However, the development of the cardiac chambers is dependent on the flow of blood through the chambers, not the pressure within the chambers (7). Therefore, it is hypothesized that decreased flow in the left ventricle causes hypoplastic left heart syndrome. In making the diagnosis, the four-chamber view is usually sufficient to demonstrate the abnormalities (Fig 4). However, the base view may be helpful in documenting the disproportionately smaller aorta in comparison to the pulmonary artery.

### Endocardial Cushion Defect

Endocardial cushion defects occur in approximately 0.19–0.56 per 1,000 live births, which represents 2%–7% of congenital heart defects (4,9). Formation of the atrioventricular canal is via growth of the endocardial cushions and proliferation of the dextrodorsal conus cushion (10). When the endocardial cushions fail to fuse, a wide range of atrioventricular septal defects occur. The complete form of endocardial cushion defect consists of a large defect involving the inferior portion of the atrial septum and the posterior portion of the ventricular septum (9,10). There is one common atrioventricular valve instead of two separate valves. Invariably, there is free regurgitation at the atrioventricular valve level. An endocardial cushion defect can be accurately diagnosed by using only the four-chamber view (Fig 5). In most instances, the base view cannot be obtained.

### Ventricular Septal Defect

The interventricular septum separates the right ventricle from the left ventricle and is composed of both muscular and membranous tissue. A normal interventricular septum extends from the cardiac apex to the atrial septum (9). Formation of the interventricular septum begins at approximately 28 days gestation when the median muscular ridge begins to invaginate. The free edge of



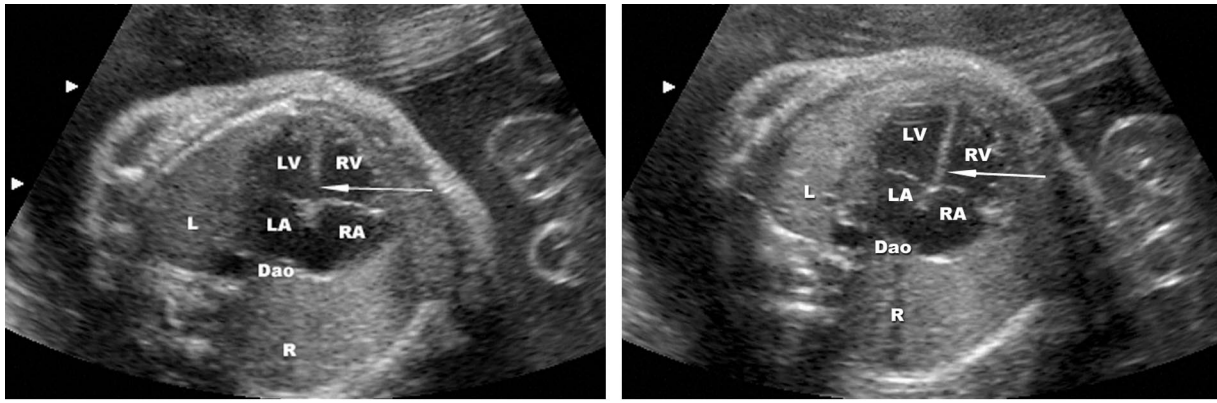
**Figure 6.** VSD in a fetus with a breech presentation. **(a)** Transabdominal US image (four-chamber view) shows leftward deviation of the axis with the apex (arrowhead) shifted to the left. A large VSD (arrow) is also present, which connects the left ventricle to the right ventricle. **(b)** Color Doppler image (four-chamber view) shows free flow between the ventricles through a large VSD (arrow). **(c)** Transabdominal US image (base view) shows a normal anatomic relationship between the aorta and the pulmonary artery (see **[d]** for a diagram). **(d)** Transabdominal US image (base view) shows normal crossing of the pulmonary artery (*PA*) over the ascending aorta (*Aao*).

the muscular septum fuses with the membranous septum formed by the endocardial cushions at approximately 49 days gestation (11). A ventricular septal defect (VSD) results from maldevelopment of the embryonic muscular septum, maldevelopment of the endocardial cushions, or excess resorption of myocardial tissue in the muscular septum (11).

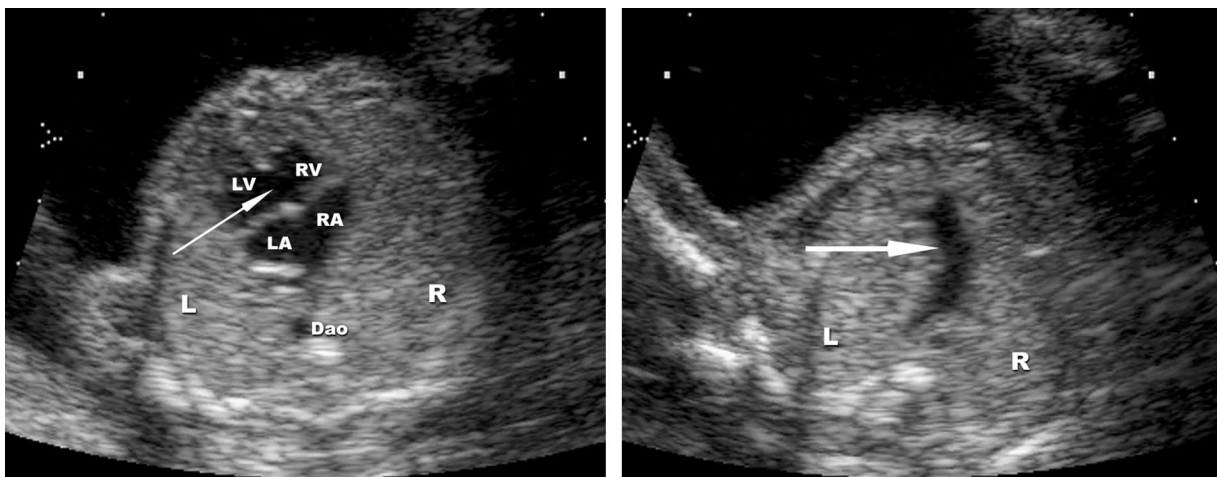
VSD is one of the most common cardiac anomalies, accounting for 20%–40% of congenital heart defects (9). The prevalence of VSD ranges from 1.5 to 3.5 per 1,000 live births

(9,12). A VSD results in an abnormal hemodynamic communication between the right and left ventricles, causing a left-to-right shunt. A large VSD is easily diagnosed on the four-chamber view alone. However, color Doppler US may be needed to demonstrate smaller defects (Fig 6), and some may not be detected until after birth. Fetal position is a major factor in the detection of VSD. For accurate diagnosis, it is critical to approximate the ideal 90° insonation of the interventricular septum. When the ultrasound beam is directly at 0° relative to the septum, a pseudo-VSD may result. This is easily resolved by changing the angle of insonation (Fig 7).





**Figure 7.** Pseudo-VSD in a fetus with a cephalic presentation. *Dao* = descending aorta. **(a)** Transabdominal US image (four-chamber view) shows a pseudo-VSD (arrow), which is caused by insonation of the interventricular septum by the ultrasound beam at a parallel angle. **(b)** Four-chamber view obtained in the same patient with a better angle of insonation shows an intact ventricular septum (arrow).



**Figure 8.** Persistent truncus arteriosus in a fetus with a cephalic presentation. **(a)** Transabdominal US image (four-chamber view) shows a VSD (arrow). *Dao* = descending aorta. **(b)** Transabdominal US image (base view) shows a single trunk (arrow) overriding both ventricles.

### Persistent Truncus Arteriosus

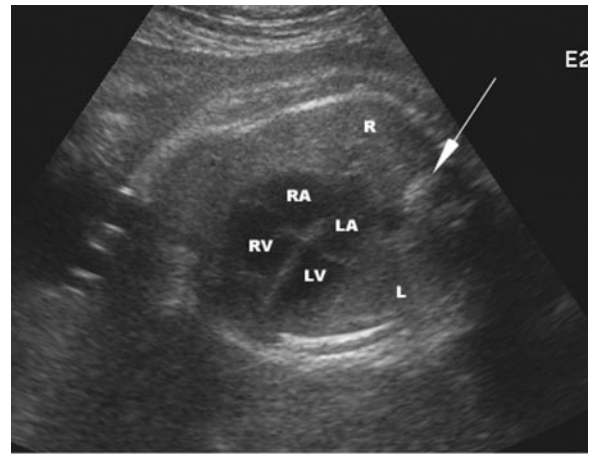
Persistent truncus arteriosus accounts for approximately 1%–2% of congenital heart defects, occurring in approximately 0.08–0.16 per 1,000 live births (13). A persistent truncus arteriosus develops when the truncoconal ridges fail to fuse and descend toward the aortic sac (4,13,14). It is characterized by a single overriding arterial trunk that feeds both the aorta and the pulmonary artery. The undivided truncus receives blood from both ventricles. A VSD is almost always present (4,13).

During embryonic formation, the truncus arteriosus is created by fusion of two opposing ridges

in the cephalic portion of the truncus, which grow caudally and in the direction of the aortic sac. The ridges twist around each other, dividing the truncus into an anterior pulmonary section and a posterior aortic section (15).

This diagnosis may not be apparent on the four-chamber view alone. However, several attempts at obtaining a base view will fail to reveal normal crossing of the great vessels. Instead, a single vessel is seen with several branches connecting with the pulmonary vessels and aorta (Fig 8).

**Figure 9.** Complete transposition of the great arteries in a fetus with a cephalic presentation. **(a)** Transabdominal US image (four-chamber view) shows a normal appearance of the four chambers. Arrow = spine. **(b)** Transabdominal US image (base view) shows D-transposition of the great arteries (see **[c]** for a diagram). **(c)** Transabdominal US image (base view) shows D-transposition of the great arteries. The aorta (*Ao*) arises from the right ventricle, and the pulmonary artery (*PA*) arises from the left ventricle. Also, the pulmonary artery and the aorta are parallel, instead of demonstrating the normal crossing of the pulmonary artery over the aorta.



b.



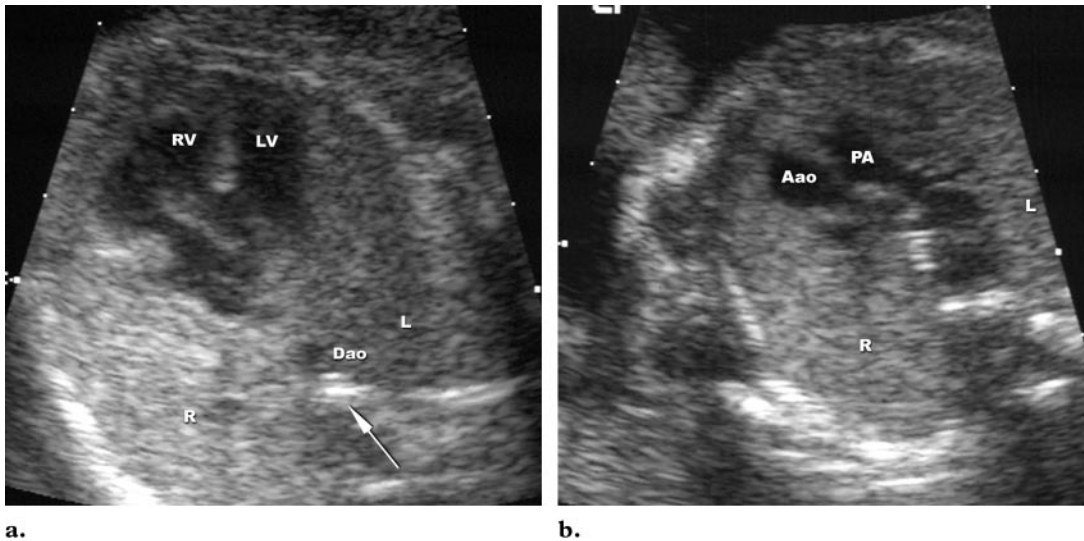
c.

### Complete Transposition of the Great Arteries

Complete transposition of the great arteries occurs in 0.2–0.4 per 1,000 live births, which represents 2.5%–5% of congenital heart defects (16). Normally, the truncus arteriosus divides into a pulmonary artery arising from the right ventricle and an aorta arising from the left ventricle. This occurs by the caudal and spiral growth of the conal truncal ridge, which is usually complete by the end of the 4th week after conception (15). If the septum grows downward without spiraling, the

normal relationship between the ventricles and great arteries is disrupted. This causes the aorta to arise from the right ventricle and the pulmonary artery to arise from the left ventricle. The great arteries are oriented parallel to each other and do not cross (4,16).

In diagnosis of this anomaly, correct orientation is essential. Only when the aorta is seen to arise definitely from the right ventricle and the pulmonary artery is seen to arise definitely from the left ventricle can one be confident of the diagnosis. The base view of the fetal heart is needed to confirm the diagnosis by demonstrating that the great vessels do not cross (Fig 9; see also Movie 2 at [radiographics.rsna.org/cgi/content/full/22/5/1125/DC1](http://radiographics.rsna.org/cgi/content/full/22/5/1125/DC1)).



**Figure 10.** Double-outlet right ventricle in a fetus with a breech presentation. **(a)** Transabdominal US image (four-chamber view) shows a normal appearance of the ventricles. Arrow = spine, *Dao* = descending aorta. **(b)** Transabdominal US image (base view) shows that the ascending aorta (*Aao*) and pulmonary artery (*PA*) are parallel and both arise from the right ventricle.

### Double-Outlet Right Ventricle

Double-outlet right ventricle is a spectrum of cardiac lesions that occur when both great arteries arise partially or completely from the right ventricle. A VSD is almost always present (17).

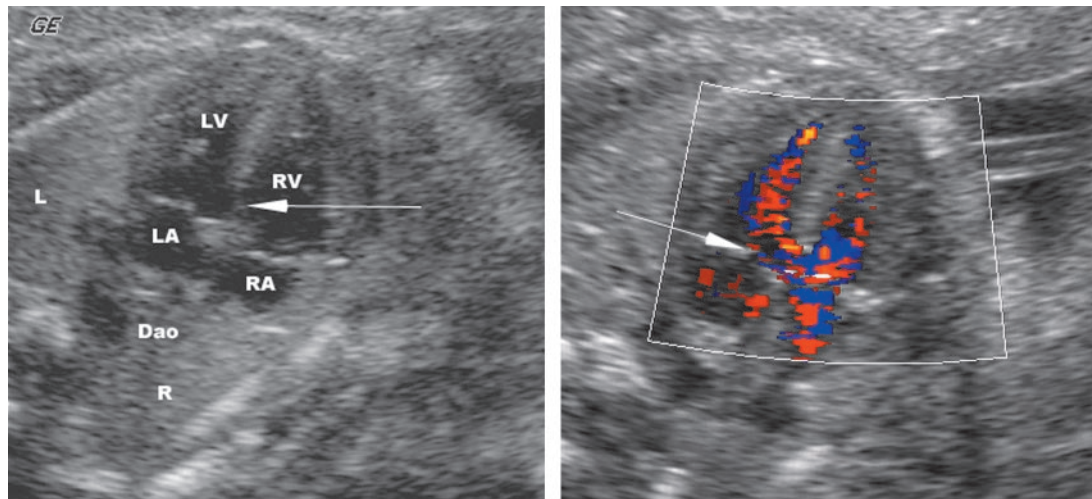
Double-outlet right ventricle occurs in 0.08–0.16 per 1,000 live births, which represents 1%–2% of congenital heart defects (4). A double-outlet right ventricle results from several different defects caused by abnormal spiraling of the truncus arteriosus and developmental arrest of membranous interventricular septum formation (17).

A base view of the fetal heart is required to make the diagnosis of double-outlet right ventricle. The four-chamber view may be proportionate in appearance, but the great vessels are not seen to cross at the base. Instead, both vessels are seen to arise from the right ventricle and assume a parallel course. Double-outlet right ventricle needs to be differentiated from transposition of the great vessels by showing that both vessels arise on the same side of the ventricular septum (Fig 10).

### Tetralogy of Fallot

Tetralogy of Fallot has four classic features: a VSD, an overriding aorta, pulmonary artery stenosis, and right ventricular hypertrophy. Owing to the shunts that exist in the fetal circulation, the right ventricular hypertrophy may not be seen in utero (18). The VSD associated with tetralogy of Fallot is usually subaortic and attributed to the overriding aorta. The prevalence of tetralogy of Fallot is approximately 0.24–0.56 per 1,000 live births, which represents approximately 3%–7% of congenital heart defects (4).

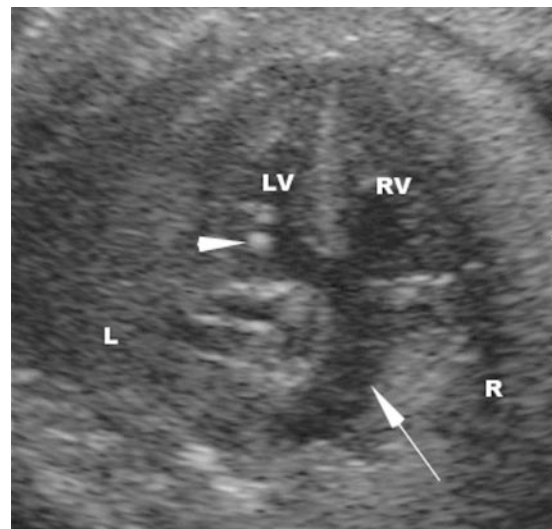
Tetralogy of Fallot is caused by unequal division of the conus resulting from anterior displacement of the truncoconal system. This divides the conus into a smaller anterior right ventricular portion and a larger posterior left ventricular portion, causing the overriding aorta and pulmonary artery stenosis (15).



a.

**Figure 11.** Tetralogy of Fallot in a fetus with a cephalic presentation. **(a)** Transabdominal US image (four-chamber view) shows a VSD (arrow). *Dao* = descending aorta. **(b)** Color Doppler image (four-chamber view) shows free flow through the VSD (arrow). **(c)** Transabdominal US image (attempted base view) shows the aorta (arrow) projecting over the interventricular septum. An echogenic intracardiac focus (arrowhead) is seen in the left ventricle. The normal orientation of the base view could not be achieved, and this result suggested the diagnosis of tetralogy of Fallot.

b.



c.

The diagnosis of tetralogy of Fallot is suspected when a large VSD leads into a great vessel that straddles the interventricular septum. The pulmonary artery may not be easily demonstrated, since the predominant feature of the anomaly is usually the overriding aorta. Again, the main reason for suspecting this anomaly is failure to demonstrate the normal crossing of the great vessels at the base of the heart (Fig 11; see also Movie 3 at [radiographics.rsna.org/cgi/content/full/22/5/1125/DC1](http://radiographics.rsna.org/cgi/content/full/22/5/1125/DC1)).

### Conclusions

Evaluation of the fetal heart is an essential part of a routine prenatal US examination. Adequate visualization of fetal cardiac anatomy is dependent on many factors, such as the ability and experience of the sonographer, maternal body habitus, fetal activity, and fetal position. In cases where the fetus is in a suboptimal position, adequate four-chamber and base views of the heart

may not be obtained during the initial examination. The patient should then be rescanned at another time when the fetus may be in a position to allow better imaging. If any abnormalities are suspected, the patient should be referred to a specialist for a full cardiac evaluation.

A four-chamber view of the heart satisfies the minimum requirement of the guidelines of both the American College of Radiology and the American Institute of Ultrasound in Medicine. However, anomalies involving the outflow tracts of the aorta and pulmonary artery are not visualized on this view. By adding the base view of the heart, the outflow tracts can be visualized, thus increasing not only the sensitivity of detection of cardiac anomalies but also the accuracy of diagnosis.

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