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Review

Introduction to fetal echocardiography

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ABSTRACT

Congenital heart disease is the most common congenital malformation and has significant impact on postnatal outcomes. Prenatal diagnosis is associated with improved infant morbidity and mortality. Advancements in fetal echocardiography and robust screening practices have significantly increased accuracy of prenatal diagnosis of congenital heart disease and other cardiovascular pathologies. Therefore, it is very important to develop a comprehensive fetal echocardiographic screening protocol as part of the prenatal cardiology assessment. This review is geared towards the novice perinatal cardiologist or fetal sonographer. We will highlight indications for fetal echocardiography, guide the reader through a basic screening fetal echocardiogram utilizing the segmental approach, and discuss delivery planning for various cardiac disorders, incorporating practices from our institution.

1. Introduction

Congenital heart defects are the most common birth defects and occur in approximately 1 in 100 births in the United States [1,2]. Congenital heart disease is a leading cause of perinatal demise as well as neonatal morbidity and mortality [3]. The heart is one of the first organs to differentiate in the fetus. At approximately 3 weeks gestation, the primitive heart tube forms. Over the next several weeks, the heart tube undergoes looping, chamber septation, outlet septation, and finally connects into the paired aortic arch system. The mature heart is formed by 8 weeks gestation [4]. Transabdominal ultrasound can be performed reliably after 12 weeks gestation and congenital heart defects can be consistently identified as early as 14 weeks [4]. Fetal echocardiography plays a pivotal role in the detection of congenital heart disease and is typically performed in the second trimester between 18 and 22 weeks gestation [5]. Advances in fetal echocardiography over the last few decades have seen progression from 2-dimensional, 3 and 4-dimensional echocardiography and are now delving into fetal intelligent navigation echocardiography.

The concept of fetal cardiac echocardiography originated in the 1960s. Following the development of M-mode echocardiography in 1954 by Edler and Hertz, Wang reported first use in the fetal heart in 1964 [6,7]. Over the next 15 years, B-mode (or 2-dimensional echocardiography) was developed and multiple studies were published in 1980 documenting use of this modality on the fetal heart [8–10]. During this decade, ultrasound was used to identify fetuses with congenital heart defects and arrhythmias [2]. Allan et al. performed 2-dimensional fetal echocardiography on 200 fetuses and compared their findings to autopsy specimens in order to optimize views for fetal cardiac scanning [9]. Spectral Doppler followed by color Doppler was introduced in fetal echocardiography in the 1980s. In the early 2000s, 3-dimensional echocardiography was applied to fetal cardiac scanning, allowing for more robust imaging of structural cardiac lesions and flow hemodynamics. More recently, volumetric sonography with 4-dimensional scanning has been used to further improve our ability to identify complex intracardiac malformations and can shorten scan times due to full volume acquisitions. However, this modality may be limited by the operator's ability to process the volumetric data. Fetal intelligent navigation echocardiography is a novel approach that can automatically display the nine standard fetal echocardiography views after a 4-dimensional volume has been acquired. This technology remains experimental but holds significant promise for the future of perinatal cardiac screening. Today, fetal echocardiography may detect approximately 87% of major forms of congenital heart disease, but has limitations on

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Table 1
Common indications for fetal echocardiogram.
Adapted and modified from Donofrio MT, Moon-Grady AJ, Hornberger LK, et al. Diagnosis and treatment of fetal cardiac disease, a scientific statement from the American Heart Association. Circulation 2014;129:2183–2242

Maternal indications	Fetal indications
Pregestational diabetes mellitus Gestational diabetes diagnosed in first trimester and/or hemoglobin A1C > 6%	Abnormal obstetrical ultrasound with suspected congenital cardiac or extracardiac anomaly
1st or 2nd degree relative with congenital heart defect	Increased nuchal thickness
1st or 2nd degree relative with genetic syndrome associated with congenital heart defects	Abnormal genetic testing
Maternal lupus or Sjogren's syndrome with SSA/SSB+	Arrhythmia
Medications during pregnancy: • ACE inhibitor • Retinoic acid • NSAIDs • Anticonvulsants • Lithium • Vitamin A • SSRI	Monochorionic twinning, increased risk associated with twin-twin transfusion syndrome.
Assisted reproduction technologies	Placental, umbilical cord or intra-abdominal venous abnormality
Poorly controlled phenylketonuria	Tricuspid regurgitation
Consanguinity	Hydrops
Maternal Rubella infection or other infection associated with suspected fetal myocarditis	Aberrant right subclavian artery

minor forms of congenital heart disease [11]. Accurate prenatal diagnosis of congenital heart disease allows fetal cardiologists to assist with prenatal management in collaboration with maternal fetal medicine, determine need for fetal intervention, and optimize postnatal delivery planning.

In this paper, we will discuss the indications for fetal echocardiography, review the basics of performing the initial screening fetal echocardiogram, and provide delivery planning guidelines. This review is directed towards the novice perinatal cardiologist and fetal sonographer. Imaging of abnormal fetal hearts, advanced fetal cardiac imaging modalities, and fetal cardiac interventions are outside of the scope of this article. Readers are encouraged to see complete guidelines for a fetal echocardiogram by the American Heart Association, American Society of Echocardiography, American Institute of Ultrasound in Medicine, and International Society of Ultrasound in Obstetrics and Gynecology [5,12–14].

2. Indications

There are several maternal, familial, and pregnancy risk factors associated with increased incidence of congenital heart disease that guide referrals for a fetal echocardiogram. However, most fetuses that will be diagnosed with congenital heart disease are first identified for referral based on abnormal obstetrical ultrasound alone [13]. Based on elevated level of risk for fetal congenital heart disease; commonly accepted indications for fetal echocardiogram have been developed and included in multiple guidelines. Table 1 summarizes common maternal and fetal indications for fetal echocardiogram that are associated with presence of congenital heart disease in more than 1% of live births [4,7,12].

3. Preparation for fetal echocardiogram

3.1. Scheduling

As previously stated, the initial fetal echocardiogram is typically performed at age 18–22 weeks though may be performed earlier or later depending on indication and family planning. A perinatal nurse coordinator is an essential part of the perinatal program and may be the first contact a family has with your program. A perinatal nurse coordinator is important for obtaining all obstetrical and maternal fetal medicine records for review, scheduling timing of the visit based on institutional protocol and medical necessity, giving the patient and family a point person for questions or concerns, communication of results back to the referring provider, and coordinating delivery plans for those patients with complex congenital heart disease.

3.2. Technical aspects of the perinatal cardiology lab

Traditionally, fetal echocardiograms may be performed in one of two different perinatal lab designs:

- fetal echocardiograms are performed within a women's or fetal care center along with maternal fetal medicine and/or obstetrical services.
- fetal echocardiograms are performed within a pediatric cardiology office that is separate from maternal fetal medicine and/or obstetrical services.

Each option has inherent advantages. The fetal care center model places the pregnant mother as the center point and brings all providers to her, including obstetrical, maternal fetal medicine, perinatal cardiology with fetal echocardiography, genetic counseling, and consultation with surgical or pediatric sub-specialists. This model requires all members of the care team to come to a common clinical space and perform perinatal counseling. This works extremely well when practitioners are all employed by a single entity such as a hospital or university, but may be challenging in other scenarios. Alternatively, fetal echocardiograms can be performed within a pediatric cardiology clinic that is separate from the obstetrical services. This requires the pregnant mother to travel to multiple clinic locations (obstetrics, maternal fetal medicine, perinatal cardiology, and cardiovascular surgical center, if indicated). In either scenario, perinatal cardiology clinics require an appropriately sized and equipped room for scanning, a physician workspace for viewing images and performing documentation, a nurse coordinator workspace, and a family counseling area. Whenever possible, try to keep pregnant women separate from pediatric patients by using separate waiting rooms, scanning pregnant women in a separate clinic location or scheduling at alternative times.

Another important aspect of the perinatal cardiology clinic is the ultrasound equipment. The ultrasound machine must have capability to perform two-dimensional imaging, M-mode, pulse-wave Doppler, and color Doppler. Three and four-dimensional imaging probes along with post-processing software may also be beneficial. Probes used include

sector and curvilinear styles with frequencies between 1 and 9 MHz depending on the mother's body habitus. The highest possible transducer frequency should be used. When performing a fetal echocardiogram, the sonographer should optimize their system settings to maintain high frame rate, narrow the field of view, use the shallowest imaging depth possible, utilize a single focal zone, use low persistence and narrow the color Doppler box. The heart should fill one-third to one-half of the screen. Limit fetal exposure to Doppler and harmonics by following the ALARA principle (As Low As Reasonably Achievable), including limiting acoustic output, performing exams for appropriate indications, and avoiding unnecessarily prolonged scanning [5,14].

4. Performing a complete fetal echocardiogram

Table 2 summaries the essential components of the initial complete fetal echocardiogram adapted from current guidelines [5,12–14].

Table 2
Components of a complete fetal echocardiogram.

Segmental approach	Color Doppler	Pulsed Doppler
Establish fetal number and fetal lie	Systemic Veins	Atrioventricular valves
Visceral situs		Semilunar valves
	• Superior and inferior vena cava	Pulmonary veins
• Side of liver, stomach, descending aorta and inferior vena cava	• Ductus venosus	Ductus arteriosus
• Cardiac apex position and axis	Pulmonary veins	Systemic veins
Atria		Aortic isthmus
	• At least one right and one left	Branch pulmonary arteries
• Situs	Atrial septum and foramen ovale	Middle cerebral artery
• Septal anatomy	Atrioventricular valves	Umbilical artery and vein
• Systemic venous connections	Ventricular septum	Ductus venosus
• Pulmonary venous connections	Semilunar valves	Abnormal structure or flow suspected
Atrioventricular valves	Ductal arch	
	Aortic arch	
• Anatomy, size and function		
Ventricles		
• Position		
• Atrioventricular connections		
• Morphology		
• Size		
• Qualitative systolic function		
• Septal anatomy		
• Pericardium		
Semilunar valves		
• Anatomy, size and function		
• Outflow tracts		
Great arteries		
• Ventricular connections		
• Size, flow and patency		
• Pulmonary artery bifurcation		
Ductus arteriosus		
2-Dimensional Imaging	Cardiac measurements	Other measurements
Four-chamber view	Aortic and pulmonary valve annulus diameters in systole	Biometry
Left ventricular outflow tract		
Right ventricular outflow tract	Mitral and tricuspid valve annulus diameters in diastole	• Cardiothoracic ratio
Branch pulmonary artery bifurcation		• Femur length
Three-vessel views	Right and left ventricular widths and lengths	• Biparietal diameter
	Thicknesses of ventricular free walls and interventricular septum	• Head circumference
• with pulmonary artery bifurcation		• Abdominal circumference
• trachea view with ductal arch	Aortic transverse arch and isthmus diameters	
Short axis views	Main and branch pulmonary artery diameters	Heart rate and rhythm
	Ductus arteriosus diameter	
• ventricles	Transverse atrial dimensions	• Doppler or M-mode technique
• outflow tracts		• Detailed assessment of atrial and ventricular contractions if arrhythmia detected
Long axis view		
Aortic arch		
Ductal arch		Quantitative ventricular function assessment if indicated

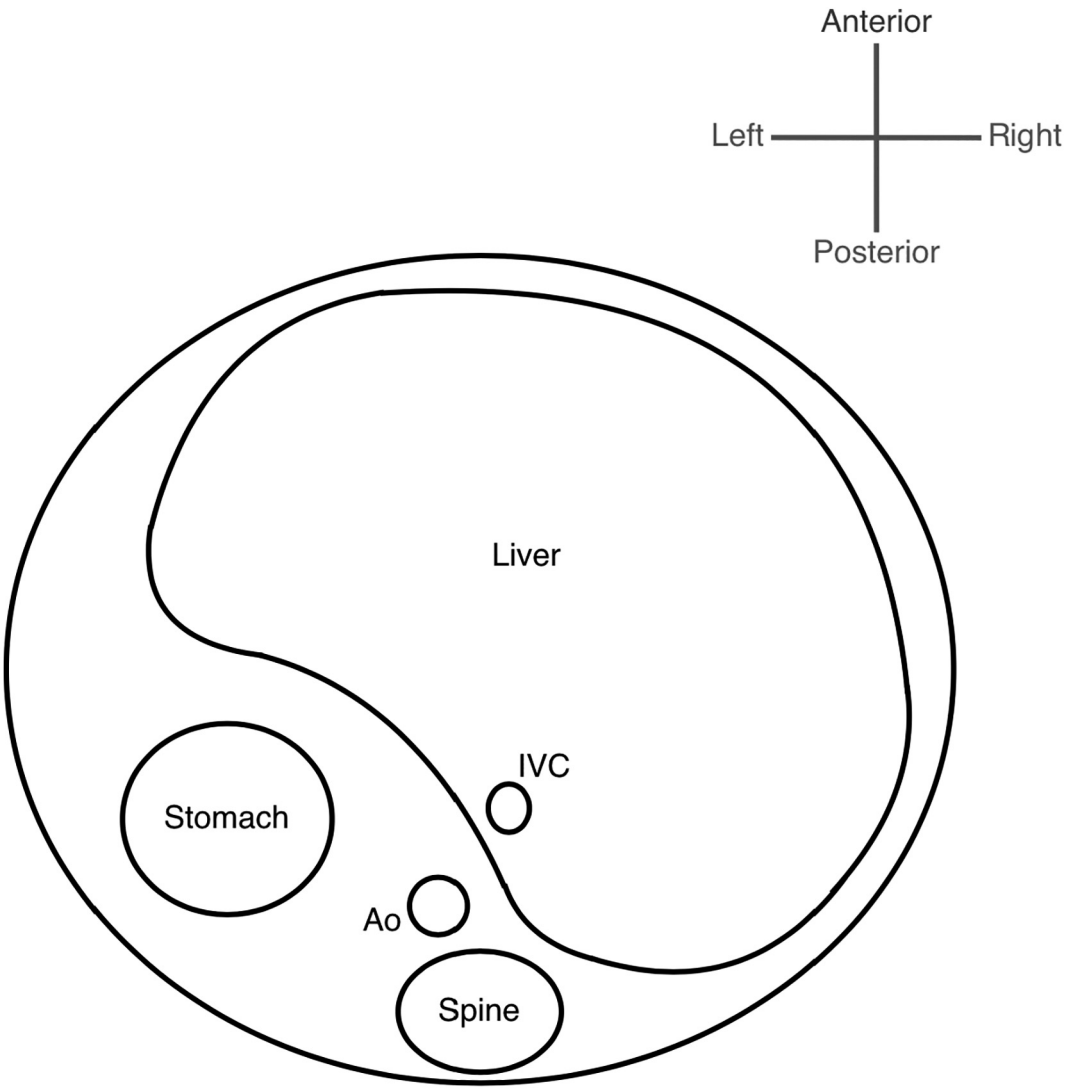

Fig. 1. Diagram of transverse view of the fetal abdomen at level of the stomach demonstrating visceral situs solitus. Ao, aorta; IVC, inferior vena cava.

4.1. Establishing situs

First, the sonographer should perform a sweep of mother's abdomen to determine number of fetuses present and what position they are in, vertex, breech, or transverse. In our institution, we perform an inferior to superior sweep of mother's abdomen with transducer indicator to the sonographer's right (maternal left). Based on the position of the fetal spine, the right and left side of the fetus can be determined. For instance, if the fetus is breech (head up) with spine to maternal left, then the left side of the fetus will be on the right side of the screen. If the fetus is cephalic with spine to maternal left then the fetal left side will be on the left side of the screen. This will allow determination of visceral situs and in which direction the cardiac apex is pointing. An alternative method, which may be particularly helpful in transverse lie, begins with positioning baby horizontally on the screen with head to the right. It is ok to flip the screen in this instance. The probe is then rotated 90° clockwise to obtain a transverse plane through the fetal chest. If the reviewer's left hand is placed over the image with the palm over the fetal spine and fingers pointing to the fetal sternum, the thumb will point to the left side of the fetus [15]. The situs sweep should be performed through transverse sections of the fetal abdomen and chest to determine visceral situs solitus, cardiac apex direction, and axis of the heart (Figs. 1 and 2). Normal visceral situs solitus is defined by a right sided liver and left sided stomach. The aorta should be located on the left side of the spine and the inferior vena cava on the right. The heart should sit primarily in the left thorax with apex pointing left. The normal position is called levocardia. A normal cardiac apex is 45 ± 20° from midline. Abnormalities of cardiac axis are associated with increased risk of cardiac anomalies [14].

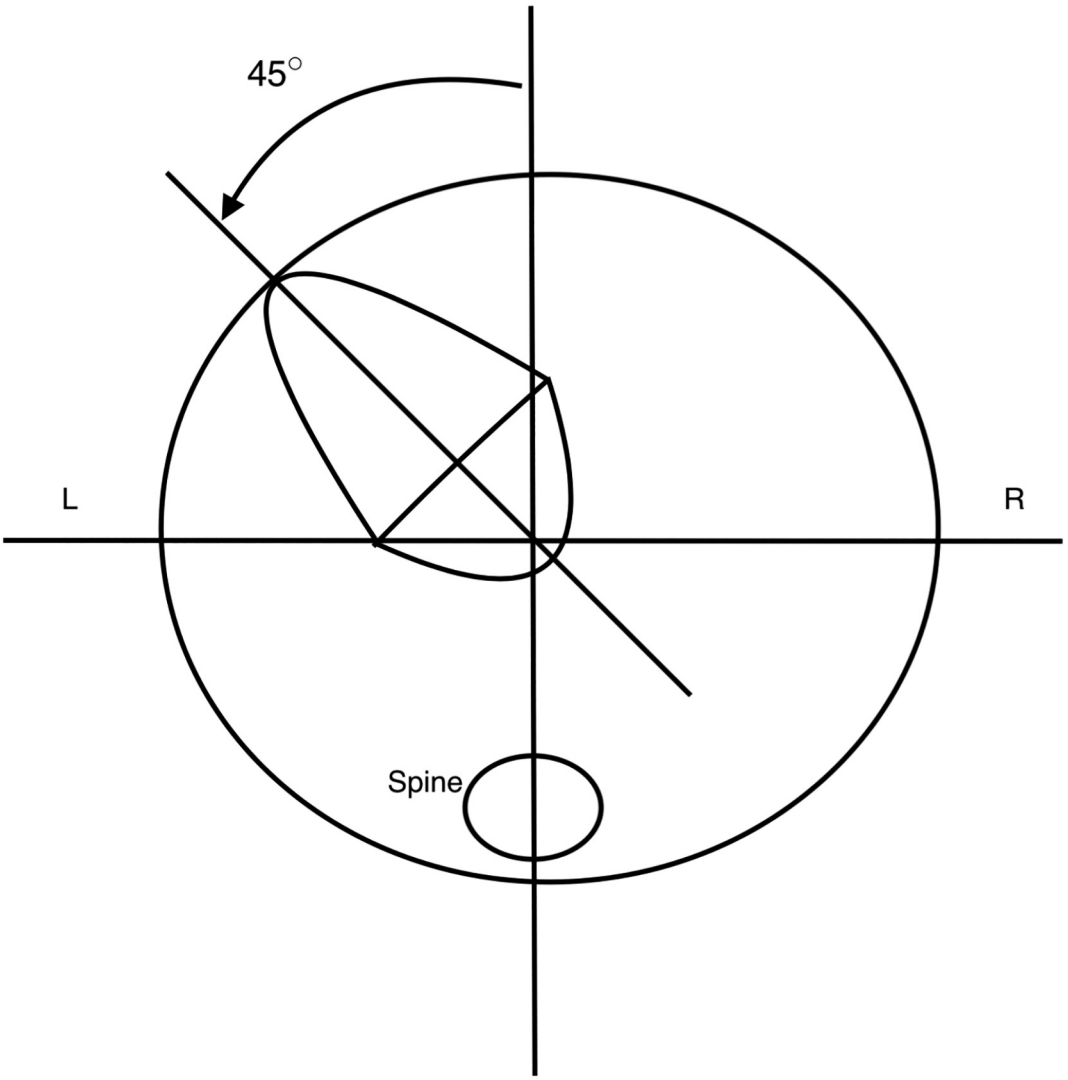

Fig. 2. Diagram of transverse view of the fetal chest demonstrating levocardia and normal cardiac axis. L, left; R, right.

4.2. Fetal biometry

For this portion of the exam, measurements of extracardiac fetal structures and vessels are performed to determine estimated fetal weight, if size of fetus is appropriate for gestational age and evaluate for evidence of fetal distress or hydrops. To obtain biparietal diameter, an axial section of the fetal head is obtained at the level of the cavum septum pellucidum. The diameter is obtained by measuring from inner edge of the fetal skull to outer edge. In this image, the fetal head circumference can also be measured (Fig. 3). In the fetal brain, color, and pulsed wave Doppler interrogation of the middle cerebral artery is performed. The middle cerebral artery is located between the eyes and ears of the fetus coursing towards the Circle of Willis. The Doppler pattern is traced to obtain peak systolic velocity, mean systolic velocity, and end diastolic velocity from which the pulsatility index and systolic/diastolic ratio can be calculated (Fig. 4). There are established normal values by gestational age for comparison [16]. Abnormalities should be noted as elevated peak systolic velocity for gestational age is suggestive of fetal anemia and decreased pulsatility index may be present in fetuses with congenital heart disease.

Cardiothoracic ratio is performed in the transverse plane of the chest at level of the four-chamber view. In the correct plane, one should be able to demonstrate a complete set of ribs on each side of the thorax and no abdominal content should be in the image (Fig. 5). The circumference or area of the thorax and heart, respectively, can be used. Images for measurement are obtained in diastole. The circumference or area of the heart is divided by the circumference or area of the thorax, respectively, to obtain a ratio. A normal cardiothoracic area ratio is

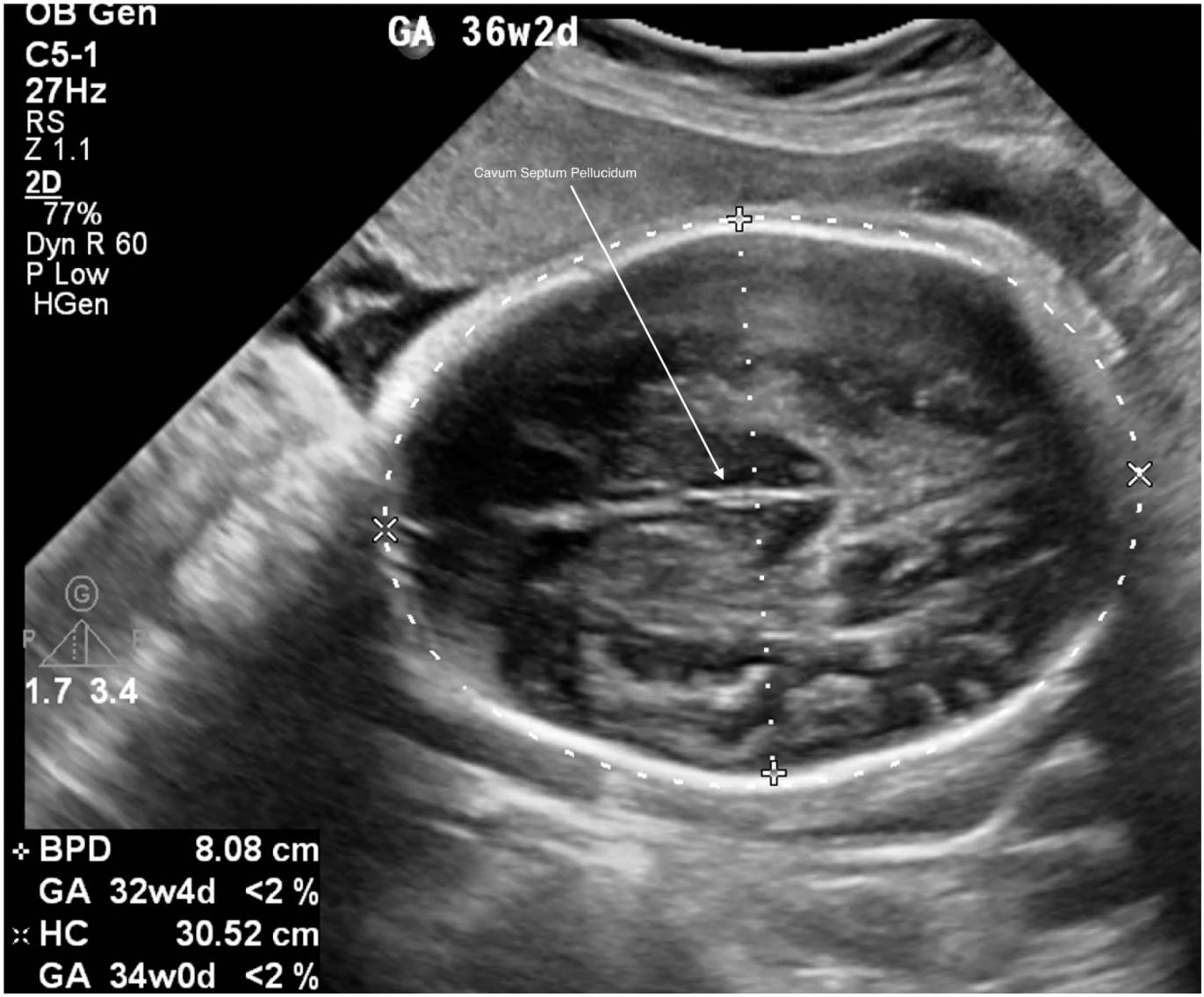

Fig. 3. Fetal cranium. BPD, biparietal diameter; HC, head circumference.

typically between 0.25 and 0.35. An abnormal ratio warrants further investigation. Additional measurements needed to evaluate fetal growth and estimate fetal weight include a femur length and abdominal circumference.

Pulsed wave Doppler interrogation of the umbilical artery, umbilical vein, and ductus venosus should be performed. In the fetal abdomen, the ductus venosus can be located with color Doppler by visualizing a focal area of flow acceleration between the umbilical vein and inferior vena cava. Pulsed-wave Doppler is obtained to determine the systolic/diastolic ratio. The umbilical cord is viewed outside the fetus in short axis and pulsed Doppler of both the umbilical artery and umbilical vein may be obtained in tandem (Fig. 6). For the umbilical artery, the peak systolic velocity, mean systolic velocity and end diastolic velocity are measured to obtain the pulsatility index and systolic/diastolic ratio. It should be noted that low resistance in the ductus venosus and umbilical arteries and presence of pulsatility in the umbilical vein are very abnormal and warrant further investigation.

4.3. Fetal cardiac views

In this section, we will review the different cardiac views obtained in a basic complete fetal echocardiogram and what components should be evaluated and measured for each view. Detailed evaluation of individual congenital heart disease and advanced cardiac functional assessment is outside the scope of this review. A transverse view of the fetal chest is obtained at the level of the atrioventricular valves to image

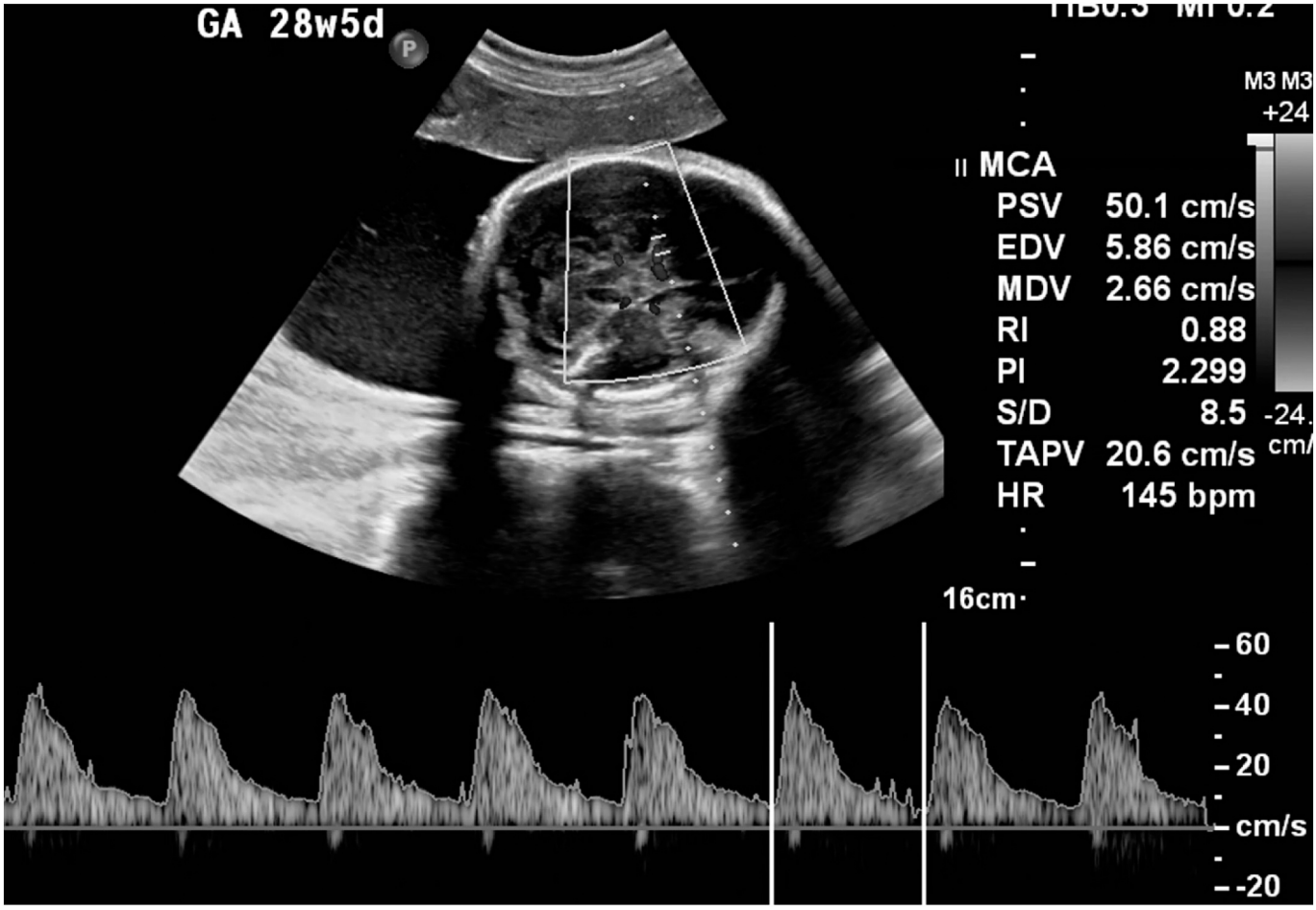

Fig. 4. Normal middle cerebral artery Doppler pattern.

a 4-chamber axial view of the heart. In this view, we can qualitatively assess cardiac chamber sizes, direction of atrial septal bowing, biventricular systolic function, and assess for pericardial disease (Fig. 7). Morphology of the ventricles can be evaluated in multiple ways. The right ventricle will connect to a more apically offset atrioventricular valve. If the atrioventricular valves are not offset, this is suggestive of an endocardial cushion defect. The right ventricle myocardium is more trabeculated and the right ventricle contains a moderator band. This muscular band can be seen near the right ventricular apex in Fig. 7. The left ventricle myocardium is compact and smooth walled and the left ventricle is "bullet-shaped". A normal left sided atrioventricular valve will not have septal attachments. The left ventricle has 2 distinct papillary muscles that are better visualized in a short axis. Identifying the correct ventricular morphology is especially important in diagnosing L-looped or levo-transposition of the great arteries. This disease is an example of incorrect looping resulting in the right atrium draining to a right-sided morphological left ventricle and the left atrium draining to a left-sided morphological right ventricle. Additionally, there is ventricular arterial discordance in this condition.

After evaluating the four chamber view at the level of the atrioventricular valves, sweep towards the fetal head to view outflow tracts and then to the great vessels crossing. This sweep is critical in detecting conotruncal heart defects such as tetralogy of Fallot and D-looped or dextro-transposition of the great arteries. Color Doppler interrogation of the pulmonary veins (Fig. 8), atrial septum, atrioventricular valves, interventricular septum, and semilunar valves is performed in this view. Angling to achieve the interventricular septal position nearly perpendicular to the transducer allows for more accurate evaluation for ventricular septal defects. Pulsed wave Doppler should be obtained through the atrioventricular valves, at least one pulmonary vein from each side and the semilunar valves. Measurements should be taken of atrioventricular valve annulus diameters (Fig. 9) and ventricular chamber lengths and widths. The aortic valve annulus may be measured in this view or in a long axis.

While still in a transverse plane through the chest, sweep towards the fetal head and move probe slightly up the fetal chest to obtain a 3 vessel view in grayscale and with color Doppler. The three vessels from fetal right to left are the superior vena cava, aorta, and pulmonary

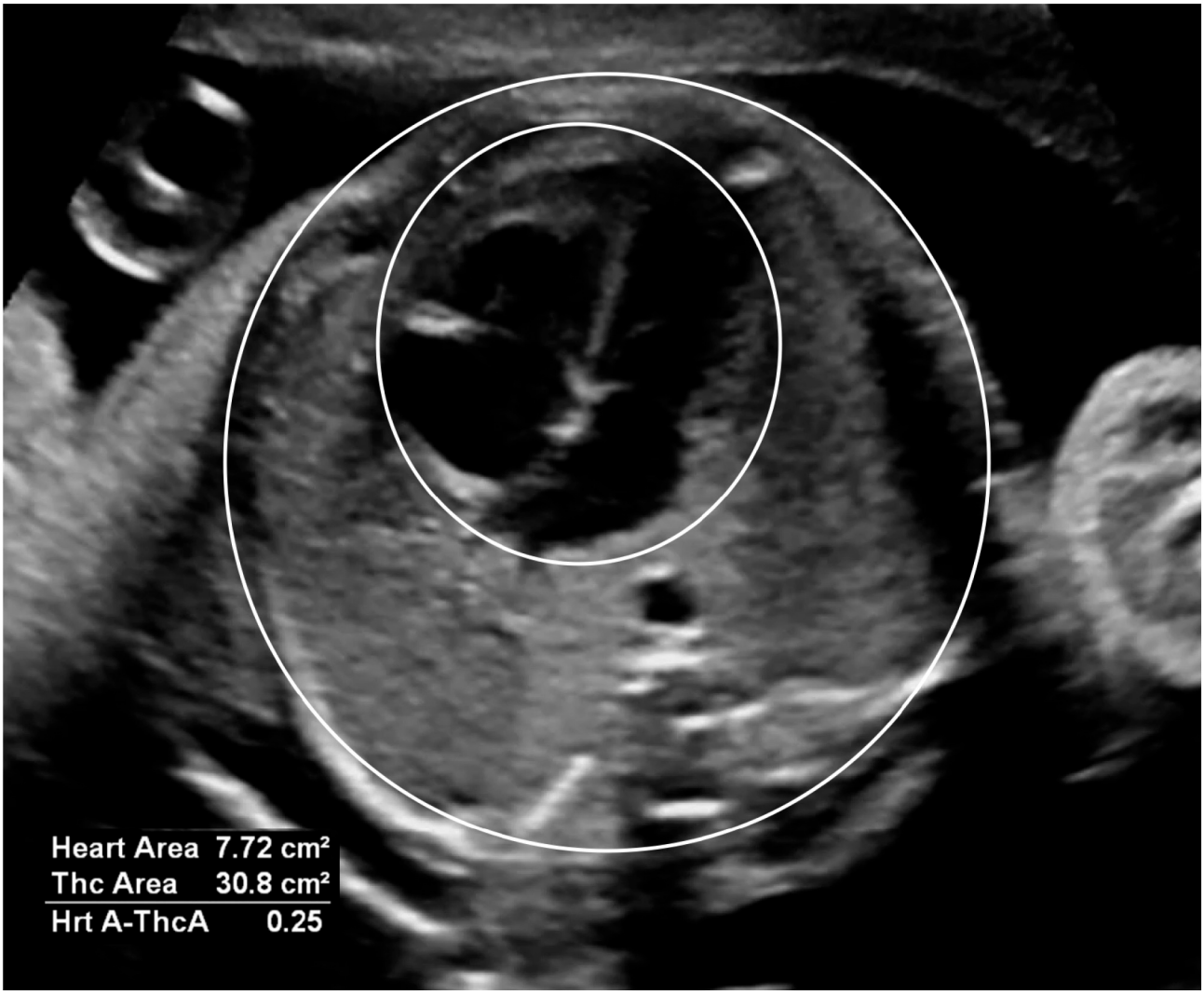

Fig. 5. Cardiothoracic area ratio. Thc, thoracic; Hrt A, heart area; ThcA, thoracic area.

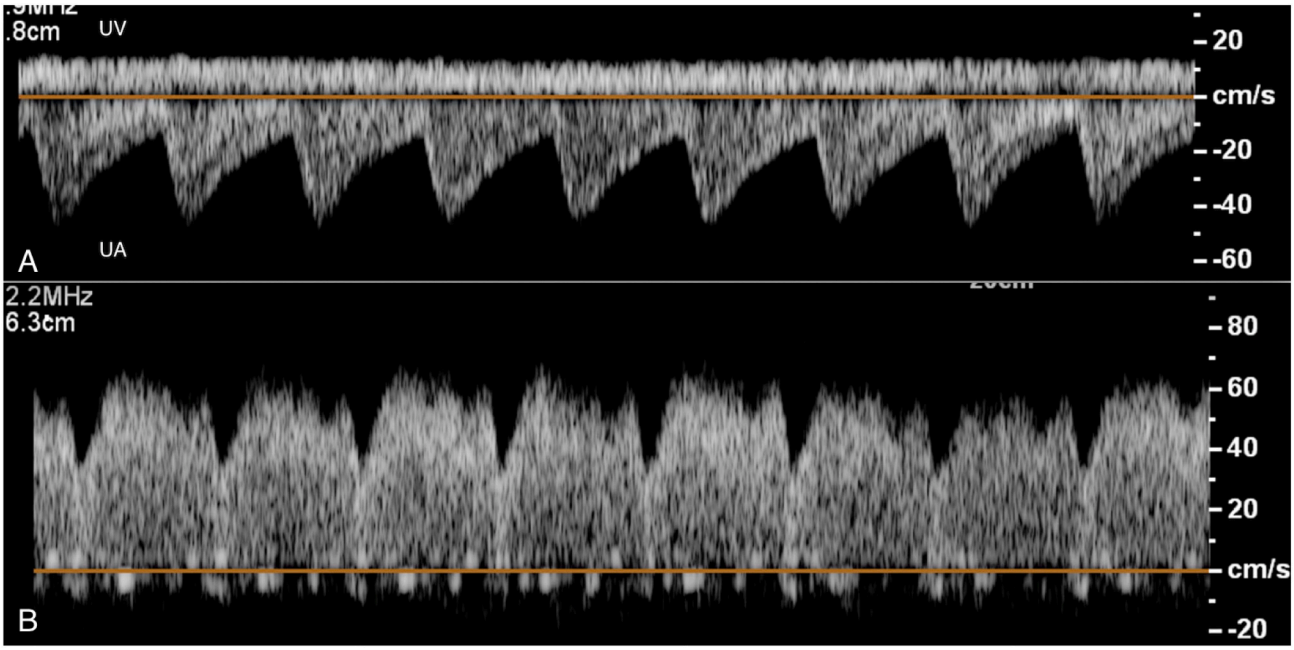

Fig. 6. (A) Normal umbilical vein and umbilical artery Doppler patterns. (B) Ductus venosus Doppler pattern. UV, umbilical vein; UA, umbilical artery.

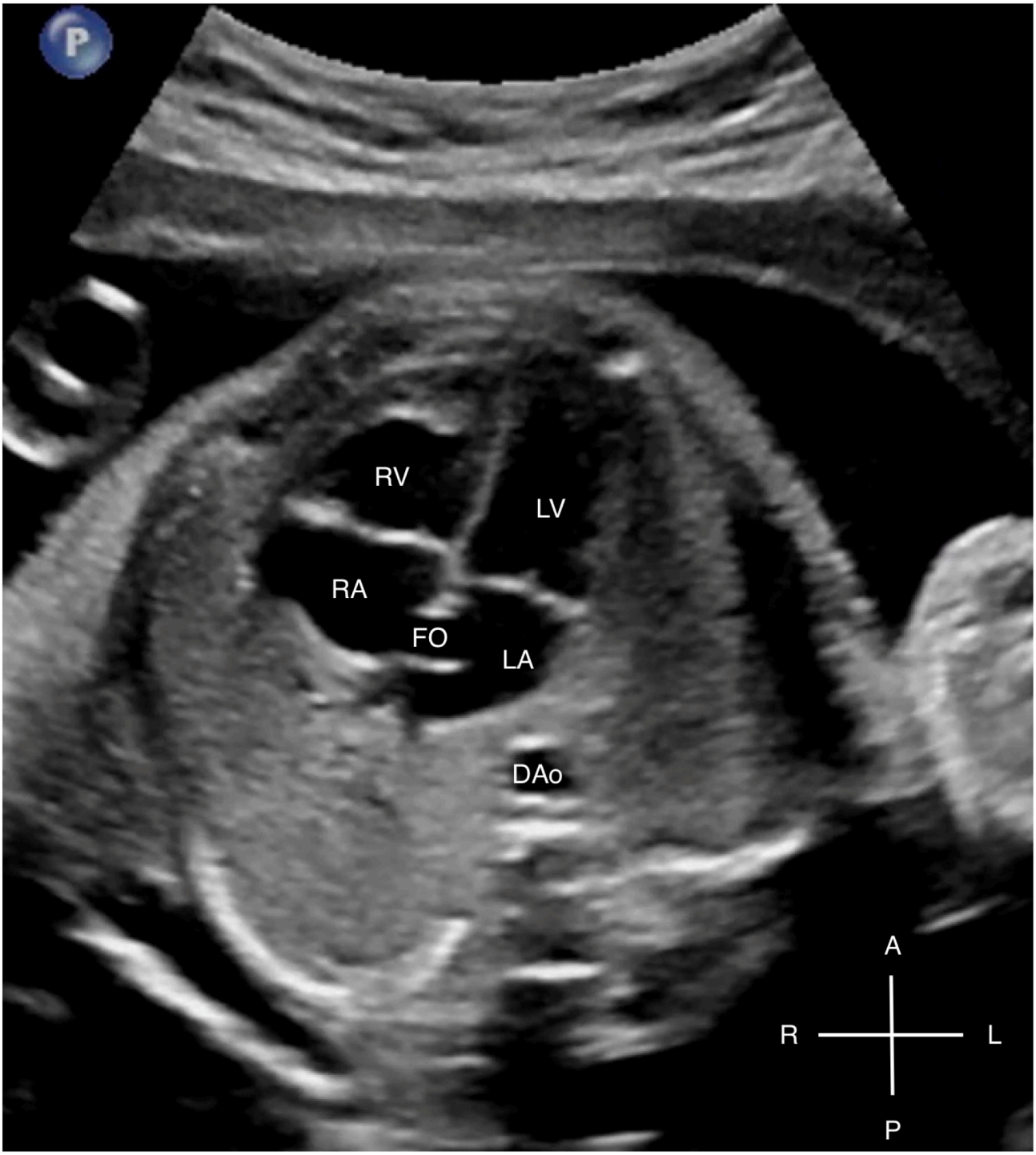

Fig. 7. Four-chamber view of the fetal heart. DAo, descending aorta; LA, left atrium; FO, foramen ovale; RA, right atrium; LV, left ventricle; RV, right ventricle; R, right; A, anterior; L, left; P, posterior.

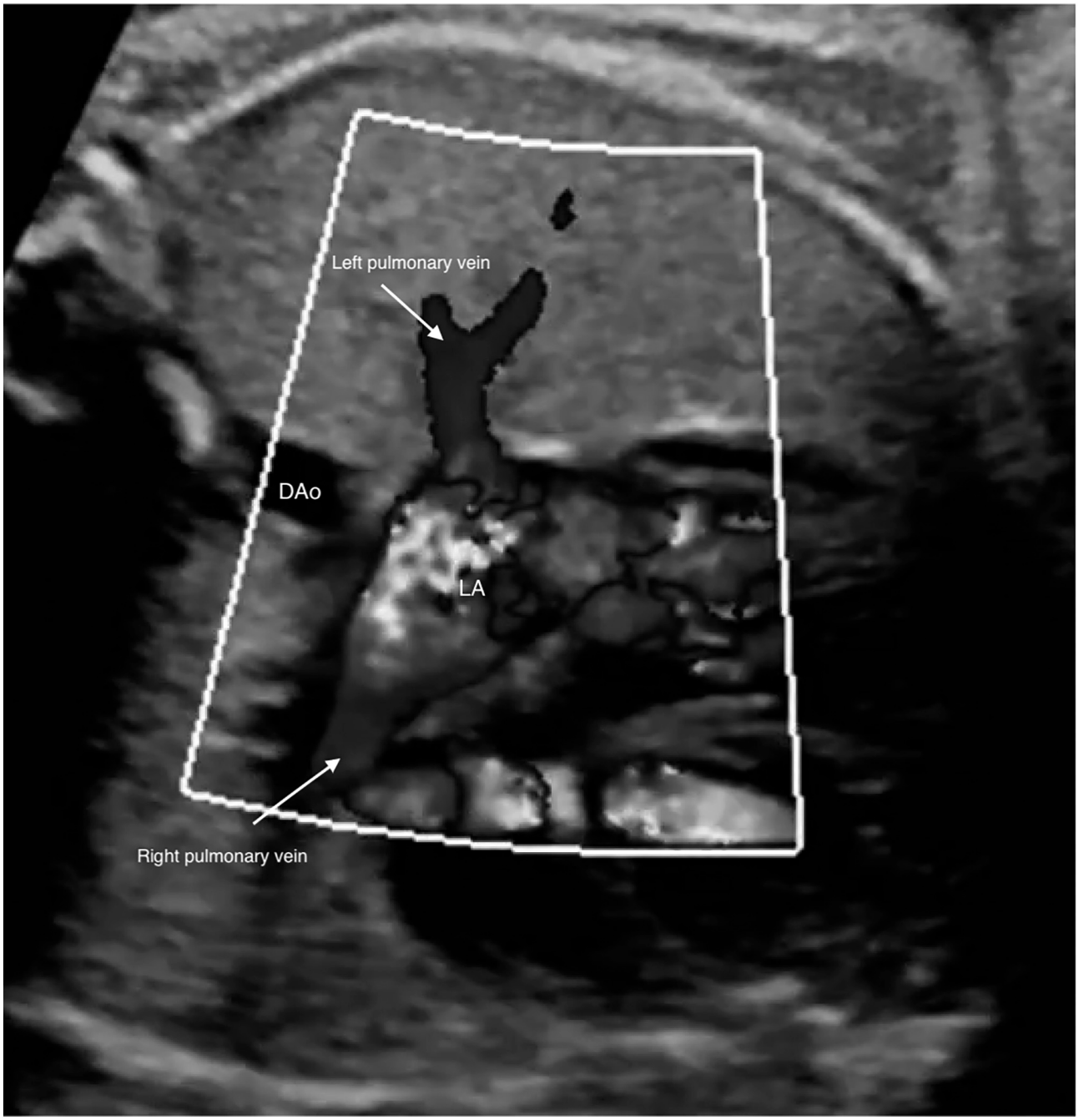

Fig. 8. Four-chamber view of the fetal heart showing color Doppler of a right and left pulmonary vein. DAo, descending aorta; LA, left atrium.

artery. The three vessels should be shown at the level of the pulmonary artery bifurcation and again at the level of the ductal arch. Diameter of the ductus arteriosus may be measured here. When the ductal arch is displayed, the trachea can be visualized between the superior vena cava and aorta. This is commonly called the 3-vessel trachea view. In the normal left aortic arch, the aortic and ductal arches come together in a "V" formation to the left of the trachea (Fig. 10). The side of the trachea that the aortic arch is on determines arch sidedness. Arch size

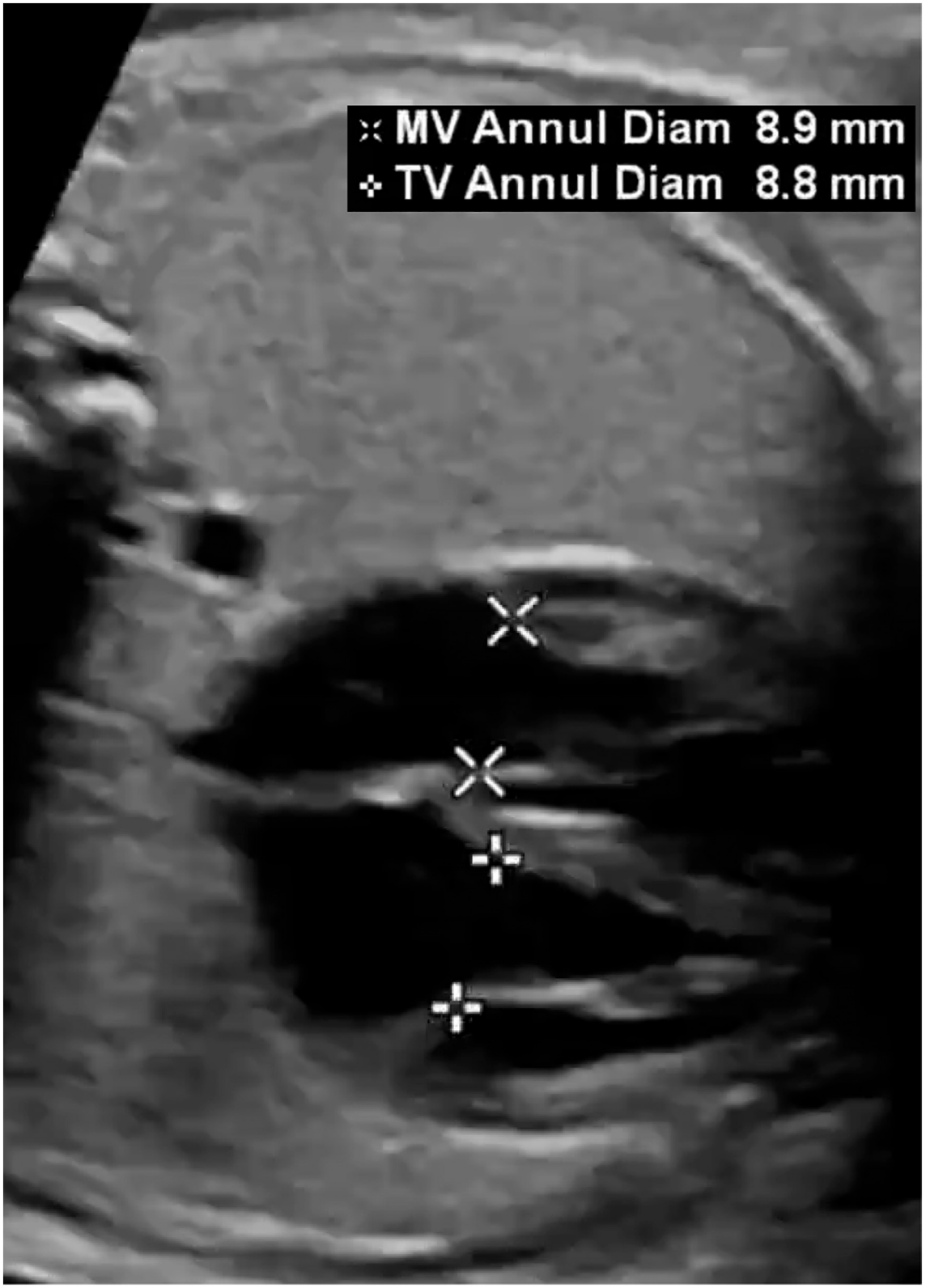

Fig. 9. Four-chamber view of the fetal heart in diastole with atrioventricular valve annulus dimensions noted. MV, mitral valve; Annul, annulus; Diam, diameter; TV, tricuspid valve.

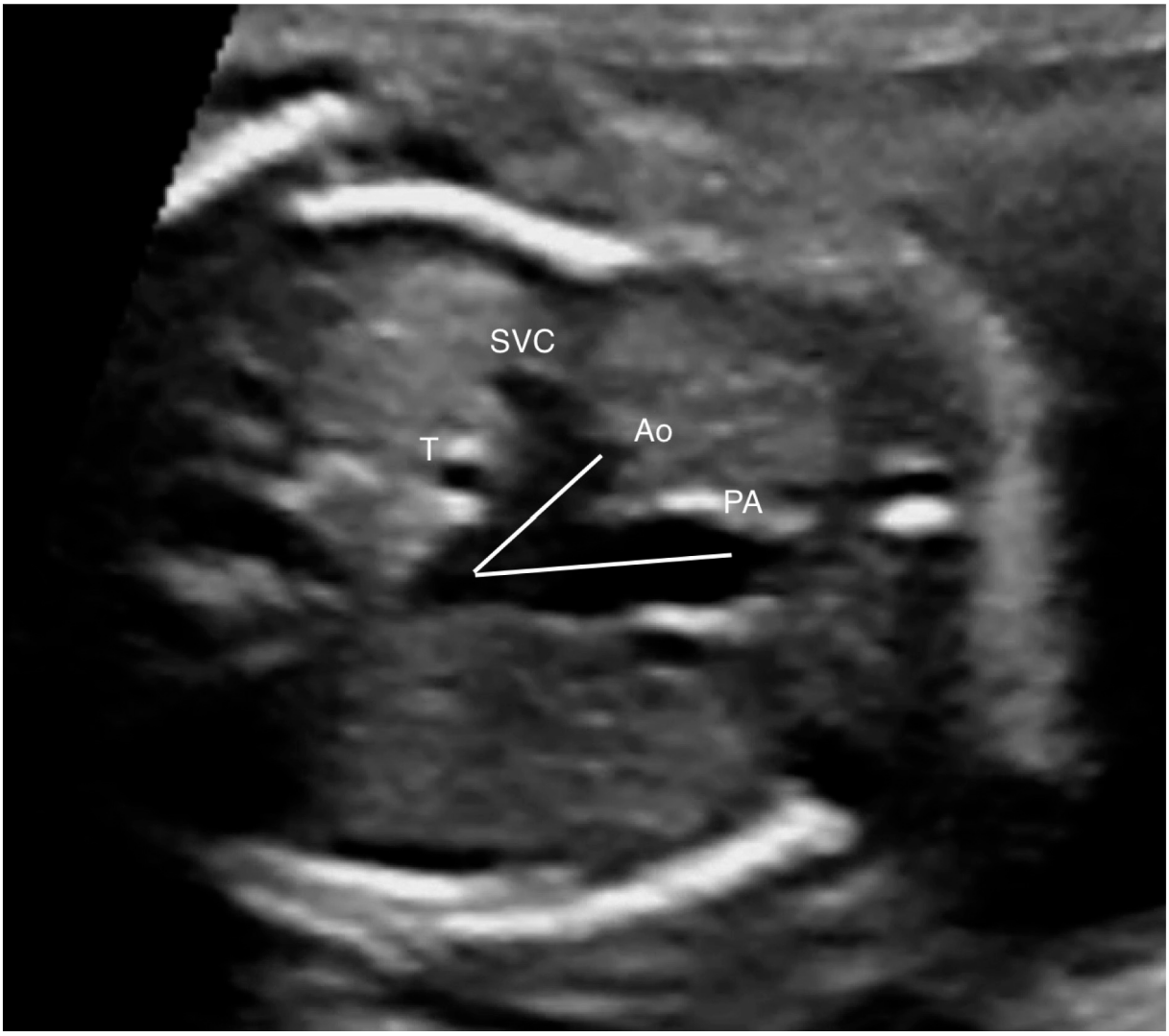

Fig. 10. 3 vessel tracheal view. Aortic arch and pulmonary trunk/ductal arch form a 'V' configuration. PA, pulmonary artery; Ao, Aorta; T, trachea; SVC, superior vena cava.

discrepancy and vascular rings can be optimally assessed in this view.

From the four-chamber view, a long axis view can be obtained by moving the probe cephalad in relation to the fetus and rotating the probe indicator towards the fetal right shoulder until the left ventricular outflow tract is visualized (Fig. 11). Then, continue to sweep cephalad until the right ventricular outflow tract is displayed. In this view, sweep in both grayscale and color to define outflow anatomy and evaluate for ventricular septal defects. This is an optimal view to measure pulmonary valve and aortic valve annulus diameters. By rotating the probe indicator to the fetus's left shoulder, a short axis view can be obtained. More anteriorly, the pulmonary artery bifurcation can be visualized (Fig. 12). If desired, this is a good imaging plane to measure dimensions of the main and branch pulmonary arteries. Pulse wave Doppler through the main pulmonary artery is typically performed. Scanning more posteriorly, a short axis view of the ventricles can be displayed (Fig. 13). Here, we can again qualitatively assess ventricular size and systolic function. Color Doppler assessment of the interventricular septum is performed to evaluate for ventricular septal defects. As an alternative to the 4-chamber view, M-mode through the ventricles can be obtained in short axis to measure ventricular free wall and interventricular septal thicknesses.

The superior and inferior vena cava are imaged in a sagittal plane through the chest and abdomen, commonly called the "bicaval view." The transducer is positioned along the posterior aspect of the right atrium parallel to where the cava enter (Fig. 14). The superior and inferior vena cavae are evaluated in grayscale and with color Doppler. Pulsed wave Doppler interrogation may be performed if clinically indicated. Angling posteriorly and leftward to bring the left atrium and atrial septum into view allows for additional color Doppler assessment of the atrial septum.

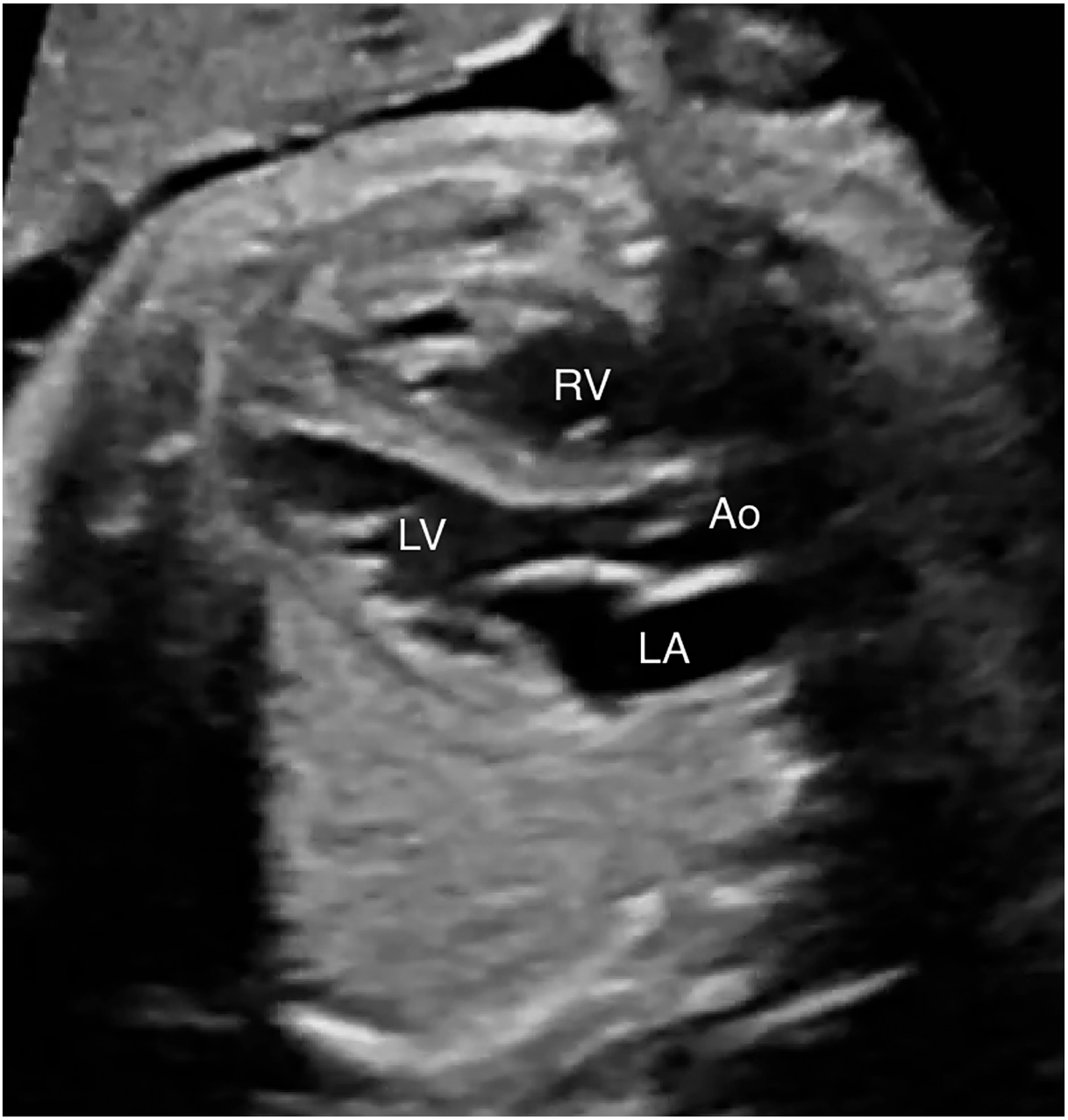

Fig. 11. Long axis view of the fetal heart. Aov, aortic valve; LA, left atrium; LV, left ventricle; RV, right ventricle.

Also in the sagittal plane, the ductal arch is imaged by aligning the transducer along the right ventricular outflow tract and main pulmonary artery. This arch has a less rounded appearance compared to the aortic arch and is frequently likened to a "hockey stick" (Fig. 15). A minimal sweep with the probe angled from anterior fetal right to posterior fetal left will bring in the classic "candy cane" aortic arch view (Fig. 16). Alternatively, the aortic arch can be obtained by rotating the transducer approximately 90° from the 3-vessel view. An important distinction between the two arches is that head and neck vessels arise from the aortic arch while the ductal arch is smooth. The arches are each evaluated in grayscale and color Doppler. Evaluate for antegrade flow in both arches. Pulsed Doppler of the arches and dimensions of the ascending aorta, transverse arch, isthmus and descending aorta may be measured as indicated.

The normal heart rate for the fetus is between 120 and 180 beats/min [13]. The normal heart rhythm is atrial mediated with one to one atrioventricular conduction. This can be demonstrated in multiple ways. A common method is to perform pulsed Doppler simultaneously

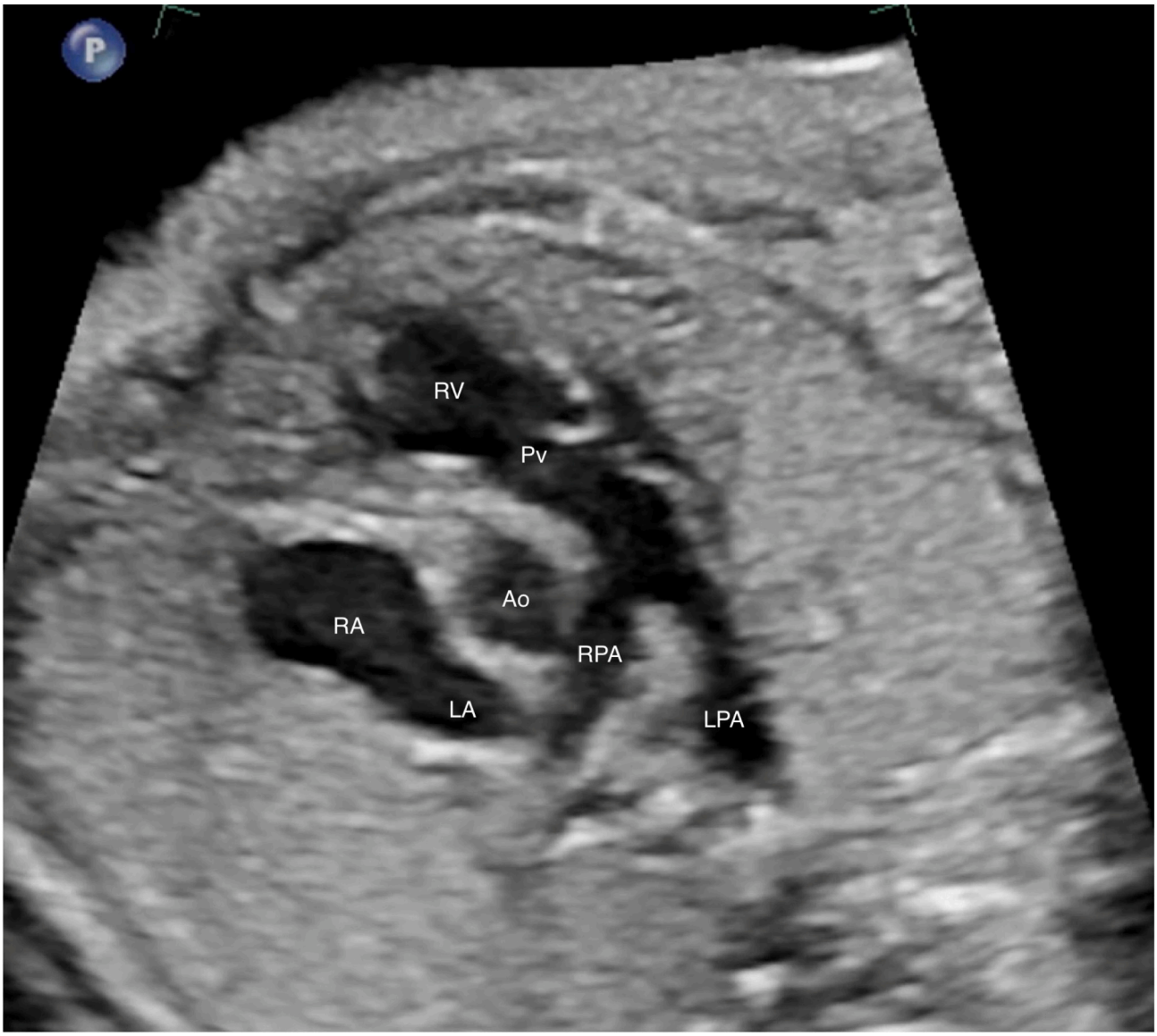

Fig. 12. Short axis view of the fetal heart at level of pulmonary artery bifurcation. LPA, left pulmonary artery; RPA, right pulmonary artery; Ao, aorta; LA, left atrium; RA, right atrium; RV, right ventricle; PV, pulmonary valve.

through an atrioventricular valve and a semilunar valve (Fig. 17A). To calculate a heart rate, the time between each inflow or outflow tracing can be measured on the machine in milliseconds and that number divided by 60,000. Many machines will do this calculation for the scanner. An outflow Doppler pattern should occur after every inflow Doppler pattern and conversely, an inflow before every outflow, if the patient is in an atrial mediated or sinus rhythm. Similarly, simultaneous pulsed Doppler of a pulmonary vein and artery can also be used to assess rate and rhythm (Fig. 17C). It should be noted that this Doppler pattern may be especially helpful in calculating ventricular-atrial and atrial-ventricular intervals in the classification of arrhythmias. Further discussion of arrhythmia evaluation is outside the scope of this review.

M-mode assessment can be performed in a 4-chamber view by aligning the cursor through a ventricle and contralateral atria. The ventricular and atrial contractions are evident on the M-mode tracing and can be utilized to assess for normal conduction and calculate a heart rate (Fig. 17B).

5. Limitations of fetal echocardiography

While fetal echocardiography has greatly increased the detection rate of fetal congenital heart disease, there are several limitations. Some limitations are inherent to the fetal cardiovascular system. For example, the large fetal ductal arteriosus may mask aortic arch obstruction and

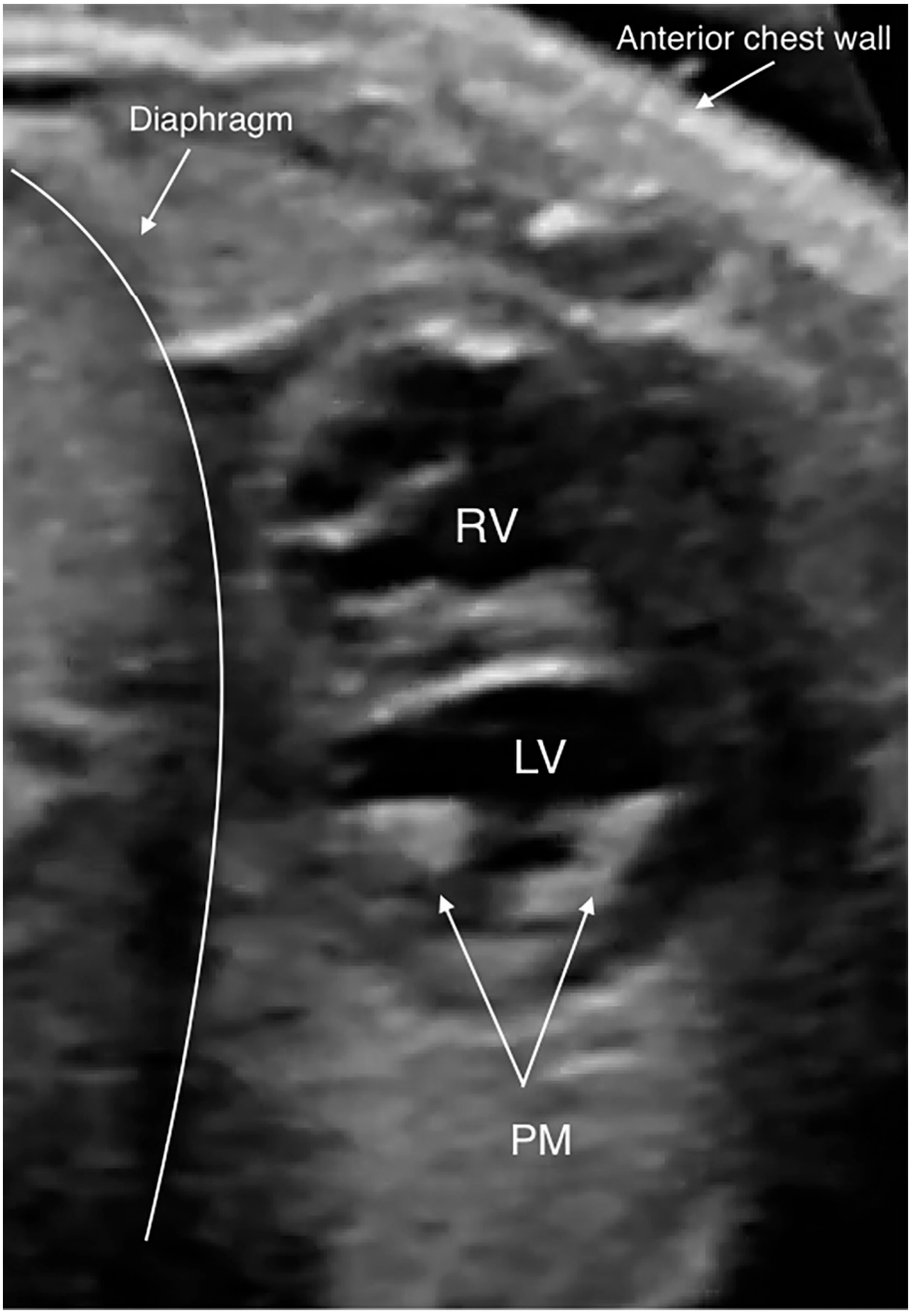

Fig. 13. Short axis view of the fetal heart at level of ventricles. PM, papillary muscles; LV, left ventricle; RV, right ventricle.

atrial septal defects can be difficult to differentiate from the stretched foramen ovale present prenatally. Small defects such as bicuspid aortic valve, coronary anomalies, partial anomalous pulmonary venous return, and small atrial or ventricular septal defects are not reliably evaluated by fetal echocardiography. Congenital conditions with later onset of phenotype such as cardiomyopathies cannot be ruled out prenatally. Additionally, fetal echocardiography may be limited by fetal position or maternal body habitus providing for suboptimal views.

6. Follow-up fetal echocardiograms

If congenital heart disease or fetal arrhythmia is detected or the fetus has a lesion that may predispose them to develop heart failure and chamber dilation, serial fetal echocardiography is recommended. While most serious congenital heart disease can be detected by 18–20 weeks, valvular stenosis and insufficiency may be difficult to detect prior to 30 weeks [17]. Additionally, many congenital heart lesions are

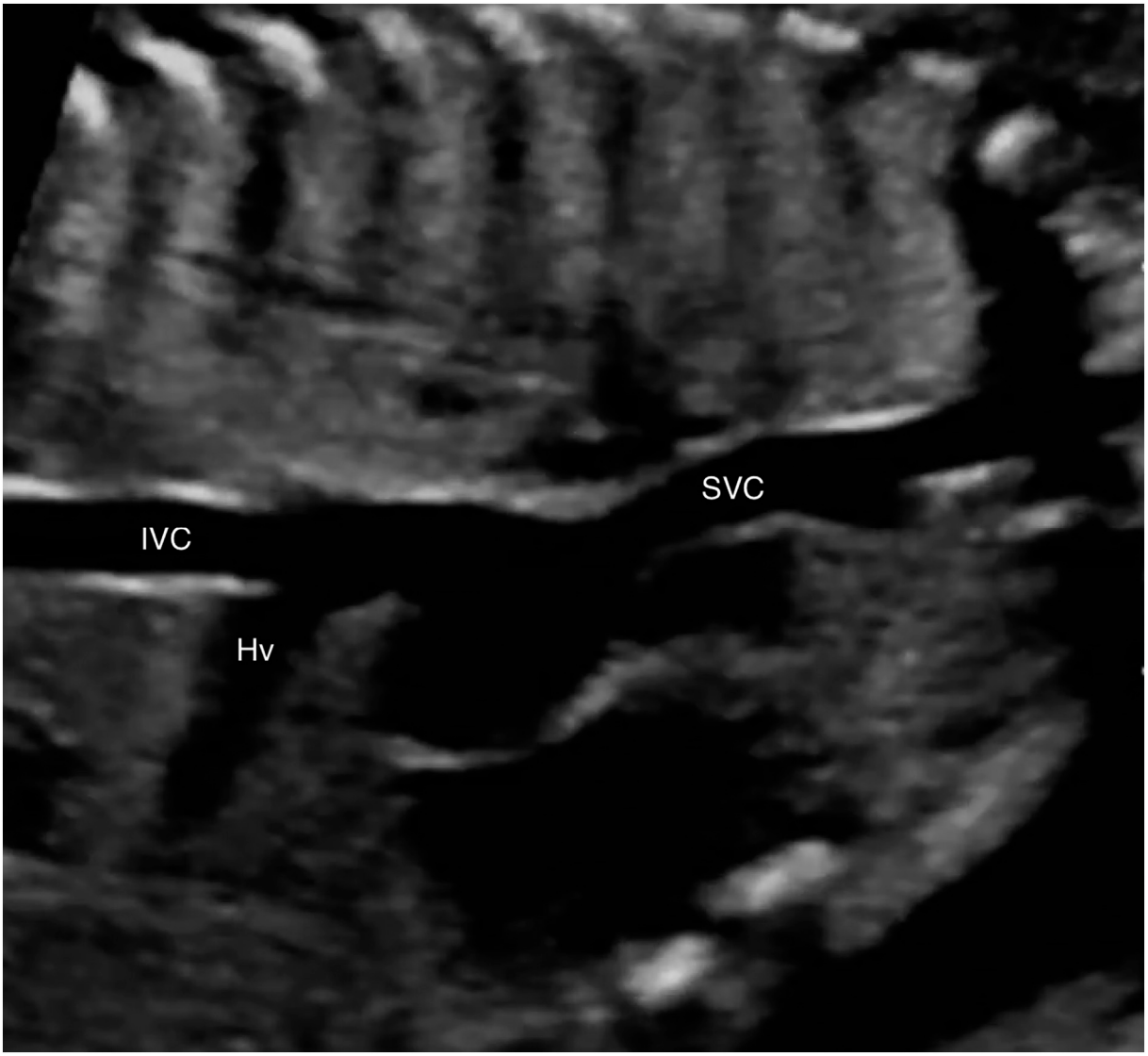

Fig. 14. Fetal bicaval view. SVC, superior vena cava; RA, right atrium; Hv, hepatic vein; IVC, inferior vena cava.

progressive during pregnancy and development of more severe valvular dysfunction or hypoplasia of chambers, arteries or valves can warrant change in delivery planning and treatment options. Additionally, some lesions may benefit from fetal intervention if caught early enough, such as severe aortic stenosis prior to progression to hypoplastic left heart syndrome. Timing of follow up is dependent on individual diagnoses and expected progression of that lesion.

7. Delivery planning

Fetal echocardiography is an essential tool for delivery planning in prenatally diagnosed congenital heart disease. It has high sensitivity and specificity predicting whether a newborn will require standard care in the delivery room or intervention immediately. With the close collaboration of the care team (which should include the maternal-fetal medicine specialist, neonatology, and cardiology), and continued monitoring with fetal echocardiography, a specialized delivery plan should be in place well before the expected due date. It is important to ensure that a baby is not separated from their mother nor a burden placed on the family by delivering at a facility far from their home if it is not necessary. The following scenarios are examples of when delivery at a local hospital is acceptable [5].

- A fetus with congenital heart disease where palliative care is planned. This may include patients with a fatal chromosome anomaly or multi system organ disease where surgery or heart transplantation is contraindicated. The family may deliver the baby at the institution of their choice with appropriate support services and palliative care in place.
- Infants with non-cyanotic congenital heart disease that are not

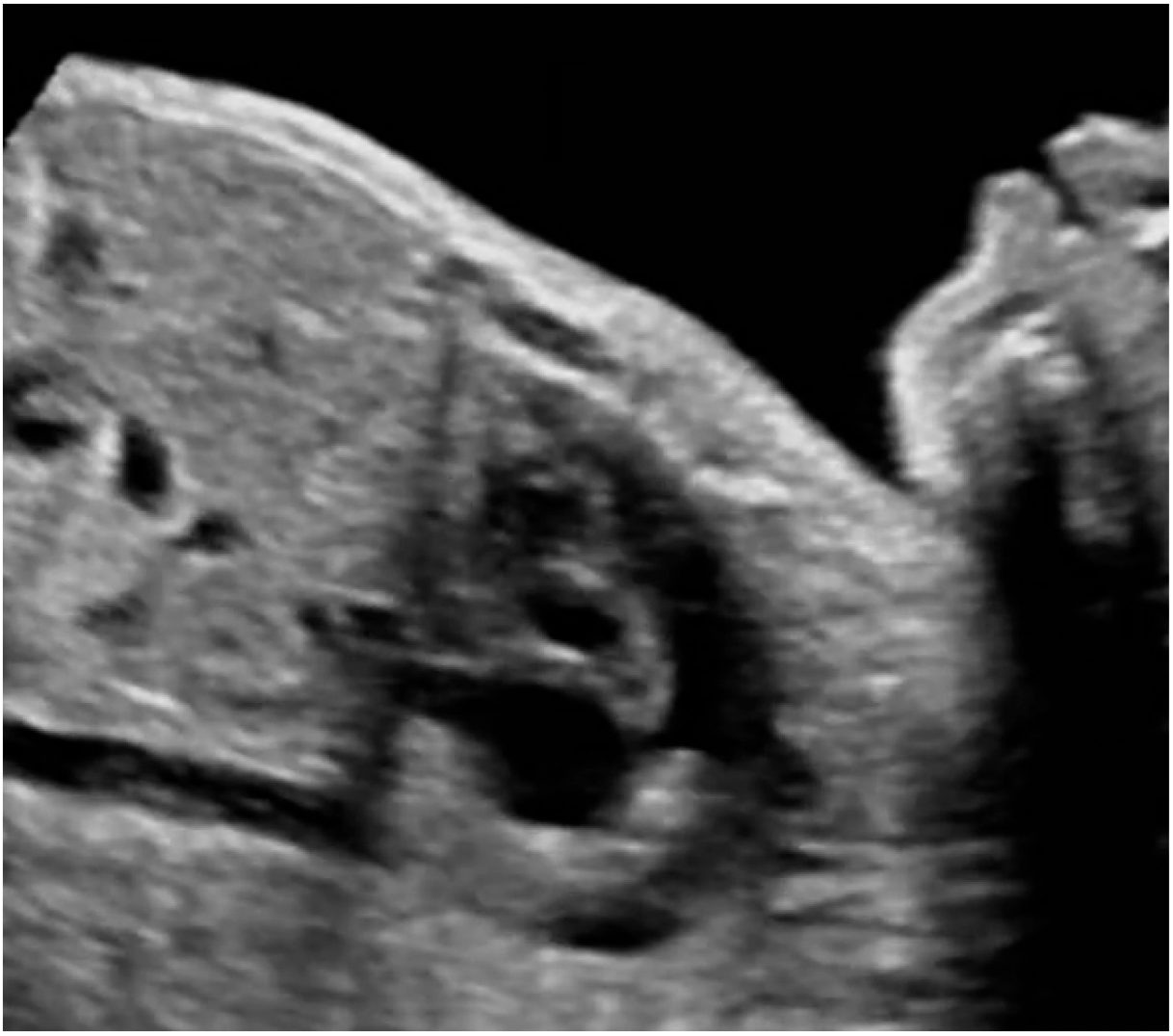

Fig. 15. Fetal ductal arch.

expected to have hemodynamic instability in the newborn period. This includes atrial and ventricular septal defects, balanced endocardial cushion defects and tetralogy of Fallot with minimal outflow obstruction and adequately sized pulmonary arteries. Depending on the lesion, inpatient cardiology consultation and echocardiogram can be performed after delivery or the baby may follow up with outpatient cardiology after discharge.

Infants with cyanotic congenital heart disease and ductal dependent lesions but no predicted hemodynamic instability in the immediate postnatal period should be delivered at an institution with a neonatologist in attendance and the ability to perform pediatric transthoracic echocardiograms [5]. The infant should be transferred to the neonatal or cardiac intensive care unit from the delivery room for umbilical line placement and initiation of prostaglandin infusion. If necessary, the baby can then be transferred to a tertiary care institution with pediatric cardiothoracic surgical services. Examples of this type of congenital heart disease include hypoplastic left heart syndrome, critical coarctation of the aorta, interrupted aortic arch, and tetralogy of Fallot. Institutions may consider induction near term. These infants can be born via vaginal delivery unless contraindicated. However, if there is potential for hemodynamic instability soon after delivery, the delivery should be coordinated at a tertiary care center with pediatric catheterization services, ECMO, pediatric cardiothoracic surgery, and in-house cardiologists. This includes d-transposition of the great arteries or hypoplastic left heart syndrome with restrictive atrial septum, intractable fetal arrhythmia, and congenital heart block with congestive heart failure. In order to coordinate care adequately, these pregnancies should be induced at 38–39 weeks gestation and cesarean section may be considered if necessary. The cardiologist and neonatologist should be present for delivery. If hemodynamic instability is expected to develop after the umbilical cord is clamped, the baby should be delivered

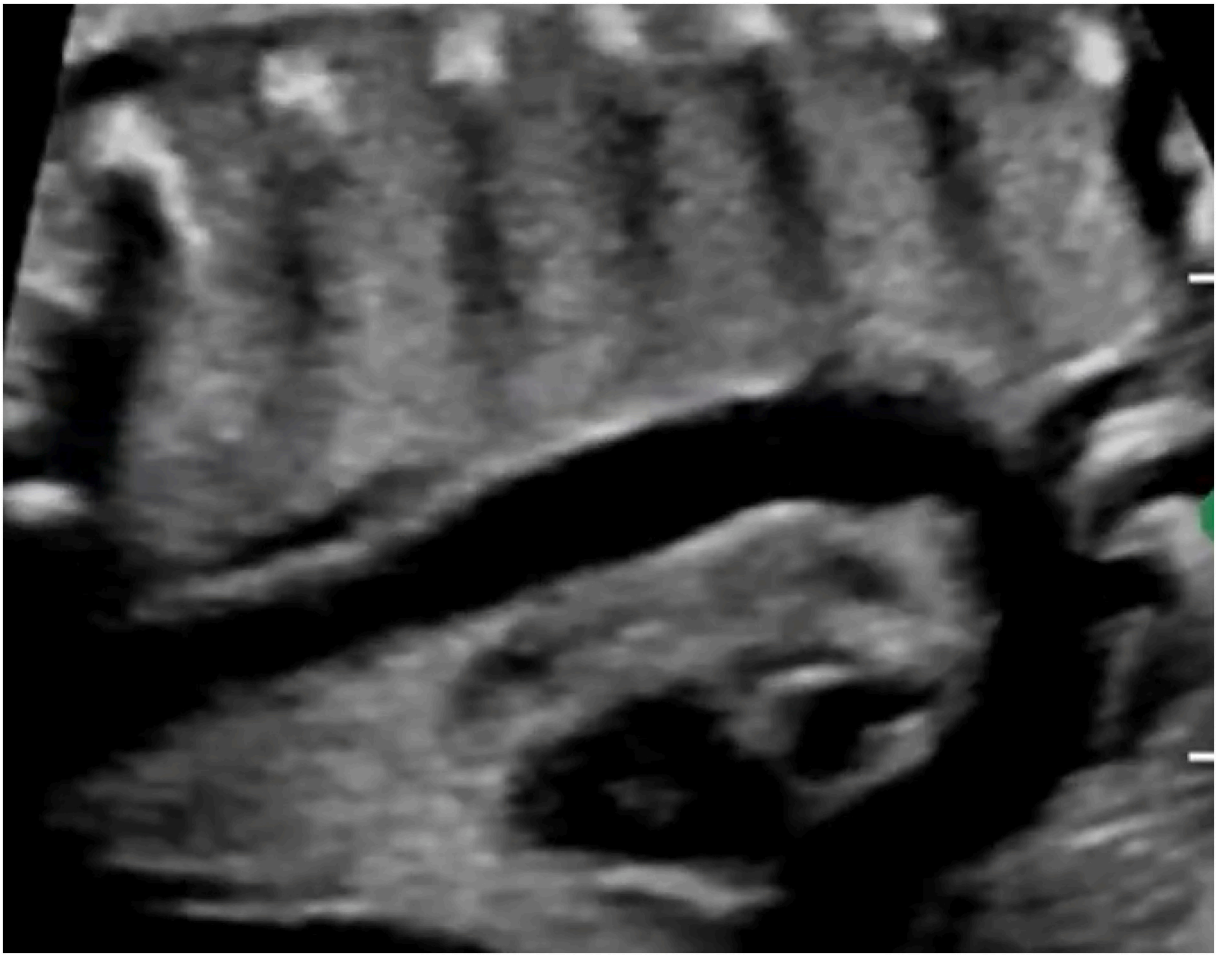

Fig. 16. Fetal aortic arch.

via cesarean section in close proximity to or in the catheterization lab or cardiovascular operating room for immediate intervention with appropriate personnel available at the delivery [5].

And finally, if neonatal heart transplantation is anticipated, the fetus can be listed at 35 weeks gestation and delivery induced or cesarean section scheduled once a heart is available. The baby would be delivered at a tertiary care institution with an appropriate cardiac care team present for delivery [5].

At our institution, the parents of fetuses diagnosed with complex congenital heart disease meet with the cardiothoracic surgeons and tour the cardiac intensive care unit at approximately 32 weeks gestation. If they will be delivering an affiliated institution and have not already done so, we facilitate transfer of general obstetrician care around 34–36 weeks gestation. Additionally, a perinatal nurse coordinator is available as a point of contact for the family when coordinating maternal fetal medicine and fetal cardiology appointments, other subspecialty consultations, and delivery planning. Additionally, they will accompany the mother to her appointments and assist with communication between the cardiologist, obstetricians and delivery hospital. If the family lives a significant distance from the tertiary institution where they plan to deliver, the institution may consider organizing local accommodations for the mother to stay near the delivery hospital preceding anticipated date of delivery. It is helpful to provide families with materials noting cardiac diagnosis with diagram, delivery recommendations and cardiologist's contact information to keep on their person. This is not only valuable to the family but in the event they have preterm labor or deliver at an unplanned institution, this documentation may help facilitate efficient and prompt coordination of care and appropriate treatment.

8. Conclusion

Fetal echocardiography is an important component in improving morbidity and mortality in infants with congenital heart disease. Prenatal diagnosis of severe congenital heart disease is associated with improved preoperative clinical status and improved postoperative survival [18,19]. With thorough fetal echocardiographic screening and more accurate prenatal diagnosis of congenital heart disease, we can better coordinate care between maternal-fetal medicine and pediatric cardiology and arrange appropriate care at and immediately after

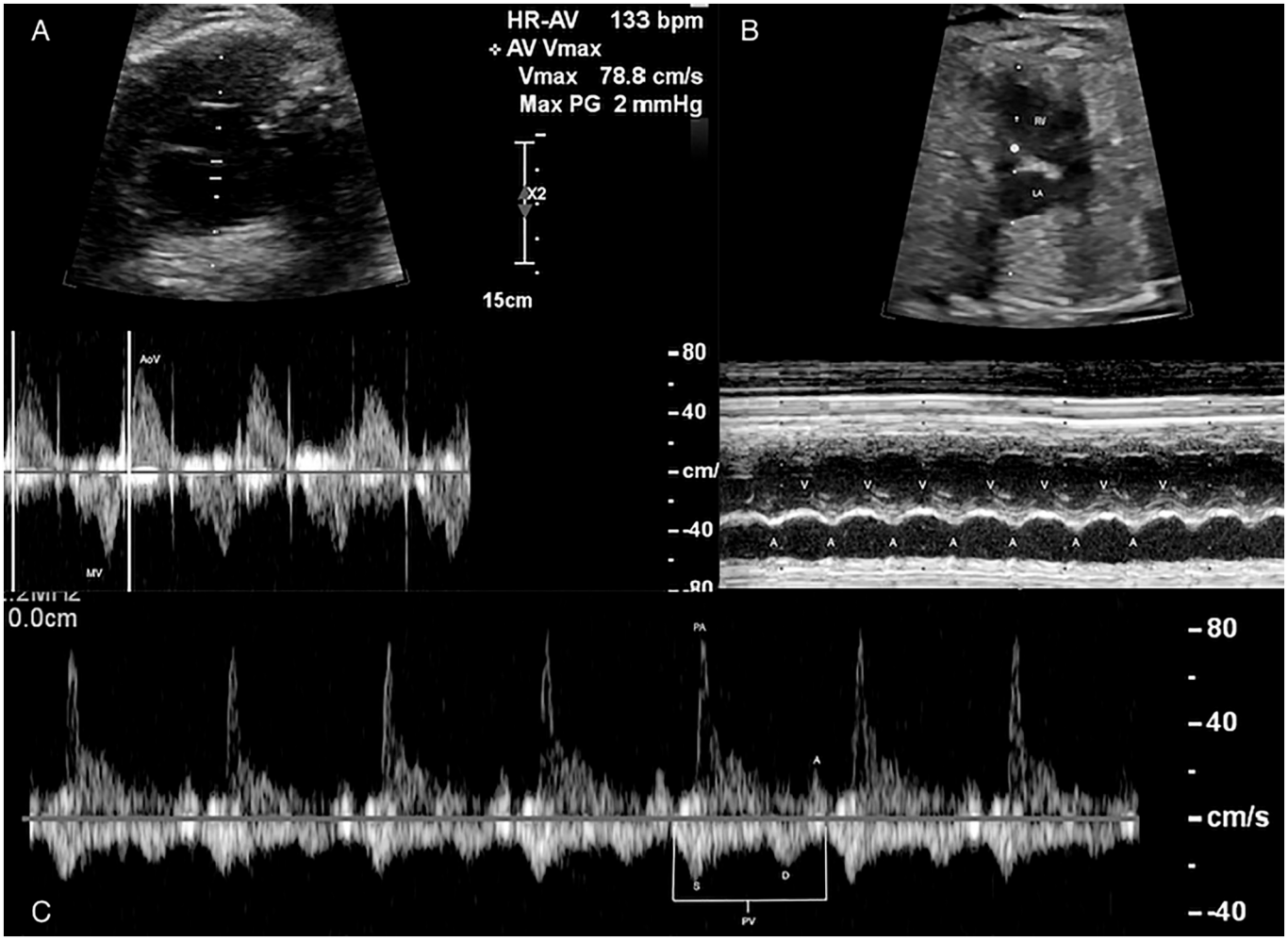

Fig. 17. (A) Pulsed-wave Doppler through mitral inflow and aortic outflow. (B) M-mode through right ventricle and left atrium. (C) Simultaneous pulsed-wave Doppler of pulmonary artery and pulmonary vein with S, D and A waves labeled. AoV, aortic valve; MV, mitral valve; V, ventricle; A, atrium; PA, pulmonary artery; PV, pulmonary vein.

delivery. This will minimize hemodynamic instability, ensure the infant is in optimal health prior to surgical intervention, and prevent delays in treatment.

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Declaration of competing interest

Authors have no conflicts of interest to declare.

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