CHAPTER 33

Coarctation of the Aorta

COARCTATION OF THE AORTA

Definition, Spectrum of Disease, and Incidence

Coarctation of the aorta, a common anomaly found in about 5% to 8% of newborns and infants with congenital heart disease (1), involves narrowing of the aortic arch, typically located at the isthmic region between the left subclavian artery and the ductus arteriosus (2) (Fig. 33.1). Alternatively, coarctation of the aorta may involve a long segment of the aortic arch, termed *tubular hypoplasia of the aortic arch*. Coarctation of the aorta occurs more commonly in boys, with a male-to-female ratio of 1.27 to 1.74:1 (3). The anomaly has a fairly high recurrence risk, between 2% and 6% for a previously affected child and 4% for an affected mother (4,5).

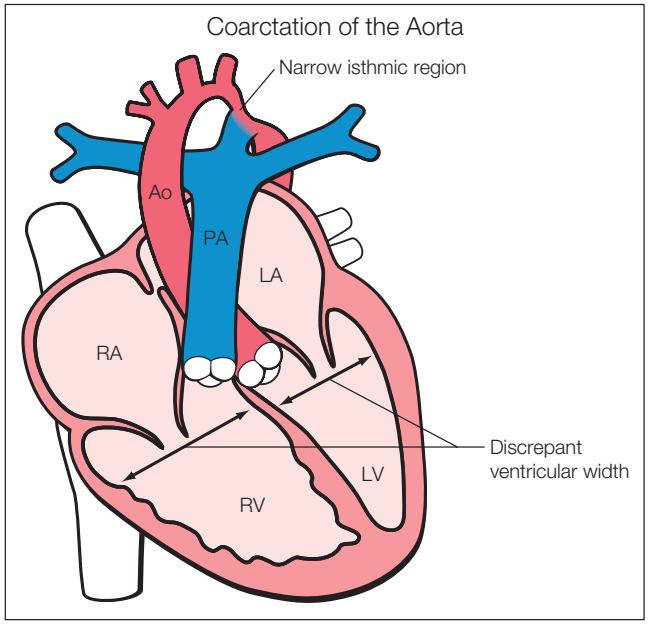

Figure 33.1: Schematic drawing of coarctation of the aorta. See text for details. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle.

Table 33.1 • Cardiac Anomalies Associated with Coarctation of the Aorta or Tubular Aortic Hypoplasia

- Unbalanced atrioventricular septal defect with a narrow left ventricle
- Hypoplastic left heart syndrome
- Double-outlet right ventricle
- Tricuspid atresia with ventricular septal defect and malposition of great vessels (type II)
- Corrected transposition of the great arteries
- Double-inlet ventricle (single ventricle)

Chromosomal and extracardiac abnormalities are commonly associated with coarctation of the aorta.

The embryologic origin of coarctation of the aorta is complex and not well understood, with two proposed concepts: the ductus tissue theory proposes that aortic constriction is due to migration of ductal smooth muscle cell into the aorta (6), whereas the hemodynamic theory proposes that coarctation results from reduced flow through the aortic arch in fetal life (7). Coarctation of the aorta can be classified as simple when it occurs without important intracardiac lesions and complex when it occurs in association with significant intracardiac pathology. When associated with hypoplastic left heart syndrome and aortic atresia, hypoplastic aortic arch should not be classified as coarctation of the aorta but rather as part of the main cardiac anomaly. Table 33.1 lists cardiac malformations that may include aortic coarctation or tubular aortic hypoplasia as part of the main cardiac anomaly. Figure 33.2 demonstrates coarctation of the aorta in an anatomic specimen of a fetal heart.

Ultrasound Findings

Grayscale

Ventricular disproportion is the leading sign for the suspicion of coarctation of the aorta in the four-chamber view. In

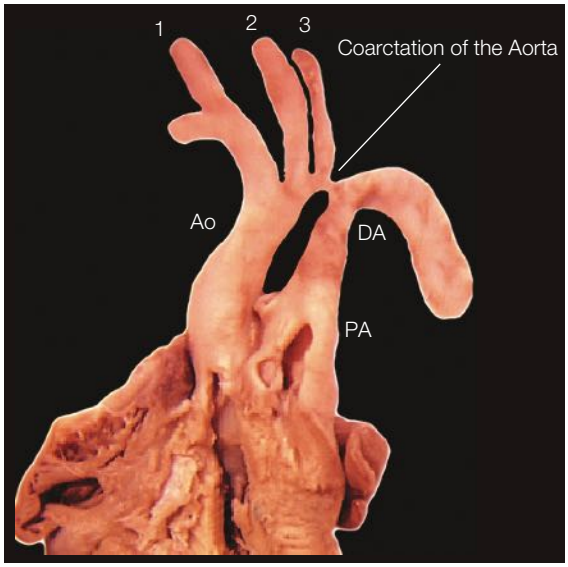

Figure 33.2: Anatomic specimen of a fetal heart with coarctation of the aorta. Note the presence of aortic narrowing at the level of the aortic isthmus (labeled coarctation of the aorta). 1,2 and 3 refer to the three arterial branches arising from the aortic arch (see Figure 33.8A) Ao, aorta; DA, ductus arteriosus; PA, pulmonary artery.

this view, the left ventricle appears narrow when compared to the right ventricle (8-10) (Figs. 33.3 and 33.4). The ratio of right ventricular and left ventricular width, reported as 1.19 in normal fetuses, is 1.69 in fetuses with coarctation of the aorta (11). Conversely to hypoplastic left heart syndrome, in coarctation of the aorta, the contractility of the left ventricle is normal and the mitral valve is patent (see Chapter 32). Aortic coarctation in the fetus is occasionally seen in association with a persistent left superior vena cava (LSVC) (Fig. 33.5). In this association, a cross section of the LSVC can be seen at the level of the four-chamber view, bulging into the external border of the left atrium and resulting in narrowing of the inflow of the mitral valve (Fig. 33.5) (see Chapter 42). The five-chamber view shows the ascending aorta with typically a normal diameter. The aortic root can be narrow on occasions, especially when a perimembranous ventricular septal defect and/or aortic stenosis are present. The aortic valve is occasionally bicuspid, an anatomic finding that is difficult to diagnose antenatally. In the three-vessel-trachea view, the diameter of the transverse aortic arch, in a tangential section, is narrow when compared to the diameter of the main pulmonary artery (11), and this narrowing of the transverse aortic arch is most impressive in the isthmus region (Figs. 33.6 and 33.7). In the diagnosis of aortic coarctation, the three-vessel-trachea view depicting the great vessel disproportion is more specific than ventricular disproportion alone, noted on a four-chamber view. Occasionally in this plane also a persistent LSVC can be found left to the pulmonary artery (Fig. 33.7E,F). Once coarctation of the aorta is suspected in the transverse planes (four-chamber and three-vessel-trachea views), a longitudinal view of the aortic arch should be attempted. In this plane, the length

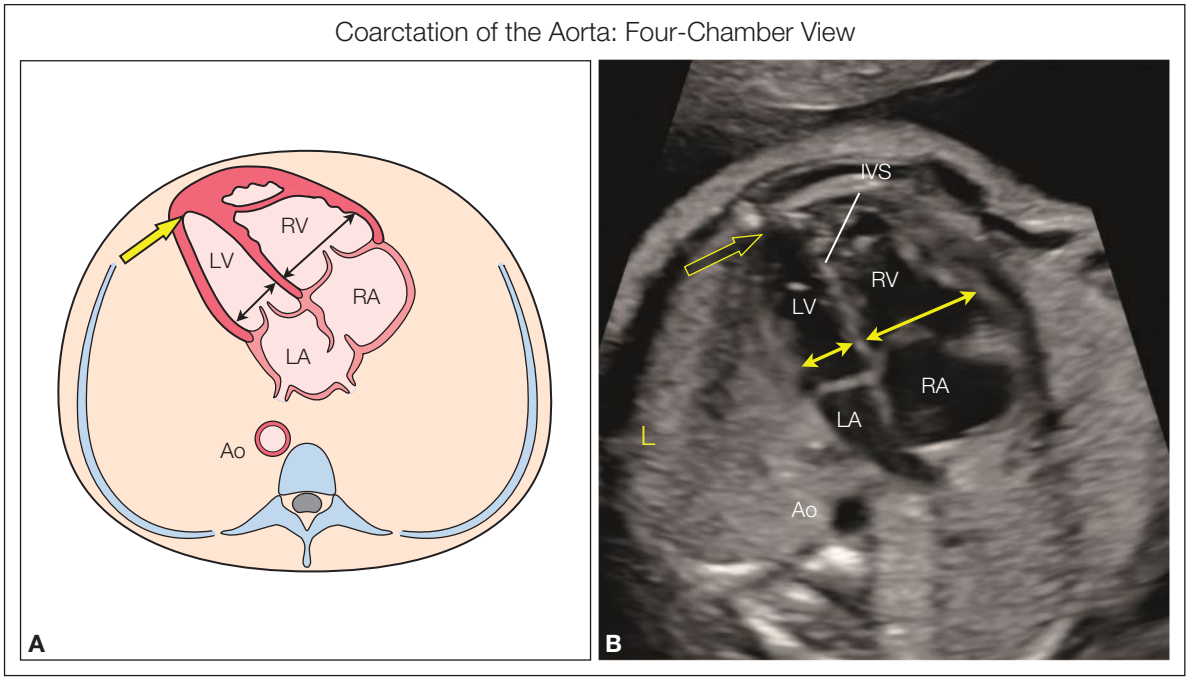

Figure 33.3: Schematic drawing (A) and corresponding ultrasound image (B) of a four-chamber view of a fetal heart with coarctation of the aorta, shown in grayscale (B). Primary clue for the presence of coarctation of the aorta is ventricular disproportion (double arrows, A, B), with the left ventricle (LV) smaller in width when compared to the right ventricle (RV). Another clue, which differentiates coarctation of the aorta from hypoplastic left heart syndrome, is that the LV is apex forming in coarctation (open arrow). Color Doppler is recommended to demonstrate normal filling of both ventricles in diastole (see text for details). Ao, descending aorta; IVS, interventricular septum; L, left; LA, left atrium; RA, right atrium.

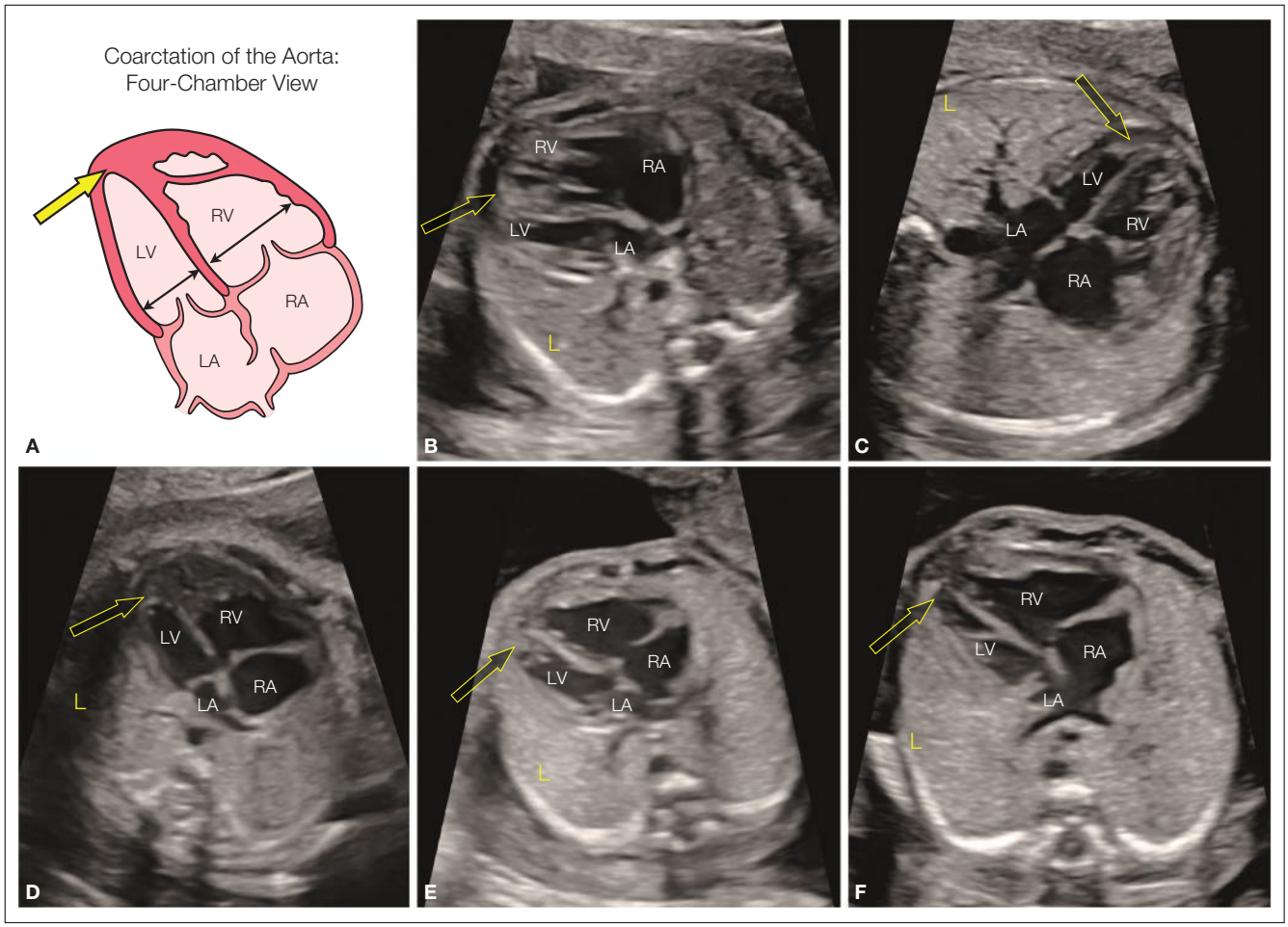

Figure 33.4: Schematic drawing (A) and corresponding four-chamber views in grayscale in five fetuses (B-F) with coarctation of the aorta. Note the two four-chamber view markers of coarctation of the aorta in grayscale: (1) ventricular disproportion and (2) apex-forming left ventricle (LV) (open arrow). L, left; LA, left atrium; RA, right atrium; RV, right ventricle.

and degree of the narrowing can be better assessed, and the junction of the aortic isthmus and the ductus arteriosus with the descending aorta is better evaluated (Figs. 33.8 and 33.9, see section "Color Doppler"). In this longitudinal view of the aortic arch, the narrowing is commonly located between the left subclavian artery and the origin of the ductus arteriosus. The aortic arch appears narrow and occasionally tortuous, termed *contraductal shelf*, an important clue for the presence of coarctation of the aorta. In the presence of severe coarctation of the aorta, the transverse arch between the left common and left subclavian arteries (Figs. 33.8 and 33.9) is elongated and narrow, and the left subclavian artery is found to arise at the junction of the ductus arteriosus with the descending aorta. *Z*-Scores for the measurements of the size of the aortic isthmus, the transverse arch, and the angle between the aortic isthmus and the ductus arteriosus were proposed to improve the accurate description of this cardiac anomaly (12-14).

Color Doppler

Color Doppler helps in differentiating coarctation of the aorta from other cardiac abnormalities and in demonstrating the narrow isthmic region. Applied to the four-chamber view, color Doppler confirms normal left ventricular filling in diastole and thus differentiates aortic coarctation from hypoplastic left heart syndrome (Figs. 33.10 and 33.11A,C). At the level of the five-chamber view, color Doppler demonstrates forward flow across the aortic valve (Fig. 33.11B,D) and a perimembranous ventricular septal defect when present in association with coarctation of the aorta. Often, the ascending aorta appears narrow at its root. In the three-vessel-trachea view or the transverse aortic arch view, a narrow transverse aortic arch can be recognized, which progressively becomes smaller in diameter as it approaches the aortic isthmus (Figs. 33.12 and 33.13). Despite the narrowing in the isthmic region, typically blood flow velocities are not increased, and color Doppler aliasing is not present in

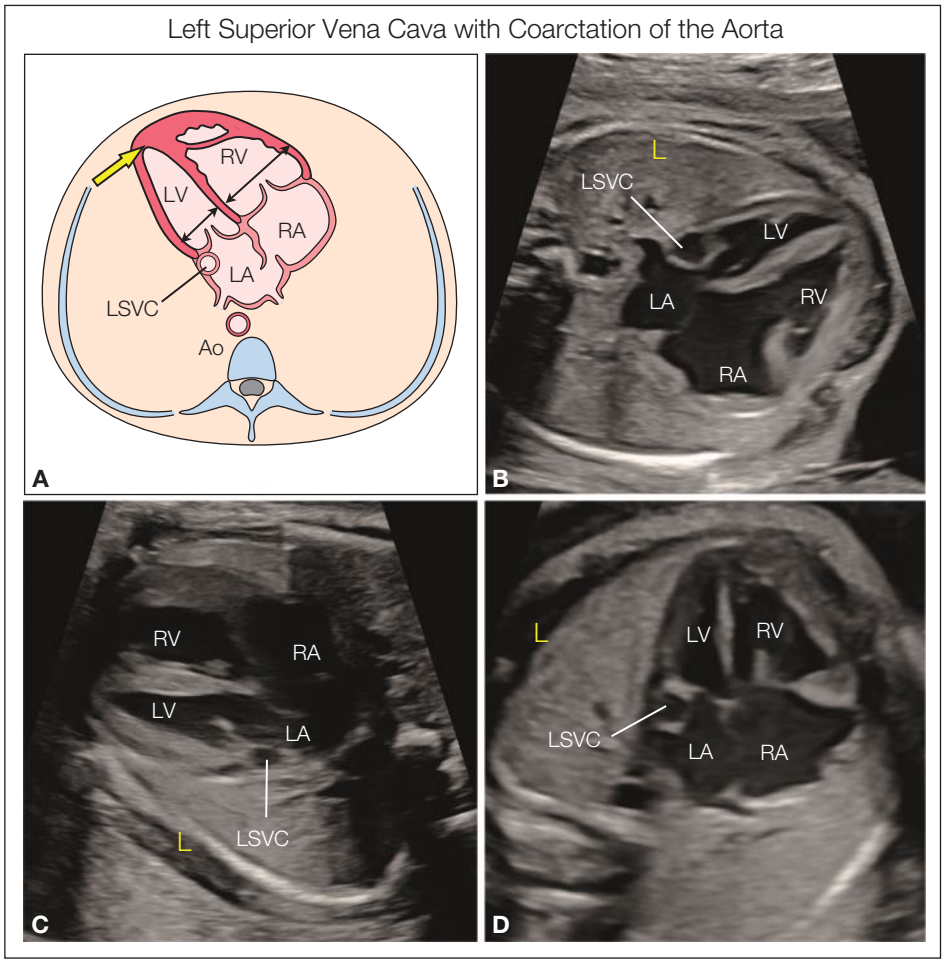

Figure 33.5: Schematic drawing (A) and corresponding four-chamber views in grayscale in three fetuses (B-D) with ventricular disproportion and coarctation of the aorta in combination with the presence of a persistent left superior vena cava (LSVC). The width of the left ventricle (LV) is smaller in comparison to the right ventricle (RV) (double arrows in A). A cross section of the LSVC can be seen at the left border of the left atrium (LA). The combination of coarctation of the aorta with LSVC is very typical, but the diagnosis should be supported by the evaluation of the great vessels. L, left; RA, right atrium.

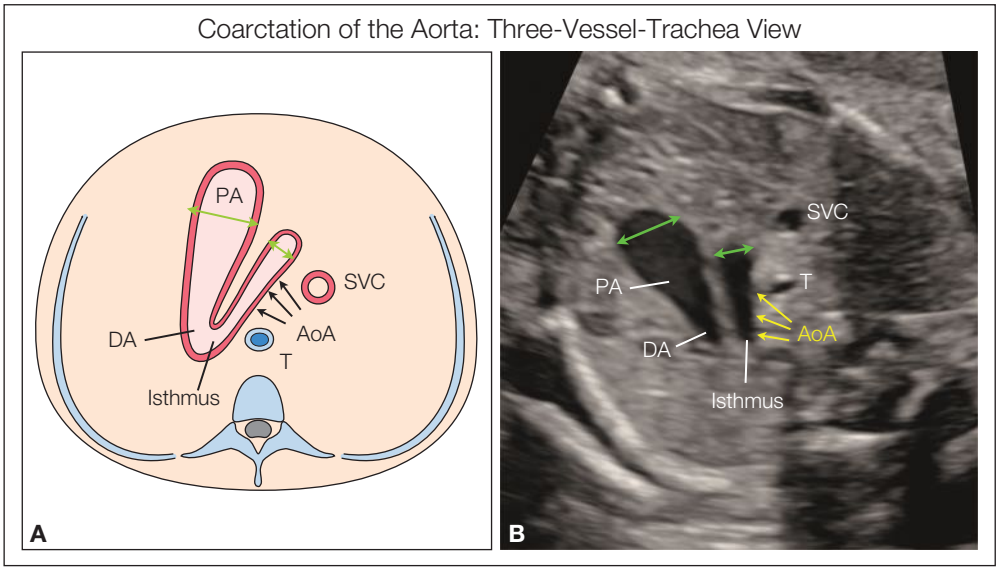

Figure 33.6: Schematic drawing (A) and corresponding ultrasound image (B) of a three-vessel-trachea view in a fetus with moderate coarctation of the aorta. The transverse aortic arch (AoA) is narrow when compared to the size of the pulmonary artery (PA) and ductal arch (DA) (double arrows). This finding complements ventricular disproportion in the four-chamber view and raises the suspicion for aortic coarctation or tubular hypoplasia of the AoA. Color Doppler further supports the diagnosis of moderate aortic coarctation by demonstrating antegrade flow in the AoA (see text for details). L, left; SVC, superior vena cava; T, trachea.

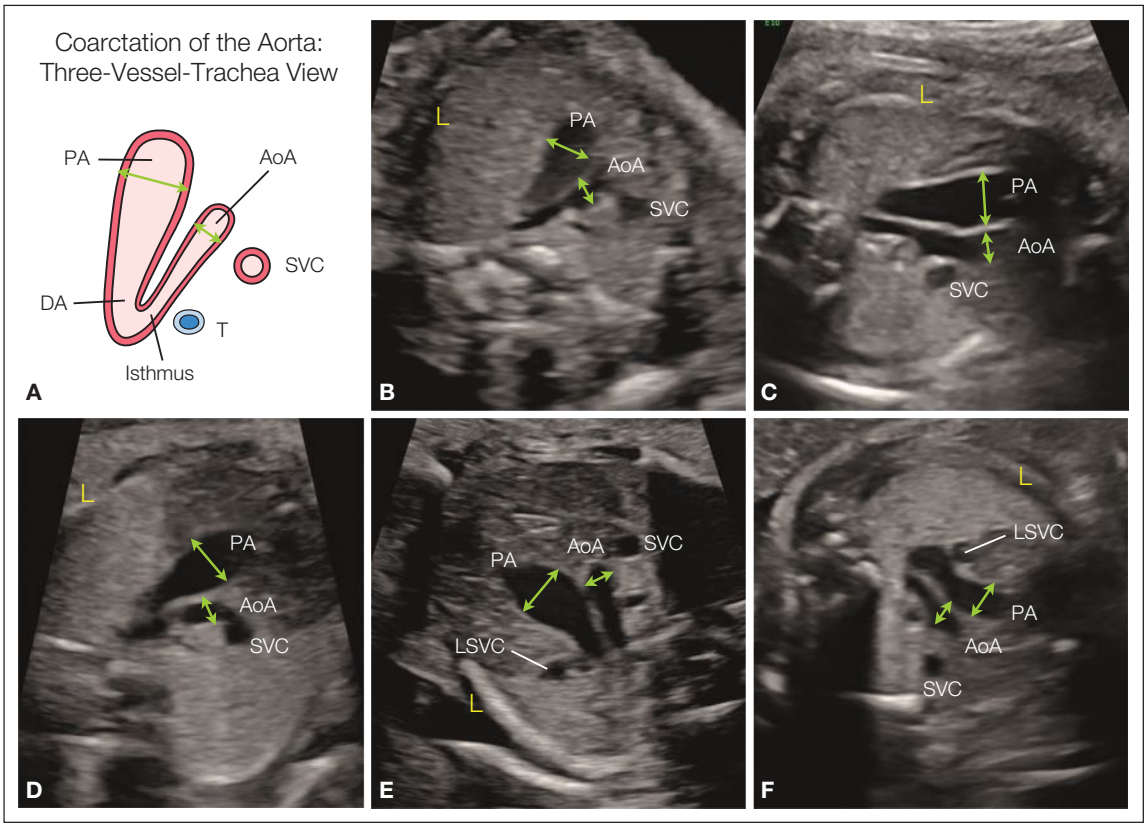

Figure 33.7: Schematic drawing (A) and corresponding ultrasound images of the three-vessel-trachea view in grayscale in five fetuses (B-F) with coarctation of the aorta with different insonation angles. Ultrasound images show size variations of the narrow transverse aortic arch (AoA) when compared to the size of the pulmonary artery (PA) and ductal arch (DA) (double arrows). Note that fetuses E and F also have a persistent left superior vena cava (LSVC), a common association with coarctation of the aorta. L, left; SVC, superior vena cava; T, trachea.

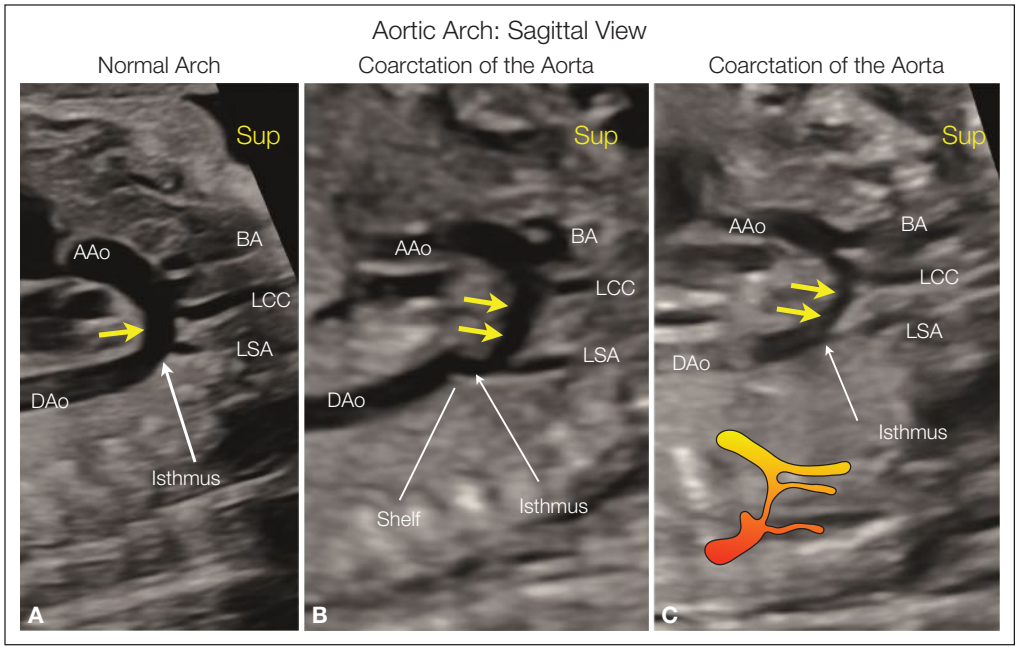

Figure 33.8: Sagittal views of the aortic arch in three fetuses, one with normal aortic arch (A) and two fetuses (B,C) with coarctation of the aorta. Schematic drawing of the arch in fetus C is also shown. Fetus B has the coarctation mainly localized to the isthmic region, whereas fetus C has tubular hypoplasia of the aortic arch. In fetus B, the connection of the isthmus with the descending aorta (DAo) shows a characteristic shelf. Note the origin of the left subclavian artery (LSA) from the transverse aortic arch in the normal fetus (A) in comparison to its more distal origin in the isthmic region in both affected fetuses (B,C). The schematic drawing (C) illustrates the abnormal shape of the arch. Yellow arrows point to the short distal aortic arch in the normal fetus (A), compared to a longer distal arch in aortic coarctation in fetuses B and C. Often, it is difficult to get the aortic arch plane, demonstrating the aortic narrowing, in grayscale and the use of color Doppler may be of help. AAo, ascending aorta; BA, brachiocephalic artery; DAo, descending aorta; LCC, left common carotid artery; Sup, superior.

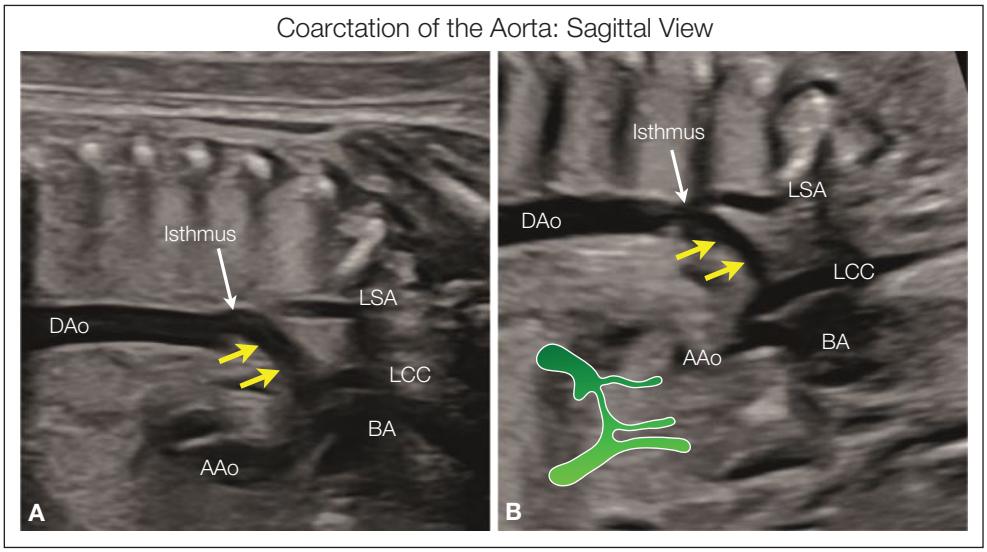

Figure 33.9: Sagittal view from a dorsal insonation in two fetuses, one with moderate coarctation of the aorta (A) and the other (B) with tubular hypoplasia of the aortic arch. Schematic drawing of the arch in fetus B is also shown, demonstrating its abnormal shape. The dorsal insonation reveals a better view of the isthmic region of the aortic arch. Yellow arrows show the stretched distal aortic arch along with the more distally arising left subclavian artery, almost from the isthmic and ductus arteriosus region. AAo, ascending aorta; BA, brachiocephalic artery; DAo, descending aorta; LCC, left common carotid artery.

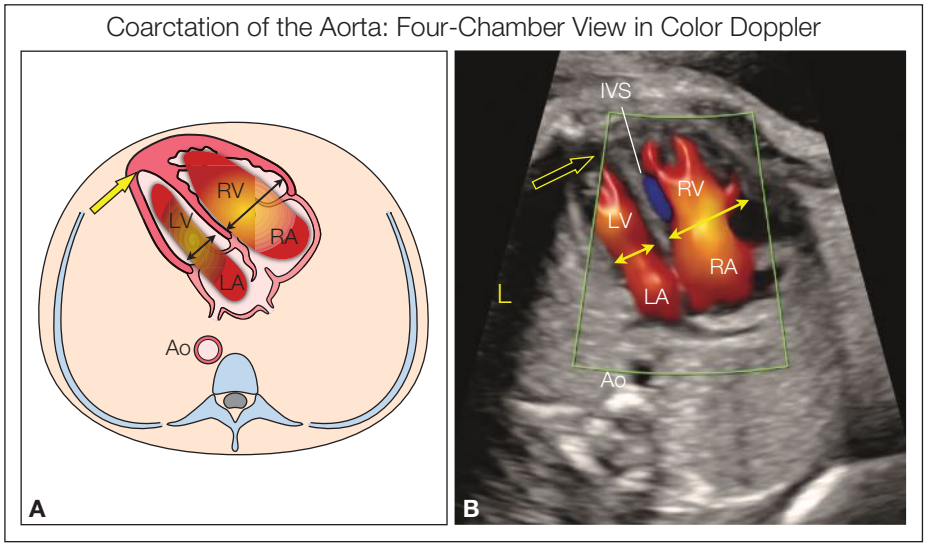

Figure 33.10: Schematic drawing (A) and corresponding ultrasound image (B) of a four-chamber view of a fetal heart with coarctation of the aorta, shown in color Doppler in diastole. The typical narrow width of the left ventricle (LV) in comparison to that of the right ventricle (RV) (double arrows) is demonstrated in color Doppler along with normal filling of both LV and RV in diastole, confirming patency of the atrioventricular valves. Note that the LV is apex forming in coarctation of the aorta (open arrows). Ao, descending aorta; IVS, interventricular septum; L, left; LA, left atrium; RA, right atrium

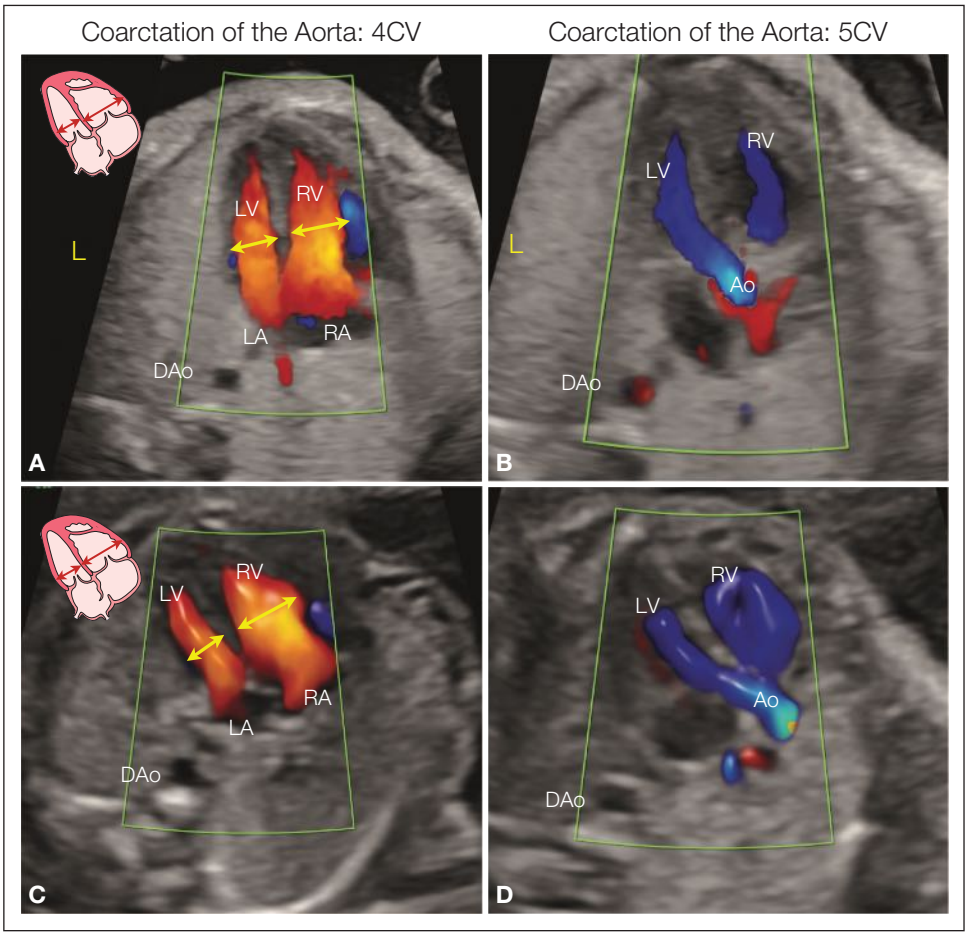

Figure 33.11: Color Doppler at the level of the four-chamber view (4CV) (A,C) and five-chamber view (5CV) (B,D) in two fetuses with coarctation of the aorta. Note (A,C) the narrow width of the left ventricle (LV) in comparison to that of the right ventricle (RV) (double arrows) and the normal filling of both LV and RV during diastole. In the five-chamber view (B,D), the ascending aorta (Ao) is visualized in normal size and normal antegrade flow. These color Doppler features are important to differentiate aortic coarctation from hypoplastic left heart syndrome as both presents with ventricular disproportion on grayscale imaging (see Chapter 32). DAo, descending aorta; L, left; LA, left atrium; RA, right atrium.

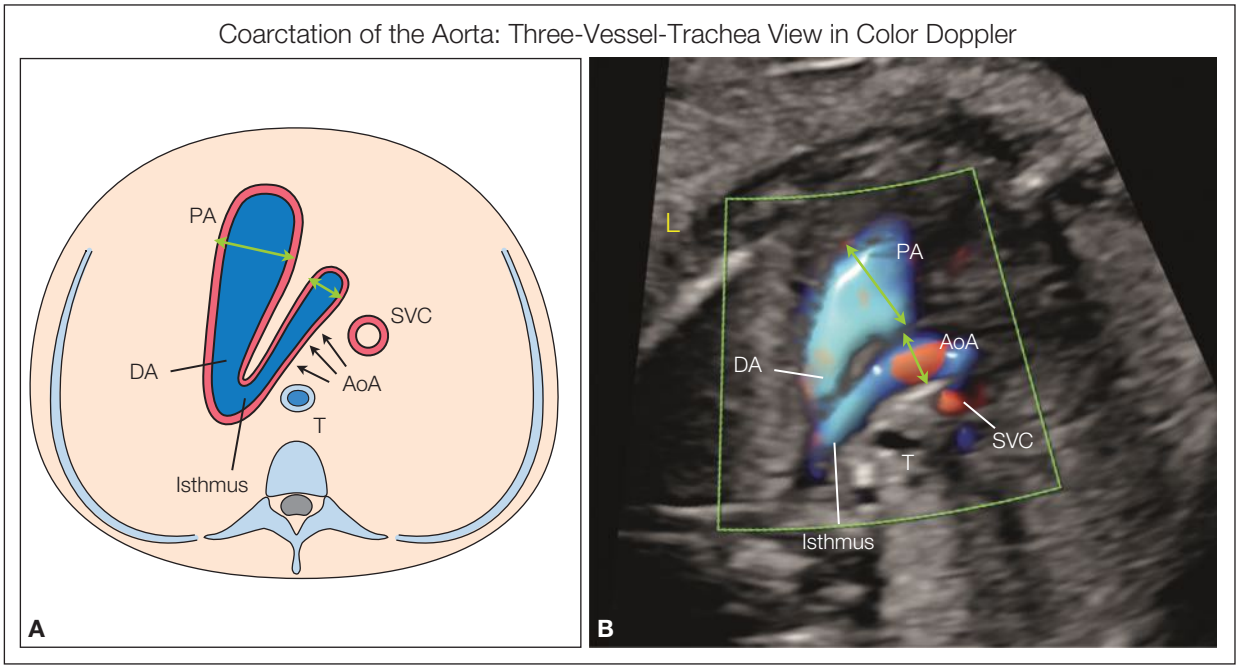

Figure 33.12: Schematic drawing (A) and corresponding ultrasound image (B) in color Doppler at the level of the three-vessel-trachea view in a fetus with coarctation of the aorta. Color Doppler shows the narrow transverse aortic arch (AoA) and isthmus in comparison to the pulmonary artery (PA) (double arrows) with antegrade flow across both AoA and PA, conversely to hypoplastic left heart syndrome (see Chapter 32) where flow in the AoA is retrograde. DA, ductus arteriosus; SVC, superior vena cava; T, trachea; L, left.

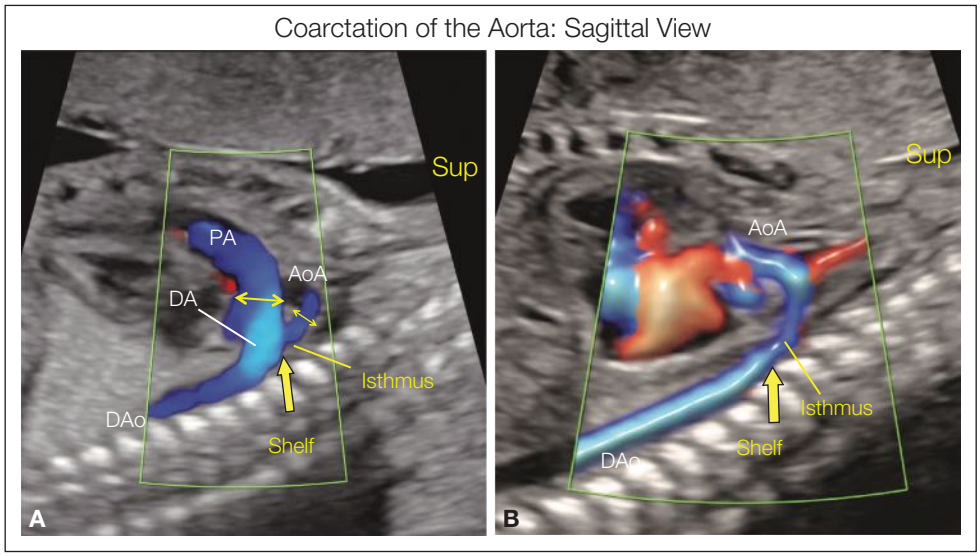

Figure 33.14: Color Doppler in a sagittal view of the aortic arch in two fetuses (A,B) with coarctation of the aorta. In fetus A, note the difference in diameter of the narrow distal aortic arch (AoA) and isthmus in comparison with the pulmonary artery (PA) and ductus arteriosus (DA) (double arrows). Yellow arrow in A points to the shelf. In fetus B, the tubular hypoplasia of the AoA is well seen, which is often difficult to visualize on grayscale. DAo, descending aorta; Sup, superior.

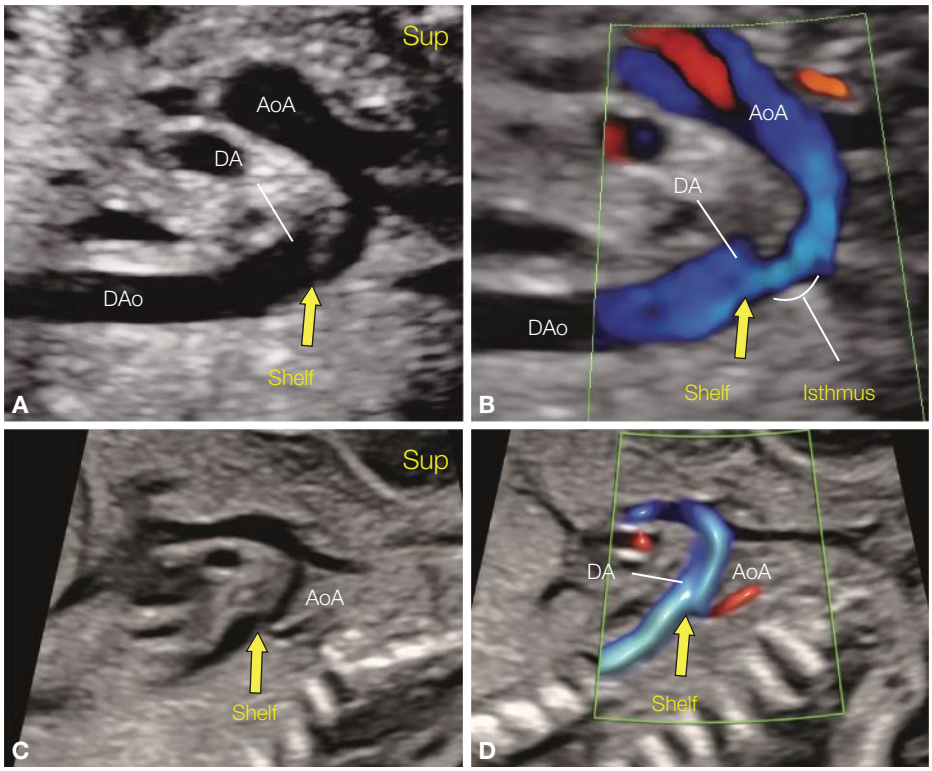

Figure 33.15: Sagittal views in grayscale (A,C) and color Doppler (B,D) of the transverse aortic arch (AoA) in two fetuses with coarctation of the aorta showing the narrow isthmic region (isthmus). Note the typical shelf (yellow arrow) in the region of the junction of the AoA and the ductus arteriosus (DA) into the descending aorta (DAo). Sup, superior.

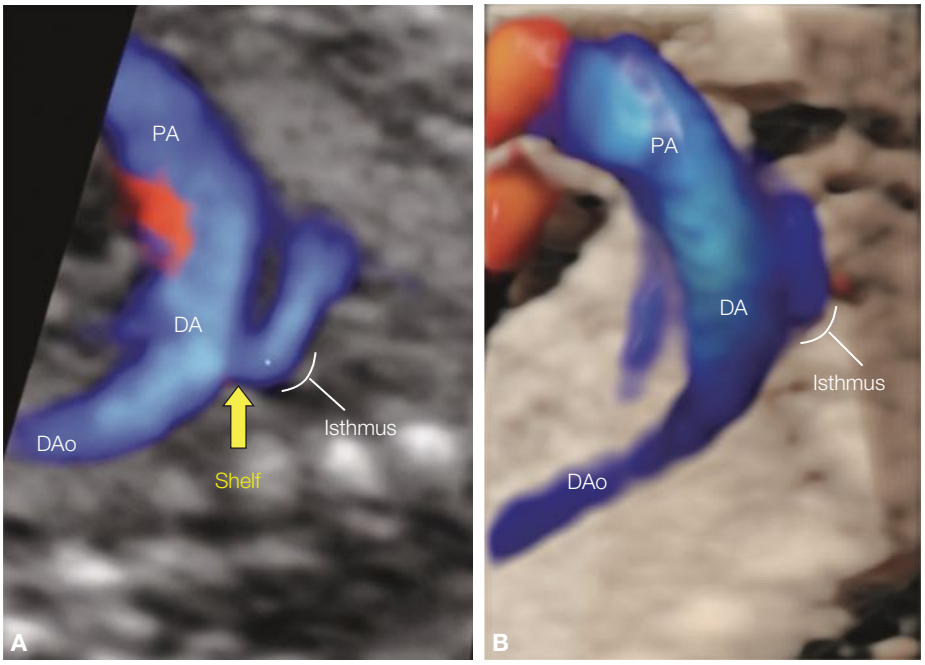

Figure 33.20: 3D ultrasound volume obtained in color spatiotemporal image correlation (STIC) of a fetus with coarctation of the aorta (A). Volume manipulation (A) allowed the retrieval of an oblique view of the ductal arch (DA) at the level of the junction with the aortic isthmus. Note (A) the narrow aortic isthmus with the typical shelf (shelf sign) at the transition into the descending aorta (DAo). Flow in the isthmus is antegrade in contrast to retrograde flow commonly noted in hypoplastic left heart syndrome (see Chapter 32). 3D glass-body mode (B) rendering in another fetus with aortic coarctation shows similar features. PA, pulmonary artery.

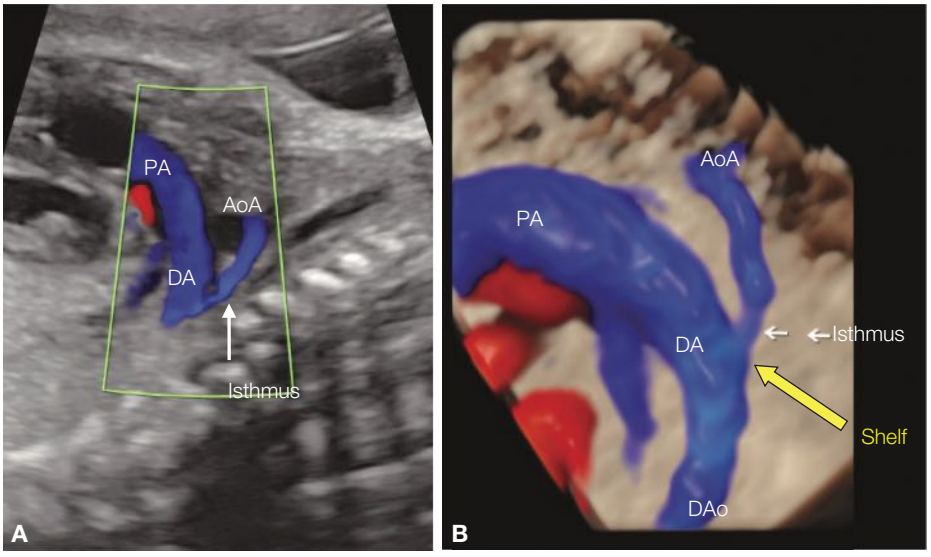

Figure 33.21: Sagittal view of the aortic arch in color Doppler (A) and the corresponding 3D ultrasound glass-body mode (B) in a fetus with coarctation of the aorta and tubular hypoplasia of the aortic arch. Note (A,B) the narrow transverse aortic arch (AoA) and its junction with the ductus arteriosus (DA) (shelf). DAo, descending aorta; PA, pulmonary artery.

involve variations in brachiocephalic anatomy and in berry aneurysms of the circle of Willis, which may lead to intracerebral bleeding. Berry aneurysms of the cerebral circulation have been reported in up to 3% to 5% of patients with aortic coarctation. Nonvascular-associated anomalies involve multiple organ systems, such as the genitourinary, musculoskeletal, gastrointestinal, and others, and may be present in up to 30% of children with coarctation of the aorta (27). Chromosomal abnormalities are commonly associated with aortic coarctation diagnosed prenatally, with a reported aneuploidy rate up to 35% in a retrospective referral-based review, and Turner syndrome representing the most common abnormality (27). Aortic coarctation can also be found in association with other chromosomal aberrations such as trisomy 13 or 18, especially when coarctation is associated with multiple extracardiac malformations.

Differential Diagnosis

Coarctation of the aorta is difficult to detect prenatally, with a high false-positive and false-negative diagnosis (9,10,28,29). Indeed, precise diagnosis of coarctation of the aorta during fetal life remains difficult because coarctation of the aorta may present several weeks after closure of the arterial duct and, thus, sequential echocardiography is recommended (30).

In a retrospective study including 108 fetuses with suspected coarctation of the aorta, postnatal confirmation was performed in 51% of neonates (29). The presence of aortic arch hypoplasia on prenatal ultrasound yielded the highest correlation, and affected neonates presented also with significantly lower *Z*-scores of the ascending aorta and of the aortic isthmus (29). Earlier gestational age at referral was also positively correlated with neonatal aortic coarctation (29). In this cohort, no single prenatal parameter was sufficiently accurate in diagnosis, but a scoring system permitted better identification of affected fetuses. In a single-center study evaluating the association between fetal echocardiographic measurements and the need for postnatal intervention in a cohort of 107 fetuses with prenatally diagnosed coarctation of the aorta, variables associated with intervention comprised smaller ascending aorta and transverse arch, earlier gestational age at diagnosis, and the additional finding of a higher peak systolic velocity in the ascending aorta on spectral Doppler (31).

A systemic review and meta-analysis on prenatal ultrasound risk factors for the diagnosis of coarctation of the aorta also showed that detailed fetal echocardiography can stratify the risk for aortic coarctation in fetuses with a suspected diagnosis and that prenatal detection rate may improve when a multiple criteria prediction model is adopted (28). Prospective and validated objective models for risk assessment are needed, however, in order to ascertain the actual performance of prenatal ultrasound in the diagnosis of coarctation of the aorta (28). Until then, the prenatal diagnosis of coarctation of the aorta will continue to be associated with high false-positive and false-negative rates.

The most common differential diagnosis for coarctation of the aorta includes hypoplastic left heart syndrome (HLHS) and type A interrupted aortic arch (see Chapters 32 and 34). Assessing ventricular contractility on grayscale ultrasound and flow across the mitral valve on color Doppler can help differentiate HLHS from coarctation (Table 32.1). Documenting flow across the aortic arch on color Doppler in longitudinal views can help differentiate coarctation from interrupted arch (no flow across). The presence of severe fetal growth restriction can also present with a narrow aortic isthmus due to blood shunting and, thus, be mistaken for coarctation of the aorta. The presence of a redundant foramen ovale flap, also described as aneurysm of the foramen ovale and defined by a foramen ovale flap that herniates into the left atrium for more than 50% of the left atrial diameter, may fully mimic coarctation of the aorta with ventricular disproportion and narrowing of the aortic isthmus with reverse isthmic flow in some cases (32) (Figs. 19.15 and 19.16 in Chapter 19). Other conditions associated with discrepant ventricular dimensions with a diminutive-appearing left ventricle are listed in **Table 33.2** and **Figure 33.22**.

Table 33.2 • Conditions Associated with Ventricular Size Disproportion with an Increased Right Ventricle–to–Left Ventricle Ratio (RV/LV)

- Aortic coarctation
- Hypoplastic left heart syndrome
- Physiologic condition in some fetuses in late gestation >32 weeks
- Transient RV > LV seen before 18 weeks (i.e., trisomy 21)
- Interruption of aortic arch
- Atrioventricular septal defect (unbalanced) with small left ventricle
- Mitral atresia with ventricular septal defect
- Total anomalous pulmonary venous connection
- Left superior vena cava (with or without coarctation)
- Double-outlet right ventricle
- Absent pulmonary valve syndrome
- Ebstein anomaly/tricuspid dysplasia
- Corrected transposition of the great arteries (left-sided smaller right ventricle misinterpreted as small left ventricle)
- Left congenital diaphragmatic hernia
- Peripheral arteriovenous fistula with RV volume overload (vein of Galen aneurysm and others)
- Severe tricuspid insufficiency in various fetal conditions
- Aneurysm of the foramen ovale

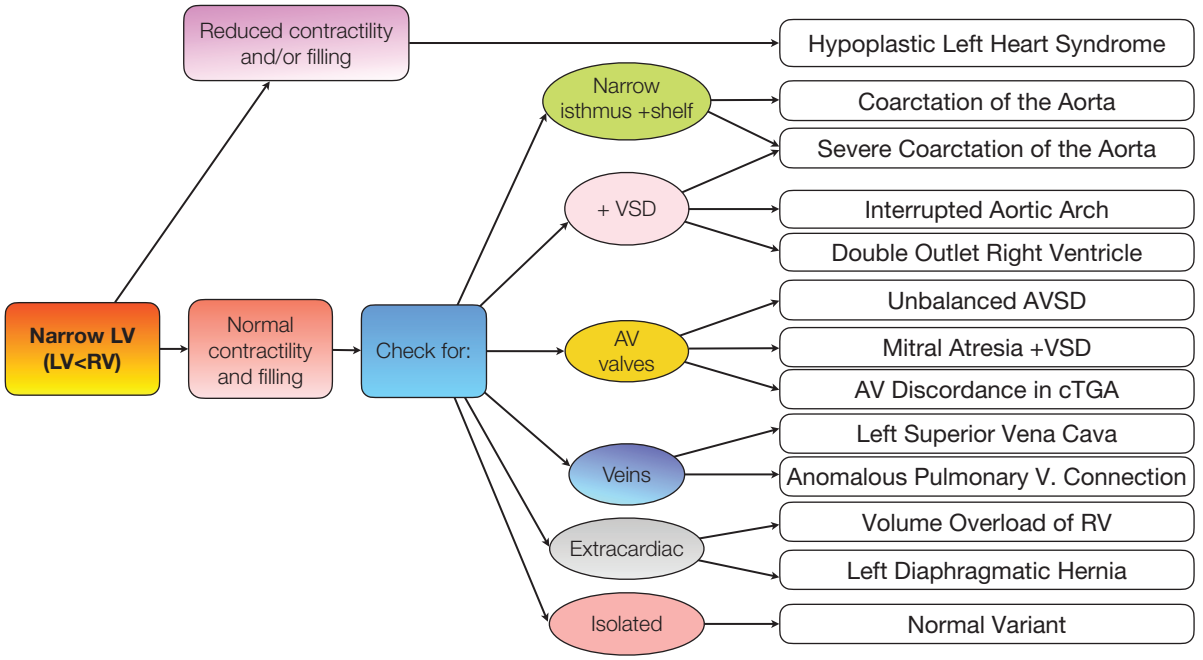

Figure 33.22: Systematic workup and differential diagnosis of ventricular disproportion with a small left ventricle (LV) in comparison to the right ventricle (RV). Keep in mind that some diagnoses are not exclusive but can occur together, such as in aortic coarctation with a left superior vena cava (LSVC) or in combination with an unbalanced atrioventricular septal defect (AVSD). AV, atrioventricular; cTGA, corrected transposition of the great arteries; V, venous; VSD, ventricular septal defect.

The presence of fetal ventricular size disproportion in the absence of coarctation of the aorta postnatally seems to have increased associated morbidity. In a study involving 46 fetuses with such findings, postnatal course of fetuses with ventricular size disproportion was complicated by prenatally undetected congenital defects in 46% and pulmonary or transition problems in 35% of neonates. Proper fetal monitoring and incorporating the risks for additional morbidity and neonatal complications in prenatal counseling is, therefore, warranted (33).

Prognosis and Outcome

The in utero course of a fetus with coarctation of the aorta is generally uneventful. We recommend serial ultrasound examinations every 4 to 6 weeks to observe the development of the transverse arch and the progression of the coarctation. Prenatal diagnosis of aortic coarctation should lead to the delivery of the neonate at a tertiary center with immediate availability of neonatology and pediatric cardiology services. Prostaglandin infusion should be commenced immediately after delivery to maintain ductal patency. Prenatal diagnosis of coarctation of the aorta has been shown to improve neonatal outcome (34).

Long-term studies of outcome following prenatal diagnosis of coarctation of the aorta and surgical repair in infancy are not currently available. Available data on long-term follow-up, however, show that chronic hypertension, surgery site complications (aneurysm, stricture), and coronary artery disease play a significant role in the long-term outcome (35,36). Following a successful repair, lifelong ongoing surveillance of the coarctation site, ascending aorta, aortic valve, blood pressure, and left ventricular function is thus required (35). Overall, the prognosis following successful repair of simple coarctation without extracardiac anomalies in childhood should be considered excellent.

Similar to other cardiac lesions, the prognosis of coarctation of the aorta when detected prenatally seems to be worse than reported in postnatal series, probably due to selection bias and associated malformations (27). An overall adjusted survival rate of 79% was reported in a prenatal series after excluding pregnancy terminations and extracardiac and chromosomal abnormalities (27). In a study comparing neonatal outcome between the prenatal versus the postnatal diagnosis of aortic coarctation, the neonates with prenatally diagnosed coarctation had smaller left heart structures than the neonates with coarctation diagnosed after the first week of age, were more likely to require extensive arch reconstruction under cardiopulmonary bypass, and had longer hospital stays (37). The presence of fetal growth restriction had a significant negative effect on survival (27). Complex coarctation, with its association with other cardiac anomalies, has a worse prognosis.

Approach to Diagnosis

Figure 33.23 displays the systematic approach to diagnosis in a fetus with suspected coarctation of the aorta.

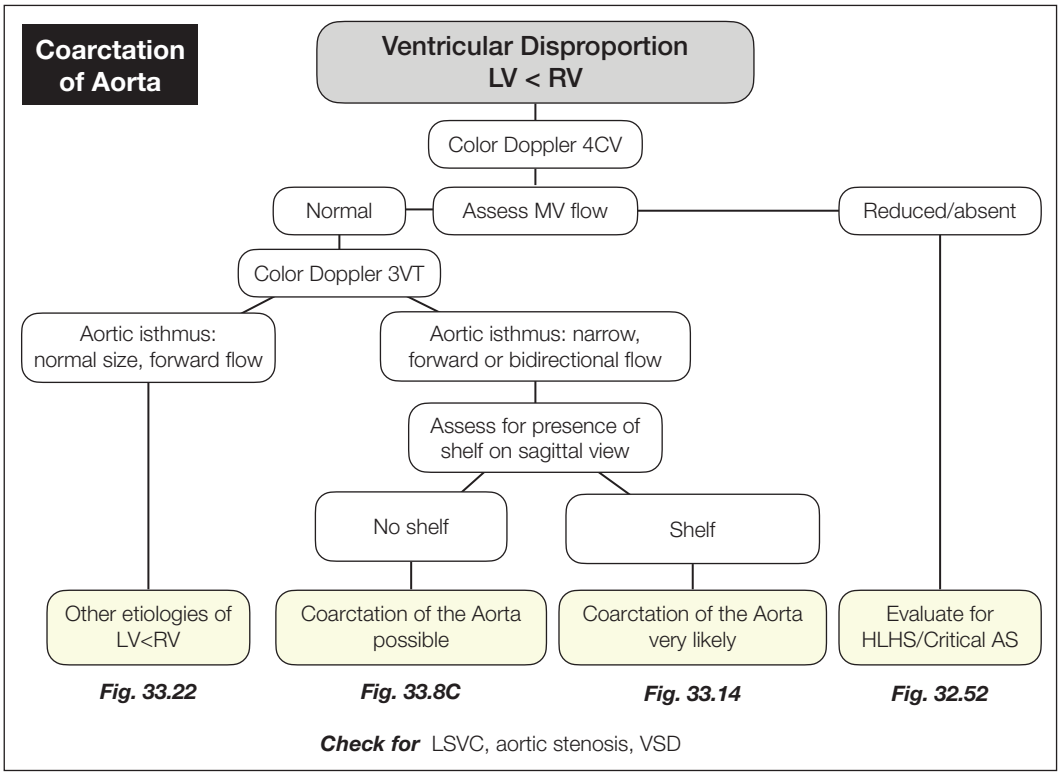

Figure 33.23: Approach to diagnosis in a fetus with suspected coarctation of the aorta. See text for details. 3VT, three-vessel-trachea; 4CV, four-chamber view; AS, aortic stenosis; HLHS, hypoplastic left heart syndrome; LSVC, left superior vena cava; LV, left ventricle; MV, mitral valve; RV, right ventricle; VSD, ventricular septal defect.

KEY POINTS ■ Coarctation of the Aorta

- Coarctation of the aorta involves narrowing of the aortic arch, typically located at the isthmic region, between the left subclavian artery and the ductus arteriosus.
- When a long segment of the aortic arch is narrowed, the descriptive term is tubular hypoplasia of the aortic arch.
- In aortic coarctation, the four-chamber view is abnormal, where the left ventricle appears narrow when compared to the right ventricle.
- The three-vessel-trachea view, depicting great vessel disproportion, is more specific than ventricular disproportion alone on a four-chamber view in the diagnosis of aortic coarctation.
- Longitudinal view of the aortic arch demonstrates tortuosity and narrowing between the left subclavian artery and the origin of the ductus arteriosus (coarctation shelf) in aortic coarctation.
- In aortic coarctation, associated cardiac abnormalities are common, with a large ventricular septal defect representing the most common associated lesion.
- Various left-sided lesions are commonly associated with aortic coarctation, including bicuspid aortic valve, aortic stenosis at the valvular and subvalvular levels, and mitral stenosis.
- The presence of multiple left-sided obstructive cardiac lesions with coarctation of the aorta has been referred to as Shone syndrome.
- The presence of a persistent LSVC has been associated with coarctation of the aorta.
- Associated extracardiac malformations are common in aortic coarctation.
- Berry aneurysms of the cerebral circulation have been reported in up to 3% to 5% of patients with coarctation.
- In aortic coarctation, chromosomal abnormalities are common, with Turner syndrome representing the most common abnormality.
- Chronic hypertension, surgery site complications (aneurysm, stricture), and coronary artery disease play a significant role in the long-term outcome in coarctation of the aorta.