# Coarctation of the Aorta

# 21

## Contents


21.1 **Anatomy and Haemodynamics** ..... 283

21.2 **2D Echocardiography** ..... 285

21.3 **Colour Doppler Echocardiography** ..... 286

21.3.1 Coarctation of the Aorta in Neonates with a Patent Ductus Arteriosus ..... 287

21.3.2 Coarctation of the Aorta in a Neonates with Restrictive or Closed Ductus Arteriosus ..... 288

21.3.3 Coarctation of the Aorta in Older Children ..... 288

21.4 **Pulsed Wave and Continuous Wave Doppler** ..... 290

21.4.1 Flow Measurements of the Aorta ..... 290

21.4.2 Doppler Flow Measurements in Systemic Arteries ..... 293

**References** ..... 295


## 21.1 Anatomy and Haemodynamics

Obstruction of the aortic arch may present as discrete coarctation, tubular hypoplasia of aortic arch segments, aortic atresia or interruption of the aortic arch. This chapter deals with discrete coarctation and tubular hypoplasia, while interruption of the aortic arch and aortic arch atresia are addressed in Chap. 22.

Coarctation of the aorta describes a circumscribed stenosis, which is typically located close to the junction of the ductus arteriosus with the descending aorta. The substrate of the stenosis is an intraluminal shelf that frequently contains ductal tissue extending into the aortic wall (Elzenga and Gittenberger-de Groot 1983; Elzenga et al. 1986; Ho and Anderson 1979). The intraluminal shelf is often accompanied by some infolding of the aortic lumen. In older children the obstruction is frequently aggravated by intimal proliferation (Elzenga and Gittenberger-de Groot 1983). According to the exact location of the stenosis in relation to the ductal insertion, the obstruction is classified as preductal, paraductal (juxtaductal) or postductal coarctation (Fig. 21.1). Most coarctations presenting in the neonatal period are preductal coarctations, while a gradual shift of the stenosis from a preductal to a postductal position is noted in older patients (Elzenga and Gittenberger-de Groot 1983). Coarctation is frequently associated with gradual tapering of the aortic diameter in the isthmus, the aortic region between the origin of the left subclavian artery and the insertion of the ductus arteriosus. Morphologically discrete coarctations have to be distinguished from tubular hypoplasia referring to tubular narrowing of a distinct area of the arch. The latter is much less frequent although it may coexist with discrete coarctation (Ho and Anderson 1979).

The haemodynamic effects of coarctation are determined by the degree of the stenosis, patency of the ductus arteriosus or timing of its closure, formation of collateral circulation and last but not least presence of associated cardiovascular anomalies. In neonates with severe coarctation, spontaneous closure of the ductus will result in critical

**Electronic supplementary material** The online version of this chapter (doi:10.1007/978-3-319-42919-9_21) contains supplementary material, which is available to authorized users.



M. Hofbeck et al., *Doppler Echocardiography in Infancy and Childhood*, DOI 10.1007/978-3-319-42919-9_21

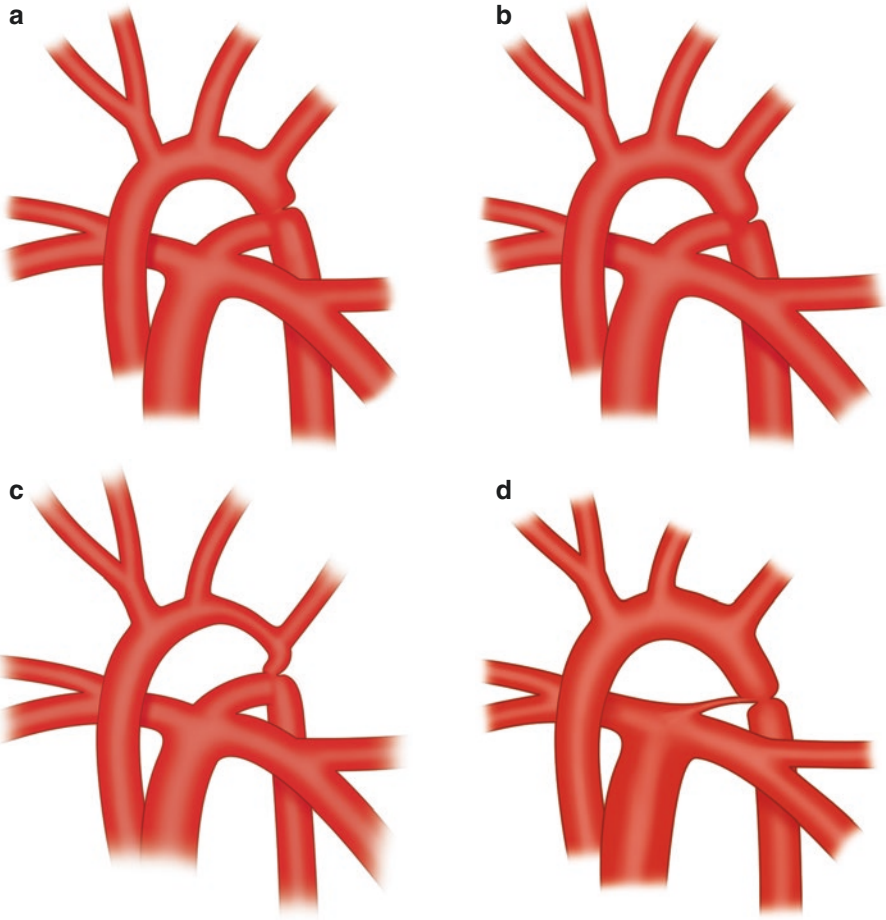


**Fig. 21.1** Aortic coarctation can be described according to the location of the obstruction relative to the ductus arteriosus: in preductal coarctation the obstruction is localized proximal (**a**); in juxtaductal coarctation the obstruction is localized just opposite to the aortic end of the ductus arteriosus (**b**). In neonates aortic coarctation is frequently associated with tubular hypoplasia of parts of the aortic arch (**c**). In older children and in adolescents, the ductus arteriosus is usually closed (**d**). When the coarctation is localized distal to the aortic insertion of the ductal ligament, it is classified as postductal coarctation (**d**)

reduction of perfusion of the descending aorta. Children become symptomatic with reduced pulses and blood pressure on the lower limbs, impaired renal and hepatic function as well as reduced intestinal perfusion. The clinical picture with depressed cardiovascular function, coagulation disorder due to hepatic dysfunction, disseminated intravascular coagulation and renal failure frequently resembles sepsis, which is the primary differential diagnosis. Infants with severe coarctation often develop pulmonary hypertension resulting in right ventricular dilatation and tricuspid regurgitation. They may present with severe congestive heart failure due to reduced right and left ventricular function. Severe coarctation remains a cardiovascular malformation associated with significant morbidity and mortality in the neonatal period (Fesseha et al. 2005; Franklin et al. 2002). Less commonly infants with isolated severe coarctation may present beyond the neonatal period with severely decreased left ventricular function (Wiegand et al. 2015). Therefore coarctation of the aorta has to be included in the differential diagnosis of possible causes of dilated cardiomyopathy.

If coarctation of the aorta is less severe and if stenosis progresses slowly, children and adolescents frequently remain asymptomatic (Ing et al. 1996). Slow progression of the severity of coarctation in older children allows formation of collateral vessels providing adequate perfusion of the lower part of the body (Perloff 2010). This applies even in the presence of subtotal stenosis or acquired atresia of the isthmic region (Eicken et al. 2008; Glancy and Roberts 2002). These patients however develop brachiocephalic hypertension in the prestenotic vascular bed (Eicken et al. 2006; Ing et al. 1996; Perloff 2010). Symptoms and

complications in these patients are sequelae of brachiocephalic hypertension resulting in premature arteriosclerotic changes of the ascending aorta, coronary arteries and cerebral vessels (Perloff 2010). Therefore patients with isolated coarctation are at risk to experience myocardial infarction, rupture or dissection of the aorta and cerebrovascular events at a relatively young age (Perloff 2010). In the majority of children and adolescents with isolated coarctation, the left ventricle will be able to cope with the requirement of brachiocephalic hypertension, and cardiac examination reveals only mild or moderate concentric left ventricular hypertrophy.

Coarctation of the aorta is frequently associated with cardiovascular anomalies, especially if patients present in the neonatal period (Shinebourne and Elseed 1974). Associated anomalies include ventricular septal defects, patent ductus arteriosus, hypoplasia of the aortic arch, bicuspid aortic valve, aortic stenosis and mitral valve anomalies (Bove et al. 1993; Brouwer et al. 1996; Shone et al. 1963; Teo et al. 2011). Coarctation of the aorta can be also encountered in complex malformations such as transposition of the great arteries, double outlet right ventricle and functionally univentricular hearts. On the other hand, coarctation of the aorta is rarely associated with right-sided obstructive lesions (e.g. pulmonary stenosis, tetralogy of Fallot) and right aortic arch (Ismat et al. 2006; Perdreau et al. 2014; Teo et al. 2011). In the German PAN study, coarctation accounted for 3.6 % of infants diagnosed with congenital heart disease (Lindinger et al. 2010).

## 21.2 2D Echocardiography

Since coarctation of the aorta is frequently associated with other congenital cardiovascular anomalies, evaluation of the cardiac anatomy requires a systematic approach including evaluation of the cardiac connections, size of cardiac chambers and evaluation for possible valvular anomalies.

Severe isolated coarctation in the neonatal period can cause both left heart and right heart failure resulting in pulmonary hypertension and right ventricular dilatation (Fig. 21.2; Videos 21.01, 21.02 and 21.03). *Dilatation of the right atrium, right ventricle and pulmonary artery in a newborn should always prompt careful search for possible coarctation.*

Imaging of the aortic arch can be achieved from a high right parasternal window as well as from the suprasternal notch (Smallhorn et al. 1983; Smallhorn et al. 1982). Extension of the child's neck by elevation of the shoulders (e.g. by a small towel) is extremely helpful to improve imaging quality in these planes. Evaluation of the aortic arch should start in the suprasternal or parasternal short axis of the ascending aorta with determination of the laterality of the aortic arch (see Chaps. 22 and 23). It will be a rare exception to find coarctation of the aorta in the presence of right aortic arch (Ismat et al. 2006).

The best imaging plane for visualization of coarctation is the suprasternal long-axis view obtained from the suprasternal notch (Smallhorn et al. 1983). In newborns the aortic arch can be displayed form the right parasternal view as well, since the ribs and the sternum are not yet ossified at this age (Marek et al. 2009). The suprasternal long-axis view of the aortic arch is important to describe the origin of the head and neck arteries and to detect possible hypoplasia of segments of the aortic arch proximal to the isthmic region (Fig. 21.3; Videos 21.04, 21.05 and 21.06). Most frequently hypoplasia will be present distal to the origin of the left common carotid artery. Stenoses that are located distally may be missed in the long-axis view of the aortic arch however (Video 21.07). The "ductal view", obtained from a left parasternal sagittal window, is the best approach for visualization of the distal aortic arch, the aortic isthmus and the proximal descending aorta (Lai et al. 2006). Therefore the ductal view is a good plane to visualize the shelf of circumscribed coarctations and to describe the morphology of the isthmic region (Fig. 21.4; Videos 21.08 and 21.09).

Two-dimensional echocardiography should focus to clarify the anatomy of the aortic arch and the isthmus. Echocardiographic diagnosis of coarctation requires optimal imaging conditions (including sedation of uncooperative patients and elevation of the shoulders) and optimal settings of the echo machine to visualize the stenosis. Assessment of the aortic arch should include description of the origin of the brachiocephalic vessels; measurement of the aortic diameters at the origin of the innominate artery, the transverse arch, at the level of the left subclavian artery and isthmus; as well as visualization of the shelf of a possible circumscribed coarctation (Fig. 21.5; Videos 21.08 and 21.09). The localization of the stenosis in relation to the ductus arteriosus is described defining the stenosis as preductal, juxtaductal (paraductal) or postductal (Video 21.10).

*Since a significant gradient across the aortic isthmus may be absent in the presence of a patent ductus arteriosus or in patients with reduced left ventricular function, diagnosis of aortic coarctation should not rely on detection of the stenosis by Doppler interrogation alone (*Videos 21.11 and 21.12).

In rare cases coarctation is complicated by aberrant origin of the right subclavian artery from the descending aorta. The aberrant subclavian artery usually takes its origin just below the coarctation or from the area of isthmic stenosis. *Preoperative detection of aberrant subclavian artery origin is extremely important in the planning of the surgical procedure:* anomalies of the right subclavian artery affect intraoperative invasive blood pressure measurements obtained by a right radial artery line, especially if the aberrant subclavian

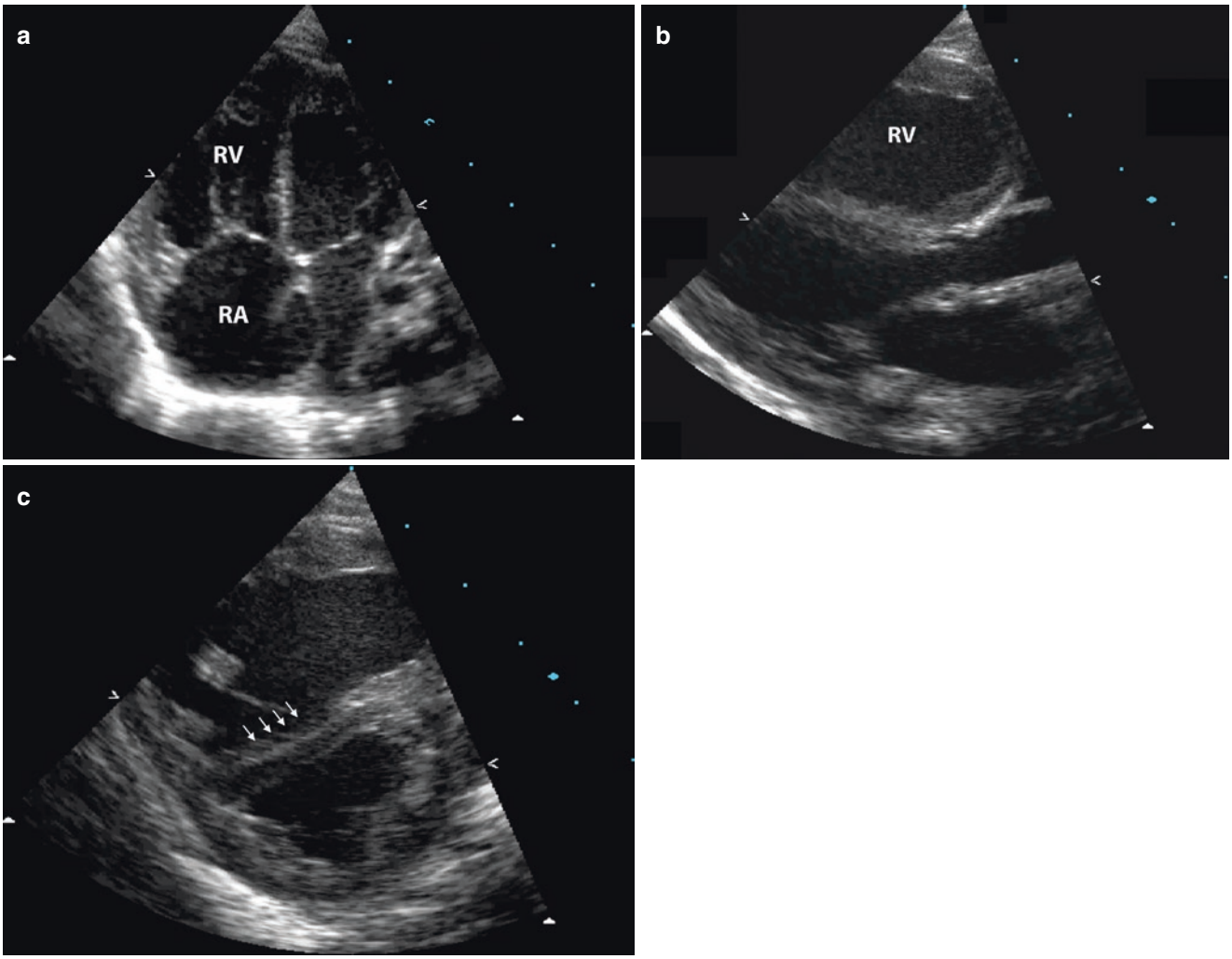


**Fig. 21.2** The apical four-chamber view (**a**) and the parasternal long-axis view (**b**) in a newborn with isolated severe coarctation show significant dilatation of right atrium (*RA*) and right ventricle (*RV*). Leftward shift of the ventricular septum (*arrows*), indicating elevation of right ventricular pressure, is apparent in the parasternal short-axis view (**c**)

artery originates distal of the stenosis. Furthermore unexpected aberrant origin of the subclavian artery from the descending aorta confronts the surgeon with an additional vessel originating from the isthmic region that complicates correct identification of the vascular structures and prolongs the surgical procedure. In the worst case, this may prolong cross-clamping time of the aorta increasing the risk of spinal damage. Presently the majority of neonates with coarctation are sent to the operating room based on the findings of echocardiography. *Therefore anomalies of the right subclavian artery, such as aberrant origin from the descending aorta, should be excluded in the preoperative assessment of any patient with coarctation of the aorta by careful investigation of the innominate artery.* Visualization of bifurcation of the innominate artery into the right subclavian and right common carotid artery excludes subclavian artery anomalies (see also Chaps. 22 and 23). The rare cases of coarctation in the presence of right aortic arch are frequently associated with anomalies of the left subclavian artery (Ismat et al. 2006; Perdreau et al. 2014).

Echocardiography including pulsed and continuous wave Doppler is the most important tool in the evaluation of the postoperative result following surgical correction of coarctation in neonates and infants. 2D echocardiography allows visualization of the anastomosis following resection of coarctation as well as assessment of residual hypoplasia of aortic arch segments (Fig. 21.6; Videos 21.13 and 21.14).

## 21.3 Colour Doppler Echocardiography

Colour Doppler echocardiography is very helpful for the diagnosis of coarctation improving visualization of the aortic arch and origin of the brachiocephalic vessels, visualization

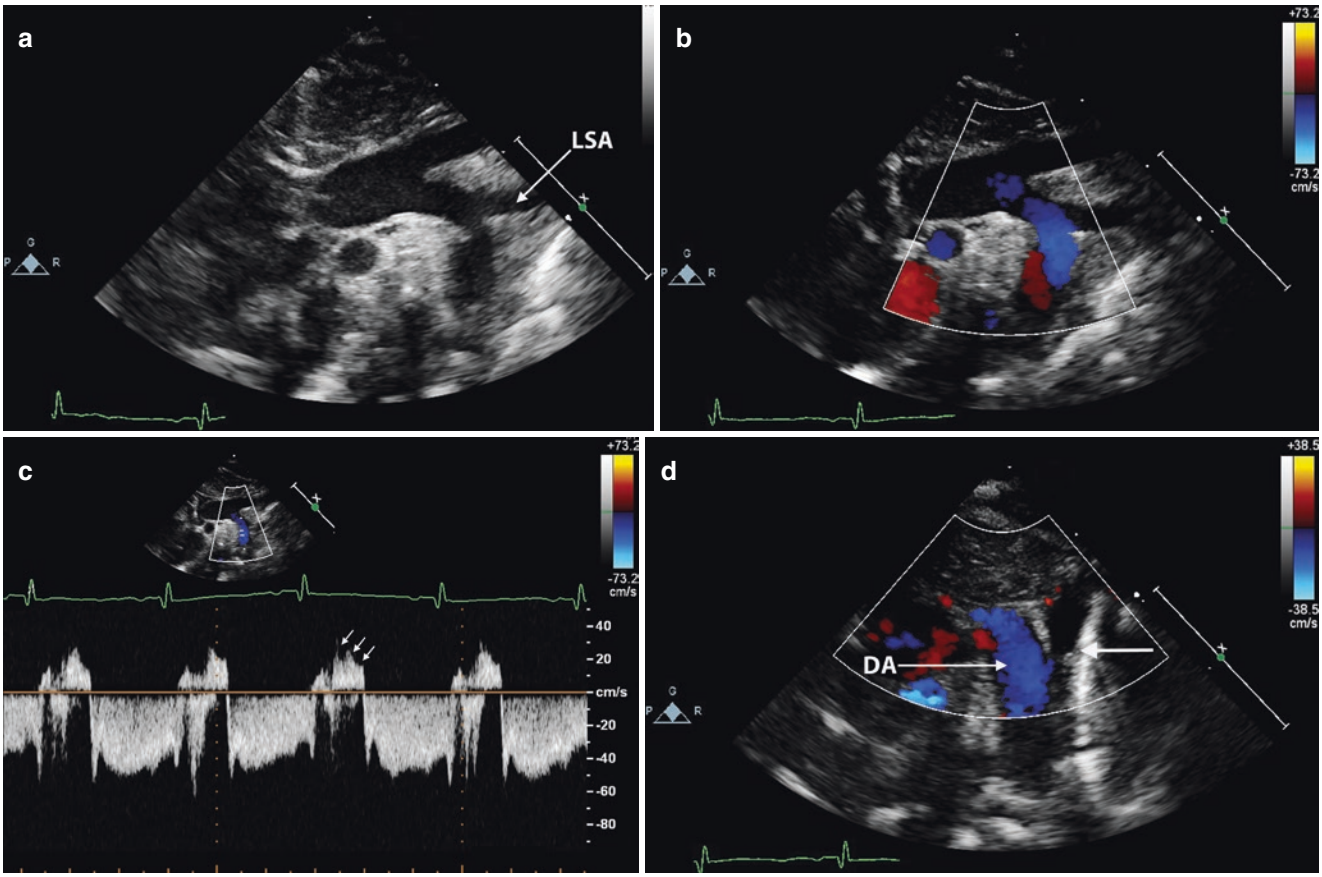


**Fig. 21.3** In a newborn with coarctation and widely patent ductus arteriosus, the long-axis view of the aortic arch (**a**) shows narrowing of the aorta distal to the left subclavian artery (*LSA*). Colour Doppler fails to show acceleration of flow in the aortic isthmus (**b**). PW Doppler reveals abnormal flow in the isthmus with reverse flow in systole (*arrows*) and accelerated antegrade flow in diastole (**c**). The ductal view (**d**) shows the large ductus arteriosus (*DA*) and significant narrowing of the aortic isthmus (*arrow*)

of hypoplastic segments and identification of circumscribed stenoses. However colour Doppler has some significant limitations and shortcuts and requires careful interpretation in different haemodynamic situations.

### 21.3.1 Coarctation of the Aorta in Neonates with a Patent Ductus Arteriosus

In the presence of a widely patent ductus arteriosus, colour Doppler frequently reveals no acceleration and no variance across a hypoplastic aortic arch segment or a circumscribed coarctation (Figs. 21.3 and 21.5; Videos 21.11 and 21.12). This is due to the fact that the neonatal right ventricle, in the presence of a widely patent ductus arteriosus, is able to maintain systemic pressure in the descending aorta. Therefore no pressure gradient is present between the aortic arch and the descending aorta. *As long as the ductus arteriosus is widely patent, absence of accelerated flow during colour Doppler interrogation is completely insufficient to exclude severe coarctation*. In these cases diagnosis of possible coarctation has to rely on meticulous examination of the isthmic region by 2D echo. Flow patterns in the ductus arteriosus depend on the severity of coarctation as well as on the diameter of the ductus. If the ductus is widely open in the neonate, it will exhibit bidirectional shunting. This consists of right to left shunt during systole and left to right shunt during diastole reflecting the fact that the right ventricle during systole contributes to the perfusion of the descending aorta (Figs. 21.5 and 21.7; Video 21.12). Doppler interrogation of the ductus arteriosus should include frame-by-frame analysis of colour Doppler as well as spectral and continuous Doppler interrogation. The "ductal view" is ideal both to visualize or to exclude a patent ductus arteriosus and to perform Doppler interrogation of the ductus if patent.

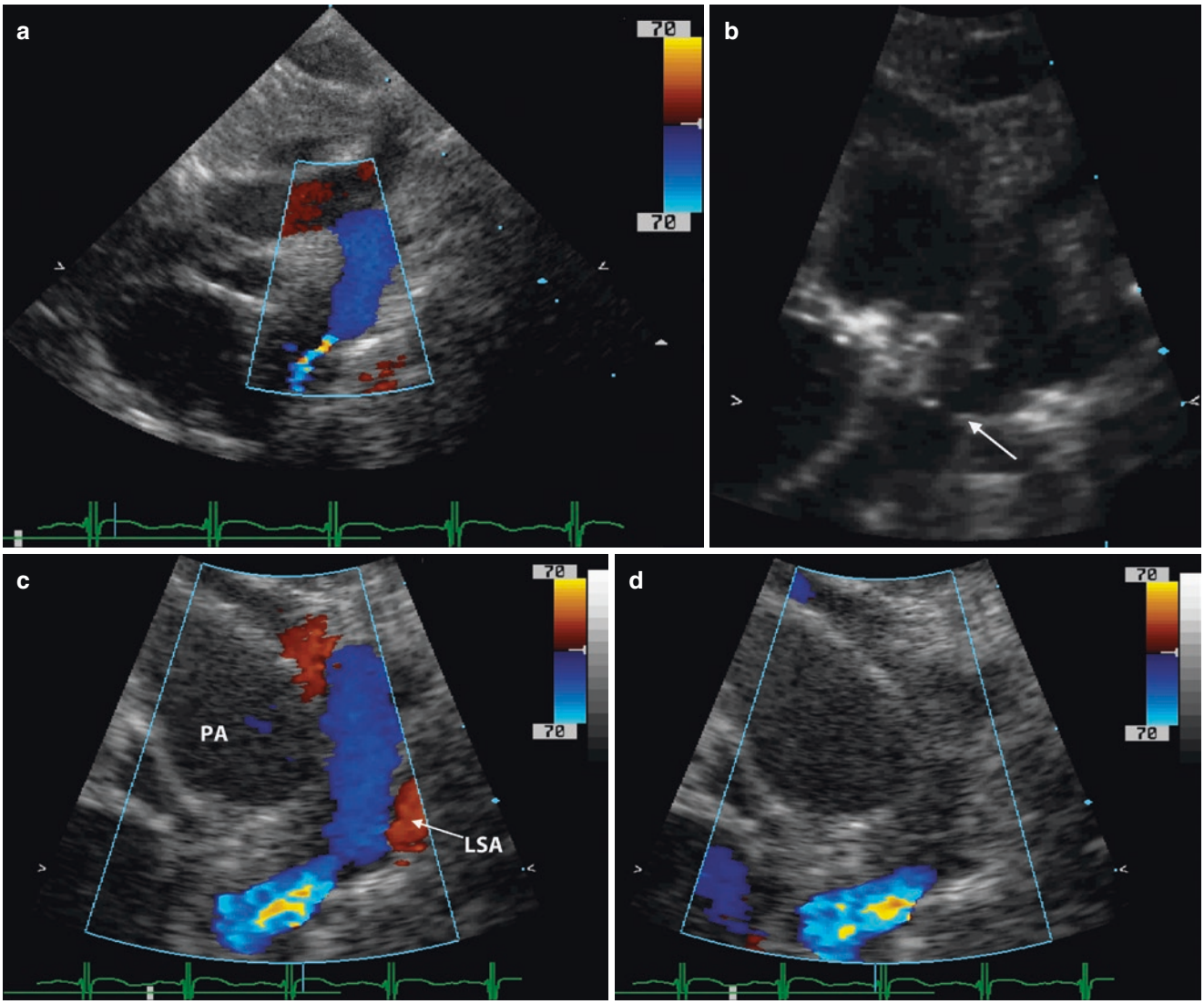


**Fig. 21.4** Colour Doppler in the suprasternal long-axis view of a newborn shows a very tight distal coarctation with minimal residual flow (**a**). Magnification of the aortic isthmus in the ductal view (**b**) reveals the circumscribed shelf of coarctation (*arrow*). Colour Doppler in systole (**c**) and diastole (**d**) reveals continuous systolic-diastolic flow across the obstruction. *PA* pulmonary artery, *LSA* left subclavian artery

### 21.3.2 Coarctation of the Aorta in Neonates with Restrictive or Closed Ductus Arteriosus

In the presence of severe coarctation, perfusion of the descending aorta depends on patency of the ductus arteriosus. Constriction of the ductus in this situation results in an increasing velocity of the right to left shunt due to an increasing pressure gradient between the pulmonary artery and the descending aorta. Colour Doppler analysis in the ductal view shows variance and aliasing during systolic right to left shunting. If the ductus arteriosus is closed, colour Doppler reveals accelerated systolic and diastolic flow across the coarctation as well as variance at the site of obstruction (Fig. 21.4; Videos 21.07 and 21.09). Visualization of the coarctation may be difficult if it is located quite distal to the left subclavian artery (Wiegand et al. 2015). Furthermore in children with critical stenosis and left ventricular failure, there may be only minimal flow across the coarctation, which makes it difficult to visualize the obstruction by colour Doppler (Fig. 21.4). In both situations, the obstruction may be missed in the suprasternal or parasternal long-axis view of the aortic arch (Wiegand et al. 2015). Visualization of distal coarctations is easier in the left parasternal long-axis plane ("ductal view") (Video 21.09).

### 21.3.3 Coarctation of the Aorta in Older Children

In older children with coarctation and a closed ductus, the haemodynamics are usually well adapted to the presence of the stenosis. The parasternal long- and short-axis views may

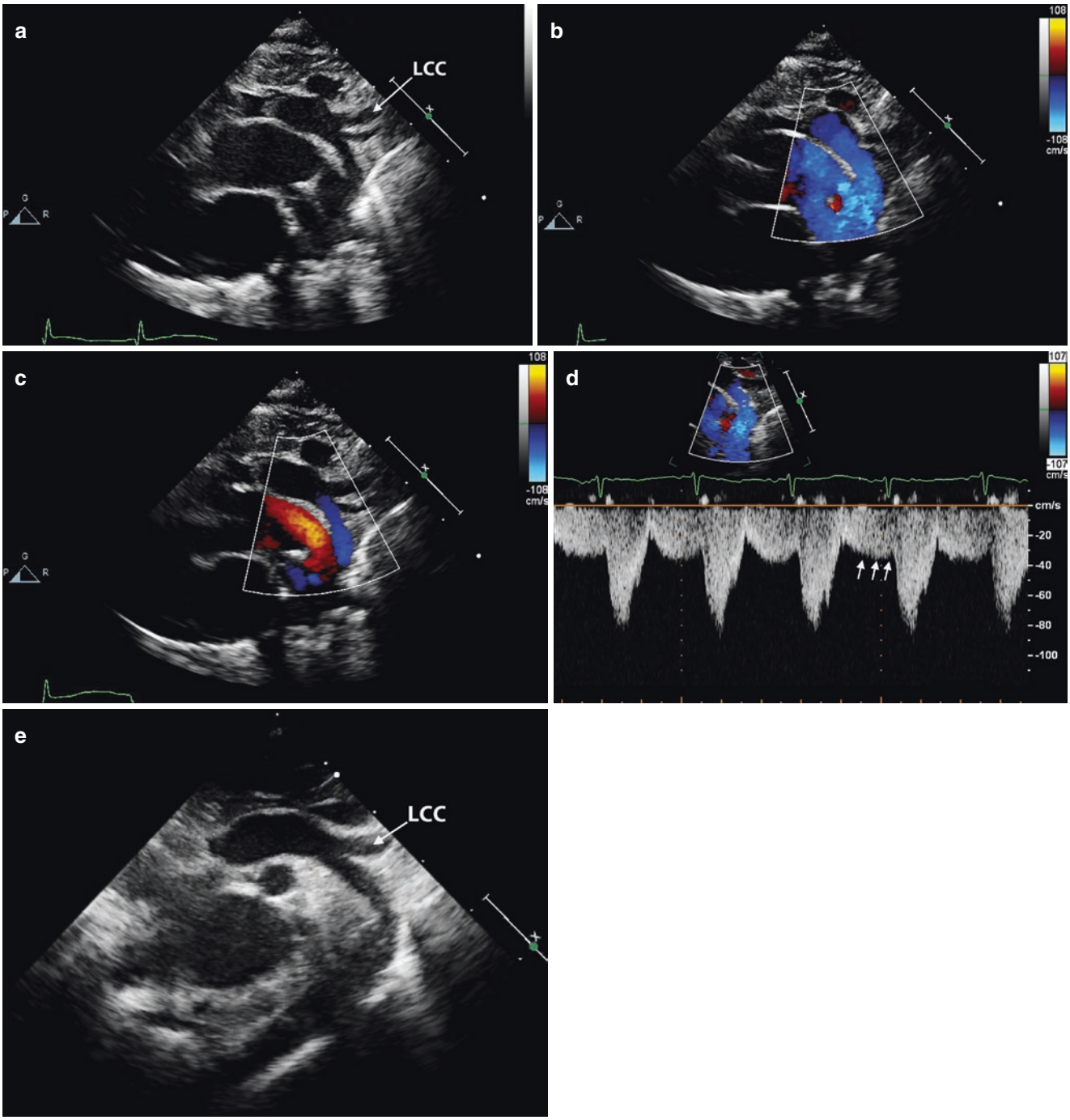


**Fig. 21.5** The suprasternal long-axis view (**a**) in a newborn with transposition and VSD shows hypoplasia of the aortic arch distal to the left common carotid artery (*LCC*). Colour Doppler displays laminar flow across the aortic arch and right to left shunting across the ductus arteriosus (DA) in systole (**b**), while there is left to right ductal shunting in diastole (**c**). PW Doppler of the distal arch (**d**) shows normal systolic flow velocity, while there is some increase in diastolic flow (*arrows*). The right parasternal long-axis view in another newborn (**e**) shows pronounced, long-segment hypoplasia of the aortic arch starting even proximal to the left common carotid artery (*LCC*)

be without signs of significant left ventricular hypertrophy even in the presence of significant stenosis (Video 21.15). The aortic arch in these patients frequently has normal dimensions in the prestenotic segments (Fig. 21.8; Videos 21.16 and 21.17). Since it is not uncommon that stenoses in children and adolescents are located quite distally, they may be difficult to detect by colour Doppler from the suprasternal window (Video 21.17). Frequently it is possible to obtain sufficient visualization of the stenosis in the "ductal view" (Fig. 21.9; Videos 21.18 and 21.19). To obtain adequate imaging in this view, it is necessary to place patients in a left oblique position, if they are older than 3 or 4 years of age. In older children, adolescents and adults, echocardiography may be unable to provide sufficient imaging of the complete

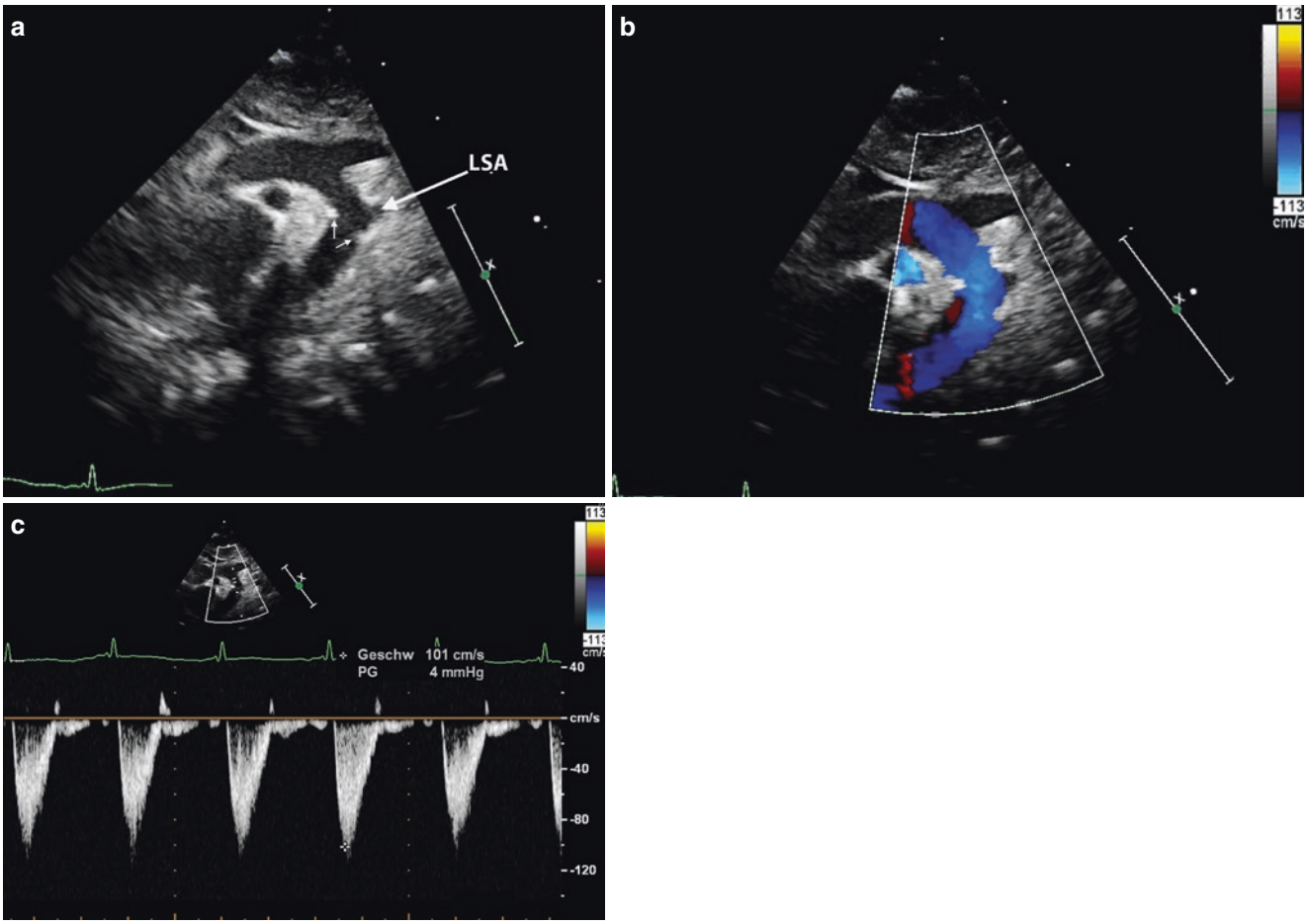


**Fig. 21.6** The suprasternal long-axis view in an infant shows a normal-size aortic arch following resection of coarctation and end-to-end anastomosis of the aorta (**a**). The sutures of the anastomosis (*arrows*) are visible distal to the left subclavian artery (*LSA*). Colour Doppler shows laminar flow (**b**), while normal flow velocity is confirmed by PW Doppler of the distal aortic arch (**c**)

anatomy. In these age groups, it is frequently necessary to complete the diagnosis by additional imaging modalities such as cardiac MRI or CT-thorax (Eichhorn and Ley 2007; Teo et al. 2011). These latter imaging techniques can display the complete anatomy of the aortic arch and descending aorta as well as collateral circulation (Fig. 21.10).

## 21.4 Pulsed Wave and Continuous Wave Doppler

In the diagnosis of coarctation in neonates and infants, both Doppler flow measurements of the aorta and of peripheral systemic arteries provide valuable information.

### 21.4.1 Flow Measurements of the Aorta

Doppler interrogation of the proximal and distal aortic arch as well as of the isthmic region can be performed from the high right subclavicular window and in the suprasternal long-axis view of the aortic arch. Flow velocities below 2–2.5 m/s can be measured by spectral Doppler. If velocities exceed 2–2.5 m/s, continuous wave Doppler has to be used for quantification of blood flow across the stenosis. Pulsed wave Doppler is advantageous as compared to CW Doppler, as it can measure blood flow at defined locations of the aortic arch with the possibility to detect circumscribed acceleration or abnormalities of flow (Figs. 21.3 and 21.5). For recording of flow in the distal aortic arch, the sample volume of the pulsed wave Doppler should be placed distal to the origin of the left subclavian artery. For quantification of flow across the coarctation, the sample volume should be placed in the stenosis, trying to get good alignment of the Doppler beam with the jet. In neonates with distally located coarctation and in older children, the suprasternal long-axis view may not be suitable to visualize the aortic isthmus well enough to allow good alignment of the Doppler beam with the jet of the stenosis. In these cases it may be advantageous to use the left parasternal sagittal plane ("ductal view"). Doppler interrogation of the aortic isthmus for verification or exclusion of coarctation in the neonatal period has to take

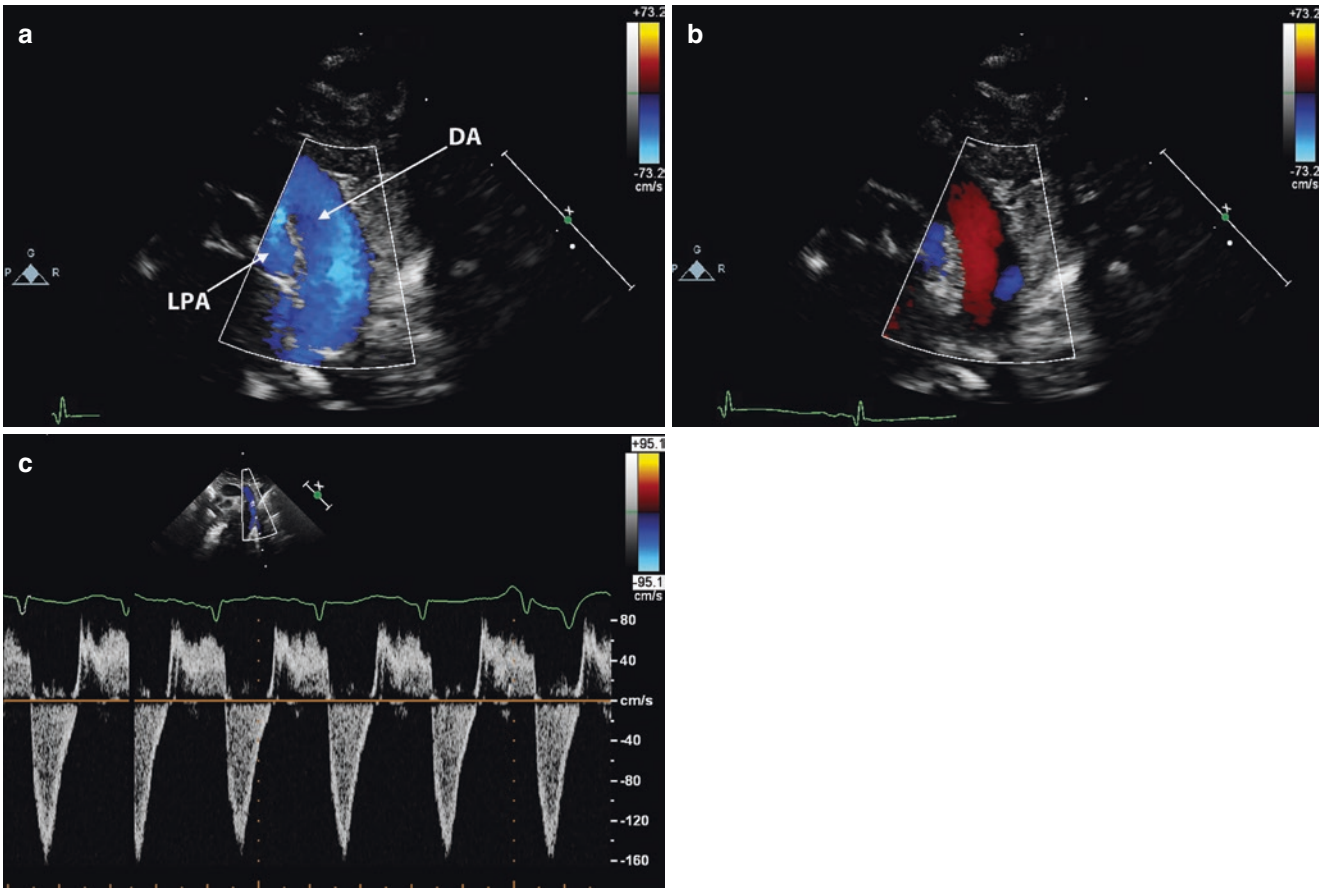


**Fig. 21.7** Colour Doppler in the ductal view of a neonate with severe coarctation and widely patent ductus arteriosus (*DA*) shows right to left ductal shunting in systole (**a**) and left to right shunting in diastole (**b**). Bidirectional flow is confirmed by PW Doppler interrogation of the ductus arteriosus (**c**). *LPA* left pulmonary artery

into consideration the limitations already addressed in the previous section.

- Neonates with a widely patent ductus arteriosus have equal blood pressures in the ascending and descending aorta associated with absence of a gradient across the stenosis. Therefore spectral Doppler will be unable to detect significant acceleration of blood flow across the isthmic region (Fig. 21.5). Nevertheless spectral Doppler examinations frequently are able to detect abnormalities of flow in the distal aortic arch. These abnormalities include a relative increase in diastolic flow velocities frequently associated with a relative decrease in systolic flow velocities (Figs. 21.3 and 21.5). Progressive closure of the ductus results in an increasing flow velocity and gradient over the ductus arteriosus as well as the coarctation. Doppler interrogation of the ductus arteriosus is best accomplished in the left parasternal long-axis ("ductal view"). A widely patent ductus shows bidirectional shunting with right to left shunting during systole and left to right shunting during diastole (Fig. 21.7).
- In neonates and infants with significant coarctation and closed ductus arteriosus, both the systolic and the diastolic pressures in the proximal aorta will exceed pressure in the descending aorta resulting in accelerated flow velocities both in systole and diastole. This results in a continuous flow pattern across the stenosis both in PW and CW Doppler interrogation. The maximum systolic velocity correlates with the severity of the stenosis. However this holds true only if the function of the left ventricle is well preserved. *Neonates with critical coarctation and severe left ventricular failure may be unable to generate a significant pressure gradient despite severe stenosis.* Furthermore if there is only very little flow across the stenosis, it may be difficult to record a reliable Doppler signal. Therefore optimal placement of the sample volume within the area of stenosis is mandatory in these patients. Neonates with severe coarctation frequently develop pulmonary hypertension. Doppler interrogation of tricuspid regurgitation is a valuable tool for the detection and quantification of pulmonary hypertension. If an adequate signal of tricuspid regurgitation is

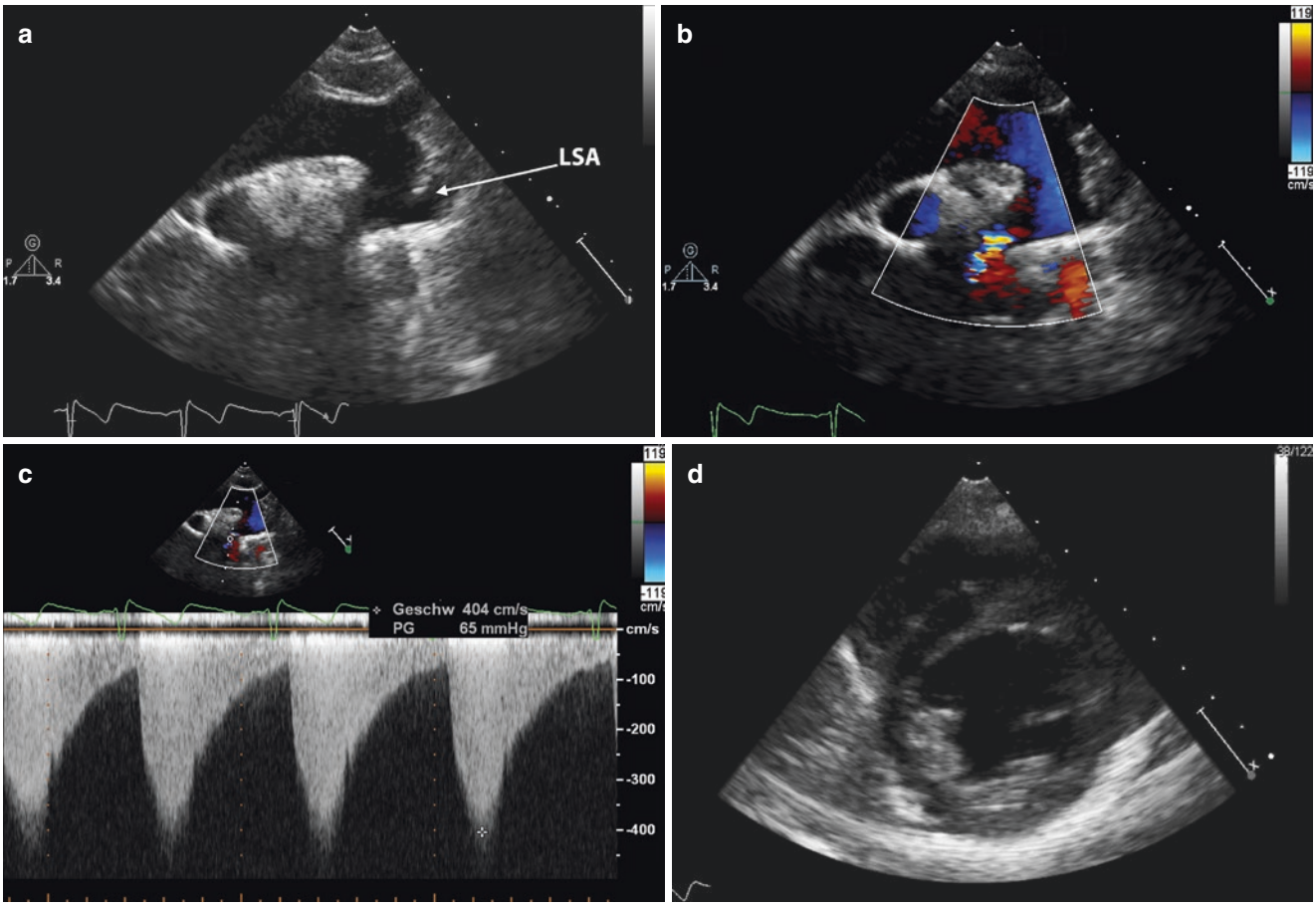


**Fig. 21.8** The suprasternal long-axis view in a 9-year-old patient (**a**) shows normal diameters of the aortic arch and severe coarctation distal to the left subclavian artery (*LSA*). Colour Doppler reveals acceleration of flow in the aortic isthmus due to severe coarctation, while the descending aorta is not well displayed (**b**). CW Doppler shows accelerated flow both in systole and diastole (**c**). The parasternal short-axis view does not reveal significant hypertrophy of the left ventricle (**d**)

displayed by pulsed wave or continuous wave Doppler, right ventricular systolic pressure can be determined based on the simplified Bernoulli equation (see Chap. 1).

- In older children Doppler interrogation of isthmic coarctation is sometimes difficult, since the stenosis may be located quite distally (Figs. 21.8 and 21.9). Since imaging of the isthmic region and the upper descending aorta becomes increasingly problematic with age, it may be difficult or impossible to visualize the area of maximal narrowing and to achieve optimal alignment of the Doppler beam with the isthmus and descending aorta. Doppler interrogation of the aortic isthmus can be attempted from the suprasternal notch and in the left parasternal sagittal view (Fig. 21.9). Imaging in the latter view ("ductal view") can be improved, if the patient is brought into a rather steep left lateral position.

In patients with coarctation beyond infancy, the ductus is usually closed or, if patent, it is without haemodynamic significance. In the presence of a severe stenosis, both systolic and diastolic blood pressures in the prestenotic aortic segment exceed pressures in the descending aorta. Therefore Doppler interrogation reveals both systolic and diastolic acceleration of flow due to the continuous pressure gradient throughout the cardiac cycle resulting in a typical sawtooth pattern across the stenosis (Figs. 21.8 and 21.9). Mild coarctations present with accelerated flow limited to systole (Fig. 21.11). Since flow velocities of severe coarctations beyond infancy usually exceed 2 m/s, Doppler interrogation requires CW Doppler. The peak instantaneous gradient across the stenosis can be calculated with the simplified Bernoulli equation (Snider et al. 1997). This results however in some overestimation of the maximal gradient as compared to the peak-to-peak gradient obtained at cardiac catheterization (Aldousany et al. 1990; Marx and Allen 1986). *If the simplified Bernoulli equation is applied, significant overestimation of the gradient has to be expected in the presence of accelerated flow in the prestenotic segment of the aorta.* This applies specifically for patients with left heart obstructive lesions (subaortic stenosis, aortic stenosis) or hypoplasia of

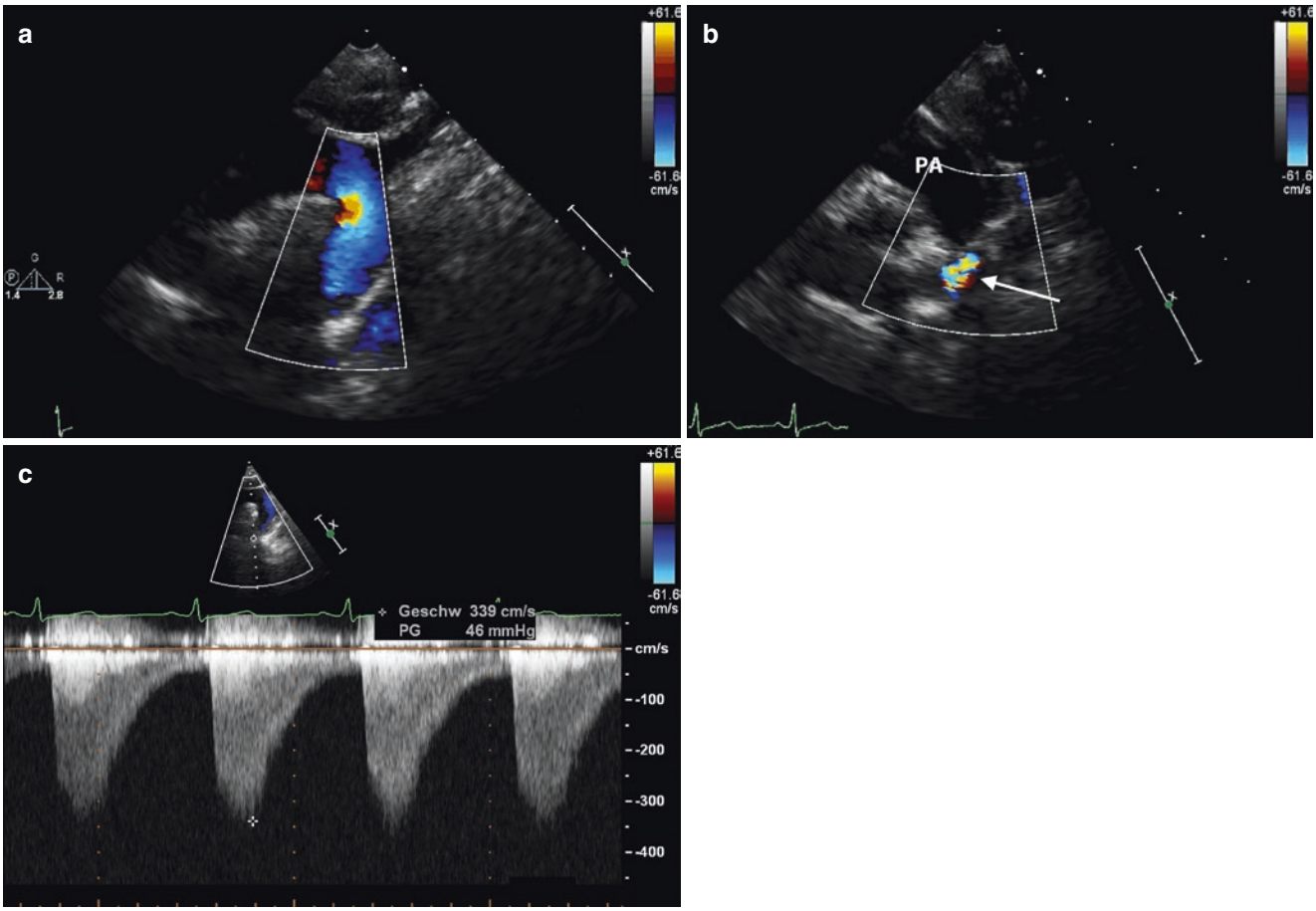


**Fig. 21.9** Colour Doppler in the suprasternal long-axis view of a 16-year-old patient fails to display isthmic coarctation (**a**). Acceleration of blood in the isthmus (*arrow*) due to severe coarctation becomes apparent in the ductal view (**b**), which was obtained with the patient in a left lateral position. Systolic and diastolic acceleration of flow is confirmed by CW Doppler (**c**). *PA* pulmonary artery

the proximal aortic arch (Snider et al. 1997). If the prestenotic velocity exceeds 1 m/s, overestimation of the gradient can be avoided by the use of the expanded Bernoulli equation.

$$\text{Gradient of coarctation} = 4\left(V_2^2 - V_1^2\right)$$

The systolic velocity proximal to the stenosis ($V_1$) is measured by PW Doppler and the systolic velocity across the stenosis ($V_2$) by CW Doppler. Application of the modified (expanded) Bernoulli equation results in a good correlation with invasive gradients obtained at cardiac catheterization (Aldousany et al. 1990; Marx and Allen 1986).

If the recorded Doppler gradient appears inadequately low, compared to appearance of the stenosis, two possibilities should be considered:

- In adolescents or young adults, severe coarctation may progress to complete atresia of the isthmus (Glancy and Roberts 2002). Without flow across the isthmus, there will be no gradient.

Adolescents and young adults with severe coarctation may develop significant collateral vessels, which are effective enough to reduce the pressure gradient across the isthmus.

### 21.4.2 Doppler Flow Measurements in Systemic Arteries

In neonates and young infants, the haemodynamic relevance of coarctation of the aorta can be assessed by Doppler flow measurements in pre- and poststenotic reference arteries (Deeg 2015): prestenotic reference arteries are the cerebral arteries such as the anterior cerebral artery; poststenotic reference arteries are abdominal arteries such as the coeliac trunk (Deeg 1989; Deeg et al. 1987).

The anterior cerebral artery can be measured in median sagittal sections in front of the third ventricle (Fig. 21.12). The anterior cerebral artery is one of the branches of the internal carotid artery which originates from the prestenotic part of the aorta. Pulsed wave Doppler interrogation reveals a pulsatile flow profile with high peak systolic flow

**Fig. 21.10** Cardiac MRI in a young adult shows previously undiagnosed severe coarctation of the aorta (*arrow*) distal to the origin of the left subclavian artery

velocities. The flow in the coeliac trunk can be measured either in sagittal sections through the midline or in coronal sections of the upper abdomen. The coeliac trunk originates as the first abdominal branch from the abdominal part of the descending aorta (Fig. 21.12). In healthy infants a pulsatile systolic-diastolic forward flow can be found. In the individual patient, peak systolic flow velocities in the coeliac trunk are always higher than peak systolic flow velocities in the anterior cerebral arteries (Deeg 2015).

In patients with coarctation, perfusion of the abdominal arteries decreases dramatically following closure of the ductus arteriosus (Deeg 1989; Deeg et al. 1987). The flow profile in the abdominal arteries is no longer pulsatile and changes into a venous-like flow (Fig. 21.12). The peak systolic flow velocities in the coeliac artery fall significantly under the peak systolic flow velocities in the anterior cerebral artery (Fig. 21.12). The decreased perfusion of the abdominal arteries may cause necrotizing enterocolitis, oliguria and anuria as well as disseminated intravascular coagulation. Surgical correction of the coarctation results in normalization of flow in the coeliac trunk, which can be used to confirm the haemodynamic success of the procedure (Fig. 21.12).

Reopening of the ductus arteriosus by administration of prostaglandin $E_1$, which is performed to stabilize critically ill neonates prior to surgery, results in improvement of flow to the descending aorta and normalization of systolic peak flow as well as time average flow velocities in the abdominal arteries (Deeg 2015). Diastolic flow velocities in this situation depend on pulmonary vascular resistance: immediately after birth in the presence of elevated pulmonary vascular resistance, diastolic flow velocities are normal. Decreasing pulmonary vascular resistance results in diastolic run-off from the aorta to the pulmonary artery (leakage of the aortic Windkessel). According to the amount of diastolic run-off, PW Doppler of systemic arteries shows marked decrease and, in more severe cases, even negative diastolic flow (Fig. 21.13).

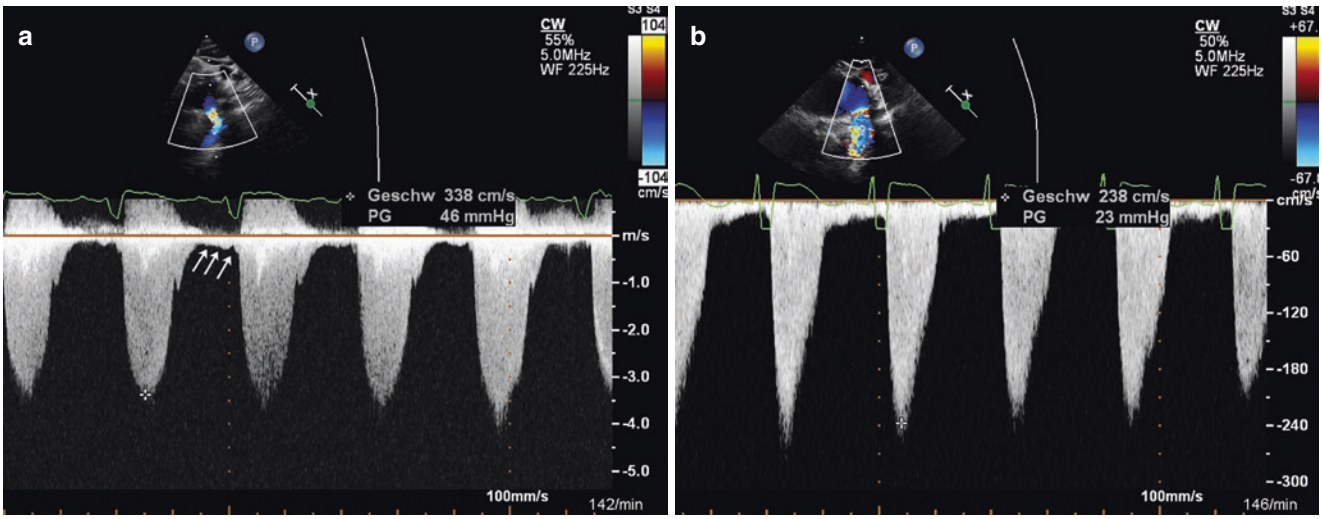


**Fig. 21.11** CW Doppler of the aortic isthmus in an infant following reconstruction of a hypoplastic aortic arch shows significant acceleration of systolic flow with a peak velocity of 338 cm/s (**a**). There is only mild acceleration of flow in diastole (*arrows*). Repeat examination immediately after balloon dilatation of moderate recoarctation with no residual gradient shows reduction of systolic flow velocities to 238 cm/s (**b**)

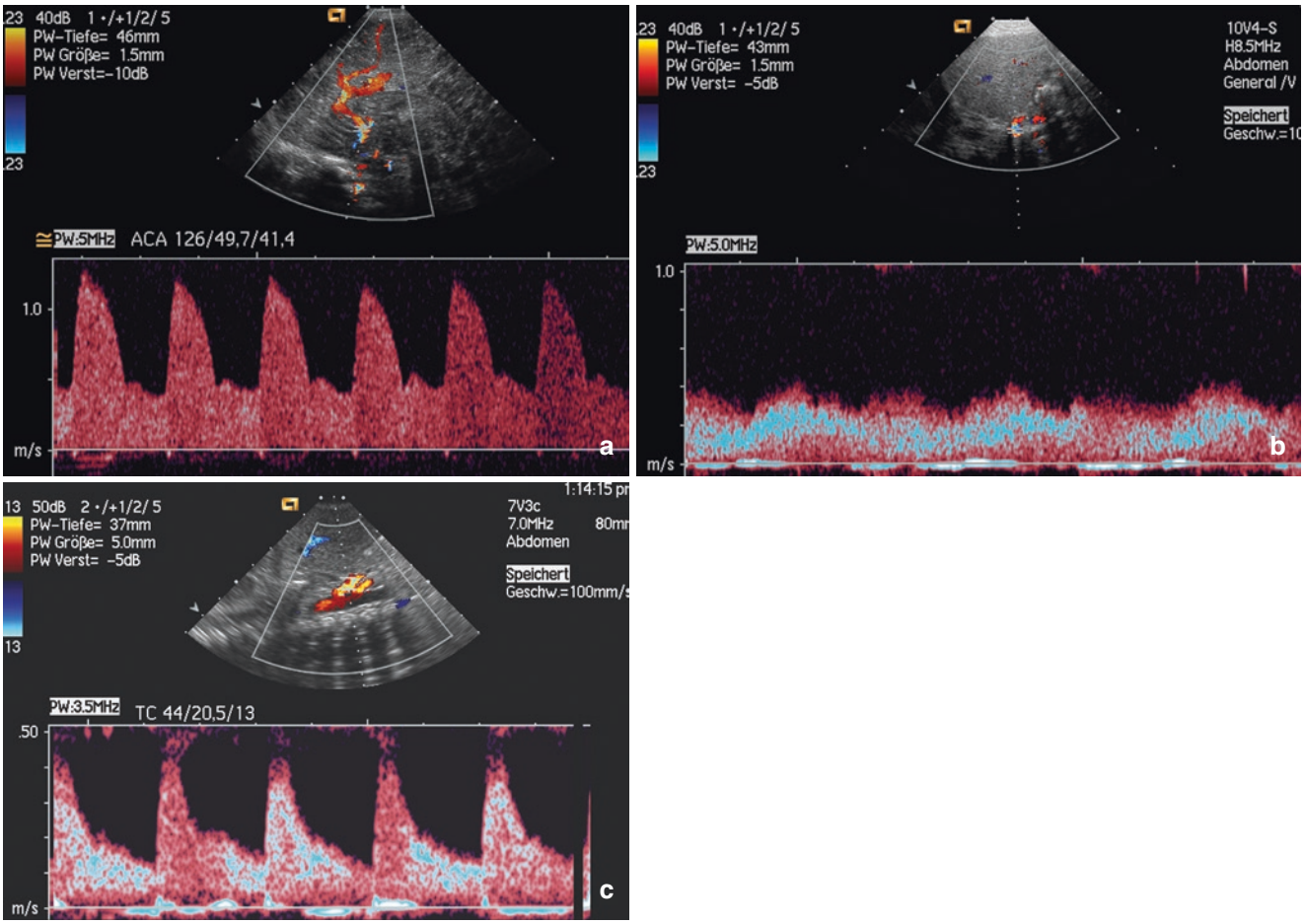


**Fig. 21.12** Doppler sonographic flow measurement in the anterior cerebral artery in a neonate with coarctation of the aorta following spontaneous closure of the ductus arteriosus (**a**). The sample volume of the pulsed Doppler is placed in the anterior cerebral artery in a midline sagittal section. The flow profile shows pulsatile flow with increased peak systolic flow velocity of 126 cm/s. Flow measurement in the coeliac trunk is performed in a midline sagittal section through the upper abdomen. Pulsed wave Doppler shows non-pulsatile, venous-like flow pattern with decreased peak systolic flow velocity (40 cm/s), which is significantly lower than the peak systolic flow velocity in the anterior cerebral artery (**b**). Repeat examination following operation reveals a pulsatile flow profile with increase of the peak systolic flow velocity to 44 cm/s and an increase of the time average flow velocity (**c**)

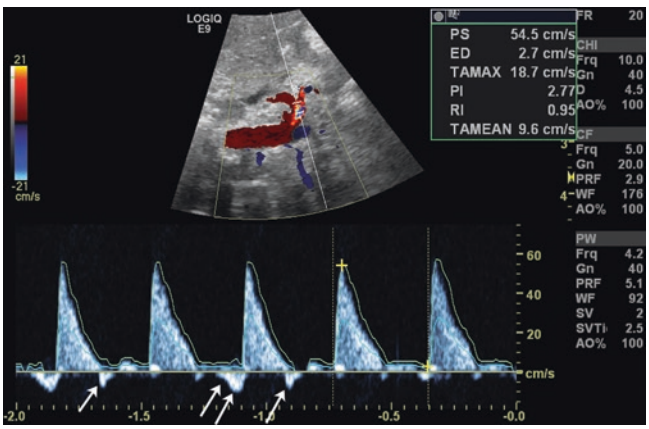


**Fig. 21.13** Doppler sonographic flow measurement in the coeliac trunk of a prematurely born infant with Down syndrome, atrioventricular septum defect, hypoplastic aortic arch, coarctation of the aorta and a widely patent ductus arteriosus. The flow spectrum shows a pulsatile flow with reduced, sometimes absent or even retrograde (*arrows*) diastolic flow and an elevated resistive index

## References

Aldousany AW, DiSessa TG et al (1990) Significance of the Doppler-derived gradient across a residual aortic coarctation. Pediatr Cardiol 11(1):8–14

Bove EL, Minich LL et al (1993) The management of severe subaortic stenosis, ventricular septal defect, and aortic arch obstruction in the neonate. J Thorac Cardiovasc Surg 105(2):289–295; discussion 295–6

Brouwer RM, Cromme-Dijkhuis AH et al (1996) Decision making for the surgical management of aortic coarctation associated with ventricular septal defect. J Thorac Cardiovasc Surg 111(1):168–175

Deeg KH (1989) Dopplersonographische flussmessungen bei herzerkrankungen. In: Deeg KH (ed) Zerebrale dopplersonographie im kinderalter. Springer, Berlin

Deeg KH (2015) Cardiovascular diseases which influence the flow in the extracardial arteries. In: Deeg KH, Rupprecht T, Hofbeck M (eds) Doppler sonography in infancy and childhood. Springer, Heidelberg/New York

Deeg KH, Wehner S et al (1987) Dopplersonographische Flussmessung in der Arteria cerebri anterior und im Truncus coeliacus bei Säuglingen mit Coarctationssyndrom im Vergleich zu gesunden Säuglingen. Klin Padiatr 199(6):411–423

Eichhorn JG, Ley S (2007) Angeborene Fehlbildungen der Aorta im Kindes- und Jugendalter. Radiologe 47(11):974–981

Eicken A, Pensl U et al (2006) The fate of systemic blood pressure in patients after effectively stented coarctation. Eur Heart J 27(9):1100–1105

Eicken A, Kaemmerer H et al (2008) Treatment of aortic isthmus atresia with a covered stent. Catheter Cardiovasc Interv 72(6):844–846

Elzenga NJ, Gittenberger-de Groot AC (1983) Localised coarctation of the aorta. An age dependent spectrum. Br Heart J 49(4):317–323

Elzenga NJ, Gittenberger-de Groot AC et al (1986) Coarctation and other obstructive aortic arch anomalies: their relationship to the ductus arteriosus. Int J Cardiol 13(3):289–308

Fesseha AK, Eidem BW et al (2005) Neonates with aortic coarctation and cardiogenic shock: presentation and outcomes. Ann Thorac Surg 79(5):1650–1655

Franklin O, Burch M et al (2002) Prenatal diagnosis of coarctation of the aorta improves survival and reduces morbidity. Heart 87(1):67–69

Glancy DL, Roberts WC (2002) Not congenital atresia of the aortic isthmus, but acquired complete occlusion in congenital aortic coarctation. Catheter Cardiovasc Interv 56(1):103–104; author reply 105

Ho SY, Anderson RH (1979) Coarctation, tubular hypoplasia, and the ductus arteriosus. Histological study of 35 specimens. Br Heart J 41(3):268–274

Ing FF, Starc TJ et al (1996) Early diagnosis of coarctation of the aorta in children: a continuing dilemma. Pediatrics 98(3 Pt 1):378–382

Ismat FA, Weinberg PM et al (2006) Right aortic arch and coarctation: a rare association. Congenit Heart Dis 1(5):217–223

Lai WW, Geva T et al (2006) Guidelines and standards for performance of a pediatric echocardiogram: a report from the Task Force of the Pediatric Council of the American Society of Echocardiography. J Am Soc Echocardiogr Off Publ Am Soc Echocardiogr 19(12):1413–1430

Lindinger A, Schwedler G et al (2010) Prevalence of congenital heart defects in newborns in Germany: results of the first registration year of the PAN Study (July 2006 to June 2007). Klin Padiatr 222(5): 321–326

Marek J, Fenton M et al (2009) Aortic arch anomalies: Coarctation of the aorta and interrupted aortic arch. In: Lai WW, Mertens LL, Cohen MS, Geva T (eds) Echocardiography in pediatric and congenital heart disease. Blackwell Publishing Ltd, Chichester

Marx GR, Allen HD (1986) Accuracy and pitfalls of Doppler evaluation of the pressure gradient in aortic coarctation. J Am Coll Cardiol 7(6):1379–1385

Perdreau E, Houyel L et al (2014) Tetralogy of Fallot with coarctation of the aorta: a newly recognised developmental and anatomic syndrome. Cardiol Young 24(4):714–720

Perloff JK (2010) The variant associations of aortic isthmic coarctation. Am J Cardiol 106(7):1038–1041

Shinebourne EA, Elseed AM (1974) Relation between fetal flow patterns, coarctation of the aorta, and pulmonary blood flow. Br Heart J 36(5):492–498

Shone JD, Sellers RD et al (1963) The developmental complex of "parachute mitral valve," supravalvular ring of left atrium, subaortic stenosis, and coarctation of aorta. Am J Cardiol 11:714–725

Smallhorn JF, Huhta JC et al (1982) Suprasternal cross-sectional echocardiography in assessment of patient ducts arteriosus. Br Heart J 48(4):321–330

Smallhorn JF, Huhta JC et al (1983) Cross-sectional echocardiographic assessment of coarctation in the sick neonate and infant. Br Heart J 50(4):349–361

Snider RA, Serwer GA et al (1997) Echocardiography in pediatric heart disease. Mosby, St. Louis

Teo LL, Cannell T et al (2011) Prevalence of associated cardiovascular abnormalities in 500 patients with aortic coarctation referred for cardiovascular magnetic resonance imaging to a tertiary center. Pediatr Cardiol 32(8):1120–1127

Wiegand G, Schlensak C et al (2015) Pitfalls in echocardiography: coarctation of the aorta presenting as dilated cardiomyopathy (DCM). Ultraschall Med 37(5):482–486