Patent Ductus Arteriosus

5

Contents

5.1 Anatomy and Haemodynamics

Although patent ductus arteriosus is basically a rather simple anomaly, representing persistence of a normal fetal structure, it may present in a variety of different anatomical variations, associated either with normal cardiac anatomy or with more or less complex congenital heart disease. While the common and most frequent patent ductus arteriosus is a left-sided structure, variations of its manifestation can be explained based on the hypothetical model of bilateral aortic arches proposed by J. Edwards (1948; Moes and Freedom 1992). According to this model of bilateral aortic arches, the ductus arteriosus in patients with a normal left aortic arch develops from the left-sided distal sixth aortic arch (Fig. 5.1). This left-sided ductus connects the aorta distal to the left subclavian artery with the main pulmonary artery just above the origin of the left pulmonary artery. According to the aortic arch model, there are three further possibilities of ductal patency: in patients with left aortic arch, patency of the right-sided distal sixth aortic arch results in a right ductus arteriosus originating from the base of the innominate artery and connecting to the right pulmonary artery (Fig. 5.2). In patients with right aortic arch, the same applies in a mirror image fashion with the possibility of a left ductus arteriosus originating from the base of a left-sided innominate artery and a right-sided ductus arteriosus originating from the undersurface of the right aortic arch (Fig. 5.2).

In the vast majority of cases however, the ductus arteriosus is left sided in the presence of a left aortic arch. According to the German PAN study, isolated persistent ductus arteriosus accounts for 4.3 % of congenital heart defects, detected in the first year of life (Lindinger et al. 2010; Schwedler et al. 2011). The anatomic variations of ductus arteriosus mentioned above are rare and frequently occur in conjunction with significant other congenital cardiac malformations. This applies also to patients with bilateral persistence of ductus arteriosus. In these cases, both pulmonary arteries are frequently perfused separately by a left and a right ductus arteriosus in the absence of a pulmonary artery bifurcation

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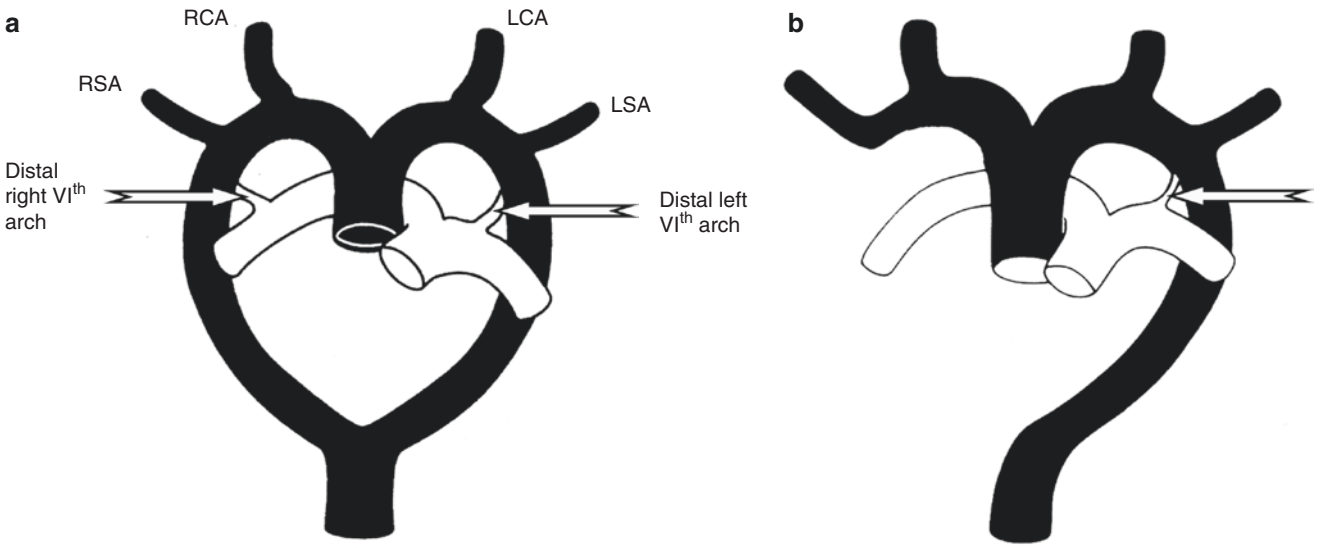

Fig. 5.1 Scheme of the hypothetical double aortic arch model according to Edwards (**a**). The development of the normal left aortic arch is explained by regression of the right dorsal aortic root (**b**). The distal part of the right sixth aortic arch disappears, while the distal part of the left sixth aortic arch becomes the left-sided ductus arteriosus, originating from the inner curvature of the aortic arch (*arrow*). *RSA*, *LSA* right, left subclavian artery, *RCA*, *LCA* right, left carotid artery (With kind permission modified according to Chap. 43 of the textbook "Neonatal heart disease", editors Freedom, Benson, Smallhorn, Springer Verlag 1992)

(Formigari et al. 1992; Freedom et al. 1984; Kwan-Wong et al. 2010; Peirone et al. 2002).

The ductus arteriosus is a vascular structure of vital importance for the circulation in the fetus. While 60 % of the fetal cardiac output is ejected from the right ventricle to the main pulmonary artery, only 8 % pass the lungs. The remainder of the blood is directed from the pulmonary artery via the ductus to the descending aorta. Premature closure of the ductus in utero in the presence of normal cardiac anatomy results in a maximal increase of right ventricular afterload. In this situation the fetus develops significant right ventricular dilatation, tricuspid regurgitation, fetal hydrops and frequently fetal death (Musewe and Olley 1992).

Congenital absence of the ductus arteriosus may occur in conjunction with specific complex forms of congenital heart disease like tetralogy of Fallot, pulmonary atresia and VSD with collateral lung perfusion by major aortopulmonary collateral arteries, absent pulmonary valve syndrome or truncus arteriosus. Congenital absence of the ductus or early intrauterine closure does not impede the fetal circulation in these patients, since these congenital cardiac defects have in common a large ventricular septal defect, allowing decompression of the right ventricle to the systemic circulation during intrauterine life.

As already mentioned, patency of the ductus arteriosus is of vital importance for the fetus with normal cardiac anatomy. Closure of the ductus following birth is initiated by significant haemodynamic changes. Due to inflation of the lungs immediately after birth, there is a dramatic decrease in pulmonary vascular resistance and a dramatic increase in pulmonary blood flow resulting in an immediate decrease of right to left shunting across the ductus arteriosus. The ductus arteriosus closes shortly after birth in response to increasing oxygen tension, decreasing levels of prostaglandin and prostacyclin and increasing levels of vasoactive substances (Musewe and Olley 1992). Normally complete functional closure of the ductus occurs within the first few days of life, followed by anatomical obliteration within the following weeks. In healthy neonates, closure of the ductus arteriosus is observed by the fourth day of life, and ductal patency beyond this is abnormal (Reller et al. 1990). Ductal patency depends on factors stimulating either patency or ductal closure. Factors involved in maintaining ductal patency are vasodilating prostaglandins and NO. Ductal closure is stimulated by a significant increase in PO_2.

Persistent patency of the ductus arteriosus may be encountered in three different situations:

Persistent ductus arteriosus in term newborns
Persistent ductus arteriosus in preterm newborns
Persistent ductus arteriosus in children with congenital heart disease
  Right-sided obstructive lesions
  Left-sided obstructive lesions

1. The haemodynamic consequences of patent ductus arteriosus in term newborns depend on its size and on systemic and pulmonary vascular resistance. With a widely patent ductus, the pulmonary vascular bed continues to be exposed to systemic blood pressure and is confronted with a significantly increased pulmonary blood flow. A large ductus

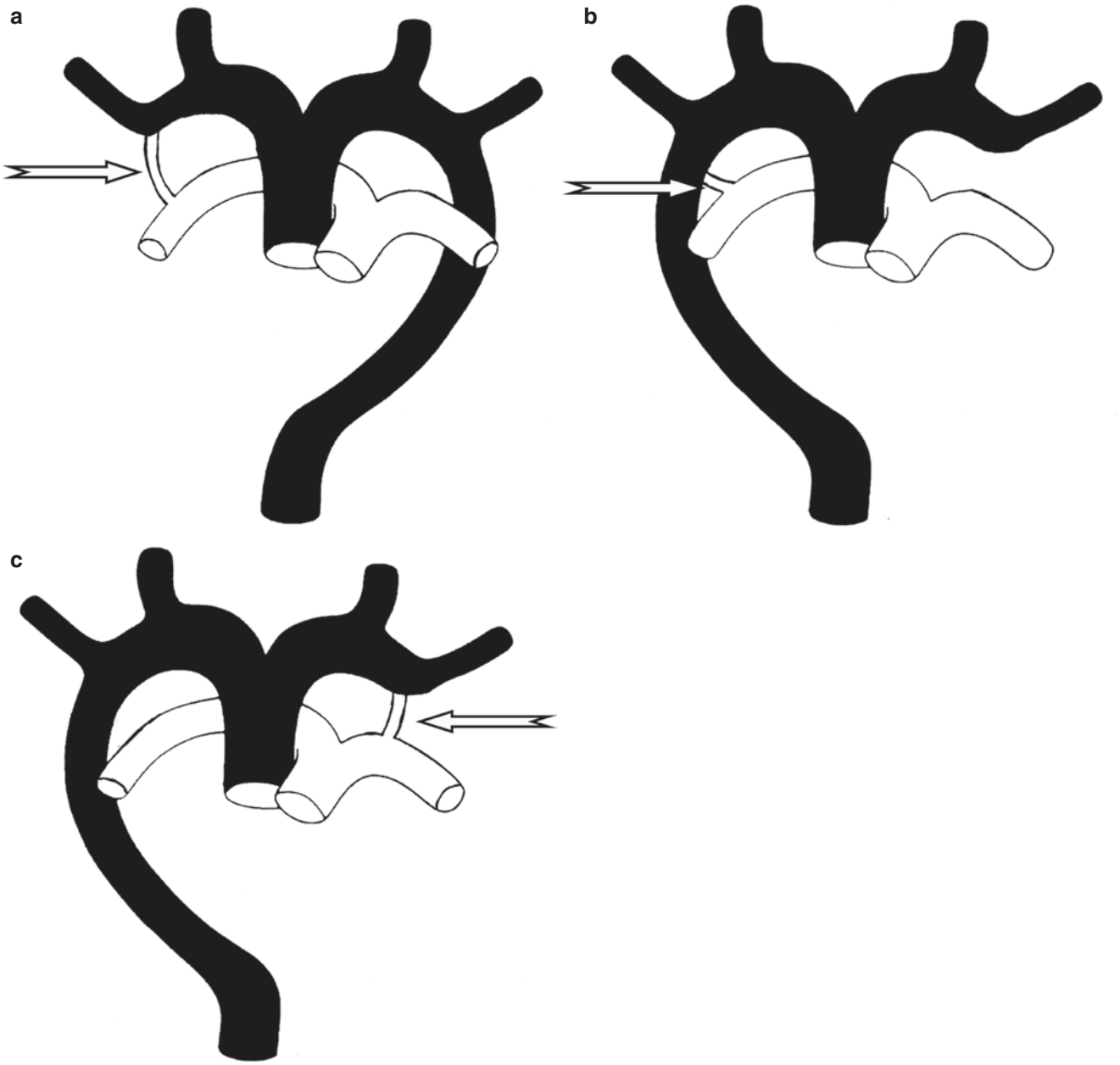

Fig. 5.2 Persistence of the distal part of the right sixth aortic arch in a patient with left aortic arch results in a right ductus arteriosus (*arrow*), connecting the base of the right innominate artery with the right pulmonary artery (**a**). In patients with right aortic arch, persistence of the distal right sixth aortic arch results in a ductus arteriosus originating from the undersurface of the aortic arch (**b**). Development of a left-sided ductus arteriosus in right aortic arch, originating from the base of the left-sided innominate artery, is the result of persistence of the distal left sixth aortic arch (**c**). The presence of bilaterally patent ductus is possible both in patients with right and left aortic arch (With kind permission modified according to Chap. 43 of the textbook "Neonatal heart disease", editors Freedom, Benson, Smallhorn, Springer Verlag 1992)

arteriosus allows equalization of pressures in the aorta and pulmonary artery. Elevated pulmonary artery pressure in combination with increased blood flow exposes the pulmonary vascular bed to increased shear stress, resulting in progressive pulmonary vascular damage. In term newborns, this refers to ductus with a minimal diameter of >3 mm. Medium-size ductus with a diameter of 2–3 mm does not allow equalization of pressures in the aorta and pulmonary artery, while they still allow significant LR-shunting. LR-shunting via a patent ductus arteriosus results in significant volume load of the pulmonary arteries, the left atrium and the left ventricle. Small ducts with a diameter of <2 mm neither expose the pulmonary arteries to elevated pressure nor do they allow significant LR-shunting.

2. Persistent patency of the ductus arteriosus is a problem frequently encountered in premature infants with structurally normal hearts (Pees and Obladen 2005). However the true incidence of patent ductus had been unknown

prior to the introduction of echocardiography and its Doppler modalities into the routine assessment of these children. In healthy premature infants of 30–37 weeks of gestation, timing of ductal closure is comparable to term newborns and occurs normally within the first 4 days of life (Reller et al. 1988). Among these patients, even respiratory distress syndrome does not increase the incidence of persistent patent ductus arteriosus significantly (Reller et al. 1990). This is quite different among infants with very low birth weight. Especially among infants who are born before 26 weeks of gestation, the patency rate of ductus arteriosus has been found to be as high as 65 % (Costeloe et al. 2000). In these children, patency of the ductus contributes significantly to respiratory complications, aggravates respiratory distress syndrome by impairment of lung function and promotes development of chronic lung disease. In addition significant LR-shunting and diastolic run-off from the systemic circulation in the presence of a large ductus result in reduced perfusion of the postductal organs including the gastrointestinal tract and the kidneys (Coombs et al. 1990; Deeg et al. 1987). Ischaemia due to hypoperfusion of the gastrointestinal tract is a significant risk factor of necrotizing enterocolitis (van de Bor et al. 1988). Furthermore patency of the ductus arteriosus in premature children is associated with a higher incidence of intraventricular haemorrhage, which may be attributed to alterations of cerebral blood flow. There is still debate however about the details of their causal relationship (Cooke et al. 2003; Fowlie and Davis 2002; Maier 2005; Osborn et al. 2003).

3. In the context of congenital heart disease associated with right-sided obstructive lesions and diminished pulmonary blood flow, the ductus arteriosus may function as a source of collateral pulmonary perfusion. This applies to patients with pulmonary atresia and intact ventricular septum, critical pulmonary stenosis, tetralogy of Fallot and pulmonary atresia with VSD, tricuspid atresia with pulmonary stenosis and other forms of complex congenital heart disease with critical reduction of antegrade pulmonary blood flow. If the malformation is associated with significant reduction of pulmonary blood flow during fetal life, the ductus is usually long and tortuous and originates with an acute angle from the aortic arch, indicating preferential LR-shunting already in fetal life (Santos et al. 1980). Following spontaneous closure of the ductus in the neonatal period, patients experience critical reduction of collateral pulmonary blood flow, associated with life-threatening hypoxaemia. This can be reverted by reopening of the ductus arteriosus through administration of prostaglandin E_1, a medical treatment that has revolutionarized treatment of patients with duct-dependent congenital heart disease (Musewe and Olley 1992). In critical left heart obstructions, the ductus arteriosus bypasses the left heart in fetal life: in these patients, the right ventricle takes care of the entire cardiac output via the main pulmonary artery and the ductus, thereby supporting the systemic circulation (Rychik et al. 2000; Rychik et al. 1996). Progressive constriction of the ductus arteriosus following birth deprives the systemic circulation of its source of perfusion. In synchrony with the progressive constriction of the ductus, there is a gradual decrease of systemic output with incipient shock and acidosis. In patients with critical coarctation, ductal constriction results in reduced perfusion of the lower half of the body. In this situation again administration of prostaglandin E_1 has been established as a very effective emergency treatment. For patients requiring ductal patency for a longer time period, ductal stenting by interventional cardiac catheterization has been introduced both in the setting of ductal dependency of the pulmonary and the systemic circulation (Akintuerk et al. 2002; Schranz et al. 2010).

Last but not least, it has to be mentioned that patent ductus arteriosus may be associated with a variety of other congenital cardiac malformations. In the context of septal defects like VSD and AVSD, additional shunting via the patent ductus arteriosus may aggravate the deleterious haemodynamic effects of the intracardiac defects.

5.2 Two-Dimensional Echocardiography

Indirect signs, associated with patent ductus arteriosus that can be detected by 2D-echocardiography, refer to the sequelae of LR-shunting (Skinner 2000a). In patients with large- or medium-size ductus, the apical four-chamber view will show significant increase in size of the left atrium and left ventricle (Fig. 5.3, Video 5.1). The increase in left atrial size can be verified by M-mode echocardiography in the

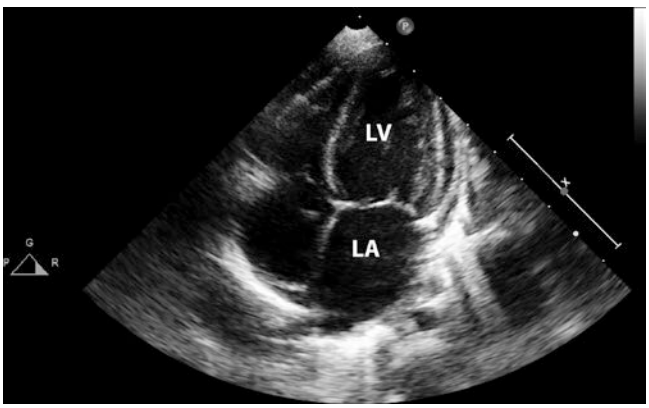

Fig. 5.3 Apical four-chamber view in a 12-month-old infant with medium-size ductus arteriosus shows moderate dilatation of left atrium (LA) and left ventricle (LV) due to significant left to right shunting

parasternal long axis view (Fig. 5.4). Normally the diameter of the aortic root in this plane equalizes the diameter of the left atrium behind the aorta. An increase of the LA:AO ratio > 1,4:1 reflects significant left atrial enlargement (Skinner 2000a).

Two-dimensional evaluation of the ductus should aim for visualization of the whole length of the duct. It has to be kept in mind, however, that especially small ducts may escape the detection of echocardiography without the support of colour Doppler. Several imaging planes are valuable in the detection and assessment of the normal left-sided patent ductus arteriosus in the setting of a left aortic arch. Frequently but not always, large ducts can be displayed in the suprasternal long axis view of the aortic arch (Fig. 5.5). In this plane, the ductus connects to the undersurface of the aortic arch (Fig. 5.6). In the parasternal short axis view, the ductus arteriosus can be visualized in a plane just slightly superior to the pulmonary bifurcation (Video 5.2). A large ductus in the neonate has a diameter similar to the diameter of the left and right pulmonary artery and appears as a third vessel originating from the bifurcation and connecting to the descending aorta (Fig. 5.7, Video 5.3). The detection of smaller ductus in this plane will require CDE (Fig. 5.8).

The most valuable plane for visualization and assessment of the ductus is the left parasternal sagittal plane, *the so-called ductal view*, *which is obtained with the transducer placed in the second left intercostal space*. This plane shows the transition of the distal aortic arch to the descending aorta (the aortic isthmus) as well as the space between the left pulmonary artery and the aorta – the space that is occupied by the ductus arteriosus, if it remains patent (Lai et al. 2009; 2009). In the majority of cases, this plane allows to display the entire length of a persistently patent ductus (Fig. 5.9, Videos 5.4 and 5.5). Assessment of the whole length of the ductus is important, since in the majority of cases, the ductus is not a tubular structure but almost always has an area of constriction, which is frequently located at the pulmonary end (Krichenko et al. 1989) (Video 5.6). Furthermore the "ductal view" is the ideal plane to visualize devices (Fig. 5.10), implanted for interventional occlusion (Video 5.7) or maintenance of patency of the ductus (Videos 5.8 and 5.9) in the cath lab (Rauch et al. 2001). Although the presence of a large- or medium-size ductus can be detected with the help of colour Doppler in the subcostal planes, these planes do not play a

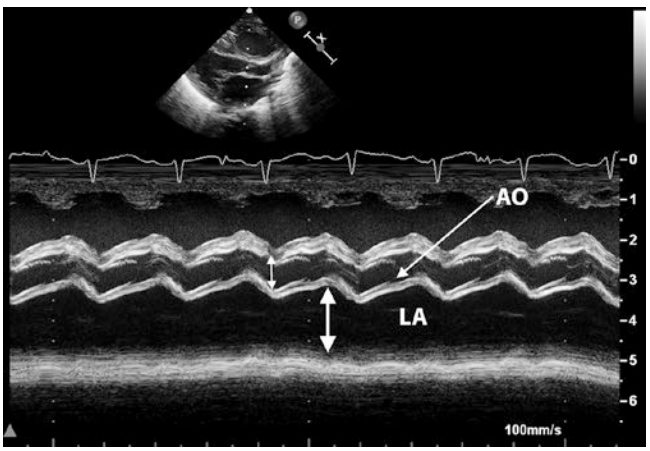

Fig. 5.4 M-mode echocardiogram in the parasternal long axis in a small infant with large ductus arteriosus shows significant dilatation (*broad arrow*) of the left atrium (*LA*) compared to the aortic root (*AO*, *small arrow*) with a LA:AO ratio of 1,8: 1

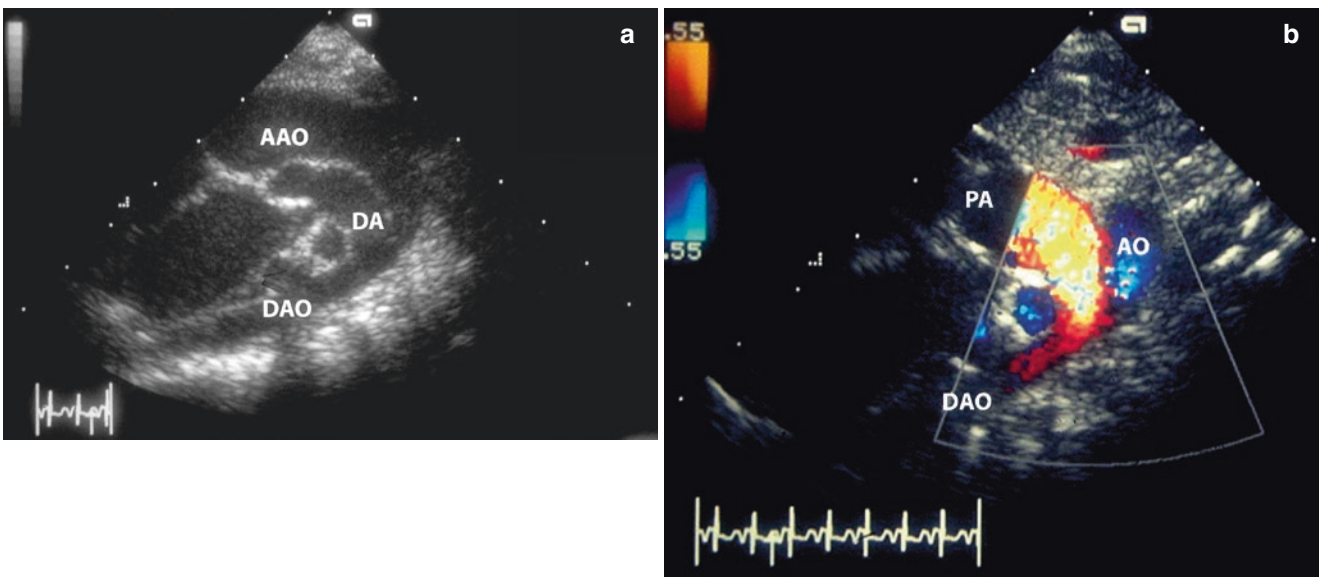

Fig. 5.5 Suprasternal long axis in a 6-week-old infant with a large ductus (**a**). Colour Doppler in the ductal view shows significant left to right shunting across the duct during diastole (**b**). *AAO* ascending aorta, *DAO* descending aorta, *DA* ductus arteriosus. *PA* pulmonary artery

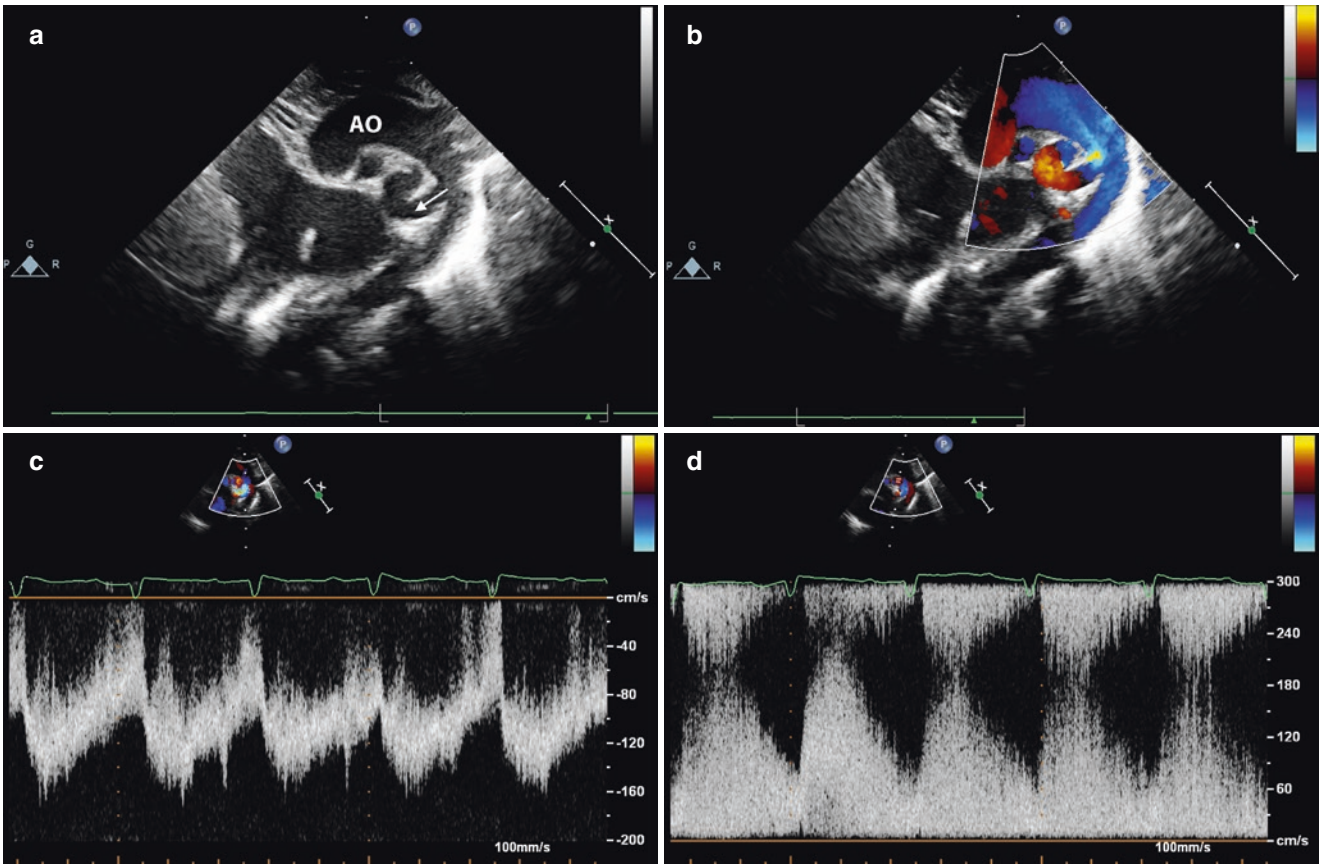

Fig. 5.6 Suprasternal long axis showing the ductus arteriosus (*arrow*) in a neonate with cyanotic congenital heart disease (**a**) originating from the undersurface of the aortic arch (*AO*). Although there is exclusive LR-shunting, colour Doppler shows different flow directions due to the tortuosity of the duct (**b**). This is confirmed by PW Doppler interrogation showing different flow directions according to different sites of interrogation (**c**, **d**)

significant role in the two-dimensional assessment of this structure.

5.3 Colour Doppler Echocardiography

Colour Doppler echocardiography is the most sensitive and most specific imaging modality to detect patency of the ductus arteriosus in infants and children (Musewe and Olley 1992). In the majority of cases, the ductus arteriosus is a left-sided structure connecting the undersurface of the aortic arch to the roof of the left pulmonary artery close to its origin from the bifurcation. The most sensitive plane to visualize the ductus by colour Doppler is the "ductal view". Since this plane shows the distal aortic arch and the left pulmonary artery in a rather parallel position, normal blood flow in both vessels, which is directed away from the transducer, is coded in blue (Fig. 5.10). LR-shunting via a patent ductus arteriosus in this plane is oriented towards the transducer and depicted in red (Videos 5.4 and 5.5). Since the direction of blood flow in the ductus differs entirely from normal blood flow in the pulmonary artery, colour Doppler interrogation in this plane is extremely sensitive and will detect even minimal shunting in the presence of a diminutive ("silent") ductus arteriosus (Fig. 5.11). "Silent ducts" are characterized by the fact that they are too small to create a typical cardiac murmur, which can be detected on auscultation (Bennhagen and Benson 2003; Cantinotti et al. 2014).

In the neonatal period, while pulmonary resistance is still elevated, CDE reveals bidirectional shunting across the ductus with RL-shunting during systole and LR-shunting during diastole (Fig. 5.7, Video 5.3). With decreasing pulmonary vascular resistance, the velocity and duration of RL-shunting diminish resulting in continuous LR-shunting. The latter is the normal flow pattern in children with isolated ductus arteriosus after the first week of life. As long as pulmonary artery pressure is elevated in the neonatal period, shunting across the ductus may occur at low flow velocities requiring a significant reduction in the scale (prf-rate) of colour Doppler interrogation. With decreasing pulmonary pressure and resistance, velocity of LR-shunting increases resulting in turbulence and aliasing (Fig. 5.5).

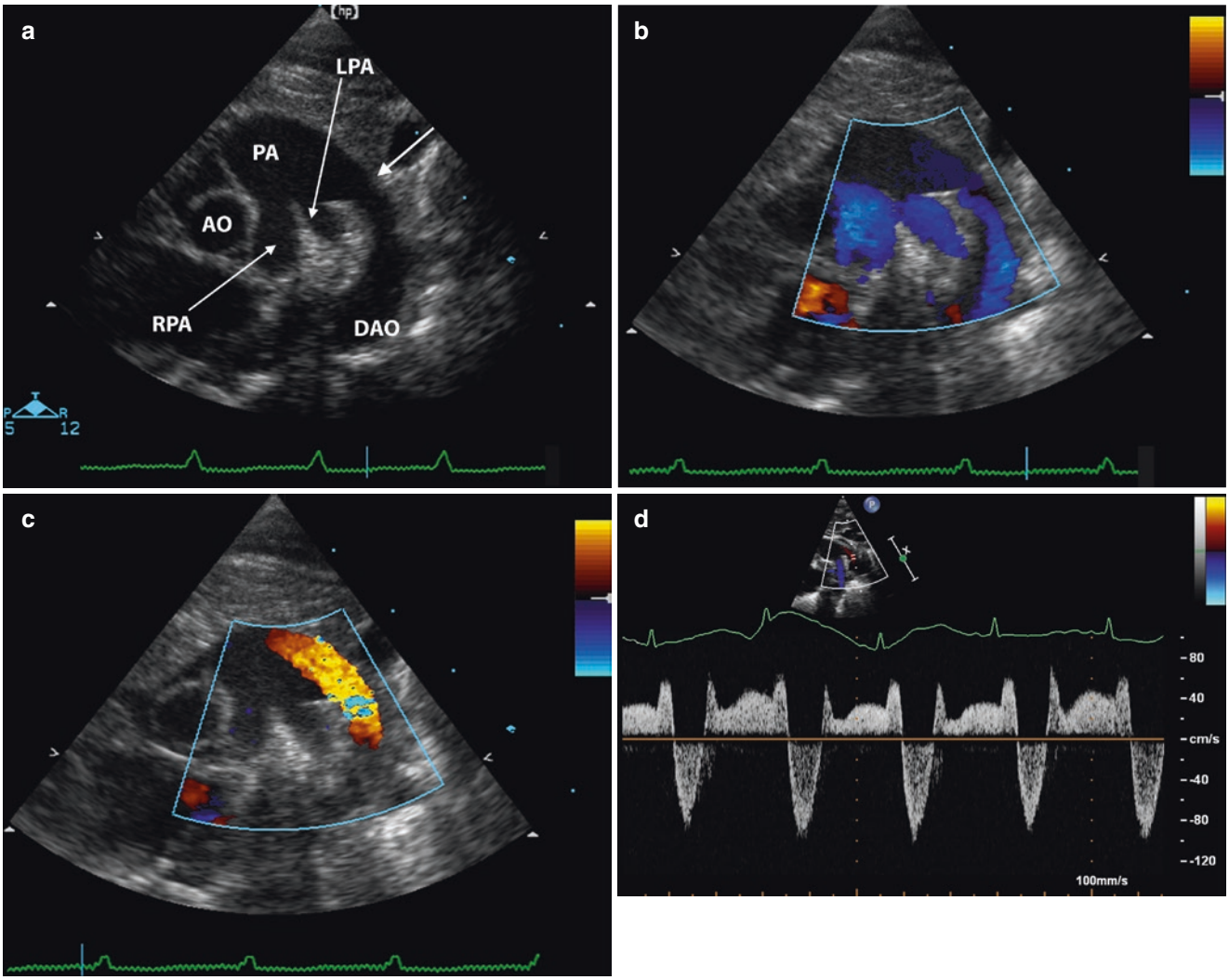

Fig. 5.7 This slightly angulated parasternal short axis in a neonate shows a large ductus (*arrow*) connecting the pulmonary artery (*PA*) with the descending aorta (*DAO*). *AO* ascending aorta, *RPA*, *LPA* right, left pulmonary artery. Colour Doppler shows bidirectional shunting with right to left shunting in systole (**b**) and left to right shunting in diastole (**c**). Bidirectional shunting is confirmed by PW Doppler (**d**)

Another scanning plane that allows good visualization of a left-sided duct is the high parasternal short axis. In a plane slightly superior to the pulmonary bifurcation, the duct can be displayed as a third vessel connecting the bifurcation to the descending aorta (Figs. 5.7 and 5.8, Videos 5.2 and 5.3).

The amount of shunting can be estimated by the size of colour Doppler flow entering the pulmonary artery. In the presence of a large ductus, LR-shunting extends retrogradely into the main pulmonary artery down to the level of the pulmonary valve. A tiny ductus arteriosus appears as a tiny flame in the pulmonary bifurcation, which does not persist over the entire cardiac cycle (Fig. 5.11).

There are several differential diagnoses that have to be excluded by careful visualization of the duct in the ductal view:

1. Coronary fistulae may enter the main pulmonary artery and will result in colour Doppler inflow originating from their connection to the pulmonary artery (Fig. 5.11). Differentiation of this vascular structure is given by the fact that it is impossible to connect this vascular structure to the descending aorta (Video 5.10).
2. Small aortopulmonary collateral arteries may originate from the descending aorta and connect to central or peripheral pulmonary arteries. They can be discriminated from a small ductus, since they do not show direct inflow into the pulmonary arteries at the bifurcation (Fig. 5.11, Video 5.11).
3. Pulmonary valvular stenoses (especially mild stenoses) may cause turbulent flow in the main pulmonary artery. In the parasternal short axis, this may imitate retrograde flow at the aortic side of the main pulmonary artery.

In patients with right aortic arch, the ductus arteriosus may present as a right-sided ductus, originating from the

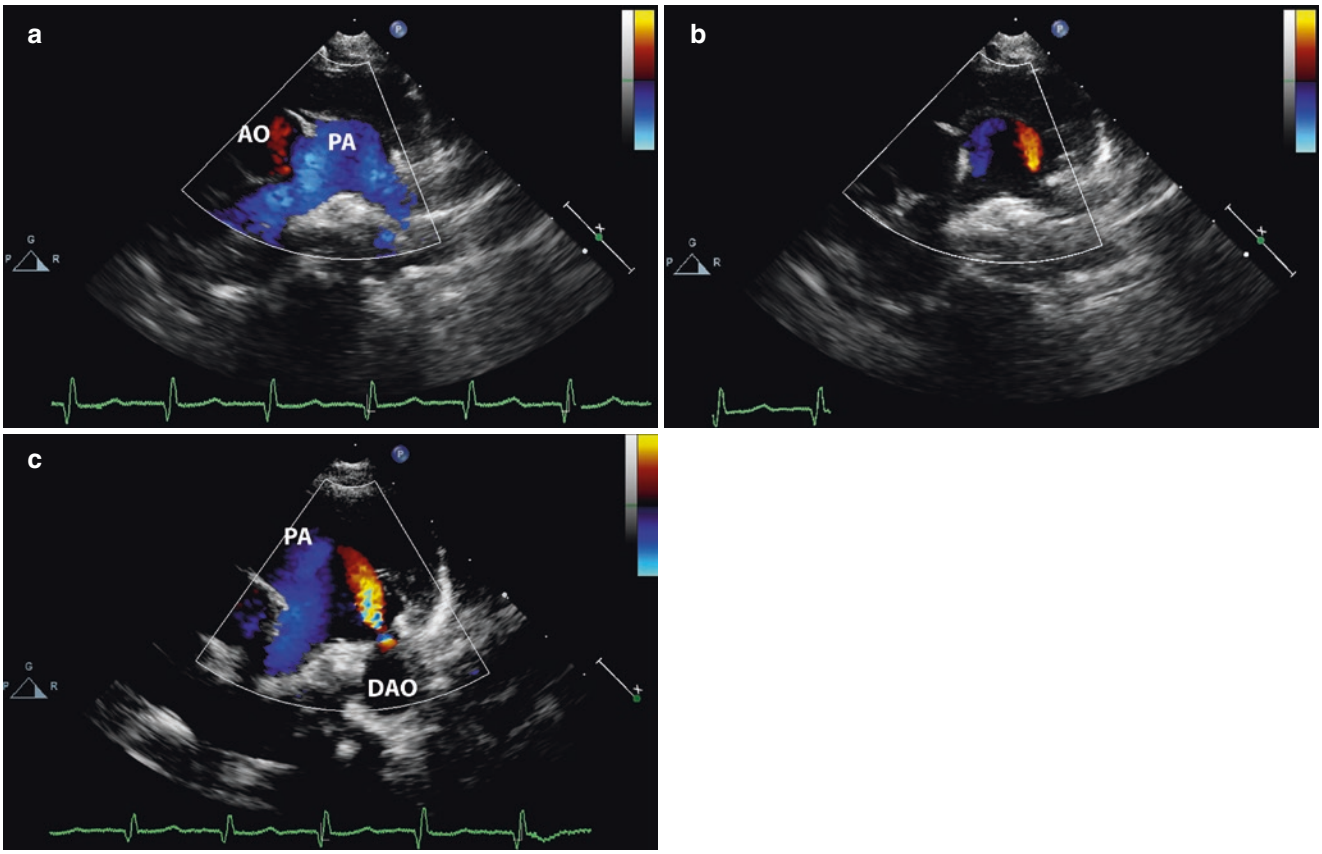

Fig. 5.8 Colour Doppler examination in this parasternal short axis view appears normal (**a**). Slight cranial angulation reveals diastolic inflow of a small ductus (**b**). Further cranial tilt (**c**) visualizes the ductus connecting the descending aorta (*DAO*) with the pulmonary artery (*PA*)

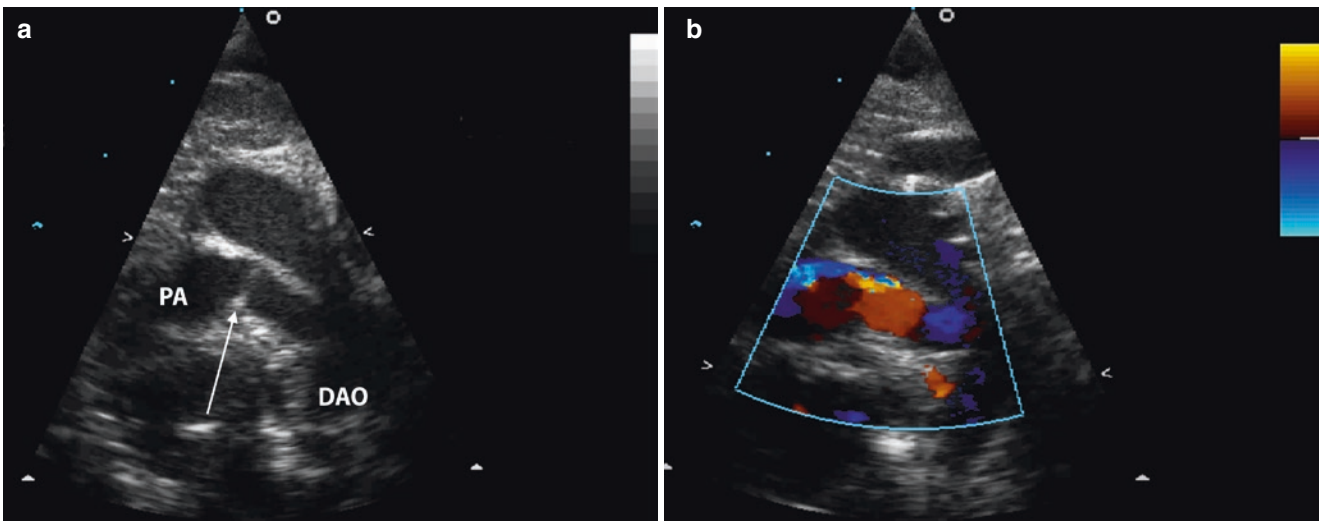

Fig. 5.9 The ductal view in a neonate shows a large ductus connecting the pulmonary artery (*PA*) to the descending aorta (*DAO*). The large ductus is partially obstructed by a membrane (*arrow*) at the pulmonary end (**a**). This is confirmed by colour Doppler revealing shunting limited to the cranial part of the ductus (**b**)

undersurface of the aortic arch. In the majority of cases, this anatomic variant will be found in the presence of complex congenital heart disease. The ductus in this situation is usually well depicted during evaluation of the right aortic arch in the suprasternal long axis.

Another variant is patent ductus arteriosus originating from the base of the innominate artery, usually presenting as a left-sided duct in the presence of a right aortic arch (Fig. 5.2). In these cases, the ductus is well displayed by examination of the innominate artery: longitudinal imaging

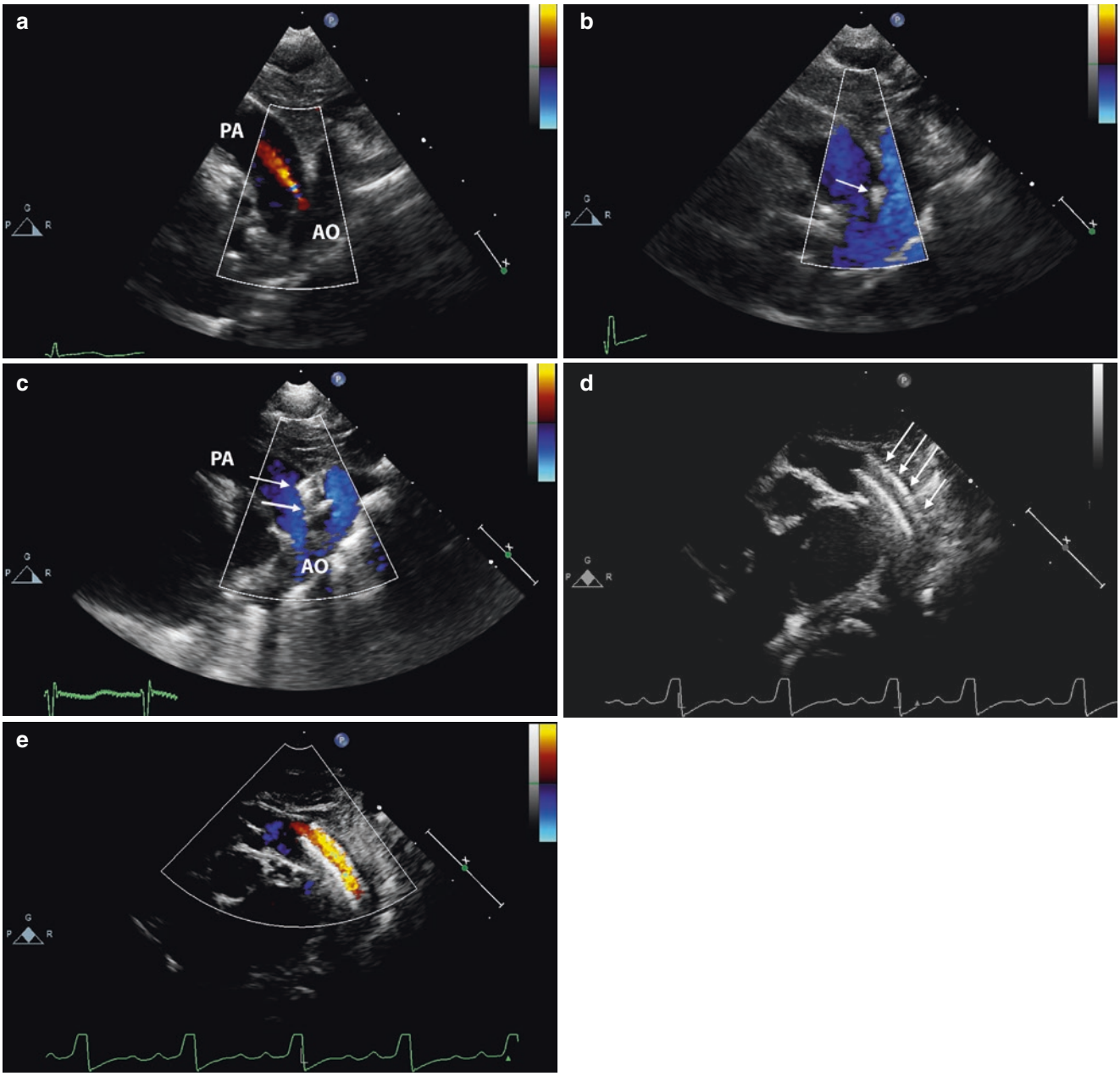

Fig. 5.10 Colour Doppler in the "ductal view" (**a**) shows a small ductus arteriosus with a small amount of left to right shunting. *AO* aorta, *PA* pulmonary artery. Following interventional occlusion with a detachable coil (*arrow*), the ductus is completely occluded (**b**). The "ductal view" in a different patient shows (**c**) complete occlusion by an Amplatzer^{R} duct occluder (*arrows*). Following interventional stent placement to maintain patency of the duct in a neonate with pulmonary atresia, the stent (*arrows*) is seen in projection on the ductus (**d**), with confirmation of patency by colour Doppler (**e**)

of the innominate artery, in addition to normal branching into subclavian and carotid artery, demonstrates the origin of a third vessel, which is oriented in a caudal direction (Fig. 5.12). The caudal course of this vessel connecting to the pulmonary artery can be displayed in parasternal long and short axis planes (Fig. 5.12). *It is important to remember that the pulmonary artery connected to this ductus may miss a connection to the pulmonary bifurcation and may be completely dependent on ductal patency* (Fig. 5.13). Spontaneous closure of the ductus results in isolation and unilateral absence of the respective pulmonary artery (Koga et al. 2010; Ten Harkel et al. 2002).

The latter situation is frequently the case in patients with bilateral ducts (Formigari et al. 1992; Freedom et al. 1984; Kwan-Wong et al. 2010; Peirone et al. 2002). In these patients, one ductus originates from the undersurface of the aortic arch, while the contralateral ductus originates from the base of the innominate artery (Fig. 5.2). CDE in the high parasternal short axis demonstrates bilateral ductus connecting to the hilar pulmonary arteries (Fig. 5.12). The pulmonary bifurca-

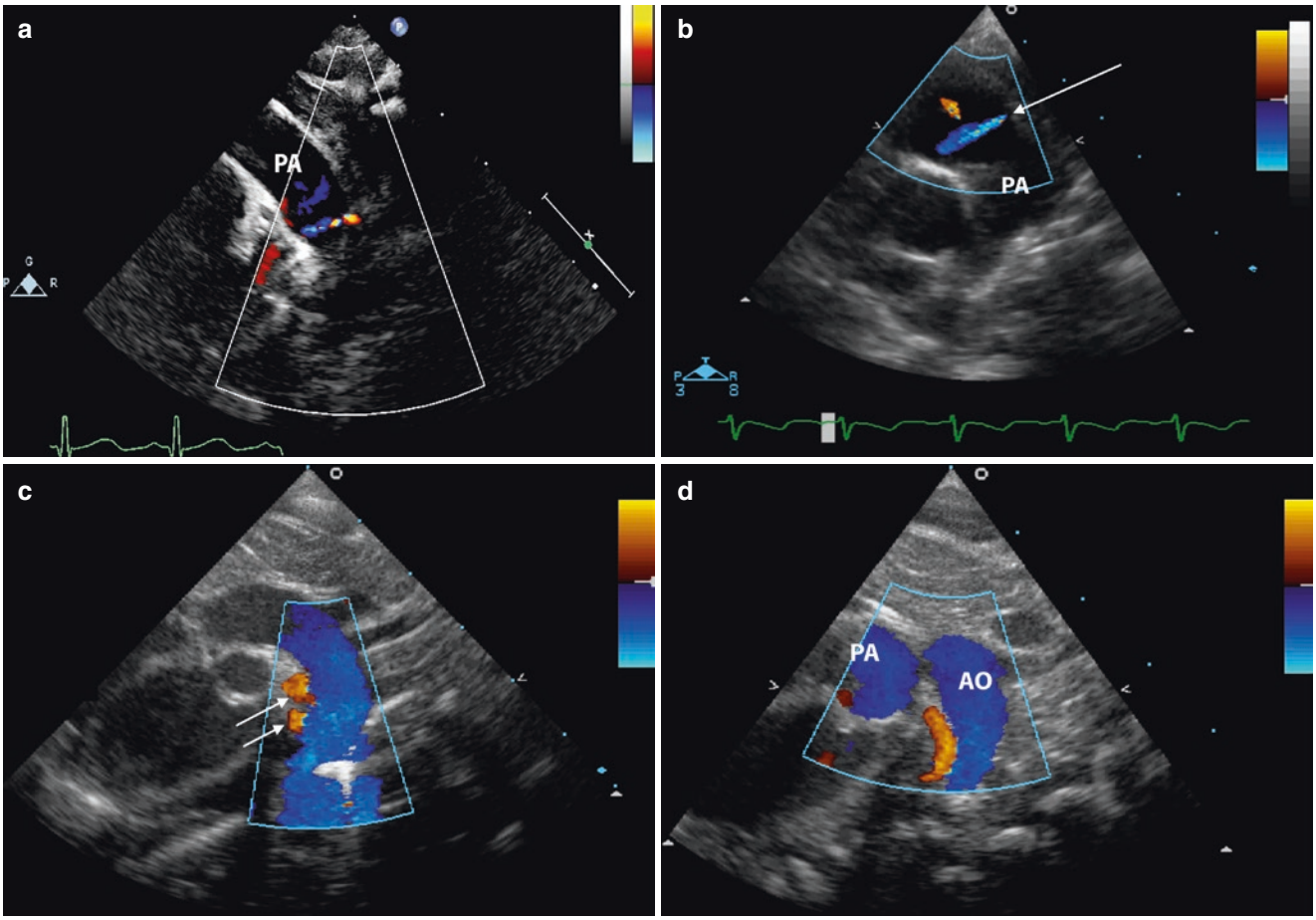

Fig. 5.11 Colour Doppler in the ductal view (**a**) showing a very tiny ("silent") ductus arteriosus resulting in very limited, eccentric diastolic left to right shunting into the pulmonary artery (*PA*). The parasternal short axis in this infant shows diastolic inflow from the lateral wall (*arrow*) of the main pulmonary artery (*PA*), originating from a small coronary artery fistula (**b**). The suprasternal long axis of the aortic arch in an infant shows two small vessels originating from the undersurface of the arch representing small aortopulmonary collateral arteries (**c**); they can be differentiated from a tiny patent duct in the "ductal view", since they do not produce inflow into the roof of the main pulmonary artery (**d**). *PA* pulmonary artery, *AO* aorta

tion in this situation is usually absent. In the majority of children, bilateral ductal perfusion of pulmonary arteries will be associated with complex congenital heart disease (Formigari et al. 1992; Freedom et al. 1984; Kwan-Wong et al. 2010; Peirone et al. 2002).

5.4 Pulsed Wave and Continuous Wave Doppler Sonography

In the neonatal period, PW Doppler is the optimal tool to verify direction and duration of shunting across the ductus. Due to the elevated pulmonary vascular resistance, flow velocities across the ductus are low and can be well depicted by pulsed wave Doppler interrogation. Depending on the underlying haemodynamics, three different flow patterns can be encountered in the neonatal period:

1. Patients with pulmonary hypertension and significantly elevated pulmonary vascular resistance will exhibit exclusive RL-shunting, since pulmonary artery pressure exceeds aortic pressure in systole as well as in diastole. This flow pattern, which is observed only on rare occasions, is always pathologic and requires immediate clarification of its underlying pathomechanism (Skinner 2000b). Possible underlying causes are persistent pulmonary hypertension of the newborn, pulmonary hypertension in children with diaphragmatic hernia, pulmonary vein stenosis or total anomalous pulmonary venous connection with obstructive pulmonary veins. Exclusive RL-shunting may also occur in neonates with hypoplastic left heart syndrome or critical coarctation in the presence of a narrow, restrictive ductus arteriosus.
2. In patients with congenital heart disease and pulmonary hypertension, the ductus presents with a bidirectional

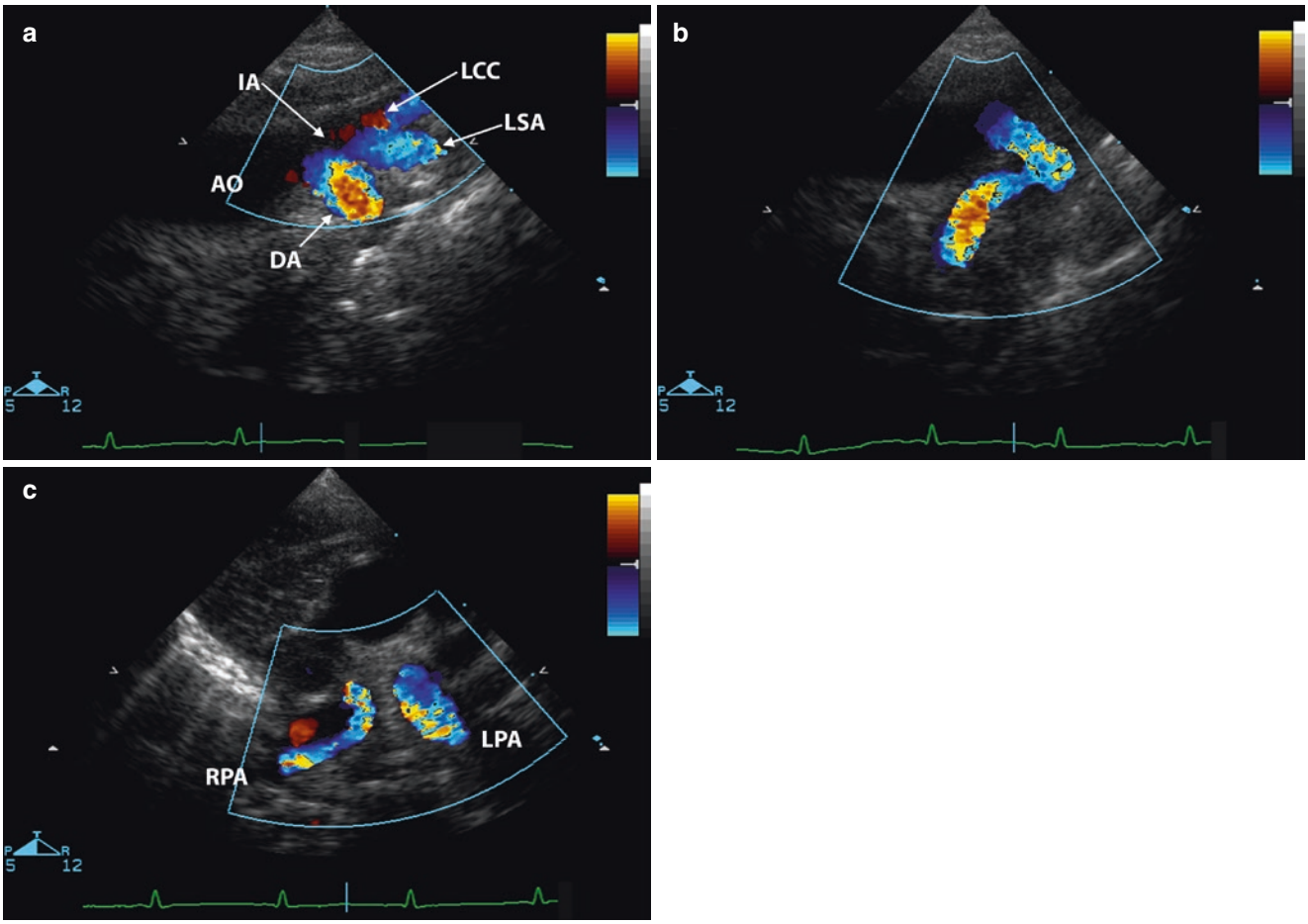

Fig. 5.12 High parasternal short axis view (**a**) in a patient with univentricular heart and right aortic arch shows a left innominate artery (*IA*) branching into left subclavian (*LSA*) and left common carotid artery (*LCC*). Colour Doppler shows a left-sided ductus (*DA*) originating from the base of the innominate artery. Caudal tilt of the transducer visualizes the course of ductus down the left hilum (**b**). Further caudal tilt (**c**) shows absence of a pulmonary bifurcation with perfusion of the left pulmonary artery (*LPA*) by the left-sided duct and separate perfusion of the right pulmonary artery (*RPA*) by a right-sided ductus originating from the undersurface of the right aortic arch (not visualized in this plane)

flow pattern (Popat and Kluckow 2012; Rychik et al. 1996, 2000; Skinner 2000b): during systole RL-shunting occurs from the pulmonary artery into the aorta, while flow reversal in diastole due to low diastolic pulmonary artery pressures results in LR-shunting (Fig. 5.7). In the neonate without associated congenital cardiac malformation or extracardiac disease, this flow pattern will be present only during a very limited time period followed by gradual transition into exclusive LR-shunting (Popat and Kluckow 2012; Skinner 2000a, b).

3. If the ductus arteriosus remains patent in the term neonate without associated cardiac or extracardiac disease, rapid decrease in pulmonary vascular resistance results in exclusive LR-shunting during the cardiac cycle with a maximal flow velocity during systole (Skinner 2000b) (Fig. 5.14).

Since the ductus arteriosus represents a communication between aorta and pulmonary artery, flow velocities can be used to estimate systolic pressure differences between both circulations. Following measurement of the maximal flow velocities across the ductus, the systolic gradient between systemic and pulmonary circulation can be calculated based on the simplified Bernoulli equation (Musewe et al. 1987, 1990):

$$\text{Systolic gradient Aorta / Pulmonary artery} = 4v^2$$

Based on the pressure difference, the pulmonary artery pressure can be calculated noninvasively following oscillometric measurement of the patient's blood pressure:

$$\text{Systolic blood pressure} - \text{Pressure gradient} = \text{Systolic pressure pulmonary artery}$$

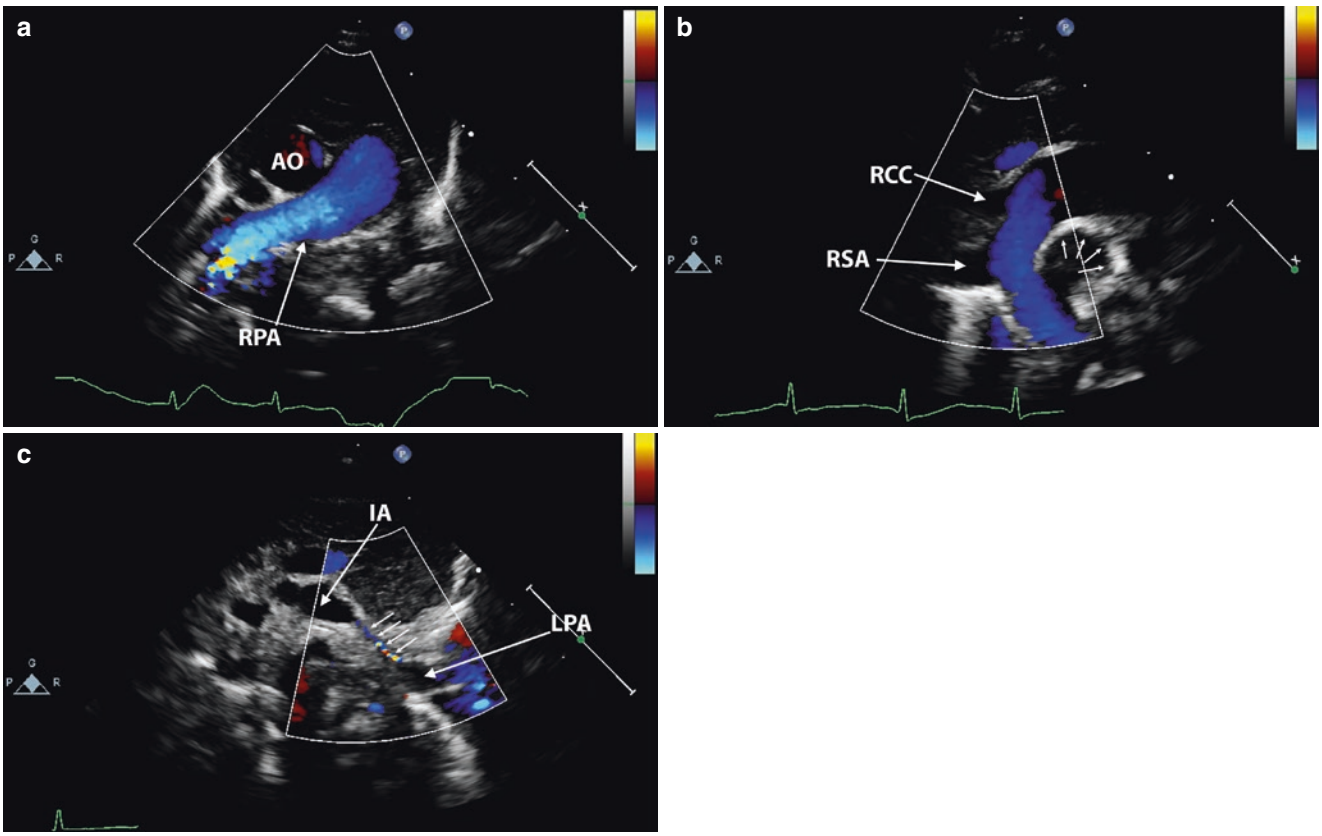

Fig. 5.13 High parasternal short axis view in a neonate with structurally normal heart showing normal retroaortic course (*AO*) of the right (*RPA*) and absence of the left pulmonary artery. The suprasternal long axis in this patient (**b**) shows a right aortic arch with origin of the right common carotid (*RCC*) and right subclavian artery (*RSA*). A large diverticulum originating from the undersurface of the aortic arch represents the remnant of a right-sided ductus (*arrows*). The high parasternal short axis (**c**) shows an almost completely occluded left-sided ductus arteriosus (*arrows*) connecting the base of the innominate artery (IA) to the left pulmonary artery in the hilum (*LPA*)

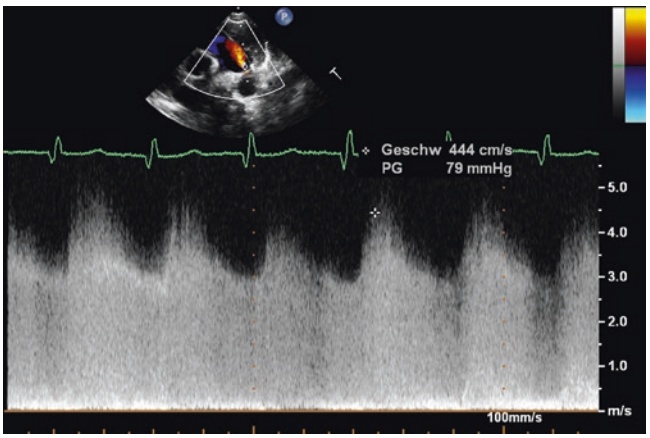

Fig. 5.14 CW Doppler interrogation shows continuous left to right shunting in a child with medium-size ductus arteriosus. According to the systolic pressure gradient of 79 mmHg, systolic pulmonary artery pressure is not elevated

A large ductus that results in significant elevation of the pulmonary artery pressure can be detected based on a low pressure gradient between both circulations with relatively low flow velocities in the duct. As a rule of thumb, flow velocities exceeding 4 m/s in children beyond the age of 8 weeks speak against pulmonary artery hypertension. On the other hand, flow velocities below 3 m/s are highly suspicious for some elevation of pulmonary artery pressure. These patients should be assessed carefully, if elevation can be attributed to the size of the ductus arteriosus or if other factors are detectable that might contribute to the pathology like pulmonary vascular disease, e.g. in association with bronchopulmonary dysplasia or upper airway obstruction.

Large- or medium-size ducts, resulting in significant LR-shunting, are associated with significant changes in flow patterns of systemic arteries:

Shunting across the ductus arteriosus results in an increased blood flow across the aortic isthmus both in systole and in diastole with systolic flow velocities that may well exceed 2 m/s (Fig. 5.15). Careful examination of the aortic arch and isthmus by 2D echocardiography is required in these patients to rule out isthmic coarctation.

Doppler interrogation of the descending aorta distal to the ductus arteriosus reveals retrograde diastolic flow due to

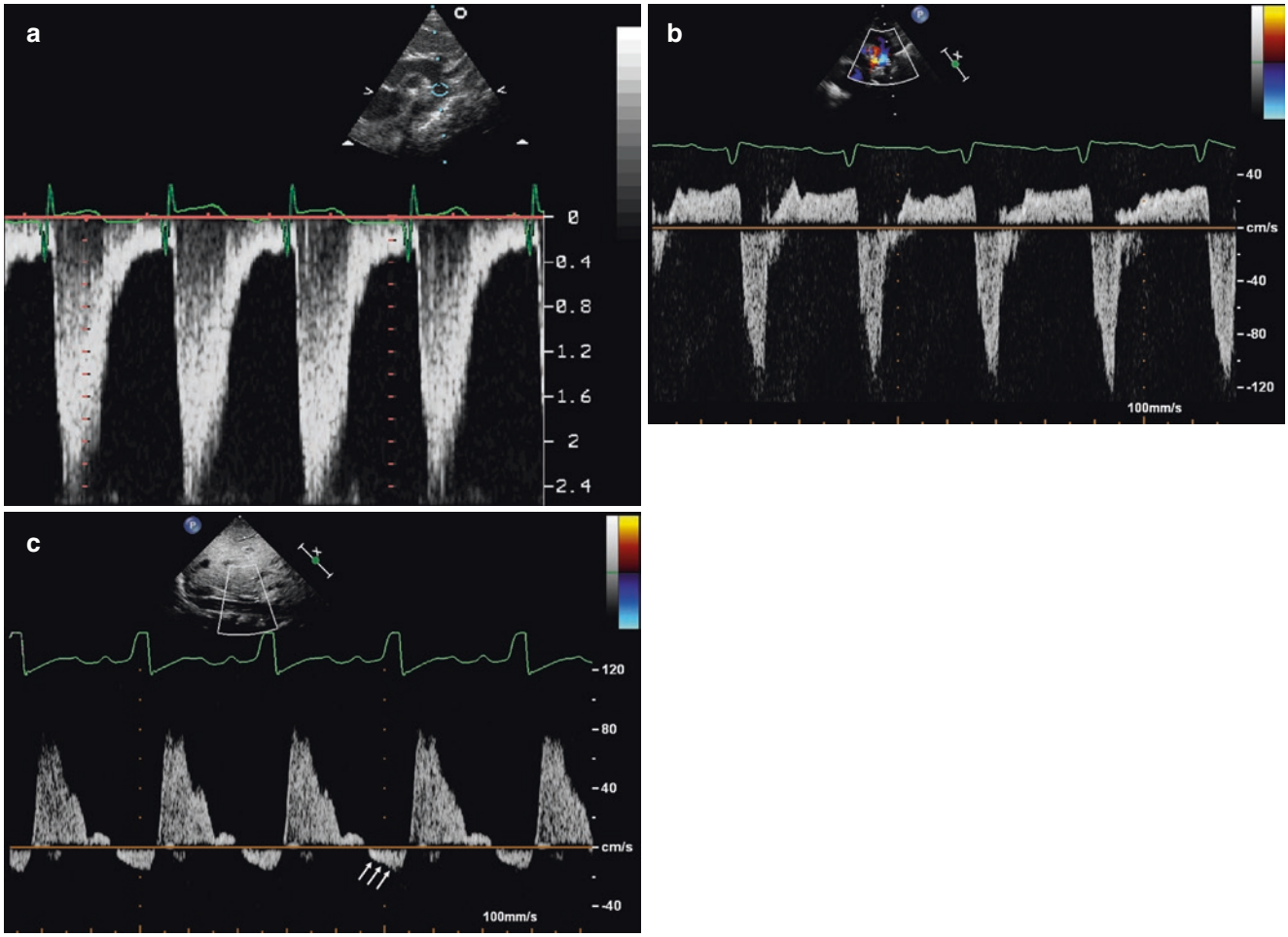

Fig. 5.15 In this infant with large ductus arteriosus, PW Doppler interrogation of the aortic arch shows significant elevation of systolic (2,4 m/s) and diastolic flow velocity despite the absence of obstruction (coarctation) of the aorta, which can explained by significant increase of flow (**a**). PW Doppler interrogation of the descending aorta distal to the large ductus shows diastolic backward flow due to the diastolic run-off across the ductus into the pulmonary arteriosus (**b**). The haemodynamic significance of this diastolic run-off is confirmed by a pronounced negative diastolic flow (*arrows*) in the coeliac artery (**c**)

run-off from the systemic circulation (Fig. 5.15). These changes are comparable to flow patterns in children with aortic regurgitation, aortopulmonary window or truncus arteriosus and remain restricted however to the descending aorta and not to the aortic arch.

Diastolic run-off from the aorta results in decreased diastolic flow in systemic arteries (Fig. 5.15). In neonates and small infants, these changes can be verified by Doppler examination of cerebral and abdominal arteries. Assessment of diastolic flow allows semiquantitative estimation of the haemodynamic relevance of the ductus arteriosus (Deeg et al. 1987): while preservation of diastolic antegrade flow is associated with limited haemodynamic effect of the duct, zero flow during diastole represents moderate and negative diastolic flow and significant haemodynamic relevance of the ductus arteriosus (Chapter 25).

References

Akintuerk H, Michel-Behnke I et al (2002) Stenting of the arterial duct and banding of the pulmonary arteries: basis for combined Norwood stage I and II repair in hypoplastic left heart. Circulation 105(9):1099–1103

Bennhagen RG, Benson LN (2003) Silent and audible persistent ductus arteriosus: an angiographic study. Pediatr Cardiol 24(1):27–30

Cantinotti M, Assanta N et al (2014) Controversies in the definition and management of insignificant left-to-right shunts. Heart 100(3):200–205

Cooke L, Steer P et al (2003) Indomethacin for asymptomatic patent ductus arteriosus in preterm infants. Cochrane Database Syst Rev (2):CD003745

Coombs RC, Morgan ME et al (1990) Gut blood flow velocities in the newborn: effects of patent ductus arteriosus and parenteral indomethacin. Arch Dis Child 65(10 Spec No):1067–1071

Costeloe K, Hennessy E et al (2000) The EPICure study: outcomes to discharge from hospital for infants born at the threshold of viability. Pediatrics 106(4):659–671

Deeg KH, Gerstner R et al (1987) Doppler sonographic detection of reduced flow velocity in the celiac trunk of the newborn infant with patent ductus arteriosus Botalli compared to a healthy control group. Monatsschr Kinderheilkd 135(1):24–29

Edwards JE (1948) Anomalies of the derivatives of the aortic arch system. Med Clin North Am 32:925–949

Formigari R, Vairo U et al (1992) Prevalence of bilateral patent ductus arteriosus in patients with pulmonic valve atresia and asplenia syndrome. Am J Cardiol 70(13):1219–1220

Fowlie PW, Davis PG (2002) Prophylactic intravenous indomethacin for preventing mortality and morbidity in preterm infants. Cochrane Database Syst Rev (3):CD000174

Freedom RM, Moes CA et al (1984) Bilateral ductus arteriosus (or remnant): an analysis of 27 patients. Am J Cardiol 53(7):884–891

Koga H, Hidaka T et al (2010) Age-related clinical characteristics of isolated congenital unilateral absence of a pulmonary artery. Pediatr Cardiol 31(8):1186–1190

Krichenko A, Benson LN et al (1989) Angiographic classification of the isolated, persistently patent ductus arteriosus and implications for percutaneous catheter occlusion. Am J Cardiol 63(12):877–880

Kwan-Wong C, Kramer L et al (2010) Echocardiographic diagnosis of bilateral ductus with discontinuous branch pulmonary arteries and heterotaxia syndrome. Pediatr Cardiol 31(6):917–918

Lai WW, Ko HH (2009) The normal pediatric echocardiogram. In: Lai WW, Mertens LL, Cohen MS, Geva T (eds) Echocardiography in pediatric and congenital heart disease. Blackwell Publishing Ltd, Chichester

Lai WW, Mertens LL et al (2009) Echocardiography in pediatric and congenital heart disease. Blackwell Publishing Ltd, Chichester

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

Maier RF (2005) Persisting ductus arteriosus and intraventricular hemorrhage. In: Obladen M, Koehne P (eds) Interventions for persisting ductus arteriosus in the preterm infant. Springer Medizin Verlag, Heidelberg, pp 35–38

Moes CAF, Freedom RM (1992) Rings, slings, and other things: vascular structures contributing to a neonatal "noose". In: Freedom RM, Benson LN, Smallhorn JF (eds) Neonatal heart disease. Springer, Berlin/Heidelberg/New York

Musewe NN, Olley PM (1992) Patent ductus arteriosus. In: Freedom RM, Benson LN, Smallhorn JF (eds) Neonatal heart disease. Springer, Berlin/Heidelberg/New York

Musewe NN, Poppe D et al (1990) Doppler echocardiographic measurement of pulmonary artery pressure from ductal Doppler velocities in the newborn. J Am Coll Cardiol 15(2):446–456

Musewe NN, Smallhorn JF et al (1987) Validation of Doppler-derived pulmonary arterial pressure in patients with ductus arteriosus under different hemodynamic states. Circulation 76(5): 1081–1091

Osborn DA, Evans N et al (2003) Hemodynamic and antecedent risk factors of early and late periventricular/intraventricular hemorrhage in premature infants. Pediatrics 112(1 Pt 1):33–39

Pees C, Obladen M (2005) Epidemiology of persisting ductus in preterm infants. In: Obladen M, Koehne P (eds) Interventions for persisting ductus arteriosus in the preterm infant. Springer Medizin Verlag, Heidelberg, pp 19–23

Peirone A, Abdullah MM et al (2002) Echocardiographic evaluation, management and outcomes of bilateral arterial ducts and complex congenital heart disease: 16 years' experience. Cardiol Young 12(3):272–277

Popat H, Kluckow M (2012) Noninvasive assessment of the early transitional circulation in healthy term infants. Neonatology 101(3): 166–171

Rauch R, Koch A et al (2001) Echocardiographic findings before and after interventional occlusion of persistently patent ductus arteriosus with the amplatzer duct occluder. Ultraschall Med 22(6): 279–283

Reller MD, Colasurdo MA et al (1990) The timing of spontaneous closure of the ductus arteriosus in infants with respiratory distress syndrome. Am J Cardiol 66(1):75–78

Reller MD, Ziegler ML et al (1988) Duration of ductal shunting in healthy preterm infants: an echocardiographic color flow Doppler study. J Pediatr 112(3):441–446

Rychik J, Bush DM et al (2000) Assessment of pulmonary/systemic blood flow ratio after first-stage palliation for hypoplastic left heart syndrome: development of a new index with the use of doppler echocardiography. J Thorac Cardiovasc Surg 120(1):81–87

Rychik J, Gullquist SD et al (1996) Doppler echocardiographic analysis of flow in the ductus arteriosus of infants with hypoplastic left heart syndrome: relationship of flow patterns to systemic oxygenation and size of interatrial communication. J Am Soc Echocardiogr 9(2):166–173

Santos MA, Moll JN et al (1980) Development of the ductus arteriosus in right ventricular outflow tract obstruction. Circulation 62(4): 818–822

Schranz D, Michel-Behnke I et al (2010) Stent implantation of the arterial duct in newborns with a truly duct-dependent pulmonary circulation: a single-center experience with emphasis on aspects of the interventional technique. J Interv Cardiol 23(6):581–588

Schwedler G, Lindinger A et al (2011) Frequency and spectrum of congenital heart defects among live births in Germany : a study of the Competence Network for Congenital Heart Defects. Clin Res Cardiol Off J German Cardiac Soc 100(12):1111–1117

Skinner J (2000a) Ductal shunting. In: Skinner J, Alverson D, Hunter S (eds) Echocardiography for the neonatologist. Churchill Livingstone, London

Skinner J (2000b) Pulmonary arterial pressure. In: Skinner J, Alverson D, Hunter S (eds) Echocardiography for the neonatologist. Churchill Livingstone, London

Ten Harkel AD, Blom NA et al (2002) Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. Chest 122(4):1471–1477

van de Bor M, Verloove-Vanhorick SP et al (1988) Patent ductus arteriosus in a cohort of 1338 preterm infants: a collaborative study. Paediatr Perinat Epidemiol 2(4):328–336