Aortopulmonary Window

6

Contents

6.1 Anatomy and Haemodynamics

Aortopulmonary window (APW) is a rare congenital cardiovascular malformation which is characterized by a connection between the ascending aorta and the main pulmonary artery (Anderson et al. 2010; Ho et al. 1994). APW is differentiated from truncus arteriosus communis (see Chap. 13) by the fact that patients with APW have two separate ventricular outflow tracts and semilunar valves, while the latter is characterized by a common truncal valve (Anderson et al. 2008). The incidence of APW has been quoted as 0.59 % of congenital heart defects (Anderson et al. 2008; Kutsche and Van Mierop 1987). Classification of APWs is descriptive according to the localization within the main pulmonary artery and extension towards the semilunar valves and the pulmonary bifurcation (Ho et al. 1994). Proximal defects have only a minor rim towards the semilunar valves (Fig. 6.1). Intermediate-type defects are located halfway between the pulmonary valve and bifurcation. The distal type extends up to the origin of the right pulmonary artery from the bifurcation. This type may also be associated with aberrant origin of the right pulmonary artery from the ascending aorta. The confluent type extends over the length of the main pulmonary artery without well-developed margins at the lower and superior end of the defect. Embryologic development of APW has been explained as the result of failing closure of the embryonic aortopulmonary foramen (Anderson et al. 2010, 2012).

Haemodynamically APW causes left to right shunting at the level of the great arteries, resulting in volume load of the pulmonary artery, left atrium and left ventricle. Furthermore large defects allow equalization of systemic and pulmonary artery pressures with consecutive pulmonary hypertension. In the majority of cases, APW is associated with additional significant congenital cardiac malformations (Apitz et al. 2007; Bagtharia et al. 2004; Ho et al. 1994; Konstantinov et al. 2006; Kutsche and Van Mierop 1987). Those malformations include interrupted aortic arch (12–13 %), coarctation of the aorta (10 %), right aortic arch (9–10 %), tetralogy

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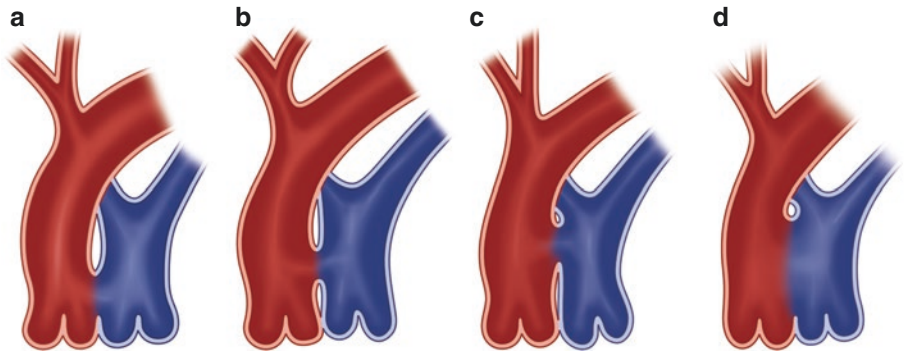

Fig. 6.1 Diagram showing the different types of aortopulmonary windows including the proximal (**a**), intermediate (**b**) and distal type (**c**). In the confluent type, the whole length of the pulmonary trunk is involved (**d**) (Redrawn with kind permission after (Ho et al. 1994))

of Fallot (5–6 %) and aberrant origin of a coronary artery from the pulmonary artery (5 %). Quite frequent are ventricular septal defects (17 %), atrial septal defects and patent ductus arteriosus (36–38 %). In rare cases, APW may be associated with pulmonary atresia and VSD, tetralogy of Fallot, aortic atresia, tricuspid atresia and transposition of the great arteries (Apitz et al. 2007; Bagtharia et al. 2004; Kiran et al. 2008; Kutsche and Van Mierop 1987). The combination of distal aortopulmonary window with aortic origin of the right pulmonary artery, patent ductus arteriosus and interrupted aortic arch has been termed as "Berry syndrome" (Alva-Espinosa et al. 1995; Berry et al. 1982). *The association of APW with significant additional congenital heart disease has important diagnostic implications*, *since APW may remain undetected*, *if echocardiography focuses exclusively on the more obvious associated cardiac malformation* (Kiran et al. 2008). This is especially true for large ventricular septal defects: without a systematic approach of echocardiographic examination, the APW may easily avoid detection during preoperative evaluation, since the apparent findings can be attributed to the interventricular communication (Apitz et al. 2007; Kiran et al. 2008).

The prognosis of APW depends upon timely detection and surgery before irreversible pulmonary vascular damage has developed (Bagtharia et al. 2004). Large defects require surgical closure within the first 3 months of life.

Diagnosis of aortopulmonary window is facilitated, if several general rules are kept in mind:

1. APW belongs to the differential diagnosis of malformations associated with severe left atrial and left ventricular volume load including large VSD and large patent ductus arteriosus.
2. APW is part of the differential diagnosis of significant diastolic run-off from the aorta including widely patent ductus arteriosus, truncus arteriosus communis, large arteriovenous fistulae, aorto-left ventricular tunnels, significant aortic valve regurgitation and ruptured aneurysms of sinus valsalvae.
3. APW window is frequently associated with aortic arch abnormalities including interruption of the aortic arch, coarctation and right aortic arch.
4. Patients with interrupted aortic arch type A and intact ventricular septum very likely will have APW.
5. Aberrant origin of a coronary artery from the pulmonary artery, associated with large APW, will be asymptomatic and undetectable by ECG changes prior to surgical repair. It has to be excluded always in the preoperative assessment, since this anomaly would result in vital threat of the patient following surgical closure of the APW.

6.2 Two-Dimensional Echocardiography

Significant dilatation of the left atrium and left ventricle is encountered in infants with large APW (Videos 6.1 and 6.2). The optimal plane to verify enlargement of the left atrium and left ventricle is the apical four-chamber view (Fig. 6.2). Visualization of the APW is well possible by 2D echocardiography (Balaji et al. 1991). It is best accomplished in the parasternal short axis of the great arteries (Video 6.3). In the presence of a large APW, the aorta and main pulmonary artery do not appear as two separate structures but rather resemble to a horizontal figure 8 (Fig. 6.3, Video 6.4). Frequently there is an increased echogenicity of the borders of the defect termed T-artifacts (Balaji et al. 1991). Colour Doppler in this plane should be used to confirm shunting across the defect and to rule out artificial dropout (Fig. 6.4, Videos 6.5 and 6.6).

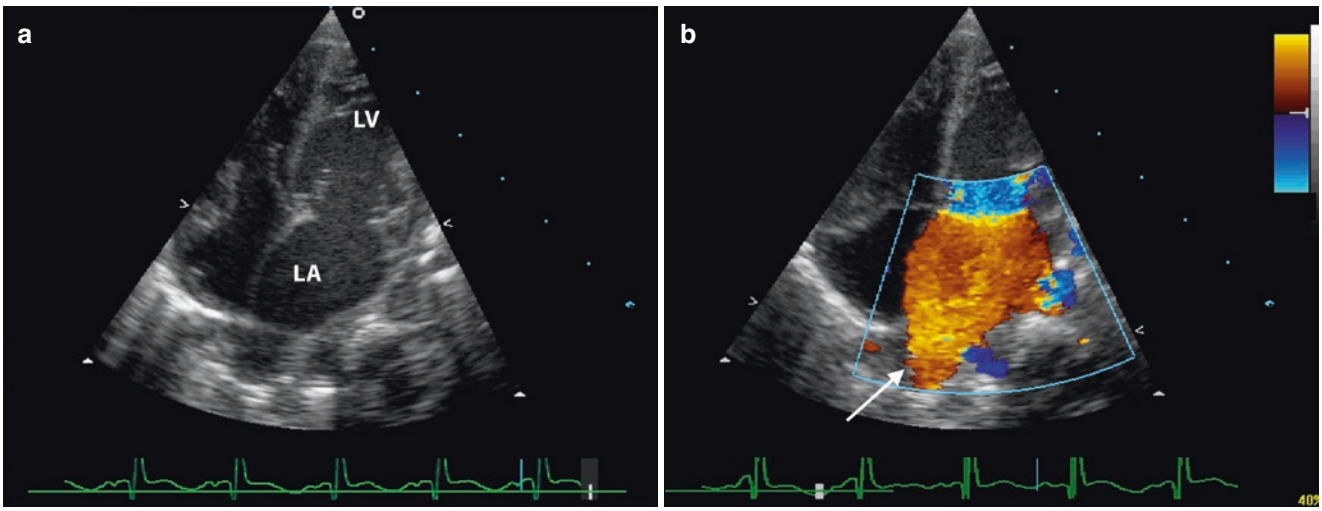

Fig. 6.2 Four-chamber view in a 4-week-old patient showing significant enlargement of the left atrium (*LA*) and ventricle (*LV*) due to significant shunting of a large AP window (**a**); colour Doppler shows enlarged pulmonary veins (*arrow*) and increased flow to the left atrium (**b**)

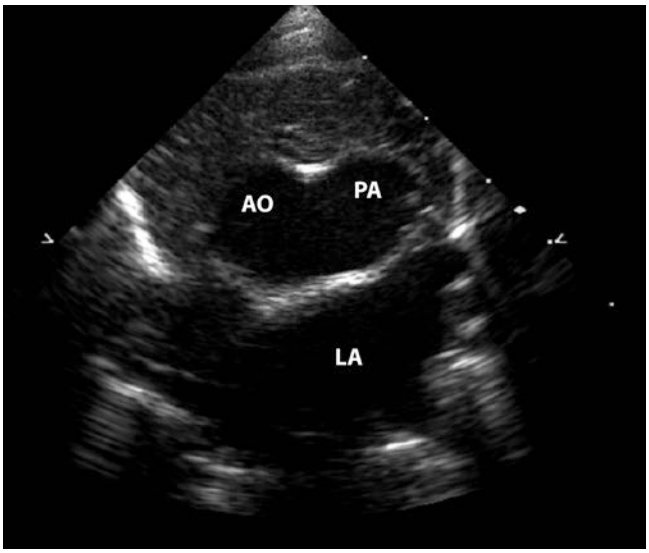

Fig. 6.3 Parasternal short axis (**a**) showing a large APW between the aorta (*AO*) and pulmonary artery (*PA*)

It should be kept in mind however that the diagnosis even of a large APW may be missed (Fig. 6.5) if the defect is located ventrally and if the echo plane in the parasternal short axis is oriented too far posteriorly (Apitz et al. 2007). Special attention should be paid to the distal extension of the APW and to its relation to the origin of the right pulmonary artery.

Distal as well as confluent type defects may extend up to the origin of the right pulmonary artery from the bifurcation (Fig. 6.6) and may be associated with origin of the right pulmonary artery from the right lateral aspect of the ascending aorta (Alva-Espinosa et al. 1995; Berry et al. 1982; Ho et al. 1994). The subcostal coronal and RAO views are helpful to define the inferior borders of the defect and to clarify its distance to the level of the aortic and the pulmonary valve (Fig. 6.7 and 6.8, Videos 6.7 and 6.8).

6.3 Colour Doppler Echocardiography

Although the diagnosis of APW can be established primarily by 2D echocardiography, colour Doppler is important to confirm shunting across the defect (Apitz et al. 2007; Horimi et al. 1992). Colour Doppler interrogation of the defect is best accomplished in the parasternal short axis, but it can be performed in the subcostal coronal and subcostal RAO view as well (Fig. 6.4 and 6.6, Videos 6.5, 6.6 and 6.8). Although shunting across the APW in these planes occurs at an unfavourable angle for Doppler interrogation, high blood flow velocities and turbulent flow facilitate detection of the defect. Shunting across the APW depends upon the size of the defect and on the resistances of systemic and pulmonary circulation. In the presence of an elevated pulmonary vascular resistance, the velocity of shunting will be low, and colour Doppler may be of limited help to confirm or exclude the suspected defect. Much easier is the situation in the presence of a small restrictive APW. In these cases, there is a significant gradient between systemic and pulmonary circulation, and colour Doppler interrogation will detect a highly turbulent and accelerated flow from the aorta into the pulmonary artery.

Since large- or medium-size APW results in significant diastolic run-off from the ascending aorta to the pulmonary artery, colour Doppler examination of the aortic arch and the descending aorta in patients with large APW reveals significant retrograde flow directed towards the ascending aorta in diastole (Fig. 6.9, Video 6.9). On the other hand, the resulting regurgitant flow in the aortic arch results in an increase of systolic flow velocities as well (Fig. 6.9). Careful

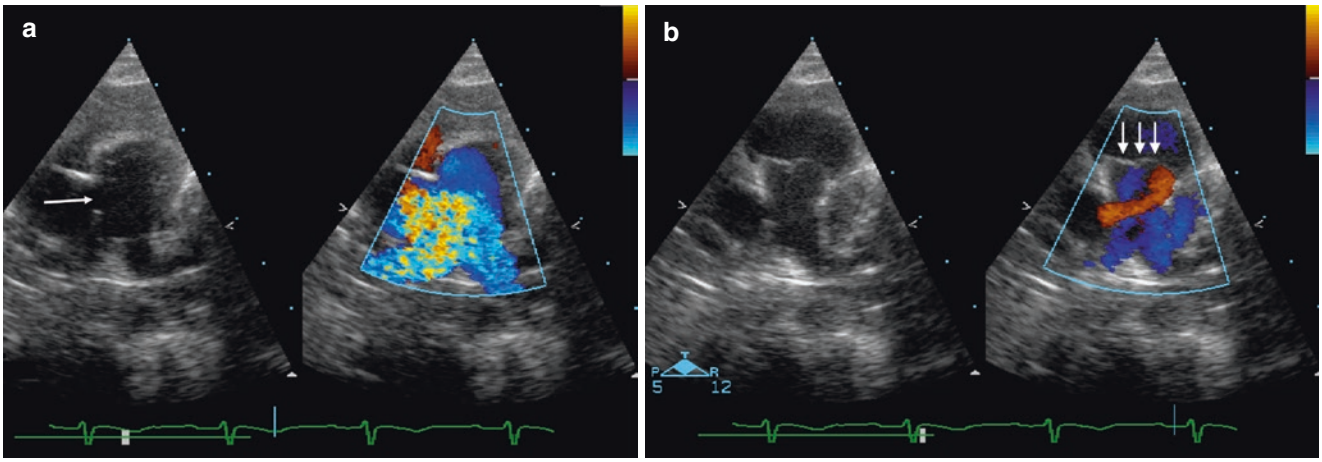

Fig. 6.4 Colour Doppler in an infant with medium-size intermediate AP window (*arrow*) shows LR-shunting both in systole (**a**) and in diastole (**b**). In diastole the jet extends down to the level of the closed pulmonary valve (*arrows*)

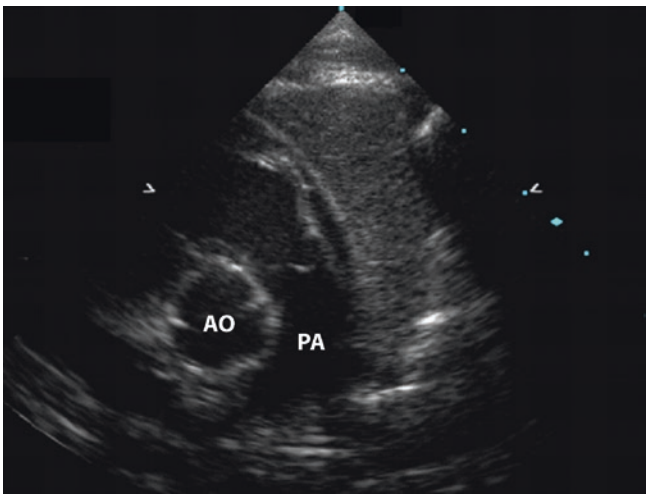

Fig. 6.5 In a patient with large AP window, the parasternal short axis at the level of the aortic root (*AO*), obtained with a posterior orientation of the transducer, apparently displays a normal pulmonary artery (*PA*) and no defect

inspection of the aortic isthmus by 2D echocardiography is required therefore to exclude anatomic obstruction.

APW can be associated with anomalous origin of a coronary artery from the pulmonary artery (Bagtharia et al. 2004). Colour Doppler of the aortic root is helpful to ensure normal origin of both coronary arteries (Fig. 6.10).

Furthermore colour Doppler plays a significant role in the diagnosis and confirmation of associated cardiovascular malformations. These include atrial and ventricular septal defects, coarctation of the aorta and interruption of the aortic arch, abnormalities of the head and neck vessels as well as patent ductus arteriosus (Fig. 6.11).

6.4 Pulsed Wave and Continuous Wave Doppler

PW Doppler and CW Doppler do not play a very prominent role in the diagnosis of large APW. In children with large APW, there is turbulent flow in the main and in the branch pulmonary arteries due to the influx through the defect. Since large defects cause equalization of blood in the systemic and pulmonary circulation, LR-shunting will occur without a significant gradient at flow velocities of 1 – 2 m/s. Flow velocities in this range can be depicted by pulsed wave Doppler. Continuous wave Doppler is required for those rare instances of small, restrictive APW, which are associated with significantly elevated flow velocities, due to a substantial gradient between systemic and pulmonary circulation.

In patients with restrictive AP window, pulmonary artery pressure can be calculated noninvasively according to the formula:

Systolic pressure pulmonary artery = Systolic blood pressure – pressure gradient across APW

Reliable noninvasive quantification of systolic pulmonary artery pressure is possible only by exact determination of the maximal velocity across the APW, which requires as a precondition a reasonable insonation angle of the interrogating Doppler beam and the jet across the defect.

PW Doppler evaluation of diastolic flow in the systemic circulation is a valuable tool to assess the haemodynamic significance of LR-shunting. Interrogation of flow in the descending aorta in the presence of a large APW shows retrograde flow directed towards the ascending aorta similar to

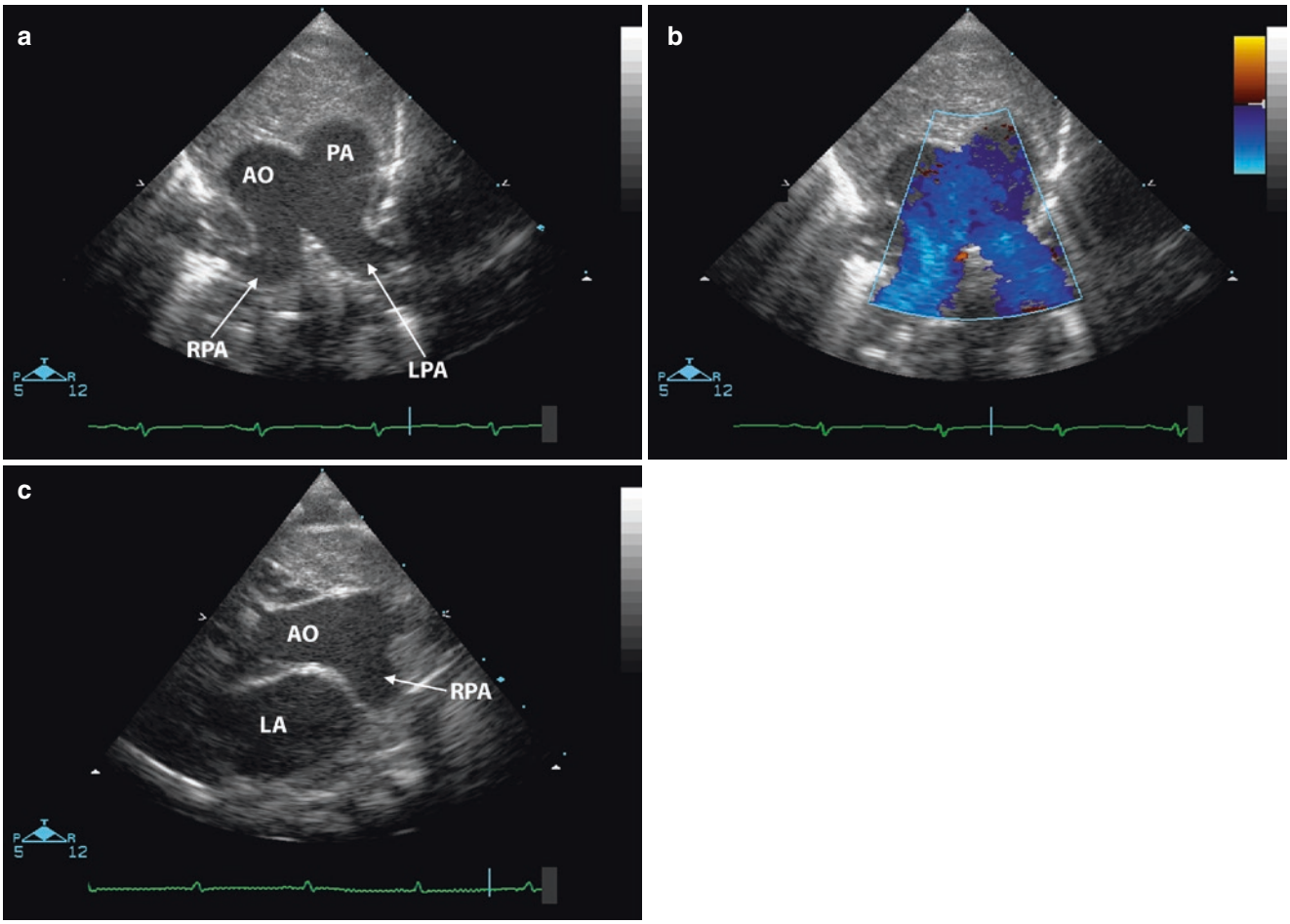

Fig. 6.6 Parasternal short axis view at the level of the pulmonary bifurcation (**a**) shows a large distal AP window with origin of the RPA from the aorta (*AO*) and LPA from the pulmonary artery (*PA*). Colour Doppler shows low velocity flow across the defect (**b**). The parasternal long axis (**c**) depicts the direct communication of the ascending aorta (*AO*) with the right pulmonary artery (*RPA*)

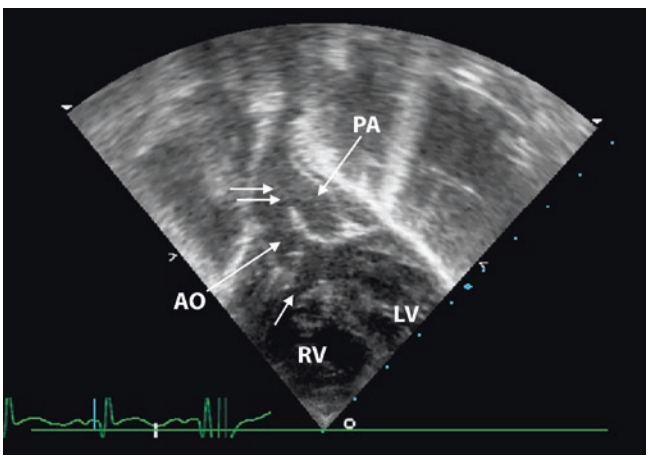

Fig. 6.7 In this infant with large distal type AP window, the subcostal coronal plane (**a**) shows the large defect (*small arrows*) between the aorta (*AO*) and pulmonary artery (*PA*). In addition the infant has a perimembranous VSD (*single arrow*). *RV/LV* right, left ventricle

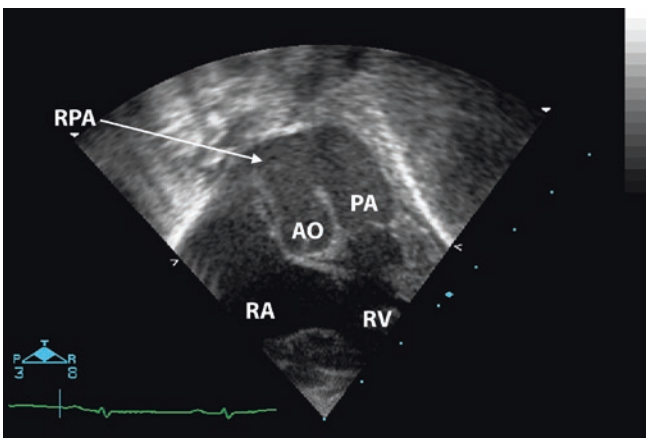

Fig. 6.8 Subcostal RAO view in a patient with large distal AP window shows extension of the defect into the bifurcation connecting the proximal RPA with the posterior wall of the aorta. *RA/RV* right atrium, right ventricle

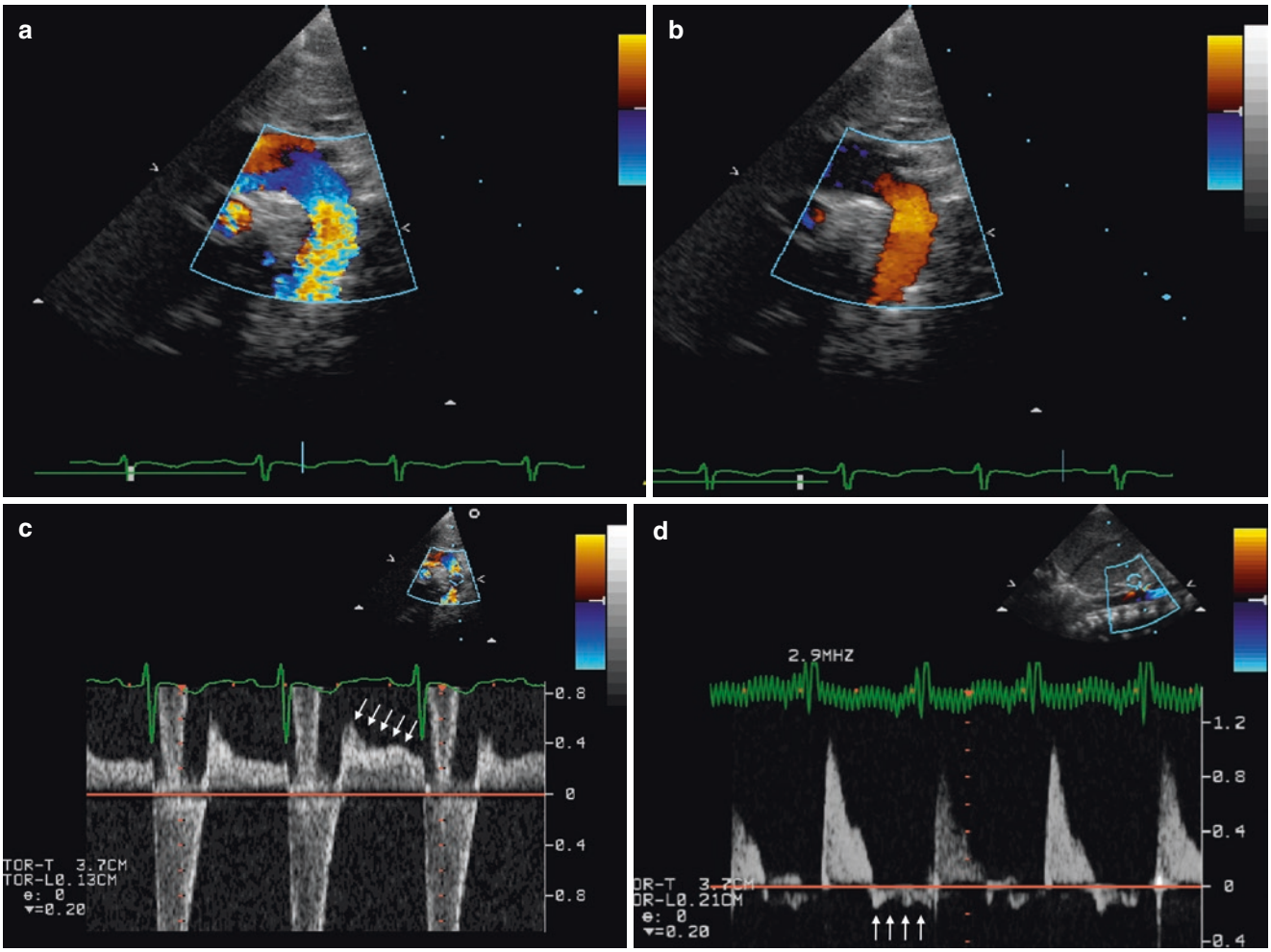

Fig. 6.9 Suprasternal long axis in a patient with large AP window. Colour Doppler reveals accelerated and turbulent flow in the aortic isthmus during systole despite the absence of coarctation (**a**). The diastolic frame reveals marked retrograde flow due to diastolic run-off from the ascending aorta (**b**). Confirmation of significant retrograde diastolic flow (*arrows*) is obtained by PW Doppler interrogation of the descending aorta (**c**). Diastolic retrograde flow (*arrows*) is also apparent in the PW Doppler of the coeliac artery (**d**)

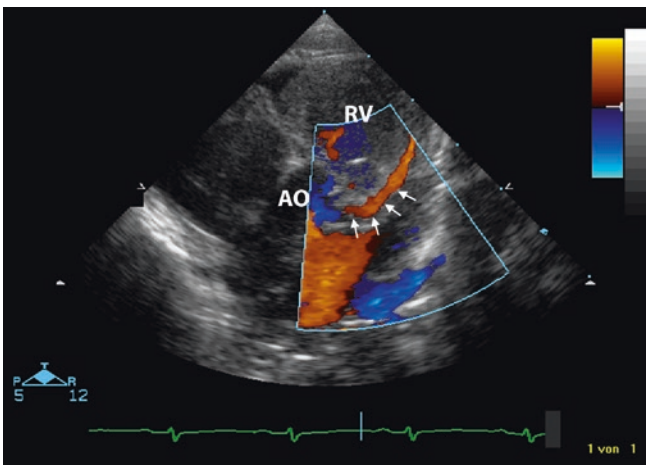

Fig. 6.10 Normal origin of the left coronary artery (*arrows*) demonstrated by colour Doppler examination of the aortic root (*AO*) in the parasternal short axis view. *RV* right ventricle

patients with significant aortic regurgitation (Fig. 6.9). This flow pattern may show considerable increase of diastolic run-off during the first few weeks of life, due to physiologic decrease in pulmonary vascular resistance with consecutive increase of LR-shunting in this time period. Diastolic run-off from the aorta to the pulmonary arteries in patients with large APW is also apparent at Doppler examination of systemic arteries: leakage from the aortic "Windkessel" results in diminished or even negative diastolic flow in abdominal arteries (e.g. the coeliac artery) or cerebral vessels (e.g. the anterior cerebral artery) (Fig. 6.9).

While Doppler interrogation of tricuspid regurgitation is usually not required to confirm elevated right ventricular pressures in children with large APW, since pulmonary hypertension can always be assumed in the presence of a large defect, it may be valuable in patients with medium-size restrictive defects, if there is uncertainty regarding right

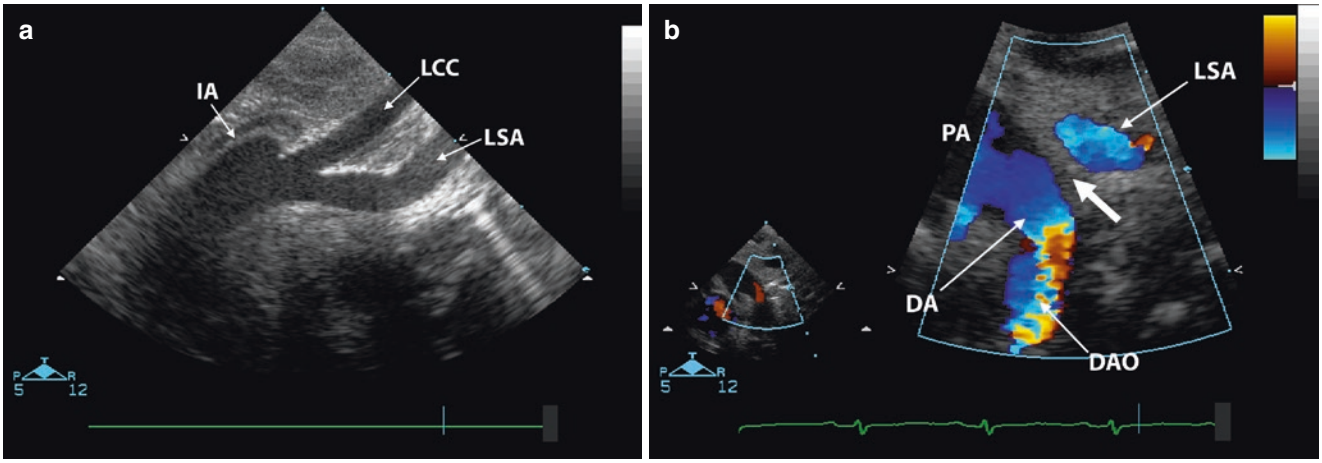

Fig. 6.11 Suprasternal long axis in this neonate with large AP window shows interruption of the aortic arch (IAA type A) distal to the left subclavian artery (**a**). Colour Doppler in the ductal view exhibits ductal perfusion of the descending aorta (*DAO*) and confirms interruption of the arch (*large arrow*) distal to the left subclavian artery (**b**). *IA* innominate artery, *LCC* left common carotid artery, *LSA* left subclavian artery, *PA* pulmonary artery, *DA* ductus arteriosus

ventricular or pulmonary artery pressures. Pulsed wave or continuous wave Doppler interrogation of tricuspid regurgitation requires regurgitant flow with a jet that is powerful enough to create an acceptable signal (see Chap. 1). Another possibility for noninvasive determination of systolic pulmonary artery pressure is provided in the presence of a patent ductus arteriosus. The gradient across the ductus can be used for calculation of systolic pulmonary artery pressure according to the same formula mentioned above (see Chap. 5).

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