Aortopulmonary Window

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G. Deepak Gowda and B. C. Hamsini

High Yield Facts

- Aortopulmonary window (APW) is a rare, surgically correctable congenital cardiac anomaly representing 0.2–0.3% of all congenital cardiac lesions.
- Echocardiography is generally diagnostic and has replaced angiography as the gold standard.
- Patients typically present with symptoms of congestive heart failure (tachypnea, diaphoresis, failure to thrive, or recurrent respiratory infections).
- Operative treatment is indicated as soon as the diagnosis is established, regardless of the patient's age.
- Unless there is a right-to-left shunt despite oxygen administration, no patients with APW should be denied the operation.
- Elective repair is advised before age 3 months.
- Long-term outcome after operative correction is excellent regardless of age or pulmonary vascular resistance.

Introduction

The aortopulmonary window (APW) is a communication between the ascending aorta and the pulmonary trunk and/or right pulmonary branch, in the presence of both separate sigmoid valve planes, which differentiates it from truncus arteriosus (TA). APW is a rare, surgically correctable congenital cardiac anomaly. This lesion represents between 0.2% and 0.3% of all congenital cardiac lesions [1, 2]. It was first described by Eliotson in a clinicopathologic discussion given at St. Thomas' Hospital in London in 1830 [3]. Dodds and Hoyle gave the first clinical diagnosis in 1949 [4]. Gross did first successful ligation in 1952 [5]. Subsequent classification was done by Mori et al. in 1978 [6] and by Richardson and associates in 1979 [7]. This lesion is associated with rapid progression of pulmonary arterial hypertension (PAH) unless it is corrected surgically.

Embryology

The beginning of the formation of the aortopulmonary septum can be traced to the fifth week of development, when pairs of opposing ridges appear in the truncus, called cushions or truncus swellings. They grow towards the aortic sac, twisting counterclockwise around each other, foreshadowing the spiral course of the future septum [8]. After complete fusion, the ridges form the aortopulmonary septum, dividing the truncus into separate aortic and pulmonary channels. Neural crest cells migrate to the outflow region of the heart contributing to the formation of the aortopulmonary septum. There are many proposed mechanisms explaining outflow tract defects, including direct insults to the truncus swellings, insults to neural crest cells that disrupt signaling to truncus swellings or insults to neural crest cells that disrupt their contribution to fusion. All proposed mechanisms can potentially disrupt the whole process of outflow tract formation. Abnormalities in neural crest migration and signaling are linked to velocardiofacial syndrome, because neural crest cells also contribute to the craniofacial development. Unlike other conotruncal malformations, such as truncus arteriosus or interrupted aortic arch (IAA), the risk of associated chromosomal abnormalities in APW, including 22q11 deletion, seems low [2].

G. D. Gowda (✉)
Department of CardioThoracic and Vascular Surgery, Star Pinnacle Heart Centre, Health city, Visakhapatnam, Andhra Pradesh, India
e-mail: drdeepakgowda@gmail.com

B. C. Hamsini
Department of Radiology, Sri Sathya Sai Institute of Higher Medical Sciences, Puttaparthy, Andhra Pradesh, India

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Classification

Several classifications have been proposed for APW. The most often used is that of Mori et al. [6], which divides it into type I, or proximal (70%) (the defect is circular, located in a zone equidistant between the sigmoid valve plane and the pulmonary bifurcation); type II, or distal (25%) (of spiral form, it affects the trunk and origin of the right pulmonary artery), and type III (5%) (complete defect of the aortopulmonary septum).

Ho and associates [9] modified this classification in a way that is more useful to interventional cardiologists considering transcatheter device closure of APW. They keep the basic terminology from Mori and associates as defined above but add additional description. Proximal defects are noted to have little inferior rim separating the APW from the semilunar valves. Distal defects are noted to have a well-formed inferior rim but little superior rim. Total defects are called confluent defects with little superior and inferior rims. Finally, intermediate defects are defects with adequate superior and inferior rims; this type is obviously the group most suited for device closure.

Classification of APW as proposed by Richardson et al. [7] includes (A) Type I is a "typical" APW between the posteromedial wall of the ascending aorta and the main pulmonary artery. (B) Type II is located between the posterior wall of the ascending aorta and the origin of the right pulmonary artery. (C) Type III defects are defined as an anomalous origin of the right pulmonary artery from the posterolateral wall of the ascending aorta.

Classification of APW used by the Society of Thoracic Surgeons [10] includes (A) Type I—proximal defect; (B) Type II—distal defect; (C) Type III—total defect; and (D) Type IV—intermediate defect (as described by Ho et al. [9]). In proximal extension of the APW, the ostium of the right coronary artery may come from the APW or originate in close proximity to it.

Associated Conditions

APW can be associated with other congenital cardiac defects in 25–65% of cases. The associated conditions include patent ductus arteriosus (PDA), left superior vena cava, right aortic arch, ventricular septal defect (VSD), patent foramen ovale (PFO), supramitral ring, mitral regurgitation and interrupted aortic arch (IAA) mostly type A [11]. As reported by Braunlin and coworkers IAA is associated with 43% of cases of APW and coarctation of aorta in 14% cases [12]. Sometimes APW can be associated with tetralogy of fallot and anomalous origin of coronary artery. Anomalous origin of right pulmonary artery from ascending aorta is a rare congenital lesion with a high mortality and morbidity. The natural history of this lesion is that of progressive heart failure with a 70% mortality rate at 1 year compared with an 84% survival rate at 1 year with surgical repair. The combination of an APW, aortic origin of the right pulmonary artery, intact ventricular septum, and interruption or coarctation of the aortic arch is termed the *Berry syndrome*.

Pathophysiology

The lesion, if large, resembles a large PDA. When pulmonary vascular resistance (PVR) falls the patient develops severe congestive heart failure that often causes early death. If they survive, pulmonary vascular disease occurs [13–15]. Occasionally, if the window is small or there is stenosis of one or both pulmonary arteries, neither heart failure nor pulmonary vascular disease occurs. Under 1 year of age there is usually a large left-to-right shunt and pulmonary and aortic systolic pressures are equal or almost equal, but PVR is usually but not always near normal [14]. In older children pulmonary hypertension is still present, but shunts vary in size, depending on PVR. This lesion resembles a large ventricular septal defect in the time course of pulmonary vascular disease.

Clinical Features

Clinical manifestations in patients with an aortopulmonary window depend on the size of the defect and associated cardiac anomalies.

Symptoms

Patients typically present with symptoms of congestive heart failure (tachypnea, diaphoresis, failure to thrive, or recurrent respiratory infections).

Signs

On examination, the left precordium is prominent because of marked cardiomegaly. Loud continuous murmur with systolic accentuation which is heard best at the left upper sternal border. The second heart sound may be accentuated and narrowly split. In a large shunt, the diastolic blood pressure is usually reduced, with a concomitant widened pulse pressure and, on occasion, a water-hammer pulse may be present.

Diagnosis

Chest radiograph and electrocardiogram (ECG) show evidence of left and right ventricular enlargement and increased pulmonary blood flow. Left atrial enlargement as a result of increased pulmonary blood flow is usually prominent. Echocardiography (Fig. 95.1) is generally diagnostic and has

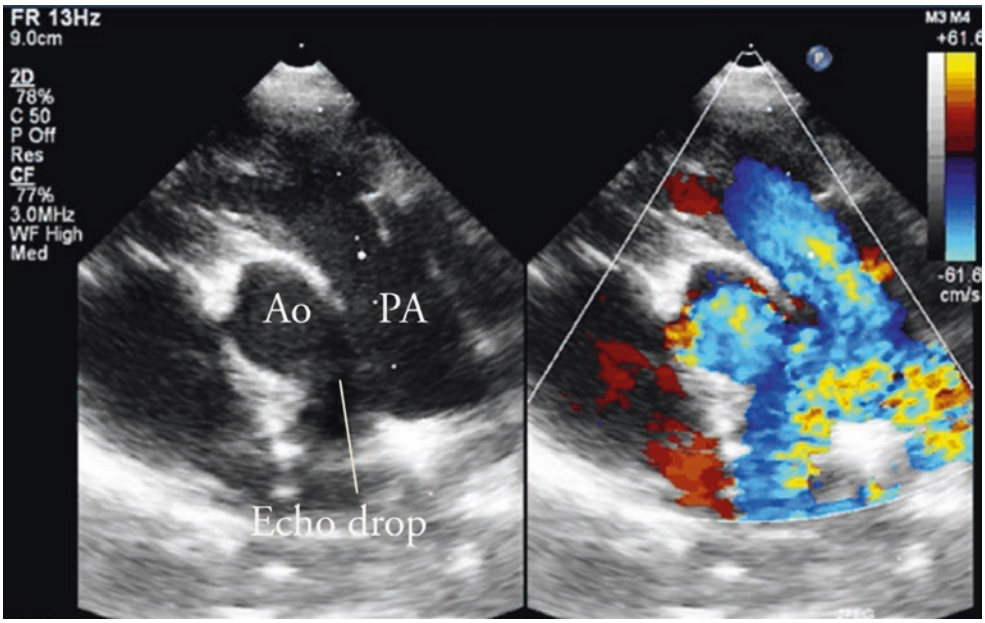

Fig. 95.1 2D echocardiography in modified short axis view shows echo dropout between aorta and pulmonary artery. *Ao* aorta, *PA* pulmonary artery

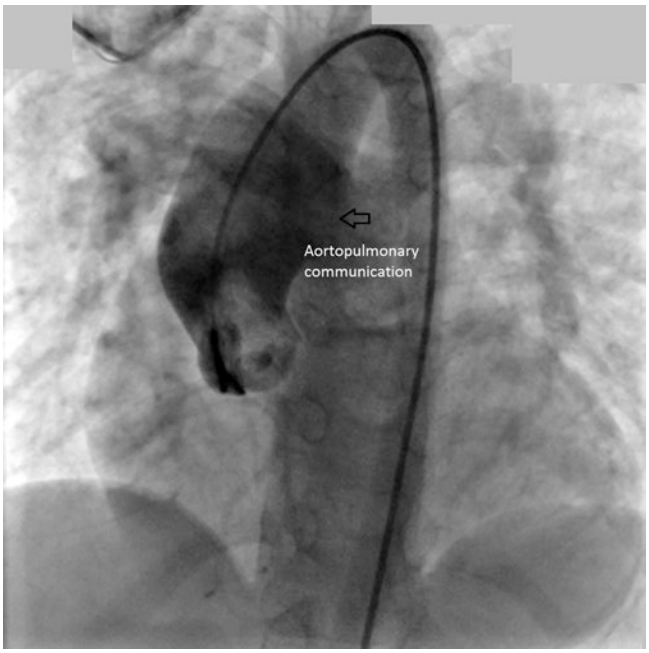

Fig. 95.2 Catheterization study (catheter course from descending aorta to arch and ascending aorta) showing aortopulmonary communication

replaced angiography as the gold standard [16]. The most useful echocardiographic views to delineate the margins of the window include the suprasternal long axis, the subcostal coronal plane through the pulmonary trunk, and the high parasternal short plane cephalad to the aortic valve.

Magnetic resonance imaging is an emerging technique that is capable of clarifying anatomy with great detail [17].

Cardiac catheterization (Fig. 95.2) is indicated to determine operability in children diagnosed beyond infancy with features suggestive of PAH and also in cases of associated coronary abnormality to delineate the coronary anatomy. The differential diagnosis of APW includes PDA, truncus arteriosus, VSD with aortic insufficiency, and ruptured sinus of Valsalva aneurysm.

Natural History

Patients with large APW generally do not survive beyond childhood. Of untreated patients, 40% die within the first year of life [18]. Untreated patients generally develop pulmonary vascular obstructive disease.

Only 32 patients over 20 years of age have been described [2, 4, 5, 11, 15, 19–27]. The oldest was 58 years of age [15], had pulmonary vascular disease, and died from an unassociated aortic valve lesion. Patients over 10 years of age seldom have associated complex heart disease, although they may have simpler lesions such as a bicuspid aortic valve, coarctation of the aorta or a VSD. They usually have pulmonary vascular disease unless the defect is very small.

Infective endocarditis is rare [23, 27].

In part the natural history depends on the severity of associated lesions. Furthermore, because an APW is rare, it is particularly difficult to estimate the natural history. Because of the poor prognosis associated with this condition, it is recommended that all patients with APW undergo repair.

Management

Medical and Interventional Therapy

The medical therapy for an APW is the same as for a large PDA. There have been reports on successful transcatheter closure of APW [25, 28, 29] but the success using this

approach requires secure positioning of the device to occlude the orifice of the APW without impinging on the ostium of the left coronary artery. Following device closure, patients need to be followed for the development of aortic regurgitation at the time of the *procedure as well as long-term.*

Surgical

Indications

Operative treatment is indicated as soon as the diagnosis is established, regardless of the patient's age. Unless there is a right-to-left shunt despite oxygen administration, no patients with APW should be denied the operation [11]. Elective repair is advised before age 3 months.

Technique of Operation

There are several surgical techniques described with success (Table 95.1) but the most widely accepted optimal approach is the one described by Johansson and colleagues (sandwich type repair) [36].

Table 95.1 Evolution of surgery for aortopulmonary window

Year	Surgeon	Method
1948	Gross [5]	Ligation
1953	Scott and Sabiston [30]	Division without cardiopulmonary bypass
1957	Cooley et al. [31]	Division with cardiopulmonary bypass
1966	Putnam and Gross [32]	Transpulmonary patch closure
1968	Negre et al. [33]	Anterior sandwich patch closure
1968	Wright et al. [34]	Transaortic suture closure
1969	Deverall et al. [35]	Transaortic patch closure
1978	Johansson et al. [36] Aberg [37]	Single patch sandwich type repair
1987	Shatapathy et al. [38]	Pulmonary artery flap closure
1992	Matsuki et al. [39]	Anterior pulmonary artery flap closure

APW is approached through standard midline sternotomy. Diagnosis confirmed when separate semilunar valve "anuli" can usually be verified by finding a dimple between the two great arteries where they arise from the heart (Fig. 95.3a) which differentiates it from truncus arteriosus.

Cardiopulmonary bypass is established using bicaval venous and distal aortic cannulation. Bilateral pulmonary arteries are snared and kept. Most cases are dealt under moderate hypothermia unless the infant weighs less than 2.5 kg wherein deep hypothermic circulatory arrest may be used. Aorta is cross clamped and antegrade cardioplegia is delivered after snaring the bilateral branch pulmonary arteries. Left atrium can be vented through the PFO after right atriotomy. An incision is made in the anterior wall of the communication between the aorta and the pulmonary trunk (Fig. 95.3b). A PTFE patch is then sutured to the posterior wall of the defect. The suture line is subsequently carried anteriorly to incorporate the patch in the closure of the arteriotomy.

In the transaortic approach [35] for the repair of APW, a longitudinal aortotomy is made in the ascending aorta. For simple proximal (type I) defects, closure is readily accomplished using a polyester, PTFE or pericardial patch. Care must be taken to identify the left coronary orifice. For distal (type II) defects, a more extensive patch must be fashioned. When there is considerable unroofing of the right pulmonary artery, the creation of an intra-aortic baffle and closing the aortotomy with an elliptical patch may suffice. Total (type III) and intermediate type defects are repaired using similar techniques.

In pulmonary flap technique [38], an anterior flap of pulmonary artery is incised in continuity with the aorta along the anterior border of the window. The anterior flap is then sewn to the posterior margin of the APW, thereby closing the defect in the aorta. The resultant defect in the pulmonary artery is then closed with a patch (autologous or heterologous).

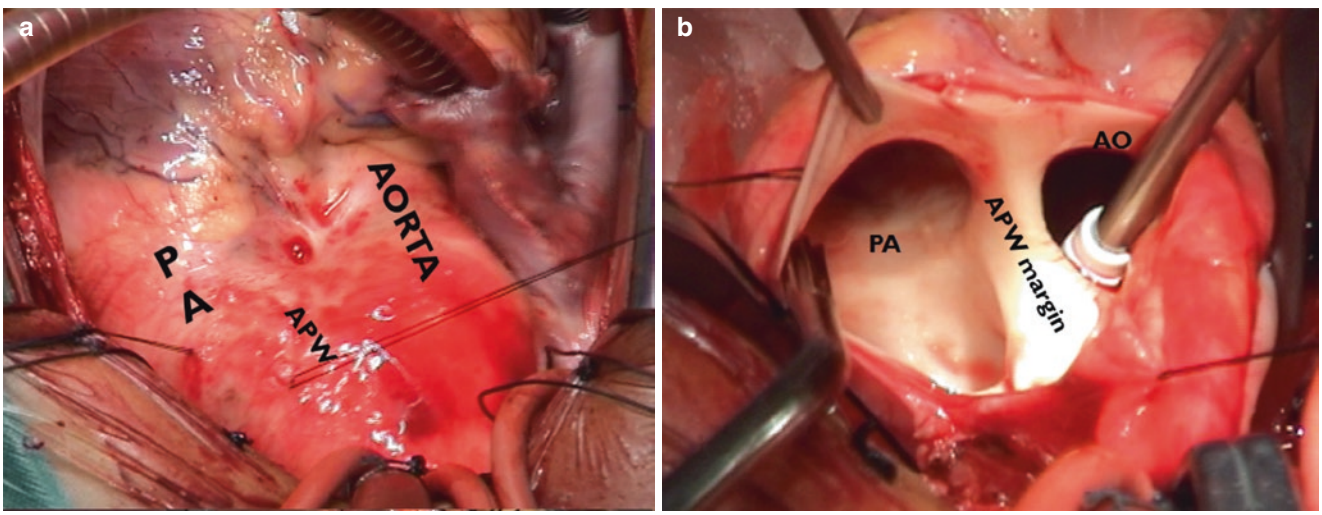

Fig. 95.3 Operative photograph showing (**a**) large sized aortopulmonary window (APW) externally (**b**) after opening the APW

Kitagawa and associates [40] reported an alternative technique to repair a distal APW with extensive unroofing of the right pulmonary artery. In this approach, the ascending aorta is transected at the distal extent of the window, and the right pulmonary artery is excised along with a strip of posterior aorta in continuity with the window and pulmonary trunk. The defect in the pulmonary artery is then repaired either primarily or using a patch, and the aorta is reconnected primarily. This technique has the advantage of avoiding any potential problems resulting from an intra-aortic baffle.

Associated cardiac anomalies are repaired in a same setting as far as possible.

Postoperative Management

Intraoperative transesophageal echocardiography is useful for confirming complete closure of the defect and ensuring that the pulmonary arteries have not been narrowed. Management of PAH post operatively in patients presenting later in life is the most crucial part. PA catheter is best suited in managing such patients. Inhaled nitric oxide, phosphodiesterase inhibitors, endothelin receptor antagonist, fentanyl and muscle relaxants, and mild hyperventilation with 100% oxygen can be used in preventing pulmonary hypertensive crisis.

Results

The results of surgical treatment of APW demonstrate low risk, even if the APW is associated with major cardiac anomalies. Surgical options are tailored to the anatomy and the size of the defect. Whenever possible, continuity ligation, division and suture should be done with cardiopulmonary bypass on standby. This approach reduces the potential complications of cardiopulmonary bypass, avoids blood requirements, and reduces the hospital stay, thus leading to lower morbidity. Long-term outcome after operative correction is excellent regardless of age or PVR [11]. For patients with complex associated anomalies, the prognosis is determined largely by the presence of associated anomalies.

Conclusion

APW is a rare but well identified and surgically correctable anomaly. Operative repair should be offered as soon as the diagnosis is established, regardless of the patient's age. Irreversible PAH with a right to left shunt despite oxygen administration is the only contraindication for surgery. Various surgical techniques can be applied depending on the size of the communication. Associated arch anomalies may require technically challenging approaches and surgical strategies. Early and long-term outcomes after surgical correction are excellent regardless of age or PVR.

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