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Surgical Repair of Aortopulmonary Window: Thirty-Seven Years of Experience

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Abstract An aortopulmonary window (APW) is a communication between the ascending aorta and the pulmonary trunk in the presence of two separate semilunar valves. In order to increase our understanding about the surgical management of this rare lesion and its long-term results, we describe our experience over a 37-year period. Between 1968 and 2005, 18 patients were diagnosed with APW. Seventeen underwent surgical correction. Age at operation ranged from 22 days to 22 years (median, 0.20 years). Follow-up ranged from 2 weeks to 28.6 years (median, 11.0 years). Surgical closure was achieved using a single patch in 7 patients (41.2%) double patch in 4 (23.5%), primary closure in 3 (17.6%), clip in 2 (11.8%), and ligation in 1 (5.9%). Complex APW was present in 8 patients (44.4%). One patient was treated nonsurgically. There were no early or late deaths after surgery. Both primary closure and patch closure gave excellent long-term results. Sporadic postoperative complications were only associated with complex lesions. One patient who was treated conservatively died (of pulmonary hypertension) 21 years after diagnosis. Repair of APW is ideally performed in the first months of life, before irreversible PHT has developed. Various surgical repair techniques in this series of patients gave excellent short-term and long-term results, without significant hemodynamic sequelae.

Keywords Aortopulmonary window · Aortopulmonary fenestration · Congenital heart defect · Pediatric cardiac surgery

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Introduction

An aortopulmonary window (APW) is a congenital cardiac anomaly characterized by an abnormal communication between the main pulmonary artery (PA) and the ascending aorta (Fig. 1). The presence of two distinct semilunar valves distinguishes this anomaly from persistent truncus arteriosus. Various other terms have been applied to this anomaly, including aortopulmonary fistula, aortic septal defect, aorticopulmonary septal defect, and aortopulmonary fenestration. An APW is usually a single large defect, located just above the semilunar valves [23]. The defect may occasionally be found more distally overlying the origin of the right pulmonary artery. This variability in morphology is reflected in several classification schemes [12, 18, 19], reviewed by Jacobs et al. [13]. The current standard nomenclature for APW involves the terms proximal, distal, total, and intermediate defects [13].

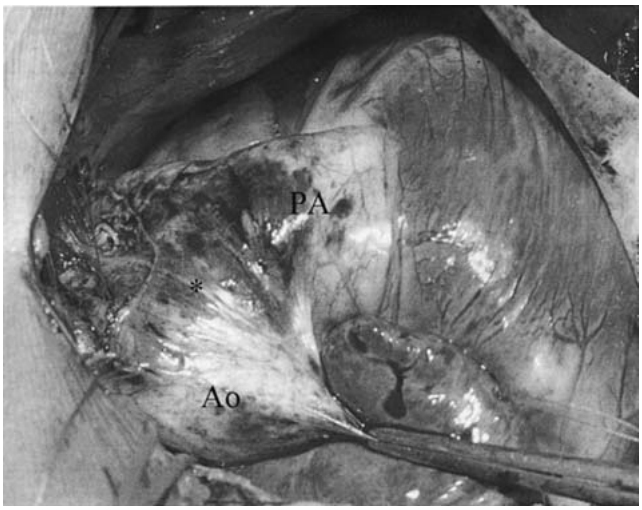

Fig. 1 Typical view of an Aortopulmonary window (*) between the ascending aorta Ao and the pulmonary trunk as seen by the surgeon. PA; pulmonary artery

APW can be found as an isolated defect, but in 47–77% of cases it is associated with other cardiac anomalies, most commonly interrupted aortic arch or coarctation of the aorta, ventricular septal defect, tetralogy of Fallot, and anomalous origin of the coronary artery. Patent ductus arteriosus occurs less frequently [2, 5, 14]. A *simple* APW is defined as having no associated cardiac defects, or simple defects requiring no or only minor intervention, such as patency of the ductus arteriosus, right aortic arch, or atrial septal defect. A *complex* APW, on the other hand, has one or more associated complex cardiac malformations or anomalies requiring more complex repair, such as interrupted aortic arch or ventricular septal defect.

Since APW is a rare lesion, occurring in 0.1–0.2% of patients with congenital heart disease, surgical series rarely include more than 20 patients [2]. Consequently, long-term postoperative studies are rare. Here, we review our experience over a 37-year time period.

Patients and Methods

Methods

Between 1968 and 2005, 18 patients were diagnosed with APW at the Center for Congenital Heart Disease in Amsterdam and Leiden, The Netherlands (Table 1). Data were obtained from review of their charts and communication with the patients' physicians.

The morphology of the defect was classified according to the most recent nomenclature, reported by Jacobs et al. [13]. Type I is a proximal APW located just above the sinus of Valsalva, a few millimeters above the semilunar valve. Type II is a distal APW located in the uppermost position of the ascending aorta. Type III is a total defect involving the majority of the ascending aorta. An intermediate defect is comparable to a type III defect but has adequate superior and inferior rims.

Patients

Seventeen patients underwent surgical correction of APW and associated anomalies. One patient with severe mental retardation was treated conservatively for unknown reasons. Demographic and diagnostic characteristics are summarized in Table 1. The male-to-female ratio was 1.6:1. Signs of congestive heart failure were noted in 16 patients (88.9%), frequent airway infections in 5 (27.8%), and cyanosis in 3 (16.7%). Age at operation ranged from 8 days to 22 years (medial, 2.4 months); 14 patients (82%) were younger than one year of age. Weight at operation ranged from 1.9 to 48.6 kg (median, 4.0). Type I APW was present in 8 patients (44.4%), whereas type II was found in 6 patients. Type III and intermediate defects were both found in 1 patient each. In 2 patients, the morphology of the defect could not be specified from the charts.

Associated Cardiac Anomalies

Simple APW was present in 10 patients (55.6%). An isolated defect was identified in 5 of them, and the other 5 patients presented with minor associated lesions, including right aortic arch ($n = 1$), patent foramen ovale ($n = 4$), and persistent ductus arteriosus ($n = 3$). Eight patients (44.4%) had complex APW, including 3 with interrupted ($n = 2$) or hypoplastic aortic arch ($n = 1$), 1 with discrete subvalvar and valvar aortic stenosis, and 1 with a malalignment ventricular septal defect. A perimembranous ventricular septal defect was identified in 5 other patients. The interrupted aortic arches were both type A, located distal to the left subclavian artery. One of these patients presented with an associated aortic origin of the right pulmonary artery, patent ductus arteriosus, atrial septal defect, and ventricular septal defect (thus resembling the anatomy of Berry syndrome, in which the ventricular sepum is intact [3]). The other patient with interrupted aortic arch also had a persistent left vena cava superior. Associated noncardiac anomalies were found in 3 patients, including Klinefelter syndrome in 1 and VACTERL association in 2 patients.

Table 1 Clinical characteristics, surgical techniques, and outcomes of the patients with aortopulmonary window

No. Patient	Year of surgery or diagnosis	Age at operation (years)	APW location	Diagnosis	Associated lesions	Surgical technique	Follow-up (years)	Comments
1	1968	—	Unknown	CATH	PDA	Conservatively	19.7	Irreversible PHT
2	1976	21.9	Total	CATH	Isolated	Single patch	28.6	Severe PHT, rest-APW
3	1980	1.21	Unknown	CATH	Isolated	Single patch	24.1	
4	1982	0.44	Proximal	CATH	RAA, PFO	Primary	22.2	
5	1987	0.37	Proximal	CATH	VSD, overriding aorta	Single patch	18.0	
6	1990	0.26	Proximal	ECHO	PFO	Primary	14.9	Klinefelter syndrome
7	1990	0.03	Proximal	Surgery	Hypoplastic aortic arch, VSD	Primary	14.6	Phrenic nerve paralysis
8	1991	0.07	Distal	ECHO	ASD, VSD, RAA	Ligation	13.1	PA stenosis
9	1992	0.31	Proximal	ECHO + CATH	Isolated	Double patch	12.2	
10	1995	0.1	Distal	ECHO + CATH	ASD, VSD	Single patch	9.8	VACTERL, cardiac tamponade
11	1998	0.07	Distal	ECHO + CATH	IAA-A, VSD, ASD	Double patch	6.3	Rhythm disturbance
12	1998	2.5	Proximal	ECHO + CATH	Isolated	Single patch	Lost F/U	Sternal wound Infection
13	1998	7.82	Proximal	Surgery	AoSv, AoSs	Single patch	6.9	
14	2001	0.02	Intermediate	ECHO	IAA-A	Clip	3.1	
15	2002	0.06	Distal	ECHO	PFO	Double patch	3.2	Identical twin
16	2003	0.16	Proximal	ECHO	PFO, PDA	Clip	1.7	
17	2004	0.05	Distal	ECHO	VSD, ASD, PDA	Single patch	0.12	VACTERL
18	2005	0.20	Proximal	ECHO	Isolated	Double patch	0.4	

CATH, cardiac catheterization; ECHO, echocardiography; PDA, patent ductus arteriosus; RAA, right aortic arch; PFO, patent foramen ovale; VSD, ventricular septal defect; ASE, atrial septal defect; IAA-A, type A interrupted aortic arch; AoSv, valvar aortic stenosis; AoSs, subvalvar aortic stenosis; PHT, pulmonary hypertension; APW, aortopulmonary window; PA, pulmonary artery; VACTERL, vertebral, anorectal, cardiac, tracheoesophageal, renal, radiai, and limb abnormalites; F/U, follow-up

Preoperative Studies

Preoperative cardiac catheterization was performed in 8 patients. The first three patients were operated on the basis of catheterization data alone since echocardiography was not yet available. Conversely, the last five patients were operated on solely on the basis of echocardiographic evaluation. Interestingly, echocardiography failed to correctly diagnose the defect in two cases. Cardiac catheterization was mainly carried out if uncertainty about the diagnosis existed or if major associated lesions required further hemodynamic assessment.

Genetic Research

Genetic research was performed in 5 patients. In one patient with VACTERL association, an inversion of chromosome 9 was found. Three patients, 2 of whom had an interrupted aortic arch type A, were investigated for 22q11 deletion, which could not be demonstrated. However, one of them was diagnosed with Klinefelter syndrome. One patient presented with elevated serum calcium levels, but genetic analysis of chromosome 7 revealed no abnormalities so that a diagnosis of Williams syndrome could not be made.

Surgical Techniques

The surgical techniques employed varied considerably over time, reflecting the advances in pediatric cardiac surgery. They included ligation without the use of cardiopulmonary bypass (CPB) in one patient (7%), closure by means of a clip under CPB in two patients (11.8%), primary closure under CPB in three patients (17.6%), single patch closure under CPB in seven patients (41.2%), and double patch closure under CPB in four patients (23.5%). The defect was approached either through an aortotomy (seven patients) or by dissecting both great arteries at the window site (seven patients). No pulmonary arteriotomy was used. Associated cardiac anomalies were always corrected in the same setting.

Early and Long-Term Results

Follow-up ranged from 5 months to 28.6 years (median 11.0 years). There were no early deaths. In one mentally retarded patient, APW was diagnosed at an age of 3 weeks. However, for unknown reasons the decision was made against surgical treatment and he died of Eisenmenger syndrome and consequent heart failure 21 years after the diagnosis.

One patient was operated on at the age of 21.9 years. Since severe pulmonary hypertension (mean pulmonary artery pressure 70 mmHg) had already developed by that time, initial patch closure of the defect was not tolerated. A new opening, 10 mm in diameter, was created in the patch in the same surgical setting to relieve pulmonary hypertension. Two patients required early reintervention, one due to cardiac tamponade and the other as a cause of diaphragmatic paralysis.

The 17 patients who successfully underwent surgery are all alive, and all but 1 are currently in New York Heart Association (NYHA) functional class I. One patient with a partially closed APW and preoperative pulmonary hypertension was in NYHA class II. Closure by ligation (1 patient) was associated with development of a transient mild pulmonary artery stenosis due to torsion of the pulmonary arteries, which resolved spontaneously during follow-up. Turbulent right and left pulmonary artery flow with pressure gradients of less then 20 mmHg was noted in 6 patients. Both primary and patch closure gave excellent long-term results.

Discussion

An aortopulmonary window was first described in an autopsy study by Elliotson in 1830 [9]. Since then, several classification schemes have been developed to describe the variability in size, location, and morphology of the defect [12, 13, 18, 19]. The current standard nomenclature involves the terms proximal, distal, total, and intermediate defects. Aortic origin of the right pulmonary artery, or hemitruncus, is described as a separate entity [1, 13]. Most APWs are located a few millimeters above the semilunar valves, thus comprising the proximal defects [13, 18]. This was confirmed in our study, in which approximately 45% of patients showed a similar lesion. In this study, complex defects, defined as APW associated with major cardiac lesions, were also seen in approximately 45% of patients.

The differential diagnosis includes patent ductus arteriosus with pulmonary hypertension. Although echocardiography has made a significant contribution to the early recognition of APW and is the diagnostic tool of choice, it may fail to correctly diagnose the defect [8, 16, 22]. In two of our patients, the true diagnosis was not disclosed until the operation. In both cases, this was related to unalertness of the examiner to this defect and its associated cardiac anomalies. Cardiac catheterization is currently only recommended to evaluate the degree of pulmonary vascular disease in patients older than 6 months of age [10, 16].

The abnormal communication between the ascending aorta and the main pulmonary artery causes a significant left-to-right shunting. Symptoms of congestive heart failure appear during early infancy; occasionally, there is minimal cyanosis. If left untreated, 40% of patients will eventually succumb to the sequelae of pulmonary hypertension and Eisenmenger syndrome [21]. Since irreversible pulmonary disease may occur at an early age, surgical closure of the defect is ideally performed in the first months of life [16, 21, 22] The retrospective design of this study resulted in a relatively high median age at surgery of 2.4 months. Currently, surgery is often performed in the neonatal period; three of the last five patients in this series were operated on in the first 3 weeks of life. Concerning the surgical technique, most centers recommend transaortic closure of APW, either by direct suture or by patch closure [16, 22]. Small, hemodynamically unimportant defects may be closed by direct suture; larger defects should be closed with a pericardial or synthetic patch. Both methods of repair showed excellent short-term and long-term results in this series of patients, without significant hemodynamic sequelae. The patch methods in general have a much more favorable outcome with less distortion of the aorta and pulmonary artery compared to primary closure or ligation. The transaortic route is preferred to the transpulmonary method because it offers better visualization of the coronary ostia and the aortic valve leaflets, and at the same time, it allows right pulmonary artery reconstruction if necessary [1, 6, 11, 16, l7, 22]. Furthermore, in one study, repair by way of a pulmonary arteriotomy was associated with a significantly higher probability of reintervention [11]. Today mortality is predominantly related to major associated cardiac lesions, particularly interruption of the aortic arch, and mainly occurs before surgical repair [2, 11].

The pathogenesis of APW has not been resolved completely. Abnormalities of the cardiac neural crest axis have been implicated in many conotruncal anomalies, based on the association of such lesions with 22q11 deletions [7, 15, 20]. However, in neural crest ablation studies in mice, no APWs were reported

[4]. Furthermore, there appears to be no association between 22q11 deletion syndrome and the occurrence of aortopulmonary septal defects [14, 20]. This syndrome often coexists with interrupted aortic arch type B (IAA-B), whereas APW is predominantly associated with IAA-A. Therefore, cardiac neural crest does not seem to play a major role in the pathogenesis of APW [14]. An interesting finding in our study was the association of APW with the VACTERL association in two patients. This association has been described in earlier reports, but our knowledge about the pathogenetic relation between these two conditions is incomplete [2, 4]. APW is only rarely associated with genetic lesions, as exemplified by one of our patients who was born as one of identical twins, whereas her sister had no congenital heart defects.

In conclusion, APW shows a varied morphology, with proximal defects being the most frequent. Various surgical repair techniques in this series of patients gave excellent short-term and long-term results, without significant hemodynamic sequela.

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