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Original Article

Diagnosis, management, and results of treatment for aortopulmonary window

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Abstract The aortopulmonary window is a communication between the ascending aorta and the pulmonary trunk in the presence of two separate arterial valves. This uncommon congenital anomaly is reported rarely in the literature. We present here our experience with 16 patients, emphasizing the importance of early closure of the defect by a transaortic approach.

We performed surgery on 16 patients over a period of 13 years using a transaortic approach under cardiopulmonary bypass. The median age of the patients at the time of operation was 6.5 months, with a range from 1 month to 11 years. Preoperative pulmonary arterial systolic pressure ranged from 30 to 100 mmHg. Associated cardiac anomalies were present in 7 of the patients, and were repaired at the same stage. The defect was between the ascending aorta and the proximal pulmonary trunk in 13 patients, and between the ascending aorta and the distal pulmonary trunk, with overriding of the orifice of the right pulmonary artery, in 3 patients. For closure, we used a patch of 0.4 mm Gore-Tex in 11, and gluteraldehyde-treated autologous pericardium in 5 of the patients.

One patient died during surgery. The mean follow-up period for the surviving 15 patients was 52.2 months, with a range from 12 to 130 months. All the patients were in good condition during the follow-up, and no residual defects have been detected.

Aortopulmonary window is a rare congenital cardiac anomaly, which can be repaired with very good operative results if surgery is performed before the development of irreversible pulmonary hypertension. We advise early correction of the defect with a transaortic patch, repairing all associated cardiac anomalies at the time of diagnosis.

Keywords: Aortopulmonary fenestration; pulmonary hypertension; transthoracic echocardiography

THE AORTOPULMONARY WINDOW, CHARACTERIZED by a communication between the ascending aorta and the pulmonary trunk, is a rare congenital cardiac anomaly. It represents 0.15–0.6% of all congenital cardiac malformations,[1,2] and is associated with other cardiac anomalies in one-quarter to half of cases.[1,3] Patients with the isolated form usually present with symptoms due to increased flow of blood to the lungs as the main clinical feature, but if associated with other cardiac anomalies, the clinical findings change according to the additional defect. Surgical closure is the accepted means of correction. Good operative results depend on the timing of operation, preferably undertaken in early infancy, and on the severity of the associated anomalies of the heart and great vessels. Delay of surgical correction leads to pulmonary hypertension.[4] To date, experience with only about 300 cases has been published, appearing for the most part as description of isolated case reports.[5,6] There are very few described surgical series based on more than 10 patients. We report

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Table 1. Preoperative characteristics and follow-up details of the patients.

p	Year of operation	Age and weight at operation	APW location	Diagnosis	PA pressure at operation*	Oxygen study	Patch material	Concomittant anomalies/ concomittant procedure	Follow-up interval	Comment
1	1987	3/13 kg	Distal	ECHO + CATH	75	Yes	Gore-Tex	–	130 months	NYHA class-1, no medication
2	1988	6/17 kg	Distal	ECHO + CATH	70	Yes	Gore-Tex	–	108 months	NYHA class-1, on digital
3	1991	7 months/5.8 kg	Intermediate	ECHO + CATH	88		Gore-Tex	–	110 months	NYHA class-1, no medication
4	1995	9 months/5.7 kg	Intermediate	ECHO	60		Gore-Tex	PFO-MR/Primary PFO Repair + mitral plasty	84 months	Mild MR, NYHA class-1, on digital
5	1997	21 months/10.5 kg	Intermediate	ECHO + CATH	56	Yes	Gore-Tex	Subaortic ridge/ Subaortic ridge resection	36 months	Mild AR, NYHA class-1, no medication
6	1997	6 months/5.2 kg	Intermediate	ECHO + CATH	65		Gore-Tex	–	49 months	NYHA class-1, no medication
7	1997	4 months/5.5 kg	Intermediate	ECHO	70		Gore-Tex	–	51 months	NYHA class-1, no medication
8	1998	3 months/4.8 kg	Intermediate	ECHO	55		Pericard	–	45 months	NYHA class-1, no medication
9	1998	4 months/5.2 kg	Intermediate	ECHO	40		Gore-Tex	IAA-PDA/repair of IAA	41 months	NYHA class-1, no medication
10	1999	1 month/3.7 kg	Intermediate	ECHO	48		Pericard	ASD/primary ASD closure	26 months	NYHA class-1, no medication
11	1999	15 months/11 kg	Intermediate	ECHO + CATH	30	Yes	Pericard	PAD/PAD ligation	21 months	NYHA class-1, no medication
12	2000	7 months/4.1 kg	Intermediate	ECHO	60		Gore-Tex	–	28 months	NYHA class-1, no medication
13	2000	11/33 kg	Intermediate	ECHO + CATH	100	Yes	Gore-Tex	–	29 months	NYHA class-2, on digital, diuretic and ACE-inhibitor
14	2001	1.5 months/4 kg	Distal	ECHO + CATH	60		Gore-Tex	–	13 months	NYHA class-1, no medication
15	2001	2.5 months/4.4 kg	Intermediate	ECHO	58		Pericard	ASD/ASD closure with pericardial patch	12 months	NYHA class-1, no medication
16	2002	1 month/2.2 kg	Intermediate	ECHO	80		Pericard	TOF/TOF total correction	Dead postoperative 1st day	Dead due to a possible pulmonary hypertensive crisis

*Pulmonary arterial systolic pressure (mmHg). Abbreviations: CATH: cardiac catheterization; PFO: patent foramen ovale; MR: mitral regurgitation; AR: aortic regurgitation; IAA: interrupted aortic arch; PAD: patent arterial duct; TOF: tetralogy of Fallot

here our experience with 16 patients undergoing surgery over a period of 13 years.

Patients and methods

Between April 1987, and May 2000, we undertook surgical repair in 16 patients presenting with aortopulmonary window. The preoperative characteristics of the patients are summarized in Table 1. The median age of the patients at the time of operation was 6.5 months, with a range from 1 month to 11 years. The median weight was 5.35 kg, with a range from 2.2 to 33 kg. The diagnosis was made by transthoracic echocardiography in 8 patients, with cardiac catheterization as an added diagnostic procedure in 8 patients. The location of the defect was between the ascending aorta and the proximal pulmonary trunk in 13 patients, and between the ascending aorta and the distal pulmonary trunk, with overriding of the orifice of the right pulmonary artery, in 3 patients (Fig. 1). In all defects between the ascending aorta and the proximal pulmonary trunk, there was a considerable rim between the inferior edge of the defect and the arterial valves. In 9 of the patients, the window was an isolated defect, but additional congenital anomalies were diagnosed respectively in 7 of the patients. These were a patent oval foramen and mitral regurgitation, a subaortic ridge, interruption of the aortic arch with a patent arterial duct, tetrology of Fallot, patency of the arterial duct, and an atrial septal defect in 2 patients. The median systolic pulmonary arterial pressure at the time of diagnosis was 60 mmHg, within a range from 30 to 100 mmHg. All the patients underwent surgery via a transaortic approach. The follow-up of the patients was done by clinical examination and echocardiographic studies. The mean period of follow-up after the procedure is 52.22 months, with a range from 12 to 130 months.

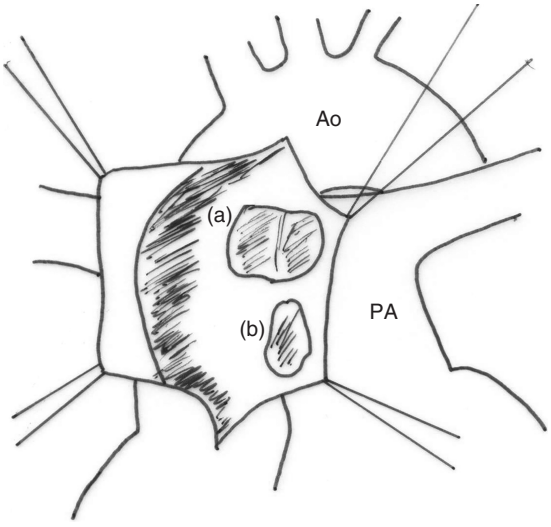

Figure 1.
Schematic representation of the two different types of aortopulmonary window defects encountered in our 16 patients as viewed from the interior of the aorta. The distal defect (a), between the ascending aorta and the distal pulmonary trunk, has overriding the origin of the right pulmonary artery, while the intermediate defect (b) provides a communication between the ascending aorta and the proximal pulmonary trunk. Ao: aorta; PA: pulmonary trunk.

Preoperative evaluation

All the patients under one-year-old were referred for operation following the diagnosis of aortopulmonary window, regardless of the degree of pulmonary hypertension. Patients presenting after one year of life underwent oxygen studies in the catheterization laboratory. Pulmonary vascular resistance below 6 units/m^2, or a fall in the pulmonary vascular resistance to this level following inhalation of 100% oxygen, were accepted as suitable criterions for operation.

Surgical technique

All patients underwent repair via a transaortic approach and under cardiopulmonary bypass. The left and right pulmonary arteries were controlled with snares at the initiation of cardiopulmonary bypass, and the patient was cooled to 28°. The aorta was cross-clamped, and the heart was arrested using blood cardioplegia, the cardioplegia being repeated every 20 min. The aorta was opened by an anterior vertical incision. After observing the orifices of the coronary arteries and the aortic valve, the aortopulmonary window was located and closed with a patch of 0.4 mm thick Gore-Tex in 11 patients, and with gluteraldehyde-treated autologous pericardial patch in the other 5 patients, using a continuous polypropylene suture. Following the closure of the aortotomy, and de-airing of the heart, the patient was weaned from cardiopulmonary bypass. The concomittant cardiac anomalies were repaired at the same stage. The details of the operative procedures are listed in Table 1.

Postoperative care

All the patients were kept fully sedated, using a combination of a muscle relaxant and fentanyl, and hyperventilated during the night following the operative procedure to prevent any pulmonary hypertensive crisis. The weaning from the ventilator was achieved in the first postoperative day in all patients, with close monitoring of the cardiac and pulmonary states. Diuretics and digoxin were given to patients who had presented with symptoms of congestive cardiac failure before the operation. These drugs were reevaluated in

the postoperative period, and stopped subsequent to regression of the symptoms of heart failure.

Results

Mortality and morbidity

We lost 1 patient during the perioperative period. This patient, severely stressed before the operation, underwent closure of the aortopulmonary window and concomitant repair of tetralogy of Fallot. After an uneventful postoperative night, the patient suddenly deteriorated, and died during the first postoperative day due to a possible pulmonary hypertensive crisis. Another patient was taken back to the operating room in the evening of the operation due to excessive drainage from the chest tubes. In the operating room, leakage from the aortotomy was repaired with 2 simple sutures, and the patient subsequently did well. All the remaining patients had an uneventful postoperative course, and were discharged from the hospital in good condition.

Late follow-up

All the patients underwent regular follow-up by echocardiographic examination and clinical investigation. The follow-up is complete, with a mean period of 52.2 months, and a range from 12 to 130 months. All patients were alive at the time of follow-up. All but one was in excellent functional capacity, and undertaking normal daily activities. Of these patients, 11 were not using any medication, while 3 of them remained on digoxin, which had not been discontinued because of persisting cardiomegaly. The patient who underwent operation at the age of 11 was observed to have some limitations in his daily activities, and was receiving digoxin, diuretics, and inhibition of the angiotensin converting enzyme. Despite this, the patient was following his studies normally. We believe that this patient has established pulmonary vasculature disease because of the delayed timing of the operation. Residual defects were not observed in any of the patients.

Discussion

Aortopulmonary window, first described as an autopsy finding by Elliotson in 1830,[7] is a communication between the ascending aorta and the pulmonary trunk in the presence of separate arterial valves, this finding distinguishing the entity from common arterial trunk.[1] There are several different schemes proposed for classification of the defects. Richardson et al.[8] distinguished three types of defects, proximal defects, distal defects including the origin of the right pulmonary artery, and cases in which the right pulmonary artery arose exclusively from the aorta. Ho et al.,[9] having studied 25 autopsied specimens, argued in favour of a more descriptive approach, excluding cases of anomalous origin of the right pulmonary artery from the aorta as a subset of aortopulmonary window. Thus, based on their experience, Ho et al. described 4 types of window. These were described as proximal, intermediate, distal, and confluent defects. Proximal defects had little or no inferior rim separating the window from the arterial valves at the level of the sinutubular junctions. Generous arterial walls surrounded intermediate defects. Distal defects had a well-formed inferior rim distancing them from the arterial valves, but had little or no superior rim. Confluent defects had practically no inferior or superior rims. It is this classification suggested by Ho et al.[9] that we have used to categorize our patients. Embryologically, aortopulmonary window represents incomplete separation of the intrapericardial arterial trunks. These defects result from abnormal septation of the distal outflow tract during the embryonic period, coupled in some cases with abnormal fusion with the aortopulmonary septum, the wedge of tissue that separates the fourth and sixth arches in the posterior wall of the aortic sac.[10]

Patients with aortopulmonary window usually present with early congestive heart failure due to the left-to-right shunting. A continuous murmur, thought to be characteristic of the disease, may not be heard in all patients.[1] An isolated window can be diagnosed with precision by transthoracic echocardiography (Fig. 2). Catheterization can be helpful if the echocardiographic diagnosis remains uncertain, or if the window is accompanied by other cardiac

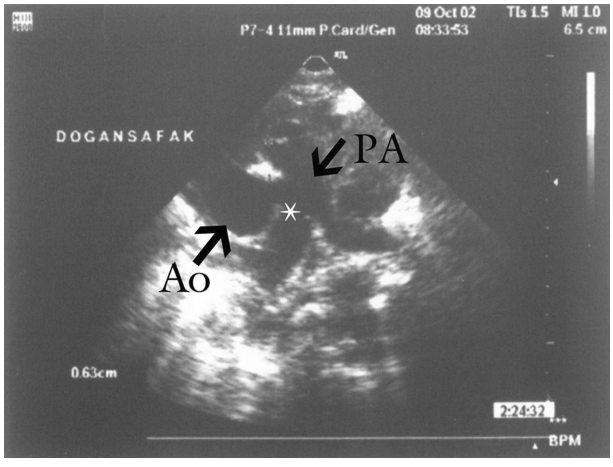

Figure 2.
Typical view of an Aortopulmonary window (*) between the ascending aorta and the pulmonary trunk as demonstrated by transthoracic echocardiography. Ao: aorta; PA: pulmonary trunk.

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defects, as well as in patients with suspected severe pulmonary hypertension, where the invasive study is needed determine the reversibility of the pulmonary hypertension. As already discussed, the window is often associated with other cardiac defects. The most frequent are interruption of the aortic arch, patency of the arterial duct, coronary arterial anomalies, and tetralogy of Fallot.[11] In these cases, the clinical picture varies according to the associated defect. Some patients as old as 58 at the time of their surgery have been described,[12] but nowadays patients should undergo surgery early in the infancy prior to the development of irreversible pulmonary hypertension.[4]

Surgical repair usually consists of closure using a patch. Since the first correction, made by Gross in 1952 through a left thoracotomy,[13] numerous techniques have been described using either thoracotomy or sternotomy incisions, with or without cardiopulmonary bypass, and by transpulmonary or transaortic approaches. Of these techniques, it is now recognized that simple ligation of the window is fated to cause unacceptable pulmonary arterial distortion,[14] while the transpulmonary approach is reported to result in a higher need for reintervention.[15] Thus, those working in experienced centres now advocate transaortic repair.[15] Although closure through the pulmonary trunk using an autologous pulmonary arterial patch has been described with good follow-up results in recent years,[16,17] we believe that this approach may complicate the operative process when compared with the transaortic route. Transcatheter closure is also reported for small defects, or for closure of any residual defects after the operation.[18]

We have always preferred the transaortic approach, and we have never tried the other approaches. We believe that this approach permits safer closure, with better exposure of the defect. Our experience endorses recent reports,[15] and we believe that the fact that we have not detected any residual defects during the period of follow-up justifies our approach. For the patch, we use either gluteraldehyde-treated autologous pericardium, or Gore-Tex of 0.4 mm thickness. We have not detected any difference between these two materials over our period of long-term follow-up, but we think that Gore-Tex patch may be more suitable, as it is a more solid material than the pericardium to work under systemic pressures, therefore preventing any possible late aneurysmal transformation. Simple transthoracic echocardiography is now usually sufficient for decisions regarding operability, and the patient should now undergo surgery during the same period of hospitalization. Half of our patients underwent operative repair subsequent to echocardiographic diagnosis. The majority of the patients undergoing catheterization were seen at the beginning of our experience, and the intervention was undertaken with the aim of confirming the echocardiographic diagnosis, or to study the pulmonary vascular bed in patients older than 1 year of age. Of our 16 patients, 10 underwent surgery during the first year of life, with the remaining patients only having a surgical procedure after careful study by the pediatric cardiologist to confirm the reversibility of pulmonary hypertension. We think that the co-existence of other cardiac defects changes the surgical risk depending on the associated defect when compared to isolated cases, but the co-existence of aortopulmonary window does not change the surgical outcome of the associated anomaly, and should be repaired at the same stage.

Thus, we conclude that closure of an aortopulmonary window is a safe surgical procedure, which can be carried out with very low morbidity and low rates of mortality. Early closure by the transaortic approach offers complete correction to the child, with a subsequently normal life expectancy, and should be performed at the time of diagnosis, preferentially early in the infancy. Patients with defects diagnosed at older ages should be studied carefully before the repair to assess the level of pulmonary hypertension, and surgical closure should be attempted only in appropriate cases. The association of other cardiac anomalies changes the clinical picture and the surgical risk, but correction at a single stage should be offered to these patients when seen in experienced centres.

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