Distal type of aortopulmonary window

Report of 4 cases

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SUMMARY We have studied 14 patients with aortopulmonary window (10 male and 4 female, age range 1 month to 41 years). Four of these had a distal defect with characteristic haemodynamic and angiographic features. Aortopulmonary window may be classified into 3 types: type I (proximal) defects occur in the proximal part of aortopulmonary septum; type II (distal) defects occur in the distal part of the aortopulmonary septum adjacent to the right pulmonary artery; the type III defect is a combination of types I and II.

In type I, injection of contrast media into the aortic root opacifies the main pulmonary trunk and then both pulmonary arteries. In type II, the right pulmonary artery is preferentially opacified simulating the finding of right pulmonary artery arising from the ascending aorta. In one case of type II, injection into the right ventricle showed preferential flow to the left pulmonary artery, because of the large shunt of unopacified blood into the right pulmonary artery, but in both types I and II the left and right pulmonary arteries are usually opacified simultaneously after injection into the main pulmonary trunk.

In type I either transaortic or transpulmonary closure is the appropriate surgical procedure. In types II and III, the transaortic approach provides better exposure and facilitates the operative repair.

Aortopulmonary window is a relatively rare congenital anomaly involving the great arteries. Patients with this anomaly in whom there is an increased pulmonary flow and pulmonary hypertension usually develop symptoms in the neonatal period or in early infancy, and without operation have a uniformly poor prognosis. The high mortality rate in these babies is the result of congestive heart failure and of pulmonary complications caused by an excessive left-to-right shunt through the aortopulmonary window. Since cure is effected by surgical closure of the defect, precise preoperative recognition of the type and the location of the window is essential.

In the majority of cases, the defect is in the left lateral wall of the ascending aorta and communicates with the right lateral wall of the pulmonary trunk; the window is then anterior to the ostium of the right pulmonary artery when viewed from the lumen of the pulmonary trunk. In another type, however, there is a distal communication between the ascending aorta and the junctional area of the

right pulmonary artery and the pulmonary trunk (Kimoto *et al.*, 1957; Nakaya, 1963; Wright *et al.*, 1968).

In a 19-year period from 1958 to 1976, 14 patients with aortopulmonary window were studied at the Heart Institute of Japan (Table 1), and 4 of the 14 had the distal type of communication. The purpose of this report is to describe 4 cases of the distal type of aortopulmonary window and to discuss diagnostic and surgical problems in 3 varieties of this defect.

Case reports

Clinical data are summarised in Table 2.

CASE 1

J.A., a 6-month old girl, was first seen at the age of 30 days after an episode of unconsciousness, with opisthotonus. She was then an underdeveloped, poorly nourished infant, and had failed to thrive. There was no cyanosis, but she had been tachypnoeic and dyspnoeic from birth. The clinical diagnosis was ventricular septal defect and per-

Table 1 Cases of aortopulmonary window seen at the Heart Institute of Japan, Tokyo Women's Medical College, from 1958 to 1967

Anatomical classification of aortopulmonary window	Case No.	Age		Medical death	Alive—unoperated	Operation			
		Male cases	Female cases			Ligation	Division	Transpulmonary closure	Transaortic closure
Type I: proximal defect	10	4 m 13 y 7 m 13 y 5 y 5 y 5 y 6 y	21 y 41 y	1	3	1	2	1	2
Type II: distal defect	4	1 y 5 m 1 y 7 m	30 d 50 d	1					3
Type III: total defect	0								
Total	14	10	4	2	3	1	2	1	5

Table 2 Clinical findings in distal type of aortopulmonary window

Case No.	Age	Sex	Presenting features	Signs	Chest x-ray		ECG QRS axis	Course
					CTR	Pulmonary vascularity		
1	30 d	Female	Underdeveloped, poorly nourished, tachypnoea, failure to thrive	Bounding pulses, pansystolic murmur along left sternal border, diastolic murmur at apex, and accentuation of 2nd sound	60%	Increased	+170° RVH	Died at age of 6 months; pneumonia and CHF
2	17 m	Male	Tachypnoea, failure to thrive; at 8 m, bronchopneumonia and CHF	CHF with oedema, hepatomegaly, and ascites; ejection murmur along left sternal border, systolic and diastolic murmurs at apex, and accentuation of 2nd sound	64%	Pulmonary venous congestion +	+95° LAH LVH ST-T changes (II, III, aVF V5-7)	Well after surgical repair of aortopulmonary window and later mitral valve replacement
3	19 m	Male	Recurrent respiratory infection; tachypnoea slow weight gain	Ejection murmur along left sternal border, diastolic murmur at apex, and accentuation of 2nd sound	56%	Increased	+95° CVH	Well after surgical repair
4	50 d	Female	Tachypnoea, failure to thrive; hepatomegaly, sweating	Bounding pulses, ejection murmur along left sternal border, and accentuation of 2nd sound	64%	Increased	+35° CVH	Died at home 11 months after successful surgical repair: acute respiratory infection

sistent ductus arteriosus. Administration of digitalis and diuretics resulted in a temporary clinical improvement. She was discharged after 1 month in hospital but was readmitted 4 months later with pneumonia and congestive failure and died at the age of 6 months.

At necropsy, the external appearance of the heart indicated biventricular hypertrophy and dilatation and the apex was formed by the right ventricle. The ascending aorta and the pulmonary trunk appeared to have a normal relation (Fig. 1a). When the anterior wall of the pulmonary trunk was opened longitudinally, the window between the ascending aorta and the proximal portion of the right pulmonary artery was seen within the orifice of the

right pulmonary artery (Fig. 1b). From the aortic side, the window was seen 5 mm above the ostium of the left coronary artery on the left posterior aspect of the aortic wall (Fig. 1c).

CASE 2

E.M., a 17-month-old boy, was the second child of healthy parents and was born after an uneventful gestation and delivery. There was consanguinity on the paternal side. The infant had been tachypnoeic from birth and had failed to thrive. He was admitted to another hospital at the age of 8 months with cough, fever, and dyspnoea. The diagnosis then was bronchopneumonia and congenital heart disease. Since he failed to improve and had a per-

Fig. 1 Case 1. (a) The ascending aorta and the pulmonary trunk are normally related. (b) Upon opening the pulmonary trunk longitudinally on the anterior wall, the window between the ascending aorta and the pulmonary artery is seen through the orifice of the right pulmonary artery. (c) The communication is between the left posterior aspect of the ascending aorta and the origin of the right pulmonary artery from the pulmonary trunk.

sistent cough and congestive heart failure despite medical treatment, he was referred to the Heart Institute of Japan at the age of 17 months.

On examination, he was a fretful male infant weighing 6.5 kg. He was dyspnoeic but not cyanosed, and was in heart failure with oedema of the extremities and face, hepatomegaly, and ascites. The clinical diagnosis was persistent ductus arteriosus, mitral regurgitation, and endocardial fibroelastosis. His condition improved after treatment with digitalis.

Cardiac catheterisation and angiographic data are summarised in Table 3. The right ventricular and main pulmonary arterial pressures were at systemic level, but the catheter could not be passed into the aorta or the pulmonary arterial branches. A selective injection of contrast into the pulmonary trunk showed early opacification of the ascending aorta through the aortopulmonary window (Fig. 2a), and later films showed a huge left atrium which

emptied slowly. The aortogram clearly showed an aortopulmonary communication (Fig. 2b); though the right pulmonary artery was preferentially opacified, the previous pulmonary arteriogram had shown both pulmonary arteries. Aortopulmonary window of distal type was diagnosed.

At operation with the combined use of surface and perfusion hypothermia, a communication was found between the ascending aorta and the origin of the right pulmonary artery. Through a longitudinal aortotomy, the defect measuring 8 mm in diameter was closed with a 'teflon' patch. Since there was systolic expansion of the left atrium with 40 mmHg *v* wave, mitral annuloplasty was performed. The left atrial pressure after this procedure was 16 mmHg.

He made an uneventful recovery and remained well until 3 years after operation, but mitral regurgitation then reappeared with gradual deterioration of symptoms. At a second operation 3 years after the

Table 3 Cardiac catheterisation data

Age	Case 2		Case 3		2 y 8 m		After tolazoline		Postoperative (2 y 10 m)		Case 4	
	1 y 7 m		1 y 7 m								2 m	
Site	S/D (M) (mmHg)	O ₂ sat (%)	S/D (M) (mmHg)	O ₂ sat (%)	S/D (M) (mmHg)	O ₂ sat (%)	S/D (M) (mmHg)	O ₂ sat (%)	S/D (M) (mmHg)	O ₂ sat (%)	S/D (M) (mmHg)	O ₂ sat (%)
SVC			7/3 (4)	65.5	7/2 (4)	64.0	7/2 (4)	62.0	(5)	65.0	6/3 (4)	38.5
IVC	10/6 (7)	50.5	5/2 (3)	65.5	6/2 (3)	63.0	7/2 (4)	64.5	(5)	74.0	7/1 (4)	38.5
RA	11/5 (6)	45.0	6/2 (3)	66.5	6/2 (3)	69.0	7/1 (4)	52.5	(5)	70.0	6/1 (4)	37.0
RV	90/10 (45)	47.5	110/10 (60)	52.5	102/11 (51)	62.0	120/11 (59)	61.0	43/5	64.0	92.9 (41)	
PA trunk	93/56 (75)	52.0	115/75 (105)	56.5			122/71 (73)	70.5	44/13 (28)	68.0	87.9 (31)	37.0
Right PA					102/56 (80)	94.0	128/65 (88)	93.2	39/15 (25)	68.0	72/32 (47)	57.0
Left PA	93/56 (75)	50.0			98/59 (75)	69.0	120/72 (85)	70.5	44/13 (28)	64.0		
LV	95/5 (35)	94.0									92/48 (68)	81.5
Ascending ao	90/50 (80)	83.0	96/53 (77)	94.0							107/53 (75)	91.5
Descending ao							80/56 (67)	95.7

Fig. 2 Case 2. (a) Injection into pulmonary trunk shows early opacification of the aorta through the aortopulmonary window. (b) Aortic injection of dye shows selective opacification of the right pulmonary artery.

first, the mitral valve was replaced with a Hancock xenograft valve. He is now well.

CASE 3

D.N., a 19-month-old boy, was referred with recurrent respiratory infections and tachypnoea. At 2 months of age, he had been found to have a heart murmur. His growth and development were poor and he was cyanosed.

Right heart catheterisation (Table 3) then showed severe pulmonary hypertension (115/75 mmHg, mean 105 mmHg), but the catheter could not be passed into either branch of the pulmonary artery. A right ventriculogram opacified the pul-

monary trunk and left pulmonary artery clearly, but there was only faint opacification of the right pulmonary artery (Fig. 3a). Later films showed the left heart and the ascending aorta, but then the right pulmonary artery opacified more densely than the left pulmonary artery. Possible diagnoses were the distal type of aortopulmonary window, or persistent ductus arteriosus with anomalous origin of the right pulmonary artery from the ascending aorta. Operation was refused and the patient was followed up as an outpatient.

He was readmitted at the age of 2 years and 8 months for repeat catheterisation (Table 3). A catheter was passed from the right saphenous vein

Fig. 3 Case 3. (a) Right ventricular injection shows preferential flow to the left pulmonary artery. (b) Injection into the pulmonary trunk opacifies the left and right pulmonary arteries simultaneously. (c) On the aortogram, the right pulmonary artery opacifies preferentially, simulating origin of the right pulmonary artery from the ascending aorta.

to the right ventricle and both pulmonary arteries. When contrast was injected into the pulmonary trunk, both right and left pulmonary arteries opacified and there was also faint opacification of the ascending aorta (Fig. 3b). The aortogram showed preferential opacification of the right pulmonary artery, simulating the appearance of right pulmonary artery arising from ascending aorta (Fig. 3c). There was also a prominent increase in oxygen saturation in the right pulmonary artery. The calculated left-to-right shunt was 60 per cent with pulmonary to systemic flow ratio 2.5, and resistance ratio 0.40. After injection of tolazoline (1 mg/kg) into the pulmonary artery, there was little change in the magnitude of the shunt (62%), the flow ratio (2.6), or the resistance ratio (0.38).

The findings were compatible with the distal type of aortopulmonary window.

At operation using combined surface and core cooling, the diagnosis of aortopulmonary window was confirmed. The defect (20 × 25 mm) was closed with a patch by the transaortic route. Postoperative right heart catheterisation showed only slightly raised right ventricular and pulmonary arterial pressures (43/5 and 44/13 mmHg, respectively).

CASE 4

S.M., a 1-month-old baby girl, was born at full-term after an uncomplicated gestation. Her birth-weight was 3640 g, and no perinatal abnormality was observed. At 3 days of age, she was noted to be tachypnoeic, to cry weakly, and to have poor

appetite, and was admitted to hospital for tube feeding. At the age of 50 days, she was transferred to the Heart Institute of Japan for further evaluation.

Right heart catheterisation (Table 3) showed right ventricular and pulmonary arterial pressures equal to systemic, and a large left-to-right shunt into the pulmonary trunk. Since the catheter could be advanced into the descending aorta easily and repeatedly, the diagnosis of persistent ductus arteriosus was made.

At operation, the left chest was entered through the third interspace, but only a closed ligamentum arteriosum was found. When the pericardium was opened, the aortopulmonary window was found. Since the exposure was inadequate for its repair, only bilateral pulmonary arterial banding was performed. At reoperation one week later, using combined surface and perfusion hypothermia, the aortopulmonary window, which communicated with the most proximal portion of the right pulmonary artery, was closed with a patch by the transaortic route. The patient made an uneventful recovery and remained well until 11 months after operation, when she died suddenly as a result of an acute respiratory infection.

Discussion

Aortopulmonary window is a relatively rare congenital anomaly which presents with a serious clinical picture, particularly in early infancy. Corrective operation should be performed before the development of irreversible obstructive changes in the pulmonary vascular bed. It is clear that successful surgical correction of the defect depends upon pre-operative recognition of its exact anatomical location. The clinical features of aortopulmonary window with pulmonary hypertension resemble those of persistent ductus or ventricular septal defect with pulmonary hypertension, and differentiation is occasionally difficult.

Neufeld *et al.* (1962) observed a 25 per cent incidence of associated anomalies. These include persistent ductus arteriosus, ventricular septal defect, mitral regurgitation, aortic arch anomalies including right aortic arch, subaortic stenosis, tetralogy of Fallot, and coronary arterial anomalies (Cooley, 1957; Kimoto *et al.*, 1957; Taguchi *et al.*, 1959; Morrow *et al.*, 1962; Putnam and Gross, 1966; Coleman *et al.*, 1967; Hurwitz *et al.*, 1967; Deverall *et al.*, 1969; Agius *et al.*, 1970). In cases with these associated cardiac anomalies, diagnosis is often difficult. At cardiac catheterisation, the catheter course and angiographic appearances may be diagnostic (Bosher and McCue, 1962; Morrow *et al.*, 1962).

The size of defect is variable, ranging from 0.5 to 6 cm in diameter. The defect is usually single, but cases with two coexisting defects have also been reported (Baronofsky *et al.*, 1960). The defect is usually between the ascending aorta and the pulmonary trunk, and may take the form of a fenestration or a narrow channel. However, there are two other types of defect which differ in their anatomical site. We propose, therefore, that aortopulmonary window should be classified into three types: type I, proximal defect; type II, distal defect; and type III, total defect (Fig. 4). The 4 cases included in this report and 3 other cases reported by Kimoto *et al.* (1957), Nakaya *et al.* (1963), and Wright *et al.* (1968) are of type II. Anatomically, type II is a communication between the left posterolateral wall of the ascending aorta and the junctional portion of the right pulmonary artery and the pulmonary trunk. A total defect (type III) involves the entire length of the pulmonary trunk from immediately above both semilunar valves to the level of the pulmonary bifurcation and the proximal portion of the right pulmonary artery. In this type the aortic and pulmonary valves are separated haemodynamically and anatomically, a crucial distinguishing feature from persistent truncus arteriosus. In type III aortopulmonary window,

Fig. 4 Anatomical classification of aortopulmonary window.

the semilunar valves are completely separated by the conus septum, which is reduced or absent in persistent truncus arteriosus (Van Praagh and Van Praagh, 1965). No classification of aortopulmonary window has been reported hitherto except by Collett and Edwards (1949), who described it as a type of persistent truncus arteriosus.

Separation of the truncus arteriosus into the ascending aorta and the pulmonary trunk in the fetus is achieved by formation of the truncal septum, which is derived distally from the aortopulmonary septum originating from the posterior wall of the aortic sac between the fourth and the sixth arch, and proximally by the fusion of dextro-superior and sinistroinferior truncus swellings (Van Mierop, 1968). We suggest that the proximal defect (type I) may be caused by defective development of the proximal truncal septum from the two truncus swellings, and the distal defect (type II) by failure of proper fusion between the aortopulmonary septum and the proximal truncal septum. The total defect (type III) may result from defective growth of both the distal and proximal truncal septum.

Retrograde aortography is the most useful investigation in determining the precise location of the defect in patients with aortopulmonary window.

In type I (proximal) defect, aortic root injection opacifies the pulmonary trunk and then both pulmonary arteries (Fig. 5a, b). However, in type II (distal) defect, aortography preferentially opacifies the right pulmonary artery, and the pulmonary trunk and the left pulmonary artery are only faintly opacified. This angiographic feature may help to differentiate type II from type I defect. Preferential opacification of the right pulmonary artery by retrograde aortography can also be seen in anomalous origin of the right pulmonary artery from the ascending aorta, which must be considered in the differential diagnosis of type II defect. At right heart catheterisation, passage of the catheter into the right pulmonary artery or opacification of both pulmonary arteries after injection of contrast medium into either the pulmonary trunk or the right ventricle, should rule out anomalous right pulmonary artery arising from the ascending aorta. In our case 2, injection of dye into the pulmonary trunk was followed by faint opacification of the ascending aorta as a result of a small right-to-left shunt, the window was not crossed during right heart catheterisation, and the correct diagnosis was only established after retrograde aortography.

Diagnosis was particularly difficult in our case 3:

Fig. 5 (a) and (b) In type I (proximal defect), aortic root injection opacifies the pulmonary trunk and then both right and left pulmonary arteries.

during the first catheterisation study the catheter was not passed into either right or left pulmonary artery, and the right ventricular injection opacified only the left pulmonary artery, since the contrast was diluted by an enormous shunt of unopacified blood from the aorta into the right pulmonary artery. Anomalous origin of the right pulmonary artery from the ascending aorta was not excluded until the second catheterisation. The diagnosis of aortopulmonary window was then established by the successful passage of the catheter into the right pulmonary artery and opacification of both pulmonary arteries on the pulmonary arteriogram.

It is necessary to differentiate an atypical persistent ductus arteriosus situated more medially than usual from aortopulmonary window. In persistent ductus, the communication between the systemic and pulmonary circulations is not at the level of the ascending aorta but between the aortic arch and the bifurcation of the pulmonary artery.

Repair of aortopulmonary window has been accomplished by simple ligation, division, or closure of the defect through either a transaortic or a transpulmonary approach. It is agreed that simple ligation is effective in curing small aortopulmonary windows, but re-establishment of the communication has been reported after simple ligation of large defects (Neufeld *et al.*, 1962). For this reason, complete division seems to be superior to ligation. However, serious complications of this procedure have also been reported, including narrowing of the lumen of the great arteries, stenosis of the orifice of the left coronary artery adjacent to the proximal margin of the defect, and accidental injury to the semilunar valves.

In order to avoid these complications, it is recommended that either a transpulmonary (Putnam and Gross, 1966) or transaortic approach should be used (Wright *et al.*, 1968; Deverall *et al.*, 1969). Comparing these two methods, Deverall and Wright stressed the greater safety of aortotomy, compared with an incision into a dilated and thin-walled pulmonary trunk. Proper choice of approach is especially important in type II (distal) defect, in which the transpulmonary approach fails to expose the entire extent of the defect unless the incision is extended further into the right pulmonary artery. The transaortic approach, on the other hand, always gives an excellent view of the whole defect. Moreover, transpulmonary closure of the defect is technically difficult, because the window is situated on the anterior wall of the right pulmonary artery and can only be seen through the orifice of the latter. In the case described by Putnam and Gross (1966), necropsy showed narrowing of the proximal portion of the right pulmonary artery in addition to obstruc-

tive vascular changes. The transaortic approach should, therefore, be used, especially for type II defects, because it not only affords an excellent exposure of the defect, but also provides a safeguard against accidental injury to a semilunar valve or the left coronary ostium.

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