Aorticopulmonary septal defect

An experience with 17 patients

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Seventeen patients with aorticopulmonary septal defect were studied over a period of 15 years. This report includes clinical, electrocardiographic, radiographic, cardiac catheterization, and angiocardigraphic data on the patients. Clinical examination usually showed a large left-to-right shunt and significant pulmonary hypertension. A definitive diagnosis was made by cardiac catheterization and angiography in 12 patients. The presence of a systolic ejection click in 5 patients and a right aortic arch in 3 was a diagnostic help and may differentiate aorticopulmonary septal defect from persistent ductus arteriosus. The catheter position at the time of haemodynamic study was also of diagnostic importance. Associated lesions were present in 9 patients, and corrected in 4 patients. Twelve patients underwent surgery: complete bypass and moderate total body hypothermia were used in 11 patients. Four patients underwent postoperative haemodynamic studies.

Aorticopulmonary septal defect is a broad communication between the left side of the ascending aorta and the right wall of the pulmonary trunk. The aortic and pulmonary valves are both present and usually the ventricular septum is intact.

Many cases are not initially diagnosed until operation or necropsy, particularly when the defect coexists with another cardiac anomaly. The defect has no length and it is usually large in diameter so that pulmonary hypertension results; if operation is delayed, pulmonary vascular disease may complicate operative closure.

By 1968 (Marquis, 1968) less than 100 cases had been well documented in the English language reports. Subsequent case reports have emphasized coexistent conditions and operative repair of the defect (Agius, Rushworth, and Connolly, 1970; Deverall *et al.*, 1969).

This paper describes 17 patients with aorticopulmonary septal defect studied at the University of Minnesota Medical Center. We will discuss the patients as a group, noting differences and similarities and emphasizing the presence of coexistent lesions and operative features where appropriate.

Patients and methods

Nine female patients and eight male patients were

studied. Their ages at the time of definitive diagnosis ranged from 12 days to 35 years (Table 1). Eleven were diagnosed by cardiac catheterization or angiography. Three were diagnosed at operation: 1 during exploratory thoractomy and 2 during an operation to correct a persistent ductus arteriosus. Three infants died in congestive cardiac failure before a diagnosis could be made; at necropsy it was observed that each infant had an aorticopulmonary septal defect.

Nine patients had major coexisting cardiac condition, such as interruption of the aortic arch, tetralogy of Fallot, ventricular septal defect, and persistent ductus arteriosus (Table 1).

Clinical features (see Table 2)

Seven patients were cyanotic. A pulse pressure greater than 50 mmHg was measured in 5 patients. The first heart sound was normal in all of the patients; the second heart sound was accentuated in 14 patients and normal in the other 3. In 5 patients, a systolic ejection click was heard along the left sternal border. In 13 patients an ejection systolic murmur was heard at the upper left sternal border. In 3 of these a grade 2/6 early diastolic murmur of pulmonary regurgitation coexisted. In another patient a grade 3/6 holosystolic murmur was described as present along the upper left sternal border. The only auscultatory abnormality in 1 patient was a decrescendo diastolic murmur present at the upper left sternal border. Two patients had a

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TABLE 1 *Conditions coexisting with aorticopulmonary septal defect*

<i>Lesions</i>	<i>No. of cases</i>
Persistent ductus arteriosus	2
Persistent ductus arteriosus and atrial septal defect	1
Interruption of aortic arch	2
Tetralogy of Fallot	2
Ventricular septal defect	2

machinery-type murmur (typical of persistent ductus arteriosus) heard best at the upper left sternal border. No murmur was heard in a 12-day-old patient with a coexistent interrupted aortic arch and congestive cardiac failure.

Electrocardiographic features (see Table 2)

The QRS axis was between $+30$ and $+120^\circ$ in 14 patients, indeterminate in the frontal plane in 2, and $+150^\circ$ in the remaining patient. The P waves were normal in all but 2 patients who showed right atrial enlargement. PR and QRS intervals were normal. There were no characteristic QRS abnormalities. Four patients, each with a large pulmonary blood flow, showed a deep Q wave in lead V6. The electrocardiograms showed a variable pattern of ventricular hypertrophy. Isolated right ventricular hypertrophy was present in 3 young infants and 2 older patients with pulmonary vascular disease. Biventricular hypertrophy or left ventricular hypertrophy was present in 12 older infants, children, and adults, and was associated with an increased volume of pulmonary blood flow in the 10 in whom it was measured. T waves appeared normal in 15 patients. The other 2, each less than 1 month of age, showed inverted T waves in the left precordial leads.

Radiographic features

Generalized cardiomegaly including left atrial enlargement and increased pulmonary vascularity were present in each of the 17 patients. The aortic knuckle did not appear enlarged. The aortic arch was on the right in 3 patients, 1 with isolated aorticopulmonary septal defect, 1 with a coexistent ventricular septal defect, and 1 with a coexistent tetralogy of Fallot.

Cardiac catheterization and angiographic features (see Table 2)

Cardiac catheterization was performed in 14 patients: the pulmonary artery pressure was raised

to systemic levels in 12 patients; 1 patient was catheterized at 4 years of age and again at 20 years of age and the pulmonary artery pressure was the same on both occasions (45 mmHg); and in the last patient the pulmonary arterial pressure was normal. A large left-to-right shunt was found in 11 patients, a bidirectional shunt was found in a patient with severe pulmonary vascular disease, and a right-to-left shunt in both patients with coexistent interruption of the aortic arch.

The catheter was passed from the pulmonary artery through the aorticopulmonary septal defect to the ascending aorta in 4 patients; in 2 patients the catheter was manipulated into the right carotid artery. The diagnosis was confirmed by selective aortography in 5 patients. In another patient the diagnosis of aorticopulmonary septal defect and interruption of the aortic arch was made by retrograde aortography. Another example of coexistent aorticopulmonary septal defect and interruption of the aortic arch is shown in the Fig.

Operative experience

Twelve patients underwent cardiac operation. The aorticopulmonary septal defect was corrected in 11 patients; the other patient was found to have a coexistent interrupted aortic arch and his chest was closed. Complete cardiopulmonary bypass and moderate total body hypothermia were used in all patients. The aorticopulmonary defect was divided in 9 patients and the defect was closed by the transpulmonary route in the other 2 patients.

Four patients had associated lesions corrected at the same time as the aorticopulmonary septal defect was repaired. Through a ventriculotomy an associated ventricular septal defect was closed with a patch in 2 patients. In one of these patients, the left pulmonary artery appeared to originate from the aorta and was transplanted to the main pulmonary artery at the time of operation. A ventricular septal defect was closed and an infundibular resection performed in a third patient. In the fourth patient the right coronary artery originated from the aorticopulmonary window and was incorporated into the aorta at the time of operation.

Four patients died during operation. One was the patient with a coexistent interrupted aortic arch, one had an isolated aorticopulmonary septal defect, the third had an associated ventricular defect, and the last had a right coronary artery rising from the aorticopulmonary defect.

Patient follow-up

Five patients did not undergo an operation and 3 have died: 1 of them, with an associated atrial septal defect, died before cardiac catheterization

TABLE 2 *Clinical, electrocardiographic, and haemodynamic features in 17 patients with aorticopulmonary septal defect*

Condition	Clinical features						
	Case No.	Sex	Age at diagnosis	Cyanosis	Congestive cardiac failure	Ejection click	Systolic murmur
Isolated aorticopulmonary septal defect	1	F	7½ mth	o	+	+	3/6 ejection LSB
	2	F	8 mth	o	o	o	4/6 ejection LSB
	3	F	2½ yr	+	+	o	3/6 ejection LSB
	4	F	3½ yr	+	+	+	2/6 ejection LSB
	5	M	8 yr	+	o	o	3/6 ejection LSB
	6	M	10 yr	o	o	o	3/6 ejection LSB
	7	M	25 yr	o	o	o	2-3/6 machinery
Isolated aorticopulmonary septal defect and ventricular septal defect	8	F	35 yr	+	o	o	o
Isolated aorticopulmonary septal defect and persistent ductus arteriosus	9	F	1 yr	o	+	o	3/6 ejection LSB
	10	F	8 yr	o	+	+	3/6 ejection LSB
Isolated aorticopulmonary septal defect and persistent ductus arteriosus	11	M	11 mth	o	+	o	2/6 systolic
	12	F	4 yr	+	+	+	2-3/6 machinery
Isolated aorticopulmonary septal defect and persistent ductus arteriosus and atrial septal defect	13	M	2 wk	+	+	o	4/6 ejection LSB
Isolated aorticopulmonary septal defect and interrupted aortic arch	14	M	12 dy	o	+	o	o
	15	M	5½ yr	+	+	+	2-3/6 ejection LSB
Isolated aorticopulmonary septal defect and tetralogy of Fallot	16	F	7 yr	o	+	o	3/6 holosystolic LSB
	17	M	3 mth	o	+	o	5/6 ejection LSB

+ = present; o = absent; $\bar{m}$ = mean pressure; LSB = left sternal border.

could be performed; another patient who had associated tetralogy of Fallot died during cardiac catheterization; the third patient, with isolated aorticopulmonary septal defect, died at 3 years of age from atelectasis and pneumonia. An adult patient with isolated aorticopulmonary septal defect refused surgical intervention. She was last seen at 35 years of age and was cyanotic because of a right-to-left shunt secondary to pulmonary vascular disease. The fifth patient had associated interrupted aortic arch and persistent ductus arteriosus. He is 8 years of age and shows normal growth and development. Though mildly cyanotic, he has no signs of

congestive cardiac failure.

The 8 patients are asymptomatic 2 to 8 years after their operations: 6 have a residual grade 1 to 2/6 ejection systolic murmur at the upper left sternal border. Four patients have no residual shunts by catheterization; the pulmonary artery pressure had decreased in each, the mean pressures being 25 mmHg in 2 patients and 15 mmHg in the other 2.

One major postoperative complication occurred. The patient who underwent ligation and the subsequent division of the aorticopulmonary septal defect developed right hemiplegia soon after the

TABLE 2 (cont'd)

<i>Diastolic murmur</i>	<i>Electrocardiographic features</i>		<i>Cardiac catheterization findings</i>					
	<i>RVH</i>	<i>LVH</i>	<i>Systemic arterial pressure (mmHg)</i>		<i>Pulmonary arterial pressure (mmHg)</i>		<i>Systolic blood flow (l./min per m²)</i>	<i>Pulmonary blood flow (l./min per m²)</i>
o	+	o	116/65	$\overline{75}$	116/60	$\overline{70}$	4.3	11.8
o	o	+	Not measured		75/40	$\overline{68}$	3.0	9.5
o	o	+	104/48	$\overline{77}$	90/56	$\overline{67}$	4.5	22.7
3/6 early blowing LSB	+	+	95/55	$\overline{75}$	95/55	$\overline{72}$	3.5	7.5
o	o	+	105/50	$\overline{78}$	105/55	$\overline{75}$	4.5	10.8
2-3/6 blowing LSB	o	+	90/50	$\overline{75}$	95/50	$\overline{75}$	3.0	10.4
2-3/6 machinery	o	+	110/76	$\overline{94}$	45/20	$\overline{40}$	2.8	3.5
2/6 blowing LSB	+	o	105/60	$\overline{80}$	105/60	$\overline{80}$	3.5	3.5
o	o	+	No catheterization					
2/6 blowing LSB	+	+	105/75	$\overline{85}$	105/75	$\overline{85}$	3.4	6.9
	+	+	80/50	$\overline{60}$	80/45	$\overline{57}$	2.7	8.0
2-3/6 machinery	+	+	110/50	$\overline{75}$	28/30	$\overline{22}$	4.2	15.6
o	+	o	No catheterization					
o	+	o	125/60	$\overline{80}$	120/60	$\overline{85}$	Not measured	
o	+	o	130/75	$\overline{95}$	130/75	$\overline{95}$	—	9.2
1/6 diastolic apex	+	+	105/50	$\overline{77}$	100/55	$\overline{75}$	4.2	20
o	+	+	No catheterization					

second operation. The condition improved, however, and 4 years after the operation there were only mild neurological findings.

Comment

An initial diagnosis of aorticopulmonary septal defect is often made at operation or necropsy, both in published cases and in our own. Though the first clinical diagnosis was reported in 1949 by Dadds and Hoyle, the common occurrence of pulmonary hypertension, the clinical features similar to persistent ductus arteriosus, and the high incidence of associated lesions make the diagnosis difficult.

Once suspected, however, a diagnosis can be readily confirmed by cardiac catheterization and angiography.

Clinical features, related primarily to congestive cardiac failure, are not specific for aorticopulmonary septal defect. The findings on physical examination are often similar to those of ventricular septal defect or a persistent ductus arteriosus. The presence of a wide pulse pressure and accompanying prominent peripheral pulses help to distinguish aorticopulmonary septal defect from ventricular septal defect and indicate the presence of communication between the aorta and right side of the heart.

FIG. Angiograms in Case 15. Coexistent aorticopulmonary septal defect and interruption of aortic arch. A) Aortogram. Anteroposterior view. Contrast injected into ascending aorta. Prompt opacification of pulmonary arterial system from ascending aorta. Ascending aorta ends blindly (arrow). Faint opacification of descending aorta. B) Right ventriculogram. Lateral view. Opacification of right ventricle and pulmonary trunk. Dense filling of descending aorta through persistent ductus arteriosus. Faint opacification of ascending aorta through aorticopulmonary septal defect.

The presence of an ejection click and the absence of a diastolic component of the murmur tend to distinguish this condition from persistent ductus arteriosus. In addition, the aortic knuckle was not enlarged in patients with aorticopulmonary septal defect, whereas it is frequently enlarged in patients with persistent ductus arteriosus. The presence of a murmur of pulmonary insufficiency was found in 4 of our patients, and in our experience this is unusual in patients with persistent ductus arteriosus at a young age.

That the aortic arch may be on the right in aorticopulmonary septal defect has been reported (Deverall *et al.*, 1969; Somerville, 1959; Scott and Sabiston, 1953). A right aortic arch was present in 3 of our patients, one of whom had tetralogy of Fallot. This frequency may serve as a diagnostic feature in patients with a left-to-right shunt at the level of the great vessels. Aorticopulmonary septal defect resembles persistent truncus arteriosus in which a high incidence of right aortic arch is found also.

Neither the electrocardiogram nor the thoracic

radiograph are characteristic for aorticopulmonary septal defect, but they do help in the assessment of pulmonary vascular changes.

At cardiac catheterization in patients with an aorticopulmonary septal defect, a shunt at the pulmonary artery level is usually found and the pulmonary artery pressure is usually raised to systemic levels. In 2 of our patients, however, the peak systolic pressures were 45 and 25 mmHg, respectively. Thus, pulmonary hypertension does not invariably coexist with aorticopulmonary septal defect. In 1 of these 2 patients, a continuous murmur was present, reflecting the pressure difference between the aorta and pulmonary artery throughout the cardiac cycle.

Several anomalies have been associated with aorticopulmonary septal defect (Neufeld *et al.*, 1962; Hurwitz, Ruttenberg, and Fonkalsrud, 1967; Somerville, 1959). In our series, 9 of 17 patients (53%) had associated major cardiovascular lesions (Table 1). The most common associated lesion was persistent ductus arteriosus (3 patients). This was an isolated association in 2 patients. After the ductus

had been divided in these patients, persistence of the preoperative symptoms led to further investigation and a diagnosis of aorticopulmonary septal defect. In the third patient with associated persistent ductus arteriosus, a coexistent atrial septal defect was found.

There were 2 patients with associated interrupted aortic arch. This association has been reported in only 4 previous instances (Chiemmongkoltip, Moulder, and Cassels, 1971).

The association with tetralogy of Fallot is very uncommon (Cooley, McNamara, and Latson, 1957). In our 2 patients with this association, the large shunt at the pulmonary arterial level allowed high pulmonary artery pressures and the severe infundibular obstruction was masked at cardiac catheterization. Similar observations were present in Case 3 of Cooley *et al.* (1957). In one of our patients, after closure of the aorticopulmonary septal defect and repair of the intracardiac lesion, the pulmonary artery pressure fell, but was still slightly raised and a moderate gradient was present across the pulmonary outflow tract.

In one of our patients, the right coronary artery originated from the aorticopulmonary fistula and was incorporated into the aorta during the surgical correction. Anomalous coronary arteries arising from the defect, or from a persistent ductus arteriosus, or from the pulmonary artery, have been described in at least 2 other instances (Morrow, Greenfield, and Braunwald, 1962; Schumacker, 1957).

Coarctation of the aorta, membranous subaortic stenosis, pulmonary arteriovenous fistula, atrial septal defect, and persistent left superior vena cava are other coexisting lesions which have been described with aorticopulmonary septal defect (Neufeld *et al.*, 1962).

The aorticopulmonary septal defect is similar, haemodynamically, to persistent ductus arteriosus, but the operative management is more complicated. Aorticopulmonary septal defect resembles a short 'fat' ductus, since the walls are thin and friable. Usually the defect is large, the posterior aspect is thinned out, and the fistula is under high pressure.

Cooley *et al.* (1957) first described the method of division of the lesion using complete cardiopulmonary bypass.

Although operation using cardiopulmonary bypass facilitates the approach to the lesion, it certainly does not obviate the necessity for dividing the fistula. Schumaker (1957) suggested exploration through the pulmonary artery; this was successfully completed in at least one patient. Wright, Freeman, and Johnston (1968) employed a transaortic approach successfully in one patient. Deverall *et al.* (1969) also used the transaortic approach in 3 patients and

they were successful in each instance. They were impressed with this method, partly because it allowed an easy approach and offered a clear view of the defect. Fistula dissection is not necessary using the transaortic approach; the arteriotomy is in a normal aortic wall, not in a thin-walled vessel.

When the aorticopulmonary septal defect is repaired and there are associated abnormalities of the coronary arteries, these arteries need to be transplanted into the aorta. Joining a cuff of associated arterial wall to the coronary artery facilitates the transplantation (Burroughs *et al.*, 1962).

References

- Agius, P. V., Rushworth, A., and Connolly, N. (1970). Anomalous origin of left coronary artery from pulmonary artery associated with an aorto-pulmonary septal defect. *British Heart Journal*, **32**, 708.
- Burroughs, J. T., Schmutz, K. J., Linder, F., and Nuehaus, G. (1962). Anomalous origin of the right coronary artery with aorticopulmonary window and ventricular septal defect. Report of a case with complete operative correction. *Journal of Cardiovascular Surgery*, **3**, 142.
- Chiemmongkoltip, P., Moulder, P. V., and Cassels, D. E. (1971). Interruption of the aortic arch with aorticopulmonary septal defect and intact ventricular septum in a teenage girl. *Chest*, **60**, 324.
- Cooley, D. A., McNamara, D. G., and Latson, J. R. (1957). Aorticopulmonary septal defect. Diagnosis and surgical treatment. *Surgery*, **42**, 101.
- Dadds, J. H., and Hoyle, C. (1949). Congenital aortic septal defect. *British Heart Journal*, **11**, 390.
- Deverall, P. B., Aberdeen, E., Bonham-Carter, R. E., and Waterston, D. J. (1969). Aortopulmonary window. *Journal of Thoracic and Cardiovascular Surgery*, **57**, 479.
- Hurwitz, R. A., Ruttenberg, H. D., and Fonkalsrud, E. (1967). Aorto-pulmonary window, ventricular septal defect and mesoversion. Surgical correction in an infant. *American Journal of Cardiology*, **20**, 566.
- Marquis, R. M. (1968). Aortopulmonary window. In *Paediatric Cardiology*, p. 286. Ed. by H. Watson. Lloyd-Luke, London.
- Morrow, A. G., Greenfield, L. J., and Braunwald, E. (1962). Congenital aortopulmonary septal defect. Clinical and hemodynamic findings, surgical technic, and results of operative correction. *Circulation*, **25**, 463.
- Neufeld, H. N., Lester, R. G., Adams, P., Anderson, R. C., Lillehei, C. W., and Edwards, J. E. (1962). Aorticopulmonary septal defect. *American Journal of Cardiology*, **9**, 12.
- Schumaker, H. B. (1957). In discussion of Cooley, McNamara and Latson, Aorticopulmonary septal defect. Diagnosis and surgical treatment. *Surgery*, **42**, 120.
- Scott, H. W., and Sabiston, D. C. (1953). Surgical treatment for congenital aorticopulmonary fistula. Experimental and clinical aspects. *Journal of Thoracic Surgery*, **25**, 26.
- Somerville, J. (1959). Aortopulmonary septal defect, five cases treated by operation. *Guy's Hospital Reports*, **108**, 177.
- Wright, J. S., Freeman, R., and Johnston, J. B. (1968). Aortopulmonary fenestration. A technique of surgical management. *Journal of Thoracic and Cardiovascular Surgery*, **55**, 280.

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