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Management of pulmonary atresia

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Based on experience with 48 patients, a new simple classification of pulmonary atresia, according to pulmonary artery development is suggested. The need for extensive investigation including biplane aortography and right ventricular angiocardiography is emphasized. Adequate display of the anatomy is required to plan the surgical management. Early palliative procedures are advised for hypoxia and later radical correction is possible in a large number of these patients.

Pulmonary atresia is usually associated with hypoxia and disability in early life. Survival depends upon the adequacy of the collateral blood supply to the lungs, a persistent duct, or a 'man-made' shunt. Infants with pulmonary atresia may occasionally be in thriving difficulties due to an excessive pulmonary blood flow when the duct or anomalous collateral vessels from the descending aorta are large. This is uncommon, and in the majority of patients the collateral arterial development is inadequate to prevent hypoxia during the first decade of life. Surgical treatment is, therefore, indicated in most patients with pulmonary atresia during childhood.

The principles of surgical treatment depend upon the anatomy and extent of the pulmonary atresia which must be clearly delineated before surgical management is planned. This may be difficult in those with a poor pulmonary blood flow and often extensive careful angiocardiography is required.

Views on the classification and management of pulmonary atresia are based on a series of 48 consecutive patients with pulmonary atresia referred to the National Heart Hospital during the past 5 years.

Terminology

Pulmonary atresia[1] should be defined as a complete obstruction between the right ventricle and pulmonary arteries. In those patients with pulmonary atresia where there is no development of the main pulmonary arteries and large vessels arise from the aorta to perfuse distal pulmonary arteries or the lungs direct, the similarity to true truncus arteriosus (common trunk) is obvious. However, the essential feature that distinguishes the patient with pulmonary atresia from truncus is that there is a completely obstructed right ventricular outflow tract which is separate from and anterior to the aortic root (Fig. 1) in the absence of transposition of the great arteries. When a form of transposition is present, the blind pouch is medial or posterior to the aortic root. The atretic outflow must be seen before the condition is classified as pulmonary atresia. This author considers that the use of such terms as Fallot variant, types 1–4 truncus, and pseudotruncus does little to help in the understanding or treatment of this difficult group of patients who should be classified together as variants of pulmonary atresia. Having accepted the concept of pulmonary atresia as a separate entity from other lesions that it may resemble, it is suggested that the wide and strange variations are classified according to the extent of the atresia and the degree of pulmonary artery development beyond the atretic segment.

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[1] Atresia means occlusion of a natural channel of the body. *The Shorter Oxford English Dictionary*, 3rd ed., The Clarendon Press, Oxford.

Investigation

For identification and classification of pulmonary atresia, good angiography is necessary. A complete investigation of a patient with suspected pulmonary atresia should show not only the anatomy of the ventricular septal defect and confirm the presence of pulmonary valve atresia (Fig. 2) which may also involve the infundibulum, but should also display the position of large systemic collateral arteries and the site and patency of 'man-made' shunts or a duct. Both aortic and right ventricular injections are necessary (Ongley *et al.*, 1966; Somerville, 1969), and biplane still film angiography has given the most satisfactory results. Cine-angiography has been found to

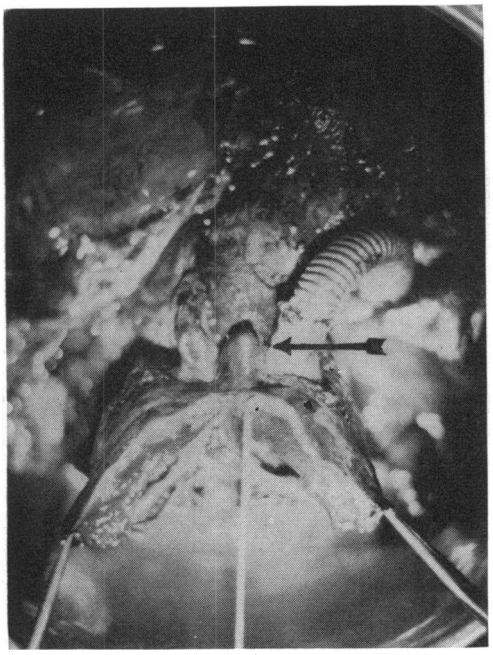


FIG. 1 *Photograph of necropsy specimen from Case 43. Arrow indicates the dome of atresia of right ventricular outflow tract in which a probe has been placed.*

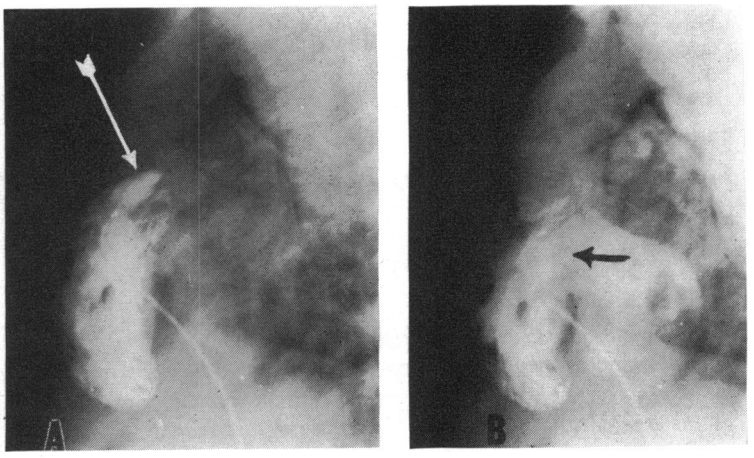


FIG. 2 *Lateral view of right ventricular angiocardiogram from Case 25, showing (A) atretic pulmonary valve (arrow) lying anterior to the aortic root, (B) ventricular septal defect (arrow) in Fallot position with aortic override.*

be adequate for identifying the atretic valve and anatomy of the ventricular septal defect, but the definition has not been good enough to distinguish systemic from pulmonary arteries and delineate their anatomy. A right ventricular injection of contrast medium of 1 ml./kg. is given to show the ventricular septal defect, and in order to identify the blind atretic pouch with certainty the tip of the catheter is placed below the region of the outflow tract (Fig. 2) rather than in the conventional position in the right ventricular body towards the apex. If a right ventricular outflow tract injection is given, the dose of contrast medium should be reduced to $\frac{1}{2}$–$\frac{3}{4}$ ml./kg., and the injection pressure should also be reduced.

Provided aortography is also done, an angiocardiographic cover time of the right ventricular injection need be only 1–2 sec. In the small group of patients with pulmonary atresia and intact ventricular septum, a longer angiocardiographic cover time should be used to enable clear definition of coronary sinusoids or fistulous communications, and if the test injection has shown the cavity of the right ventricle to be small, less than 1 ml./kg. of contrast medium is used at lower pressure than is given in a conventional right ventricular angiocardiogram.

Aortography is used routinely in the investigation of pulmonary atresia to display the anatomy of the collaterals, duct, surgical anastomoses, associated aortic valve regurgitation, aortic arch anomalies, and pulmonary artery development distal to the atretic area (Rees and Somerville, 1969). In many patients the catheter can be passed directly from the right ventricle into the ascending aorta, but in 20 per cent of patients with pulmonary atresia the aortic root may lie in a horizontal plane, making it difficult for the catheter to enter the ascending aorta from the right ventricle. When this occurs retrograde aortography should be performed either by Seldinger technique or through a femoral arteriotomy. The choice of technique depends on the age of the patient and experience of the investigator. The approach from the leg has been found to be preferable to brachial arteriotomy as selective injection of large collateral 'bronchial' arteries arising from the descending aorta may be needed to differentiate pulmonary artery development from additional large systemic arteries entering the lung (Fig. 3). When the contrast medium is injected into the ascending aorta the origin of these vessels arising from the arch or lower down may be obscured and selective injections into the vessels as well as routine ascending aortography may be necessary to be certain of the anatomy.

In patients with clinical and radiological evidence of a good pulmonary blood flow, a large injection of contrast (1½–2 ml./kg. body weight) is given into the aorta. The pulmonary

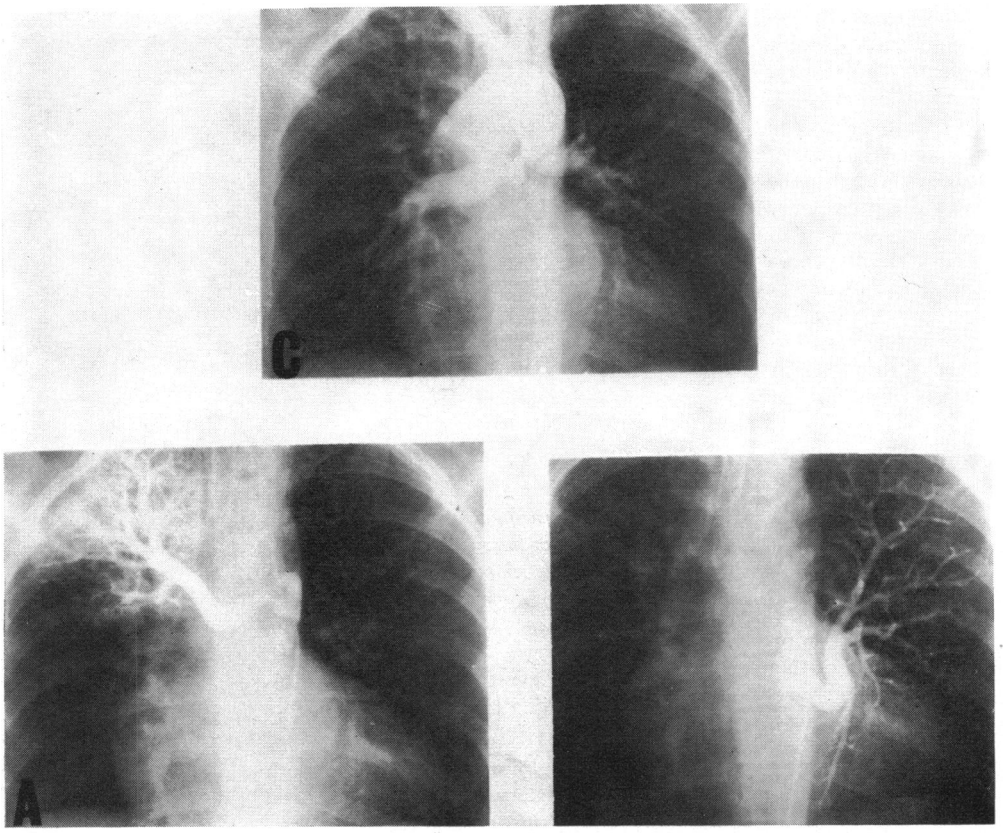
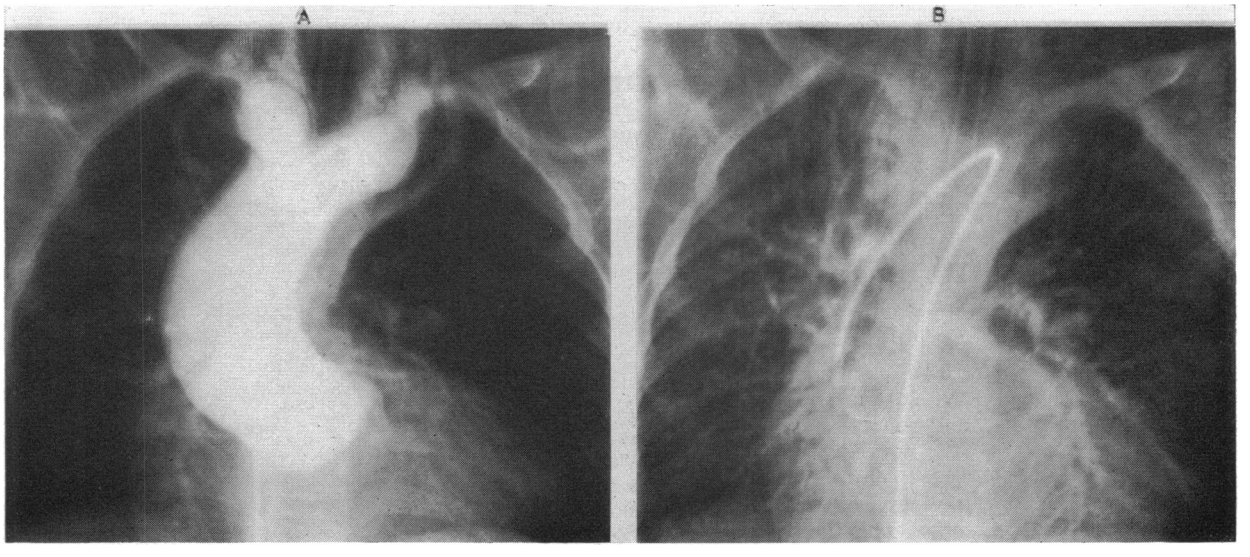


FIG. 3 *Selective injection of arteries arising from the descending aorta to perfuse (A) right upper lobe, (B) left upper lobe and lingula, and (C) pulmonary arteries filled through duct shown from a contrast injection into the arch of the aorta (Case 39).*

FIG. 4 *Retrograde aortogram from Case 32 showing (A) double aortic arch, (B) faintly opacified pulmonary arteries in late films taken 5 seconds after the injection.*



vessels opacify quickly under these circumstances and 3–4 seconds of angiocardiographic cover time are enough to display pulmonary artery anatomy. If, however, the patient is deeply cyanosed with poor pulmonary blood flow, a long angiocardiographic cover time of up to 8 seconds should be given in order to see the 'delayed' opacification of the pulmonary arteries (Fig. 4). This is important even in those patients who have had a shunt procedure. A continuous murmur does not necessarily mean a functioning shunt, as the noise may arise from collaterals which tend to develop when there has been thrombosis in the pulmonary artery at the site of attempted operation.

In patients with an intact ventricular septum, the catheter is passed across the atrial septum into the left ventricle, and left ventricular angiocardiography rather than aortography is done. As patients with pulmonary atresia and intact ventricular septum do not have bizarre systemic vessels perfusing the lungs, adequate information is obtained from the left ventricular angiogram. As part of the investigation, routine oxygen saturations are measured and numerous estimations of systemic arterial saturation should be made as this varies with the changes in arterial blood pressure. In older children, gas collection over a 5-minute period is obtained in order to calculate pulmonary blood flow using the following formula.

$$\frac{\text{Oxygen consumption}}{\text{Oxygen content Pulm. venous—Oxygen content Aorta}}$$

Classification

For clinical use, it is suggested that pulmonary atresia is classified according to the extent of the atresia and development of the pulmonary arteries distal to the atresia, as it is upon this that surgical management depends. A simple classification is as follows (Fig. 5):

1 Type 1 (Fig. 5A)—Atresia of pulmonary valve. Beyond there is complete pulmonary artery development.

2 Type 2 (Fig. 5B)—Atresia of pulmonary valve and main pulmonary artery. Right and left pulmonary arteries are present, which may be separate or joined (Fig. 6).

3 Type 3 (Fig. 5C)—Atresia of pulmonary valve, main pulmonary artery, and one main branch. Either right or left pulmonary artery is present and enters the lung normally. In the lung which does not have a main branch pulmonary artery the distal pulmonary artery plexus is perfused by collaterals. The angiocardiogram from Case 19 shows Type 3 pulmonary atresia (Fig. 7).

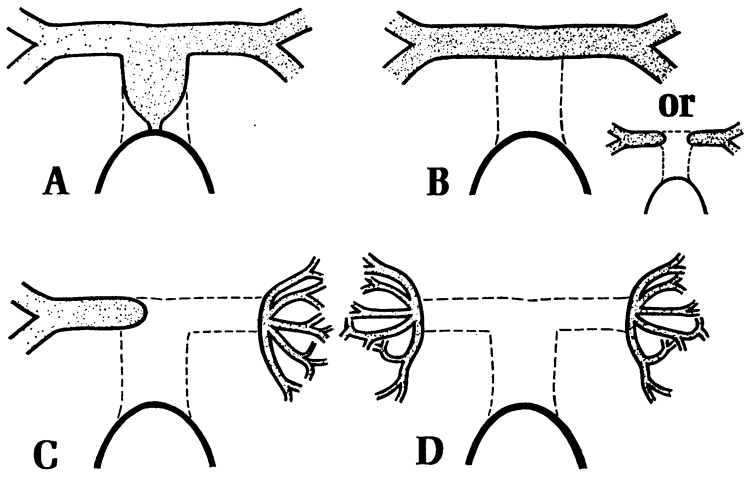


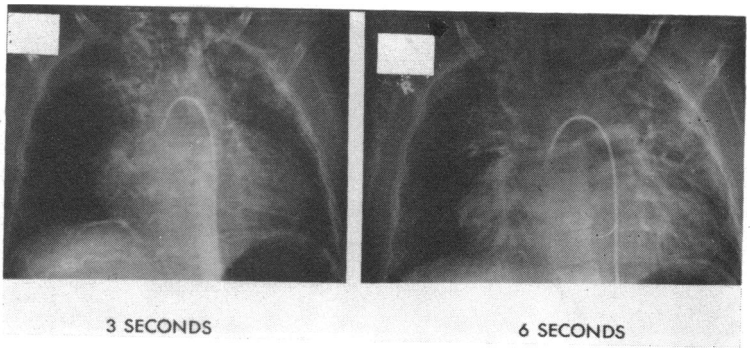
FIG. 5 *Diagram to show simple classification of pulmonary atresia based on the degree of pulmonary artery development distal to the atretic segment. (A) Type 1, (B) Type 2, (C) Type 3, (D) Type 4.*

4 Type 4 (Fig. 5D)—Atresia of the pulmonary valve, main pulmonary artery, and both main pulmonary artery branches. The distal pulmonary artery plexus is perfused by collateral vessels entering the hila of the lungs (Fig. 8).

Using this simple classification, the 48 patients were classified irrespective of any additional lesions as follows: Type 1 (29 patients); Type 2 (13 patients); Type 3 (3 patients); and Type 4 (3 patients).

Patients with pulmonary atresia and intact ventricular septum appear from the one example in this series (Case 23) and in other

FIG. 6 *Type 2 pulmonary atresia with development of the right and left pulmonary artery opacified on the late films from retrograde aortogram from Case 17. At 3 seconds only collaterals from aortic branches were filled.*



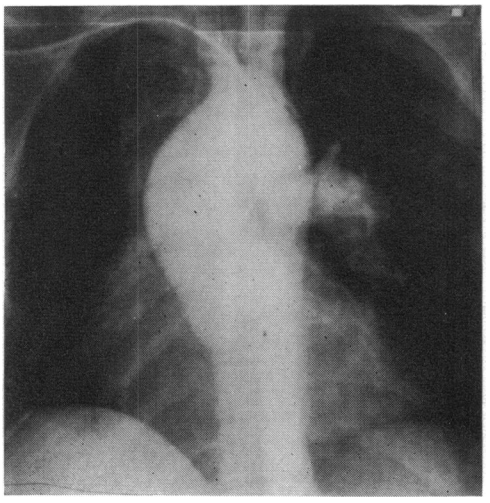


FIG. 7 *Angiocardiogram from Case 19 showing Type 3 pulmonary atresia. There is no right pulmonary artery visualized but a large left pulmonary artery has been filled through a duct.*

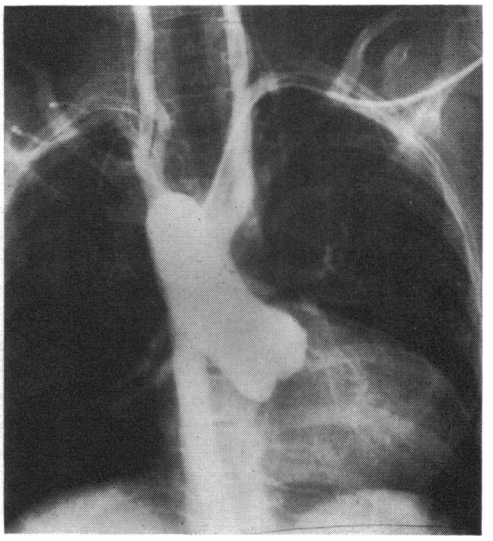
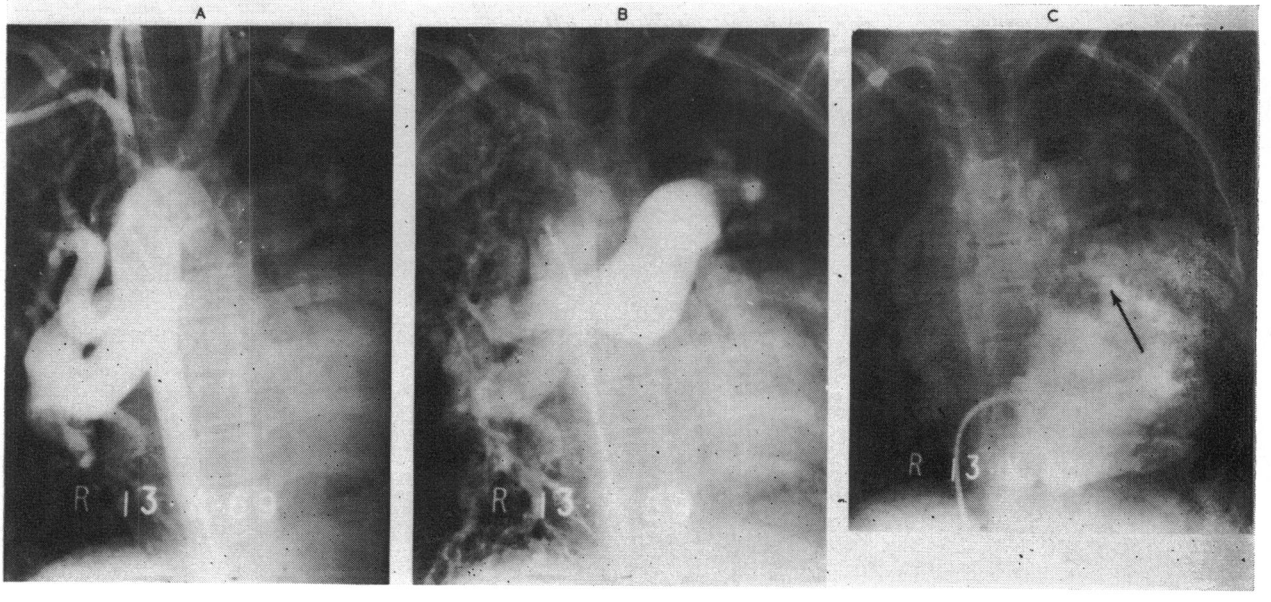


FIG. 8 *Aortogram from Case 24, showing Type 4 pulmonary atresia and right aortic arch with only a small hilar pulmonary artery present in left lung which perfused left upper lobe and probably the lingular; absence of the main and right and left pulmonary arteries was confirmed by bilateral thoracotomies.*

studies (R. Radley-Smith, personal communication) to have complete pulmonary artery development beyond the atresia, thus always falling into the category of Type 1. This type of pulmonary atresia with intact ventricular septum more commonly presents in infancy and has different problems and a different natural history from the commoner form with large ventricular septal defect. It is considered that this special type of pulmonary atresia should be discussed separately.

This simple classification of pulmonary atresia based on pulmonary artery develop-

FIG. 9 *Huge systemic artery from descending aorta giving off arteries to perfuse (A) the right lung, (B) crossing the hilum to perfuse the left lung, (C) atretic right ventricular outflow tract shown (arrow) by right ventricular angiocardiogram. From Case 40.*



ment is not altered by the presence of the large systemic arteries arising from the descending aorta or aortic arch main branches which perfuse the lungs direct or through pulmonary artery branch remnants. These arteries which profoundly affect physiology and management usually arise close to the duct area or lower down, and were found in 10 patients in various forms (Fig. 3 and 9) which will be discussed elsewhere (Jefferson, Rees, and Somerville, 1970. The atretic right ventricular outflow tract was clearly shown either by angiocardiography (Fig. 9C) or at necropsy (Fig. 1) in all. Three patients with these large vessels had Type 3 pulmonary atresia, 6 had Type 2, and 1 had Type 1 pulmonary atresia with complete development of the main and right and left pulmonary arteries. It is the type of pulmonary atresia with large arteries arising direct from the descending aorta which has been previously classified as Type 4 truncus (Collett and Edwards, 1949).

Additional cardiovascular abnormalities

Only major abnormalities have been noted (Table). Forty-four patients had a large ventricular septal defect of the Fallot type; in one of these (Case 38) the defect was larger, extending posteriorly under the septal cusp of the tricuspid valve. Three patients with transposition had single ventricle (Cases 31, 36, and 45) and one (Case 23) had no defect in the ventricular septum. The duct persisted after 6 months in 10 patients. In at least a further 8 patients the duct had been suspected or shown to be open in the neonatal period and spontaneous closure was documented later.

Left superior vena cava, noted radiologically, at catheter, or at operation was found in 11 patients (24%). Absence of the right superior vena cava was found at the time of total correction (Case 30) but was not suspected from previous investigations. The atrial septum was patent to the exploring catheter tip in 79 per cent, but the incidence of true atrial septal defect is not known.

Anomalous pulmonary veins from the right upper and middle lobes entering the superior vena cava in 3 (Cases 21, 44, and 48) were found at thoracotomy and were unsuspected from investigations.

Transposition of the great arteries was present in 7 patients (Fig. 10), it was 'corrected' in 2 (Cases 10 and 47), and in a probable third patient (Case 22). One other patient (Case 4) had an aortic root in the normal position but the aortic valve faced anteriorly.

Twenty-one patients (40%) had a right aortic arch, including one with a double arch (Case 32) and another with dextrocardia and a left arch. Dextrocardia was present in 3 patients, 2 of whom had transposition of the great arteries.

A single coronary artery was found in one (Case 5) and aortic regurgitation with a bicuspid aortic valve was seen in another (Case 37). Bicuspid aortic valve was suspected from the appearance of the aortogram in a further 2 patients.

Stenoses of pulmonary artery sub-branches

FIG. 10 *Angiocardiogram from patient with corrected transposition associated with pulmonary atresia. (A) early frames showing aortic root displaced to left and inverted ventricular anatomy, (B) late films showing shadowy opacification of pulmonary arteries.*

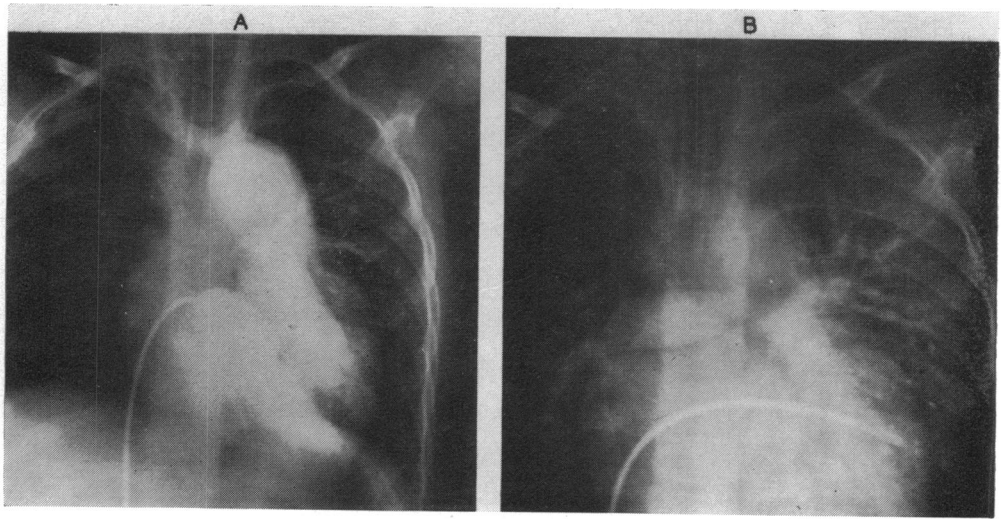


TABLE *Clinical data, surgical management, and type of pulmonary atresia in 48 patients*

Case No.	*Sex*	*Age (yr.)*	*Hb (g./100 ml.)*	*Right aorta*	*Persistent ductus arteriosus*	*Other abnormalities*	*Palliative operations*							
							Age (yr.)	*1st op.*	*Age (yr.)*	*2nd op.*	*Age (yr.)*	*3rd op.*	*Plan*	*Type 1–4*
1	M	14	21	–	–	—	3	Lt. Blalock	12	Waterston	—	—	Awaiting total corr.	1
2	M	15	20·5	–	–	—	2	Lt. Blalock	—	—	—	—	Total corr. 14 yr.	1
3	F	6	19·8	–	–	—	6/12	Lt. Blalock	5	Waterston	—	—	Awaiting total corr.	2
4	F	8	21·3	–	–	Aortic valve	1	Rt. Blalock	4	Lt. Blalock	—	—	Total corr.	1
5	F	3	22	+	–	Single coron art.	2	Waterston	—	—	—	—	? Total corr. later	2
6	M	14	17·4	–	+	—	—	—	—	—	—	—	No operation	2
7	M	2	22·8	+	–	Mongol	2	Waterston	—	—	—	Died, peric. tamp.		1
8	F	6	18·4	–	–	—	6/12	Rt. Blalock	2	Lt. Blalock	6	Waterston	? Total corr.	1
9	M	6	20·2	+	–	—	6	Attempted rt. Waterston	6½	Lt. Blalock	—	—	None	4
10	F	17	22·4	+	–	Corr. transp.	6	Rt. Blalock	17	Waterston	—	—	? Total corr. later	1
11	F	6	17·8	+	–	—	1	Rt. Blalock	6	Waterston	—	—	Awaiting total corr.	1
12	M	6	18·4	+	–	—	6	Waterston	—	—	—	—	Awaiting total corr.	2
13	M	8	19·2	+	–	—	—	Paediatrician considering shunt						1
14	M	14	16·8	–	–	—	—	—	—	—	—	—	? Awaiting total corr.	2
15	F	15	24·2	–	—	—	13	Waterston	—	—	—	—	Total corr., 14	1
16	M	10	18·4	+	–	—	—	—	—	—	—	—	Total corr., 10	1
17	F	14	21·8	–	–	—	4	Rt. Blalock	10	Lt. Blalock	12	Attempted Waterston		2
18	F	9	19·4	+	–	—	3	Rt. Blalock	—	—	—	—	Total corr., 10	1
19	F	15	18·6	–	+	—	—	—	—	—	—	—	Awaiting rt. thoracotomy	3
20	F	13	22·6	–	–	—	4	Lt. Blalock	13	Waterston	—	—	Total corr., 14	1
21	F	6	20·2	–	+	Anom. rt. pulm. vein	6/12	Rt. Blalock	6	Waterston	—	—	Awaiting total corr.	1
22	M	6	19·6	–	+	Transp. gt. art.	6	Lt. Waterston	—	—	—	—	—	2
23	F	8	18·2	–	–	No VSD	5/12	Lt. Blalock	8	Pulmonary valvotomy	—	—	—	1
24	F	14	18·6	+	–	—	13	Rt. thoracot.	13	Lt. Blalock	—	—	—	4
25	M	12	15·0	–	–	—	—	—	—	No operation planned				2
26	M	4	17·2	+	+	—	4/12	Waterston	4	Lt. Blalock	—	—	Awaiting total corr.	1
27	M	7	20·1	–	–	—	4	Rt. Blalock	7	Waterston	—	—	? Awaiting total corr.	2
28	M	9	24·4	+	–	Transp. gt. art.	9	Lt. Waterston	—	—	—	—	—	1
29	F	4	21·1	–	+	—	4	—	—	—	—	—	Awaiting shunt	1
30	F	12	18·0	–	+	Abs. rt. sup. vena cava, pulm. art. sten.	—	—	—	—	—	—	Total corr.	1
31	M	6	24·0	+	+	Transp. gt. art., sing. vent.	6	Waterston	—	—	—	—	—	1
32	M	6	17·4	+	–	Double aorta	6	Waterston	—	—	—	—	Awaiting total corr.	2
33	M	10	20·1			—	10	Waterston	—	—	—	—	—	1
34	M	13	18·2	+		—	—	—	—	—	—	—	—	3

TABLE—*continued*

Case No.	Sex	Age (yr.)	Hb (g./100 ml.)	Right aorta	Persistent ductus arteriosus	Other abnormalities	Palliative operations							
							Age (yr.)	1st op.	Age (yr.)	2nd op.	Age (yr.)	3rd op.	Plan	Type 1–4
35	F	8	20·2	–	–	Rubella; multiple defects	2	Lt. Blalock	8	Waterston	—	—	—	1
36	F	6	21·8	+	+	Dextrocardia; transp. gt. art., sing. vent.	6	Potts	—	—	—	—	—	1
37	M	9	24·2	–	–	Aortic regurg.	1	Rt. Blalock	5	Lt. Blalock	9	Waterston	Awaiting total corr.	1
38	M	13	20·0	–	–	—	4	Lt. Blalock	13	Waterston	—	—	Total corr.	1
39	F	14	17·6	–	–	—	—	—	—	—	—	—	Awaiting total corr.	2
40	F	13	15·1	+	–	—	—	—	—	—	—	—	—	2
41	M	3	16	+	–	—	—	—	—	—	—	—	Awaiting thoracotomy	4
42	M	10	22·6	+	–	—	10	Waterston	—	—	—	—	Awaiting total corr.	2
43	F	6	18·4	–	+	—	6	Attempted Waterston		Died, thoracotomy	—	—	—	3
44	M	8	20·2	+	–	Anom. rt. upper pulm. veins	8	Waterston	—	—	—	—	Awaiting total corr.	1
45	F	12	18·4	–	–	Transp. gt. art., sing. vent.	1	Rt. Blalock	2	Potts	—	—	Awaiting total corr.	1
46	M	6	24·4	–	–	Dextrocardia, lt. aorta	6	Lt. Waterston	—	Died, lung haemorrhage, peric. tamponade			—	1
47	F	12	18·6	+	–	Dextrocardia, ? corr. transp. gt. art.	6	Lt. Blalock	—	—	—	—	—	1
48	F	14	25·2	–	–	Anom. rt. upper veins	2	Thoracotomy	14	Waterston	—	—	Awaiting total corr.	1

were common at the point where the sixth arch joined up with the pulmonary plexus in the hilum. In one patient (Case 30) there were more proximal stenoses in the main pulmonary artery branches (Fig. 11), resembling those seen in the rubella syndrome which could have been an aetiological factor in this patient. Infundibular hypertrophy and organic stenosis were commonly present beneath the pulmonary valve atresia, and in 5 patients the narrow area was 1–1·5 cm. in length. No patient with infundibular atresia and an open deformed pulmonary valve was included in this series, as this situation is probably acquired in severe Fallot and not present from birth.

Lesions outside the cardiovascular system

Congenital abnormalities outside the cardiovascular system were not specifically searched for though routinely a late film was taken after the angiocardiogram in order to see the kidneys. Congenital abnormalities of the kidney were found in 5 patients.

One patient was a mongol (Case 7), and one had rubella syndrome (Case 35), which was suspected but unproven in another child (Case 30).

Principles of management

Following complete demonstration of the anatomy, the management was planned. In the sick hypoxic infant and young child a right ventricular angiocardiogram only was often done before an emergency shunt procedure, and sometimes when the routine chest radiograph suggested the presence of a right pulmonary artery; no preliminary investigation was undertaken before thoracotomy.

The indications for a shunt procedure were severe hypoxia and effort intolerance with haemoglobin levels of 18·6–26 g./100 ml. In patients with Type 1 or Type 2 pulmonary

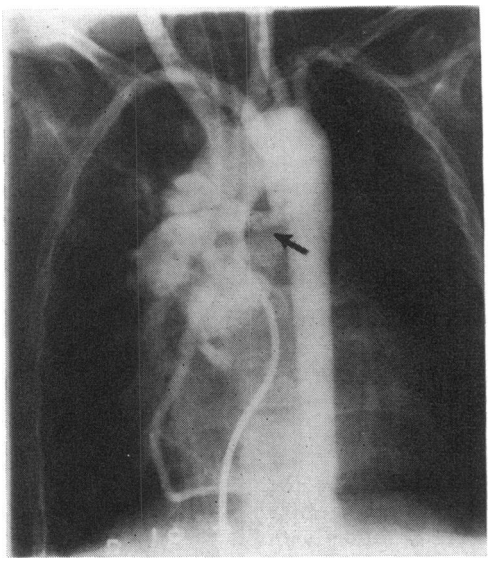


FIG. 11 *Aortogram from Case 30 showing persistent duct (arrow) preferentially filling the right pulmonary arteries with small distal pulmonary artery branches.*

atresia, where it was considered that radical corrective surgery would be possible at some time, a Waterston anastomosis was the shunt of choice as it was the easiest to close at the time of total correction (Somerville *et al.*, 1969). When this had already been done and the child still had an important hypoxic problem, a left Blalock anastomosis was performed. Five patients had required an emergency shunt before the age of 1 year and a further 16 had the first shunt before 6 years. In patients with transposition of the great arteries complicating pulmonary atresia, a modified Waterston or Potts anastomosis was done.

In patients classified as Type 3 and 4 pulmonary atresia, a right thoracotomy was done first in the hope of finding a hilar pulmonary artery suitable for Waterston's anastomosis. If nothing could be achieved, the other side of the chest was opened at a later date. This approach was successful in establishing a beneficial shunt in 2 patients (Cases 9 and 24), but no shunt could be achieved after right thoracotomy alone in Case 43. This policy was not recommended unless the patient was extremely disabled with haemoglobin above 19 g./100 ml.

There were 35 patients with Type 1 or 2 pulmonary atresia without any form of transposition, in whom radical corrective surgery using a homograft reconstruction of the atretic segment (Ross and Somerville, 1966) or fascia lata valve (Ionescu and Ross, 1969) was technically possible. Ideally, this would not be undertaken until after age 8 years or in patients with haemoglobin levels above 18·5 g. When patients presented at the right age, with ideal anatomy but with a high haemoglobin, a preliminary shunt was performed 6–12 months before total correction in order to develop the left side of the heart and reduce the haematocrit. This policy was followed in 5 patients who have since had successful surgery with homograft replacement (Ross and Somerville, 1968). Full details of technique and follow-up of these operations are discussed elsewhere (Ross and Somerville, 1970. Two patients (Cases 18 and 30) had total correction at age 10 and 12 years without a previous shunt procedure. In the first there were problems from left heart failure and large remaining systemic collaterals, but not in the second whose large duct was closed at the time of radical correction.

There remains a group of patients with pulmonary atresia who survive to adolescence without extreme hypoxia or without needing a palliative shunt procedure. Eight patients were in this clinical category; 6 had large systemic vessels perfusing the lungs and 2 had a moderate size duct. The disability in the 2 with a large duct increased considerably at puberty after a period of rapid growth and total correction had been advised.

The 6 patients with a good or large pulmonary blood flow present difficult problems in management. Theoretically it would be possible to do complete correction in some as there appear to be vessels behaving as pulmonary arteries which could be connected to the right ventricle (Case 39). However, the symptoms in this group are mild, the resting arterial oxygen saturation varies from 82–87 per cent, and the haemoglobin is only slightly raised so radical surgery is not at present recommended.

Discussion

The terminology referring to pulmonary atresia has not been uniform nor has it contributed to the understanding of the condition. It is accepted that, when pulmonary atresia is associated with a large ventricular septal defect as in 47 of the 48 patients, and is without transposition, it may resemble Fallot's tetralogy both in development and physiology. Unfortunately, the use of the term 'Fallot variant' for this combination of lesions does not draw attention either to the varying development of the pulmonary arteries distal to the atresia, or to the important large collateral arteries which may arise from the descending aorta to perfuse the lungs. These large strange

arteries present in 25 per cent of patients with pulmonary atresia without transposition are not seen in Fallot and differ from the commoner small, spidery collaterals often found in both extreme Fallot as well as in pulmonary atresia, and which also develop after occlusion of a main pulmonary artery branch.

Despite the embryological resemblance of pulmonary atresia to common truncus, the term 'pseudotruncus' for pulmonary atresia is inappropriate and should be abandoned. First, because the term 'pseudo' means 'false' (*The Shorter Oxford English Dictionary*) and the condition of pulmonary atresia is as much an entity as coarctation or aortic stenosis. Furthermore, a term like 'pseudotruncus' does not highlight the basic abnormality which is complete obstruction between the right ventricle and the pulmonary arteries. In those patients with no central pulmonary artery development one might be tempted to use 'pseudotruncus', but this author like others (Van Praagh and Van Praagh, 1965; Miller *et al.*, 1968) considers that it should be classified as a variant of pulmonary atresia and not of truncus. This variation is termed Type 4 pulmonary atresia in the present classification. Collett and Edwards' (1949) description of this anomaly as Type 4 truncus is also considered to be misleading and should be discontinued, for in all patients in this series, whether they had large arteries arising from the descending aorta or spidery collaterals filling only hypoplastic peripheral lung arteries, the atretic outflow tract of the right ventricle was clearly seen (Fig. 1). Whether the condition of Type 4 truncus without a blind right ventricular outflow tract really exists is in doubt. The temptation to accept this condition after inadequate angiocardiography must be resisted.

Since the therapy for pulmonary atresia depends mainly on the degree of pulmonary artery development distal to the atresia, it is logical to classify pulmonary atresia according to this simple principle. The presence of other lesions does not influence the classification though it may influence the surgical approach.

Many of the problems of classifying and managing pulmonary atresia arise because of inadequate investigation. For instance, an injection of contrast medium into the body of the right ventricle may pass quickly through the large ventricular septal defect and the atretic outflow may not be opacified, thus encouraging the erroneous diagnosis of common trunk. The angiocardiographic cover time may not be long enough, or adequate volumes of contrast medium may not have been injected into the aorta to opacify the pulmonary arteries. Five patients in this series had been declared as inoperable after the usual routine right ventricular angiocardiography, including cineangiography in 3, had failed to reveal pulmonary arteries and one had had an aortogram covering only 2 seconds. All were subsequently shown to have pulmonary arteries suitable for a shunt procedure, and 2 have had successful radical correction. Thus, an aggressive approach to the investigation of pulmonary atresia is justified before the condition is deemed inoperable. It is not suggested that extensive investigation is carried out in sick infants with pulmonary atresia where the problem is to get them to survive to an age where radical correction is possible.

Palliative operations are required in many patients with pulmonary atresia before the ideal age is reached for radical corrective surgery. Spontaneous closure of a duct with cessation of a continuous murmur in an infant with pulmonary atresia gives rise to extreme hypoxia, and is probably the cause of so-called cyanotic attacks in these patients; an emergency shunt procedure is required, as in 5 of the present group. Miller and his colleagues (1968) recommend a Waterston anastomosis (ascending aorta to right pulmonary artery) as the shunt of choice in the infant. This would be our operation of choice in a child over 6 months as it is easy to close at the time of radical surgery. It has a disadvantage in the younger infant or neonate, in that it is easy to make the stoma too large, causing death from pulmonary oedema, and if it kinks resulting in sole perfusion of the right lung there will be a lack of development of the rest of the pulmonary arteries, as occurred in Case 26. This creates difficulties for further palliative or radical corrective surgery. The presence of unsuspected anomalous pulmonary veins in 3 patients made the siting of the Waterston anastomosis difficult as it had to be made anterior to the superior vena cava, which predisposed to anterior kinking of the pulmonary artery (Somerville *et al.*, 1969).

Radical corrective procedures for pulmonary atresia include closure of the ventricular septal defect with opening of the atretic segment or bypassing the atretic area with a tubular graft (Rastelli *et al.*, 1965), or reconstruction of the outflow tract with a homograft aortic valve. Recently, Ionescu and Ross (1969) have reconstructed the outflow tract in Fallot and pulmonary atresia using autologous fascia lata. Construction of a pulmonary valve is desirable in order to prevent serious pulmonary regurgitation which overloads the already hypertrophied right ventricle and causes chronic right ventricular failure.

The difficult problem of the patient with pulmonary atresia and an increased pulmonary blood flow which may lead to congestive failure in the early weeks of life can be managed by antifailure therapy (Miller *et al.*, 1968), or may be helped by ligating or creating a stenosis on the large collateral arteries arising from the descending aorta. Care must be taken not to ligate the large duct keeping some patients with pulmonary atresia alive. At a later age these patients present a problem in management as it would be possible technically to achieve complete correction, though most are relatively well. The danger of leaving this state is that irreversible pulmonary vascular disease may develop and the risks of cerebral complications with an open ventricular septal defect remain. However, until the long-term fate of these different grafts is known, it is preferred not to recommend radical surgery in this type of patient who is reasonably well.

Other congenital lesions inside and outside the cardiovascular system complicating pulmonary atresia have probably been underestimated in this series as it does not include infants who may have died from complex problems. Persistent duct probably complicates pulmonary atresia more commonly than 20 per cent estimated in this series not made up of infants. The duct may close in the first year before we see them.

From this study, it has been concluded that no patient with pulmonary atresia should be labelled as inoperable without extensive investigation, and if that fails to reveal pulmonary arteries, a search at bilateral thoracotomy should be made. This attitude frequently has been rewarded by the finding of a pulmonary artery suitable for a shunt procedure and in some of these 'hopeless' cases, radical correction has ultimately been possible.

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