Congenital "Absence" and Anomalous Origin of the Main Pulmonary Arteries

Variations of the Same Congenital Defect

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Anomalous origin of a pulmonary artery and "absence" of a pulmonary artery are both rare congenital malformations which, until the advent of angiocardiology and cardiac surgery, had rarely been diagnosed. During the past 15 years both conditions have been described with increasing frequency (Pool, Vogel, and Blount, 1962).

In presenting one patient with anomalous origin of the right pulmonary artery from the ascending aorta, and three with "absence" of the left pulmonary artery complicating tetralogy of Fallot, who were seen at the Cardiac Clinic, Groote Schuur Hospital, during the past 15 years, it is our object to draw attention to these defects and to discuss the embryological aspects, clinical presentation, and management of such patients.

ANOMALOUS ORIGIN OF RIGHT PULMONARY ARTERY FROM ASCENDING AORTA

Patient 1. L.P., a Cape Coloured girl, was admitted to the Cardiac Unit for investigation at the age of 6 months. Since the age of 1 week she had suffered from repeated attacks of right-sided pneumonia and at the age of 6 weeks had first developed congestive cardiac failure. Despite almost continuous treatment thereafter, she failed to thrive and her condition remained precarious.

Examination revealed an ill child with broncho-pneumonia, variable cyanosis, and congestive cardiac failure. A systolic murmur was heard along the left sternal border, and a mid-diastolic murmur at the mitral area. The second heart sound was loud and normally split. A chest radiograph (Fig. 1) showed an enlarged heart with pulmonary plethora—more marked on the right side than on the left, and broncho-pneumonic

consolidation in the right upper lobe. The electrocardiogram indicated right atrial and right ventricular hypertrophy.

She was thought to have a large left-to-right shunt through a ventricular septal defect, and cardiac catheterization was done to confirm the diagnosis with a view to operation. A catheter introduced into the right saphenous vein was passed to the main pulmonary artery; it could then be repeatedly passed into the left pulmonary artery and through a ductus arteriosus into the descending aorta; the right pulmonary artery could not be entered.

The findings at cardiac catheterization are shown in Fig. 2. Equal systolic pressures were recorded in both ventricles, the main pulmonary artery, and throughout the length of the aorta. Blood sampling revealed arterial oxygen desaturation and bidirectional shunting via the persistent ductus arteriosus.

Selective main pulmonary artery cine-angiocardiology (Fig. 3) showed filling of the left pulmonary artery and aorta with a reflux back to the aortic valve. The right pulmonary artery did not opacify. When contrast medium was injected into the left ventricle (Fig. 4) no ventricular septal defect was seen, but the right pulmonary artery filled synchronously with the ascending aorta. A later phase of this angiocardiology demonstrated right-to-left shunting of blood through the ductus arteriosus, a negative shadow impinging on the contrast medium as it passed the ductus arteriosus.

Published descriptions of only two similar cases of anomalous origin of the right pulmonary artery from the aorta (Caro, Lermenda, and Lyons, 1957; Armer, Shumacker, and Klatte, 1961) submitted to operation could be found. In both patients the ductus arteriosus was divided and the right pulmonary artery was disconnected from the aorta and joined to the main pulmonary artery by means of a graft.

After intensive hospital care our patient was submitted to operation at the age of 7½ months and the right pulmonary artery was successfully anastomosed to the pulmonary trunk, using cardiopulmonary bypass and

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FIG. 1.—Cardiomegaly and right-sided pulmonary plethora (Patient 1).

FIG. 2.—Diagrams showing the findings at cardiac catheterization (Patient 1).

hypothermia; the total duration of the bypass was 260 minutes. After operation she did well for 36 hours, then unfortunately developed acute respiratory distress and died.

Necropsy. On macroscopical examination of the heart and lungs both ventricles were found to be hypertrophied. No intracardiac defects were found. The aorta and main pulmonary artery were normally situated, the closure of the aorta was intact, as was the anastomosis of the right pulmonary artery to the main pulmonary artery. Both ends of the divided ductus arteriosus were intact.

Histology. All the arteries and arterioles of the right lung, except the main branches, showed a much thickened media in which elastic laminae were abundant. In the left lung minimal medial hypertrophy was present in the pulmonary arteries. All the pulmonary veins were conspicuously dilated. Patchy pneumonic consolidation and lobular atelectasis were found in both lungs. A mononuclear exudate with small patches of hyaline material was present in the alveoli. The pneumonic consolidation may have been due to a viral infection, or to the result of prolonged cardiac bypass in a young infant (Dodrill, 1958). Apart from congestion all the other organs were normal.

FIG. 3.—Selective main pulmonary artery angiogram (postero-anterior view), showing filling of the left pulmonary artery and the aorta via a persistent ductus arteriosus. There is no filling of the right pulmonary artery (Patient 1).

FIG. 4.—Selective left ventricular angiogram (left anterior oblique view) showing filling of the right pulmonary artery from the ascending aorta. The right-to-left shunting via the ductus arteriosus is shown as a negative shadow impinging on the contrast medium in the aorta (Patient 1).

"ABSENT" LEFT PULMONARY ARTERY

Three cases of tetralogy of Fallot in which "absence" of a left pulmonary artery was found have been seen in the Cardiac Clinic since 1951.

Patient 2. J.G. A severely cyanosed and disabled Coloured boy of 10 years was investigated in 1951 and found to have Fallot's tetralogy, a right-sided aorta, and "absence" of the left pulmonary artery on angiography. He underwent a closed pulmonary valvotomy, but died during the operation. Unfortunately, at

necropsy the lungs were not specifically examined for vascular changes, anomalous connexion of the left pulmonary artery, or collateral supply to the left lung.

Patient 3. C. LeG., a White boy, was first seen at the age of 4 in 1957 because of dyspnoea on exertion. He was found to have grade 1 cyanosis and a systolic murmur maximal at the 4th left intercostal space on examination. Cardiac catheterization confirmed the clinical diagnosis of Fallot's tetralogy, but the "absent" left pulmonary artery was not detected until he was

operated upon in 1961. The aorta was normal in position.

At operation the ventricular septal defect was closed with an ivalon patch and the pulmonary infundibular stenosis corrected by means of a teflon roof. No attempt was made to connect the left lung to the pulmonary artery.

After operation this boy did extremely well and now has only mild pulmonary incompetence. Angiocardiography in 1962 showed a normal pulmonary outflow tract, no ventricular septal defect, and "absence" of the left main pulmonary artery.

Patient 4. I.S., a White woman of 20, was first referred to the Cardiac Clinic with cyanosis and diminished effort tolerance in 1962. At cardiac catheterization the diagnosis of Fallot's tetralogy was confirmed, and "absence" of the left pulmonary artery was demonstrated by angiocardiography (Fig. 5). The aorta was normal in position.

FIG. 5.—Selective right ventricular angiocardiogram (postero-anterior view) showing infundibular and pulmonary valvar stenosis, a large right pulmonary artery, and "absence" of the left pulmonary artery (Patient 4).

She underwent operation for correction of her defects in 1962. The ventricular septal defect was closed with an ivalon patch, and the unfundibulum and conspicuously narrowed pulmonary valve ring were enlarged by means of a teflon gusset extending into the main pulmonary artery. No attempt was made to make a connexion between the main pulmonary artery and the left lung. After operation she did poorly, with persistent hypertension (75/7 mm. Hg) and severe pulmonary incompetence leading to tricuspid incompetence, intractable right heart failure, and death after 2 years. The septal defect had been completely closed.

DISCUSSION

Most of those who have written about absence or anomalous origin of a pulmonary artery treat these defects as unrelated entities. Furthermore, amongst the few patients with anomalous origin of a pulmonary artery are included those in whom the anomalous vessel arose from the arch of the aorta and not from the ascending aorta (Caro *et al.*, 1957; Findlay and Maier, 1951) (Fig. 6). Careful necropsy examination of patients with "absence" of a pulmonary artery has revealed an obliterated vessel connecting the innominate artery with the proximal end of a normally developed intrapulmonary artery in the disconnected lung (McKim and Wigglesworth, 1954; Andersen, Char, and Adams, 1958). It is apparent, therefore, that anomalous origin of a pulmonary artery from the arch of the aorta and so-called absence of a pulmonary artery are variants of the same embryological fault (Pool *et al.*, 1962; Andersen *et al.*, 1958). Recently Cucci, Doyle, and Lewis (1964) have proposed an ontogenetic theory by which all these pulmonary artery defects may be explained on the basis of abnormal septation of the truncus arteriosus.

Normally the trunco-conal ridges develop diametrically opposite to one another (Fig. 7A). They grow into the lumen of the truncus arteriosus and conus to fuse with each other. If the trunco-conal ridges do not arise in their normal diametrically opposed positions, the result will be unequal partitioning of the truncus and conus. Abnormal positioning of one or other of the trunco-conal ridges may occur at the cephalic end, the conal end, or uniformly throughout the truncus and conus (Cucci *et al.*, 1964). The grosser defects involving the base of the conus range from persistent truncus arteriosus to Fallot's tetralogy.

If defective siting of the trunco-conal ridges involves only the cephalic end of the truncus and the ridges are normally sited in the conus, the trunco-conal septum and the muscular ventricular septum will be correctly aligned and will fuse so that no cardiac defect will occur, and the two outflow tracts will be equal and normal. In anomalous origin of the right pulmonary artery it is suggested that the right trunco-conal ridge arises more dorsally than normal (Fig. 7B), and so becomes interposed between the origins of the pulmonary arteries. The right pulmonary artery then arises from the aorta and there is almost equal division of the truncus arteriosus and conus (Cucci *et al.*, 1964).

On the other hand, dislocation of the left trunco-conal ridge to a position dorsal to the origin of the left pulmonary artery (Fig. 7C) will cause unequal

FIG. 6.—Drawings illustrating the two sites of origin of anomalous right pulmonary artery. AO—aorta, MPA—main pulmonary artery, IA—innominate artery, RPA and LPA—right and left pulmonary arteries, respectively.

division of the truncus. A small pulmonary trunk then results and the left pulmonary artery will be connected to the ascending aorta. Such unequal division of the truncus arteriosus is unlikely to occur without dislocation of the trunco-conal ridge in the conus as well, and the only recorded example of anomalous origin of the left pulmonary artery from the ascending aorta occurred in a still-born child who had a right-sided aorta overriding a ventricular septal defect (Šikl, 1952).

When discussing the malformations, “absent pulmonary artery” and “anomalous origin of a pulmonary artery from the arch of the aorta”, it is necessary to review the embryological development of the pulmonary arteries.

Three anlagen are involved in the formation of the pulmonary arteries (Cucci *et al.*, 1964): (i) the pulmonary trunk derived from the truncus arteriosus as a result of its septation; (ii) the proximal segments of the paired 6th aortic arches; and (iii) the intrapulmonary arteries, which develop from the primitive vessels in the lung buds.

In Fig. 8 an embryological model of the paired 4th and 6th aortic arches is shown. Each 6th arch is composed of a proximal segment arising from the truncus arteriosus and a distal segment arising from the dorsal aorta on its respective side. These segments fuse together to complete the arches, and the intrapulmonary arteries connect with the arches proximally.

Interruption of the right dorsal aorta between the

subclavian artery and the unpaired aorta, and the distal segment of the right 6th arch between its connexion with the intrapulmonary artery and the right dorsal aorta occurs normally between the 31st and the 36th day of foetal life. At the same time, the proximal segment of the left 6th arch becomes incorporated into the main pulmonary artery (Fig. 9A), resulting in the distal segment of the left 6th arch—the ductus arteriosus—becoming aligned with the pulmonary trunk (Andersen *et al.*, 1958). Thus, a shorter and more direct route between the pulmonary trunk and the left dorsal aorta is established, and as the left pulmonary arch carries the greater blood flow, it becomes dominant in the same way as the left dorsal aorta. The involution that takes place on the right side is, therefore, the result of the diminished flow that occurs on the right side.

In cases of so-called “absence” of a pulmonary artery (Fig. 9B), the defect is the result of involution of the proximal segment of the affected 6th arch with persistence of the distal segment during foetal life. In the majority of cases the distal segment, being a ductal structure, involutes at birth, leaving the intrapulmonary artery disconnected from both the pulmonary and systemic circulations and the affected lung becomes dependent on the development of a collateral circulation from the bronchial arteries. In those few cases where the distal segment remains patent, anomalous origin of the pulmonary artery from the arch of the aorta results.

The reason for persistence of the distal segment

FIG. 7.—Cephalic view of the truncus arteriosus at the level of the sixth paired arch—with its septum, formed by the out-growth and coalescence of the right (R) and the left (L) trunco-conal ridges. (A) The septum in the normal position. (B) The right ridge emerges from a more dorsal position and the right sixth arch is thus incorporated into the ascending aorta. (C) The dorso-rotation of the left ridge causes the left sixth arch to remain connected with the aorta. (Reproduced from Cucci *et al.* (1964), *Circulation*, 29, 127, with the permission of the author and publisher.)

and involution of the proximal segment of a pulmonary (6th) arch is considered to be due to greater flow occurring through the distal segment in such cases. Cucci *et al.* (1964) have proposed that this situation probably occurs only when both ends of the affected pulmonary arch are connected to the systemic circulation, and they consider that the initial defect is identical with that of anomalous origin of a pulmonary artery from the ascending aorta. Thus, whether the eventual defect will be anomalous origin of a pulmonary artery from the ascending aorta, or interruption of the proximal segment of the pulmonary arch, depends upon whether flow is greater through the proximal or the distal segment of a pulmonary arch that has remained connected proximally to the aorta as a result of incorrect septation of the truncus arteriosus. In support of this theory is the fact that

FIG. 8.—Model of the fourth and sixth aortic arches in the human embryo. (1) Proximal segment of the right sixth arch. (2) Proximal segment of the left sixth arch. (3) Distal segment of the right sixth arch. (4) Distal segment of the left sixth arch. (5) Right intrapulmonary artery. (6) Left intrapulmonary artery. T.A.=truncus arteriosus; R.D.A. and L.D.A.=right and left dorsal aorta.

anomalous origin of the right pulmonary artery and "absence" of the right pulmonary artery are not associated with defects of the conus, whereas "absence" of the left pulmonary artery is commonly associated with defects of the conus, e.g. tetralogy of Fallot. Cases of isolated "absence" of the left pulmonary artery have, however, been reported (Ferencz, 1961).

Clinical Manifestations and Management. Isolated interruption of a pulmonary artery is compatible with life. The majority of patients so far described have been adults, the oldest being 63 years of age at the time of the report (Pool *et al.*, 1962). Since several were asymptomatic or suffered only slight diminution in effort tolerance, it is possible that many cases go undiscovered. Haemoptysis occurs in about 10 per cent of patients surviving infancy and is probably due to the increased flow via the bronchial collateral vessels (Pool *et al.*,

FIG. 9.—Models demonstrating (A) the normal development of a left aortic arch and the pulmonary arteries; (B) Involution of the proximal segment of the right sixth arch with persistence of the distal segment. The symbols used are the same as in Fig. 8.

1962), atherosclerotic branches of which may rupture (Findlay and Maier, 1951).

Pulmonary hypertension occurs in 19 per cent of cases and is usually associated with a defect allowing a left-to-right shunt. Persistence of the ductus arteriosus is the commonest associated defect and in these cases pulmonary hypertension is invariable, causing severe symptoms in infancy and early death from right heart failure if the associated defect is not corrected (Pool *et al.*, 1962).

The lung lacking a connexion with the main pulmonary artery takes part almost normally in ventilation. Oxygen uptake from such a lung, however, is rarely more than 6 per cent of the total, since it receives a supply of fully saturated blood either through the persistent distal segment of the pulmonary arch or via the bronchial collateral vessels (Pool *et al.*, 1962; Ferencz, 1961; Oakley, Glick, and McCredie, 1963). The lives of such patients are therefore imperilled if their one correctly connected lung becomes infected or injured.

Clinical examination is unrewarding in patients with isolated interruption of a pulmonary artery. In a few instances a systolic murmur to the left of the sternum has been heard and some diminution of the affected hemithorax has been noted. The diagnosis, however, becomes apparent on radiological investigation (Fig. 10). Schneiderman (1958) listed the radiological features as follows.

(i) Increased translucency of the affected lung with diminished vascular markings.

(ii) Diminished size of the affected hemithorax, as evidenced by narrowed intercostal spaces.

(iii) Mediastinal displacement towards the affected side.

(iv) Absence of the "hilar comma" shadow of the descending main branches of the pulmonary artery on the affected side (Danielius, 1942).

In Fallot's tetralogy these signs should be particularly looked for in the presence of a right-sided aorta. Emanuel and Pattinson (1956) drew attention to the fact that in 60 per cent of patients where "absence" of a left pulmonary artery complicated Fallot's tetralogy a right-sided aortic arch was found. It was absent in 2 of our 3 patients.

Where tetralogy of Fallot is complicated by having one lung disconnected, the prognosis has been extremely poor. Pool *et al.* (1962), reviewing 8 patients with tetralogy of Fallot and proximal interruption of the left pulmonary artery who underwent "total correction", found that only one had survived operation. They suggested that this was because of a greatly increased pulmonary resistance as a result of a reduced pulmonary vascular bed. The ability of the vascular bed in the one connected lung to cope with the total pulmonary flow after operation must depend on the degree of pulmonary stenosis present before operation, as this determines

FIG. 10.—The left hemithorax is smaller than the right and the vascular markings are diminished in the left lung (Patient 4).

whether flow is normal, almost normal, or reduced. In isolated cases of "absence" of a pulmonary artery, the unaffected lung has been able to cope with the entire cardiac output (Ferencz, 1961). In our experience the pulmonary vascular resistance was raised to 27 per cent of systemic resistance in our fourth patient after operation. Severe pulmonary valvar and infundibular stenosis was present before operation, and she died after the operation. Our third patient had only moderate infundibular stenosis before surgery with a normal pulmonary vascular resistance, and he has done very well since operation.

Venous or pulmonary angiography will demonstrate the "absence" of a pulmonary artery. It is important, however, to search and demonstrate if possible either a persistent distal arch connexion, or a bronchial collateral supply to the affected lung by aortic angiography. If the former is present, it is confirmatory evidence of congenital interruption of the affected pulmonary arch. Persistent distal arch connexion does not occur in severe stenosis or in thrombo-embolic obstruction of the pulmonary artery. Pulmonary angiography alone may be misleading. We have misdiagnosed "absence" of the left pulmonary artery on angiocardiographic grounds in two patients. At operation, or repeat selective angiocardiography, a normal left pulmonary artery was found. The pitfalls in angiocardiographic diagnosis of pulmonary artery

obstruction have recently been discussed by Massumi, Rios, and Donohoe (1965).

Where proximal interruption of a pulmonary artery is discovered, the surgeon should be prepared to connect the normal but disconnected intrapulmonary artery to the main pulmonary artery (Andersen *et al.*, 1958) by means of a graft. Lillehei (1965) has effected such a connexion in one patient. In those patients where the distal segment has remained patent, it may be possible to disconnect this from the innominate artery and connect it to the main pulmonary artery.

Anomalous origin of the pulmonary artery from the aorta should be suspected when a small infant presents with signs of a large left-to-right shunt, variable cyanosis associated with frequent attacks of pneumonia, and a radiographic appearance of cardiomegaly and unilateral pulmonary plethora (Fig. 1). Pulmonary angiography will demonstrate absence of a pulmonary artery, which will be shown to arise from the aorta on selective left ventricular or aortic angiography.

Of the 13 published cases all but 2 died in infancy. Progressive and intractable pulmonary hypertension develop and increasingly severe changes in the small pulmonary arteries have been found with advancing age (Griffiths, Levine, and Andersen, 1962). All patients have died following division of the persistent ductus arteriosus, which is an integral part of this anomaly (Armer *et al.*, 1961; DuShane

et al., 1960). It is, therefore, imperative to recognize this lethal condition because it may be corrected by operation at an early age.

SUMMARY

One patient with anomalous origin of the right pulmonary artery from the aorta and 3 with "absent" left pulmonary artery complicating tetralogy of Fallot have been presented.

The embryological development of the pulmonary arteries and the current concepts of the disturbance in the septation of the truncus arteriosus, which probably gives rise to such defects, have been reviewed.

It is considered that anomalous origin and "absence" of a pulmonary artery are different manifestations of defective septation of the truncus arteriosus resulting in the affected pulmonary (6th) arch remaining connected to the aorta proximally. The involutionary changes occurring in this arch determine the final defect: if the proximal segment remains patent and the distal segment involutes, anomalous origin from the ascending aorta results; if the distal segment remains patent and the proximal segment involutes, anomalous origin from the arch of the aorta results. If both the distal and proximal segments involute, the affected pulmonary artery is "absent" or, correctly speaking, disconnected.

The importance of early recognition and surgical correction of anomalous origin of a pulmonary artery is emphasized. Where there is proximal interruption of a pulmonary arch it should be possible to connect the intrapulmonary artery to the main pulmonary artery. By doing so the danger of hæmoptysis from the affected lung will be avoided. The mortality in patients with tetralogy of Fallot complicated by this defect should be reduced and the patient will benefit if both lungs participate in oxygen uptake.

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