Anomalous Origin of One Pulmonary Artery from the Ascending Aorta

Diagnostic, Physiological and Surgical Considerations

By John F. Keane, M.D., David Maltz, M.D., William F. Bernhard, M.D., Robert D. Corwin, M.D., and Alexander S. Nadas, M.D.

SUMMARY

Six patients with origin of one pulmonary artery from the ascending aorta are presented. Two are alive: a 21-year-old female, unoperated, with bilateral pulmonary vascular obstructive disease (PVO) and a 4-year-old girl who underwent successful correction at age 5 months. In addition, the data are reviewed from previous reports of 44 patients with this anomaly. Among the total of 50 patients, 42 had congestive heart failure in infancy. The right pulmonary artery arose anomalously in 44. A patent ductus arteriosus was present in 38 cases. At cardiac catheterization, pressure in both pulmonary arteries was generally at systemic level and the diagnosis was confirmed by aortography. Histologically, no significant PVO was observed in the majority of 18 infants who died, while significant changes, mainly in the "systemic" lung, were observed in 6 of 10 older children.

Twenty-two patients have been operated on with 13 survivors. Anastomosis of the aberrant pulmonary artery to the main pulmonary artery directly or using a graft has been successful in 12 of 18 children. Corrective surgery should be performed as early as possible to prevent PVO.

Additional Indexing Words:
Hemitruncus arteriosus
Surgical correction of anomalous pulmonary artery

The ANOMALY consisting of one pulmonary artery arising from the ascending aorta while the other originates normally from the right ventricle is a rare congenital cardiac malformation. To date, there have been 44 reported cases in the literature. In the past, most patients have died by three months of age from congestive heart failure but in recent years, due to early recognition and improved surgical techniques, successful results have been reported in 12 patients.1-12

It is the purpose of this presentation to report 6 additional patients with this anomaly and to review the 44 other cases from the literature. In addition, diagnostic features will be reviewed, together with our attempt to evaluate flow and resistance measurements in both lungs, and surgical techniques will be commented on.

Material and Methods

Between 1949 and 1973, six patients with one pulmonary artery originating from the ascending aorta while the other originated normally have been seen among approximately 12,000 patients with congenital heart disease at the Children's Hospital Medical Center, Boston and their clinical course has been reviewed.

Results

Four patients died in early infancy and pertinent data concerning them are presented in table 1. Since none of them underwent cardiac catheterization, the diagnoses were made at autopsy. The remaining two patients are alive and their case histories are now presented in more detail.

Case 5

This female was born in 1952, the product of a 36 week gestation in a gravida 6, para 6 (4 living, one premature death at 24 hours, one stillbirth), 26-year-old mother who had been taking stillbestrol during the last trimester. The child was admitted to the Children's Hospital Medical Center at age 3 months
## Table 1

### Data of Four Patients Who Died

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Clinical features</th>
<th>ECG</th>
<th>Heart size</th>
<th>Pulmonary blood flow</th>
<th>Age at death</th>
<th>Anomalous pulmonary artery</th>
<th>Associated anomalies</th>
<th>Pulmonary vascular changes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>Male</td>
<td>Bronchiolitis at 9 weeks; grade III/VI systolic murmur; CHF and cyanosis at 12 weeks</td>
<td>BVH</td>
<td>CE</td>
<td>Increased bilaterally</td>
<td>3 months</td>
<td>Right</td>
<td>Patent ductus arteriosus; Bicuspid aortic valve</td>
<td>Dilatation, tortuosity &amp; thickening of media of pulmonary arteries; cellular &amp; acellular intimal &amp; fibrous proliferation of pulmonary arteries &amp; arterioles (grade 3 changes)</td>
</tr>
<tr>
<td>Case 2</td>
<td>Female</td>
<td>Cyanotic episode at 5 days; cyanosis and grade II systolic murmur at 10 weeks</td>
<td>RVH</td>
<td>Normal</td>
<td>Decreased</td>
<td>4 months</td>
<td>Left</td>
<td>Tetralogy of Fallot; Bicuspid pulmonary valve; Right aortic arch; Bilobed right lung</td>
<td>Normal</td>
</tr>
<tr>
<td>Case 3</td>
<td>Male</td>
<td>Cyanosis at 1 day; CHF at 2 days; no murmurs</td>
<td>RVH</td>
<td>CE</td>
<td>Increased bilaterally</td>
<td>3 days</td>
<td>Right</td>
<td>Patent ductus arteriosus; Preductal coarctation; Ventricular septal defect</td>
<td>Normal</td>
</tr>
<tr>
<td>Case 4</td>
<td>Female</td>
<td>CHF at 1 day</td>
<td>BVH</td>
<td>CE</td>
<td>Increased bilaterally</td>
<td>3 months</td>
<td>Right</td>
<td>None</td>
<td>Arterioles show uniform persistence of fetal pattern more transitional in right than left lung -- normal for age</td>
</tr>
</tbody>
</table>

Abbreviations: CHF = Congestive heart failure; RVH = Right ventricular hypertrophy; ECG = Electrocardiogram; CE = Cardiac enlargement; BVH = Biventricular hypertrophy; PAT = Supraventricular tachycardia.
with progressive congestive heart failure without cyanosis. A grade III/VI systolic murmur was audible at the left sternal border. The electrocardiogram revealed biventricular hypertrophy and the chest X-ray demonstrated cardiomegaly with bilaterally increased pulmonary vasculature. She responded satisfactorily to anticongestives and antibiotics. Mild, generalized cyanosis was first observed at 1 year, gradually increased and at age 6 years (1958) was accompanied by increasing exercise intolerance. At that time, a grade II/VI systolic murmur was described at the left upper sternal border, the electrocardiogram revealed right ventricular hypertrophy and a chest X-ray (fig. 1) demonstrated mild cardiomegaly with increased pulmonary vasculature in the right lung field only. At cardiac catheterization at that time (table 2), only the left pulmonary artery, in which pressure was at systemic level, could be entered from the right ventricle. A patent ductus arteriosus, shunting exclusively right to left was also identified.

Symptomatology remained unchanged through the next several years. At age 14 (1966), a lung scan (fig. 2) following intravenous injection of iodine211 labelled macroaggregates of albumin, demonstrated uptake in the left lung but almost none in the right lung. At age 18 years (1970), minor episodes of hemoptysis appeared and as the diagnosis was uncertain, cardiac catheterization was repeated (table 2). It was shown that pressure in both pulmonary arteries was at systemic level, exclusive right to left shunting through the ductus continued and the right pulmonary artery was seen to arise from the proximal ascending aorta (fig. 3). The situation was felt inoperable and currently, at age 21 years, the patient’s clinical status remains unchanged and she is attending college.

![Figure 1](image-url)

**Figure 1**

Chest X-ray of case 5 at age 6 years, showing mild cardiac enlargement with increased pulmonary blood flow in the right lung only.

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Catheterization Data, Case 5 L.S.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Age 6 Yrs. (1958)</td>
</tr>
<tr>
<td>-----------------------------------</td>
<td>-----------------</td>
</tr>
<tr>
<td><strong>Pressure</strong> <strong>(mm Hg)</strong></td>
<td>RA</td>
</tr>
<tr>
<td></td>
<td>MPA</td>
</tr>
<tr>
<td></td>
<td>LPA</td>
</tr>
<tr>
<td></td>
<td>RPA</td>
</tr>
<tr>
<td></td>
<td>D.AO</td>
</tr>
<tr>
<td><strong>Oxygen saturation (%)</strong></td>
<td>RA</td>
</tr>
<tr>
<td></td>
<td>MPA</td>
</tr>
<tr>
<td></td>
<td>LPA</td>
</tr>
<tr>
<td></td>
<td>RPA</td>
</tr>
<tr>
<td></td>
<td>D.AO</td>
</tr>
<tr>
<td><strong>Flow (L/min/m²)</strong></td>
<td>Systemic</td>
</tr>
<tr>
<td></td>
<td>Left lung</td>
</tr>
<tr>
<td></td>
<td>Right lung</td>
</tr>
<tr>
<td><strong>Resistance (units/m²)</strong></td>
<td>Systemic</td>
</tr>
<tr>
<td></td>
<td>Left lung</td>
</tr>
<tr>
<td></td>
<td>Right lung</td>
</tr>
</tbody>
</table>

Abbreviations: RA = Right atrium; MPA = Main pulmonary artery; LPA = Left pulmonary artery; RPA = Right pulmonary artery; D.AO = Descending aorta.

Mean pressure values in parenthesis.
ANOMALOUS ORIGIN OF PA FROM AORTA

Case 6

This female patient was first seen at age 4 months (April 1969) at the Rhode Island Hospital in congestive heart failure. A grade II/VI systolic murmur was audible together with an apical diastolic flow rumble of similar intensity. The electrocardiogram revealed right atrial and biventricular hypertrophy and the chest X-ray (fig. 4) showed cardiac enlargement with increased pulmonary vasculature bilaterally. At cardiac catheterization (table 3), the pressure in the normally arising left pulmonary artery was found to be almost at systemic level. A patent ductus with a large amount of left to right shunting was identified and left ventricular angiography revealed the right pulmonary artery to arise from the posterior wall of the proximal ascending aorta (fig. 5). At surgery, at age 5 months (May 1969) at Children’s Hospital Medical Center, the right pulmonary artery was detached from the aorta and anastomosed to the main pulmonary artery using a segment of Teflon graft (10 mm diameter) placed anterior to the aorta. The patent ductus was divided. At cardiac catheterization two weeks postoperatively (table 3), the pulmonary artery pressure was normal and the prosthetic graft patent. The patient has grown normally and is asymptomatic. The electrocardiogram is normal, as is the chest X-ray. At a repeat catheterization at the Rhode Island Hospital 4 years postoperatively (table 3), the main pulmonary artery pressure was at the upper limits of normal and a peak systolic gradient of 7 mm Hg was present across the patent (fig. 6) graft.

Discussion

Clinical Profile

Forty-four patients with origin of one pulmonary artery from the ascending aorta while the other originated normally have been reported in the literature since Fraentzel first described the anomaly.
in 1968.12 The addition of our 6 cases now raises the total to 50. Of this group, 28 were female. Age at diagnosis ranged from two days to 25 years (median three months), the diagnosis being made at catheterization in 27 instances and at autopsy in the remaining 23. Congestive heart failure was present in 42 and cyanosis was evident in 28. Electrocardiographically, right ventricular hypertrophy was evident in 40, right atrial hypertrophy in 16, and additional left ventricular hypertrophy in 11. Left ventricular hypertrophy alone was rare, being observed in only two patients. On X-ray, cardiomegaly was evident in 42.

In terms of anatomy, the right pulmonary artery arose from the aorta in 44 patients. The commonest associated cardiac defect was a patent ductus arteriosus, this being present in 38 cases. A ventricular septal defect was present in four patients. Tetralogy of Fallot was present in six; among these the left pulmonary artery was found to arise anomalously in four patients (references 7, 14, 15, and our case 2) and the right in two.11 An embryological explanation for this anomaly has been proposed.16 The right and left pulmonary arteries arise from the proximal segments of the paired

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**Table 3**

*Catheterization Data, Case 6 C.B.*

<table>
<thead>
<tr>
<th></th>
<th>Age 4 Mon. (Pre-op)</th>
<th>Age 5 Mon. (Post-op)</th>
<th>Age 4 Yrs. (1973)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pressure</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(mm Hg)</td>
<td>RA</td>
<td>MPA</td>
<td>D.AO</td>
</tr>
<tr>
<td></td>
<td>(6.5)</td>
<td>83/35(50)</td>
<td>103/38(68)</td>
</tr>
<tr>
<td></td>
<td>(4.0)</td>
<td>30/10(20)</td>
<td>100/30(70)</td>
</tr>
<tr>
<td>Oxygen</td>
<td>RA</td>
<td>MPA</td>
<td>D.AO</td>
</tr>
<tr>
<td>saturation (%)</td>
<td>56</td>
<td>67</td>
<td>81</td>
</tr>
<tr>
<td></td>
<td>67</td>
<td>63</td>
<td>81</td>
</tr>
<tr>
<td></td>
<td>87</td>
<td>97</td>
<td>?</td>
</tr>
<tr>
<td>Flow</td>
<td>Systemic</td>
<td>Left lung</td>
<td>Right lung</td>
</tr>
<tr>
<td>(L/min/m²)</td>
<td>3.0</td>
<td>12.5</td>
<td>?</td>
</tr>
<tr>
<td></td>
<td>5.5</td>
<td>2.7</td>
<td>2.7 ≥ 5.4</td>
</tr>
<tr>
<td>Resistance</td>
<td>Systemic</td>
<td>Left lung</td>
<td>Right lung</td>
</tr>
<tr>
<td>(units/m²)</td>
<td>18.7</td>
<td>12.0</td>
<td>?</td>
</tr>
<tr>
<td></td>
<td>6.0</td>
<td>6.0</td>
<td>2.8 ≥ 1.4</td>
</tr>
</tbody>
</table>

Abbreviations: RA = Right atrium; MPA = Main pulmonary artery; LPA = Left pulmonary artery; D.AO = Descending aorta.

Mean pressure values in parenthesis.

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Figure 5

*Left ventricular preoperative cineangiogram at age 4 months, demonstrating origin of right pulmonary artery from ascending aorta.*

Figure 6

*Right ventricular postoperative cineangiogram showing patency of main to right pulmonary artery graft.*
sixth aortic arches. The conotruncal ridges, which arise diametrically opposite each other, fuse, thus partitioning the lumen of the truncus arteriosus. If these ridges do not arise directly opposite each other, unequal partitioning of the truncus results. Thus, should the right truncal ridge emerge from a more dorsal position than normal, the proximal right sixth aortic arch becomes incorporated into the ascending aorta and the right pulmonary artery then arises from the ascending aorta.

Diagnosis

Patients with origin of one pulmonary artery from the ascending aorta while the other originates normally usually present in early infancy when the normal postnatal drop in pulmonary vascular resistance occurs and the consequent aortic run-off through the "systemic" lung increases. The cardiac findings are nonspecific and suggest pulmonary artery hypertension with increased pulmonary blood flow and increased pulmonary vascularity in the "systemic" lung.1,17 A lung scan with venous injection of radionuclide18 reveals uptake only by the lung supplied by the normally arising pulmonary artery. The differential vascularity on X-ray, together with the lung scan findings, make the diagnosis likely, the only alternative being unilateral pulmonary atresia with large bronchial collateral or ductal flow. At cardiac catheterization, the diagnosis is established by aortography or left ventricular angiography in the absence of a ventricular septal defect which identifies the anomalously arising pulmonary artery. The pressure in this vessel is usually at systemic level. From the right ventricle only one pulmonary artery can be entered and pressure in this vessel is usually also significantly elevated. If a patent ductus is present, a small left to right shunt may be found in younger children whereas in older patients shunting is bidirectional or exclusively right to left.

Physiology

The lung supplied by the pulmonary artery originating from the right ventricle receives the total systemic venous return, which is the effective pulmonary flow. This can be measured using either the Fick or indicator dilution19 technique. The left ventricular output consists of the systemic venous return together with the pulmonary venous return of the "systemic" lung. The left ventricular output may be measured by angiographic volume analysis,20 as in our case 5 or by dye dilution technique.19 When the effective pulmonary flow is subtracted from this value, one is left with the pulmonary flow through the "systemic" lung. However, it should be mentioned that measurements using large film angiography are subject to error which may be as high as 30%. In our case 5, this would increase right lung flow to 1.8 L/min/m² and reduce resistance, this latter value still remaining, however, at systemic level (table 2).

Resistances may be calculated in the usual manner but the values obtained should be halved for comparison with resistances when both lungs are present (tables 2 and 3). The reason for this is that total pulmonary resistance represents the resistance of two lungs in parallel and the resistance across each must be twice the resistance if flow is equally distributed.8

Pulmonary Vascular Obstruction

Histological examination of the pulmonary vascular bed was described in 28 patients. No significant obstructive changes were observed in either lung among the majority of the 18 infants who died under six months of age.

Among the 10 older patients, no significant pulmonary vascular changes were noted in four, aged 8 months, 2 years, 2 years and 4 years. Of the remaining six patients, three, aged 7 months, 8 months and 23 years, had significant vascular obstruction more pronounced in the "systemic" lung.1,3,21 Two children,17 aged 6 months and 2 years, had similar changes in both lungs, these being more advanced in the older child. The remaining patient, aged 19 months, had more pronounced vascular obstructive changes in the normally supplied lung.22 The differential changes observed in some of these older children may be related to the fact that the "systemic" lung receives an increased volume of highly saturated blood at systemic pressure whereas the other lung is perfused with an increased amount of unsaturated blood often at a slightly lower pressure.

Surgical Considerations

Twenty-two patients have been operated upon with 13 survivors. By far the most successful procedure consists of anastomosing the aberrant pulmonary artery to the main pulmonary artery either directly or with a graft. This technique has been successful in 12 of 18 patients.

Bandling of the aberrant pulmonary artery was performed in one infant with a ventricular septal defect. No initial relief of cardiac failure was accomplished and immediate and successful re-anastomosis and ventricular septal defect closure was required.4 Other procedures, including ligation of a patent ductus only in two patients23,24 and ligation of both the ductus and anomalous pulmonary artery in another patient,15 were unsuccessful.

The major contraindication to surgery is the presence of severe pulmonary vascular obstructive disease in both lungs. This finding prevented operation
in our case 5. Survival following surgery in a 7-year-old patient with moderate vascular obstruction and bidirectional shunting across a patent ductus arteriosus has been reported, however. The remainder of those patients who survived surgery had insignificant pulmonary vascular obstructive disease. Exclusive right to left shunting across a patent ductus generally precludes surgery although repair in a 3-week-old infant with filling of the descending aorta through the duct has been recently reported. Corrective surgery should be performed as early as possible to prevent development of pulmonary vascular obstructive disease.

Acknowledgment

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References

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