Origin of the Right Pulmonary Artery from the Ascending Aorta

Report of a Surgically Corrected Case

By ROBERT M. ARMER, M.D., HARRIS B. SHUMACKER, M.D., AND EUGENE C. KLATTE, M.D.

The occurrence of a form of congenital heart disease in which there is an anomalous origin of the right pulmonary artery from the ascending aorta, with or without an associated patent ductus arteriosus, but no anomalous pulmonary venous drainage or intracardiac anomaly, is rare. Eight cases have been found on review. The purpose of this paper is to describe a surgically corrected case.

Review of Literature

Findlay and Maier, in 1951, reviewed the literature concerning anomalies of the pulmonary vessels and reported one case of their own that fits the category under discussion. She was a 4-month-old Negro girl with intermittent cyanosis, dyspnea, and fever. Her congestive heart failure improved initially with medical treatment but she soon died in severe failure and with extensive pneumonia. At autopsy there was a greatly enlarged heart but no intracardiac defect. Just distal to the aortic valve a large vessel originated from the left side of the aorta and coursed behind the esophagus to enter the right lung. There was no right pulmonary artery branch from the main pulmonary artery, and the ligamentum arteriosum was in the normal location. Right pneumonectomy and division of the anomalous pulmonary artery were proposed in retrospect as possible methods of treatment. Maier discussed the same patient in 1954 and suggested that an essentially normal distribution of the blood flow to both lungs might have resulted from a surgical transfer of the proximal end of the anomalous artery from the aorta to the side of the main pulmonary artery.

Sikl described the autopsy findings in a 4-month-old infant with this anomaly. There was an associated patent ductus arteriosus.

In May of 1960, DuShane et al. reported such a malformation in a 2-month-old infant. This baby entered the hospital in severe congestive heart failure that did not improve with medical management. Venous cardiac catheterization demonstrated a bidirectional shunt via a patent ductus arteriosus. The right pulmonary artery could not be entered with the catheter. Angiocardiography was not performed. The infant did reasonably well for a few hours following division and suture of the patent ductus arteriosus but then deteriorated rapidly and died the day after the operation. The true situation was not appreciated until the postmortem examination. Edwards, in discussing embryologic considerations, thought that the origin of the right pulmonary artery from the ascending aorta possibly represented an abnormality in the evolution of the sixth right aortic arch, that is, persistence of its distal portion rather than its proximal part. Wagenvoort et al. reported the autopsy study in another very young infant with this malformation.

Levine and Griffiths are preparing a paper describing three cases discovered at autopsy. None had significant patency of the ductus arteriosus. These children were 3½ and 6½ months, and 2 3/4 years of age. One is the previously mentioned case of Findlay and Maier.

Vlad and Lambert correctly diagnosed a case in an infant 4 months old by cardiac...
catheterization and angiocardiography. The ductus was closed and the right pulmonary artery was ligated, following which there was dramatic relief of congestive heart failure but unfortunately the baby died 4 weeks later of pneumonia. Another infant of theirs died at 2½ months of age and was found at postmortem study to have this anomaly with a large patent ductus arteriosus.

**Case Report**

**Clinical Findings**

The patient was a 10-month-old white boy. The complaints were poor weight gain, repeated respiratory infections, and mild cyanosis associated with crying.

He was the first child of a 20-year-old mother. The course of the pregnancy and delivery were unknown, maternal serology was negative, and the infant's birth weight was 5 pounds 8 ounces. He was said to have breathed and cried spontaneously at birth. A heart murmur, systolic in time, maximal in the pulmonic area, and associated with a snapping pulmonic second sound was first detected at 3 weeks of age. At 6 weeks of age it was first noted that the infant was mildly cyanotic at times of stress. Repeated respiratory infections were treated with intramuscular antibiotics. At no time was the infant considered to be in obvious congestive heart failure, but his growth and development were retarded.

When hospitalized he was 10 months old and weighed 14 pounds 4 ounces. The respiratory rate was 36 per minute and not labored. The skin color was grayish but not obviously cyanotic. The thumbnails were slightly clubbed. He was small, poorly developed, poorly nourished, and unable to sit without support. The lungs were clear. The heart rate was 136 per minute and regular. There was a forward bulge of the precordium and a left parasternal right ventricular lift. The left border of cardiac dullness was midway between the midclavicular line and the anterior axillary line. Only an insignificant, vibratory type, grade I (on the basis of 5) systolic murmur was heard maximal at the lower left sternal border. There were no thrills. The pulmonic second sound was single, obviously accentuated, and snapping in character. The liver margin was sharp and palpable two fingerbreadths below the right costal margin in the right midclavicular line. There was no edema. The femoral pulses were normal. Blood pressure in the right arm was 90/45 mm. Hg and 105/7 mm. Hg in the right leg.

Urine analysis was normal. The hemoglobin was 17.9 Gm. per cent, hematoctrit value 50 per cent, and total white blood cell count 7,800 per mm.³ with 26 per cent neutrophils and 74 per cent lymphocytes.

Radiographically (figs. 1 and 2) there was enlargement of both ventricles and the left atrium (fig. 1). A disproportionate increase of pulmonary vasculature in the right as compared with the left lung was not appreciated. There was incomplete right bundle-branch block and right ventricular hypertrophy on the electrocardiogram (fig. 3). The clinical impression was patent ductus arteriosus with moderate to severe pulmonary hypertension.

**Preoperative Physiologic Studies and Cineangiocardiography**

Under general sedation a venous cardiac catheterization by the saphenous route was carried out at 12 months of age (table 1). The catheter passed easily through a patent ductus arteriosus into the descending aorta. The left pulmonary
Figure 3
Top. Preoperative electrocardiogram January 4, 1960. There is incomplete right bundle-branch block and right ventricular hypertrophy. Bottom. Postoperative study June 1, 1960. There has been a return toward normal with a change to a rSr' QRS configuration in V1 and disappearance of the deep S wave in V5.

artery was entered repeatedly with the catheter, but the right pulmonary artery could not be located. This was not thought to be unusual at the time. The blood oxygen determinations were extremely erratic when the infant was breathing room air. Pressures in the pulmonary artery and the descending aorta were equal, 103/61 and 101/59 mm. Hg respectively. The sampling and pressures were then repeated with the patient breathing pure oxygen. Systemic pressure was unaltered, but there was a decrease in the pulmonary artery pressure creating a systolic pressure gradient across the ductus arteriosus of 20 mm. Hg (fig. 4). A left-to-right shunt with an increase of 3.1 volumes per cent oxygen content from the right ventricle to the pulmonary artery was found.

Figure 4
DA, descending aorta; PA, pulmonary artery; RV, right ventricle; RFA, right femoral artery. Top. Pressure tracings February 16, 1960 prior to surgery. The descending aorta was entered via the patent ductus arteriosus. During breathing of room air the systemic and pulmonary artery pressures were equal. Middle. After breathing of pure oxygen. The systemic pressure is unaltered but the pulmonary artery pressure has decreased creating a peak systolic pressure gradient across the patent ductus arteriosus of 20 mm. Hg. Bottom. Three months after surgery.

Fully saturated blood was obtained from the descending aorta, indicating no reversal of the flow while oxygen was being administered. The left atrium was entered through a patent foramen ovale but no shunt was found in either direction.

Selective cineangiocardiography was performed at the time of the cardiac catheterization. Serial filming at 30 frames per second was done during the injection of 90 per cent Hypaque-M* into the patent ductus arteriosus, the main pulmonary artery, the right ventricle, and the left atrium. The injection of contrast medium into the right ventricle opacified that chamber, the main pulmonary artery, the left pulmonary artery, the ductus ar-

*Hypaque brand of diatrizoic acid with methylglucamine concentrate: Winthrop Laboratories, New York 18, New York.

Circulation, Volume XXIV, September 1961
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Table 1

<table>
<thead>
<tr>
<th></th>
<th>Preoperative study</th>
<th>Pure oxygen</th>
<th>Postoperative study</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Age 12 months</td>
<td>(Pressure, mm Hg)</td>
<td>(Content of O₂ vol. %)</td>
</tr>
<tr>
<td>Room air</td>
<td></td>
<td>(Pressure, mm Hg)</td>
<td>(Content of O₂ vol. %)</td>
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<tr>
<td>IVC</td>
<td>3.6</td>
<td>14.5</td>
<td>10.4</td>
</tr>
<tr>
<td>SVC</td>
<td>3.1</td>
<td>15.6</td>
<td>11.6</td>
</tr>
<tr>
<td>RA</td>
<td>5.4</td>
<td>1.0</td>
<td>11.9</td>
</tr>
<tr>
<td>LA</td>
<td>5.7</td>
<td>14.6</td>
<td>16.5 (97% saturation)</td>
</tr>
<tr>
<td>RV</td>
<td>101/5</td>
<td>82/9</td>
<td>11.5</td>
</tr>
<tr>
<td>PA</td>
<td>103/61</td>
<td>81/50</td>
<td>11.3</td>
</tr>
<tr>
<td>DA</td>
<td>101/59</td>
<td>101/59</td>
<td>22.2 (100% saturation)</td>
</tr>
<tr>
<td>RFA</td>
<td>104/58</td>
<td>—</td>
<td>16.3 (97% saturation)</td>
</tr>
</tbody>
</table>

Explanation of symbols: IVC, inferior vena cava; SVC, superior vena cava; RA, right atrium; LA, left atrium; RV, right ventricle; PA, pulmonary artery; DA, descending aorta; RFA, right femoral artery.

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The right pulmonary artery, however, did not fill (fig. 5). Following the injection into the left atrium a normal left atrium and ventricle with intact septa were noted. There was simultaneous opacification of the ascending aorta and the right pulmonary artery and its branches.

To clarify better the origin of the blood flow to the right lung a retrograde arterial cineangiogram by way of the right brachial artery was carried out. The catheter tip was positioned just distal to the aortic valve for the injection of contrast medium. It was conclusively demonstrated that the right pulmonary artery arose as a single vessel from the posterior aspect of the ascending aorta (fig. 6).

Once the diagnosis was known examination showed minimal but definite signs of early clubbing and faint cyanosis of the nailbeds, more pronounced in the left arm than in the right.

Operative Procedure

At surgery a T-shaped sternal incision was made. The ascending aorta was enlarged and appeared dilated beyond the aortic valve. A large right pulmonary artery arose from the posterior surface of the aorta. Its diameter was judged to be 12 or 13 mm., approximately the size of the aorta at that level. Just distal to the origin of the left subclavian artery the aorta narrowed somewhat to a diameter of 6 or 7 mm. The ductus arteriosus extended from the end of the pulmonary trunk to the narrowed portion of the distal aortic arch. It was approximately the same size as the aorta at that location. The pulmonary trunk then continued as the left pulmonary artery (fig. 7). Systolic pressures in each of these structures were essentially equal. After dissection of the various vessels the right pulmonary artery was clamped and its point of origin from the aorta was closed with sutures. The ductus arteriosus was divided between clamps and both ends were sutured. Finally, the anterior portion of the main pulmonary trunk was clamped with a curved vascular clamp in such a way as to permit a longitudinal incision, and a stretch-weave Dacron graft with a diameter of 13 mm. was interpolated anterior to the aorta end-to-side between the distal end of the right pulmonary artery and the pulmonary trunk. The surgery was well tolerated.

Postoperative Course

The postoperative course was uneventful. Although the child had not been in respiratory distress prior to surgery, it was soon evident that he breathed a great deal easier after surgery. He was discharged on the fourteenth day after the operation.

Six weeks following surgery, at the age of 14 months, the patient weighed 18 pounds 14 ounces, a postoperative gain of 3 pounds 14 ounces. He had improved in all respects. The heart was slightly enlarged to the left by percussion but not overactive. There was a faint systolic murmur in the pulmonic area. The pulmonary second sound was single and regarded as being at the upper limits of normal in intensity.

The cardiac status was re-evaluated 3 months after surgery when the infant was 15 months old. On the postoperative radiographs the con-
gestive changes in the lungs had cleared. No unusual chamber enlargement was noted but there was a prominent bulge along the right superior paramediastinal area (fig. 2). The electrocardiogram had returned toward normal (fig. 3). The findings at right heart catheterization were regarded as within normal limits except for mild elevation of the right ventricular pressure (32/5 mm. Hg). The pulmonary artery pressure was 28/7 mm. Hg (fig. 4). Again there was no evidence of an intracardiac shunt (table 1). Cineangiography proved the surgical anastomosis to be widely patent (fig. 8).

Discussion

Although anomalous origin of the right pulmonary artery from the ascending aorta with or without a patent ductus arteriosus is uncommonly encountered, its recognition is essential for proper patient management, since
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Left. Anomalous origin of the right pulmonary artery from the posterior aspect of the ascending aorta with an associated patent ductus arteriosus. Right. Method of surgical correction.

it is a surgically correctable condition. Symptoms tend to appear early and to be severe. There are two features that might aid one in suspecting the diagnosis. If radiographic study shows a disproportionate or unilateral increase in pulmonary vasculature, this condition should be considered. If a patent ductus arteriosus were present, a difference in color and clubbing in the arms might be evident. Angiocardiography is of prime importance and the only certain method of delineating the anomaly.

While the prognosis in the case presented must remain uncertain pending long-term observation, there is reason to believe the outcome will be good. The excellent response of the pulmonary vascular bed in a 1-year-old child, in whom the pressure previously was equal to systemic, is very encouraging. Fortunately it was possible to utilize a graft of such size that it should permit good perfusion of the right lung despite growth.

As far as we can ascertain this is the first case in which a graft has been used successfully in the pulmonary arterial tree. The problem has, however, been subjected to experimental study. Robinson, Glotzer, Gilbert, and Hurwitt showed that aortic homografts in the pulmonary artery are subject to severe degenerative alterations. Moore and his associates in our laboratory demonstrated that plastic tubular grafts work well when interposed in the pulmonary arteries. After surgery in our patient, Sarrvage, Rudolf, and Gross reported on the experimental utilization of pericardial grafts for pulmonary artery replacement.

A survey of the literature has revealed no other case of successful complete surgical correction of this defect.

Summary

A case of anomalous origin of the right pulmonary artery from the ascending aorta with an associated patent ductus arteriosus has been described. Successful complete surgical correction was accomplished after physiologic and cineangiographic studies had established the diagnosis. Postoperative evaluation demonstrated a return to nearly normal physiologic values. The unparalleled value of selective angiography in the anatomic assessment of cardiovascular lesions is emphasized.

Addendum

Additional reference: Caro et al.* reported a 23-year-old man with aortic origin of the right pulmonary artery and patent ductus arteriosus. The anomalous pulmonary vessel was stenotic at its origin, which

may have accounted for the absence of symptoms until the age of 17 years. Diagnosis was established by cardiac catheterization and angiocardiography. Surgical correction by an Ivalon graft with end-to-end anastomosis to the right pulmonary artery and end-to-side anastomosis to the main pulmonary artery was accomplished; however, the patient died 5 hours after completion of surgery.

Our patient has been examined 15 months after surgery. He has grown and developed normally and is asymptomatic. A soft systolic murmur persists at the base of the heart, but examination otherwise is normal.

References

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Circulation. 1961;24:662-668
doi: 10.1161/01.CIR.24.3.662

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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