Aortic Origin of the Right Pulmonary Artery

Surgical Repair Without a Graft

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SUMMARY
Surgical repair of an anomalous right pulmonary artery arising from the ascending aorta is rare. Angiocardiography is the procedure of choice in establishing the diagnosis. Surgical correction of the defect is mandatory; otherwise most of these children die in refractory heart failure. The present paper reports the first successful anatomic repair without a graft in a 2-month-old baby. Five months after operation the boy is asymptomatic and demonstrating normal growth and development.

Additional Indexing Words:
Anomalous right pulmonary artery

A SINGLE anomalous vessel that arises from the ascending aorta to supply the right lung is an uncommon form of congenital heart disease; when it does occur, frequently associated defects are patent ductus arteriosus, anatomically patent foramen ovale, or both. Seven cases of this anomaly were reported prior to 1960; however, as a result of the increasing use of angiocardiography, 24 more cases including Stanton’s cases (personal communication) have now been reported. Since nearly all infants with this anomaly die of congestive heart failure and pneumonia before they are 1 year old, surgical therapy is urgent. Five patients have been subjected to complete surgical repair. Two of these, died in the immediate postoperative period but three survived. The present paper reports the first successful anastomosis of the right pulmonary artery to the main pulmonary artery without a graft.

Report of Case
G.J., a 7-month-old Caucasian boy, was first referred to the Riley Hospital when 2 months of age because of cyanosis and labored breathing. He was the third of three boys born to a 28-year-old mother. The prenatal history was uncomplicated, and the child was born by a normal vaginal delivery with a birth weight of 8 lb. 9 oz. When discharged with the mother at 4 days of age, the infant was not noted to be cyanotic or to have abnormal breathing. Except for a slight cough, he apparently did well until 6 weeks of age when his breathing became labored and he “turned blue” during one of his feedings. The baby was taken to the local hospital where a murmur was first heard, and an electrocardiogram and chest films were said to indicate congenital heart disease. The child was started on digoxin (Lanoxin) between this visit to the local hospital and his first referral to the Riley Hospital. He continued to have faster and more labored breathing, but there were no further cyanotic episodes.

On physical examination the child was irritable, pale, and minimally cyanotic. He appeared well-nourished and weighed 10 lb. 2 oz. The respirations were 70/min with mild retraction and a brassy cough. Auscultation revealed scattered rhonchi throughout the chest and rales in the lung bases. The left hemithorax was larger than the right with a precordial bulge. The
heart rate was 140/min, and the pulses were somewhat increased in volume. Cardiac dullness was percussed to the anterior axillary line, and there was increased left parasternal activity. A thrill was palpable at the left sternal border. The first heart sound was loud. The second heart sound was partially obscured by a long, harsh, grade IV/VI systolic murmur with maximal intensity at the left sternal border. A mid-diastolic filling murmur was heard at the apex. The liver which was palpable 3 cm below the right costal margin had a blunt edge. The spleen was not palpable. No clubbing of the nails or edema was noted.

Laboratory examination revealed normal urine; hemoglobin values of 18.2 g/100 ml; 7,500 leukocytes with 23% polymorphonuclear leukocytes and 76% lymphocytes; hematocrit 52%; and blood urea nitrogen of 7 mg/100 ml. The electrocardiogram showed large P waves and evidence of biventricular hypertrophy. The R wave in aV_R was 11.5 mm, and a qR complex was noted in V_1 with the q 3 mm and the R 22 mm. The chest roentgenogram revealed a markedly enlarged heart with a prominent main pulmonary artery and increased pulmonary vasculature (fig. 1).

The clinical impression was a large ventricular septal defect or a persistent truncus arteriosus.

After admission, the infant showed progressive cardiac failure with listlessness and frequent

Figure 1

Preoperative frontal chest roentgenogram demonstrates a markedly enlarged heart with prominent main pulmonary artery and increased pulmonary vasculature.

Figure 2

(A) Cineroentgenogram in the frontal position with the injecting catheter tip in the ascending aorta, showing anomalous right pulmonary artery filling from the aorta. (B) Drawing illustrating figure 2A.
vomiting. The dose of Lanoxin was increased, and diuretics were added. Because of unresponsiveness to medical management, cardiac catheterization was performed with the aid of local anesthesia. Left heart catheterization was done via the right brachial artery, and right heart catheterization was done via the right saphenous vein. The oxygen data showed no evidence of a right-to-left shunt. Right ventricular pressure was 100/10 mm Hg, and the left ventricular pressure was 90/8 mm Hg. The pulmonary artery was not entered, and a patent ductus was not demonstrated but could not be excluded. Cineangiograms with injection of contrast medium into the right ventricle disclosed filling of the left pulmonary artery and the left pulmonary tree; no filling of the right lung was seen after this injection. Left ventricular injection revealed a vessel arising from the ascending aorta which filled the right pulmonary arterial tree. The ventricular septum was intact. The aortogram showed that the right pulmonary artery arose from the ascending aorta (fig. 2).

Surgery was performed 2 days following the catheterization. Under general anesthesia, a mediastinotomy incision was made and the pericardium was opened vertically. The right pulmonary artery, approximately 1 cm in diameter, arose from the ascending aorta about 1 cm above the aortic valve (fig. 3). Further dissection and inspection revealed a patent ductus arteriosus which was doubly ligated and divided. The aorta and pulmonary artery were then dissected free. A partial occluding clamp was placed across the right side of the aorta to include the origin of the right pulmonary artery. The right pulmonary artery was dissected from the ascending aorta and the ascending aorta was sutured with 5-0 Dacron. The right pulmonary artery was then passed behind the ascending aorta and anastomosed end-to-side to the right side of the main pulmonary artery, with a running everting mattress suture posteriorly and an interrupted everting mattress suture anteriorly (fig. 4). After release of all clamps, there appeared to be good flow of blood from the main pulmonary artery into the right pulmonary artery. The pressures, measured immediately following the procedure, were approximately 30 mm Hg in the pulmonary artery and approximately 60 mm Hg in the systemic circulation. The pericardium was closed with interrupted sutures and the sternum was closed with interrupted encircling Dacron sutures.

Figure 3
The anomalous right pulmonary artery arising from the ascending aorta as seen at surgery.

Figure 4
The complete surgical repair. The ductus arteriosus has been doubly ligated and divided and the right pulmonary artery is anastomosed to the main pulmonary artery behind the ascending aorta.
When the child was seen 1 month following surgery, the parents reported that the child's color and respirations were normal and that he was eating without difficulty. He weighed 12 lb. 10 oz. The pulses were within normal limits, and there was no cyanosis. The first heart sound was still prominent, and a grade III/VI ejection systolic murmur was present at the base. No diastolic filling murmur was heard.

When the boy was readmitted 5 months postoperatively for functional evaluation of the surgical repair, the parents stated that the boy "is perfect." His growth and development were normal, and he was above the 90th percentile in height and weight. On physical examination the child was alert and active without distress. The blood pressure was 100 by palpation, and the point of maximal impulse was within normal limits. A grade III/VI ejection systolic murmur was still present at the left sternal border with slight increase in the second component of the second sound. Results of the rest of the examination were within normal limits. The electrocardiogram showed only minimal right ventricular hypertrophy. The x-rays revealed a decrease in the heart size to the upper limits of normal (fig. 5).

Figure 5
Postoperative frontal chest roentgenogram demonstrates normal lung vasculature and heart size at the upper limits of normal.

Figure 6
Postoperative frontal cineroentgenogram with the injecting catheter tip in the main pulmonary artery shows filling of right and left pulmonary arteries. The right pulmonary artery is slightly less filled than the left.

Figure 7
Lung scan illustrates slightly diminished count over the right lung field. Note: The area in upper left is the site of injection of radioactive material.
At catheterization the left ventricular pressure and the descending aortic pressure were normal. The right ventricular pressure was 36/6 mm Hg. The right pulmonary artery mean pressure was 13 mm Hg as compared to the mean of the main pulmonary artery and the left pulmonary artery of 19 mm Hg. Cineangiography, with injection in the main pulmonary artery demonstrated a minimal delay in filling of the right pulmonary artery with a little stenosis at the anastomotic site (fig. 6). The site of excision of the right pulmonary artery from the ascending aorta, as shown by aortography, seemed essentially normal. A lung scan done the day following repeat catheterization indicated slightly better filling of the left than of the right lung (fig. 7).

Discussion

The signs and symptoms related to this abnormality are not diagnostic. They suggest a large left-to-right shunt, usually with mild or intermittent cyanosis from a smaller right-to-left shunt through a patent ductus arteriosus or a patent foramen ovale. If there is a considerable right-to-left shunt through the patent ductus arteriosus, differential cyanosis may be noted with the left upper extremity being more cyanotic than the right. Congestive heart failure unresponsive to medical management characteristically appears in the first year.

Angiocardiography is the diagnostic procedure of choice. Right and left heart catheterizations are necessary to delineate clearly the origin of this anomalous vessel and to demonstrate associated cardiovascular defects which may condition the surgical treatment. The cases of Bopp, Fraenkel, and Jew and Gross are not included in the cases reviewed here because of this latter consideration.

Definitive diagnosis and surgical repair are now possible. Findlay and Maier initially suggested ligation of the right pulmonary artery and pneumonectomy. In three cases, however, ligation was unsuccessful. Later Maier proposed use of a graft to anastomose the anomalous right pulmonary artery to the main pulmonary artery. This was first accomplished by Caro and associates with an Ivalon graft, but the patient died 5 hours after operation. The successful use of a Dacron graft in a 1-year-old boy and a Teflon graft in a 4½-year-old boy were reported by Armer and associates, and Redo and associates, respectively. Griffiths and associates reasoned that surgical correction of this anomaly could be accomplished by anastomosing the anomalous right pulmonary artery to the main pulmonary artery. That this is surgically possible has been demonstrated both by our case and those of Stanton. It is further suggested that this is the treatment of choice when it is technically possible. When direct surgical anastomosis is not possible, an interposed graft of sufficient caliber to provide adequate blood flow as the child grows would be the treatment of choice.

The data presented in the case report indicate near normal pressures in the pulmonary arteries after operation with an estimated left lung resistance of less than 40% of that prior to the repair. When surgical correction is performed before the patient is 1 year of age, a return of pulmonary pressures to near normal can be expected.

The embryological origin of the anomalous right pulmonary artery is unclear. Misplaced attachment of the sixth aortic arch or retardation of this arch as the truncus divides was suggested by Schneiderman. The finding, in the cases of Porter and Turpin and their associates of a fibrous band connecting the right pulmonary artery to the main pulmonary artery lends support to this concept. Wagenvoort and associates hypothesized that the anomalous vessel was composed, proximally to distally, of the right dorsal aorta, right patent ductus arteriosus, and right pulmonary artery. Rosenberg and associates suggested two major types. Type I is based on the observations of Wagenvoort and associates while type II is based on those of Schneiderman. Since type I tends to have a smaller lumen and thus a decreased flow to the right lung, these patients would not be expected to have failure from a large left-to-right shunt; patients with the type II anomaly may be more likely to succumb from congestive heart failure secondary to a large left-to-right shunt through a large luminal anomalous right pulmonary artery. The case reported herein can best be classified as type II.
References
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