Diagnosis and Surgical Treatment of Aortopulmonary Fenestration

By Lewis H. Bosher, Jr., M.D., and Carolyn Moore McCue, M.D.

AORTOPULMONARY FENESTRATION or partial persistent truncus arteriosus is undoubtedly not as rare a congenital lesion as earlier statistics have suggested. This defect is also called aortic septal defect or aortopulmonary window.

In 1949 Perelman and Putschar1 were able to assemble only 14 cases from the literature. Skall-Jensen2 reported 64 additional patients collected between 1949 and 1956 inclusive. In only 10 of these had the diagnosis been definitely established prior to operation or death. In 1948 Gross3 achieved the first successful closure of an aortopulmonary fenestration by means of a tape ligature, a method which he himself considered hazardous and uncertain. In 1953 Scott and Sabiston4 reported successful division and suture of this lesion in a 10-month-old infant with a clamp technic. Fletcher and associates5 added a third successful case in 1954 performed in a similar manner.

Cooley et al.6 advanced the surgical treatment of this condition with a description in 1957 of three successfully managed cases, one performed under hypothermia and two under cardiopulmonary bypass. At that presentation, 16 surgical cases were mentioned with nine complete closures, three incomplete closures, and four operative deaths.

Skall-Jensen2 in his review added three more completely successful surgical cases, two partially successful ones and three operative deaths. Baronofsky et al.7 recently reported an unusual case with two congenital aortopulmonary septal defects, 1 cm. and 3 cm. in size, respectively, both of which he was able to divide with a clamp technic.

A few additional cases have been reported in a large series of patients with congenital heart disease operated upon by cardiopulmonary bypass.

Embryologically the defect appears during the fifth to the eighth week of intrauterine development through an incomplete formation of the truncoconal septum. The opening is usually large, although it may vary from 5 to 30 mm. in diameter. Its proximal margin is located about 5 to 10 mm. distal to the aortic valve and therefore presents within a few millimeters of the edge of the myocardium. The hemodynamic changes associated with the lesion are severe. In contrast to patent ductus, bacterial endarteritis rarely occurs. A few aneurysms of the pulmonary artery have been reported. The mortality in infancy is high, and relatively few such individuals attain adulthood.

We have observed five patients with this defect. Surgery was attempted in three of these, and in two the outcome was successful. The correct diagnosis was made preoperatively in all three.

The important clinical findings observed in these five cases are tabulated in table 1. The symptoms that characterize the condition are frequent respiratory infections, failure to gain weight and develop normally, dyspnea on exertion, and excessive fatigue. Death in infancy usually results from pulmonary infection and heart failure. Clinical differentiation from other large left-to-right shunts may be difficult, and confusion with interventricular septal defect and patent ductus arteriosus is common. Our patients exhibited only a systolic murmur, but in at least half of the reported cases either a continuous or diphasic systolic and diastolic murmur has

From the Division of Thoracic and Cardiovascular Surgery, Department of Surgery, and from the Department of Pediatrics, Medical College of Virginia, and the Congenital Heart Clinic of the Virginia State Health Department, Richmond, Virginia.
been heard. Careful localization of the systolic murmur is helpful, since in aortopulmonary fenestration the maximum intensity is usually found in the third and fourth left intercostal spaces in contrast to the murmur of patent ductus, which is loudest in the second interspace or higher.

Since the opening is usually large, pressure in the main pulmonary artery is equivalent to that in the ascending aorta. Pulmonary vascular resistance is high and in some cases rises progressively. Hemodynamically, the defect resembles the high pressure patent ductus, and a continuous murmur is therefore frequently absent. Pulmonary hypertension is reflected in the loud, banging pulmonary second sound. The electrocardiogram often shows right ventricular hypertrophy and strain. Characteristically, radiologic examination reveals right and left ventricular enlargement, pulmonary artery dilatation, prominence of the pulmonary vascular markings, and various degrees of increased pulmonary artery pulsation. The ascending aorta is enlarged in aortopulmonary fenestration, and this segment is seen to pulsate strongly. In patent ductus arteriosus the enlargement extends to the first portion of the descending aorta and prominent pulsation can be observed in the arch.

A 30-year-old married woman (case 4), mother of two children, presented a somewhat different clinical picture. The only complaint was excessive fatigue. Moderate generalized cyanosis with clubbing was evident. A faint systolic murmur was audible in the fourth left intercostal space. The heart was small, and the peripheral lung fields were clear. There was moderate polycythemia with a red blood cell count of 5.8. The electrocardiogram showed right axis deviation. Cardiac catheterization demonstrated severe pulmonary hypertension and a moderate increase in oxygen saturation in the pulmonary artery. Confirmation of the suspected aortopulmonary fenestration was obtained by retrograde aortography with a catheter passed through the defect into the pulmonary artery (fig. 1). A moderate right-to-left shunt was demon-
strated as the dye streamed back through the defect into the aorta. Surgery was not advised for this patient, since it was believed that pulmonary vascular changes were advanced.

One other patient observed was not subjected to surgery. This was a white male infant who had an imperforate anus repaired shortly after birth. Other congenital defects were a clubbed foot and hypospadias. Cardiac failure developed at 6 weeks of age and was relieved only temporarily by digitalis. A faint systolic murmur was heard in the left parasternal region, and a gallop rhythm and precordial shock were present. The failure never could be controlled adequately and unfortunately he died before retrograde aortography could be carried out. Terminally there was mild cyanosis. The electrocardiogram showed severe right ventricular strain. He weighed 8.9 pounds at birth and 7.9 pounds at death. Postmortem examination revealed a 15 mm. defect located approximately \( \frac{1}{2} \) centimeter beyond the aortic leaflets (fig. 2).

In all four patients subjected to cardiac catheterization, an increase in oxygen saturation was demonstrated at the pulmonary artery level (table 2). Differentiation from a patent ductus arteriosus, however, can usually not be made unless the catheter is passed through the aortic septal defect and the more medial or anterior position of the catheter demonstrated by fluoroscopy or by passage of the catheter into one of the arch vessels. The catheter through a patent ductus arteriosus assumes a more posterior position as viewed in the lateral projection and subtends the arch. In three of four patients, retrograde aortography with the catheter positioned in the ascending aorta clearly demonstrated the defect (fig. 3). In one infant it was possible to achieve adequate visualization by remote injection from the left brachial artery (fig. 4). Gasul, Fell, and Casas\(^9\) first utilized retrograde aortography to establish the diagnosis of aortopulmonary fenestration.

In the presence of some right-to-left shunting comparative oxygen saturation determinations from the right brachial, left brachial, and femoral arteries may yield diagnostic information. Identical values will be found in these locations in aortopulmonary fenestration, whereas in patent ductus, the femoral artery and, in some instances, the left brachial artery saturations will be lower. The difference is enhanced when the patient is made to breathe low oxygen mixtures. Similar information is provided by dye-dilution curves sampled from these sites.

The surgery of aortopulmonary fenestration has progressed through several stages. Ligation or suture ligation, first successfully

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**Figure 1**

Patient M. T. Chest film: Note small heart, prominent pulmonary artery, and avascular peripheral lung fields. Aortogram: Injection made into main pulmonary artery and right ventricle via catheter passed through defect showing defect and some right-to-left shunting.

**Figure 2**

Specimen of 2½ month old infant (B. II.) dying in heart failure with pulmonary complications. Defect 15 mm. in diameter.
AORTOPULMONARY FENESTRATION

employing by Gross, was recognized as potentially hazardous and likely to result in incomplete closure. It is certainly not appropriate for a large defect. Successful employment of this technic, however, has been reported by Varco and Davis. Bailey achieved nearly complete closure of a large defect by placing two rows of parallel mattress sutures without division. A distinct advance was made with the report by Scott and Sabiston of the successful division and suture of this defect by use of thin Potts vascular clamps. Scott, Kirklin, and Baronofsky reported additional cases managed by this technic. However, the dissection required for adequate isolation of the defect and the application of clamps to vessels under high pressure may be extremely difficult and dangerous. The defect is large and constitutes a window opening between the pulmonary artery and the aorta, both of which may be markedly dilated. The posterior wall of the defect is often thinned out. Only a few millimeters separate the proximal margin of the defect from the edge of the myocardium. In a number of instances, aggressive dissection has led to laceration of one of the vessels with resulting fatal hemorrhage. Ross and Cooley et al. described the successful use of hypothermia with inflow occlusion in order to deflate these vessels and to facilitate the application of the clamps.

Shumway and Lewis demonstrated the feasibility of closing through the pulmonary artery an experimentally created aortopulmonary window under inflow occlusion and hypothermia. We are aware of only one clinical case that has been managed in precisely this manner. Kirklin and Devloo have employed a similar technic in the closure of previously constructed Potts anastomoses during definitive correction of tetralogy of Fallot.

The application of cardiopulmonary bypass to this problem has seemed a further logical advance. Minimal dissection is required prior to severance of the fenestration. Major hazards introduced are the interruption of coronary flow and the great likelihood of inducing coronary air embolism unless special precautions are taken. We have modified our technic with each successive case and will probably manage the next case in the following manner. Shortly after initiating cardiopulmonary bypass, the distal ascending aorta is clamped. The apex of the left ventricle is quickly cannulated with a catheter and pulmonary ventilation suspended. Selective hypothermia of the myocardium is achieved with ice-saline, and the aortopulmonary window severed. The pulmonary artery defect is closed first, either longitudinally or transversely, with care to exclude all air by releasing the superior vena caval tape just prior to completing the closure. The aorta is then closed transversely and again the superior vena caval tape is released to facilitate washing all air out of the left ventricle and ascending aorta. The heart is rewarmed by releasing the aortic occluding clamp, and
Table 2

Cardiac Catheterization (Diagnostic)

<table>
<thead>
<tr>
<th></th>
<th>J. G.</th>
<th>B. P.</th>
<th>K. C.</th>
<th>M. T.</th>
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<tr>
<td>Pressures</td>
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<tr>
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<td>48%</td>
<td>72%</td>
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<td>92</td>
<td>99</td>
<td>93</td>
<td>88.91</td>
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Pulmonary to systemic blood flow ratio 4.3:1 1:1 2.1:1 1:1

after restoration of normal beat and contractility the ventricular vent is removed.

As an alternative to the above technic the transpulmonary approach to the defect might be expected to simplify its closure, especially when performed on cardiopulmonary bypass.

Surgical Case Reports

Case 1

J. G., age 4 years (table 1). This child had a murmur recognized at 1 month of age and frequent respiratory infections, but no cyanosis or dyspnea. At 4 years of age she weighed only 26 pounds.

Cardiac catheterization* was performed in September 1956 and revealed severe pulmonary hypertension and a large left-to-right shunt at the pulmonary artery level. Retrograde aortography in November 1956 clearly showed an aortopulmonary fenestration. Surgery was performed in March 1957 by use of extracorporeal circulation. Under normothermic conditions the heart was arrested with potassium citrate, the fenestration severed, and the aorta closed longitudinally with two rows of continuous silk sutures (fig. 5). The clamp occluding the aorta was then released after 23 minutes of myocardial ischemia, following which the pulmonary artery was closed longitudinally in a similar fashion. Resumption of normal rhythm was followed by ventricular fibrillation, but the heart was defibrillated readily. Her immediate postoperative state was good. During the night after operation she was digitalized because of moderate hypotension. On the fourth postoperative day she suffered a sudden transient left hemiparesis, which cleared completely after 36 hours and was thought to be caused by a small embolus from the suture line.

At the end of 1 week, chylothorax developed and persisted under conservative management for 2 weeks. Reexploration and ligation of the thoracic duct was followed by complete recovery. Four years later, repeat cardiac catheterization reveals normal hemodynamics and normal pulmonary artery pressures (fig. 6). The pulmonic second sound is still somewhat loud, and there is mild cardiomegaly on the chest film. Her general condition is excellent.

Case 2

B. P., age 15 months (table 1). In the newborn nursery this baby was found to have a loud murmur and intermittent cyanosis with dyspnea. The cyanosis diminished over the first few months of life, but frequent respiratory infections were a real problem. Digitalis appeared to be beneficial. He grew slowly and weighed about 16 pounds at 1 year of age. The electrocardiogram, which showed a right axis and mild right strain at 3 weeks of age, developed a more definite left ventricular pattern during the first year and by the time of surgery there were prominent Q waves, huge complexes, and a left ventricular strain pattern.

Cardiac catheterization performed at age 13 months revealed severe pulmonary hypertension and a left-to-right shunt at the pulmonary artery level. Retrograde aortography from the left brachial artery disclosed an aortopulmonary fenestration. Surgery was performed in August 1960 by use of extracorporeal circulation under normothermia. The aorta was clamped, the fenestration severed, and the aorta closed longitudinally with continuous silk suture. Flow was restored after 12 minutes of myocardial ischemia and the pulmonary artery was then sutured longitudinally. Heart beat returned promptly but did not seem to be vigorous. At the conclusion of the operative

*Performed by Dr. H. Page Mauck, Jr.
procedure there was evidence of pulmonary congestion. Moderate hypotension ensued which did not respond to digitalization. Urine output was adequate, however. Because of the pulmonary congestion, the child was placed on the Bennett respirator. Ten hours after operation he had a mild generalized convulsion. The circulation deteriorated. A tracheostomy was performed but the child died approximately 12 hours after surgery. Postmortem examination showed pulmonary congestion and cerebral edema. Cardiac failure due to coronary air embolism could not be excluded.

Case 3

K. C., age 3 years (table 1). This acyanotic girl had a loud murmur at 6 weeks of age, but surprisingly little exercise intolerance. The murmur diminished over the first 2 years of life, and the striking feature was the banging pulmonary second sound, thrill, and shock at the second left intercostal space, with cardiomegaly and pulmonary congestion. Her growth was strikingly below normal; she weighed 23½ pounds at 3½ years of age.

The child was first catheterized at age 2. The results were inconclusive, since the pulmonary artery was not entered, but suggested an atrioventricular canal. Repeat catheterization studies at age 3 disclosed severe pulmonary hypertension and a left-to-right shunt at the pulmonary artery level. The pulmonary artery saturation was 81 per cent, but in a medial position in the main pulmonary artery, a value of 93 per cent was obtained, suggesting aortopulmonary fenestration. Retrograde aortography confirmed this suspicion and clearly demonstrated the defect.

Surgery was carried out in January 1961 under extracorporeal circulation. After clamping the aorta, the heart was arrested by selective hypothermia using external cooling. The defect was severed and the aorta closed transversely with two rows of continuous suture. The pulmonary artery was then closed longitudinally. The left ventricle was vented with a catheter to assure removal of all air before releasing the aortic clamp after 28 minutes of occlusion. Normal sinus rhythm resumed. Her condition postoperatively was satisfactory. She was digitalized soon after surgery. Thrombocytopenia of 6,000 developed on the fourth postoperative day but responded rapidly to prednisolone (Meticortelone) and her further course was uncomplicated. Her present condition is excellent.

Summary and Conclusions

The clinical picture of aortopulmonary fenestration has been reviewed and the important role of cardiac catheterization and retrograde aortography in the early diagnosis of this lesion emphasized.

The history of the surgical treatment leading to the routine use of extracorporeal circulation has been summarized.

Three patients have been treated surgically, two of whom survived. Coronary air embolism may have contributed to cardiac failure in the patient who died and certain precautions necessary to prevent this complication have been outlined.

References


4. Scott, H. W., Jr., and Sabiston, D. C., Jr.

On Permanent Patency of the Mouth of the Aorta, or Inadequacy of the Aortic Valves
By DOMINIC JOHN CORRIGAN, M.D.

One of the Physicians to the Charitable Infirmary, Jervis Street, Dublin; Lecturer on the Theory and Practice of Medicine; Consulting Physician to St. Patrick's College, Maynooth

The disease to which the above name is given has not, so far as I am aware, been described in any of the works on diseases of the heart. The object of the present paper is to supply that deficiency. The disease is not uncommon. It forms a considerable proportion of cases of deranged action of the heart, and it deserves attention from its peculiar signs, its progress, and its treatment. The pathological essence of the disease consists in inefficiency of the valvular apparatus at the mouth of the aorta, in consequence of which the blood sent into the aorta regurgitates into the ventricle. This regurgitation, and the signs by which it is denoted, are not necessarily connected with one particular change of structure in the valvular apparatus, and hence the name Permanent Patency of the Mouth of the Aorta, or Inadequacy of the Aortic Valves, has been chosen as simply expressing such a state of the parts as permits the regurgitation to occur.

Circulation, Volume XXV, March 1962

[Image: On Permanent Patency of the Mouth of the Aorta, or Inadequacy of the Aortic Valves by Dominic John Corrigan, M.D.]
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_Circulation._ 1962;25:456-462
doi: 10.1161/01.CIR.25.3.456
_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

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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