Congenital Aortopulmonary Septal Defect
Clinical and Hemodynamic Findings, Surgical Technic,
and Results of Operative Correction

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The management of patients with congenital communications between the ascending aorta and the main pulmonary artery is a challenging problem, not only in the establishment of the correct diagnosis preoperatively, but in subsequent surgical treatment as well. Although often described as a rare malformation, aortopulmonary septal defect will probably be encountered in any large group of patients with acyanotic forms of congenital heart disease that is subjected to detailed diagnosis assessment or operation. Six patients with aortopulmonary septal defects have been studied at the National Heart Institute and in five the lesion was subsequently corrected. Since the success of operative treatment, for this lesion in particular, is largely dependent upon preoperative recognition and appropriate preparation, the diagnostic features of the malformation in these patients as well as the technic and results of closure are presented in detail.

Clinical and Hemodynamic Findings

The pertinent features of the histories and clinical examinations of the six patients are summarized in table 1. All were males and their ages ranged from 4 to 14 years. A heart murmur had been known to be present since birth or early infancy in four of the patients and, with the exception of S. D., each had experienced progressive exertional dyspnea, fatigability, and frequent respiratory infections. Only patient J. S., however, had had congestive heart failure; this occurred at the age of 1 year, before an associated patent ductus had been closed. Four of the six patients evidenced essentially normal growth and development while two were well below average height and weight.

On physical examination, prominence of the left precordium and a systolic thrill along the left sternal border were usually present. The second heart sound over the pulmonic area was always found to be accentuated. On auscultation, and by phonocardiography, no patient was found to have a continuous murmur. A systolic murmur, of the ejection type, however, was always audible and recordable along the left sternal border (fig. 1). In S. S., the patient with associated subaortic stenosis, the second sound exhibited paradoxical splitting. A diastolic murmur was never heard at the base of the heart in any patient but at the apex a low-pitched, mid-diastolic rumbling murmur was recorded in two of them. In patient S. S., subaortic stenosis was evidenced by prominence of the systolic murmur and thrill to the right as well as to the left of the sternum. The blood pressures, as recorded by sphygmomanometry, were not abnormal. The heart of every patient was found to be enlarged on roentgenographic examination. The characteristic changes (fig. 2) were enlargement of both ventricles, the left atrium and the pulmonary artery, and evidence of increased pulmonary vascular markings. The electrocardiographic findings are listed in table 1 and a typical tracing, showing left and right ventricular hypertrophy with right axis deviation, is reproduced in figure 3.

Cardiac catheterization was carried out in each patient and a summary of the hemodynamic data is presented in table 2. Severe right ventricular and pulmonary arterial hypertension was found to be present in every instance. By means of inhaled inert gas tests utilizing N₂O or Kr⁸⁵ a left-to-right shunt, terminating in the pulmonary artery, was demonstrated in every patient. In five patients (S. D. excepted) the left-to-right shunt was large and the pulmonary to systemic flow ratios ranged from 1.7:1 to 2.8:1 and averaged 2.3:1. By relating these flow ratios to the mean pulmonary and systemic arterial pressures the ratio of pulmonary to total systemic vascular resistance could be calculated. These relationships were similar in the five patients, ranging from 31 to 44 per cent and averaging 39 per cent. The presence of pulmonic regurgitation was demonstrated in three of the six patients by positive N₂O or Kr⁸⁵ tests in the right ventricular outflow tract. In contrast to the blood pressures recorded...
<table>
<thead>
<tr>
<th>Patient</th>
<th>Age, years</th>
<th>Sex</th>
<th>History</th>
<th>Physical signs</th>
<th>Electrocardiogram</th>
<th>Associated anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>H.C.</td>
<td>14</td>
<td>M</td>
<td>Frequent respiratory infections, cardiomegaly noted 1 year of age, progressive dyspnea from age 6 years; previously explored for ductus</td>
<td>Systolic murmur max. in pulmonic area, thrill 2-3 left ICS, short diastolic murmur; normal development; BP 120/50</td>
<td>LVH, semivertical axis</td>
<td>None</td>
</tr>
<tr>
<td>G.L.</td>
<td>7</td>
<td>M</td>
<td>Frequent respiratory infections, exertional dyspnea, fatigability</td>
<td>Precordial bulge, systolic murmur and thrill left 3-4 ICS, diastolic murmur and thrill; normal height, underweight; BP 110/60</td>
<td>RVH and strain, right atrial enlargement</td>
<td>Right aortic arch, right coronary artery arose from defect</td>
</tr>
<tr>
<td>S.S.</td>
<td>10</td>
<td>M</td>
<td>Dyspnea on exertion, excess fatigability, episodes of vertigo</td>
<td>Precordial bulge, systolic murmur and thrill left 3rd ICS, no diastolic murmur, BP 110/70; markedly underdeveloped</td>
<td>LVH and strain, wandering pacemaker</td>
<td>Persistent left superior vena cava, subaortic stenosis</td>
</tr>
<tr>
<td>G.B.</td>
<td>7</td>
<td>M</td>
<td>Murmur first heard age 3 years, dyspnea on exertion and fatigability</td>
<td>Pectus carinatum, systolic murmur 2nd left ICS, systolic gallop, no diastolic murmur; markedly underdeveloped; BP 110/50</td>
<td>LVH, RVH, right ventricular conduction defect</td>
<td>None</td>
</tr>
<tr>
<td>J.S.</td>
<td>4</td>
<td>M</td>
<td>Congestive heart failure with an infection, age 1 year; ductus closed age 2 years; dyspnea and fatigability</td>
<td>Pansystolic murmur at apex, diastolic rumble; normal development; BP 140/70</td>
<td>LVH, right ventricular conduction defect</td>
<td>Patent ductus</td>
</tr>
<tr>
<td>S.D.</td>
<td>10</td>
<td>M</td>
<td>Cyanosis first evidence of heart disease, discovered at 6 years; no failure or limitation of activity</td>
<td>Slight generalized cyanosis, no clubbing; grade 2 early systolic murmur, maximal 4th left ICS, no diastolic murmur, well developed, BP 110/80</td>
<td>RVH, right axis</td>
<td>None</td>
</tr>
</tbody>
</table>
by sphygmomanometry, the femoral arterial diastolic pressure was abnormally low in four of the six patients. Patient S. D. was found to have an arterial oxygen saturation of 82 per cent and in him a large right-to-left shunt was evident on arterial indicator-dilution curves recorded after right heart and pulmonary artery injection. In the other patients arterial oxygen saturation was normal and only a left-to-right shunt was evident in similar curves.

Retrograde aortic catheterization was also performed in each patient. In four studies the aortopulmonary defect was crossed by the catheter and in three it was passed into the right ventricle as well. Pressures were recorded as the catheter tip was withdrawn across the defect and only small differences in the mean pressures in the central aorta and main pulmonary artery were evident. Indicator-dilution curves were recorded with femoral or brachial arterial sampling following injections at various sites in the aorta in five patients. By this technic the origin of the left-to-right shunt was correctly localized to the ascending aorta in each of them (fig. 4). In two patients injections of dissolved Kr\(^{85}\) were also made and by recording the appearance time of the isotope in expired air the shunt was again shown to originate from the proximal rather than the distal aorta (fig. 5).

Selective angiography was carried out in every patient at the time of retrograde aortic catheterization. The lesion was demonstrated by this means in all patients, and selected films from the angiographic studies in three patients are reproduced in figures 6, 7, and 8. In patient S. S. the

**Figure 1**

*Phonoangiograms of patients J. S. (above) and S. S. (below). S\(_1\) and S\(_2\) refer to the first and second heart sounds. P\(_2\) and A\(_2\) indicate the sounds of pulmonic and aortic valve closure. S. M., systolic murmur.*

**Figure 2**

*Preoperative (top) and postoperative (bottom) chest x-rays of patient G. B. Before operation generalized cardiac enlargement with prominence of the left atrium and both ventricles is evident and the pulmonary vascular markings are prominent. Following correction the size and configuration of the heart are normal.*

presence of a systolic pressure gradient of 85 mm. Hg within the left ventricle was demonstrated when this chamber was catheterized from the aorta and to localize the area of subaortic stenosis, as well as to demonstrate the defect, the angiocardiogram was carried out with left ventricular injection (fig. 8). In patient S. D., with predominant right-to-left shunt, the communication was best visualized in films exposed after right ventricular injection (fig. 7).
Surgical Treatment

The aortopulmonary septal defect was successfully closed by division and suture in each of the five patients in whom a large left-to-right shunt was present. Operation was not carried out in S. D. because of the evidence, obtained at catheterization, that the predominant circulatory shunt was right-to-left and that pulmonary vascular resistance exceeded systemic vascular resistance. The opening between the great vessels is a form of persistent truncus arteriosus and results from a partial failure of partitioning of the truncus arteriosus. Thus, in contrast to ductus arteriosus, the communication is one without intrinsic length. Therefore, it was not possible in any patient to isolate the defect primarily between clamps and in each division was accomplished by an open technic.

A complete median sternotomy was made in every patient. In the first two (H. C. and G. L.) operations were performed during a period of inflow occlusion after the induction of general hypothermia while in the latter three patients total cardiopulmonary bypass was utilized.

After the pericardium is incised, the adventitia is dissected from the anterior aspects of the aorta and pulmonary artery as shown in figure 9. Both vessels are freed by division of the pericardial reflections between the aorta and pulmonary artery above and below the communication; posteriorly the plane between the right pulmonary artery and the aorta is also developed (fig. 10 A). This dissection is made after the vena caval and femoral cannulations have been made, so that bypass can be instituted immediately in the event of injury to either vessel. When these preparations have been completed, bypass is begun. A curved vascular clamp is then passed behind the fistula and placed to occlude it on the aortic side as shown in figure 10 B. The main pulmonary artery is then opened anteriorly and, after the origin of the right pulmonary artery is thereby visualized, the posterior wall of the pulmonary artery is divided. The resulting opening in the main pulmonary artery usually extends into the right main branch and is closed with two rows of continuous sutures as shown in figure 11 A. The aorta is similarly closed, distal to the clamp, and this suture line is sometimes covered with Teflon felt to reinforce it (fig. 11, B and C).

When the operation was performed with hypothermia, the dissection described above was made and, after occlusion of the venae cavae, clamps were placed on the pulmonary artery side of the fistula and on the ascending aorta as shown in figure 12. The defect was then divided, allowing the aorta to be rotated to the right and drawn anteriorly, permitting the application of a third clamp to the opening in it (fig. 13). After restoration of flow the openings in the great vessels were closed. The diameters of the defects in the five patients ranged from 15 to 20 mm.

In two patients associated malformations were present at the time of operation. In G. L. the right coronary artery, which was of large size, was found to arise abnormally high and to originate not from the aorta but from the anterior wall of the pulmonary artery near this end of the defect (fig. 14). Temporary occlusion of the coro-
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in Six Patients with Aortopulmonary Septal Defect

<table>
<thead>
<tr>
<th>Inert gas, index, and site</th>
<th>Pulmonary to systemic flow ratio</th>
<th>Pulmonary to systemic resistance ratio</th>
<th>Interval months</th>
<th>Pressures, systolic/diastolic, (mean), mm. Hg</th>
<th>Preop/postop cardiothoracic ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>PA 42%</td>
<td>1.7:1</td>
<td>44%</td>
<td>8</td>
<td>(2) 37/3</td>
<td>0.64/0.55</td>
</tr>
<tr>
<td>RV 0%</td>
<td></td>
<td></td>
<td></td>
<td>(3) 105/5</td>
<td></td>
</tr>
<tr>
<td>PA 66%</td>
<td>2.8:1</td>
<td>31%</td>
<td></td>
<td>(2) 39/3</td>
<td>0.64/0.55</td>
</tr>
<tr>
<td>RV 31%</td>
<td></td>
<td></td>
<td></td>
<td>(1) 39/4</td>
<td>0.56/0.47</td>
</tr>
<tr>
<td>PA 50%</td>
<td>1.9:1</td>
<td>43%</td>
<td>13</td>
<td>(2) 39/3</td>
<td>0.64/0.55</td>
</tr>
<tr>
<td>RV 36%</td>
<td></td>
<td></td>
<td></td>
<td>(1) 39/4</td>
<td>0.64/0.55</td>
</tr>
<tr>
<td>PA 68%</td>
<td>2.8:1</td>
<td>36%</td>
<td>7</td>
<td>37/10</td>
<td>0.74/0.66</td>
</tr>
<tr>
<td>RV 39%</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PA 55%</td>
<td>2.2:1</td>
<td>42%</td>
<td>At Oper.</td>
<td>39/4</td>
<td>0.74/0.66</td>
</tr>
<tr>
<td>RV 0%</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>95/55</td>
</tr>
<tr>
<td>PA 14%</td>
<td>Large R→L and small L→R shunt by indicator dilution.</td>
<td></td>
<td></td>
<td></td>
<td>Not operated upon</td>
</tr>
<tr>
<td>RV 8%</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Results

All of the five patients did well postoperatively, striking gains in height and weight have been evidenced by all, and all are free of symptoms of cardiovascular disease. Soft systolic murmurs are still audible at the base of the heart in the three boys without residual lesions. In G. L., in whom the pulmonary artery was constricted, a systolic murmur of grade 3/6 intensity is present over the pulmonary area, and S. S. still presents the characteristic thrill and murmur of congenital aortic stenosis. No striking electrocardiographic changes were noted although decreases in electromotive force over the right precordium were evident in several of the patients. By x-ray the heart of every patient has been found significantly smaller, and the average decrease in the cardiothoracic ratio is 18 per cent.

Cardiac catheterization was carried out postoperatively in four patients and in the other patient pressure measurements and indicator-dilution curves were recorded at the conclusion of the operation. The data obtained at these studies are included in table 2. In patient G. L. a systolic gradient of 79 mm. Hg was present across the constriction in the main pulmonary artery. In the other four patients the right ventricular and pulmonary arterial pressures were found to have fallen...
Figure 3
The electrocardiogram of patient G. L. Right ventricular hypertrophy and strain as well as right atrial enlargement are evident.

Discussion
A detailed analysis of the published reports concerning aortopulmonary septal defect reveals that the malformation was first recognized by John Elliotson, the Professor of Practice at the University of London, and was described by him in a clinical lecture at St. Thomas’ Hospital in 1830. A total of 71 patients with this lesion, including those in the present report, has been described since that date. It should be noted, however, that in a number of the early papers the pathologic descriptions are unillustrated and, by modern standards, vague; it is likely that a certain number of allied malformations, such as more severe forms of persistent truncus arteriosus, are included. Similarly, eight of these patients were operated upon for patent ductus and, when a ductus was not found, a thrill in the pulmonary artery was attributed to aortopulmonary septal defect although the presence of the communication was not confirmed by pericardiotomy and inspection of the great vessels.

Aortopulmonary septal defect has been recognized from infancy to the age of 44 years but the majority of patients have been less than 20 years of age. The sex of 65 patients was noted; 37 were males and 28 females. Twenty-one patients died without attempted surgical treatment; eight of these...
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Figure 4
Indicator-dilution curves recorded with right brachial artery sampling following injections in the ascending aorta (left) and distal aortic arch (right) in patient G. B. In the curve recorded after proximal injection a small primary curve (1) results from dye passing directly into the brachial artery, a large amount of dye appears later after having been shunted through the pulmonary circulation (2) and a third peak due to systemic recirculation is also evident (3). When the injection was made in the distal aortic arch the large initial deflection indicates dye regurgitated back to the origin of the innominate artery and the small break in the descending limb of the curve (oblique arrow) represents the small fraction of dye shunted into the pulmonary circulation.

Figure 5
Time-concentration curves of Kr85 in expired air following its injection in the proximal (left) and distal aorta (right) in patient G. B. The immediate appearance of the isotope following proximal injection indicates that blood from the ascending aorta is being shunted into the pulmonary circulation.

deads were before the age of 2 years, the others at various times up to the age of 40 years. Death usually resulted from congestive heart failure but endocarditis was described in two patients and in another the pulmonary artery ruptured. The infants who died did not have defects of significantly larger size than patients surviving into later life. All patients, however, who had communications 1 cm. or larger in diameter died before the age of 20 years. Numerous other congenital malformations have been noted to occur in association with aortopulmonary septal defect. The most common of these have been patent foramen ovale, right aortic arch, and patent ductus arteriosus. Less common accompanying cardiovascular lesions have been various valvular deformities, ventricular septal defect, postductal coarctation, and the tetralogy of Fallot.

Although the physical findings in aortopulmonary septal defect are often described as...
mimicking those of patent ductus arteriosus, it is noteworthy that among the 56 patients

in whom the auscultatory findings were reported, a continuous murmur was present in only nine. The usual finding was a basal sys-
The results of cardiac catheterization have been reported in 28 patients in whom the diagnosis was subsequently confirmed by operation, autopsy, angiography, or by passage of the catheter through the defect. In the majority, the presence of a left-to-right shunt was demonstrated by oximetry but the data presented are insufficient to permit conclusions concerning the relationship between pulmonary and systemic flows and resistances. The average level of the pulmonary arterial or right ventricular systolic pressures in the 28 patients in whom it was measured was 78 mm. Hg and it exceeded 40 mm. Hg in 24 of them. In many of the patients catheterized, however, the correct diagnosis was not appre-
Figure 10

Mobilization of the aorta and right pulmonary artery (A) after the great vessels have been isolated. B. Division of the main pulmonary artery during cardiopulmonary bypass and after the placement of a clamp on the aorta.

Various methods have been utilized in the closure of aortopulmonary defects. The first correction was performed by Gross in 1948 and in this patient ligation of the communication was carried out. Scott and Sabiston were able to divide such a defect between clamps and successfully suture the openings in the great vessels. The method of division and suture during cardiopulmonary bypass was described by Cooley in 1957. There have been reports of 26 attempted corrections of aortopulmonary septal defect including the patients described herein. Ligation alone was employed in seven patients, none of whom died, but complete closure was apparently achieved in only three of them. Closure of the fistula with sutures, without division, has also been attempted in seven patients with one death; but whether or not complete closure was achieved in the survivors is difficult to ascertain. In five patients division and suture of the communication between clamps was attempted but was successfully completed in only three instances. Complete closure with survival was accomplished in all eight patients (Cooley’s three patients and the five described herein) in whom an open technique, with either hypothermia or cardiopulmonary bypass, was employed.
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Figure 11
The opening in the main pulmonary artery is closed without the placement of a clamp (A) and the repair is completed as shown in B and C.

Conclusions
From an assessment of the information furnished by the patients described, and that derived from previous reports, certain conclusions can be reached concerning the clinical, hemodynamic, and diagnostic findings and the surgical management of aortopulmonary septal defect. The malformation is a relatively unusual one, but has been encountered with increasing frequency as greater numbers of patients with congenital heart disease have been subjected to detailed study or operation. The majority of untreated patients will die during childhood or early adult life unless surgical treatment is undertaken. Contrary to the concept held by many, the murmur of aortopulmonary septal defect is rarely continuous, and a basal systolic murmur is most common. The lesion results in a large left-to-right circulatory shunt and almost invariably in pulmonary hypertension of severe degree. Irreversible pulmonary vascular changes are
When the great vessels have been separated, the aorta is drawn anteriorly and to the right, permitting the placement of a clamp across the opening in it. After restoration of flow the openings in the aorta and pulmonary artery are sutured distal to the clamps.

relatively uncommon, however, since the shunt remained left-to-right in five of six patients and in them the pulmonary artery pressure fell strikingly when the defect was closed.

The diagnosis of aortopulmonary septal defect should be suspected whenever a large shunt into the pulmonary artery is demonstrated at catheterization. The malformation can most easily be distinguished from patent ductus and persistent truncus arteriosus by the application of indicator or isotope dilution technics and selective angiography with left heart or aortic injection.

It would appear that surgical correction is indicated in most patients in whom the presence of an aortopulmonary defect is demonstrated and division and suture of the communication during cardiopulmonary bypass has been shown to be the surgical method of choice.

Summary

The clinical, hemodynamic, and angiographic findings in six patients with aortopulmonary septal defect are described. A basal systolic murmur was present in each of the patients and in none was a continuous murmur audible or recordable. Five of the patients had experienced diminished cardiac reserve and at right heart catheterization a
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large left-to-right shunt and severe pulmonary hypertension were found. In the sixth patient the predominant circulatory shunt was right-to-left. The diagnosis was established preoperatively in all patients with the aid of indicator-dilution curves, injections of Kr⁸⁵ and selective angiography.

The defect was successfully divided and closed at open operation in the five patients with predominant left-to-right shunts. Two of them were operated upon with the aid of general hypothermia and three with cardio-pulmonary bypass. Postoperatively all patients have been improved and the pulmonary artery pressure has fallen markedly. The clinical picture, hemodynamic findings, and surgical results in 65 previously reported patients are also summarized.

Acknowledgment

The authors wish to express their appreciation to Dr. Henry P. Goldberg, who referred two of the patients described and has assisted in their postoperative management.

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Figure 15

Records of left ventricular (LV) and brachial arterial (BA) pressures recorded in patient S. S. with the aortopulmonary defect patent (left) and occluded (right). A diminution in the systolic pressure gradient is seen to occur when the defect is clamped and left ventricular output thereby reduced. The systemic arterial diastolic pressure also rises markedly.

References


MORROW, GREENFIELD, BRAUNWALD


