The Surgical Treatment of Ventricular Septal Defect in Infancy

The Technic and Results of Pulmonary Artery Constriction

By Andrew G. Morrow, M.D., and Nina S. Braunwald, M.D.

In patients with ventricular septal defect who are more than 3 or 4 years of age, an open corrective operation may be carried out with minimal risk if the lesion is not complicated by the presence of severe pulmonary hypertension. With surgical techniques presently available, however, the closure of a ventricular septal defect in an infant or very young child is associated with considerable hazard. In 41 children under the age of 2 operated upon by Cooley the mortality was 39 per cent and in Kirklin’s experience in 1956, quoted by Keith, 70 per cent of children in this age group died after open repair. Thus, it seems clear that elective operations for ventricular septal defect should, whenever possible, be deferred until the child has reached optimal age and size.

Although isolated ventricular septal defect is generally considered to be one of the more benign congenital cardiovascular malformations, a certain proportion of infants with this lesion develop severe symptomatology and in this group the early mortality is high. Keith, for example, observed a series of 111 symptomatic children with ventricular septal defect; more than one third died in the first year of life. Seventeen patients who evidenced heart failure were reported by Morgan et al. and, of these, 10 died before the age of 1 year. Marquis found that ventricular septal defect was the most common cause of death from congenital heart disease among children less than 3 years of age.

It is thus apparent that a large number of infants with ventricular septal defect will not survive with nonoperative management and yet should not be subjected to corrective operation because of the prohibitive risk associated with this procedure at the present time. Death in such infants usually results from congestive heart failure often complicated by repeated and severe pulmonary infections. Both are clearly attributable to excessive pulmonary blood flow. Muller and Dammann reasoned that the artificial production of pulmonic stenosis should be beneficial, since children with ventricular septal defect and pulmonary stenosis (e.g., tetralogy of Fallot) ordinarily do well through early childhood. A modification of the operative procedure described by Muller and Dammann has been utilized at the National Heart Institute in the surgical treatment of 13 infants with ventricular septal defect. The criteria utilized in selecting patients for pulmonary artery constriction, the operative method employed, and the results of the procedure are described in the present report.

Clinical and Hemodynamic Findings

The 13 children ranged in age from 3 to 22 months (mean 6.5 months) and weighed from 2.6 to 9.8 Kg. (mean 4.7 Kg.). Each had failed to grow and develop normally, and the weight of every patient was below the third percentile in comparison with normal children of the same age and sex. All had experienced repeated episodes of pneumonia and congestive heart failure and the majority of the children had spent most of their lives in hospitals. All were receiving digitalis when first seen. On physical examination they appeared chronically ill and markedly underdeveloped. Gross cardiac enlargement was evident in each, and the typical precordial systolic thrill and murmur of ventricular septal defect were present. The second heart sound in the pulmonary area was always accentuated.

Right heart catheterization was carried out
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Table 1
Summary of Hemodynamic Data in 13 Children Undergoing Pulmonary Artery Constriction*

<table>
<thead>
<tr>
<th>Patient</th>
<th>Preoperative catheterization</th>
<th>Operative pressures after constriction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>Weight (Kg.)</td>
<td>PA* S/D mean</td>
</tr>
<tr>
<td>R.B.</td>
<td>3.9</td>
<td>75/10, 45</td>
</tr>
<tr>
<td>C.C.</td>
<td>5.6</td>
<td>60/10, 28</td>
</tr>
<tr>
<td>S.D.</td>
<td>5.1</td>
<td>85/40, 60</td>
</tr>
<tr>
<td>R.F.</td>
<td>4.2</td>
<td>65/26, 40</td>
</tr>
<tr>
<td>J.M.</td>
<td>9.2</td>
<td>60/20, 40</td>
</tr>
<tr>
<td>L.R.</td>
<td>2.6</td>
<td>—</td>
</tr>
<tr>
<td>W.V.</td>
<td>9.8</td>
<td>55/24, 30</td>
</tr>
<tr>
<td>K.W.</td>
<td>4.8</td>
<td>50/16, 40</td>
</tr>
<tr>
<td>J.P.</td>
<td>3.6</td>
<td>105/55, 80</td>
</tr>
<tr>
<td>F.P.</td>
<td>3.5</td>
<td>75/20, 50</td>
</tr>
<tr>
<td>J.J.</td>
<td>3.5</td>
<td>65/25, 40</td>
</tr>
<tr>
<td>S.G.</td>
<td>3.2</td>
<td>100/60, 68</td>
</tr>
<tr>
<td>R.M.</td>
<td>2.7</td>
<td>40/15, 20</td>
</tr>
</tbody>
</table>

*PA, RV, and FA refer to pulmonary arterial, right ventricular, and femoral arterial pressures, respectively. The systolic (S), diastolic (D), and mean pulmonary arterial pressures are listed. All pressures are in mm. Hg.
†Operative pressure measurement, no preoperative catheterization.

in 12 of the 13 children; these data are summarized in Table 1. In every instance the presence of a left-to-right shunt entering the right ventricle was demonstrated by the nitrous oxide test, serial blood oxygen determinations, or indicator-dilution curves. Pulmonary arterial and right ventricular hypertension was uniformly found, and the ratio between the pulmonary arterial and systemic arterial pressures was 66 per cent or more in nine of the 10 patients in whom both measurements were made.

In every patient it was the opinion of the staff and the referring pediatrician that unless immediate operative treatment was undertaken the infant would not survive to an age at which the ventricular septal defect could be closed.

Operative Technic

Anesthesia is ordinarily induced with cyclopropane and maintained with nitrous oxide and oxygen while respiration is controlled with succinylcholine. The saphenous vein is cannulated at the ankle for the administration of fluids and blood. The patient is placed on the table in the supine position with the left side slightly elevated. A left anterolateral thoracotomy through the fourth intercostal space is employed (fig. 1). The lung is first retracted anteriorly and the region of the ductus arteriosus dissected. It is usually difficult to determine the patency of a small ductus but the structure is always ligated. The pericardium is then incised anteriorly, exposing the main pulmonary artery and right ventricle. The pleural reflection between the aorta and main pulmonary artery is incised, and the pulmonary artery is freed until an angled clamp can be passed beneath it. A strip of Nylon cloth 1 cm. in width is then drawn about the vessel. Control pressure measurements are then made in the right ventricle and pulmonary artery by means of 20-gauge needles attached through sterile connecting tubes to equisensitive Statham pressure transducers. The cloth band is then tightened with a clamp until a vigorous thrill is palpable distal to it (fig. 2). While this degree of constriction is maintained by the clamp, the right ventricle and pulmonary arterial pressures are again measured. The constriction is adjusted until the mean pulmonary artery pressure is reduced to approximately 20 to 30 mm. Hg. The systolic gradient between the right ventricle and distal pulmonary artery will ordinarily be 30 to 50 mm. Hg at this time. When the desirable degree of constriction is achieved the heart action is observed for several minutes and if no detrimental effect is apparent the tape is sutured to itself beneath the clamp (fig. 3). Care must be taken that the sutures do not pierce the wall of the pulmonary artery. Following completion of the constriction, confirmatory pressure measurements are again made. The final operative pressure measurements

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in all patients are listed in table 1, and representative tracings obtained in patients C.C. and S.G. are reproduced in figures 4 and 5. Postoperative management is usually not difficult. The usual precautions must be taken to insure that tracheobronchial secretions are evacuated and oral feedings are begun 12 hours after operation.

Results

Of the 13 infants with ventricular septal defect subjected to pulmonary artery constriction 12 are living and well. The one death occurred in patient P.P. She appeared to tolerate the operative procedure well but died suddenly 36 hours afterward. It was considered likely that she aspirated a feeding but this could not be established at autopsy and no specific cause for her death was apparent. The presence of a large ventricular septal defect was confirmed at this examination.

The 12 surviving patients have been followed for periods of 9 months to 4 years. With one exception each has shown striking improvement in growth and development and all have increased exercise tolerance. The weight of each patient before operation and at successive clinic visits has been plotted by percentiles according to the anthropometric charts constructed at the Children's Medical Center, Boston. These data for the 10 children who have been followed for 20 months or longer are reproduced in figure 6. With the exception of patient R.M. each has shown excellent weight gain. Improvement following operation has occurred at a variable rate but in general maximum benefit has become apparent only after a period of 6 to 12 months.

Clinical evidences of congestive heart failure have disappeared in every patient postoperatively, and the administration of digitalis has been discontinued in the majority of them. The children have all experienced upper respiratory infections but none has had pneumonia. It was considered that cyanosis might become apparent as the children became older but this has not been evident in any thus far. Every patient continues to have the murmur of ventricular septal defect and in addition a loud ejection murmur and often a thrill are present over the pulmonary area. The hearts of all patients are still enlarged on x-ray examination. None has, as yet, been catheterized postoperatively.

Clinical Example

A brief clinical summary of patient R.F. is presented to illustrate the typical findings and the course of a patient subjected to pulmonary artery constriction.

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Figure 3

When the optimal degree of constriction has been achieved the band is sutured to itself beneath the clamp and the pressures are again measured. The degree of constriction can be increased, if necessary, by the placement of additional sutures.

R.F. (02-01-10) was a male infant whose birth weight was 2.5 Kg. He was apparently well until the age of 2½ months, when severe dyspnea with feedings noted and he developed a chronic cough. At this time a heart murmur was heard and congestive heart failure was evident. He was said to have been cyanotic on several occasions and had been hospitalized repeatedly for severe respiratory infections. He had gained weight poorly since birth and when admitted to the National Heart Institute at the age of 5½ months he weighed only 3.3 Kg.

On examination he was obviously cæchetic (fig. 7A). The heart was enlarged, and a systolic thrill was felt in the third left intercostal space. The second heart sound in the pulmonary area was loud and a grade 3/6 systolic murmur was audible all over the precordium and was loudest along the left sternal border. There were moist rales at both lung bases and the liver was palpable 4 cm. below the costal margin. By x-ray the heart was seen to be grossly enlarged and the pulmonary vascularity was markedly increased. The electrocardiogram revealed right axis deviation and right ventricular hypertrophy. At right heart catheterization (table 1) there was found to be severe right ventricular and pulmonary arterial hypertension. Nitrous oxide indices of 66, 65, and 8 per cent in the pulmonary artery, right ventricle, and right atrium, respectively, indicated that a large left-to-right shunt entered the right ventricle.

The child was considered to be an ideal candidate for pulmonary artery constriction and the operation was performed in December 1957. A ductus 2 mm. in diameter, and questionably patent, was found and ligated. The pulmonary artery was constricted until the mean pressure distally was reduced to 30 mm. Hg and a systolic gradient of similar magnitude had been produced. Postoperatively his course was smooth and he immediately began to take much larger feedings; in the first 12 postoperative weeks he gained 1.1 Kg. After 1 year his development and weight gain continued to be rapid (fig. 7B) and by the age of 2½ years both his height and weight were greater than the fiftieth percentile on the anthropometric chart. He has never evidenced cyanosis, dyspnea, or fatigability on unrestricted activity. The typical ejection murmur of pulmonic stenosis is audible. The electrocardiogram and relative size of the heart x-ray have remained essentially unchanged, although some decrease in pulmonary vascularity is apparent.

Discussion

The prognosis in a newborn infant with a ventricular septal defect is, in general, dependent upon the size of the communication and the pulmonary vascular resistance. If the defect is small the shunt and the resultant extra burden on the left ventricle will usually not be sufficient to cause symptoms or failure. If, on the other hand, the defect is of large size and approximates the aortic orifice, it will offer little resistance to the flow of left ventricular blood, and the magnitude of the shunt is limited only by the resistance in the pulmonary vascular bed. If the circulation of an infant with a large ventricular septal defect can adjust to the increased pulmonary blood flow soon after birth, presumably by the
development of increased pulmonary vascular resistance, survival is possible. If such adjustment is not made failure will certainly ensue and the risk of death within the first months of life will be high. The operation of pulmonary artery constriction artificially produces increased resistance to right ventricular ejection but does so proximally and in a reversible manner. It is likely that the medial hypertrophy of the pulmonary arterioles is related to increased pulmonary blood flow and high pulmonary arterial pressure. Thus pulmonary artery constriction may also be found helpful in preventing or ameliorating the development of these changes, considered by some to be irreversible.

When operative treatment is contemplated in an acyanotic infant presenting congestive failure, first consideration must be given to the establishment of the correct diagnosis. If the shunt is primarily into the right atrium rather than into the right ventricle, as in the incomplete form of persistent atrioventricular canal, pulmonary artery constriction will be of little benefit, since reduction of the magnitude of the shunt could result only at the expense of right ventricular failure with elevation of the right ventricular end-diastolic and right atrial pressures. These considerations may explain why patient R.M. has not experienced so great benefit as the other patients. In addition to a ventricular septal defect he also was shown to have an intratral communication and a moderate-sized shunt into the right atrium. Thus cardiac catheterization would seem to be indicated preoperatively in every patient. When the diagnosis of ventricular septal defect is made by catheterization, the results of the study are usually of little further aid in determining the necessity for pulmonary artery constriction, and this decision must rest largely on clinical considerations.

The selection of patients for the operation is usually not difficult. The procedure ordinarily is indicated in an infant with ventricular septal defect under the age of 12 to 18 months when weight gain is poor, or at a standstill, and when failure and infection have not responded to intensive pediatric management over a prolonged period of hospitalization. When failure responds to digitalization and respiratory infections can be effectively controlled by the use of antibiotics, operative treatment can likely be deferred until an open corrective procedure can be performed with relative safety.

It has been suggested that the correct degree of constriction of the pulmonary artery can be determined by inspection alone. In our experience the mere appearance of the narrowed segment may be quite misleading, and physiologic control has been found to be essential. When the first pulmonary artery constrictions were performed in this clinic, attempts were made to regulate the degree of constriction not only by the measurement of proximal and distal pressures but also by ear oximetry. It was considered that optimal constriction might be the point at which a
small right-to-left shunt first occurred, signaled by a slight reduction in peripheral arterial oxygen saturation. This technic was found unsatisfactory and was abandoned because of the great respiratory variations recorded by the oximeter; pressure measurements alone are now used.

The immediate object of the operation is to reduce pulmonary blood flow, and this of course is necessarily accompanied by a fall in pulmonary arterial pressure. Since the flow through the pulmonary vascular bed is directly proportional to the difference between the pulmonary arterial and left atrial pressures, the reduction in flow can easily be estimated. In the present series the average reduction in mean pulmonary arterial pressure was about 15 mm. Hg and the average reduction in pulmonary flow was approximately 33 per cent. To accomplish a constriction of this degree, which by experience seems satisfactory, the area of the lumen of the artery must be narrowed to nearly one third of its original diameter. It should be emphasized that the pressure measurements must be made while the action of the heart is vigorous. Often in the course of the operation, particularly when the pulmonary artery is being freed or when the band is initially tightened, the heart will slow and its contractions become feeble. Under these circumstances small doses of intracardiac calcium chloride (0.5 to 1.0 ml. of 10 per cent solution) ordinarily restore vigorous heart action, and this drug was necessary in many of the patients described. The above considerations would seem to indicate that the pulmonary artery, under optimal conditions, should be narrowed sufficiently to effect a reduction of 30 to 40 per cent in the mean pulmonary arterial pressure. This will ordinarily result in a systolic gradient between the ventricle, and distal pulmonary artery of about 50 mm. Hg and the diameter of the artery will be reduced by about two thirds.

The operation described is, of course, a palliative one and a later definitive procedure will be necessary. Both experimental studies and isolated clinical reports indicate that the stenosis produced by constriction of the pulmonary artery by a cloth band can be
relieved without undue difficulty at the time the ventricular septal defect is closed.

The gratifying clinical benefit evidenced by the children described in this report is reinforced by similar results in other clinics.10-12 It would seem that pulmonary artery constriction will remain the surgical procedure of choice in infants with ventricular septal defect and heart failure until the techniques of the open operation are sufficiently refined to permit an initial corrective operation at an acceptable risk.

Summary

Thirteen infants with ventricular septal defect, pulmonary hypertension, and severe congestive heart failure were treated by pulmonary artery constriction. The 12 children who survived operation have been followed from 1 to 4 years and have evidenced striking improvement in growth, weight gain, and exercise tolerance. None has had pneumonia or heart failure after operation. Considerations in the selection of patients for pulmonary artery constriction, the surgical method employed, and its hemodynamic control are described.

References


The effect of infused fluids in producing the morbid sound is at once proved by the injection of water into the thorax of a dead body; in which case it will be found that the sound elicited by percussion will be obscure over the portion of the cavity occupied by the injected liquid.—From On Percussion of the Chest. Published in 1761. Translated by John Forbes, M.D. In: Classics of Medicine and Surgery. New York, Dover Publications, Inc., 1959, p. 129.
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ANDREW G. MORROW and NINA S. BRAUNWALD

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