Pulmonic Stenosis with Intact Ventricular Septum

Treatment Utilizing Extracorporeal Circulation

By Dwight C. McGoon, M.D., and John W. Kirklin, M.D.

Pulmonic stenosis unassociated with a ventricular septal defect of significant size is often a complex abnormality. The stenosis may be valvar, infundibular, or both, and may be associated with an atrial septal defect or even with a small ventricular septal defect. Preoperative studies cannot accurately predict these variations. The surgical approach selected for such patients should permit the correction of each of the cardiac defects encountered. That this can be accomplished by means of extracorporeal circulation and open cardiotomy is demonstrated by the presented series of 10 cases without operative mortality.

PULMONIC stenosis may be valvar, sub-valvar, or both valvar and subvalvar. Although often occurring as an isolated defect, it may be associated with certain other intracardiac abnormalities. These several possible combinations of defects have resulted in difficulties of classification and in discrepancies of terminology.

Definitions

To a considerable degree, matters of classification and terminology are arbitrary, but it is important that all terms be clearly defined. The distinction between valvar pulmonary stenosis and subvalvar or infundibular pulmonic stenosis is now generally recognized.

In the tetralogy of Fallot, there is, in addition to pulmonic stenosis, a ventricular septal defect of a size that approximates that of the aortic orifice. At the opposite extreme is the heart in which there is pulmonic stenosis and an intact ventricular septum. Difficulty in classification is posed by the heart with pulmonic stenosis, but with a very small ventricular septal defect. Since patients with this combination seen at the Mayo Clinic have clinically resembled those patients with pulmonic stenosis and intact ventricular septum, it appears wise from a practical standpoint to classify these in the same group. They might be designated as having "pulmonic stenosis with essentially intact ventricular septum." Efforts to distinguish these hearts from those with so-called "tetralogy of Fallot" on the basis of the presence or absence of dextroposition of the aorta, or the nature of the pulmonic stenosis, have not been satisfactory.

The present study deals with pulmonic stenosis either with an intact ventricular septum or with a very small ventricular septal defect. In this type of cardiac deformity there also may or may not be an intracardiac communication that permits usually a right-to-left shunt, and less commonly a left-to-right shunt.1 2

Characteristics of the Pulmonic Stenosis

It is believed of primary importance to re-emphasize that infundibular stenosis is frequently encountered in those patients having pulmonic stenosis with an intact ventricular septum. Previously, several reports4 5 have been made of patients having isolated valvar pulmonic stenosis and an intact ventricular septum, with the apparent exclusion of any such patients also having infundibular pulmonic stenosis. The misconception might thereby arise that in all such patients the pulmonic stenosis is exclusively valvar. On the other hand, even the nonsurgical literature reports an incidence of infundibular stenosis as high as 14% to 25 per cent8 in these patients, either alone or in combination with valvular stenosis; in certain surgical series where infundibular stenosis has been looked for carefully, an equally high or higher incidence has been found.9 10 Infundibular stenosis occurred with similar frequency in our total experience with the indirect technic of pulmonic valvotomy.

Thirty-two patients classified as having

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TABLE 1.—Incidence of Association of Valvular and Infundibular Stenosis in Pulmonic Stenosis with Presumably Intact Ventricular Septum

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Valvular stenosis unassociated with significant infundibular stenosis</th>
<th>Combined valvular and significant infundibular stenosis</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Patients</td>
<td>Per cent of total</td>
<td>Patients</td>
</tr>
<tr>
<td>Less than 15</td>
<td>14</td>
<td>78</td>
<td>4</td>
</tr>
<tr>
<td>15 or more</td>
<td>5</td>
<td>36</td>
<td>9</td>
</tr>
<tr>
<td>All ages</td>
<td>19</td>
<td>59</td>
<td>13</td>
</tr>
</tbody>
</table>

Pulmonic stenosis with intact ventricular septum have undergone operation at the Mayo Clinic by the closed technic with transventricular approach to the pulmonic valve. The first 12 have been previously reported in detail. In 13 of the 32 patients (41 per cent) the pulmonic stenosis was combined with significant infundibular stenosis (table 1), and in 5 of these 13 the infundibular stenosis was severe.* Numerous additional authors have recognized the occurrence of infundibular stenosis in this type of cardiac anomaly. It should be realized, however, that only pressure determinations before and after pulmonic valvotomy can accurately detect certain associated infundibular stenoses that might otherwise have been overlooked.

Recently a distinction has been recognized between the infundibular stenosis as found in this group having an intact ventricular septum and the infundibular stenosis associated with a large ventricular septal defect. The possibility has been suggested that muscular hypertrophy of the right ventricular outflow tract, secondary to pulmonic valvular stenosis, may cause obstruction from its own bulkiness and sphincterlike action, and thus perpetuate a vicious cycle of stenosis, hypertrophy, and more stenosis.

Furthermore, it is of interest that infundibular stenosis in association with valvular pulmonic stenosis apparently occurs with greater frequency in adults than in children, suggesting that time is required for the development of obstructing muscular hypertrophy of the infundibulum. This is again demonstrated by our series of patients operated on by the closed approach. As shown in table 1, the incidence of significant infundibular stenosis in addition to valvular stenosis increased from 22 per cent in children to 64 per cent in patients 15 years of age and older. Yet the infundibular stenosis encountered in this group of patients cannot be entirely the result of secondary muscular hypertrophy, for it may occur, as in cases 2 and 6 in table 2, in hearts with normal pulmonary valves.

Feasibility of Complete Surgical Correction

Pulmonic stenosis with essentially intact ventricular septum may thus consist of valvular or of infundibular stenosis, or of both, and may be associated with an atrial septal defect, or a small ventricular septal defect, or both. If it can be demonstrated that all these abnormalities are correctable with little or no additional operative risk, obviously this should be preferable to only partial correction.

A total of 10 patients with this condition have been operated on by means of extracorporeal circulation and open cardiotomy at the Mayo Clinic prior to January 1, 1957. A summary of the essential data in these cases is presented in table 2.

All patients were free of peripheral edema at the time of operation. All had a loud coarse systolic murmur and a palpable thrill in the pulmonic area. The second pulmonic sound was obscure or absent in all except case 8. The electrocardiograms and roentgenograms showed alterations commensurate with the anatomic and hemodynamic findings.

It should be noted from table 2 that the preoperative right ventricular systolic pressures ranged from 83 to 227 mm. Hg and that the systolic pressure gradient between the right ventricle and the pulmonary artery ranged from 52 to 210 mm. Hg. At operation the pulmonic stenosis was found to be exclusively valvular in only 2 patients and entirely infundibular in 3 patients; both infundibular and valvular stenosis were found in the remaining 5 patients.

* Infundibular stenosis is considered significant when the low right ventricular systolic pressure is more than 20 mm. Hg greater than that in the infundibular zone, and severe when it is more than 40 mm. Hg greater.
<table>
<thead>
<tr>
<th>Case, sex, age (years)</th>
<th>N.Y. heart class</th>
<th>Cyanosis</th>
<th>Preoperative heart catheterization</th>
<th>Operative diagnosis</th>
<th>Atriotomy for repair of atrial septal defect</th>
<th>Pulmonary arteriotomy for valvotomy</th>
<th>Ventriculotomy for resection of infundibular stenosis</th>
<th>Postcorrection systolic pressure at operation (mm. Hg)</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1, F, 44</td>
<td>III D</td>
<td>+</td>
<td>Rt. ventricle, systolic pressure (mm. Hg) 208 198 12 31 84</td>
<td>Valvular and infundibular stenosis; probe-patent foramen ovale</td>
<td>0</td>
<td>+</td>
<td>60 25</td>
<td>Died 3 mo. p.o. after saddle embolus</td>
<td>Excellent</td>
</tr>
<tr>
<td>2, F, 11</td>
<td>III D</td>
<td>+</td>
<td>Rt. ventricle, systolic pressure (mm. Hg) 180 19 19</td>
<td>Infundibular stenosis, probe-patent foramen ovale</td>
<td>0</td>
<td>0</td>
<td>20 20</td>
<td>Excellent</td>
<td></td>
</tr>
<tr>
<td>3, F, 33</td>
<td>III D</td>
<td>+</td>
<td>Pulmonary artery, systolic pressure (mm. Hg) 210 178 13 25 85</td>
<td>Valvular and infundibular stenosis; 8-mm. defect in foramen ovale</td>
<td>+</td>
<td>0</td>
<td>50 20</td>
<td>Excellent</td>
<td></td>
</tr>
<tr>
<td>4, M, 10</td>
<td>II B</td>
<td>0</td>
<td>Rt. to-left shunt, per cent (O₂ saturation data) 98 28 34 0* 98</td>
<td>Infundibular stenosis; bicuspid nonobstructing pulmonic valve; ventricular septal defect 5 mm.</td>
<td>0</td>
<td>0</td>
<td>49 24</td>
<td>Excellent</td>
<td></td>
</tr>
<tr>
<td>5, F, 8</td>
<td>III C</td>
<td>+</td>
<td>Peripheral arteriovenous saturation, per cent 175</td>
<td>Valvular and infundibular stenosis; atrial septal defect 8 mm. and 2 additional fenestrations</td>
<td>0</td>
<td>0</td>
<td>30 14</td>
<td>Excellent</td>
<td></td>
</tr>
<tr>
<td>6, F, 39</td>
<td>II C</td>
<td>0</td>
<td>Ventricular septal defect 4 mm. 153</td>
<td>Infundibular stenosis; ventricular septal defect 4 mm.</td>
<td>0</td>
<td>0</td>
<td>22 —</td>
<td>Excellent</td>
<td></td>
</tr>
<tr>
<td>7, M, 40</td>
<td>II C</td>
<td>+</td>
<td>Ventricular septal defect 2 small openings in foramen ovale 97 62 18 6 94</td>
<td>Valvular and infundibular stenosis; 2 small openings in foramen ovale</td>
<td>+</td>
<td>+</td>
<td>—</td>
<td>—</td>
<td>Excellent</td>
</tr>
<tr>
<td>8, F, 4</td>
<td>I B</td>
<td>0</td>
<td>Atrial septal defect, 30 × 15 mm.; valvular pulmonic stenosis 83 31 0* 99</td>
<td>Valvular and infundibular stenosis; 2 small openings in foramen ovale</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>25 20</td>
<td>Excellent</td>
</tr>
<tr>
<td>9, M, 14</td>
<td>III C</td>
<td>+</td>
<td>Ventricular septal defect 10 mm. and 3 or 4 fenestrations 121</td>
<td>Valvular and infundibular stenosis; atrial septal defect 10 mm. and 3 or 4 fenestrations</td>
<td>+</td>
<td>+</td>
<td>+†</td>
<td>35 25</td>
<td>Excellent</td>
</tr>
<tr>
<td>10, M, 38</td>
<td>IV D</td>
<td>0</td>
<td>Valvular-pulmonic stenosis 227 17 0 93.4</td>
<td>Valvular-pulmonic stenosis</td>
<td>0</td>
<td>+</td>
<td>45 35</td>
<td>Excellent</td>
<td></td>
</tr>
</tbody>
</table>

* These 2 patients had significant left-to-right shunts.
† An Ivalon diamond-shaped prosthesis in outflow tract incision.
Small atrial septal defects, located in the foramen ovale, were present in 3 instances, and each was closed through an open right atriotomy. Two patients had a relatively large atrial septal defect, each of which was similarly closed. In addition, 2 instances of a probe-patent but valvular-competent foramen ovale were noted but were not treated.

Very small ventricular septal defects were present in 2 hearts; in both of these the pulmonic stenosis was infundibular, without valvular obstruction. In case 4, the defect lay just inferior to the infundibular stenosis, and in case 6, just superior to it. Both ventricular septal defects were closed through the open right ventriculotomy.

Each of the 7 stenotic pulmonic valves could be precisely opened under direct vision. In 6 cases the valve was approached through an incision in the main pulmonary artery, as shown in the inset of figure 1. The 1 remaining stenotic valve was incised through the right ventriculotomy.

Resection of the infundibular stenosis under direct vision was performed in each case through a right ventriculotomy. Of extreme interest is the fact that during 2 of the 5 operations (cases 1 and 5) on hearts with associated infundibular and valvular stenosis, the presence of the infundibular stenosis was not suspected during the pulmonic valvotomy through the pulmonary artery. Only after normal blood flow through the heart and lungs had been resumed, and extracorporeal circulation had been discontinued, did measurements of right ventricular and pulmonary arterial pressures
demonstrate infundibular stenosis. It was then necessary to re-establish extracorporeal circulation, open the right ventricle, and resect the infundibular stenosis.

In the ninth patient of this series, the infundibular stenosis was subvalvular and could not be relieved by simple excision of redundant muscular and endocardial tissues, and thus resembled the condition in a number of patients previously encountered in our tetralogy series. This patient was treated by a technic that has been employed in certain patients with tetralogy. Even after excision of all possible redundant infundibular tissues, it became obvious that closure of the infundibular portion of the right ventricular incision would result in residual infundibular stenosis. Consequently this portion of the incision was not closed, but rather hemostasis and continuity of the right ventricular wall were accomplished by the insertion of a diamond-shaped piece of compressed Ivalon. This was sutured carefully about its periphery to the epicardial edge of the ventriculotomy wound, as depicted in figure 1. The technic is somewhat analogous to the pyloromyotomy that is employed in the treatment of hypertrophic pyloric stenosis.

Especially to be noted from table 2 are the pressure measurements recorded at operation following relief of the pulmonic stenosis. The systolic pressure gradients between the right ventricle and the pulmonary artery range from 0 to 35 mm. Hg, with an average of 16 mm. Hg.

There were no operative deaths in the series, and the only serious postoperative complication occurred in the first patient. She died 3 months after cardiac operation, shortly after the removal of an acute saddle embolus of the aorta. Just prior to this event she had made rapid progress following a stormy convalescence.

The remaining 9 patients have had excellent results. They were dismissed from the hospital 10 to 18 days after operation. Only 1 diastolic murmur (limited to early diastole) has been detected postoperatively, and indeed, 2 cases (cases 2 and 6) have shown complete absence of all murmurs. The remainder show systolic pulmonic murmurs. The electrocardiographic pattern of right bundle-branch block, complete or incomplete, was consistently encountered postoperatively.

Technic of Operation

The surgical technic employed in these patients requires little description beyond that previously presented. The basic principles and technics for whole-body perfusion utilized for these operations have been described.20, 21

Atrial septal defects are closed through a right atriotomy. The stenotic pulmonic valve is repaired through an incision in the main pulmonary artery by radial incisions made with scissors to produce a tricuspid or bicuspid valve (fig. 1).

Ordinarily, infundibular stenosis may be relieved simply by excision of redundant infundibular muscle and thickened endocardium with the scalpel and scissors. As in 1 instance in the series reported here, however, the insertion of an Ivalon prosthesis to relieve a high infundibular stenosis may be required.

There are, of course, many details that require attention in the operative and postoperative management of these patients, perhaps the most important of which are accurate blood replacement and frugality in the administration of fluids.

Discussion

Three general types of surgical technic for the relief of pulmonic stenosis with intact ventricular septum are currently applicable, namely, an indirect approach, a direct approach with use of hypothermia, and a direct approach with use of extracorporeal circulation. The first utilizes an indirect or blind access to the valvular stenosis, either by way of the right ventricle as introduced by Brock and by Sellors, or by way of the pulmonary artery as introduced by Pettersson. Cutting and dilating instruments are passed through the stenotic pulmonic valve. The low mortality rate of this procedure and the excellent postoperative subjective improvement are gratifying.

However, objective postoperative studies have shown less than the desired reduction of the pressure gradient between right ventricle and pulmonary artery in the majority of these cases. The prognostic significance of this result will not be fully appreciated until prolonged follow-up studies are available. Furthermore, associated anomalies, including atrial septal defect, ventricular septal defect,
and infundibular stenosis, cannot be readily repaired when this technic is used.

In our experience with this indirect procedure it has been particularly those patients exhibiting cyanosis preoperatively who have had the less satisfactory results. A mortality rate of 36 per cent of the 14 patients in the cyanotic group is compared to 0 per cent of the 18 in the acyanotic group. Even significant reduction of right ventricular pressures, resulted only in 7 of the 9 surviving cyanotic patients, in a satisfactory relief of cyanosis in only 3. For these reasons we have now abandoned the indirect approach to the pulmonary valve in patients who are cyanotic.

Swan and his associates have presented their experience in which hypothermia and circulatory arrest are instituted to enable direct-vision valvuloplasty through a pulmonary arteriotomy. In their reported series, a “total cure” was accomplished in 11 of the 12 patients—an excellent record.

An argument might be made against the use of hypothermia in the treatment of pulmonic stenosis, however, because satisfactory infundibular resection by this technic would seem both difficult and dangerous. Since, as shown, infundibular stenosis is frequently encountered in pulmonic stenosis with an intact ventricular septum and often cannot be accurately predicted preoperatively, one must accept the possibility of incomplete relief of pulmonic stenosis in this sizable group if the technic employing hypothermia is used.

It is true that in our experience and that of others, in some patients infundibular stenosis that was demonstrated at cardiac catheterization during the first few weeks after valvotomy regressed completely in the 12 to 18 months after operation. Nonetheless, there is evidence that severe residual infundibular stenosis adds to the hazards of the postoperative period, and it is on this account that one may have some concern as to the routine choice of hypothermia for the management of these cases.

The third approach to the surgical treatment of these patients utilizes extracorporeal circul-

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* A residual right-to-left shunt of less than 15 per cent, or a resting arterial oxygen saturation of more than 92 per cent.

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SUMMARY

Pulmonic stenosis with presumably intact ventricular septum is frequently a complex abnormality, for the stenosis may be valvular or infundibular and may be associated with an atrial septal defect or even with a small ventricular septal defect.

The feasibility of complete correction of these several possible associated cardiac abnormalities, by means of extracorporeal circulation and open cardiotomy, is exemplified by the presented series of 10 cases without operative mortality, in 9 of which an excellent result was obtained. One patient died 3 months after operation.

SUMMARIO IN INTERLINGUA

Stenose pulmonic con septo ventricular apparentemente intacte es frequentemente un anormalitate complexe, proque le stenose pote esser valvular o infundibular e pote esser associate con un defecto atrio-septal o mesmo con un parve defecto ventriculo-septal.

Le practicabilitate del correction complete de iste varie anormalitates cardiac de occurrentia associate possibile, effectuate per medio de circulatio extracorporee e cardiotomia aperte, es exemplificate per le hic
REFERENCES


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