The Reopened Ventricular Septal Defect
A Syndrome Following Unsuccessful Closure of Interventricular Septal Defects Particularly in Association with Infundibular Stenosis

By Harold W. March, M.D., Frank Gerbode, M.D., and Herbert N. Hultgren, M.D.

Since the first open intracardiac repair of interventricular septal defects and tetralogy of Fallot, rapid progress has been made in operative techniques and in extracorporeal perfusion technology. Operative risk has steadily declined, and recent communications indicate that in ventricular septal defects uncomplicated by pulmonary hypertension the mortality rate is approximately 2 per cent. In tetralogy of Fallot the mortality risk is in the vicinity of 12 to 16 per cent, but this incidence is also diminishing.

The results after successful closure of a ventricular septal defect and the correction of Fallot’s tetralogy are gratifying. Symptoms disappear or are markedly alleviated. The precordium becomes quiet, and the typical murmurs cease or are dramatically modified. Cyanosis disappears. Radiographically, there may be a reduction in heart size, and the plethora of the lung fields regresses. It soon became apparent, however, that the therapeutic benefits of complete closure were not always achieved because of disruption of the repair, and the technic was refined. Although the first disruptions were reported in attempted closure of uncomplicated ventricular septal defects and continue to be described in more recent reports pertaining to the treatment of this lesion, it has been the experience in this laboratory that the persistence of residual defects after surgery is rare in uncomplicated ventricular septal defects but that it is particularly a problem in the repair of tetralogy of Fallot. Similar opinions have been expressed elsewhere, and we believe that this point deserves particular emphasis; for the clinical consequences of incomplete or impermanent closure of a ventricular septal defect, during a procedure in which infundibular stenosis has been successfully relieved, are appreciable and at times alarming.

In some instances the left-to-right shunt through the lungs may be huge, and both the pulmonary circulation and left ventricle are presented abruptly with unaccustomed loads. Two deaths after progressive hypotension following repair for tetralogy of Fallot have been ascribed, at least in part, to incomplete ventricular septal defect repair. It is the purpose of this paper to describe more fully the clinical and hemodynamic consequences of unsuccessful closure of ventricular septal defect, particularly in association with infundibular stenosis, and to discuss the modifications of surgical technics that show promise of minimizing the incidence of unsuccessful closure.

Material and Methods
The material of the study consists of seven patients who underwent open intracardiac repair at Stanford University Hospital between March 1956 and December 1959. Six of the seven patients had ventricular septal defects complicated by infundibular stenosis or a hypertrophied crista supraventricularis. The seventh patient had an uncomplicated ventricular septal defect. All patients had extensive preoperative clinical and catheterization studies. Postoperative clinical information, cardiac roentgenograms, and electrocardiograms were reviewed. The diagnosis of persisting left-to-right shunt was made by the recognition of a distinctive clinical syndrome and was confirmed by
cardiac catheterization. In each instance a second operative closure was performed and the outcome was evaluated by careful and repeated observations.

Case Reports

Case 1

D.S. was an 11-year-old girl in whom a heart murmur was heard at age 2. In December 1957, at age 10½, she was admitted for study, having experienced subnormal growth and easily induced dyspnea. Severe exertion was not tolerated. There was no cyanosis at rest or mild exercise.

On examination there was a "pigeon-breast" deformity of the sternum. There was a systolic heave at the left sternal border, and a palpable thrill was present low along the sternum. A grade 4/4 full systolic murmur was heard in this area. It was well transmitted over the precordium and back. There was a diastolic rumble at the apex. The second sound at the pulmonic area was not widely split and not accentuated.

Roentgenograms revealed biventricular enlargement, a prominent main pulmonary artery, hypertensive lung fields, a right-sided aortic arch, and an anomalous left subclavian artery. The electrocardiogram was compatible with right ventricular hypertrophy. The clinical and laboratory data were consistent with the catheterization findings elsewhere (August 1954) of an interventricular septal defect. There was mild elevation of the pulmonary artery pressure, and pulmonary stenosis was absent.

The patient underwent cardiopulmonary bypass surgery on March 6, 1958. After potassium arrest and right ventriculotomy, an interventricular septal defect measuring 2.5 cm. in diameter was repaired with a continuous suture line reinforced by interrupted sutures. The defect had a fibrous wall on the right ventricular side of the aortic valve, and a fibrous approximation was possible, but access was difficult because of a hypertrophied eisra supraventricularis. During the next 48 hours there were persisting hypotension, pallor, and restlessness, in spite of blood replacement and hydrocortisone. On March 8, cardiac tamponade was diagnosed. The sternotomy incision was reopened, the pericardium evacuated of 500 ml. of sanguineous fluid, and the blood loss was replaced. The vital signs then stabilized and the patient's condition improved, but she continued to be dyspneic at rest. The precordium remained active. A left sternal border thrill and a loud systolic murmur were present low along the sternum. There was a left ventricular heave but no third sound or rumble. The second sound was widely split, coinciding with the appearance of a pattern of right bundle-branch block in the electrocardiogram. The liver was 3 or 4 cm. down. On recatheterization, it was confirmed that a left-to-right shunt was still present, and the values for pulmonary blood flow and pressures were practically identical to those of the original study 4 years before. On April 10, 4½ weeks after the original procedure, repair was attempted once more. The upper two thirds of the repair was intact, but the lower one third adjacent to the tricuspid valve had come loose. The hypertrophied bar of eisra supraventricularis was removed for better access and an Ivalon sponge was sutured to the repair with purse string sutures, additionally reinforced with interrupted sutures. The repair appeared firm and the postoperative course was uneventful. The precordium was now quiet and the intense murmur and thrill were gone. The patient was discharged and has been followed without evidence of further reappearance of the shunt.

Case 2

J.L., a 13-year-old boy with a heart murmur heard at birth, was first studied in October 1956. He had experienced normal growth and development, and was able to participate in competitive sports without difficulty. There was no history of cyanosis. On examination he was a husky lad with an obvious left sternal border thrill and a grade 4/4 pansystolic murmur heard all over the precordium but maximal at the pulmonic area. Pulmonic closure could not be heard but was recorded as a delayed, faint sound on the phonocardiogram. There was prominence of the left hemithorax, which was hyperactive. Radiographically, the left ventricle was not enlarged but there was some prominence of the right ventricular silhouette as well as of the central pulmonary vessels. There was suggestive evidence of increased pulmonary blood flow. The electrocardiogram was indicative of right ventricular hypertrophy. Cardiac catheterization established the diagnosis of infundibular stenosis, and an interventricular septal defect with left-to-right shunt (table 1).

He underwent open heart surgery on March 25, 1958. The right ventricle was incised, and a large oval defect measuring 2.25 cm. in transverse diameter was located in the muscular portion of the septum just above the tricuspid valve and beneath the orifice of the aorta. The lowermost corner of the defect was first approximated with interrupted sutures of 3-0 silk, after which an Ivalon prosthesis was interposed and secured by three mattress sutures passing through the tissues on each side. About 9 Gm. of muscle were removed from the outflow tract, and additional muscle masses were excised below the area of stenosis.

On the third postoperative day, sinus tachycardia was present and a pansystolic murmur

Circulation, Volume XXIV, August 1961
Table 1
Cardiac Catheterization in Case 2

<table>
<thead>
<tr>
<th></th>
<th>Preoperative—October 16, 1956</th>
<th>Postoperative—April 24, 1958</th>
</tr>
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<tbody>
<tr>
<td>O₂ ml./100 ml. blood</td>
<td></td>
<td></td>
</tr>
<tr>
<td>SVC</td>
<td>14.6</td>
<td>9.5</td>
</tr>
<tr>
<td>IVC</td>
<td>15.1</td>
<td></td>
</tr>
<tr>
<td>RA</td>
<td>14.5, 15.2</td>
<td>9.9, 9.8</td>
</tr>
<tr>
<td>RV</td>
<td>16.5, 16.6</td>
<td>15.5, 15.7</td>
</tr>
<tr>
<td>PA</td>
<td>16.9, 17.2</td>
<td>15.3</td>
</tr>
<tr>
<td>BA</td>
<td>19.4 (91%)</td>
<td>15.9 (90%)</td>
</tr>
<tr>
<td>Capacity</td>
<td></td>
<td>21.2</td>
</tr>
<tr>
<td>Pulmonary-systemic flow ratio</td>
<td>1.9/1</td>
<td>6.0/1</td>
</tr>
<tr>
<td>Pressures, mm. Hg</td>
<td></td>
<td></td>
</tr>
<tr>
<td>RA</td>
<td>2, mean</td>
<td>13, mean</td>
</tr>
<tr>
<td>RV infund.</td>
<td>19/6</td>
<td></td>
</tr>
<tr>
<td>RV</td>
<td>110/6</td>
<td>55/11</td>
</tr>
<tr>
<td>PA</td>
<td>23/10</td>
<td>56/27</td>
</tr>
<tr>
<td>PA mean</td>
<td>18</td>
<td>40</td>
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<tr>
<td>PA wedge, mean</td>
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<td>24</td>
</tr>
<tr>
<td>BA</td>
<td>142/87</td>
<td>131/65</td>
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<tr>
<td>Estimated O₂ consumption</td>
<td>230</td>
<td>288</td>
</tr>
<tr>
<td>Pulmonary A-V difference</td>
<td>ml./100 ml.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>1.9</td>
<td>1.0*</td>
</tr>
<tr>
<td>Pulmonary blood flow, L/min.</td>
<td>12.0</td>
<td>29.0†</td>
</tr>
<tr>
<td>Pulmonary vascular resistance units</td>
<td>1.1</td>
<td>0.5†</td>
</tr>
</tbody>
</table>

*1.0 ml./100 ml. for the pulmonary A-V difference is arbitrarily assumed, since the true oxygen content of pulmonary artery blood probably cannot be determined under these circumstances.

†These values for pulmonary blood flow and vascular resistance were derived from the assumed pulmonary A-V difference of 1.0 ml./100 ml.

of maximal intensity could be heard along the lower left sternal border. During the following 2 weeks he appeared to be doing well except for a persisting tachycardia. On April 10 he was noted to be dyspneic at rest. The pulse rate was 96, and the liver was large and tender. A thrill and an intense systolic murmur were present along the lower left sternal border, and a distinct third sound was now present at the apex. X-rays showed an increase of heart size, and the lung fields were diffusely engorged, suggesting pulmonary edema. On April 16 he was pale and breathless. The precordium was hyperdynamic, and the pulse was small and rapid. Rales were present at the right base. That evening he coughed up 40 ml. of frothy red sputum and was tachypneic and anxious. The neck veins were full, and systolic pulsations were present. Moist rales were again heard. He was treated for congestive failure with digoxin, diuretics, oxygen, and hypnotics. By April 21 his symptoms had improved and there was radiographic clearing of the pulmonary edema. He was recatheterized at this time. The pulmonary stenosis had been completely relieved but there was a 29-liter pulmonary blood flow indicating that the interventricular septal defect was again patent. A second repair was undertaken on May 6. The medial margin of the repair had separated. The Ivalon prosthesis had remained adherent on the right side, but was free on the left. The defect now had a firm fibrous rim that could be approximated with continuous sutures and closed with interrupted stitches. The closure line was then covered by compressed Ivalon fastened down by continuous over-and-over sutures. On the third postoperative day he became progressively dyspneic and the pulse was rapid. The sternotomy was reopened and a liter of blood removed from the thorax. The pericardium was evacuated of clot and 200 ml. of blood. There was immediate improvement and the postoperative course was subsequently uneventful. The precordium was now quiet and there was no evidence of congestive failure. In follow-up examinations he has maintained his improvement and now feels entirely well.

Case 3
F.H., a 37-year-old woman, had a heart murmur from birth as well as an episode of polyarthritis at age 12. She had always been on activity restriction. On her initial admission she complained of exertional dyspnea. There had been no cyanosis.

On examination there was a right ventricular lift and a left ventricular heave outside the mid-clavicular line. A thrill and a grade 4/4 full systolic murmur were present at the lower left sternal border. At the apex a distinct third sound was present. The second sound was widely split to auscultation and on the phonocardiogram. Cardiac views showed biventricular enlargement, large main pulmonary vessels, and hypervascular peripheral lung fields. The electrocardiogram was indicative of right bundle-branch block. Cardiac catheterization, January 10, 1958, demonstrated the presence of infundibular stenosis with a left-to-right shunt through an interventricular septal defect (table 2).

The patient underwent operative repair on March 11, 1958. After potassium arrest on cardiopulmonary bypass, a ventriculotomy was made and 6 Gm. of muscle were removed from the infundibulum. An interventricular septal defect measuring 15 mm. in transverse diameter was located adjacent to the aortic valve. The defect had a fibrous edge. An Ivalon prosthesis was sewn into place with interrupted 3-0 silk sutures; this was further reinforced by sewing a redundant
Table 2
Cardiac Catheterization in Case 3

<table>
<thead>
<tr>
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<tr>
<td>O₂ ml./100 ml. blood</td>
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</tr>
<tr>
<td>SVC</td>
<td>10.1</td>
</tr>
<tr>
<td>IVC</td>
<td>8.1</td>
</tr>
<tr>
<td>RA</td>
<td>8.9, 9.4</td>
</tr>
<tr>
<td>RV</td>
<td>11.7, 11.8, 12.9</td>
</tr>
<tr>
<td>PA</td>
<td>12.6, 12.9</td>
</tr>
<tr>
<td>BA</td>
<td>14.0 (92%)</td>
</tr>
<tr>
<td>Capacity</td>
<td>15.3</td>
</tr>
<tr>
<td>Pulmonary-systemic flow ratio</td>
<td>3.4/1</td>
</tr>
<tr>
<td>Pressures, mm. Hg</td>
<td></td>
</tr>
<tr>
<td>RA</td>
<td>5, mean</td>
</tr>
<tr>
<td>RV infund.</td>
<td>30/2</td>
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<tr>
<td>RV</td>
<td>117/4</td>
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<tr>
<td>PA</td>
<td>29/12</td>
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<tr>
<td>PA mean</td>
<td>22</td>
</tr>
<tr>
<td>BA</td>
<td>100/49</td>
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<tr>
<td>Estimated O₂ consumption</td>
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<tr>
<td>ml./min.</td>
<td>197</td>
</tr>
<tr>
<td>Pulmonary A-V difference</td>
<td></td>
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<tr>
<td>ml./min.</td>
<td>1.4</td>
</tr>
<tr>
<td>Pulmonary blood flow, L./min.</td>
<td>14.0</td>
</tr>
<tr>
<td>Pulmonary vascular resistance</td>
<td></td>
</tr>
<tr>
<td>units</td>
<td>1.2</td>
</tr>
</tbody>
</table>

leaf of the tricuspid valve was not over the repair. On March 13 there was an abrupt onset of tachycardia, rate 200, accompanied by pallor, sweating, and hypotension. Pericardial tamponade was feared, and the patient was re-explored by opening the sternotomy incision. There was no free blood but the right upper lobe was collapsed and the pulmonary artery was distended. Two-hundred and fifty milliliters of blood were removed from this vessel, and the patient was rapidly digitalized. Her general condition improved and she was now in atrial fibrillation. Definite orthopnea was present. On March 19 her jugular veins were tense, the liver was four fingerbreadths below the costal margin; a loud systolic murmur along the left sternal border and right basilar rales were noted. Her failure was controlled with chlorothiazide and further digitalization. On March 26 sinus rhythm was restored with quinidine. By April 4, at the time of discharge, she felt improved, but there were venous systolic pulsation and a 4/4 pansystolic murmur along the left sternal border followed in diastole by a blowing murmur of pulmonary insufficiency.

She was re-examined on June 2, 1958. In the interim she had experienced breathlessness on slight exertion and angina on hill climbing. The veins were flat at this time, but there were distinct right and left ventricular heaves and a maximal intensity pansystolic murmur and thrill low along the left sternal border. The second sound was loud, and a soft diastolic murmur of pulmonary insufficiency was heard. The liver was not enlarged. The electrocardiogram continued to show right bundle-branch block. Cardiac catheterization indicated that the infundibular stenosis had been removed successfully but that a large left-to-right shunt persisted. On September 17, reclosure was performed. The inferior portion of the defect was again patent just above the attachment of the tricuspid leaflet. There was dense surrounding scar tissue and the Ivalon could not be identified. The defect could be firmly closed by interrupted sutures. The patient’s postoperative course was uneventful, and her precordium was quiet. In follow-up examination she has felt much improved, and it is apparent that her defect has remained closed.

Case 4
RiJo is a 15-year-old boy in whom a heart murmur was heard at birth. His growth and development were somewhat subnormal and he had experienced a gradual decrease in exercise tolerance. On examination there was a marked
thrill along the left sternal border accompanied by a grade 4/4 systolic murmur, which was well transmitted to the apex, aortic area, neck, and axillae. The second sound components were broadly separated. The electrocardiogram pattern was indicative of incomplete right bundle-branch block. X-rays were suggestive of right ventricular enlargement, and the pulmonary vascular markings were increased, suggesting a left-to-right shunt. Cardiac catheterization done elsewhere 2 years before was indicative of infundibular stenosis with a small left-to-right shunt (table 3).

Cardiopulmonary bypass without cardiac arrest was performed on March 31, 1959. A ventriculotomy was made, and 4 Gm. of muscle were removed from the infundibulum. An interventricular septal defect was located just proximal to the aortic valve. With ventricular systole the defect appeared as a split with a linear measurement of 3 to 4 mm. A woven Teflon graft was sewn over the defect with mattress sutures, care being taken to include a good quantity of surrounding muscle in the sutures. Access was difficult because of the large crista supraventricularis. There was slight oozing across the porous fabric of the graft, which appeared to be in good position.

The immediate postoperative course was uneventful, but on April 5 a thrill and loud systolic murmur were again present. There were a persistent tachycardia, rales at the right base, and a four-fingerbreadth tender liver. On x-ray examination right pleural effusion was identified. Recatheterization on April 21 indicated a persisting gradient across the pulmonary valve but the pulmonary artery pressure had become normal. Evidence of a left-to-right interventricular shunt was still present.

On April 29 reclosure was performed. The partially detached Teflon graft was reaproximated through an atriotomy incision, and then the ventriculotomy was reopened for the purpose of reinforcing the closure with a Teflon sponge. On the first postoperative day after signs of tamponade had appeared, the sternotomy had to be reopened and clots removed from the pericardium. The subsequent course was marked by recurrent right pleural effusion and fever. The patient improved after repeated aspiration of sterile fluid, and there was gradual defervescence. He then became asymptomatic and was discharged on May 25. On follow-up examination, he has remained well; there has been no evidence of further disruption of his interventricular septal defect repair.

Case 5

B.L.M., a 17-year-old boy, was admitted for open-heart surgery on October 8, 1958. A murmur had been detected at 3 months of age. The patient had been a frail child who ate poorly and gained weight slowly. There had been intermittent cyanosis but the time of onset was not known. Frequent pneumonia and episodes of subacute bacterial endocarditis had made numerous hospitalizations necessary. Cardiac catheterization in September 1952 indicated the presence of a bidirectional shunt at ventricular level and pulmonary stenosis.

On examination the patient was small and thin. Moderate cyanosis and clubbing were present. There was slight, left precordial bulge, and the apex impulse was within the midsternal line. At the third left intercostal space there was a grade II blowing systolic murmur, and the second sound could not be heard in the pulmonary area. A right ventricular hypertrophy pattern was present in the electrocardiogram. Cardiac views demonstrated slight right ventricular prominence and slightly diminished pulmonary vascularity. A selective angiogram confirmed the presence of an interventricular septal defect with pulmonary stenosis and a right-to-left shunt. The packed cell volume was 61; hemoglobin, 19.1 Gm.

On October 15 at open-heart surgery a conical valvular stenosis with a 4-mm. orifice was found. This was opened in both directions, producing a bicuspid valve. Five grams of infundibular muscle were removed, and an interventricular septal defect measuring 14 mm. in diameter was identified just above the insertion of the tricuspid valve. The defect was sutured mattress-fashion, the edges being apposed with four sutures; this was reinforced posteriorly by a flap of redundant tricuspid valve. An Ivalon prosthesis was then anchored over the linear repair with continuous over-and-over sutures.

The first postoperative week was marked by dyspnea, tachypnea, and mild cyanosis. There were some neck vein distention, a 3-em. heparin-megaly, and basal rales. The electrocardiogram now showed a pattern of right bundle-branch block and x-rays demonstrated congested lung fields and free fluid in the bases. The venous pressure was 26 cm. of saline, and the circulation time was 33 seconds. In spite of digitalization and other measures, heart failure persisted. There was an irregular fever, which reached 39.4 C. in the third postoperative week. Blood cultures were positive for Staphylococcus albus, coagulase positive and Bacillus subtilis, but these were thought to be contaminants. The clinical situation was grave. Examination now revealed pulsating neck veins and a pansystolic murmur of maximal intensity along the left sternal border, over the precordium and back. The spleen and liver were enlarged to the umbilicus. Repeat catheterization on December 9 showed persistence of a left-to-
right shunt at ventricular level and pulmonary stenosis.

A second open-heart procedure was performed on December 23, and a hole was identified in the lowermost portion of the repair where the Ivalon had become detached laterally and posteriorly from the tricuspid corner. The defect was again repaired with mattress sutures, the Ivalon being repositioned. The closure was further reinforced by a suture placed deep in the tricuspid valve and encompassing the defect. More muscle was removed from the infundibular region. Sections from some necrotic muscle at the edge of the defect contained clumps of gram-positive cocci and endocardium from the region of the sutures showed acute and chronic endocarditis. There was now less respiratory distress, the venous engorgement was diminished, the liver became smaller, and the precordium was quieter, a much softer systolic murmur now being present along the left sternal border. But the temperature continued to rise to 39.9° C., *Staph. aureus* appeared in blood cultures, and the right shoulder became swollen and tender. On Jan. 4, 1959, the right shoulder joint was opened and drained and osteomyelitis was found in the humerus. X-rays also revealed mottled luencies in the acromial process. The patient received penicillin, chloramphenicol, novobiocin and streptomycin, but fever to 40.5° C. continued and the blood cultures remained positive for *Staph. aureus*. On January 31 the doses of chloramphenicol and novobiocin were increased to 2.0 Gm., and vancomycin was started. The patient then became afebrile. The blood cultures were sterile until February 19, 3 days after the discontinuation of all antibiotic treatment. The previous regimen of antibiotics was begun once more, and the temperature became normal again on March 8. Bacitracin was administered for 4 days, but was discontinued because of a blood urea of 108 mg. In addition, a precordial thrill was again palpated, loud systolic and diastolic murmurs were heard along the left sternal border, and the liver was larger. On April 11 chills and a temperature of 39.5° C. were accompanied by pain in the right ankle, and the blood cultures again contained *Staph. aureus*. Erythromycin, 8 Gm., and rifocetin 4 Gm. were started. The patient became afebrile again and the cultures were sterile; but the general condition was poor. Hypotension and oliguria were present, and the blood pressure was maintained with the help of norepinephrine. On April 21 pulsating neck veins, large liver, right ventricular heave, and a maximal left sternal border systolic murmur indicated a reopened interventricular septal defect and tricuspid insufficiency. This was confirmed by catheterization on May 11. The following day an attempt was made to close the defect again. This was accomplished but postoperatively circulatory failure was intractable, and the patient died within 24 hours.

At autopsy the heart weighed 520 Gm. The right ventricle was hypertrophied and dilated. The pulmonic valve was bicuspid, and the cusps were deformed and folded back upon themselves. The recent interventricular septal defect repair was intact. On the left ventricular side of the defect a small vegetation was identified. Infarrets were present in the spleen and left kidney, and thrombi were present in the iliac veins and moderate-sized pulmonary arteries. There were edema of the lungs, congestion of other viscera, and ascites.

**Case 6**

Roja, a 26-year-old man, was first admitted on March 3, 1959. In spite of a history of heart murmur from birth, there were no symptoms or limitation of activity, no cyanosis, or disability. On examination there were a slight right ventricular heave and a thrill at the lower left sternal border. There was a maximal intensity, full systolic murmur in this position. The second sound was heard better at the aortic area, and no splitting could be detected. Cardiac catheterization indicated the presence of a left-to-right shunt at the ventricular level and infundibular stenosis. The electrocardiogram pattern was right ventricular hypertrophy but there was only questionable chamber enlargement radiographically. The pulmonary artery was dilated but the vascularity of the lung fields was normal.

Open-heart surgery was performed on May 28. There was a 2.0-cm. defect in the membranous septum and both infundibular and valvular stenosis were present. The defect was closed with six ventral sutures. The closure was reinforced by suturing Ivalon sponge over the defect with interrupted mattress sutures. Pulmonary valvotomy was done, and infundibular muscle was excised. An Ivalon prosthesis was used to enlarge the outflow tract; this was reinforced with pericardium.

A loud systolic murmur persisted at the left sternal border, and a diastolic flow murmur appeared at the apex. The precordium was now hyperdynamic, and there were jugular pulsations. Edema appeared in the saerum and lower extremities. In spite of digitalization and other measures to control congestive failure, the patient was dyspneic, uncomfortable, and had persisting tachycardia. Radiographically, the heart was now much enlarged. The clinical picture suggested reopening of the ventricular septal defect with a large left-to-right flow, tricuspid insufficiency, and congestive heart failure. On June 15, during catheterization, ventricular fibrillation occurred abruptly, and resuscitation was effected by cardiac massage.
and electrical defibrillation. During the next 24 hours there were persisting tachycardia and hypotension, and 3,750 ml. of blood were lost through the drainage tubes in the chest. The chest was reopened, and an additional 1,500 ml. of blood were evacuated, after which a bleeding intercostal artery was identified and ligated. The patient’s condition then improved, but he was febrile and a heavy growth of *Aerobacter aerogenes* was present in the fluid from the chest tubes. On examination, the precordium was heaving, and jugular and hepatic pulsations were marked. The systolic murmur with maximal intensity at the left sternal border was followed by a loud diastolic rumble, suggesting a tricuspid flow murmur. The systolic murmur could easily be heard over the liver. There were ascites and pronounced edema. Fever continued in spite of multiple antibiotic therapy, and *Aerobacter* was cultured routinely from persistently draining chest tubes. Roentgenograms indicated the persistence of an extensive empyema cavity. An open drainage was done on July 27. The patient subsequently became afibrile and was discharged for interim nursing care. He was readmitted in October, at which time physical findings were essentially unchanged, and a second attempt was made to close the defect. Portions of the previously placed Ivalon were still in place but the defect was widely open. Closure was effected with five mattress sutures, and a muscle flap from the anterior portion of the septum was used for reinforcement. Several sutures were taken in the patulous tricuspid ring. Postoperatively there was profuse drainage of blood, and the right hemithorax was repeatedly evacuated. A total of 6 liters of blood was withdrawn or drained. Oliguria and hypotensive, in spite of vigorous replacement therapy, the patient died on the fourth postoperative day.

At autopsy, the heart weighed 650 Gm.; there was notable enlargement of the right ventricle. The pulmonic valve was normal but just distal to the valve, there was a fibrous ring. The circumference was 3.5 cm. The aorta arose anteriorly to and to the right of the right ventricular outflow tract. The Ivalon gusset in the outflow tract was intact. The recent interventricular septal defect closure was secure. At the apex the myocardium was greatly thinned and replaced by fibrous tissue. There were small mural thrombi attached to the endocardium of both ventricles, and an embolus was present in the left coronary artery. The lungs were edematous, hemorrhagic, and atelectatic, and generalized visceral congestion was present.

**Case 7**

M.M., was an 8½-year-old girl with a congenital heart murmur. A diagnosis of interventricular septal defect had been established by cardiac catheterization at the age of 10 months. No significant gradient was present across the pulmonary valve. There was no cyanosis, but exertional dyspnea was present, the child did not gain weight normally, and frequent respiratory infections occurred. For 3 years prior to admission the patient had been on a low-salt diet and digitalis because of “pulmonary congestion.” On examination the veins were flat, and there was no cyanosis. A right ventricular heave was present, and there was a thrill and a loud pansystolic murmur at the third left interspace along the sternal edge. The second sound was loud. On March 20, 1956, at open-heart surgery, a 15-mm. interventricular septal defect was located in the membranous septum; it was sutured with interrupted silk. Postoperatively a loud systolic murmur persisted; this could also be heard in the neck. The second sound was accentuated in the pulmonic area. Radiographically, the heart was larger than preoperatively, and there was venous engorgement. The patient was seen again in December 1957, at which time she was thought to be less dyspneic. The precordium was active, however, and a maximal intensity, left sternal border systolic murmur was present. The second sound was loud though not audibly split, and a diastolic rumble was heard at the apex. In April 1959, her findings were similar. X-rays now showed biventricular enlargement and a prominent left atrium. There was increased pulmonary vascularity suggestive of a persisting left-to-right shunt. Surgery was again performed in August 1959 and the reopened defect was identified. Closure was effected by means of interrupted mattress sutures reinforced by compressed Ivalon. In the immediate postoperative period, a third sound was heard at the apex and the liver was enlarged. The patient was digitalized and placed on a salt-free diet. The liver regressed in size, and the patient improved rapidly. At the time of discharge the precordium was quiet, and a residual grade II systolic murmur was heard at the left sternal border.

**Discussion**

**Description of Patient and Preoperative Status**

The case reports include seven patients ranging in age from 8½ to 35 years. They all had ventricular septal defects. Six had complicating infundibular stenosis or a greatly hypertrophied crista supraventricularis. One patient was cyanotic. Two of the patients with pulmonary stenosis were asymptomatic. The remainder of the group had degrees of disability ranging from mild exertional dyspnea to easily induced fatigue, breathlessness, or cyanosis. Three patients had experienced sub-
normal growth and development. None had experienced congestive heart failure preoperatively although one child had been on salt restriction and digitalis because of "pulmonary congestion.'"

**Description of Defects and Technic of Closure**

All the defects were located in the region of the membranous septum, but were not necessarily delimited by the position of this structure. They were most commonly bordered by the insertion of the tricuspid valve below and by the aortic valve in front. Although one of the defects was described as a 3 to 4 mm. slit in systole, the remainder varied from 1.5 to 2.5 cm. in diameter. Characteristically, the defects lacked a firm fibrous rim, the edge of the defect consisting instead of cardiac muscle.

The defects were repaired by conventional methods, and an Ivalon prosthesis was usually employed to close the defect or reinforce the closure. The infundibular stenosis was relieved by resection of muscle in the outflow tract and crista supraventricularis.

**Time of Detection of Persisting Shunt**

This could not be ascertained precisely in each instance. In three cases findings suggestive of a persisting shunt were present on the first to third postoperative day. In the other cases, the significant auscultatory observations were not made until the fifth to eighth day. Significantly, in no case did earlier auscultatory findings, indicating the closure of a shunt, precede notes reporting the reappearance of a shunt. It is probable that in all cases the defect was either never completely closed or that the reopening occurred within the first few postoperative days. This has also been the experience elsewhere.10, 11

**The Syndrome of Persisting Shunt**

It will be noted above that, preoperatively, the symptomatology of these patients was quite variable, ranging from no symptoms to easily induced dyspnea. In none, however, was dyspnea present at rest, and none had orthopnea or objective evidence of heart failure. This relatively benign situation was profoundly altered by the persistence postoper-avely of a left-to-right shunt. Sinus tachycardia and tachypnea were the rule, and these became increasingly significant after the first 72 hours, when fluid balance had been stabilized and when postoperative pain or fever had been excluded. The patient was usually dyspneic and orthopneic. The pulses were small. The precordium was hyperactive. There was a right ventricular heave; occasionally, a left ventricular heave. At this time a thrill and a full systolic murmur were present at the lower sternal border. If right bundle-branch block had occurred, the second sound was broadly split. A third sound or diastolic flow murmur, not heard preoperatively, could be detected in three instances. All these find-
The heart is moderately enlarged and the lung fields suggest a small increase in pulmonary blood flow. Right. Postoperatively there is distinct broadening of the cardiac silhouette and the pulmonary vasculature is more prominent, suggesting an increase in left-to-right flow after failure to close the defect at a time when the infundibular stenosis had been removed. The right atrium and superior vena cava appear dilated in this patient, who had signs of tricuspid insufficiency.

The most striking changes appeared in the five patients who had had significant infundibular gradients as well as ventricular septal defects preoperatively. The pulmonary stenosis had been adequately relieved in these instances and, in some, the left-to-right shunt now seemed to be greater than before surgery. This was suggested by the strikingly dynamic precordial movements, by the appearance for the first time of an apical third sound or flow murmur, or by the murmur of pulmonary insufficiency. Notable too was the appearance of tricuspid insufficiency in at least three of this group. This was signified by systolic jugular pulsations followed by rapid diastolic collapse, and by the pronounced evidence of right-sided failure, which included high venous pressure, hepatomegaly, edema, and ascites. Because of the loud murmur of the ventricular septal defect, the presence of a discrete murmur of tricuspid insufficiency was difficult to establish but was identified in at least one instance where it was characteristically louder on inspiration. In this individual the liver was pulsatile. Pulmonary congestion also occurred, and one patient had frank pulmonary edema indicating that left ventricular failure is a distinct part of this syndrome. Five of the patients had to be treated intensively with digitalis, salt restriction, diuretics, and, occasionally, oxygen. One patient had atrial fibrillation which was later converted to sinus rhythm.

Effects of the Persisting Shunt on the X-ray and Electrocardiogram

The cardiac silhouette regularly increased in size in the presence of a persisting left-to-right shunt. This was quite striking since, by comparison, the preoperative increases in the cardiac silhouette were usually modest. Figures 1 and 2 illustrate this point. There were also notable changes in the lung fields. For example, in both figures the lungs are more vascular postoperatively, indicating greater pulmonary blood flow. There is also hilar engorgement and diffuse haziness in figure 1, which was taken after the patient had experienced pulmonary edema. In figure 2, there is dilatation of the right atrium and superior vena cava, and this film was taken at a time when clinical and hemodynamic evidence of tricuspid insufficiency was present. In oblique views some enlargement of the left atrium is also apparent.

Electrocardiographic alterations are more difficult to assess, since the electrocardiogram is often altered by surgery alone. Electrocardiograms before and after operation were available in six cases. In one instance the preoperative tracing remained that of complete right bundle-branch block postoperatively. Complete right bundle-branch block appeared in three cases where patterns of right ventricular hypertrophy were originally present and in two cases where, preoperatively, the QRS duration was less than 0.12 second and an rsr' or rsR' complex was present in V1.

Cardiac Catheterization Data

Preoperatively left-to-right shunts in the patients with ventricular septal defects complicated by infundibular stenosis were
not large, and in only one instance did the pulmonary to systemic flow ratio exceed 2.3/1. One patient was cyanotic and had no net left-to-right shunt preoperatively. Systolic pressure in the right ventricle ranged between 79 and 117 mm. Hg, and the peak systolic gradient between the main right ventricular chamber and the pulmonary artery ranged between 65 and 87 mm. Hg, indicating moderate infundibular stenosis. With one exception the right ventricular diastolic and mean right atrial pressures were normal.

Postoperative cardiac catheterization invariably disclosed the presence of a left-to-right shunt, whereas the infundibular gradient had been eliminated or reduced. Of the six patients with preoperative gradients, postoperative catheter data showed no gradient in two, and residual systolic gradients of 49 and 13 mm. Hg, respectively, in two others. In the two remaining cases, observations at second operation suggested that the obstruction had been successfully eliminated.

The pulmonary to systemic flow ratios were either essentially unchanged or greater postoperatively. At times the increase in pulmonary blood flow was striking. For example, in table 1 are the data on an 11-year-old boy with so-called acyanotic tetralogy. He had a modest left-to-right shunt preoperatively, the pulmonary to systemic flow ratio being just less than 2/1, and there was a peak systolic gradient of 87 mm. Hg between the right ventricle and pulmonary artery. Postoperatively this gradient was eliminated. At the same time the pulmonary blood flow has become very large, but it can only be approximated under these circumstances because of the difficulty in sampling true mixed venous blood. If a pulmonary arteriovenous difference of 10 ml. per 100 ml. is assumed, the estimated pulmonary blood flow is 29 liters per minute. The mild arterial desaturation is probably due to hyperventilation induced by presedation with secobarbital and meperidine. The pulmonary wedge pressure was 24 mm. Hg, and although a flow gradient across the mitral valve cannot be excluded, it is probable that there was also an elevation of left ventricular diastolic pressure. Right ventricular diastolic pressure was clearly elevated. Right atrial pressure curves of course reflected these ventricular events, but at times they also offered graphic evidence of tricuspid insufficiency. This is illustrated in figures 3 and 4.

**Reoperation to Close the Persisting Shunt**

In all instances a second attempt was made to close the defect. The second operations were performed at a minimum of 4½ weeks after the first and in all but one instance within 6 months. In one case a third closure was attempted 5 months after the second operation. There were two postoperative deaths. One was due to intractable intrathoracic hemorrhage, oliguria, and hypotension in a patient who had had severe tricuspid insufficiency and heart failure after the second operation. The second death occurred in a patient who had staphylocoecal endocarditis at the site of his repair, osteomyelitis, and heart failure. This patient survived a second operation but died in heart failure after the third operation.
The most constant finding at reoperation was the detachment of the Ivalon and Teflon prostheses. There was an apparent failure of tissue to invade the material and create a firm union when it was used as a supporting plug. In addition, where the material had been retained by sutures and became firm, it tended to fracture at the suture line, suggesting brittleness. There was also a single Teflon cloth failure apparently due to inadequate invasion of the material from the margin of the repair.

Improvements in Surgical Technic

During the past 2 years, we have repaired the ventricular defect with interrupted sutures of number 3-0 silk. The repair is then covered with a crimped Dacron patch held in place with interrupted sutures. Great care is taken with the lower margin, where recurrences are prone to occur. Here some of the redundant tricuspid valve is often drawn in with the first suture. Since using this method, recurrences have virtually disappeared.

Potassium arrest was abandoned approximately 2 years ago in favor of general body hypothermia in conjunction with extracorporeal circulation. For tetralogy of Fallot or complicated interventricular defects, temperatures of 20 to 24° C. are reached through the use of a heat-exchange unit incorporated in the lung. At approximately 26° C., the heart fibrillates, and at lower temperatures, it often will cease beating. When necessary, the aorta is cross-clamped for short periods of time. Left atrial decompression is routinely employed with a catheter placed in the right side of the left atrium.

General body hypothermia has been combined with extracorporeal circulation in over 240 cardiac procedures of which 110 were interventricular defects of tetralogy of Fallot. Permanent heart block has virtually disappeared since employing the above technique. In only one operation for tetralogy of Fallot was death probably caused by this complication, and there is only one living patient with permanent block.

Conclusions

Disruption of a ventricular septal defect repair is particularly, though not exclusively, a complication in the open cardiotomy treatment of patients with coexisting infundibular stenosis. The disruption occurs within the first few days and may usher in profound circulatory disturbances. As a consequence patients who were relatively asymptomatic exhibit symptoms and signs of biventricular failure and tricuspid insufficiency. An anatomic basis for this is provided by the successful elimination of, or significant reduction of, the infundibular obstruction. The example cited above and in table 1 indicates how, under these circumstances, a patient who had had a relatively small left-to-right shunt may develop an extremely large pulmonary blood flow approximating 30 liters per minute. Such a flow is clearly larger than would be customary for a ventricular septal defect in adults. The estimated pulmonary vascular resistance of 0.5 units is low; this suggests the failure of the pulmonary vasculature to regulate flow through vasoconstriction.

However, tremendous increases in pulmonary blood flow were not always present and cannot be solely responsible for the production of this syndrome. In the case represented in table 3, for example, the pulmonary blood flow has increased by a third only, probably because residual infundibular obstruction was present, and because the pulmonary vascular resistance increased. The patient whose data are shown in table 2 exhibited the features of this syndrome even though her repair was only partially disrupted so that she had the
REOPENED VENTRICULAR SEPTAL DEFECT

combined benefits of an actual reduction in pulmonary flow and of the complete elimination of infundibular stenosis. In such individuals it would appear that something had happened relatively acutely so that the heart was now unable to handle a work load that for many years it had supported with few ill effects.

In this regard some evidence has accumulated to suggest that the methods employed for cardiac arrest at surgery and ventriculotomy itself may have an adverse effect on the myocardium. In most of these repairs potassium arrest was used. It has recently been shown in acute experiments that potassium arrest depresses left ventricular function considerably. Furthermore, the effects of right ventriculotomy done in the conventional way from base to apex along the long axis of the ventricle are not entirely benign. Ventricular function curves and high-speed cinefilms before and after right ventriculotomy in experimental animals have shown appreciable depression of function and alterations in the sequential contraction pattern of the right ventricle. These alterations in function can persist after a number of months. Such a ventricle may perform adequately when not stressed but could fail when presented with a persisting left-to-right shunt postoperatively.

The tendency for disruption to occur in patients with infundibular stenosis is understandable. The defect is in close relation to the area of stenosis and to the thickened, overgrown crista supraventricularis, making exposure suboptimal. In spite of resection of the obstructing muscle elements, the edges of the defect may be difficult to bring together easily without tension. Moreover, the tough fibrous edge, which is the rule in uncomplicated ventricular septal defects, is not present, and the margin of the defect consists of soft cardiac muscle, which is not so suitable for holding sutures.

A major disappointment has been the failure of Ivalon and Teflon prostheses in our hands. We have observed disruptions of the suture line and failure of the material to be overgrown by tissues, the requisite of a firm repair. There has also been some evidence of brittleness of the material. These experiences have led to the adoption of the present method, which consists in direct suture of the defect with interrupted silk, followed by the application of a crimped Dacron patch over the repair with interrupted sutures. Potassium arrest has been discontinued, and cardiac arrest or ventricular fibrillation is now induced by total body hypothermia routinely in the treatment of tetralogies and large ventricular septal defects. The present approach seems promising, and there have been no recurrences or deaths in the last 25 cases of tetralogy of Fallot.

Summary

The persistence of a left-to-right shunt due to the unsuccessful repair of a ventricular septal defect associated with infundibular stenosis has been described.
A characteristic clinical picture has been reported. The main features include congestive heart failure, tricuspid insufficiency, and increased pulmonary blood flow.

The severity of the symptoms appears to be due to the persistence of a left-to-right shunt at a time when the myocardium has been affected adversely by such operative insults as potassium arrest and ventriculotomy.

The particular difficulties attending the repair of a ventricular septal defect with infundibular stenosis include the proximity of the defect to the hypertrophied muscle mass, the soft muscular margin of such a defect, and the tension on the suture line. Ivalon and Teflon prostheses have been disappointing in their failure to maintain the integrity of the repair.

All of the patients were reoperated upon. Five survived and their defects are now closed. The second repairs were abetted by the development of a fibrous scar around the margin of the defect. The five survivors are in good health and there is no evidence of a remaining shunt.

As a consequence of these experiences, surgical technics were modified. The defects are now closed by direct suture with interrupted silk, followed by the application of a crimped Dacron patch over the repair. The results have been encouraging and since the new technic has been adopted, there have been virtually no recurrences.

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The Reopened Ventricular Septal Defect: A Syndrome Following Unsuccessful Closure of Interventricular Septal Defects Particularly in Association with Infundibular Stenosis

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Circulation. 1961;24:250-262
doi: 10.1161/01.CIR.24.2.250

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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