Syndrome of Left Ventricular–Right Atrial Shunt Resulting from High Interventricular Septal Defect Associated with Defective Septal Leaflet of the Tricuspid Valve

By Mildred Stahlman, M.D., Samuel Kaplan, M.D., James A. Helmsworth, M.D., Leland C. Clark, Ph.D. and H. William Scott, Jr., M.D.

Two similar cases of unusual congenital anomalies of the heart are presented, in which there existed a direct communication between the left ventricle and the right auricle through a defect involving the membranous portion of the interventricular septum with defective formation of the medial leaflet of the tricuspid valve. Data obtained at catheterization, operative and postmortem findings are presented in each case. The differential diagnosis of this type of lesion is discussed.

Because of recent advances in the surgical treatment of defects of the cardiac septa it has become of considerable practical importance to be able to recognize such defects by their clinical manifestations and to localize them accurately by current diagnostic technics. It is usually not difficult to differentiate defects of the atrial septum from ventricular septal defects. However, we have recently observed two children with manifestations of a large left-to-right intracardiac shunt erroneously diagnosed by clinical and catheterization data in each instance as atrial septal defect. Surgical closure under direct vision by open cardiomyotomy was attempted in each case but without survival. At autopsy, each child was found to have an unusual form of high interventricular septal defect associated with a malformation of the septal leaflet of the tricuspid valve.

Case Reports

Case 1.* M. Z., a 4 year old white boy, had been under medical observation most of his life. He was the result of an uncomplicated full term pregnancy, delivery was spontaneous and his birth weight was 5 pounds 8 ounces. At the age of 7 weeks he developed a severe respiratory infection characterized by pyrexia, dyspnea and noisy respirations with a prolonged expiratory phase. Three further similar episodes of respiratory infection occurred during the first year of life. At the age of 1 year he had been treated in the Cincinnati Children's Hospital for bronchopneumonia, tonsillitis and left otitis media. Frequent attacks of bronchitis occurred during the second and third years of life complicated by the development of bronchopneumonia on three occasions. One year before death, congestive cardiac failure occurred during a bout of bronchopneumonia. He was treated with digitalis and mercurial diuretics and maintained on a daily dose of 0.05 mg. digitoxin.

The patient's exercise tolerance was always poor but diminished during his fourth year so that he could walk a distance of only 25 yards. He slept poorly because of orthopnea and a persistent nonproductive cough. Cyanosis was not noted at any time, even during the periods of bronchopneumonia and congestive heart failure.

The boy's weight was 26½ pounds, his height 38 inches and he appeared to have been chronically ill. The venous pressure was increased, causing jugular distension 5 cm. above the sternal angle and the venous pulse exhibited a prominent intrinsic "c" wave. The liver edge, which did not pulsate, was 6 cm. below the right costal margin. There was no peripheral edema and there were no adventitious sounds in the lungs. The pulse was small, 90 per minute and regular. The blood pressure in both upper extremities was 94/50 and 110/60 mm. Hg in both legs.

The precordial area bulged markedly. The cardiac impulse, which was tapping in nature, was maximal in the sixth intercostal space in the left anterior...
axillary line. A coarse systolic thrill was palpable down the left sternal border. A sternal lift was both visible and palpable. A harsh grade IV systolic murmur was heard maximally in the second left parasternal space but radiated down the left sternal border, to the apex and to the back between the scapulae. The second heart sound was widely split but there were no diastolic murmurs. The electrocardiogram showed evidence of incomplete right bundle branch block (RsR' in V1). Roentgenographic studies of the chest showed cardiac enlargement mainly involving the right ventricle. The pulmonary artery segment was prominent, the aortic knob inconspicuous and pulmonary over-vascularity was evident (fig. 1). There were no abnormal pulsations of the intrapulmonary vessels. Examination of the peripheral blood, urine and erythrocyte sedimentation rate gave normal values. The total blood volume was estimated to be 990 cc. (T-1824 method).

Cardiac catheterization five months before death gave the following results:

<table>
<thead>
<tr>
<th>Pressure Systolic/Diastolic Mean mm. Hg</th>
<th>Oxygen Content (vols. %)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary Capillary</td>
<td>—</td>
</tr>
<tr>
<td>Pulmonary Artery</td>
<td>92/38</td>
</tr>
<tr>
<td>Right Ventriecle (outflow tract)</td>
<td>92/10</td>
</tr>
<tr>
<td>Right Ventriecle (mid)</td>
<td>12/7</td>
</tr>
<tr>
<td>Right Atrium (near tricuspid valve)</td>
<td>12/7</td>
</tr>
<tr>
<td>Right Atrium (mid)</td>
<td>12/7</td>
</tr>
<tr>
<td>Superior Vena Cava</td>
<td>—</td>
</tr>
</tbody>
</table>

Arter. O2 sat. 97 per cent.
Pulmonary artery flow 5.8 L. per minute.
Left-to-right shunt 3.1 L. per minute.
Pulmon. arteriolar resistance 604 dyne second cm.⁻²
Rt. ventric. work 6.6 Kg. M./m.²/minute.
Rt. atrial pressure curve showed a prominent "c" wave.

The studies suggested the presence of an atrial septal defect. The increased venous pressure with prominent systolic venous pulsations was thought to be due to associated functional tricuspid insufficiency.

It was planned to close the defect surgically with the aid of the Clark extracorporeal pump oxygenator. The patient was anesthetized with Pentothal sodium following which oxygen and ether were given through a closed endotracheal system. Bags of cracked ice were applied around the trunk and extremities to produce a mild hypothermic stage and the operation was begun after the rectal temperature had been reduced to 90 F. A right anterolateral incision was made and the thoracic cavity entered through the fourth intercostal space. The catheter system for venous drainage included three plastic tubes with multiple perforations. Two of these were inserted via the saphenous veins into the inferior vena cava. The superior vena cava was cannulated through the azygos vein. Arterialized blood was returned through a single cannula placed in the right subclavian artery with its tip in the ascending aorta. The entire extracorporeal system was filled with freshly collected whole blood.

After an intravenous injection of heparin (3 mg. per Kg.) extracorporeal circulation was started and slowly increased to between 1,200 and 1,400 cc. per minute. The superior and inferior vena cava were then occluded. The right atrial wall was opened through an incision extending from the insertion of the superior vena cava to the inferior vena cava (fig. 2). Although the right atrium appeared empty before cardiotomy, the entire area became flooded with a large volume of coronary venous blood within a few minutes. Because of the delay in returning this blood to the extracorporeal circuit, the arterial blood pressure fell to shock levels and did not return to the preoperative level for a period of 50 minutes. The right atrial cardiotomy allowed closure of the complex defect by sutures placed under direct vision. Despite difficulty with the overwhelming coronary venous blood, which was not less than 250 cc. per minute, the right atrium was open for only 17 minutes and the extracorporeal circulation was terminated after a total of 33 minutes.
After the intravenous infusion of protamine sulfate was started, all cannulas were withdrawn, the pleural space was drained and the chest wall closed in the standard manner. Although the boy was able to move his extremities and take sips of water eight hours after operation, he remained anuric and died 16 hours after cardiotomy.

At autopsy the major findings were in the heart. The preoperative diagnosis of atrial septal defect was not confirmed. The basic cardiac anomaly was a high ventricular septal defect of the membranous septum associated with an anomaly of the septal leaflet of the tricuspid valve (Fig. 3). The ventricular septal defect had been closed by silk sutures but after their removal, the defect measured 1.5 cm. in diameter. The septal leaflet of the tricuspid valve was very short and incomplete, allowing a direct communication between the left ventricular and right atrial cavities. The right ventricular wall measured 7 mm. and the left 13 mm. in thickness. The circumference of the cardiac valves in millimeters was as follows: tricuspid 105, mitral 85, aortic 60 and pulmonary 60. The myocardium was of normal color and consistency and the pulmonary veins, coronary sinus and venae cavae appeared normal. The ductus arteriosus was obliterated and the aorta was normal. The other positive findings included 90 cc. of blood in the right pleural cavity, some atelectasis of the dorsal portion of both lungs, a small recent infarct of the upper pole of the right kidney, mucosal hemorrhages in the stomach and acute focal colitis. Because the brain was not examined, the possibility of cerebral air embolism could not be excluded.

Case 2. M. H. W. III, a 4 year old white boy, was the result of a full term pregnancy uncomplicated by maternal illness. Delivery was spontaneous and no cyanosis was observed at birth. A cardiac murmur was noted by his attending pediatrician in early infancy. Feudings were taken poorly and his growth pattern was below normal standards. He vomited frequently throughout his first year of life. At the age of 2 years, the boy developed a severe acute respiratory infection characterized by fever, dyspnea and cough which was diagnosed as bronchopneumonia and required hospitalization. Cyanosis was said to be present at this time and he was treated with penicillin and oxygen with disappearance of cyanosis and respiratory symptoms. Subsequently, he showed no cyanosis on exercise and his exercise tolerance was good, although he was said to tire a bit sooner than his siblings. At no time had he shown signs or symptoms of cardiac failure.

On July 28, 1952, at the age of 21/2 years, he was admitted to Vanderbilt University Hospital for the first time and cardiac catheterization was unsuccessfully attempted. He was discharged and readmitted on Nov. 5, 1952. At this time cardiac catheterization was successfully performed. Physical examination showed a weight of 25 pounds and height of 34 inches. He was an energetic, wiry little boy who was in no distress. There was no evidence of cyanosis of the mucous membrane or clubbing of the fingers. No edema was present and the liver was not palpable. The pulse was full in all extremities and averaged 85 per minute. The blood pressure in both arms was 98/76 mm. Hg and in both legs 104/70. There was a marked precordial bulge associated with a mild pigeon breast deformity. The precordium was overactive and the apical impulse was in the left fifth intercostal space at the anterior axillary line. There was a coarse systolic thrill easily felt over the left lower precordium. A harsh grade IV systolic
murmur was best heard in the left fourth and fifth intercostal spaces parasternally and it was widely transmitted over both left and right sides of the chest, anteriorly, but poorly transmitted to the back. Diastole was clear. The pulmonary second sound was moderately accentuated. There was a regular sinus rhythm. The remainder of the examination was essentially negative.

The electrocardiogram showed right ventricular enlargement and inverted T1, TVL and T of V1 through V6. There was depression of S-T1, S-T of aVF, V1 and V4. Roentgenographic studies of the chest showed a bizarre configuration. There was fullness of the pulmonary artery region and the right ventricle was markedly enlarged. The right border of the cardiac shadow projected far into the right lung field as a large smooth crescent and pulsations in this region were diminished. The vascularity of the lungs was increased. Examination of the blood, urine and erythrocyte sedimentation rate were within normal limits. Cardiac catheterization was performed and while an attempt was being made to turn the catheter tip into the left pulmonary artery, the sinus mechanism was replaced by atrial fibrillation with a ventricular rate of 200 beats per minute. As this mechanism persisted, the procedure was discontinued with a minimum of catheterization data having been obtained. These data appear below:

<table>
<thead>
<tr>
<th>Pressure (mm. Hg)</th>
<th>O2 Content Vol. %</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>S/D Mean</td>
</tr>
<tr>
<td>Rt. Pulmonary Artery</td>
<td>65/30 42 11.05</td>
</tr>
<tr>
<td>Rt. Venticle (outflow)</td>
<td>65/0 25 11.6</td>
</tr>
<tr>
<td>Rt. Atrium (mid)</td>
<td>±0              11.25</td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>— —             8.0</td>
</tr>
<tr>
<td>Capacity</td>
<td>— —             15.2</td>
</tr>
</tbody>
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These data suggested the presence of a left-to-right shunt into the right atrium and were interpreted as most probably the result of an atrial septal defect. The episode of atrial fibrillation was

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**Fig. 4.** Case 2. Teleoroentgenogram with barium swallow showing cardiac enlargement with extreme prominence of right atrium.

**Fig. 5.** Case 2. Sketches made at operation and autopsy showing ventricular septal defect with anomalous septal leaflet of tricuspid valve as viewed through open right atrium. Diagram shows relations in frontal cross-section.
treated with intramuscular quinidine and the mechanism reverted to a sinus one within 18 hours. The patient was discharged to return at a later date for surgical repair of his defect.

He was admitted for the third time on Jan. 5, 1954. He had been well between admissions and physical examination was essentially unchanged. The heart was greatly enlarged with a tremendous right atrium (fig. 4). He weighed 29½ pounds and was 38 inches tall. Electrocardiogram and cardiac fluoroscopy were unremarkable. The patient was digitalized preoperatively and on Jan. 8, 1954 operation was performed in the hope that an atrial septal defect might be closed. Anesthesia was induced with cyclopropane and hypothermia induced by immersing the patient in ice water until his body temperature had fallen to 28 C. A transverse anterior intercostal incision was placed in the fourth intercostal space with transection of the sternum. The heart was so large, the right atrium in particular, that it was necessary to divide the costal cartilages of the fifth, sixth and seventh ribs in order to expose the inferior vena cava. There was no evidence of any anomaly of venous return. Palpation through the enormous right atrium revealed a vigorous systolic thrill and a jet of blood was ejected into the right atrium with each systole. Both venae cavae were occluded with tapes. A Satinsky clamp occluded the pulmonary artery and aorta and the right atrium was opened widely. The atrial septum was intact. A jet of blood was seen regurgitating from the region of the tricuspid valve. Examination of this area revealed a 1 by 2 cm. defect which was interpreted by the surgeon as a patent ostium primum. The defect lay immediately below the annulus of the tricuspid ring and seemed to be in continuity with the tricuspid valve itself. Closure of the defect was carried out with continuous suture of 3-0 silk. After closing the defect the atrium was flooded with saline. Potts ductus clamps were applied to the atrial incision. The aortic clamps were removed and the superior vena cava was opened allowing blood to return to the heart. The total time of inflow stasis was 10 minutes. Heart action had been slow but regular during the period of open cardiotomy. Heart action continued to be feeble but regular for about one minute after the venae cavae had been opened, when ventricular fibrillation supervened. Prolonged cardiac massage, defibrillation with electric shocks and intracardiac potassium were used to no avail.

The significant postmortem findings were limited to the heart. The heart weighed 160 Gm. The right atrium was greatly enlarged and showed a recent surgical incision closed with silk. The great vessels appeared quite normal in position and appearance. The foramen ovale was patent at its anterior margin with a lumen of approximately 0.5 cm. by 1 cm. The right ventricle measured 5 mm. in thickness, as did the left. The tricuspid valve measured 8 cm. in circumference. There was a defect in the ventricular septum approximately 1 cm. by 2 cm. in size. Adjacent to this defect the septal leaflet of the tricuspid valve appeared deformed, being small, thickened and nodular (fig. 5). On the left side of the heart the interventricular septal defect presented in the aortic outflow tract high in the membranous portion of the left ventricular septum just beneath the aortic valve and between two adjacent commissures of the aortic cusps. On the right side the defect was associated with the septal leaflet of the tricuspid valve and presented just below the annulus of the valve. Several chordae tendineae of the valve leaflet were attached below the defect. There was a small amount of air in the coronary arteries.

**Comment**

These two cases illustrate an unusual type of abnormal communication between cardiac chambers. Whereas the usual type of defect in the membranous portion of the ventricular septum allows a shunt between the left ventricle and the right ventricle, in each of these patients the shunt through the interventricular septal defect was directed into the right atrium. Cardiac catheterization data in each case indicated the presence of a shunt of highly oxygenated blood entering the right atrium. Diagnostic considerations before operation included atrial septal defect, anomalous pulmonary veins draining into the right atrium and interventricular septal defect with tricuspid insufficiency and regurgitation of shunted blood back into the right atrium. Another anomaly which may result in a shunt of arterial blood into the right atrium is congenital aneurysm of a sinus of Valsalva of the aortic valve with erosion through the atrial wall and establishment of an aorticoatrial fistula.

In each of these cases the shunt was interpreted before operation as indicating an atrial septal defect. In retrospect there are several points which should have raised considerable question as to the correctness of this diagnosis. The presence of a loud, coarse systolic murmur accompanied by a readily palpable systolic thrill certainly indicated something other than uncomplicated atrial septal defect. Signs of tricuspid insufficiency were present in one case and in the other there was tremendous enlargement of the right atrium. Right ventricular and pulmonary arterial
hypertension was present to a considerable degree in both instances and in one patient approached systemic levels of pressure. This degree of right-sided hypertension is uncommonly encountered in atrial septal defects in young children. It would seem logical to suspect the presence of a left ventricular-right atrial shunt resulting from a high interventricular septal defect with a defective septal leaflet of the tricuspid valve in a patient presenting catheterization evidence of a shunt of highly oxygenated blood entering the right atrium and the clinical findings of a coarse systolic murmur and thrill over the midportion of the heart. The rare instances of aorticcoatrial shunt resulting from rupture of an aneurysm of a sinus of Valsalva into the right atrium might be differentiated from this syndrome by the presence of a continuous murmur over the base of the heart in most patients with aorticcoatrial shunts.

It was technically possible to close this form of ventricular septal defect through a right atrial approach in both of these cases, although the defective septal leaflet of the tricuspid valve was irreparable. Lack of understanding of the nature of the anomaly certainly did not facilitate its surgical handling in either instance. There have been several case reports of this type of ventricular defect, with minor modifications, which might lend themselves to surgical closure through a right atrial approach.2,3,4,5 These have been reviewed recently by Perry, Burchell and Edwards.6

The embryologic origin of such a congenital anomaly is of some interest. Anatomically, the membranous portion of the ventricular septum of the adult heart has a segment which lies between the floor of the right atrium and the aortic cone of the left ventricle. This is the so-called atrioventricular part of the membranous septum, which was formed from the right tuberces of the endocardial cushions of the atrioventricular canal and the conus septum.7 An arrest in the development of this portion of the septum might give rise to this combination of atrioventricular communication with defective formation of the tricuspid valve.

SUMMARY

Two unusual cases of a congenital communication between the left ventricle and the right atrium, with defective medial leaflet of the tricuspid valve, are reported. In both cases the diagnosis was misinterpreted as interatrial septal defect from data obtained at cardiac catheterization. In both cases, surgical closure was attempted with fatal results; post-mortem studies revealed the true nature of the defect.

SUMMARIO IN INTERLINGUA

Es presentate duo simile casos de inusual anormalitates del cardui. Existeva in illos un communication directe inter le ventriculo sinistre e le auriculo dextere via un defecto in le portion membranose del septo inter-ventricular. Isto eseva associate con un formation defective del foliolo medial del valvula tricuspid. In ambe casos datos es presentate que eseva obtenite per catheterisation, durante le operation, e al autopsia. Es discutite le diagnose differential de iste typo de lesion.

REFERENCES

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