Obstruction of Left Ventricular Outlet in Association with Ventricular Septal Defect

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A ventricular septal defect either as an isolated malformation or as part of many developmental complexes is the most common single developmental lesion of the heart. In an overwhelming percentage of cases, hearts bearing a ventricular septal defect whether or not other conditions are associated have no additional intracardiac lesion that causes obstruction to the normal egress of blood from the left ventricle.

In a pathologic collection of 722 hearts bearing malformations, we have observed 10 cases in which a ventricular septal defect was associated with obstruction of the left ventricular outlet. The purpose of this communication is to describe the varied anatomic arrangements encountered in these 10 cases and to report on their clinical features as well as the objective functional studies, when done.

One additional case that was observed clinically and at operation is also reported. This is done since it serves to make more nearly complete the classification of those anatomic arrangements in which the common denominators are ventricular septal defect and obstruction to left ventricular outlet.

The hemodynamic derangements that obstruction of the left ventricular outlet adds to a ventricular septal defect depend on the anatomic interrelationship between the ventricular septal defect and the site of obstruction and on the size of the defect itself. When the defect is large, the factor that determines the pressures in the chambers of the heart and the systemic circulation is the position of obstruction with respect to the interventricular communication. When the defect is small, it does not of itself affect the hemodynamics significantly.

In an introductory manner, it is to be emphasized that in some of the anatomic patterns the site of left ventricular obstruction lies proximal to the ventricular septal defect, while in others this relationship is reversed. In the former situation, the right ventricle and aorta lie in the same compartment, which is separated from the left ventricle by the site of left ventricular stenosis. Here, on anatomic grounds, it would be anticipated that the right ventricle and aorta would exhibit essentially similar systolic pressures, while the systolic pressure in the left ventricle would be of a higher order.

In the situation wherein the site of left ventricular obstruction lies distal to the ventricular septal defect, the 2 ventricles share a systolic compartment. The zone of obstruction lies between this systolic compartment and the aorta. Here, on anatomic grounds, it would be anticipated that the 2 ventricles would exhibit essentially equal systolic pressures, while the aortic pressure would be of a lower order.

These cases are presented in the framework of a classification based upon size of defect and site of obstruction. Some of the hemodynamic considerations are derived from direct measurements of pressure and saturation. In other instances, in which only limited or no direct measurements were made, certain assumptions based upon established physiologic principles are justified from the anatomic arrangements.

The varied arrangements of obstruction to left ventricular outlet coexisting with a ven-
tricular septal defect which we have encountered are illustrated in figures 1, 2, and 3. Some of these cases have been reported previously.\(^1\)\(^2\) They are again mentioned in this communication in the context of a classification (table 1).

I. Right Ventricle Freely in Communication with the Aorta or Left Ventricle

1. Right Ventricle and Aorta in Free Communication

   a. Subaortic Stenosis Below a Large Ventricular Septal Defect (Case 1)

   Clinical Features. A diabetic boy who was first seen at the Mayo Clinic in 1951 at the age of 4\(\frac{1}{2}\) years had been known to have a cardiac murmur from infancy. Examination in 1951 revealed a continuous murmur in the left infraclavicular fossa and a loud systolic murmur and thrill at the lower left sternal border. The thoracic roentgenogram revealed cardiomegaly with normal pulmonary arterial and aortic segments, and the pulmonary vasculature was normal. The electrocardiogram was interpreted as showing right ventricular hypertrophy. The working clinical diagnosis was patent ductus arteriosus. However, because some of the findings were unusual for classic patent ductus arteriosus, cardiac catheterization was advised. It was refused by the parents.

   Exploratory thoracotomy was recommended in the hope that the lesion was a patent ductus arteriosus, correction of which would lead to definitive cure. At operation on May 3, 1952, the ductus arteriosus was found to be closed. No further exploration was done.

   After an uneventful postoperative recovery the patient was followed over the next 8 years; his diabetes and cardiac lesion caused him little difficulty. In 1958, at 12 years of age and following a respiratory illness, the patient had signs of congestive heart failure and returned for care. He was admitted to the hospital on February 18, 1958, and treated with digoxin and diuretics; the congestive failure was relieved rapidly. Examination at that time revealed a thrill and a grade IV systolic murmur (graded on a basis of I to IV) at the lower left sternal border. The murmur was widely transmitted; it had a more musical quality at the apex, and extended to the axilla. At the base there was a grade-I diastolic murmur which with the systolic murmur gave a continuous bruit. The second sound in the pulmonary area was single and not accentuated. The thoracic roentgenogram (fig. 4a) showed moderate cardiomegaly and normal pulmonary vascular markings. The electrocardiogram (fig. 4b) showed sinus rhythm, a rate of 100 per minute, a P-R interval of 0.2 second, incomplete right bundle-branch block with a QRS complex lasting 0.11 second and an axis of \(+120^\circ\), and was interpreted as showing right ventricular hypertrophy, digitalis effect, and no evidence of left ventricular hypertrophy.
Ventricular septal defect in association with obstruction to the left ventricular outlet. Right and left ventricles in free communication. a. Subaortic stenosis above a ventricular septal defect (cases 4 to 8). In this complex, reported by Becu and associates, intra-cardiac defects are associated with obstruction of the aortic arch and a patent ductus arteriosus communicating with the descending aorta. (Reproduced with permission from: Becu, L. M., Tauxe, W. N., DuShane, J. W., and Edwards, J. E.: A complex of congenital cardiac anomalies: Ventricular septal defect, biventricular origin of the pulmonary trunk, and subaortic stenosis. Am. Heart J. 50: 901, 1955.) b. Anomalous chordal insertion of the anterior mitral leaflet causing obstruction of the left ventricular outlet in association with a ventricular septal defect of the type in the complex known as persistent common atroventricular canal (case 9). In this case the peculiar chordal insertion of the cleft anterior mitral leaflet caused obstruction in the left ventricular outlet. The letter “A” appears on each half of the cleft anterior mitral leaflet. c. Subaortic stenosis opposite a ventricular septal defect (case 10). No functional obstruction to the left ventricular outlet exists when the ventricular septal defect is present. As shown in the insert, when the defect was closed with a prosthetic patch (Ivalon) the existing mass of muscle in the left ventricular outlet was effective in causing left ventricular obstruction.

Cardiac catheterization was done on March 28, 1958 (table 2), and the patient was dismissed from the hospital without evidence of congestive heart failure. He was to return for evaluation in several weeks. About 3 weeks later he became suddenly ill with fever and prostration. He was admitted to the hospital in his home community but died shortly thereafter (on April 13, 1958) in congestive heart failure.

Pathologic Features. Necropsy performed by one of us (J.E.E.) disclosed in the upper part of the outflow tract of the left ventricle a severe degree of subaortic stenosis (fig. 5a) in which the lumen of the tract was reduced to a diameter of only 7 mm. The zone of obstruction was characterized by a cylindrical deposit of dense fibrous tissue involving the anterior wall of the left ventricle, the ventricular septum, and the ventricular aspect of the anterior leaflet of the mitral valve. The zone of obstruction extended for about 8 mm. in a supero-inferior direction. Its uppermost aspect lay about 1 cm. below the lower part of the aortic valve.

Immediately above the site of subaortic stenosis was a ventricular septal defect measuring 1.2 cm. in diameter (fig. 5b). On the left the defect lay beneath the posterior and right aortic cusps. On the right it lay immediately postero-inferior to the crista supraventricularis (fig. 5c).

In the left ventricle just above the upper edge of the ventricular septal defect and just below the aortic valve a membrane of fibrous tissue protruded horizontally into the lumen. At the level of this horizontal membrane the caliber of the left ventricular outflow tract was reduced to 1.0 cm.

The aortic valve orifice measured 2.2 cm. in diameter. The aortic cusps were irregularly thickened and moderately friable. Representative histologic examination of the aortic valve showed a picture of healing bacterial endocarditis.

The left ventricular wall was markedly hypertrophied, measuring 1.5 cm. in thickness. The right ventricle, though less thickened than the left, was hypertrophied and 0.9 cm. thick.

The valves other than the aortic were normal.

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LEFT VENTRICULAR OBSTRUCTION

The foramen ovale and ductus arteriosus were closed. The coronary arteries were normal.

Histologic examination of the lungs showed edema, hemosiderosis, and medial hypertrophy of the arterioles and muscular arteries. No intimal lesions were identified in these vessels.

Comment. At catheterization of the right side of the heart a large left-to-right shunt at ventricular level was noted. There was close approximation of the systolic pressures in the right ventricle, pulmonary artery, and aorta—the findings usually associated with an un-complicated large ventricular septal defect.

An area of subaortic stenosis was situated below the ventricular defect. Thus it may be assumed that the pressure in the left ventricle was elevated over that in the systolic compartment composed of the right ventricle, pulmonary artery, and aorta. This assumption of elevated left ventricular pressure is supported by the finding of undue left ventricular hypertrophy in the specimen.

In the face of clinical findings and the data obtained when the right side of the heart was catheterized, which were consistent with a large ventricular septal defect, it would be unusual to proceed with catheterization of the left side of the heart. However, if this had been done the presence of the subaortic stenosis probably would have been detected.

b. Origin of Both Great Vessels from the Right Ventricle with a Small Ventricular Septal Defect as the Only Route of Egress from the Left Ventricle (Case 2)

Clinical Features. The patient was first seen at the Mayo Clinic in 1948 at the age of 2½ years. A heart murmur had been noted at birth. Symptomatically, the patient had previously had 2 transient episodes of cyanosis, and became short of breath and perspired profusely with slight exertion. Physical examination revealed a thrill and a loud systolic murmur at the lower left sternal border. The second pulmonic sound was single. Ecg oximetry showed a systemic arterial saturation of 85 per cent at rest and 77 per cent with exercise.

The radiologic configuration of the heart and electrocardiographic changes were consistent with the diagnosis of tetralogy of Fallot. However, the murmur was not considered typical of this condition.

Three years later, in 1951, at another center, the heart was catheterized. The findings (table 3) were consistent with a diagnosis of tetralogy of Fallot. Therefore a Brock valvotomy ("closed" pulmonary valvotomy and infundibular resection) was done in 1951.

The patient was next seen at the Mayo Clinic in 1958 at the age of 13 years. He had no visible cyanosis and only slight limitation of exercise. Physical examination revealed a thrill at the lower left sternal border and a very loud, grade-IV systolic murmur heard in the same area and extending to the base, axilla, apex, and posterior part of the thorax. Diastole was clear. The second sound in the pulmonic area was single and of normal intensity. A thoracic roentgenogram showed moderate cardiomegaly, an upturned apex, a concave pulmonary segment, and normal pulmonary vascular markings (fig. 6a). The electrocardiogram showed sinus rhythm, a rate of 75 per minute, and an axis of –130°; it was interpreted as showing right ventricular hypertrophy and no evidence of left ventricular hypertrophy (fig. 6b).
Evidence for elevated left ventricular pressure related to obstruction at the small ventricular septal defect was additionally supported by a prominent jet lesion on the anterolateral aspect of the right ventricle opposite the ventricular septal defect and just beneath the aortic valve. The atrial septum showed a very small defect based upon a short valve of the foramen ovale.

Histologic examination revealed normal to thinner-than-normal arterioles in the lungs and widened arterioles in the visceral pleura, suggesting an enlarged collateral system to the lungs.

Comment. In this condition the right ventricle and aorta are in the same hemodynamic vascular compartment, which may be termed “right ventricular-aortic compartment.” The presence or absence of pulmonic stenosis determines whether the pulmonary artery is in free communication with this compartment.

A case similar in some respects to this was reported by Edwards and associates in 1952. For completeness in reporting our material it is included here as case 3. These authors described a heart in which both great vessels arose from the right ventricle and were associated with a small membranous ventricular septal defect that was sealed by the adhesion of the anterior mitral leaflet to its edges. There was a tiny accessory opening in the anterior mitral leaflet that corresponded in position with the adhesion of the mitral valve to the septal defect, thus allowing communication between the left atrium and the right ventricle. This opening and a small patency

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### Table 2

**Synopsis of Cardiac Catheterization Data in Case 1 (Subaortic Stenosis Below a Large Ventricular Septal Defect)**

<table>
<thead>
<tr>
<th>Site of sampling</th>
<th>Pressure, mm. Hg</th>
<th>Oxygen saturation, per cent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>—</td>
<td>63</td>
</tr>
<tr>
<td>Right atrium</td>
<td>21/8</td>
<td>58</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>107/11.24</td>
<td>79-88</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>117/49</td>
<td>89-91</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>117/59</td>
<td>100</td>
</tr>
</tbody>
</table>

*Pressure and saturation data obtained during administration of 100 per cent oxygen.

†Saturations determined with cuvette oximeter. Dye-dilution curves from the right side of the heart with sampling at the radial artery revealed a right-to-left shunt of about 5 per cent. The left-to-right shunt calculated from saturation data was about 54 per cent.

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### Table 3

**Synopsis of Cardiac Catheterization Data in Case 2**

<table>
<thead>
<tr>
<th>Chamber</th>
<th>Pressure, mm. Hg</th>
<th>Oxygen saturation, per cent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>—</td>
<td>60</td>
</tr>
<tr>
<td>Right atrium</td>
<td>0 to −4</td>
<td>63</td>
</tr>
<tr>
<td>Right ventricle at apex</td>
<td>8/0</td>
<td>74</td>
</tr>
<tr>
<td>Right ventricle at pulmonary valve</td>
<td>100/8</td>
<td>73</td>
</tr>
<tr>
<td>Pulmonary veins</td>
<td>4/−2</td>
<td>99</td>
</tr>
<tr>
<td>Aorta via right ventricle</td>
<td>62/54</td>
<td>84</td>
</tr>
</tbody>
</table>

*Cardiac catheterization done at the University of Minnesota Hospitals in Minneapolis in 1951 and presented here with their permission.

†Cuvette method.
at the foramen ovale were the only routes of egress from the left side of the heart. There was no infundibular or valvular pulmonary stenosis. Thus the right ventricle, aorta, and pulmonary artery were in the same systolic compartment. The left ventricle was separated completely from this compartment. It is reasonable to assume, therefore, that equal systemic systolic pressures had existed in the right ventricle, pulmonary artery, and aorta, while the left atrial pressure and left ventricular pressure were markedly elevated above normal.

In the case reported herein (case 2) the left ventricle and pulmonary artery were separated from the right ventricular-aortic compartment by a very small ventricular septal defect and by infundibular and valvular pulmonary stenosis respectively. These anatomic considerations make understandable the facts demonstrated by cardiac catheterization that the right ventricular and aortic systolic pressures were equal and that a pressure gradient existed across the pulmonary valve. It is assumed that in the left ventricle, since its only outlet was a small ventricular septal defect, the systolic pressure was greater than in either the right ventricle or the aorta. The hypertrophy of the left ventricle and the jet lesions in the right ventricle opposite the ventricular septal defect support this view.

The anatomic arrangements are such that catheterization of the right side of the heart yields findings similar to those in tetralogy of Fallot.

It is of interest to postulate that differentiation of this situation might have been accomplished by special technics (catheter position or angiography) that would have demonstrated that the aortic valve lay more to the right and at a more superior plane than it does in tetralogy of Fallot. Further, selective angiography of the left atrium or left ventricle might have allowed the left ventricular obstruction to have been suspected, especially if it were combined with angiography of the right side of the heart. The former might have revealed the small ventricular septal defect with perhaps delayed emptying of the left ventricle. The latter might have demonstrated the abnormal origin of the aorta.

*Subaortic stenosis below a large ventricular septal defect (case 1). a. Posteroanterior roentgenogram of thorax. b. Electrocardiogram.*

Figure 4

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2. Right Ventricle and Left Ventricle in Free Communication

a. Subaortic Stenosis Above a Ventricular Septal Defect (Cases 4 to 6)

A group of five cases was reported from the Mayo Clinic in 1955 by Beeu and associates. These portrayed an anatomic complex of a ventricular septal defect situated anteriorly in the right ventricular outflow tract (not involving the membranous septum), subaortic stenosis resulting from a muscular ridge lying across the outflow tract of the left ventricle, obstructive anomalies of the aortic arch system (interruption of the aortic arch or coarctation), and patent ductus arteriosus. A similar case was noted by Taussig. A diagrammatic representation is shown in figure 2a.

Comment. In the 5 cases reported by Beeu and associates each infant died during its first month of life. None had cardiac catheterization.

In this situation both ventricles and the pulmonary artery are in the same systolic compartment. Subaortic stenosis separates the ascending aorta from this compartment; coarctation or interruption of the arch separates the ascending aorta from the descending aorta. A patent ductus arteriosus connects the pulmonary artery with the descending aorta. Thus it can be assumed that equal systolic

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Figure 5

Subaortic stenosis below a large ventricular septal defect (case 1). a. The left ventricle viewed from below. In the outflow tract (point of arrow) is a constricting fibrous ring representing the lower portion of the subaortic stenosis. L.V. = left ventricle. L.A. = left atrium. The anterior leaflet of the mitral valve is photographed from the perspective of its lower edge. b. The subaortic portion of the left ventricle and the aortic valve opened. The subaortic stenosis lies above the point of the arrow. The ventricular septal defect (D.) lies above the subaortic stenosis. Vegetations of bacterial endocarditis are present upon the aortic valve. Between the aortic valve and the ventricular septal defect a semicircular shelf of fibrous tissue protrudes into the subaortic tract. A.M. = anterior leaflet of mitral valve. c. Right ventricle and pulmonary valve. The ventricular septal defect (D.) lies below the crista supraventricularis and above the papillary muscle of the conus (P.M.C.). P.V. = pulmonary valve. R.V. = inflow portion of right ventricle.
pressures existed in the right ventricle, left ventricle, pulmonary artery, and descending aorta, while the pressure in the ascending aorta was lower than in these chambers or vascular segments.

b. Anomalous Chordal Insertion of a Cleft Anterior Mitral Leaflet Causing Obstruction of Left Ventricular Outflow in Association with a Ventricular Septal Defect of the Persistent Common Atrioventricular Canal Type (Case 9)

Clinical History. A 26-month-old girl was referred to the Mayo Clinic for cardiac evaluation in 1958. She had a heart murmur known from 7 weeks of age. During the first 15 months of life she had gained poorly and had frequent episodes described as pneumonia. Since the age of about 16 months her general condition had improved. She had gained weight more rapidly and the respiratory infections occurred less often. There was no history of exertional dyspnea, cyanosis or anoxic episodes.

Physical examination showed a very small but vigorous female child. Her heart was moderately overactive. There was no thrill. There was a grade II systolic murmur maximal at the second left interspace with extension to midprecordium and fading toward the apex. There was a grade II diastolic murmur maximal at the second and third left interspaces. The second pulmonic sound was moderately accentuated and split. The blood pressure in the brachial artery determined by the cuff method was 120 mm. Hg systolic and 80 mm. diastolic. The femoral pulsations were easily palpable bilaterally.

A thoracic roentgenogram (fig. 8a) showed moderate cardiomegaly with a full pulmonary arterial segment and increased pulmonary vascular markings. The electrocardiogram (fig. 8b) showed sinus rhythm, a rate of 120 per minute, and an axis of $-70^\circ$. The changes were interpreted as those of right ventricular hypertrophy, with evidence of left ventricular overload for age. The latter was thought to be present because of the counterclockwise frontal loop in the frontal plane with marked left axis deviation of the mean QRS vector in so young a patient. Although the electrocardiogram in itself was suggestive of persistent common atrioventricular canal, the total clinical picture was considered more consistent with that of the usual ventricular septal defect with associated pulmonary valvular insufficiency. The patient was referred for surgical repair with the aid of extracorporeal circulation.

At operation, before the heart was opened, the pressure in millimeters of mercury was 108/54 in the pulmonary artery and 106/10 in the right ventricle, while the cuff pressure in the brachial artery was 105/80. A small patent ductus arteriosus was

Figure 6

Origin of both great vessels from the right ventricle with a small ventricular septal defect as the only route of egress from the left ventricle (case 2). a. Posteroanterior roentgenogram of thorax. b. Electrocardiogram.
ligated. Palpation over the left atrium revealed no thrill of mitral insufficiency. On opening the right atrium, no atrial septal defect or tricuspid insufficiency was noted.

Through an incision in the right ventricle, a ventricular septal defect in the position of the common atrioventricular canal was noted. This defect lay under a slightly eleft septal tricuspid leaflet. The defect was repaired with a Teflon prosthesis.

When the heart had resumed action following the repair, the systolic pressure was 90 to 95 mm. Hg in the pulmonary artery and right ventricle, and 95 to 100 in the aorta and left ventricle. Thus no fall in pulmonary pressure followed repair. After extracorporeal circulation, the patient's condition was precarious and about 1 hour postoperatively she died.

Pathologic Features. A ventricular septal defect was present beneath the anterior leaflet of the mitral valve and the septal leaflet of the tricuspid valve. On the right side the defect extended to the crista supraventricularis. While the anterior extent of the defect was not unusual, the fact that it extended farther under the septal leaflet of the tricuspid valve than usual placed it in the category of an interventricular communication in persistent common atrioventricular canal (fig. 9a). The septal leaflet of the tricuspid valve had a small cleft. Viewed from the left side (fig. 9b) the defect lay beneath the anterior leaflet of the mitral valve. The mitral valve was the site of gross abnormality with a cleft involving the anterior leaflet. The chordal attachments of the mitral valve were abnormal. There were 2 sets of papillary muscles and the usual posteromedial and anterolateral sets of chordae. In addition to these chordae there were chordae that ran from the edges of the cleft of the anterior leaflet of the mitral valve and inserted into the ventricular septum. This made the anterior half of the anterior mitral leaflet relatively immobile and caused the approach to the outflow tract of the left ventricle to be relatively confined. The confined area lay between the ventricular septum, the anterolateral commissure of the mitral valve, the anterior half of the anterior mitral cusp, and the chordae that joined the anterior half of the anterior mitral cusp to the ventricular septum. The lower part of the outflow tract was thereby compromised, measuring only about 1 cm. in diameter.

The aortic arch system was anomalous. Five branches arose from it. The first branch was the right common carotid artery, the second the left common carotid artery, the third the left vertebral, and the fourth the left subclavian; each of these arose from the most superior aspect of the aortic

**Figure 7**

*Origin of both great vessels from the right ventricle with a small ventricular septal defect as the only route of egress from the left ventricle (case 2). a. Outflow portion of right ventricle (R.V.). The aorta rises exclusively from the right ventricle beside the pulmonary trunk (P.T.). The subpulmonary portion of the outflow tract is somewhat narrowed, in part by the presence of the hypertrophied crista supraventricularis (C.S.). The position of the narrow ventricular septal defect is marked by the presence of a probe, opposite the ventricular septal defect and immediately beneath the aortic valve there is endocardial thickening of the right ventricle representing a reaction to the impact of the jetlike stream coming from the small ventricular septal defect. b. Left atrium and left ventricle. Marked left ventricular hypertrophy. There is anatomic evidence of the obstructive nature of the small ventricular septal defect (defect not shown in this perspective).*
arch. Arising as the fifth branch of the arch, and from the posterior wall of it, was the right subclavian artery, which passed behind the esophagus to reach the right arm.

Immediately distal to the origin of the right subclavian artery was a site of typical coarctation at which the aortic lumen measured 3 mm. in diameter. Distal to the coarctation was a slightly patent ductus arteriosus, which had a lumen of 3 mm. as it joined the aorta.

Histologic examination of the lungs revealed marked (grade IV) changes of hypertensive pulmonary vascular disease, characterized by the formation ofplexiform lesions and generalized arterial dilatation.4

Comment. In this case the insertion of the anterior mitral leaflet resulted in obstruction to left ventricular outflow above the ventricular septal defect. This anatomic arrangement creates an intracardiac functional situation which resembles that in the cases reported by Beeu and co-workers.2 The right ventricle, pulmonary artery and left ventricle were in the same systolic compartment, while the left ventricular outflow tract and the ascending aorta were separated from the intracardiac systolic compartment by the anomalously inserted anterior mitral leaflet. The descending aorta was separated from the ascending aorta by a coarctation and from the pulmonary artery by a very small patent ductus arteriosus.

Thus it may be assumed that the systolic pressure was uniform in the right ventricle, pulmonary artery and left ventricle, decreased in the ascending aorta, and still further decreased in the descending aorta.

c. Subaortic Stenosis Opposite a Ventricular Septal Defect (Case 10)

In our collection is another case portraying subaortic stenosis opposite a ventricular defect but without obstruction of the aorta itself.

Clinical History. A 3½-year-old girl was referred to the Mayo Clinic in 1958. She was known to have had a murmur from 2 months of age. The parents had noticed that she had always be-
Figure 9
Anomalous chordal insertion of the anterior mitral leaflet causing obstruction of left ventricular outflow in association with a large ventricular septal defect (case 9). a. Right atrium and ventricle. The septal leaflet of the tricuspid valve (S) has a small cleft above the ventricular septal defect (D) of the type seen in persistent common atrioventricular canal. There is hypertrophy of the right ventricle. The atrial septum is intact. b. Left atrium and left ventricle. A ventricular septal defect of the type seen in persistent common atrioventricular canal lies opposite and below a cleft in the anterior mitral leaflet (A). The anterior mitral leaflet has both normal and abnormal chordal attachments. The normal attachments are to the papillary muscles. The abnormal attachments run from the edges of the cleft in the leaflet to the ventricular septum. The peculiar chordal attachments of the anterior half of the cleft anterior leaflet cause obstruction to the left ventricular outflow.

come easily fatigued and manifested grunting respirations with slight exertion. Cyanosis had never been observed.

At 1 year of age, cardiac catheterization performed at another center had revealed a left-to-right shunt at the ventricular level, with pressures in millimeters of mercury as follows: right ventricle 70/0-11, pulmonary artery 65/25, left ventricle 86/0-12, and aorta 86/50. The catheter entered the aorta and left ventricle from the right ventricle. During the next 2 years the patient had gained fairly well but continued to fatigue easily.

At the age of 3½ years physical examination revealed an overactive heart, a grade-III systolic murmur maximal at the fourth left interspace with extension to the second left interspace and the apex. There was an apical diastolic inflow rumble. The second sound in the pulmonic area was split and the second component was moderately accentuated.

The thoracic roentgenogram (fig. 10a) revealed moderate cardiomegaly, an enlarged pulmonary artery segment, and markedly increased pulmonary vascular markings. The electrocardiogram (fig. 10b) showed sinus rhythm, a rate of 109 per minute, a P-R interval of 0.20 second, an axis of −110°, right ventricular hypertrophy, and left ventricular hypertrophy.

The clinical diagnosis was ventricular septal defect with pulmonary hypertension and a large left-to-right shunt. Operation was advised.

During thoracotomy but before any repair, the pressure was 79/6 in the right ventricle and 95/41 in a systemic artery.

With the aid of extracorporeal circulation and asystole, 2 ventricular septal defects were identified. One was in the usual location, lying posteroinferior to the crista supraventricularis and extending under the tricuspid ring along almost the entire extent of its septal leaflet. This defect measured 1.6 cm. in diameter. A second defect in the crista supraventricularis was represented by 2 small openings, one measuring 2 mm. and the other 4 mm. in diameter. Muscular tissue was present between this defect and the pulmonary valve. The large defect was closed by placement of an Ivalon sponge. The smaller (muscular) defects were closed by direct suture. After the right ventricle had been closed and the heart had resumed its function the pressure was 52/7 in the right ventricle and 85/36 in a systemic artery.

Immediately following the surgical procedure flow (containing probe). The obstruction is caused by the chordae holding the leaflet taut near the ventricular septum. There is left ventricular hypertrophy. The atrial septum is intact.
complete heart block was noted. All measures failed to maintain an adequate systemic blood pressure, and death occurred 24 hours postoperatively.

Pathologic Features. Necropsy revealed the repaired defects described. From the left ventricular aspect a mass of muscle was seen protruding from the upper part of the muscular ventricular septum and adjacent anterior left ventricular wall. The mass lay at the level of the larger ventricular septal defect, and so, slightly below the aortic valve. It protruded into the left ventricular outflow tract and narrowed it to a diameter of 1.0 cm. (fig. 11). The position of the protruding mass was such that repair of the ventricular septal defect with the prosthetic patch caused the outflow tract to become obstructed. Apparently, before the patch was placed, blood could be diverted around the mass in the space of the septal defect and so reach the aorta without barrier (fig. 2c).

The smaller defects of the ventricular septum that were observed and closed during the operation opened into the left ventricle through the proximal portion of the protruding mass described.

The left ventricle and right ventricle were hypertrophied, their walls being 1.2 and 0.8 cm. thick respectively. The left atrium was dilated and its endocardium was uniformly thickened. The valves were normal. The ductus arteriosus was closed.

Histologic sections of the lungs showed mild medial hypertrophy and no significant intimal lesions.

Comment. In this case there was subaortic stenosis opposite a ventricular septal defect. Unobstructed flow to the aorta from the left ventricle was considered to have been present before the larger defect was closed. Before closure, the hemodynamic aspects were similar to those frequently encountered in uncomplicated ventricular septal defects.

When the defect was closed the detour allowing free egress from the left ventricular chamber was abolished and obstruction to left ventricular outflow resulted. Although the pressure measured in the peripheral systemic circuit following repair of the defect was at first within normal range, it may be assumed from the anatomic details that postoperatively the left ventricular pressure was elevated above that measured in the aorta.

II. Right Ventricle Not in Free Communication with the Left Ventricle or Aorta

1. Coexisting Right Ventricular Infundibular Stenosis and Subaortic Stenosis Associated with Small Ventricular Septal Defect (Case 11)

In 1957 a 13-year-old girl was referred to the Mayo Clinic for cardiac evaluation and treatment. She was known to have had a heart murmur from birth. Her growth and development had been normal. During the previous year she had had easy fatigability, dyspnea with slight exertion, and periods of orthopnea. There was no history of cyanosis or anoxic episodes.
Physical examination revealed a large 13-year-old girl with normal color. There was a systolic thrill in the suprasternal notch, a grade-II, harsh stenotic type of systolic murmur in the first right interspace with transmission to the neck, and a grade-III, harsh systolic murmur maximal in the third left interspace with extension to midprecordium and the apex. Neither the pulmonic nor the aortic second sound was audible. Diastole was clear. The femoral pulsations were normal. The blood pressure in the arms was 90/70.

The thoracic roentgenogram (fig. 12a) showed marked cardiomegaly. The pulmonary vascular markings were considered to be within normal range. The electrocardiogram (fig. 12b) showed sinus rhythm, a rate of 84 per minute, and right axis of +120°. There was evidence of combined overloading of both the right and the left ventricle.

A clinical diagnosis of combined pulmonary and aortic stenosis was made and surgical repair recommended.

At the time of thoracotomy but prior to any repair, the pressures in millimeters of mercury were: pulmonary artery 20/13, just below the pulmonary valve in the right ventricle 20/6, low in the right ventricle 170/6, left ventricle 188/9, and ascending aorta 108/61.

Under extracorporeal circulation, right ventriculotomy was performed. The right ventricular wall was markedly hypertrophied, being about 2.5 cm. thick. There was obvious infundibular stenosis, with an orifice 5 mm. in diameter, and a small infundibular chamber about 1 cm. in length beneath a normal pulmonary valve. There was a small ventricular septal defect about 5 mm. in diameter situated well behind the crista supraventricularis just at the angle between the anterior and septal leaflets of the tricuspid valve.

The site of the aortic valve prosthesis was determined and the ventricle septed and the anterior wall into the left ventricular cavity opposite the larger ventricular septal defect. b. The left ventricle in essentially the same perspective as shown in a but with the prosthesis used to close the larger ventricular septal defect removed. The mass of muscle seen in the subaortic position (Su.S.) lies opposite the ventricular septal defect. c. Right ventricular aspect of the larger ventricular septal defect after the prosthesis has been removed. The mass of muscle protruding from the anterior septal wall of the left ventricle (Su.S.) is shown in its position opposite the larger ventricular septal defect. Before closure of the larger ventricular septal defect blood could be diverted around the mass of muscle to reach the aorta. After closure of the defect with the prosthetic device this route was eliminated and subaortic stenosis followed as illustrated in figure 2c.
of infundibular pulmonary stenosis was resected and the ventricular septal defect closed by direct suture.

On opening of the ascending aorta the aortic valve was found to be normal. Immediately beneath this valve was a tough but essentially membranous subaortic stenotic area with an orifice measuring about 6 mm. in diameter. Through the aortic valve an incision was made in the subaortic membrane.

When the heart had resumed normal action after repair of the defects the pressure in the left ventricle was 120/10, while the aortic pressure was 112/49. The pressure in the pulmonary artery was 35/14, while that low in the right ventricle was 41/6.

The patient had an uneventful postoperative recovery. When last seen 1 year postoperatively she had no symptoms referable to the cardiovascular system. She had a grade II to III systolic murmur in the second, third, and fourth left interspaces with extension to the neck, and a moderately loud, grade I to II diastolic murmur maximal in the third and fourth left interspaces. The clinical impression was that of aortic insufficiency, which may have been the result of gradual scarring of the aortic leaflets.

Comment. When a ventricular septal defect is tiny it contributes little to any hemodynamic situation. In this case the elevated pressures in the ventricular chambers were the result of the stenotic outflow lesions. The pressure generated in each ventricular cavity depended only upon its relationship to the resistance offered by the stenotic lesion.

Although the pressures in the right and left ventricles prior to repair were almost of equal magnitude, this was considered to be fortuitous, for the ventricular septal defect was so small that free communication did not exist between the ventricular chambers.

Discussion

The cases presented herein are examples of ventricular septal defects associated with obstruction to the left ventricular outlet. The advent of surgical repair for ventricular septal defects has made the recognition of the complexes reported of practical significance. For, if the obstruction is unrecognized, repair of
the ventricular septal defect may leave the patient with obstruction to the left ventricular outlet or, in rare instances, may create obstruction that was not present preoperatively.

Clinically, isolated obstruction of left ventricular outflow is suspected when a murmur suggesting a stenotic lesion is heard in the aortic area, especially when the electrocardiogram shows evidence of left ventricular hypertrophy of the "systolic overload" type.  

In all but one (case 11) of the cases presented herein the murmurs were not recognized as those usually associated with obstruction to left ventricular outflow. In cases 1, 2, and 9 the lesions producing obstruction to left ventricular outflow were not related to the aortic valve, so that the murmurs produced were neither in the location nor of the quality of a usual aortic or subaortic stenosis.

In cases 4 to 8, wherein the subaortic stenosis was above the ventricular septal defect, the stenotic lesions were more closely related to the aortic valves. Perhaps in these cases phonocardiography, had it been done, would have revealed the "classic" diamond-shaped murmur of aortic stenosis.

In case 10 the obstruction was not present prior to operation. Thus, a murmur related to left ventricular obstruction could not be expected to exist preoperatively.

In cases 1, 2, and 11, left ventricular outflow was obstructed, and thus left ventricular overload patterns might be logically anticipated in the electrocardiogram. However, such patterns did not occur consistently. Only the case of coexisting right ventricular infundibular and subaortic stenosis associated with a very small ventricular septal defect (case 11) exhibited this electrocardiographic characteristic.

In cases 4 to 8 and 10 the obstruction to left ventricular outflow lay above the ventricular defect. The obstructing lesion in these anatomic arrangements did not result in left ventricular "systolic overload," for the obstruction could be bypassed through the ventricular septal defect and lead to a large left-to-right shunt.

In these cases electrocardiographic patterns were encountered that were like those in simple large ventricular septal defect with large left-to-right shunt. The observations are in concert with the hemodynamic features peculiar to these cases.

In cases 1 and 2, wherein the right ventricle was in free communication with the aorta, cardiac catheterization revealed equal right ventricular and systemic arterial systolic pressures, a relationship usually associated with an uncomplicated large ventricular septal defect or tetralogy of Fallot. With this anatomic arrangement, catheterization of the left side of the heart would be expected to demonstrate the obstructing lesion. However, since neither the auscultatory nor the electrocardiographic findings caused a suspicion of left ventricular obstruction, left ventricular pressures were not determined.

In cases 4 to 8, wherein the site of obstruction to the left ventricular outlet lay above a large ventricular septal defect and a large patent ductus arteriosus communicated with the descending aorta, one expects that there would be no differential in pressure between the 2 ventricles, the pulmonary artery and the descending aorta; but a differential would exist between the cardiac systolic compartment and the ascending aorta. Measurements of pressure in both the ascending and the descending aorta would be necessary to reveal the presence of the subaortic stenosis and the coarctation.

In the exceptional instance of case 10, obstruction is only potential, and preoperative studies of pressure would not indicate the presence of left ventricular obstruction.

Thus it can be seen that the preoperative diagnosis of left ventricular obstruction associated with a ventricular septal defect may prove not only difficult in some cases but impossible in others. While the auscultatory, roentgenographic, electrocardiographic, or cardiac-catheterization findings obtained by usual methods may readily establish the presence of a ventricular defect, they may not reveal that additional obstruction to the left ventricular outlet exists.
Summary

In a pathologic collection of 722 congenital heart lesions we have observed 10 cases in which a ventricular septal defect was associated with obstruction to normal left ventricular outflow. These cases are presented in the framework of a classification relating hemodynamics to the position of the obstructing lesion and the size of the ventricular septal defect. One additional case observed clinically and at operation is also reported to make more nearly complete the classification of the anatomic arrangements in which the common denominators are ventricular septal defect and obstruction to normal ventricular outflow.

Summario in Interlingua

In un collection pathologic de 722 congenite lesions cardiac, nos ha observate 10 casos in que un defecto ventriculo-septal esseva associate con un obstruction del normal effluxo sinistro-ventricular. Iste casos es presentate intra un cadro de classification que relaciona le hemodynamica al sito del lesion obstructive e al dimensiones del defecto ventriculo-septal. Es etiam reportate un caso additional que esseva observate clinica- e chirurgicamente pro render plus complete le classification del configurationes anatomic in que denominatres comun es defecto ventriculo-septal e obstruction al normal effluxo ventricular.

References


Obstruction of Left Ventricular Outlet in Association with Ventricular Septal Defect

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