Abnormal Communication between the Aorta and Left Ventricle

Aortico-Left Ventricular Tunnel

By Robert N. Cooley, M.D., Leonard C. Harris, M.D., and Alvin E. Rodin, M.D.

LEVY et al. described "aortico-left ventricular tunnel" as an abnormal communication that begins in the ascending aorta above the level of the coronary arteries, bypasses the aortic valve, and terminates in the left ventricle, resulting in aortic insufficiency. The authors described three cases and stated that four similar cases had been previously described. A review of the original papers suggests, however, that in three instances the history and anatomic findings were more compatible with rupture of a congenital aneurysm of a sinus of Valsalva than with an aortico-left ventricular tunnel. Levy et al. made the point that in their three cases, and in another previously described by Edwards, the tunnel originated or joined the ascending aorta above the sinus of Valsalva and the sinuses were normal. Furthermore, evidence of the presence of the tunnel appeared during infancy, suggesting that it was present at birth and not the result of a postnatal rupture.

An abnormal communication between the aorta and the left ventricle may also be secondary to a rupture of either a congenital or acquired aneurysm of a sinus of Valsalva. Congenital aneurysms usually come to the attention of the physician only after rupture into one of the adjacent cardiac chambers, although the diagnosis prior to rupture can be made by angiocardiography. Up to 1961 Sakakibara and Konno found 52 well-documented cases of aneurysm of the sinuses of Valsalva with rupture, and in three of these, an aneurysm of the right sinus had ruptured into the left ventricle. The average age of rupture, as ascertained by history and physical examination, of the entire 52 cases was 32 years, and the youngest with rupture into the left ventricle was 38 years. Heiner et al., however, described a patient who developed a continuous cardiac murmur between the first and ninth weeks of life and in whom at autopsy an aneurysmal channel extended from the left sinus of Valsalva to the left atrial appendage. The histologic structure of the channel was quite compatible with a rupture of an aneurysm of a sinus of Valsalva. Also, Jones and Langley believed that in five of 25 cases of congenital aneurysm of the sinuses of Valsalva a rupture had occurred before birth, but details concerning these cases are lacking. Certainly, cases with convincing evidence of rupture of the aneurysm during the first year of life are extremely rare.

The case presented here is considered as a probable example of an aortico-left ventricular tunnel as described by Levy et al. in that the channel appears to have been well formed at birth and different in histologic structure from a rupture of an aneurysm of a sinus of Valsalva. On the other hand, the channel communicated with an aneurysm of the right sinus of Valsalva instead of the ascending aorta, and there were small aneurysms of the posterior and left sinuses. We are reporting our case because of its rarity; also, this condition is susceptible to surgical correction and can be readily diagnosed by clinical and radiologic studies.

Case Report

This white male infant was first seen at the age of

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of three days because of a heart murmur detected by a physician shortly after birth. The mother was tall but proportionately built, the father was of normal height, and there were no stigmata of the Marfan syndrome in the parents.

Physical examination showed a vigorous baby

**Figure 1**

Electrocardiograms at three days and seven months of age. Progressive left ventricular hypertrophy is indicated.

**Figure 2**

A. Chest film exposed on third day of life showing enlargement of the left ventricle. The ascending aorta is also dilated. B. Chest film exposed at 16 months. The left ventricular enlargement has been progressive and is now very marked, and there is also marked dilatation of the ascending aorta.

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in no distress. The brachial and femoral pulses were abnormally full, and the blood pressure by the flush method was 75 mm. Hg in the right arm. The maximum cardiac impulse was forceful and in the sixth intercostal space in the midclavicular line. A grade-IV/VI harsh to-and-fro murmur was heard best on both sides of the midsternum. The second sound in the pulmonary area was single and of normal intensity. The electrocardiogram (fig. 1) showed evidence of left ventricular hypertrophy. Chest film (fig. 2A) showed cardiac enlargement and a contour compatible with left ventricular enlargement.

On a return visit at four months of age, the murmurs were accompanied by a thrill to the right and left of the sternum at the third intercostal space. An ejection type systolic murmur was best heard in the aortic area. The harsh diastolic murmur was best heard in both the aortic and pulmonary areas and continued past mid-diastole (fig. 3). The clinical impression was aortic stenosis and insufficiency with an alternate possibility of a fistula between a coronary artery and the right ventricle.

Exertional dyspnea appeared at 7 months of age, and the electrocardiogram showed progression of left ventricular hypertrophy (fig. 1). Right heart catheterization showed a mild systolic gradient across the outflow tract of the right ventricle (table 1). A catheter was then inserted into the ascending aorta, and an attempt was made to pass it in a retrograde fashion into the left ventricle under fluoroscopic vision and the

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<tr>
<td>Systolic</td>
<td>Pulmonary capillary</td>
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<td>Diastolic</td>
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<td>Hydrogen inhalation test: In pulmonary artery, negative</td>
<td>Dye-dilution curves: In right atrium, slow washout slope compatible with aortic insufficiency</td>
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| Figure 3 |

Phonocardiogram recorded at the pulmonary area with pulse tracing recorded over the axillary artery. There is an ejection click (EC), an ejection systolic murmur (SM), and a long diastolic murmur (DM).

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Figure 4

A. Reproduction of a single 16-mm. movie frame of the retrograde aortogram exposed in the right posterior oblique view. The tip of the catheter is in the tunnel and marked reflux of the contrast substance into the left ventricle is seen. B. Line drawing of A.

A ascending aorta was quite large. The impression after these studies was aneurysm of a sinus of Valsalva with rupture into the left ventricle.

Dyspnea and fatigue increased gradually, and at 16 months of age an operation was performed. Inspection of the exterior of the heart revealed an aneurysmal dilatation of the right coronary sinus of Valsalva apparently involving the origin of the right coronary artery. A second bulge or aneurysm was seen in the region of the outflow tract of the right ventricle. A thrill was present in both of these areas. The circulation was maintained by a pump oxygenator, and after the ascending aorta was opened, an opening about 1 cm. in diameter was seen close to the orifice of the right coronary artery. With a probe placed in the coronary artery, the aneurysmal sac was sutured, apparently without compromising the lumen of the artery. After closure of the ascending aorta and the administration of calcium chloride and epinephrine, the heart resumed a satisfactory beat, but this lasted only about 15 minutes and was followed by cardiac arrest. Resuscitation was of no avail.

An autopsy was performed. The major findings were limited to the heart. The pleural space contained 200 ml. of unclotted blood. The pericardial sac had been opened at the prior surgery. The great vessels of the mediastinum had a normal origin and pattern of distribution. The heart was huge and weighed 200 Gm. as compared to a normal range of 48 Gm. for this age. A 4.0-cm. L-shaped sutured incision ran down the anterior

Figure 5

Anterior surface of the heart. The aneurysm (A) is continuous with a tunnel (T) which runs to the left and disappears behind the right ventricular outlet (O). The applicator protrudes through cut end of the right coronary artery (C). Below this is a blind recess (R) in the epicardial surface.
surface of the ascending aorta to the valve ring. At the lower end of this incision a 2.0-cm. aneurysmal bulge was quite evident on the external surface (fig. 5). The inferior margin of the aneurysm continued as a tunnel, which ran to the left, inferiorly, and then posterior to the right ventricular outflow tract. The tunnel produced an obvious smooth indentation of the posterolateral wall of the infundibular region of the right ventricle (fig. 6). The tunnel measured 3.0 cm. in length and 1.0 cm. in diameter. It opened into the left ventricle, just anterior and to the right of the aortic outlet (fig. 7). Both the tunnel and its openings into the aneurysm above and the left ventricle below had a smooth lining. In addition, marked hypertrophy of the left ventricular musculature resulted in bulging of the interventricular septum into the right ventricle.

When the initial portion of the ascending aorta was opened, the left and posterior sinuses of Valsalva were found to be 2 to 3 times their normal size (fig. 8). The aortic cusps were firm, moderately thickened, and irregular with resultant aortic stenosis. The valve opening was rigid and measured only 2 by 6 mm. The mouth of the aneurysm was situated at the right sinus of Valsalva and was obliterated by sutures. Although the right coronary artery took its origin immediately posterior to the sutured mouth of the aneurysm, its lumen had not been compromised.

Both coronary arteries had a normal pattern of origin and distribution. A patent ductus arteriosus 2 mm. wide was present. The ascending aorta was dilated and measured 3 cm. in diameter. On the epicardial surface between the aneurysm to the left and above, and the right coronary artery to the right and above, was a 5.0-mm. blind saccular indentation (fig. 5).

Microscopic sections of the myocardium and coronary arteries were not unusual. Sections of the ascending aorta, aortic cusps, aneurysm and tunnel revealed a large amount of acid mucopolysaccharides as exhibited by alcian-blue and toluidine-blue stains. In the ascending aorta the material was located in cystlike spaces within the elastic media (fig. 9). The wall of the aortico-left ventricular tunnel resembled the ascending aorta in having an elastic media but only for its proximal half (fig. 10). The distal point of entrance of the tunnel into the left ventricle showed fibrous thickening of the endocardium, which extended for a short distance into the underlying myocardium.

**Discussion**

The present case and the three cases of aortico-left ventricular tunnel described by

*Figure 6*

Anterior surface of the heart. The tunnel (T) begins at the lower border of the aneurysm (A) and runs posterolateral to the right ventricular infundibulum (I) where it produces a discernible bulge (B).

*Figure 7*

Aortic valve from below. The opening of the tunnel (T) is seen to the right of and immediately below the aortic valve opening (V) which is flanked by dilated left and posterior sinuses of Valsalva (D).
Aortico-left ventricular tunnel vessels do not contain contrast substance, and it was readily apparent that the contrast substance was in a cavity that communicates with the aorta.

Right-sided cardiac catheterization revealed a normal or mildly elevated right ventricular pressure. In two cases there was a small or moderate gradient across the pulmonary valve.

Levy et al. present a recognizable clinical profile. All of the patients were male. A murmur was heard in all during the first weeks or months of life. Typically, the murmurs were not continuous. The systolic murmur was loud and harsh, the diastolic murmur was long and likewise rather harsh, and both murmurs were well heard at the aortic area and were transmitted down along the left sternal border. An ejection click has been heard or recorded on the phonocardiogram in three of the four cases. The heart was enlarged, and the electrocardiogram showed left axis deviation and progressively increasing left ventricular hypertrophy.

The roentgenogram showed marked left ventricular enlargement, a dilated aorta, and normal pulmonary arterial markings. Retrograde aortography confirmed the dilatation of the aorta, and there was concomitant marked reflux of the contrast substance into the left ventricle. The most striking finding was filling of a rounded or oval-shaped cavity in the region of the pulmonary trunk and adjacent to the aorta. The first supposition may be that the pulmonary artery has filled with contrast substance, but the more distal pulmonary arter-

**Figure 8**

Aortic valve from above. The narrow valve opening (V) is flanked by thickened, fused cusps (F). The left and posterior sinuses of Valsalva are dilated (D). The larger applicator has been introduced into the mouth of the aneurysm (A). The smaller applicator is inserted into the opening of the right coronary artery (C).

**Figure 9**

Media of the ascending aorta. There are numerous lighter staining areas which, by special stain, contain acid mucopolysaccharides. Hematoxylin and eosin stain; × 250.

**Figure 10**

Mid-portion of wall of tunnel showing termination of elastic tissue (stained black). Verhoeff's elastic-tissue stain; × 50.
indicating either stenosis, as in one case cited by Levy et al., or pressure on the posterior wall of the infundibulum due to bulging of the tunnel into the outflow tract.

An associated feature of considerable importance in our case was the presence of aortic stenosis. With use of the transaortic route to the left ventricle, the catheter presumably passed through the tunnel rather than through the stenotic aortic valve. Thus, interpretation of the pressures obtained was entirely misleading in reference to function of the aortic valve. Severity of the aortic stenosis was masked because the tunnel permitted ejected blood to bypass the aortic valve. This is an important point in surgical correction. The presence of severe aortic stenosis in our case was probably a determining factor in the fatal outcome. After closure of the ostium of the tunnel, the severe aortic valve stenosis prevented an adequate left ventricular output, and left ventricular failure promptly followed. Consequently, the possibility of a coexistent aortic stenosis must be kept in mind and valvulotomy may be indicated.

Levy et al. suggested that an aortico-left ventricular tunnel may represent an anomalous coronary artery opening into the left ventricle, even though they emphasized that the tunnel originates above the level of the coronary arteries. This explanation might well apply to our case, in which the tunnel originated in the right sinus of Valsalva near the origin of the right coronary artery. Anomalous coronary arteries, however, usually have a normal pattern of distribution in spite of their abnormal origin. In the present case we would have to postulate a very unlikely course posterior to the pulmonary infundibulum of the right ventricle. In addition, the presence of elastic media in the tunnel wall makes a coronary arterial origin seem unlikely.

Another possible explanation of the origin of the tunnel is that an aneurysm of a sinus of Valsalva may have ruptured into the left ventricle as suggested by fibrosis of the endocardium and underlying myocardium at the junction between the tunnel and ventricle. The fact, however, that there was no sudden change in the clinical picture and that the opening of the tunnel was quite smooth would suggest that the rupture, if it did occur, must have taken place during fetal life. Another possible explanation is related to maldevelopment of the bulbar ridges associated with a division of the primitive truncus arteriosus into the aortic and pulmonary valves. Associated with the presence of excessive acid mucopolysaccharides, the bulbar ridges and the endocardial cushions may develop large cystic areas, which in turn could rupture into the sinus of Valsalva and the left ventricle. In the case of Heiner et al. of “aortic-left atrial communication” polysaccharides were found in the aneurysmal wall and adjacent portion of the aorta, and in this regard were similar to cases of the Marfan syndrome. A similar collection of mucopolysaccharides was a feature of our case. Also, dilatation of all the sinuses of Valsalva, as found in our case, has been described as a feature of the Marfan syndrome. Neither our case, however, nor that of Heiner et al. had a family history or other stigmata of the syndrome. Whether this hypothesis is applicable to other cases of aortico-left ventricular tunnel is undetermined, since studies for acid mucopolysaccharides are not reported.

**Summary**

A case is presented of a 16-month-old white boy with a communication or tunnel between the right sinus of Valsalva and the outflow tract of the left ventricle. The presence of a murmur at birth and the subsequent clinical course suggest that the tunnel was congenital. The physical, electrocardiographic, phonocardiographic, and aortographic findings were quite similar to those of “aortico-left ventricular tunnel” as described by Levy et al. The tunnel was occluded surgically, but failure to appreciate the presence of severe aortic stenosis contributed to a fatal outcome.

Postmortem study was carried out, and the relationship of the findings of aortico-left ventricular tunnel to aneurysm of the sinuses of Valsalva and the Marfan syndrome is briefly discussed.
References

The Question, The Answer, and Communication

Experiments have two great uses—a use in discovery and verification, and a use in tuition. They were long ago defined as the investigator's language addressed to Nature, to which she sends intelligible replies. These replies, however, usually reach the questioner in whispers too feeble for the public ear. But after the discoverer comes the teacher, whose function is so to exalt and modify the experiments of his predecessor as to render them fit for public presentation.—JOHN TYNDALL. Six Lectures on Light, Lecture 1.
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