AORTIC INSUFFICIENCY that results from 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 is rare. Although clinical findings are similar, this condition may be distinguished anatomically from ruptured aortic sinus aneurysm because the abnormal communication which is to be presented here arises above the level of the coronary arteries (fig. 1).

The purpose of this report is to record three cases of this condition, which we have chosen to call "aortico–left ventricular tunnel." All three patients were considered clinically as having congenital abnormalities of the aortic valve. Aortography was interpreted as showing aortic insufficiency. In one case, aortic sinus aneurysm was additionally diagnosed and, in another, a fistula arising in a coronary artery was suspected. When, in one case, the exact anatomical situation was determined by operation (fig. 2) and subsequent necropsy, it became obvious that the same fundamental problem was also present in the other two patients, and that a definitive operative procedure was available to them. Therefore, the latter two patients were recalled for operation, the abnormal communication was closed and, in each, a cure was effected. To our knowledge, only four other cases of this entity have been reported; in each instance, the diagnosis had been made only at necropsy.1–4 Successful correction has not been reported previously.

The manifestations in the three patients were so similar that the case findings justifiably may be presented in composite form to yield a profile of this condition.

Clinical Observations

History and Physical Findings

Each of our three patients was male; general body development had appeared normal. At the time of operation, their ages were 5, 10, and 3 years, respectively. Cyanosis was not a feature except in one patient (case 1) in whom it appeared in early infancy during a period of cardiac failure. Two patients had decreased exercise tolerance. Of interest is that a cardiac murmur had been detected at an early age in each, namely, at one week, one day, and nine months, respectively.

Physical examination revealed a prominent precordial bulge in two patients. Cardiac enlargement, on the basis of the position of the apex, was readily apparent. In each patient, there was a systolic thrill maximal over the "aortic area," and a diastolic thrill maximal along the left sternal border. There were long grade 3 to 4 (on the basis of 1 to 4) harsh systolic and grade 3 to 4 blowing diastolic murmurs in the corresponding areas (fig. 3). These murmurs were separate and widely transmitted, with no resemblance to the machinery murmur of a patent ductus arteriosus. In two cases, the clinical records make men-
A reported example of aortico-left ventricular tunnel. The ascending aorta (A.A.) is grossly dilated. The aortic valve (A.V.), which is viewed from above, is bicuspid and slightly stenotic. Above the valve is the aortic ostium (arrows) of an aortico-left ventricular tunnel. Shortly after its origin, the abnormal channel shows a saccular aneurysm (An.). Beyond the aneurysm, the tunnel (T.) extends downward toward the left ventricle. (Case of Edwards. Specimen submitted to Edwards by N. G. B. McLetchie.)

Electrocardiographic Findings

The mean QRS axis was of the left-axis-deviation type in cases 1 and 2, and was normal in case 3. Left ventricular hypertrophy was observed in each case at the time of initial evaluation. In cases 1 and 2, however, interval examinations revealed progressive left ventricular hypertrophy and strain (fig. 4). Vectorcardiograms in two cases were also interpreted as showing left ventricular hypertrophy.

Radiological Findings

Thoracic roentgenograms (fig. 5) in each case showed moderate to severe cardiomegaly.
in posteroanterior projections. The configuration of the cardiac silhouette was left ventricular in type; the left ventricle was seen to extend posteriorly to the barium-filled esophagus in lateral projection, suggesting considerable enlargement. There was no left atrial enlargement. The pulmonary vasculature was normal. The most characteristic finding was an unusual prominence of the right cardiac border owing to marked dilatation of the ascending aorta. There was deviation of the trachea to the right, as might occur as a result of a large aortic arch. Studies in lateral projection showed a density in the retrosternal space which was interpreted as a sign of enlargement of the ascending aorta. Roentgenoscopically, marked increased pulsations of the left ventricle and of the aorta were observed. The pulmonary arterial segment was not prominent. The impression from roentgenoscopy was that of aortic valvular incompetence with marked dilatation of the ascending aorta and of the aortic arch.

Selective angiograms\(^5,6\) (fig. 6) showed diffuse massive dilatation of the ascending aorta, a picture resembling the aneurysmal dilatation seen in Marfan's syndrome. Dilatation was fusiform and involved the ascending aorta and the aortic arch. In lateral films, the aorta occupied the retrosternal space, being somewhat more anterior than normal, a feature occasionally observed in cases of marked left ventricular enlargement. The aortic valve appeared to be at a normal level. After radiopaque material was placed into the aorta, there was immediate and concentrated opacification of the left ventricular chamber which was enlarged. The left ventricle had a relatively thick wall and was seen to contract very forcefully; this indicated aortic incompetence with increased stroke volume. There was no sign of mitral insufficiency. In each patient, anteroposterior projection of the selective aortogram revealed an area of density in the region of the pulmonary trunk, but no contrast medium appeared in the pulmonary arteries. This excluded an extracardiac shunt. In lateral films, density related to the pulmo-

**Figure 3**

Phonocardiograms. Upper, case 1. Phonocardiogram recorded from the second left intercostal space parasternally. Intense "diamond-shaped" systolic murmur (SM) is evident. In addition to the systolic murmur, a loud diastolic murmur (DM) is recorded. The latter begins immediately after the second sound and is in continuity with the next systolic murmur. There is a short interruption between the systolic murmur and the onset of the diastolic murmur, an interruption which creates a pattern different from that of a typical machinery continuous murmur. (Paper speed, 75 mm/sec.) Center and Lower, case 3. Distinct systolic and diastolic murmurs. A = second left intercostal space parasternally; P = second right intercostal space parasternally.
Electrocardiograms. Upper left and center, case 1. Electrocardiograms taken at an interval of three years. Upper left (July, 1958, age two years). Left ventricular hypertrophy with early signs of left ventricular "strain," the latter exhibited by the elevated S-T segment in lead V1. Upper center (August, 1961, age five years). Further progression of left ventricular "strain" indicated by changes in the ST-T complexes of lead V6. Upper right, case 3 (age three years). Evidence of marked left ventricular hypertrophy. *N* = one-fifth standardization. Lower, case 2 (age 10 years). Electrocardiograms over a period of eight years, showing progressive changes from borderline to marked left ventricular hypertrophy with negative or biphasic T waves over the left precordial leads (1958, 1960).

Pathological Physiology

Right-sided cardiac catheterization was performed in two patients (cases 2 and 3), in each at the age of one year. No evidence of rotation of the heart which, in turn, had resulted from the marked left ventricular enlargement.
a left-to-right shunt was obtained in either case. In one patient (case 2), the pulmonary arterial pressure was normal, being 14 mm Hg systolic and 6 mm Hg diastolic. In the other patient (case 3), the right ventricular and the pulmonary arterial systolic pressures were each 38 mm Hg. By other studies, evidence of aortic and pulmonary stenosis was obtained in case 1. In this patient, pressures obtained by transthoracic, percutaneous, left-sided cardiac catheterization were as follows: left ventricle, 175 mm Hg systolic and 0, diastolic; aorta, 105 mm Hg systolic, and 65, diastolic (fig. 7); left atrium, 12 mm. Hg systolic and 2, diastolic. No unusual configuration of the left atrial pressure curves was evident. In this patient, at the time of operation (while the thorax was open), pressures indicating pulmonary stenosis were recorded. The right ventricular pressures were 70 mm Hg systolic and 0 diastolic, while in the pulmonary trunk, the systolic pressure was 17 mm Hg and the diastolic, 9 mm Hg.

**Pathological Observations**

The pathological anatomical findings were derived from operative observations in each case and from necropsy, additionally, in case 1. The following is a composite description of the information obtained from these sources.

After the pericardium was opened, it was apparent that the origin of the ascending aorta was grossly and uniformly dilated. From the anterior aspect of the ascending aorta and above the levels of coronary arterial ori-
Figure 6

Selective angiocardiograms. Upper, case 1. Selective left ventriculograms. Upper left, posteroanterior view. Upper right, lateral projection. Marked dilatation of ascending aorta (A) is seen, with an unusual opacification (arrow) along the medial aortic border. The latter, in lateral projection, bulges along the anterior aspect of the aortic shadow (upper arrow) and above the level of the aortic valve (AV). The aortic valve is dome-shaped and eccentrically located. Center, case 2. Center left, antero-posterior selective aortogram. The tip of the catheter is above the aortic valve (AV).
gins, a wide vessel-like structure, covered by visceral pericardium, arose. This structure, tubular in cases 1 and 2 (fig. 8, left), was about 1.5-cm. wide. In case 3, the origin of this structure resembled a saccular aneurysm closely applied to the aorta (fig. 8, right).

The abnormal structure descended to the right of the pulmonary trunk and, from the exterior, was lost from view as it appeared to penetrate the basal aspect of the ventricular septum.

From within the aorta, the abnormal channel was represented by an ostium about 1.5 cm. in diameter. This ostium lay either just superior to the right coronary arterial origin or above and slightly to the left of it (fig. 9). In cases 1 and 2, the coronary arterial ostia were distinct from the abnormal opening in the aorta, while in case 3 the right coronary artery arose from the lower aspect of the aneurysmally dilated origin of the abnormal channel.

Considering the region where the abnormal channel appeared to penetrate the ventricular septum, it was apparent from the pathological specimen (case 1) that the channel did enter the ventricular septum. It was that upward prolongation of the ventricular septum that lies to the right of the aortic root which received the channel (fig. 10). From this area, the channel tunneled through that part of the ventricular septum which forms the posterior wall of the right ventricular infundibulum. It then terminated by communicating with the

outflow tract of the left ventricle just inferior to the commissure between the left and right aortic cusps. As the tunnel passed through the ventricular septum, it narrowed somewhat; but throughout its intramural extent, it maintained a diameter of at least 1 cm. Throughout its extent, the lining of the tunnel was yellow to gray in color.

Of special note is the fact that, as the tunnel passed behind the right ventricular infundibulum, it caused the posterior wall of the infundibulum to bulge into the right ventricular cavity. Over the bulge, the endocardium of the right ventricle was thickened. It was suggested that this distortion of the right ventricular infundibulum might have been responsible for some obstruction to right ventricular outflow.

In cases 2 and 3, the aortic valves appeared normal when observed at the time of aortot-

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**Figure 7**

Case 1. Pressure curves obtained at rest during percutaneous left-sided cardiac catheterization. Pressure differences between the aorta (left) and the left ventricle (right) indicate a systolic gradient of 70 mm. Hg across the aortic valve.

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*The opacified tunnel (between right-sided arrows) extends from the medial border of the aorta. There is simultaneous opacification of the left ventricle (LV) from regurgitant flow. Center middle, lateral projection of selective aortogram corresponding to anteroposterior position shown center left. The tip of the catheter is above the aortic valve. An unusual bulge along the anterior aspect of the aorta is opacified. This is accompanied by retrograde filling of the left ventricle. Center right, selective left ventriculogram. The tip of the catheter has been passed through the aortic valve and lies in the outflow tract of the left ventricle. When radiopaque media are injected into the ventricle, the aortico-left ventricular tunnel (arrow) is well opacified. Illustrations indicate that blood flows through the tunnel during systole as well as during diastole. Lower, case 3, posterocentral and lateral projections of selective aortograms demonstrating the aortico-left ventricular tunnel. The origin of the tunnel was unusually dilated (see fig. 8, right), appearing as an aneurysmal-like formation against the medial aspect of the ascending aorta. Early filling of left ventricle. In the lateral projection, the tunnel is in relation to the anterior aspect of the ascending aorta.*
omy. In case 1, the aortic valve was congenitally bicuspid and appeared slightly stenotic. In the same case, the pulmonary valve showed minor degrees of commissural fusion, lesions which may have contributed to the demonstrated pulmonary stenosis. Nevertheless, some of this functional derangement, as suggested, may have been caused by the distortion of the wall of the right ventricular infundibulum imparted by the tunnel passing through the ventricular septum.

In case 1, the necropsy revealed hypertrophy of the left ventricular wall and enlargement of the left ventricular cavity. These changes were not different from those which are observed in common types of aortic valvular insufficiency. The right and left coronary arteries were demonstrated to be present and to follow usual patterns of origin and distribution. Histological examination was made of the entire length of the wall of the tunnel in case 1 (fig. 11). At its origin from the aorta, the outer layer of the tunnel was composed chiefly of elastic fibers which appeared to be an extension into the wall of the tunnel from the aortic media. This layer persisted until the tunnel penetrated the ventricular septum. Beyond that, the wall was composed of nonspecific, hyalinized collagen containing irregularly distributed elastic fibers. In this region, the wall of the tunnel bore resemblance to that of a myocardial sinusoid. Throughout the entire length of the tunnel, the inner aspect of the wall was composed of irregularly deposited connective tissue, some cellular and some hyalinized. The inner layer showed variable amounts of elastic tissue; in some areas these were exceedingly numerous and closely packed.

Representative examples of the ascending aorta at zones removed from the origin of the tunnel showed no histological changes of significance. No features which are exhibited in Marfan's syndrome or in so-called idiopathic dilatation of the aorta were encountered.

In case 3, at operation, a specimen for histological examination had been obtained of the junction of the aorta and the tunnel. The histological picture was like that observed at a comparable area in case 1.

Circulation, Volume XXVII, April 1963
Operative Treatment

Each of the three patients was operated upon by means of extracorporeal circulation. Selective coronary perfusion9 was not used in the first case (which was followed by a fatality), but was utilized in cases 2 and 3. The operative approach in each instance was through a midline sternotomy. The ascending aorta was greatly dilated and a to-and-fro thrill was readily apparent at the aortic origin. The duration of cardiac arrest was 60, 42, and 62 minutes in cases 1, 2, and 3, respectively.

From the exterior, a vessel appeared to arise from the ascending aorta above the levels of the coronary arteries. With the aortic lumen then exposed, the opening of a channel was identified in the anterior wall of the ascending aorta above the right coronary artery. Probing of the channel readily led to the left ventricular cavity.

In each instance, the aortic ostium of the abnormal communication was closed by multiple interrupted mattress sutures of silk, followed by a running over-and-over continuous stitch as a second layer. In cases 2 and 3, no further procedure was done, while in case 1 it was evident that the aortic valve was bicuspoid and stenotic. The commissure, representing the center of the conjoined cusp, was divided to effect a greater opening of the aortic valve. In the same case, in whom pulmonary
stenosis also had been identified, the pulmonary valve was exposed through the pulmonary trunk after completion of the aortic phase of the operation. The pulmonary valve appeared stenotic on the basis of fusion of the cusps; this process was relieved by division of the commissures. Following closure of the heart and before completion of the operation, it was noted that the pressure differential between the right ventricle and the pulmonary artery had fallen materially to 50 mm. Hg systolic in the right ventricle and 40 mm. Hg systolic in the pulmonary trunk. Left ventricular obstruction appeared to have been overcome since, after the aorta was closed, the aortic pressure was 105 mm. Hg systolic.
and 75 mm. Hg diastolic, while the left ventricular pressure was recorded as 100 systolic and 5 diastolic.

The patients in cases 2 and 3 experienced an uncomplicated postoperative course, while the first patient developed postoperative complications. In the early phase of the postoperative period, the patient’s (case 1) condition appeared satisfactory. Electrocardiograms revealed short episodes of ventricular extrasystole. Approximately 12 hours after operation, the systemic blood pressure dropped; the patient became apprehensive and cyanotic. Vasopressor agents were administered and tracheostomy was performed. Although these measures were followed by temporary improvement, the patient’s condition gradually deteriorated and cardiac arrest occurred several hours later.

Comment

Two theories concerning the fundamental nature of the aortico–left ventricular tunnel may be presented. One view is that it is an acquired lesion; the other is that it is a congenital anomaly. On the theory that the process is acquired, the lesion might be viewed as a rupture of the aorta leading to a dissecting hematoma which finds its way through the base of the ventricular septum and finally into the left ventricle. In case 1, the presence of aortic stenosis would support such a theory, for it is recognized that in rare cases of aortic valvular stenosis a rupture of the ascending aorta may appear as a complication, and varying degrees of dissecting hematoma may follow.10,11 In cases 2 and 3, the aortic valve did not appear stenotic when observed at operation. Also, the recorded pressures across the aortic valve excluded the presence of aortic stenosis in the second and third cases.

Continuing with a consideration of the possibility of the aortico–left ventricular tunnel’s representing an acquired rupture, one must consider the questions of Marfan’s syndrome and so-called idiopathic dilatation of the aorta. Each of these conditions may become complicated by dissecting aneurysm of the aorta. Tissue examination of the aorta, which was done in cases 1 and 3, did not reveal any of the histological features of these conditions.8 The marked dilatation of the ascending aorta and of the aortic arch, which characterized our cases, must be accepted as a response to the functional derangement caused by the tunnel (aortic insufficiency) rather than as a manifestation of intrinsic disease of the aorta.

The similarity in appearance of the abnormal process among the three cases and the very early presence of abnormal physical signs strongly suggest a congenital basis.

Demonstration of continuation of aortic media into the wall of the tunnel suggests that it is an anomalous vessel rather than an acquired tract. Although the histological nature of the wall is not an exact counterpart of that of a coronary artery, the possibility remains strong that the aortico–left ventricular tunnel is, in effect, an accessory coronary artery which communicates, through an enlarged myocardial sinusoid, with the left ventricular cavity.

The aneurysmal nature of the origin of the tunnel in case 3 deserves some mention. Figure 1, which represents a case previously observed by Edwards, shows a saccular aneurysm of the tunnel shortly beyond its origin. It is possible in that case, as well as in case 3 of this presentation, that the aneurysm was an acquired complication of a congenital malformation—the aortico–left ventricular tunnel. It will be recalled that secondary aneurysm formation is not uncommon in those anomalous coronary arteries that communicate with cardiac chambers.12-14

As pointed out earlier, the tunnel should not be confused with aortic sinus aneurysm, since the aortic origin of the tunnel is above the level of the coronary arterial ostia. True aortic sinus (Valsalva) aneurysms arise below the coronary ostia.15 In keeping with this distinction is the observation that the origin of an aortico–left ventricular tunnel lies above the level of the heart and may be visualized roentgenographically as an abnormal extracardiac shadow. True aortic sinus aneurysms
lie within the roentgenographic shadow of the heart and do not lend an abnormal shape to the cardiac silhouette.

The clinical differential diagnosis of aortico-left ventricular tunnel includes aortic valvular incompetence and ruptured aneurysm of an aortic sinus (Valsalva). A specific diagnosis may be derived from radiological studies. In an aortico-left ventricular tunnel, the ascending aorta seems to be dilated in a fusiform aneurysmal fashion; this usually is not seen in aortic valvular incompetence, where the dilatation is more cylindrical. The aneurysmal dilatation in aortico-left ventricular tunnel is quite similar to the aneurysmal dilatation encountered in Marfan’s syndrome.

In the aortogram, visualization of the round, double-density shadow above the aortic sinuses (in the region of the pulmonary trunk), coupled with lack of radiopaque material entering the pulmonary arteries, should establish the correct diagnosis of aortico-left ventricular tunnel and eliminate aortic sinus aneurysm. Such findings also exclude the diagnosis of isolated aortic valvular incompetence.

The angiographic appearance of aortico-left ventricular tunnel might be confused with an aneurysm of a left coronary artery. Demonstration of both coronary arteries quite apart from the abnormal channel should serve to distinguish aortico-left ventricular tunnel from aneurysmal coronary artery.

Summary

To be distinguished from aneurysm of an aortic sinus (Valsalva) is the rare condition in which an abnormal channel begins in the ascending aorta (above the right coronary artery), bypasses the aortic valve, and leads through the ventricular septum into the left ventricular cavity. This entity, which has not previously been diagnosed during life, to our knowledge, is herein named aortico-left ventricular tunnel.

The cases of three patients (aged 3, 5, and 10 years) with this rare condition are described. The clinical, electrocardiographic, and radiological features were similar in each case and resembled those in aortic valvular insufficiency.

The diagnosis may be established clinically through selective aortography. This reveals that the origin of the abnormal communication lies in the anterior wall of the ascending aorta above the levels of the coronary arterial origins. The abnormal communication leads to the left ventricle. The condition is distinguished from aortic sinus aneurysm by its origin above the level of the aortic sinuses.

Identical anatomical findings and the presence of abnormal physical findings since infancy in each of the three patients suggest that this peculiar condition is a congenital malformation.

References


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Aortico-Left Ventricular Tunnel
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