Arteriovenous Aneurysm of Phrenic Vessels Simulating Cardiac Fistula

By IAN R. GRAY, M.D. and H. PARRY WILLIAMS, M.D.

WHEN a continuous murmur is heard in the chest of a child one usually thinks of a patent ductus arteriosus. This diagnosis is unlikely, however, if the murmur is not loudest in the left infraclavicular region, and some other explanation must then be sought. This communication is about a child with such a murmur and details the investigations that led to the diagnosis of phrenic arteriovenous aneurysm. This would appear to be the first reported example of such an aneurysm.

Case Report

The patient was referred to the hospital at the age of 4 years and 8 months because a murmur had been discovered when she visited her doctor because of a respiratory infection. She had always been healthy and active with no sign of cardiac disability. There was no cyanosis and no clubbing of the fingers. The pulse was regular and normal in quality; the blood pressure was 90/65 mm. Hg. There was a continuous thrill and murmur maximal at the apex that was quite widely transmitted medially and downwards. The heart sounds were normal and no other murmurs were heard. A provisional diagnosis of coronary arteriovenous aneurysm was made.

Phonocardiogram showed the continuous murmur (fig. 1). The heart sounds were hard to demonstrate but were not obviously abnormal. The murmur seemed to be loudest toward the end of systole as in patent ductus arteriosus.

Radiologic examination of the chest showed no cardiac enlargement and no evidence of increase in pulmonary blood flow (fig. 2). Two significant abnormalities were detected only after aortography had revealed the diagnosis. These were a shadow along the upper third of the left cardiac border corresponding to the dilated pericardio-phrenic vein and rounded indentations of the gastric air bubble at the site of the diaphragmatic aneurysm. An electrocardiogram was normal.

Cardiac catheterization (table 1) showed no shunt into the right atrium or ventricle or into the pulmonary artery, and the pulmonary arterial and right ventricular pressures were normal. The oxygen saturation of the blood from the superior vena cava and the upper right atrium was considerably higher than that from the inferior vena cava.

Retrograde aortography was then undertaken as the site of the shunt was still uncertain. The dye filled a convoluted collection of vessels in the region of the left diaphragm about the junction of its middle and outer thirds (fig. 3). These abnormal vessels were supplied by an artery of medium size that arose from the abdominal aorta at the site of the left phrenic artery. The vein that drained them pierced the diaphragm, ran up the left border of the heart along the course of the pericardio-phrenic vessels, and ended in the region of the left innominate vein (fig. 4). The venous drainage of the arteriovenous communication explains the unexpectedly high oxygen saturation found in the superior vena cava and right atrial blood samples.

Discussion

This case illustrates the difficulty of clinical diagnosis when a continuous murmur is found in an unusual site. It also emphasizes how a correct conclusion can be reached by modern technics of investigation. A continuous murmur is only generated when a pressure difference exists throughout the entire cardiac cycle.

Table 1

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure (mm. Hg) (mean in brackets)</th>
<th>Oxygen content ml. per cent</th>
<th>Oxygen saturation (per cent)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>—</td>
<td>15.6</td>
<td>84</td>
</tr>
<tr>
<td>Right atrium</td>
<td>High</td>
<td>15.5</td>
<td>83.5</td>
</tr>
<tr>
<td></td>
<td>Mid. (—4)</td>
<td>14.5</td>
<td>78</td>
</tr>
<tr>
<td></td>
<td>Low</td>
<td>14.7</td>
<td>79</td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>—</td>
<td>13.3</td>
<td>71.5</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>10/—4 (4)</td>
<td>14.0</td>
<td>75</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>10/2 (5)</td>
<td>13.4</td>
<td>72</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>70/55 (60)</td>
<td>15.9</td>
<td>85.5</td>
</tr>
</tbody>
</table>

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between the two vessels or chambers that are in abnormal communication, and such a murmur usually occurs when the flow is under high pressure. Hence, the site of origin of a continuous murmur is usually to be found in relation to either a systemic artery or a pulmonary artery with an abnormally high pressure.

The majority of continuous murmurs heard over the chest are due to a left-to-right shunt, and the commonest example is patent ductus arteriosus. Many of the other conditions with left-to-right shunt causing a continuous murmur have, like patent ductus, signs of a leak-back from the aorta and evidence of left ventricular hypertrophy. In such cases the location of the murmur may help in diagnosis. In patent ductus the murmur is heard best in the first and second left intercostal spaces. It tends to be loudest rather lower than this when there is an aortopulmonary septal defect, and lower still with ruptured aneurysm of the sinus of Valsalva and a coronary arteriovenous aneurysm. The murmur in these conditions is usually loudest near the midline and it is unusual for it to be maximal as far to the left as in the case described. A continuous murmur loudest in the left axilla was, however, reported in an infant with a communication between the left coronary sinus of Valsalva and left atrial appendage.1

Neill and Mounsey2 drew attention to the position in the cardiac cycle of continuous murmurs due to different causes. They pointed out that the crescendo of the murmur is later in systole in systemic and pulmonary arteriovenous fistulas than when the shunt is from the aorta. In four of five patients with communications between the coronary arteries and the right ventricle Gasul et al.3 found the crescendo in diastole. The site and timing of a continuous murmur often point to the probable diagnosis, and this is of great value in planning appropriate investigations.

Cardiac catheterization may indicate the site of the left-to-right shunt by increased oxygen saturation of the blood in the appropriate chamber or vessel. Such a rise is found

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**Figure 1**
High-frequency phonocardiogram of patient J.E., with phrenic arteriovenous aneurysm, recorded from the fourth left intercostal space just lateral to the sternum, with electrocardiogram (lead I). This shows a continuous murmur loudest toward the end of systole. The first and second heart sounds (1 and 2) are difficult to identify.

**Figure 2**
Skiagram of chest. Attention is drawn to the linear shadow along the upper third of the left cardiac border corresponding to the dilated left pericardiophrenic vein (see fig. 4), and the indentations of the gastric air bubble at the site of the diaphragmatic aneurysm.
in the pulmonary artery in patent ductus and aortopulmonary septal defect, and in the right atrium or ventricle in ruptured aneurysm of the sinus of Valsalva and coronary arteriovenous aneurysm. When the shunt is into one of the larger veins, as in the case described, sampling from the appropriate vessel proximally and distally may help in accurate localization. The passage of the catheter through the defect may be diagnostic: this frequently happens in patent ductus and aortopulmonary septal defect and has also occurred in ruptured aneurysm of the sinus of Valsalva. The shunt can be demonstrated by aortography in most cases, and this procedure is of the greatest value.

A special group of conditions with cyanosis should be mentioned in which a continuous murmur may be heard. These are firstly, the pulmonary arteriovenous communications, in which a continuous murmur may be present over an aneurysm in the lung, and secondly pulmonary atresia and persistent trunca, in which a single arterial trunk arising from the heart supplies blood to the lungs through a patent ductus or dilated bronchial arteries. Systemic arteriovenous communications may be congenital or caused by trauma. The acquired communications will not be discussed but hemodynamic changes are similar. Congenital arteriovenous communications are rare. The lower limbs are most frequently affected and the upper limbs, scalp, face, and cranial contents less often. Such communications are very rarely found in the trunk, and when in the chest wall they tend to be mistaken for patent ductus. Maier and Stout reported an arteriovenous fistula in the left pectoralis major muscle that was supplied by the internal mammary artery, and Wells and Hurt described an arteriovenous communication between the left internal mammary artery and vein. Two further examples of particular interest are the patient described by Brain and Kauntze with a communication

Figure 3
Aortogram, arterial phase. The convoluted vessels of the diaphragmatic aneurysm are shown to be filled by an artery that arises from the aorta and that corresponds in position and course with the left phrenic artery.

Figure 4
Aortogram, venous phase. The left pericardiophrenic vein is seen to drain the aneurysm and, passing up the left cardiac border, to disappear in the region of the left innominate vein.
between the intercostal arteries and pulmonary veins, and the neonate with a fistulous communication between the left internal mammary artery and the ductus venosus.8

There seems to be no simple embryologic explanation of the development of congenital arteriovenous aneurysm. The arteries and veins in the embryo are differentiated from a common capillary plexus, and persistent direct arteriovenous communication might well be expected to occur more frequently than is the case. The histologic features of the abnormal vessels are variable, and the distinction between artery and vein may be impossible. The observation that the artery distal to a large arteriovenous communication develops the histologic features of the vein, and the vein proximal to a communication comes to resemble an artery,9 suggests that histologic distinction is less clear than might be supposed.

That cardiac enlargement and even congestive cardiac failure may occur in patients with large arteriovenous fistulas is well established, and considerable attention has been paid to the mechanisms by which this takes place. Lewis and Drury10 pointed out the similarity of the changes in peripheral circulation in arteriovenous communications and aortic regurgitation. They did not find increase in the venous pressure in their patients and postulated therefore that the cardiac output was not increased. They ascribed the enlargement of the heart to deficient nutrition of the myocardium. An outstanding experimental study by Holmana established that in large shunts the venous filling pressure rises, with increase in cardiac output, increase in pulse pressure, and tachycardia. He also showed that the total blood volume increases in direct proportion to the amount diverted through the fistula. These observations have since been confirmed in man by various investigators.11-13 It is thus evident that the cardiac consequences of arteriovenous fistula are similar to those that can occur in any condition in which the output is constantly increased.

The diminished arterial resistance in the neighborhood of an arteriovenous communication has been shown to cause a progressive increase in the amount of blood diverted from the general circulation.9 The hemodynamic consequences are likely to increase with time, giving rise to cardiac enlargement and, ultimately, to failure. The risk of infective endarteritis is a further hazard. Successful surgical treatment of arteriovenous communications invariably restores the hemodynamic situation to normal with diminution in the size of the heart, and, in advanced cases, disappearance of signs of congestive failure. Congenital arteriovenous communications often present a formidable surgical problem as the lesion may be extensive and involve numerous vessels. In spite of the difficulties it seems to us that surgical correction should be considered in all cases as early as possible.

Summary

A child is described with an arteriovenous communication between the left phrenic artery and the left pericardio-phrenic vein. The differential diagnosis of continuous murmurs in the chest is discussed in the light of this case. The hemodynamic consequences of arteriovenous communications and indications for their surgical treatment are briefly outlined.

Acknowledgment

We would like to thank Dr. R. Laverty, who referred the case, and Dr. J. C. A. Raison, who carried out aortography. We are also grateful to Miss J. Brookes for technical assistance in the investigations and to Miss A. L. Hebden, who is responsible for the illustrations.

References


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Galvani and the Electrophysiology of Muscular Contraction

Soon afterwards, Galvani was strongly convinced of the existence of animal electricity which was discharged by metallic ares, but he was a thorough investigator and required proof, “for it is easy in experimentation to be deceived, and to think one has seen and discovered what one has desired to see and discover.” (Commentary, Part Three). The new observations, made by him on frogs and other animals, led him to believe that a fluid, similar but not identical with the electrical fluid, might flow through the are. He called this phenomenon electricitatis excursus. His theory was finally summarized as follows (Commentary, Part Four), “It would perhaps not be an inept hypothesis and conjecture, nor altogether deviating from the truth, which should compare a muscle fiber to a small Leyden jar, or other similar electric body, charged with two opposite kinds of electricity; but should liken the nerve to the conductor, and therefore compare the whole muscle with the assemblage of Leyden jars.—Giulio Pupilli. **Commentary on the Effect of Electricity on Muscular Motion.** BY LUIGI GALVANI. Translated by Robert Montraville Green, M.D., Cambridge, Massachusetts, Elizabeth Licht, Publisher, 1953, p. xii.
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Circulation. 1962;26:434-438
doi: 10.1161/01.CIR.26.3.434

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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