Continuous Murmur Due to the Combination of Rheumatic Mitral Stenosis and a Rare Type of Anomalous Pulmonary Venous Drainage

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SUMMARY
A patient is described in whom a continuous murmur was caused by the combination of rheumatic mitral stenosis and a rare form of partial anomalous pulmonary venous drainage. In this case, the left superior pulmonary vein drained into the left atrium and also into a large anomalous vein which then drained to the right atrium via the innominate vein and superior vena cava. At surgery, mitral commissurotomy and then ligation of the anomalous vein were performed with a good result; the continuous murmur was no longer audible after operation. The embryology, physiology, auscultatory findings and surgical consideration are discussed.

Additional Indexing Words:
Mitral commissurotomy    Ligation of anomalous vein

Continuous murmurs are generated by the continuous flow of blood through a channel, across which a significant pressure gradient exists throughout the cardiac cycle. Patent ductus arteriosus is the best known example of this situation, and the flow is from the systemic to pulmonary circulation. Continuous murmurs may also be associated with arterial to venous shunts (e.g., rupture of aneurysm of sinus of Valsalva into the right atrium), coronary, pulmonary, or systemic arteriovenous fistulæ, systemic or pulmonary arterial narrowing, and increased peripheral or collateral flow. Ross and associates have described a continuous murmur in patients with the combination of atrial septal defect and left atrial hypertension. The purpose of this communication is to describe an additional cause of a continuous murmur in a patient with rheumatic mitral stenosis and a rare form of partial anomalous pulmonary venous drainage with an extrapericardial connection between the left and right atria.

Report of Case
C. B., a 28-year-old Caucasian male, was hospitalized at ages 13 and 23 with acute rheumatic fever. Nevertheless, he was well and active until 2 months prior to admission when he noted marked dyspnea on exertion, a persistent dry cough, and fatigue. Three weeks prior to admission, he was admitted to another hospital with tachycardia and severe shortness of breath. He responded to treatment for congestive heart failure and was referred to Johns Hopkins Hospital for cardiac evaluation.

On physical examination, he was well developed and of average height. His blood pressure was 120/75 and his pulse 80 and regular. There was no clubbing or cyanosis, jugular venous distention, peripheral edema, or hepatomegaly. The arterial pulses were of normal quality. No precordial thrill or lift was palpated. The first heart sound was accentuated at the apex and was preceded by a low-pitched presystolic rumble. The second sound was split normally, and the second or pulmonic component was mildly

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accentuated. An opening snap was not heard. A grade 1/VI systolic murmur was heard along the lower left sternal border. A grade III/VI low-pitched continuous murmur was heard best in the left supraclavicular area but was also audible anteriorly in the first and second left intercostal spaces. This murmur could not be obliterated by neck compression, rotation of the head, or assumption of the upright position. The continuous murmur did not vary significantly in intensity with respiration. The Valsalva maneuver resulted in diminution of the heart sounds and continuous murmur.

**Laboratory Studies**

The hematocrit was 48%, and urinalysis was normal. The electrocardiogram showed sinus rhythm and diphasic P waves in lead V1, with a 2-mm negative deflection, suggesting left atrial enlargement. Chest x-rays showed moderate cardiomegaly and left atrial enlargement, an abnormal pulmonary venous pattern with lower zone constriction and upper zone dilatation, prominence of the pulmonary arterial shadows, a reticular pattern to the lung fields, and discoid atelectasis. There was no suggestion of a left superior vena caval shadow along the upper mediastinum. A phonocardiogram confirmed the presence of a continuous murmur and apical presystolic murmur. Normal breathing had no effect on the intensity or character of the continuous murmur (fig. 1).

**Cardiac Catheterization**

The results of right heart, transseptal, and retrograde left heart catheterization are diagrammed in figure 2. With the platinum electrode in the pulmonary artery, the hydrogen test was positive 2 sec after inhalation. A step-up in oxygen saturation was demonstrated at the level of the superior vena cava (SVC), and with the platinum electrode in the SVC, the hydrogen test was positive 1 sec after inhalation. The atrial septum was probed, but an atrial septal defect was not found; a transseptal left atrial catheterization was then performed. The left atrial pressure was elevated with a mean pressure of 15 mm Hg. A mitral valve gradient of 13 mm Hg was calculated from simultaneous left atrial and left ventricular pressure tracings. A left atriogram showed the mitral valve to be thickened and stenotic but mobile and noncalcified. Indocyanine green dye was injected through the transseptal catheter into the left atrium and was detected in

**Figure 1**

Phonocardiogram. The continuous murmur is recorded best above the left clavicle. The phase of respiration has no significant effect on the intensity of the continuous murmur.

**Figure 2**

Heart diagram of findings at cardiac catheterization and surgery. Pressures are given for the cardiac chambers and great vessels; the numbers in parentheses indicate the mean pressures. The right heart oxygen saturations are indicated by the numbers with percent signs.
Figure 3

Illustration of anatomical findings at surgery. The patient is in the right lateral decubitus position. The anomalous vein is anterior to the pulmonary artery and connects the left superior pulmonary vein to the innominate vein. The left subclavian and left jugular veins also drain into the innominate vein.

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the femoral artery 7 sec later. A recirculation hump then appeared, indicating a left-to-right shunt. After injection of indocyanine green dye into the left ventricle or aorta, with sampling in the pulmonary artery, there was no early appearance of dye. To define precisely the level of the left-to-right shunt, a catheter was passed from the SVC into a vein which entered the SVC horizontally from the left at the level of the right subclavian vein. Oxygen saturation in this vessel was 94%, but the catheter could not be advanced to the left atrium from it. The ratio of pulmonary to systemic blood flow was approximately 2:1.

Twenty-four hours after catheterization, the patient developed fever and pleuritic left chest pain. Chest x-rays showed left lower lobe atelectasis; a lung scan was negative. Intravenous heparin therapy was begun but was discontinued 24 hours later when dizziness with left-sided paresthesia and mild weakness developed. These symptoms abated over the following 2 weeks and he underwent cardiac surgery. The preoperative diagnosis was mitral stenosis and anomalous pulmonary venous drainage with an extrapericardial connection between the left and right atria. The latter diagnosis was made because of (1) the continuous murmur, (2) the left-to-right shunt at the level of the SVC, (3) the recirculation noted on left atrium to femoral artery dye curve, and (4) no evidence for an atrial septal defect or a shunt at the ventricular or aortic level.

Surgical Technique

A left posterolateral incision was carried through the bed of the excised fifth rib. Lateral retraction of the lung revealed a large left superior pulmonary vein (fig. 3). This tense 2-cm vein bifurcated near the hilum into a superior and inferior division. The superior branch, or anomalous vein, drained into the innominate vein. A prominent continuous thrill was felt on palpation of the anomalous vein. The inferior branch entered the left atrium directly with no coronary sinus communication.

The left atrium was then digitally explored via the atrial appendage. No thrombus was noted. The tightly stenotic mitral valve was minimally thickened with slight nodularity of the mural leaflet. From the left atrium, the surgeon's finger could be introduced into the left superior pulmonary vein. The left inferior pulmonary vein entered the atrium normally. Mitral commissurotomy was accomplished with the transventricular dilator. The valve orifice was dilated to 4 cm and resulted in the development of minimal mitral regurgitation.

Subsequent occlusion of the anomalous vein obliterated the thrill and no pressure gradient could be demonstrated between the pulmonary vein and left atrium. The anomalous vein was then doubly ligated and divided. The chest was closed in the standard manner. Microscopic examination of the resected atrial appendage revealed no Aschoff bodies.

Postoperative Physical Examination

The continuous murmur was no longer audible.

Discussion

Developmental anomalies of the pulmonary and systemic venous systems have been reviewed by others.2-5 The early embryologic systemic venous system is represented by the left and right horns of the sinus venosus. Into these drain the vitelline, umbilical, azygos, and cardinal veins. The right vitelline vein persists later as the inferior vena cava (IVC) and the right common cardinal vein persists as the superior vena cava (SVC). The coronary sinus develops from the left horn of the sinus venosus and the left common cardinal vein is normally obliterated and represented by the ligament of Marshall. The left anterior cardiac vein may, however, persist as a left superior vena cava (LSVC) and when present usually drains into the coronary sinus and thence right atrium. The precursors of the pulmonary veins begin as a capillary plexus which serves the foregut as well as the developing lungs. This capillary plexus has connections with the cardinal venous system, but these are usually lost as the pulmonary venous connection occurs with the left atrium. Part of the pulmonary venous system may retain its connections with the cardinal venous system and fail to connect with the left atrium. Such is the case in anomalous pulmonary venous connections where one or more pulmonary veins drain into the SVC, IVC, coronary sinus, or a persistent LSVC, and subsequently the right atrium.6 In the present case, the superior pulmonary vein was connected to the left atrium and to a large anomalous vein which led to the innominate vein (fig. 3). The anomalous vein was anterior to the pulmonary artery. A ligament of Marshall could not be identified and the coronary sinus appeared normal. The most likely embryologic explanation is that the anomalous connection represents persistence
of a primitive connection with the cardinal system as well as the development of the definitive connection with the left atrium. A patient with a similar anatomic venous drainage was described in 1916 by McCotter.7 A coronary sinus abnormality could become important if the ostium of the coronary sinus into the right atrium were closed and coronary sinus blood must reach the right atrium by way of the anomalous vein. Simple ligature of the anomalous vein would then be tragic.

Several cases of mitral stenosis with partial anomalous pulmonary venous drainage (APVD) have now been described.8-19 The diagnosis of accompanying partial APVD has been made unexpectedly at the time of catheterization or operation. Aldridge and Wigle18 have described the differential pulmonary capillary wedge pressures as a clue to the diagnosis. The normally draining lung reflects the high left atrial pressure, while the wedge pressure from anomalously drained lung reflects right atrial pressure. The association of partial APVD and rheumatic mitral stenosis appears to be no more than coincidental, since partial APVD occurs in approximately one out of 350 persons.20 Only one case of mitral stenosis has been reported with partial APVD which involved an extrapericardial connection between left atrium and right atrium.21 In that case, a vertical venous connection, arising just where the left pulmonary vein entered the left atrium, connected the pulmonary veins to the innominate vein. There was no atrial septal defect, and after mitral commissurotomy the abnormal vein was ligated. Neither a diastolic nor a continuous murmur was heard prior to surgery.

The case described herein is the first one in which a continuous murmur has been recognized to result from the association of mitral stenosis and partial APVD. The murmur is assumed to be generated by the continuous flow of blood from the pulmonary venous system toward the right atrium through the anomalous venous connection. This assumption is supported by the finding of a continuous thrill over the anomalous vessel at the time of surgery. Keith's group22 described a continuous murmur occurring in four of 58 cases of total APVD. In all four cases, the drainage was to the left innominate vein, and the murmur was heard in the second left intercostal space and sounded like a venous hum. Venables and co-workers23 described two cases of total APVD with continuous murmurs due to constrictions in the ascending vertical pulmonary venous trunks. Ross1 and Aykent24 and their associates have reported continuous murmurs in patients with the combination of left atrial hypertension and a small atrial septal defect. The murmurs were heard best at the lower right sternal border and were reported to increase with inspiration. Ross's group1 reported in one of their cases that the interatrial pressure gradient increased with inspiration following a lag of two or three cardiac cycles, probably due to delayed transmission, to the left heart, of the inspiratory augmentation of right ventricular filling.25 In the present case, there was no distinct change with respiration in the intensity of the continuous murmur by auscultation or phonocardiography. The most likely explanation is that in the present case, because of the large size of the anomalous vein and shunt, changes in the intensity of the murmur might not be expected with small incremental increases in flow.

Since operation, the patient has noted disappearance of dyspnea and fatigue. His prognosis should be the same as other patients with mitral stenosis, since the anomalous vein has been ligated and divided, and a left-to-right shunt no longer exists.

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