Interatrial Communication and Left Atrial Hypertension
A Cause of Continuous Murmur

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The detection of a continuous murmur is of considerable importance in clinical diagnosis, since the number of cardiac malformations accompanied by this physical finding are limited. Most commonly, a continuous murmur results from a shunt between the aorta and the right side of the heart or pulmonary artery and is produced by lesions such as patent ductus arteriosus, aortopulmonary septal defect, and ruptured aneurysm of the sinus of Valsalva. Less commonly, continuous murmurs may occur with systemic, coronary, or pulmonary arteriovenous fistulae, anomalous pulmonary venous drainage, and dilated collateral vessels, within the lung or in the chest wall;1 dilated mammary vessels may also be responsible for a continuous precordial souffle during pregnancy. It is the purpose of this report to describe the findings in three patients with a previously unrecognized cause of continuous murmur, interatrial communication and left atrial hypertension.

Clinical Summaries

Case 1
This 36-year-old woman (no. 04-60-59) was admitted to the National Heart Institute because of dyspnea and palpitations. There was no history of rheumatic fever and she had been told of an enlarged heart for the first time at the age of 28. A heart murmur was subsequently noted, and 2 years prior to admission, a cardiac catheterization was performed at another hospital because of progressive dyspnea. The pulmonary artery pressure was 30/12 mm Hg, the right atrial pressure was 4 mm Hg, and the mean pressure in the left atrium, determined by transseptal puncture, was 20 mm Hg. She was considered to have mitral stenosis, and at mitral commissurotomy the valve orifice was estimated to be 0.5 by 0.8 cm2, and a

Figure 1
Phonocardiograms recorded during inspiration (INSPIR.) top, and expiration (EXPIR.) bottom, in patient 1. S1, first heart sound; A2, aortic valve closure; P2, pulmonic valve closure; 4 L.I.C.S., fourth left intercostal space; S.M., systolic component of the continuous murmur; D.M., diastolic component. Note the diminution of the diastolic component of the murmur during expiration. Time lines, 0.04 second.

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small amount of mitral regurgitation was palpable. It was thought that a satisfactory opening was achieved by finger fracture of the valve. One year later dyspnea and palpitations-recurred, and several weeks prior to the patient's admission to the National Heart Institute, a continuous murmur was noted for the first time by a physician who had examined the patient on numerous occasions previously.

On physical examination there was a right ventricular lift. The second heart sound was widely split, did not vary with respiration, and the pulmonic component was slightly accentuated. At the apex there was an opening snap of the mitral valve and a grade III/VI presystolic rumbling murmur. A grade III/VI continuous murmur was audible in the fourth and fifth interspaces to the right of the sternum and over the xiphoid process. This murmur increased in intensity on inspiration (fig. 1), and was abolished by the Valsalva maneuver. The electrocardiogram showed right atrial enlargement and right ventricular hypertrophy. Roentgenographic and fluoroscopic examinations revealed left atrial enlargement slightly exaggerated pulmonary vascular markings, and calcification in the region of the mitral valve.

At cardiac catheterization, the mean pressure in the right atrium was 4 mm. Hg, the right ventricular pressure was 40/7 mm. Hg, and the pulmonary artery pressure 40/15 mm. Hg. The intracardiac phonocardiogram (Phono.) and intracardiac pressure tracings as the Telco catheter was withdrawn from the right ventricle (RV) into the right atrium (RA). The continuous murmur becomes apparent as the catheter is withdrawn across the tricuspid valve into the RA. Time lines, 0.10 second.

Simultaneous recordings of intracardiac phonocardiogram (Phono.) and intracardiac pressure tracings as the Telco catheter was withdrawn from the right ventricle (RV) into the right atrium (RA). The continuous murmur becomes apparent as the catheter is withdrawn across the tricuspid valve into the RA. Time lines, 0.10 second.
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The interatrial septum was identified (fig. 4). This defect was caudad to the fossa ovalis, measured 7.5 mm. in diameter, and had a thickened, fibrotic rim. The mitral valve orifice measured approximately 1 cm., and a small degree of mitral regurgitation was noted. The valve leaflets were thickened, the commissures fused, and the chordae tendineae shortened. A mitral commissurotomy was performed, and the defect in the atrial septum was closed by direct suture. The patient's postoperative course has been uneventful, and the continuous murmur is no longer audible.

Case 2

This 10-year old boy (no. 04-63-50) had a history of cyanosis and heart failure during infancy. At the age of 7 months a chest roentgenogram showed pulmonary congestion and cardiac en-

Figure 3

Top. Simultaneous recordings, from above downward, of electrocardiogram, pneumogram (Pneumo.), interatrial phonocardiogram (Phono.), left atrial pressure (LA) and right atrial pressure (RA) in patient 1. The timing of expiration (EXP.) and inspiration (INSP.) is shown at the bottom of the tracing. Time lines, 0.10 second. Note that during late inspiration the interatrial pressure gradient increases. Bottom. Similar recordings during inspiration followed by a Valsalva maneuver. Note the reduction of the interatrial pressure gradient during the Valsalva maneuver. Time lines, 0.04 second.
The electrocardiogram revealed right ventricular hypertrophy. Reduced activity and slow weight gain were noted during childhood. At the age of 9 years he developed increasing dyspnea and cyanosis.

On admission, the child was found to be physically underdeveloped and to exhibit moderate cyanosis and clubbing of the digits. There were a precordial bulge and a right ventricular lift. The second heart sound was accentuated and single, and there was an ejection sound in the second left intercostal space. A grade II/VI rumbling diastolic murmur was heard along the lower left sternal border, and a grade II/VI systolic ejection murmur, which increased in intensity with inspiration, was noted over the lower sternum. A grade IV/VI rough continuous murmur, which increased in intensity with inspiration and became markedly diminished during forced expiration, was audible in the fourth right and left intercostal spaces and over the right posterior chest (fig. 5).

Roentgenographic and fluoroscopic examinations of the chest disclosed marked enlargement of the right ventricle and increased pulmonary vascular markings, but no enlargement of the left atrium. The electrocardiogram showed right axis deviation, right bundle-branch block, right atrial enlargement, and right ventricular hypertrophy.

At cardiac catheterization the mean pressure in the right atrium was 4 mm. Hg, the right ventricular pressure was 96/5 mm. Hg, and the pulmonary arterial pressure 95/60 mm. Hg. The pressure in the brachial artery was 96/70 mm. Hg, and the arterial oxygen saturation was 82 per cent. The inhaled Krypton\textsuperscript{85} test disclosed a large left-to-right shunt entering the right atrium. Indicator-dilution curves confirmed the presence of a left-to-right shunt, and in addition showed that a right-to-left shunt originated from the right ventricle. At transseptal puncture of the left atrium, the mean left atrial pressure was 17 mm. Hg and there was a large pressure gradient across the interatrial septum (fig. 6). The intracardiac phonocardiogram localized the continuous murmur to the right atrium. A selective angiogram, with injection into the left atrium, revealed mitral atresia and a small atrial septal defect, a single ventricle, and transposition of the great vessels (fig. 7).

**Case 3**

This 48-year old woman (no. 00-87-65) was admitted to the National Heart Institute because of dyspnea and edema. A heart murmur had been present since an episode of rheumatic fever in early childhood. At 18 years of age she developed congestive heart failure necessitating treatment with digitalis. Three years before admission she noted increasing shortness of breath, ankle edema, and chest pain suggestive of angina pectoris. One year later a continuous cardiac murmur was heard for the first time, and cardiac catheterization was performed at another hospital. An increase in the oxygen content of blood obtained from the right atrium compared to that in the vena cavae was found, and a sinus of Valsalva aneurysm with rupture into the right atrium was suspected.

On admission the patient was found to be chronically ill and emaciated, and both ventricles were enlarged to palpation. There was a continuous thrill in the right midclavicular line in the third and fourth intercostal spaces. The second heart sound at the pulmonic area was accentuated, finely split, and the splitting did not vary with respiration. A grade IV/VI continuous murmur was heard along the right sternal border in the fourth, fifth, and sixth intercostal spaces. In addition, a grade IV/VI basal ejection murmur and a grade III/VI apical mid-diastolic rumble were audible.

The electrocardiogram showed atrial fibrillation, right axis deviation, and right ventricular hypertrophy. Roentgenographic and fluoroscopic examinations of the chest revealed biventricular enlargement and a greatly dilated left atrium. At cardiac catheterization, the mean right atrial pressure was
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6 mm. Hg; the right ventricular pressure, 45/10 mm. Hg; and the pulmonary arterial pressure, 45/20 mm. Hg. Serial oxygen samples and the inhaled nitrous oxide test demonstrated that a left-to-right shunt entered the right atrium, resulting in a pulmonary-to-systemic blood-flow ratio of 2.4/1.0. A retrograde aortogram was performed but immediately after injection of the contrast substance, ventricular fibrillation occurred, and all efforts at resuscitation were unsuccessful.

At autopsy, the heart weighed 370 Gm. The left atrium was thin-walled and greatly enlarged. The mitral valve admitted a fingertip, and the valve cusps were thickened and calcified, with fusion of both commissures. In addition, there was a defect in the interatrial septum of the ostium secundum type that measured 1.0 by 0.5 cm. (fig. 8). The right atrium was also enlarged, and the right ventricle was dilated and hypertrophied.

Discussion

The anatomic and hemodynamic studies in these three patients suggested that in each the continuous murmur resulted from the flow of blood across the interatrial communication throughout the cardiac cycle. The demonstration of a continuous pressure gradient between the atria supported this concept, which was confirmed in two patients by the localization of the continuous murmur to the right atrium by means of intracardiac phonocardiography. The left atrial hypertension responsible for the sustained left-to-right shunt resulted from the combination of obstruction at the mitral valve and the unusually small defect in the atrial septum. Although continuous murmurs are not audible in patients with the usual forms of atrial septal defect, in the patients described herein the small size of the defects, the significant interatrial pressure gradients and the relatively large left-to-right

Figure 5

Phonocardiograms recorded during various phases of respiration in patient 2. Note the increased intensity of the murmur during inspiration and its diminution during held expiration.
Continuous pressure tracing recorded as the catheter was withdrawn from the left atrium (LA) to the right atrium (RA), demonstrating the gradient between these chambers. Time lines, 0.10 second.

Selective angiocardiogram with left atrial injection exposed in the lateral projection (left) and frontal projection (right) in patient 2. L.A., left atrium; R.A., right atrium. The arrows delineate the jet of contrast material traversing the interatrial communication.

Shunts, undoubtedly, resulted in the turbulent flow responsible for the continuous murmurs. The continuous murmurs in these patients had several distinctive features that should permit their differentiation from the continu-
ous murmurs associated with shunts originating from a systemic artery. First of all, the murmurs were localized to the lower sternal area and were louder on the right than on the left side. Secondly, the murmurs increased in intensity during inspiration and, thirdly, there was marked reduction or abolition of the murmurs by the Valsalva maneuver. The pressure tracings recorded simultaneously from the right and left atria in patient 1 provide a possible explanation for the variations in the intensity of the murmurs observed with respiratory maneuvers. It was noted that during late inspiration the pressure in the left atrium increased more than that in the right atrium (fig. 3). Recent experiments have shown that the inspiratory augmentation of right ventricular filling is transmitted to the left heart after a delay of two or three cardiac cycles. Since, in these patients, neither the atrial defect nor the mitral orifice provided an adequate outlet for this augmentation of inflow into the left atrium, elevation of left atrial pressure, increase of the interatrial pressure gradient, and augmentation of the murmur ensued. In contrast, the forced expiration of the Valsalva maneuver resulted in a disproportionate rise of the right atrial pressure with abolition of the interatrial pressure gradient and the murmur. The Valsalva maneuver should prove particularly useful in the clinical differentiation of a continuous murmur through an interatrial communication from one originating in a systemic artery, since it is unlikely that in the latter type the high pressure gradient could be abolished in this manner. Additional physical signs that should be helpful in establishing a clinical diagnosis were present in the two patients in whom mitral stenosis was associated with the interatrial communication. These were the typical diastolic rumbling murmurs at the apex and the fixed splitting of the second heart sound during the respiratory cycle.

Although the hemodynamic basis for the continuous murmur was similar in each of the three patients, the anatomic defects were somewhat different in each instance. In patient 2, the entire pulmonary venous return traversed a relatively small interatrial communication. Thus, the combination of mitral atresia and atrial septal defect is an additional cause for continuous murmurs in patients with cyanotic congenital heart disease. Patient 3 may be designated an example of the Lutembacher syndrome, although the small size of the atrial septal defect is unusual and a continuous murmur is not considered to be characteristic of this malformation. In patient 1 the history suggests that the continuous murmur became audible only when restenosis of the mitral valve resulted in left atrial hypertension and in a pressure gradient between the atria; furthermore, the position and morphology of the interatrial communication suggested that it was acquired at the time of the initial transseptal left atrial puncture. It is remarkable, perhaps, that this complication has not been encountered more frequently following left atrial puncture in patients with left atrial hypertension. Thus far it has not been observed following the

Figure 8
Photograph of postmortem specimen showing the interatrial septum viewed from the left atrium in patient 3. The small defect in the septum is apparent.
more than 800 transseptal left atrial punctures performed in this laboratory.

Summary

The combination of left atrial hypertension and a small atrial septal defect was observed in three patients and was found to be responsible for a continuous heart murmur. Mitral stenosis was present in two patients, and mitral atresia in the third. The continuous murmurs were loudest over the lower sternum, were augmented by inspiration, and were reduced or abolished by the Valsalva maneuver. Cardiac catheterization studies demonstrated the presence of a left-to-right shunt at the atrial level in each instance. The continuous murmurs were localized to the right atrium by intracardiac phonocardiography in two patients, and alterations in the interatrial pressure gradient during respiratory maneuvers were documented.

References


Fear of the New

The mind delights in a static environment, and if there is any change to be itself the source of it. Change from without, interfering as it must with the sovereignty of the individual, seems in its very essence to be repulsive and an object of fear. A little self-examination tells us pretty easily how deeply rooted in the mind is the fear of the new, and how simple it is when fear is afoot to block the path of the new idea by unbelief and call it scepticism, and by misunderstanding and call it suspended judgement. The only way to the serene sanity which is the scientific mind—but how difficult consistently to follow—is to give to every fresh idea its one intense moment of cool but imaginative attention before venturing to mark it for rejection or suspense, as alas nine times out of ten we must do. In this traffic it is above all necessary not to be heavy-handed with ideas. It is the function of notions in science to be useful, to be interesting, to be verifiable, and to acquire value from any one of these qualities. Scientific notions have little to gain as science from being forced into relation with that formidable abstraction 'general truth.' Any such relation is only too apt to discourage the getting rid of the superseded and the absorption of the new which make up the very metabolism of the mind.—The Collected Papers of Wilfred Trotter, F.R.S. London, Oxford University Press, 1946, pp. 28-29.
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Circulation. 1963;28:853-860
doi: 10.1161/01.CIR.28.5.853

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

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