

Sequential heart murmurs in a 43-year-old man with congenital heart disease

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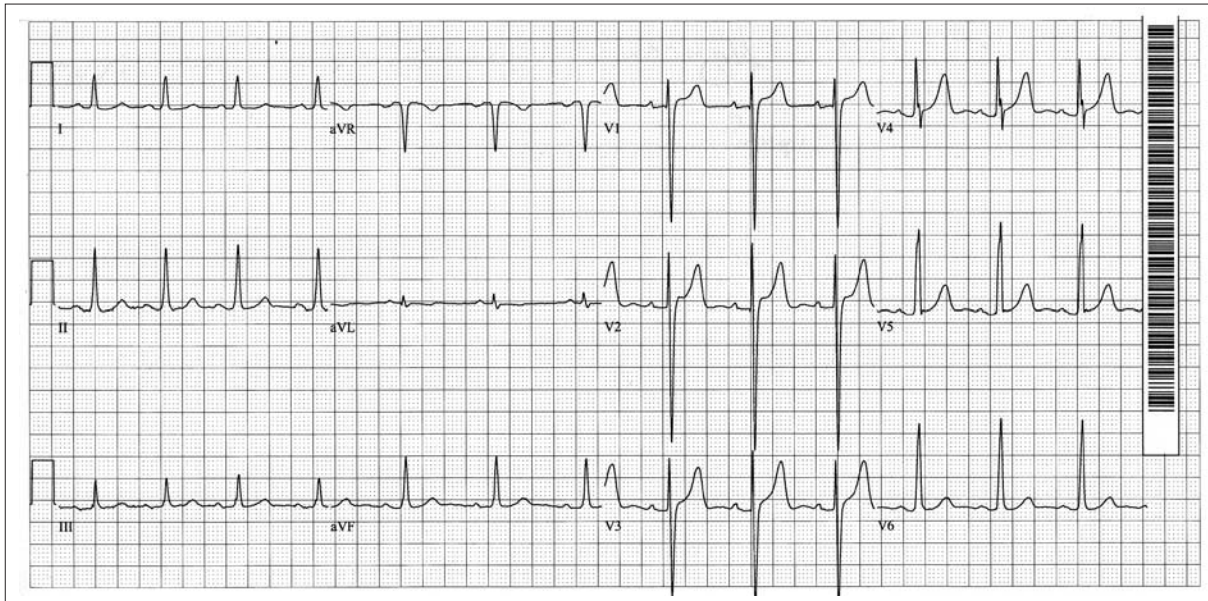


Figure. Electrocardiogram recorded on admission at age 43 years. The rhythm is normal sinus. The voltage of the QRS complexes meets several criteria for left ventricular enlargement: $SV_1 + RV_5$ or $RV_6 > 35$ mm (1); $SV_2 + RV_5$ or $RV_6 > 45$ mm (2); $SV_1 + RaVL > 22$ mm when $TV_1 \geq 2$ mm (3). The electrocardiogram otherwise is normal.

Within a week of this patient's birth, a physician discovered a loud systolic murmur. When the patient was 14 years old, the murmur disappeared. He developed a second murmur when in his mid-30s. At age 43, some 6 months before admission, he noted dyspnea on exertion, which became worse during the month prior to his hospital admission. He was never cyanotic.

His pulse was 80 beats per minute and regular; his blood pressure, 152/72 mm Hg; and his respiratory rate, 18 breaths per minute. Breath sounds were normal. There was no cyanosis, clubbing, edema, or sign of infective endocarditis. The carotid pulses were fuller and brisker than normal. The left ventricular impulse was prominent with the patient on his left side. No gallop could be heard. A continuous murmur with a peak in intensity in systole and another in diastole was loudest in the third left intercostal space. The admission electrocardiogram showed left ventricular enlargement (*Figure*) (1–3).

A loud murmur that is heard soon after birth and subsequently disappears is usually due to a ventricular septal defect that subsequently closes spontaneously. The patient remembers

being told that he had a hole between the pumping chambers of his heart.

Ventricular septal defect is the most common congenital cardiovascular malformation that manifests itself at the time of birth. (Bicuspid aortic valves and mitral valves that subsequently prolapse are more common but usually function normally at birth and for many years thereafter [4].) Complete spontaneous closure of ventricular septal defects is common (5). The prevalence of spontaneous closure may be as high as 75% to 80%, with most closures occurring in the first 2 years of life (6). A few spontaneous closures occur in adolescence or even adulthood (5).

Closure occurs by endocardial fibrous tissue proliferation, the result of turbulent flow through the lesion. The fibrous

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Table. Cardiac catheterization data

Chamber	Pressure (mm Hg)	Blood oxygen saturation (%)
Superior vena cava	NA	68
Inferior vena cava	NA	73
Right atrium	A = 19; V = 14; mean = 13	73
Right ventricle	58/15	83
Pulmonary artery	50/26; mean = 36	83
Pulmonary arterial wedge	A = 24; V = 22; mean = 19	NA
Left ventricle	131/18	NA
Aorta	131/68; mean = 93;	97
pulmonary/systemic flow = 1.9/1		

NA indicates not available.

tissue closes muscular ventricular septal defects directly and closes membranous defects by attaching the septal leaflet of the tricuspid valve to the margins of the defect (5).

The murmur that developed in the patient's mid-30s and was heard by us on admission was continuous, had peak intensities both in systole and in diastole, and was best heard in the third left intercostal space. Together with the brisk carotid pulses and the wide systemic arterial pulse pressure, the murmur suggested an aorto-right ventricular shunt. The spontaneous development of such a shunt in adulthood is usually due to rupture of a congenital sinus of Valsalva aneurysm. This was confirmed by echocardiography and by cardiac angiography and catheterization (Table), each of which demonstrated a shunt from the right sinus of Valsalva into the outflow tract of the right ventricle.

Congenital aneurysms of the sinuses of Valsalva are uncommon. Approximately two thirds arise from the right sinus and usually rupture into the right ventricle or, less commonly, into the right atrium (7, 8). About one quarter arise from the non-coronary sinus and rupture into the right atrium. Congenital aneurysms of the left sinus are rare. Some 75% to 80% of patients are males (7, 8).

The congenital fault is a weakness at the junction of the aortic media and the wall of the sinus of Valsalva (8). The aneurysm develops later, and the rupture comes much later, with the average age at rupture being in the early 30s (7, 8). Some unruptured aneurysms are discovered by chance on an echocardiogram done for another reason; a few cause symptoms or signs by pressure on other structures; and a minority never rupture (8). Most rupture, and then the aneurysm resembles a windsock with a large opening from the sinus of Valsalva and a narrow opening in the chamber into which it has ruptured.

The rupture may be accompanied by chest pain and dyspnea lasting days to weeks or may be asymptomatic, as in our patient (8). The left-to-right shunt is usually of moderate size, as it was in our patient, whose pulmonary-to-systemic flow ratio was 1.9:1. Most patients eventually develop symptoms, and if the defect is not closed, full-blown congestive heart failure and death usually ensue.

The sinus of Valsalva aneurysm often distorts the attachment of the aortic valvular leaflet of the involved sinus. Aortic regurgitation may result but usually is mild. This was the case in our patient, in whom the aortic end of the aneurysm was closed by a patch after the aneurysmal tissue was excised. The aortic valve was not disturbed, and a postoperative echocardiogram showed only mild aortic regurgitation. The patient also continued to have good left ventricular systolic function postoperatively.

Ventricular septal defects are present in some 40% of patients who rupture a sinus of Valsalva aneurysm and are more common when the aneurysm involves the right sinus (7). Many of the ventricular septal malformations involve the outlet septum, and in those patients progressive aortic regurgitation may occur due to prolapse of a cusp into the defect.

Other congenital cardiovascular defects are relatively uncommon in patients with ruptured sinus of Valsalva aneurysms. To our knowledge, this is the first instance in which a coexisting ventricular septal defect closed spontaneously before the aneurysm ruptured, and perhaps before it developed.

The ventricular septal defect and the ruptured sinus of Valsalva aneurysm each caused increased blood flow through both ventricles and the left atrium. With each malformation, however, most of the extra work was performed by the left ventricle. Thus, the electrocardiographic finding of isolated left ventricular hypertrophy was not unexpected.

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