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A 23-year-old man with a continuous heart murmur

A 23-YEAR-OLD man presented to his family physician seeking treatment for a “cold.” On examination, the physician heard a loud murmur over the precordium and referred the patient to a cardiologist for evaluation. Transthoracic echocardiography revealed bilateral ventricular dilatation, a tricuspid aortic valve with moderate to severe aortic regurgitation, and left-to-right shunting between the aorta and right ventricle. He was referred to The Cleveland Clinic and admitted for further workup.

The patient denied any chest pain, dyspnea at rest or on exertion, palpitations, diaphoresis, dizziness, or near syncope. As an infant, he had undergone cardiac catheterization for the evaluation of a loud murmur, and his parents were told that he had “a hole in his heart” but that no further evaluation or treatment was necessary. The patient had not seen a physician within the past 15 years.

On examination, his blood pressure was 180/50 mm Hg and his pulse was 92. The lungs were clear to auscultation. A grade 5/6 continuous murmur was most prominent over the third left intercostal space at the sternal edge, but was heard well over the entire precordium. A palpable thrill was present along the left sternal border. All peripheral pulses were bounding. The jugular venous pressure was not elevated.

■ THE DIFFERENTIAL DIAGNOSIS

1 What is the most likely diagnosis?

- Atrial septal defect
- Ventricular septal defect
- Coarctation of the aorta
- Ruptured sinus of Valsalva aneurysm

Atrial septal defect is associated with a systolic ejection murmur, most prominent over the second or third intercostal space at the left sternal border, and a fixed split S₂. The murmur is created by increased blood flow through the main pulmonary artery and not by left-to-right shunting across the defect.

Ventricular septal defect is associated with a holosystolic murmur heard best along the left lower sternal border. The murmur is due to left-to-right shunting across the defect.

Coarctation of the aorta may be associated with a continuous murmur thought to be caused by increased blood flow through the intercostal vessels. Aortic regurgitation may accompany coarctation if a bicuspid aortic valve is present. However, coarctation is not associated with left-to-right shunting.

Ruptured sinus of Valsalva aneurysm is the diagnosis. Transesophageal echocardiography revealed that our patient had a right coronary sinus of Valsalva aneurysm that had ruptured into the right ventricle.

■ SINUS OF VALSALVA ANEURYSM

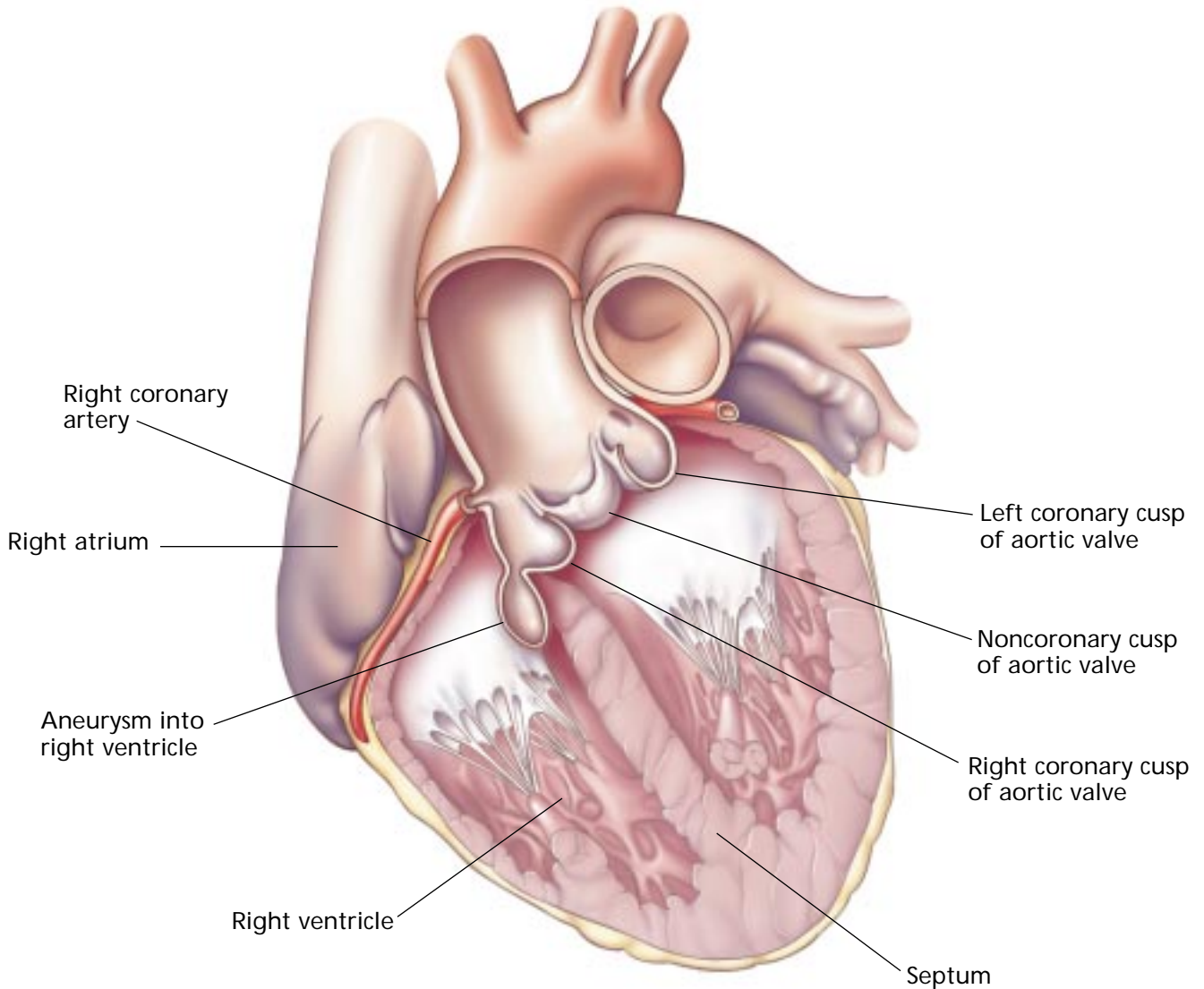
The sinuses of Valsalva are dilatations in the aortic wall immediately superior to the attachments of the three aortic valve cusps (FIGURE 1). The sinuses are named according to their relationship with the coronary arteries: ie, the right coronary sinus, the left coronary sinus, and the noncoronary sinus.¹ Aneurysms of the sinuses of Valsalva occur where a lack of fusion exists between the aortic media and the annulus fibrosus of the aortic valve.²

The right coronary sinus is the most common site of aneurysm formation.³ Most aneurysms originating from the right coronary

The problem is three times more common in men than in women



■ Aneurysm of the sinus of Valsalva



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FIGURE 1. Aneurysm of the sinus of Valsalva occurs where a lack of fusion exists between the aortic media and the annulus fibrosis of the aortic valve. Most aneurysms that originate in the right coronary sinus rupture into the right ventricle, producing left-to-right shunting, as seen in **FIGURE 2**.

sinus rupture into the right ventricle, producing left-to-right shunting. Right coronary sinus aneurysms may also rupture into the right atrium. Noncoronary sinus aneurysms generally rupture into the right atrium. Left coronary sinus aneurysms are extremely rare,

but they may rupture into the pericardium, resulting in cardiac tamponade and death if not quickly recognized.¹ Sinus of Valsalva aneurysms appear to be congenital and are three times more common in men than in women.⁴

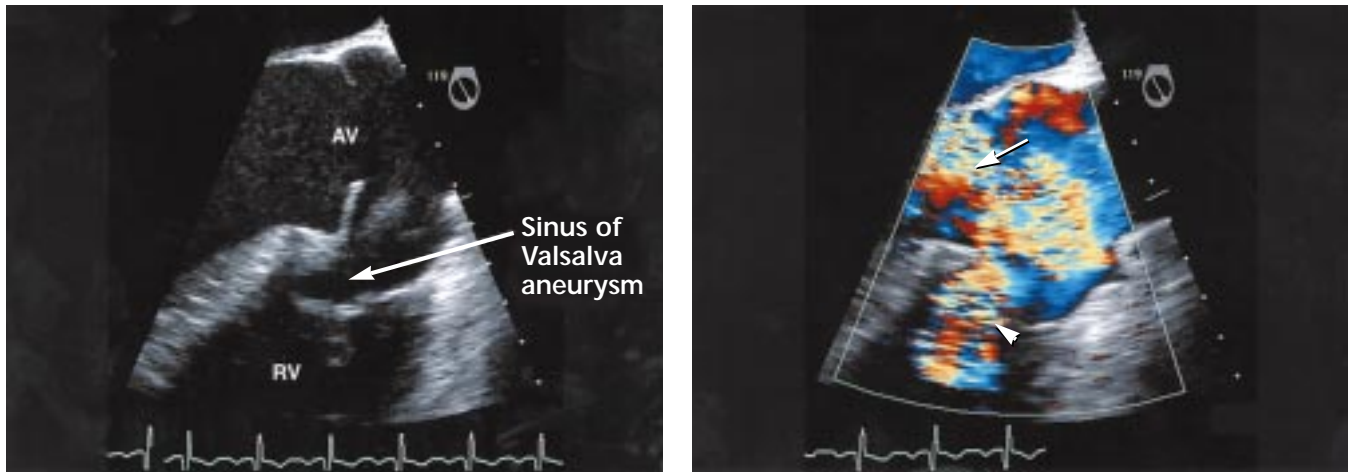


FIGURE 2. The transesophageal echocardiogram (TEE) at left shows the ruptured sinus of Valsalva aneurysm, while the TEE with Doppler at right shows left-to-right shunting (arrowhead) between the aorta and the right ventricle and aortic regurgitation (arrow). AV aortic valve, RV right ventricle.

■ TYPICAL PRESENTATION

Sinus of Valsalva aneurysms have one of three basic pathologic patterns.¹

Unruptured aneurysms usually produce no symptoms and are often incidentally found during cardiac catheterization or echocardiographic examination, or at autopsy. However, the aneurysm may compress the interventricular septum, resulting in complete heart block with subsequent dizziness or syncope. Coronary artery compression may also occur, producing myocardial ischemia and chest pain. Occasionally, a patient with an unruptured sinus of Valsalva aneurysm presents with symptoms related to chronic aortic regurgitation, including dyspnea on exertion, orthopnea, paroxysmal nocturnal dyspnea, and syncope. Aortic regurgitation occurs as progressive aneurysmal enlargement produces dilatation of the aortic cusp.

A small perforation that slowly enlarges may remain asymptomatic for several years because of hemodynamic adjustment. Eventually, however, symptoms related to volume overload, such as dyspnea and exercise intolerance, develop as the degree of shunting increases.

An aneurysm that acutely ruptures is often heralded by the sudden onset of dyspnea and severe chest pain.⁵ Following this initial symptomatic period, the patient may become asymptomatic even without treatment, as the body adjusts hemodynamically to the left-to-

right shunting. However, without treatment, symptoms of congestive heart failure eventually result as the shunting and volume overload overwhelm the compensatory mechanisms. The second and third decades of life are the average age for sinus of Valsalva aneurysms to rupture.⁴

■ PHYSICAL EXAMINATION

In general, unruptured sinus of Valsalva aneurysms produce no murmurs. However, in rare cases an unruptured aneurysm produces a continuous murmur due to blood flow in and out of the aneurysmal pouch. The murmur of aortic regurgitation may be heard if aortic cusp dilatation occurs. Eventually, the patient shows the classic physical signs of chronic aortic regurgitation.

2 Physical findings associated with chronic aortic regurgitation include all of the following except which one?

- Pulsations of the capillary bed when the nail bed is compressed (Quincke sign)
- Visible pulsations of the uvula (Müller sign)
- Up-and-down head bobbing (De Musset sign)
- “Pistol shots” heard over the femoral artery (Traube sign)
- A decrease in murmur intensity with isometric handgrip

The murmur may be very loud and is best heard over the lower left sternal border

The first four signs have been found in association with chronic aortic regurgitation. These findings are due to a large stroke volume and widened pulse pressure. They are not specific for aortic regurgitation and may occur in any chronic high-flow state.

Isometric handgrip increases arterial pressure, left ventricular systolic pressure, and left ventricular diastolic pressure. The elevation in arterial pressure increases the flow gradient for aortic regurgitation, thereby *increasing* the intensity of the murmur.

Once a sinus of Valsalva aneurysm ruptures, an ensuing continuous murmur can be heard. The continuous murmur may be extremely loud, is usually best heard along the lower left sternal border, and is often accompanied by a palpable thrill along the left sternal border. Due to left-to-right shunting and subsequent volume overload, a left-sided or right-sided third heart sound may be heard.

3 All of the following are causes of continuous murmurs except which one?

- Patent ductus arteriosus
- Coronary artery-to-right heart fistula
- Bicuspid aortic valve with resultant aortic stenosis and aortic regurgitation
- Venous hum
- Mammary souffle

A continuous murmur is defined as a murmur that begins in systole and continues through the second heart sound and into part or all of diastole.⁶ The murmur is created when blood flows from a region of higher pressure or resistance into one of lower pressure or resistance without interruption between systole and diastole.⁶

Patent ductus arteriosus classically produces a continuous murmur that has been described as “machine-like.” Caused by blood flowing from the aorta to the left pulmonary artery, the murmur is loudest over the second intercostal space at the left sternal border.

Coronary artery-to-right heart fistulas produce a continuous murmur that may be heard best at several different locations over the precordium, depending on which coronary artery and vessel or chamber is involved.⁷

Venous hum is a continuous murmur that

is believed to result from turbulent blood flow as the subclavian and internal jugular veins join to form the brachiocephalic veins.⁷ This murmur is heard in almost all children and in many young adults. The venous hum is best heard slightly superior to the clavicle, either between the insertions of the sternocleidomastoid muscle or medial to the muscle. Although it may be heard on either side, it is better heard on the right side. The murmur is best heard with the stethoscope bell, while using very light pressure. Elevating and rotating the chin away from the side of auscultation will often accentuate the murmur. Pressure on the neck superior to the site of auscultation will eliminate the murmur, allowing differentiation between a venous hum and arterial or thyroid bruits, which will not be eliminated when pressure is applied above the stethoscope.⁷

Mammary souffle is a continuous arterial murmur created by increased blood flow to the breast during pregnancy and lactation. The murmur generally begins in the second or third trimester of pregnancy and usually resolves by the second postpartum month. The mammary souffle is heard best along the left sternal border and may be eliminated by firm pressure with the stethoscope.⁷

Bicuspid aortic valve with resultant aortic stenosis and aortic regurgitation does not cause a continuous murmur but rather a to-and-fro murmur, as blood flows in opposite directions during the cardiac cycles, as opposed to a continuous murmur in which blood flows in the same direction.⁸ Also, the murmur combination of aortic stenosis and regurgitation does not occur through the second heart sound, as the murmur of aortic stenosis ceases prior to the A₂ component of the second heart sound.

■ SIGNS OF RUPTURED SINUS OF VALSALVA ANEURYSM

When a sinus of Valsalva aneurysm ruptures, the pulses are generally bounding, with a rapid rise and bisferious contour due to the large stroke volume. The jugular venous pressure waveform is usually normal with small and slowly progressive shunting. If shunting occurs secondary to sudden aneurysmal rupture, the a

TEE is the diagnostic test of choice



and v waves may become prominent in the setting of congestive heart failure.¹

The electrocardiogram is generally normal in cases of unruptured sinus of Valsalva aneurysms. However, compression of the AV node or bundle of His may result in atrioventricular conduction disturbances, such as complete heart block. Once a sinus of Valsalva aneurysm ruptures, electrocardiographic signs of biatrial enlargement and biventricular hypertrophy or enlargement may ensue. The chest radiograph may also reveal signs of chamber enlargement in cases of sinus of Valsalva aneurysm rupture, as well as findings associated with congestive heart failure.

■ DIAGNOSIS

Unruptured aneurysms are generally found incidentally during cardiovascular diagnostic studies or at autopsy. Ruptured sinus of Valsalva aneurysm should be strongly suspected in any young patient with a continuous murmur and a history of chest pain or signs and symptoms of congestive heart failure, or both.

The preliminary diagnostic workup includes electrocardiography and chest radiography, which often reveal nonspecific findings.

Transthoracic echocardiography often reveals aneurysmal formation with left-to-right shunting and chamber dilatation and hypertrophy.

Transesophageal echocardiography (TEE) is the diagnostic test of choice and provides more precise anatomic detail (FIGURE 2) when compared to the transthoracic technique. TEE also better visualizes associated congenital abnormalities such as ventricular septal defect, atrial septal defect, bicuspid aortic valve, and coarctation of the aorta. Left-sided cardiac catheterization will often reveal left-to-right

shunting at the ventricular or atrial level.

During the diagnostic workup, infectious endocarditis needs to be searched for and ruled out. Endocarditis may develop either on an unruptured aneurysm, precipitating its rupture, or on an already ruptured sinus of Valsalva aneurysm.⁵

■ TREATMENT

Preoperative management involves relieving the signs and symptoms of congestive heart failure caused by volume overload. Also, concurrent cardiac arrhythmias and infectious endocarditis need to be treated.

Surgical repair of the aneurysm is the definitive treatment. At surgery, the aneurysm is resected and the aortic wall is reunited with the heart.⁹ Attempts should be made to preserve the aortic valve, if possible. However, aortic valve replacement may be necessary. Following surgical repair, patients have an excellent prognosis with a normal life expectancy.⁵

Our patient underwent resection of the sinus of Valsalva aneurysm and resuspension of the aortic valve. A patent foramen ovale, which had been detected on TEE, was also closed, and a small ventricular septal defect was discovered and closed. The defect was the likely cause of the patient's cardiac workup as an infant. He did well in the immediate postoperative period, with only 1+ to 2+ aortic regurgitation detected on transthoracic echocardiography prior to discharge. However, repeat transthoracic echocardiographic examinations revealed an increasing severity of aortic regurgitation. The patient subsequently underwent aortic valve replacement and was discharged after an uncomplicated postoperative course.

Always rule out endocarditis during the workup

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