Aortic Valve Regurgitation that Resolved
after a Ruptured Coronary Sinus Aneurysm Was Patched

Sinus of Valsalva aneurysms appear to be rare. They occur most frequently in the right sinus of Valsalva (52%) and the noncoronary sinus (33%). More of these aneurysms originate from the right coronary cusp than from the noncoronary cusp. Surgical intervention is usually recommended when symptoms become evident.

We report the case of a 34-year-old woman who presented with a congenital, ruptured sinus of Valsalva aneurysm that originated from the noncoronary cusp. Moderate aortic regurgitation was associated with this lesion. Simple, direct patch closure of the ruptured aneurysm resolved the patient’s left-to-right shunt and was associated with decreased aortic regurgitation to a degree that valve replacement was not necessary. Only trace residual aortic regurgitation was evident after 3 months, and the patient remained free of symptoms after 6 months.

Our observations support the idea that substantial runoff blood flow in the immediate supra-annular region can be responsible for aortic regurgitation in the absence of a notable structural defect in the aortic valve, and that restoring physiologic flow in this region and equalizing aortic-cusp closure pressure can largely or completely resolve aortic insufficiency. Accordingly, valve replacement may not be necessary in all cases of ruptured sinus of Valsalva aneurysms with associated aortic valve regurgitation. (Tex Heart Inst J 2013;40(4):489-92)

Sinus of Valsalva aneurysm (SVA) was first described in 1839. This lesion is encountered during 0.14% to 0.96% of all open-heart procedures and in 0.09% of postmortem studies. Right coronary cusp aneurysms are the most frequent form of SVA (52% of all SVA cases), and noncoronary cusp aneurysms are diagnosed in 33% of SVA cases.

We present the case of a woman who had a congenital, ruptured SVA of the noncoronary cusp and moderate aortic regurgitation (AR). We discuss our simple, direct patch closure of the aneurysm, and we present our reasoning as to how this treatment eliminated the patient’s AR without the need to replace the aortic valve.

Case Report

In 2011, an active, normotensive, previously healthy 34-year-old black woman with new palpitations presented to a cardiologist. Cardiac auscultation revealed a poorly defined systolic and diastolic murmur, and a transthoracic echocardiogram (TTE) with color-flow Doppler showed moderate AR and ill-defined continuous flow within the right atrium. No definitive diagnosis was made at that time because the patient deferred further evaluation. Three months later, she emergently presented with palpitations, gradually progressive paroxysmal nocturnal dyspnea, and moderate exertional dyspnea. Her examination results were notable for clear lung fields and no clinical signs of volume overload. Cardiac auscultation revealed a loud, distinctive, continuous, machinery-like murmur that was heard best at the upper sternal border.

The patient was referred to our institution for definitive diagnosis. The earlier TTE was evaluated. A new transesophageal echocardiogram (TEE) showed a 3 x 7-mm, narrow, windsock-like, congenital noncoronary SVA with a distal rupture site (Figs. 1A and 1B). The aneurysm originated adjacent to the right fibrous trigone region. The aneurysm protruded into the right atrium, adjacent to the septal leaflet of the tricuspid valve. There was continuous flow from the aorta to the right atrium through the ruptured SVA, confirmed by continuous-wave Doppler (Fig. 1C). The TEE also revealed moderate aortic regurgitation (Fig. 2). Initially, the primary mechanism for
AR appeared to be subtle sclerosis and deficiency of the tip of the right coronary cusp (Figs. 3 and 4). It was decided to proceed with surgical repair.3-6 The SVA was closed by direct patch repair with autologous pericardium via a transaortic approach during cardiopulmonary bypass (CPB) with bicaval cannulation and cross-clamping of the ascending aorta. Particular care was taken not to distort the normal structure of the noncoronary cusp. Immediately after CPB was stopped, a postoperative TEE obtained with the patient under general endotracheal anesthesia showed resolution of the left-to-right shunt and no significant AR (Fig. 5). The patient's postoperative recovery was uneventful. Three months after her discharge from the hospital, a TTE confirmed trace residual AR. The patient remained free of symptoms at the 6-month evaluation.

**Discussion**

The first successful surgical repair of SVA was reported by Lillehei and colleagues,6 and the first percutaneous repair was reported by Cullen and coworkers.7 Congenital SVAs result from a structural anomaly of the distal bulbar septum8; acquired SVAs have several possible causes, including primary aortic cystic medial necrosis or secondary cystic medial necrosis caused by syphilis, atherosclerosis, or bacterial endocarditis.

Current knowledge regarding the natural history of ruptured SVAs has come from early surgical experience9 and isolated case reports,3,10 and the consensus is to proceed with surgical repair when heart-failure symptoms are present. However, in cases of unruptured SVA, the optimal type and timing of surgical intervention are less clear. Early repair could be indicated if there is superimposed infection, malignant arrhythmia, ostial coronary artery obstruction, or right ventricular outflow tract obstruction. In our institution's previous series,3 SVA repair was performed concomitantly with surgical aortic valve replacement in up to 58% of cases.

Frequently, SVA rupture is an acute, catastrophic event that necessitates urgent repair to reverse severe acute heart failure. An unusual aspect of our patient's clinical course is the remarkably subacute presentation. A murmur had been detected before our evaluation, and vague heart-failure symptoms and palpitations developed and worsened during the 3 months from the time of initial presentation to the time of definitive diagno-
sis by TEE. The lack of acute symptoms was probably related to the unusually small caliber of the SVA.

Also noteworthy is that moderate aortic valve regurgitation, as seen on preoperative TEE, was corrected by simple patch closure of the SVA defect alone. Aortic valve regurgitation is frequently associated with SVA (prevalence, up to 44% in our previous SVA case series3), as are ventricular septal defect, bicuspid aortic valve, endocarditis, Marfan syndrome, infundibular pulmonary artery stenosis, isolated aortic stenosis, and combined aortic stenosis and insufficiency. In addition, the auscultation findings suggested several possible diagnoses: continuous murmur with patent ductus arteriosus, coronary arteriovenous fistula, pulmonary arteriovenous fistula, and aortopulmonary window, which should also be considered in patients who have a continuous murmur.

In our patient, postoperative TEE revealed a marked reduction of AR after simple patch closure of the SVA defect. The pre-repair AR might have resulted from aortic valve cusp malcoaptation caused by unbalanced diastolic closing pressure of the opposing cusps. It is conceivable that blood flow from the ruptured SVA produced a low-pressure zone (the Venturi effect) over the noncoronary cusp and distorted its closure characteristics, as evidenced by the TEE findings. After repair, equalized aortic-cusp closure pressure would be expected, and this might explain the correction of the AR. These observations support the idea that substantial runoff blood flow in the immediate supra-annular region (Figs. 1 and 4) can be responsible for AR in the absence of other notable structural defects in the aortic valve. There is probably a considerable difference between the continuous ruptured-SVA runoff and the coronary runoff that occurs more cranially and possibly at a lower flow rate. Furthermore, restoring physiologic flow in this region appeared to resolve the patient’s aortic insufficiency. The residual trace AR was probably due to the initially observed subtle sclerosis of the right coronary cusp. Therefore, valve replacement (and the attendant increased risks) might not be necessary in all operations for ruptured SVA with associated aortic valve regurgitation.

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