

Aneurysms of the Sinuses of Valsalva

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Key Words

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Abstract

Sinus of Valsalva aneurysms are rare cardiac anomalies which may be acquired or congenital, most commonly involving the right or noncoronary sinuses. The congenital aneurysms are more common and often caused by weakness at the junction of the aortic media and the annulus fibrosus. Acquired aneurysms are caused by conditions affecting the aortic wall, such as infections (syphilis, bacterial endocarditis, or tuberculosis), trauma, or connective tissue disorders. Unruptured aneurysms are usually found incidentally during diagnostic studies. More commonly, sinus of Valsalva aneurysms are diagnosed after clinical sequelae of rupture. Diagnosis of sinus of Valsalva aneurysm is facilitated by echocardiography, contrast aortography, and more recently, magnetic resonance imaging. Repair is generally required for ruptured aneurysms; unruptured aneurysms encroaching on nearby structures, causing myocardial ischemia, or having the potential to rupture warrant repair. A review of the literature is presented focusing on anatomy,

clinical presentation of ruptured and unruptured aneurysms, noninvasive diagnostic modalities, and techniques for repair of this anomaly.

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Introduction

Sinus of Valsalva aneurysms (SVA) are rare cardiac anomalies. Detailed illustrations of the sinuses of Valsalva were first published in 1740, in Antonio Maria Valsalva's *Opera*, after the death of this skilled Italian anatomist and pathologist [1]. An aneurysm of the sinus of Valsalva is defined as dilatation of one of the three aortic sinuses between the aortic valve annulus and the sinotubular junction or supra-aortic ridge. The first reported case of SVA rupture, 'bursting into the right ventricle,' was by Hope [2]. One year later, Thurnam [3] described multiple examples of unruptured SVA. Recently Ring et al. [4] proposed a four-level hierarchy of nomenclature for SVA, aortic root aneurysms, aortic dissections and their repair.

SVA may be acquired or congenital. The congenital aneurysm is more common and most often caused by weakness at the junction of the aortic media and the annulus fibrosus. In acquired aneurysms, secondary degeneration of the elastic connective tissue occurs from atherosclerosis or infection. Aneurysms originate most com-

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monly in the right coronary sinus or the noncoronary sinus and, rarely, the left coronary sinus [5]. The anatomic location of the aneurysm determines the chamber into which the aneurysm may rupture. Aneurysmal ruptures into all cardiac chambers, as well as extracardiac perforations, have been described [6, 7].

Embryology

Edwards and Burchell [8] described the structural defect of a SVA as a lack of continuity between the aortic media and the aortic annulus, thereby setting the stage for subsequent avulsion and aneurysm formation. The congenital aneurysm usually begins as a blind diverticulum that originates from a localized site in one of the sinuses of Valsalva as the result of sustained high pressure in the aortic root on the weakened tissue. Although the congenital fault is present at birth, the aneurysm usually takes several years to develop as a finger-like extension with perforation frequently occurring at the apex of the aneurysm [9].

Other congenital anomalies associated with ruptured SVA, particularly aneurysms that rupture into the right ventricle, include ventricular septal defects (VSD) (30–60%), bicuspid aortic valve (10%), aortic insufficiency, pulmonary stenosis, coarctation of the aorta, atrial septal defect, and subvalvular aneurysms [10, 11]. The association between the right and noncoronary SVA and VSD has been proposed to result from incomplete fusion between the right and left distal bulbous septum in the fetus, the base of which forms the right and noncoronary sinuses of Valsalva. The resulting congenital weakness along the aortic annulus might be closely related to defects in the membranous ventricular septum [6, 12].

Epidemiology

SVA are very uncommon, with an incidence ranging from 0.1 to 3.5% of all congenital heart defects; such aneurysms account for only 0.14% of all open heart surgical procedures [7]. As the aneurysms are frequently clinically silent, their exact prevalence is unknown, however an autopsy study of 8,138 individuals suggests a prevalence of 0.09% in the general population [13]. A family history is generally lacking, although Johnson et al. described a case of two brothers with SVA that perforated into the right atrium [14]. SVA have a marked male preponder-

ance (4:1) and their incidence is higher in Asian populations [15]. The incidence of associated VSD is also higher in eastern countries. The supracristal VSD (type I) is the predominant type seen in Asian countries, whereas perimembranous VSD (type II) is often seen in western countries [16].

Congenital aneurysms consist of a localized burrowing of part of the aortic sinus into the neighboring myocardium, one of the cardiac chambers, or the free pericardium. Such aneurysms of the sinuses of Valsalva tend to be single, although, rare reports of congenital involvement of more than one sinus have been described [17].

Acquired Aneurysms

Acquired, or noncongenital, SVA occur less frequently than congenital aneurysms. An acquired aneurysm is caused by conditions affecting the aortic wall, such as infection (syphilis, bacterial/fungal endocarditis, or tuberculosis) [18, 19], degenerative disease (atherosclerosis, connective tissue disorders, or cystic medial necrosis), or thoracic trauma [20]. Howard et al. [21] reported a patient with dilatation of all three sinuses of Valsalva coexisting with cystic medial necrosis of the aortic root. Congenital weakness of the aortic annulus is implicated in the subgroup of patients with multiple aneurysms and a hereditary connective disorder, such as Ehlers-Danlos or Marfan syndrome [22, 23], resulting in diffuse enlargement of the sinuses rather than focal aneurysmal dilatation of a segment of a sinus, should be sought. There have been case reports suggesting Takayasu's arteritis [24] or Behcet's disease [25] as the cause of aneurysm formation.

Rarely, these aneurysms may be acquired iatrogenically, during aortic valve replacement (AVR) [26] or after removal of extensive aortic valve calcifications [27], with subsequent formation of a hematoma, which represents a pseudoaneurysm. Such pseudoaneurysms may dissect the interventricular septum, bulging into both the left and right ventricles [28].

Several differentiating features have been described that are frequently present in acquired versus congenital aneurysms: they are often extracardiac, may originate from any sinus, and may extend superiorly; they are usually associated with acquired heart infections and rarely with other congenital cardiac defects [29]. Aneurysms of the left sinus of Valsalva, which are the least common, are frequently acquired, especially if they rupture into the left heart [20, 26]. One explanation for extremely rare

involvement of the left coronary sinus in congenital aneurysms is that the left coronary artery is supported by the sinus wall that runs intramurally just after its origin [30].

Unruptured Aneurysms

Unruptured SVA usually remain asymptomatic and undetected. Previous reports based on necropsy and cardiac surgery findings estimated that 20% of SVA were unruptured [5]. However, with the advent of echocardiography and other noninvasive imaging modalities, unruptured SVA are diagnosed more frequently. Rarely, an aneurysm may present before rupture with a continuous murmur due to flow in and out of the intact aneurysmal pouch [31]. Exertional dyspnea, palpitations, and angina-like chest pain have been reported in patients with unruptured aneurysm of the sinus of Valsalva [32, 33], although the aneurysm may not be causally related to the symptoms.

Multiple case reports describe unruptured SVA causing significant anatomic and physiologic derangements. Significant arrhythmias may also be caused by unruptured aneurysms, including ventricular tachycardia [34], atrial fibrillation [35], and complete heart block through extension into the interventricular septum and compression of the AV node and His bundle [36].

Myocardial ischemia due to obstruction of coronary flow by a SVA has been frequently reported. The first such report, by Chipps [37], involved a 39-year-old man in whom myocardial infarction resulted from compression of the left main coronary artery by a syphilitic isolated aneurysm of the left sinus of Valsalva. A large SVA may serve as a potential site of thrombus formation. Cases of SVA presenting with thromboembolic cerebrovascular accidents or other systemic emboli have been described in the literature [38].

Ruptured Aneurysms

Although the rate of discovery has increased with the widespread availability of echocardiography, SVA are usually diagnosed in the setting of clinical sequelae of a rupture. A substantial number of ruptures develop well after puberty and between 20 and 40 years of age, with a mean age at operation of 35–39 years and a range of 13–74 years [7, 39]. A smaller number of ruptures occur in infancy or childhood [40] as well as in late adulthood. In

fact, an 80-year-old man with ruptured SVA into the right outflow tract was recently described [41].

The physiologic consequences of rupture depend upon the rapidity of the rupture, the size of the ruptured orifice, and the chamber into which rupture occurs. Ruptured SVA may present a diagnostic dilemma because of their diverse clinical presentations [42]. Two clinical patterns prevail in patients with ruptured SVA: the acute rupture of a large SVA and the gradual progression of a small perforation. Acute, large rupture may present with dramatic onset of marked substernal chest pain, upper abdominal pain or severe dyspnea with sudden hemodynamic collapse. Symptoms often follow physical stress, with acute dyspnea and chest pain developing after heavy exertion [43]. Blunt chest trauma or iatrogenic trauma during percutaneous catheterization have been reported as potential triggers for rupture [20]. Progressive dyspnea, fatigue, orthopnea, and paroxysmal nocturnal dyspnea are often seen after acute, large rupture, as there is little time for the heart to adapt to volume overload of the left and/or right side of the heart. Regardless of the location of a ruptured aneurysm, resulting in left-to-right or left-to-left shunt, volume overload of the left heart inevitably occurs. Acute symptoms of heart failure may last for hours or days, followed by progressive improvement or worsening of symptoms.

Small perforations of aneurysms present more insidiously; patients may remain asymptomatic for months or years until symptoms of congestive heart failure develop. In patients with small perforations that either do not increase or increase in size very gradually, mild dyspnea may precede symptoms of failure. These patients often present with asymptomatic continuous murmurs [44] or symptoms of infective endocarditis [45].

The clinical presentation and the hemodynamic effects of a ruptured SVA also depend on the receiving chamber. Ruptured aneurysms originate most frequently from the right coronary sinus (65–85%), less frequently from the noncoronary sinus (10–30%), and rarely from the left coronary sinus (<5%) [6, 7, 39]. The right ventricle is the most common receiving chamber (about 80–90%), due to rupture of either right or noncoronary SVA [7].

Sudden cardiac death due to ruptured or unruptured SVA may result from tamponade, myocardial ischemia, conduction disturbances and/or arrhythmias. Rupture into the pericardial space, a very rare complication (2% of noncoronary SVA ruptures), almost invariably leads to fatal cardiac tamponade [46]. Sudden death may also occur if the rupture causes compression of the ostium of the

left main coronary artery, resulting in myocardial ischemia and arrhythmic death [47]. Compression of the His bundle occurs when the ruptured SVA penetrates the base of the interventricular septum and results in atrioventricular conduction defects and arrhythmias [28, 48]. Conduction disturbances are the result of the direct pressure as well as hemorrhage or inflammation in conduction tissue by the expanding aneurysm. Choudhary et al. [49] reviewed 26 reported cases of aneurysms dissecting into interventricular septum; more than 50% of patients had congestive heart failure and complete atrioventricular block. A ruptured SVA may precipitate ventricular fibrillation because of an abrupt change in circulatory dynamics [50].

Aortic valve abnormalities and aortic insufficiency are common (25–45%) in patients with ruptured aneurysms. VSD frequently coexists in these patients and is a predisposing factor for aortic insufficiency [10]. In patients with coexisting VSD, aortic insufficiency is caused by several mechanisms: lack of supporting tissue near aortic cusps; Bernoulli effect during systole, which tends to pull the related cusp into the VSD; shunt flow through the ruptured aneurysm puts traction on the aortic cusps in diastole. Similarly, in patients without a VSD, shunt flow through the ruptured SVA may put traction on the aortic cusps in diastole resulting in aortic regurgitation [51].

Right and Noncoronary Sinus of Valsalva Aneurysms

Aneurysms arising from the right coronary sinus, which is normally adjacent to the right ventricular outflow tract, usually rupture into the right ventricle (75%), less commonly into the right atrium (19%), left atrium/ventricle (6%), and very rarely into the pulmonary trunk (less than 1% of cases) [7, 52]. Unruptured aneurysms of the right sinus of Valsalva commonly protrude into the right ventricular outflow tract and cause obstruction [53]. When this occurs, patients may present with symptoms of acute right-sided heart failure. Unruptured aneurysms of the right sinus may distort pulmonic valve leaflets causing pulmonary and tricuspid insufficiency [54], may obstruct right coronary ostia causing myocardial ischemia, or cause conduction disturbances due to compression of the interventricular septum.

Most noncoronary SVA rupture into the right atrium [7, 39]. All other possible variations, ruptures into right and left ventricles, left atrium, as well as pericardium,

have been described. Harkness et al. [55] reported higher number of SVA originating from the noncoronary sinus versus right coronary sinus, which may reflect a difference in the racial composition of western population as well as higher number of acquired aneurysms.

Left Sinus of Valsalva Aneurysms

Left SVA are commonly detected prior to their rupture [7]. Unruptured aneurysms of left sinus of Valsalva are usually asymptomatic. When they become symptomatic, they often present as an acute coronary syndrome or even cardiogenic shock by involving the origin of left coronary artery, either by direct compression or by spontaneous dissection [56]. There is more risk of myocardial ischemia with left SVA than with right or noncoronary sinus aneurysms, possibly because aneurysm of the left coronary sinus can protrude between the left atrium and pulmonary trunk and compress the trunk or branches of the left coronary artery [57].

Ruptures of left SVA typically occur into the left ventricle or left atrium [58]. Clinically these patients may present with symptoms of acute aortic regurgitation as well as coronary insufficiency and paroxysmal ventricular fibrillation. Since the right-sided cardiac chambers are not in close proximity with the left sinus, rupture of left SVA into right atrium is very rare [59]. Rare cases of rupture into pulmonary artery creating aortopulmonary tunnel [60] or into pericardium causing cardiac tamponade have been reported.

Diagnosis

Previously healthy individuals with small perforations of SVA may present with an asymptomatic continuous murmur, a hallmark of a ruptured aneurysm into the right side of the heart [57]. Hope described ‘a very loud, superficial sawing murmur prolonged continuously over the first and second heart sounds’ [2]. The classic murmur is described in 40–95% patients presenting with an aneurysm, as a loud, machinery-like, continuous murmur, varying in intensity with systole and diastole, best heard at the base of the heart [11, 14, 39]. The murmur does not peak around the second heart sound, as does the murmur of patent ductus arteriosus. The intensity of the murmur may be diminished around the second heart sound, only to increase again in diastole, creating a ‘to-and-fro’ cadence.

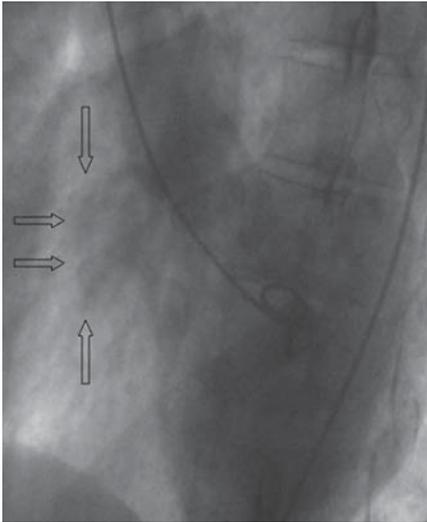


Fig. 1. Left ventriculogram (LAO cranial projection); contrast dye is visualized in the right ventricular cavity (arrows).

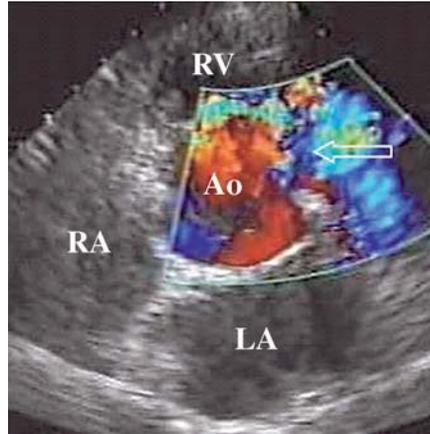


Fig. 2. Transthoracic color Doppler echocardiography in parasternal short-axis view. Ruptured right SVA into the right ventricular outflow tract is shown (arrow). Abbreviations: Ao = aorta; LA = left atrium; RA = right atrium; RV = right ventricle.

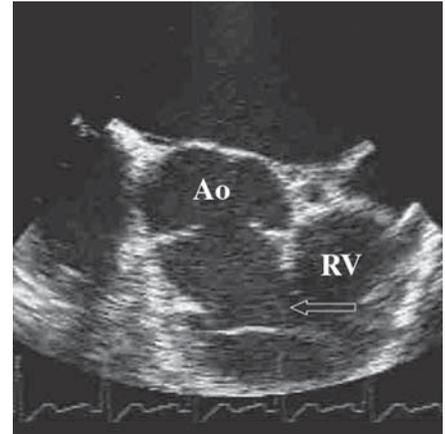


Fig. 3. TEE in short-axis view. Ruptured SVA into the right ventricular outflow tract is shown (arrow). Abbreviations: Ao = aorta; RV = right ventricle.

The electrocardiogram usually shows voltage criteria for left ventricular hypertrophy and ST-T wave abnormalities [57]. Chest X-ray may reveal a bulge to the right of the caval shadow in the postero-anterior view and appear as an excessive anterior-superior bulge in the left anterior oblique view [61].

Prior to the advent of noninvasive imaging modalities, SVA were diagnosed by cardiac catheterization. In 1967, De Bakey et al. [62] published a series of 35 cases of fistulized SVA diagnosed by aortography. Cardiac catheterization demonstrates a finger-like projection of the ruptured aneurysm into the receiving chamber ('wind-sock' image), and dye may be visualized in the receiving chamber (fig. 1). The presence of aortic insufficiency may be evaluated by contrast aortography. Right heart catheterization allows determination of pulmonary pressure and resistance and permits quantitative shunt assessment [57].

In 1974, Rothbaum et al. [63] reported echocardiographic findings of ruptured SVA and, since then, the usefulness of two-dimensional and Doppler echocardiography in the assessment of such aneurysms has been well documented [64, 65]. Two-dimensional echocardiography with color flow imaging and spectral Doppler interrogation allows accurate imaging of ruptured or unruptured SVA as well as the presence of associated abnormalities. By using multiple views, it is possible to evaluate the size and location of the aneurysmal sac. The paraster-

nal long- and short-axis views (at the level of the aortic root) are considered best to visualize aneurysms of sinuses of Valsalva (fig. 2). In ruptured aneurysms, color flow imaging with pulsed and continuous-wave Doppler demonstrates a continuous turbulent systolic and diastolic flow into the receiving chamber, most commonly the right ventricle or right atrium [66, 67]. Volume overload of the affected chambers will be detected due to increased blood flow associated with significantly sized shunts. Also, tricuspid valve fluttering and diastolic pulmonic valve opening may be seen in cases of ruptures into the right heart [68]. Detailed imaging of short-axis of the left ventricular outflow tract and aortic root helps to differentiate the aneurysm of VSD from the aneurysm of sinus of Valsalva above the aortic annulus. Imaging of parasternal short-axis will also help differentiate SVA from a coronary arteriovenous fistula, where a dilated coronary artery may be seen near origin from the aorta. Surgical repair may be performed in selected patients based on two-dimensional and Doppler echocardiography findings, without need for cardiac catheterization [69].

Transesophageal echocardiography (TEE) is highly sensitive for the identification and location of the ruptured aneurysm (fig. 3) [70, 71]. TEE may prove more helpful in delineating small ruptures as it usually allows better visualization of both the aneurysm and its rupture. Also, TEE may provide further information to differentiate SVA from the more common aneurysm of a VSD or

coronary arteriovenous fistula. TEE is invaluable in differentiating between isolated perforation of a SVA and perforation of an aneurysm in association with a VSD [72]. TEE has been shown to be more useful in assessing associated congenital lesions, but is not superior to transthoracic echocardiography or aortography in assessing the degree of aortic insufficiency or left-to-right shunting [73].

Use of contrast-enhanced helical CT scan has not been widely used thus far to diagnose SVA [74]. When the diagnosis of the ruptured SVA is uncertain, even with a combination of echocardiography and contrast aortography, magnetic resonance imaging (MRI) has been shown to be very useful. MRI, including cine MRI, may define the three-dimensional anatomy of the aneurysm more precisely, including variations of flow direction during cardiac cycle [75]. Compared with contrast aortography, MRI may also provide better evaluation of aneurysm wall thickness or presence of thrombus, and better detection of small perforations.

Currently, transthoracic color Doppler echocardiography is the initial imaging technique of choice to delineate ruptured or unruptured SVA. We recommend TEE to be the next test of choice once SVA is suspected, especially when the aneurysm is small and other congenital defects may be present. Cardiac catheterization should be used to confirm the diagnosis of the SVA, to evaluate the hemodynamic significance of the rupture, associated cardiac anomalies and, perhaps most importantly, to define coronary anatomy. We suggest further imaging with cardiac MRI and/or CT as the confirmative diagnostic tool in cases when multiple congenital abnormalities are present and better visualization of regional anatomy (as well as detection of thrombus) is needed. MRI and CT imaging, in addition to TEE, may provide surgeons with a better 3D appreciation prior to approaching the surgical repair.

Treatment

The mainstay of treatment for ruptured SVA is surgical intervention, since prognosis of patients without urgent surgical repair is very poor [76]. Early aggressive treatment is recommended for ruptured SVA in order to prevent endocarditis or enlargement of the lesion, which might necessitate more extensive repair [7]. Because of recent advances in diagnostic technique, the number of patients undergoing surgery has been increasing, including patients with unruptured aneurysms.

The first surgical repair of SVA was described independently by Morrow et al. [77] and Bigelow and Barnes [78] in the mid-1950s using hypothermia and inflow occlusion. The conventional surgical repair under cardiopulmonary bypass was first attempted by Lillehei et al. [79] and since then has been commonly used. Depending on the size and location of the aneurysm, the site of rupture, and associated cardiac anomalies, primary suture closure, patch closure, or aortic root replacement have been used in the operative repair of the aneurysm.

In early studies, the repair was approached through the cardiac chamber into which rupture occurred [80]. In the current surgical era, dual chamber exposure has been favored by most surgical centers [55, 81]. A combined approach, in which both the involved chamber and aortic root are opened, allows meticulous closure of the rupture, better evaluation of coexistent anomalies, avoidance of distortion of the aortic valve cusps, and reduction in the incidence of late aortic insufficiency [81]. In addition, aortotomy allows selective perfusion of the coronary ostia in larger patients. Several investigators have suggested that direct closure with sutures may be associated with a higher incidence of aneurysm recurrence [55, 81, 82]. Many centers have been using patch closure for all SVA repairs, regardless of size, to avoid tension on the annulus, especially in complex cases.

Surgical repair is highly successful with low operative risk. The life expectancy of patients who have successful repair of a ruptured aneurysm approximates that of the healthy population (10 year survival rates of 90–95%) [39, 81], although survival rates may be worse in predominantly western patient populations [55]. In one of the largest series involving 129 patients, the perioperative mortality was 3.9% and was associated with presence of preexisting sepsis and endocarditis [7]. In cases with severe infection, complex procedures such as aortic root replacement or Ross procedure may be necessary. Late complications were due to prosthetic valve malfunction, prosthetic valve endocarditis, aneurysm recurrence, and anticoagulation-related bleeding. The risk for recurrent fistula or VSD is minimal in the current surgical era.

Aortic regurgitation frequently complicates the pathology and presentation of patients with SVA. The incidence of aortic regurgitation has been reported between 30 and 50%, with rates of AVR between 50 and 80% patients [55, 81]. In cases of moderate to severe aortic regurgitation, retracted, thickened, or bicuspid aortic valve leaflets, AVR is usually required. Patients with aortic regurgitation have not been shown to have poor outcome, however, patients with a SVA coexisting with a VSD plus

associated aortic regurgitation appear to have a worse prognosis [83]. Early postoperative aortic insufficiency at discharge from hospital is a predictor of long-term aortic insufficiency [84]. Patients with Marfans syndrome and SVA may have worse long-term outcome after repair [7]. Diagnosis of chronic connective tissue disorders should be investigated prior to surgery, as aortic root replacement may be needed in such patients [85].

With the improvement in cardiac imaging and availability of various percutaneous catheter devices, nonsurgical closure of various intracardiac defects has become a new alternative to open surgical repair. Percutaneous closure of congenital ruptured aneurysm was first described by Cullen et al. [86]. Arora et al. [87] described eight patients who underwent percutaneous repair. One patient required surgical repair due to intermittent hemolysis, one patient died of progressive congestive heart failure, while other six patients were asymptomatic at 2–96 months following repair. Since 1994, multiple case reports have been published utilizing the available percutaneous closure devices, demonstrating their feasibility and effectiveness, especially with the Amplatzer device [88]. As percutaneous catheter closure is a new procedure, long-term follow-up data and management will need to be established.

The indications for repair of unruptured SVA are controversial as reports of these patients are limited. In the presence of complications such as right ventricular outflow tract obstruction, arrhythmias or infection, surgical repair is indicated [34, 36, 53]. It is generally recommended to repair large SVA to avoid later complications of rupture, myocardial ischemia, and right ventricular outflow tract obstruction. The natural history of small aneurysms of sinuses of Valsalva is unclear. One case report

described a patient with an unruptured aneurysm being followed for 13 years without surgery [89]. Other investigators reported a case with rapid expansion of the aneurysm which led to left coronary artery compression and death from a massive myocardial infarction [90]. Criteria for surgical intervention in asymptomatic unruptured aneurysms await definition, however, progressive enlargement of an aneurysm on serial evaluations should be considered as an indication for repair.

Conclusions

SVA are rare cardiac anomalies which may be acquired or congenital, most commonly involving the right or noncoronary sinuses. The congenital aneurysms are more common and often caused by deficiency of normal elastic tissue, resulting in weakness at the junction of the aortic media and the annulus fibrosus. The need for invasive imaging has been reduced by transthoracic and TEE echocardiography that allow to determine: (1) sinus of origin, (2) size of the aneurysm, (3) which chamber the aneurysm ruptures into, (4) degree of aortic valve regurgitation, and (5) associated congenital anomalies. Repair is generally required for ruptured aneurysms; unruptured aneurysms invading nearby structures, causing myocardial ischemia, conduction disturbances, or having the potential to rupture warrant repair. It is suggested that repair of ruptured SVA should be performed with the use of patch repair, through an aortotomy with or without cardiotomy. An early surgical repair of SVA can prevent potentially life-threatening complications, and life expectancy after adequate repair approximates that of the healthy population.

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