



**UNRUPTURED ANEURYSM OF RIGHT SINUS OF VALSALVA ERODING  
INTERVENTRICULAR SEPTUM MANIFESTING AS PERMANENT COMPLETE  
HEART BLOCK.**

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**ABSTRACT**

We present a patient with bilateral unruptured sinus of Valsalva aneurysms (SVA) involving both the left and right coronary sinuses. The large right sinus aneurysm eroding into the interventricular septum and almost destroying it completely producing a permanent complete heart block. The small left sinus aneurysm was very small causing no mass effects.

**KEYWORDS:** complete heart block; aneurysm of sinus of valsalva; interventricular septum; pacemaker; cardiopulmonary bypass.

**CASE REPORT**

A 57-year-old male patient presented with complaints of recurrent episodes of dizzy spells for 6months and two episodes of syncope on the day of admission, suggestive of Stokes-Adams attacks. The patient also had NYHA grade 3 dyspnoea for 3 months. There was no significant past medical illness. On admission, he was experiencing bradycardia with a heart rate of 45 beats/min. Blood pressure was 110/66 mm Hg in the right upper limb in a supine posture. Auscultation revealed a short early diastolic murmur in the aortic area. The rest of the examination was unremarkable. Resting ECG confirmed complete AV block with a slow regular broad complex ventricular escape rhythm (Figure 1). Temporary transvenous pacing was performed by right femoral vein puncture. The patient did not spontaneously reverted to sinus rhythm. Transthoracic echocardiography revealed mild aortic regurgitation. There were two aneurysms (Figure 2) arising from the right and left aortic sinuses. The right sinus aneurysm was large and penetrating into the interventricular septum and the left sinus aneurysm was smaller (video 1 and Figure 2). Computerised Tomogram demonstrated the presence of two unruptured sinus of Valsalva aneurysms (SVAs) arising from the right and left coronary sinuses (Figure 3). The right coronary sinus aneurysm was large burrowing into the interventricular septum. Conventional coronary angiogram showed normal coronaries and left ventricular angiogram showed a dilated left ventricle with normal function. The patient was operated and repair of the right sinus aneurysm with pericardial patch closure done

(figure 4 and 5) under full cardiopulmonary bypass. The patient made an uncomplicated recovery and remains well. Intra-operatively interventricular septum was found to be moth eaten appearance and near completely destroyed so permanent pacemaker was inserted later. The presence of bilateral unruptured SVAs involving left and right sinuses, manifesting as a permanent complete heart block, makes this a very unusual presentation of an uncommon disease.

**LEGENDS TO FIGURES**

**Figure 1-** Twelve-lead electrocardiogram at presentation showing complete heart block with an irregular ventricular escape rhythm of right bundle branch block morphology with a heart rate of 45 beats/min.

**Figure 2-** Transthoracic Echo PLAX view showing unruptured right sinus of Valsalva aneurysm penetrating into interventricular septum.

**Figure 3-** Aneurysm from right coronary sinus measuring 6.9 x 4 cm extending into interventricular septum with no evidence of leakage and any chamber connection. Small 1x 0.8 cm similar aneurysm from left coronary sinus is also seen.

**Figure 4 –** Intraoperative view showing tricuspid aortic valves and aneurysm of right coronary sinus

**Figure 5 –** pericardial patch closure of mouth of aneurysm.

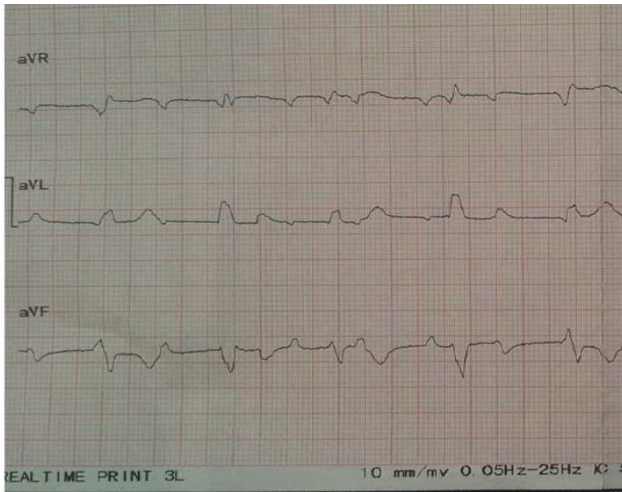


Figure 1

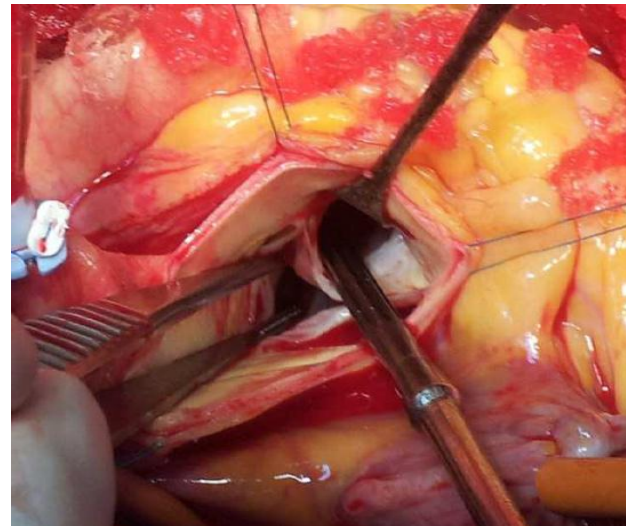


Figure 4



Figure 2

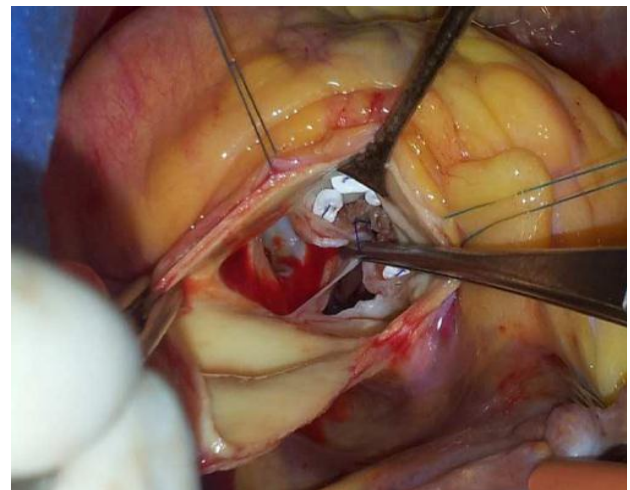


Figure 5



Figure 3

## DISCUSSION

SVA is a rare congenital anomaly caused by a defect of continuity between the aortic media and the annulus fibrosis.<sup>[1]</sup> SVA was first described by Hope in 1837. Although the true incidence is unknown, unruptured SVAs are rarely encountered clinically. Among patients undergoing cardiopulmonary bypass, the incidence of SVA has been reported to be 0.1% to 1.5%.<sup>[1]</sup> The anomaly is more common in the Oriental population compared to Caucasians. SVAs are most frequently located in the right coronary sinus (67% to 77%) with 15% originating in the non-coronary sinus and rare reports of isolated left SVAs.<sup>[3]</sup>

The majority of cases of SVA are congenital. Involvement of multiple coronary sinuses is rare and often associated with a secondary etiology.<sup>[4]</sup> Secondary causes of SVA include atherosclerotic disease, Marfan's syndrome, syphilis, bacterial endocarditis, and cystic medial necrosis trauma (deceleration injury). Rare association with polycystic kidney, Behcet's disease and tuberculosis has also been reported.<sup>[1]</sup> A review of the literature revealed very rare case reports of SVAs arising

from the right and left sinuses of Valsalva<sup>[4]</sup> and triple aneurysms arising from all three sinuses<sup>[5]</sup>

The most common clinical manifestation of SVA is rupture into the right ventricle, although rupture into the right atrium, left ventricle, interventricular septum, pleural space, pulmonary artery and pericardium can also occur.<sup>[1]</sup>

The pattern of presentation does, however, vary widely and unruptured aneurysms are often 'silent'. Unruptured ASOV can cause significant anatomic and physiological derangement and may present with evidence of right ventricular outflow tract obstruction, aortic insufficiency, conduction abnormalities or coronary artery compression.<sup>[6]</sup> Dissection of the interventricular septum (IVS) is a rare complication of ASOV but. Rare manifestations include cerebrovascular accidents, myocardial ischemia, mitral incompetence, right ventricular outflow tract obstruction, left ventricular outflow tract obstruction and atrial fibrillation.<sup>[7]</sup>

Cardiac conduction disturbance due to SVA can occur at several levels<sup>[8-9]</sup> including sinoatrial conduction disruption and various levels of his bundle block. The mechanism for septal extension has been ascribed to simple progression, intraseptal rupture with haematoma and subsequent pseudo aneurysm and bacterial endocarditis. The presence of atrioventricular block, however, does not necessarily mean septal extension. In the most frequent type of aneurysm of the sinus of Valsalva (right coronary sinus) the proximity of the conduction tissue may lead to conduction disturbances. Transient atrioventricular block and persistent complete atrioventricular block are also reported.

Before the advent of modern diagnostic techniques, the diagnosis of unruptured sinus of Valsalva aneurysm was rarely made on a living patient, and most of the reports<sup>[10]</sup> were based on autopsy or on cases found serendipitously at cardiac surgery. Diagnosis of unruptured ASOV has evolved from post-mortem findings in 1956 by Lee *et al.*, to preoperative diagnosis.<sup>[11]</sup> The first preoperative diagnosis of this condition was reported by Engel *et al.*, in 1980. Lewis and Agathangelou demonstrated on echocardiography, the continuity between the echo free space with the upper septum and the aortic root at the level of the right coronary sinus.<sup>[12]</sup>

Although the implications and natural history of an unruptured ASOV are not clear, surgery forms the only definitive treatment and has prophylactic value in preventing complications such as AR and development of conduction defects. Conservative treatment is possible if AR is absent or mild and if the aneurysm is not increasing further. During surgery, resection of the pouch is unnecessary and may damage the aortic valve and IVS. Closure of the mouth of aneurysm with either direct sutures or with a patch is a simple and effective

treatment when operated upon early. Aortic valve repair or replacement is necessary when operated at a late stage. Surgical treatment is required in nearly all cases with the long term results being favourable; one large series showed a 95% 20 year survival.<sup>[13]</sup>

In our patient, the unruptured right SVA had extended into the interventricular septum and destruction by mass effect on conduction tissue had compromised normal atrioventricular (A-V) node/His bundle function resulting in complete heart block of permanent type.

## CONCLUSION

We conclude from this case and a review of the literature that the presence of bilateral unruptured SVAs involving the right and left sinuses is rare. Moreover, presentation as a permanent complete heart block is very rare and merits reporting.

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