

## Ruptured Sinus of Valsalva Masquerading as Rheumatic Heart Disease

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Ruptured sinus of Valsalva is a rare lesion in pediatric age group. We are reporting a twelve year old child with hyperdynamic circulation being diagnosed and treated as Rheumatic heart disease.

**Key words:** *Hyperdynamic Circulation, Rheumatic heart disease, Ruptured sinus of Valsalva.*

**R**uptured sinus of Valsalva (RSOV) is a rare lesion that usually originates in the right or noncoronary aortic sinus and communicates with a cardiac chamber, frequently right sided, producing an aorto-cardiac fistula. The presentation may range from an asymptomatic murmur to acute cardiogenic shock and death. The mean age for the onset of symptoms owing to sudden rupture of the aneurysms is 31 year. RSOV is extremely rare in pediatric age group [1]. We report a child with ruptured sinus of Valsalva who was symptomatic for 6 years before diagnosis.

### CASE REPORT

A 12-year-old male child was referred to us with a diagnosis of Rheumatic heart disease with refractory congestive heart failure. His chief complaints were undue awareness of heart beat, and breathlessness on physical exertion since the age of 6 years, which was gradually progressive. Two months before presentation, his condition had suddenly worsened and now he was dyspneic even at rest and had developed generalized body swelling. On clinical examination, he was markedly breathless at rest, had head bobbing, with a regular large volume collapsing pulse of 120 bpm. Blood pressure was 130/40 mmHg in both arms and 210/40 in both lower limbs. A 4/6 continuous murmur with maximum intensity in systole at the left sternal border was present. Bilateral basal creptitations with significant expiratory wheeze was audible. Liver was palpable 7 cm below costal margin in the midclavicular line. Investigations revealed a hemoglobin of 9.5 g/dL, leucocytosis with polymorphonuclear cell dominance, ESR-20mm, CRP was negative and liver and renal function tests were within normal limits. On X-ray chest there was cardiomegaly with pulmonary plethora. ECG showed

sinus rhythm with normal axis, P pulmonale and biventricular hypertrophy. Transthoracic echocardiography (TEE) showed right atrial and right ventricular enlargement, perimembranous VSD measuring 5mm with prolapse of aortic valve cusp, dilated ruptured right sinus of Valsalva into right ventricle, and enlarged LV with preserved systolic function. Aortic regurgitation and left to right shunt from right sinus of Valsalva to right ventricle was evident on color flow examination. Doppler study showed continuous flow at RSOV. He was managed as acute left ventricular failure with diuretics, digitalis, humidified oxygen and calcium channel blockers. Patient showed improvement but still had dyspnoea (NYHA grade 3). After stabilization, he was referred for surgical management.

### DISCUSSION

Aneurysms of the sinus of Valsalva account for only 1% of congenital cardiac anomalies. Of these aneurysms, 70% arise from the right sinus of Valsalva. Most of the remainder arises from the noncoronary sinus, and, 5% from the left coronary sinus. [2]. It is produced by mural deficiencies of an aortic sinus that perforate into a cardiac chamber resulting in an aorto-cardiac fistula. The fistula develops either as a “windsock” deformity or simple fistulous connection due to the venturi effect produced by VSD flow. Clinical presentation is usually within the third decade of life. There are few case reports in children but presentation can range from infancy to seventh decade [2-4]. Our case was asymptomatic till six years of age when he started developing gradually increasing breathlessness on physical exertion but suddenly deteriorated at twelve years of age when he developed severe respiratory distress with signs of hyperkinetic circulation. The patient was diagnosed as a case of

rheumatic fever with severe aortic regurgitation. Gomez, *et al.* [5] reported a similar case of ruptured aneurysm of the sinus of Valsalva in a 12-year-old boy diagnosed with ventricular septal defect in the neonatal period [6]. Dattilo, *et al.* [6] reported a chance finding of an aneurysm of the right sinus of Valsalva in an 11-year-old Italian child with a ventricular septal defect and a pericardial effusion. Diwedi, *et al.* [7] reported an extremely rare case of a 6-year-old child with left Sinus of Valsalva aneurysm opening in right atrium [7].

The lesion is five times more common in Asians [2-4]. Various reports indicate a male preponderance in Asians [2-4]. Upon presentation, approximately 80% to 85% of patients are symptomatic with dyspnea, pain, palpitations, or fatigue. Majority will have gradual onset, while 1/4<sup>th</sup> will have acute onset of their symptoms [2-4]. The right coronary sinus is most commonly affected as in our case, followed by the noncoronary sinus. Rupture of the aneurysm most commonly occurs into the right ventricle followed by the right atrium but it may also rupture into the left ventricle, the interventricular septum and the pericardial space[8].

Associated lesions are common in patients with congenital, ruptured sinus of Valsalva aneurysm [2]. Aortic regurgitation occurs in 30% to 75% of patients, and ventricular septal defects, either subarterial or perimembranous, occur with a comparable incidence of 30% to 50% [2]. In the present case the gradual onset of symptoms at six years of age may be due to slow but progressive development of aortic regurgitation which may be on account of increasing prolapse of aortic cusp in the perimembranous VSD or the increasing size of aneurysm producing valvular dysfunction. The marked systolic BP discrepancy between the upper limb and lower limb can be explained by Hill's sign and indicates severe regurgitation in this case [9].

The gold standard for diagnosis of this lesion has traditionally been cardiac catheterization and aortography. With the advent of newer generation ultrasound machines, echocardiography has taken centre-stage for diagnostic confirmation. In the vast majority of cases, it can totally supplant the need for angiography [10]. Surgery should be done as soon as rupture of sinus of Valsalva aneurysm is diagnosed because without surgery most cases will die of intractable congestive heart failure.

A clinician should always ponder on the unusual etiology of a common aortic valvular lesion like acute aortic regurgitation; in this case report, the etiology was a rupture of the right sinus of Valsalva aneurysm into the right ventricle.

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