

# Right Sinus of Valsalva Fistula to the Right Ventricle as a Cause of Severe Heart Failure

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## Abstract

Fistula arising from ruptured sinus of Valsalva aneurysm is an uncommon cause of congestive heart failure, and it is even rarer in the absence of an aneurysm. We present the case of a female patient with acute idiopathic rupture of the right sinus of Valsalva to the right ventricle and atrium in the absence of typical aneurysm. This anomaly resulted in a left-to-right shunt leading to rapidly progressive heart failure. [*Indian J Chest Dis Allied Sci* 2018;60:35-36]

**Key words:** Sinus of Valsalva, Fistula, Rupture, Heart failure.

## Introduction

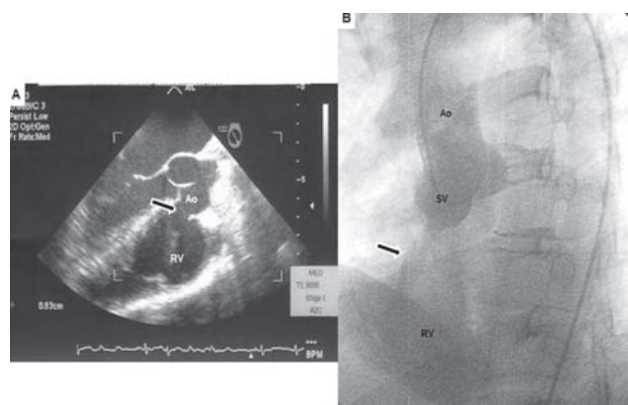
Fistula arising from ruptured sinus of Valsalva aneurysm (SVA) is an uncommon cause of congestive heart failure and it is even rarer in the absence of aneurysm. Sinus of Valsalva fistulas (SVFs) are an uncommon congenital or acquired cardiac anomaly.<sup>1</sup> Sinus of Valsalva (SV) anomalies were first described in 1840 in the case of a 33-year-old man with a sudden onset of a “breaking” feeling in the heart, accompanied by syncope, palpitations, dyspnoea and death within 11 weeks.<sup>2</sup> This condition is more common in males from Asia and Eastern Europe.<sup>3</sup> We present here a case of a female patient with acute idiopathic rupture of the right SV to the right ventricle and atrium in the absence of typical aneurysm. This anomaly resulted in a left-to-right shunt leading to rapidly progressive heart failure.

## Case Report

A 35-year-old female patient with a history of pathological hypothyroidism under endocrinology follow-up and dyspnoea of functional class II the long time came to us. Two weeks before hospitalisation, the patient claimed to have sudden onset of New York Heart Association (NYHA) functional class III-IV dyspnoea, angina pectoris, and palpitations that a few days later progressed to lower limb oedema. At the time of hospitalisation, she showed signs of right congestive heart failure with 2/3 jugular ingurgitation, painful hepatomegaly, and 3/6 lower limb oedema. Vital signs showed a heart rate of 115 beats per minute (bpm) and a blood pressure of 100/70 mmHg. Heart auscultation revealed increased second heart sound, right R3, and continuous grade 4/6 murmur audible in the left parasternal region, third intercostal space and mesocardium, with no preferential irradiation. The electrocardiogram showed sinus tachycardia. Laboratory tests were within normal ranges. Chest

radiograph revealed a normal cardiothoracic ratio and increased prominence of the main pulmonary artery and mildly increased pulmonary vascularity. The transthoracic echocardiogram showed a high-speed left-to-right shunt in the upper part of the interventricular septum, under the aortic valve. Measurement between pulmonary and systemic flows (QP/QS) was 2.4/1, with normal right cavities and preserved left ventricle systolic function. As a result of these findings, the assessment was completed with a transesophageal echocardiography (TEE), that revealed a left-to-right shunt between the right SV and the under-surface of the right ventricle, with an 8-mm diameter break in continuity into the implantation site of the tricuspid septal valve (Figure 1A).

Cardiac catheterisation showed contrast passage from the aorta to the right ventricle with a QP/QS of 2.2/1 (Figure1B).



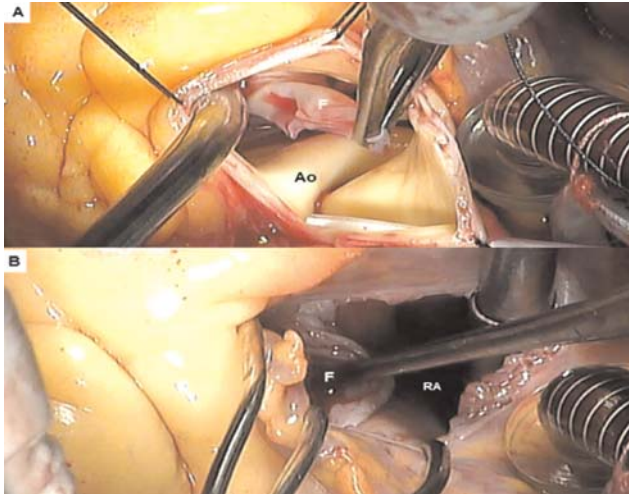
**Figure 1.** (A) Transesophageal echocardiography (TEE) showing left-to-right shunt between the right SV and the under-surface of the right ventricle (arrow) and (B) cardiac catheterisation evidenced the contrast passage from the aorta to the right ventricle (arrow).

**Definition of abbreviations:** Ao=Aorta; RV=Right ventricle; SV=Sinus of Valsalva.

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After being diagnosed with SVF to the right ventricle, the patient had surgery. Surgery showed an 8-mm hole in the right coronary sinus leading to the right cavities (the fistulous tract straddled the line between the right ventricle and atrium) (Figure 2 A,B). Simple closure was performed on the hole (intra-operative TEE without shunt), with uncomplicated progress at the time of discharge.



**Figure 2.** Photos of the surgery showing an 8-mm hole in the coronary sinus leading to the right cavities, (A) View from the aorta and (B) view from the right cavities.

*Definition of abbreviations:* Ao=Aorta; RA=Right atrium; F=Fistula.

## Discussion

Sinus of Valsalva fistula is a very rare condition, with an incidence of about 0.1% to 3.5% of all congenital heart malformations.<sup>4</sup> Anatomically, SV anomalies are classified as: (1) aneurysm; (2) aneurysm with fistula (rupture), and (3) fistula.<sup>5</sup> Most congenital SV anomalies affects the right SV and then non-coronary sinus; those in the left SV are usually acquired<sup>5</sup> and can result from trauma, endocarditis, syphilis, Behcet disease, Marfan syndrome, and senile dilation<sup>6</sup>; 76% of SVF are caused by SVA, and few cases of SVF without aneurysm have been reported, as in our case.

Aetiopathogenesis is simple in the absence of other SV disorders.<sup>7</sup> In contrast, SV are related to a cardiac loop formation defect during embryonic development associated with long-lasting high pressure periods typical of the aorta.<sup>8</sup> The left coronary cusp is not derived from bulbar septum, and therefore, congenital left SV aneurysms do not occur.<sup>9</sup> The SV are 3 small dilations on the wall of the aorta just above the aortic valves. The anatomical arrangement of the aortic sinus of Valsalva with respect to heart chambers may explains the haemodynamic changes arising from a ruptured sinus.<sup>10</sup>

The gold standard in imaging diagnosis is angiography, even though in 90% of cases initial diagnosis is provided by the TEE; cardiac magnetic resonance and multi-slice computed tomography can also be useful.<sup>3</sup>

Sinus of Valsalva rupture needs to be surgically corrected.<sup>11</sup> Survival in patients with non-surgical ruptured SV is 3.9 years and the post-operative survival rate after 10 and 20 years is of about 90%. Therefore, surgery is mandatory in these cases.<sup>12</sup> Surgical options include aortotomy, opening of heart cavities or a combination, as well as simple closure of the fistulous hole or use of patches depending on the size of the defect. Defects larger than 0.8 cm should be closed with patches.<sup>9</sup> In our case, the patient had an 8-mm defect; thus, simple closure was done, with uncomplicated progress and no immediate or remote post-operative complications.

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