

# An Unusual Case of Acute Onset of Cardiac Failure

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## ABSTRACT

**Rupture of the aortic sinus of Valsalva is one of the rare causes of acute dyspnoea. Modern advances have enabled diagnosis to be made easily with echocardiography as illustrated in this case. The treatment of choice is surgery with excellent prognosis if detected early. Therefore a high index of suspicion is required to diagnose this potentially threatening but yet treatable condition.**

**Keywords: aneurysm, continuous murmur, echocardiography, rupture, sinus of Valsalva, ventricular septal defect**

## INTRODUCTION

Acute dyspnoea is a common presentation in our daily clinical practice. It may be the initial presentation of several conditions, including sudden cardiopulmonary events, acute decompensation of chronic cardiopulmonary diseases, or a compensatory respiratory response to acute metabolic events. In many cases, the diagnosis may not be apparent on initial assessment. We present a case in which the cause of acute breathlessness was unusual.

## CASE REPORT

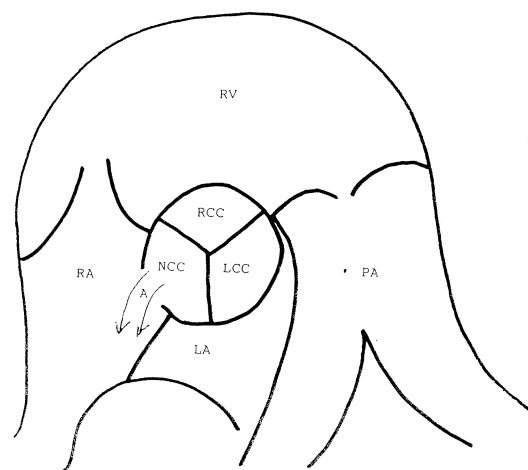
A 43-year-old man was hospitalised for a sudden onset of cardiac failure during his visit to China for 11 days. He remained symptomatic upon his return with complaints of orthopnoea and exertional dyspnoea. On admission, the patient remained stable and was not in overt cardiac failure. A continuous murmur was detected at the left sternal edge. An echocardiogram done showed ruptured aortic aneurysm of sinus Valsalva (ASOV) to the right atrium with mild aortic regurgitation (Fig 1). This was confirmed by cardiac catheterisation which showed ruptured sinus into the right atrium with left-to-right shunt QP:QS 3.8:1 and a step-up in oxygen saturation in the right atrium. Subsequently, the patient underwent surgery. At surgery, a non-coronary sinus of Valsalva aneurysm measuring 1 cm in diameter was found tunneling through the right ventricle and opening into the right atrium via the annulus of tricuspid valve. The defect was closed surgically with dacron patch. Follow-up of the patient with serial

echocardiography at 4 months and 16 months post-surgery showed trivial aortic regurgitation with no subsequent complications.

## DISCUSSION

ASOVs are rare congenital anomalies accounting for 0.3%-3.5% of all congenital heart disease<sup>(1-3)</sup>. This condition comprises of a thin-walled, tubular outpouching, located most frequently at the right coronary sinus or the adjacent half of the non-coronary sinus. It has a higher preponderance in the Oriental male population<sup>(4-6)</sup>. Prior to rupture, patients with ASOV are usually asymptomatic. Rupture of the ASOV is uncommon during infancy, and majority of patients present between 20 and 40 years of age<sup>(7)</sup>.

Most of the ASOVs are congenital. The aetiology is due to failure of fusion between the aortic media and the heart at the level of annulus fibrosus of the aortic valve<sup>(8)</sup>. Less commonly, it may be acquired from bacterial endocarditis or syphilis<sup>(9)</sup>. Rupture may occur spontaneously or following physical exertion, road traffic accidents



**Fig 1** - Schematic diagram of a transthoracic echocardiographic parasternal short-axis view of the ruptured aneurysm sinus of Valsalva. Arrows indicate flow from the site of rupture at the non-coronary cusp to the right atrium.

RCC	right coronary cusp
LCC	left coronary cusp
NCC	non-coronary cusp
RA	right atrium
LA	left atrium
RV	right ventricle
PA	pulmonary artery

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and cardiac catheterisation. Most of the aneurysms involve the right coronary sinus (67%), with the remaining ones occurring in the non-coronary sinus (25%) and left coronary sinus (8%)<sup>(10)</sup>.

Sakakibara and Konno classified the lesion into 4 main categories, based on the location of the aneurysm and fistula (Table I)<sup>(11)</sup>. The sites of rupture involve the right ventricle (57%), right atrium (35%), left ventricle (4%), and left atrium (2%)<sup>(10)</sup>. Interestingly, fistula between the right coronary sinus and right ventricle occurred more frequently among Orientals. Whereas, among Caucasians, communication between the right coronary sinus and right atrium is more common<sup>(12)</sup>.

Approximately 55% of these patients may be associated with ventricular septal defect (VSD), especially in children<sup>(13-15)</sup>. Of note, subarterial VSD occurs mainly in Orientals and perimembranous type in Caucasians<sup>(7)</sup>. Valvular lesions, such as aortic regurgitation, pulmonary and aortic stenosis, are other common associations. Rarely, the condition

It is evident now that echocardiography as well as cardiac catheterisation<sup>(16)</sup> can accurately diagnose ASOV. It is able to delineate the sinus involved, the chamber of exit for the fistula, the magnitude and associated anomalies, and any evidence of endocarditis vegetations. Doppler measurement can further compute the right ventricular systolic pressure with precision. In cases whereby the aneurysm arises from the right coronary sinus and ruptures into the right ventricular outflow tract, pulmonary stenosis may be missed during echocardiography. The loud continuous signal at the same location caused by the rupture of aneurysms makes doppler identification of a separate gradient of pulmonary stenosis difficult. Similarly, a small VSD can be missed in the presence of continuous signal from the right ventricular outflow tract<sup>(16,17)</sup>. Colour flow imaging may be particularly helpful in these cases. Where transthoracic echocardiography provides inadequate visualisation of important structures, transesophageal echocardiography often provides additional useful information.

The treatment of choice in rupture of ASOV is surgery. However, late aortic regurgitation still presents a problem especially in those with right ASOV to right ventricle fistula with associated subarterial VSD. The long-term prognosis after surgery for ruptured ASOV, especially when detected early, is excellent, with minimal risk of recurrence. Life expectancy after repair approaches that of normal population<sup>(7)</sup>. Thus, a high index of suspicion is necessary to diagnose this serious but treatable condition.

**Table I - Classification of congenital aneurysm of sinus of Valsalva\***

Type	Site	Fistula	A/w VSD
I	Left portion of right sinus	Right ventricular outflow tract	Frequent
II	Mid portion right coronary cusp	Right ventricle	Uncommon
III	Posterior portion right coronary cusp	Right atrium, sometimes right ventricle	Uncommon
IV	Right portion non coronary cusp	Right atrium	Uncommon

A/w VSD associated with ventricular septal defect.

\* Adapted with permission from Sakakibara and Konno

may be associated with coarctation of aorta, patent ductus arteriosus, tricuspid incompetence, atrial septal defect and tetralogy of Fallot. Among Orientals, there is a lower association rate with cardiac anomalies other than VSD<sup>(6)</sup>.

Following rupture, acute symptoms inadvertently occur in 40% of patients<sup>(7)</sup>. These include fatigue, exertional dyspnoea, palpitations, angina and syncope. Ruptured ASOV often produces a continuous murmur, as was the case in our patient. Differential diagnosis include patent ductus arteriosus, coronary arteriovenous fistula or VSD with aortic regurgitation. Symptoms occur less frequently when subarterial VSD is present. In contrast, symptoms are more frequent if aortic regurgitation is present<sup>(8)</sup>. Rapid progression of cardiac failure requires immediate treatment. Clinical suspicion should be high and with the availability of echocardiography, ASOV can be readily detected and treated promptly.

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