# **Ruptured Sinus of Valsalva Aneurysm Initially Misdiagnosed as Ventricular Septal Defect by Echocardiography**

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Aneurysms of sinus of valsalva are rare cardiac lesions and most of them are in congenital origin. The malformation consists of a separation or lack of fusion between the media of the aorta and the annulus fibrosis of the aortic valve. The structure becomes aneurysmal and may rupture to form a fistula. We present a case of ruptured sinus of valasalva aneurysm in a 25-year-old man. The diagnosis was made by echocardiography and confirmed at operation.

## Introduction

Ruptured sinus of valsalva aneurysm (RSVA) is a rare entity with a varied clinical presentation. Because of rapid hemodynamic deterioration, early diagnosis is mandatory and often lifesaving. It can be congenital or acquired and is sometimes associated with other cardiac lesions such as ventricular septal defect (VSD), membranous subaortic stenosis, aortc regurgitation, bicuspid aortic valve and coarctation.<sup>1,2</sup> Echocardiography is a powerful diagnostic tool for detection of the lesion.<sup>1</sup> We present a case of isolated ruptured sinus of valsalva aneurysm in a 25- year- old man which was initially misdiagnosed as VSD by echocardiography.

## **Case report**

A 25-year-old man was referred to our echocardiography laboratory for transesophageal study. Few days ago he was seen by a general cardiologist because of his recent onset, progressive dyspnea and was suspected of

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Cardiovascular Research Center, Shahid Faghihi Hospital, Zand Blvd., Shiraz, Iran. Tel/Fax: +98-711-2343529 E-mail: grezaian@gmail.com having VSD by transthaoracic echocardiography. Systolic trill and a grade IV/ VI continuous murmur were detected along the left lower sternal border during the cardiac examination. There were no pulmonic rales or sign of peripheral edema. Electrocardiogram was normal. A chest radiograph demonstrated a slightly enlarged cardiac silhouette and increased pulmonary arterial flow. Transthoracic and transesophageal echocardiographies showed mild left ventricular dilatation, LVEF =55% and a discontinuity in the wall of right coronary sinus. Color-flow Doppler revealed a continuous turbulent flow accentuated in diastole between the right sinus of valsalva and the right ventricular outflow tract (Fig. 1 A-D). The diagnosis was confirmed in the operating room where he was successfully operated upon.

## Discussion

Ruptured sinus of valsalva aneurysm is an uncommon condition with protean clinical manifestation. Rupture is often with an abrupt onset and the presentation may range from an asymptomatic murmur to acute cardiogenic shock and death. The right coronary sinus is



Figure1. Illustration of various echocardiographic features of RSVA.

- **A**. Transthoracic echocardiography demonstrating a ruptured aneurysm. **B**. Color flow image showing blood flow from the aorta to RVOT.
- C. Continuous wave Doppler image showing continuous flow
- accentuated in diastole.

**D.** Transesophageal echocardiography showing a ruptured aneurysm.

most common site (approximately 73%) and the noncoronary sinus being the second most common site (approximately 20%). In 6%, the left coronary sinus is involved.<sup>3</sup> The receiving chamber of a right aortic sinus aortocardiac fistula is usually the right ventricle but noncoronary sinus aortocardiac fistula drains into right atrium.<sup>4</sup> Althogh transesophageal echocardiography is the most reliable method to confirm the diagnosis, the following pitfalls must be taken into account:

 A few patients are misdiagnosed as having isolated ventricular septal defect as in our patient. In this situation, the characteristic Doppler spectrum showing continuous high jet velocity accentuated in diastole as opposed to a high velocity systolic and low velocity diastolic flow which differentiates the two conditions.

- 2 If both conditions coexist, then the ventricular septal defect jet may be hidden in the ruptured sinus of valsalva aneurysm jet and coexisting ventricular septal defect could be missed.
- 3 Coronary arteriovenous fistula with continuous color flow between the aorta and right ventricle or the right atrium may be misdiagnosedasrupturedsinusofvalsalvaaneurysm.

In conclusion although transthoracic echocardiography can initially be used as an screening test, transesophageal echocardiography is mandatory for a more definitive and accurate diagnosis of <sup>5</sup> ruptured sinus of valsalva aneurysm.

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