

# Undiagnosed Sinus Venosus Atrial Septal Defect Giving Rise to Eisenmenger Syndrome: A Case Report

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## Abstract

Atrial septal defects are communications of variable size that occur in the heart and allow interatrial shunting. We report a 49-year-old lady with sinus venosus type ASD, diagnosed by trans-esophageal echocardiography.

**Keywords:** Sinus Venosus Atrial Septal Defect; Eisenmenger Syndrome; Iran

## Introduction

The defects of inter-atrial septum are classified according to their embryonic origin into ostium secundum, ostium primum, sinus venosus (SVC and IVC form) and coronary sinus types.<sup>1,2</sup> Atrial septal defects (ASDs) are common, accounting for 6-10% of all cardiac anomalies encountered and 4 in every 100,000 people.<sup>3</sup> Isolated secundum ASDs are the most common type which account for approximately 70% of all ASDs, and are often easily diagnosed on Trans-Thoracic-Echocardiography (TTE).<sup>3</sup>

However, sinus venous defects vary in that the atrial septum is intact except in the superior portion, the most one adjacent to the superior vena cava and may co-exist with partial anomalous pulmonary venous connections. Diagnosis is very difficult on trans-thoracic echocardiography while trans-esophageal echocardiography (TEE) may contribute to the diagnosis of sinus venous defects and assessment of associated anomalies. Sinus venosus atrial septal defects (SVD) in the strict sense cannot be called ASD. This is because there is no defect in any of the embryological inter-atrial septal component.<sup>4</sup> There is no direct communication between RA and LA, instead there is a window or passage of communication between pulmonary vein (PV) and SVC. The right upper pulmonary vein is usually the culprit. Sometimes,

more than one PV communicates with SVC.<sup>4</sup>

## Case Report

A 49 year old woman from UAE was admitted in the hospital due to respiratory distress and cyanosis. She had a history of unknown congenital heart disease from several years ago and was under medical treatment for right sided heart failure only with digoxin and lasix, without a known cause for right sided heart failure.

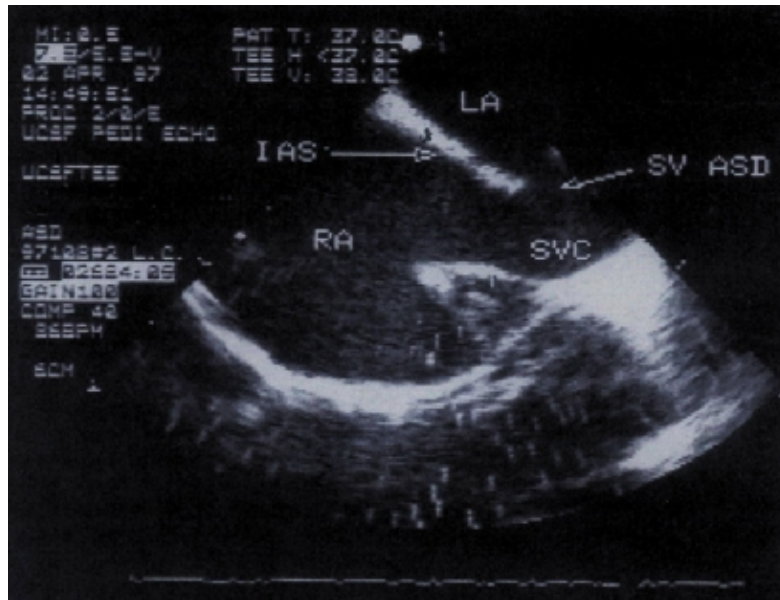
First, physical examination showed cyanosis of the upper extremity and mild edema in the lower extremity and fine basilar rales at the lungs but the heart sounds were muffle due to morbid obesity.

CXR showed cardiomegaly and pulmonary congestion. ECG showed sinus tachycardia, extreme axis deviation, right atrium enlargement, and right ventricular dilation and hypertrophy.

TTE showed a poor echo window due to morbid obesity, preserved left ventricle ejection fraction (approximately 50%), reduced right ventricle ejection fraction, dilated right ventricle, severe tricuspid regurgitation, right atrium enlargement, pulmonary artery systolic pressure approximately 130 mmHg, dilated pulmonary artery, pulmonary insufficiency, and pulmonary capillary wedge pressure approximately 24 mmHg, moderate pericardial effusion.

After stabilizing the general condition in CCU, the patient underwent TEE which in addition to previous data on TTE showed ASD sinus venosus type (SVC form with bidirectional shunt; Figure 1). The ASD flow was viewed on classic bicaval view.

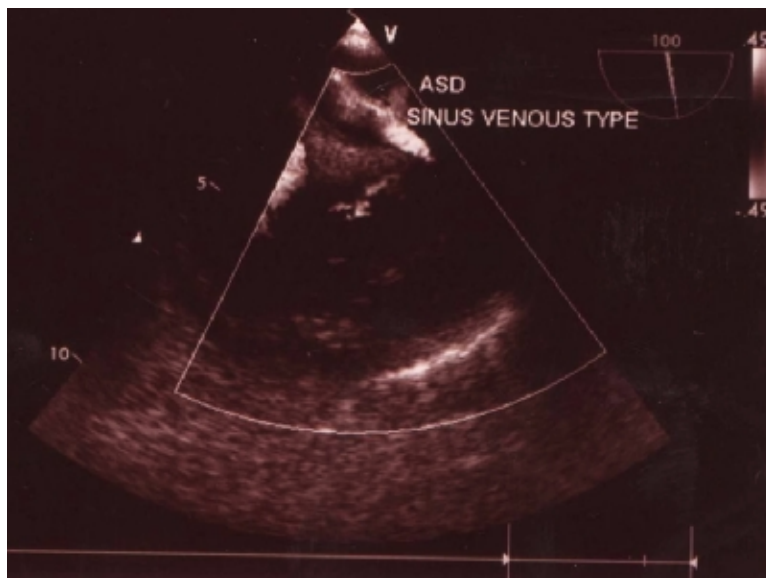
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**Fig. 1:** Longitudinal plane of interatrial septum showing the typical appearance of sinus venous defect (SV ASD), IAS=interatrial septum, LA=left atrium, RA=right atrium, SVC=superior vena cava (Courtesy of W. C. Miller-Hance, MD, and I. A. Muhiudeen Russell, MD, PhD, *Anesth Analg* 1997;85:1214-7)

(With the probe relatively high within the esophagus, a vertical plane allows both atrial and the inter-atrial septum to be recorded; this plane is called the bicaval view and also records the entrance of the superior vena cava in to the right atrium).<sup>5</sup> In this manner, the impression of Eisenmenger syndrome secondary to undiagnosed SVD was established (Figure 2).

The patient underwent exercise stress test with pulse oximetry, which showed low functional capacity (less than 4 METS) and decrease in oxygen saturation at pulse oximetry after exercise, and no ST-T changes. She was discharged from hospital with medical treatment and due to irreversible PAH, no intervention was indicated for her.



**Fig. 2:** Sinus venous type ASD with bidirect

## Discussion

Sinus venosus atrial septal defect is under-diagnosed with trans-thoracic echocardiography because of its posterior (far field) location. TEE should be ideally suited to diagnose SVD, given the proximity of the transducer to the defect.<sup>6</sup> TEE is accurate for the diagnosis of SVD should be undertaken in any patient with unexplained dilation of the right side of the heart. The best view on TEE for recognition of ASD sinus venosus (SVC type) is bicaval view.

It is advisable to search for primary and secondary cause of right side heart failure in a patient with right side heart failure and severe TR. Patients suspected with having Pulmonary Artery Hypertension (PAH) should undergo diagnostic testing. A useful assessment for these patients is exercise testing with pulse oximetry. Patients with little or no pulmonary vascular disease should continue to shunt left to right during exercise, and thus their pulse oximetry should be normal throughout. Patients with bidirectional shunting may have normal resting pulse oximetry but typically it will fall during exercise, reflecting shunt

reversal. Those with advanced pulmonary vascular disease will have hypoxemia at rest, which should worsen significantly with any level of exercise activity.<sup>7</sup> The goal of diagnostic testing is to confirm that PAH exists and to identify its underlying cause.

Echocardiogram is suggestive of pulmonary hypertension. In patients in whom there are sufficient cardiac anomalies on TTE, explaining PAH does not require further diagnostic testing but if there is no evidence of cardiac anomaly on TTE to clarify PAH, TEE should be performed. Patients with PAH should have exercise testing performed to determine the functional severity of the disease which will guide therapy decision. Cardiac catheterization for diagnostic purpose should not be required before surgical correction.<sup>5</sup> Right heart catheterization should be considered in patients to confirm pulmonary vaso-reactivity to a short acting vasodilatation.<sup>8</sup> In our patient, in spite of several hospital admissions and several TTEs, no definite cause was confirmed as a cause of severe TR and right-sided heart failure.

**Conflict of interest:** None declared.

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