

Inadvertent Diversion of Superior Vena Cava to the Left Atrium- A Diagnosis Made by Bubble Contrast Echocardiography

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ABSTRACT

Sinus venosus Atrial Septal Defect (ASD) is the abnormal drainage of pulmonary veins to right atrium, which can present with symptoms of pulmonary hypertension. Sinus venosus ASD presents a diagnostic challenge considering low detection rates using transthoracic echocardiography. We report a case of a 36-year-old female in whom the sinus venosus ASD was wrongly diagnosed as primary pulmonary hypertension. Also, the role of transoesophageal and bubble contrast saline echocardiography in detecting an adequate closure of sinus venosus ASD after the surgical repair was presented. Based on transthoracic echocardiography and ECG findings, the patient was diagnosed with primary pulmonary hypertension. Because of the persistent symptoms and failure to improve with Bosentan, Sildenafil and diuretics, the patient was advised Transoesophageal Echocardiography (TEE), which revealed superior venacava sinus venosus ASD. The patient had surgical closure of the defect but she developed cyanosis and hypoxemia after the surgery. The bubble contrast saline echocardiography was performed for detecting adequate closure of the defect. The bubble contrast echocardiography showed simultaneous opacification of all the four chambers of heart. Thereby, a repeat surgery was done and patient experienced improvement in her symptoms. The present case concludes that bubble contrast saline echocardiography can be used to detect the presence of intra and extra-cardiac shunt.

Keywords: Endocardium, Pulmonary hypertension, Sinus venosus atrial septal defect, Transoesophageal

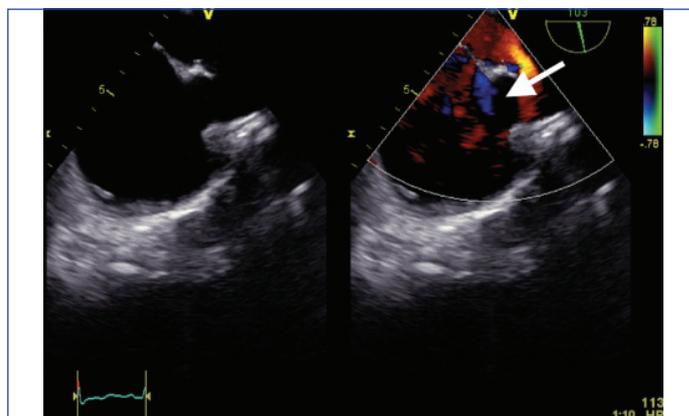
CASE REPORT

A 36-year-old female with previously diagnosed primary pulmonary artery hypertension presented with chief complaints of shortness of breath and easy fatigability since 8-10 years. Her previous ECG and 2-dimensional echocardiography reports were suggestive of pulmonary artery hypertension. She was taking Sildenafil, Bosentan and diuretics but there was no improvement in her symptoms. A repeat 2D transthoracic echocardiography was done in the hospital and showed dilated right atrium and right ventricle, severe tricuspid regurgitation and pulmonary artery systolic pressure of 100 mm Hg. There was no Echo dropout seen at inter-atrial septum. A subcostal view could not be obtained as the patient was obese.

Because of the persistent symptoms, patient was advised TEE to rule out ASD. TEE revealed superior vena cava type of sinus venosus ASD with a prominent left to right shunt with dilated right atrium and right ventricle and an anomalous right upper pulmonary venous drainage into the right atria [Table/Fig-1].

Patient was planned for surgical ASD closure. After the surgery, her physical examination and vitals were normal on day 1. However, on day 2 she developed cyanosis and started desaturating with SpO₂ of 76% on oxygen. The patient's arterial blood gas analysis reported hypoxemia with PaO₂ of 52 mmHg. The differentials for this clinical setting included pulmonary causes like atelectasis, pneumonia, or pleural collection and cardiac causes like residual ASD lesion, additional ASD lesion, and iatrogenic right to left shunt. To rule out the pulmonary causes, a chest X-ray was done but that was unrevealing.

Further, a 2-dimensional transthoracic bubble contrast saline echocardiogram was performed to rule out residual or additional ASD lesion. Bubble contrast echocardiography revealed simultaneous opacification of all four chambers of heart [Table/Fig-2]. TEE was done to assess the cause of bilateral opacification of atrium and ventricles. TEE showed a disrupted septal patch that was diverting the blood from

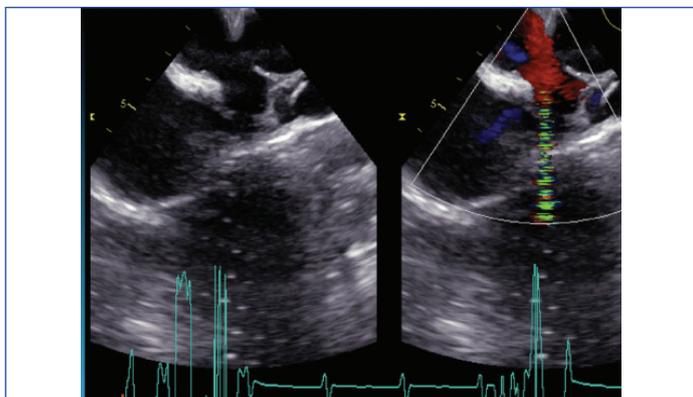


[Table/Fig-1]: Transoesophageal echocardiography suggestive of superior venacava type sinus venosus atrial septal defect.



[Table/Fig-2]: 2D transthoracic agitated saline contrast echocardiography showing bilateral simultaneous opacification of both atrium and ventricles.

superior vena cava directly to left atrium [Table/Fig-3] and because of this; the patient had cyanosis and decreased oxygen saturation. Patient was re-operated for ASD patch closure. TEE and bubble contrast were done again postoperatively. There was no negative contrast in right



[Table/Fig-3]: Transthoracic echocardiography suggestive of disrupted septal patch diverting superior vena cava flow to left atrium.

atrium and bubble saline contrast showed no bilateral opacification of atrium and ventricles. The patient was followed up a week after the patch closure and she had improved clinically with relief in her symptoms and her pulmonary artery systolic pressure decreased to 28 mmHg.

DISCUSSION

Sinus venosus defect is a type of ASD and an abnormal pulmonary venous drainage of pulmonary vein to right atrium. The defect can occur near the superior vena cava or inferior vena cava but former is more common. Sinus venosus constitutes 2-10% of all the ASDs [1]. Sinus venosus differs from other ASDs in that atrial septum is intact however, the defect is in the insertion of superior vena cava to right atrium. Sinus venosus can have wide range of presentations from being benign including minimal functional limitation with exercise to being severe with clinical presentations of exertional dyspnoea, atrial fibrillation, recurrent pulmonary infections, paradoxical emboli, and right heart failure.

Sinus venosus ASD should be considered as a differential in presentation with unexplained right atrial and ventricular dilation. Diagnosis of sinus venosus ASD is more difficult than other forms of ASD. The detection rates with transthoracic echocardiography are low ranging from 12-44% because of posterior location (detection rates are best using subcostal view) [2,3]. Therefore, to diagnose sinus venosus there is requirement of imaging modalities like TEE, Magnetic resonance imaging and computed tomographic scanning [4]. TEE seems to be a feasible imaging modality in developing countries in patients with unexplained right-sided dilation. The 2D Echo was unable to detect sinus venosus ASD in the patient. Also, because of the obesity, subcostal view could not be obtained. Thus, she was advised TEE, which revealed a sinus venosus defect.

The treatment of sinus venosus ASD is surgical repair with patch closure (single or double) or Warden procedure depending on the size of superior vena cava and site of drainage of pulmonary veins into superior vena cava [5]. The surgical repair should be done before two decades, as later repair is associated with dismal prognosis. However, it is possible that the defect is not repaired adequately and patient may have persistent symptoms. The patient was not diagnosed during early period thus she had delayed repair. After her

surgical repair, she developed hypoxemia and cyanosis. Akyuz M et al., reported the iatrogenic diversion of superior vena cava to left atrium after the surgical repair of ASD [6].

The use of bubble contrast saline echocardiography is known since 1968 however it is an underutilised imaging modality [7]. Bubble contrast saline echocardiography is a non invasive imaging test that uses imaging ultrasound combined with injection of microbubble agitated saline contrast to assess the intracardial blood flow and delineate the endocardium [8]. Bubble contrast echocardiography is an imaging modality that can be used not only for detecting patent foramen ovale but also for detection of intracardiac shunts, anomalous connection of systemic veins to left atrium, detection of shunt reversal due to elevated pulmonary artery pressure (Eisenmenger's syndrome), intrapulmonary (extracardiac) shunts, systemic to pulmonary venous connections, pericardiocentesis and other clinical applications [9]. This case presents the utility of a non invasive imaging bubble contrast echocardiography in detecting residual ASD lesion after the surgery. If it shows simultaneous opacification of all four chambers of heart, it implies that defect has not been repaired adequately and thus, a repeat operation is needed.

CONCLUSION

Sinus venosus ASD can be misdiagnosed and treated as pulmonary hypertension because of low detection with TTE. TEE is an important tool for diagnosis and this condition needs surgical repair. Earlier repair is associated with better prognosis and life expectancy. Bubble contrast echocardiography is an imaging tool that can be used postoperatively to detect if there is residual lesion and thus a repeat operation can be done as and when needed.

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