



Published in final edited form as:

Echocardiography. 2013 July ; 30(6): E182–E183. doi:10.1111/echo.12191.

A Sinus Venosus Atrial Septal Defect Is Diagnosed by Echocardiography with an Unusual Bubble Study

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Keywords

Eisenmenger's syndrome; sinus venosus atrial septal defect; contrast echocardiography

A 68-year-old man with a history of shortness of breath and hypoxia was undergoing workup for severe pulmonary hypertension. He was referred to pulmonary hypertension clinic for the first time. An echocardiography (iE33, Philips Medical Systems, Bothell, WA, USA) with agitated saline was performed. The calculated pulmonary artery pressure was 113 mmHg which was associated with right ventricular hypertrophy and dilatation. On contrast injection with agitated saline, the bubbles surprisingly filled the left heart rapidly before the right side (Fig. 1 and movie clips S1 and S2). Based on this bubble study, a diagnosis of Eisenmenger's syndrome from a sinus venosus atrial septal defect was made and later confirmed on cardiac magnetic resonance imaging (Avanto, Siemens Medical Solutions, Erlangen, Germany) (Fig. 2).

The fact that the bubbles first appear on the left side suggests that the shunt occurs above the level of the atria. Because of high right-sided pressures in this case, flow from the superior vena cava (SVC) preferentially travels through the defect to the right superior pulmonary vein and left atrium.

A residual left SVC with a direct connection to the left atrium via an unroofed coronary sinus was also considered as it would have also shown a similar filling pattern with agitated saline. This defect was excluded, however, because the agitated saline was serendipitously injected into the right hand.

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Supporting Information

Additional Supporting Information may be found in the online version of this article:

In this case, the shunt was not diagnosed until late in life. He had been treated for years with vasodilators for presumed primary pulmonary hypertension. Unfortunately, he died 3 months later from severe Eisenmenger's syndrome. In this case, the initial echocardiography with agitated saline was essential in making the diagnosis, but because of the large right-to-left shunt, further bubble studies should be avoided in patients with known Eisenmenger's syndrome.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

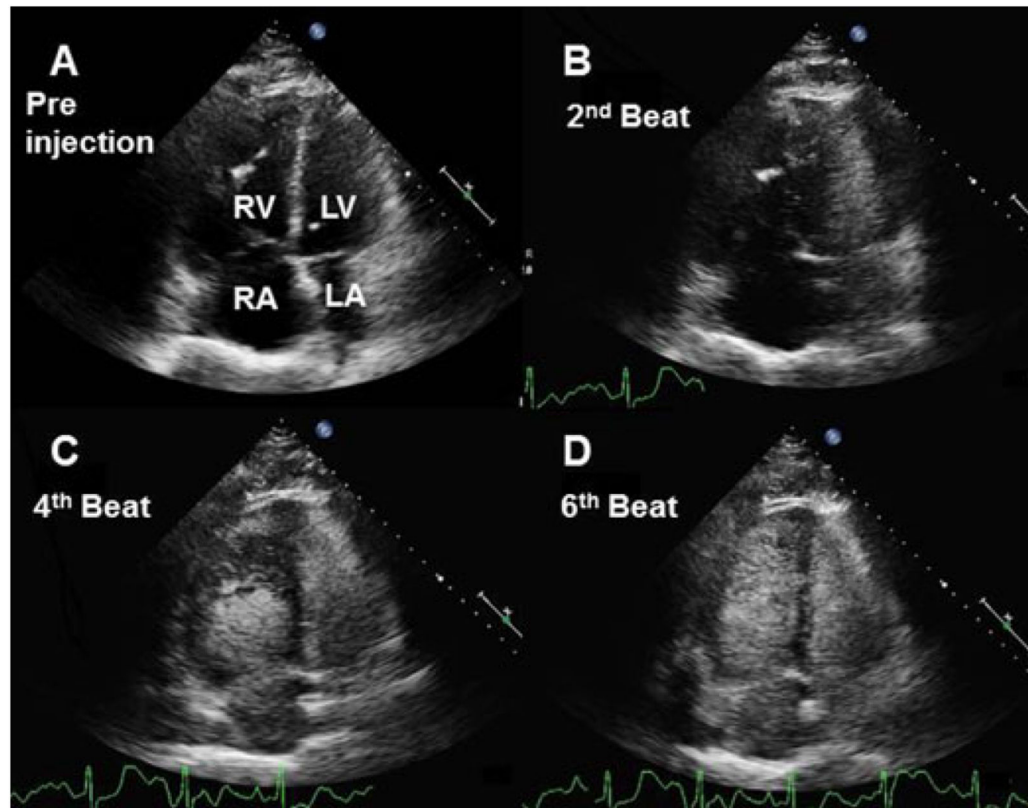


Figure 1.

Echocardiography with agitated saline injection in a patient with a sinus venosus atrial septal defect. **A.** shows a four-chamber view prior to the injection of agitated saline, highlighting the right ventricular enlargement and hypertrophy associated with severe pulmonary hypertension. **B.-D.** show the progress of the bubbles after the 2nd, 4th, and 6th beats, respectively. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.



Figure 2. The MRI of the sinus venosus atrial septal defect. LA = left atrium; RA = right atrium; SVC = superior vena cava.