

# Diagnosis of the superior sinus venosus defect by intracardiac echocardiography

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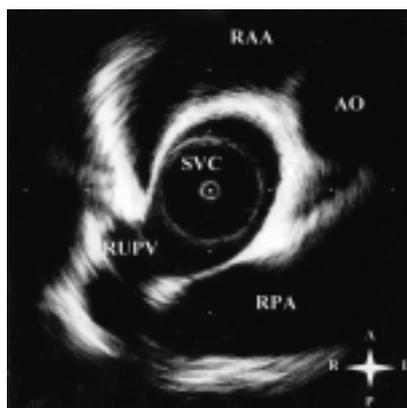
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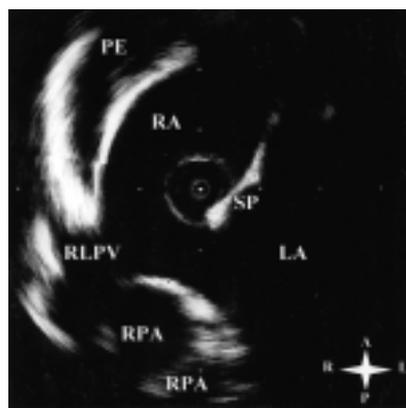
Direct embryological evidence and the clinical diagnosis of the superior sinus venosus defect are still, at present, a matter of debate<sup>1</sup>. Despite prior descriptions of the diagnostic echocardiographic and angiographic features, its location makes correct identification and diagnosis during life difficult, and both false positive and negative findings still occur.

We report the unique anatomical intracardiac echocardiographic features of the superior sinus venosus defect and of the anomalous pulmonary venous drainage in a

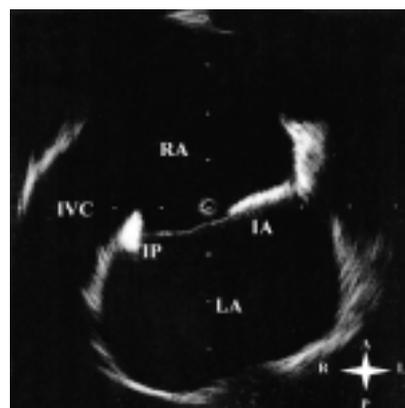
72-year-old man. The patient, suffering from systemic hypertension and atrial fibrillation was admitted to our center for effort dyspnea and a recent transient ischemic attack. Transthoracic echocardiography revealed an increased volume of the left and right ventricles, a middle pulmonary hypertension, and a middle mitral and tricuspid valve regurgitation. A left-to-right shunt was present at the atrial level. Subsequent transesophageal echocardiography confirmed the left-to-right shunt, suggesting but not proving the presence of



**Figure 1.** Transverse imaging view at the level of the great vessel plane. The short axis of the superior vena cava (SVC) and of the ascending aorta (AO) are viewed, whereas the right pulmonary artery (RPA) is resected along its long axis. Anterior to the SVC, the right atrial auricle (RAA), similar in appearance to Snoopy's nose may be seen. The SVC is larger than a normal one and it is abnormally shifted leftwards. The right upper pulmonary vein (RUPV) enters the right-posterior free wall of the SVC, resulting in a teardrop appearance (16.6 mm) instead of the normally round appearance.



**Figure 2.** Longitudinal view at the level of the superior vena cava-right atrium junction plane, which highlights the sinus venosus defect (arrowhead, 19.2 mm) lying exterior to the fossa ovalis, with the orifice of the superior vena cava having a biatrial connection and overriding the superior-posterior rim (SP) of the fossa. It is apparent that the interatrial communication is the result of a defect of the most posterior and superior common infolding wall between the attachments of the superior vena cava to the right atrium (RA) and of the right pulmonary veins to the left atrium (LA). The connection of the right lower pulmonary vein (RLPV) to the cava-atrial junction is also clearly visible. PE = pericardial effusion; RPA = right pulmonary artery.



**Figure 3.** Longitudinal view at the fossa ovalis level, generating a 4-chamber plane in a familiar apex-up orientation. This view allows clear images of both atria (RA, LA), of the inferior vena cava (IVC) and of the true interatrial septum. Within this extensive region (71.6 mm), the floor of the fossa ovalis (30.5 mm) is easily identifiable as a distinct component of the interatrial septum, characterized by a relatively thin and intact membranous flap valve within the thicker muscular septum. The inferior-posterior rim (IP) of the fossa continues directly into the wall of the IVC, whereas the inferior-anterior rim (IA) continues with the muscular atrioventricular septum. Other abbreviations as in figure 2.

a superior vena cava interatrial defect. The patient underwent intracardiac echocardiographic evaluation which clarified the type of defect and demonstrated a superior sinus venosus interatrial defect (Figs. 1-3). The patient was referred to the cardiac surgeon and was successfully submitted to surgical correction. The intracardiac echocardiographic findings were confirmed intraoperatively.

The following anatomical criteria are the most important ones for the diagnosis of a superior sinus venosus defect. Firstly, the abnormal location of the superior vena cava attached to both atria, with its orifice overriding the intact muscular superior-posterior rim of the fossa ovalis (this is an anatomical prerequisite criterion and the key to diagnosis). Secondly, the abnormal location of the defect exterior to the fossa ovalis (archetypal septa), at the level of the most posterior and superior infolding of the wall of the two atria, and incorporating a wedge of extracardiac space and adipose tissue. This criterion allows one to distinguish between secundum atrial septal defects and interatrial communications such as the coronary sinus defect and the atrioventricular septal defect of an ostium primum variety, and the sinus venosus defect.

Thirdly, the connections of the right pulmonary vein might also be abnormal, but an anomalous pulmonary venous drainage is not held to be a basic anatomical feature because it can be associated with secundum atrial septal defect or it can appear as an isolated defect associated with an intact atrial septum.

As our case shows and as confirmed by a recent study<sup>2</sup>, standard echocardiographic studies often failed to identify these anatomical criteria. In this setting, due to the proximity of the transducer to cardiac structures, intracardiac echocardiography could be a highly accurate tool enabling correct diagnosis of a superior sinus venosus defect in adult populations.

## References

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