Sinus Venosus Defect - A Case Series

Estelle Abela, Luise Reichmuth, Alex Borg, Herbert Felice, Maryanne Caruana

AIM

Right heart chamber dilatation on cardiac imaging should raise the possibility of an intracardiac shunt at atrial level. We present two cases of patients who were diagnosed with a sinus venosus defect (SVD) in adult life.

Methodology

Patient 1 is a 72-year-old man with multiple comorbidities who had been known to have dilated right heart chambers for many years, presented with peripheral oedema and breathlessness. Patient 2 is a 32-year-old previously healthy man who was noted to have an ejection systolic murmur at a medical test prior to enrolment in an athletics club. Both patients underwent echocardiography and computed tomography.

Results

Retrospective reassessment of the CT pulmonary angiogram of Patient 1 confirmed the presence of anomalous drainage of two right upper pulmonary veins into the superior vena cava (SVC). A subsequent transoesophageal echocardiogram (TOE) demonstrated the defect high in the atrial septum and showed bidirectional shunting on colour Doppler. Catheter studies showed a Qp:Qs of <1, confirming reversal of shunting and an Eisenmenger physiology. Transthoracic echocardiography of Patient 2 showed dilated right heart chambers. Cardiac CT confirmed the presence of a variant of SVD (cavopulmonary window). The patient was put forward for surgical correction.

Discussion

A sinus venosus defect (SVD) is one form of interatrial communication. It is related to the insertion of the superior or inferior vena cava into the right atrium (RA) and is usually accompanied by partial anomalous pulmonary venous drainage (PAPVD). SVDs are notoriously easy to miss on transthoracic echocardiography (TTE) due to their location.

Conclusion

Timely diagnosis of SVD relies on a high index of suspicion and usually requires further imaging following an initial TTE. Surgical correction to close the defect and re-route all pulmonary venous drainage to the left atrium is recommended in haemodynamically significant shunts, as long as pulmonary pressures are not prohibitively elevated.