Sinus venosus ASD

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A 32-year-old man presented at the cardiology department for a routine check-up. He had no symptoms, particularly no cardio- logical symptoms. Clinical examination and ECG revealed no abnormalities. Transthoracic echocardiography showed a mild dilatation of the right heart.

Further functional evaluation of the right heart was performed with MRI to evaluate the presence of a left to right shunt as a possible cause of right heart dilatation. We used the phase-contrast technique to measure the flow in the left ventricular outflow tract ($Q_s = 4.6$ l/min) and right ventricular outflow tract ($Q_p = 13$ l/min). The $Q_p/Q_s$ ratio measured 2.88, indicating a severe left to right shunt.

Anatomical work up included a cardiac CT. It showed a dilated right heart (Fig. A) with a large defect in the upper atrial septum (Fig. B) contiguous with the superior vena cava, which is a sinus venosus type of atrial septal defect (SVA SD). There was also an anomalous drainage of the right upper lobe pulmonary vein in the superior vena cava (Fig. C).

Comment

There are 4 types of ASD: septum primum, septum secundum, patent foramen ovale and sinus venosus defect (SVASD). SVASD is located posterior to the foramen ovale and near the superior vena cava. Atrial septal defects lead to volume overload of the right heart with right heart dilatation and eventually pulmonary hypertension as a complication.

SVASD is a rare type of ASD and is almost always associated with anomalous connection of the right upper pulmonary vein into the superior vena cava, which aggravates the left to right shunt.

Diagnosis can be made by several diagnostic imaging techniques. Echocardiography is a good and non-invasive option but for better anatomical detail cardiac CT or cardiac MRI can be used. MRI also had the benefit to give functional information in addition to anatomical information.

The mainstay of the therapy is surgical correction. Repair of SVASD is more complex than repair of the other types of ASD. A patch will redirect the blood flow from the right upper lobe pulmonary vein into the left atrium with closure of the interatrial communication and also correcting the anomalous pulmonary venous drainage. Adults with a left to right shunt of more than 2 benefit from surgical repair.

Reference


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