Case 4542

Sinus Venosus Atrial Septal Defect associated with Partial Anomalous Pulmonary Venous Return

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Patient: 20 years, male

Clinical History:

Asymptomatic 20 years old with a PA chest X-ray showing enlargement of the main pulmonary artery convexity, increased pulmonary vascularity and right ventricle enlargement (fig.1).

Imaging Findings:

Transthoracic echocardiography suggested the ASD (atrial septal defect) showing a right atrium enlargement although no communication between the right and left atrium was depicted. MRI examination revealed enlargement of the right chambers (fig.2), high septal communication between LA (left atrium) and RA (right atrium) with L-R flow (fig.3) as well as PAPVR (partial anomalous pulmonary venous return) of the right superior pulmonary vein to the SVC (superior vena cava) root through a defect in the septum that normally separates them (fig.4,5). Flow quantification using velocity encoded (VENC) cine phase contrast at the level of the ascending aorta and the main pulmonary artery revealed the higher pulmonary output (Qp) in comparison with the systemic one (Qs) (fig. 6). The calculated ratio Qp/Qs was 2, expressing the L-R shunt.

Discussion:

PAPVR is a condition that behaves as an ASD and is usually asymptomatic during the first years of life. Anomalous pulmonary venous return to the SVC in association with ASD of the sinus venosus (very high in the atrial septum, near the insertion of the SVC) is the most frequent type of PAPVR. There is a defect in the septum that separates the SVC from the superior right pulmonary vein. It is often difficult to detect the pulmonary vein confluence or the combined congenital anomaly by echocardiography. Catheter based angiography is an invasive method that may be required in case of doubt. Multislice CT is a another promising non-invasive technique that may provide useful information on anatomical abnormalities. Cardiovascular MRI is an emerging technique that can non-invasively 1) define ASD anatomy of the defect, 2) size the defect, which is a valuable preoperative information, 3) define the anatomy of pulmonary veins in sinus venosus defects, 4) accurately quantify Qp/Qs avoiding invasive catheterization studies, 5) predict the successful closure of the defect, 6) evaluate the function of the ventricles. The condition requires surgical closure of the SVASD (sinus venosus ASD) with a patch that incorporates all the pulmonary veins to the left atrium in patients with significant shunt. Operation for SVASD is associated with low morbidity and mortality, and postoperative subjective clinical improvement occurs irrespective of age at surgery. Postoperative atrial fibrillation appears to be related to older age at operation. SVASD repair achieves survival similar to that of a matched population and should be considered whenever repair may impact survival or symptoms. Other types of PAPVR are the drainage of the superior pulmonary vein into the SVC without ASD (at slightly higher point), right pulmonary vein into the right atrium (this condition is often associated with ASD), anomalous return of the right pulmonary vein into the inferior vena cava (IVC) (scimitar syndrome, often accompanied by hypoplasia of
the right lung), anomalous left pulmonary vein return to the innominate vein through the persistence of a vertical vein and other more rare kinds.

**Differential Diagnosis List:** Sinus Venosus ASD associated with PAPVR

**Final Diagnosis:** Sinus Venosus ASD associated with PAPVR

**References:**

J. Bogaert, S. Dymarkowski, A.M.Tylor Clinical Cardiac MRI.
Description: PA chest X-ray showing enlargement of the main pulmonary artery convexity, increased pulmonary vascularity and right ventricle enlargement (fig.1). Origin:
Figure 2

Description: Axial b-SSFP shows dilatation of right ventricle and atrium

Origin:
Description: Axial b-SSFP shows a defect in the portion of the atrial septum between the LA and the SVC-RA junction Origin:
Description: Axial b-SSFP shows anomalous return of right upper pulmonary vein to SVC.

Origin:
Description: Sagital b-SSFP shows the defect between LA and RA Origin:
Figure 6

Description: Results of VENC cine PC, show Qp/Qs ~2

Origin: