Cardiac Imaging

Sinus Venosus Defect: Right Heart Catheterisation and Computed Tomographic Scan

Georgios Leventopoulos, Georgios Giannopoulos, Spiridon Deftereos, Nikolaos Kouvousis, Andreas Kaoukis, Vlassios Pyrgakis

Department of Cardiology, "G. Gennimatas" Hospital, Athens, Greece

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Address:
Georgios Leventopoulos

53 Fokidos St.
115 27 Athens, Greece
e-mail: levent2669@hotmail.com

The *sinus venosus* type atrial septal defect is a rare congenital cardiopathy that is mostly associated with partial anomalous pulmonary venous return (PAPVR). Pulmonary hypertension is the clinical sequela if this cardiac defect remains unrepaired. Right heart catheterisation is a procedure for evaluating the haemodynamic severity of the cardiopathy, measuring pulmonary resistance and determining the appropriate medical therapy.

A 63-year-old female presented with orthopnoea and dyspnoea. Her symptoms were attributed to a narrow regular QRS tachycardia of 150 bpm, which was not terminated by adenosine administration. Rate control was achieved by a short acting β-blocker (esmolol). The ECG demonstrated sinus rhythm and many signs indicative of right ventricular hypertrophy, such as right axis deviation, R/S ratio >1 in V_1, positive peaked P waves best seen in lead II, and an inverted P in V_1. An atrial septal defect was suspected, based on clinical examination and ECG. Marked dilation and pressure overload of the right cardiac chambers were revealed by transthoracic ultrasonography. Right ventricular systolic pressure was estimated at around 90 mmHg. A *sinus venosus* defect was suspected, combined with partial anomalous pulmonary venous return. These findings were confirmed by a transoesophageal examination. Bidirectional atrial shunting implied the development of Eisenmenger syndrome and the right upper pulmonary vein was abnormally connected to the right atrium.

Right heart catheterisation was performed in order to measure pressures in the right cardiac chambers and the pulmonary vasculature, and to further evaluate any anatomical defects. The values were consistent with those derived from the echocardiographic study. Oxygen saturation was measured in different anatomical regions. These results provided more evidence of PAPVR. Specifically, oxygen saturation was unexpectedly elevated (87.5%) in the high right atrium region, suggesting a mixture of low and highly saturated blood coming from the superior *vena cava* and the right upper pulmonary vein, respectively. The route of the abnormal pulmonary vein was depicted by contrast angiography (Figure 1A). A vasoreactivity test was also done using the standard procedure of administering adenosine in gradually increasing doses. No decline in mean pulmonary pressure was recorded, indicating irreversible lesions in the pulmonary arterioles. Multi-slice computed tomography was performed. The multi-planar reformations and the three-dimensional reconstructed images depicted the precise anatomy of the atrial septal defect and the abnormal return of the right upper pulmonary vein to the right (Figures 1B, 1C, 1D).[1]

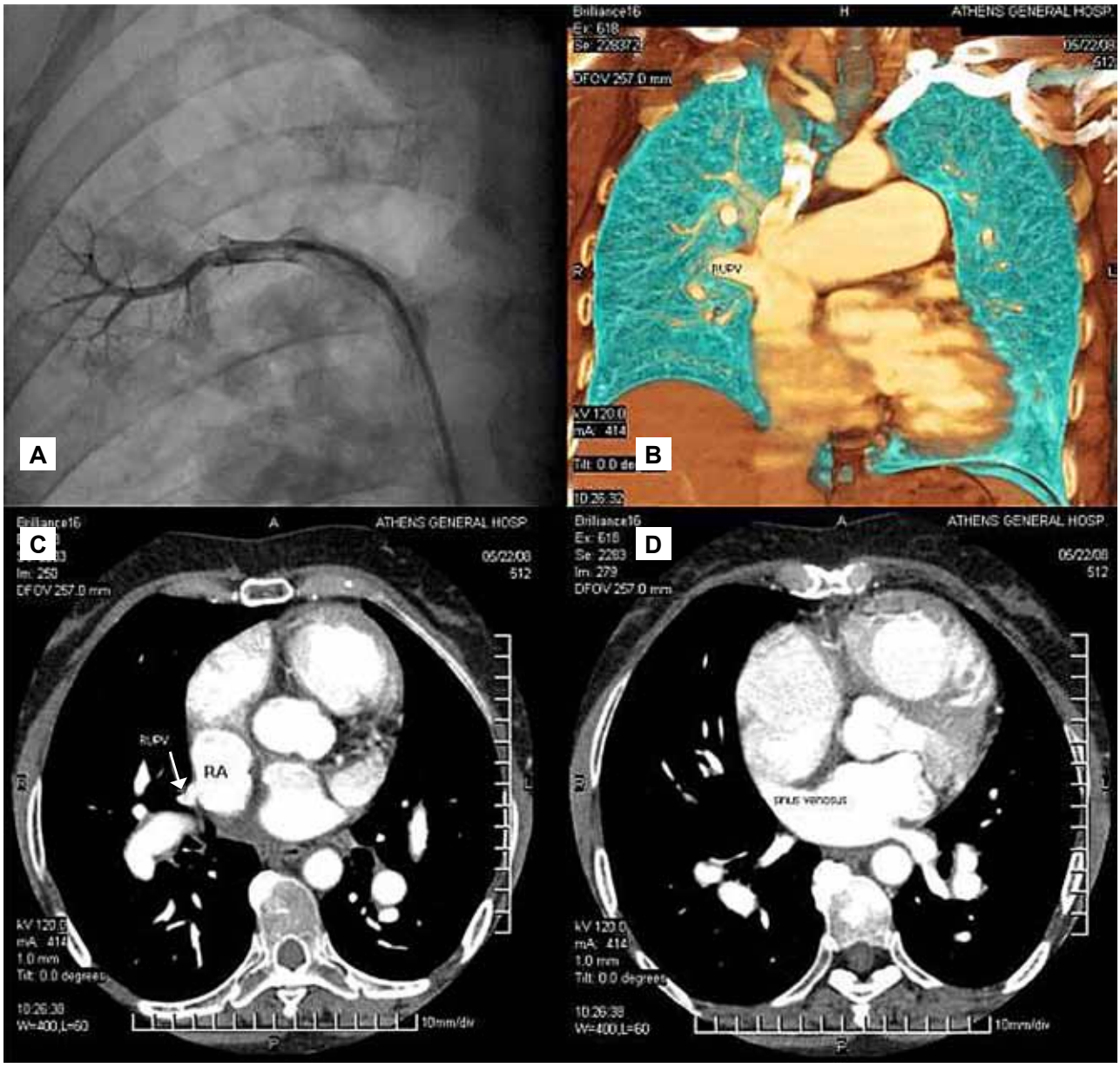

Figure 1. Abnormal return of right upper pulmonary vein and *sinus venosus* defect. The route of the right upper pulmonary vein (RUPV) is seen in the right heart catheterisation (A) and the cardiac computed tomographic scan (B). Abnormal return of the pulmonary vein into the right atrium (RA, arrow) and the size of the *sinus venosus* defect are illustrated in images C and D, respectively.

Sinus venosus defect is the rarest of the three most common types of atrial septal defect (<10% of all cases). Its anatomical site precludes the possibility of transcatheter device closure. This congenital cardiopathy is commonly associated with total or partial anomalous pulmonary venous connection.[2] This malformation contributes to additional left-to-right shunting. Thus, the development of Eisenmenger syndrome occurs earlier than with an isolated atrial septal defect. However, as a result of the PAPVR, cyanosis—the clinical component of Eisemenger—is not evident in such cases because of the well-saturated blood mixture via the intra-atrial shunting. A vasoreactivity response test is a useful test to determine safely whether the patient has a positive acute response to vasodilating agents.[3] In this case, the patient did not present a vasodilating response, as no decline in mean pulmonary pressure was observed during adenosine administration because the pulmonary arterioles have lost their functional vasodilating properties.

Any surgical correction of the defect would have

worsened the patient's clinical status. This group of non-responders can be started on p.o. bosentan, and bosentan was given to our patient. *Per os* administration facilitates patient compliance, while there are optimistic results from the BREATHE 5 study concerning the long-term use of bosentan in Eisenmenger patients and clinical improvement.[4]

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