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Multiple atrial septal defects with concomitant partial anomalous pulmonary venous return on cardiac computed tomography

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Short title: Multiple ASD with PAPVR

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We present a case of multiple atrial septal defect (ASD) with concomitant anomalous pulmonary venous return on cardiac computed tomography images.

A 42-year old female patient undergoing a scheduled diagnosis at the laboratory of computed tomography due to deterioration of exercise tolerance observed for many years and dyspnoea during exercise, currently NYHA class II/III. Her medical history also reveals mixed hyperlipidaemia and nicotinism.

Cardiac computed tomography (CCT) was performed using a protocol for assessing cardiac chamber morphology with maximum saturation of the left atrium with a contrast agent. With regard to the interatrial septum, a double defect has been revealed with pronounced signs of interatrial shunt. The first observed defect in the form of ostium secundum ASD (ASD II) had a size of 0.8 x 0.7 cm. The second observed defect in the form of sinus venosus ASD (SV ASD) was located at the opening of superior vena cava into the right atrium, and it had a size of 1.0 x 0.8 cm. Moreover, CCT revealed anomalous pulmonary venous return. The image showed 4 right pulmonary veins - 2 right upper pulmonary veins (RUPV1 and RUPV2), right intermediate pulmonary vein (RIPV) and right lower pulmonary vein (RLPV); and 2 left pulmonary veins - left upper pulmonary vein (LUPV) and left lower pulmonary vein (LLPV). RUPV1 and RUPV2 opened to the superior vena cava (SVC), RIPV at the border of the superior vena cava and right atrium; RLPV, LUPV and LLPV opened typically into the left atrium. The examination also revealed dilation of the right atrium, right ventricle and pulmonary veins. Left ventricular ejection fraction estimated by tomography was 72%, and right ventricular ejection fraction was 46%.

Atrial septal defects are the most common congenital heart defects in adults, representing about 40% of congenital heart defects diagnosed in patients over 40 years old [1]. The most common is single ASD, which in the majority of cases (70%) is located in the mid part of the atrial septum (ostium secundum ASD). SV ASD is rare and occurs in 5-10%
of ASD cases. Multiple ASDs are very rare [2]. ASD is typically asymptomatic at young age and becomes symptomatic around 40 years of age. ASD symptoms increase with age [3]. If ASD is diagnosed, closure should be considered, optimally percutaneous [4].

The rate of partial anomalous pulmonary venous return (PAPVR) in the population is 0.4-0.7%. The probability of its occurrence is 10-fold higher in the presence of ASD. It usually (60-90%) affects right pulmonary veins with a typical additional abnormal opening of a single pulmonary vein beside the left atrium. PAPVR is often asymptomatic, while symptoms of overload of the right cardiac chambers occur in the presence of large shunts or in the event of concomitant ASD [5].

Summing up, the presented case of multiple ASD with PAPVR represents one possible anatomical variant of congenital heart defect of this type - a very rare variant from the epidemiological point of view.
References


**Figure 1** Cardiac computed tomography

A. Axial reconstruction. Double atrial septal defect: ostium secundum defect (white arrow) and sinus venosus defect (black arrow).

B. VRT reconstruction. Atrial septal sinus venosus defect (black arrow).

C. Axial reconstruction. Opening of the right upper pulmonary vein 1 into the superior vena cava (black arrow).

D. Axial reconstruction. Opening of the right upper pulmonary vein 2 into the superior vena cava (black arrow).

E. Axial reconstruction. Opening of the right intermediate pulmonary vein on the border of the superior vena cava and right atrium (black arrow).

F. MIP reconstruction. Typically opening of the right lower pulmonary vein (black arrow pointing right), opening of the left upper pulmonary vein (black arrow pointing downwards) and opening of the left lower pulmonary vein (white arrow pointing upwards) into the left atrium.