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STUDIUM PRZYPADKU / *CLINICAL VIGNETTE*

Cor triatriatum and a rare form of partial anomalous pulmonary venous drainage

Serce trójprzedsionkowe i rzadka postać częściowego nieprawidłowego spływu żył płucnych

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An eutrophic five-month-old infant boy presented with systolic murmur without congestive heart failure. Echocardiography established cor triatriatum sinistrum (Fig. 1) with mild obstruction (mean Doppler gradient 4.5 mm Hg) between the upper-proximal and lower-distal parts of the left atrium (LA). A 4 mm secundum atrial septal defect (ASD) was also described between the upper LA and right atrium (RA) with left-to-right shunt, as well as partial anomalous pulmonary venous drainage (PAPVD) of the left lower pulmonary vein (PV) to the dilated coronary sinus (CS) (Fig. 2). Echocardiographic signs of significant right ventricular volume overload were present. Computed tomography angiography confirmed cor triatriatum sinistrum (Fig. 3) and the PV drainage was described as follows: both right PVs and left upper PV to upper LA part, and confirmed PAPVD of left lower PV to CS. A surprising new finding was the presence of a connecting vein between the left upper and left lower PVs (Fig. 4). At the age of nine months the patient underwent surgical correction (membrane excision, CS unroofing, and ASD closure) without complications. Cor triatriatum sinistrum is an uncommon congenital cardiac anomaly accounting for 0.1–0.4% of all congenital heart defects, where the LA is divided by a membrane. Usually the PVs drain into the upper LA part; while the lower is the "true" LA connected to the left ventricle. One or more defects may be found in the membrane, as well as an ASD to RA. Associated congenital heart anomalies are reported in 50% of cases, like tetralogy of Fallot, ventricular septal defects, PAPVD, etc. Clinical manifestation of cor triatratum sinistrum is variable from asymptomatic to congestive heart failure, and even pulmonary oedema, according to the degree of flow obstruction between the upper and lower LA parts or due to associated defects, i.e. left-to-right shunting via ASD or PAPVD, as in our patient. PAPVD associated in this setting is quite frequent (9–25%). It is usually described to course to RA, to superior vena cava (SVC) directly or via vertical vein, or very rarely, via persistent L-SVC to CS. On the other hand, the PV drainage to CS, combined with an anomalous connecting vein between the upper and lower left PVs, is to our knowledge the first ever reported variation of this anomaly.

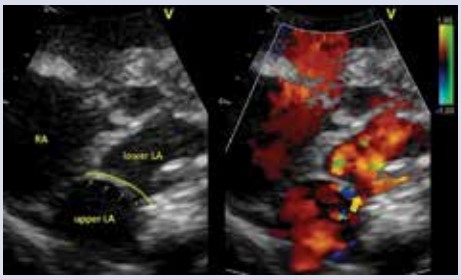

Figure 1. Echocardiography. Cor triatriatum sinistrum (arrow); LA — left atrium; RA — right atrium

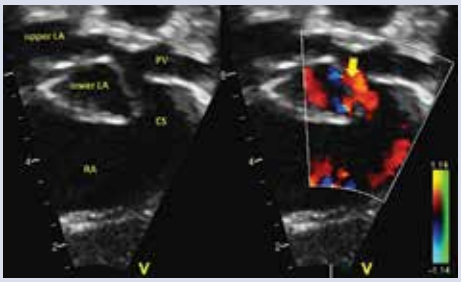

Figure 2. Echocardiography. Anomalous pulmonary venous drainage to dilated coronary sinus (CS) (arrow); LA — left atrium; PV — pulmonary vein; RA — right atrium

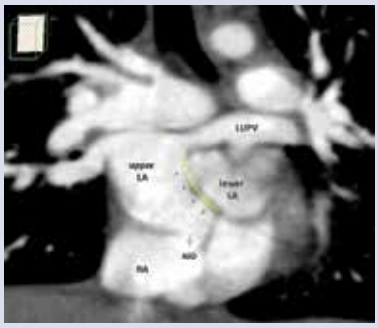

Figure 3. Computed tomography angiography. Cor triatriatum sinistrum; ASD — atrial septal defect; LUPV — left upper pulmonary vein; LA — left atrium; RA — right atrium

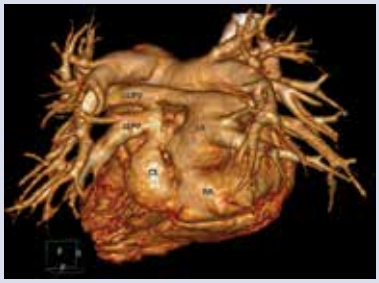

Figure 4. Computed tomography angiographic three-dimensional image. Pulmonary venous drainage and the connecting vein between left upper and left lower pulmonary veins (LUPV, LLPV); CS — coronary sinus; LA — left atrium; RA — right atrium; *Connecting vein between LLPV and LUPV

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Conflict of interest: *none declared*