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STUDIUM PRZYPADKU / *CLINICAL VIGNETTE*

Interruption of the fourth aortic arch and a persistent fifth aortic arch with coarctation: a rare morphological finding

Przerwanie czwartego łuku aorty i przetrwały piąty łuk aorty z koarktacją: rzadkie znalezisko morfologiczne

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A eutrophic six-month-old boy was admitted to our institution due to bronchitis. Precordial systolic murmur, weak arterial pulsation on the lower extremities, and a systolic non-invasive blood pressure difference of 35 mm Hg between upper and lower extremities were found. Echocardiography showed normal left-sided branching pattern of the 'superior' aortic arch with a blind-ending, and the 'inferior' aortic arch connecting to the descending aorta with coarctation (with isthmus diameter 2.6 mm and continuous peak Doppler gradient 60 mm Hg) (Fig. 1A, B). Additional mild left atrial and left ventricular dilatation and moderate mitral insufficiency were present. Computed tomography angiography confirmed the diagnosis — interruption of the fourth aortic arch and a persistent fifth aortic arch with coarctation (Fig. 2). Surgical resection of the coarctation and extended end-to-end anastomosis was performed with an uneventful postoperative course. Persistent fifth aortic arch is an extremely rare anomaly first described by Van Praaghs in 1969 (Am J Cardiol, 1969; 24: 279–282) and is defined by two parallel aortic arches coursing above each other. Two morphological variations may occur. The first is a systemic-to-systemic connection between the ascending and descending aorta. This applies in 76% of cases, and this type was present in our patient. The second variation is a systemic-to-pulmonary connection between ascending aorta and left pulmonary artery. This anomaly is often associated with other congenital heart defects, most frequently with coarctation of the aorta or interruption of the aortic arch (in 38%) — both of these were present in our case. This anomaly has also been described with other defects such as pulmonary atresia, tetralogy of Fallot, tricuspid atresia, ventricular septal defect and persistent ductus arteriosus. Clinical manifestation depends on the severity of the associated anomalies. An isolated form without aortic narrowing has no clinical manifestation. If combined with severe coarctation, it can manifest already in the neonatal period with congestive heart failure, cardiogenic shock and/or multiorgan failure. Mild coarctation may be detected during the first months of life, as was the case in our patient, or it can be found accidentally in an adult patient with systemic arterial hypertension. Treatment is surgical in symptomatic patients — coarctation resection and end-to-end anastomosis; while other techniques use a patch or grafts. Stent implantation may be considered in adult patients.

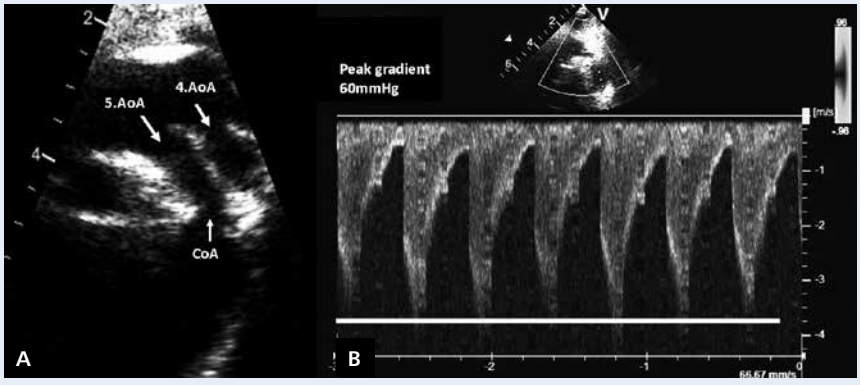

Figure 1. Transthoracic echocardiography; **A**. Two-dimensional image of 'superior' (fourth) aortic arch with interruption and 'lower' (persistent fifth) aortic arch with coarctation; **B**. Peak Doppler gradient at the coarctation; 4.AoA — fourth aortic arch; 5.AoA — persistent fifth aortic arch; COA — coarctation of the fifth aortic arch

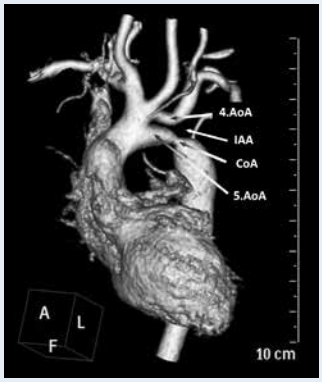

Figure 2. Computed tomography-angiographic three-dimensional image — interruption of fourth aortic arch (4.AoA) and a persistent fifth aortic arch (5.AoA) with coarctation; IAA — interruption of the fourth aortic arch; COA — coarctation of the fifth aortic arch

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Conflict of interest: *none declared*