

Scimitar syndrome and a common origin of the brachiocephalic trunk and left common carotid artery: a unique coexistence

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Partial anomalous pulmonary venous return (PAPVR) is a rare congenital heart defect (prevalence of 1–3/100 000 live births, with a female predominance). It involves abnormal venous return from the right lung directly or indirectly to the right atrium, usually to the superior vena cava or, less frequently, to the right atrium or inferior vena cava (IVC). The PAPVR anomaly with venous return directly into the IVC is called scimitar syndrome. Most commonly, PAPVR-associated abnormalities include hypoplasia of the right lung and pulmonary artery, and dextrocardia. Since the first report of PAPVR in 1836 and coinage of the term “scimitar syndrome” for its variant by Catherine Neill in 1960, numerous papers have described this anomaly along with its variations. The developmental errors leading to the scimitar syndrome are not clearly understood so far.

A common origin of the brachiocephalic trunk (the so-called innominate artery) and the left common carotid artery, known as the bovine arch, bicarotid trunk, or common brachiocephalic trunk, is the most common variation of aortic arch branching, with a prevalence between 7% and 27%.¹ The coincidence of PAPVR with a common origin of innominate and carotid arteries (COIC) has been reported only once so far.²

A 46-year-old man with previously diagnosed scimitar syndrome and dextrocardia was admitted to our center for further evaluation. He

complained of reduced exercise tolerance. Chest X-ray confirmed dextrocardia and also showed enlarged pulmonary trunk and asymmetric distribution of the pulmonary blood flow with increased blood flow in the right lung (Supplementary material, *Figure S1*). Transthoracic echocardiography (TTE) revealed mildly increased diameter of the right ventricle, right atrial enlargement (area of 24 cm²), and increased diameter of the pulmonary trunk (29 mm), without other signs of pulmonary hypertension. The diameter and function of the left ventricle were normal. Transesophageal echocardiography (TEE) showed a typical return of the left pulmonary veins, while the assessment of the right pulmonary veins was ambiguous. No atrial septal defect was visualized. Chest computed tomography (CT) and cardiac magnetic resonance (CMR) revealed anomalous venous return from the right lung into the IVC, with right lung hypoplasia (*FIGURE 1A* and Supplementary material, *Figure S2*). Additionally, the presence of COIC was visualized (*FIGURE 1B* and *1C*). The right-to-left shunting ratio (Qp:Qs) was 1.7. Based on the cardiopulmonary exercise test, the oxygen uptake was 22.7 ml/min/kg (74% of value according to age and sex). Conservative treatment with further clinical vigilance was chosen.

The so-called scimitar sign (a curved shadow resembling a scimitar) is present on chest X-rays of fewer than a half of patients with PAPVR,³ therefore multimodality imaging is

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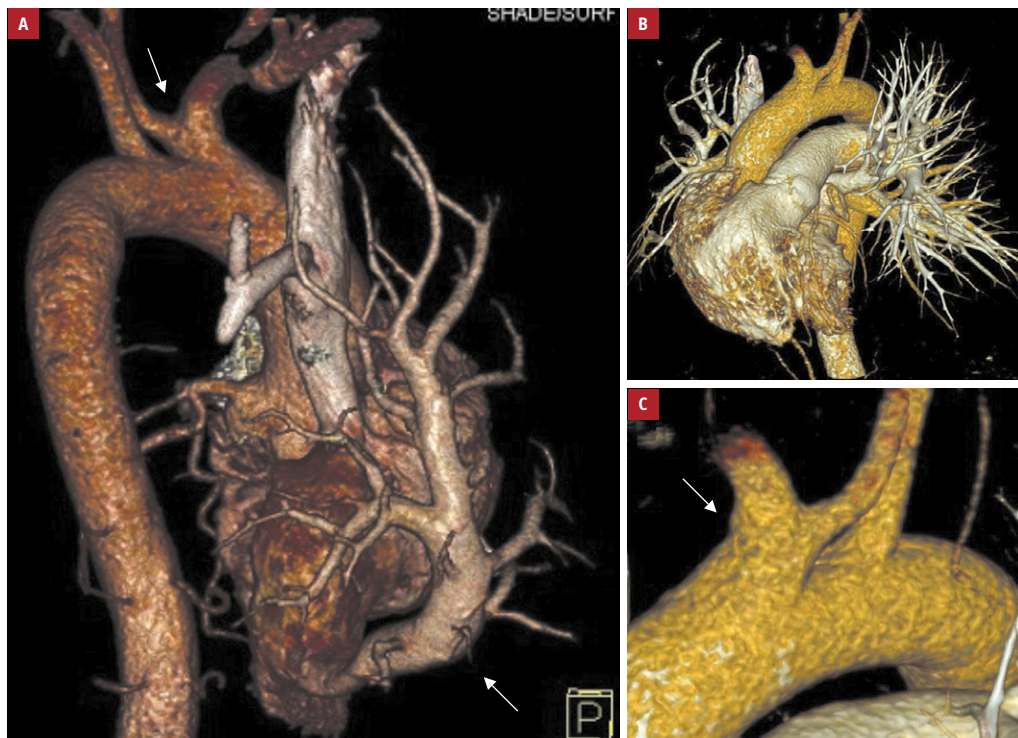


FIGURE 1 Computed tomography; **A** – the right pulmonary vein draining into the inferior vena cava (bottom arrow) and a common origin of the innominate and carotid arteries (top arrow); posterior view; **B** – the aortic arch; posteriorolateral view; **C** – enlargement of the image from panel **B** with a focus on the common origin of the innominate and carotid arteries (arrow)

recommended. Reliable assessment of pulmonary venous return by TTE may be difficult. By contrast, TEE offers a sensitive and specific evaluation. A high radiation dose during CT poses a limitation to use this modality in children and pregnant women, while the use of CMR is precluded in patients with metallic prostheses. Otherwise, both modalities have excellent value in depicting anatomical and cardiovascular relations. Indications for surgical corrections include hemodynamically significant left-to-right shunting ($Q_p:Q_s > 2$), right heart failure, or recurrent pulmonary infections.⁴ From a practical point of view, the presence of COIC may have possible advantages or disadvantages in cases of cardiac operations requiring cerebral perfusion (described in detail elsewhere)⁵. Briefly, the presence of COIC may be advantageous during the repair of aortic coarctation and aortic arch surgeries. Conversely, COIC may compromise cerebral blood flow during placement of a modified Blalock–Taussig shunt.⁵

SUPPLEMENTARY MATERIAL

Supplementary material is available at www.mp.pl/kardiologiapolska.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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