

Cardiovascular MR angiography in partial anomalous pulmonary venous connection

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DESCRIPTION

A 17-year-old girl was assessed for exercise-induced hypertension. Her medical and family history were unremarkable. After fast running during school sport she had felt unwell with headache for about an hour. Subsequently, she presented to her paediatrician who measured elevated blood pressures and decided on further diagnostic work up. The 24 h blood pressure measurements were normal, but bicycle ergometry was stopped when the patient reached a high systolic/diastolic blood pressure of 256/96 mm Hg at 2.4 W/kg. Chest radiography and subsequent MRI showed an aberrant right pulmonary vein (an infracardiac type of partial anomalous pulmonary venous connection), whereas the lung was normal (no Scimitar syndrome). At that time the patient's symptoms were minor, and she and her parents decided against diagnostic/interventional catheter angiography.

Ten years later the patient opted for further diagnostics since she now had exercise-induced dyspnoea. A temporally resolved contrast-enhanced cardiovascular MR angiography was performed. According to phase contrast imaging, the pulmonary-to-systemic flow ratio was elevated to 1.4-to-1, indicating left-to-right shunting. On the right side several small lung veins drained into an abnormal vertical vein (figure 1A, arrows) with anomalous venous return into the inferior vena cava (figure 1A, B, *). Two right pulmonary veins drained independently into the left atrium, whereas two other tortuous right pulmonary veins extended from the left atrium to the vertical vein (figure 1B, arrowheads; online supplementary figure S2; video 1). Effectively, this represented dual drainage from the vertical vein into



Video 1 Cardiovascular MR angiography, rotating volume rendering. A time-resolved three-dimensional MR angiography was performed using a dynamic TWIST (time-resolved angiography with interleaved stochastic trajectories) sequence during intravenous injection of gadolinium chelates. The rotating volume rendering was produced after subtracting a pulmonary arterial contrast phase from a systemic arterial contrast phase. In this way the right atrium, right ventricle and the pulmonary arteries are excluded. The video showing normal anatomy of the left pulmonary veins. On the right side a large abnormal pulmonary vertical vein is connected to the inferior vena cava. Two of four right pulmonary veins drain independently into the left atrium. Two other tortuous right pulmonary veins extend from the left atrium to the vertical vein.

the inferior vena cava and into the left atrium. This vascular anatomy allowed for interventional occlusion of the distal vertical vein by an Amplatzer

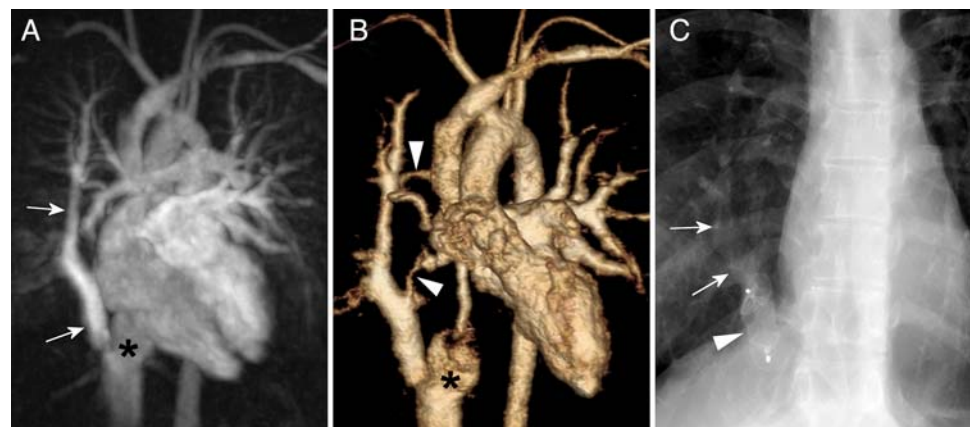


Figure 1 (A) Maximum intensity projection of the MR angiography (MRA). (B) Volume rendering of the MRA. The right atrium, right ventricle and the pulmonary arteries were excluded by subtracting the pulmonary arterial phase from the systemic arterial phase. A rotating volume rendering is supplied in video 1. (C) Chest radiography after occlusion of the distal vertical vein by a vascular plug. See full text for the description of the figure symbols ("*", arrows, and arrowheads).



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vascular plug (figure 1C, arrowhead)¹ by paediatric cardiologists after a balloon occlusion test had been successful. Subsequently, the patient was well and without dyspnoea.

In general, the indication for surgical or interventional treatment versus conservative treatment in partial anomalous pulmonary venous connection depends on individual factors, such as a patient's symptoms.^{1 2} Cardiovascular MR angiography is useful for non-invasive assessment of the associated left-to-right shunting

and for depicting the anomalous pulmonary vasculature three-dimensionally.^{2 3} Similar cases may be recognised, particularly by exercise-induced dyspnoea, where conditions with left-to-right shunting and subsequently increased right ventricular output are among the several differential diagnoses. Chest X-ray may give a hint when the aberrant vein is identified as a well-demarcated vertical shadowing (normally all pulmonary vessels are directed towards the central hilus).

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ In partial anomalous pulmonary venous connection some but not all pulmonary veins of the left or right lung recirculate oxygenated blood towards the right of the heart.
- ▶ Cardiovascular MR angiography can measure the associated left-to-right shunting and can depict the anomalous vasculature three-dimensionally.
- ▶ The clinical decision for surgical/interventional versus conservative treatment depends on individual factors, including a patient's symptoms.

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