A puzzling congenital heart disease: a late diagnosis of left atrial isomerism

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Clinical Case

• 41 years-old male, previously living abroad.

• Only known information:
  • Blalock-Thomas-Taussig shunt in the first days of live.
  • Ventricular septal defect closure and implantation of a conduct between the subpulmonary ventricle and the pulmonary artery – in 1992 (with 15 yo)

• Unknown cardiac anatomy
Clinical Case

Visceral and cardiac situs solitus, with mesocardia and levoapex; AV discordance; severe regurgitation of the systemic AV valve with systolic disfunction of the systemic RV.
Cardiac Magnetic Resonance

- Abdominal situs inversus
  - liver medial and a polylobulated spleen
- Aorta was anterior and to the right of the pulmonary artery. Right-sided aortic arch.
- Ambiguous atrial situs.
- Left-sided atrium received the venous drainage of 4 pulmonary veins and was connected to the morphological systemic RV.
- Right-sided atrium received the systemic venous drainage and was connected to the subpulmonary morphological LV.
- VA discordance and hypoplastic main pulmonary artery.
AngioCT

- Both lungs were bilobated;
- Main bronchi were hyparterial;
- Atrial appendages morphology were similar (chicken-wing);
- Polysplenia and right upper quadrant stomach.
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• Heterotaxy syndrome
  • with a dextro-transposition of the great arteries and pulmonary atresia,
  • palliated with a left-ventricle to pulmonary artery conduit
Heterotaxy Syndrome

• “An abnormal symmetry of certain viscera and veins (lungs, liver, vena cava) and situs discordance between the various segments of the heart”.

• 3–6% CHD. The cardiac condition with the highest familial recurrence rate.

• Puzzling Cardiac Anomalies
  • Importance of multimodality imaging techniques: