Clinical Image

Ostium secundum Atrial Septal Defect: Is Cyanosis Possible after Complete Surgical Correction?

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CLINICAL IMAGE

A 40 year-old female presented to the cardiology clinic complaining of progressive fatigue and effort dyspnea. Her past history was only notable for the diagnosis of an ostium secundum atrial septal defect (ASD) at the age of 14, referred for closure at the age of 16. Surgery proceeded with no complications but the patient started to complain of tiredness a few years later. Transthoracic echocardiograms performed thereafter were unremarkable, namely for residual septal defect, right chambers dilatation and elevated pulmonary artery pressures. At the age of 28 she had a full-term uneventful pregnancy. Recently she was admitted to the hospital for a community-acquired pneumonia with persistent hypoxemia, being discharged under ambulatory oxygen therapy. Both pulmonary computed tomography (CT) and V/Q lung scan were negative for the presence of pulmonary embolism.

Physical examination revealed central cyanosis and digital clubbing (Figure 1, A). In attempting to exclude an abnormal cardiac systemic venous return an additional cardiac CT angiography was performed. This exam displayed complete abnormal inferior caval vein topography, with an almost entire drainage of the vessel into the left atrium (*) (Figure B,C). There was also a prominent azygos vein and arch (arrow) (Figure D). A transesophageal echocardiogram conducted later on confirmed these findings, depicting also a small residual ASD defect during agitated saline injection into a distal vein of the inferior vena cava.

Figure 1 A: prominent digital clubbing; B–C: computed tomography (axial and sagittal views) showing abnormal inferior vena cava diversion into the left atrium; D: computed tomography (coronal and axial views) showing prominent azygos vein and arch; (E) transesophageal echocardiogram with a small residual IAS defect. (F) Schematic view (adapted from 3) illustrating when the valve of the inferior vena cava is wrongly taken for repair of the defect, draining the inferior vena cava into the left atrium.

the forearm (arrowhead) (Figure E). Afterwards the patient underwent a complete surgical correction with inferior vena cava (IVC) repositioning and residual ASC suture with a pericardial patch. Recovery was uneventful and hypoxemia correction was completely achieved.

**IVC diversion into the left atrium during surgical atrial septal defect correction by direct suture was first described in 1957 [1].** It may stem from IVC Eustachian valve inclusion into the left atrium when the surgeon is not able to look inside the atrial cavity, which may not happen with the routine use of cardiopulmonary bypass (Figure F) [2,3]. Notwithstanding the uneventful immediate postoperative course following the abnormal diversion, this condition usually subsequently presents in adult patients with insidious onset of cyanosis, effort dyspnea, finger clubbing and polycythemia [4]. As this was the case, progressive insidious symptoms came about eventually also owing to prominent azygos venous system development, returning blood to the right atrium.

Early detection of this condition may not only avoid complications related to cyanosis and persistent hypoxemia, but also prevent the ominous consequences of paradoxical embolism [5,6]. Nevertheless its recognition at transthoracic echocardiography should be difficult both without clinical suspicion and right to left shunt measurements. Either cardiac CT angiography or cardiac magnetic resonance imaging may alternatively depict this acquired rare defect [5].

**REFERENCES**