IMAGES in PAEDIATRIC CARDIOLOGY

Shiina Y,* Slavik S,* Uemura H, McCarthy KP,** Yen Ho S.** The inferior caval vein draining into the left atrial cavity – a rare case. Images Paediatr Cardiol 2011;13(4):1-5**
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Introduction

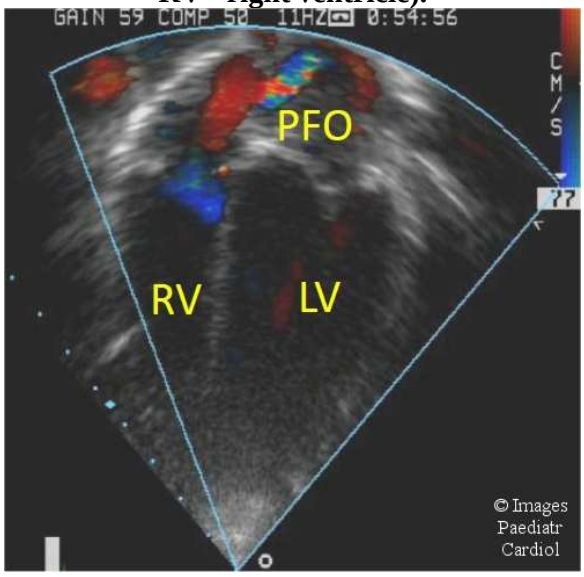
The inferior vena cava (IVC) draining into the left atrium (LA) is exceedingly rare in the setting of the usual atrial arrangement (situs solitus).[1] We describe a patient with this unique anomaly, and its repair.

Case report

A female patient aged 9 years initially developed significant exertional dyspnoea and desaturation. These symptoms, together with clubbing of fingers and toes, prompted referral for cardiovascular assessment. On physical examination her height and weight were at the 3rd centile for age (124cm and 23kg, respectively). Oxygen saturation was 91% at rest and fell to 87% with exercise. Normal volume and symmetrical peripheral pulses were present. Abdominal examination excluded hepatosplenomegaly. Chest auscultation revealed normal first and second heart sounds with no murmurs. Symmetrical air entry was heard into both lungs without added sounds. Chest x-ray and 12-lead electrocardiogram did not show any abnormal findings.

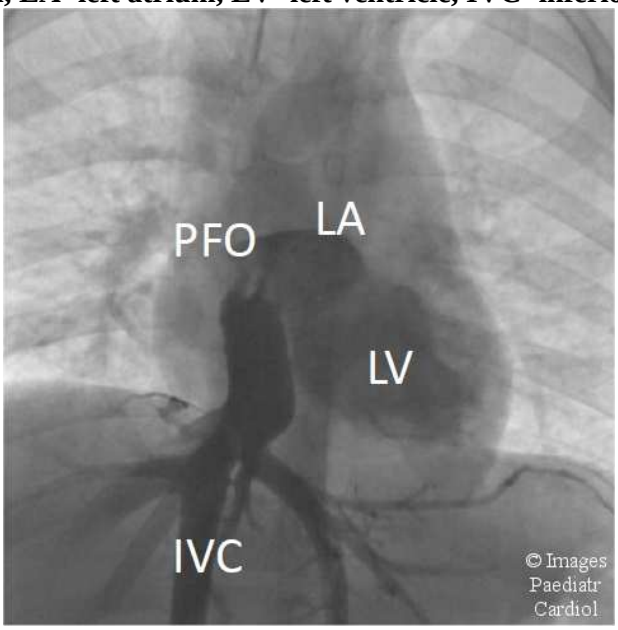
Transthoracic echocardiogram showed situs solitus with atrio-ventricular and ventriculo-arterial concordance. The IVC appeared to drain into the LA through a restrictive tunnel, although details of this route remained unclear (Figure 1).

Figure1: Transthoracic echocardiography before surgical repair. Right to left shunt through PFO was detected from the 4chamber view. (PFO=patent oval foramen, LV=left ventricle, RV= right ventricle).



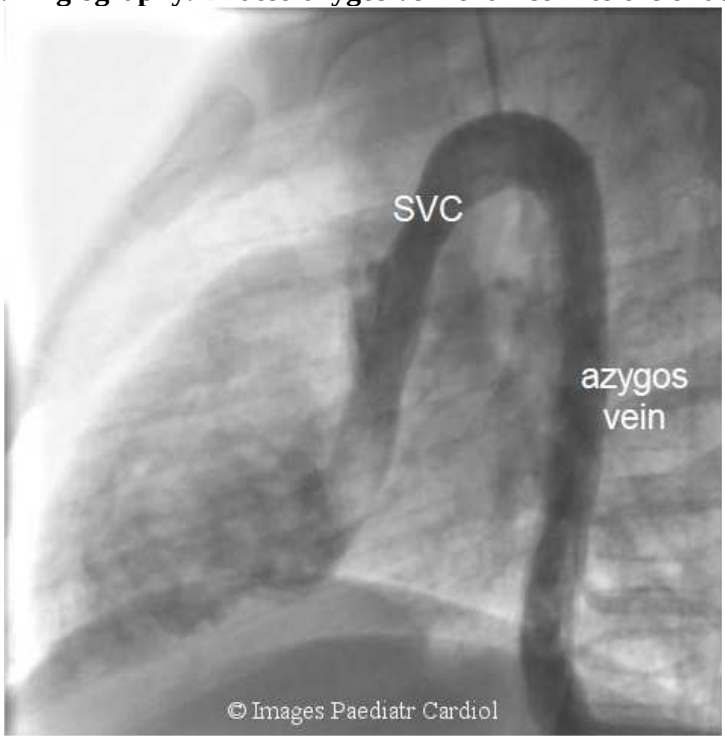
On angiography, contrast medium passed directly from the IVC into the LA through a 2mm wide opening (Figure 2). The mean IVC blood pressure was 12mmHg and mean LA pressure was 6mmHg.

Figure 2: Angiography. A catheter was passed along the IVC directly into the LA. The IVC accepted the hepatic veins and then drained into the LA through the PFO (PFO=patent oval foramen, LA=left atrium, LV=left ventricle, IVC=inferior vena).



A dilated azygos vein (9mm) drained venous blood from the lower body into a dilated (12.4mm) superior vena cava (SVC) (Figure 3).

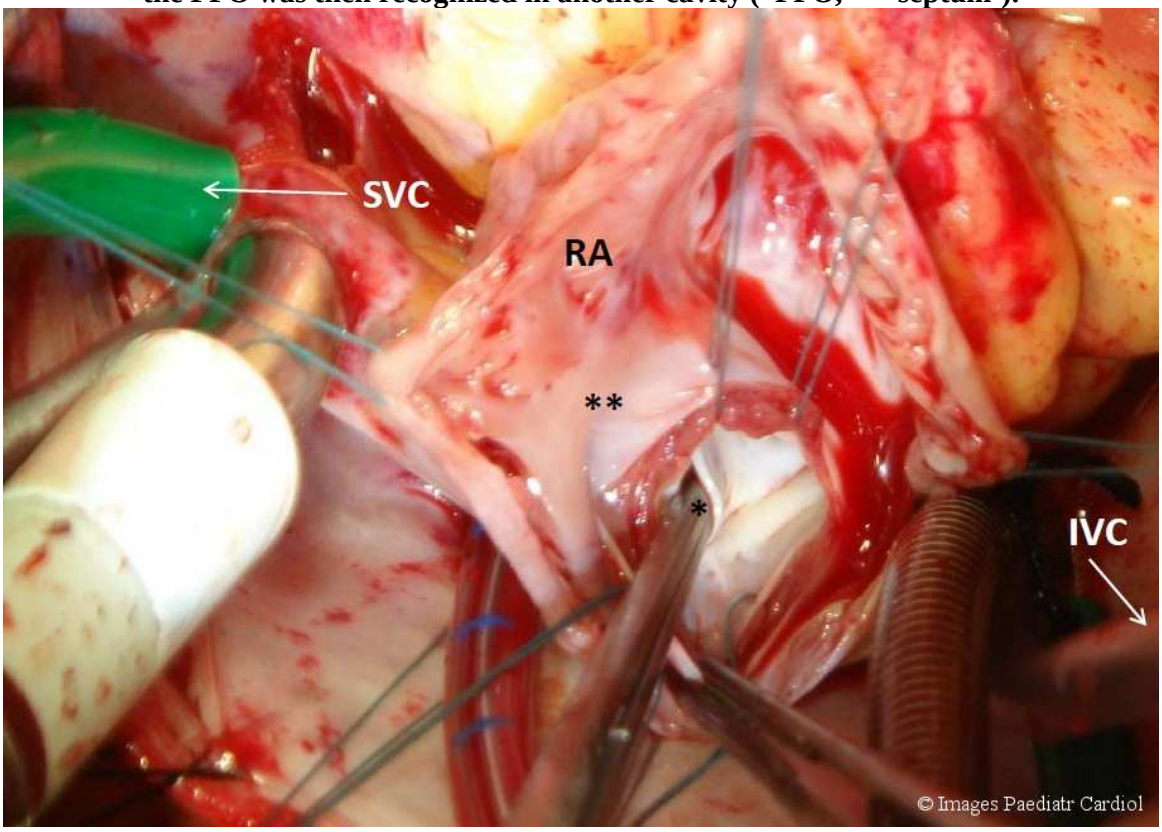
Figure 3: Angiography. Dilated azygos vein drained into the dilated SVC.



At surgery via a medial sternotomy, the epicardial aspect of the heart showed moderate hypoplasia of the right atrium (RA) and right ventricle. With the SVC and IVC directly cannulated, cardiopulmonary bypass and cardiac arrest were established in a standard fashion. When the right

atrial appendage was opened along the atrio-ventricular groove, the SVC orifice was readily identified but not the IVC orifice. There was no identifiable oval fossa present and the atrial septum was intact. The triangle of Koch was determined from the whitish colouration of the membranous septum/central fibrous body at its apex, although there was no well-formed coronary sinus or tendon of Todaro visible. Only a small orifice of the middle cardiac vein was present at the anticipated site of the coronary sinus. A longitudinal incision in the atrial septum between the putative triangle of Koch and the terminal crest revealed a cavity behind it receiving the IVC (Figure 4).

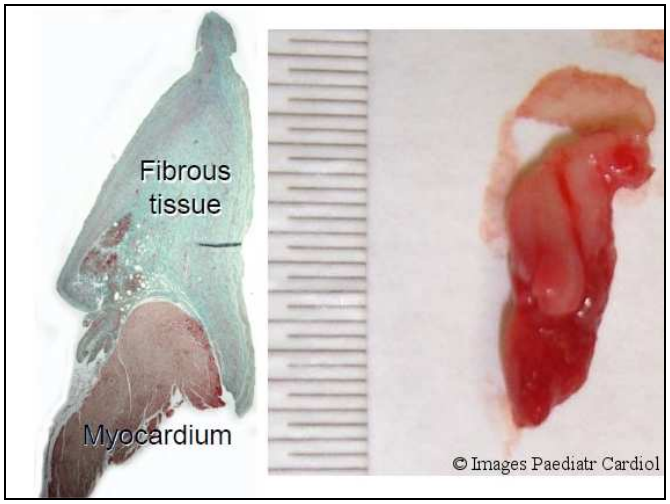
Figure 4: Intraoperative views. The left-hand side is towards the SVC and the right-hand side is towards the IVC. The huge Eustachian valve, a thick muscular layer, has been resected, and the PFO was then recognized in another cavity (*PFO, ** 'septum').



No coronary sinus orifice could be identified in this cavity. The abnormal septum between RA and the cavity receiving the IVC had the macroscopic appearance of a thick muscular layer with endothelium on both sides. A thick fibrous membrane with the appearance of flap mechanism of a patent fossa ovalis (PFO) lined the other side of the cavity. The flap mechanism, however, was different from a typical PFO, because the upper margin was formed by a membranous structure rather than an muscular infold as seen in the normally structured heart. The PFO was closed with direct sutures. The abnormal muscular septum was resected as much as possible allowing the IVC to open widely into the right atrial cavity leading to the vestibule of the tricuspid valve. The area of the putative triangle of Koch was entirely preserved; the abnormal muscular septum and the fibrous oval fossa membrane fused at the expected site of the tendon of Todaro.

On cross section, the resected septum was 4mm at its thickest, tapering to 2mm. Histology revealed that the thick end was composed of atrial myocardium covered with a thin layer of endocardium. Toward the thin end, the myocardium was capped with thick deposition of fibrous tissue (Figure 5).

Figure 5: Histology. 'Septum' was resected and stained with Masson's trichrome (x16magnification). Toward the thin end, the myocardium was capped with thick deposition of fibrous tissue.



This patient had a smooth and uneventful post-operative course. Peripheral oxygen saturation normalized and her shortness of breath improved at 3 months follow-up. During follow up of 5 years, no evidence of arrhythmia or IVC obstruction was noted.

Discussion

Most case reports regarding such anomalous connection of the IVC to the LA indicated an abnormally large right valve of the sinus venous, the Eustachian valve, coexisting with an atrial septal defect or PFO.[2,3] The persistence of a large Eustachian valve in various forms is a recognized entity and, when coexisting with an interatrial defect, can lead to a right to left shunt resulting in a clinically detectable cyanosis. This right valve of the sinus venosus contributes to the inferior border of the atrial septum. When extensive, it can almost divide the RA into two chambers resulting in right-sided cor triatriatum.[4]

During early embryologic development, the right and left venous valves separate the systemic sinus venosus from the primary part of the RA. The left venous valve subsequently fuses with the developing atrial septum. As the systemic sinus venosus becomes incorporated into the RA, the right venous valve diminishes. Its rudiments usually become the crista terminalis, the valve of the IVC (Eustachian valve) and the valve of the coronary sinus (Thebesian valve).[5] However, abnormal regression of the right valve of the sinus venosus may be present occasionally. In our case, muscularisation instead of the normal absorption possibly occurred at the portion of the right valve of the sinus venosus resulting in the large Eustachian valve diverting the blood stream from the IVC into the LA through the PFO. Considering the histological composition of the abnormal septum, it is possible that initially there was a communication between the IVC cavity and the RA as cor triatriatum dextrum but the opening became obliterated by deposition of fibrous tissue at its margins.

Regarding surgical repair, some case reports indicated that the prominent Eustachian valve was intraoperatively misinterpreted as an atrial septal defect and surgically closed, even though there was an experienced cardiac surgical team at work .[6] Detailed imaging of the IVC drainage anatomy using angiography, CT and/or magnetic resonance imaging is therefore mandatory in cases where clinical and echocardiographic suspicion of drainage anomaly is present.

Conclusions

We present a rare congenital malformation involving anomalous drainage of the IVC into the LA. A high level of clinical suspicion in otherwise unexplained cyanosis and detailed imaging of the abnormal drainage anatomy can lead to accurate diagnosis and successful surgical treatment.

References

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