Atrial septal defect closure complicated by anomalous inferior vena cava return to the left atrium: a case report of a 5-year-old child

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Received 8 August 2018; accepted 16 May 2019; online publish-ahead-of-print 24 May 2019

Background
The anatomy of the sinus venosus atrial septal defect (ASD) of the inferior vena caval type is complex. Limited rate of complications during its closure has been described. One of the unusual complications, with few case reports, is the iatrogenic diversion of the inferior vena cava (IVC) to the left atrium (LA).

Case summary
We report the case of a 5-year-old boy who underwent previous surgical closure of sinus venosus ASD of the inferior vena caval type aged 2 years. Three years after surgery, he was diagnosed as having iatrogenic diversion of the IVC to the LA.

Discussion
Cardiologists and cardiac surgeons should be aware of this rare but significant complication following routine ASD surgical repair and vigilant follow-up should be performed routinely.

Keywords
Case report • Atrial septal defect • Inferior vena cava • Left atrium • Surgical ASD closure

Learning points
• Cardiologists and cardiac surgeons should be aware of iatrogenic diversion of the inferior vena cava (IVC) as a rare but important complication of surgical atrial septal defect repair. They should have high suspicion of this iatrogenic diversion of IVC to left atrium when saturations are low or low normal.
• Non-invasive imagines are excellent tools to confirm the diagnosis. Long-standing cyanosis requires detailed workup and careful evaluation.

Introduction
An atrial septal defect (ASD) of the inferior vena caval type has a complex anatomy for the surgeon. Limited rate of complications during its closure has been described. Arrhythmias and patch dehiscence appear to be the main long-term complications after ASD closure. Right to left shunting is an infrequent complication of repaired acyanotic congenital heart disease. However, iatrogenic diversion of the inferior vena cava (IVC) to the left atrium (LA) has been reported. A communication of the IVC directly with the LA is seen due to the absence of lower margin of the defect. During
surgery, the Eustachian valve could erroneously be considered as the margin of the ASD, and this complication has particularly been described before the era of cardiopulmonary bypass.\(^2\) We report a case of sinus venosus ASD repair of the inferior vena caval type with an unintentional diversion of the inferior vena caval blood to the LA, diagnosed incidentally 3 years following surgery.

**Timeline**

| Event                  | Date               |
|------------------------|--------------------|
| Diagnosed to have atrial septal defect (ASD) | At 1 year of age |
| Surgical closure of ASD | At 2 years of age  |
| Incidental finding of desaturation/hypoxia on a routine anaesthetic assessment prior to planned dental procedure (92%) | At 5 years of age—March 2018 |
| Inferior vena cava drainage to the left atrium seen on echocardiography and computed tomography angiography scan | At 5 years of age—March 2018 |
| Complete surgical repair—saturation 100% | At 5 years of age—June 2018 |
| Discharged home | At 5 years of age—July 2018 |
| Latest follow-up | At 5 years of age—July 2018 |

**Case presentation**

**Patient information**

A 5-year-old boy was referred to the Children’s heart centre at AUBMC after detecting a saturation of 92% while being screened by the anaesthesia team preceding a dental procedure. Patient was asymptomatic, denied any chest discomfort, dyspnoea, episodes of altered mental status, and other symptoms of hyperviscosity. His medical history, reported by the parents, revealed open-heart surgery for ASD closure in another country at the age of 2 years. Parents informed the medical staff that he had a smooth post-operative course and was discharged home after few days; however, no documentation was available. Follow-up after his surgical closure was done routinely showing good results and no abnormalities.

His assessment in our clinic revealed \(O_2\) saturation ranging from 90% to 94%, blood pressure 105/68, and respiratory rate 25. The patient had no dysmorphic features. His physical exam revealed minimal central cyanosis, normal apical impulse no right ventricular heaves or thrills, normal first and second heart sounds, and absence clubbing.

**Diagnostic assessment**

The electrocardiogram and chest X-ray were within normal limits. Echocardiogram showed a small fenestration through the ASD patch with a left to right shunt across it. Normal atrioventricular and normal ventriculoarterial concordance, left and right normal ventricular function and dimensions, no secondary signs of pulmonary hypertension. The superior vena cava (SVC) was draining into the right atrium. The pulmonary veins were draining normally into the LA, and the estimated pulmonary systolic and diastolic pressures were normal. However, the IVC flow was not directed into the right atrium (Figures 1 and 2, Supplementary material online, Video S1), and we suspected that the drainage was directed to LA. Pulmonary to systemic flow ratio was 0.6. A chest computed tomography was requested and the result confirmed our diagnosis (Figure 3). A multidisciplinary meeting was held with the surgeon and a decision was taken to perform a surgical repair.

**Interventions**

Transoesophageal echocardiographies are performed routinely at our institution prior and after cardiothoracic surgeries looking for

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optimal surgical results. A transoesophageal study was done in the operating room immediately before surgery confirming our findings. A bicaval view was also taken showing the above mentioned findings (Figure 4). Our patient had two central venous lines, one in his jugular vein and the second in the femoral vein. Central pressures were taken showing SVC pressure of 7 mmHg as compared to an IVC pressure of 13 mmHg. Saline was injected in the femoral venous line during the transthoracic echocardiography allowing the visualization of micro-bubbles in the LA and subsequently in the other chambers (Figures 5 and 6, Supplementary material online, Video S2). The intra-operative finding of aberrant IVC drainage to the LA was confirmed, and the IVC was re-implanted into the right atrium. The post-operative course was smooth, and the patient was discharged 5 days later having an SpO2 of 100%. One week later, during a routine follow-up, the patient was recovering well, fully saturated and had good surgical result confirmed by echocardiography.

Discussion

Atrial septal defects originate at particular sites in the atrial septum and are named according to their embryonic origin. The secundum type ASD occurs in the central part of the atrial septum (fossa ovalis) as a result of deficient growth of septum secundum, deficient valve tissue, ectopic, or excessive resorption of septum primum. The sinus venosus type occurs outside the margins of the fossa ovalis in relationship to the venous connections of the right atrium in relationship to the IVC or SVC. The IVC-type sinus venosus ASD, occur inferior...
to the fossa ovalis in relation of the IVC and the right lower pulmonary venous orifice. Others include those in the region of endocardial cushion (primum ASD) and in the ostium of coronary sinus (coronary sinus ASD).  

Similar earlier cases have been reported with iatrogenic diversion of IVC flow to the LA after surgical closure of sinus venosus ASD of the IVC type. This complication was more frequent before the use of cardiopulmonary bypass due to time limitations. The anatomy of the ASD of the IVC type was first described by Rokitansky. He specified that this defect does not have lower septal margin; therefore, the ASD of the IVC type was first described by Rokitansky.4 He specified that this defect does not have lower septal margin; therefore, the opening of the IVC is directed partly into the LA. In addition, Bedford et al reported that three cases of the deviation of the IVC to the LA during a surgical closure of a low lying secundum ASD occurring by direct suture using circulatory arrest under hypothermia. Bjork et al described the operation and management of a case after diversion of the IVC into the LA post-open repair of an ASD. The complication described by Bedford et al. and Bjork et al occurred during or immediately after operation. Mustard, Shuntoh et al, Desnick et al, and Effler and Groves also described this complication that had an unremarkable post-operative course and insidious onset of dyspnoea mainly with exercise and cyanosis. Ross and Johnson stated that under tension the repair may disrupt post-operatively.

Partial diversion of the IVC to the LA can result in reduced level of cyanosis. Paradoxical embolism is also one of the consequences of this complication. Alanbaei et al, Fouty et al, and Munet et al described these finding in adults who had surgical closure of an ASD during childhood. Some of them were misdiagnosed to have Eisenmenger’s syndrome.

With the advances of cardiac imaging and non-invasive modalities, the diagnosis of this condition is being diagnosed accurately can be done accurately without the need for contrast echocardiogram or cardiac catheterization and at an earlier stage. The incidental finding of desaturation while screening our asymptomatic patient for a dental procedure is interesting. In our case, transcatheter closure was possible by inserting a stent from the IVC into the LA and then into the right atrium via the fenestration. However, we decided to go for surgical repair considering the child’s young age, the complication rate, and the need for a lifetime anticoagulation therapy.

**Conclusion**

In conclusion, cardiologists and cardiac surgeons should be aware of this rare but significant complication following a routine ASD surgical repair. It is also important to have a high index of suspicion of this iatrogenic diversion of IVC to the LA when saturations are low or low normal. Non-invasive imagines are excellent tools to confirm the diagnosis. Long-standing cyanosis requires detailed workup and careful point by point analysis.

**Supplementary material**

Supplementary material is available at European Heart Journal - Case Reports online.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of Interest:** none declared.

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