

Iatrogenic cyanosis in children, a pitfall of atrial septal defect repair

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ABSTRACT

Today surgical repair of different types of atrial septal defect (ASD) is considered as relatively a simple and safe procedure with minor to nil morbidity and mortality. When a complication occurs after ASD repair, is associated with considerable morbidity and mortality, which is much higher than primary surgical procedure. We report two patients that underwent ASD repair with caval inflow occlusion without CPB with late presentations of iatrogenic diversion of inferior vena cava to left atrium and subsequent complications of right to left shunting.

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Introduction

Atrial septal defects (ASDs) constitute 10% of all congenital cardiac anomalies with a female preponderance. Secundum type of ASD forms seventy percent of cases. ASD is a defect of variable size and is usually located at the fossa ovalis position but sometimes extends posteriorly or towards the inferior vena cava (IVC) and characteristically presents with left to right shunts (1). Anecdotal reports of surgical ASD closure dates back to 1948 that operation has been performed with indirect palpation of ASD rims until 1952 that procedure was done using caval inflow occlusion technique with systemic hypothermia, and during 70 years of operation experience, surgery is usually

performed with minor mortality or morbidity (1). Major surgical complications can include bleeding, dysrhythmia, infection, pneumothorax, pleural effusions and pericardial effusions but are usually benign and temporary. ASD can be surgically closed with either direct suturing technique or using a pericardial or synthetic patch. Although surgical closure of secundum ASD is a safe procedure with near-zero mortality, nevertheless, iatrogenic diversion of the IVC blood stream to the LA has been described as a rare complication of repaired ASD causing right to left shunting (2,3). This complication occurs when the Eustachian valve is wrongly considered as the lower rim of the septal defect and this event has mostly been

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reported before the invention of cardiopulmonary bypass (CPB).

Herein, we report two patients that underwent ASD repair with caval inflow occlusion without CPB, with iatrogenic diversion of inferior vena cava to left atrium and subsequent complications of right to left shunting.

Case Reports

The first case is an 8 years old boy who had been undergone surgical repair of ASD at the age 2. The ASD repair has been performed with caval inflow occlusion without CPB and he was discharged home with uneventful course. Post-operative transthoracic echocardiography showed no considerable abnormalities. Six years later after surgery, the patient presented with cyanosis and clubbing. Transthoracic echocardiography revealed residual ASD and partial anomalous connection (PAPVC) of right pulmonary veins (RPVs) to the right atrium (RA) (Figure 1)

Cardiac catheterization showed that IVC flow was directed to the left atrium (LA), along with the presence of PAPVC and moderate pulmonary hypertension (PH). The patient scheduled for redo reoperation and complete diversion of IVC to RA and RPVs to LA was performed with success.

The second case is a twenty years old female with previous surgical history of ASD closure when she was 6. The operation has been performed with caval inflow occlusion without CPB. Post-operative transthoracic echocardiography has revealed residual shunt with recommendation of further follow-up. Eight years later, the patient was referred to cardiologist for dyspnea of exertion, cyanosis and clubbing. Contrast echocardiography showed bubble pacification of LA and LV from IVC which made the suspicion of inadvertent diversion of inferior vena cava to left atrium during the first procedure. Cardiac MRI also proved the diagnosis and showed the predominance of IVC flow to LA rather than RA (Figure 2). The patient was taken to the operation room for surgical revision of previous repair. Redo operation was performed with complete revision of previous repair for diversion of IVC to RA with successful result.

Discussion

Intracardiac repair of a simple secundum ASD is reasonably straightforward. The right atrium (RA) incision is done transversely or longitudinally and ASD repair is done by either direct suturing of ASD rims or applying a synthetic or pericardial patch, according to the defect size and the preference of surgeon (4).

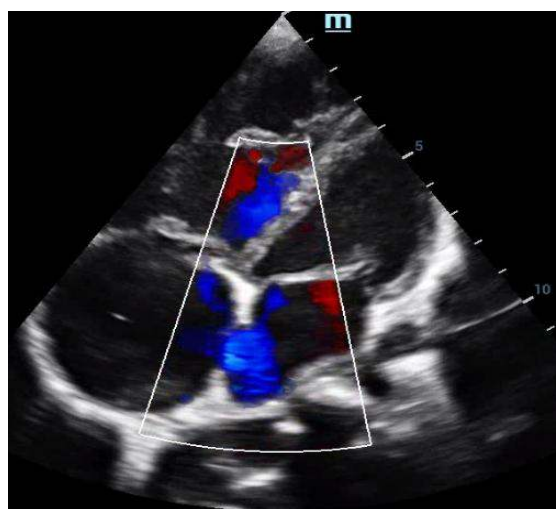


Figure 1: Transthoracic echocardiography showing diversion of IVC flow to left atrium.



Figure 2: Cardiac MRI showing the predominance of IVC flow to LA rather than RA.

Simple repair of ASD is commonly applied for smaller defects, whereas larger defects are repaired with patch. Primary closure has the likelihood of suture dehiscence and reopening of defect, whereas patch repair could be associated with residual shunt (5, 6). Surgical complication after ASD repair is associated with considerable morbidity and mortality, which is much higher than primary surgical ASD repair.

Anatomically in cases of large absence of the fossa ovalis floor, a larger fossa ovalis defect is present. In severe absence of fossa ovalis tissue, the defect may be confluent with the entrance of the IVC. The Eustachian valve of the IVC then overhangs the septal defect and must not be mistaken for its lower margin at operation. It is important for the surgeon to distinguish this type of ASD because if the large Eustachian valve is erroneously taken as the inferior edge of ASD to the upper septal margin, the IVC might then be connected partially or completely to the LA (7).

Inadvertent iatrogenic diversion of inferior vena cava to left atrium during ASD repair is a very rare complication. This complication is first described by Bjork in 1958 after ASD repair when hypothermia had been used with caval inflow occlusion to RA (8). Such complications would have been more common before the invention of cardiopulmonary bypass (CPB) because the patients used to be operated with inflow occlusion and systemic hypothermia (1).

Ross and Johnson reported that a distortion of the IVC orifice during caval occlusion by the snare in combination with a large Eustachian valve may lead to error in defect closure and iatrogenic right to left shunt (9, 10). Complete or partial diversion of the inferior vena cava to left atrium leads to fall in oxygen saturation, cyanosis and possible further paradoxical embolism.

We described two cases of ostium secundum ASD repair surgery with inflow occlusion technique which remained undiagnosed early after surgery with late presentation of cyanosis and pulmonary hypertension.

Few such cases have been reported after few years of surgery also (10). Whenever oxygen desaturation is noticed immediately after ASD repair or presence of unexpected pulmonary hypertension during follow-up this complication should be suspected. Intra-operative TEE is a helpful diagnostic tool for early detection and correction of this complication without morbidity and mortality. Cardiac MRI is also an accurate imaging modality to evaluate the cardiac anatomy, function and flow stream for assessment of intra-cardiac repair and possible complications.

In conclusion, great care should be taken during ASD repair to avoid confusing the Eustachian valve of the inferior vena cava with the remnant of the floor of the fossa ovalis. Such an uncommon error results in connecting the IVC to the LA, which can occur when the operation is done under circulatory arrest and without IVC cannula, when direct caval cannulation is used or during caval inflow occlusion. Careful echocardiographic study before and after ASD repair by the expert cardiologist is also an important point to prevent this complication.

Competing Interests

The authors declare that they have no competing interests.

Authors' Contributions

All authors have made substantial contribution to concept this paper.

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