Iatrogenic diversion of inferior vena cava to the left atrium presented as recurrent foetal loss: a case report

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Background
Iatrogenic diversion of inferior vena cava (IVC) to the left atrium (LA) after atrial septal defect repair (ASD) is an unusual complication. It rarely occurred nowadays due to trans-oesophageal echocardiography (TEE) check during surgery, but there are still few numbers of patients who survived from an old operation during childhood and reached adulthood undiagnosed.

Case summary
We present a 27-year-old female post ASD repair in childhood with a unique presentation of recurrent abortion in adulthood besides exertional dyspnoea. A full workup of investigations was normal except for haemoglobin of 21 and oxygen saturation of 70%. TEE revealed abnormal drainage of IVC to the LA. Surgical correction was done to release the IVC opening to drain in the right atrium and the oxygen saturation reached 99% after weaning from the bypass machine. The postoperative course was uneventful, and the patient was discharged 5 days later. Two years later, she got pregnant twice and completed her pregnancies to term with well-developed infants.

Discussion
The diversion of the IVC may be either complete or partial. Such complications may result in intraoperative death on the table or may present as early desaturation, shortness of breath, cyanosis, and clubbing, or it may present with such symptoms in adulthood. Rarely, it may present with cerebral stroke. Our case presented with the unexpected presentation of recurrent abortion. So, even if it is rare, echocardiography should be considered as a workup for recurrent abortion in a patient with a history of congenital heart surgery.

Keywords
Inferior vena cava • Atrial septal defect • Polycythaemia • Abortion • Case report

ESC Curriculum
2.2 Echocardiography • 7.5 Cardiac surgery • 9.7 Adult congenital heart disease

Learning points
- Diversion of IVC to the LA is an uncommon complication of ASD repair and presents as early desaturation, shortness of breath, and cyanosis.
- Aside of all the common presentations, we suggest that recurrent abortions may be a form of manifestation of IVC diversion.
- Hypoxaemia will increase Red Blood Cells (RBC) production and blood viscosity; the RBCs will lodge in placenta microvessels, hence placental thrombosis and abortion.
Introduction

Atrial septal defect (ASD) is the third most common congenital heart anomaly, and surgical repair is considered relatively simple and safe. Nevertheless, various complications have been reported including arrhythmias, heart block, mediastinal bleeding, infective endocarditis, and transient ischaemic attacks or strokes. On very rare occasions, erroneous diversion of the inferior vena cava (IVC) to the left atrium (LA) could occur during surgery especially when there is no inferior rim of the ASD. Such cases can be missed due to the absence of routine intraoperative trans-oesophageal echocardiography (TEE). These cases are either presented early with desaturation and death intraoperatively or late with exertional dyspnoea and cyanosis. We report a case of diversion of IVC to the LA presented with a unique late complication of recurrent abortion. The patient was successfully managed by surgical intervention.

Timeline

| Patient | Age | Event |
|---------|-----|-------|
| 3 years old | Atrial septal defect closure surgery. |
| 3–9 years old | Limitation of activity associated with cyanosis. |
| 9 years old | Seen by a doctor and advised her to have another operation, but the family refused another surgery. |
| 9–22 years old | She tolerated her symptoms with limitation of activity and cyanosis. |
| 22 years old | She got married. |
| 22–27 years old | Had five recurrent abortions. Investigation showed haemoglobin 21 and low oxygen saturation of 70%. |
| 27 years old | Echocardiography revealed abnormal drainage of inferior vena cava (IVC) to the right atrium. |
| 27 years old | Surgical repair was done by using vascular patch to close the defect in the septum thus making the IVC orifice in the right atrium. |
| Post-operative period | Was uneventful and patient was discharged 5 days later. |
| 3 months later follow-up | Echocardiography revealed normal drainage of IVC to the right atrium. Oxygen saturation 99% and haemoglobin dropped to 13.5. |
| 28 years old | Got pregnant and had birth to a term with well-developed infant of 3500 g weight. |
| 29 years old | Got pregnant again and had birth to a term with well-developed infant of 3700 g weight. |
| 30 years old | Routine follow up showed unlimited exercise tolerance and oxygen saturation 99%. |

Case report

A 27-year-old female patient with a known history of ASD closure was referred to our department due to abnormal drainage of IVC to the LA.

Since her operation at the age of 3, she had a gradual limitation of activity associated with cyanosis. At the age of 9, she was advised to have another operation, but unfortunately, her family refused. Over the years, the patient tolerated her symptoms until she got married 5 years before presentation. Since then, she has had five recurrent abortions. Each pregnancy reached 10–13 weeks gestational age and then intrauterine death occurred. The last abortion was 6 months before presentation despite anticoagulant therapy including enoxaparin and aspirin.

A full workup of investigations was done including chromosomal analysis, thrombophilia, protein C, protein S, anti-phospholipid, anti-cardiolipin, toxoplasma antibody, and genetic study. All her investigations were normal except for the presence of polycythaemia with haemoglobin 21 g/dL and low oxygen saturation.

TEE revealed a small diameter of right atrium RA (2.5 cm) and right ventricle RV (2.0 cm). Both LA and left ventricle (LV) were dilated with an end-systolic diameter of 6 and 6.9 cm, respectively. Abnormal drainage of IVC to the LA was seen. No evidence of intra atrial shunt was detected. This was verified by injection of a 10 cc contrast air bubble through the femoral vein (Figure 1, see Supplementary material online, Video 1). Right-sided heart catheterization confirmed the above findings with decreased RA and RV pressure and increase LA pressure (Figure 2, see Supplementary material online, Video 2).

When presented to our department, she had central cyanosis with oxygen saturation of 75%. Blood pressure was normal with a regular pulse of 75 beats per minute. Heart and chest examinations were normal except for a 2/6 systolic ejection murmur without fixed splitting of S2. Besides central cyanosis and finger clubbing, her general examination was unremarkable. Routine laboratory investigations were all normal except for a high haemoglobin level of 21 g/dL, haematoctrit of 63%, and a high red blood cell of 7.5 million cells per micro-liter. Chest x-ray showed cardiomegaly with decreased pulmonary vascular markings.

In view of the present findings, a decision was made to perform surgery. The heart was exposed through a median sternotomy. After dissection of adhesions, cardiopulmonary bypass was instituted through cannulation of the aorta, superior vena cava, and right femoral vein. Exposure of the RA revealed a small cavity with no evidence of inter-atrial communication, and the orifice of IVC was not seen.

The septum was incised along the previous surgical incision, and the orifice of IVC was seen draining completely into the LA (Figure 3). A polytetrafluoroethylene patch was used to close the intra-atrial septum making the IVC orifice in the RA.

After surgical correction, oxygen saturation reached 99%. TEE showed no residual shunts. Cardiac dimensions were as follows: RA (3.5 cm), RV

Figure 1 Apical four chambers view of trans-esophageal echocardiography demonstrating appearance of agitated saline in the left atrium and subsequently in the left ventricle without appearance in the right chambers of the heart.
Iatrogenic diversion of IVC to the LA (2.9 cm), LA (4.8), and LV was 6 cm. Both operative and postoperative recovery were uneventful. The patient was discharged after 5 days with normal oxygen saturation. Follow-up echocardiography revealed normal drainage of IVC to the RA (Figure 4, see Supplementary material online, Video 3).

The patient had two full-term pregnancies 1 and 2 years later after the surgery with normal delivery of well-developed infants weighing 3500 and 3700 g, respectively. During that period, no history of abortion was encountered.

Discussion

Diversion of IVC to the LA is a very rare complication of surgical ASD closure. This could occur when there is no inferior rim of the ASD, and the eustachian valve, which is the embryonic remnant of the IVC valve, could be mistaken for the lower margin of the ASD (Figure 5B). The use of intraoperative TEE helps in the detection and immediate repair of this complication. The presentation of this complication may vary according to the type of diversion. A complete diversion of IVC to the LA may happen as a result. This may cause acute arterial desaturation which may be noted intraoperatively and if left uncorrected may result in immediate demise of the patient. Or it may be a partial diversion with part of IVC in the LA and the other part in the RA, where patients may present with digital clubbing, exertional dyspnoea, and secondary polycythaemia.

Diversion of IVC to the LA results in right to left shunt, with deoxygenated blood bypassing the RA, RV, and the lung to the LA. Thus, the mixed deoxygenated and oxygenated blood in the LA leads to a decrease in oxygen saturation. Subsequently, the patient became hypoxic and cyanosed, leading to an increase in the production of erythropoietin, red cell production, and increased blood viscosity with a significant risk of paradoxical embolism and cerebral strokes.

Our case presented with recurrent unexplained abortions during the first trimester of pregnancy. Each pregnancy reached 10–13 weeks gestational age and then intrauterine death occurred, and the last abortion was 6 months before presentation. She was fully investigated for the aetiology of abortions including chromosomal analysis, thrombophilia, and genetic study, but all investigations were normal. The patient was prescribed anticoagulant therapy (enoxaparin and aspirin) during the pregnancy with no response, with subsequent abortion.

Chronic maternal hypoxia retards foetal growth and leads to the delivery of a newborn with intrauterine growth restriction, while polycythaemia will increase the risk of thrombosis and abortion.

The patient presented with a haematocrit of 65 and a high red blood cell count, explaining the cause of repeated abortions, as elevated haematocrit leads to high viscosity, so when blood enters the placenta microcirculation, RBCs lodge in the microvessels leading to placental thrombosis and eventually abortion (Figure 5A).

The right to left shunt resulted in small RA and RV with dilated LA, a follow-up TEE 6 months after the surgery showed normalization of the previously dilated LA, small RA, and RV with no gradient across the tricuspid valve.

One year after discharge from the hospital, the patient was completely asymptomatic with unlimited exercise activity and increase oxygen saturation up to 99%. She became pregnant twice within 2 years after the surgery, delivering full-grown newborns with no history of abortions.
Figure 5 (A) Normal red blood cells count and normal blood viscosity circulate smoothly in the placenta microcirculation, while in patients with polycythaemia high red blood cell count and high viscosity make it difficult to pass through the placenta microcirculation, therefore lodge inside the microvessels and occlude them. (B1) Atrial septal defect repair with no inferior rim: the inferior vena cava orifice is very close to the atrial septal defect repair making it the lower margin of the atrial septal defect repair. (B2) The patch closes the atrial septal defect repair with its lower margin taking the inferior vena cava orifice and making the inferior vena cava drain in the left atrium. (B3) The patch closes the atrial septal defect repair with its lower margin left to the inferior vena cava orifice, making it draining in the right atrium.

Conclusion

The diversion of IVC to the LA after ASD repair is a serious complication. It could lead to multiple abortions among female patients. To avoid such complications, surgeons should be cautious about the inferior margin of the ASD during repair and use intraoperative TEE routinely.

Supplementary material

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

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

Consent: The authors confirm that written consent for submission and publication of this case report, including images and associated text, has been obtained from the patient in line with COPE guidance.

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