Atresia of the inferior vena cava in a patient undergoing mitral and tricuspid valve surgery

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INTRODUCTION

The interrupted inferior vena cava (IVC) with azygos continuation is a rare congenital anomaly with a prevalence of approximately 0.3% in otherwise healthy individuals and 0.6–2% in patients with congenital cardiovascular defects. The anomaly is frequently associated with a recurrent deep venous thrombosis, sick sinus syndrome and/or the polysplenia syndrome and is typically detected in early adulthood.

The interrupted IVC is usually terminated above the renal vein and below the hepatic vein, and systemic venous flow is accomplished by the dilated azygos and hemiazygos veins that eventually drain into the superior vena cava (SVC), intrathoracic or anomalous intrahepatic veins.

The anomaly, if unknown, can cause procedural complications during interventional procedures. Therefore, it is of utmost importance to be aware of the IVC anomaly prior to the performance of any such procedure.

We report on a patient with the interrupted IVC during a minimally invasive intervention to replace a diseased mitral valve with concomitant tricuspid insufficiency. In the literature, there is no such case but one, in which a 22-year-old woman with severe mitral stenosis and severe tricuspid regurgitation in the presence of the interrupted IVC had successful retrograde non-transseptal balloon mitral valvotomy performed.

CASE REPORT

A 61-year-old man presented with dyspnoea New York Heart Association Class III–IV and oedema in both legs and episodes of presyncope. Echocardiography revealed a degenerative disease with mitral valve regurgitation III° with annulus dilatation and a restrictive posterior mitral valve leaflet. There was a concomitant tricuspid valve regurgitation III° and a left ventricular ejection fraction of 35%.

The patient was scheduled to undergo minimally invasive surgery. For this purpose, we forwarded the venous wire through the femoral vein under transoesophageal echocardiography (TOE) control. At 20–25 cm, at the level of the diaphragm, we were not able to further advance the wire, failing to reach the right atrium or ventricle. We were not able to capture the reason for our inability to advance the catheter during the intraoperative TOE. Fluoroscopy with a mobile C-arm, which may prove useful in these cases, was not available.

Subsequently, we decided to proceed with an open sternotomy. Arterial cannulation was performed via the ascending aorta and venous cannulation via the SVC and the residual IVC (2–3 mm), which drained into the suprahepatic veins to establish cardiopulmonary bypass. Mitral valve disease together with a retraction of the anterior and posterior leaflets required a replacement with a 33-mm biological valve. Further, the annulus of the tricuspid valve was dilated, and an open annuloplasty ring (34 mm) was implanted. TOE demonstrated fully functional and competent mitral- and tricuspid valves (Video 1).

To further elucidate the reasons for our inability to advance the venous wire into the right atrium or ventricle, computed tomography (CT) was performed. It demonstrated atresia of the IVC (Fig. 1) but no signs of abdominal thrombosis or tumour. Multislice CT (Somatom force) of the thoraco-abdominal region revealed atresia of the inferior vena cava.
revealed atresia of the IVC at the diaphragm level with pronounced paravertebral venous collaterals with the no-persistent left-sided SVC and draining via enlarged azygos or hemiazygos veins into the SVC (Fig. 2). Reconstruction of the abdominal and thoracic venous system in a serial axial CT showed a normal infrarenal IVC with venous flow from the inferior body regions drained via large paravertebral collaterals into the dilated azygos and hemiazygos veins and the SVC, while a segment connecting the IVC to the hepatic vein and right atrium was missing. The hepatic veins drained directly into the right atrium.

**DISCUSSION**

Although being infrequent, the asymptomatic interrupted IVC presents a serious obstacle to successful cardiac interventions, which, ideally, needs to be known prior to the intervention. Non-invasive imaging, such as multislice CT and magnetic resonance imaging, is the most reliable technique for the identification of this anomaly; however, their routine use prior to cardiac intervention has not been established. Saito et al. [1] reported that, using a plain chest X-ray, an abnormal right paravertebral pleural line may be identified representing the dilated azygos vein. Others have described the identification of an interrupted IVC using echocardiography [2, 3]. Pantin et al. [2] reported that upon intraoperative TOE, a severely enlarged azygos vein, dilated hepatic veins draining via a common vein into the right atrium and an invisible IVC can be identified in patients with the interrupted IVC undergoing bypass surgery. Because of the close proximity of the enlarged azygos vein and the descending thoracic aorta, colour flow Doppler may help to reveal the vascular nature of the structure and to recognize the missing communication between the aorta and the azygos vein. Pulse-wave Doppler may, furthermore, allow to the detection of blood flow when it takes an opposite direction in the descending aorta versus the azygos vein and a lack of a connection between the hepatic veins and the distal IVC. These findings were also observed by Kuroda et al. [4] describing an abnormal connection of the IVC to the right atrium (RA) identified by TOE and Kuzumi et al. [5] who described an abnormal connection of the azygos vein to the SVC. Finally, Mihmanli et al. [3] were able to detect an interrupted IVC with azygos continuation by abdominal ultrasound.

**CONCLUSION**

In conclusion, we caution surgeons that, although rarely seen, an interrupted IVC may prohibit venous access to the right atrium or ventricle and suggest that a preoperative chest X-ray and the echocardiography may allow the timely detection of such cases.

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REFERENCES


