Absent Superior Vena Cava in Tetralogy of Fallot

Abstract

Absent superior vena cava (SVC) is an asymptomatic congenital systemic venous anomaly which is rarely detected and compatible with normal life. Undiagnosed absent SVC may cause problems during cardiac catheterization or cardiac surgery. We present our surgical experience in a patient with tetralogy of Fallot who had undiagnosed absent SVC.

Keywords: Azygous continuation, superior vena cava interruption, tetralogy of Fallot

Introduction

In systemic venous anomalies of upper body, systemic veins drain into the right atrium through azygous or hemiazygous or both, depending on the presence or absence of an innominate vein. There will be only one systemic vein (inferior vena cava [IVC]) connecting to the right atrium in the pericardial cavity.

Case Report

A 2-year-old female child presented to our hospital with a history of cyanosis. Her electrocardiograph, Chest X-ray [Figure 4], and two-dimensional echocardiogram showed tetralogy of Fallot with infundibular and valvular pulmonary stenosis and good-sized pulmonary arteries. She was taken up for intracardiac repair.

bilateral During surgery, superior vena cava (SVC) was found to be absent [Figure 1]. Innominate vein was present, but course could not be traced within the pericardial cavity. Cardiopulmonary bypass (CPB) was established with aortic, innominate, and IVC cannulation. As this was a surprise on the table, we cooled the patient to 18°C and opened right atrium under short circulatory arrest. Inspection inside both atria did not show any systemic venous drainage other than IVC which was snugged for total CPB.

CPB flows were restarted, and during rewarming, intracardiac repair (infundibular resection and ventricular septal defect closure with Gortex(pTFE) patch) was done through the right atrial approach. The patient was weaned off CPB, extubated same day, and discharged on day 3.

Computerized tomography (CT)-cardiac was done to confirm absent SVC which showed drainage into IVC [Figure 2].

Discussion

Absent SVC is the rarest asymptomatic anomaly in venous system characterized by innominate draining IVC through azygous. If innominate vein is absent, it can take azygous route on the right side and hemiazygous on the left side.

SVC is formed by the right cranial cardinal vein and the right common cardinal vein. The azygous system is contributed by the more cranial portion of the right (azygous) and left (hemiazygous) supracardinal veins. Any anomalies in this complex development may lead to abnormal vessel development. (Hypothesis of SVC and IVC interruption. Figure 3).^[1] This anomaly may not cause significant problem to anesthetic management except for introduction of a Swan–Ganz catheter from above.

On reviewing the literature, 11 cases of SVC interruption were reported and some of them were diagnosed during cardiac catheterization.^[2-4] Looking back in our case, innominate and IVC cannulation achieved excellent drainage for CPB. We could have done the same with one cannula alone in IVC.

Although this rare anomaly is found incidentally in asymptomatic individuals, it is important in terms of the clinical implication. An incidental finding of this condition may confuse the surgeon during

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Figure 1: Absence of bilateral superior vena cava



Figure 3: Hypothesis of superior vena cava and inferior vena cava interruption

venous cannulation for CPB, but now, we can say that it makes the cannulation simple by achieving total CPB with a single IVC cannulation.^[5] Bidirectional Glenn cannot be done here, as SVC is absent, whereas connecting IVC to pulmonary artery will achieve total cavopulmonary connection during single-ventricle palliation.

If innominate vein is present, it can be cannulated for bicaval drainage but may not be necessary in all cases. During cardiac transplant, if recipient has this anomaly, we need one IVC anastomosis only. If donor heart has this anomaly, the recipient's SVC can be anastomosed to the right atrial appendage as in Warden procedure.

Conclusion

A totally absent SVC is a very rare anomaly. Since it is usually asymptomatic, the incidence in the general population may be higher than detected because of benign nature of this SVC anomaly. The knowledge of the systemic venous anatomy has to be accurate before cardiac surgery.



Figure 2: Computerized tomography cardiac-superior vena cava interruption which showed drainage into inferior vena cava



Figure 4: Chest X-ray

The possibility of this rare SVC anomaly should also be kept in mind when unexpected difficulties arise in cardiac catheterization. If doubts emerge regarding systemic venous drainage during echocardiography, cardiac catheterization, CT angiography, or magnetic resonance imaging angiography may be considered for accurate preoperative diagnosis so that we can plan innominate and IVC cannulation separately or single venous cannulation (IVC) and proceed with the procedure without any added risk.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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