

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.

During surgery, bilateral 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

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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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