

# An Unusual Case of Uninterrupted Inferior Vena Cava with Accessory Hemizygous Channel: An Incidental Finding in a Child

## Abstract

Azygos or hemizygous continuation of inferior vena cava (IVC) is diagnosed in the presence of intrahepatic interruption of IVC. We report a case of a 4-year-old, male child presenting with a history of poor weight gain. A detailed evaluation of the child revealed a diagnosis of hemizygous continuation of uninterrupted, but severely obstructed, IVC. This incidental finding has rarely been reported in the literature.

**Keywords:** Accessory hemizygous channel, obstructed inferior vena cava, uninterrupted inferior vena cava

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## Introduction

Interruption of the inferior vena cava (IVC) is rare, with a reported incidence of 1:5000 based on prenatal ultrasound screening. In the majority of the cases, it presents as an isolated anomaly, although it may be associated with cardiac or splenic abnormalities and often with left atrial isomerism. Azygos continuation of IVC is commonly associated with congenital heart defects like a complete atrioventricular septal defect, double outlet right ventricle, ventricular outflow tract obstructions, total anomalous pulmonary vein drainage, complete heart block, and viscera-cardiac heterotaxy.<sup>[1]</sup>

The prevalence of azygos continuation of the IVC approaches 0.6–2% in association with congenital heart defects and less than 0.3% when isolated.<sup>[2]</sup>

Hemizygous or azygous continuation of an obstructed IVC is still a rare anomaly. We report such a case of hemizygous continuation of the IVC in the presence of a severely obstructed IVC without atrial isomerism.

## Case Report

A 4-year-old, male child weighing 10 kg (birth weight 2.5 kg) reported with a history of poor weight gain and failure to thrive (body mass index [BMI] 12.5). There was no history of recurrent respiratory tract

infection, cyanosis, dyspnea, edema, and abdominal distension. Antenatal, natal, and postnatal periods were uneventful. Family and socioeconomic history were normal. On examination, his vitals were unremarkable, and saturation was 97% in all four limbs on room air. Auscultation revealed a short systolic murmur all over the precordium. Respiratory, abdominal, and neurological examinations were unremarkable.

Transthoracic echocardiogram revealed situs solitus, levocardia, right-sided IVC, and left-sided aorta on the subcostal short-axis view; however, the direct drainage of IVC into the right atrium could not be confirmed on subcostal long-axis view [Figure 1a]. An upward draining venous channel was detected in the subcostal long-axis view on the left side of the descending aorta [Figure 1b and Supplementary Video 1]. Right superior vena cava (RSVC) was draining into the right atrium. There was persistent left superior vena cava (LSVC) draining into the coronary sinus (CS) with no innominate bridging vein and normal pulmonary venous drainage and no intracardiac defect. The rest of the cardiac anatomy was unexceptional on echocardiography.

The chest radiograph and ultrasound of the abdomen excluded the presence of atrial isomerism with utmost certainty. There was no evidence of Budd–Chiari malformation or hepatic vena cava syndrome (HVCS) on the abdominal ultrasound. To determine the IVC drainage into the right atrium, we

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contemplated the options of computerized tomographic angiography (CTA) or cardiac catheterization angiography. Although an invasive procedure, cardiac catheterization was still preferred over CTA to obtain hemodynamic data, which were not possible with a CTA.

Diagnostic cardiac catheterization was done under conscious sedation, which showed severely obstructed intrahepatic part of IVC that was draining into right atrium [Figure 2a and Supplementary Video 2], and there was a hemizygous continuation of IVC that was draining into the right atrium via LSVC and CS [Figure 2b and Supplementary Video 2]. There was a mean pressure difference of 6 mmHg between the right atrium and IVC. The pressure difference was measured using a 4 French multipurpose A2 catheter (Cordis Corporation) by the pullback method through the right femoral venous route. Cardiac catheterization was performed to confirm the anatomy and establish the diagnosis.

It is unlikely that this incidental and rare anatomical anomaly had any pathological concerns for the patient in view of the failure to thrive and poor weight gain. The parents were counseled and the patient was advised to consult a pediatrician for a detailed evaluation of poor weight gain to exclude other causes of failure to thrive, namely, nutritional deficiencies, collagen vascular diseases (including serum antinuclear antibody and rheumatoid factor), celiac disease, cystic fibrosis, gastroesophageal reflux, and intestinal parasites. The child is under regular follow-up with the pediatrician.

The child later underwent detailed nutritional and serological work up with the pediatrician and was diagnosed with celiac disease. His serum test was positive for antitransglutaminase immunoglobulin A (IgA). He is being managed with proper dietary modifications, including gluten-free diet and nutritional supplementation and, currently, under bimonthly follow-up with adequate weight gain.

## Discussion

Congenital malformations and abnormalities of IVC and its tributaries have been rarely reported.

During embryological development, IVC is composed of the hepatic, prerenal, renal, and postrenal segments,

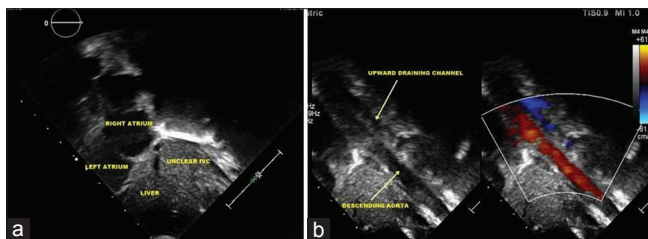


Figure 1: (a) Unclear drainage of inferior vena cava (IVC) on transthoracic echocardiogram in the subcostal long-axis view. (b) Transthoracic echocardiogram showing upward draining venous channel on the left side of the descending aorta in the subcostal long-axis view

which undergo segmental fusion, regression, and midline anastomosis. Failure of the hepatic and prerenal segments to fuse results in infrahepatic interruption of IVC, the most common developmental anomaly of the IVC. The infrahepatic IVC may continue either as the azygos vein or hemizygous vein to the LSVC.<sup>[3-5]</sup>

This vascular anomaly is most frequently associated with visceral heterotaxy, atrial septal defects, left atrial isomerism (polysplenia syndrome), and right atrial isomerism (asplenia syndrome).<sup>[6,7]</sup>

Although most of these isolated anomalies are incidental findings, they may have clinical implications in certain situations. The dilated azygos vein may be misdiagnosed as a mediastinal mass on chest radiography.<sup>[8]</sup>

It is hypothesized that the symptoms and prognosis are related to associated congenital heart disease.

There are several reports of interrupted IVC with azygos or hemizygous continuation in literature, but this combination of congenital IVC malformation, in which there was patent but severely obstructed intrahepatic part of IVC along with accessory hemizygous channel with no evidence of isomerism, has been infrequently reported in the literature until now. Most of the cases are supposed to be asymptomatic and are detected incidentally.

A thorough literature search revealed a case of widely patent IVC with azygos continuation.<sup>[9]</sup>

Obstructed IVC mandates an escape route for the blood from the lower part of the body to be diverted via the persistence of an embryonal anastomosis between the primordia of IVC and azygos or hemizygous system.

In our case, the patient had no signs of obstructed IVC, namely, ascites or pedal edema; hence, he was advised for follow-up at regular intervals.

Palliative implantation of a stent in the IVC may be an option that can be anticipated for maintaining its patency at later stages if symptomatology of IVC obstruction develops.

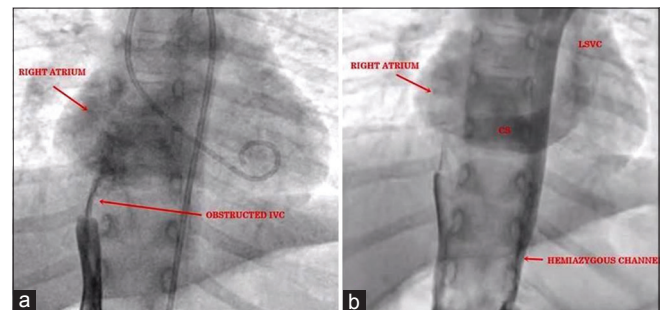


Figure 2: (a) Angiographic image in anteroposterior view showing drainage of obstructed inferior vena cava (IVC) into the right atrium. (b) Angiographic image in anteroposterior view showing drainage of the hemizygous venous channel into the right atrium via left superior vena cava (LSVC) and coronary sinus (CS)

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### Conflicts of interest

There are no conflicts of interest.

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