Interrupted Inferior Vena Cava Syndrome

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Abstract

We report a case of interrupted inferior vena cava (IVC) as a rare developmental defect. Inferior vena cava interruption is usually accompanied with azygos and hemiazygos continuation, and is asymptomatic. Consequently, venous blood from the caudal part of the body reaches the heart via the azygous vein and superior vena cava. A 50 year old female who came for routine health check-up was found to have pulmonary hypertension on two dimensional echocardiography. On further investigations she also had restriction on pulmonary function test. When computed tomography pulmonary angiography was done, showed dilated azygous vein without pulmonary embolism. Computed tomography of the abdomen demonstrated interrupted inferior vena cava. Such patients are at increased risk of deep vein thrombosis and pulmonary embolism.

Case Report

A 50 year old female came for routine health check up. The only clinical symptoms she had was dyspnea on exertion class I Medical Research Council dyspnea scale. Patient had two normal pregnancy without any complications. She gave history of diabetes mellitus since 18 years and was receiving insulin therapy since six years. Family history was non contributory. General examination revealed mild lower limb pedal edema. Rest general examination was normal. Pulse - 88/min, BP – 130/90 mmHg, SpO₂ - 96% on room air. Respiratory Rate - 20/ min. Respiratory system examination revealed bilateral equal breath sounds. Rest of the systemic examination was also normal. The electrocardiogram was normal. Two dimensional echocardiography revealed mildly dilated right atrium (42mm) and dilated right ventricle (32mm), mild tricuspid regurgitation, mild pulmonary hypertension (pulmonary artery systolic pressure = 38 mmHg), no left ventricular regional wall motion abnormality, no diastolic dysfunction, no left ventricular hypertrophy and normal left ventricular systolic function (left ventricular ejection fraction 55%). Pulmonary function test revealed mild restriction. She was advised computed tomography pulmonary angiography which revealed dilated azygous vein and no evidence of pulmonary embolism. Computed tomography of abdomen revealed inferior vena cava continuing superiorly as the azygous vein, entering the posterior mediastinum through the hiatus in the diaphragm for the aorta, where it appeared slightly compressed (Figures 4 and 5).

Embryologically, the morphologic features of the inferior vena cava are the result of a dynamic process of development, regression, anastomosis, and replacement of 3 paired venous blood conduits (the posterior cardinal, supracardinal, and subcardinal veins of the primary system) starting in the fifth week. When the development of the venous system is complete, it is possible to define the embryologic origins of all its components. In particular, the superior vena cava originates from the right superior cardinal vein. the azygos system is formed from the more cranial portion of the right (azygos) and left (hemiazygos) supracardinal veins. Blood from the subdiaphragmatic portion of the embryo is channeled into a single inferior vena cava, flowing to the paramedian right. In caudal to cranial order, these segments are the iliac veins from the posterior cardinal veins, the subrenal (postrenal) segment from the right supracardinal veins, the renal segment from anastomosis between the right supracardinal and subcardinal veins, the suprarenal (prerenal) segment from the right supracardinal vein, the hepatic vein. Systemic venous flow beyond this point is accomodated by the dilated azygos and hemiazygos veins, which eventually empty into the superior vena cava through a dilated azygos arch.² Our patient demonstrated the inferior vena cava continuing superiorly as the azygous vein, entering the posterior mediastinum through the hiatus in the diaphragm for the aorta, where it appeared slightly compressed (Figures 4 and 5).

Discussion

Infrahepatic interruption of the inferior vena cava with azygous continuation is a rare congenital anomaly seen in 0.6–2% of patients with congenital heart disease¹ and in less than 0.3% of otherwise normal individuals. This developmental anomaly results in termination of the inferior vena cava below the hepatic vein. Systemic venous flow beyond this point is accommodated by the dilated azygos and hemiazygos veins, which eventually empty into the superior vena cava through a dilated azygos arch.² Our patient demonstrated the inferior vena cava continuing superiorly as the azygous vein, entering the posterior mediastinum through the hiatus in the diaphragm for the aorta, where it appeared slightly compressed (Figures 4 and 5).

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Interrupted inferior vena cava is associated with congenital heart disease in approximately 85% of cases, and frequently with the polysplenia syndrome. Our patient demonstrated polysplenia. Other associated cardiac anomalies include atrial septal defect, pulmonary stenosis or atresia, double-outflow right ventricle, atrioventricular canal, cor biloculare, and anomalously connecting pulmonary disease. Also documented is the presence of mirror-image dextrocardia, dextroversion, situs inversus, partial inversion of the abdominal viscera, and bilateral superior vena cava. In our patient two dimensional echocardiography showed mildly dilated right atrium and right ventricle, mild pulmonary hypertension with normal left ventricular parameters.

Failure of the hepatic and prerenal segments to fuse is the most common developmental anomaly of the inferior vena cava and results in infrahepatic inferior vena cava interruption. The infrahepatic inferior vena cava may continue as the azygos vein or may continue as the hemiazygos vein to the left superior vena cava, intrathoracic veins or anomalous intrahepatic veins. The dilated azygos vein may be misinterpreted as a paracardiac or mediastinal mass on chest radiography. Our patient presented for a routine health check up. During investigations, pulmonary function tests revealed mild restriction and two dimensional echo revealed mild pulmonary hypertension. Further investigations by means of computed tomography pulmonary angiogram and computed tomography of abdomen and pelvis permitted the confirmation of the diagnosis. Although, the patient as yet had not manifested any complications related to this anomaly, this diagnosis is important as such it may be associated with deep vein thrombosis of the lower limbs, bilateral venous insufficiency or with sick sinus syndrome. The other potential issues or complications such patients may face include procedural difficulties during right heart catheterization, electrophysiological studies, cardiopulmonary bypass surgery, femoral vein catheter advancement, inferior vena cava filter placement, and temporary pacing through the transfemoral route.

**Conclusion**

To our knowledge this is the first report in which further investigation of mild restriction on pulmonary function test and mild pulmonary hypertension during routine health check led to the diagnosis of interrupted inferior vena cava syndrome. The knowledge of this congenital anomaly is important so that deep vein thrombosis measures may be advised during high risk situations and also alternative sites be used for cardiac catheterization other than the femoral vein.
References


