Interrupted Inferior Vena Cava with Atrial Septal Defect

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Abstract

Interrupted intrahepatic inferior vena cava (IVC) is a rare vascular anomaly that can occur in isolation or with various cardiac anomalies like atrial septal defect (ASD), atrio-ventricular canal, anomalous pulmonary veins, double outflow right ventricle, pulmonary stenosis or atresia, and sick sinus syndrome. It may be detected antenatally or incidentally in adulthood. The anomaly may be associated with increased risk of deep vein thrombosis, and can have an impact on interventional cardiac procedures and surgical procedures. Here, we report a baby with antenatally diagnosed IVC interruption with an ASD that was confirmed postnatally by echocardiography. The baby remained clinically stable and was advised follow-up and periodic monitoring for the development of rhythm abnormalities in the future.

Introduction

Interrupted intrahepatic inferior vena cava (IVC) is a rare vascular anomaly that is found in 0.6-2% of individuals with congenital heart disease and 0.3% of otherwise normal individuals [1]. It may be associated with various cardiac anomalies like atrial septal defect (ASD), atrio-ventricular canal, anomalous pulmonary veins, double outflow right ventricle, pulmonary stenosis or atresia, and sick sinus syndrome [1,2].

This anomaly may be detected antenatally [3] or incidentally in adulthood [1,4]. Here, we report a prenatally diagnosed IVC interruption with an ASD, confirmed postnatally by echocardiography.

Case Report

A 30 years old primigravida, during antenatal ultrasonography at 22 weeks gestation, was found to have an interrupted IVC connected to theazygos vein, the superior vena cava (SVC) connected to coronary sinus and an ostium secundum ASD. No other abnormalities were detected within the heart or other organs. Biometry was consistent with foetal age and karyotype was not done. The hepatic veins and ductus venosus reached the right atrium normally. The mother later developed gestational hypertension and intrahepatic cholestasis of pregnancy (ICP). A male baby weighing 2.8 kg was delivered by emergency caesarean section at 37 weeks. Clinical evaluation of the cardiovascular system was unremarkable. Electrocardiography was unremarkable and showed sinus rhythm with normal atrio-ventricular conduction. Echocardiography confirmed the antenatal diagnosis of inferior vena cava with azygos continuation to superior vena cava and dilated coronary sinus, a 7 mm ostium secundum ASD dilated right atrium and right ventricle with normal pulmonary veins and normal right and left ventricular function. No other cardiac or extra cardiac abnormalities were detected. The patient was advised follow-up and periodic monitoring for the development of rhythm abnormalities in the future.

Discussion

IVC embryogenesis is a complicated process involving three paired veins: the posterior cardinal, supracardinal, and sub cardinal veins of the primary system in the fifth week. The SVC originates from the right superior cardinal vein. The azygos system is formed from the more cranial portion of the right and the hemiazygos from the left supracardinal veins. Blood from the sub diaphragmatic portion of the embryo is channeled into a single IVC, flowing to the paramedian right. Five embryologic segments contribute to its composition. Failure of the formation of the most cranial segment, the hepatic, results in interrupted IVC [5,6].

Interruption of the inferior vena cava with azygos continuation represents the most common abnormality involving these veins

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This malformation is strongly associated with the presence of the polysplenia syndrome [7] and atrial isomerism [8,9].

With an isolated interrupted IVC, patient is usually asymptomatic and the vascular anomaly itself does not mandate any treatment [2]. Postnatal survival has been related to the severity of the associated heart disease [7,9]. The anomaly may be associated with increased risk of deep vein thrombosis [10].

This finding also can have an impact on interventional procedures (IVC filter placement). There can be procedural difficulties during right heart catheterisation, electrophysiological studies, cardiopulmonary bypass surgery, femoral vein catheterisation, IVC filter placement, and temporary pacing through the transfemoral route [1] and surgical procedures like oesophagectomy and liver transplants [2]. Awareness of the anomaly before initiating the procedure can help in deciding an alternate strategy, and avoid undue delay.

This case report is among those few where diagnosis of an interruption of IVC with azygos continuation was made antenatally and confirmed postnatally. Most of the previous reports of isolated IVC interruption have been in adults [10].

Prenatal preparation for babies with this anomaly should address possible cardiac and extra cardiac malformations and counseling to the parents about the same and also about the possibility of development of sick sinus syndrome and increased risk for venous thrombosis in the future. A careful a postnatal evaluation and follow-up is hence necessary.

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