Prenatal diagnosis of isolated interrupted inferior vena cava with azygos continuation to superior vena cava

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ABSTRACT

Absence of inferior vena cava is an uncommon congenital abnormality. It is usually associated with other structural anomalies, typically left isomerism. We report a case of interrupted inferior vena cava with azygos continuation diagnosed as an isolated finding during routine prenatal ultrasound scan, confirmed by post-natal echocardiography. Detailed ultrasound examination of the fetal anatomy failed to demonstrate other anomalies. The neonatal course of this fetus was uneventful.

Keywords: Azygos continuation, fetal diagnosis, interrupted inferior vena cava

INTRODUCTION

Azygos continuation of the inferior vena cava (IVC) (also known as absence of the hepatic segment of the IVC with azygos continuation) is an uncommon vascular anomaly, with prevalence of about 0.2-3%.[1] The incidence of this anomaly approximates 0.6% in patients with congenital heart defects including atrio-ventricular canal, anomalously connecting pulmonary veins, double outflow right ventricle, large atrial septal defect, pulmonary stenosis or atresia, and sick sinus syndrome.[2,3] This is often associated with visceral and cardiac malpositions typically associated with left isomerism.[4,5] The most common pattern is azygos continuation with normal position of the heart and abdominal viscera. There is no evident relationship between the subtypes of this anomaly and the intracardiac defects that may be present.[6] This anomaly can be detected in the prenatal period[7] or incidentally in the elderly.[8]

Isolated interrupted inferior vena cava with azygos continuation has been rarely described,[6,9] especially in prenatal series. We report the prenatal diagnosis of isolated IVC interruption, confirmed by post-natal echocardiography.

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Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/0974-2069.126558

CASE REPORT

A 38-year-old second gravida was referred to us for fetal echocardiography at 25 weeks in view dilated superior vena cava (SVC) and suspicion of interrupted IVC with persistent right umbilical vein. Biometry was consistent with fetal age. No other abnormalities were detected within the heart or other organs. Fetal karyotype was normal. Fetal echocardiography showed the presence of a vessel posterior to the descending aorta in the four-chamber view ("Double vessel sign," Figure 1). The next clue came from the three-vessel view, which showed a dilated, prominent SVC [Figure 2]. In the bi-caval view, the intra-hepatic portion of the inferior vena cava was found to be interrupted [Figure 3]. Rostral to this point, a thinner vessel with venous flow was detected as a continuation of the infra-hepatic portion of the IVC. On tracing this vessel cranially, its connection with the superior vena cava was documented [Figure 4]. The hepatic veins and ductus venosus reached the right atrium normally. No other anomalies of the heart were detected.

A female baby weighing 2.8 Kg was delivered by elective caesarian section at 38 weeks. Clinical evaluation of the cardio-vascular system was unremarkable. A detailed cardiac evaluation was performed at 6 weeks post-natal age. ECG was unremarkable and showed sinus rhythm with normal atrio-ventricular conduction. Echocardiography confirmed the prenatal diagnosis of isolated inferior vena cava with azygos continuation to superior vena cava [Figure 5]. No other cardiac/extracardiac abnormalities were detected. The patient was advised follow-up and

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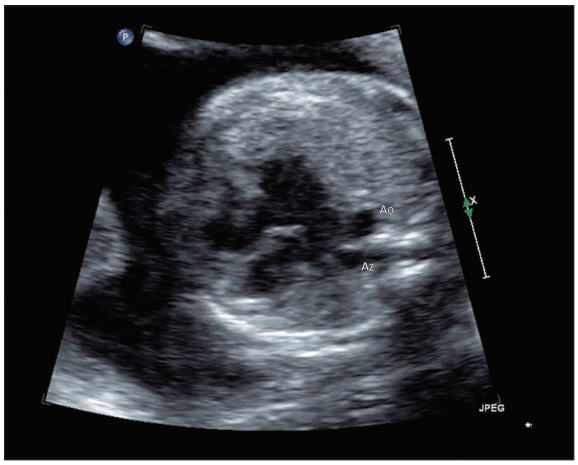

Figure 1: Four chamber view showing a dilated azygos vein (Az) posterior to the descending aorta (Ao) = "Double vessel sign."

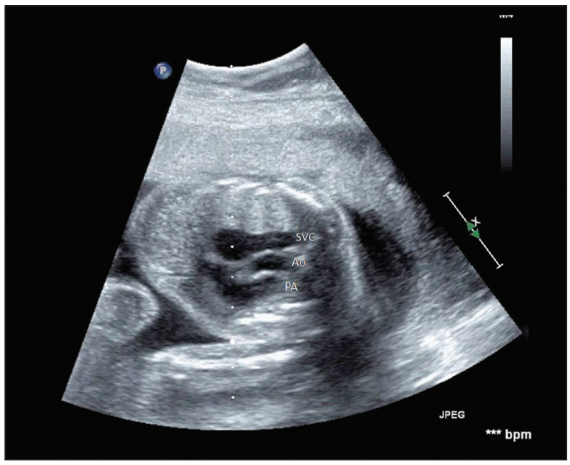

Figure 2: Three-vessel view showing a dilated superior vena cava. SVC = Superior vena cava; Ao = Aorta; PA = Pulmonary artery

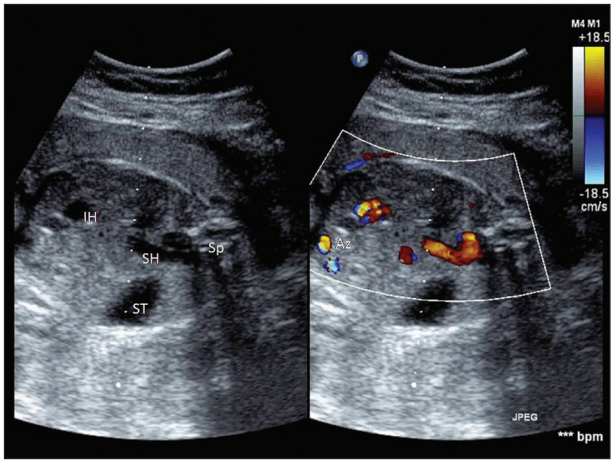

Figure 3: Bi-caval view showing the section of liver in the sagittal plane. There is a gap between the supra-hepatic (SH) and infra-hepatic (IH) portions of the IVC, suggesting interruption of the intrahepatic portion of the IVC. A dilated azygos vein (Az) is also seen posterior to the infra-hepatic IVC. SH = Supra-hepatic IVC; IH = Infra-hepatic IVC; Sp = Spine; ST =Stomach

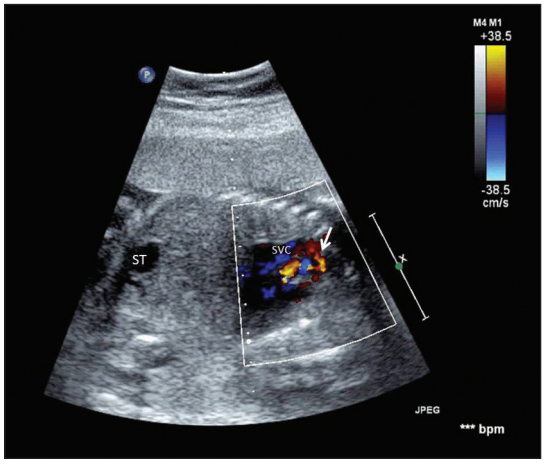

Figure 4: Three-vessel view showing azygos vein joining the SVC (arrow). SVC = Superior vena cava; ST = stomach

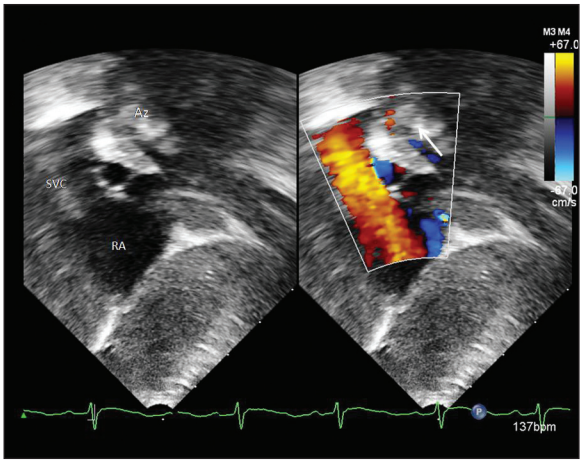

Figure 5: Post-natal echocardiography sub-xiphoid view showing a dilated SVC with the venous channel (azygos vein) running parallel to the descending aorta (arrow) and joining the SVC. SVC = superior vena cava; RA = right atrium; Az = azygos vein

periodic monitoring for the development of any rhythm anomalies in future.

DISCUSSION

The IVC is made up of four segments during the embryonic period, and failure of the formation of hepatic segment results in an interrupted IVC. The embryogenesis of IVC is a complex process involving the formation of several anastomoses between three paired embryonic veins. Interruption of the inferior vena cava with azygos continuation represents the most common abnormality involving these veins.[10] This malformation is considered to be an excellent marker for the presence of atrial isomerism[11] and the polysplenia syndrome.[4] Symptoms and prognosis are related to associated congenital heart disease.[2,3] This finding also can have an impact on interventional procedures (IVC filter placement) and surgical procedures (esophagectomy, liver transplants). According to Sheley and colleagues,[12] post-natal survival was related to the severity of the associated heart disease.

In their series, six fetuses with complex cardiac anomalies did not survive, while those with isolated IVC interruption (n = 1) or with associated atrial septal defect (n = 2) survived. In a series reported by Phoon *et al.*, of a total of 18 infants with left atrial isomerism diagnosed prenatally, all but four (78%) died in the neonatal period. In the same study, 15 mothers elected to terminate the pregnancy and there were five still births.

In the situation of an isolated interrupted IVC, patient is usually asymptomatic and the vascular anomaly itself does not mandate any treatment. In some case, venous insufficiency of lower extremities is seen and may have increased risk of deep venous thrombosis in future.[13,14]

The unique aspect of this case report is the diagnosis of an isolated interruption of IVC with azygos continuation by fetal echocardiography. Most of the previous reports of isolated IVC interruption have been in adults.[2,8,13] We feel the bi-caval view is extremely useful in demonstrating this anomaly along with demonstration of the azygos continuation to the SVC. Pre-natal counseling for this vascular anomaly should include the likelihood of possible development of sick sinus syndrome in future and the increased risk for venous thrombosis. A careful search for other structural anomalies and a post-natal evaluation and follow-up is hence mandatory.

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How to cite this article: Giang DC, Rajeesh G, Vaidyanathan B. Prenatal diagnosis of isolated interrupted inferior vena cava with azygos continuation to superior vena cava. Ann Pediatr Card 2014;7:49-51.

Source of Support: Nil, **Conflict of Interest:** None declared