Rare Case of Bilateral Superior Vena Cava, Persistent Left Superior Vena Cava Draining to Coronary Sinus, Absent Bridging Vein, Interrupted Inferior Vena Cava with Azygos Vein Continuation to Right Superior Vena Cava, Situs Inversus and Pulmonary Hypertension in a Neonate, A Case Report

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Clinical Image

Figure 1: Echocardiogram: 2D and colour Doppler acquisitions showing left SVC (LSVC) draining to coronary sinus (CS). (LA: Left Atrium).

Figure 2: Echocardiogram: 2D and colour Doppler acquisitions showing hepatic veins (HV) draining to right atrium (RA) and no inferior vena cava's flow could be appreciated.

Figure 3: Echocardiogram: 2D and colour Doppler acquisitions showing right SVC (RSVC) draining to right atrium (RA). LA: left atrium.

We report a rare case of multiple congenital anomalies as described above. The baby girl was born at term after uneventful pregnancy. The cardiology service was consulted at birth because of abnormal fetal echocardiography. Her clinical examination was unremarkable apart from mild cyanosis (oxygen saturation was 87%). Chest and abdominal x-ray showed levocardia, normal bronchi and abdominal situs inversus.

The echocardiography revealed abdominal situs inversus, interrupted inferior vena cava (IVC), bilateral superior vena cava (SVC) with no bridging vein, left SVC draining to coronary sinus, right ventricular (RV) dilatation, and patent ductus arteriosus (PDA). Serial echocardiography's showed progressive RV dilatation and spontaneous closure of PDA (Figures 1-3).

She underwent diagnostic cardiac catheterization which revealed high pulmonary pressure, left SVC draining to coronary sinus, absent bridging vein and interrupted IVC with azygos vein continuation to the right SVC (Figures 4,5).
Figure 4: Angiogram, AP projection showing the venous catheter course confirming interrupted IVC with azygos vein (AV) continuation to Right SVC. The catheter was advanced to right atrium (RA), coronary sinus and the tip was kept in the persistent left SVC (LSVC). The angiogram showing good size persistent LSVC draining to coronary sinus (CS) and to right atrium (RA) with no evidence of bridging vein seen.

Figure 5: Angiogram, AP projection showing the venous catheter course confirming interrupted IVC with azygos vein continuation to Right SVC. The catheter was further advanced to right ventricle and pulmonary artery. The angiogram showing normal size and confluent branch pulmonary arteries.