



A case of supracardiac total anomalous pulmonary venous return

Kamel R, Abdel Salam SA, Badr I
Fetal Medicine Unit, Cairo University , Egypt., Cairo, Egypt

Objective

To present a case of supracardiac total anomalous pulmonary venous return.

Methods

This is a case report.

Results

A 23-year old woman (G1P0) was referred to fetal medicine unit at Cairo University for detailed fetal cardiac assessment. Fetal cardiac examination was carried out using 2 dimensional and 3 dimensional ultrasound (Glass body and color STIC mode) as well as color Doppler using an abdominal volume probe (Voluson E10, GE Healthcare, Zipf, Austria). Ultrasound examination at 25 weeks of pregnancy revealed single ventricle of left morphology with common AV canal and double outlet morphology of both great vessels with parallel course. The ascending aorta was cephalad to the main pulmonary artery which showed relatively small size compared to the ascending aorta denoting evidence of pulmonary stenosis. Right sided aortic arch was also noted without evidence of coarctation. Left SVC directly draining into the left sided atrium with absent right SVC. A retro cardiac common pulmonary vein draining all right and left pulmonary veins and then ascending on the right aspect of the mediastinum passing anterior to right lung hilum (in front of right pulmonary artery and right main bronchus) in a vertical pathway to drain into innominate vein (Color Doppler showed opposite flow direction between the left SVC and the ascending vertical vein). - Dextrocardia with situs ambiguous. The stomach was seen on the left side with central liver and ambiguous distribution of umbilical vein into portal sinus. Both aorta and IVC were seen on the right side (juxtaped). Most of small bowel loops on the contra lateral aspect of the stomach suggesting gut mal rotation. This combination of complex congenital heart disease in association with situs ambiguous and juxtaped aorta and IVC as well as dextrocardia is in favor of the diagnosis of right atrial isomerism with more likely associated gut mal rotation based upon its great incidence in cases of heterotaxy syndromes (approximately 30%) as well as the discordant location of small bowel loops and stomach. Parents refused termination and a male baby (3200 g) was born with fair general condition and systemic saturation reaching 90% denoting mild form of pulmonary stenosis and unobstructed TAPVD.

Conclusion

Owing to absent pulmonary venous obstruction, unrestricted shunting between systemic and pulmonary aspects of the heart (due to complete mixing of blood at atrial and ventricular levels attributed to common AV canal and single ventricle physiology), common AV valve regurgitation and mild form of pulmonary stenosis that did not cause significant reduction of pulmonary arterial blood flow with relative decrease in size of pulmonary branches, the postnatal course was uneventful and the surgeon asked for staged repair, initial correction of TAPVD without need for systemic to pulmonary arterial shunt, then univentricular repair.

