

Three dimensional Spatio Temporal Imaging Correlation (STIC) in the diagnosis of isolated infra-cardiac total anomalous pulmonary venous connection (TAPVC) in fetal life

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Abstract

Isolated TAPVC is difficult to diagnose on a prenatal ultrasound as standard obstetric screening views are often normal. We present a case of isolated infra-cardiac TAPVC highlighting the utility of STIC imaging in the diagnosis of TAPVC

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STIC in prenatal TAPVC diagnosis

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Total anomalous pulmonary venous connection (TAPVC) is a critical congenital heart disease (CHD) and failure to recognise the condition in the first few days of life can result in significant morbidity and mortality. However, the standard obstetric screening views of the heart are often normal in TAPVC resulting in missed prenatal diagnosis(1). With improvement in resolution of ultrasound imaging and the advent of advanced imaging modalities including spatio-temporal imaging correlation (STIC), the condition is increasingly recognised on prenatal ultrasound(2). We report a fetus with infra-cardiac TAPVC in whom STIC imaging permitted excellent visualisation of the infra-cardiac drainage as well as the potential site of obstruction.

A 24 year old prime gravida mother was referred to us for a detailed evaluation after coarctation of aorta was suspected on the anomaly scan. The gestational age was 26 weeks at the time of assessment. An anomaly scan had been performed 3 weeks earlier. This reported ventricular disproportion with a small left ventricle and a suspicion of coarctation of aorta. The parents were no-consanguineous. Both the parents did not report any chronic medical illness and there was no family history of congenital heart diseases. The mother was not on any medications except iron and folic acid supplements. The nuchal translucency was reported to be normal on a 12 week ultrasound and the combined first trimester screening revealed a low risk for chromosomal aneuploidies.

A detailed fetal evaluation was performed. The imaging was performed on a Philips Epiq Elite ultrasound machine using a V9-2 volume transducer. The fetal growth and fetal as well as maternal Doppler evaluation was normal. There were no important extra-cardiac anomalies with usual arrangement of the abdominal organs. The cardiac mass was in the left side of the chest. There was a single right sided superior caval vein which drained into the right atrium. The inferior caval vein was intact and drained into the right atrium. The pulmonary venous drainage to the left atrium (LA) could not be reliably imaged. The atrioventricular connections were concordant. There was ventricular asymmetry with a smaller left ventricle (LV) (Fig 1 A). However, the LV formed the apex of the cardiac mass. The ventriculoarterial connections were concordant. The outflows were unconstructed. The aortic arch was left sided with normal branching pattern. There was no posterior shelf or substrate for coarctation. The pulmonary veins appeared to drain through a descending vertical vein (Fig 1 B) into the portal venous system suggesting infra-cardiac total anomalous pulmonary venous connection (TAPVC). Three dimensional (3D) colour Doppler imaging was obtained by using spatial-temporal imaging correlation (STIC) and the obtained image was rendered to demonstrate the pulmonary venous drainage. This showed flow acceleration at the site of drainage of the vertical vein to the left portal vein close to the ductus venosus suggesting possible obstruction (Fig 2 and Video 1).

The family were counselled about the probable need for emergency surgical repair soon after birth and the need to plan delivery in a pediatric cardiac surgical facility. The family were also counselled about the excellent long term outcomes after TAPVC repair

TAPVC is a rare CHD with an incidence of approximately 8 per 100,000 live births(3). Neonates with the obstructed form of the disease often become very symptomatic in the first few days of life. Prenatal diagnosis probably has the highest impact in TAPVC among all critical CHD. During prenatal life, only 10% of the combined cardiac output flows through the pulmonary veins. Hence, the pulmonary veins are small and often difficult to image. The diagnosis of TAPVC in prenatal ultrasound was traditionally established by a combination of “soft findings” including mild ventricular disproportion with a smaller LV, increased retrocardiac space between the LA and the descending aorta and the visualisation of an additional vessel on the three chamber view in cases of supra-cardiac TAPVC(4-6). In a review of 424 children with TAPVC operated between 1998 and 2004 in all cardiac centres in United Kingdom, only 1.9% were diagnosed prenatally(7). Ganesan and colleagues reviewed their experience with prenatal diagnosis in 26 cases of TAPVC of which only 4 were isolated TAPVC while 22 were associated with right isomerism. They described the utility of pulse Doppler evaluation of the pulmonary veins in addition to two dimensional imaging in establishing the diagnosis(1).

3D fetal echocardiography using STIC has been shown to be superior to conventional two dimensional ultrasound in the diagnosis of anomalous pulmonary veins. Zhang and colleagues reported their experience with prenatal diagnosis of TAPVC. Among 31 fetuses with adequate quality 2D and 3D imaging in whom the prenatal suspicion of TAPVC was confirmed by postnatal ultrasound or fetal autopsy, the diagnosis was identified only on 3D echocardiogram in 5 cases (16%) establishing the additional utility of STIC imaging(8). Although the diagnosis of TAPVC was established by 2 ultrasound in our case, STIC allowed us to delineate the course of the anomalous venous drainage and also identify the site of potential obstruction.

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Legends

Figure 1 A: Four chamber view of the fetal heart demonstrating mild ventricular disproportion with the left ventricle smaller than the right ventricle. The left ventricle, however, still forms the apex of the heart (LV- left ventricle, RV- right ventricle)

Figure 1 B: Color compare imaging of the fetal thorax and abdomen demonstrating the descending vertical vein (green arrow) draining into the abdomen

Figure 2: Still image of 4D STIC imaging demonstrating the drainage of the vertical vein into the portal system as well as the site of obstruction of the vertical vein at its drainage into the portal system (IVC- inferior caval vein)

Video 1: 4D STIC imaging demonstrating the anomalous pulmonary venous drainage to the portal system. There is flow acceleration at the site of entry into the portal vein which represent a potential site of pulmonary venous obstruction



