Demonstration of circular shunt in fetal Ebstein anomaly

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ABSTRACT

Ebstein's anomaly was diagnosed in a fetus at 24 weeks of gestation. There was significant cardiomegaly and severe tricuspid regurgitation (TR). There was functional pulmonary atresia with severe pulmonary regurgitation (PR) and this was causing a circular shunt. There was no fetal hydrops.

Keywords: Circular shunt, ebstein anomaly, fetal echocardiography

CASE HISTORY

Ebstein's anomaly was diagnosed in a fetus at 24 weeks of gestation [Video 1]. The cardiothoracic ratio was 70% [Figure 1]. The functional right ventricle (RV) was small with a large atrialized chamber. Tricuspid regurgitation (TR) was severe. The pulmonary valve annulus was well-formed, but leaflets were thickened. There was no antegrade flow across the pulmonary valve. There was flow reversal in the ductus arteriosus which was regurgitating into functional RV (pulmonary regurgitation) [Figure 2 and Video 2]. The severe pulmonary regurgitation (PR) was causing a circular shunt where the blood was flowing back from the pulmonary artery to the RV, from the RV to the right atrium due to the significant TR, and from the right atrium to the left atrium through the patent foramen ovale (PFO) [Video 3]. Continuous wave Doppler across the right ventricular outflow demonstrated both systolic and diastolic PR [Figure 3]. There was no fetal hydrops [Figure 4].

DISCUSSION

A 'circular' shunt is defined as a condition in which blood originating in one cardiac chamber is shunted through the heart to return to the original chamber without having passed through a capillary bed.[1] Circular shunt

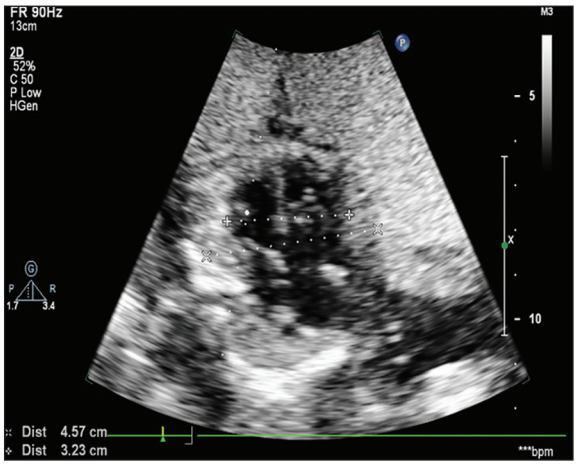

Figure 1: The cardiothoracic ratio has been measured as the ratio of the largest cardiac dimension to the dimension of the thorax

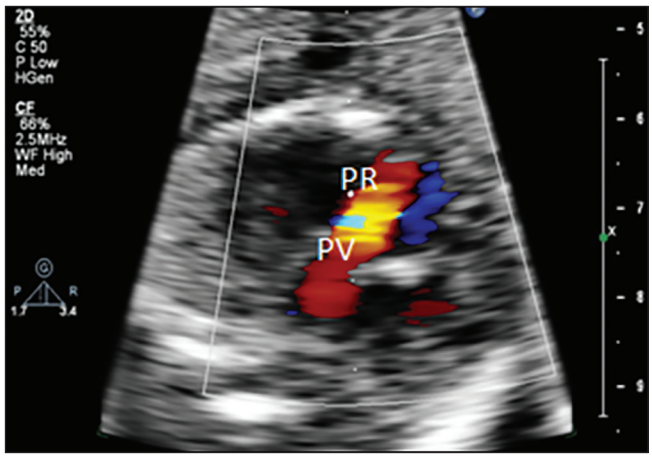

Figure 2: Severe pulmonary regurgitation (PR) across the pulmonary valve (PV) noted

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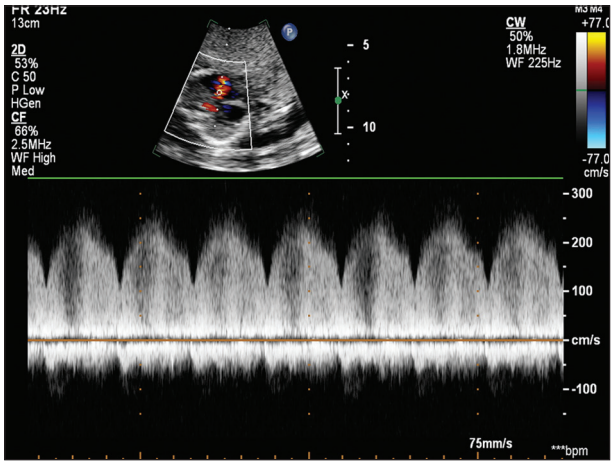

Figure 3: Continuous wave Doppler across the PV demonstrating systolic and diastolic PR

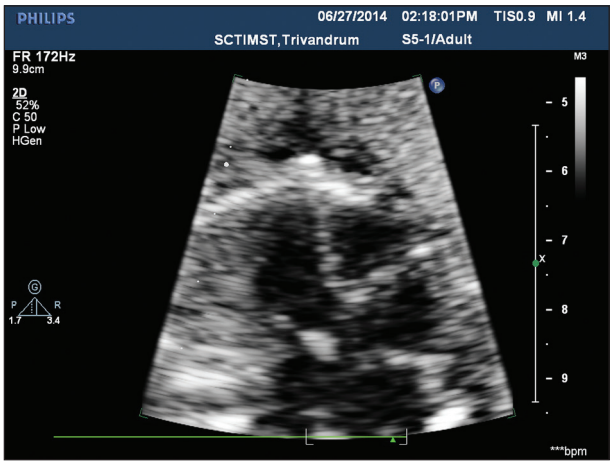

Figure 4: Four-chamber view of the fetal heart showing apical displacement of tricuspid valve. There is no hydrops

has been described in newborns in various congenital heart diseases like:

a. Ventricular septal defect with severe pulmonary stenosis, TR, and PFO,[1]
b. Pulmonary atresia with intact ventricular septum, PR (post-valvotomy), TR, and PFO,[2]
c. Severe Ebstein's anomaly with PR, TR, PFO, and patent ductus arteriosus (PDA).[3] Circular shunt in neonatal Ebstein anomaly results in:
    i. systemic steal and low cardiac output when pulmonary vascular resistance falls and
    ii. left ventricular volume load and heart failure.[4]

Functional pulmonary atresia and severe PR has been noted in severe fetal Ebstein anomaly, but the hemodynamic consequences of circular shunt *in utero* has not been well-studied.[5] Since the pulmonary vascular resistance is high in the fetus, circular shunt may not cause systemic steal *in utero*, but it can lead to ventricular volume load and heart failure. Neonatal Ebsteins anomaly with PR causing systemic steal has been managed with pulmonary vasodilators to reduce pulmonary vascular resistance. Cautious use of prostaglandin E1 (PGE1) is advised as it can increase pulmonary artery pressure and worsen PR and TR and worsen heart failure. Ductal ligation[6] and main pulmonary artery ligation with placement of aortopulmonary shunt is an option for the sickest newborns who fail medical management. There is no consensus for the treatment of fetal Ebstein with PR. For our patient, we planned watchful expectancy with early termination of pregnancy if hydrops develops. Nature of the disease and prognosis were explained to the parents and advised follow-up scan for which they did not attend.

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