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Prenatal diagnosis of left-sided Ebstein's anomaly with congenitally corrected transposition of great arteries and early fetal growth restriction: A rare case report

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Abstract

We report a rare and complex case of left-sided Ebstein's anomaly associated with congenitally corrected transposition of the great arteries (ccTGA), persistent left superior vena cava (PLSVC), and early fetal growth restriction (FGR), diagnosed prenatally at 25 weeks and 4 days of gestation. The patient, a 28-year-old gravida 2 para 1 woman with gestational diabetes mellitus and chronic pancreatitis, was referred following a targeted anomaly scan suggesting a congenital heart defect. Detailed fetal echocardiography confirmed atrioventricular and ventriculoarterial discordance, with apical displacement of the tricuspid valve on the left side, consistent with ccTGA and left-sided Ebstein anomaly. Persistent left SVC and inlet VSD were also noted. The fetus showed signs of early FGR, despite well-controlled maternal glycemia. The case emphasizes the importance of detailed fetal cardiac imaging and multidisciplinary counseling for prenatal diagnosis and management of complex congenital heart anomalies.

Keywords: Ebstein's anomaly; Congenitally corrected transposition; Prenatal diagnosis; Fetal echocardiography; Fetal growth restriction; Persistent left SVC.

Introduction

Ebstein's anomaly is a rare congenital malformation of the tricuspid valve and right ventricle, accounting for approximately 0.5-1% of all congenital heart defects, with an estimated incidence of 1 in 20,000 live births [1]. It is characterized by apical displacement of the septal and posterior tricuspid valve leaflets, leading to atrialization of the right ventricle and varying degrees of tricuspid regurgitation [2]. The left-sided variant, as seen in this case, is exceptionally rare and typically occurs in the context of congenitally corrected transposition of the great arteries (ccTGA), where both atrioventricular and ventriculoarterial discordance are present [3]. Prenatal diagnosis of Ebstein's anomaly is crucial, as the clinical spectrum ranges from asymptomatic to severe cardiomegaly and hydrops fetalis [4]. The presence of double superior vena cava (SVC), found in approximately 0.3% of the general population and up to 11% in those with congenital heart disease, adds further complexity to the fetal cardiovascular anatomy [5]. Advances in fetal echocardiography have significantly improved the detection of such anomalies, enabling early counseling and perinatal planning [3,4,6].

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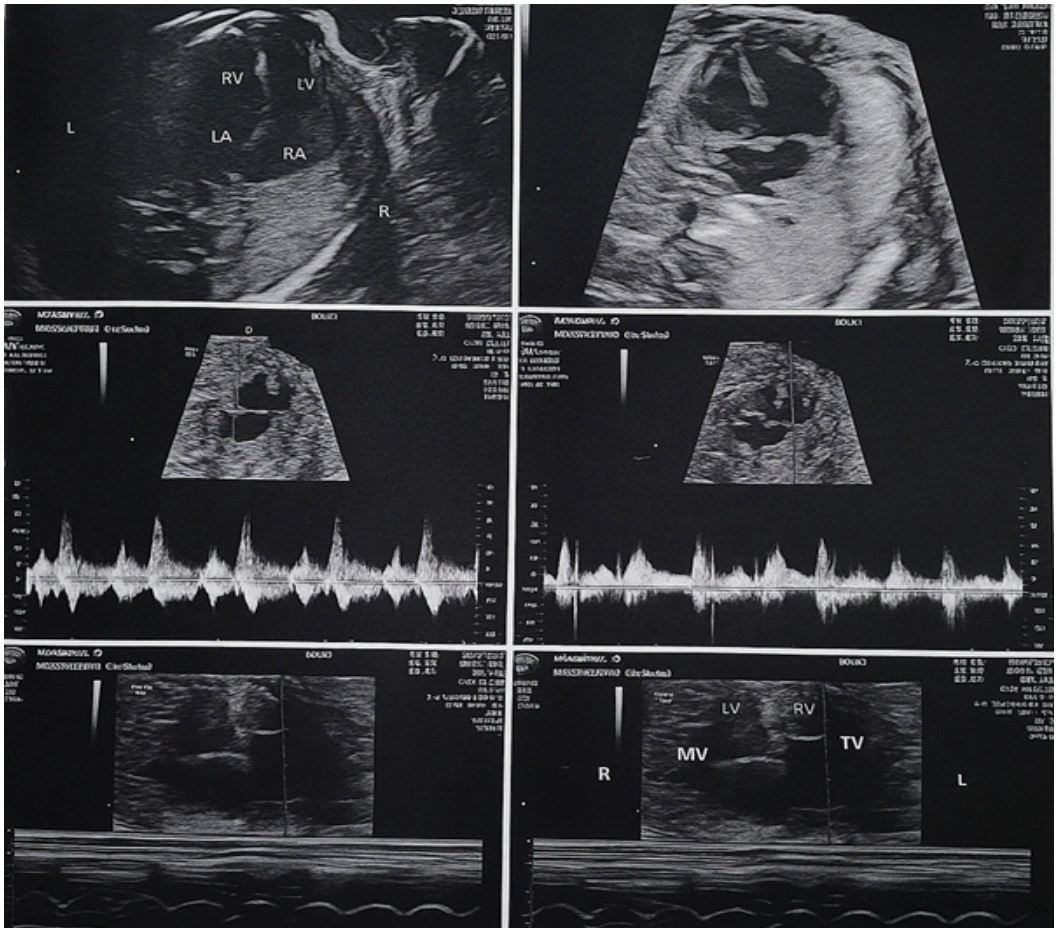

Figure 1: Second trimester ultra scan.

Case presentation

A 28-year-old gravida 2, para 1, abortion 1, living 1 (G2P1A1L1) woman at 25 weeks and 4 days gestation was referred from a primary health center for further evaluation of a suspected fetal congenital anomaly. Her obstetric history included one previous full-term vaginal delivery and a spontaneous abortion at 12 weeks of gestation. The ongoing pregnancy was non-consanguineous. She was a known case of gestational diabetes mellitus (GDM), well-managed with insulin therapy, and had a background of chronic pancreatitis. Her glycemic control was optimal, with a recent HbA1c of 5.4%.

A detailed second-trimester targeted anomaly scan, along with fetal echocardiography, revealed complex cardiac abnormalities. The fetal cranium showed normal ossification with preserved midline structures, including the falx cerebri, cavum septum pellucidum, and bilateral thalami. Both lateral ventricles and posterior fossa structures were within normal limits. No craniofacial anomalies were detected, with a well-formed facial profile and normal visualization of the orbits, nose, and lips. Cardiac evaluation revealed a severe constellation of findings. The cardiac apex was directed to the left, but there was evidence of atrioventricular discordance. The morphological right ventricle was located on the left side and was connected to the left atrium, and the tricuspid valve was significantly displaced toward the apex, consistent with a left-sided Ebstein anomaly. A morphological left ventricle on the right side was connected to the right atrium.

Additionally, an inlet-type ventricular septal defect (VSD) was found. Ventriculo-arterial discordance was also noted: the aorta arose from the morphological right ventricle, while the pulmonary artery originated from the morphological left ventricle. Pulmonary venous return was appropriately directed to the left atrium, and systemic venous return was preserved with both superior vena cavae (right and persistent left) and the inferior vena cava draining into the right atrium. The dilated coronary sinus appeared due to drainage from the left SVC. Despite these abnormalities, the overall cardiac size and rhythm were within normal parameters, and ductus venosus flow was visualized.

Assessment of extracardiac structures showed a structurally intact thorax and abdomen. The stomach bubble was identified below the diaphragm, and the gallbladder and bowel appeared normal in echogenicity without evidence of mass lesions or calcifications. Both kidneys and the urinary bladder were normal. Skeletal survey showed normal spine curvature and ossification, with intact overlying skin. Limbs were appropriately formed, and digits were visualized bilaterally. Fetal biometric measurements correlated with the gestational age but revealed signs of early-onset fetal growth restriction (FGR), necessitating closer surveillance. Given the complexity of the intracardiac findings, a diagnosis of congenitally corrected transposition of the great arteries (ccTGA) with a left-sided Ebstein anomaly and double SVC was made. In light of these findings, genetic evaluation through amniocentesis with chromosomal microarray analysis (CMA) was recommended to investigate potential underlying genetic etiologies. A pediatric cardiology consultation was also

advised to assist in perinatal counseling and to plan for potential postnatal surgical intervention. Ultrasound images highlighted the complex cardiac anatomy with clear visualization of the malformed tricuspid valve and vessel orientations.

Discussion

Ebstein's abnormality with ccTGA is a rare and complicated deformity. Left-sided Ebstein anomaly is even rarer due to the embryological inversion of the ventricles in ccTGA. In this case, both atrioventricular and ventriculo-arterial discordance contributed to the unique anatomical presentation. The presence of double SVC is a further rarity [1,7,8]. Prenatal detection allows optimized planning for delivery at a tertiary cardiac center, with potential surgical intervention in early neonatal life. Advances in fetal echocardiography have improved early diagnosis of such anomalies [6,9].

A recent multicenter study emphasized the predictive role of cardiothoracic ratio (CTR), lung-to-head ratio (LHR), and fetal growth status in fetuses with severe Ebstein anomaly. Findings showed that a CTR >0.8 and LHR <0.25 were strongly associated with perinatal mortality [9-11]. In our case, early FGR was noted, warranting close surveillance and multidisciplinary planning. The anatomical spectrum of Ebstein anomaly is broad. Coacci et al. (2024) highlighted how features such as unguarded tricuspid orifice, severe tricuspid regurgitation, and RV myocardial thinning significantly correlate with adverse fetal outcomes [10]. Our case presented with apical displacement of the tricuspid valve on the left, associated with ccTGA and inlet VSD—features that amplify circulatory complexity [2,12]. While ccTGA can maintain physiologic circulation prenatally, long-term outcomes remain guarded. The morphological right ventricle, functioning as the systemic ventricle, is prone to eventual failure, especially in the presence of valve abnormalities. The left-sided tricuspid valve in ccTGA is particularly susceptible to regurgitation and dysfunction [3,7,13].

The identification of double SVC, albeit often incidental, is clinically relevant. It may complicate cardiac interventions and central venous access and is associated with other congenital anomalies in up to 11% of cases [8]. From a maternal perspective, gestational diabetes mellitus (GDM) has been linked to congenital heart defects, although the precise mechanisms remain under study. Good glycemic control (HbA1c 5.4%) may have mitigated additional risks in this case, though chronic pancreatitis represents a compounding metabolic factor [11,12]. Importantly, novel prenatal therapies are being explored. A recent study reported the use of maternal NSAIDs to promote ductal constriction in fetuses with circular shunt physiology from severe Ebstein anomaly [14]. Despite the improved perinatal outcomes, the risk of oligohydramnios remains substantial [15]. Postnatal management should include advanced imaging like echocardiography and cardiac MRI to confirm anatomy and guide interventions. Depending on the severity of tricuspid regurgitation and ventricular function, early surgical repair, such as the cone procedure or systemic ventricular support, may be warranted [1,5].

Conclusion

This case underscores the capability of prenatal imaging to identify and characterize rare fetal cardiac anomalies. The combination of left-sided Ebstein anomaly, ccTGA, and double SVC has significant implications for neonatal prognosis and management, warranting multidisciplinary prenatal counseling and planning.

Declarations

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Conflicts of interest: The authors have no conflicts of interest to declare.

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