21 years old, 1st pregnancy - Obstetric ultrasound 18w: left heart axis deviation + suspicion of cardiac malformation

Referred to fetal cardiology

Fetal echocardiogram (20w): cardiomegaly, left axis deviation, normal atrioventricular connection and no signs of HF (cardiovascular profile score of 10 points).

4 chamber view: chamber discrepancy. A shift towards the LVOT, exposed what appeared to be a VSD (arrow), thickened aortic valve with severe insufficiency (asterisk; figure 1).

Detailed short axis view depicted an intact IVS, a perimembranous aneurysmal septum protruding to the RVOT (figure 2) favoring obstruction, but not interfering with the pulmonary valve movement.

Regurgitant aortic flow seemed to originate mostly out of the aortic valve, through a tunnel (arrow) surrounding the aortic annulus (figure 3 and 4) resembling a double ring around the aortic annulus – Cockade sign. Final diagnostic: AORTIC-LEFT VENTRICLE TUNNEL.

Regular follow-up on Fetal Cardiology.

30W: hydrops, normal flow in the ductus venous, umbilical artery and vein, shortening fraction >28%), severe LV dilation with preserved function, intermittent diastole in umbilical artery and absence of diastolic flow on median cerebral artery.

Neonatal management

Surgery was performed at 48 hours of life.

The tunnel was closed with two heterologous pericardial patches on both sides. A severely dysplastic aortic valve was observed. No aortic valve intervention or tunnel plication was performed. Cardiopulmonary bypass total time was 120min. Transoesophageal echocardiogram showed mild aortic regurgitation and moderate biventricular dysfunction. The postoperative period was uneventful, and the baby was discharged 19 days after surgery.

LEARNING POINTS

- Although challenging, ALVT diagnosis has been shifting to earlier gestational ages, which improves survival rates.
- Most of the cases of antenatal diagnosis describe challenges in differential diagnosis with DORV: visualization of a large perimembranous VSD with septal deviation towards RVOT.
- Prenatal diagnosis is crucial to inform the parents about the possibility of progression during fetal life, the incapability of predicting of such an outcome and the high chance of requiring neonatal surgical intervention.

Abbreviations: W: week; HF: heart failure; LVOT: left ventricle outflow tract; VSD: ventricle septal defect; IVS: interventricular septum; RVOT: right ventricular outflow; DORV: double outlet right ventricle; ALVT: aortic-left ventricle tunnel; LV: left ventricle