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Prenatal Diagnosis of an uncommon form of a hypoplastic left heart syndrome variant

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BACKGROUND

- Mitral valve dysplasia syndrome (MVDS) is a rare form of congenital heart disease, similar to hypoplastic left heart syndrome (HLHS).
- Prenatal identification is important for counseling, delivery planning and postnatal management.

CASE

- A 39 year-old woman underwent fetal echocardiography at ~34 weeks gestation for evaluation of fetal cardiomegaly and hydrops revealing: biatrial enlargement, mild-to-moderate mitral and tricuspid valve insufficiency, echobright mitral valve apparatus, biventricular dilation, severe LV fibroelastosis (EFE) and systolic dysfunction, mild hypoplasia of aortic valve annulus and aortic arch, and a thick restrictive atrial septum (RAS) with left to right flow (Fig 2).

DECISION MAKING

- Although critical aortic stenosis was considered initially, MVDS seemed more likely given above characteristic findings.
- Delivery planning included elective C-section with standby catheterization laboratory and ECMO teams, given restrictive atrial septum and cardiac dysfunction.
- Patient was listed for transplant as biventricular or single ventricle repair were deemed unfeasible, given valvar insufficiency, LV dysfunction and EFE (Fig 1,3).

Fig 1. Gross pathological specimen of the explanted heart confirming the prenatal diagnosis of a dysplastic mitral valve with short leaflets and chordae and significant left ventricular fibroelastosis (EFE).

Fig 2. Fetal echocardiographic images showing: Severe left ventricular (LV) fibroelastosis (EFE) and a thick atrial septum, Left atrial (LA) and left ventricular dilation with echobright mitral valve (MV)apparatus and Mild-to-moderate mitral regurgitation (MR).

Fig 3. Post-natal transthoracic subcostal image showing biatrial enlargement with a thick and restrictive atrial septum. Parasternal long axis image showing severe LV fibroelastosis (EFE), dysplastic mitral valve with short leaflets and chordae and a hypoplastic aortic valve (AoV) annulus. (RA – right atrium, PFO – patent foramen ovale).