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Goyal, Anmol, "Prenatal diagnosis of an uncommon form of a hypoplastic left heart syndrome variant." (2021). *Research Days*. 5.

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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).

Mitral Valve Dysplasia Syndrome: An important differential in fetuses with hypoplastic left sided structures

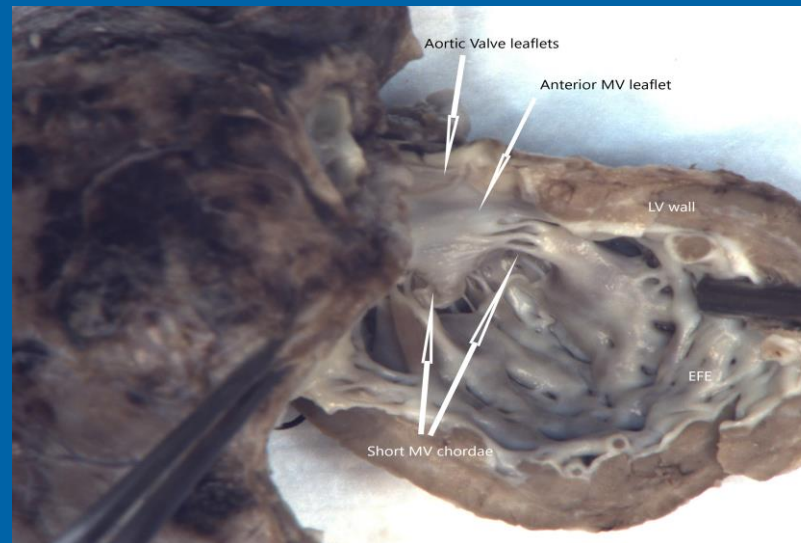
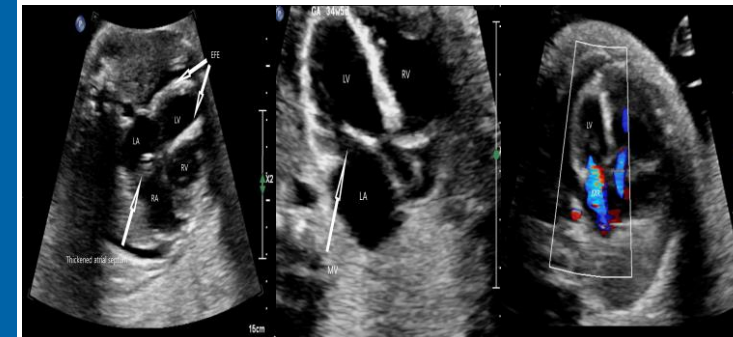


Fig 1. Gross pathological specimen of the explanted heart confirming the prenatal diagnosis of a dysplastic mitral valve with short leaflets and chordae and significant left ventricular fibroelastosis (EFE).

FIGURE 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).

FIGURE 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].