Prenatal diagnosis of an uncommon form of a hypoplastic left heart syndrome variant.

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

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).

CONCLUSION

- MVDS is uncommon but should be considered with HLHS differential, in presence of a normal or dilated left atrium and ventricle, LV dysfunction and EFE, RAS and aortic valve and arch hypoplasia.
- Planned delivery and immediate postnatal atrial septostomy is warranted and cardiac transplantation is often the only therapeutic option.

DISCLOSURE INFORMATION

- Authors have no disclosures to report

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