

## PW2-3

### Aortic Stenosis and Severe Mitral Regurgitation in the Fetus Resulting in Giant Left Atrium and Hydrops: Pathophysiology, Outcomes, and Preliminary Experience with Prenatal Cardiac Intervention

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#### Objectives.

To review anatomic, physiologic, and clinical features of fetuses and neonates with severe mitral regurgitation (MR) in conjunction with aortic stenosis (AS), and left ventricular (LV) and left atrial (LA) dilation, and present preliminary results of prenatal intervention for this condition.

#### Background.

Severe fetal valvar AS with an abnormal mitral valve (MV) and MR can lead to left heart dilation, with consequent compression of the right ventricle (RV) (figure 1a,b); hydrops and low cardiac output is often associated.

Methods. Retrospective review of fetuses diagnosed with AS, severe MR, and LA dilation (2002-2009) and neonates with the same combination of abnormalities (1988-2009).

#### Results.

Fourteen fetuses and 7 neonates were investigated. Eleven fetuses had severe hydrops; all had polyhydramnios and a structurally abnormal MV, abnormal MV inflow pattern, a restrictive/intact atrial septum, retrograde flow in the transverse aortic arch, and compression of the right heart. The mean indexed RV output was  $326 \pm 160$  mL/kg/min, substantially lower than the normal average fetal combined ventricular output of  $550 \pm 150$  mL/kg/min. Ten fetuses underwent prenatal cardiac intervention: aortic valvuloplasty (n=8) and/or atrial septal dilation/stenting (n=5). Seven of these, and 11 overall, were liveborn. Nine patients died (median age 6 days), 2 patients are currently alive. All 7 patients diagnosed in the neonatal period died (median age 1 day).

#### Conclusions.

AS associated with significant MR in the fetus can cause severe LA enlargement, leading to low cardiac output and hydrops. Despite the potential advantages of early prenatal diagnosis and both fetal and neonatal interventions, this rare complex of anomalies carries a poor prognosis.

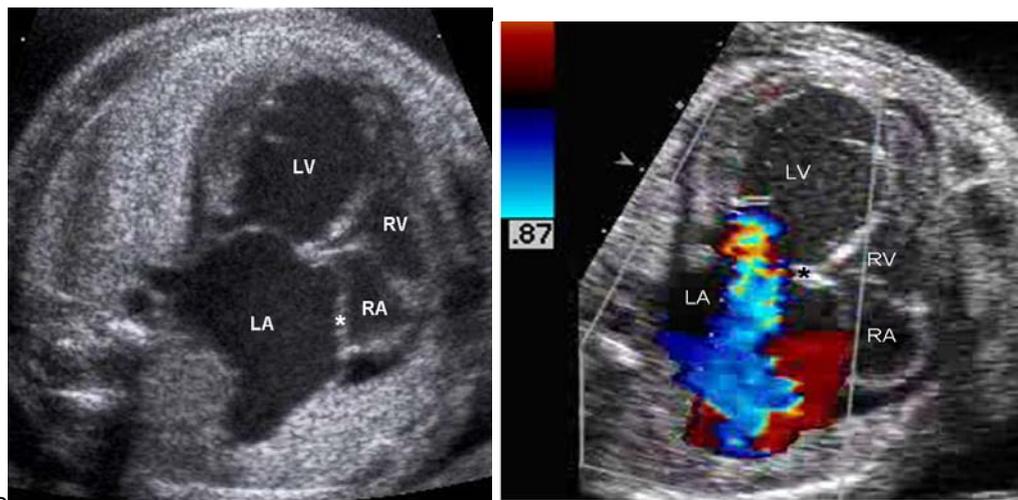


Figure 1a, b

LV, left ventricle, LA, left atrium, RA, right atrium, RV, right ventricle, \*bright anterior mitral valve leaflet