

**Postnatal left ventricular performance in prenatally treated patients with critical aortic stenosis and endocardial fibroelastosis**

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Intrauterine treatment of fetuses with critical aortic stenosis (AS) and evolving hypoplastic left heart syndrome may save the left ventricle (LV) for a postnatal biventricular circulation. However, the long-term fate of these LVs with varying degrees of endocardial fibroelastosis (EFE) is unknown. The purpose of this study was to assess LV performance in surviving infants and young children, who underwent fetal aortic valvuloplasty

**Patients and methods:** Between 12/2001 and 1/2011 29 fetal aortic valvuloplasties have been performed in 27 patients with technical success in 19 patients (2 patients still in-utero). Ten patients went on to a biventricular circulation with 2 infant deaths. So 8 patients formed the study group with a median follow-up of 48,4 months (12 – 71 months). Their median fetal pre-interventional LV long-axis z-score was 0,54 (-0,99 – 2,1) Clinical records, echocardiographic or angiographic data were reviewed retrospectively to assess LV performance

**Results:** 6/8 neonates had an aortic balloon dilatation, 1 neonate a surgical aortic valvotomy in the first week of life. During the follow-up period, all but 1 patient (age 4 years) needed a Ross-Konno Operation (partial EFE resection in 1 patient) at a median age of 15 days (7d – 49 months). All patients are without any clinical symptoms, but still have evidence of EFE by echo or MRI and are on ACE-inhibitor therapy, 2 additionally on antiarrhythmic therapy. 1 patient needed a pacemaker due to postoperative complete AV block. LVEDD z-score was >2 in 4 patients (median 2,1; range: 0,36 to 3,0), LV SF was < 28% in 3 patients (median: 30%; range 22 – 45%), evidence of elevated pulmonary artery pressure was present in 1 patient (1 year postop, 50% systemic pressure by echo), LAP (available in 4/8 patients) between 11 and 14 mmHg.

**Conclusions:** Children with biventricular circulation following successful in-utero aortic valvuloplasty show satisfactory, but not normal LV performance during short-term follow-up. In most cases multiple interventions and early aortic valve replacement were necessary. EFE resection at the time of surgery may be a strategy to further improve LV performance and positively impact the long-term fate of the LV.