Neonatal management of critical aortic stenosis after in-utero valvuloplasty: role of Ross-Konno operation

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Neonates with critical aortic stenosis (AS) after in-utero valvuloplasty usually present with borderline left ventricular size, function and varying degrees of endocardial fibroelastosis. The decision towards a biventricular circulation remains a challenge. The purpose of this study was to investigate the role of an early Ross-Konno (RK) operation on a biventricular outcome.

Between 12/2001 and 1/2012 we attempted 38 fetal aortic valvuloplasties in 35 fetuses (median GA 26+4 weeks; 21+4 to 32+1), 5 of these with advanced end stage heart failure and hydrops. The procedure was successful in 27/35 fetuses (77%), there were 3 IUDs in this group, so 24 children were live-born in 9 different centers in Austria, Germany, Poland, Denmark and Italy. According to the respective criteria of these centers, 18/24 (75%) newborns were managed towards a biventricular circulation either with aortic balloon dilation alone or followed by a RK operation.

Balloon dilation alone was effective as a first line therapy in only 5/18 newborns, two of them needed later a RK operation at 1 and 4 years respectively, 3 patients died due to left heart failure and 2 had to go on to a Norwood procedure with 1 death. The remaining 8 patients underwent a neonatal RK at a median age of 14 days (7-19 days). One child died 37 days after successful surgery due to NEC. At a median follow-up of 2.3 years (0.3-6.8 years) all children are biventricular, with sufficient LV function and normal pulmonary artery pressures. Children born in centers offering neonatal RK surgery had higher probability to achieve a biventricular circulation, than children born in centers without this option: 10/16 (62.5%) vs 2/8 (25%)

Conclusions: In neonates with critical AS born after successful in-utero valvuloplasty and non-effective post-partum aortic valvuloplasty, early aortic valve replacement with a RK operation appears to be effective in improving LV function and increasing the chance for a biventricular circulation.