
Children's Heart Center Linz, Department of Pediatric Cardiology (1), Linz, Austria
Children's Heart Center Linz, Department of Pediatric Cardiac Surgery, Linz, Austria (2)
Department of Prenatal Medicine, Children's and Women's Hospital Linz, Austria (3)

Background: newborns with critical aortic stenosis (CAS) suffer from severe left ventricular (LV) hypertrophy and dysfunction. Interventional aortic valve dilation usually leaves significant increased LV myocardial stress. Early aortic valve replacement may allow improved LV recovery and long-term outcome.

Aim: to review our experience with neonatal RK surgery

Patients: Between 10/2000 and 1/2013 22 children with CAS, median age: 17d (7-547d); median weight: 3,03 kg (2,18-9,2 kg) underwent early RK surgery in our institution. Hypoplastic or interrupted aortic arch was additionally corrected in 9 patients, 5 of them had a VSD closure. Severe endocardial fibroelastosis (EFE) was present in 8 neonates. Seven had a fetal aortic valve dilation, 16/22 a postnatal aortic valve dilation. A 12 mm Contegra valve was used as a right-sided conduit in 6 patients, homografts from 8-14 mm in 16 children. Median follow-up was 5,5 years (1,0-12,3 years)

Results: early mortality was 3/22 (14%), there was no late mortality. 1 patient required a pacemaker due to complete heart block, 1 patient had a mitral valve replacement. Conduit replacement was necessary in 9 patients after a median period of 2,2 years (0,6 – 11 years), 3 had a second replacement median 3,3 years (2,5-4,1years) later. Neo-aortic valves showed excellent function without gradient, no aortic regurgitation in 8, grade I in 15 patients. Aortic valves showed good growth (median z-score after 3 years: 1,92; range: -0,6-2,94), valve function remained stable, however z-scores of the aortic sinuses were significantly larger (median 2,77; range: 0,79-4,51; p<0,0001). Out of 8 patients with severe EFE and LV Dysfunction there were 2 neonatal deaths. LV SF improved significantly from median 11,8% (5-23%) to 30,7% (10-41%). PA pressures were normal in all.

Conclusion: in neonates and young children with CAS, early RK seems to be a safe and effective treatment to unload the LV and to allow recovery of LV function. Early conduit replacement may be necessary. Neo-aortic valve showed good growth and function but dilation of neo-aortic sinuses may occur and may become a concern in the future.