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Long-term Outcome of Surgical Aortic Valvulotomy in Pediatric Patients - A Retrospective Single Center Study Over 30 Years

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Introduction

Surgical aortic valvulotomy (SAV) has been part of the armamentarium of congenital heart surgery from the early days on. SAV allows to address the individual valve pathology on a lesion specific and highly controlled manner. Additionally, to opening the zones of commissural fusion, leaflet shaving, where thickened and dysplastic leaflets are thinned can be performed. After SAV there is generally a low risk of aortic regurgitation. The study objective was to report on survival and freedom from re-operation in the third decade after SAV.

Methods

A retrospective analysis of all patients aged under 18 years at time of surgery, who underwent SAV from May 1985 until April 2020 was conducted. A complete mortality follow-up until April 30th, 2020 was obtained for 98.6% (68/69) of patients. One patient was only transferred to the center for surgery and mainly followed at a non-Austrian center. Eight closed transventricular valvulotomies, which had been performed during the study period prior to introduction of balloon aortic valvuloplasty (BAV) were not included in the study population. Time-related events were assessed using Kaplan- Meier estimator. Univariable Cox-proportional hazard modelling was used to determine risk factors for mortality.

Results

From May 1985 until April 2020 69 patients (71% male, 49/69) underwent 71 SAVs. Median age at time of surgery was 0.3 years (IQR 0 - 3,5 years). 67.6% (48/71) were younger than 1 year of age and 28.2% (20/71) were neonates. Aortic valve anatomy was as follows: unicuspidal (7%; 5/71), bicuspid (85.9%; 61/71) and tricuspid (7%, 5/71). In five cases (7%) a BAV had been not feasible or unsuccessful in establishing an acceptable hemodynamic situation. Three patients (4.3%) required extracorporeal membrane oxygenation and seven early deaths (9.9%) occurred. All early deaths occurred in neonates with critical aortic stenosis. Three patients had already undergone early aortic valve re-operation (two homografts, one Ross-Konno). There were 5 late deaths and Kaplan-Meier estimated survival was 86.8% ± 4.1% at 10 years, 83% ± 4.7% at 20 years and 78.6% ± 6.2% at 30 years. Risk factors associated with mortality identified on univariable Cox-proportional hazards analysis were neonatal period (HR 17.2, 95% CI 3.7-81.2; $p < 0.001$) and endocardial fibroelastosis (HR 8.6, 95% CI 2.8-27.1; $p < 0.001$). Freedom from aortic valve re-operation (aortic valve repair and aortic valve replacement) was 58.2% ± 6.3% at 10 years, 33.9% ± 6.4% at 20 years and 27.1% ± 7.9% at 30 years.

Conclusion

Congenital aortic valve stenosis often requires intervention in early childhood. Neonates with critical aortic stenosis, who undergo urgent intervention early after birth, represent a high-risk group with high early morality. Endocardial fibroelastosis, which is common in neonates with critical aortic stenosis was found as a predictor for global mortality. Valvulotomy of any kind is a palliative procedure, and the majority of patients will require repeated re-interventions over their lifetime. However, the presented 30-year freedom from re-operation rates are encouraging results in delaying and in some cases even avoiding valve replacement after SAV.