MEDICAL UNIVERSITY OF VIENNA

Aortic Root and Ascending Aorta Replacement in Pediatric Patients

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Introduction

Aortic root and ascending aorta replacements (AARs) are scarcely performed in pediatric patients and are mostly performed in patients with connective tissue disease, such as Marfan syndrome and Loeys-Dietz syndrome. Besides the Bentall procedure, where AAR is performed using a composite valve graft, valve sparing root replacement (VSRR) techniques gained attention in the pediatric field.

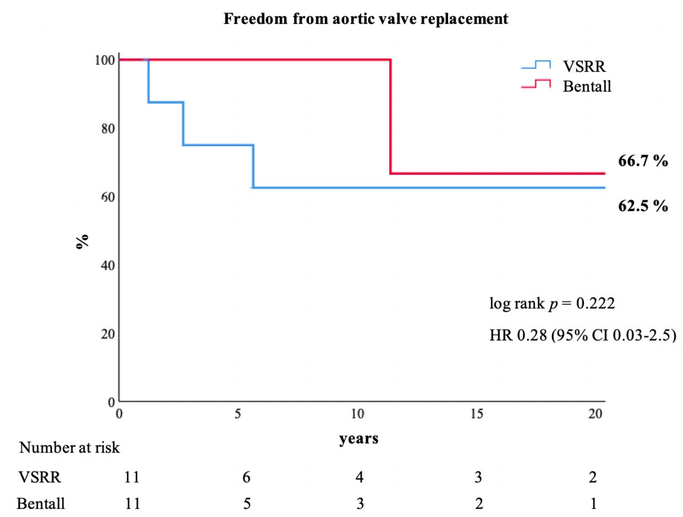
Methods

A chart review of all patients less than 18 years of age who had AAR between May 1985 and April 2020 was conducted. During the study period 20 patients underwent 22 AARs: 11 VSSR and 11 Bentall. Mortality was cross-checked with the national health insurance data base providing a mortality follow-up until April 2020.

Results

From May 1985 until April 2020, 20 patients (70% male, 45% connective tissue disease, 15% complex congenital heart disease) underwent 11 VSRRs and 11 Bentall procedures. Two patients (Loeys-Dietz syndrome and Marfan syndrome) underwent a Bentall procedure 5.6 and 1.2 years after VSRR respectively. Median age at time of operation was not different (p = 0.365) between the cohorts with 14.2 years (IQR 12.7-15.8 years) and 13.2 years (IQR 8.2-14.8 years). There were no early deaths. One patient required ECMO support after VSRR. Permanent pacemaker implantation for complete AV-block was required in three patients (VSRR: n = 2, Bentall: n = 1; $p > 0.99$). A patient with neonatal Marfan syndrome died 5.6 years after VSRR. Kaplan-Meier estimated survival at 30 years was 90% ± 9.5%.

Freedom from aortic valve re-operation was not different (p = 0.222) between the VSRR and the Bentall cohort with 62.5% ± 17.1% and 66.7% ± 27.2% at 25 years. Four VSRRs were re-operated (2 Bentall, 1 mechanical AVR, 1 decellularized homograft) and one Bentall was re-operated for pannus formation after 11.4 years (mechanical AVR). Freedom from any AAR related re-operation was 25.7% ± 19.9% at 30 years. Freedom from any AAR-related re-operation did not differ (p = 0.085) between the cohorts with 37.9% ± 19% and 66.7% ± 27.2%. One Marfan patient required early re-operation after VSRR for kinking of the RCA and a another Marfan patient was re-operated 15.8 years after VSRR for coronary artery button aneurysm.



Conclusion

Composite mechanical valve graft replacement has long been standard procedure for replacement of the aortic root, but VSRR has emerged as an option in the pediatric population with aortic root dilatation including patients after repair of cono-truncal anomalies. In patients with connective tissue disease it is important to fashion the diameter of the coronary buttons as small as possible to prevent development of coronary button aneurysm.