JOURNAL OF THE AMERICAN COLLEGE OF CARDIOLOGY
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PUBLISHED BY ELSEVIER

VOL. 67, NO. 24, 2016
ISSN 0735-1097/$36.00
http://dx.doi.org/10.1016/j.jacc.2016.04.021

Aortic Valve Replacement and the Ross Operation in Children and Young Adults

Mansour T.A. Sharabiani, PhD,[a] Dan M. Dorobantu, MD,[b,c] Alireza S. Mahani, PhD,[d] Mark Turner, PhD,[b] Andrew J. Peter Tometzki, MBChB,[b] Gianni D. Angelini, MD,[a,b] Andrew J. Parry, MBChB,[b] Massimo Caputo, MD,[b] Serban C. Stoica, MD[b]

ABSTRACT

BACKGROUND There are several options available for aortic valve replacement (AVR), with few comparative reports in the literature. The optimal choice for AVR in each age group is not clear.

OBJECTIVES The study sought to report and compare outcomes after AVR in the young using data from a national database.

METHODS AVR procedures were compared after advanced matching, both in pairs and in a 3-way manner, using a Bayesian dynamic survival model.

RESULTS A total of 1,501 patients who underwent AVR in the United Kingdom between 2000 and 2012 were included. Of these, 47.8% had a Ross procedure, 37.8% a mechanical AVR, 10.9% a bioprosthesis AVR, and 3.5% a homograft AVR, with Ross patients being significantly younger when compared to the other groups. Overall survival at 12 years was 94.6%. In children, the Ross procedure had a 12.7% higher event-free probability (death or any reintervention) at 10 years when compared to mechanical AVR ($p = 0.05$). We also compared all procedures except the homograft in a matched population of young adults, where the bioprosthesis had the lowest event-free probability of 78.8%, followed by comparable results in mechanical AVR and Ross, with 86.3% and 89.6%, respectively. Younger age was associated with mortality and pulmonary reintervention in the Ross group and with aortic reintervention in the mechanical AVR. Of all 3 options, only the patients undergoing the Ross procedure approached the survival of the general population.

CONCLUSIONS AVR in the young achieves good results, with the Ross being overall better suited for this age group, especially in children. Although freedom from aortic valve reintervention is superior after the Ross procedure, the need for homograft reinterventions is an issue to take into account. All methods have advantages and limitations, with reinterventions being an issue in the long term for all, more crucially in smaller children. (J Am Coll Cardiol 2016;67:2858–70)

Young patients with aortic valve (AoV) disease can be palliated by transcatheter or surgical methods but most will eventually require an aortic valve replacement (AVR). There are several options available for children and young adults: mechanical valve replacement (M-AVR), pulmonary autograft or the Ross procedure (R-AVR), biological heterograft (B-AVR), and homograft valves (H-AVR).

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From the [a]Department of Cardiothoracic Surgery, Imperial College, London, United Kingdom; [b]Department of Cardiothoracic Surgery, University Hospitals Bristol NHS Trust, Bristol, United Kingdom; [c]Cardiology Department, "Prof. C.C. Iliescu" Emergency Institute for Cardiovascular Diseases, Bucharest, Romania; and [d]Sentrana Inc., Washington, DC. This research was supported by National Institute for Health Research (NIHR) Bristol Cardiovascular Biomedical Research Unit and used data provided by the National Institute for Cardiovascular Outcomes Research, as part of the National Congenital Heart Disease Audit (NCHDA). The NCHDA is commissioned by the Healthcare Quality Improvement Partnership (HQIP) as part of the National Clinical Audit and Patient Outcomes Programme (NCAPOP) and within the National Health Service, United Kingdom. The views expressed are those of the authors and not necessarily those of the National Health Service, NIHR, or Department of Health. Dr. Turner has served as a proctor for Medtronic and Edwards Lifesciences. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose. Drs. Sharabiani and Dorobantu contributed equally to this work.

Manuscript received February 18, 2016; revised manuscript received March 29, 2016, accepted April 5, 2016.

Each has its uses and limitations and, more importantly, no option is perfect. There is a set of qualities an AoV substitute should have, and presently there is no choice that can achieve them all, with many factors influencing the choice and long-term results of an AVR. Data on outcomes vary, with few national and even fewer comparative studies. Multicenter studies would be best suited to describe and compare modern results. The objective of the current study is to describe early and long-term survival and freedom from reintervention in a national population of consecutive, unselected young patients, to compare the results of the main types of AVR in appropriately matched populations and to identify factors influencing outcome for each procedure type.

SEE PAGE 2871

METHODS

The National Congenital Heart Disease Audit collects validated key data on cardiac procedures from all the UK units, using a mechanism for data capture, cleaning, and validation similar to that for adult cardiac surgery (1).

Using linkage with census records at the Office of National Statistics, the audit database publicly reports survival rates at 30 days and 1 year following the index procedure online. Linkage with survival registries of Northern Ireland and Scotland cannot be done consistently with the patient's personal identification number, whereas a minority of them either have errors in their social data or are foreign. This resulted in 10.6% of patients not having data beyond 30 days, due to administrative reasons. The remaining patients have long-term follow-up from either the Office of National Statistics or from other entries in the database.

Indications for each operation were established by multidisciplinary teams at each center. Diagnosis and procedure codes from the European Pediatric Cardiac Code Short List are used for reporting of data. The quality index for key procedure fields is above 95%. The completeness and accuracy of noncritical data fields cannot be estimated without detailed patient-level data from each center, but there is no indication of systematic, persistent errors in reporting. The need for patient-level consent to participate in this retrospective study was waived by the National Institute for Cardiovascular Outcomes Research Board.

PATIENT SELECTION. All available data on patients undergoing an AoV procedure for a congenital cause between April 2000 and March 2012 were selected and anonymized. Out of these, 2,767 had an AVR.

We excluded 671 patients who were >40 years of age, as we considered that above this age degenerative disease is more prevalent. We also excluded those patients with associated complex heart abnormalities (n = 193), rheumatic fever (n = 15), unclassified AVR procedures (n = 313), and unknown age at index procedure (n = 74). The excluded complex heart abnormalities were univentricular conditions, valvar atresias, interrupted aortic arch, atrioventricular septal defect, transposition of great arteries, common arterial trunk, Fallot-type defects, severe vascular abnormalities (e.g., major aortopulmonary collaterals), and atrial isomerism. Unclassified AVR procedures were due to errors in reporting (i.e., using a general "Aortic valve replacement" code).

Reinterventions were defined as either reoperations or catheter-based procedures related to the AoV or root and to the pulmonary valve and right ventricle outflow tract (RVOT) for the Ross operation group. Not included were early reinterventions (within 30 days, considered connected to intraoperative events and not prosthesis durability) and those aorta dilation/aneurysm repairs that were related to previous conditions (e.g., Marfan syndrome). When comparing the procedures, separate calculations were made for AoV reinterventions and any reinterventions, due to the fact that the Ross procedure is at risk of both AoV and RVOT reinterventions. This was done to ensure that the comparisons between procedures can be properly interpreted, with both AoV and overall freedom from reintervention comparisons.

STATISTICAL ANALYSIS. Frequencies are given as absolute numbers and percentages, continuous values as median (interquartile range). Short-term mortality is calculated on the basis of 30-day life status. Population characteristics were compared using the Mann-Whitney *U* test, Kruskall-Wallis test, Student *t* test, and Fisher exact test. Descriptive estimates of long-term outcomes by AVR category and also for neonates are made with the Kaplan Meier method using mortality (all-cause) and reintervention, death being censoring for reintervention. Risk factor analysis for the B-AVR and H-AVR groups was performed using the log-rank test and Cox proportional hazards regression.

In addition to aggregate, nonparametric analysis of short- and long-term survival rates, we used a Bayesian dynamic survival model to perform Variable Importance Analysis and Procedure Comparison Analysis (2). The resulting dynamic HRs allowed us to

ABBREVIATIONS AND ACRONYMS

AoV = aortic valve
AVR = aortic valve replacement
B-AVR = biological heterograft
CI = confidence interval
H-AVR = homograft valve
HR = hazard ratio
M-AVR = mechanical valve replacement
R-AVR = Ross procedure
RVOT = right ventricle outflow tract