Title: The Ross operation after removal of a transcatheter aortic valve replacement in pediatric patients.

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Central Picture: Balloon-expandable TAVR prosthesis prior to removal
Central Message: The Ross procedure can be performed safely after TAVR removal in pediatric patients with good short-term outcomes. While mortality is low, morbidity is higher than the Ross performed alone.

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Introduction

The ideal choice for aortic valve replacement (AVR) in the pediatric population is unknown. Options include bioprosthetic and mechanical valves and the pulmonary autograft (Ross operation). Each contains a unique risk-benefit profile with no gold standard (1). Pediatric transcatheter aortic valve replacement (TAVR) is an intriguing option, and its use is expanding with short-term results being similar to those of surgical AVR (2). The Ross procedure, with improved autograft durability (4, sup ref 1), remains our preferred option. Currently, there are no data reporting the results of the Ross operation after TAVR removal from a native left ventricular outflow track in either adults or children. The purpose of our study is to report and better understand the safety and short-term outcomes of Ross after TAVR removal in children.

Operative Technique

All procedures were performed on cardiopulmonary bypass with mild hypothermia (Video 1). The TAVR was removed by longitudinally dividing the prosthesis and wrapping the stent frame around a clamp to separate it from the aortic root (4). Our Ross technique has been previously reported (3). To summarize, the autograft is harvest from the right ventricular outflow track and the muscular cuff is trimmed to within 2-3 mm of the pulmonary annulus. The autograft is implanted in a subannular position using interrupted monofilament sutures. If the native aortic annulus is greater than 25mm, an extra-aortic 3-4mm Dacron subannular stabilization graft is secured by passing six pledgeted sutures from within the left ventricular outflow track under the nadirs of each sinus and intercommissural triangle. The chosen graft is sized 5-6 mm larger than the size of pulmonary autograft annulus and then tied down over a dilator sized to the autograft annulus. Following autograft implantation, excess tissue above the autograft commissural posts
is resected to within 2 mm of the commissures. For patients with either a paucity of native aortic
tissue or those with enlarged aortic aneurysms, we insert a short Dacron tube graft to stabilize the
sinotubular junction of the autograft (sizes typically 24-26mm).

Cases
This study was exempted from the University of Utah IRB because case reports including less
than or equal to three patients are exempted from review and consent if all data is deidentified
and is part of standard of care.

Descriptions of native aortic valve pathology, initial catheter-based procedures, and indications
for TAVR are listed in Table 1. All three TAVR implants were balloon-expandable valves.

Patient 1: 20-year-old male received a 26mm TAVR for mixed aortic insufficiency/stenosis
(AI/AS). Twenty-four months later he developed progressive aortic root and ascending aortic
dilation with AS (mean 26 mmHg). He had partial aortomitral disruption during TAVR removal
requiring repair by resuspending the injured portion of the anterior leaflet of the mitral valve to
the annulus through the left ventricular outflow track. He received a 27mm pulmonary
homograft and a 24mm ascending aortic interposition graft for an ascending aneurysm.

Patient 2: 16-year-old male received a 29mm TAVR for AI/AS. Twenty-six months later he
developed moderate AI through a paravalvar leak with increasing left ventricular dilation (Sup
Fig 1). He received subaortic annular stabilization and a 24 mm pulmonary homograft with a
22mm ascending aortic interposition graft for an ascending aneurysm.
Patient 3: 15-year-old male received a 29mm TAVR for AI/AS. Thirty months later he developed severe AS (mean 51mm Hg) and mildly diminished left ventricular function. He received subaortic annular stabilization and a 28mm pulmonary homograft with a 22mm ascending aortic interposition graft for an ascending aneurysm.

Results

There were no operative mortalities, no neurologic events, one patient with heart block and one postoperative bleeding event. Mean ICU length of stay was 3 days (range 2-4), mean total hospital length of stay 6.3 days (range 4-12) and all patients were extubated within 24 hours of surgery. Mean cardiopulmonary bypass time was 228 minutes and mean aortic cross clamp time was 197 minutes.

One patient was empirically started on dual antiplatelet therapy postoperatively for a presumed embolic event and suffered a gastrointestinal bleed postoperative day 14 with no long-term sequelae. Another patient required implantation of a permanent pacemaker which was complicated by left subclavian vein thrombosis requiring pacemaker revision. Follow up echo and clinical status are listed in Table 2.

Discussion

We are the first to report that the Ross operation after TAVR removal from native left ventricular outflow tracks can be performed with good short-term outcomes in the pediatric population. The Ross is a well-proven operation with excellent long-term outcomes and is our preferred surgical
option for AVR in children. When performed at experienced centers, mortality and major
morbidity are low, and the autograft offers excellent hemodynamics and growth potential (1,3,4).

Current adult data suggest that AVR after TAVR removal results in significant morbidity (mitral
take intervention 32%, bleeding 12%, stroke 9%, and heart block 18%) and mortality of 13%
(5). Our limited pediatric experience of Ross after TAVR removal shows decreased overall
morbidity (although a similar incidence of heart block and mitral intervention) and mortality.
The reasons for these different experiences are speculative but likely related to the lack of
pediatric preoperative comorbidities and anatomic substrate.

Our programmatic approach to aortic valve disease has evolved. Historically, referrals for
TAVR were sent directly from referring cardiologists to the interventional cardiology team
without surgical consultation. Recently, a multidisciplinary valve clinic was initiated where all
patients undergo echo and CT evaluations with both a cardiologist and surgeon meeting with the
family to decide the best course of treatment. To date, our program has implanted 35 TAVR
valves in the pediatric population (including 13 valve-in-valves and 22 native outflow tracks)
and our short-term results have been reported (2). We currently do not have any long-term data
about the durability of the TAVR valve in children and cannot make a more meaningful long-
term comparison of TAVR vs. Ross.

To date, we have explanted 10 TAVR valves; 5 patients received the Ross operation (one was a
TAVR valve-in-valve in a bioprosthetic valve and the fifth occurred following manuscript
submission). In addition, 2 patients had a tissue AVR, 2 patients had a mechanical AVR and one
patient required a ventricular assist device and transplantation. Two patients developed endocarditis requiring TAVR removal.

Pitfalls

All of our patients had mixed AI/AS with large and pliable annuli compared to adults with mostly AS and calcified annuli. Despite this, we encountered challenges separating the TAVR from the mitral valve apparatus in all cases. Due to incomplete somatic growth, the TAVR implantation was commonly more proximal to protect the coronaries rendering the sealing cuff of the TAVR adhered to or closely abutting the anterior leaflet of the mitral valve. After the first explant we have started to transect the TAVR stent longitudinally to release tension and wrapping the remaining stent around a clamp has resulted in no mitral injuries. In contrast to adult series, our avoidance of self-expanding TAVR valves, has allowed for safe coronary button harvesting and no ascending aortic damage during prosthesis removal. However, in patients who received a TAVR in the native outflow track, we experienced aortic annular damage during removal. In such cases, additional hemostatic sutures were placed through the native remnant of the aortic wall and the autograft annulus for support.

Although of interest, our objective is not to report on a comparison of TAVR v. Ross AVR in the pediatric population. Such an analysis will require longitudinal data and follow up. That being said, our surgical bias and concerns for TAVR outcomes led to the creation of a valve clinic as described and in turn has lent to an exponential growth in our Ross program.

In conclusion, the Ross operation can be safely performed after TAVR removal with good short-term outcomes in the pediatric population. While mortality was low following Ross after TAVR removal, similar morbidities and technical challenges exist as in the adult population and more experience will be required to make meaningful comparisons.
References


2. Robertson Dm, Boucek DM, Martin MH, Gray RG, Griffiths ER, Eckhauser AW et al. Transcatheter and Surgical Aortic Valve Implantation in Children, Adolescents, and Young Adults with Congenital Heart Disease. Am J Cardiol. 2022 Jun 9; S0002-9149(22)00532-X. doi: 10.1016/j.amjcard.2022.04.056.


Supplemental References

Legend

Central Picture: Balloon-expandable TAVR prosthesis prior to removal

Table 1. Description of native aortic valve pathology, initial catheter-based intervention and indications for both TAVR and Ross operation.

Table 2. Echocardiographic and clinical patient follow-up data


Supplemental Figure 1. Preoperative echocardiography of a TAVR valve prior to removal and Ross operation. A. Color showing moderate paravalvar leak. B. Anatomical proximity of TAVR stent to the anterior leaflet of the mitral valve.
<table>
<thead>
<tr>
<th>PATIENT</th>
<th>Native AV Pathology</th>
<th>Initial AV Procedure</th>
<th>Age at Initial Procedure</th>
<th>Indication for TAVR</th>
<th>Age at TAVR</th>
<th>Indication for Ross</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Bicuspid Valve, Severe stenosis</td>
<td>Balloon valvuloplasty</td>
<td>13</td>
<td>Mixed-Severe AS (peak 103, mean 54mmHg), Moderate AI</td>
<td>17</td>
<td>Mixed AS/AI and Asc Ao Aneurysm 4.21cm</td>
</tr>
<tr>
<td>2</td>
<td>Unicuspid Valve, Severe stenosis</td>
<td>Balloon valvuloplasty</td>
<td>11</td>
<td>Severe AI, LVIDd 6.4cm</td>
<td>14</td>
<td>Moderate PVL, LV dilation 6.4cm</td>
</tr>
<tr>
<td>3</td>
<td>Bicuspid Valve, Severe stenosis</td>
<td>Balloon valvuloplasty</td>
<td>Neonate</td>
<td>Moderate AI, dilated LV, mildly decreased LV EF</td>
<td>13</td>
<td>Severe AS (peak 81, mean 51mmHg), mild AI, moderately decreased LV EF</td>
</tr>
</tbody>
</table>

ASC AO - ASCENDING AORTIC, AI - AORTIC INSUFFICIENCY, AS - AORTIC STENOSIS, AV - AORTIC VALVE, EF - EJECTION FRACTION, LVIDD - LEFT VENTRICLE INTERNAL DIAMETER IN DIASTOLE, LV - LEFT VENTRICLE, PVL - PERIVALVAR LEAK, TAVR - TRANSCATHETER AORTIC VALVE REPLACEMENT
<table>
<thead>
<tr>
<th>PATIENT</th>
<th>Echo Findings</th>
<th>Clinical Status</th>
<th>Follow-up Period</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Trivial AI, no AS, no PS, no PI,</td>
<td>Having arrhythmias and pacemaker related complications reducing exercise capacity</td>
<td>12 months</td>
</tr>
<tr>
<td></td>
<td>normal biventricular function</td>
<td>and reports decreased quality of life</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Trivial AI, no AS, no PS, no PI,</td>
<td>Doing well clinically, exercise restrictions removed</td>
<td>6 months</td>
</tr>
<tr>
<td></td>
<td>normal biventricular function</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Mild AI, no AS, no PS, no PI,</td>
<td>Complains of persistent upper/lower extremity swelling and decreased exercise</td>
<td>9 months</td>
</tr>
<tr>
<td></td>
<td>moderately decreased LV function</td>
<td>capacity, concern for genetic cardiomyopathy</td>
<td></td>
</tr>
</tbody>
</table>

AI- Aortic insufficiency, AS- Aortic stenosis, LV- Left ventricle, PI- Pulmonary insufficiency, PS- Pulmonary stenosis