Intermediate results following complex biventricular repair of left ventricular outflow tract obstruction in neonates and infants

Bahaaldin Alsoufi a,*, Zohair Al-Halees a, Cedric Manlhiot b, Abid Awan a, Mamdouh Al-Ahmadi a, Brian W. McCrindle b, Mansoor Al-Joufan a, Charles C. Canver a

a King Faisal Heart Institute, King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia
b Labatt Family Heart Center, the Hospital for Sick Children and the University of Toronto, Toronto, Ontario, Canada

Received 16 October 2009; received in revised form 6 February 2010; accepted 12 February 2010; Available online 10 April 2010

Abstract

Objectives: Although the majority of infants with severe left ventricular outflow tract obstruction (LVOTO) can be managed with balloon or surgical aortic valvotomy, a more complex biventricular repair may be required in a subset of infants with multi-level obstruction, failure of or complication to prior intervention. In the presence of normal left ventricle size and inflow, the Ross procedure is applied in patients with/without ventricular septal defect (VSD), while the Yasui procedure is applied only in those with VSD. We report mid-term outcomes in a single institution.

Methods: Thirty-five consecutive infants with severe LVOTO underwent complex biventricular repair using the Ross (n = 21) or Yasui (n = 14) procedure. Outcomes were studied using univariate and multivariable parametric models. Results: The Ross procedure was done at a median age of 88 days (8—353 days), in 8/21 (38%) neonates. As many as 12/21 (57%) had prior catheter and/or surgical intervention. Concomitant procedures included arch reconstruction (n = 4/21, 19%) and mitral valve repair (n = 6/21, 29%). In addition, 14/21 (67%) had annular enlargement (modified Ross—Konno). Haemodynamic manifestation was isolated obstruction (n = 10/21, 48%) or mixed obstruction/regurgitation (n = 11/21, 52%). Survival was 81% at 1 month, 70% at 1 year and 63% at 5 years. In multivariable regression models, factors associated with increased risk of mortality included neonatal surgery (p = 0.007), mitral valve repair (p = 0.02), longer cross-clamp time (p = 0.003), and postoperative extracorporeal membrane oxygenator (ECMO) (p = 0.004). Freedom from any cardiac re-operation was 86% at 5 years. The Yasui procedure was done at a median age of 36 days (7—207 days), 6/14 (43%) in neonates. As many as 13/14 procedures (93%) were primary and one procedure followed a prior single-stage Norwood palliation. A total of 10/14 (71%) had critical aortic stenosis and 4/14 (29%) had atresia. All patients had VSD, and 11/14 (79%) required concomitant arch reconstruction. Survival was 79% at 1 month and 5 years while 5-year freedom from re-operation was 57%. Conclusions: Complex biventricular repair can be performed in neonates and infants with severe LVOTO with mid-term survival that is equivalent to that published following balloon or surgical aortic valvotomy. Associated lesions are significant factors that influence outcome and proper patient selection may further improve survival. In neonates with concomitant arch obstruction and VSD, the Yasui operation may be associated with lower early mortality risk. Neonates with concomitant mitral valve pathology may be better served with single ventricle palliation strategy.

© 2010 European Association for Cardio-Thoracic Surgery. Published by Elsevier B.V. All rights reserved.

Keywords: Aortic stenosis; Yasui procedure; Ross procedure; Congenital heart disease

1. Introduction

Critical left ventricular outflow tract obstruction (LVOTO) in the neonate and infant includes a continuous and diverse spectrum of anatomic diagnoses ranging from hypoplastic left heart complex to isolated aortic valvular stenosis with otherwise normally formed left heart structures. Depending on related anatomic features, there are multiple single ventricle and biventricular management strategies available for patients with critical LVOTO, all of which with significant mortality and requirement for future interventions [1—3]. The majority of biventricular repair strategy candidates can be managed with either balloon or surgical aortic valvotomy. However, a small subset of children with complex LVOT anatomy and severe obstruction, infants who fail or develop significant regurgitation following a prior intervention may require a more complicated surgical repair strategy [1—5]. Several reports in the literature describe the use of the Ross or the Yasui procedure to accomplish complex biventricular repair in neonates and infants with multi-level LVOTO.

The Yasui procedure was initially described as a strategy to repair neonates with aortic atresia, ventricular septal...
defect (VSD) and well-formed left heart structures [6]. Several technical modifications were developed and the use of the Yasui procedure expanded to be employed in selected patients with severe aortic stenosis and VSD, with or without aortic arch obstruction [7—11].

The Ross procedure was initially described for aortic valve replacement in young adults; however, it achieved a significant popularity as the aortic valve replacement of choice in children [12—20]. Increased experience with the Ross procedure allowed surgeons to apply it in small infants and neonates with various complex congenital heart deformities with encouraging early results [12—20].

We aim, in the current series, to describe intermediate results at a single institution following the Ross and Yasui procedures for biventricular repair of severe LVOTO in neonates and infants. In addition, we examine patients’ characteristics, and anatomic and operative variables that significantly influence outcomes.

2. Patients and methods

2.1. Inclusion criteria

From 1993 to 2008, 35 consecutive infants with LVOTO underwent complex biventricular repair: Ross procedure (n = 21) or Yasui procedure (n = 14) at the King Faisal Specialist Hospital and Research Center in Riyadh, Saudi Arabia. Patients were identified using the hospital surgical database. Clinical, operative and outcome data were extracted from their medical records. Approval of this study was obtained from the Research Ethics Board at our institution and requirement for individual consent was waived for this observational study.

2.2. Operative details

Midline sternotomy was performed and standard cardiopulmonary bypass and myocardial protection techniques were used in all cases. When arch reconstruction was necessary, deep hypothermic circulatory arrest was employed. Most recently, selective cerebral perfusion via a cannula placed into the innominate artery was used during arch reconstruction in selected cases.

The Ross operative technique used has been described earlier [19,20]. The pulmonary autograft was implanted as a full root with coronary transfer in all cases. The autograft muscle cuff was trimmed with sutures placed almost directly at the base of the cusps. The proximal suture line was performed using running polypropylene sutures. The proximal suture line was not reinforced, so as not to limit growth. In patients with narrow left ventricular outflow tract (LVOT), a modified Ross—Konno technique was used. In those patients, the fibrous annulus of the aortic valve was often divided, and the cut was partially taken down to the septum. Part of the septum was cored out to completely open up the opening was extended proximally to the ascending aorta. Following that, the main pulmonary artery was connected to the left ventricle to the pulmonary artery. RV—PA continuity was established with a homograft in nine patients and a Contegra bovine jugular valved conduit in five patients.

The immediate postoperative results were assessed in all patients in the operating room by means of trans-oesophageal echocardiography.

2.3. Follow-up

Late outcomes were determined from recent outpatient visits at King Faisal Specialist Hospital and Research Center or from direct correspondence with patients’ families. Follow-up was complete in 88% of the patients and partial in 9% with one patient missing. Median follow-up duration for hospital survivors was 5.7 years and ranged up to 12.7 years.

2.4. Statistical analysis

Data are presented as means with standard deviation, medians with minimum and maximum and frequencies as appropriate. As the patients who underwent the Ross or Yasui procedures were not analogous and as our purpose was not to compare the results between the two operations, outcomes were described separately for the Ross and the Yasui procedures without comparative analyses. For Ross patients only, comparisons between those patients who underwent a primary procedure versus a secondary procedure were done using Student’s unpaired t-test and Fisher’s exact test. Separate time-dependent outcomes (death and re-operation) after each procedure were parametrically modelled. Parametric probability estimates for time-dependent outcomes use models based on multiple, overlapping, phases of risk (available for use with the SAS system at http://www.clevelandclinic.org/heartcenter/hazard) [21]. Competing risk analysis was performed to model the probability over time of each of two mutually exclusive end points: cardiac re-operation and death without re-operation, the remainder of patients being alive and free from cardiac
re-operation. Both univariate and multivariable analyses were performed. Variable selection in multivariable models was done using a stepwise regression approach (criteria of $p < 0.05$ to enter the model). Reported are parameter estimates and their associated standard error, which represent the change in hazard with the presence (vs absence) of the factor or the change in hazard with each increase of 1 unit in the factor. A positive parameter estimate represents an increase in risk while a negative parameter estimate represents a decrease in risk. Effects of covariates on the probability of outcomes in competing risk models are given as parameter estimates and standard error. Clinical relevance of identified covariates on likelihood of selected outcomes was established by solving the regression equations for multiple ‘typical’ test patients. All statistical analyses were performed using SAS statistical software v9.1 (The SAS Institute, Cary, NC, USA).

3. Results

3.1. Patient characteristics for the Ross procedure ($n = 21$)

The Ross procedure was performed at a median age of 88 days (range 8—353 days). There were 15 males (71%). At time of surgery, 11 patients (52%) were under 3 months of age, including eight neonates less than 31 days old (38%). Haemodynamic manifestation just prior to the operation was stenosis in 10 patients (48%) and mixed stenosis and regurgitation in 11 patients (52%). Four patients (19%) had associated VSD and four (19%) had arch obstruction.

The Ross operation was done as a primary procedure in nine patients (43%) while the remaining had undergone a prior intervention: nine (43%) balloon aortic valvotomy, five (24%) surgical interventions including four surgical valvotomy (one of them with VSD closure and coarctation repair) and one prior interrupted arch repair with VSD closure. Two patients had undergone both catheter and surgical interventions.

At time of the Ross procedure, nine patients (43%) required concomitant cardiac surgery including mitral valve repair ($n = 6$, 29%), aortic arch reconstruction ($n = 4$, 19%) and VSD closure ($n = 2$, 10%). LVOT enlargement with the modified Ross—Konno technique was done in 14 patients (67%). Median cardiopulmonary bypass time was 161 min (range 95—419 min) and median aortic cross-clamp time was 100 min (range 55—189 min).

Patients who underwent a primary Ross procedure were younger, included more neonates, and required relatively more concomitant operations and longer surgery than those undergoing a secondary Ross procedure following a prior catheter or surgical intervention (Table 1).

3.2. Outcomes following the Ross procedure

Median mechanical ventilation requirements for hospital survivors were 3 days (range 1—11 days) and their median hospital stay was 11 days (range 7—35 days). There were a total of eight mortalities including six deaths prior to hospital discharge (29%). Survival was 81% at 1 month, 70% at 1 year and 63% at 5 years. Two patients required three cardiac re-operations: one patient required mitral valve replacement and tricuspid valve repair, and the other patient required re-operation for supraaortic ascending aorta and recurrent arch obstruction followed by a second re-operation for homograft change, after which he expired. Freedom from cardiac re-operation was 86% at 5 years.

The hazard function for time-related transition to cardiac re-operation was characterised by the presence of a constant hazard throughout the years after surgery. The hazard function for time-related transition to death without cardiac re-operation was characterised by a high early hazard phase in the immediate postoperative period but relatively stable survival thereafter. The competing risks for the two events showed that 5 years following the Ross procedure, approximately 39% of patients have died, 13% have undergone cardiac re-operation and 48% were alive and free from cardiac re-operation (Fig. 1).

The association between multiple demographic, operative and postoperative variables on survival was studied. Several factors were associated with increased mortality (Table 2). In univariate analysis, several associated factors were identified. Younger age at time of surgery was associated with higher mortality, especially neonates of whom only 25% are alive at the last follow-up. Primary Ross was also associated with an increased mortality with only 33% alive at last follow-up compared to 83% in those who had a prior palliative intervention. Concomitant surgery at time of the Ross procedure was also associated with lower survival.

### Table 1

<table>
<thead>
<tr>
<th>Primary ($N = 9$)</th>
<th>Secondary ($N = 12$)</th>
<th>$p$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Demographics</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gender (male)</td>
<td>4/9 (44%)</td>
<td>11/12 (92%)</td>
</tr>
<tr>
<td>Median age at surgery (days)</td>
<td>15 (8—274)</td>
<td>119 (20—353)</td>
</tr>
<tr>
<td>&lt;90 days</td>
<td>7/9 (78%)</td>
<td>4/12 (33%)</td>
</tr>
<tr>
<td>&lt;31 days</td>
<td>7/9 (78%)</td>
<td>1/12 (8%)</td>
</tr>
<tr>
<td>Diagnosis and clinical history</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Subaortic stenosis</td>
<td>4/9 (44%)</td>
<td>5/12 (45%)</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>2/9 (22%)</td>
<td>2/12 (17%)</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>1/9 (11%)</td>
<td>1/12 (8%)</td>
</tr>
<tr>
<td>Hypoplastic aortic arch</td>
<td>1/9 (11%)</td>
<td>0/12 (0%)</td>
</tr>
<tr>
<td>Interrupted aortic arch</td>
<td>2/9 (22%)</td>
<td>1/12 (8%)</td>
</tr>
<tr>
<td>Pathology (stenosis vs mixed)</td>
<td>6/9 (67%)</td>
<td>4/12 (33%)</td>
</tr>
<tr>
<td>Previous catheterisation</td>
<td>NA</td>
<td>9/12 (75%)</td>
</tr>
<tr>
<td>Previous surgery</td>
<td>NA</td>
<td>5/12 (42%)</td>
</tr>
<tr>
<td>Surgical variables</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Additional surgeries</td>
<td>6/9 (67%)</td>
<td>3/12 (25%)</td>
</tr>
<tr>
<td>Mitral valve repair</td>
<td>3/9 (33%)</td>
<td>3/12 (25%)</td>
</tr>
<tr>
<td>Arch reconstruction</td>
<td>3/9 (33%)</td>
<td>1/12 (8%)</td>
</tr>
<tr>
<td>Modified Ross—Konno</td>
<td>6/9 (67%)</td>
<td>8/12 (67%)</td>
</tr>
<tr>
<td>Cardiopulmonary bypass time (min)</td>
<td>201 (107—419)</td>
<td>154 (95—220)</td>
</tr>
<tr>
<td>Cross-clamp time (min)</td>
<td>113 (73—189)</td>
<td>92 (55—132)</td>
</tr>
<tr>
<td>Postoperative ECMO</td>
<td>2/9 (22%)</td>
<td>1/12 (8%)</td>
</tr>
<tr>
<td>Duration ventilation (days)</td>
<td>3 (1—62)</td>
<td>4 (1—11)</td>
</tr>
<tr>
<td>Duration hospitalisation (days)</td>
<td>9 (7—62)</td>
<td>16 (7—35)</td>
</tr>
<tr>
<td>Total death</td>
<td>6/9 (67%)</td>
<td>2/12 (17%)</td>
</tr>
</tbody>
</table>
Only 33% of infants who had a simultaneous mitral valve repair and 25% of those who had simultaneous arch reconstruction survived. On the contrary, annular enlargement (modified Ross—Konno procedure) was not associated with diminished survival ($p = 0.45$). Longer bypass and cross-clamp durations were also associated with higher mortality while the requirement for postoperative extracorporeal membrane oxygenator support (ECMO) was universally fatal. On multivariable analysis, significant factors associated with increased mortality were neonatal surgery ($p = 0.007$), concomitant mitral valve repair ($p = 0.02$), longer cross-clamp time ($p = 0.003$) and postoperative ECMO ($p = 0.004$).

The higher early hazard of mortality in neonates is depicted in Fig. 2 and the higher hazard in patients requiring concomitant mitral valve repair is shown in Fig. 3. The effect of longer cross-clamp time, a surrogate for complexity of the repair, is demonstrated in Fig. 4 while Fig. 5 portrays the risk-adjusted survival adjusted for a hypothetical high-risk patient with a combination of unfavourable characteristics versus a hypothetical low-risk patient not having those characteristics.

### Table 2
Factors associated with mortality following the Ross procedure.

<table>
<thead>
<tr>
<th>Factor</th>
<th>Univariate PE (SE)</th>
<th>p</th>
<th>Multivariable PE (SE)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender (female)</td>
<td>+3.16 (1.04)</td>
<td>0.003</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Younger age at surgery (per 10 days)</td>
<td>+0.05 (0.03)</td>
<td>0.10</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at surgery &lt;31 days</td>
<td>+2.24 (0.85)</td>
<td>0.009</td>
<td>+6.17 (2.29)</td>
<td>0.007</td>
</tr>
<tr>
<td>Primary Ross (vs secondary)</td>
<td>+1.73 (0.80)</td>
<td>0.03</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Concomitant surgery</td>
<td>+1.70 (0.85)</td>
<td>0.05</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Concomitant mitral valve repair</td>
<td>+2.08 (0.92)</td>
<td>0.03</td>
<td>+6.30 (2.66)</td>
<td>0.02</td>
</tr>
<tr>
<td>Concomitant arch reconstruction</td>
<td>+2.94 (1.42)</td>
<td>0.04</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Longer bypass time (per 10 min)</td>
<td>+0.34 (0.07)</td>
<td>&lt;0.001</td>
<td>+1.24 (0.42)</td>
<td>0.003</td>
</tr>
<tr>
<td>Longer cross-clamp time (per 10 min)</td>
<td>+0.70 (0.15)</td>
<td>&lt;0.001</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Circulatory arrest requirement</td>
<td>+2.34 (1.31)</td>
<td>0.08</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Postoperative ECMO</td>
<td>+11.30 (1.76)</td>
<td>&lt;0.001</td>
<td>+15.89 (5.41)</td>
<td>0.004</td>
</tr>
</tbody>
</table>

* Reported are parameter estimate and their associated standard error which represent the change in hazard with the presence (vs absence) of risk factor or the change in hazard with each increase of 1 unit in the risk factor. A positive parameter estimate represents an increase in risk while a negative parameter estimate represents a decrease in risk.

### 3.3. Patient characteristics for the Yasui procedure ($n = 14$)

The Yasui procedure was performed at a median age of 36 days (range 7—207 days). There were seven males (50%). At time of surgery, 10 patients (71%) were under 3 months of age including six neonates less than 31 days old (43%). Ten patients (71%) had aortic stenosis while the remaining four (29%) had aortic atresia. All patients had associated VSD and 12 (86%) had arch obstruction including five (36%) who had interrupted aortic arch. All but one of the procedures (93%) were primary with one patient who underwent a prior first-stage Norwood single ventricle palliative surgery.

At the time of the Yasui procedure, 11 patients (79%) required concomitant aortic arch reconstruction but none required mitral valve intervention. Median cardiopulmonary bypass time was 171 min (range 110—342 min); median aortic cross-clamp time was 95 min (range 65—124) and median circulatory arrest time was 46 min (range 5—100 min).

### 3.4. Outcomes following the Yasui procedure

Median mechanical ventilation requirements for hospital survivors was six days (range 3—30) and their median hospital
stay was 16 days (range 10–75). There were a total of four mortalities including three (21%) prior to hospital discharge. Survival was 79% at 1 month and at 5 years after the procedure. There was one late mortality at 11 years, which was a result of complications to RV–PA conduit change. Five patients required six cardiac re-operations including: RV–PA conduit change \( (n = 6) \), tricuspid valve repair \( (n = 3) \), recurrent arch obstruction repair \( (n = 1) \) and mitral valve repair \( (n = 1) \). Freedom from cardiac re-operation was 86% at 1 year and 57% at 5 years.

The hazard function for time-related transition to cardiac re-operation was characterised by the presence of a late hazard that increases as years since surgery performed. The hazard function for time-related transition to death without cardiac re-operation was characterised by a high early hazard in the immediate postoperative period but relatively stable survival thereafter. The competing risks for the two events showed that 5 years following the Yasui procedure, approximately 25% of patients have died, 38% have undergone cardiac re-operation and 37% were alive and free from re-operation.

The effect of multiple demographic, operative and postoperative variables on survival was studied. None of the variables was significantly associated with mortality, likely due to the small sample size. Of note, none of the four patients with aortic atresia died while all the mortalities were in the group of patients with the diagnosis of critical aortic stenosis.

4. Discussion

Critical LVOTO in the neonate and infant includes a continuous and diverse spectrum of anatomic diagnoses ranging from hypoplastic left heart complex to isolated aortic valvular stenosis with otherwise normally formed left heart structures [1–3].
The degree of left ventricular hypoplasia, the size of the mitral valve, the presence of endocardial fibroelastosis (EFE) and left ventricular function generally establish whether neonates and infants with critical LVOTO are managed with a univentricular staged palliation strategy or a biventricular approach [1—3].

Biventricular repair strategy largely involves either balloon or surgical aortic valvotomy with a reported 5-year survival with either one of those treatment modalities around 70% [1—3]. While the majority of candidates for biventricular repair can be managed with those approaches, a more complicated surgical repair may be necessary in about 4% of those patients with multi-level LVOTO and failure of or complication to prior intervention [2,3,7]. The surgical approach varies based on specific morphologic characteristics and associated lesions.

The Yasui operation was originally described in 1987 as a strategy to repair 2 neonates with aortic atresia and VSD [6]. Subsequently, further modifications were described by several surgeons and a number of small series were reported from different centres with encouraging intermediate outcomes [7—11]. The Yasui procedure can be performed as a primary operation or can be staged after initial Norwood-type first-stage palliation. While some centres reported very good outcomes with the staged approach, our institutional policy is to offer primary repair in suitable biventricular candidates. Advantages of primary repair include early restoration of normal physiology of a circulation in series, elimination of prolonged cyanosis and volume load effects of the shunts, decreased numbers of operations and decreased cost. Several other centres have similarly reported excellent results with primary Yasui repair. Our dataset cannot address the superiority other centres have similarly reported excellent results with the Rastelli procedure where we perform routine mechanical support for post-operative low cardiac output in neonates and infants with critical LVOTO, especially that ECMO ischaemia and the development of EFE, which further impairs the ventricular function and affects the function of the mitral valve. In addition, reduced antegrade flow through the aortic valve will predispose to underdevelopment of the left heart structures and hypoplasia of the mitral valve, left ventricle and aortic arch.

Patients with critical LVOTO, concomitant arch hypoplasia/obstruction and mitral valve pathology, either stenosis or regurgitation due to EFE, represent a subgroup of patients with a more severe form of LVOTO and worse preoperative left ventricular dysfunction. The requirement for concomitant arch reconstruction, mitral valve repair and, consequently, longer surgery and requirement for circulatory arrest were all associated with increased mortality in our current series and in other reports from different institutions. It seems that those complex surgeries are poorly tolerated in infants with critical LVOTO, especially that ECMO mechanical support for post-operative low cardiac output in this population is associated with a dismal prognosis.

On the other hand, the necessity for annular enlargement technique using the modified Ross—Konno technique was not associated with an increased risk for mortality. This finding stresses that associated lesions, not the LVOT, have a more significant impact on survival.

Younger age at time of the Ross procedure, especially neonates, was significantly associated with poor survival. In our series, neonates who underwent the Ross procedure were more likely to require concomitant, more prolonged surgery. Neonates with concomitant arch and/or mitral surgery had a very high mortality while most neonates who underwent the Ross—Konno procedure for isolated LVOTO survived except for one neonate in whom surgery was done emergently after he had multiple episodes of cardiac arrest prior to skin incision. Our findings suggest that the Ross—Konno procedure can still be considered a valid option if necessary in neonates with isolated critical LVOTO.

We have identified that primary Ross was associated with higher mortality compared with secondary Ross after initial percutaneous and/or surgical palliation. This may be a surrogate to the fact that primary Ross was done in younger patients, especially those who had associated arch and mitral pathology. Of note, we did not have any patient in our series who required emergency surgery for acute aortic regurgitation complicating balloon valvotomy. This factor was identified to
be associated with increased operative mortality by Hickey et al. in a multi-institutional review of a similar patient cohort of neonates with critical LVOTO. Acute aortic regurgitation can be poorly tolerated in this subset of patients with ventricular hypertrophy and dysfunction. Therefore, careful decisions should be made prior to any intervention and those at risk of developing significant regurgitation may be better candidates for a primary Ross procedure.

The survival in our patient cohort is comparable to the 5-year survival of 70% in neonates and infants with critical aortic stenosis following balloon or surgical aortic valvotomy. However, all the factors associated with suboptimal outcomes in our series underscore the importance of appropriate patient selection in the management of neonates with critical LVOTO. Similar to patients with isolated aortic stenosis, outcomes may significantly improve with proper selection of univentricular versus biventricular management candidates, and the suitable intervention method with the lowest risk for early/emergency re-intervention requirement.

Cardiac re-operation is common following both the Ross and Yasui procedures. It seems from our small patient cohort that RV—PA conduit change is more common in the Yasui group of patients, which may be due to the non-anatomic location of the conduit, than in the Ross procedure with the more appropriate anatomic position. On the other hand, it seems that re-operation following the Ross procedure is more likely for other cardiac pathologies, likely due to the heterogeneous nature of the Ross patient population.

In summary, associated lesions, rather than LVOTO morphology, have a more significant impact on outcome in infants with critical LVOTO. Both the Yasui and Ross procedures can attain biventricular repair in a subset of patients with severe LVOTO and with results comparable to those reported for aortic valvotomy in neonates and infants with critical aortic stenosis. In patients without an associated VSD, initial palliation with balloon aortic valvotomy is indicated. Nonetheless, neonates at high risk of re-intervention due to failure or complications, such as those with complex multi-level LVOTO or a very small annulus may be better candidates for surgical intervention or primary Ross procedure. The presence of simultaneous arch obstruction and mitral valve morphology is a marker for a more severe end of the spectrum of disease and some of those patients, especially neonates, may be better served with an initial single-stage palliative treatment. In patients with VSD, when both the Ross and Yasui procedures are valid options, the Yasui procedure may be better served with an initial single-stage palliative procedure. In patients with VSD, when both the Ross and Yasui procedures are valid options, the Yasui procedure may be better served with an initial single-stage palliative treatment. In patients with VSD, when both the Ross and Yasui procedures are valid options, the Yasui procedure may be better served with an initial single-stage palliative treatment.

References


Appendix A. Conference discussion

Dr B. Brown (Birmingham, United Kingdom): So really you’re saying that the approach that you have taken when you have other lesions is far too...
complex to achieve a good outcome. So with isolated left ventricular outflow tract obstruction, you expect good results?

Dr Alsoufi: There may be other problems.

Dr Brawn: When you have other problems, then you’re better off palliating; is that correct?

Dr Alsoufi: That’s the overall picture, yes.

Dr C. Pizarro (Wilmington, Delaware, USA): Last year we presented here a similar paper, which was an analysis of the Congenital Heart Surgery Society experience, and there we found that there were basically two subgroups, the patients who underwent Yasui who tended to be older, elective, primary procedures who had a far better outcome, in whom most of the re-interventions were related to right ventricular outflow reconstruction because of conduit change and whatnot, and then the Ross patients who were younger. A lot of them underwent a rescue operation, and certainly the risk was higher undertaking that procedure at that age.

I didn’t see in the presentation, details that we would probably like to know about, the early or the neonatal Rosses. How many of these patients did have a previous aortic balloon intervention, and if you categorized them regarding, let’s say, the CHSS equation regarding adequacy and risk related to a primary repair?

Dr Alsoufi: In our study, similar to the CHSS paper, the Yasui procedure was performed electively. However, our Yasui patients were younger than those who underwent the Ross procedure, mainly because we do the Yasui procedure as a primary operation rather than following a previous palliation.

In our series, 9 infants underwent balloon aortic valvotomy prior to the Ross procedure and prior intervention did not increase the risk of subsequent Ross operation. Nonetheless, unlike the CHSS study, our series includes only one patient who underwent emergency Ross operation. This patient was very sick prior to surgery, arrested in the operating room prior to skin incision, and expired soon after the Ross procedure. He had isolated LVOT obstruction without any mitral pathology and was our only mortality in the subgroup of infants with isolated LVOTO. While this is supported by the findings in the CHSS study, our limited number of emergency Ross procedures in infants doesn’t allow us to perform any meaningful analysis in that regard.

In our series, neonatal Ross was a significant risk factor for mortality. Neonates had more concomitant surgery and many underwent primary Ross rather than secondary following a prior palliation. On multivariable analysis both neonatal and concomitant surgeries were significant risk factors for mortality.

Dr D. Metras (Marseille, France): In your paper you write, ‘Our purpose was not to compare the results between the two operations.’ But, in fact, when you see this kind of experience, one cannot help but somehow compare the results, since, in addition, it looks like some patients could have had either the Ross operation or the Yasui operation.

My first question is this: In the patients who have severe complex LV outflow tract stenosis and the VSD, what are the facts that made you choose between the Ross and the Yasui?

Dr Alsoufi: As shown in my last conclusion slide, the Ross and the Yasui procedures are usually performed on different groups of patients. However, a grey zone exists in infants with severe LVOT obstruction and a VSD. Those are the patients who could potentially undergo either option.

Our data suggest that the Yasui operation is associated with lower early risk, especially in those who require simultaneous arch reconstruction.

In infants without concomitant arch obstruction, mortality risk is comparable between the two procedures; however the Ross procedure may offer an advantage of greater freedom from cardiac re-operation, namely RV PA conduit change. This may be due to the fact that RV PA conduits in the Yasui operation are placed in a non-anatomic position as compared to the anatomic position in the Ross procedure; and probably to the fact that the Yasui patients were somehow younger in our series.

So that’s why for infants with concomitant arch obstruction, I would recommend the Yasui operation; for those with no concomitant arch obstruction, probably the Ross procedure; and for those with concomitant mitral valve pathology, a single ventricle palliation.

Dr Metras: In your neonatal Ross group, you’ve got 6 deaths in 9 patients, and, in addition, they had mitral valve repair, Konno procedure, VSD closure in addition to aortic arch problems.

In those patients, why didn’t you choose a univentricular therapy?

Dr Alsoufi: Why did we not use a univentricular palliation?

Dr Metras: Yes.

Dr Alsoufi: In retrospect, the data suggest that those neonates and young infants with complex LVOT obstruction, no VSD, concomitant arch and mitral pathology are better served with a univentricular palliation strategy.

Dr Metras: Thank you very much.