



# Ross-Konno operation for critical aortic valve stenosis or LVOTO in infants: A single institution experience

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# NO DICLOSURE



#### Introduction

- Critical aortic valve stenosis or severe left ventricular outflow tract obstruction remains a significant mortality and morbidity in neonates or infants.
- Survival after biventricular repair (Ross or Ross-Konno group) range from 63-85%.
- Younger age was associated with worse outcome.
- Primary Ross associated with a significantly higher mortality
- Staged approach may improve outcome



#### **Ross Operation in Children: 23-Year Experience From a Single Institution**



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*Conclusions.* In children older than 1 year of age, the Ross operation has excellent outcomes with no operative mortality and a low incidence of aortic insufficiency. In children younger than 1 year of age, the Ross operation is associated with higher operative mortality and a higher incidence of aortic insufficiency. Where possible, the Ross operation should be delayed beyond infancy. Poly-(p-dioxanone)-filament banding may reduce the incidence of aortic insufficiency after the Ross operation.



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# Ross and Yasui operations for complex biventricular repair in infants with critical left ventricular outflow tract obstruction $\stackrel{\bigstar}{\sim}$

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fibroelastosis. Emergency iatrogenic aortic regurgitation (P = 0.005) and co-existing abnormalities (mitral stenosis, P = 0.02; mitral regurgitation, P = 0.05; LV dysfunction, P = 0.03) were strong determinants of death. Severe postoperative ventricular dysfunction or need for extracorporeal membrane oxygenation (ECMO) conferred negligible survival. Younger age was associated with disproportionately worse late outcome (5-year survival  $44 \pm 10\%$  for neonates vs  $76 \pm 8\%$  for age >3 months, P = 0.0013). However, mitral and left ventricular dysfunction and emergency presentation were significantly more common in the younger age groups. Infants less than 3 months of age without co-existing abnormalities had acceptable late survival ( $\approx 75 \pm 20\%$ ). YASUI (N = 13): Yasui repair (median age 22 days) was usually the primary intervention (nine of 13) but occasionally followed Norwood palliation (four of 13). None was an emergency. All had a ventricular septal defect. Survival was  $69 \pm 13\%$  at 10 years, which is not significantly different from other biventricular repair strategies in neonates. Aortic atresia was associated with better survival than stenosis ( $90 \pm 12\%$  vs  $30 \pm 14\%$  at 3 years, P = 0.039). None reverted to univentricular physiology later. **Conclusions: Case selection is key for complex biventricular repair and the importance of appropriate case selection is exaggerated at young ages**. All available options should be considered before pursuing the Ross operation in the presence of co-existing functional morphological abnormalities or emergent iatrogenic aortic regurgitation. However, both the Ross and Yasui operations in children (including neonates and young infants) with favourable functional morphology offer good survival, at least matching that of other biventricular repair strategies. © 2009 European Association for Cardio-Thoracic Surgery. Published by Elsevier B.V. All rights reserved.



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Intermediate results following complex biventricular repair of left ventricular outflow tract obstruction in neonates and infants  $^{\diamond}$ 

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**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

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.

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#### Biventricular Repair in Interrupted Aortic Arch and Ventricular Septal Defect With a Small Left Ventricular Outflow Tract

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*Background.* In patients with interrupted aortic arch and ventricular septal defect (VSD) with a small left ventricular outflow tract (LVOT), either aortopulmonary amalgamation or a Ross-Konno type procedure can be performed to create stable systemic outflow. We sought to analyze factors associated with these different surgical approaches.

*Methods.* We retrospectively identified patients who underwent surgical repair for interrupted aortic arch/ VSD at our institution between 1998 and 2017. Of these, 43 patients had a small, native LVOT that was unsuitable for systemic outflow. Patient data were retrospectively collected for this cohort and analyzed.

*Results.* Aortopulmonary amalgamation was performed at 7 days (interquartile range [IQR], 5-10) in 30 patients (group I). Within group I a primary Yasui repair with ventricular septation was performed in 3 patients and a Norwood-type repair in the other 27. Of these 27, 19 underwent subsequent biventricular conversion at 9 months (IQR, 7-11). In contrast 13 patients underwent a Ross procedure at 12 days (IQR, 6-27) (group II). Compared with group I, group II patients had a smaller VSD (3.5 vs 5.1 mm, P < .001) that was more often remote from the semilunar valves (38% vs 13%, P = .02). Operative mortality occurred in 1 group I patient (4%) at the time of biventricular conversion and 2 group II patients (15%) during the Ross procedure. After a 5.2-year (IQR, 3.2-7.4) follow-up there were 2 additional mortalities in each group, all unrelated to cardiac disease.

*Conclusions.* When native LVOT in interrupted aortic arch/VSD is unsuitable for systemic outflow, size and location of the VSD can be used to tailor the surgical approach to establish biventricular circulation with favorable intermediate-term outcomes.

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#### Staged Biventricular Repair for Neonates With Left Ventricular Outflow Tract Obstruction, Ventricular Septal Defect, and Aortic Arch Obstruction

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*Background.* The purpose of this study is to evaluate clinical outcomes of neonates who underwent a Norwood operation as a first step of a planned biventricular repair and the impact of associated risk factors.

*Methods.* A retrospective cohort study was performed on all neonates (n = 44) undergoing the Norwood operation as the first stage of a biventricular (Norwood-Rastelli) repair from January 2000 to December 2012 at a single center. Multivariable analysis was performed to identify predictors of survival.

*Results.* Stage one mortality was 9%. The interstage survival for nonsyndromic and syndromic patients was 100% versus 46%, respectively (p < 0.001). Twenty-four patients (55%) underwent biventricular completion repair with no mortality. Freedom from reintervention after

biventricular completion was 53% at 6 years. The overall survival for nonsyndromic patients versus syndromic patients was 86% versus 43%, respectively (p = 0.01). Genetic syndromes and prematurity were significant predictors of interstage mortality on multivariable analysis.

*Conclusions.* Staged biventricular repair for patients with complex left ventricular outflow tract obstruction, ventricular septal defect, and aortic arch obstruction can be achieved with excellent outcomes for neonates without genetic syndromes. The staged approach is associated with longer time to reintervention after the biventricular completion.

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### Patients

- From August 2016 to May 2022: 20 patients (neonates and infants) underwent Ross-Konno operation
- Median age (min-max): 1.02 year (0.14-6.07) (1 neonate, <3 months: 2; < 6 months: 3; < 1 year: 3; < 3 years: 8; 3-6 years: 3)</li>
- Median weight (min-max): 7.75 kg (3.7-26)
- Male/female: 15/5
- Preoperative diagnosis:
  - Critical Ao valve stenosis: 8 patients
  - Critical Ao valve regurgitation/Osler: 2 patients
  - CoA-VSD-LVOTO: 4 patients
  - AA-VSD-LVOTO: 4 patients



## Patients

- Preoperative intervention:
  - Norwood palliation: 8 patients
  - Aortic valvotomy: 3 patients
  - VSD-CoA repair: 2 patients
- Cardiac function: EF < 30% (2 patients), EF < 40% (1 patient)
- Z-score of Aortic valve stenosis group (16): -4.53 (-10.5, 0.75)

### Intraoperative variables

ATCSA2

Intraoperative variables	n	%
Ross-Konno operation	16	80
Ross operation	4	20
Aortic arch reconstruction	3	15
Mitral valve repair	1	5
Conduits size (Contegra)		
18	9	45
16	6	30
14	3	15
12	3	15
ACC time (minutes)	$131 \pm 39.7$	
Bypass time (minutes)	$190 \pm 44$	
Ventilation time (hours)	168 (16.5 - 2420)	

#### Case video

- Male, 3kg, VSD-CoA severe LVOTO due to small Ao valve and posterior deviation conal septum
- Norwood-Sano operation since 1 month old
- Ross-Konno operation at 1 year old, 9kg, Contegra number 18



## Surgical techniques





### Echo follow-up

#### Mild AR

#### Mild MR



### Echo follow-up

#### **Mild RPA stenosis**

#### **Good Arch**



#### Postoperative variables

- Secondary chest closure: 13/20 (65%)
- 1 ECMO
- Early mortality: 1 neonate patient (severe AS-VSD-CoA)
- Late mortality: 1 patient with severe AS, EFE (9 months later)
- Latest echo follow-up:
  - Non AR: 2/19
  - Mild AR: 15/19
  - Mil-moderate AR: 1/19
  - Moderate AR: 1/19
- Reintervention: 1 patient (Balloon angioplasty for RPA stenosis)
- Follow-up time (months): 12 (0.5-60)
- No reoperation

### Survival (1 month: 95%; 5 year: 88.2%)

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#### Conclusions

- Ross-Konno is a reasonable choice for critical AS or severe LVOTO in small infant
- A staged approach might improve the outcome
- Close follow-up is mandatory





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