The Ross-Konno Procedure for congenital aortic stenosis

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INTRODUCTION

➢ **Congenital aortic stenosis is a complex and lifelong disease.**

Any therapeutic approach should consider a unique set of requirements:

- Growth of the child and valve
- Activity level
- Lifestyle
- Difficulty in medical compliance

Available treatment options are:

1. Balloon Aortic Valvotomy (BAV)
2. Surgical or Open Aortic Valvotomy (OV)
3. Aortic valve replacement (mechanical prosthetic valve or bioprosthetic valve)
4. **Ross or Ross-Konno procedure**
Aortic Stenosis in Children

Indication for Surgery

Decision making

– **Symptomatic patient**
  - Gradient > 40 mmHg
  - LV hypertrophy
  - ECG changes

– **Asymptomatic patient**
  - Gradient > 70 mmHg
Balloon Valvuloplasty

- Balloon aortic valvuloplasty is considered the initial treatment of choice in order to relieve the symptoms and postpone the time for future surgical intervention.
- Despite of the good short term results, mid-term results showed a substantial incidence of restenosis, severe aortic insufficiency, vascular access complication and reinterventions.

Open Valvuloplasty

• The surgical management of aortic stenosis, shows a low early and late mortality (*actuarial survival rate at 15 years 85%*), but the residual lesions continue to progress so eventually, patients require reoperation and possible valve replacement.
• Freedom from reintervention 35% at 10 years
• Freedom from AVR 54% at 10 years.

Valvotomy of any kind is a palliative procedure and reintervention remains frequent!

Aortic valve replacement

Mechanical valves are not ideal valve substitutes in children

- They have significant limitations at the time of implant due to the lack of appropriately sized prostheses for small children and neonates.
- Absence of potential for growth can result in patient-prosthesis size mismatch as the child grows and may require re-replacement.
- mechanical valves require lifetime anticoagulation with associated activity limitations, difficulties with future pregnancy, and a lifetime risk of thromboembolic and bleeding complications.

Aortic valve replacement

Homografts and bioprosthetic valves in aortic position are also problematic in children

- they do not allow growth
- their durability in the pediatric population is very limited due to the high risk of accelerated structural valve degeneration and early calcification.
- the availability of appropriate sized homografts and bioprostheses is a serious problem.

Ross Konno operation

- Ross Konno operation is a complex procedure for severe multilevel LVOTO, mainly aortic and (sub)aortic stenosis.
- It is the combination of two different procedures (with their later modifications)

1. Aortic valve replacement with Pulmonary autograft transplantation described by sir Donald Ross in 1967
2. Anterior aortoventriculoplasty, known as the Konno - Rastan procedure described by Drs S. Konno and Rastan in 1975
1. Ross procedure

- The replacement of the aortic valve with the patient's own pulmonary valve and then implantation of a pulmonary allograft in the RVOT to replace the pulmonary valve
History


➢ 1966. The first use of aortic homograft for Pulmonary Atresia.
  Ross DN et al: Correction of Pulmonary Atresia with a homograft aortic valve.
  Lancet 2: 1446-1447, 1966

➢ 1967. Ross operation with the use of aortic homograft for RVOT reconstruction.
  D.N. Ross, Replacement of aortic and mitral valves with a pulmonary autograft,
  The operation performed at Guy’s Hospital on June 8, 1967

➢ Late 80’s. New interest for Ross operation in USA. Dr. Elkins started pulmonary autotransplant program in USA.
ANATOMICAL CONSIDERATIONS FOR ROSS OPERATION

- Normal aortic and pulmonary valve are identical in size, shape and configuration
- Even when the aortic valve is deformed, the pulmonary valve is almost always normal
- Pulmonary valve is durable substitute in aortic position due to its high pressure resistance
- The two valves are placed at two different levels and the pulmonary valve can be detached from the heart without affecting other structures
Ross operation schematically
2. Konno Procedure

• “The procedure consists of: a longitudinal incision in the aortic septum placed in the midportion of the two coronary ostia,
• a vertical incision in the outflow tract of the right ventricle to join the septal incision,
• prosthetic aortic valve replacement, and
• patch reconstruction of the outflow tracts of both ventricles by means of two layers of a fusiform”

INDICATIONS FOR ROSS-KONNO

• Aortic valve disease in Pediatric population
• Complex and/or recurrent LVOTO (annular hypoplasia, subvalvar stenosis, tunnel shape LVOT)
• Contraindication to mechanical prosthesis or anticoagulation
• Complicated mechanical valve
• Bioprosthetic valve failure
• Destructive infective endocarditis
CONTRAINDICATIONS

- Pulmonary valve pathology
- **Collagen disease** (Marfan syndrome, Ehlers-Danlos syndrome, SLE, RA, Reiter syndrome)
- Significant *irreparable mitral valve pathology* that requires mechanical valve replacement (considered a relative contraindication by many surgeons)
- Anomalous origin of LAD from the Right coronary artery, crossing the RVOT near the Pulmonary valve annulus
Advantages for Ross-Konno operation

- Use of living host tissue. Superior hemodynamics from all other substitutes
- Growth in children
- Suitable for multilevel LVOT
- Superior long term durability in the aortic position
- Freedom from mechanical valve problems, particularly anticoagulation
- Resistance to endocarditis
- Better quality of life

Disadvantages for Ross-Konno operation

- Converts a single valve procedure into a double valve procedure
- Long and technically difficult procedure
- Risk of coronary compromise
- Limited availability of homograft
- Reoperations for homograft failure in the RVOT
- Possible dilatation of autograft (neoaorta) and PV insufficiency in long term FU

Ross Konno Outcomes

• The Ross-Konno procedure can be accomplished with a mortality of <5% and a low morbidity rate.
• Early complications are especially related to arrhythmias. The incidence of complete heart block varies from 0% to 6%.
• The freedom from autograft reoperation is above 90% at 10 years of follow-up.
• The freedom from homograft reoperation is between 80% and 94% at 5 years, being less favorable for small children due to earlier homograft failure.
• The choice of conduit appears to impact the need for replacement. The aortic homograft rather than pulmonary homograft and smaller homograft size are factors adversely affecting homograft longevity.

RESULTS
The fate of Pulmonary autograft and homograft in Ross Konno operation

Freedom from right ventricular outflow tract replacement (diamonds) and pulmonary autograft replacement (squares) in children with Ross-Konno procedure.

Ross Konno Results

- Ross-Konno procedure can be performed with a low mortality rate even in neonates and infants
- In patients > 18 months, survival was 98.2% at 20 years but the freedom from valve/root reoperation was 88% at 10 and 72% at 20 years.
- In patients < 18 months, survival was 81% at 20 years, but the freedom from valve/root reoperation was almost 100% at 20 years!!!

Ross Konno Results

- Overall survival at 10 years was 87%.
- Estimated freedom from autograft reoperation was 100, 96 and 67% at 5, 10 and 15 years, respectively.
- Estimated freedom from homograft reoperation was 89, 77 and 58% at 5, 10 and 15 years, respectively.

CONCLUSION

• Ross-Konno is the method of choice for complex and/or recurrent LVOTO especially in neonates and infants with significant annular and subannular hypoplasia.
• Excellent results with low morbidity and low mortality in both adolescent and pediatric patients
• The pulmonary autograft demonstrates durability without developing aortic stenosis or progressive dilatation, and there is a low incidence of aortic insufficiency development.
• Growth of pulmonary autograft demonstrated in pediatric patients, but requires further evaluation
• The choice and size of conduit for RVOT reconstruction appears to impact the need for replacement.
Thank You

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THANK YOU
Aortic valve replacement (AVR)

Indications for AVR in children

• Hypoplasia of the aortic valve annulus in the neonate
• Progressive stenosis of the aortic valve in infants and children
• Multilevel left ventricular outflow tract obstruction (LVOTO) in association with aortic valve stenosis not amenable to aortic valve repair that requires enlargement of the outflow tract
• Aortic insufficiency as a complication of percutaneous balloon aortic valvuloplasty
• Rheumatic aortic valve disease
• Aortic valve endocarditis
Conclusions:
Stenosis and regurgitation of the RV-PA conduit in adults and children following Ross AVR is infrequent.

The most logical reasons for the superior performance of the homograft in Ross patients are:
1. Orthotopic positioning
2. Older age of implant, and
3. The ability to significantly oversize the homograft in Ross patients

Right Ventricular Outflow Tract Reconstruction in Ross Patients: Does the Homograft Fare Better?
John W. Brown, Mark Ruzmetov, Mark D. Rodefeld MD and Mark W. Turrentine
Indiana University School of Medicine, Indianapolis, Indiana.
The Annals of Thoracic Surgery 2008; Volume 86, Issue 5, 1607-1612