

# Ross-Konno Procedure in Neonates: Report Of Three Patients

François Lacour-Gayet, MD, Henning Sauer, MD, Kalliopi Ntalakoura, MD, Andreas Müller, MD, Vit Razek, MD, Jochen Weil, MD, and Christoph Haun, MD

Department of Pediatric Cardiac Surgery, Eppendorf University Hospital, University of Hamburg, Hamburg, Germany

The Ross-Konno procedure, applied to neonates with severe left ventricular outflow tract (LVOT) obstruction, offers a satisfactory solution in fully releasing the LVOT gradient, and in replacing the aortic valve with a pulmonary autograft with an excellent growth potential. We reported on three recent neonatal cases.

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Severe left ventricular outflow tract obstruction (LVOTO) may complicate congenital aortic valve stenosis, interrupted aortic arch (IAA), and Shone's complex. The optimal surgical procedure in neonates remains controversial [1]. Although the Norwood procedure may be required in patients with hypoplastic left ventricle (LV), a Ross-Konno procedure should be considered in neonates with acceptable LV size [2], as reported here in 3 patients.

Between September 1999 and July 2002, 11 patients underwent a Ross-Konno procedure, performed by the same surgeon, at the Eppendorf University Hospital, Hamburg, Germany. Patients with hypoplastic left heart syndrome were excluded from the study. The median age was 84 months, including 4 infants. This report focuses on the three neonates who underwent a Ross-Konno procedure.

A 39-weeks' gestation girl weighing 3.2 kg had interrupted aortic arch (IAA) type B1 with a 5-mm bicuspid aortic valve stenosis (AVS) and left ventricular outflow tract obstruction (LVOTO) (3 mm) by malalignment of the conal septum (displaced posteriorly), as well as a large perimembranous ventricular septal defect (VSD). A modified Ross-Konno procedure was performed at 10 days of age. The conal septum was almost totally resected (Fig 1) and the pulmonary autograft was implanted directly on the inferior rim of the VSD and on the residual aortic annulus, using the muscular wall of the autograft (Fig 2). The IAA was repaired by direct anastomosis. The right ventricular outflow tract (RVOT) was reconstructed using a 12-mm composite Labcor conduit.

A 40-weeks' gestation boy weighing 3.1 kg received a

diagnosis of Shone's complex. Echocardiography revealed a critical AVS, with a LV-to-aorta gradient of 70 mm Hg, with a doming bicuspid aortic valve and a 7-mm aortic annulus diameter. The subaortic area presented a 4-mm muscular stenosis at echocardiography. The LV shortening fraction was reduced to 16%, with a nearly normal left ventricular end diastolic volume. There was severe mitral valve regurgitation associated with a mitral gradient of 7 mm Hg. There was an arch obstruction with a hypoplastic aortic arch at 4 mm; the associated aortic coarctation was reopened under prostaglandin. A balloon valvuloplasty was attempted and led to significant aortic regurgitation. The patient underwent surgery at 16 days of age. A Ross-Konno procedure was performed, and the RVOT was reconstructed using an 11-mm valved cryopreserved homograft. The aortic arch was repaired. The mitral valve presented a rare anomaly. The two normal papillary muscles were attached by a transverse chorda tendineae implanted on top of the papillary muscles. The abnormal chordae pulled the muscles together, creating both a parachute mitral valve aspect and severe mitral valve regurgitation. This T-shape transverse fibrous chorda was simply resected.

A 38-weeks' gestation girl, weighing 2.9 kg, with Pierre-Robin sequence (micrognathia and cleft palate), presented with severe AVS (gradient 60 mm Hg) of a 7-mm bicuspid domed aortic valve with moderate aortic regurgitation after balloon valvuloplasty, 2 small muscular VSDs, and foramen ovale. The girl underwent a Ross-Konno procedure because aortic valve reconstruction was impractical.

## Technique

Procedures were performed with continuous cardiopulmonary bypass (CPB), bicaval cannulation, and moderate to deep hypothermia. IAA repair was performed without circulatory arrest. The brachiocephalic artery was selectively perfused, and the flow was reduced to  $50 \text{ mL} \cdot \text{kg}^{-1} \cdot \text{min}^{-1}$ . Repeated cold crystalloid cardioplegia (Custodiol) and topical cooling were used for myocardial protection. The Ross-Konno procedure was performed following the general technical principles described previously [3]. The Ross-Konno technical modification used in patient 1, presenting with IAA type B1 and LVOT obstruction, is detailed in Figures 1 and 2.

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Address reprint requests to Dr Lacour-Gayet, Cardiothoracic Surgery Department, The Children's Hospital in Denver, 1056 East 19th Ave, Denver, CO 80218; e-mail: lacour-gayet.francois@tchden.org.

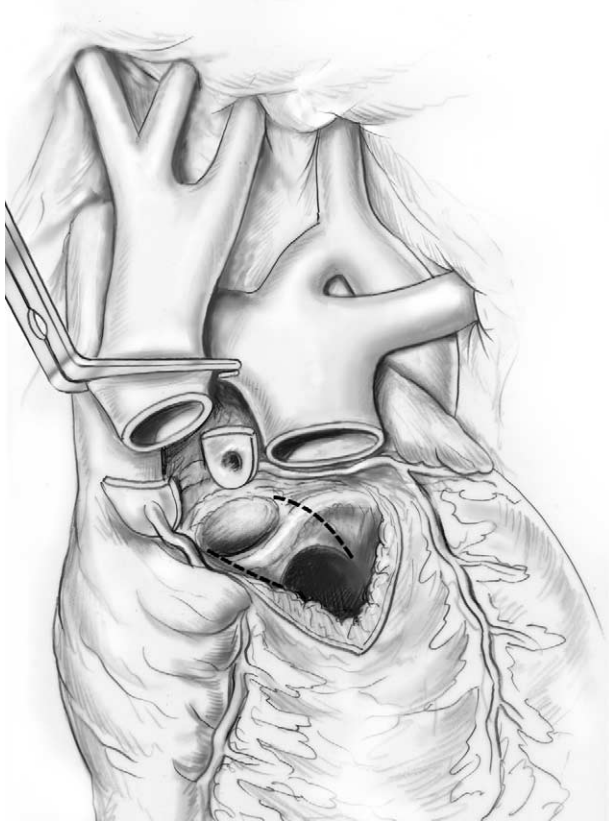


Fig 1. Technique followed for patient 1, presenting with interrupted aortic arch type B1 and left ventricular outflow tract obstruction. Note the complete resection (dashed line) of the conal septum.

## Results

### Postoperative Course

CPB time was  $248 \pm 69$  minutes (range 184 to 321 minutes), and the aortic cross-clamp time was  $144 \pm 25$  minutes (range 123 to 172 minutes). In all patients, intraoperative echocardiography showed excellent neo-aortic valve function with not more than trivial aortic valve regurgitation. All levels of obstruction were markedly reduced, with the gradient abolished. ICU stay of  $17.7 \pm 13.0$  days (median 17 days) was primarily related to infection, respiratory insufficiency, need for inotropic support  $6.0 \pm 2.6$  days (median 5 days), and delayed sternal closure  $3.7 \pm 2.9$  days (median 2 days). Length of hospital stay was  $27.3 \pm 21.5$  days (median 17 days). One patient suffered from temporary neurologic abnormalities following CPR due to hyperkalemia, need for temporary peritoneal dialysis, bilateral chylothorax, thrombolytic therapy for inferior vena cava occlusion, and moderate right pulmonary artery stenosis (apparently related to compression by the large autograft) requiring stent implantation. Another patient had a temporary left-sided phrenic nerve palsy. To date, all patients are in sinus rhythm, with no history of atrioventricular block.

### Follow-Up

All patients are alive, achieved normal growth, and are in New York Heart Association class I. For the three neonates reported here, the median follow-up was 405 days (29 days–36 months).

### Autograft Function

Serial two-dimensional echocardiography was obtained in all three patients. There was no autograft dilation or LVOTO. One patient had a mild autograft insufficiency.

### Allograft Function

One patient needed reoperation at 4 months for an early homograft failure, with massive pulmonary regurgitation. At reoperation, no more sigmoid leaflet could be found on the pulmonary homograft. Mild to moderate homograft insufficiency was present in 2 patients.

### Comment

Relief of the LV-to-aorta gradient is crucial in preserving long-term LV function in neonates presenting with various types of LVOTO. Balloon valvuloplasty can achieve good results. The risk of iatrogenic valve regurgitation, however, remains significant, as with two of our patients. Patients presenting with either IAA-LVOTO or multiple-

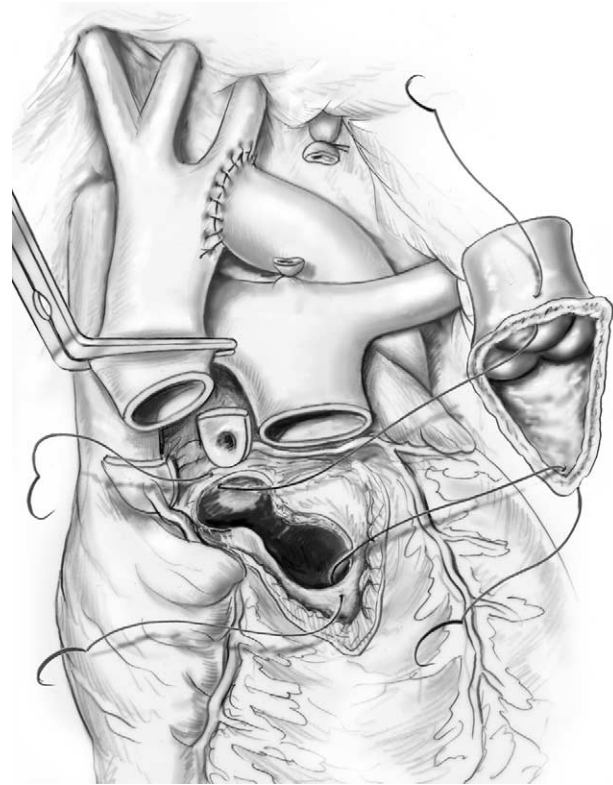


Fig 2. The left ventricular outflow tract obstruction is reconstructed using the infundibular free wall of the autograft to directly close the ventricular septal defect. The neo-aortic annulus was not reinforced, so to not limit growth. The right ventricular outflow tract was reconstructed with a right ventricle-to-pulmonary artery valved conduit, using different materials.

level stenoses cannot be treated by interventional cardiology procedures.

The surgical options in neonates are limited. The commissurotomy is the simplest and safest method. In many cases, however, the hypoplastic aortic annulus leads to a residual gradient that can rapidly become deleterious. With simple surgery or balloon valvuloplasty, subsequent corrective surgery may be required because of an important residual gradient or severe regurgitation of the aortic valve. A Ross-Konno operation is always necessary. The aortic valve replacement with a mechanical prosthesis is usually impossible, requiring an impractical aortic annulus enlargement. Aortic homografts have been used extensively [4], yet are associated with a high risk of early homograft deterioration. Apparently, the Ross-Konno procedure may be the most satisfactory option with a satisfactory growth potential of the autograft [5]. The RV-to-PA valved conduit deterioration or outgrowth may require reoperation that may be needed early as seen in 1 patient.

In IAA associated with LVOTO, several authors have suggested preserving the aortic valve, even with aortic diameters as small as 3.5 mm [6, 7]. In these patients, however, the risk of secondary LVOTO remains significant [7]. Thus, a primary Ross-Konno procedure in IAA with severe LVOTO appears reasonable, but may need confirmation by longer follow-up.

In patient 1, the LVOTO was caused by the posterior displacement of the conal septum. The infundibular septum in this patient was short. The resection through the right ventricle of this infundibular septum [8] was judged too hazardous for the aortic valve.

In multiple-level left heart stenoses (Shone's complex), the mitral valve lesion dominates both the possibility of biventricular repair and long-term prognosis. To our knowledge, the lesion found in patient 2 has not been described previously. The parachute-like lesion was cu-

riously related to an abnormal transversal chorda pulling together the top of the two papillary muscles. Resecting the transversal chordae could easily treat the abnormality. This case emphasizes the importance of intraoperative checking of the mitral valve in Shone's syndrome surgery.

## References

1. Al-Halees Z, Pieters F, Qadoura F, Shahid M, Al-Amri M, Al-Fadley F. The Ross procedure is the procedure of choice for congenital aortic valve disease. *J Thorac Cardiovasc Surg* 2002;123:437-41.
2. Ohye RG, Gomez CA, Ohye BJ, Goldberg CS, Bove EL. The Ross/Konno procedure in neonates and infants: intermediate-term survival and autograft function. *Ann Thorac Surg* 2001; 72:823-30.
3. Reddy VM, Rajasinghe HA, Teitel DF, Haas GS, Hanley FL. Aortoventriculoplasty with the pulmonary autograft: the "Ross-Konno" procedure. *J Thorac Cardiovasc Surg* 1996;111: 158-65.
4. Clarke DR. Value, viability, valves. *J Thorac Cardiovasc Surg* 2002;124:1-6.
5. Solowiejczyk DE, Bourlon F, Apfel HD, et al. Serial echocardiographic measurements of the pulmonary autograft in the aortic valve position after the Ross operation in a pediatric population using normal pulmonary artery dimensions as the reference standard. *Am J Cardiol* 2000;85:1119-23.
6. Jonas RA, Quaegebeur JM, Kirklin JW, Blackstone EH, Daicoff G. Outcomes in patients with interrupted aortic arch and ventricular septal defect. A multiinstitutional study. *Congenital Heart Surgeons Society. J Thorac Cardiovasc Surg* 1994; 107:1099-109.
7. Roussin R, Belli E, Lacour-Gayet F, et al. Aortic arch reconstruction with pulmonary autograft patch aortoplasty. *J Thorac Cardiovasc Surg* 2002;123:443-50.
8. Bove EL, Minich LL, Pridjian AK, Lupinetti FM, Snider AR, Dick M 2nd, Beekman RH. The management of severe sub-aortic stenosis, ventricular septal defect, and aortic arch obstruction in the neonate. *J Thorac Cardiovasc Surg* 1993; 105:289-95.