2014 전공의 학술세미나

Transposition of the Great Arteries

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Division of Pediatric Cardiac Surgery
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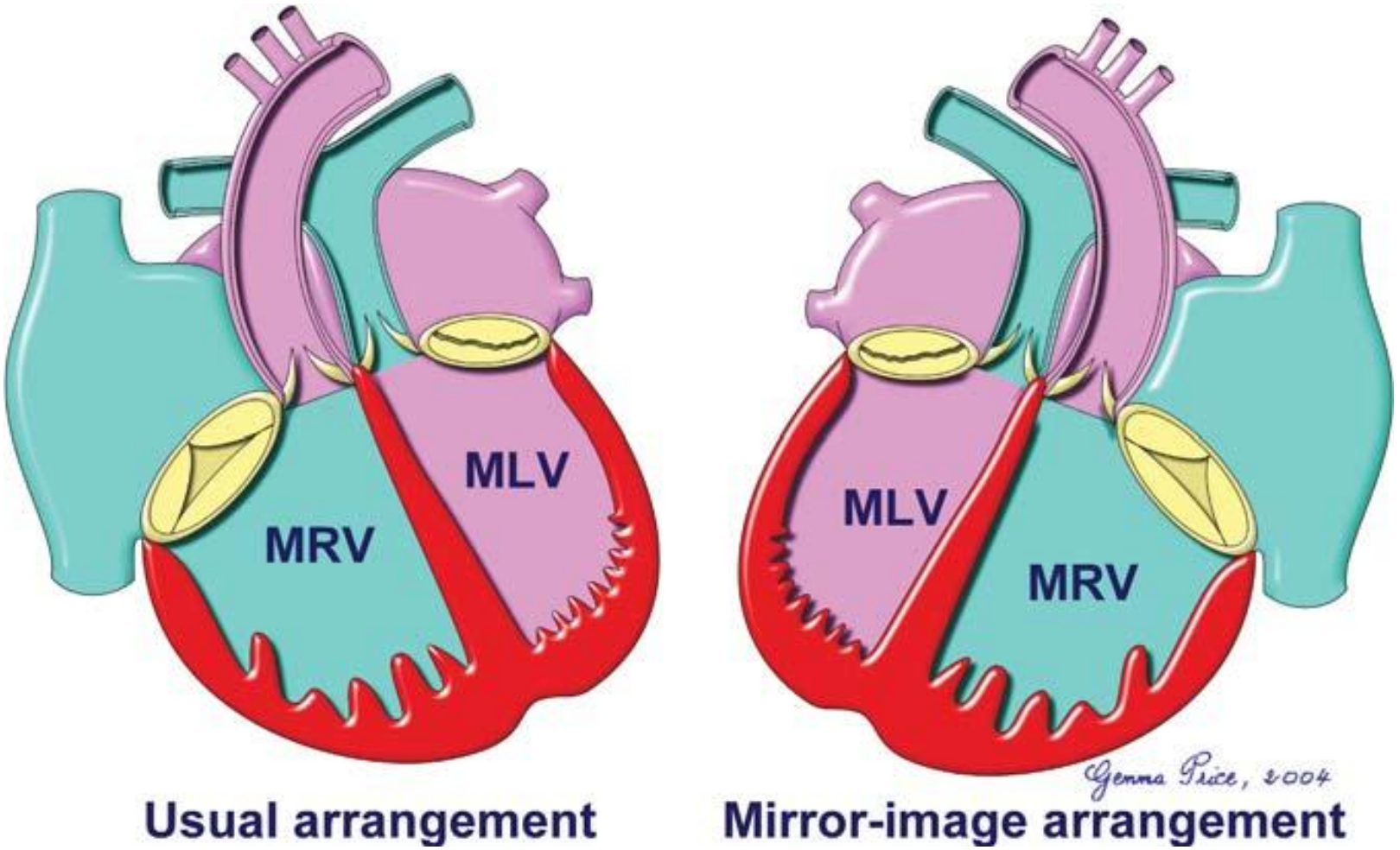
Transposition of the Great Arteries

- **John Farre (1814)**

  “transposition of aorta and pulmonary artery”

- **Concordant atrio-ventricular connections**
- **Discordant ventriculo-arterial connections**



Cardiol Young 2005;15(Suppl 1):76-87

Epidemiology

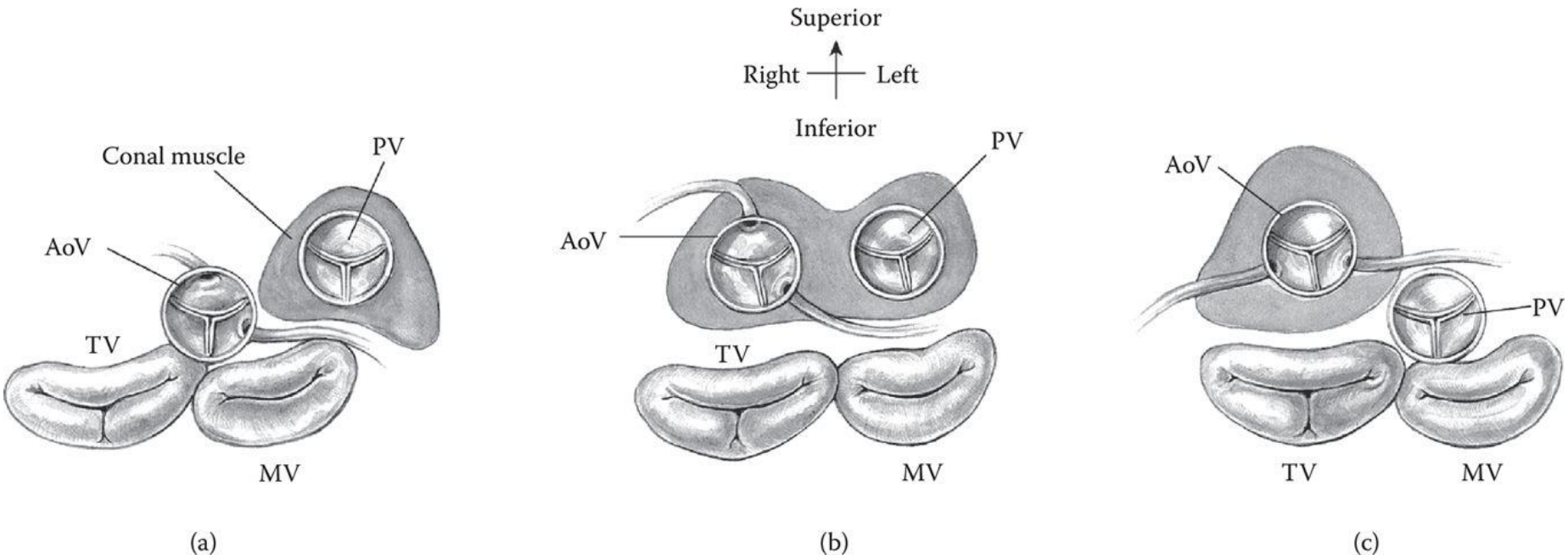
- **About 5% of all congenital heart diseases**
- **0.2 per 1000 live births**
- **If not treated, 90% of children with TGA and intact septum will die by 1 year of age.**

Embryology

- **A type of conotruncal anomaly**

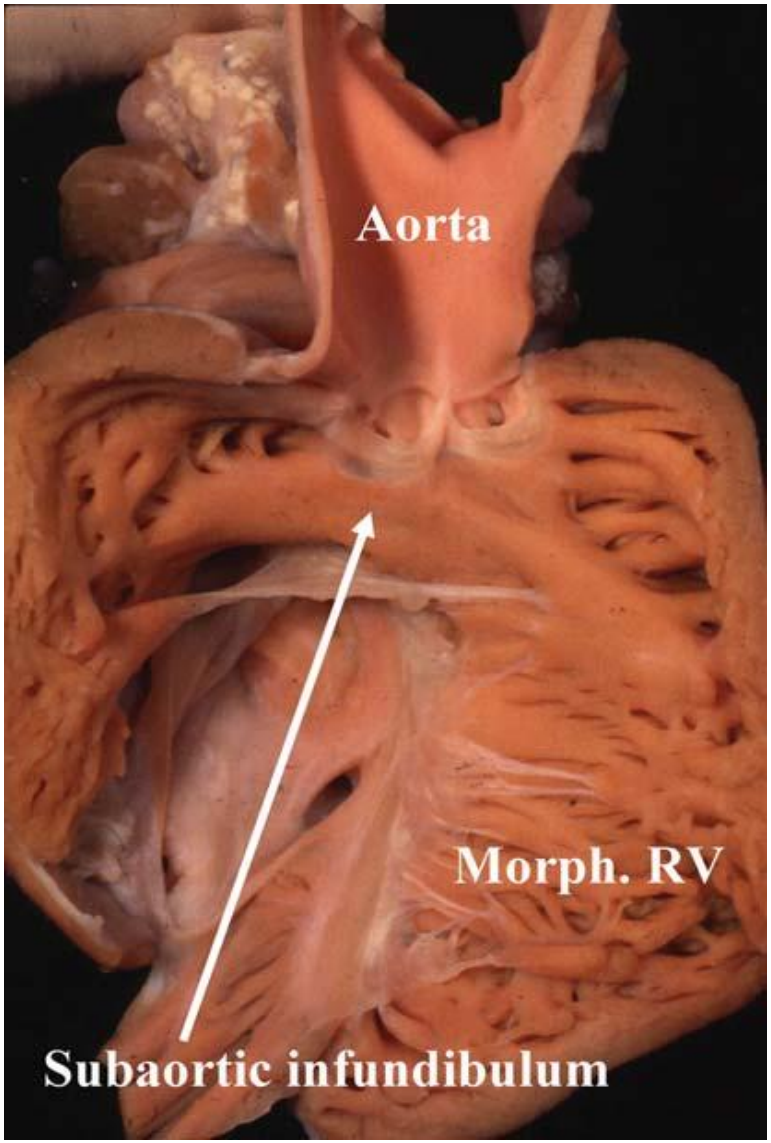
- **Deranged development of the cardiac outflow tract**

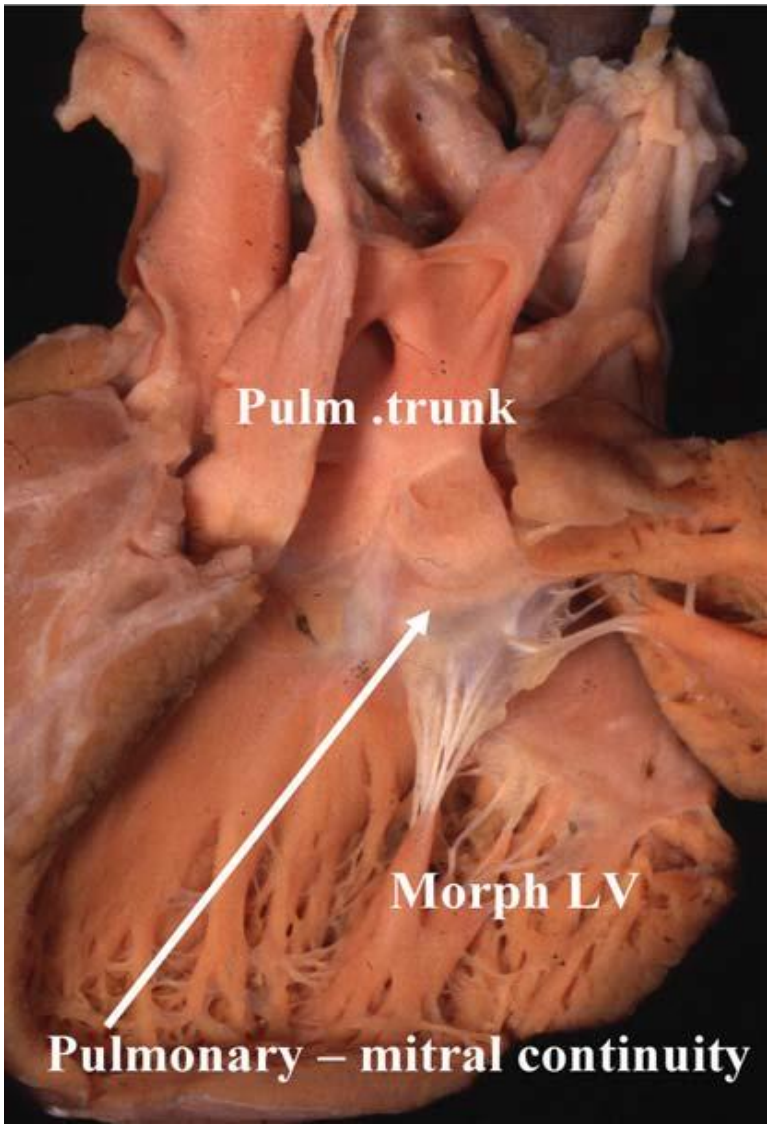
Van Praagh's Theory of Conal Underdevelopment



(a) Normal heart
(b) Double outlet right ventricle
(c) Transposition of the great arteries

Jonas RA. Comprehensive surgical management of congenital heart disease. 2nd ed. 2014





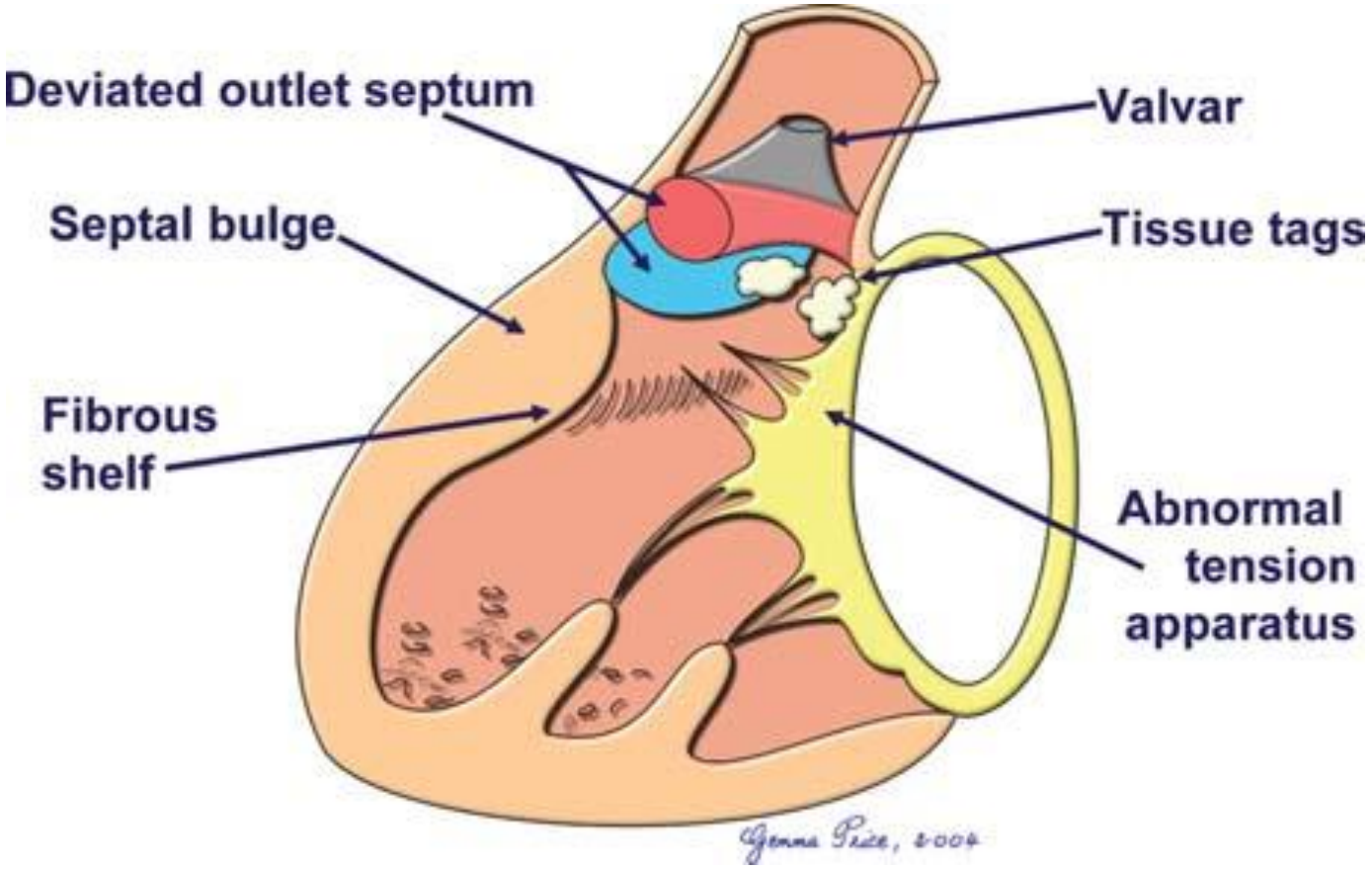
Cardiol Young 2005;15(Suppl 1):76-87

제7차 전공의 학술세미나

Associated Anomalies

- PDA
- PFO, ASD
- VSD
- LVOTO
- Coronary anomalies
- Aortic arch anomalies

LVOTO

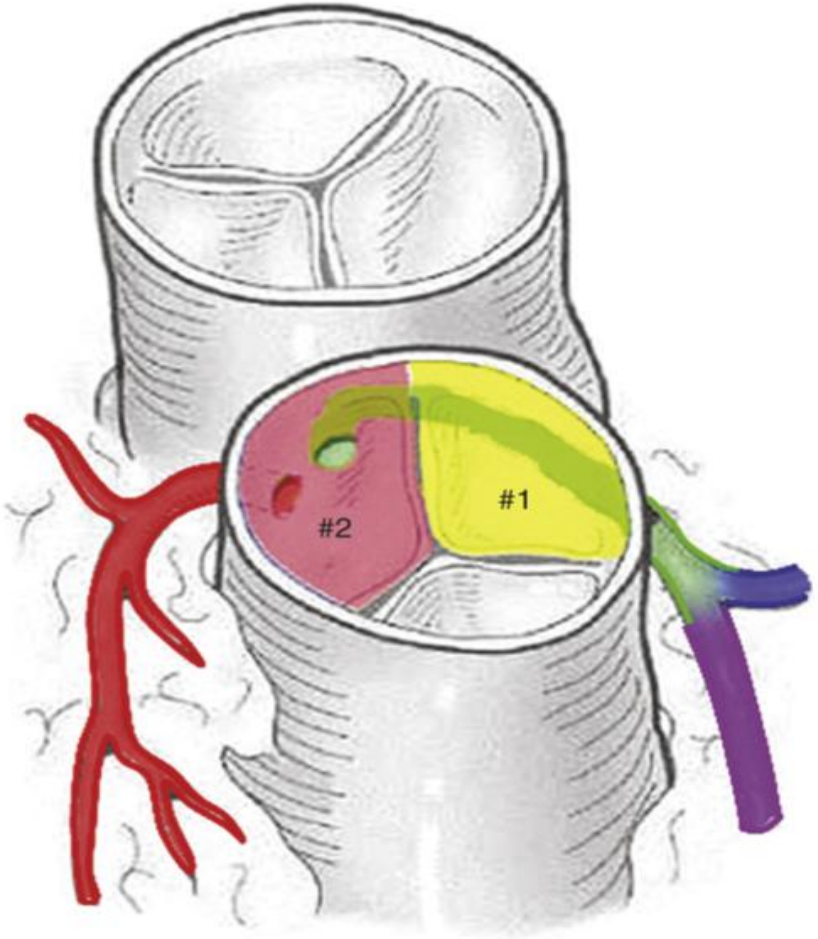


Cardiol Young 2005;15(Suppl 1):76-87

Coronary Anomalies

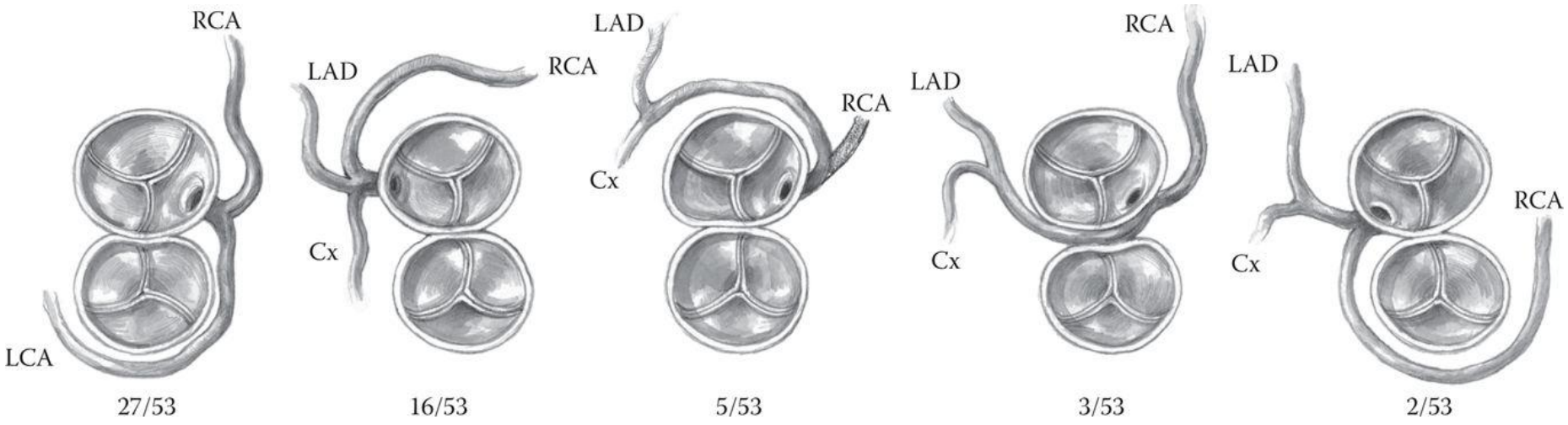
- **Intramural coronary artery**
- **Single coronary ostium**
- **Coronary ostial atresia, stenosis**

Intramural Coronary Artery



Anderson RH. Paediatric cardiology. 3rd ed. 2010

Single Coronary Artery

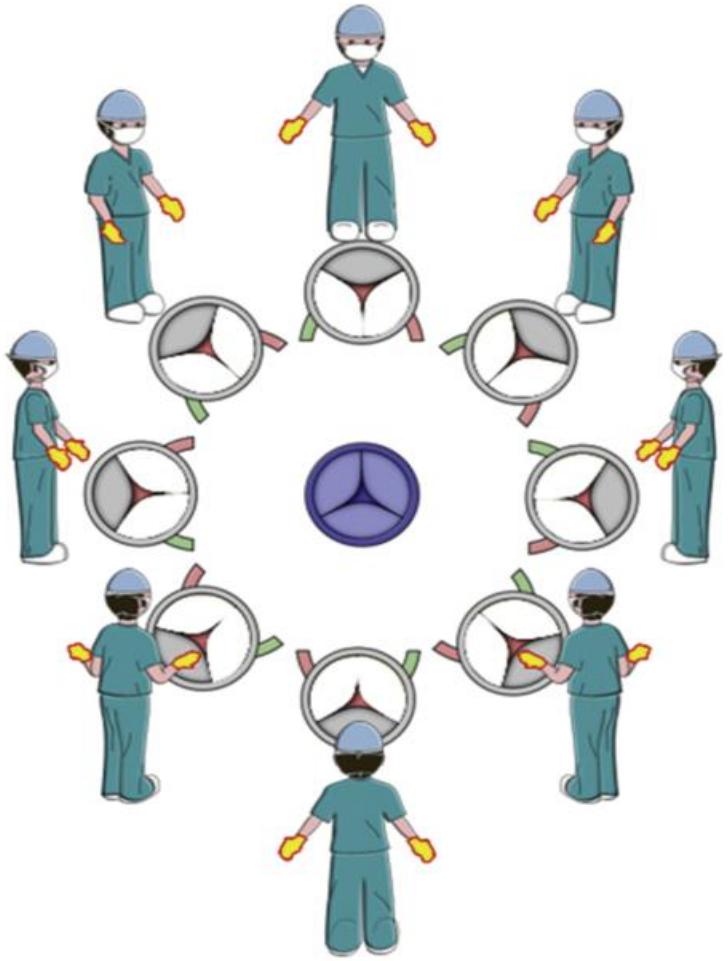


Jonas RA. Comprehensive surgical management of congenital heart disease. 2nd ed. 2014

Coronary Artery Branching Patterns

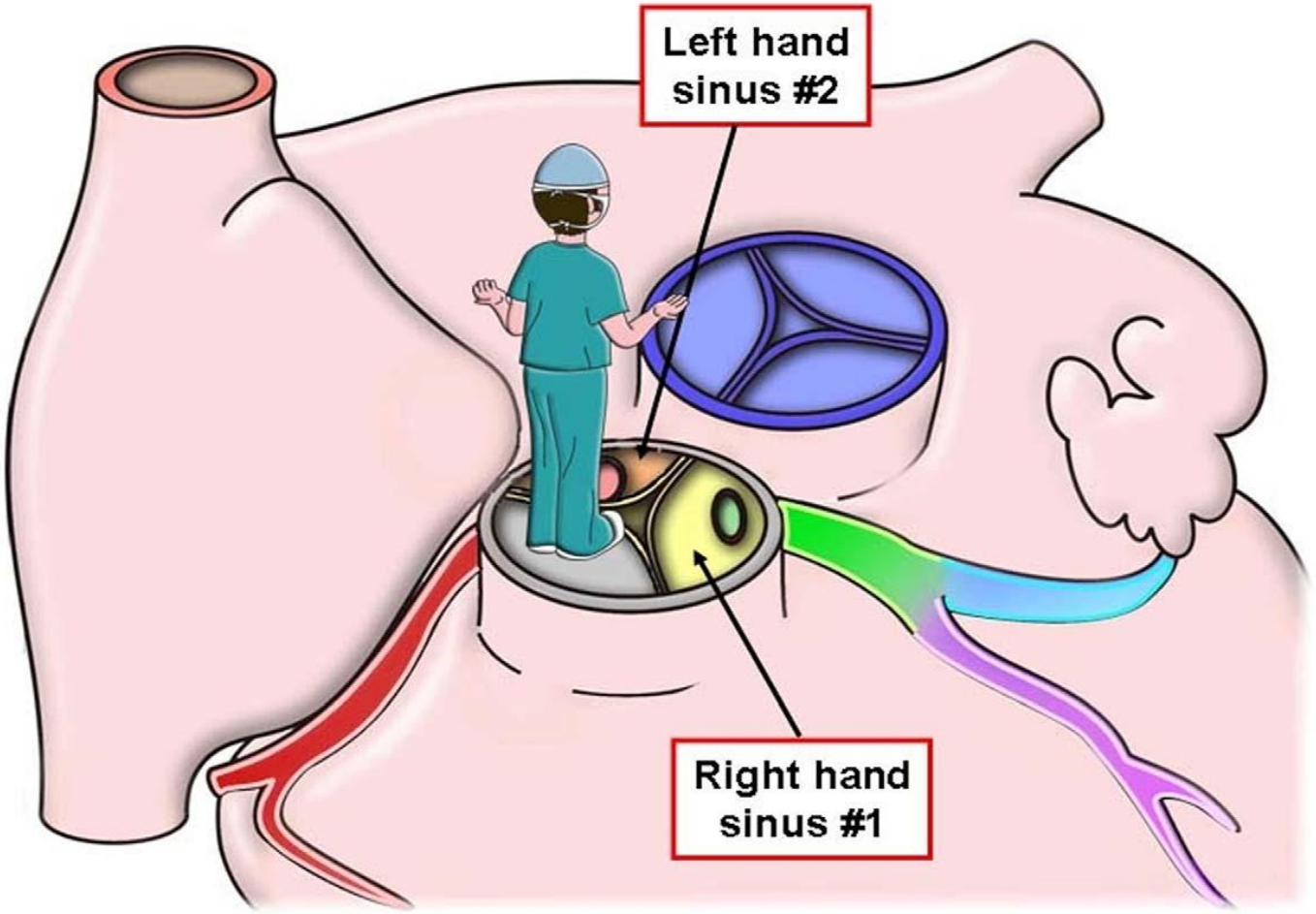
- **Leiden convention**
- **Yacoub and Radley-Smith classification**
- **Descriptive classification**

Variability of the Aorta Relative to the Pulmonary Trunk



Anderson RH. Paediatric cardiology. 3rd ed. 2010

Leiden Convention

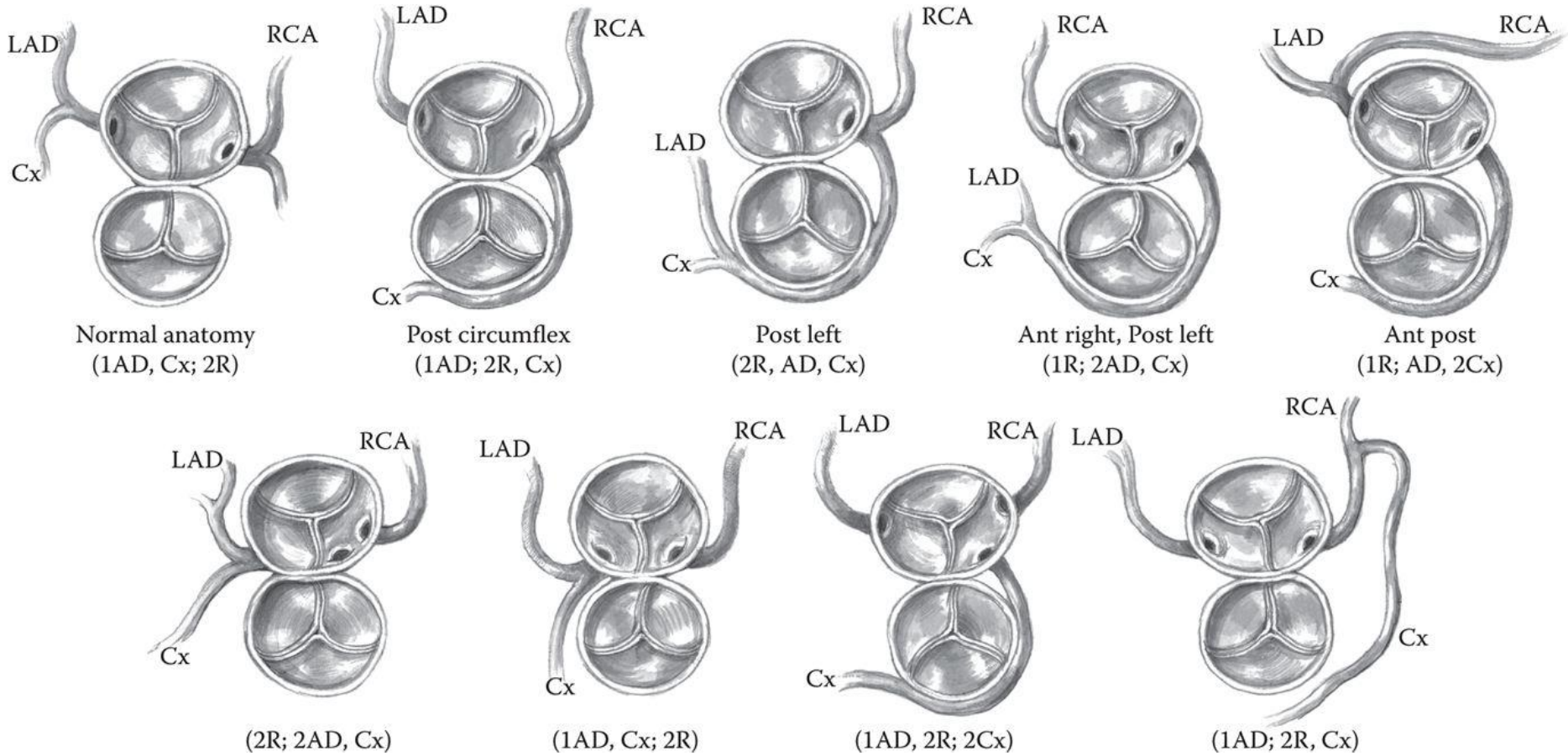


World J Pediatr Congenit Heart Surg 2011;2:9-18

Leiden Convention

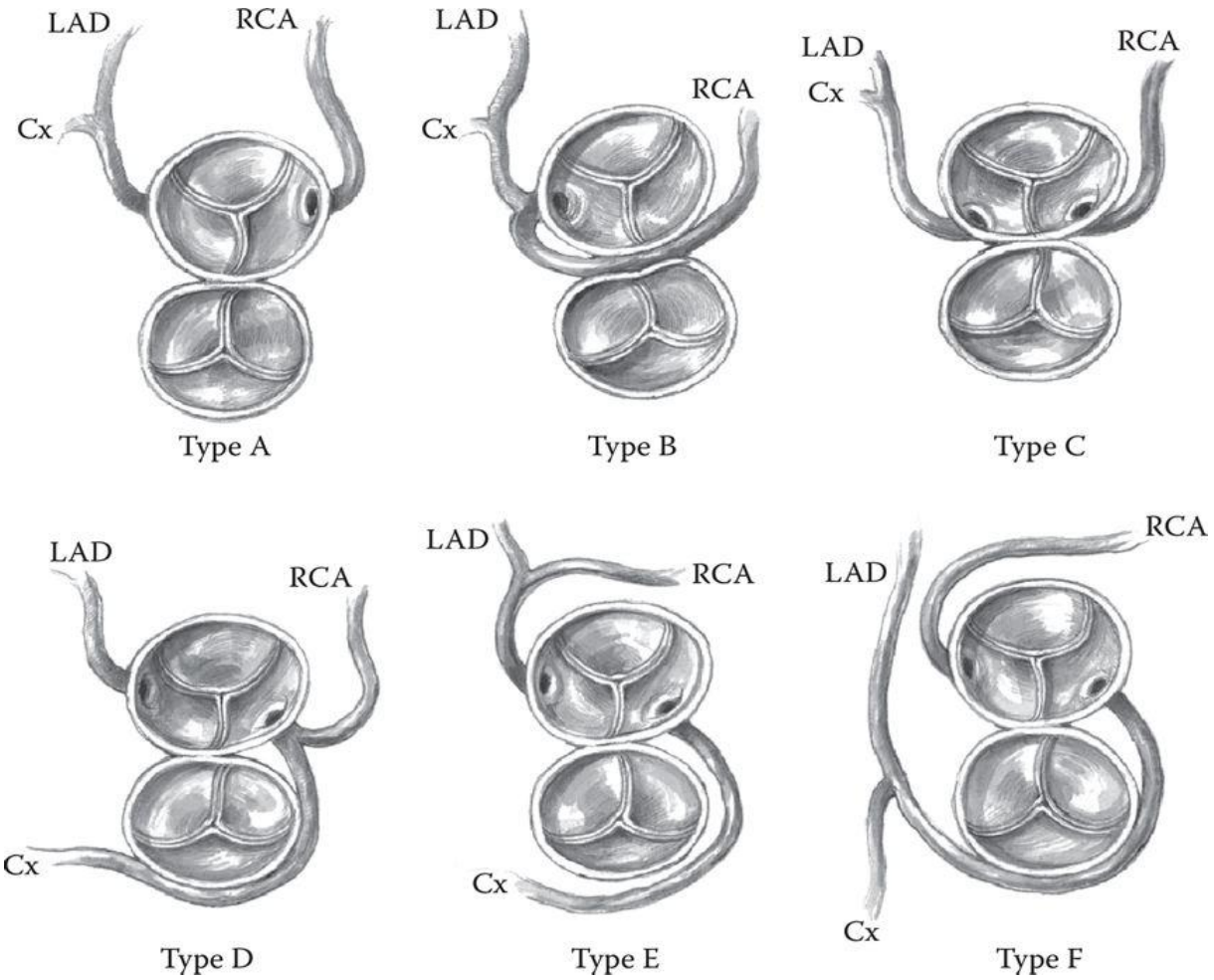
- **AD: Anterior descending artery**
- **Cx: Circumflex artery**
- **R: Right coronary artery**

Leiden Convention



Jonas RA. Comprehensive surgical management of congenital heart disease. 2nd ed. 2014

Yacoub and Radley-Smith Classification



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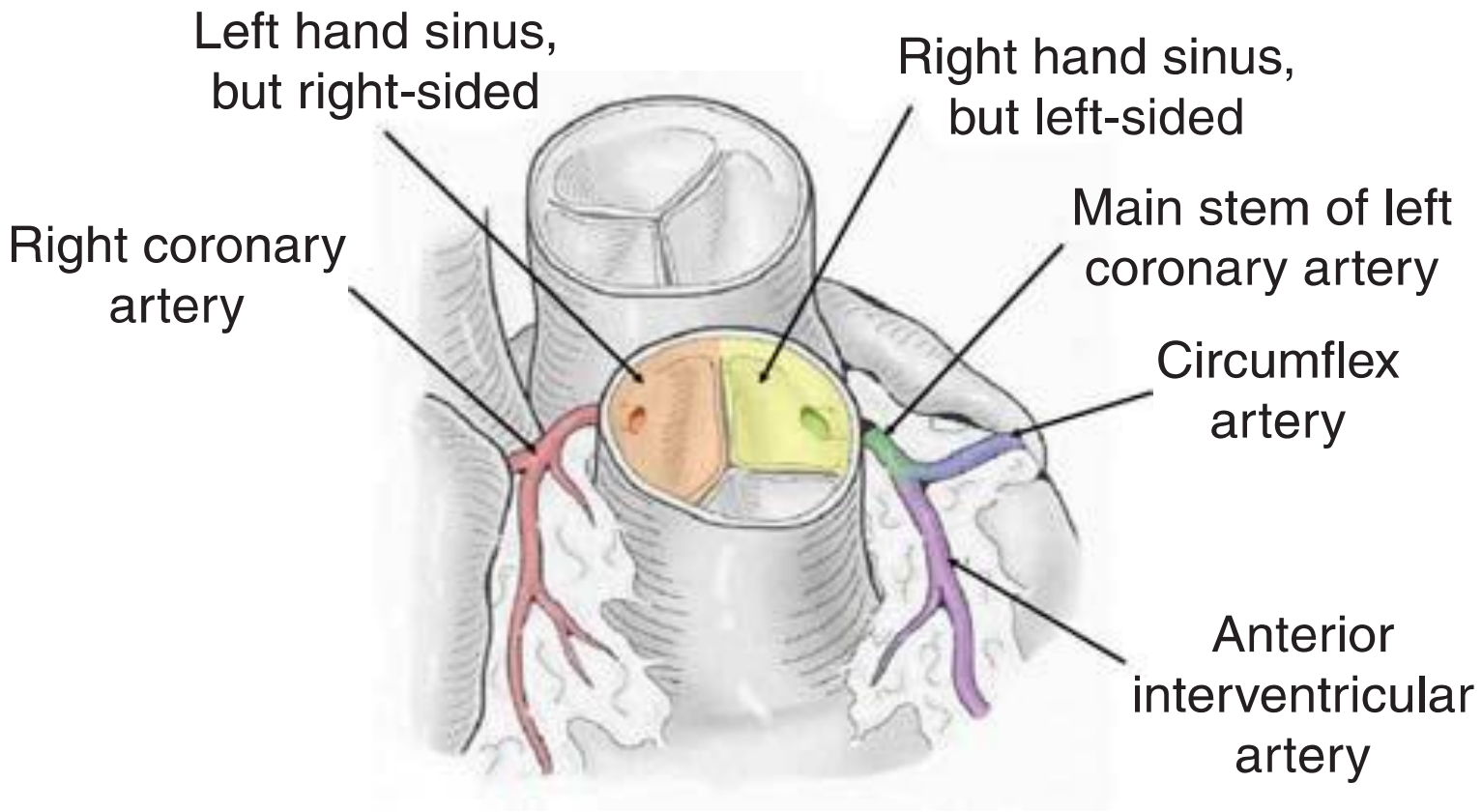
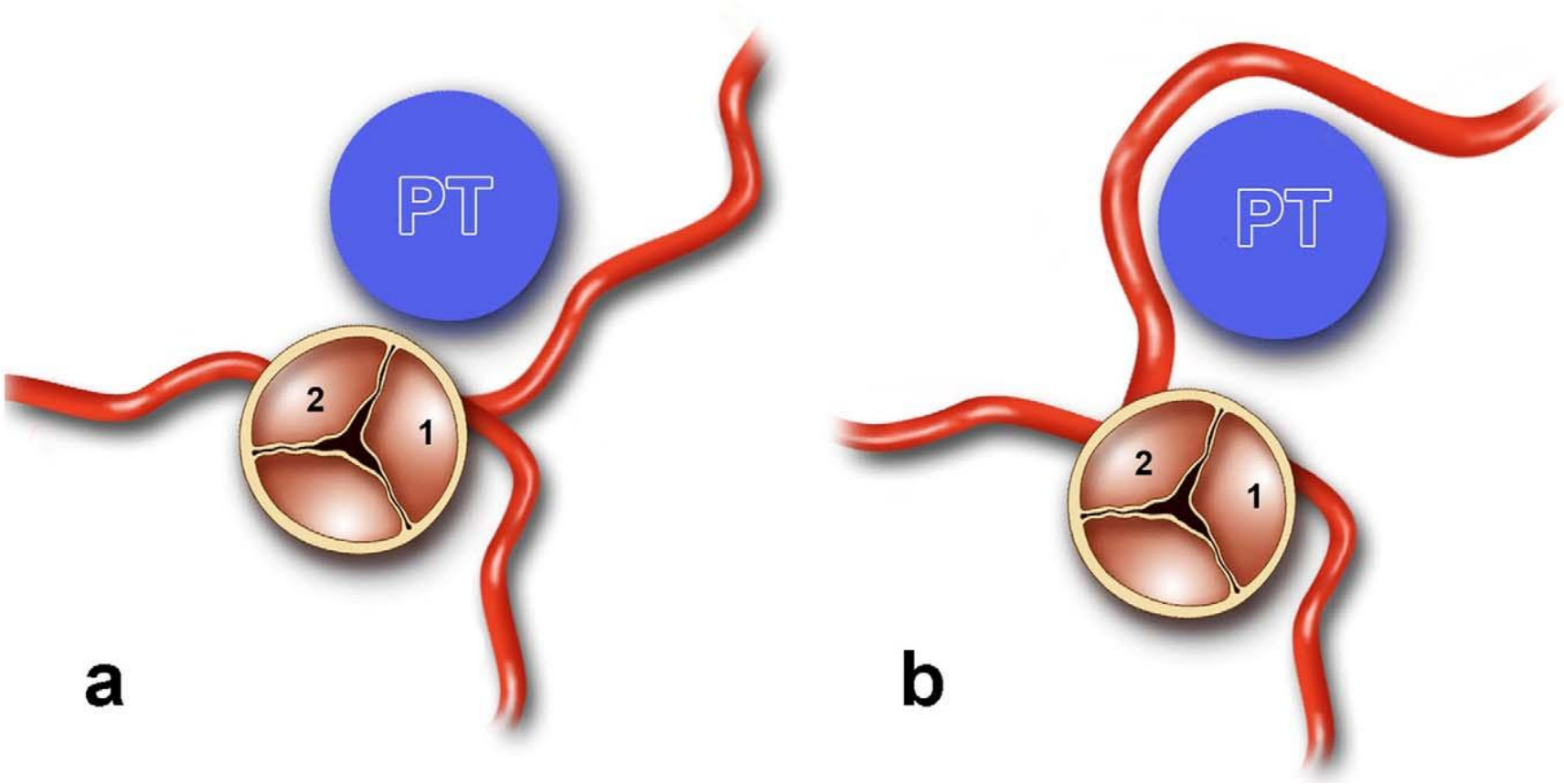
Usual Coronary Artery Pattern

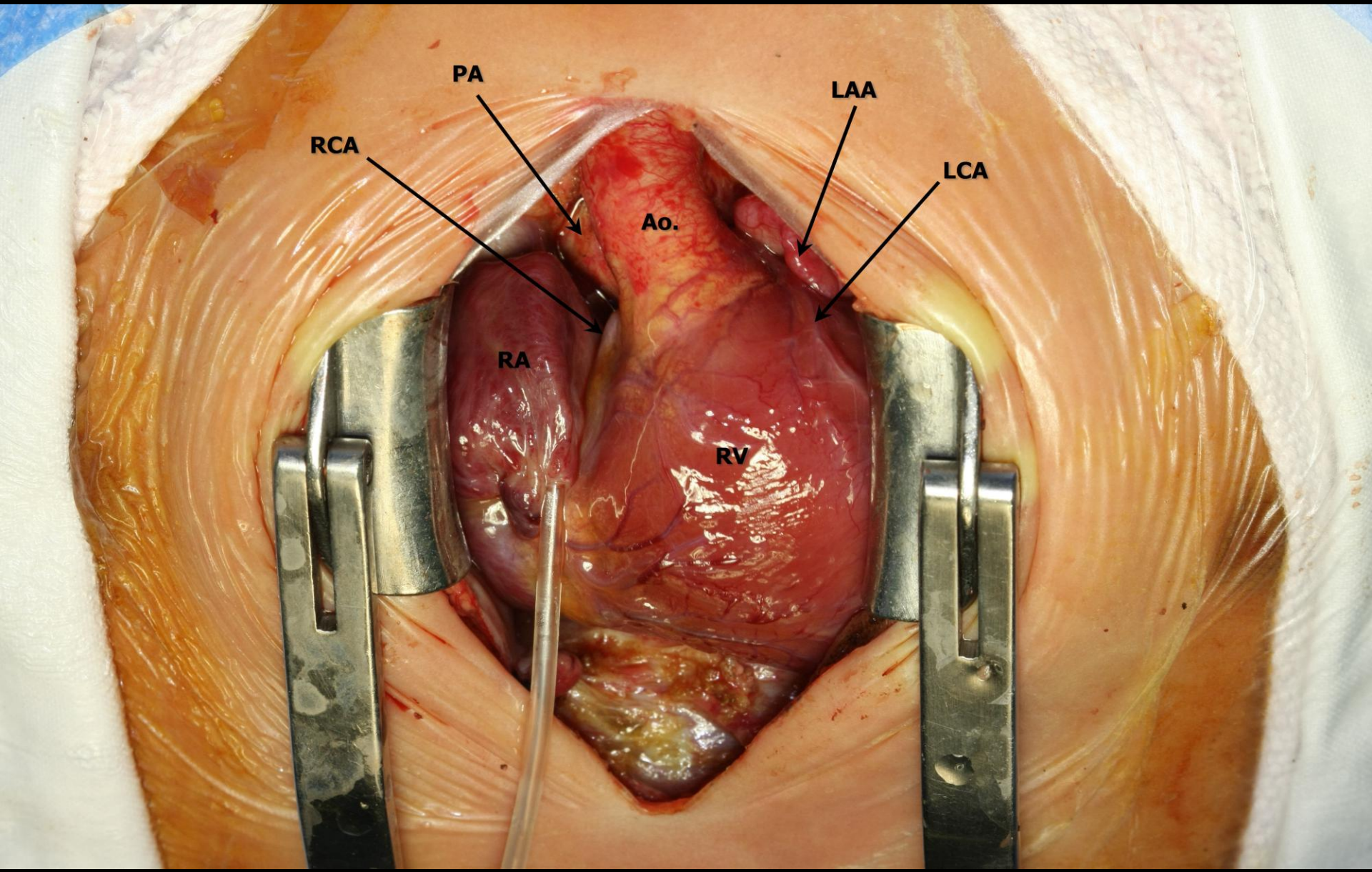


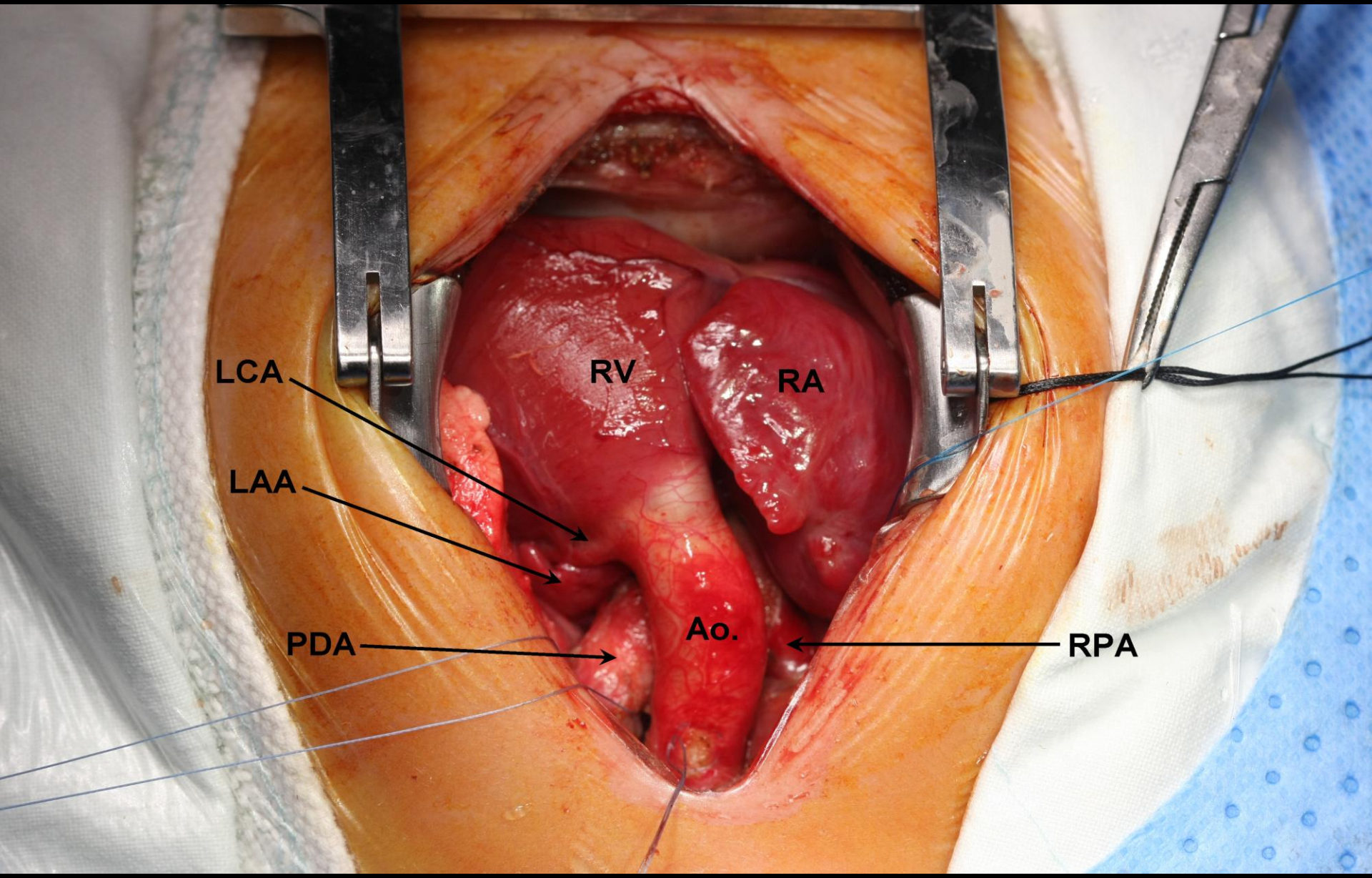
Cardiol Young 2005;15(Suppl 1):93-101

Common Coronary Artery Pattern



World J Pediatr Congenit Heart Surg 2011;2:9-18


PA
LAA
RCA
LCA
Ao.
RA
RV


LCA
RV
RA
LAA
PDA
Ao.
RPA

Pathophysiology

- **Two parallel circulations**
- **Mixing between the parallel circulations:**

  PDA, ASD, VSD
- **Rapid progression of pulmonary vascular disease**
- **LVOTO**
- **LV pressure**

Diagnosis

- **Echocardiography**

  all necessary anatomical information

- **Cardiac catheterization**

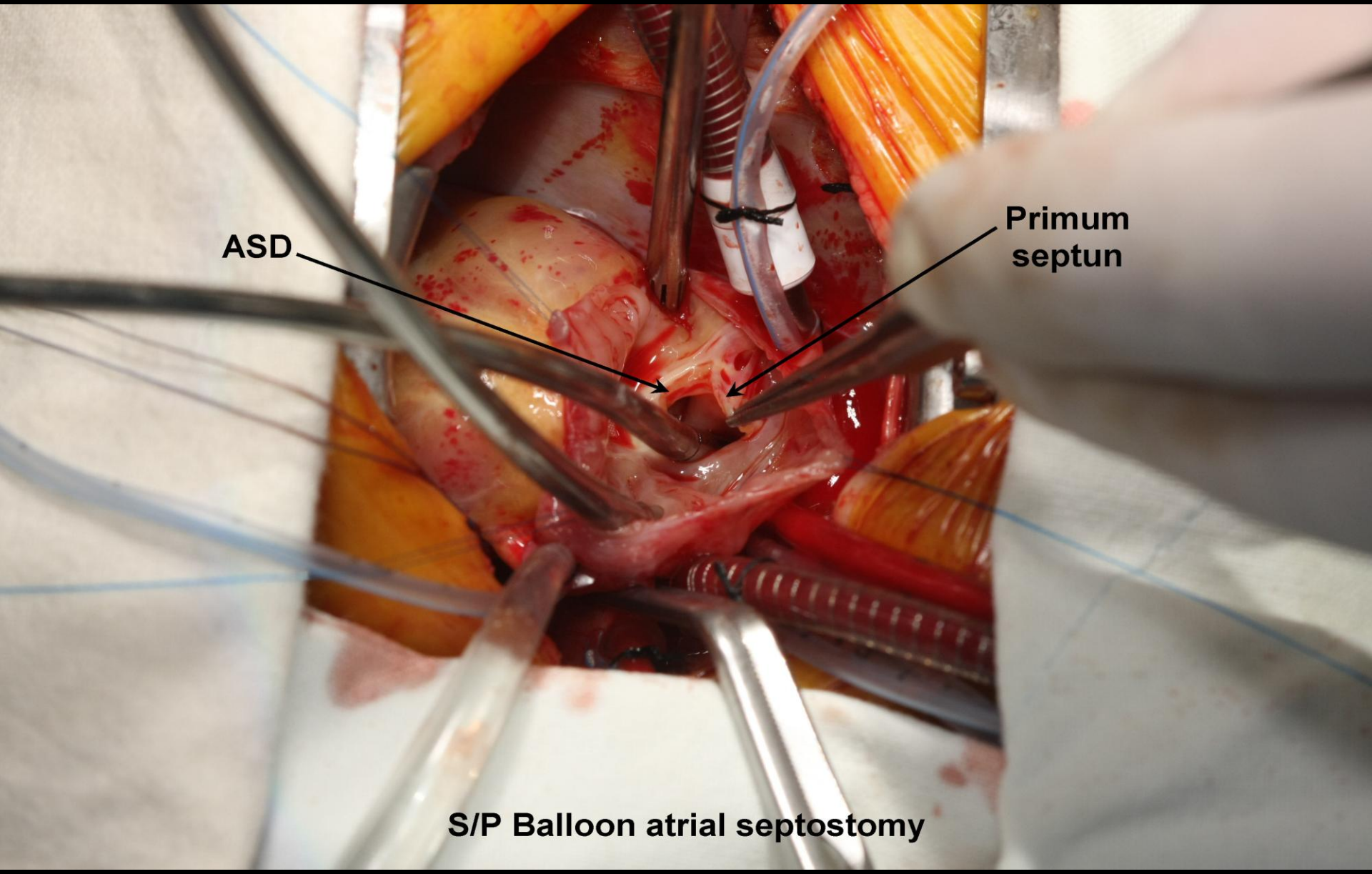
  assessment of the pulmonary vascular resistance

- **MRI**

  assessment of LV mass in the infant presenting late

Medical Management

- **Prostaglandin E_1 infusion to maintain PDA**
- **Balloon atrial septostomy**


ASD
Primum
septun
S/P Balloon atrial septostomy

Surgical Management

- Atrial switch procedure (Senning, Mustard)
- Arterial switch procedure
- Rastelli / REV / Nikaidoh procedure
- Arterial switch + arch repair
- Arterial switch after LV retraining

Timing of Arterial Switch Procedure

- **Usually in the first or second week of life**

- **Time limit of primary arterial switch procedure in children with intact ventricular septum:**
  8 weeks of age with liberal use of mechanical circulatory support

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Transposition of the Great Arteries—Outcomes and Time Interval of Early Neonatal Repair

Michael T. Cain, BS[1], Yumei Cao, PhD[2,3,4], Nancy S. Ghanayem, MD[4,5], Pippa M. Simpson, PhD[2,3,4], Katie Trapp, BS[4], Michael E. Mitchell, MD[4,6], James S. Tweddell, MD[4,6], and Ronald K. Woods, MD, PhD[4,6]

Abstract

Background: This study evaluates the relationship of morbidity and resource utilization with the timing of early neonatal repair of transposition of the great arteries and intact ventricular septum (d-TGA/IVS). **Methods:** All patients with d-TGA/IVS who underwent arterial switch in the first 14 days of life, between January 2000 and May 2011, were reviewed. Patients undergoing repair at $\leq$4 days of age were categorized as group I, 5 to 7 days as group II, and 8 to 14 days as group III. Outcomes included mortality, morbidity, and resource utilization. **Results:** Hospital survival was 69 (98.6%) of 70. The length of stay (LOS) and total charges were lowest in group I—15.5 days compared to group II—18.0 days and group III—23.5 days ($P = .005$); group I—US$128,219 compared to group II—US$141,729 and group III—US$217,427 ($P = .0006$). Using regression analysis to account for potentially confounding effects of multiple variables and treating time as a continuous variable demonstrated that age at surgery was significantly associated with total LOS ($P = .029$), hospital charges ($P = .029$) and intensive care unit charges ($P = .002$). Younger age at repair was not associated with worse outcomes for any measure of morbidity. **Conclusions:** Earlier repair of d-TGA/IVS was associated with decreased resource utilization and no detriment to clinical outcomes. Further analysis based on a larger cohort of patients is needed to verify these results that have important implications for improving the value of care.

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Earlier Arterial Switch Operation Improves Outcomes and Reduces Costs for Neonates With Transposition of the Great Arteries

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Jan M. Quaegebeur, MD, PhD,‡ Julie A. Vincent, MD,* Emile A. Bacha, MD‡
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Objectives This study sought to examine the impact of surgical timing on major morbidity and hospital reimbursement for late preterm and term infants with dextrotransposition of the great arteries (d-TGA).

Background Neonatal arterial switch operation is the standard of care for d-TGA. Little is known about the effects of age at operation on clinical outcomes or costs for these neonates.

Methods We conducted a retrospective cohort study of infants at ≥36 weeks' gestation, with d-TGA, with or without ventricular septal defects, admitted to our institution at 5 days of age or younger, between January 1, 2003 and October 1, 2012. Children with other cardiac abnormalities or other major comorbid conditions were excluded. Univariable and multivariable analyses were performed to determine the effects of age at operation on major morbidity and hospital reimbursement.

Results A total of 140 infants met inclusion criteria. Reimbursement data were available for them through January 1, 2012 ($n = 128$). The mortality rate was 1.4% ($n = 2$). Twenty percent ($n = 28$) experienced a major morbidity. The median costs were $60,000, in 2012 dollars (range: $25,000 to $549,000). The median age at operation was 5 days (range: 1 to 12 days). For every day later that surgery was performed, beyond day of life 3, the odds of major morbidity increased by 47% (range: 23% to 66%, $p < 0.001$) and costs increased by 8% (range: 5% to 11%, $p < 0.001$), after considering the effects of sex, birth weight, gestational age, year at which surgery was performed, transfer, weekend admission, insurance, surgeon, septostomy, bypass and cross-clamp times, and the presence of ventricular septal defects or abnormal coronary anatomy.

Conclusions Delay of neonatal arterial switch operation beyond 3 days is significantly associated with increased morbidity and healthcare costs. (J Am Coll Cardiol 2014;63:481–7) © 2014 by the American College of Cardiology Foundation

Primary Arterial Switch Operation for Transposition of the Great Arteries With Intact Ventricular Septum in Infants Older Than 21 Days

JOHN P. FORAN, MRCP, IAN D. SULLIVAN, FRACP, MARTIN J. ELLIOTT, FRCS, MARC R. DE LEVAL, FRCS

London, England, United Kingdom

Objectives. **The aim of this study was to assess the surgical outcome of the primary arterial switch operation (ASO) in infants 3 weeks to 2 months old.**

Background. **The surgical management of transposition of the great arteries and intact ventricular septum (TGA/IVS) beyond 2 to 3 weeks of age is controversial. Concern that regression of the left ventricular (LV) myocardial mass will render the left ventricle incapable of coping with the acutely increased work of systemic perfusion has been considered a contraindication to a primary ASO.**

Methods. **We used retrospective analysis of 37 patients 3 weeks to 2 months old and 156 patients <3 weeks old who underwent primary ASO with TGA/IVS to determine the surgical outcomes.**

Results. **Between January 1990 and December 1996, primary ASO was performed in 37 patients 21 to 61 days old (late ASO group) and 156 patients <21 days old (early ASO group) with TGA/IVS. One (2.7%, 95% confidence interval [CI] 0.07% to 14.2%) of 37 patients and 13 (8.3%, 95% CI 4.5% to 13.8%) of 156 patients died. One late death occurred in each group. Mechanical LV support was required in 1 (2.7%, 95% CI 0.07% to 14.2%) of 37 late ASO and 6 (3.8%, 95% CI 1.4% to 8.2%) of 156 early ASO group patients postoperatively. Neither death nor the need for mechanical LV support in the late ASO group patients could be attributed to LV failure. In the late ASO group, age, LV geometry, LV mass index, LV posterior wall thickness index, LV volume index, LV mass/volume ratio, patent arterial duct or pattern of coronary anatomy did not predict death, duration of postoperative ventilation or inotropic support or time in intensive care. Moreover, there was no difference in duration of ventilation, duration of inotropic support or the time spent in intensive care in comparison to a random sample of 37 neonates from the early ASO group.**

Conclusions. **Primary ASO may be appropriate treatment for infants with TGA/IVS ≤2 months old, regardless of preoperative echocardiographic variables. The upper age limit for which primary ASO is indicated in TGA/IVS is not yet defined.**

(J Am Coll Cardiol 1998;31:883–9)

Extending the Boundaries of the Primary Arterial Switch Operation in Patients With Transposition of the Great Arteries and Intact Ventricular Septum

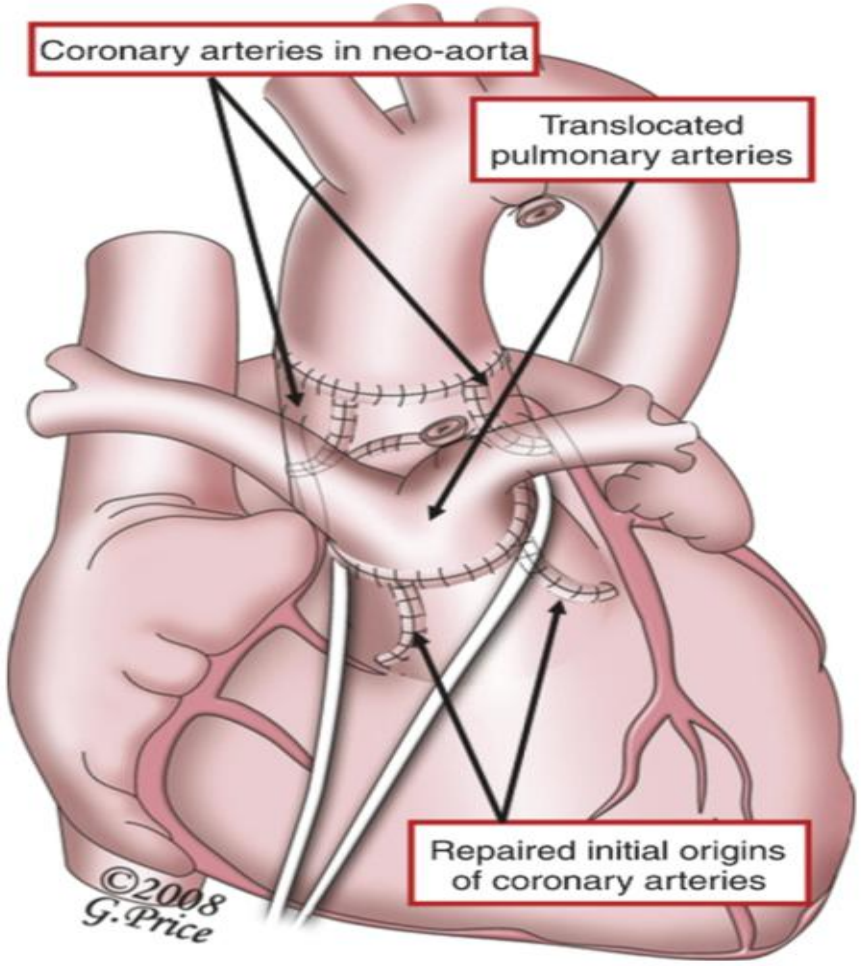
Nicholas Kang, FRACS; Marc R. de Leval, MD, FRCS; Martin Elliott, MD, FRCS; Victor Tsang, MS, MSc, FRCS; Ergin Kocyildirim, MD; Igor Sehic, MD; John Foran, FRCP; Ian Sullivan, FRACP

Background—We have previously suggested that the primary arterial switch operation is a feasible strategy for patients with transposition of the great arteries and intact ventricular septum (TGA-IVS) up to age 2 months. This study reports our current results with this approach and examines whether this policy could be extended beyond age 2 months.

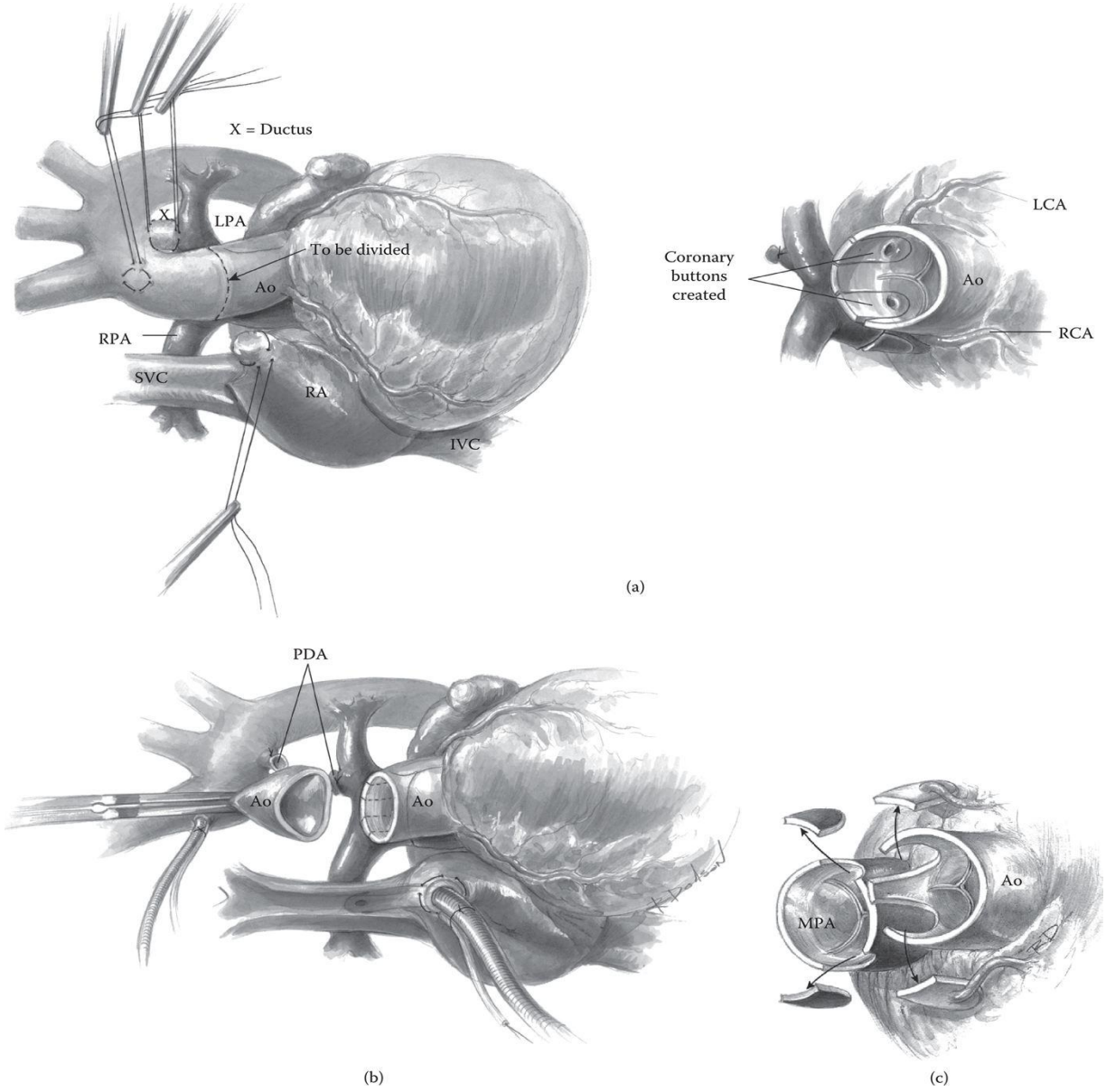
Methods and Results—380 patients who underwent arterial switch for TGA-IVS were reviewed. 275 patients were younger than 3 weeks at the time of surgery (early switch group); 105 patients were 3 weeks or older (range, 21 to 185 days) (late switch group). There was no difference in outcome in terms of in-hospital mortality (5.5% versus 3.8%) or need for mechanical circulatory support (3.6% versus 5.7%) between early and late switch groups. However, duration of postoperative ventilation (4.9 versus 7.1 days, $P=0.012$) and length of postoperative stay (12.5 versus 18.9 days, $P<0.001$) were significantly prolonged in the late switch group. Primary left ventricular failure resulting in death occurred in 2 patients in the late switch group, with no deaths in 9 patients aged 2 to 6 months.

Conclusions—This experience confirms that in TGA-IVS, the left ventricle maintains the potential for systemic work well beyond the first month of life. Consequently, neonates at high risk or late referrals can benefit from delayed arterial switch, even beyond age 2 months. However, the need for mechanical support in some of the older patients may limit the widespread adoption of such a strategy. **(*Circulation*. 2004;110[suppl II]:II-123–II-127.)**

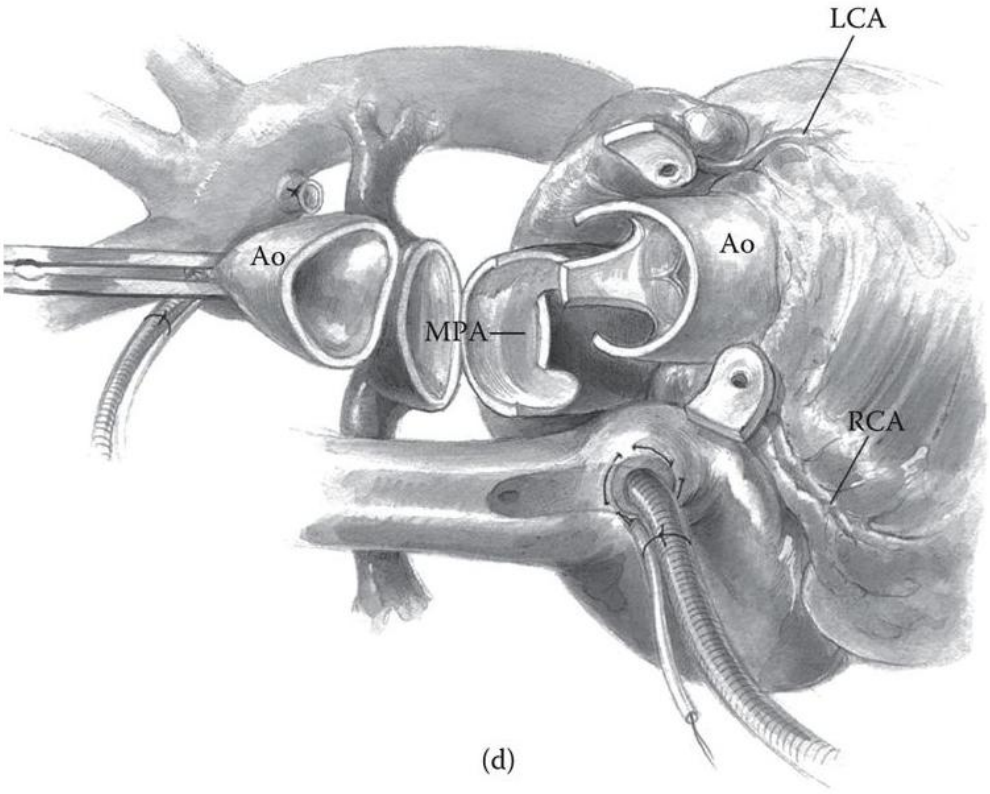
Arterial Switch Procedure

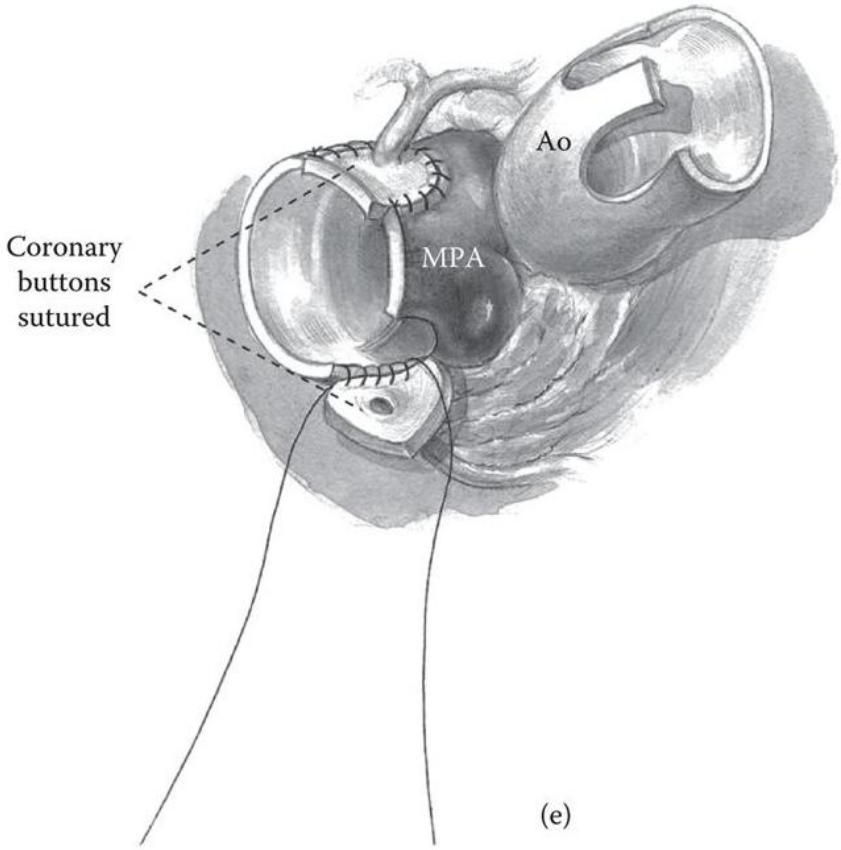


Anderson RH. Paediatric cardiology. 3rd ed. 2010



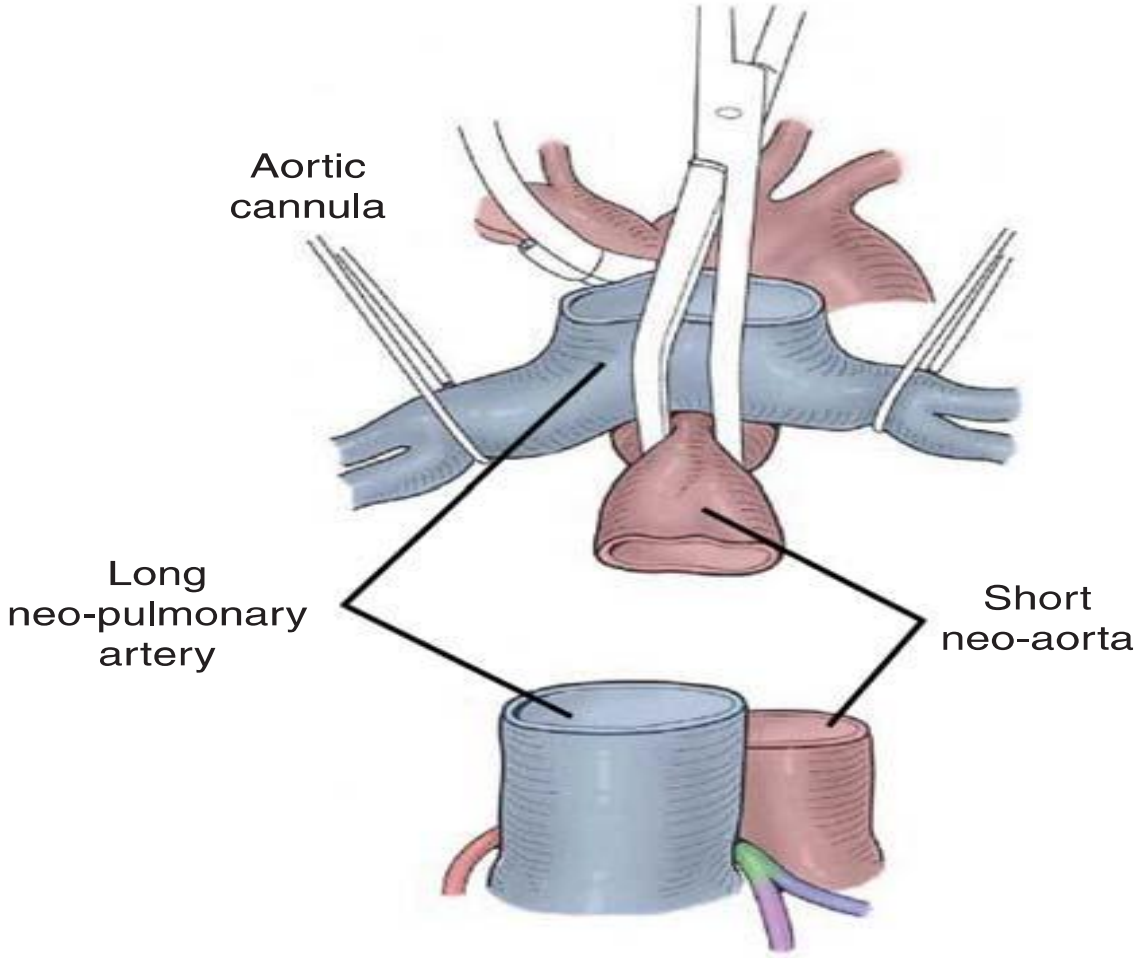
Jonas RA. Comprehensive surgical management of congenital heart disease. 2nd ed. 2014





Jonas RA. Comprehensive surgical management of congenital heart disease. 2nd ed. 2014

Lecompte Maneuver



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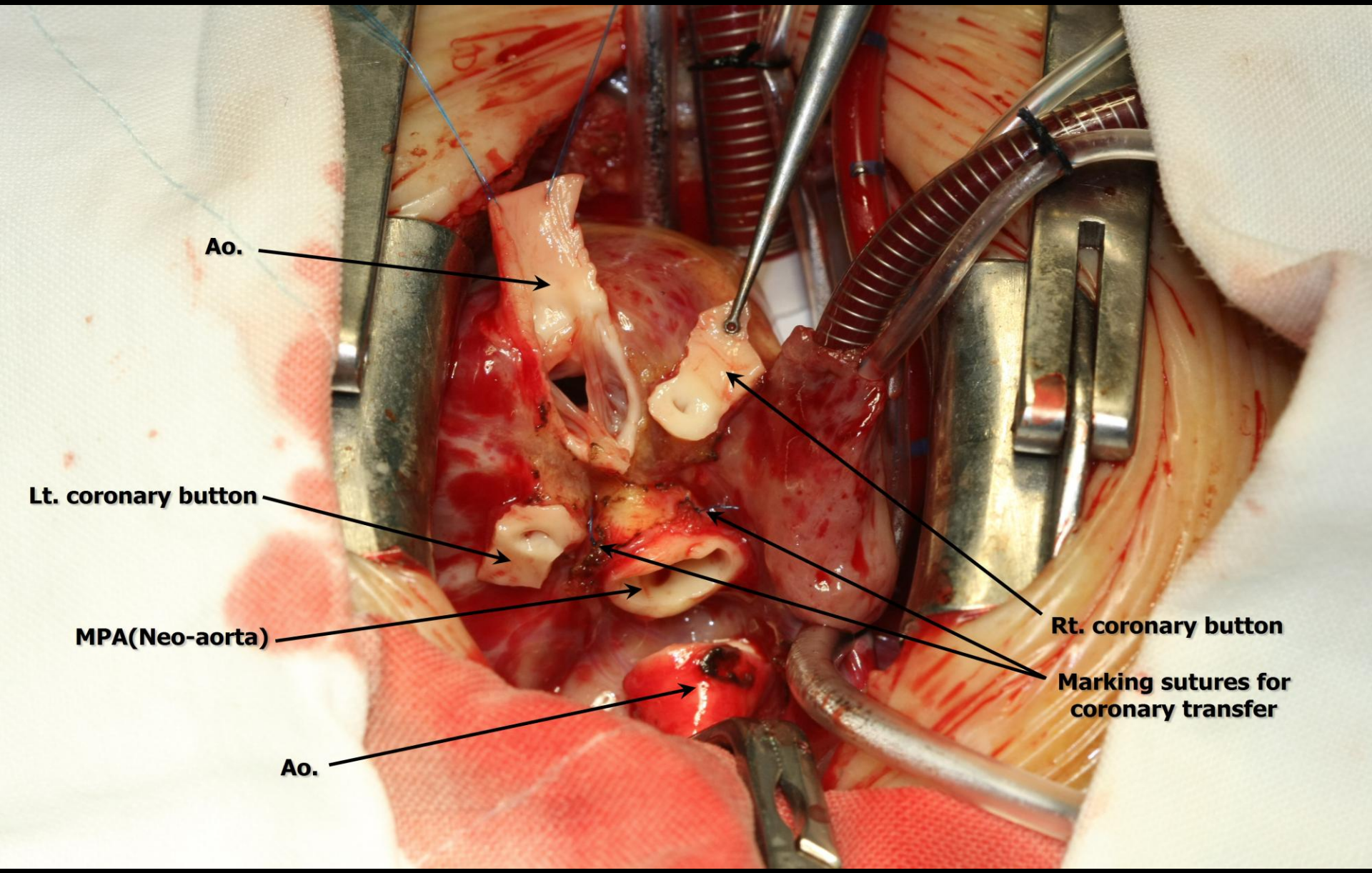
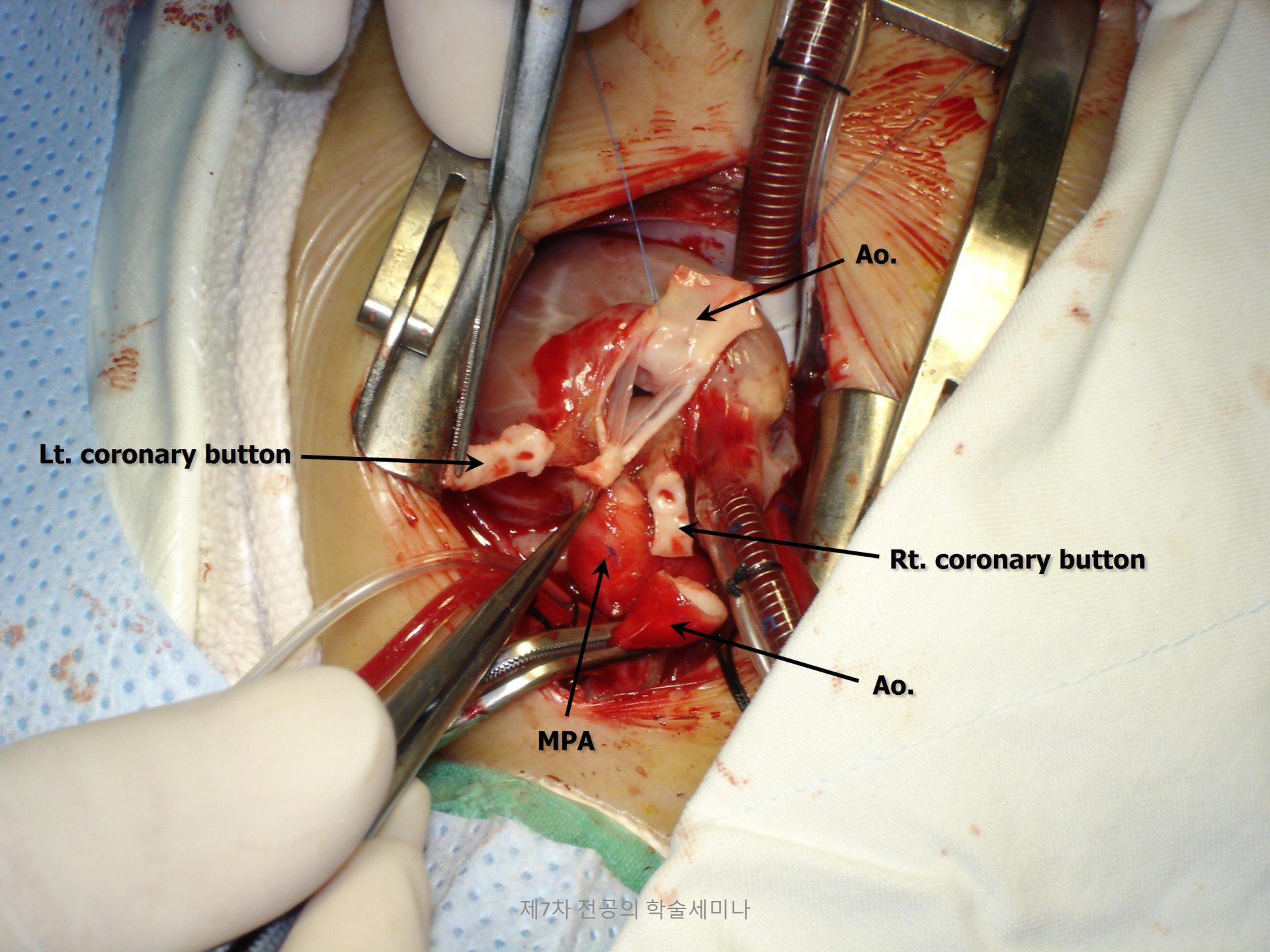
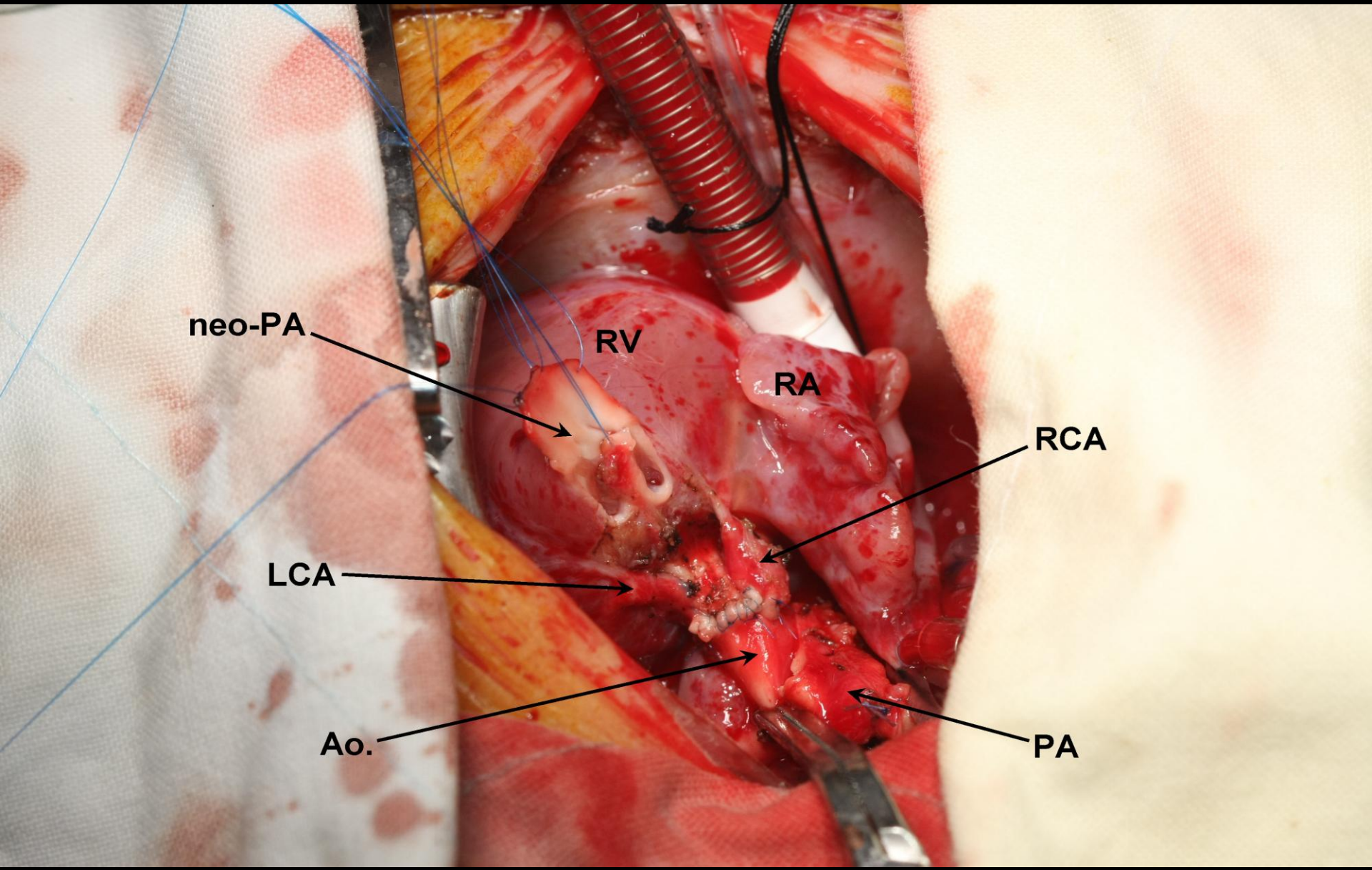
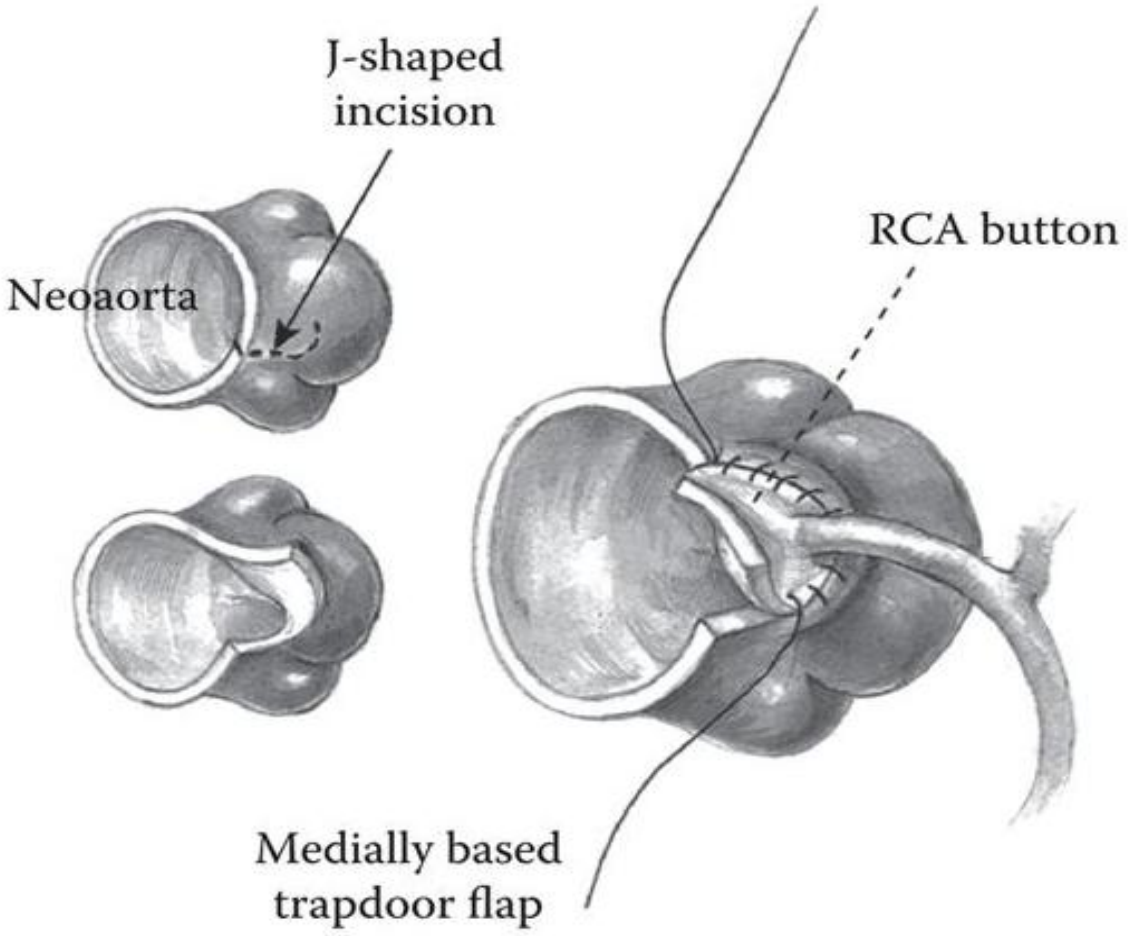

Ao.
Lt. coronary button
MPA(Neo-aorta)
Ao.
Rt. coronary button
Marking sutures for
coronary transfer


Ao.
Lt. coronary button
Rt. coronary button
Ao.
MPA

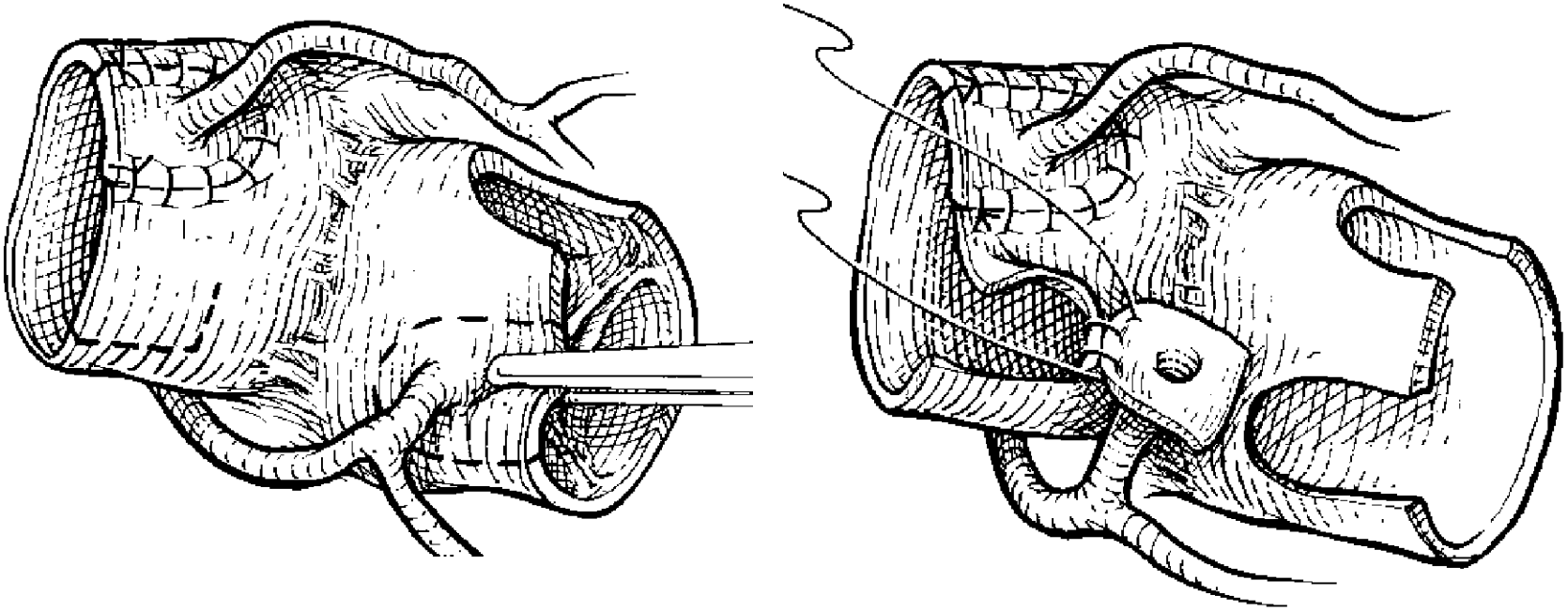

neo-PA
RV
RA
RCA
LCA
Ao.
PA

Trapdoor Technique



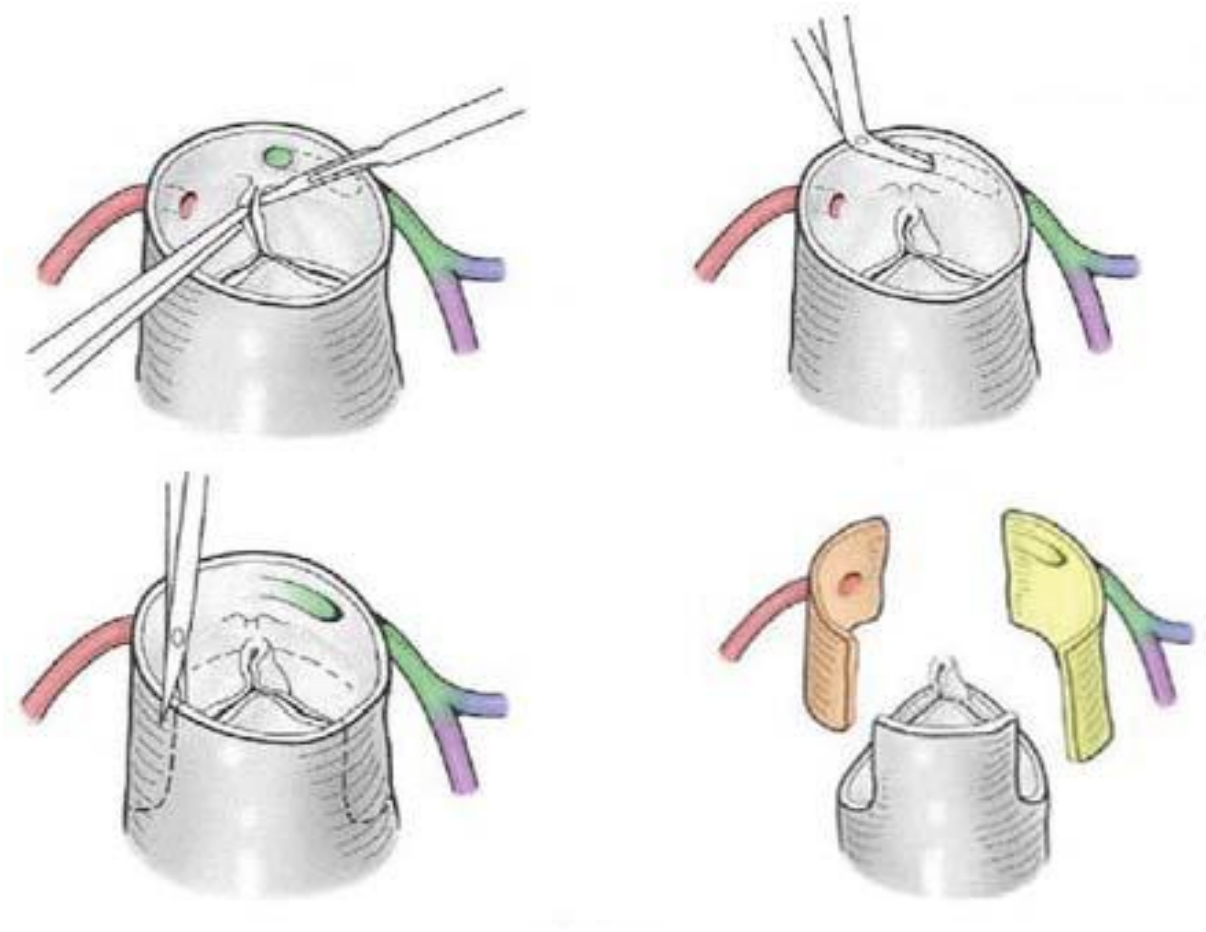
Jonas RA. Comprehensive surgical management of congenital heart disease. 2nd ed. 2014

Trapdoor Technique



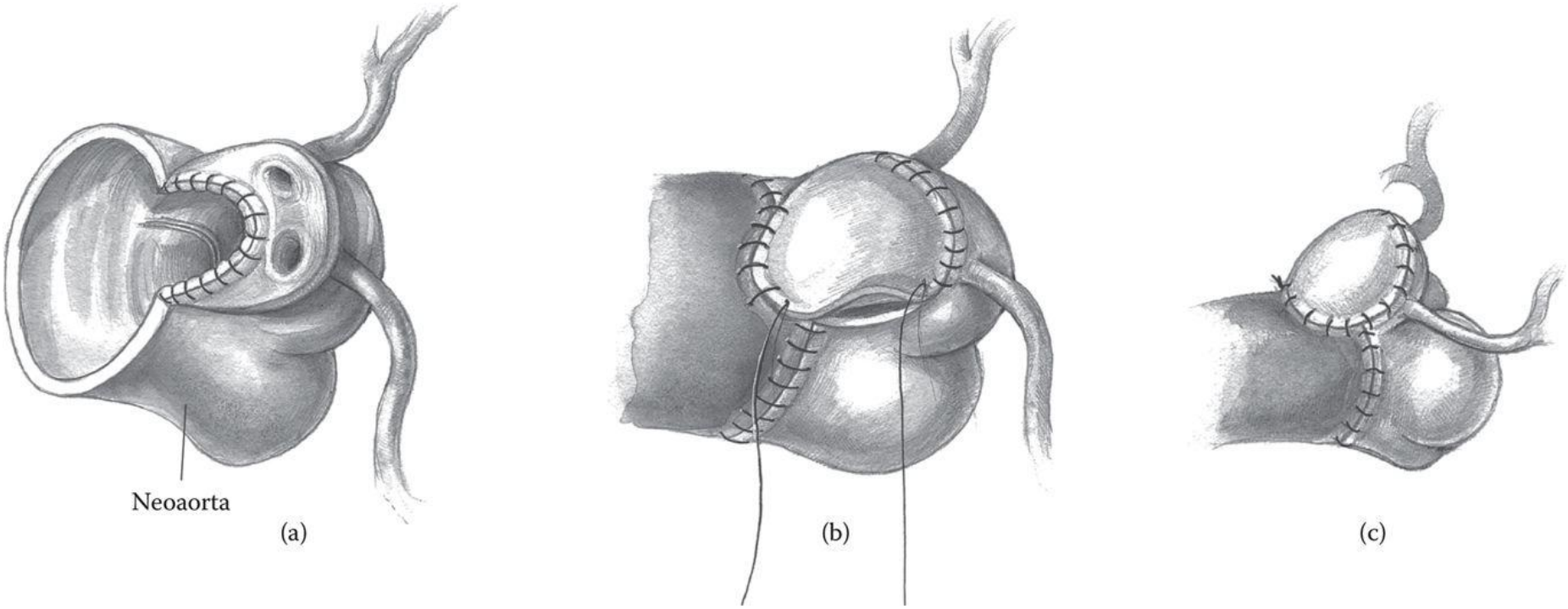
Kirklin/Barratt-Boyes Cardiac surgery. 4th ed. 2013

Intramural Coronary Artery



Cardiol Young 2005;15(Suppl 1):93-101

Single Coronary Artery Between the Aorta and Pulmonary Artery



Jonas RA. Comprehensive surgical management of congenital heart disease. 2nd ed. 2014

Coronary Artery Pattern and Outcome of Arterial Switch Operation for Transposition of the Great Arteries

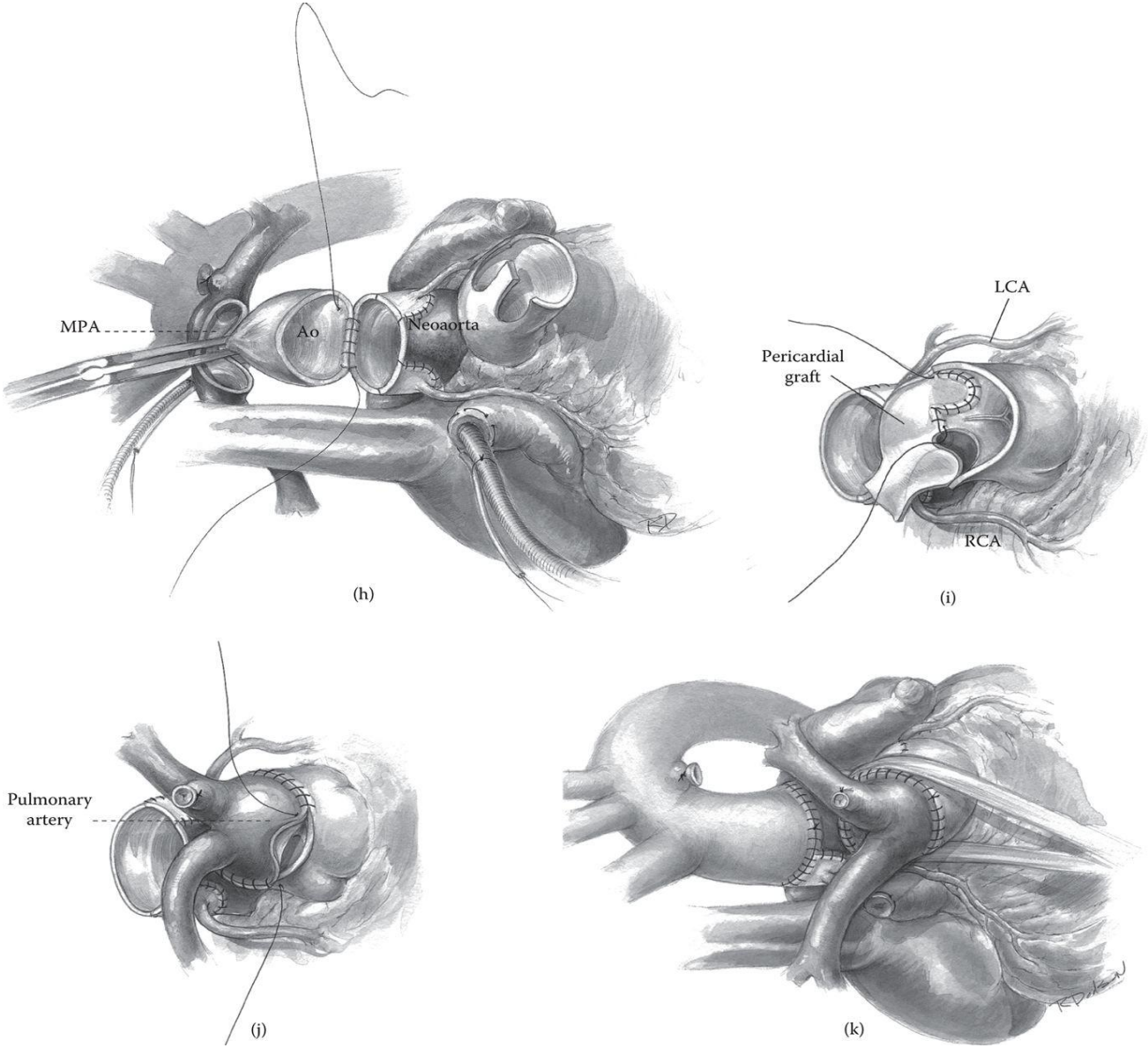
A Meta-Analysis

Sara K. Pasquali, MD; Vic Hasselblad, PhD; Jennifer S. Li, MD; David F. Kong, MD; Stephen P. Sanders, MD

Background—Prior studies of coronary pattern and outcome after arterial switch operation (ASO) for transposition of the great arteries (TGA) have been hindered by limited statistical power. This meta-analysis assesses the effect of coronary anatomy on post-ASO mortality, both overall and adjusted for time.

Methods and Results—A literature search revealed 9 independent series that reported post-ASO mortality by coronary pattern in a total of 1942 patients. Odds ratios comparing all-cause mortality in patients with usual versus variant coronary patterns were calculated and combined by use of an empirical Bayesian model. Single coronary patterns, both of which loop around the great vessels, were associated with significant mortality (OR 2.9, 95% CI 1.3 to 6.8), whereas looping patterns that arose from 2 separate ostia were not (OR 1.2, 95% CI 0.8 to 1.9). This latter group includes patients with the most common variant, circumflex from right coronary artery. Patients with an intramural coronary artery had the greatest mortality (OR 6.5, 95% CI 2.9 to 14.2). Overall, patients with any variant coronary pattern had nearly twice the mortality seen in those with the usual pattern (OR 1.7, 95% CI 1.3 to 2.4). Single ostium patterns and intramural coronary arteries remained associated with significant added mortality after adjustment for time-trend effects.

Conclusions—Over the past 2 decades, patients with common coronary variants have undergone ASO without added mortality compared with those with the usual coronary pattern. Those with intramural or single coronary arteries have significant added mortality that has persisted over time. **(*Circulation*. 2002;106:2575-2580.)**



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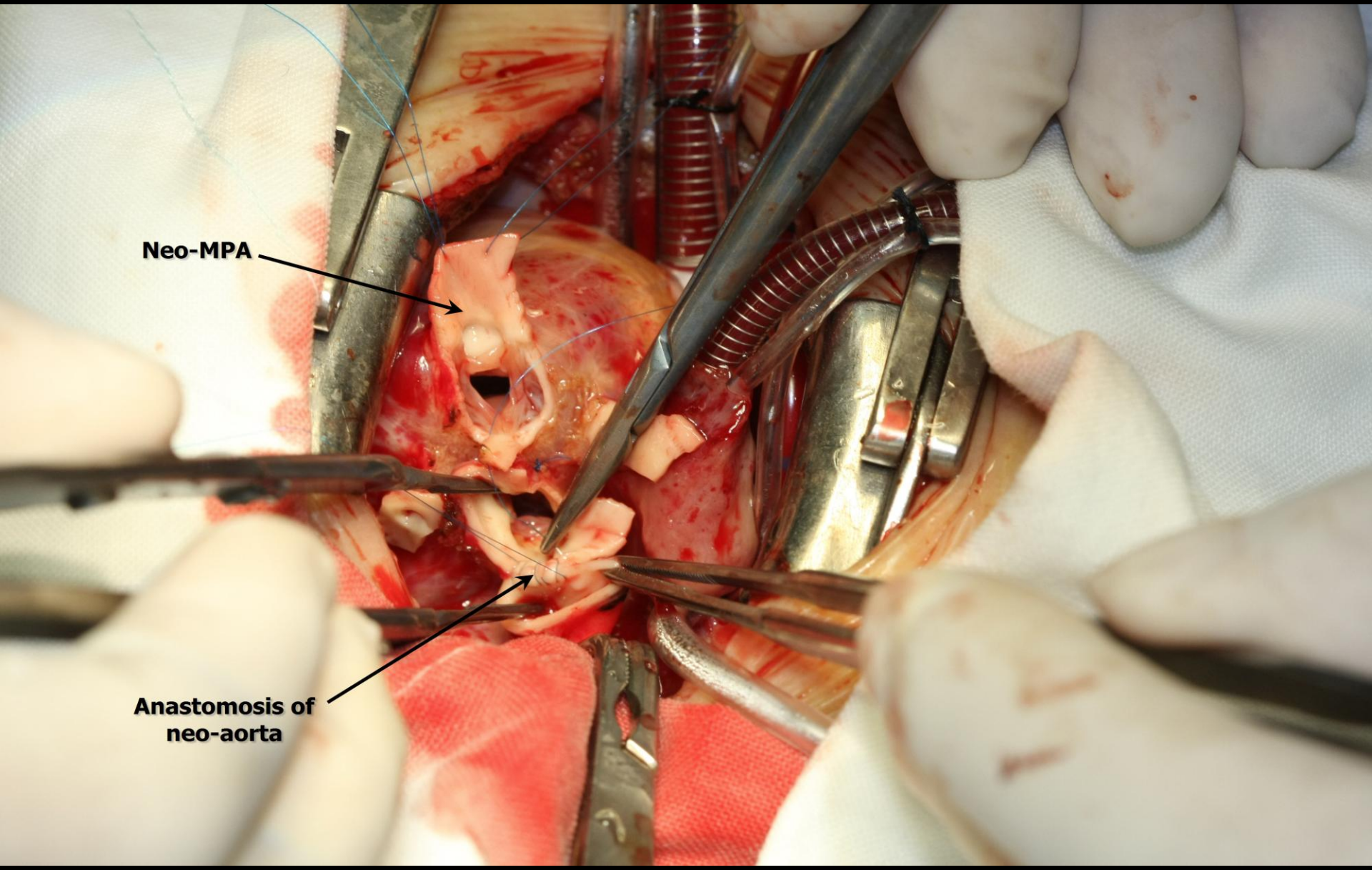
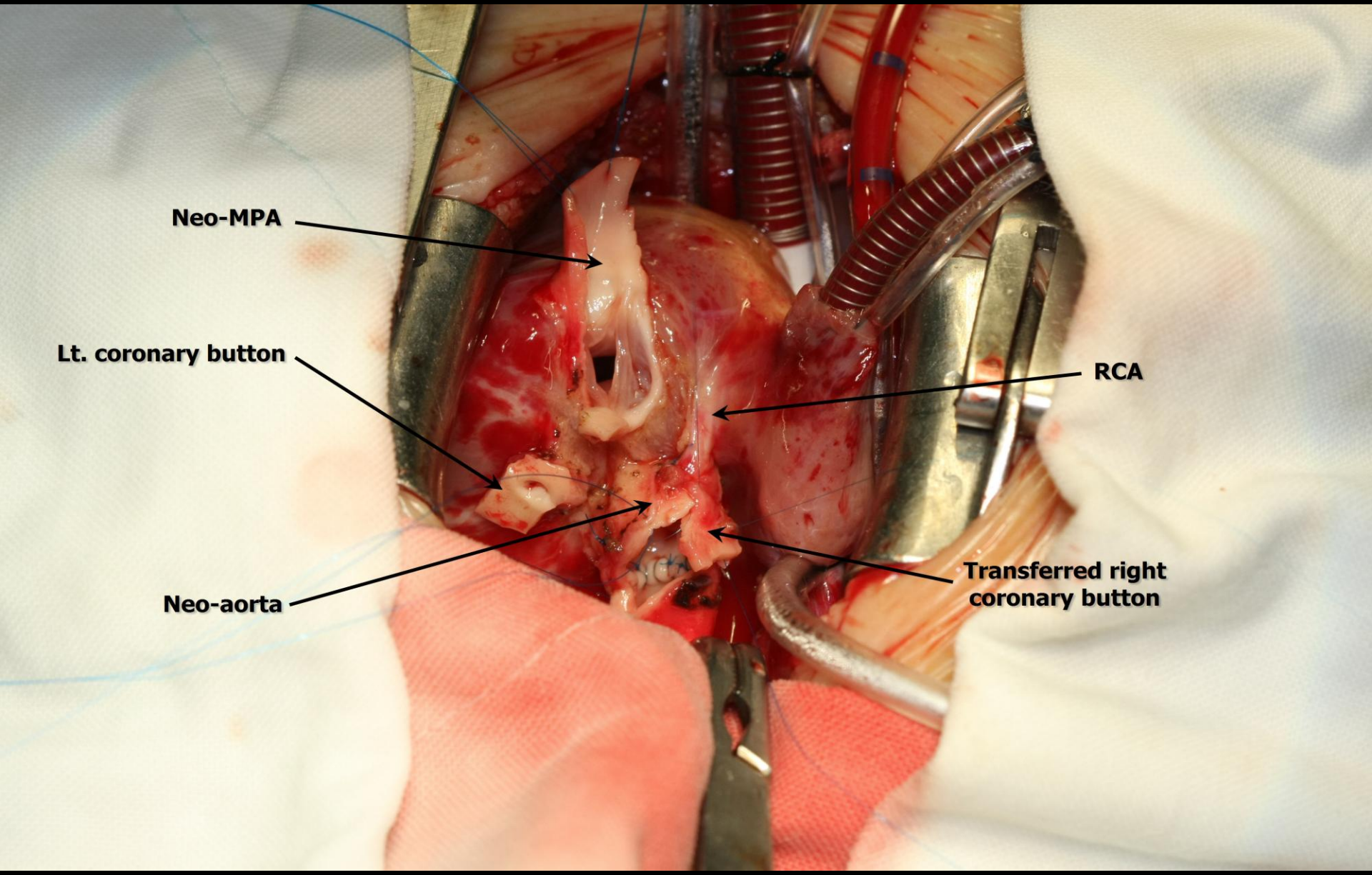
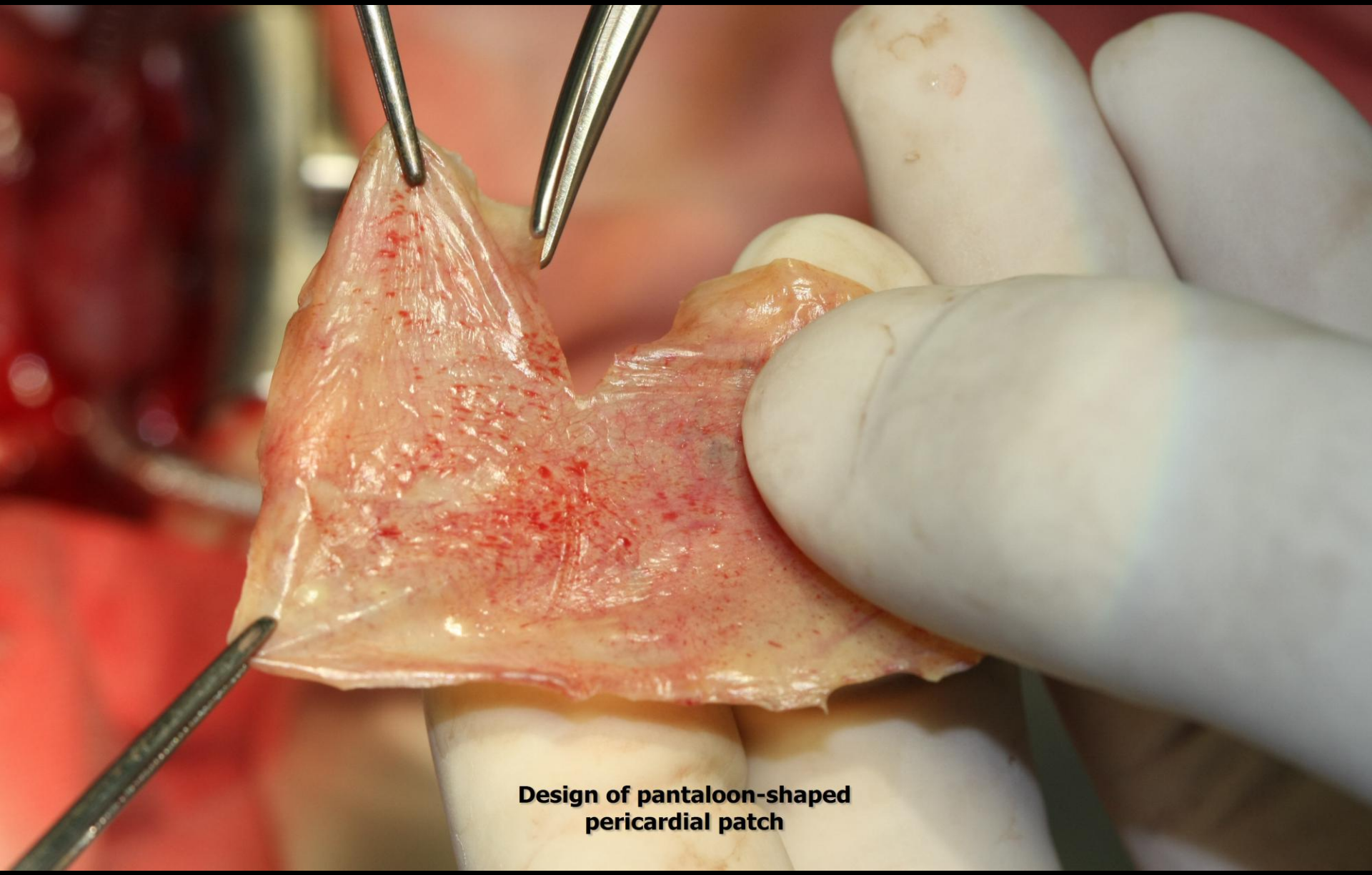
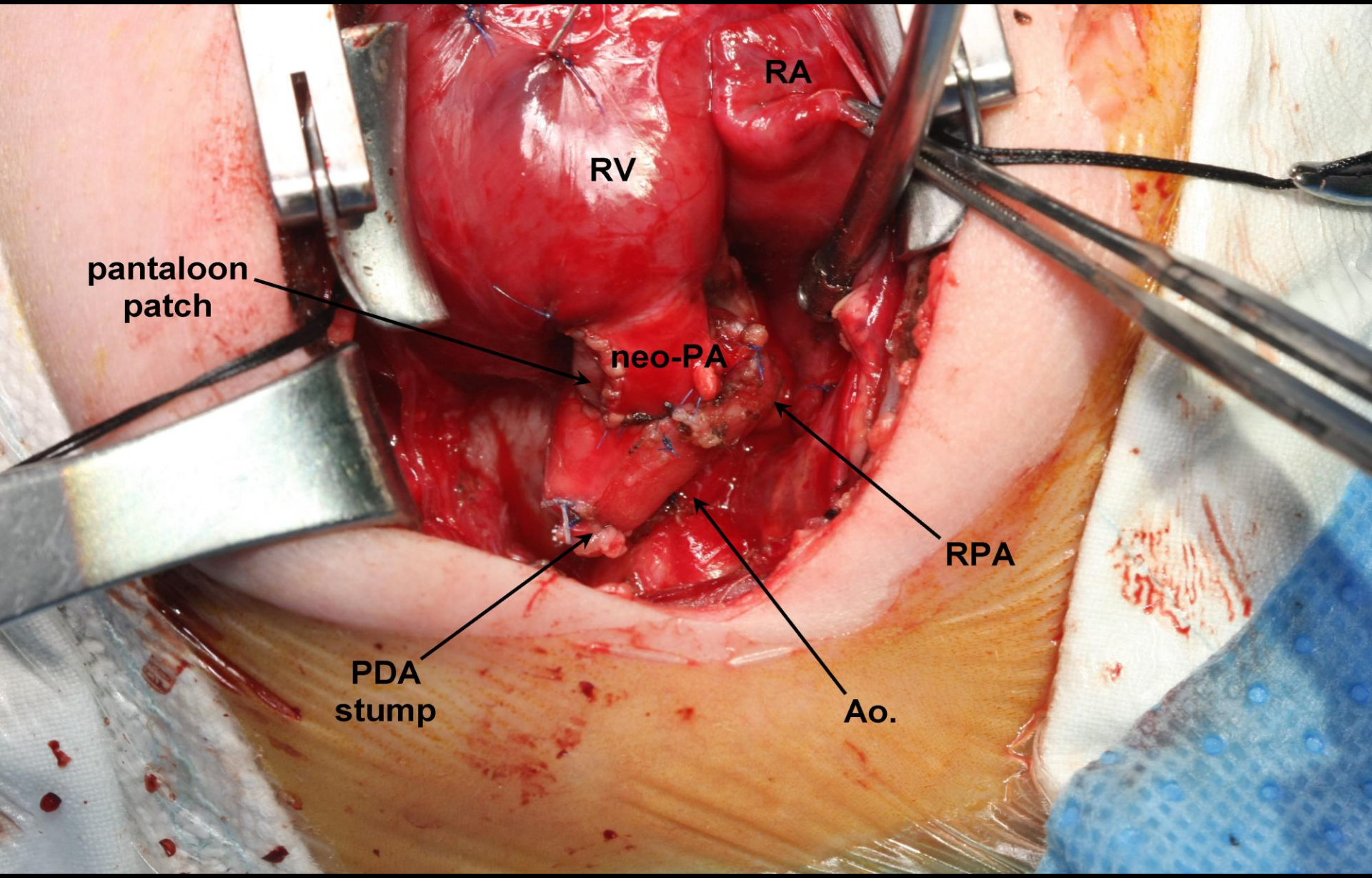

Neo-MPA
Anastomosis of
neo-aorta


Neo-MPA
Lt. coronary button
RCA
Neo-aorta
Transferred right
coronary button


Design of pantaloon-shaped
pericardial patch


RA
RV
pantaloon
patch
neo-PA
RPA
PDA
stump
Ao.

Late Outcome After Arterial Switch Operation for Transposition of the Great Arteries

J. Losay, MD; A. Touchot, MD; A. Serraf, MD; A. Litvinova, MD; V. Lambert, MD; J.D. Piot, MD; F. Lacour-Gayet, MD; A. Capderou, MD, PhD; C. Planche, MD

Background—Early and midterm results of the arterial switch operation (ASO) in transposition of the great arteries (TGA) are good, but late outcome data in large populations are still few.

Methods and Results—Twelve hundred patients had an ASO for TGA between 1982 and 1999, with prospective follow-up of 1095 survivors. Outcome measures included late death, reoperation, aortic insufficiency (AI), pulmonary stenosis (PS), and coronary anomaly. Median follow-up was 4.9 years (range 0.5 to 17 years). Late death occurred in 32 patients; survival was 88% at both 10 and 15 years. The hazard function for death declined rapidly, with no deaths after 5 years. Late mortality was correlated with reintervention and major events in the intensive care unit. Reoperation was performed in 103 patients, more often in complex TGA; the cause was mainly PS. Freedom from reintervention was 82% at 10 and 15 years, with a hazard function that declined rapidly but slowly increased after 3 years. At the last follow-up, PS was present in 3.9% of patients, and grade II or more AI was present in 3.2%, with a cumulative incidence of 9% at 15 years. Among the 278 patients who had a coronary arteriography, 8% had coronary lesions. Normal left ventricle and sinus rhythm were seen in 96.4% and 98.1%, respectively.

Conclusions—Fifteen years after ASO, late mortality was low, with no deaths after 5 years; reoperation, mainly owing to PS, occurred throughout the follow-up. AI and coronary obstruction are rare but warrant further follow-up. Good left ventricular function and sinus rhythm are maintained. **(*Circulation*. 2001;104[suppl I]:I-121-I-126.)**

Reoperative Techniques for Complications After Arterial Switch

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Background: **The purpose of this study is to review our experience with late reoperations after the arterial switch operation (ASO) and to introduce reparative solutions adapted from previous techniques.**

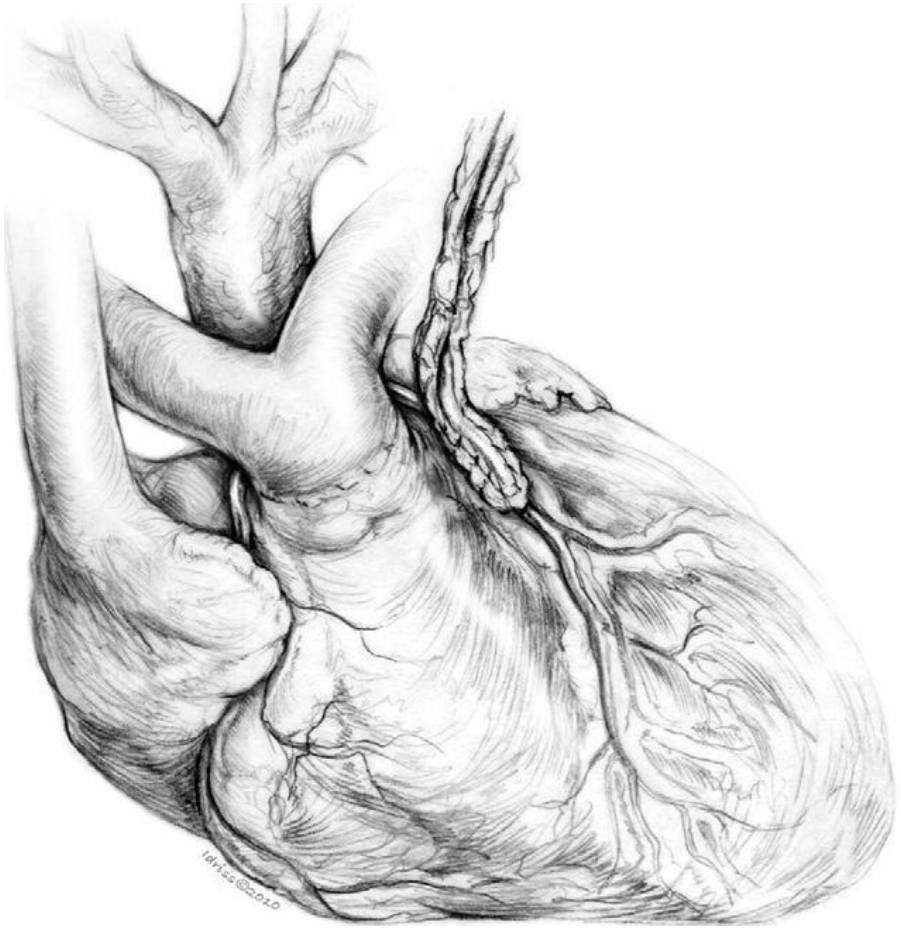
Methods. **A retrospective study was performed on 23 patients who underwent late reoperations after ASO between 1983 and 2010. Eighteen patients were from our concomitantly reported cohort of 258 ASO patients and 5 came from distant referrals.**

Results. **Twenty-seven reoperations on 23 patients were performed for lesions relating to coronary arteries (9 procedures, 7 patients), the neoaortic root (12 procedures, 10 patients), and the right ventricular outflow tract (6 procedures, 6 patients). Four patients died: 1 from an exsanguinating gastric ulcer 4 years after prosthetic valve replacement; 1 from coronary occlusion one month postoperatively from an unroofed intramural left main coronary artery; and 2 after supravalvar pulmonary artery stenosis repair complicated by coexisting left ventricular dysfunction from the original ASO.**

Conclusions. **The ASO remains the treatment of choice for transposition of the great arteries and its variants. While the incidence of late reintervention is low, a subset of patients will require operations that extend the principles of myocardial revascularization, left ventricular outflow tract reconstruction, and relief of pulmonary stenosis.**

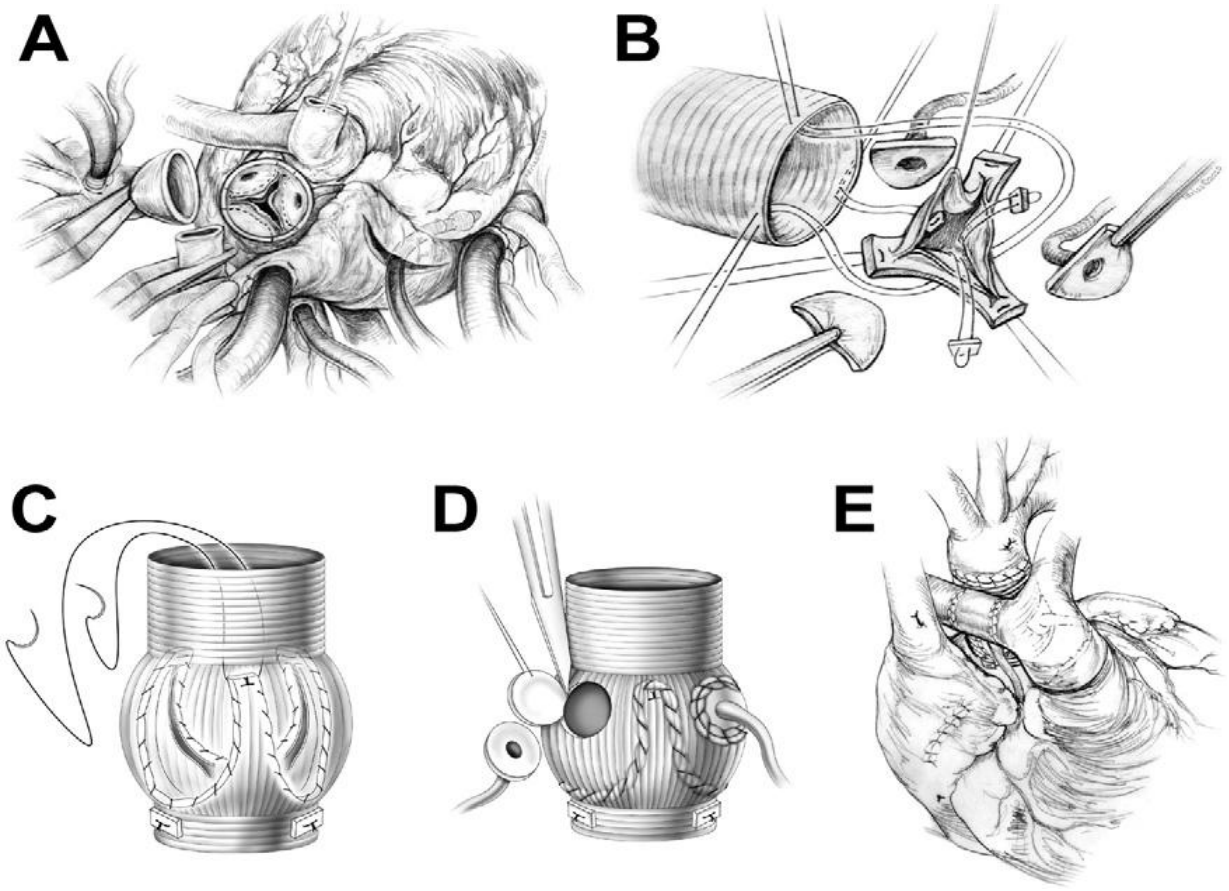
(Ann Thorac Surg 2011;92:1747–55)

CABG



Ann Thorac Surg 2011;92:1747-55

Neoaortic Valve-Sparing Operation



Ann Thorac Surg 2011;92:1747-55

Reoperation After Arterial Switch: A 27-Year Experience

Vijayakumar Raju, MD, Harold M. Burkhart, MD, Lucian A. Durham, III, MD, PhD, Benjamin W. Eidem, MD, Sabrina D. Phillips, MD, Zhuo Li, MS, Hartzell V. Schaff, MD, and Joseph A. Dearani, MD

Divisions of Cardiovascular Surgery, Pediatric Cardiology, Cardiovascular Diseases and Biomedical Statistics and Informatics, Mayo Clinic, Rochester, Minnesota

Background. **The long-term outcome and spectrum of reoperation after the arterial switch operation (ASO) has not been fully defined, and there are limited data in the literature. We reviewed our institutional experience with reoperation(s) after ASO.**

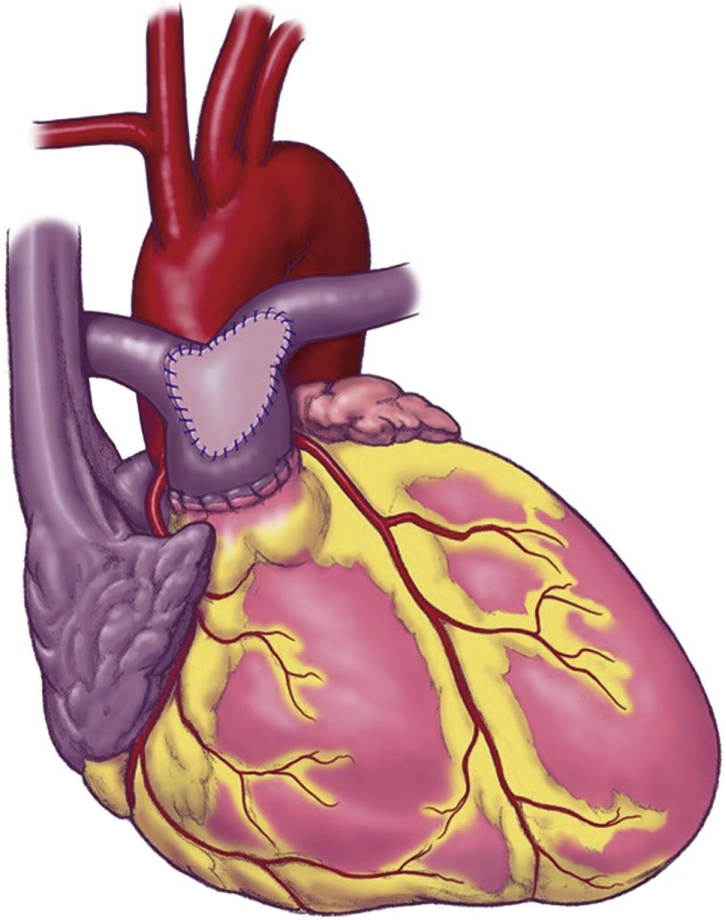
Methods. **Between January 1984 and January 2012, 32 patients (23 male) underwent reoperation(s) after ASO. Anatomy included simple transposition of the great arteries in 14, complex transposition of the great arteries in 14, and Taussig-Bing in 4. Mean age was 6.7 ± 1.4 years at first operation and 10.8 ± 13.4 years at the second operation. Isolated pathology was present in 11 (34.3%) and multiple pathologies in 21 (65.6%). Abnormalities at first reoperation were right-sided pathology in 18 (56.3%), left-sided pathology in 10 (31%), coronary artery in 3 (9%), mitral valve in 3 (9%), residual ventricular septal defect in 4 (12.5%), and recoarctation in 2 (6.3%). It was the second reoperation in 12 and the third reoperation in 3 patients.**

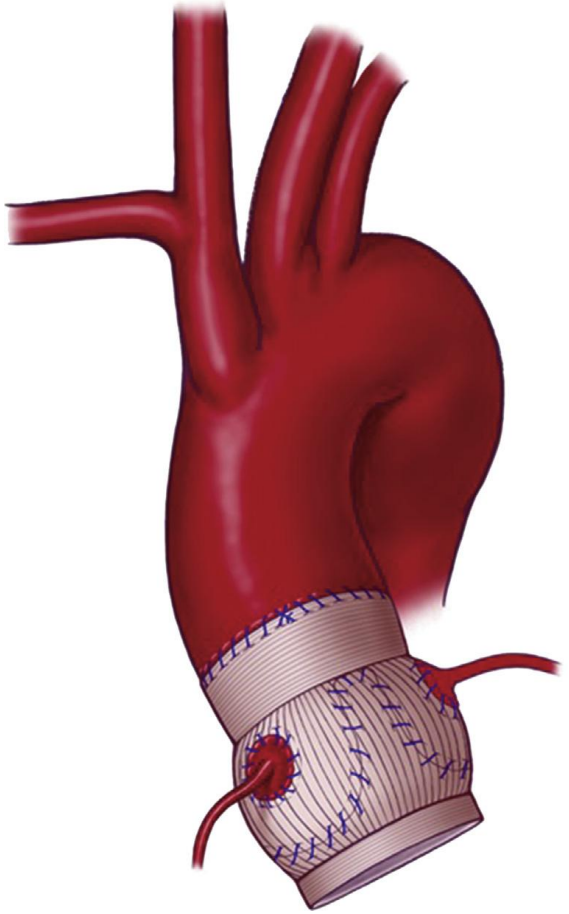
Results. **The first reoperation included pulmonary artery patch plasty in 18, aortic valve operation in 8 (4 valve replacement, 3 root replacement, and 1 repair), pulmonary valve replacement in 4, coronary artery bypass grafting in 3, and mitral valve repair in 3. Multiple reoperations occurred in 15 patients, comprising right-sided procedures (11), left-sided (2), and other (2). Pulmonary artery reconstruction occurred earlier than neoaortic intervention (5.4 ± 6.8 vs 13.8 ± 7.7 years, $p < 0.001$). There were 2 early deaths (6.2%); both patients had complex transposition of the great arteries and both were at early reoperation after ASO. Median follow-up was 14.5 years (maximum, 27 years). There were no late deaths. Freedom from reoperation at 1, 5, and 15 years was 88%, 78%, and 41%, respectively.**

Conclusions. **The most common indication for reoperation after ASO is right-sided pathology, followed by neoaortic root pathology. Late survival after ASO is excellent and risk of late reoperation is low. Life-long medical surveillance is required.**

(Ann Thorac Surg 2013;95:2105–13)

Reoperation After Arterial Switch





Ann Thorac Surg 2013;95:2105-13

CONGENITAL HEART SURGERY:
The Annals of Thoracic Surgery CME Program is located online at http://cme.ctsnetjournals.org. To take the CME activity related to this article, you must have either an STS member or an individual non-member subscription to the journal.

Long-Term Outcomes of the Neoaorta After Arterial Switch Operation for Transposition of the Great Arteries

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Department of Pediatrics, Division of Pediatric Cardiology, and Department of Internal Medicine, Division of Cardiovascular Medicine, and Department of Cardiothoracic Surgery, Medical College of Wisconsin, Milwaukee, Wisconsin

Background. After the arterial switch operation (ASO) for transposition of the great arteries (TGA), the native pulmonary root and valve function in the systemic position, and the long-term risk for neoaortic root dilation and valve regurgitation is currently undefined. The aim of this study was to determine the prevalence and progression of neoaortic root dilation and neoaortic valve regurgitation in patients with TGA repaired with the ASO.

Methods. Measurements of the neoaortic annulus, neoaortic root at the level of the sinuses of Valsalva, and the degree of neoaortic regurgitation were assessed by serial transthoracic echocardiograms on 124 patients with TGA at a median follow-up of 7.2 years (range, 1 to 23 years) after the ASO at our institution.

Results. Neoaortic root dilation with z scores of 2.5 or greater was identified in 66%, and the root diameter z score increased at an average rate of 0.08 per year over time after ASO. Freedom from neoaortic root dilation at 1, 5, 10, and 15 years after ASO was 84%, 67%, 47%, and 32%, respectively. Risk factors for root dilation include history of double-outlet right ventricle ($p = 0.003$), previous pulmonary artery banding ($p = 0.01$), and length of follow-up ($p = 0.04$). Neoaortic valve regurgitation of at least moderate degree was present in 14%. Neoaortic root dilation was a significant risk factor for neoaortic valve regurgitation ($p < 0.0001$). No patient required reintervention on the neoaorta or neoaortic valve during follow-up.

Conclusions. Progressive neoaortic root dilation is common in patients with TGA after the ASO. Continued surveillance of this population is required.

(Ann Thorac Surg 2013;95:1654–9)

TGA with VSD and LVOTO

Goals of surgery

1. **Unobstructed LV outflow tract**
2. **Unobstructed RV outflow tract**
3. **Competent aortic and pulmonary valve**

Surgical options

1. **Arterial switch ± LVOTO relief**
2. **Rastelli procedure**
3. **REV procedure**
4. **Nikaidoh procedure or its variants**
5. **Pulmonary root translocation**

The significance of pulmonary annulus size in the surgical management of transposition of the great arteries with ventricular septal defect and pulmonary stenosis

Chun Soo Park, MD,[a] Dong-Man Seo, MD, PhD,[a] Jeong-Jun Park, MD, PhD,[a] Young Hwue Kim, MD, PhD,[b] and In-Sook Park, MD[b]

Objective: Aortic translocation has received growing attention in the management of complete transposition with ventricular septal defect and pulmonary stenosis, but the criteria regarding pulmonary stenosis for selecting this option have yet to be established. The aim of this study is to evaluate the significance of pulmonary annulus size with the outcome after the arterial switch operation.

Methods: Between November 1996 and September 2008, 250 patients underwent the arterial switch operation for complete transposition. Among them, 8 patients with a pressure gradient greater than 30 mm Hg, bicuspid pulmonary valve, and an aortic Z-score of the pulmonary annulus less than 0 were included in this retrospective study. The median age was 19.1 months (range, 0.5–80.0 months). The median follow-up was 39.7 months (range 9.1–139.5 months).

Results: At latest follow-up, the Z-score of the neoaortic annulus increased from –1.50 ± 1.13 (range, –3.42 to –0.35) to 1.10 ± 1.15 (range, –0.8 to 2.10) ($P < .01$). No patient had a significant pressure gradient across the left ventricular outflow tract. There was 1 early death and there were no late deaths. Two reoperations were performed in 1 patient for neoaortic stenosis at 81 months and 110 months after the operation. Latest echocardiogram revealed grade 0 or 1 neoaortic insufficiency.

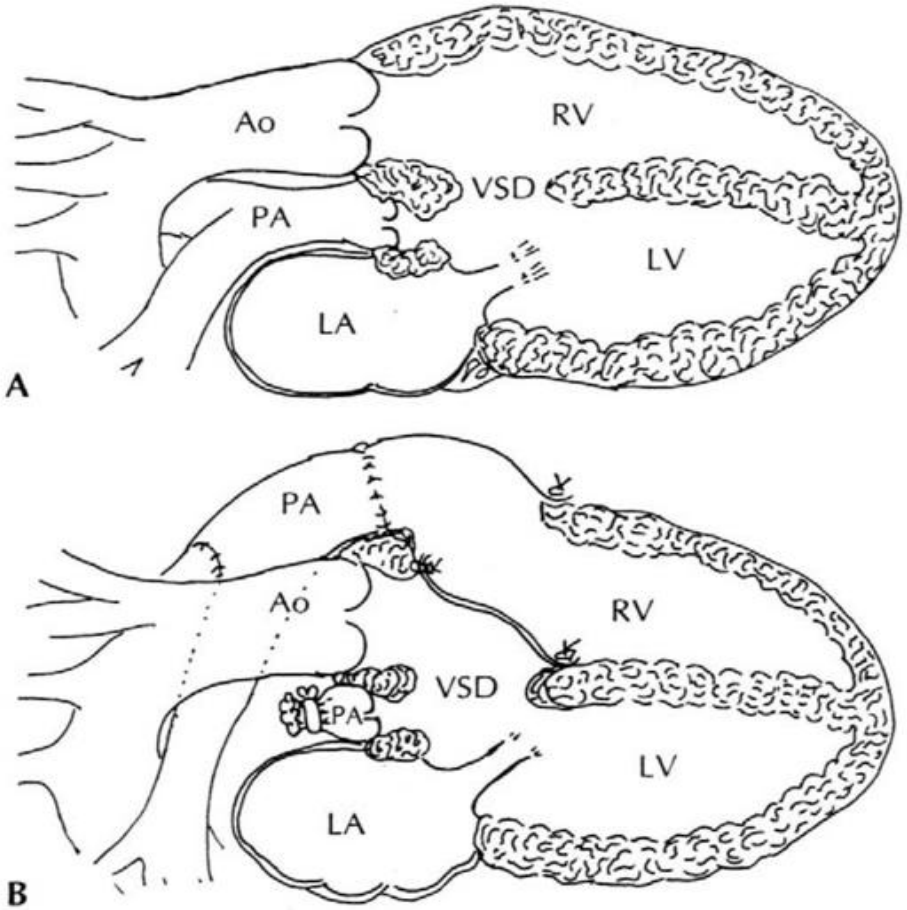
Conclusion: It was possible to extend the indication for the arterial switch operation with acceptable outcome to the patient with a Z-score of about –3 of the pulmonary annulus despite bicuspid pulmonary valve. Inasmuch as the arterial switch operation has benefits over the other options, a large-scale study is required for more reasonable triage in this group of patients. (J Thorac Cardiovasc Surg 2010;139:135-8)

Rastelli Procedure

- LV to aorta baffling ± VSD enlargement
- RV to pulmonary artery (valved) conduit

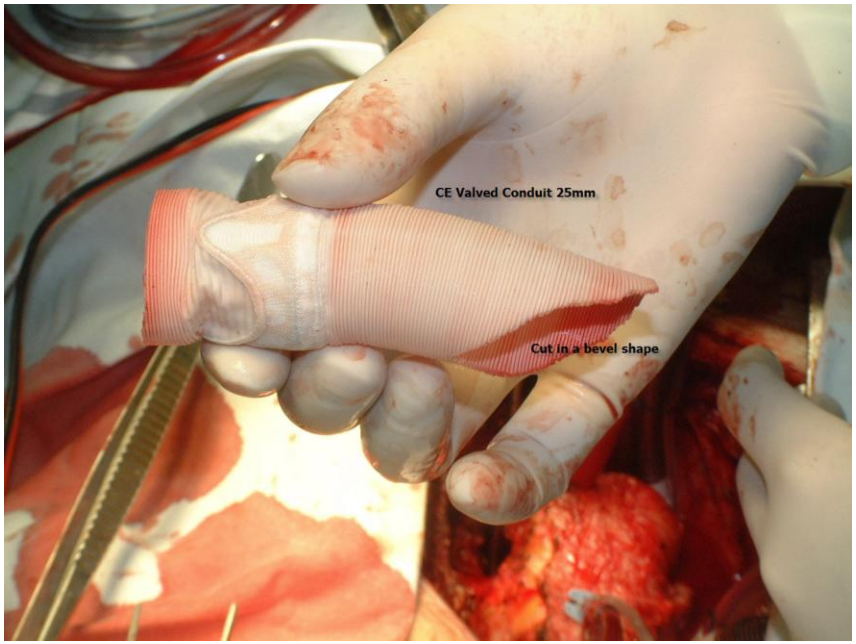
- Poor long-term survival reported
- LVOTO, RVOTO, arrhythmia, sudden death

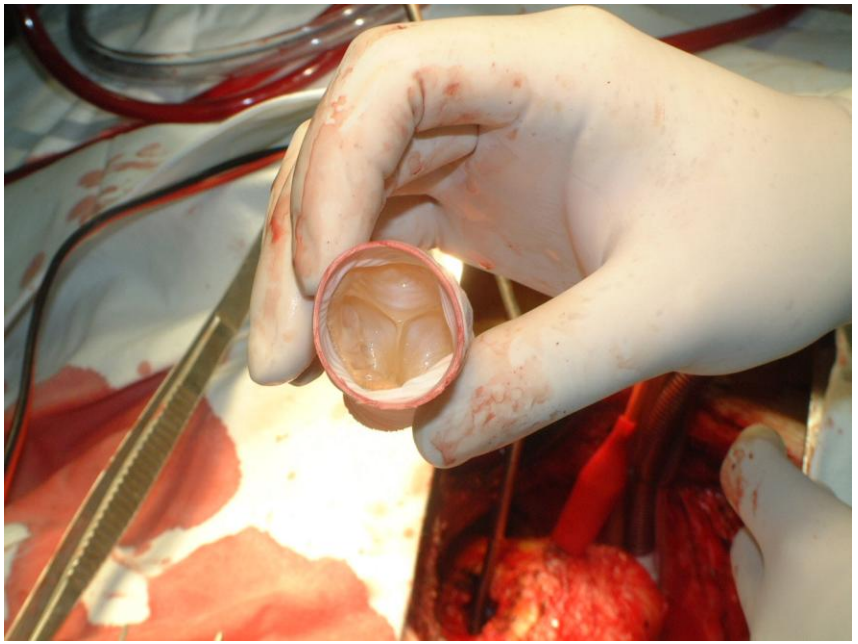
Rastelli Procedure



Ann Thorac Surg 2005;79:2089-93

RV-PA Conduit





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SURGERY FOR CONGENITAL HEART DISEASE

TWENTY-FIVE–YEAR EXPERIENCE WITH RASTELLI REPAIR FOR TRANSPOSITION OF THE GREAT ARTERIES

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Guido Oppido, MD
Jacqueline Kreutzer, MD
Kimberlee Gauvreau, ScD
Michael Freed, MD
John E. Mayer, Jr, MD
Richard Jonas, MD
Pedro J. del Nido, MD

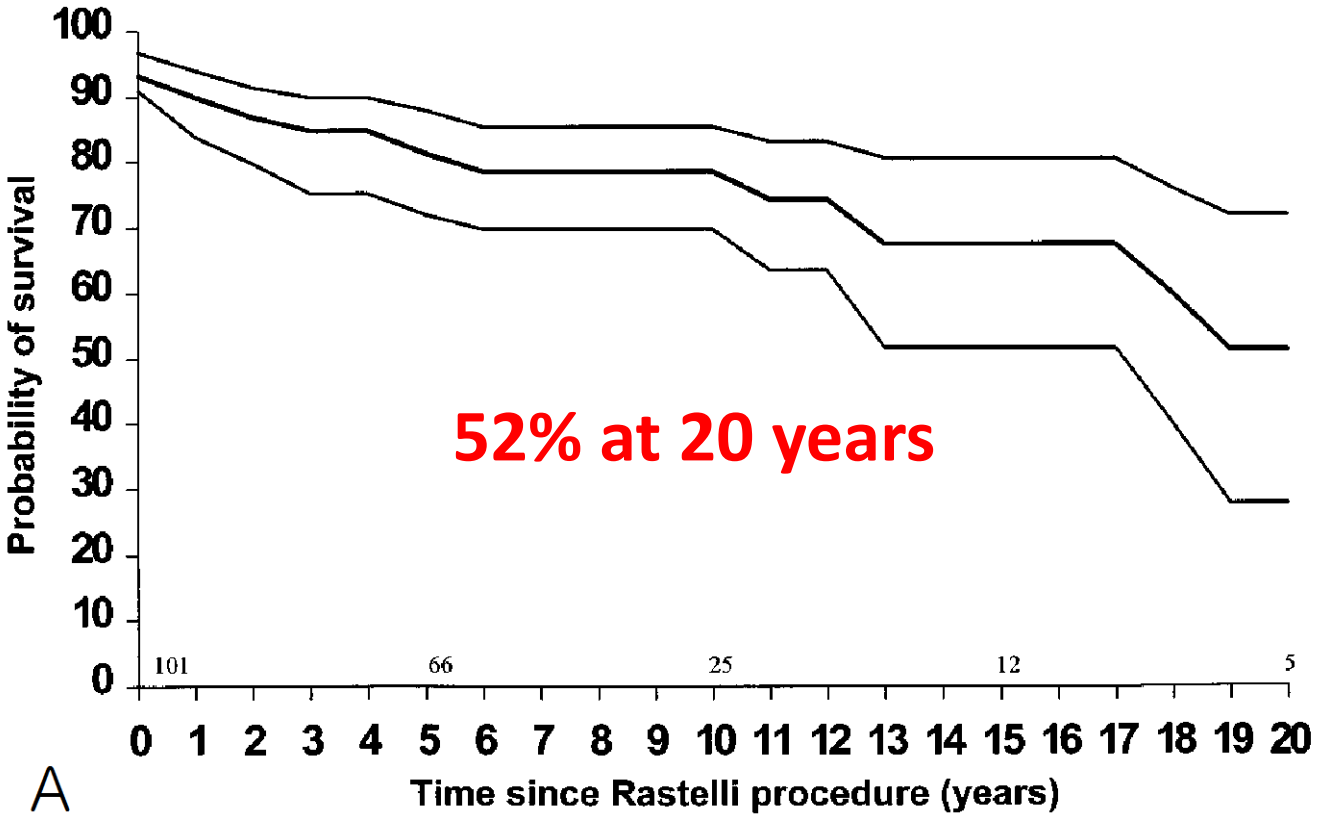
Objective: Our purpose was to describe the outcome of the Rastelli repair in D-transposition of the great arteries and to determine the risk factors associated with unfavorable events.

Methods: From March 1973 to April 1998, 101 patients with D-transposition of the great arteries and ventricular septal defect underwent a Rastelli type of repair. Median age and weight were 3.1 years (10th to 90th percentiles 0.3-9.9 years) and 12.8 kg (5.9-28.2). Pulmonary stenosis was present in 73 patients and pulmonary atresia in 18; 10 patients had no left ventricular outflow tract obstruction.

Results: There were 7 early deaths (7%) and no operative deaths in the last 7 years of the study. Risk factors for early death, by univariable analysis, included straddling tricuspid valve ($P = .04$) and longer aortic crossclamping times ($P = .04$). At a median follow-up of 8.5 years, there were 17 late deaths and 1 patient had undergone heart transplantation. Forty-four patients had

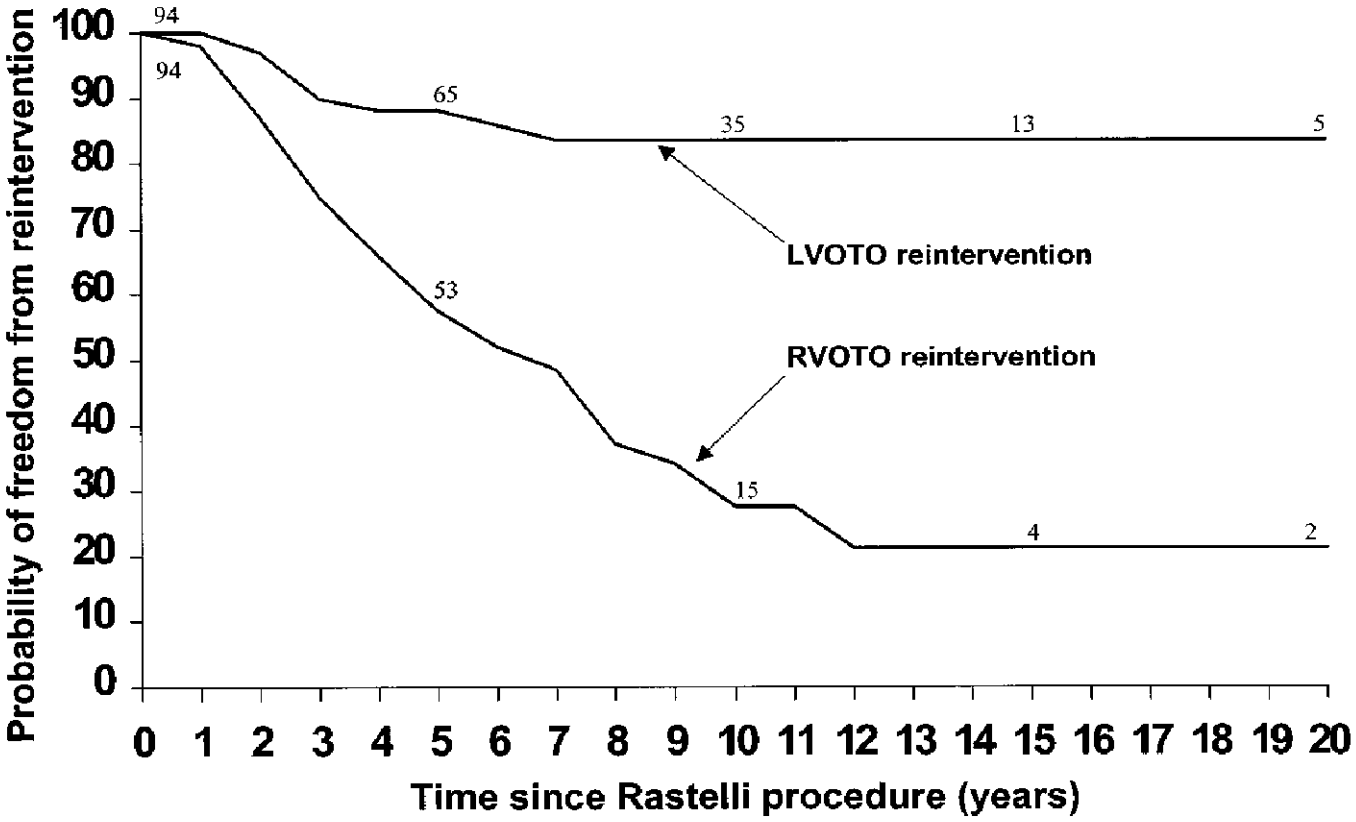
J Thorac Cardiovasc Surg 2000;120:211-23

Survival After Rastelli Procedure
Boston Experience



J Thorac Cardiovasc Surg 2000;120:211-23

Reoperation After Rastelli Procedure Boston Experience



J Thorac Cardiovasc Surg 2000;120:211-23

The Rastelli Procedure for Transposition of the Great Arteries: Resection of the Infundibular Septum Diminishes Recurrent Left Ventricular Outflow Tract Obstruction Risk

Bahaaldin Alsoufi, MD, Abid Awan, MD, Ahmad Al-Omrani, MD, Mamdouh Al-Ahmadi, MD, Charles C. Canver, MD, Ziad Bulbul, MD, Avedis Kalloghlian, MD, and Zohair Al-Halees, MD

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Background. **The Rastelli procedure is the standard surgical treatment of d-transposition of great arteries (d-TGA), ventricular septal defect (VSD), and pulmonary stenosis. Late morbidity is significant due to recurrent left ventricular outflow obstruction (LVOTO), early conduit obstruction, and arrhythmias, with troublesome late mortality. To avoid recurrent LVOTO, we routinely enlarge the VSD and resect the infundibular septum before LV baffling to the aorta. We examined the efficacy of this approach in mitigating recurrent LVOTO risk.**

Methods. **Late echocardiographic and time-related clinical results of patients undergoing the Rastelli procedure were examined. Demographics and operative variables affecting outcomes were analyzed.**

Results. **The Rastelli cohort comprised 36 patients with d-TGA, VSD, and pulmonary stenosis. Median age at operation was 2.4 years (range, 0.3 to 8.3 years). Pulmonary stenosis was present in 31 and atresia in 5. Twenty-two patients had undergone a previous aortopulmonary shunt, and 6 had an atrial septectomy. No operative or late deaths occurred. Time-related freedom from permanent pacemaker implantation, recurrent LVOTO on echocardiogram, and conduit replacement at 10 years was 82%, 100%, and 49%, respectively. Systolic function was normal in all but 3 patients and 92% were in New York Heart Association functional class I and II. None of the patients had late arrhythmias or required heart transplantation.**

Conclusions. **Early and midterm survival after the Rastelli procedure is satisfactory. Aggressive resection of the infundibular septum to enlarge the VSD has mitigated the risk of LVOTO recurrence. Late conduit obstruction remains an important source of morbidity and frequently requires reintervention.**

(Ann Thorac Surg 2009;88:137–43)

Rastelli Operation for Transposition of the Great Arteries With Ventricular Septal Defect and Pulmonary Stenosis

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Background. The optimal surgical treatment of patients with transposition of the great arteries, ventricular septal defect, and pulmonary stenosis is controversial. Although the Rastelli operation has been standard surgical management of this lesion, aortic root translocation with right ventricular outflow tract (RVOT) reconstruction (Nikaidoh) and the pulmonary artery translocation (Lecompte) or REV (réparation a l'étage ventriculaire) are surgical alternatives more recently introduced to treat this complex lesion. This report reviews our 20-year experience with the Rastelli procedure and attempts to compare our outcomes with those recently published using the Nikaidoh and REV procedures.

Methods. Between 1988 and 2008, 40 patients (median age, 4 years; range, 9 months to 17 years) underwent Rastelli operation at our institutions. The RVOT was obstructed in 32 and atretic in 8. Follow-up was available for all but one patient (mean follow-up, 8.6 ± 5.6 years). The RVOT was reconstructed with homograft (n = 25), bovine jugular vein (n = 8), nonvalved Dacron tube (n = 5), or a porcine valved conduit (n = 2). Two patients required a pacemaker.

Results. There were no early, but three late deaths and one heart transplantation 12 years postoperative the Rastelli operation. Kaplan-Meier survival was 93% at 5, 10, and 20 years. Univariate risk factors for death or transplantation included surgery before 1998 ($p = 0.03$) and concomitant noncardiac anomalies ($p = 0.001$). Sixteen patients (40%) had reoperation for right ventricular-pulmonary artery conduit stenosis (mean, 7.8 ± 3.8 years) without mortality. Freedom from conduit replacement was 86%, 74%, 63%, and 59% at 5, 10, 15, and 20 years, respectively. Multivariate analysis revealed that the risk factors of conduit replacement were younger age at operation ($p = 0.001$) and surgery before 1998 ($p < 0.001$). Two patients (5%) required reoperation for left ventricular outflow tract obstruction. At follow-up, there were no sudden unexplained deaths, and New York Heart Association functional class is I or II.

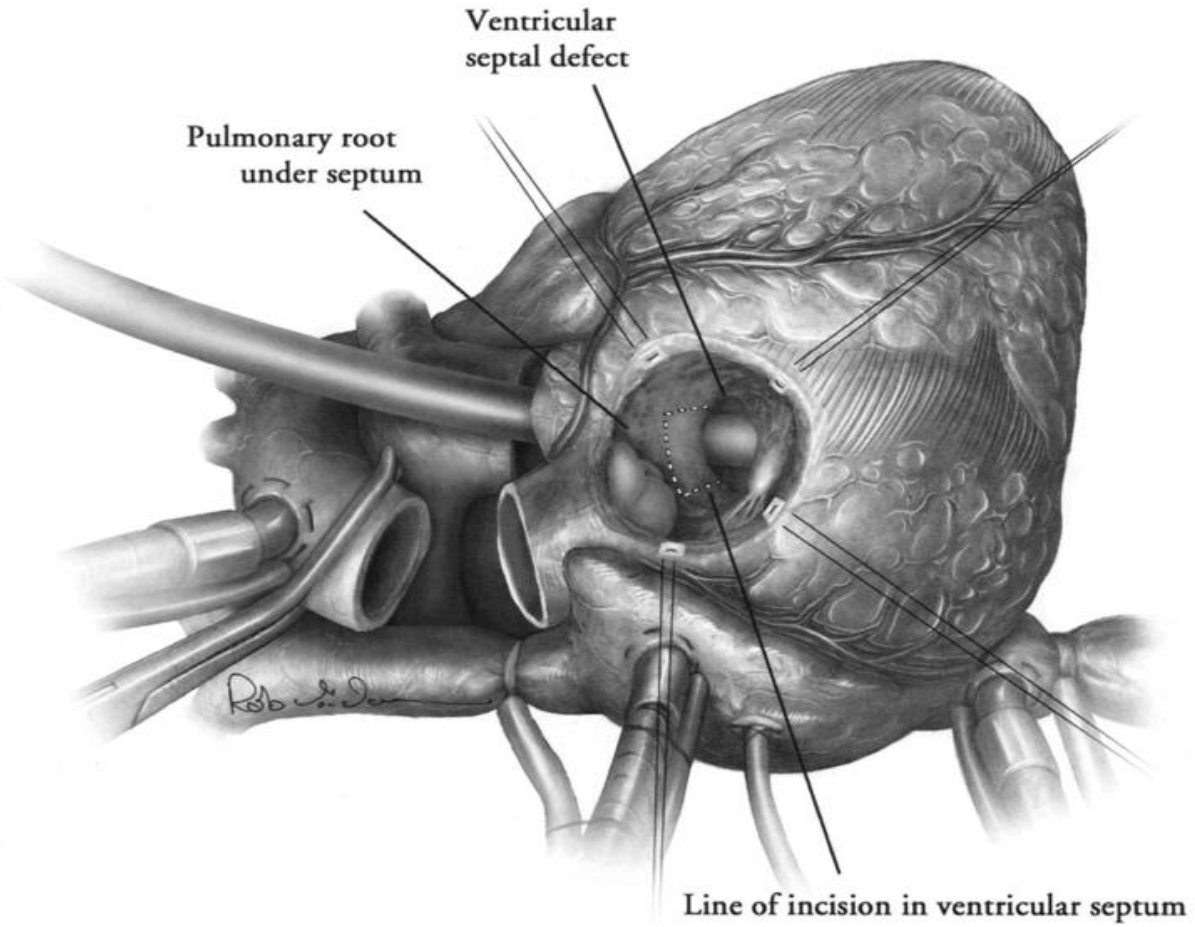
Conclusions. The Rastelli procedure is a low-risk operation with regard to early and late mortality and reoperation for left ventricular outflow tract obstruction. Conduit change operations will be required in most patients regardless of the technique of repair, but currently can be performed with low morbidity and mortality. These midterm outcomes after the Rastelli operation should serve as a basis for comparison with surgical alternatives more recently introduced for transposition of the great arteries and ventricular septal defect with RVOT obstruction.

(Ann Thorac Surg 2011;91:188–94)

REV Procedure

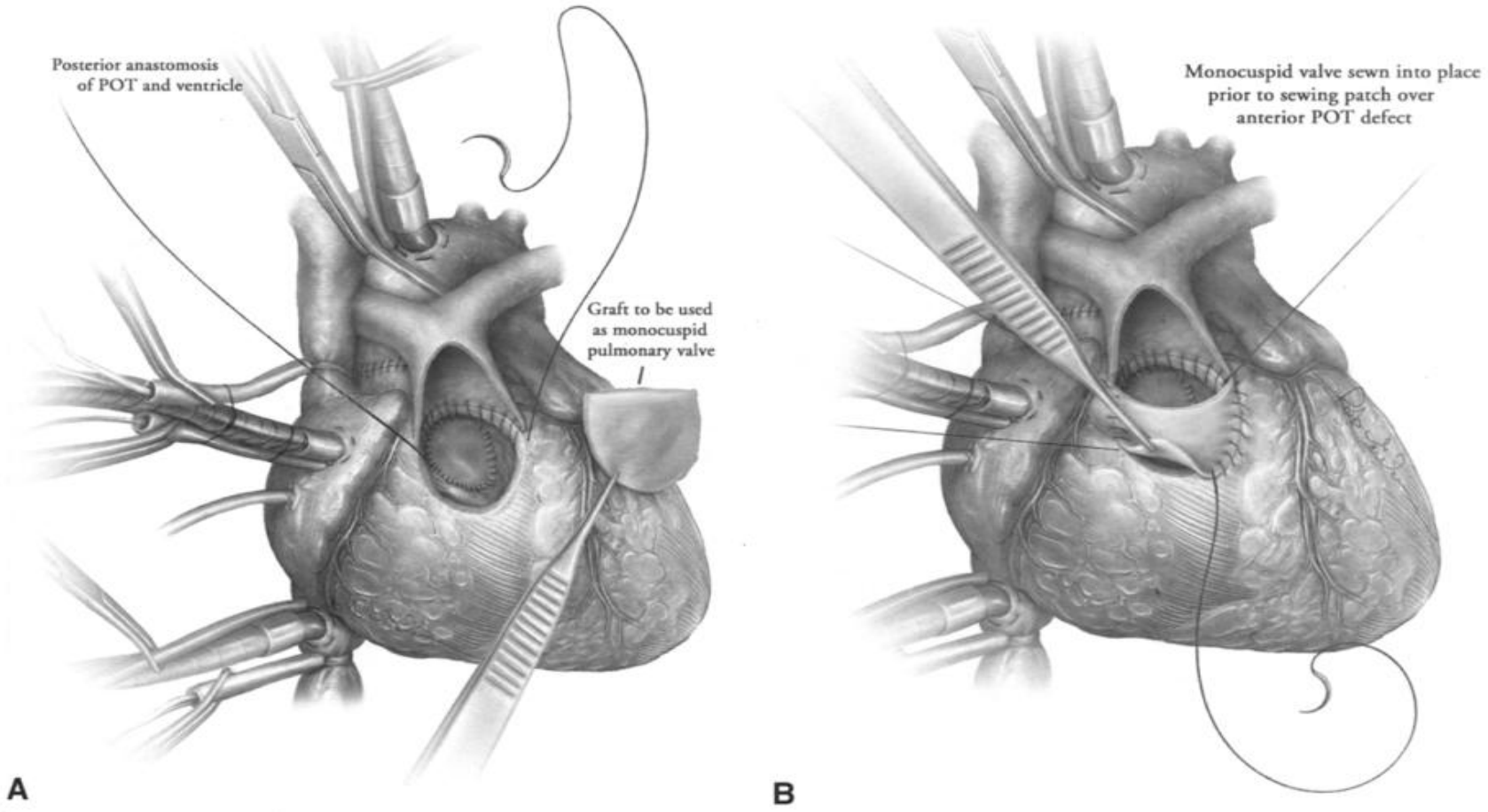
- **Aggressive VSD enlargement to prevent LVOTO**
- **Avoidance of the use a RV to PA conduit**

REV Procedure (VSD Enlargement)



Oper Tech Thorac Cardiovasc Surg 2003;8:150-9

REV Procedure (RVOT Reconstruction)



Oper Tech Thorac Cardiovasc Surg 2003;8:150-9

doi:10.1510/mmcts.2007.003046

MULTIMEDIA MANUAL OF CARDIOTHORACIC SURGERY
MMCTS

Surgery for malposition of the great arteries: the REV procedure

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Modified Lecompte Procedure for the Anomalies of Ventriculoarterial Connection

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Background. The Lecompte procedure for correcting transposition of the great arteries has an advantage because it obviates the need for an extracardiac conduit for the reconstruction of the pulmonary outflow tract. In this study, we evaluated the effectiveness and the application of the Lecompte procedure based on our experiences.

Methods. A retrospective review was conducted of the records of 45 patients who underwent the Lecompte procedure during the past 11 years to achieve direct right ventricle to pulmonary artery continuity. Mean age at operation was 2.4 ± 1.7 years (range 3.5 months to 6.9 years). The diagnoses involved anomalies of the ventriculoarterial connection with ventricular septal defect and pulmonary outflow tract obstruction, such as transposition of the great arteries, double-outlet right ventricle, and double-outlet left ventricle.

Results. Early mortality was 4.4% (2 of 45 patients) and late mortality was 4.7% (2 of 43). The mean follow-up was 4.9 ± 3.1 years. Fourteen patients (34.1% of survivors, n = 41) had pulmonary stenosis (pressure gradient above 30 mm Hg), the main reason for which was a calcified monocusp valve (n = 10, 71.4%). Eight of 45 patients (17.8%) underwent reoperation: 2 for residual ventricular septal defect, 1 for recurrent septic vegetation, and 5 for pulmonary stenosis. The cumulative survival rates were 91.1% ± 4.2% at 10 years. The actuarial probabilities of freedom from reoperation for pulmonary stenosis were 93.8% ± 4.3% and 71.4% ± 11.8% at 5 and 10 years, respectively.

Conclusions. Our review suggests that the Lecompte procedure is an effective treatment modality for anomalies of the ventriculoarterial connection with ventricular septal defect and pulmonary outflow tract obstruction. Repair in early age is possible with acceptable morbidity and mortality, but recurrent right ventricular outflow tract obstruction caused by degeneration of the monocusp valve is a problem that needs resolution.

(Ann Thorac Surg 2001;72:176–81)

Twenty Five Years Experience of Modified Lecompte Procedure for the Anomalies of Ventriculoarterial Connection with Ventricular Septal Defect and Pulmonary Stenosis

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Seoul National University Hospital, Seoul, Republic of Korea

Objective: To overcome drawbacks of the Rastelli operation, the modified Lecompte procedure create a connection from the left ventricle to the aorta free from residual obstruction due to the resection of the outlet septum and avoid the implantation of an extracardiac valved conduit. We evaluated the effectiveness of this innovative technique with analysis of our 25 years long-term results.

Methods: We reviewed the records of 50 patients who underwent the modified Lecompte procedure during the past 25 years. The median age at operation was 1.95 years (range: 0.30-12.48 years). The diagnoses involved anomalies of the ventriculoarterial connection with ventricular septal defect and pulmonary outflow tract obstruction, such as transposition of the great arteries, double outlet right ventricle, and double outlet left ventricle.

Results: There were two early deaths (4.0%). During mean follow-up of 14.2±7.9 years (range: 0.2-25 years), there were four late deaths (8.3%). Actuarial survival rate was 87.3±4.9% at 25 years. The freedom from arrhythmia and reoperation were 88.0±6.0%, and 25.2±9.4% at 25 years. The freedom from reoperation for left ventricular outflow tract obstruction and right ventricular outflow tract obstruction were 88.5±5.4%, and 49.6±9.0% at 25 years. At last follow up, 43 survivors (97.7%) are in NYHA class I.

Conclusions: The modified Lecompte procedure has an excellent longterm results for treating anomalies of ventriculoarterial connection with ventricular septal defect and pulmonary outflow tract obstruction. Early repair is possible with low mortality and morbidity in terms of arrhythmia, reoperation for right or left ventricular outflow tract obstruction, and functional class.

Nikaidoh Procedure

- Aortic translocation
- Half-turned truncal switch
- Double root translocation
- Ross-Switch-Konno

J Thorac Cardiovasc Surg. **1984** Sep;**88**(3):**365**-72.

Aortic translocation and biventricular outflow tract reconstruction. A new surgical repair for transposition of the great arteries associated with ventricular septal defect and pulmonary stenosis.

Nikaidoh H.

Abstract

Transposition of the great arteries associated with ventricular septal defect and left ventricular outflow tract obstruction remains a serious surgical challenge. A new surgical technique that utilized aortic root mobilization and transfer combined with reconstruction of the right and left ventricular outflow tracts was applied successfully in two children. This technique provides an additional type of arterial switch operation for complex transposition and is an alternative to (1) the conventional combination of Mustard (Senning) procedure with closure of the ventricular septal defect and relief of left ventricular outflow tract obstruction and (2) the Rastelli operation.

The Role of Aortic Translocation in the Management of Complex Transposition of the Great Arteries

Victor O. Morell, Jeffrey P. Jacobs, and James A. Quintessenza

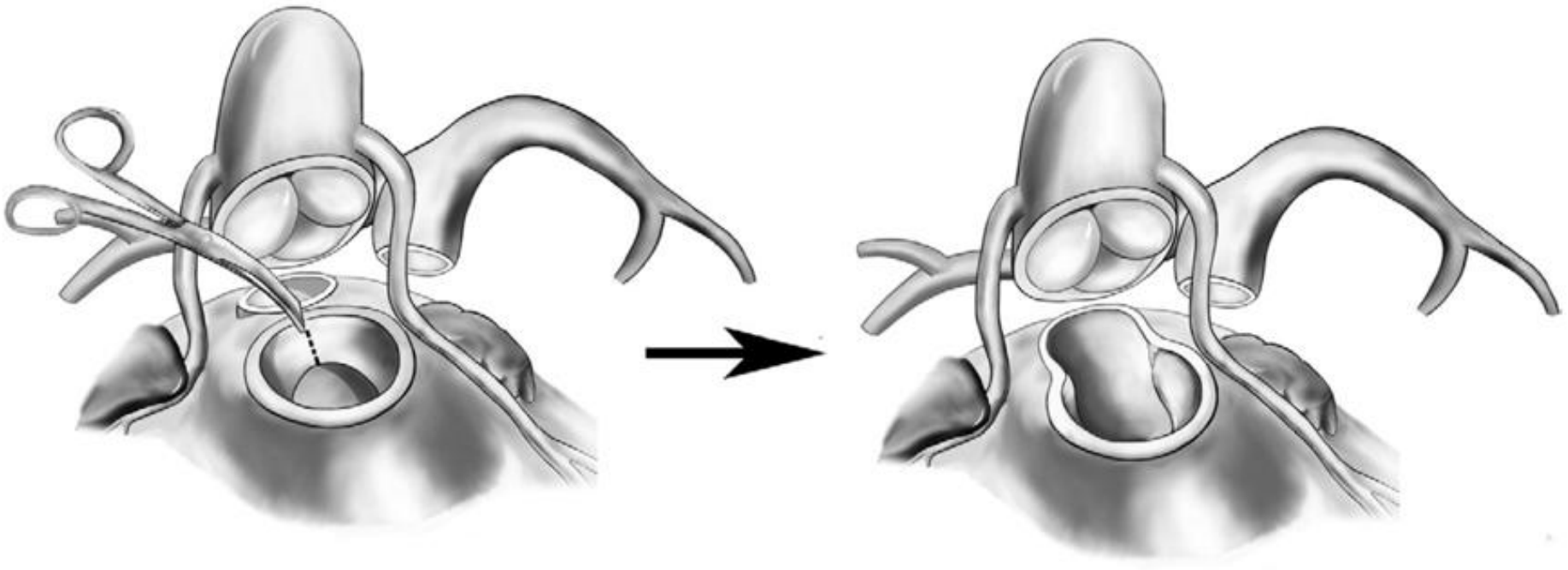
The surgical management of transposition of the great arteries with a ventricular septal defect and left ventricular outflow tract obstruction still remains a significant challenge despite major advances in pediatric cardiovascular surgery. Almost four decades after its inception, the Rastelli procedure continues to be the standard surgical technique used in the management of these patients, despite less than optimal long-term results. Aortic translocation with biventricular outflow tract reconstruction is an alternative surgical technique that creates anatomically sound connections between the ventricles and their respective great arteries, which could result in better long-term outcomes. This technique is especially useful in patients with transposition of the great arteries/ventricular septal defect/left ventricular outflow tract obstruction who have an inlet or restrictive ventricular septal defect, a small right ventricle, a straddling atrioventricular valve, or an aberrant coronary artery crossing the right ventricular outflow tract.

Pediatr Card Surg Annu 2004;7:80-4

Nikaidoh Procedure and Its Variants

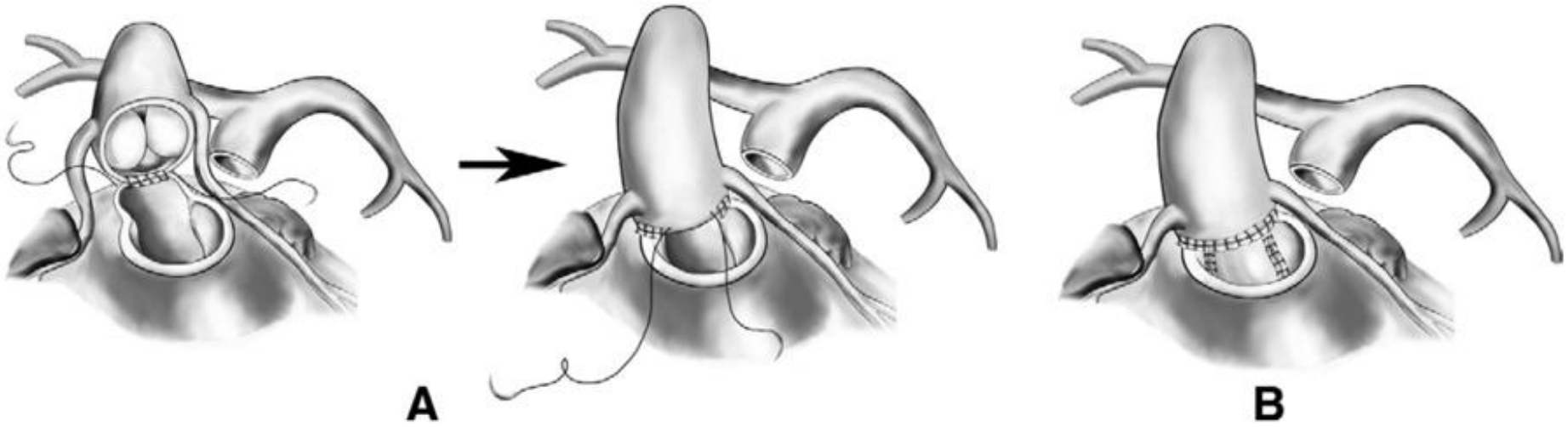
- Harvest of the aortic root from the RV
- Division of outlet septum
- Posterior translocation of the aortic root
- Reconstruction of LVOT
- Reconstruction of RVOT

Aortic Translocation
Harvest of Aortic Root and Division of Outlet Septum



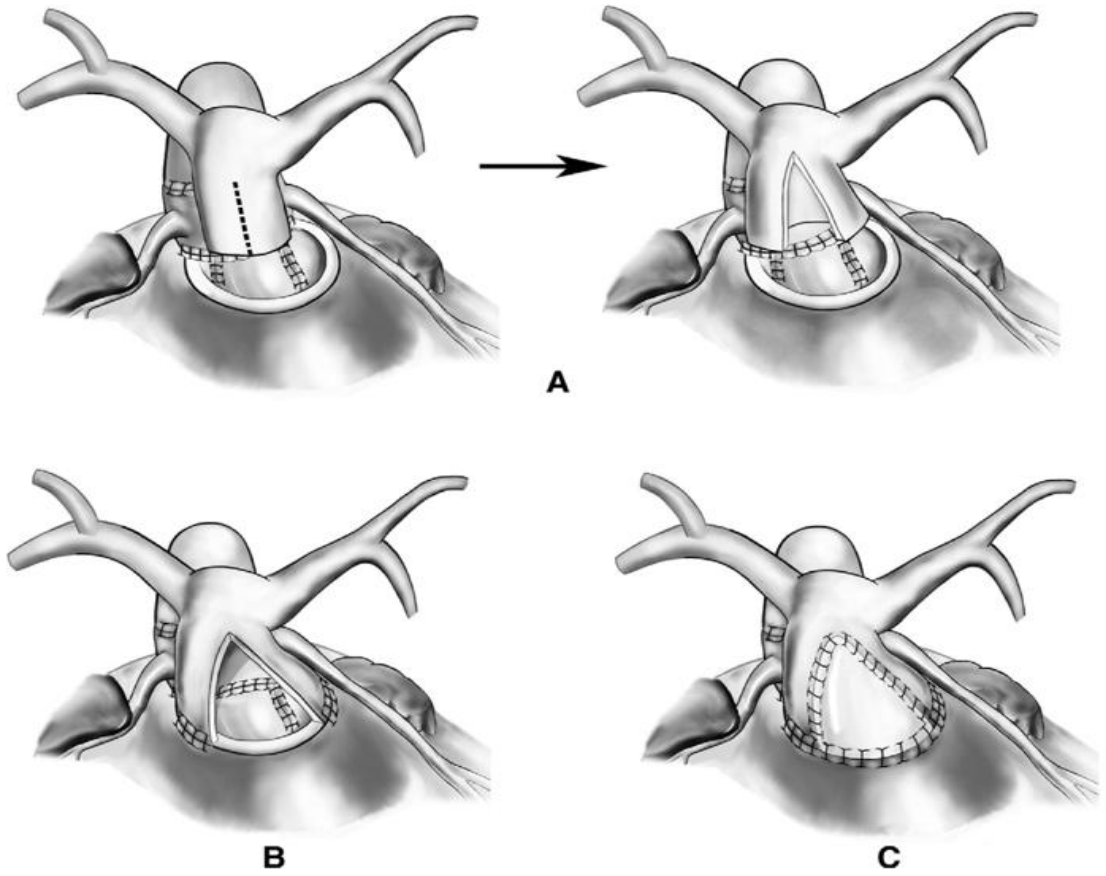
Oper Tech Thorac Cardiovasc Surg 2008;13:181-7

Aortic Translocation
Posterior Translocation of the Aortic Root and Reconstruction of LVOT



Oper Tech Thorac Cardiovasc Surg 2008;13:181-7

Aortic Translocation
Reconstruction of RVOT



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Aortic Root Translocation Plus Arterial Switch for Transposition of the Great Arteries With Left Ventricular Outflow Tract Obstruction

Intermediate-Term Results

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Objectives The goal of our study was to report our intermediate-term results with aortic root translocation plus arterial switch for d-transposition of the great arteries with left ventricular outflow tract obstruction.

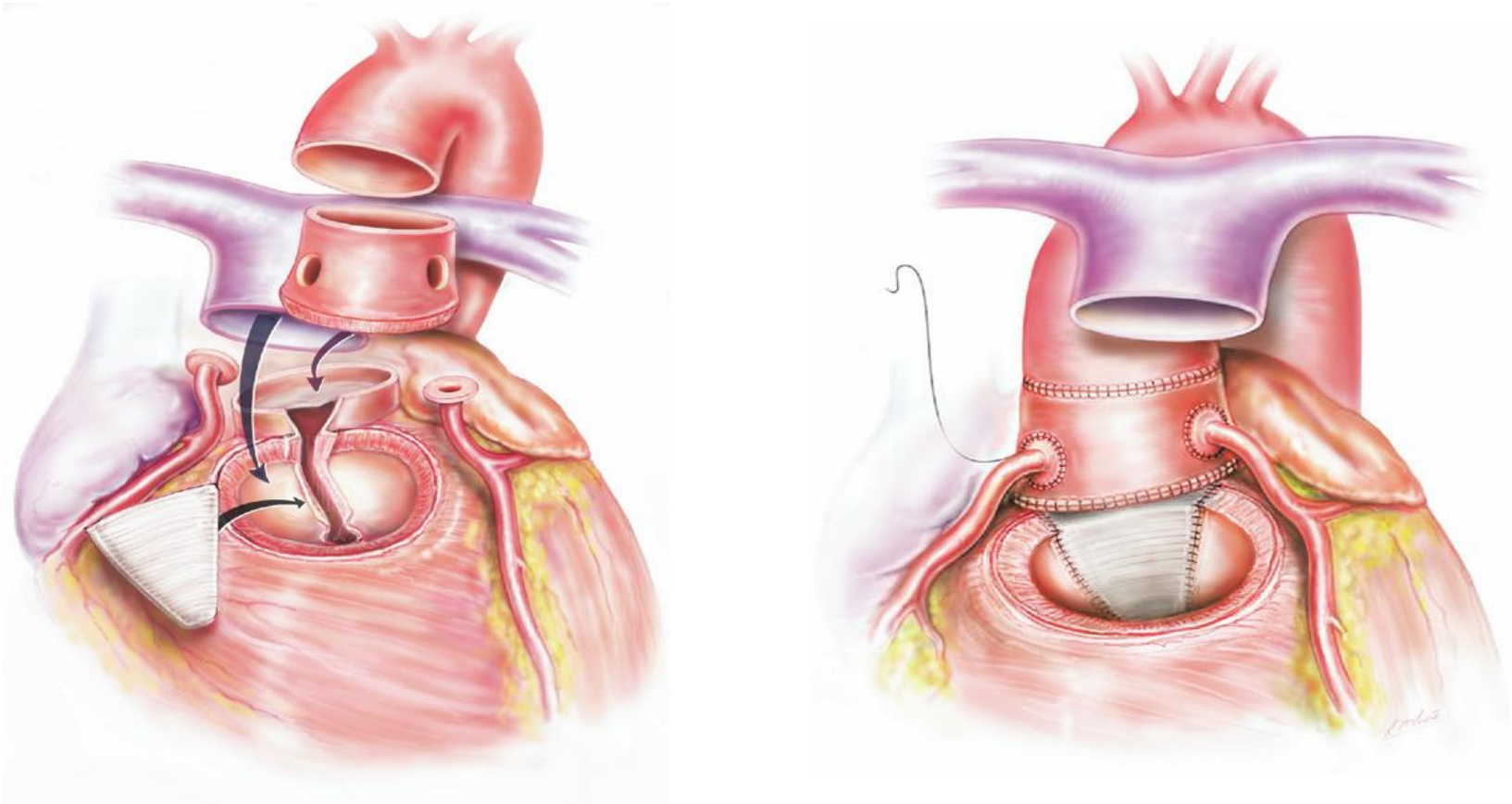
Background A d-transposition of the great arteries with left ventricular outflow tract obstruction represents a difficult surgical problem. The Rastelli procedure is the usual approach to this condition. However, recurrent left ventricular outflow tract obstruction and early conduit obstruction as well as arrhythmias and troublesome late mortality are significant limitations.

Methods From 1993 to 2005, 11 children (8 male, 3 female) ages 1 month to 11 years (median age 7 months) have undergone aortic root autograft translocation plus arterial switch to correct d-transposition of the great arteries with left ventricular outflow tract obstruction. The native aortic root was excised from the right ventricle infundibulum and inserted into the left ventricular outflow, enlarging the outflow tract by resecting the outlet septum and an appropriate-size ventricular septal defect patch. After coronary artery reimplantation, right ventricular outflow reconstruction was achieved with a homograft.

Results There were no early or late deaths. With a median follow-up of 59 months (range 2 to 137 months), 5 patients required 6 conduit replacement procedures at a median time of 53 months. Two patients required an implantable defibrillator for ventricular arrhythmias. None of the patients have developed left ventricular outflow tract obstruction.

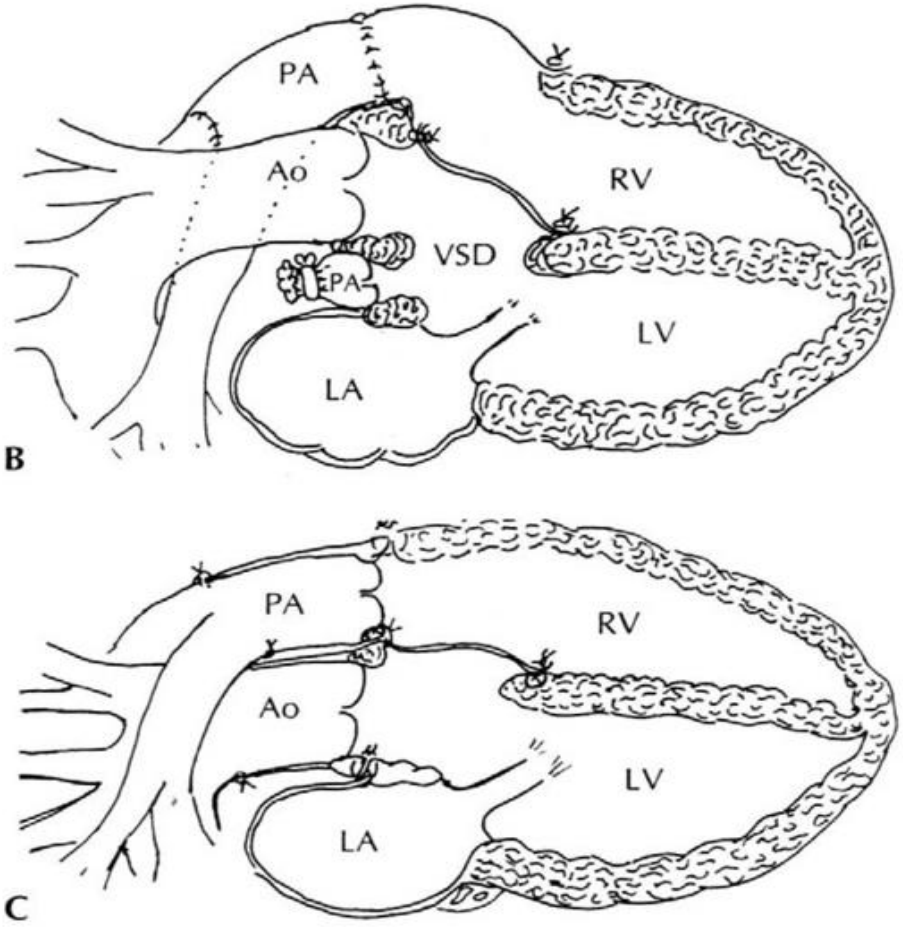
Conclusions Aortic root autograft plus arterial switch procedure is a good option for the surgical management of infants and children with d-transposition of the great arteries and left ventricular outflow tract obstruction and results in a more anatomic repair compared with Rastelli operation. Intermediate-term results indicate good relief of left ventricular outflow tract obstruction and need for conduit replacement compares favorably with the Rastelli procedure for this lesion. (J Am Coll Cardiol 2007;49:485–90) © 2007 by the American College of Cardiology Foundation

Ross-Switch-Konno



J Am Coll Cardiol 2007;49:485-90

Rastelli vs. Aortic Translocation



Ann Thorac Surg 2005;79:2089-93

The aortic translocation (Nikaidoh) procedure: Midterm results superior to the Rastelli procedure

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Objective: Midterm follow-up is analyzed after the aortic translocation (Nikaidoh) procedure, an alternative to the Rastelli procedure for ventriculoarterial discordance, ventricular septal defect, and pulmonary stenosis.

Methods: Nineteen patients underwent a Nikaidoh procedure at a median age of 3.3 years (0.9-9.3 years). The native aortic valve was translocated from the right to the left ventricular outflow tract by full (n = 6) or partial (n = 13) mobilization of the aortic root. Seven patients with partial mobilization had the right coronary artery reimplanted as a button. The conal septum was divided in 13 patients. The right ventricular outflow tract was reconstructed with either a homograft (n = 4) or a right ventricular outflow tract patch (n = 15). The median follow-up was 11.4 years (0.1-23 years), and the median age at follow-up was 17.4 years (1-30 years). Left ventricular outflow tract obstruction and aortic insufficiency were assessed by echocardiography.

Results: One patient died of right coronary arterial ischemia. All remaining patients (95%) survived. The median survival was 13.6 years (longest, 23.0 years). Seven right ventricular outflow tract reoperations were required in 5 patients (6 with obstruction and 1 with pulmonary insufficiency). No reoperations have been performed on the left ventricular outflow tract or aortic valve. No patient had any left ventricular outflow tract obstruction or aortic insufficiency more than mild (mild in 9 patients, trivial in 3 patients, and absent in 6 patients).

Conclusions: Midterm actuarial survival was 95% after the Nikaidoh procedure. Reintervention for the right ventricular outflow tract is more common when valved conduits are used versus valveless reconstruction; however, the Nikaidoh procedure provides complete freedom from important aortic insufficiency and left ventricular outflow tract obstruction.

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ORIGINAL ARTICLE

Modified Nikaidoh procedure for the correction of complex forms of transposition of the great arteries with ventricular septal defect and left ventricular outflow tract obstruction: mid-term results

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Abstract

OBJECTIVES: Different surgical techniques for the treatment of complex transposition of the great arteries (TGA) with ventricular septal defect and left ventricular outflow tract obstruction (LVOTO) have been developed, in particular the Rastelli operation, the réparation à l'étage ventriculaire procedure and the Nikaidoh procedure. The hitherto published results of the Nikaidoh procedure and its modifications compare favourably with those of other techniques; however, experience with the Nikaidoh procedure is still limited. Here, we report our institutions' early and mid-term results with modifications of the Nikaidoh procedure.

METHODS: Twenty-one patients who underwent a modified Nikaidoh procedure between 2006 and 2012 at our institution, either as aortic root translocation (n = 17) or as *en bloc* rotation of the arterial trunk (n = 4), were studied retrospectively.

RESULTS: There were 2 early and 1 mid-term deaths. The follow-up continued for a median of 2.3 years (range 0.3–6.4 years). During the follow-up, the performance of the reconstructed left ventricular outflow tract (LVOT) remained excellent: no reobstruction and no aortic valve regurgitation classified as more than mild were observed. Left ventricular function was well preserved. In 4 patients, a significant reoccurring right ventricular outflow tract obstruction due to conduit failure was observed; so far, two reoperations with conduit replacement have been necessary. The mean right ventricular outflow tract peak gradient was 24 ± 7.2 mmHg at the last follow-up in the remaining patients. No reobstruction of the right ventricular outflow tract occurred in patients with preserved pulmonary valve tissue after *en bloc* rotation.

CONCLUSIONS: The aortic translocation procedure is a valuable surgical option for patients with complex (TGA) with ventricular septal defect and LVOTO. The mid-term results document excellent performance of the reconstructed LVOT. Modifications of the Nikaidoh procedure that preserve pulmonary valve tissue may further reduce the need for right ventricular outflow tract reoperation.

Aortic root translocation with atrial switch: Another surgical option for congenitally corrected transposition of the great arteries with isolated pulmonary stenosis

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J Thorac Cardiovasc Surg 2010;139:1652-3

Anatomical Repair of Double Outlet Right Ventricle Associated with Complete Atrioventricular Septal Defect and Pulmonary Stenosis: Extending the Indications for Aortic Translocation

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J Card Surg 2012;27:231-4

Pulmonary root translocation in malposition of great arteries repair allows right ventricular outflow tract growth

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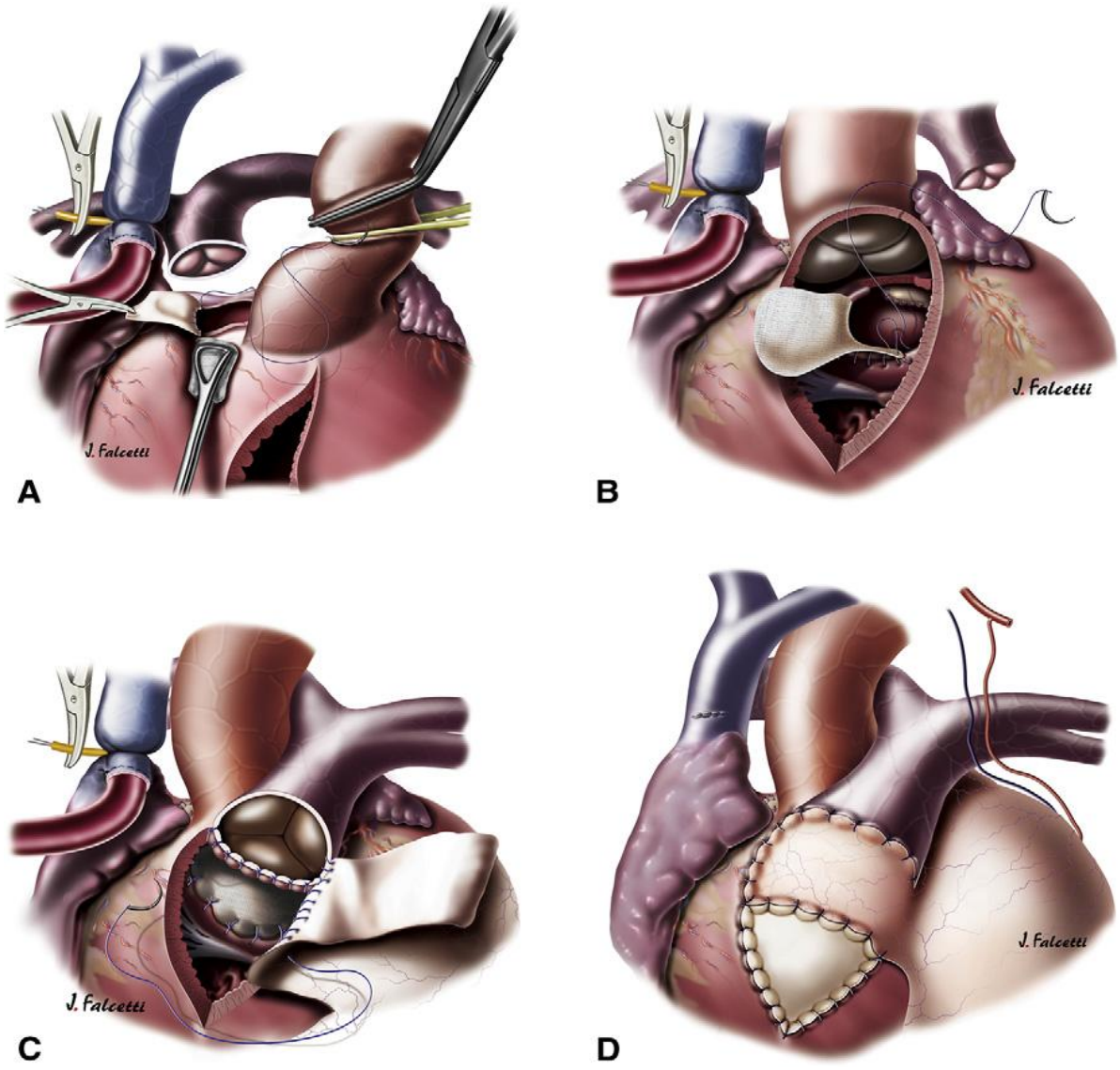
Objective: Optimal surgical treatment of patients with transposition of the great arteries (TGA), ventricular septal defect (VSD), and pulmonary stenosis (PS) remains a matter of debate. This study evaluated the clinical outcome and right ventricle outflow tract performance in the long-term follow-up of patients subjected to pulmonary root translocation (PRT) as part of their surgical repair.

Methods: From April 1994 to December 2010, we operated on 44 consecutive patients (median age, 11 months). All had malposition of the great arteries as follows: TGA with VSD and PS (n = 33); double-outlet right ventricle with subpulmonary VSD (n = 7); double-outlet right ventricle with atrioventricular septal defect (n = 1); and congenitally corrected TGA with VSD and PS (n = 3). The surgical technique consisted of PRT from the left ventricle to the right ventricle after construction of an intraventricular tunnel that diverted blood flow from the left ventricle to the aorta.

Results: The mean follow-up time was 72 ± 52.1 months. There were 3 (6.8%) early deaths and 1 (2.3%) late death. Kaplan-Meier survival was 92.8% and reintervention-free survival was 82.9% at 12 years. Repeat echocardiographic data showed nonlinear growth of the pulmonary root and good performance of the valve at 10 years. Only 4 patients required reinterventions owing to right ventricular outflow tract problems.

Conclusions: PRT is a good surgical alternative for treatment of patients with TGA complexes, VSD, and PS, with acceptable operative risk, high long-term survivals, and few reinterventions. Most patients had adequate pulmonary root growth and performance. (J Thorac Cardiovasc Surg 2012;143:1292-8)



J Thorac Cardiovasc Surg 2012;143:1292-8