Stanford Children's Health | Lucile Packard Children's Hospital Stanford

Surgical Management of Corrected Transposition

Michael Ma, MD

May 4, 2019

Stanford University School of Medicine

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Department of Cardiac Surgery

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Surgical Management of Atrioventricular and Ventriculoarterial Discordance

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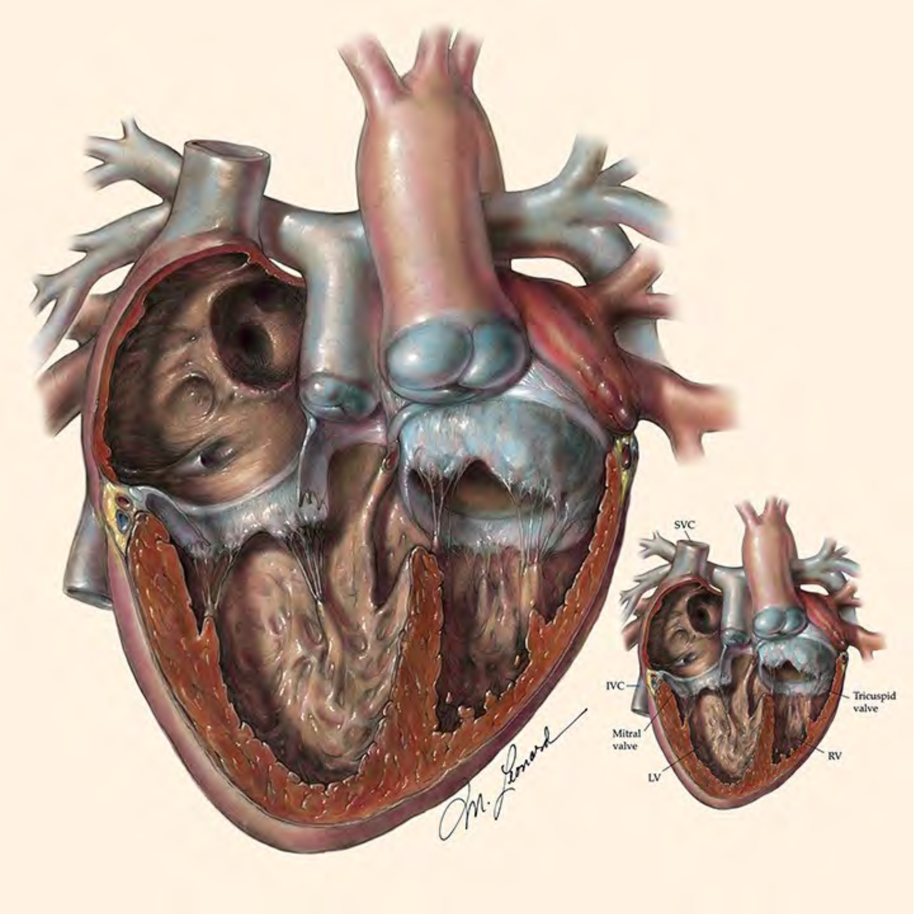
Disclosures: none

Introduction

Anatomy

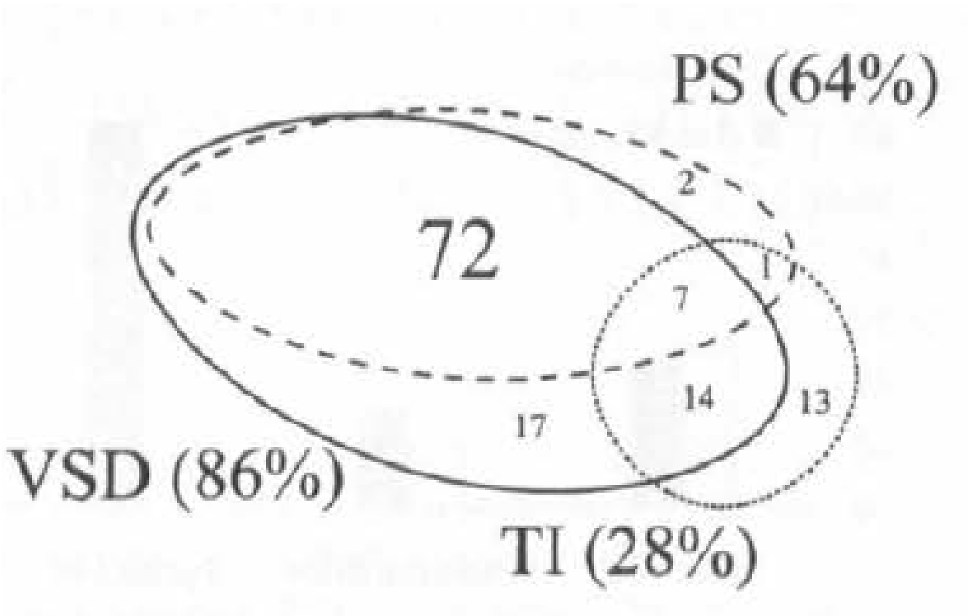
- Anderson
  - Atrio-ventricular discordance
  - Ventriculo-arterial discordance
  - RA → LV → PA
  - LA → RV → Ao
- Van Praagh
  - SLL 95%
  - IDD 5%
- <1% of Congenital Heart Disease



Associated Defects

- VSD (70-85%)
- Pulmonary stenosis (60-70%)
- Ebsteinoid tricuspid valve (20-30%)
- AV block (1-2% per year of life, 98% per year freedom from AVB)
  - IDD without increased risk

- In combination:
  - VSD and PS (50%)
  - VSD alone (20%)
  - PS alone (20%)
  - No VSD and no PS (<10%)

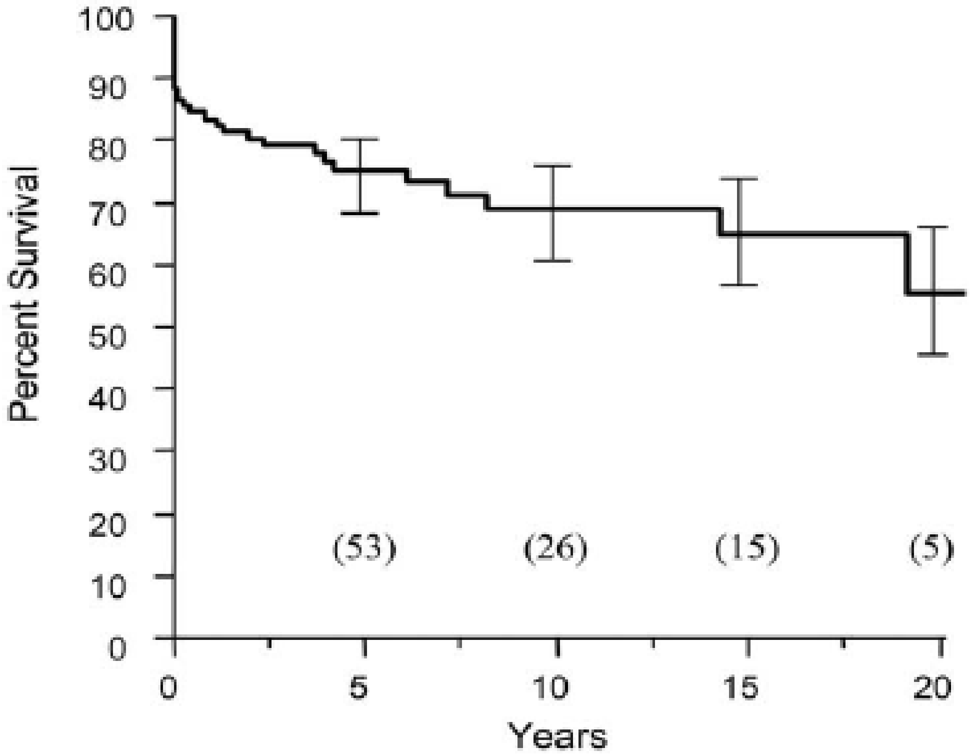


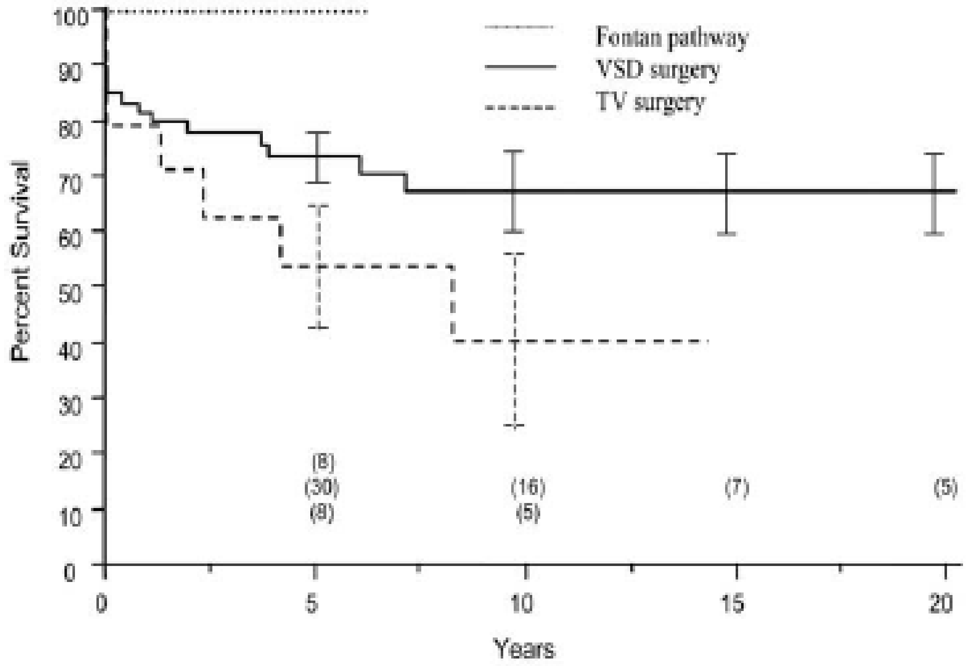
Physiologic Repair

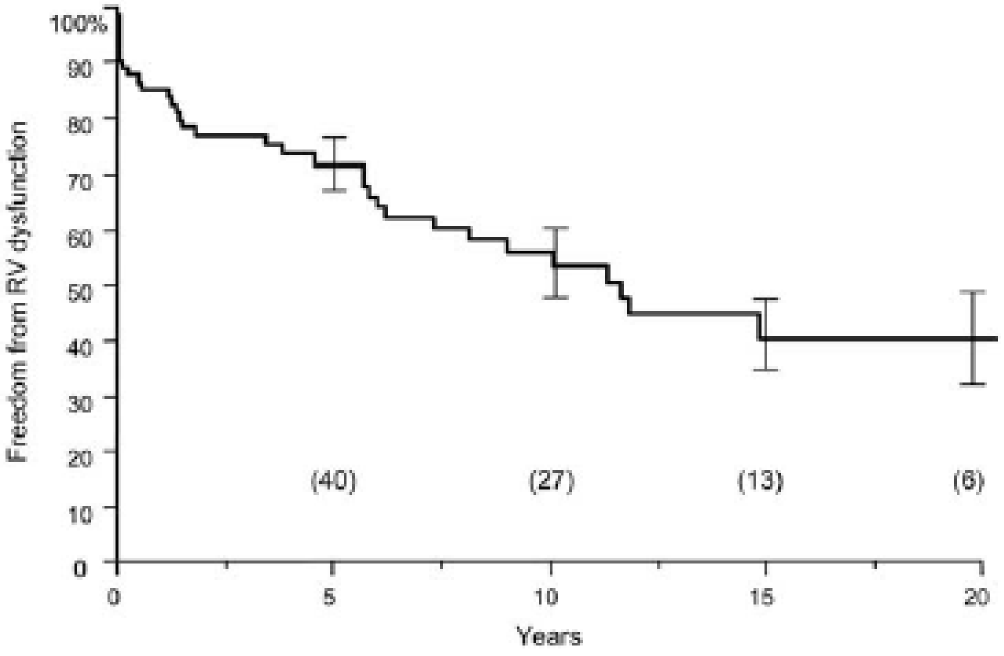
- VSD closure
- PS repair/replacement
- TR repair/replacement
- PPM

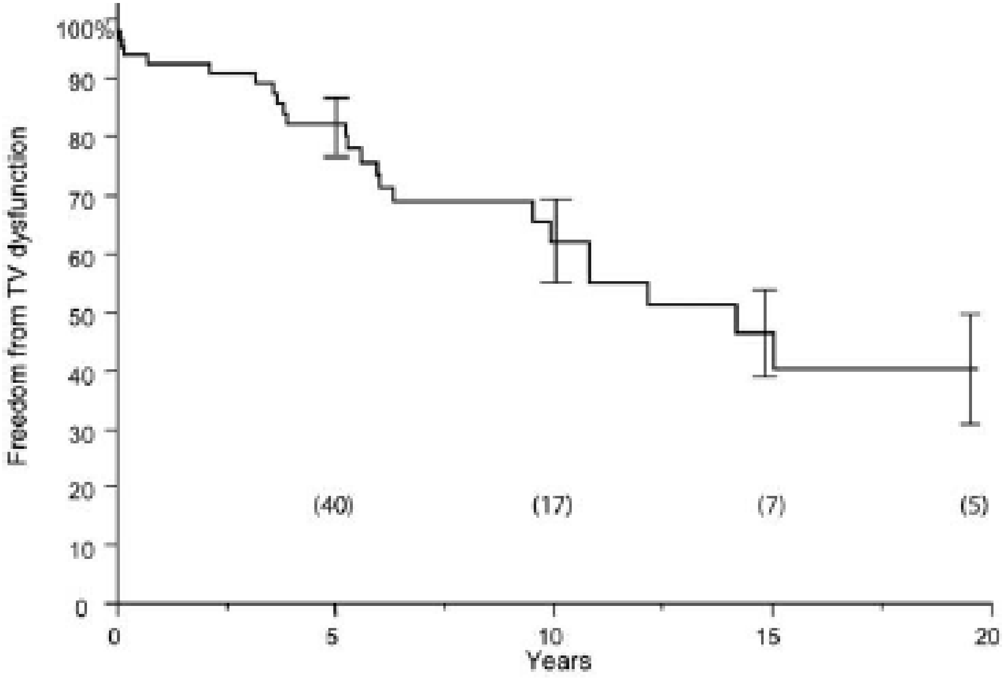
Long-term outcome of surgically treated patients with corrected transposition of the great arteries

Viktor Hraska, MD[a]
Brian W. Duncan, MD[b]
John E. Mayer, Jr, MD[c]
Michael Freed, MD[d]
Pedro J. del Nido, MD[c]
Richard A. Jonas, MD[e]






100%
90
80
70
60
50
40
30
20
10
0
Freedom from RV dysfunction
(40)
(27)
(13)
(6)
0
5
10
15
20
Years


100%
90
80
70
60
50
40
30
20
10
0
Freedom from TV dysfunction
(40)
(17)
(7)
(5)
0
5
10
15
20
Years

Stanford Children’s Management Algorithm

Stanford Children's Management Algorithm

Associated Lesion	Physiology	Early Intervention	Definitive Intervention
VSD only	• LV remains pressure-loaded • Adequate semilunar valves	PAB to control PBF	ASO, VSD closure, hemi-Mustard, SCC
VSD/PS	• LV remains pressure-loaded • Inadequate neo-Ao valve for ASO	PAB v. shunt to control PBF v. no intervention	LV-Ao baffle, hemi-Mustard, SCC; RV-PA conduit v. PA translocation
PS only	• LV loses pressure-load • Inadequate neo-Ao valve for ASO • Lack of VSD for baffle	Shunt to achieve adequate PBF v. no intervention	Best candidate for avoiding intervention
None	• LV loses pressure-load • Adequate semilunar valves	PAB for LV training	ASO, hemi-Mustard, SCC

Stanford Children's Health treatment algorithm for common anatomic variants of corrected transposition. LV, left ventricle; Ao, aorta; PAB, pulmonary artery band; PBF, pulmonary blood flow; ASO, arterial switch operation; SCC, superior cavopulmonary connection; RV, right ventricle; PA, pulmonary artery.

Stanford Children's Management Algorithm

Associated Lesion	Physiology	Early Intervention	Definitive Intervention
VSD only	• LV remains pressure-loaded • Adequate semilunar valves	PAB to control PBF	ASO, VSD closure, hemi-Mustard, SCC
VSD/PS	• LV remains pressure-loaded • Inadequate neo-Ao valve for ASO	PAB v. shunt to control PBF v. no intervention	LV-Ao baffle, hemi-Mustard, SCC; RV-PA conduit v. PA translocation
PS only	• LV loses pressure-load • Inadequate neo-Ao valve for ASO • Lack of VSD for baffle	Shunt to achieve adequate PBF v. no intervention	Best candidate for avoiding intervention
None	• LV loses pressure-load • Adequate semilunar valves	PAB for LV training	ASO, hemi-Mustard, SCC Neonatal ASO, Senning

Stanford Children's Health treatment algorithm for common anatomic variants of corrected transposition. LV, left ventricle; Ao, aorta; PAB, pulmonary artery band; PBF, pulmonary blood flow; ASO, arterial switch operation; SCC, superior cavopulmonary connection; RV, right ventricle; PA, pulmonary artery.

Hemi-Mustard/Superior Cavopulmonary Connection

- In all patients:
  - SA node preservation
  - Avoidance of SVC obstruction
  - Clamp time reduction / improvement in technical reproducibility
    - “1/2 Mustard anatomically, ¼ Mustard surgically”

- In select patients:
  - Apicocaval juxtaposition (ventricular mass and apex align with cava)
    - Situs inversus with levo/mesocardia
    - Situs solitus with dextro/mesocardia
  - Conduit preservation
  - Native PA translocation
  - Tricuspid valve competence

LV Training

Operative Technique

- PA band placement (silastic)
  - Place band on distal MPA to avoid distortion of the pulmonary (ie neo-aortic) root and valve
- Central venous line (CVP = LVEDP)
- Direct MPA or indirect (through RA) LV catheter placement
- TEE
- Ionotropic support (mild (ie dopamine 3-5 mcg/kg/min))

- PA band titration under guidance
  - Band to achieve near-systemic MPA/LVSP or heart failure
  - Monitor CVP and TEE
    - CVP < 8-10
    - MR ≤ mild
    - No regional wall motion abnormalities
    - No significant ventricular arrhythmia

ICU Management

- POD 0 – maintenance of ionotropic support, mechanical ventilation, sedation
- POD 1 – TTE to assess function, extubation
- POD 2 – TTE to assess function, gradual ionotropic downtitration
- Exercise clinical judgment in the exact timing of postoperative support de-escalation (ie take more time as needed)

Interval Assessment

- At 6-9 months
  - Catheterization
  - MRI
  - Echo

Table Scorecard of the Preparedness of a Patient for a Double-Switch Procedure

(1) Left ventricular pressure	90% of systemic pressure
(2) Left ventricular systolic function	Ejection fraction > 55%
(3) Left ventricular end diastolic pressure	Less than 12 mm Hg
(4) Mitral valve function	Mild or less insufficiency
(5) Left ventricular mass (by MRI)	65 g/m^2

MRI, magnetic resonance imaging.

Results


Left Ventricular Retraining and Double Switch in Patients With Congenitally Corrected Transposition of the Great Arteries

Ali N. Ibrahimiye, MD[1], Richard D. Mainwaring, MD[1], William L. Patrick, BS[1], Laura Downey, MD[2], Vamsi Yarlagadda, MD[3], and Frank L. Hanley, MD[1]

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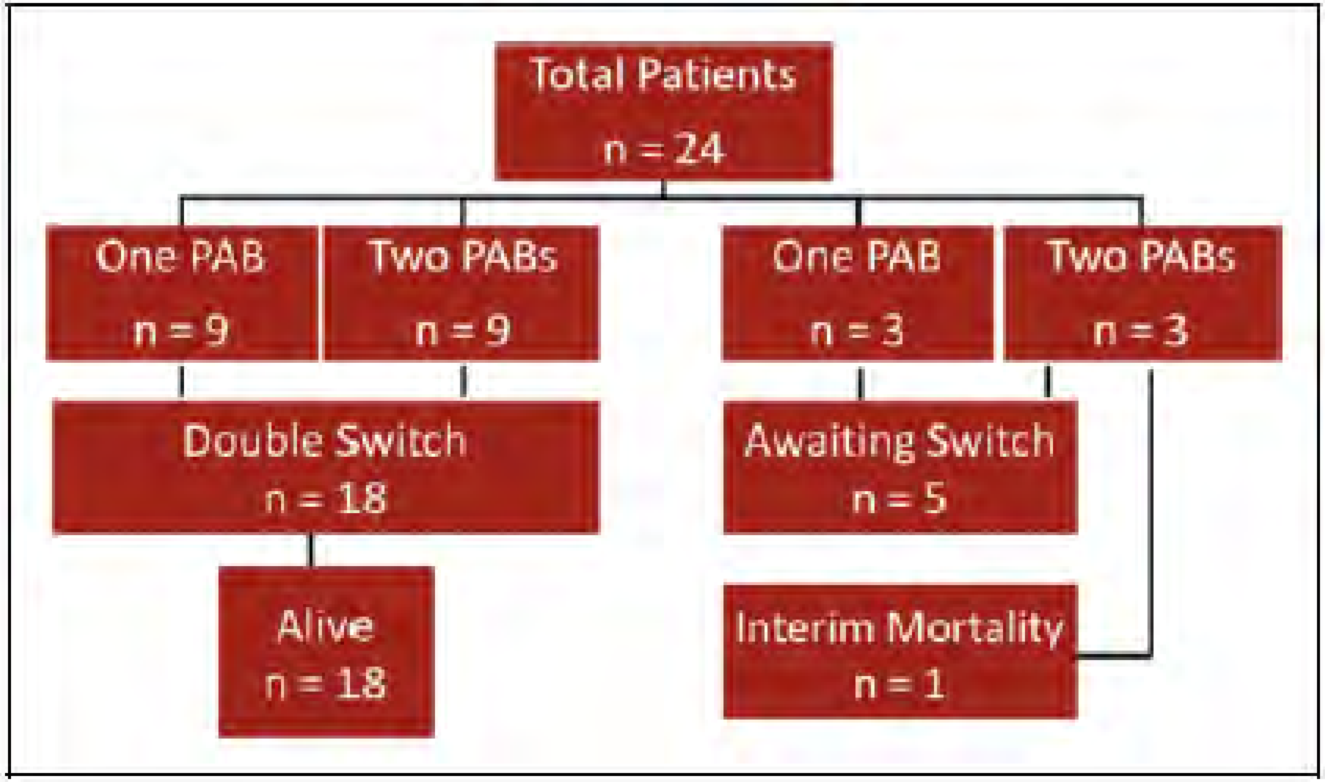
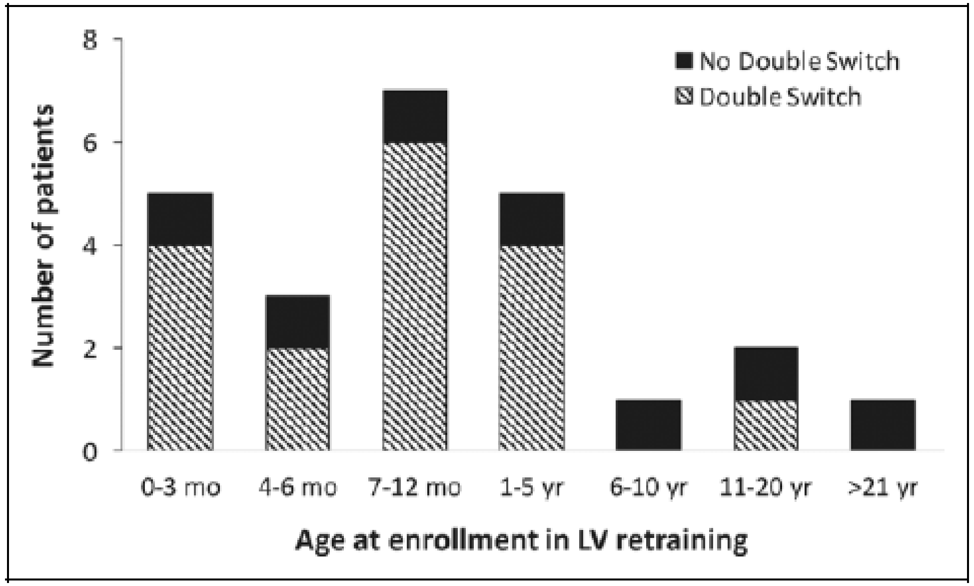
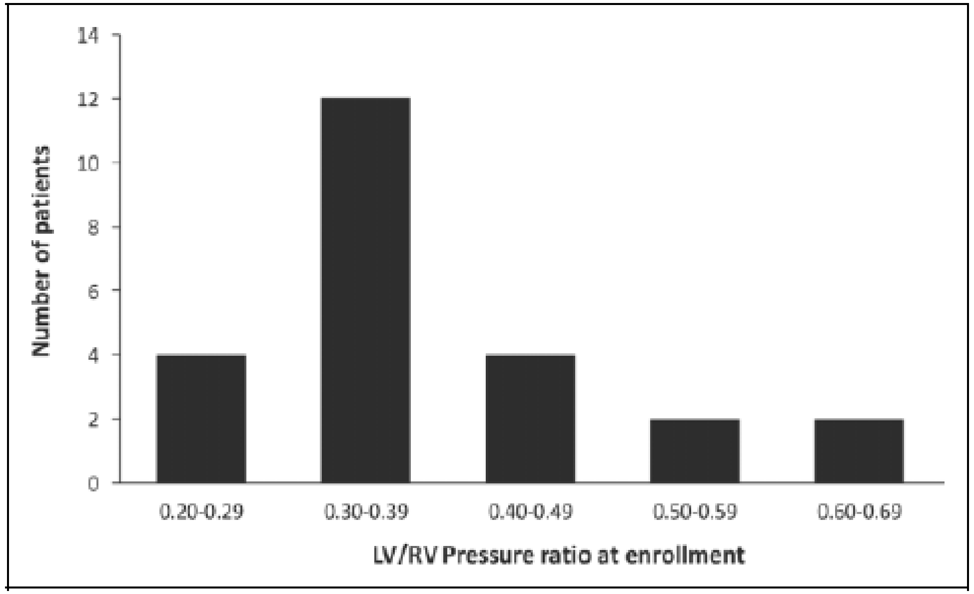
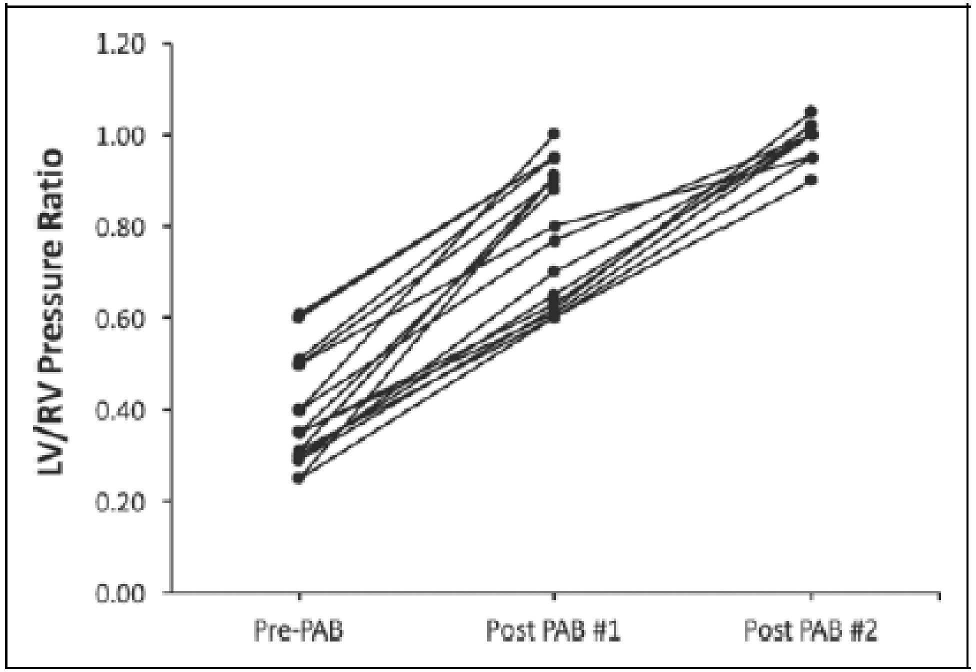

Total Patients
n = 24
One PAB
n = 9
Two PABs
n = 9
One PAB
n = 3
Two PABs
n = 3
Double Switch
n = 18
Awaiting Switch
n = 5
Alive
n = 18
Interim Mortality
n = 1

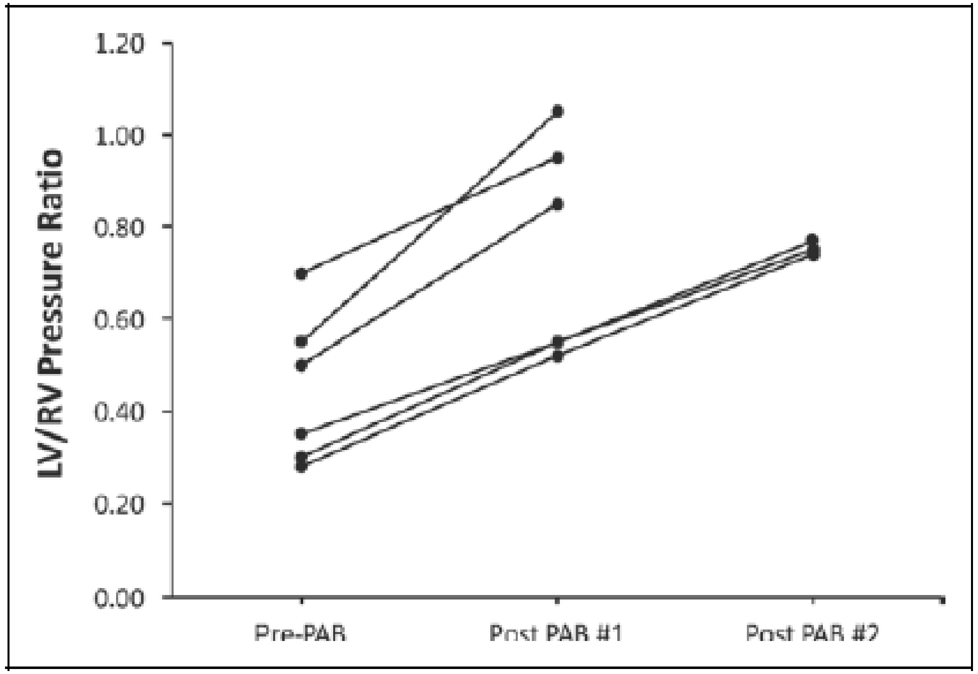


No Double Switch
Double Switch
Number of patients
0
2
4
6
8
0-3 mo
4-6 mo
7-12 mo
1-5 yr
6-10 yr
11-20 yr
>21 yr
Age at enrollment in LV retraining



Number of patients
0
2
4
6
8
10
12
14
0.20-0.29
0.30-0.39
0.40-0.49
0.50-0.59
0.60-0.69
LV/RV Pressure ratio at enrollment


1.20
1.00
0.80
0.60
0.40
0.20
0.00
LV/RV Pressure Ratio
Pre-PAB
Post PAB #1
Post PAB #2


1.20
1.00
0.80
0.60
0.40
0.20
0.00
LV/RV Pressure Ratio
Pre-PAB
Post PAB #1
Post PAB #2

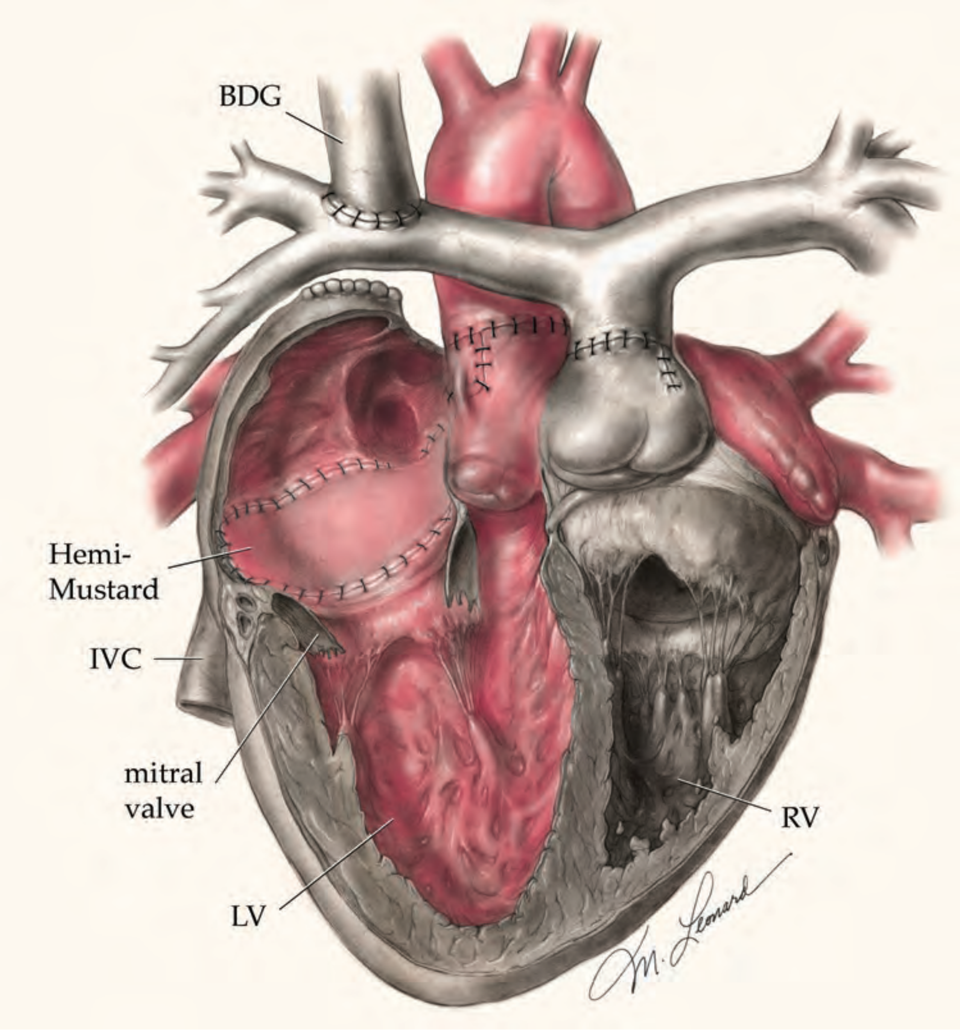
Post-operative Training

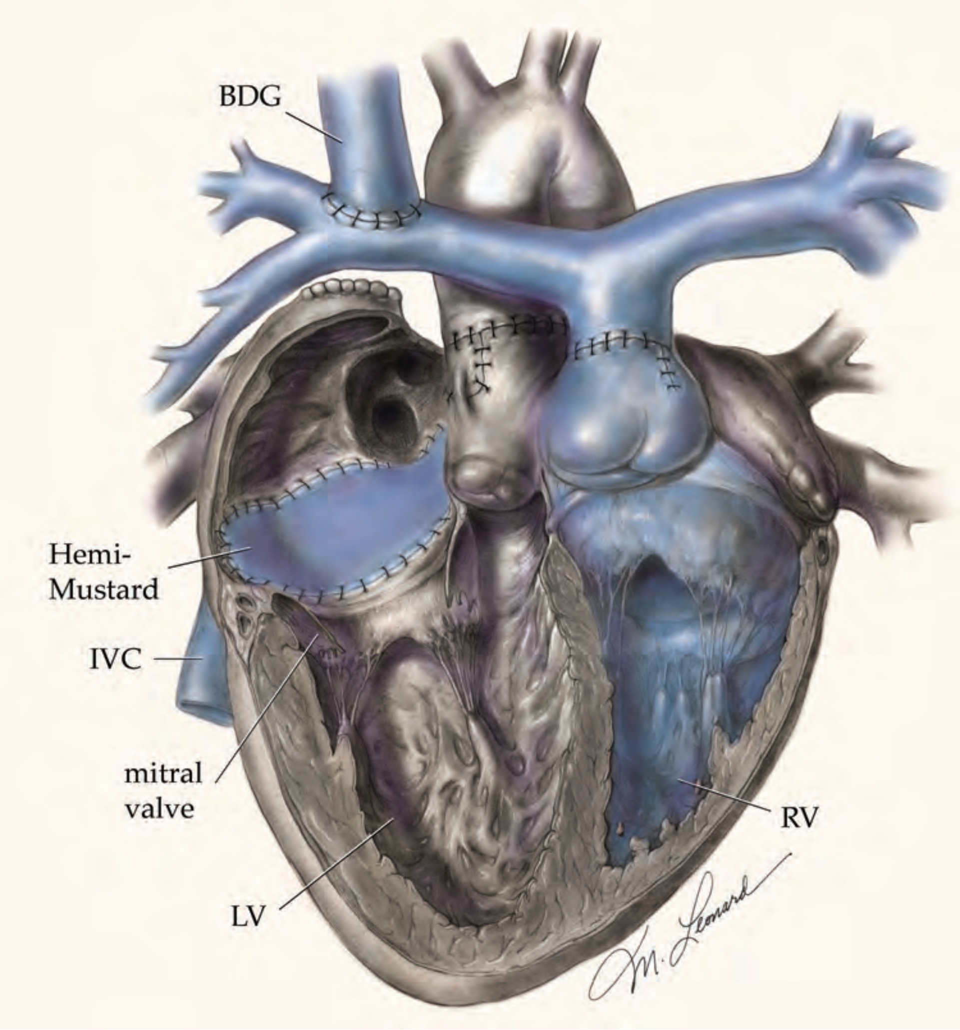
- We observed unexpected LV failure in our early experience
- Theory
  - LV training develops the ventricle to a peak systolic pressure that the PAB can simulate
  - Small MPA stump does not retain an appropriate diastolic pressure for the LV to train against in late diastole/early systole at the time of isovolumic contraction before aortic valve opening
  - LV remains undertrained for this important phase of the cardiac cycle
- Practice
  - LV must continue to train after definitive repair
  - Continue aggressive fluid restriction and diuretic therapy for one year after definitive repair

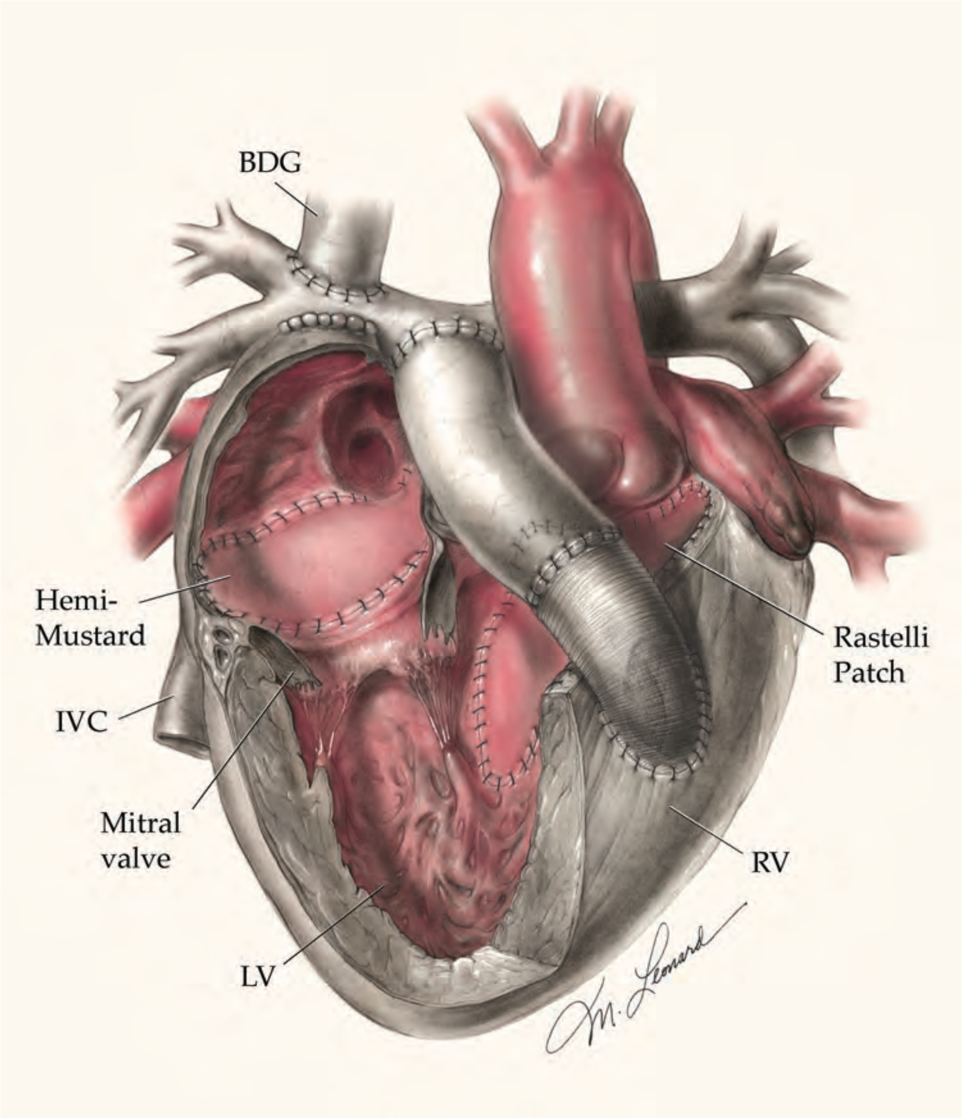
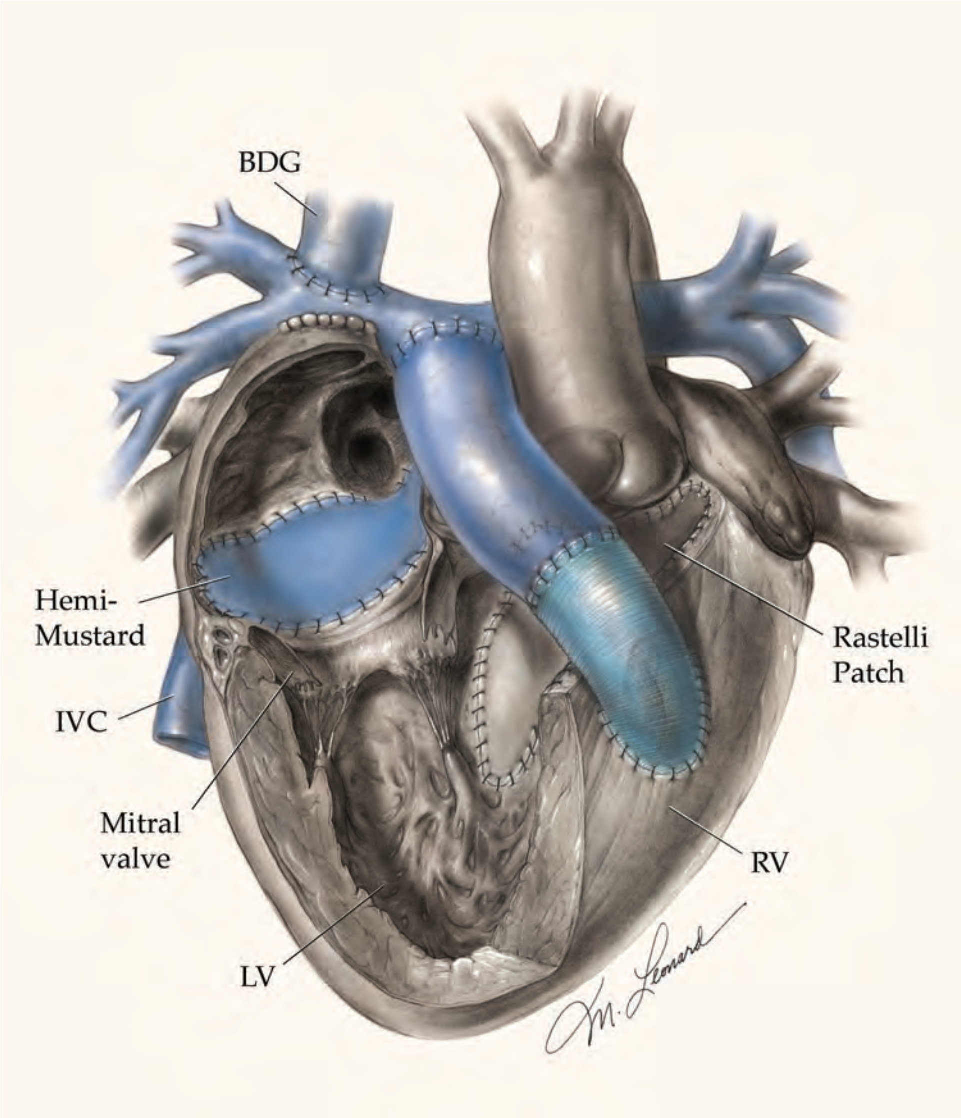
Double Switch

Operative Technique

Ventriculo-Arterial Discordance

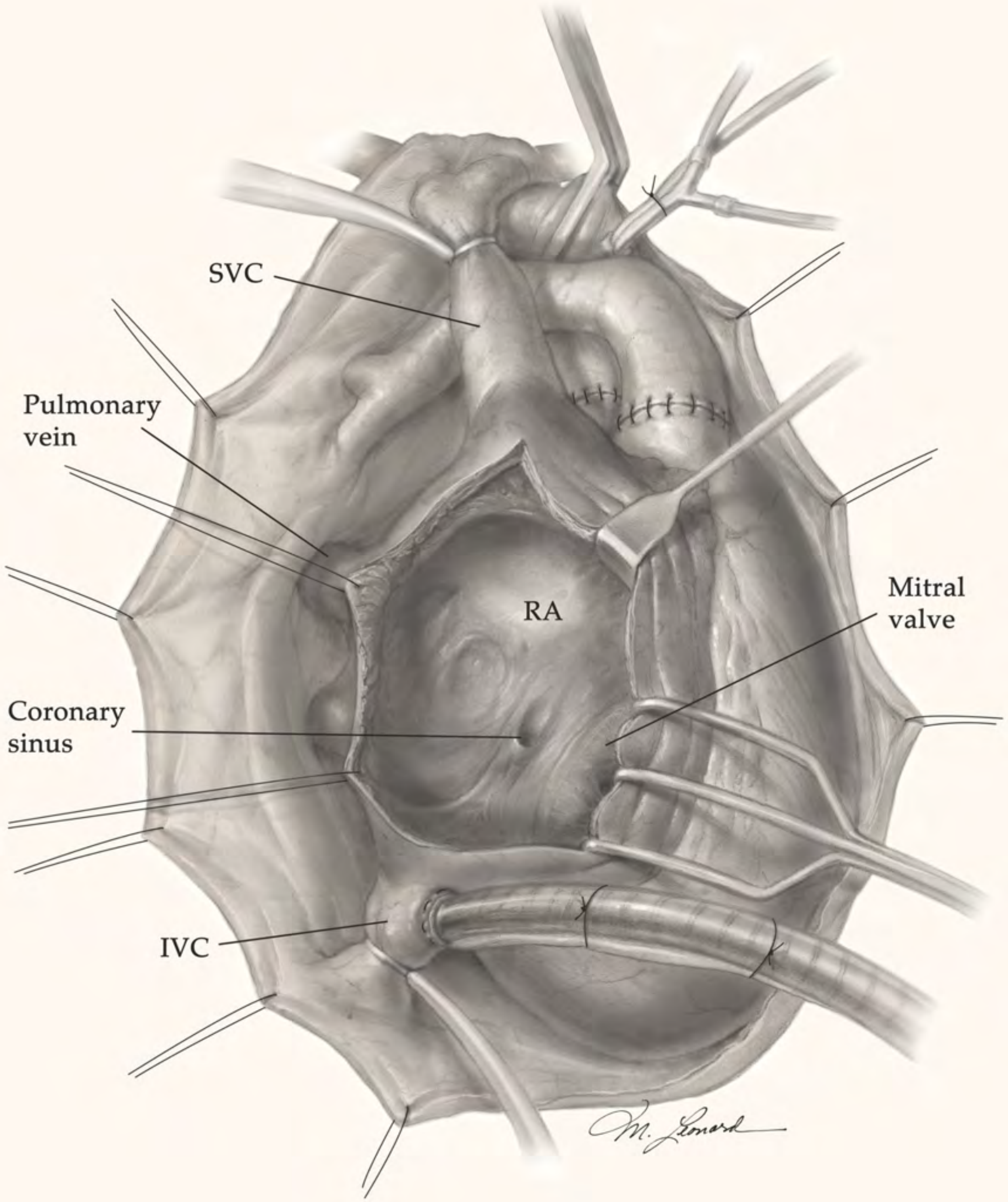

BDG
Hemi-
Mustard
IVC
mitral
valve
LV
RV
M. Leonard

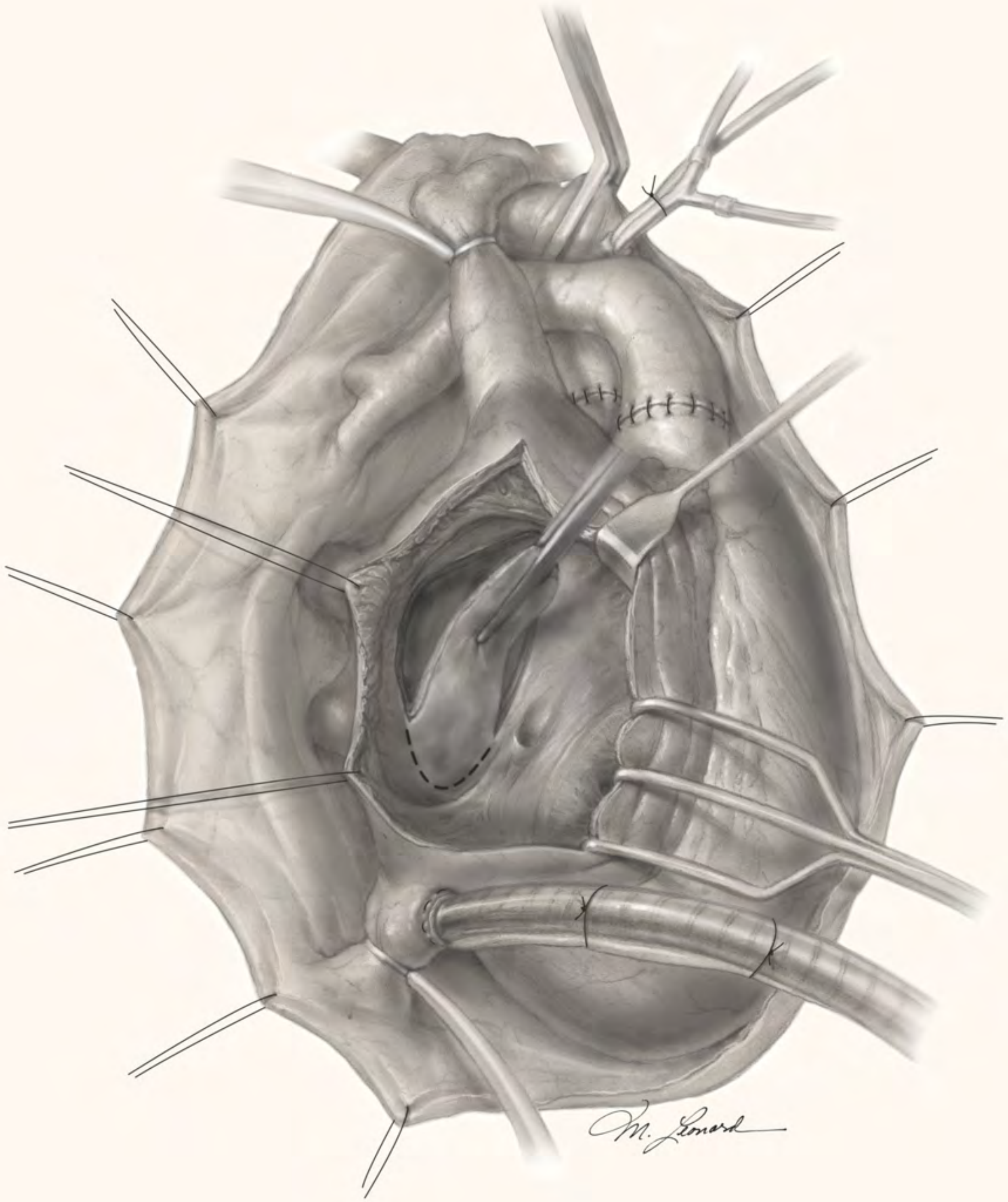

BDG
Hemi-
Mustard
IVC
mitral
valve
LV
RV
M. Leonard

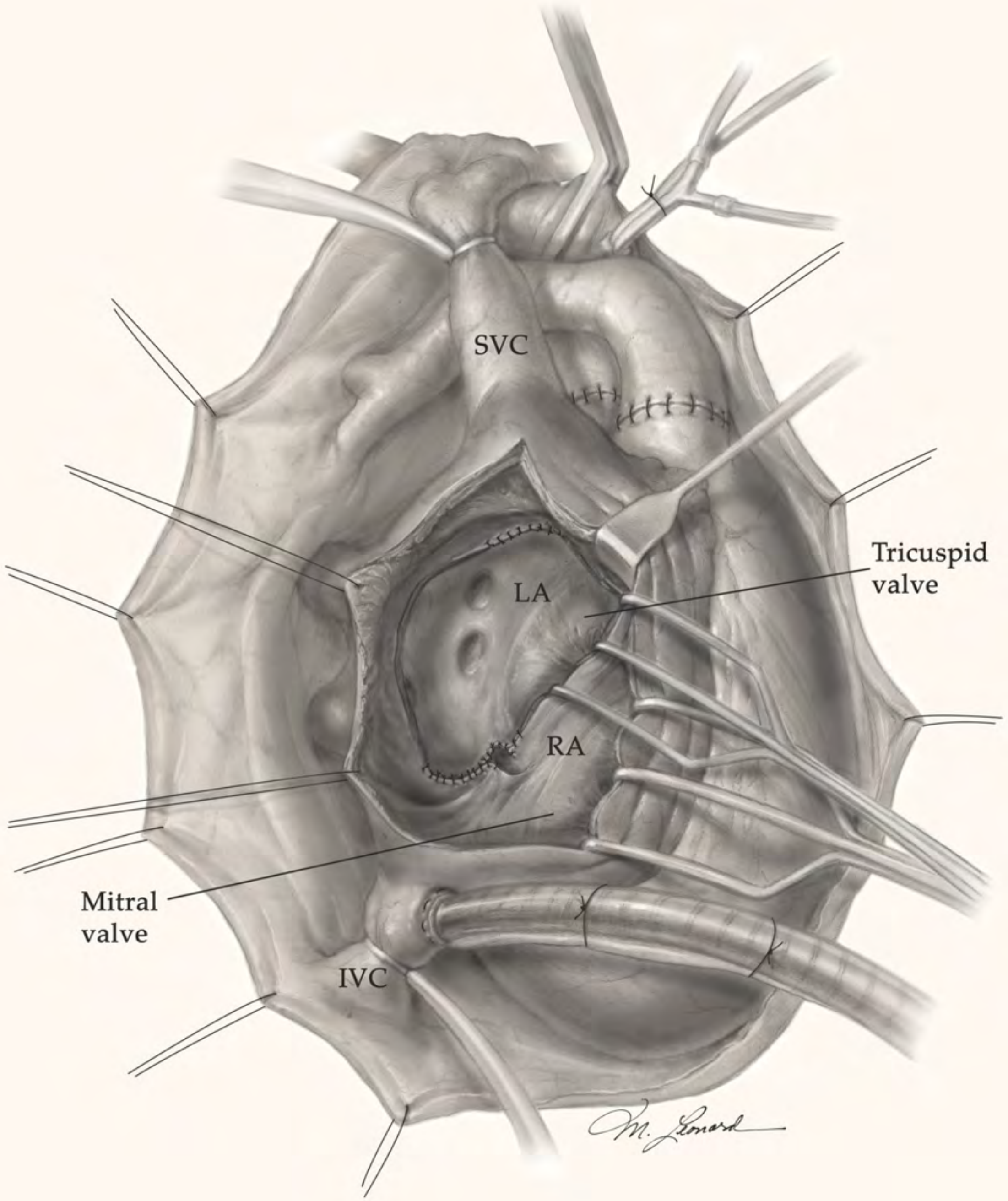

BDG
Hemi-
Mustard
IVC
Mitral
valve
LV
Rastelli
Patch
RV

BDG
Hemi-
Mustard
IVC
Mitral
valve
LV
Rastelli
Patch
RV

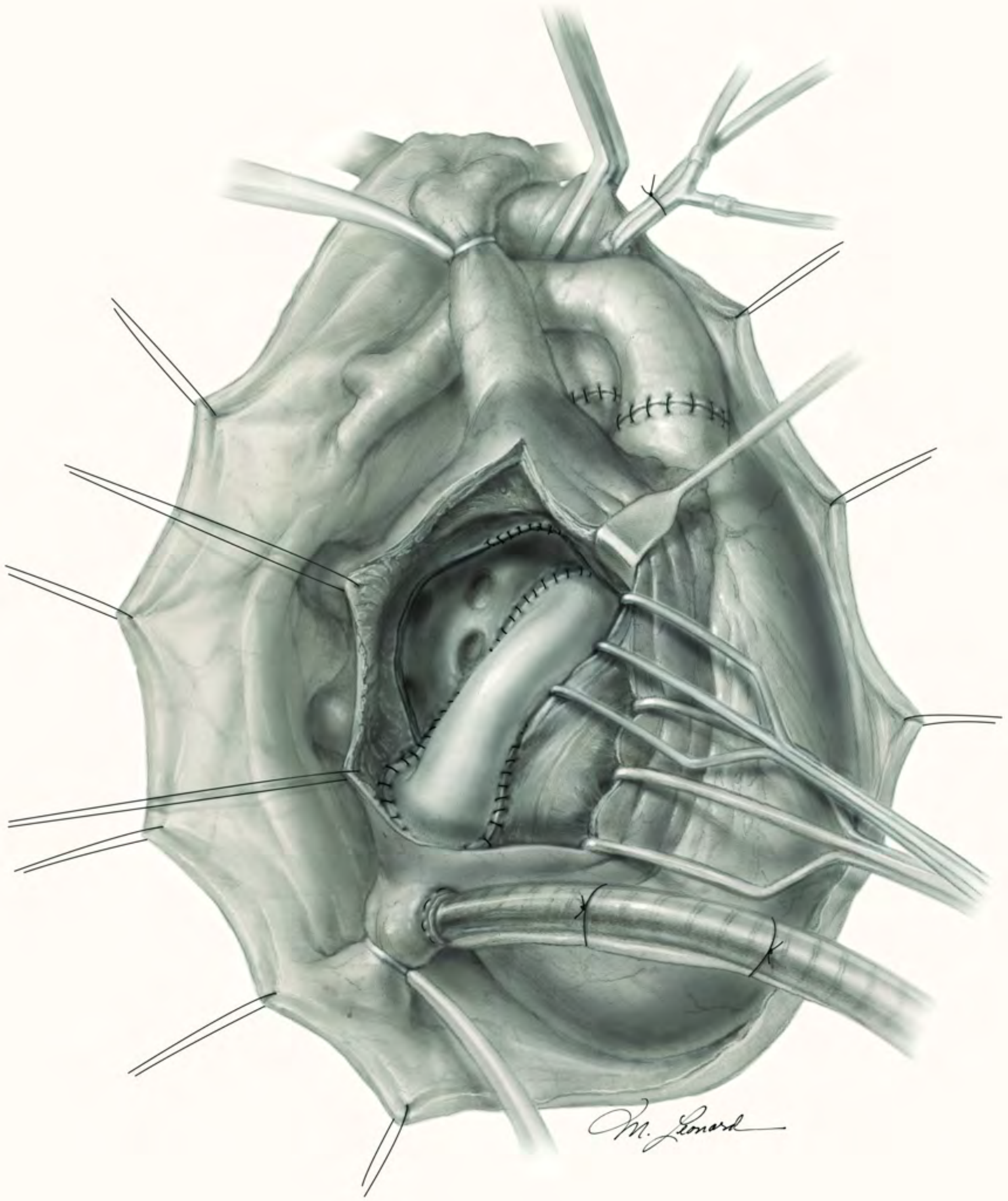
Operative Technique

Atrio-ventricular Discordance


SVC
Pulmonary vein
RA
Mitral valve
Coronary sinus
IVC

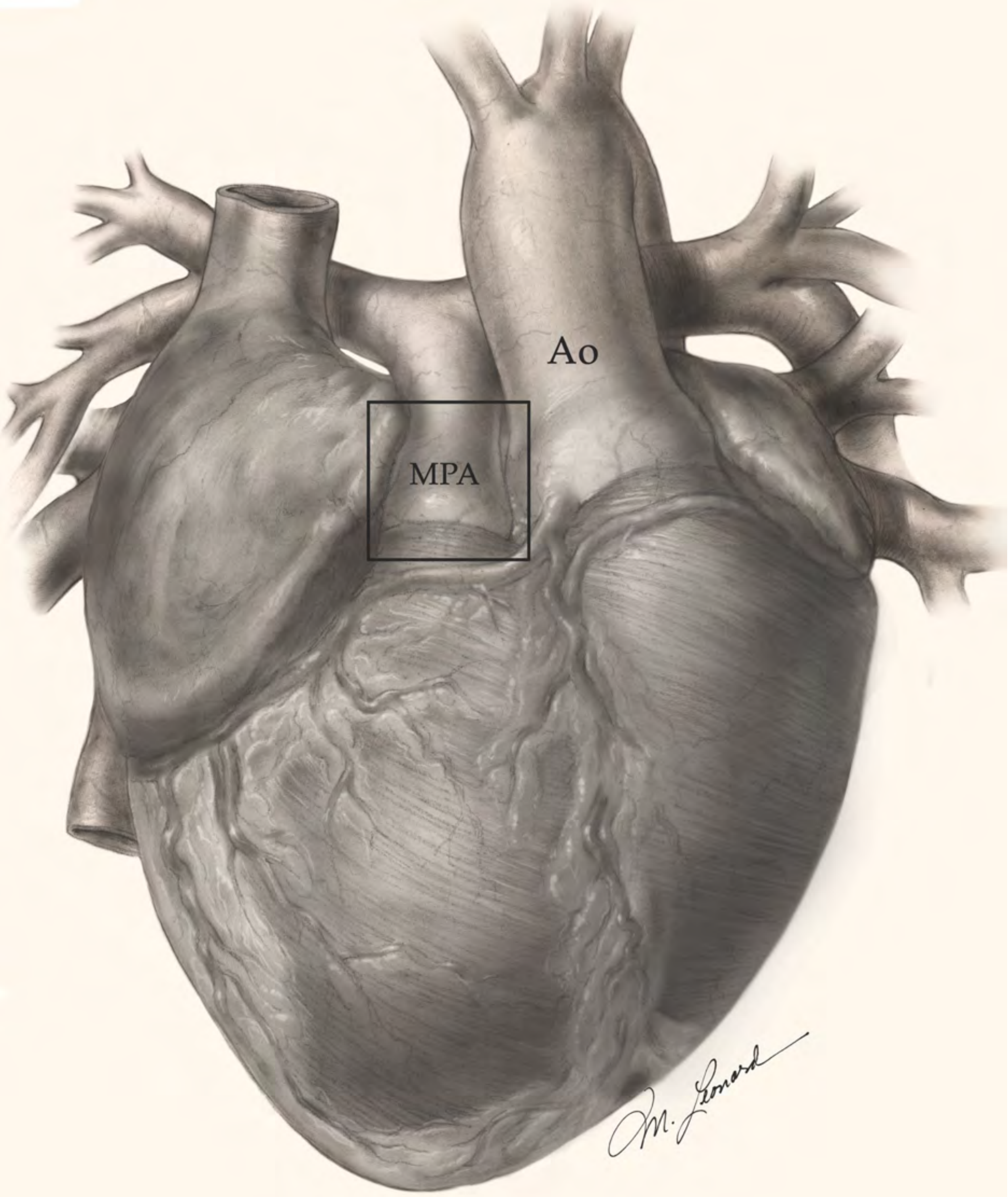

M. Leonard


SVC
Tricuspid
valve
LA
RA
Mitral
valve
IVC
M. Leonard

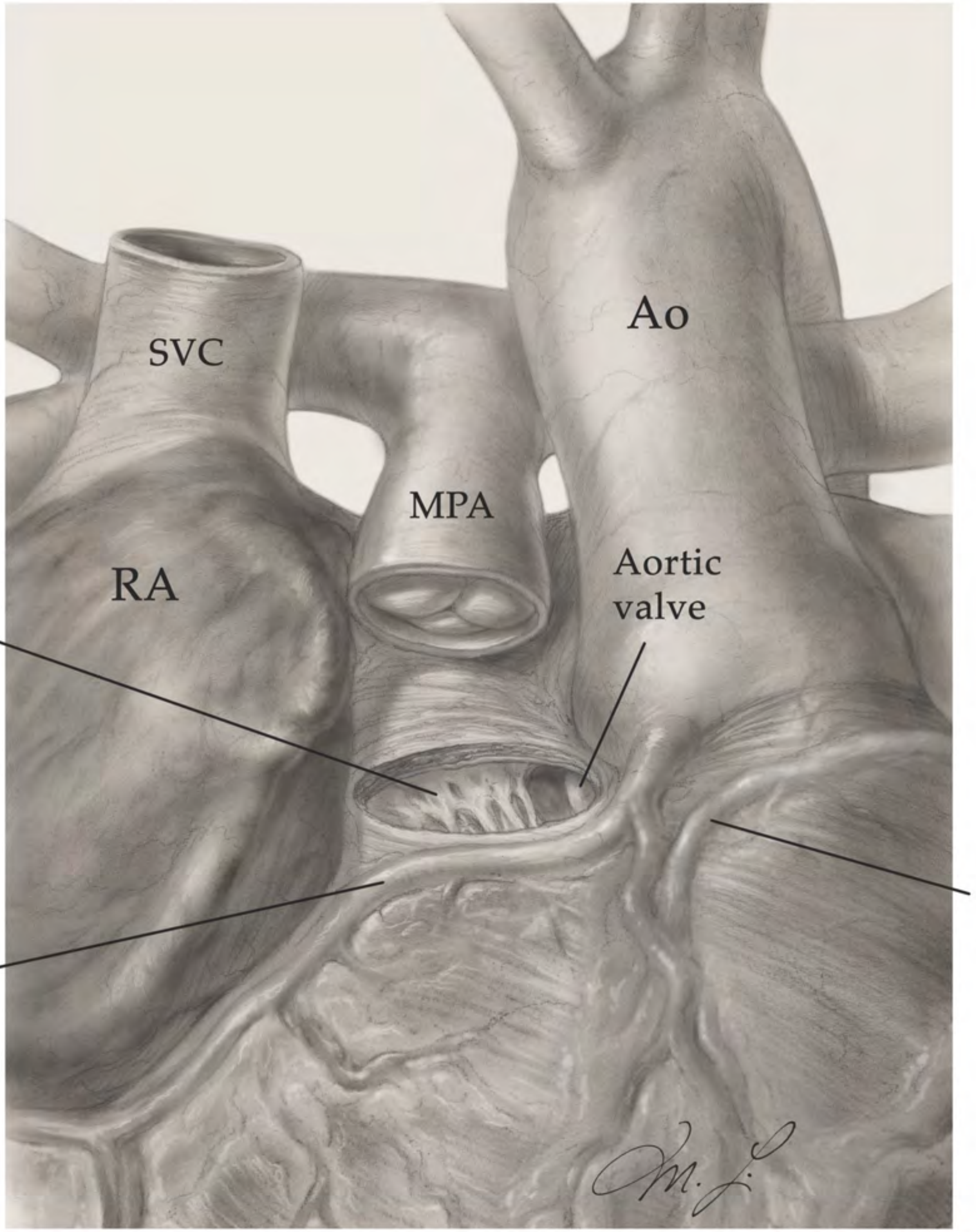


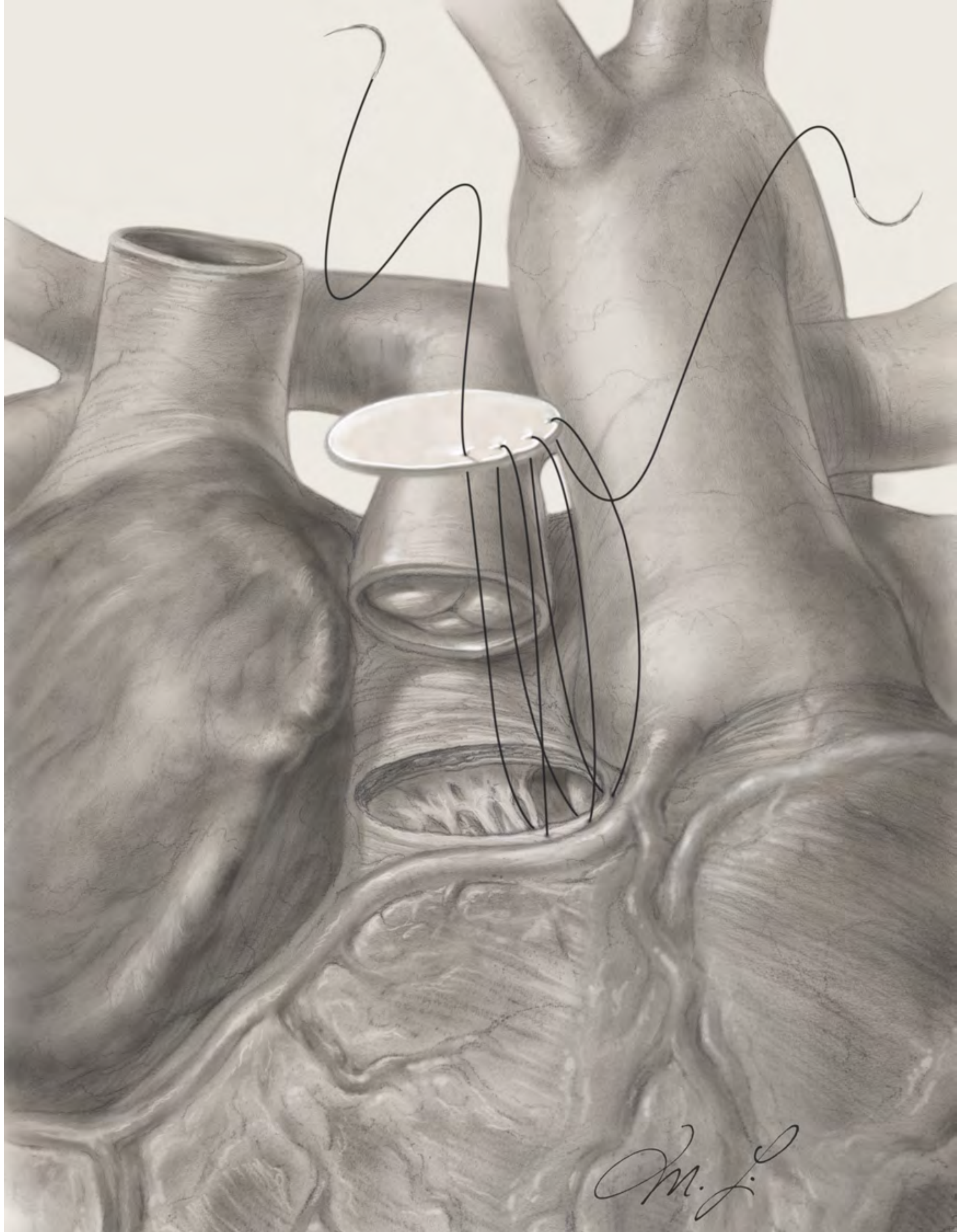
Operative Technique

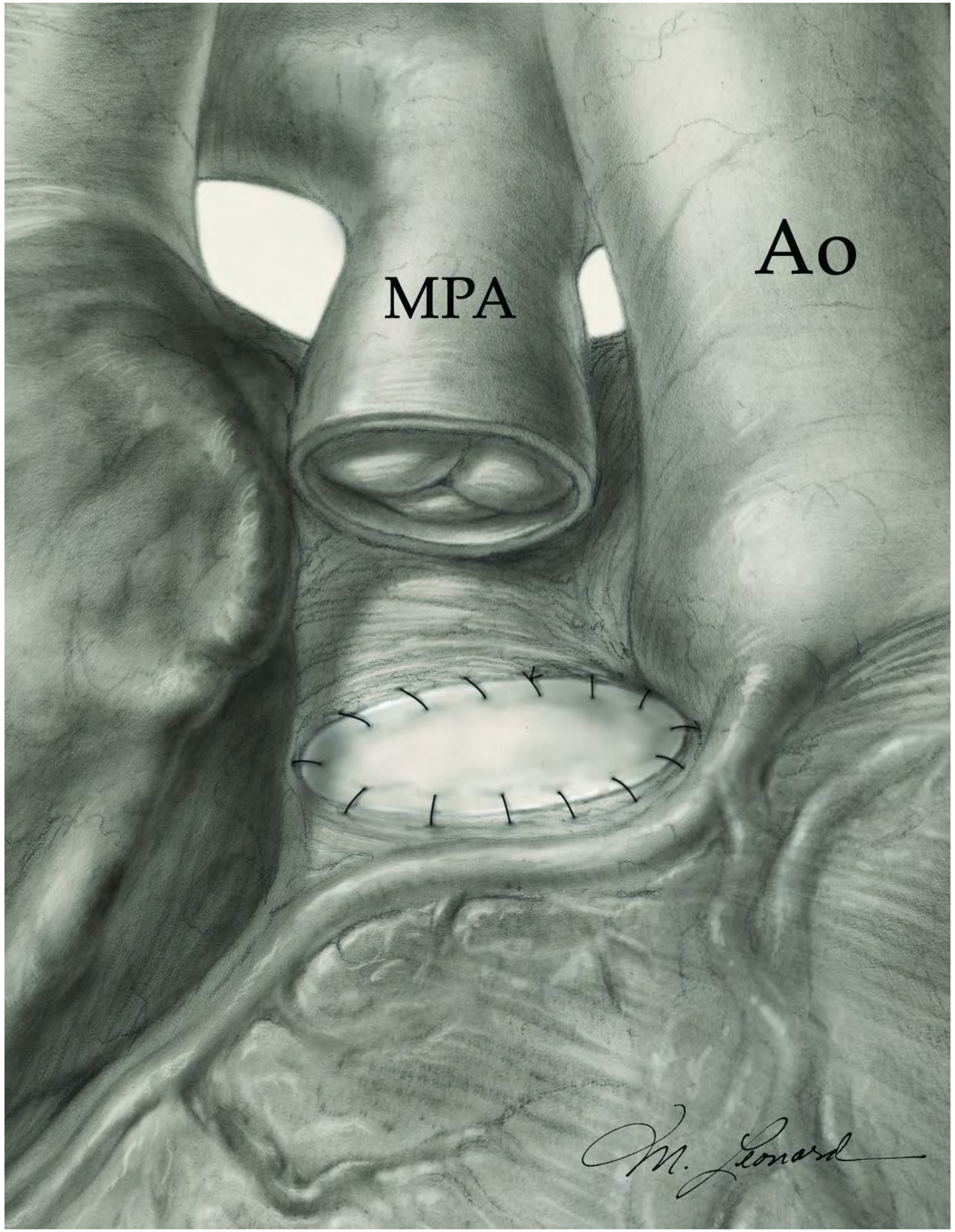
Native PA Root Translocation



Ao
MPA
M. Leonard


SVC
Ao
MPA
RA
Aortic valve
Mitral valve
Right coronary artery
Left coronary artery




MPA
Ao
M. Leonard

Results

The hemi-Mustard/bidirectional Glenn atrial switch procedure in the double-switch operation for congenitally corrected transposition of the great arteries: Rationale and midterm results

Sunil P. Malhotra, MD,[a] V. Mohan Reddy, MD,[b] Mary Qiu, BS,[b] Timothy J. Pirolli, MD,[b]
Laura Barboza, BS,[b] Olaf Reinhartz, MD,[b] and Frank L. Hanley, MD[b]

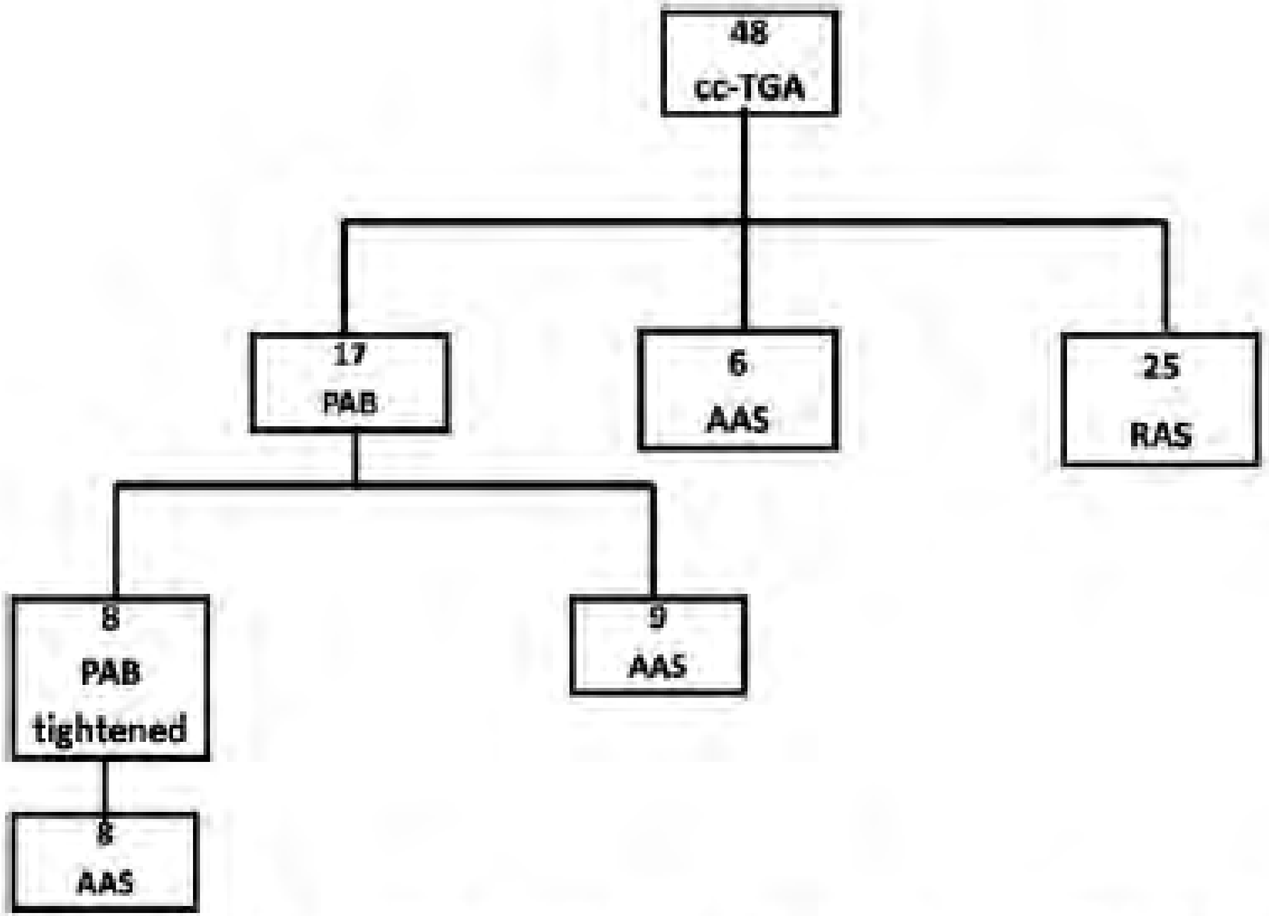


FIGURE 1. The surgical approach to anatomic repair in 48 patients. *ccTGA*, Congenitally corrected transposition of the great arteries; *PAB*, pulmonary artery banding; *AAS*, arterial–atrial switch; *RAS*, Rastelli–atrial switch.

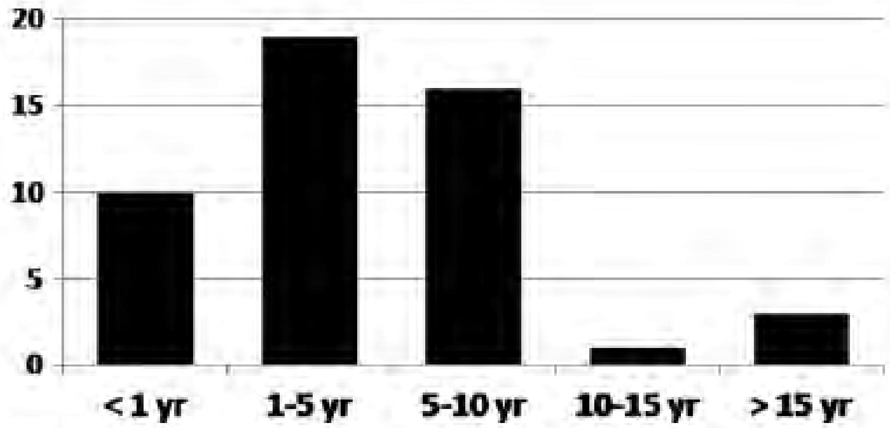
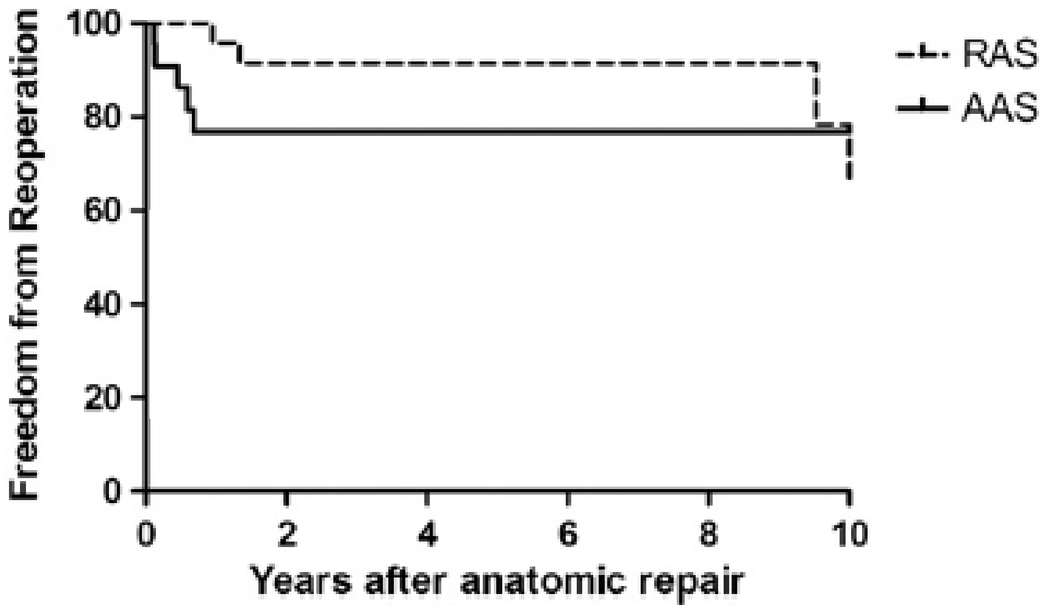
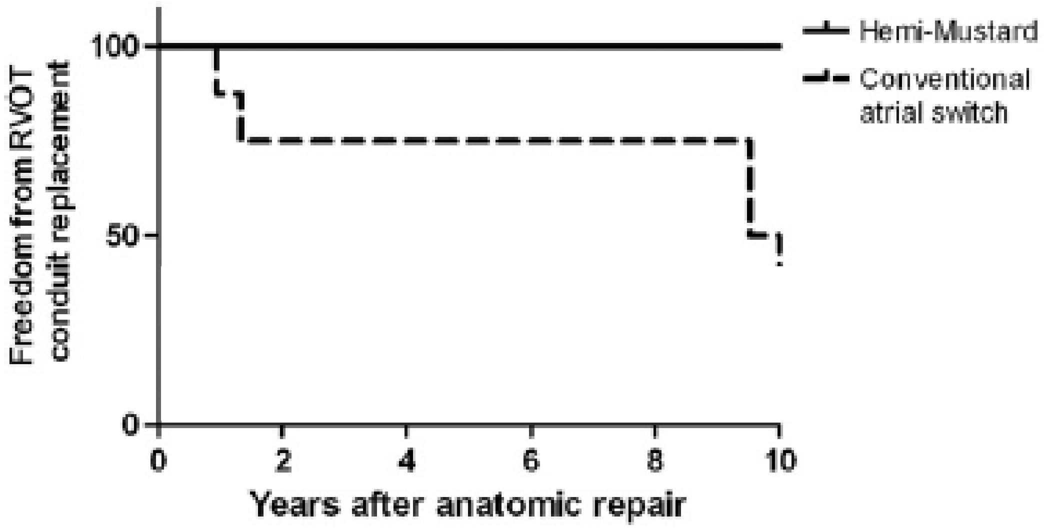
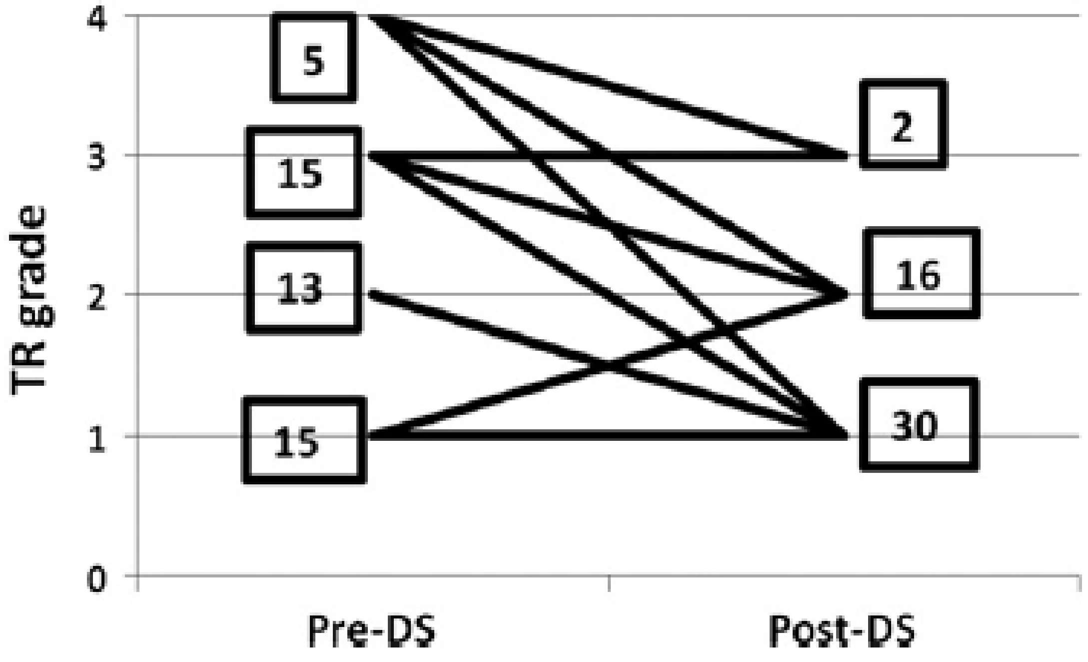


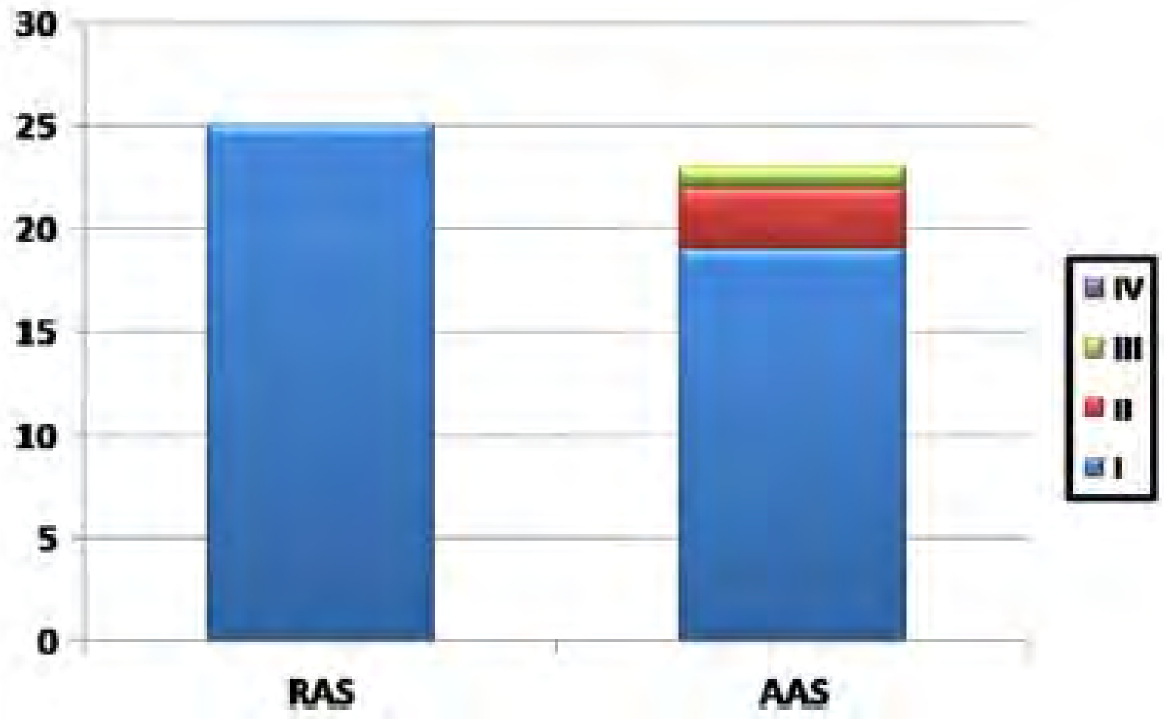
FIGURE 2. Age distribution of patients at the time of anatomic repair.


Freedom from Reoperation
100
80
60
40
20
0
0
2
4
6
8
10
Years after anatomic repair
RAS
AAS


Freedom from RVOT conduit replacement
100
50
0
0
2
4
6
8
10
Years after anatomic repair
Hemi-Mustard
Conventional atrial switch



TR grade
4
3
2
1
0
5
15
13
15
2
16
30
Pre-DS
Post-DS



30
25
20
15
10
5
0
RAS
AAS
IV
III
II
I

Update

- 98 patients
  - Arterial switch – 49
  - Rastelli – 49
  - Hemi-Mustard with Superior Cavopulmonary Connection – 77

  - Pulmonary root translocation – 3
    - Free PI without stenosis – 1
    - Mild PS/PI - 1
    - Mild PI, no PS – 1

  - Operative mortality – 3

Summary

Summary

- The initial management of corrected transposition is dependent on its commonly associated defects, and the timing of diagnosis.

- In most variations, returning the morphologic left ventricle to the systemic circulation is prioritized to reduce the known long-term morbidity and mortality of relying on systemic morphologic right ventricular support.

- Left ventricular deconditioning can be managed with an LV training algorithm that utilizes surgical pulmonary artery banding coupled with methodical and regular post-operative assessments of biventricular function through echocardiography, catheterization, and MRI.

- Ultimate surgical correction is afforded by the double (atrial and arterial-level) switch operation. A hemi-Mustard and superior cavopulmonary connection (1) reduces known atrial baffle complications, (2) improves tricuspid regurgitation, (3) extends the longevity of RV-PA conduits, and (4) enables native pulmonary artery translocation and preservation in patients with intrinsic pulmonary stenosis.

Thank You