Tetralogy of Fallot, Pulmonary Atresia, with MAPCAs
Technique for Early Complete Repair

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TOF, Pulmonary Atresia, with MAPCAs
PERSPECTIVE
GOALS
Goals of Management

Achieve completely separated two ventricle circulation

Achieve lowest possible RV pressure
Importance of PA Pressure
HOW DOES ONE ACHIEVE THE LOWEST RV PRESSURE?
• lowest PA pressure is achieved only if as many lung segments as possible are included in repair, and repair happens before PVOD develops:

  The most complete and healthiest microvascular bed can only be achieved with complete unifocalization
TIMING
Observation #1
MAPCAs are intrinsically unstable after birth

- At birth
  - All lung segments have a blood supply (true PA or MAPCA)
  - MAPCAs tend to be smooth and sinusoidal
  - Optimal health of microvasculature exists

- Loss of lung segments occurs over time due to abnormal arterial flow and pressure
  - Natural occlusion / stenosis in some
  - Obstructive vascular disease in others
Observation #2

- In TOF/PA with MAPCAs, the native pulmonary arteries, if present, do not grow after birth
These two observations support early intervention:

- waiting causes degeneration of MAPCAs
- waiting causes involution of native PAs
HYPOTHESIS
• MAPCAs degenerate when left alone: Are they useless abnormal tissue, or can this tissue be utilized?

• Based on fetal and neonatal observations and certain physiologic principles, we hypothesized that MAPCA degeneration was “environmental” and not “intrinsic”

• Thus, we hypothesized that MAPCAs were *innocent bystanders*, that their degeneration was the result of being in an abnormal environment, and therefore, moving them to the low flow, low pressure environment of the pulmonary circulation would result in long term stable vessels
NATIVE PA ARBORIZATION
Spectrum of Lung Perfusion

- 18 segments supplied by MAPCAs
- 18 segments supplied by native PAs
Spectrum of Lung Perfusion

If no unifocalization, only 80% of lung is perfused. Survival, but not ideal PVR.
Spectrum of Lung Perfusion

What about here if no unifocalization? Survival? PVR?
Spectrum of Lung Perfusion

23% pts have NO native PAs
Spectrum of Lung Perfusion

If an individual case is here, no unifocalization is needed (only 12%)
RAW MATERIAL
• I don’t care if dominant tissue is native PA or collateral: all raw material

• 20% al pts have no PAs at all, 100% raw material is collaterals

• Outcomes excellent
ISOLATED vs DUAL SUPPLY
• All isolated supply collaterals are unifocalized
• Dual supply collaterals have a specific protocol
VSD MANAGEMENT
Intraoperative Flow Study

Monitor Mean PA pressure

Roller pump

Left atrium is vigorously vented
MANAGEMENT PROTOCOL
• COMPLETE UNIFOCALIZATION AT 3-4 MONTHS

• INTRA-OP FLOW STUDY

• IF PREDICTED RV/LV < 0.5 : VSD AND CONDUIT

• IF PREDICTED RV/LV > 0.5 : SHUNT
Our Prospectively Applied Treatment Protocol

TOF/PA

True PA

12%

Hypoplastic, Normal Arborization

Surgical AP Window

88%

Abnormal Arborization or Absent PAs

Midline Complete Unifocalization

Intraoperative Flow Study

Low PA Pressure

Simultaneous Intracardiac Repair

High PA Pressure

Shunt

Staged Intracardiac Repair
Spectrum of Lung Perfusion

If an individual case is here, no unifocalization is needed (only 12%)
SURGICAL TECHNIQUE
Tetralogy of Fallot with Major Aortopulmonary Collaterals

Technique
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DATA
Early Mortality
All Patients

- 1999 – 2015: 1.9%
Complete Repair

Actuarial % Completely Repaired

RV/LV Pressure Ratio < 0.5
RV/LV Pressure

0.35 +/- .12
Mid-Late Follow Up
RV / LV Pressure Ratio

• The RV / LV pressure ratio at follow up was compared to the perioperative value to determine PA and collateral growth.

Ratio difference = -0.03
CONCLUSIONS
What is the best way to treat MAPCAs?

Management plan should provide:

- "the greatest good for as many pts as possible"

- "greatest good" defined by highest % of septation, with lowest PA pressure, and lowest mortality, with durability of repair (long term low PA pressure)
Collaterals not intrinsically unstable but are innocent bystanders

Early removal from systemic circulation

Recruit vascular supply to all lung segments
The Pulmonary Artery is like an Oak Tree
Oak tree schematic
Pulmonary artery schematic
Actuarial Survival After Unifocalization

Mortality reduced in latter half of the experience
Pulmonary Atresia with Aortopulmonary Collaterals

“Natural History”

<table>
<thead>
<tr>
<th>YEAR OF LIFE</th>
<th>MORTALITY</th>
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<tbody>
<tr>
<td>1</td>
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<tr>
<td>10</td>
<td>60%</td>
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<tr>
<td>35</td>
<td>70%</td>
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Presentation and attrition in complex PA
Bull, J Am Coll Cardiol 1995; 25:491
MOST GOOD FOR THE GREATEST NUMBER OF PTS
Surgical Principles

- Emphasis on early complete repair
- Emphasis on tissue to tissue connections
- Recruit as many collaterals as possible initially
1990 – “unconquered” lesions
No rational approach to MAPCAs
Reasons:
- peripheral PAs not surgical
- black box misconception
- collateral durability misconception
Complex Congenital Pulmonary Artery Disease

• Comes in many forms, but the final common pathophysiologic pathway is some combination of cyanosis and pulmonary hypertension

• Quality of Life and Life Expectancy are severely impacted
Historical Context

- Congenital heart Disease (CHD) is largely structural in nature
- There are dozens to hundreds of congenital cardiac defects
- The field of reconstructive surgery for CHD is about 65 years old
- Each decade of this history is know for “conquering” one or more subsets of CHD, with simpler defects first and more complex ones later
- By 1990, most defects had been addressed with rational management plans and surgical reconstructive techniques, with the exception of patients with complex pulmonary artery defects
Evolution of Staged Repair

- Serial thoracotomies → repair
  Puga 1989, Laks 1994

- Central AP window → unifoc. → Repair
  Mee 1991

- Primary RVOT conduit
  Rome 1993
Rationale for Early Complete Repair

• At birth
  • All segments have a blood supply
  • Optimal accessibility to source of PBF
  • Optimal health of microvasculature

• Loss of Lung segments
  • Natural occlusion / stenosis
  • Pulmonary hypertension
  • Iatrogenic stenosis / occlusion of collaterals

Preempts natural history; avoids palliative complications; MAPCA as intrinsically pathologic or innocent bystander
OTHER APPLICATIONS
William’s Syndrome
Right lung AP
prior to surgery
William’s Syndrome
Right lung lateral
prior to surgery
William’s Syndrome
Left lung AP
prior to surgery
William’s Syndrome
Left lung lateral
prior to surgery
Our Prospectively Applied Treatment Protocol

**MAPCAs**

**TOF/PA**
- True PA
  - Hypoplastic, Normal Arborization
    - Surgical AP Window
  - Abnormal Arborization or Absent PAs
    - MAPCAs have segmental level stenosis
      - YES
        - Staged Thoracotomy Single Lung Unifocalization
          - Staged Intracardiac Repair
      - NO
        - Midline Complete Unifocalization
          - Intraoperative Flow Study
            - Low PA Pressure
              - Simultaneous Intracardiac Repair
            - High PA Pressure
              - Shunt
              - Staged Intracardiac Repair

**OTHER**
- Single Ventricle
- Two Ventricle