Late presentation of TGA – options

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No disclosures
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Surgery of choice for TGA.IVS

Primary arterial switch operation performed in the first few days after birth
Delayed surgery in TGA - Reasons

Industrialized countries:
- Concurrent non-cardiac medical / surgical problems

Non-industrialized countries:
- **Late diagnosis & referral**
  - Lack of awareness
  - Limitations of healthcare facilities
- Low birth weight
- Neonatal sepsis
- Financial constraints
LV involution in TGA.IVS

Closure of Ductus Arteriosus, ↓ PVR
↓
↓ LV afterload
↓
↓ LV pressure, LV deconditioning
↓
↓ LV mass
Regressed LV – 2D echo
Institutional report - Congenital

An electron microscopic study of left ventricular regression in children with transposition of great arteries

Akshay Kumar Bisoia, Dhananjay Malankara,*, Sandeep Chauhanb, Sambhunath Dasb, Ruma Rays, Prasenjit Dasc

Fig. 1. Normal Z bands (arrows) with normal nucleus (N) and glycogen granules (G). Magnification, 3500×.

Fig. 3. Z-band disruption with lots of fat vacuoles (arrows). Magnification, 2800×.
Figure 1: Serial changes in indexed left ventricular mass after BAS in simple TGA. LV: left ventricle; BAS: balloon atrial septostomy.
Delayed LV involution in TGA.IVS

- Delayed fall of high neonatal PVR
- Dynamic or resectable LVOTO
- Large PDA
- Large aorto-pulmonary collaterals
- ? Restrictive ASD / PFO

Allows for safe late primary ASO
Aorto-pulmonary collaterals in TGA.IVS

Primary ASO at 3 mths of age
Age related risk for primary ASO

CHSS data  JTCVS 1988
Identifying a regressed LV

Echo parameters:
- LV shape – banana shaped
- IVS – moving with RV rather than LV
- LV posterior wall thickness < 3.0 mm
- LV mass < 35 gm/M²
- Low LV end-diastolic volume

Hemodynamic parameters
- LV / RV pressure ratio < 0.7

Intra-operative response to trial PA banding

*None of these is foolproof!*
Surgical options with regressed LV

- Atrial switch – Senning / Mustard procedure
- Re-train LV and follow with arterial switch
  - Before ASO – 2 stage ASO
    - PA band +/- BT shunt
    - PDA stenting
  - After ASO – delayed primary ASO
    - Pharmacological
    - LVAD / ECMO
Staged arterial switch – Yacoub, 1977

Introduced concept of LV ‘re-training’ for late TGA

- PA band and then ASO after several weeks

Problems:

- Adhesions, technical difficulties
- Dilatation of pulmonary root, neo-aortic regurgitation
- Post-op LV dysfunction
- Became redundant with advent of neonatal switch
Rapid 2 stage ASO – Jonas et al, 1989

1st Stage: through mid-sternotomy

- Rt. modified B.T. Shunt
- PA band

*Interval phase ‘LV hypertrophy’ 7 – 10 days*

2nd Stage:

- Takedown band and shunt
- Arterial switch
Rapid 2 stage arterial switch
Serial echo for evaluation of LV
Rapid 2-stage arterial switch

Positives:
- Anatomic correction
- Technically easier than delayed 2 stage ASO
- Less neo-aortic valve / root distortion
- Single hospital stay
- Better long term results than atrial switch
Rapid 2-stage arterial switch

Negatives:

- Unreliable beyond infancy
- Longer ICU & hospital stay, increased costs & resource utilization
- Greater morbidity & mortality
- Long term performance of re-trained LV
Rapid 2 stage arterial switch

New Delhi experience – 1991 to 1996

Patient profile:

- Total No.: 32
- Age: 2 - 60 mths (median 4 mths)
- Weight: 3 - 11 kg. (median 4 kg)
- Diagnosis: IVS - 28, small VSD - 4
- Previous BAS: 26
Serial Echocardiography for Decision Making in Rapid Two-Stage Arterial Switch Operation

Krishna S. Iyer, MCh, Rajesh Sharma, MCh, Krishna Kumar, DM, Anil Bhan, MCh, Shyam S. Kothari, DM, Anita Saxena, DM, Panangipalli Venugopal, MCh

The Cardiothoracic Sciences Centre, All India Institute of Medical Sciences, New Delhi, India
Rapid 2 stage arterial switch

Results: n = 32

- **1st stage failure**: 5
  - Takedown & Sennings: 2, Mortality: 3
- **ASO completion**: 27
  - Takedown & Sennings: 1, Mortality: 2
- **Total mortality**: 5 (15%)
- **Successful ASO**: 24 (75%)
LV function after rapid 2 stage ASO

Paucity of data

- Many reports indicated subnormal LV function
  - ↓ LV contractility
  - ↑ LV end-diast volume, ↑ LV end-syst stress

- ↑ Neo-aortic regurg, ↑ RV outflow obstruction

Severity of LV dysfunction ≡

- rapidity of LV hypertrophy
- degree of LV dilatation

Colan et al. JTCVS, 1995, Laohaprasitiporn et al. 1998
2000 – 2011, 109 pts with TGA.IVS > 1 mth age

Gr A - 78 pts (1.9 mths), adequate LV – primary ASO

Gr B - 31 pts (6 mths), regressed LV – 2 stage ASO

Early mortality: 2.6% vs 9.7% NS

Late mortality: 2.7% vs 16% p = .01

Late Aortic regurgitation: 9.8% vs 33.3% p = .01

Age > 3 mths associated with late LV dysfunction

Delayed primary ASO
(with ECMO back-up)

Davis et al – JTCVS 1993, 106, 111-115
- 18 pts
- Age: 21 – 118 days
- 1 LV assist X 2 days
- 1 death – coronary insufficiency

Dabritz et al – EJCTS 1997, 11, 112-116
- 5 pts (1 - 3 mths age)
- Trial banding of PA for 30mts to assess LV
- Successful ASO in all
## Delayed primary ASO –
GOSH, London experience

<table>
<thead>
<tr>
<th></th>
<th>Late ASO</th>
<th>Early ASO</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients</td>
<td>37</td>
<td>156</td>
</tr>
<tr>
<td>Age range</td>
<td>21- 61 days</td>
<td>2 - 21 days</td>
</tr>
<tr>
<td>Mortality</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Early</td>
<td>2.7%</td>
<td>8.3%</td>
</tr>
<tr>
<td>Late</td>
<td>2.7%</td>
<td>0.6%</td>
</tr>
<tr>
<td>ECMO</td>
<td>2.7%</td>
<td>3.8%</td>
</tr>
</tbody>
</table>

Foran et al JACC 31,883-89,1998
Midterm Outcome of Primary Arterial Switch Operation Beyond Six Weeks of Life in Children With Transposition of Great Arteries and Intact Ventricular Septum
Akshay Kumar Biso, Tameem Ahmed, Dhananjay P. Malankar, Sandeep Chauhan, Shambunath Das, Pranav Sharma, Anita Saxena and Nagendra S. Boopathy
World Journal for Pediatric and Congenital Heart Surgery 2014 5: 219

Abstract
Background: We have previously reported our experience in primary arterial switch operation (ASO) in children more than six weeks with transposition of great arteries and intact ventricular septum. The age limit for performing an ASO in these children is not yet set. In this prospective observational study, we report the midterm outcome of 109 children aged more than six weeks who underwent primary ASO. Extracorporeal life support was performed in seven children and 102 survived. Methods: A total of 109 children aged more than six weeks with transposition of great arteries and intact ventricular septum underwent primary ASO. Methods: A total of 109 children aged more than six weeks with transposition of great arteries and intact ventricular septum underwent primary ASO. Extracorporeal life support was performed in seven children and 102 survived. Results: Two late deaths occurred, and survival in the remaining 107 children was found to have a decreasing trend. Mortality was 9.2%. Conclusions: Primary ASO can be safely performed in children with regressed ventricle, irrespective of age with encouraging results. The midterm results of these children are comparable in terms of survival and freedom from complications associated with preserved ventricle.

109 pts over 6 wks
Median age : 60 days
Mortality : 9.2%
ECMO requirement : 20%

Early mortality – 9.2%    Late mortality – 2.0%
The paradox of late presenting TGA

The problem of late presenting TGA is relevant to resource limited countries

All methods of LV retraining, and ECLS are resource consuming!!
Transposition of the Great Arteries—Outcomes and Time Interval of Early Neonatal Repair

Michael T. Cain, BS¹, Yumei Cao, PhD²,³,⁴, Nancy S. Ghanayem, MD⁴,⁵, Pippa M. Simpson, PhD²,³,⁴, Katie Trapp, BS⁴, Michael E. Mitchell, MD⁴,⁶, James S. Tweddell, MD⁴,⁶, and Ronald K. Woods, MD, PhD⁴,⁶

Table 5. Resource Utilization Across Study Groups.

<table>
<thead>
<tr>
<th>Resource Utilization</th>
<th>2 – 4 days</th>
<th>5 - 7 days</th>
<th>8 – 14 days</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Length of hospital stay, days, median (IQR)</td>
<td>15.5 (13.0, 20.0)</td>
<td>18.0 (16.0, 21.0)</td>
<td>23.5 (19.0, 32.0)</td>
<td>.005</td>
</tr>
<tr>
<td>Length of ICU stay, days, median (IQR)</td>
<td>7.0 (5.0, 9.0)</td>
<td>7.0 (6.0, 10.0)</td>
<td>8.0 (7.0, 14.0)</td>
<td>.056</td>
</tr>
<tr>
<td>Postoperative length of hospital stay, days, median (IQR)</td>
<td>12.0 (9.0, 16.0)</td>
<td>12.5 (9.0, 15.0)</td>
<td>12.5 (10.0, 25.0)</td>
<td>.86</td>
</tr>
<tr>
<td>Total hospital charges (median)</td>
<td>$128,219</td>
<td>$141,729</td>
<td>$217,427</td>
<td>.002</td>
</tr>
<tr>
<td>Total ICU charges (median)</td>
<td>$110,728</td>
<td>$120,497</td>
<td>$216,039</td>
<td>.0006</td>
</tr>
</tbody>
</table>

Abbreviations: IQR, interquartile range; ICU, intensive care unit.

³ P values from univariate analysis. See text for results of regression analysis.

‘No mortality risk’
An ASO in my hospital costs $5000 - $6000

Yet the majority cannot afford this!!
Delayed primary ASO (2007 – 09) FEHI study – Intent to avoid ECMO

<table>
<thead>
<tr>
<th>Variables</th>
<th>Early switch group</th>
<th>Late switch group</th>
</tr>
</thead>
<tbody>
<tr>
<td>N = 25</td>
<td></td>
<td>N = 22</td>
</tr>
<tr>
<td>Median age (days)</td>
<td>9 (3-18)</td>
<td>55 (22-149)</td>
</tr>
<tr>
<td>Sex M/F (%)</td>
<td>72 / 28</td>
<td>90.9 / 9.1</td>
</tr>
<tr>
<td>Median weight (kg)</td>
<td>2.84</td>
<td>3.05</td>
</tr>
<tr>
<td>Preop BAS (%)</td>
<td>52</td>
<td>31.8</td>
</tr>
<tr>
<td>Coronary anatomy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1LCx, 2R</td>
<td>17 (68%)</td>
<td>15 (68%)</td>
</tr>
<tr>
<td>1L, 2RCx</td>
<td>6 (24%)</td>
<td>5 (22.7%)</td>
</tr>
<tr>
<td>Single coronary</td>
<td>1 (4%)</td>
<td>2 (9.1%)</td>
</tr>
<tr>
<td>Other</td>
<td>1 (4%)</td>
<td>0 (0%)</td>
</tr>
</tbody>
</table>
Afterload reduction: Mainstay

- Phenoxybenzamine
- Milrinone
- Nitroglycerine
- Dobutamine

Low MAP but adequate systemic perfusion

Chang AC, Towbin JA, Heart Failure in children and young adults Saunders, Philadelphia, 2006
ICU Course After Late Primary switch at 50-100 days “Highly predictable”

0-2\textsuperscript{nd} POD
Unstable

3\textsuperscript{rd} POD
More stable

POD 4-5
Sternal closed

0
20
40
60
80
100
120
140
180
200
0 12 24 36 48 60 72 84 96 108 120 132 144
Hrs post op

MAP
HR
LAP

La 18mm

MAP 29mm

La 6-7mm

POD 6-7
Extubate to CPAP

Highly predictable
Multiple pharmacologic interventions...

- **Arrived in ICU:** Dobutamine, Milrinone, Phenoxybenz
- **Inj. Atracurium**
- **Volume, Inj. Calcium**
- **Sternum closed** (POD - 5)

- **LA surge: 16 mmHg** (POD - 0) - NTG, sedation
- **Stable LA: POD - 4** MAP ~50s
- **Nor epinephrine**
- **Steroids, Calcium**
- **Extubated to N-CPAP** POD 7

**Summary:**
- Dobutamine, Milrinone, Phenoxybenz
- Inj. Atracurium
- Volume, Inj. Calcium
- Sternum closed (POD - 5)
- LA surge: 16 mmHg (POD - 0) - NTG, sedation
- Stable LA: POD - 4 MAP ~50s
- Nor epinephrine
- Steroids, Calcium
- Extubated to N-CPAP POD 7
Regressed LV
Early post-op echo
5th post-op day
Results

◆ In-hospital mortality: comparable in both groups

Early switch group: 2 / 25 (8.0%)
( 1 PAH crisis, 1 coronary dissection )

Late switch group: 1 / 22 (4.54%)
( RV diverticulum – recurrent VT )

(p = 0.6)
Outcomes with primary ASO upto 3 months (2002-2017)  N = 239

<table>
<thead>
<tr>
<th>Age</th>
<th>&lt; 3 weeks</th>
<th>&gt; 3 weeks (median – 54 days)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mortality</td>
<td>11/138 (7.9%)</td>
<td>4/101 (4.0%)</td>
</tr>
<tr>
<td>ECLS</td>
<td>2/138 (1.4%)</td>
<td>1/101 (1.0%)</td>
</tr>
</tbody>
</table>

No deaths in last 4 years
Occult pre-op multidrug resistant Gr –ve sepsis
Preop CNS events – hypoxic encephalopathy
ASO – current status

- Widely utilized
- Low mortality in large volume centers
- All coronary patterns can be addressed
- Excellent short and intermediate term outcomes
- Long term outcomes yet unclear!
Long term concerns

- Aortic root dilatation / aortic regurgitation
  - TGA.VSD, 2 stage ASO, Taussig Bing

- Coronary artery stenosis / occlusion

- Coronary ostial stenosis

- LV dysfunction

- Pulmonary artery stenosis – post LeCompte

All more likely in retrained LV
Senning operation

- Physiologic repair, leaves RV as systemic ventricle
- Applicable to all ages at presentation, low risk
- Needs low intensity intensive care, economical
- Concerns over long-term outcome
  - Arrhythmias, RV function, baffle obstruction
- Remains relevant especially in emerging economies

Is it such a bad operation after all?
Long term outcome up to 30 years after the Mustard or Senning operation: a nationwide multicentre study in Belgium

P Moons, M Gewillig, T Sluysmans, H Verhaaren, P Viart, M Massin, B Suys, W Budts, A Pasquet, D De Wolf, A Vliers
Heart 2004;90:307–313.

339 patients - Mustard – 124  Senning – 225

Early mortality - 16.5 %

Late mortality - 7.7 %
Cumulative probability of survival

Log rank = 11.46; df = 3; p = 0.0095

<table>
<thead>
<tr>
<th>Years after Mustard or Senning operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.0</td>
</tr>
<tr>
<td>n</td>
</tr>
<tr>
<td>n</td>
</tr>
<tr>
<td>n</td>
</tr>
<tr>
<td>n</td>
</tr>
</tbody>
</table>

Simple Senning
Simple Mustard
Complex Mustard
Complex Senning
Results with Senning for TGA.IVS

1995 - 2017

N = 102

Age range: 3 wks - 10 yrs

Operative mortality: 4 / 102 (3.9%) (1.8% since 2002)

Causes:
- malnutrition
- DIC
- malnutrition – sepsis x 2
- sepsis
Offsetting late RV dysfunction

Senning with loose PA band

- Allows slow LV retraining
- Minimizes septal shift
- May delay onset of TR
- Allows for ASO conversion if systemic RV fails
Non-switchable coronaries
Our current policy

- < 3 mths – primary ASO irrespective of LV parameters
- > 6 mths – Senning procedure with PA band
- 3 – 6 mths ( decision based on resource availability )
  - delayed primary if LV appears favorable
  - ASO with ECMO back-up if LV unfavorable and resources permit
  - Senning with PA band
To summarize...

- Late presentation in TGA is an important problem in resource limited countries.
- Primary ASO appears feasible up to 3 months of age with low need for mechanical support.
- Beyond early infancy the Senning procedure compares favourably with 2 stage arterial switch operation.
- Long term outcomes of ASO when available will help in our decision making.
Thank you for your attention!

The Peacock – National bird of India