The Arterial Switch Operation
Technique, Guidelines, European Results

George E. Sarris, MD, PhD
FACC, FACS, FAHA, FETCS, FCTS
Chief, Athens Heart Surgery Institute
Dept of Pediatric Cardiac Surgery
Iaso Children’s Hospital
Athens, Greece
The Jatene operation (ASO) for TGA – VSD (1975)

“Our experience was always based on anatomic studies in our laboratory of pathologic anatomy. As it relates to TGA, there were 62 specimens in the laboratory, together with other congenital cardiac lesions, that I frequently examined. After reviewing the literature on TGA, I became convinced that the problem could be solved if the coronary arteries could be transposed…”

“The first child died at the end of the operation, but that case convinced me that the operation was feasible”.

“I performed the second operation on a 40-day-old child with TGA and VSD who had severe pulmonary congestion. The child did very well.”

“Unfortunately, the other 5 cases that I subsequently performed all died because of postoperative problems. Once these postoperative problems were corrected, better results were achieved.”
Development of ASO

• Initially, most infants with TGA – IVS did not survive ASO
  – low-pressure LV not prepared for systemic pressure work

• Two stage ASO \((Yacoub \text{ et al, } \text{Lancet, 1977})\):
  – 1st stage: PAB + shunt
  – 2\textsuperscript{nd} stage: ASO

• Castaneda and Norwood: routinely successful primary neonatal ASO in TGA-IVS
ASO Technique
mostly standardized, but with critical variations
Identify coronary ostia, take coronary buttons
Take note of coronary transfer sites
MPA transection
medially based trapdoor flaps planned
Coronary transfer
(N.B. may be facilitated by performing posterior aortic anastomosis first)
LeCompte Manoeuver
and aortic anastomosis
Neo-MPA reconstruction
“pantaloon patch”
Neo MPA anastomosis
Completed operation
Technically challenging, steep learning curve
mortality risk related to experience

<table>
<thead>
<tr>
<th>No. of arterial switch repairs per institution</th>
<th>Total patients</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.</td>
</tr>
<tr>
<td>≤ 10</td>
<td>20</td>
</tr>
<tr>
<td>10–20</td>
<td>59</td>
</tr>
<tr>
<td>20–30</td>
<td>22</td>
</tr>
<tr>
<td>30–40</td>
<td>—</td>
</tr>
<tr>
<td>40–50</td>
<td>—</td>
</tr>
<tr>
<td>50</td>
<td>111</td>
</tr>
<tr>
<td>Total</td>
<td>212</td>
</tr>
</tbody>
</table>


CL, confidence limit.

* One of the nine institutions performed five operations, with one death (20%; CL = 7–93%); these were considered low-risk institutions.

† One of the four institutions performed 15 operations, with two deaths (13%; CL = 4–29%) and was considered a low-risk institution.
ASO for the TGA “family” of congenital heart defects

TGA family

- Discordant VA Connection
  - Univentricular AV connection
    - Ao from RV + SAS, AAO
  - Concordant AV connection
    - TGA IVS
    - TGA VSD (+) LVOTO
  - Discordant AV connection
    - cc TGA
    - Cc TGA VSD (+) LVOTO

- DORV
  - Taussig - Bing (sub-pulm VSD)
Impact of ASO on pediatric cardiac surgery

- Other neonatal operations
  - ALCAPA
  - Ross, Ross-Konno
  - Norwood
- Complex cono-truncal repairs
  - Nikaido
  - Double root translocation
- Double switch for ccTGA
- Small body technology
  - Premature & LBW infants
- ICU, Nursing, Benchmarking
  - LV physiology, afterload reduction, mechanical support
Rational for TGA Guidelines

• The ASO
  – has become the standard operative technique for most patients with TGA
  – is performed widely, generally with low mortality

• But
  – Wide variability in management with several controversial issues, from fetal diagnosis to long-term follow up
  – Wide variability of results
Joint EACTS and AEPC TGA Guidelines Taskforce

George E. Sarris * (Chairperson) (Greece)
Christian Balmer (Switzerland)
Pipina Bonou (Greece),
Juan V. Comas, (Spain)
Eduardo DaCruz (USA),
Luca Di Chiara (Saudi Arabia),
Roberto M. Di Donato (Saudi Arabia),
Jose Fragata (Portugal),
Tuula Eero Jokinen (Finland),
George Kirvassilis (USA),
Irene Lytrivi (USA),
Milan Milojevic (Netherlands),
Gurleen Sharland (UK),
Matthias Siepe (Germany),
Joerg Stein (Austria),
Emanuela Valsangiacomo Büchel (Switzerland),
Pascal R. Vouhe, (France).

Bringing together the specialties of the “Congenital Heart Team”
Cardiology, Anesthesia, Intensive Care and Surgery
Clinical guidelines for the management of patients with transposition of the great arteries with intact ventricular septum

The Task Force on Transposition of the Great Arteries of the European Association for Cardio-Thoracic Surgery (EACTS) and the Association for European Paediatric and Congenital Cardiology (AEPC)

Authors/Task Force Members: George E. Sarris (Chairperson) (Greece), Christian Balmer (Switzerland), Pipina Bonou (Greece), Juan V. Comas (Spain), Eduardo da Cruz (USA), Luca Di Chiara (Italy), Roberto M. Di Donato (United Arab Emirates), José Fragata (Portugal), Tuula Eero Jokinen (Finland), George Kirvasilis (USA), Irene Lytrivi (USA), Milan Miljevic (Netherlands), Gurleen Sherland (UK), Matthias Sepe (Germany), Joerg Stein (Austria), Emanuela Valangiacomo Büchel (Switzerland) and Pascal R. Vouhé (France)
Guidelines Methodology

• defining specific **issues of interest**
• reviewing the **pertinent literature**
• evaluating the **strength of evidence**
• creating **clinical practice guidelines**
Exclusions

- TGA with aortic coarctation or arch hypoplasia
- TGA with ventricular septal defect (VSD) with or without left ventricular outflow tract (LVOT) obstruction
- TGA or malposition of the great arteries associated with double-outlet right ventricle or in anatomical or functionally univentricular hearts
- Congenitally corrected transposition.
Extensive literature search

• More than 400 peer reviewed articles were selected and analyzed by **TGA Guidelines Task Force**
• **Practice recommendations** were formulated
• To each, the appropriate **Recommendation Class & Level of Evidence** was assigned
Recommendations & Evidence - glossary

**Class of Recommendation**

- **I**: Intervention is useful and effective
- **IIa**: Weight of evidence/opinion is in favor of usefulness/efficacy
- **IIb**: Usefulness/efficacy is less well established by evidence/opinion
- **III**: Intervention is not useful/effective and may be harmful

**Level of Evidence**

- **A**: Data derived from multiple randomized clinical trials or meta-analyses
- **B**: Data derived from a single randomized clinical trial or large non-randomized studies
- **C**: Consensus of opinion of the experts and/or small studies, retrospective studies, registries

Topics Examined

- Prenatal detection
- Prenatal and delivery management
- Postnatal diagnosis
- NICU management, Management of “poor mixer”
- Timing for primary arterial switch
- LV Training
- Two-stage arterial switch
- Surgical techniques – intraoperative surgical management
- Anesthesia and CPB management
- ICU management (pre- and postoperatively)
- Role of Atrial switch, LV retraining
- Long-term complications, their prevention, and management

In this presentation, recommendations for

1. Timing of ASO
2. LV training
3. Surgical technique
4. When to recommend “Atrial switch”
5. RVOTO after ASO
6. Reoperations for residual or recurrent coronary lesions
## Recommendations on Timing for ASO

<table>
<thead>
<tr>
<th>Recommendation</th>
<th>Class</th>
<th>Level</th>
<th>Ref</th>
</tr>
</thead>
<tbody>
<tr>
<td>It is recommended that a primary ASO in neonates with TGA IVS be performed from the first few days to 3 weeks of life</td>
<td>I</td>
<td>B</td>
<td>90–95</td>
</tr>
<tr>
<td>A primary ASO should be considered up until 60 days (with ECLS back-up)</td>
<td>IIa</td>
<td>B</td>
<td>7, 97–100</td>
</tr>
</tbody>
</table>

1. Primary ASO in neonates with TGV - IVS to be performed from first few days to 3 weeks
2. Primary ASO should be considered up until 60 days with ECLS back-up – IIa (B)
RECOMMENDATIONS:
LV retraining

Ventricular septal profile: alignment or L-R bulging on 2D echocardiogram is a good indicator for adequate LV training
Coronary Transfer - TGA

Normal / near normal patterns

Type A: RCA#2; CX, LAD#1
Type D: RCA, CX#2; LAD#1
Type B: LAD, CX, RCA #1
Type C: LAD, CX, RCA #2
Type E: LAD, RCA #1; CX#2

ALL PATTERNS TRANSFERABLE
But...
abnormal coronary origin traditionally was a risk factor for early and late poor outcomes
### Surgical Technique & Coronary Issues

- Although all coronary patterns in TGA are “transferable” some (such as double looping, side to side vessels and single ostium) require special surgical care (I-B)

- Large coronary buttons and adequate mobilization of proximal coronaries is recommended (I – C)

- For posterior looping (D pattern) ostium implantation should be above vessel’s suture line (I - C)

- Intra-mural course to be best addressed by unroofing & individual transfer (I-C)

- For dominantly anterior-posterior vessel patterns - Lecompte manoeuvre is indicated (I – C)
Posterior looping coronary implanted high
Juxta-commissural ostium
Ostia very close
Intramural coronary course
Juxta-commissural ostium: detach valve
Ostia very close: separate if possible, else, treat as single coronary
Intramural coronary course: unroof
Single coronary: modified Yacoub technique

FIGURE 20.6 In the extremely rare instance of a single coronary artery running between the pulmonary artery and aorta the button is rotated through 90° and roofed with pericardium.
Coronary extension
Side – by – side great vessels
When will Atrial Switch be recommendable?

<table>
<thead>
<tr>
<th>Recommendation</th>
<th>Class</th>
<th>Level</th>
<th>Ref</th>
</tr>
</thead>
<tbody>
<tr>
<td>An atrial switch should be considered in the following cases:</td>
<td>IIA</td>
<td>C</td>
<td>289, 290</td>
</tr>
<tr>
<td>Complex coronary transfer or non-facing sinus coronary origin from the non-facing sinus</td>
<td>IIA</td>
<td>C</td>
<td>291</td>
</tr>
<tr>
<td>Late referral, as an alternative to a two-stage ASO, especially in older infants and children</td>
<td>IIA</td>
<td>C</td>
<td>291</td>
</tr>
<tr>
<td>In developing countries where there is no setup for complex neonatal surgery</td>
<td>IIA</td>
<td>C</td>
<td>291</td>
</tr>
<tr>
<td>In TGA IVS with persistent pulmonary hypertension</td>
<td>IIA</td>
<td>C</td>
<td>68</td>
</tr>
</tbody>
</table>

Only Class IIA ...

- Complex coronary transfer or non facing sinus coronary origin
- Late referral..., as alternative to two stage repair in children
- Developing countries with no set-up for ASO
- TGA – IVS and persistant PHT
## Recommendations on RVOTO later after ASO

<table>
<thead>
<tr>
<th>Recommendation</th>
<th>Classa</th>
<th>Levelb</th>
<th>Refc</th>
</tr>
</thead>
<tbody>
<tr>
<td>Indications for reintervention for RVOT obstruction</td>
<td>IIa</td>
<td>B</td>
<td>293</td>
</tr>
<tr>
<td>Reoperation or intervention should be considered if evidence of significant</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>obstruction (gradient &gt; 50 mmHg) is detected during routine echo Doppler</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>evaluation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Suggested treatment</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Surgical patch angioplasty is the most effective treatment and should be</td>
<td>IIa</td>
<td>C</td>
<td>320,</td>
</tr>
<tr>
<td>considered as the treatment of choice</td>
<td></td>
<td></td>
<td>327</td>
</tr>
<tr>
<td>Balloon dilation of pulmonary stenoses</td>
<td>IIa</td>
<td>C</td>
<td></td>
</tr>
<tr>
<td>should be considered only if obstruction is confined to the valvular level</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Catheter reintervention is not indicated for supravalvular pulmonary stenosis</td>
<td>III</td>
<td>C</td>
<td></td>
</tr>
<tr>
<td>Stenting of branch pulmonary artery (bifurcation) stenoses may be considered</td>
<td>IIb</td>
<td>C</td>
<td></td>
</tr>
</tbody>
</table>

### Only Class IIa ...

- **Intervention should be considered for ECHO gradients** RV-PA > 50 mm Hg
- **Surgical patch angioplasty should be considered as best option**
- **Ballooning should be considered only for obstructions at valve level**
- **Stenting may be considered for branch, or bifurcation PA stenosis** - IIb
Reoperation for residual or recurrent coronary lesions

If there are ECG or ECHO signs suggesting ischemia, diagnostic work-up should involve CT, MRI, Angio, and Perfusion scan – thallium are indicated.

Reoperation is indicated when diagnosis of ischemia is established and a lesion is documented.

Preferable surgical technique for discrete proximal obstruction is surgical coronary-ostial patch angioplasty.
Limitations

• Guidelines and recommendations should help physicians to make decisions in their daily practice.

• However, Guidelines are no substitute for primary literature sources and particularly for clinical evaluation and judgment.

• The ultimate specific decisions regarding the care of an individual patient must be made by his, or her, responsible physician(s).
“Science tells us what we can do; Guidelines what we should do; & Registries what we are actually doing.”

Lukas Kappenberger MD
Heart Rhythm Society Policy Conference
Washington DC 2005
“Without data you’re just another person with an opinion.”

- W. Edwards Deming, Data Scientist
# Mortality vs. Procedure

## Data Filter Options

Data Selection Inactive

## Results

<table>
<thead>
<tr>
<th></th>
<th>Primary Procedure Name</th>
<th>No of operations</th>
<th>30-day Deaths</th>
<th>30-day Mortality</th>
<th>Hospital Deaths</th>
<th>Hospital Mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>VSD repair, Patch</td>
<td>36403</td>
<td>253</td>
<td>0.70%</td>
<td>300</td>
<td>0.83%</td>
</tr>
<tr>
<td>2</td>
<td>ASD repair, Patch</td>
<td>15891</td>
<td>49</td>
<td>0.31%</td>
<td>57</td>
<td>0.36%</td>
</tr>
<tr>
<td>3</td>
<td>PDA closure, Surgical</td>
<td>9834</td>
<td>234</td>
<td>2.39%</td>
<td>320</td>
<td>3.27%</td>
</tr>
<tr>
<td>4</td>
<td>TOF repair, Ventriculotomy, Transanular patch</td>
<td>8637</td>
<td>201</td>
<td>2.33%</td>
<td>219</td>
<td>2.54%</td>
</tr>
<tr>
<td>5</td>
<td>Shunt, Systemic to pulmonary, Modified Bialock-Taussig Shunt (MBTS)</td>
<td>7815</td>
<td>620</td>
<td>8.52%</td>
<td>725</td>
<td>9.96%</td>
</tr>
<tr>
<td>6</td>
<td>Arterial switch operation (ASO)</td>
<td>7372</td>
<td>370</td>
<td>5.03%</td>
<td>417</td>
<td>5.67%</td>
</tr>
<tr>
<td>7</td>
<td>AVC (AVSD) repair, Complete (CAVSD)</td>
<td>7356</td>
<td>301</td>
<td>4.11%</td>
<td>369</td>
<td>5.03%</td>
</tr>
<tr>
<td>8</td>
<td>ASD repair, Primary closure</td>
<td>7291</td>
<td>36</td>
<td>0.49%</td>
<td>40</td>
<td>0.55%</td>
</tr>
</tbody>
</table>
**TRENDS**

**DATA FILTER OPTIONS**

Primary Procedures Only:
- True

Procedures:
- (10,05) Arterial switch operation (ASO)

Patients: 7357
Operations: 7372

**RESULTS**
ASO, IVS - ASO + VSD – Neonates
1999 - 2015

N = 5543, 5.12%
N = 1698, 9.01%
p < 0.001

chi-square
TGA, IVS - ASO – Neonates

1999 – 2008
N = 2597, 5.97%

2009 - 2015
N = 2946, 4.38%
p = 0.007

chi-square
TGA, VSD - ASO + VSD – Neonates

1999 – 2008
N = 796, 8.79%

2009 - 2015
N = 902, 9.20%
p = NS

chi-square
Major complications as in STS Morbidity Score

1. Acute renal failure, requiring temporary or permanent dialysis
2. Neurological deficiency persisting at discharge
3. AV – block requiring a permanent pacemaker
4. Requirement of mechanical circulatory support
5. Phrenic nerve injury/paralysed diaphragm
6. Unplanned re-operation
TGA, IVS - ASO - major complications

1999 – 2008

- N = 2389
- 8.01%

2009 - 2015

- N = 2707
- 8.11%
- p = NS

chi-square
TGA, VSD - ASO + VSD - major complications

1999 – 2008

N = 688
13.57%

2009 – 2015

N = 769
14.75%
p = NS

chi-square
Summary of Results for ASO in Europe

1. ASO is associated with lower hospital mortality and morbidity than ASO + VSD repair

2. Hospital mortality after ASO decreased between the two study periods, but did not decrease after ASO + VSD repair and remains significantly higher

3. ASO + VSD repair is associated with higher frequency of postoperative complications without difference between the two study periods
The ASO for TGA

Future directions
Left-Sided Reoperations After Arterial Switch Operation: A European Multicenter Study

Vladimiro L. Vida, MD, PhD, Lorenza Zanotto, MD, Lucia Zanotto, PhD, and Giovanni Stellin, MD, and the European Congenital Heart Surgeons Association (ECHSA) Study Group*

Pediatric and Congenital Cardiac Surgery Unit, Department of Cardiac, Thoracic, and Vascular Sciences, and Department of Statistical Sciences, University of Padua, Padua, Italy

Background. We sought to report the frequency, types, and outcomes of left-sided reoperations (LSRs) after an arterial switch operation (ASO) for patients with D-transposition of the great arteries (D-TGA) and double-outlet right ventricle (DORV) TGA-type.

Methods. Seventeen centers belonging to the European Congenital Heart Surgeons Association (ECHSA) contributed to data collection. We included 111 patients who underwent LSRS after 7,951 ASOs (1.4%) between January 1975 and December 2010. Original diagnoses included D-TGA (n = 99) and DORV TGA-type (n = 12). Main indications for LSR were necrotic valve insufficiency (n = 52 [47%]) and coronary artery problems (CAPs) (n = 21 [19%]).

Results. Median age at reoperation was 8.2 years (interquartile range [IQR], 2.9–14 years). Seven patients died early after LSRS (6.3%): 4 patients with D-TGA (6.5%) and 3 patients with DORV TGA-type (25%) (p = 0.02). Median age at last follow-up was 16.1 years (IQR, 9.9–27.8 years). Seventeen patients (16%) required another reoperation, which was more frequent in patients with DORV-TGA type (4 of 9 [44%]) than in patients with D-TGA (13 of 95 [14%]). Late death occurred in 4 patients (4 of 104 [3.8%]). The majority of survivors were asymptomatic at last clinical examination (64 of 100 [64%]).

Conclusions. Reoperations for residual LSRS are infrequent but may become necessary late after an ASO, predominantly for necrotic valve insufficiency and CAPs. Risk at reoperation is not negligible, and DORV TGA-type anatomy, as well as procedures on the coronary arteries, were significantly associated with a higher morbidity and a lower overall survival. Recurrent reoperations after LSRS may be required.
Coronary Lesions after ASO in patients with TGA. Do we need an interventional follow up in asymptomatic patients?

An ECHSA Multi-Institutional Study

Eleftherios Protopapas (eprotopapas@gmail.com)

George Sarris
Conclusions

• The ASO, the procedure of choice is widely performed in Europe for most patients with TGA, has been refined over the years and is generally associated with very good results.

• Recently published guidelines provide evidence-based guidance for managing patients with TGA, especially regarding persistently controversial issues.

• Significant early mortality and late morbidity and complications persist for many patients with TGA, leaving room for research-based improvements.

• On-going multi-institutional studies, such as those organized by ECHSA, will hopefully lead to new knowledge and improved outcomes.
Thank you!