ΤΙ ΝΕΟΤΕΡΟ ΣΤΗΝ ΧΕΙΡΟΥΡΓΙΚΗ ΤΩΝ ΣΥΓΓΕΝΩΝ ΚΑΡΔΙΟΠΑΘΕΙΩΝ

ΦΩΤΙΟΣ Α. ΜΗΤΡΌΠΟΥΛΟΣ, MD, PHD
ΑΝΑΠΛΗΡΩΤΗΣ ΔΙΕΥΘΥΝΤΗΣ
ΚΑΡΔΙΟΧΕΙΡΟΥΡΓΙΚΗ ΚΛΙΝΙΚΗ ΠΑΙΔΩΝ ΚΑΙ ΣΥΓΓΕΝΩΝ ΚΑΡΔΙΟΠΑΘΕΙΩΝ
Hypoplastic Left Heart Syndrome

- HLHS: one of the most severe form of CHD
  - High morbidity and mortality
- Incidence: 1-2 % of all CHD
- Involved obstruction at multiple level of left heart structures.
  - Mitral stenosis to mitral atresia
  - Variable degree of LV hypoplasia
  - Aortic stenosis to aortic atresia
  - Variable degree of ascending aorta hypoplasia
HYPOPLASTIC LEFT HEART SYNDROME

Hybrid procedure VS Modified Norwood’s operation
HYPOPLASTIC LEFT HEART SYNDROME

Modified Norwood’s operation

• Aortic and arch reconstruction with/without homograft
• Atrial septectomy
• Shunt to pulmonary artery
  BT shunt or Sano shunt
• Norwood Procedure
• The first in a series of surgeries for single ventricle heart defects.
• 2013 - 2016 total procedures: 112

<table>
<thead>
<tr>
<th>Year</th>
<th>Total</th>
<th>Died</th>
</tr>
</thead>
<tbody>
<tr>
<td>2013</td>
<td>28</td>
<td>4</td>
</tr>
<tr>
<td>2014</td>
<td>28</td>
<td>7</td>
</tr>
<tr>
<td>2015</td>
<td>30</td>
<td>5</td>
</tr>
<tr>
<td>2016</td>
<td>26</td>
<td>2</td>
</tr>
</tbody>
</table>

• CHOP mortality rate: 17.0%
• STS mortality rate: 15.8%
Atrial septum ballooned or stented
Stent in ductus arteriosus
Bilateral pulmonary artery bands
Fifteen-year Single Center Experience with the “Giessen Hybrid” Approach for Hypoplastic Left Heart and Variants: Current Strategies and Outcomes


Received: 6 May 2012 Accepted: 22 August 2012 Published online: 3 September 2012 © For Author(s) 2012. This article is published with open access at Springerlink.com

Abstract. Presented is a retrospective outcome study of a 15-year single institutional experience with a contemporary cohort of patients with hypoplastic left heart syndrome and complex congenital heart disease (CHD) who underwent a “Giessen Hybrid” stage I palliation approach. Hybrid approach consists of surgical bilateral pulmonary artery banding and pericardial ductal exclusion, and/or atrial septal manipulation. A no-suture approach was developed from a trans-aortic approach at a first-line procedure. Pre-operative Atrial septum defect (ASD) and patent foramen ovale (PFO) were rare conditions. Fifteen-year follow-up mortality is reported as occurring within the staged univentricular palliation or before and after biventricular repair. Hybrid stage I was performed in 134 patients; 107 were treated by single ventricle palliation, 57 by biventricular repair (BVR), 7 received heart transplantation, and 7 were treated by surgery care, respectively. Overall, 34 children died. The Atrial septum defect and ASD are less frequent in our outcome. Two patients died during stage I (1.2 %), and the interstage I mortality was 6.7 %, and stage II mortality 9 % respectively. Stage III was up to now performed in 57 patients without mortality. At 1 year, overall survival in stage I palliation was 84 %; survival in stage II was 80 %; and survival in stage III was 77 %, with no significant impact of body weight below 2.5 kg. In conclusion, hybrid stage I fulfilled the criteria of life-saving approach. In our institution, Hybrid procedure replaced non-operative palliation with a considerable mid- and long-term survival rate. Considering interstage mortality, close surveillance is mandatory.

Keywords: Hypoplastic left heart syndrome - Hypoplastic left heart complex - Hybrid approach

Introduction

Hybrid approach expands the surgical options for patients born with hypoplastic left heart syndrome (HLHS) and newborns with multiple left heart obstructions, summarized as hypoplastic left heart complex (HLHC). Despite significant progress, surgical outcome for high-risk patients with HLHS remains suboptimal [3, 8, 11, 17, 20, 22, 29]. The hybrid palliation preserves the initial operative risk [1] and is hypothesized to improve in particular neurosurgical survival [16]; however, the outcome of this sequential approach is unknown. Improved operative outcomes have resulted in the increasing use of the surgical palliation in high-risk patients, but at the expense of considerable mortality and morbidity. [5, 6]. The early success with the hybrid approach reported by Akınobůrk et al. [1, 2] and Galawadoc et al. [9, 16] have prompted the increasing use of this strategy in order to minimize the deleterious impact of the conventional surgical intervention on high-risk patients [4, 8, 14, 26]. Moreover, aggravating factors as low birth weight and surgical complexity on survival supported the idea that a less invasive non-surgical procedure could improve the outcome in these patients [11, 22]. Therefore, we present the overall single institutional 15-year experience with an unselected cohort of HLHS and HLHC patients who underwent a hybrid sequence as initial palliation providing their mid- and long-term outcomes.
Transposition of the great arteries (TGA)

- TGA approximately 5% of all congenital heart disease
- CCTGA in approx. 0.6-1.4%

Classification
- D-TGA
  1. Simple TGA (isolated TGA): with intact ventricular septum
  2. Complex TGA : with VSD or other defects
Complete Transposition of the Great Arteries

- Ventriculoarterial discordance
- Also known as d-TGA (d = dextroposition of the bulboventricular loop)
- Aorta on the right and anterior
Arterial switch procedure

Complications: coronary stenoses with sudden death or MI, RV outflow tract distortion, dilatation of neo-aortic root with AI

Jatene 1976
Abstract

OBJECTIVES
Long-term outcomes after the arterial switch operation (ASO) for complex transposition of the great arteries (TGA) should be clarified.

METHODS
A retrospective study was conducted in patients operated on between 1982 and 1998. Overall 220 postoperative survivors, 79.1% with a ventricular septal defect, 13.2% with multiple ventricular septal defects, and 29.1% with aortic arch obstruction, were followed for 17 years (0–28 years).

RESULTS
The conditional survival rate was 96.7% [95% confidence interval (CI): 94.4–99.1] at 25 years. Late sudden death occurred in 2 asymptomatic patients. The cumulative incidence rate of death or reinterventions was 3.8% (95% CI: 2.9–4.8) at 25 years, with age at ASO <10 days and aortic regurgitation at discharge identified as independent risk factors. The cumulative incidence rate of neoaortic regurgitation was 41.6% (95% CI: 20.5–62.8) at 25 years with an aorto-pulmonary diameter mismatch at the time of the ASO, age at ASO <10 days and aortic regurgitation at discharge identified as independent risk factors. At the last follow-up, 53 patients (24.1%) had neoaortic root dilatation with an aortic sinus z-score ≥3 and 6 of them had a Bentall operation at a median delay of 14.1 years since the ASO. The only independent factors for neoaortic root dilatation were male sex and an aorto-pulmonary diameter mismatch at the time of the ASO.
From: Long-term outcomes of the arterial switch operation for transposition of the great arteries and ventricular septal defect and/or aortic arch obstruction
Interact CardioVasc Thorac Surg | © The Author 2016. Published by Oxford University Press on behalf of the European Association for Cardio-Thoracic Surgery. All rights reserved.
TRUNCUS ARTERIOSUS

Morphogenesis

• Truncus arteriosus is called also as persistent truncus arteriosus, truncus arteriosus communis, common aorticopulmonary trunk

• Chromosomal 22q11 deletion is present in a substantial number of patients with conotruncal abnormalities (about one third with truncus arteriosus)

• Many of these have additional characteristic features of DiGeorge syndrome, velocardiofacial syndrome, or conotruncal face syndrome

• As such, their natural history may be complicated by hypocalcemia, palatal abnormalities, learning disability, or other noncardiac problems.
TRUNCUS ARTERIOSUS

Pathophysiology

- A single common artery, or truncus, overlying the ventricular septum and a nonrestrictive VSD gives rise to the coronary arteries, pulmonary arteries, and ascending aorta.
- Complete mixing of systemic and pulmonary venous return at the VSD and truncal valve level results in moderate cyanosis.
- As the pulmonary vascular resistance decreases after birth, significant left-to-right shunting at the truncal valve level leads to excessive pulmonary blood flow, pulmonary hypertension, and congestive heart failure.
OPERATIVE TECHNIQUES

1. Repair with allograft valved conduit

2. Repair truncus I,II with autologous tissue
   - Barbero-Marcial technique

3. Repair of hemitruncus
   - Unifocalization of pulmonary artery

4. Repair of truncus arteriosus with IAA
Outcomes of Truncus Arteriosus Repair in Children: 35 Years of Experience From a Single Institution.
Naimo PS¹, Fricke TA¹, Yong MS², d'Udekem Y³, Kelly A⁴, Radford DJ⁵, Bullock A⁶, Weintraub RG³, Brizard CP³, Konstantinov IE⁷.

Abstract
We evaluated the long-term outcomes following repair of truncus arteriosus (TA) from a single institution. We conducted a retrospective review of children (n = 171) who underwent TA repair between 1979 and 2014. Early mortality rate was 11.7% (20/171). There were 19 late deaths. Most deaths (74%, 29/39) occurred within the first year following surgery. The 1-year mortality rate in 1979-2004 was 18% (25/136) and decreased to 11% (4/35) in 2005-2014. The overall survival rate was 73.6% at 30 years. Multivariate analysis identified postoperative extracorporeal membrane oxygenation (P = 0.003), operative weight <2.5kg (P = 0.012), prior surgical intervention (P = 0.018), and coronary artery anomaly (P = 0.037) as risk factors for early mortality. A Cox regression model identified DiGeorge syndrome (P = 0.008) as a risk factor for late mortality. Freedom from right ventricular outflow tract reoperation was 4.6% at 20 years. Concomitant truncal valve (TV) repair or replacement was undertaken in 20 patients. Additionally, 14 patients underwent late TV repair or replacement. The overall survival rate in patients who underwent TV operation was 76.9% at 20 years. A total of 19 patients had concomitant interrupted aortic arch with a survival rate of 89.5% at 20 years. Median follow-up was 19 years (mean = 17 years, range: 1-34 years). All patients were in New York Heart Association Class I/II at last follow-up. Following repair of TA, patients had good long-term functional status but had high reoperation rates. Repair of interrupted aortic arch and TV were not risk factors for mortality. Postoperative extracorporeal membrane oxygenation, operative weight <2.5kg, prior surgical intervention, and coronary artery anomaly were risk factors for early death. DiGeorge syndrome was associated with late death, most commonly from infection.
FREEDOM FROM REOPERATION AFTER TA REPAIR
TETRALOGY OF FALLOT

- 4 anatomic malformations:
  - Right Ventricular Hypertrophy
  - Pulmonary Valve Stenosis
  - Transposition of the aorta
  - Ventricular Septal Defect
Thomas-Blalock-Taussig Shunt

Vivien Thomas, Partners of the Heart, 1998 and Something the Lord Made - Best Made-for-TV Movie, 2004
Surgical Intervention

- Complete intracardiac repair of VSD and PA stenosis in the first year of life.
- Repair the VSD with a patch.
- Relief of RVOT obstruction
- In selected cases preservation of the pulmonic valve
- Tetralogy of Fallot (TOF) Repair
- To repair tetralogy of Fallot (TOF).
- 2013 - 2016 total procedures: 147
- Total: 147
- Died: 2
- CHOP mortality rate: 1.4%
- STS mortality rate: 1.1%

<table>
<thead>
<tr>
<th>Year</th>
<th>Total</th>
<th>Died</th>
</tr>
</thead>
<tbody>
<tr>
<td>2013</td>
<td>35</td>
<td>0</td>
</tr>
<tr>
<td>2014</td>
<td>38</td>
<td>1</td>
</tr>
<tr>
<td>2015</td>
<td>24</td>
<td>0</td>
</tr>
<tr>
<td>2016</td>
<td>50</td>
<td>1</td>
</tr>
</tbody>
</table>
TETRALOGY OF FALLOT AFTER REPAIR

• Severe PV regurgitation in operated patients with TOF may lead to right heart failure with:
  
  ↑ incidence of sudden death

  ↓ Exercise tolerance

  Ventricular arrhythmias

• Timely PV replacement is necessary in order to prevent the development of these complications

Gatzoulis et al, Lancet 2000
Davlouros et al, Int J cardiol2004
Introduction: Development of pulmonary insufficiency in patients with surgically corrected tetralogy of Fallot (TOF) may lead to severe right heart failure with serious consequences. We herein present our experience with pulmonary valve replacement (PVR) in these patients. Methods: From 2005-2013, 99 consecutive patients (71 males/28 females, mean age 38±8 years), underwent PVR after 7 to 40 (mean 29 ± 8) years from the initial correction. Seventy nine of the symptomatic patients presented in NYHA II, 14 in III and 2 in IV. All underwent PVR with a stented bioprosthesis valve, employing a beating heart technique with normothermic extracorporeal circulation support. Concomitant procedures included resection of aneurysmal outflow tract patches (n = 37), tricuspid valve annuloplasty (n = 36), augmentation of stenotic pulmonary arteries (n = 9), maze procedure (n = 2) and pulmonary artery stenting (n = 4). Results: There were 2 perioperative deaths (2%). One patient developed sternal dehiscence requiring rewiring. Median ICU and hospital stay was 1 and 7 days respectively. Postoperative echocardiography at 6 and 12 months showed excellent bioprosthetic valve performance, significant decrease in size of the right cardiac chambers and reduction of tricuspid regurgitation (TR) in the majority of the patients. At mean follow-up of 3.6 ± 2 years, all surviving patients remain in excellent clinical condition. Conclusion: Probability of reoperation for pulmonary insufficiency in patients with surgically corrected TOF increases with time and timely PVR by preventing the development of right heart failure is crucial for long-term survival. Current bioprosthetic valve technology in combination with the beating heart technique provides excellent immediate and short-term results. Further follow-up is necessary to evaluate long-term outcome.
HYBRID PROCEDURES FOR CONGENITAL HEART DEFECTS
BACKGROUND

• Hybrid paediatric cardiac surgery is an emerging concept that combines skills and techniques used by paediatric cardiac surgeons and interventional cardiologists.

• The goal is to provide optimal therapy by minimizing the potentially harmful effects of methods that accompany conventional surgical procedures.
CONGENITAL HYBRID APPLICATIONS

• Intraoperative balloon occlusion of BT shunts, PDAs
• Balloon dilation of critical aortic stenosis
• Intraoperative pulmonary artery stenting
• Intraoperative muscular VSD device closure
• Hybrid stage one palliation for HLHS (Hybrid Norwood operation)
• Hybrid valve implantation
SUPRA-AS / HYPOPLASTIC PAS
VSD DEVICE CLOSURE
MINIMAL INVASIVE CONGENITAL HEART SURGERY

- ASD CLOSURE
- VSD CLOSURE
- TOF REPAIR
- TV REPAIR/REPLACEMENT
- AV CANAL REPAIR
- PDA LIGATION
- DIVISION OF VASCULAR RINGS
SURGICAL APPROACHES IN MINIMAL INVASIVE CONGENITAL SURGERY

• RIGHT THORACOTOMY
• MINI RIGHT SUBMAMMARY INCISION
• AXILLARY APPROACH
• BILATERAL SUBMAMMARY INCISION
• THORACOSCOPIC TECHNIQUE
• LOWER MINI STERNOTOMY
ΠΛΕΟΝΕΚΤΗΜΑΤΑ ROSS

1. Η βαλβίδα είναι αυτόλογη
2. Το αυτομόσχευμα μπορεί να μεγαλώσει
3. Δεν χρειάζεται αντιπηκτική αγωγή
4. Δεν υπάρχει διαβαλβιδική κλίση πίεσης
5. Απουσία ήχου λειτουργίας (μηχανικό “click”)
6. Αποδεδειγμένη ανθεκτικότητα ως Νεο-Αορτική βαλβίδα (ενδοκαρδίτιδα)
7. Η αυτόλογη πνευμονική βαλβίδα είναι ιδανικού μεγέθους και στείρα
ΜΕΙΟΝΕΚΤΗΜATA ROSS

1. σύμπλοκη επέμβαση (↑ περιεγχειρητικός κίνδυνος)
2. η αρχική παθολογία μετατρέπεται σε “νόσο 2 βαλβίδων” (Rahimtoola SH: JACC, 55:2413-2426, 2010)
3. τεκμηριωμένη ανάγκη για επανεπεμβάσεις στο RV-PA μόσχευμα
4. δεν μπορεί να διενεργηθεί σε όλους τους ασθενείς
ΕΝΔΕΙΞΕΙΣ ROSS

Α. ΑΠΟΛΥΤΕΣ
• παιδιά
• γυναίκες αναπαραγωγικής ηλικίας (τεκνοποίηση)

Β. ΣΧΕΤΙΚΕΣ
• προσδόκιμο επιβίωσης >20 έτη
• αθλητική δραστηριότητα
• επιθυμία του ασθενούς
THE ROOT REPLACEMENT TECHNIQUE
### ACTUARIAL ESTIMATES FOR SURVIVAL & FREEDOM FROM AUTOGRAFT, HOMOGRRAFT REOPERATIONS

<table>
<thead>
<tr>
<th>Center-Author</th>
<th>N patients</th>
<th>Survival (%)</th>
<th>FFR Autograft (%)</th>
<th>FFR Homograft (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>10y</td>
<td>15y</td>
<td>20y</td>
</tr>
<tr>
<td>Elkins et al. JTC 2008</td>
<td>487 (a-p)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>DAVID'S ROSS data JTC 2010</td>
<td>212 (a)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sir M. YACOUB Lancet 2010</td>
<td>228/2 (a)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Juthier et al ATS 2012</td>
<td>336 (a)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kalfa et al EJCTS 2015</td>
<td>276 (&gt;18)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mookhof et al ATS 2015</td>
<td>76 (&lt;1)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dutch-German MCS</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>LUMC ATS 2017</td>
<td>105 (a-p)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
• Ross procedure
  • Median age: 95 days
    • **In hospital mortality**: 9%, 29% in neonates, 5% in infants
    • **Mechanical support**: 7%, 29% in neonates, 3% in infants

• Ross-Konno procedure
  • Median age: 63 days
    • In hospital mortality: 19%, 29% in neonates, 9% in infants
    • Mechanical support: 12%, 17% in neonates, 9% in infants

• Homograft
  • Median age: 98 days
    • In hospital mortality: 40%
    • Mechanical support: 13%, in neonates 100%
Adults with CHD in US today

2,140,000

Growing 5% per year

Cahalan MK. Anesthetic Management of Patients with Heart Disease. IARS 2003 Review Course Lectures