Surgical Strategies: 
Transplantation in the Pediatric & Adult Congenital Heart Patient

Kristine J. Guleserian, M.D.
Director of Heart Failure & Transplantation 
Nicklaus Children’s Hospital 
Professor of Surgery 
Herbert Wertheim College of Medicine 
Miami, FL

AmSECT Pediatric Perfusion Conference
October 6, 2018

Objectives

• To become familiar with the epidemiology and history of congenital heart disease (CHD).

• To recognize the growing need for care of the adult congenital heart disease (ACHD) patient.

• To identify the unique challenges in caring for the complex pediatric and ACHD patient population.

• To understand the evaluation/management of the pediatric and ACHD patient in terms of surgical intervention, catheter intervention, transplantation and mechanical support.

Congenital Heart Disease (CHD)

• Most common birth defect.
• 1% of all newborns.
• 40,000 new cases/year in United States.
• 25% require operation < 1 year of age.
• Most common cause of death in infancy from any congenital malformation.

• Adults > children w/ CHD in 2000.
• > 1.5 million known to be affected in 2010.
Congenital Heart Disease Worldwide

Impact on 15-20%

CHD births/million population WHO 2011

http://cba.musc.edu/cdbc/Weinstein2004/

Normal Heart Anatomy
Common Congenital Heart Lesions

Successful Repair Expected

Annual Adult Congenital Procedures


Epidemiology

Survival to 18 yrs of Age with Simple, Moderate or Complex CHD

Hoffman 1978
Fyler 1980
New England Ferencz 1985
Baltimore-DC

Warnes et al. J Am Coll Cardiol 2001;37:1170-75

Epidemiology

Survival to 18 yrs of Age with Simple, Moderate or Complex CHD

20,000 new patients/yr
5% increase/yr

Hoffman 1978
Kaiser Fyler 1980
New England Ferencz 1985
Baltimore-DC

Warnes et al. J Am Coll Cardiol 2001;37:1170-75
Epidemiology

Survival to 18 yrs of Age with Complex CHD

<table>
<thead>
<tr>
<th>Decade Born with CHD</th>
<th>1960-</th>
<th>1970-</th>
<th>1980-</th>
</tr>
</thead>
<tbody>
<tr>
<td>Percent Survival to 18 Years Old</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>0</td>
<td>10</td>
<td>20</td>
<td>30</td>
</tr>
<tr>
<td>0</td>
<td>10</td>
<td>20</td>
<td>30</td>
</tr>
</tbody>
</table>


Improved Outcomes in Congenital Heart Disease

- Incidence of CHD steady
- Fetal Diagnosis
- Improved Surgical/Bypass Techniques/Myocardial Protection
- Advances in NICU/CVICU Care
- Earlier Complete Repair
- Increased Early Survival
- Increased Mid-Term Survival
- Increased Long-Term Survival
- Lower Perioperative Mortality

Surgical History

August 26, 1938
- First successful surgery to correct a congenital heart defect (PDA ligation)
- Dr. Robert Gross
- Dr. John Streider
  - First attempt in 1937
- Boston, MA

Today >90% Survival to Adulthood

Daebritz, Ped Cardiol 2007;28:96-104
Surgical History

October 19, 1944

- First repair of aortic coarctation
- Dr. Clarence Crafoord
- Stockholm, Sweden

Surgical History

November 29, 1944

- First clinical use of the Blalock-Taussig shunt for tetralogy of Fallot
- Dr. Alfred Blalock
- Vivien T. Thomas
  - Surgical technician
  - Developed technique
  - Animal model of PHTN

Dr. Helen Taussig

*Blue Baby Clinic, Johns Hopkins*

Tetralogy of Fallot

"Blue Baby Syndrome"

1. Anteriorly malaligned VSD
2. Overriding aorta
3. Pulmonary stenosis
4. Right ventricular hypertrophy

25% w/ PFO, RAA
Dr. Helen Taussig, Dr. Alfred Blalock, Vivien Thomas
Blalock-Taussig-Thomas (BTT) Shunt

“Open Heart Surgery”

- Transventricular repair
- Transatrial repair

1. Transannular patch for severe pulmonary stenosis → PI
2. Right ventricle-to-pulmonary artery conduit
3. Valve-sparing technique w/ commissurotomies & intraoperative balloon angioplasty

“Blue Baby” Operation 1944

Surgical History

September 2, 1952
- 1st open heart procedure (ASD closure).
- Hypothermia w/ inflow occlusion.
  - Dr. Wilfred Bigelow, Toronto
- Dr. F. John Lewis & Dr. Mansur Taufic
  - Dr. Richard Varco
  - Dr. C. Walt Lillehei
- University of Minnesota

Surgical History

May 6, 1953

- First successful open heart surgery using CPB
  - Dr. John H. Gibbon
  - Jefferson Medical College
- 18 year-old woman w/ ASD

- First “open” ACHD operation

Early Surgical Outcomes:

Mortality

<table>
<thead>
<tr>
<th></th>
<th>Year</th>
<th>Count</th>
<th>Mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>C. Dennis</td>
<td>1952</td>
<td>2/2</td>
<td>100%</td>
</tr>
<tr>
<td>J. Gibbon</td>
<td>1953</td>
<td>5/6</td>
<td>83.3%</td>
</tr>
<tr>
<td>J. Helmsworth</td>
<td>1953</td>
<td>1/1</td>
<td>100%</td>
</tr>
<tr>
<td>D. Dodrill</td>
<td>1953</td>
<td>2/2</td>
<td>100%</td>
</tr>
<tr>
<td>G. Clowes</td>
<td>1953</td>
<td>2/2</td>
<td>100%</td>
</tr>
<tr>
<td>W. H. Mustard</td>
<td>1953-54</td>
<td>5/5</td>
<td>100%</td>
</tr>
</tbody>
</table>

Total 17/18 94.5%
Cross Circulation

1954

• First successful repairs
  – VSD
  – Tetralogy of Fallot

• University of Minnesota

VSD Repair: March 26, 1954
Mayo-Gibbon Heart Lung Bypass Machine

Surgical History

March 22, 1955
- Dr. John Kirklin
- Mayo Clinic
- VSD closure with Gibbon mechanical pump-oxygenator
- By May, 4/8 patients survived to discharge

- 94.5% → 50% mortality!!!
Surgical Pulmonary Valve Replacement

- Carpentier-Edwards PERIMOUNT Magna Ease Bovine Pericardial Bioprosthesis

Transcatheter Pulmonary Valve Replacement

**Melody™**
- Bovine jugular venous valve segment
- Platinum-Iridium Stent

**Sapien™**
- Bovine pericardial tissue leaflets

Pulmonary valve implantation
May 8, 2015

Shaun White
Olympic Snowboarder
“The Flying Tomato”
What About the Failures?
Pediatric Heart Transplantation

HLHS s/p transplant @ 12 days of age
Died 19 days later

Stephanie Fae Beauclair
“Baby Fae”

Baboon to Human Heart Transplant
Loma Linda, CA
October 26, 1984
2 days of age, 2.2 kg
s/p TDO repair w/ septal hematoma
On ECMO support
Referred for transplant

2 y/o w/ HLHS w/ rASD
s/p Norwood
s/p bidirectional Glenn
Failing Glenn physiology
Highly sensitized
Berlin Heart EXCOR®
Ventricular Assist Device (VAD)
FDA Approved December 2011
LVAD, BIVAD, Systemic VAD (single ventricle)

Parental Perspective?

Berlin Heart Systemic VAD Removal, Orthotopic Heart Transplantation w/ LPA Stent Removal, Bilateral Branch RS Reconstruction & Aortic Arch Reconstruction

July 21, 2011
October 6, 2011
Hypoplastic Left Heart Syndrome (HLHS)

Unknown Diagnosis

- Premature, 10 day-old
- Birth weight < 2.5 kg
- Mechanical ventilation
- Cardiac arrest → CPR
- Lactate 17, pH 6.5
- Creatinine 2.6, INR 2.6
- PGE1, vasopressin, epinephrine
- ECHO → HLHS, poor RV function
**Resuscitative Bilateral PA Banding Strategy**

- Prostaglandin → maintenance of ductal patency
- Balloon atrial septostomy → IAS/RAS

**Plans for conventional Stage 1 Norwood Procedure**

- Ductal stent only if not a candidate for Norwood
  - Persistent poor RV function
  - Significant AVVR
  - Persistent end-organ dysfunction
  - Intracranial hemorrhage

---

**Bilateral pulmonary artery banding for resuscitation in high-risk, single-ventricle neonates and infants: A single-center experience**

Kristine J. Galantowicz, MA,* Gregory M. Basker, MD,* Hmodes S. Shamas, MD,* Joy Macaluso, RN,*

Rong Huang, PhD,* Alan W. Nugent, MBBS,* and Joseph M. Forbes, MD*

PGE1 → Ductal Stent if no recovery
(not if vascular ring present or diminutive ascending aorta)

*From the Children’s Hospital of Philadelphia, Philadelphia, PA.

Post-Operative Course

s/p Balloon Atrial Septostomy & Resuscitative Bilateral PA Banding

- Extubated
- Off epinephrine, vasopressin
- Normalization of INR, Creatinine
- Tolerating oral feeds

- Elective Norwood Procedure & PA debanding
What if Persistent Poor Ventricular Function? Severe TR? Both?

Heart Transplantation Techniques after Hybrid Single-Ventricle Palliation

VA Sebastian, R.D.S.; Kristian J. Galvan, M.D.; Steven R. Leonard, M.D.; and Joseph R. Fehlings, M.D.

Departments of Thoracic and Cardiovascular Surgery, UT Southwestern Medical Center at Dallas, Texas, and Division of Pediatric Cardiology, Texas Children’s Hospital, Texas Children’s Hospital, and UT Southwestern Medical Center at Dallas.

HLHS/variant HLHS

Bilateral PA bands

Ductal stent

PA debanding +/- PA reconstruction

Ductal stent removal

Aortic arch reconstruction

HLHS variant HLHS

Biatrial PA branches

Ductal stent

End to end descending thoracic aortic anastomosis & Brachiocephalic vessel "island" to donor arch

PA reconstruction w/ donor PA (hilum to hilum) or donor pericardial patch

Biatrial anastomosis

What About the ACHD Patient?

Types of ACHD Patients

1. Undiagnosed or misdiagnosed CHD (w/ PHTN).

2. Previously repaired and “cured” patient (TOF, D-TGA, CC-TGA, truncus arteriosus, coarctation, complete AV canal).

1. Previously palliated patient (shunted single ventricle → completion of Fontan circulation).

2. Heart failure patients (considered for transplantation).

The Unoperated Patient w/ CHD

- Septal defects (ASD, VSD, AV Canal)
- Tetralogy of Fallot
- Sinus venosus defect +/- PAPVC
- Coarctation of aorta
- Corrected transposition (CC-TGA)
- Anomalous coronary artery (AAOCA)
- Aortic stenosis (bicuspid aortic valve)
- Scimitar syndrome

The Unoperated Cyanotic Patient

- Many reach adulthood w/ cyanotic CHD (TOF).
- Presence of aortopulmonary collaterals.
- Intraoperative nightmare.
- If well saturated → coil embolization.

- Excessive pulmonary venous return.
  - Obscuration of operative field
  - Impaired myocardial protection
  - Left-sided volume overload
  - Systemic underperfusion (cerebral, renal, mesenteric)
Adults With CHD: Challenges

- Told they were “cured”.
- Do not receive appropriate surveillance or lost to follow up.
- Community care—or no care at all.
- Not educated about their heart disease & what to expect.
- Residual lesions or sequelae of disease/operation.
- Development of co-morbidities.
- No records

CHD = A Chronic Disease

Cardiac Transplantation in Patients with Congenital Heart Disease (CHD)

- Cardiac transplantation for CHD incorporates aspects of reparative & replacement surgery.
- Intracardiac congenital malformations are replaced.
- Extracardiac malformations (congenital, acquired, or iatrogenic) → considerable challenges.
  - Anatomic variants.
  - Conduits, residual shunts, stents, etc.
  - Adhesions, pseudoaneurysm, thrombus burden, etc.
- Comprehensive operative plan = essential.

The prevalence of heart transplantation among adult recipients decreased by 28% over time (P<.001) and increased among patients with adult congenital heart disease by 41% (P<.001).

J Thorac Cardiovasc Surg 2010
Cardiac Transplantation in Patients with Congenital Heart Disease (CHD)

- **Donor team**
  - Harvest appropriate donor tissue to allow for adequate reconstruction.
  - Timing must be coordinated.

- **Recipient team (surgical-anesthesia-perfusion)**
  - Reoperative median sternotomy
  - Vascular access → safe establishment of cardiopulmonary bypass and full cardiac support.
  - AP collaterals → adequate cooling/venting.
  - Blood products → anticipated bleeding/coagulopathy.

Cardiac Transplant: Bi-atrial Anastomosis

Cardiac Transplant: Bi-caval Anastomosis
Tetralogy of Fallot w/ Pulmonary Atresia & Hypoplastic Pulmonary Arteries

Residual Shunts

Baffle Obstruction s/p Atrial Switch for D-TGA

Branch Pulmonary Artery Stenosis and Pulmonary Insufficiency s/p TOF Repair
**RVOT Pseudoaneurysm s/p TOF Repair**

**Ventricular Dysfunction & Dilation**

RAF s/p Repair With RV Diastolic Dysfunction

DORV s/p Repair With Diastolic Dysfunction

**Atriopulmonary Fontan**

Thrombus formation

*Thrombus formation, atrial dysrhythmias (atrial flutter)*

**Endocarditis**
Reoperative Right Femoral Cutdown
8mm Dacron chimney graft

Donor Heart
- Innominate vein
- Aortic arch
- Branch PAs
- Pulmonary veins
- Pericardium

Bilateral Superior Vena Cavae (B SVC)

Bilateral Superior Vena Cavae (B SVC):

L SVC $\rightarrow$ roof of LA

B SVC: L SVC $\rightarrow$ Roof of LA

Intra-atrial baffle along roof of LA

Intra-atrial baffle along floor of LA
B SVC: L SVC → Coronary Sinus
“Roofed” pathway

Bicaval Anastomosis
“Roofed” pathway

Biatrial Anastomosis
“Roofed” pathway

Bilateral Superior Vena Cavae: Modification of Recipient Cardiectomy
Heterotaxy Syndrome
Management of Ipsilateral Hepatic Veins, L IVC


Transposition of the Great Arteries (TGA)

Extracardial baffle of hepatics (or L IVC) to RA

D-TGA s/p Mustard
D-TGA s/p Senning
L-TGA

Extra Donor & Recipient Aorta

Extra donor PA

Transposition of the Great Arteries (TGA)

Sutured arteriotomy

D-TGA s/p Mustard
D-TGA s/p Senning
L-TGA

Leftward translocation of PA anastomosis


Pulmonary Artery Distortion

RSVC arteriotomy

LSVC arteriotomy

Prior femoral baffle insertion

Pulmonary Arterioplasty
Donor MPA/branch PA spatulation

Discontinuous Pulmonary Arteries

HLHS/variant HLHS s/p Hybrid Procedure

Transplant w/ Arch Reconstruction s/p Hybrid Procedure (+/- Branch PA plasty)
Situs Inversus

Donor Heart: LA Cuff Modification

Situs Inversus:
*Donor Cardectomy Modification*

Left-sided PVs oversewn

Obstructed donor LSVC-IV connection (behind great arteries) re-opened with vascular stents


Long L IVC-RA cuff

B BDG takedown

PA spatulation

Deuse T, Reitz B, J Thorac Cardiovasc Surg 2010
Situs Inversus: 
Implant Technique

- Apex rotated
- Recipient aortic stump oversewn
- End-to-side aortic anastomosis

Deuse T, Reitz B. J Thorac Cardiovasc Surg 2010

Situs Inversus: 
Transplant Technique

- Mobilization of recipient atrial septum & anastomosis to LA free wall
- Separate atriotomy anterior to L PVS = neo LA
- Closure of leftward aspect RA cuff

Mora B, Huddleston C. Curr Cardiol Rev 2011

Transplantation in Adults With Congenital Heart Disease

Abstract: Considerable progress in patients with congenital heart disease surviving into adulthood. However, improved postoperative management and thoracic surgical expertise have dramatically reduced mortality. Many centers now perform comprehensive adult congenital heart surgery. Adult congenital heart disease represents a growing population of patients being referred for heart, lung, and combined heart-lung transplantation. The group of children and young adults with complex congenital heart disease and subsequent need for transplantation is growing, as noninvasive imaging more precisely predicts outcomes and surgical expertise improves. Thus, the number of adult congenital heart disease transplant recipients expected to grow in the future.

Mora B, Huddleston C. Curr Cardiol Rev 2011
Operative Preparation is Key!!!

Utility of 3D Models

- Understand the anatomy
- Communicate the anatomy
  - Patients & families
  - Providers
  - Students
- Plan intervention
  - Cardiac catheterization
  - Cardiac surgery
- Practice/simulate intervention
  - Cardiac catheterization
  - Cardiac surgery
3D Printing Program: Library of Heart Models

D-Transposition of Great Arteries (D-TGA) w/ VSD
SynCardia Total Artificial Heart (TAH)