Surgical Home for the Patient with Congenital Heart Disease

Luis M. Zabala M.D
Associate Professor
UT Southwestern Medical Center
Children’s Health Dallas

Congenital Cardiac Anesthesia Society
Annual Meeting 2016
Colorado Springs, CO
Conflict of interest disclosure

• No financial disclosures
Learning Objectives
Session V: Adult Congenital Heart Disease

• At the end of this session, the participant will be able to:
  
  – Recognize the changing demographics of patients with CHD
  
  – Report on the improved outcomes of CHD
  
  – Demonstrate how congenital cardiac anesthesiologist can enhance the perioperative care of patients with CHD at all stages and ages
  
  – Identify some of the issues faced by adults with CHD including the patient’s perspective of living with CHD
Presentation
Objectives and Outline

20 minutes:

• Describe the current landscape of medical care for children/adults with CHD in the US

• Surgical Home model - goals

• Role of the anesthesiologist in the perioperative encounter
  • Risk assessment and basic clinical aspects
  • Major pathophysiologic pathways

• Children’s Health, Dallas
  • Role of Pediatric Cardiac Anesthesia Coordinator (CAC)
Landscape of Medical Care of Congenital Heart Disease in the US - Setting the Stage

• Birth prevalence reported to be 10 per 1000 live births and estimated incidence between 3 and 20 per 1000 live births

• **Natural history of lesions and overall survival rate have changed dramatically**

• Classification of CHD Diagnoses - Stratified by severity into 3 levels:
  – Simple, moderate and severe complexity based on cardiac lesion *(ACC/AHA)*
  – Minor, major and severe based residual lesion – functional status *(ACS NSQIP)*

• 25 – 30% of all CHD patients have associated another mayor congenital anomaly

• As life expectancy increases - also does the need for medical attention for:
  – Know natural history of the primary cardiac diagnosis
  – Comorbidities related to normal aging

“Substantial health service needs and resource utilization”
Survival: Temporal Trends in Survival Among Infants with Critical Congenital Heart Disease - Oster ME. Pediatrics 2013

Survival for persons with CCHD versus CHD: Atlanta, Georgia, 1979-2005

97%  95.4%
75.2%  68.8%

P < .0001
Estimated survival according to the era of birth.

Philip Moons et al. Circulation. 2010;122:2264-2272

Copyright © American Heart Association, Inc. All rights reserved.
Estimated survival beyond 18 years of age.

Philip Moons et al. Circulation. 2010;122:2264-2272
Trends in Hospitalization for Adults with Congenital Heart Disease in the US — Opotowsky A. et al. JACC 2009

Frequency of specific diagnosis and procedure associated with hospitalizations for ACHD

![Bar chart showing annual number of ACHD admissions in the U.S. categorized by level of defect complexity.](image)

<table>
<thead>
<tr>
<th>Year</th>
<th>1998</th>
<th>2004</th>
<th>2005</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diagnoses*</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>11,742 (1,012)</td>
<td>22,458 (1,896)</td>
<td>24,882 (1,961)</td>
</tr>
<tr>
<td>CAD†</td>
<td>8,574 (745)</td>
<td>18,440 (1,549)</td>
<td>18,788 (1,455)</td>
</tr>
<tr>
<td>Heart failure</td>
<td>7,453 (624)</td>
<td>12,939 (1,064)</td>
<td>13,604 (1,008)</td>
</tr>
<tr>
<td>Pulmonary hypertension</td>
<td>3,923 (294)</td>
<td>6,652 (552)</td>
<td>6,528 (525)</td>
</tr>
<tr>
<td>Pregnancy</td>
<td>1,706 (162)</td>
<td>2,990 (270)</td>
<td>3,431 (312)</td>
</tr>
<tr>
<td>Bacterial endocarditis</td>
<td>574 (71)</td>
<td>1,103 (116)</td>
<td>867 (99)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Procedures</th>
<th>1998</th>
<th>2004</th>
<th>2005</th>
</tr>
</thead>
<tbody>
<tr>
<td>Percutaneous ASD/PFO closure</td>
<td>134 (90)</td>
<td>4,415 (1,043)</td>
<td>3,219 (566)</td>
</tr>
<tr>
<td>Pacemaker</td>
<td>974 (150)</td>
<td>2,021 (208)</td>
<td>2,188 (236)</td>
</tr>
<tr>
<td>PCI</td>
<td>830 (103)</td>
<td>1,967 (247)</td>
<td>2,227 (239)</td>
</tr>
<tr>
<td>ICD</td>
<td>208 (45)</td>
<td>901 (130)</td>
<td>840 (109)</td>
</tr>
</tbody>
</table>

Mean hospital charges per hospitalization increased: 127% from $19,186 +-$803 to $43,496 +-$2166
Total national charges for these hospitalizations in 2005 $3.16 B
Overall Hospital Cost Estimates in Children with Congenital Heart Disease: Analysis of the 2012 Kid’s Inpatient Database

Total Cost Estimates in 2012: 6,722 M$

Faraoni D. et. Al. Pediatr Cardiol. 2015
## Financial Implications in the Care of Complex Congenital Heart Disease


### Cost of care for congenital heart surgery

<table>
<thead>
<tr>
<th>Department</th>
<th>Cost</th>
<th>% Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hospital</td>
<td>$33,658</td>
<td>79.5</td>
</tr>
<tr>
<td>Surgery</td>
<td>$4,185</td>
<td>10.0</td>
</tr>
<tr>
<td>Anesthesia</td>
<td>$1,080</td>
<td>2.6</td>
</tr>
<tr>
<td>Critical Care</td>
<td>$1,174</td>
<td>2.7</td>
</tr>
<tr>
<td>Cardiology</td>
<td>$1,250</td>
<td>3.0</td>
</tr>
<tr>
<td>Other</td>
<td>$750</td>
<td>1.9</td>
</tr>
<tr>
<td>Total</td>
<td>$42,097</td>
<td>100%</td>
</tr>
</tbody>
</table>

2011 - $92,000

“...decision makers, e.i., the surgeon, cardiologist, anesthesiologist, and intensivist are only 20% liable for the cost, yet influence the remaining 80% cost figure through their decisions”.

William J. Greeley, MD, MBA
Dallas, Texas

Reality of our Current Practice

- Children with CHD = Children with Medical Complexity
- Diagnoses are multiple and varied, generating significant cost
- Prevalence and demographics are changing
- Patients are under the continuous care of multiple pediatric specialists, and many transitioning to adult care
- Travel regionally or even nationally for diagnosis and treatment services
- Information is disorganized, fragmented and often unavailable

**Ramifications today for Anesthesia care:**
- Inadequate workup, delays, cancellations, unanticipated or prolonged admissions, multiple anesthetics, readmission, and increased cost.
- What can we do?
Surgical Home:  
A case for better Coordination/ Risk Stratification/ Cost

• ASA – Perioperative Surgical Home  
  – Patient centric / team based approach to emphasize outcome, patient satisfaction and reduce cost

• Anesthesiologist acts as coordinator of the surgical encounter:  
  – preoperative, intraoperative and postoperative  
  – inefficient resource utilization, optimize the clinical and surgical encounter process and decrease cost

• Integrating a continuous Quality Improvement (QI) process

Limited data available outlining the application of the Perioperative SH model in the pediatric population

Clinical + Encounter Optimization and Coordination
Optimization:  
Clinical and Surgical Process

• The concept: …“ an act, process, or methodology of making something (as a design, system, or decision) as fully perfect, functional, or effective as possible……” Merriam-Webster Dictionary

• In medicine: …” optimization has become an essential tool in addressing the limitations of resources and need for better medical decision-making.” Alves C. Optimization in Medicine. 2008 Spinger
Prerioperative Optimization in CHD

- Heterogeneous group of patients across all ages
- Countless permutations: physiologic variables, hematologic and hemostatic conditions, comorbidities, medications, stage of disease and known natural history
- Surgical trespass needs consideration
- Lack of standardized risk stratification
- **Optimization comes down to:**
  - **Controlling variables and unexpected outcomes:**
    - Adequate preoperative evaluation
    - Risk Assessment
    - Location of surgery and personnel
    - Planned disposition / Post Operative follow-up
Risk Assessment is Essential

- Hypoxia
- Hypercardia
- Hypotension
- Pain
- Sympathetic surgeres
- Blood Loss
- Arrhythmias
- Fluid shift

Homeostatic equilibrium of adequate circulatory function in patients with limited reserve
# Cardiac Lesions associated with highest risk of Morbidity and Mortality undergoing Anesthesia

<table>
<thead>
<tr>
<th>Cardiac lesion</th>
<th>Pathophysiologial consideration</th>
<th>Anesthetic goals</th>
<th>Risk of cardiac arrest with anesthesia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary artery hypertension</td>
<td>Catecholamine from light anesthesia, hypercarbia, hypoxemia, acidosis, hypotension, lead to right ventricular failure, low cardiac output</td>
<td>Maintain oxygenation, ventilation, adequate depth of anesthesia, pulmonary vasodilators including iNO</td>
<td>1.1 – 5.7% of anesthetics in severe pulmonary hypertension (13)</td>
</tr>
<tr>
<td>Left ventricular outflow tract obstruction</td>
<td>Tachycardia, hypovolemia, systemic hypertention, myocardial depression or hypercontractility reduce stroke volume, lead to coronary ischemia, low cardiac output</td>
<td>Maintain ventricular filling, SVR, normal to slow heart rate, normal myocardial contractility</td>
<td>16% of POCA registry (14)</td>
</tr>
<tr>
<td>Single ventricle and systemic to pulmonary artery shunt</td>
<td>Systemic and pulmonary output both ejected by single functional ventricle, SVR and PVR determines systemic cardiac output</td>
<td>Avoid hyperoxgenation, hyperventilation; maintain ventricular function</td>
<td>19% of POCA registry (14)</td>
</tr>
<tr>
<td>Dilated cardiomyopathy</td>
<td>Increased ventricular volume, ejection fraction 5-25%, cardiac output maintained by near normal stroke volume and tachycardia, very limited reserve for decreased SVR, contractility, preload</td>
<td>Avoid decrease myocardial contractility; maintain preload, and SVR</td>
<td>13% of POCA registry; 1.7% of anesthetics with dilated cardiomyopathy (14, 15)</td>
</tr>
</tbody>
</table>

POCA, pediatric perioperative cardiac registry
Modified from Gottlieb E, Andropoulos DB. Anesthesia for the patient with congenital heart disease presenting for non-cardiac surgery. Curr Opin Anesthesiol 2013, 26:318-326
Post-Operative Outcomes in Children With and Without Congenital Heart Disease Undergoing Noncardiac Surgery — Faraoni D, et. al. JACC 2016

ACS NSQIP Classification of CHD

**Minor CHD:**
Cardiac condition with or without medication and maintenance
Repair of cardiac condition with normal function and no medication

**Major CHD:**
Repair of CHD with residual hemodynamic abnormality with or without medication

**Severe CHD:**
Uncorrected cyanotic CHD
Documented Pulmonary Hypertension
Ventricular dysfunction
Listed for Heart transplantation
Cardiac Lesions associated with highest risk of Morbidity and Mortality undergoing Anesthesia in the ACHD Population
ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease

Congenital Cardiac Lesions and Perioperative Risk for Non-cardiac Surgery

High Risk
- Pulmonary hypertension, primary or secondary
- Cyanotic CHD
- New York Heart Association class III or IV
- Severe systemic ventricular dysfunction
- Severe left sided heart obstructive lesions

Moderate Risk
- Prosthetic valve or conduit
- Intracardiac shunt
- Moderate left-sided heart obstruction
- Moderate systemic ventricular dysfunction

JACC VOL. 52 NO 23, 2008
Common Pathophysiologic Pathways: At Risk of Low Cardiac Output in the Perioperative Period

- Pulmonary Hypertension
- **Single Ventricle**
- **Systemic Right Ventricle**
- **Progressive Contractile Dysfunction (Decreased SF)**
  Congenital or acquired cardiomyopathies
- Left sided obstructive lesions
- Progressive Electrophysiologic Abnormalities
- End Organ Dysfunction
Single Ventricle and the Perioperative period

• Predictable complications of the Fontan pathway:
  – Right atrial dilation
  – Atrial arrhythmias
  – Baffle leaks / Pulmonary venous obstruction and conduit obstruction

• Perioperative Optimization:
  – Maintain an adequate transpulmonary gradient
  – Prevent increases in pulmonary vascular resistance
  – Ventilation without compromising pulmonary blood flow
  – Intravascular volume optimization is essential
The Systemic Right Ventricle
The unnatural history of the systemic RV

- Adults with systemic RVs:
  - 68% - RV dysfunction
  - 64% - tricuspid regurgitation
  - 20% - tachyarrhythmias
  - Average RVEDV is 40% greater than the LVEDV.

- Perioperative Optimization:
  - Regulation of volume status, heart rate and inotropic function.
  - Sinus rhythm
  - Hypotension should be avoided
  - Concerns with paradoxical emboli during conditions of RV compromise
Substrates for Late Heart Failure in ACHD
ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease

• Severe AS and/or regurgitation, BAV and variants, subvalvar or supravalvar obstruction, superimposed coarctation
• Severe congenital mitral stenosis/regurgitation
• Unoperative ASD or partial AVSD
• CCTGA
• D-transposition after Mustard or Senning operation
• Tetralogy of Fallot with early-era surgery
• Single ventricle physiology
• Fontan surgery
American Heart Association/American College of Cardiology congestive heart failure stages.


Copyright © American Heart Association, Inc. All rights reserved.
CV Anesthesia guided Surgical Home: Perioperative Coordination Model

Diagnoses: HLHS, BDG, AVVR, Seizures, Scoliosis

Home Environment: El Paso, TX

Medical Records: 1 Job

Specialist: Neuro, GI, Ortho, Pulm, Cardio

MRI

SH or Cardiac Anesthesia Coordinated Care

Preop: Information, Clinical and Risk Assessment, Optimization, EB Protocols, Care Plan

Intraop: Right personnel, Equipment, Reduced variations

Post: Right disposition, Integrated plan, Pain mgt, Prevention of complications, Discharge

Quality Improvement & Database

Mom, Dad, 2 siblings

Medications & Diag Studies

Dallas, Texas
The goal of anesthesia-led coordination is to optimize safety of children with CHD requiring procedures under general anesthesia or sedation, by providing complete preoperative workup, risk stratification within our system, and postoperative follow-up.

We created a job description under the title: “Pediatric Cardiac Anesthesia Coordinator” that would serve as clinical leader, resource and program coordinator to all children's’ with CHD requiring anesthesia services.

Program began 2012: > 3000 Anesthesia review a year.
Current Anesthesia Coordinated Pathway
Children’s Healthcare, Dallas

Rate Limiting Step:
Incomplete workup, Coverage or disposition problem

Surgical Care
Anesthesia Care
- Evidence based
- Safety
- Intraoperative metrics

Cardiac Anesthesia Coordinator

Integration of Information
Risk Assessment
Disposition Planning (CVPACU, Floor, CVICU, PICU)
Case Assignment
Dissemination of Information
Coordination and planning for ECMO

Surgeon’s office or Preoperative Clinic

Preoperative evaluation
Laboratory Imaging testing
Med Rec consolidation
Functional assessment
### CHD Patient Anesthetic Risk Assessment

<table>
<thead>
<tr>
<th><strong>Low</strong></th>
<th><strong>Moderate</strong></th>
<th><strong>High</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>- Unrepaired Isolated lesions: Small to moderate sized/restrictive L to R PDA/ASD/VSD</td>
<td>- Mild-Mod PAH Well controlled on PO +/- O2 Tx</td>
<td>- Mod-Severe PAH (including patients on IV therapy)</td>
</tr>
<tr>
<td>- Repaired ASD/VSD</td>
<td>- Single V s/p Glenn or Fontan (with mild-mod AVVR &amp; preserved ventricular function)</td>
<td>- Single V Pre-Glenn</td>
</tr>
<tr>
<td>- Repaired TGA</td>
<td>- Moderate PS/RVOTO</td>
<td>- Single V with Severe AVVR and/or ventricular dysfunction</td>
</tr>
<tr>
<td>- Repaired AVC with trivial – mild AVVR, normal function, no ventricular outflow obstruction</td>
<td>- Moderate AS/LVOTO</td>
<td>- Severe AS/LVOTO</td>
</tr>
<tr>
<td>- Repaired TAPVR/PAPVR without pulmonary venous obstruction</td>
<td>- Cardiomyopathy w/ mild ventricular dysfunction without dysrhythmias</td>
<td>- Severe PS/RVOTO</td>
</tr>
<tr>
<td>- Mild PS/RVOTO</td>
<td>- Dysrhythmia s/p Pacemaker or ICD (stable)</td>
<td>- Unrepaired TOF/DORV/AVC/PA</td>
</tr>
</tbody>
</table>

All patients with repaired CHD are screened to assure no change in status. This includes: most recent progress note/ECHO/ECG report from primary cardiologist, most recent Cath report and/or holter as indicated, and current evaluation when patient presents to pre op.
Cardiac Anesthesia High Risk Bundle Cases
Introduction to Cardiac Anesthesia Coordinator (CAC)

Prior to introduction of the CAC, frequently procedures (surgery, radiology, IR, ENT) were requested by different services, significant fragmentation; leading to repeated exposure to anesthesia, increase risk and unwanted ICU admission.
Other Areas of Interest:

**Ventilator Weaning Protocol**

Average intubation time:
- 1.6 to 0.9 days

CICU stay
- 8.7 to 5.5 days

Average hospital stay
- 18.9 to 12.4 days
Conclusion: