Congenital Heart Surgery
Past, Present and Future

James Quintessenza, MD
Professor of Surgery
University of Kentucky Children’s Hospital
Cincinnati Children’s Hospital

Latest in CV Medicine and Surgery
April 14, 2018
Disclosures

None
Education Needs/Gaps

A major practice gap exists in the knowledge of current surgical treatment for congestive heart disease. Recent advances in surgical techniques have decreased mortality rate and greatly increase years of survival post surgery.
Learning Objectives

Upon completion of this education activity, you will be able to:
Discuss the recent trends in congenital heart surgery.
Outcomes

The desired change/result in practice is to improve surgical treatment methods used.
Today, I would like to talk about Innovation and Innovators in the Development of Surgery for Congenital Heart Disease
“There is nothing more difficult to take in hand, more perilous to conduct, nor uncertain in its success, than to take the lead in the introduction of a new order of things.

For the innovator has for enemies all of those who have done well under the old, and lukewarm defenders in all of those who may do well under the new.”

Niccolò Macchiavelli (1469–1527)
A surgeon who tries to suture a heart wound deserves to lose the esteem of his colleagues.

Performing an operation to the heart is tantamount to an act of prostitution in surgery or surgical frivolity.

http://www.aerzteblatt.de/archiv/54013/Medizingeschichte-Herznaht-wider-ethische-Bedenken
Prof. Dr.
Ludwig Rehn

1896 closure of a stab wound in the right ventricle in Frankfurt am Main by Prof. Dr. Ludwig Rehn. Patient developed empyema but eventually survived.

• First repair of a heart wound
• Dr Robert Gross @ Boston Childrens Hospital

• 1938 first successful Patent Ductus ligation

• Chief of Surgery, Dr Ladd said “Do not operate or that little girl will die”

• He ligated the PDA and was fired upon Dr Ladd’s return

• He later was rehired and named the first Ladd’s Professor of Surgery
Following several years in the laboratory working on vascular surgical techniques

1945 First successful repair of coarctation
“.. On evening rounds, we arrived at the crib of this fifteen-month old baby... I was immediately astounded by the deep cyanotic appearance of the child, much more cyanotic than any patient I had ever seen before: the lips were a deep, dark blue.. The face was suffused with dilated veins, the conjunctiva almost purple.”
• **Blalock-Thomas-Taussig Shunt** (1944)

• Aorto-Pulmonary shunt to increase blood flow to the lungs of blue babies and markedly improve blood oxygen levels.
• These developments stimulated many individuals to begin to pursue studies in cardiology and surgery for congenital heart disease
Modern Era of Congenital Heart Disease

The Quest to Enter the Heart
Pre Heart Lung Machine

- Dr Gross developed “Atrial Well” technique to close Atrial septal defects (1952)

- 30% Mortality
Hypothermia

- (1952) Use of ice cold baths to cool the patient and stop the heart as well as slow metabolism allowing surgeons to quickly open the heart and repair the defects.

Figure 2.4. Water baths for immersion hypothermia and intracardiac repair. (Courtesy of Professor H. Koyanagi of Tokyo Women’s Medical College, Japan.)
First Heart Lung Machine

- Dr John Gibbon (1952) following 20 years of lab work

- First successful closure of an Atrial Septal Defect in an 18 yr old girl on his second try

- 3 mortalities followed and he never operated again

- Moratorium on the use of his CPB machine

Figure 2.21. John Heysham Gibbon and his wife Mary Hopkinson with the heart-lung machine.
Controlled Cross Circulation

• Dr Walt Lillihei @ University of Minnesota (1953)

• Developed a technique for open heart surgery using the parent of a child as the heart lung machine

"King of Hearts"
Controlled Cross Circulation

- Repair of VSD using cross circulation with 6/8 pts surviving (75%)
- Compared to 40-50% survival with various mechanical heart lung machines at the time
- Still had to overcome considerable negative sentiment of many critics
Controlled Cross Circulation

Figure 2.8. Lillehei’s first donor cross-circulation operation.
• The next 50 years produced significant improvement in techniques, equipment and materials

• **Everything got better!**
  - Surgical technique
  - Anesthesia
  - Heart lung machines and perfusion strategy
  - Cardiology
  - Critical Care
  - Nursing and Respiratory Therapy
  - Dietitians, child life specialists, social workers and many more

• Improved outcomes!
Neonatal Repair
Transposition of the Great Arteries

- Aldo Castenada @ Boston Childrens Hospital (1980’s)
- Developed modern approach to primary repair of complex forms of CHD in newborns
Neonatal Heart Transplantation

- Dr Leonard Bailey @ Loma Linda
- Pioneered neonatal heart transplantation as primary therapy for HLHS (1985)
- Baby Fay - Baboon to Human transplant in a dying infant with HLHS
• Neonatal Transplantation
Staged Reconstruction
Hypoplastic Left Heart Syndrome

Bill Norwood
(Boston & Philadelphia)

untreated, all neonates with HLHS will die in < 4 weeks
mortality of surgery for congenital heart disease
Current Era

- Most bi-ventricular repairs have very low operative mortality and long term survival is very good
  - ASD
  - VSD
  - Tetralogy of Fallot
  - AVSD
Current Era

- Neonatal surgery for complex bi-ventricular repairs
- Arterial Switch for Transposition - Mortality 1-2%
- Long term survival approaches normal population
Current Era

• Neonatal surgery for complex bi-ventricular repairs
  • Arterial Switch for Transposition- Mortality 1-2%
  • Long term survival approaches normal population
Staged Palliation for HLHS

Stage I
Norwood
Sano or BT Shunt

Stage II
Bidirectional Glenn

Stage III
Fontan
Stage 1 Norwood Operative Mortality (complex single ventricle palliation)
Completion Fontan Surgery

Overall risks: 1-3% mortality

Complications
- Pleural effusions
- Low cardiac output
- Arrhythmias
Fontan Survival: Current Era

10 Year Survival 94-98 %
Fontan Survival: Current Era

Estimated 40 year survival @ 60%
Fontan Survival: Current Era

Estimated 40 year survival is 60%

Mortality and morbidity related to long term sequelae of chronic venous hypertension and right heart failure

Arrhythmias, heart failure, liver dysfunction, PLE, pulmonary vascular problems are continued challenges
Abstract

OBJECTIVES: Multistage palliation is the mainstay management strategy of children with hypoplastic left heart syndrome (HLHS) and related single ventricle anomalies. If this palliation strategy fails, heart transplantation (HT) is required. The results of HT in children who had a prior Norwood operation are reportedly poor due to several immune, clinical and anatomical risk factors. We report our institutional outcomes following HT in children who had a prior Norwood operation.

METHODS: Between 1994 and 2013, 107 children with congenital heart disease underwent HT. We examined early and late outcomes in our study cohort of children who had a prior Norwood operation (n = 24), and analysed risk factors affecting survival. Survival was subsequently compared with a control group of 83 children with congenital heart disease without a prior Norwood operation who received HT.

RESULTS: Twenty-four children with a prior Norwood operation underwent HT. The majority (22/24, 92%) had HLHS. Children were listed following Norwood (n = 2, 8.3%), Glenn (n = 17, 70.8%) or Fontan (n = 5, 20.8%) operation. Ten (42%) patients had panel reactive antibodies (PRAs) >10%. Median age at listing was 2.7 (range 0.4-16.8) years and median age at the time of HT was 3.0 (range 0.6-16.8) years, with the median waiting list duration of 63.5 (range 1-554) days. Hospital mortality was 1/24 (4%). Overall parametric survival estimates at 1, 5 and 10 years were 85, 65 and 52%. Survival was not affected by listing status, last palliation stage, age or high PRA. The only significant factor affecting survival was the later era in our series with significantly superior 5-year survival (100 vs 42%, P = 0.0003). Overall survival was comparable with the control group of children with congenital heart disease and no prior Norwood operation (52 vs 53% at 10 years, P = 0.97). Overall, 3 of 24 patients required retransplantation with only one late survivor.

CONCLUSIONS: Children failing multistage palliation of HLHS may require HT, often following the Glenn operation. HT results in this group are comparable with those in other children with congenital heart disease. Improvements in pretransplant management, immune suppression and outpatient care in the later era might have specifically benefited this particularly risky group of patients.

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Transplantation: Current Era Outcomes Improved for Failed Fontan with HLHS

[Graph showing patient survival rates over years since transplant, comparing two eras: 1991-2003 and 2004-2013.]
Heartmate II Ventricular Assist Device
Failed Fontan for HLHS

A new era: Use of an intracorporeal systemic ventricular assist device to support a patient with a failing Fontan circulation

David L. S. Morales, MD, Iki Adachi, MD, Jeffrey S. Heinle, MD, and Charles D. Fraser, Jr, MD, Houston, Tex

FIGURE 1. Preoperative and postoperative chest x-ray films.

15 y/o boy with failed Fontan for HLHS
The evolving role of the total artificial heart in the management of end-stage congenital heart disease and adolescents.

Ryan TD, Jefferies JL, Zafar F, Lorts A, Morales DL.

Abstract

Advances in medical therapies have yielded improvement in morbidity and a decrease in mortality for patients with congenital heart disease, both surgically palliated and uncorrected. An unintended consequence is a cohort of adolescent and adult patients with heart failure who require alternative therapies. One intriguing option is placement of a total artificial heart (TAH) either as a bridge to transplant or as a destination therapy. Of the 1091 Jarvik-7 type TAH (Symbion, CardioWest and Syncardia) placed between 1985 and 2012, only 24 have been performed in patients with congenital heart disease, and a total of 51 were placed in patients younger than 21. At our institution, the Syncardia TAH was implanted in a 19-year-old patient with cardiac allograft failure because of chronic rejection and related multisystem organ failure including need for hemodialysis. Over the next year, she was nutritionally and physically rehabilitated, as were her end organs, allowing her to come off dialysis, achieve normal renal function and eventually be successfully transplanted. Given the continued growth of adolescent and adult congenital heart disease populations with end-stage heart failure, the TAH may offer therapeutic options where previously there were few. In addition, smaller devices such as the Syncardia 50/50 will open the door for applications in smaller children. The Freedom Driver offers the chance for patients to leave the hospital with a TAH, as does the AbioCor, which is a fully implantable TAH option. In this report, we review the history of the TAH and potential applications in adolescent patients and congenital heart disease.
Mechanical Support for Fontan Patients

Bridge vs destination therapy for Fontan pts?

Single ventricle assist or total artificial heart?

For adjunctive therapy before liver dysfunction?
Stem cell therapy for CHD: towards translation*

Brody Wehman, Osama T. Siddiqui, Rachana Mishra, Sudhish Sharma, Sunjay Kaushal

Division of Cardiac Surgery, University of Maryland School of Medicine, Baltimore, Maryland, United States of America

Abstract Stem cell therapy has the optimistic goal of regenerating the myocardium as defined by re-growth of lost or destroyed myocardium. As applied to patients with heart failure, many confuse or limit the regenerative definition to just improving myocardial function and/or decreasing myocardial scar formation, which may not be the most important clinical outcome to achieve in this promising field of molecular medicine. Many different stem cell-based therapies have been tested and have demonstrated a safe and feasible profile in adult patients with heart failure, but with varied efficacious end points reported. Although not achieved as of yet, the encompassing goal to regenerate the heart is still believed to be within reach using these cell-based therapies in adult patients with heart failure, as the first-generation therapies are now being tested in different phases of clinical trials. Similar efforts to foster the translation of stem cell therapy to children with heart failure have, however, been limited. In this review, we aim to summarise the findings from pre-clinical models and clinical experiences to date that have focussed on the evaluation of stem cell therapy in children with heart failure. Finally, we present methodological considerations pertinent to the design of a stem cell-based trial for children with heart failure, as they represent a population of patients with very different sets of issues when compared with adult patients. As has been taught by many learned clinicians, children are not small adults!
HLHS Stem Cell Therapy

TICAP
Transcoronary Infusion of Cardiac Progenitor Cells in Single Ventricle pts
Improved RV fx (46.9% to 52% EF)
Decreased tricuspid valve annulus size

Circ Res. 2015 Feb 13; 116(4): 653-64
Summary

Surgery for Congenital Heart Disease has improved the life of many patients over the years. Outcomes continue to improve however patients continue to face many challenges. Increased experience and continued data collection with careful analysis will guide future developments. These efforts will benefit the patients and families we care for.
The future’s so bright
I need sun glasses
For the things we have to learn before we can do them, we usually learn by doing them

Aristotle