Long Term outcomes after surgical interventions for Congenital Heart Disease (CHD)

Lazaros Kochilas, MD, MSCR, FAAP
Professor of Pediatrics
Director of Clinical Research
The shaping of the field of Pediatric Cardiology

Concepts

Tools

Pioneers
The ductus arteriosus is ligated and divided
(Robert Gross, Boston 1938)

The ductus arteriosus is ligated and divided (Robert Gross, Boston 1938)
A “ductus” is created: the Blalock-Taussig shunt (Baltimore, 1944)
Direct vision intracardiac correction of CHD
(Lillehei, Minneapolis 1955)

Controlled cross-circulation
From Fontan to Norwood:
Staged palliation for Hypoplastic Left Heart Syndrome

Norwood, 1983
Outcomes after operations for CHD – Pediatric Cardiac Care Consortium (PCCC)

- US-based registry of cardiac interventions
  - Includes 47 US centers
  - Clinical data collected 1982 – 2011

- Linkage to long-term outcome databases through December 31, 2014 for patients entered before April 15, 2003
  - National Death Index (NDI)
  - Organ Procurement and Transplantation Network (OPTN)
Crude and risk-adjusted mortality rate decreases over time.

**Graphical Description:**
- The graph illustrates the proportion of admissions ending in death over time from 1980 to 2010, categorized by risk level.
- Each line represents a different risk category: 1, 2, 3, 4, 5 & 6.
- The overall (unweighted) proportion is also shown.

**Data Citation:**
Long-term Outcomes of CHD

**Mild CHD**
- PDA
- ASD (secundum)
- VSD (isolated)

**Moderate CHD**
- ASD (primum)
- TAPVR and PAPVR
- CAVC or PAVC
- Ebstein’s anomaly
- PS (moderate or severe)
- Sub- or supra-AS
- VSD (complex)
- CoA

**Severe CHD (2-Ventricles)**
- Tricuspid atresia
- Mitral atresia
- Double-outlet ventricle
- Pulmonary atresia (all forms)
- d-TGA
- TAC
- L-TGA
- All forms of cyanotic heart disease
- Other complex CHD

**Severe CHD (Single Ventricle)**
CHD are at increased risk for premature death across all groups

Spector et al: JACC 2018; 71:2434-2446
Long-term outcomes improve over time

* Atrioventricular canal, Transposition of the great arteries and Single Ventricle

Spector et al: JACC 2018; 71:2434-2446
Risk of death decreases by age compared to the general population until 30 years of age.
Late causes of death in patients operated for CHD

- CHD: 58.8%
- CVD: 11.1%
- Other Congenital Malformations: 4.7%
- Respiratory Diseases: 3.6%
- External Causes of Injury and Poisoning: 8.2%
- Infections: 3.4%
- Neoplasms: 2.1%
- Other: 8.2%

McCracken et al: JAHA 2018
Causes of death over time

McCracken et al: JAHA 2018
Conclusions

- Survivors with repaired CHD have higher risk for premature death than the general population.
- Most deaths are related to the CHD or cardiovascular conditions.
- External cause of death is the only cause from which CHD patients are protected.
- Most of the risk is within the first 5 years from index surgery.
- Risk of death compared to the general population decreases by age until 30 years of age.
Survival for CHD have steadily improved the last 30 years.

For most patients with CHD the worst time is already behind them.

Additional efforts are ongoing to improve the outcomes of patients with CHD to reach the full life potential of the normal population.
Other imPOSSIBLE dreams in the past...

-When Jules Verne was suggesting that one day man will go to the moon, people thought this was an insane idea.

-Reaction to Graham Bell’s invention of telephone: This device is impractical and unlikely to be of use to anyone.

-First flight’s length (105 ft for 3.5” ) was shorter than the wingspan of a Boeing 747 (211 ft 5”)!
Acknowledgements

Emory University / CHOA
• Matt Oster, MD
• Courtney McCracken, PhD
• Michael Kelleman, MS
• Kim Kihwan, MS

University of Minnesota
• James Moller, MD
• Logan Spector, PhD
• Jeremiah Menk, MS
• Brian Harvey, BA
• Susan Anderson, BS

University of Rochester, NY
• Jeffrey Vinocur, MD

Children’s Mercy Hospital, Kansas City, MO
• James St Louis, MD

PCCC
• Participating centers
• Sarah Powell & Virgil Larson
• Students

Supported by:
NIH/NHLBI (R01 HL122392-01)
NIH/NCATS (UL1TR000114)