Keeping the Beat: Pediatric Cardiac Screening and Management of Congenital Heart Disease

Beyond Survival: “Real world” outcomes of neonatal cardiac repairs
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Learning Objectives
- Review risks for developmental risks for infants born with congenital heart disease
- Neurodevelopmental outcomes in neonatal cardiac patients
- Quality of life assessment following neonatal cardiac surgery

Modern Pediatric Cardiac Surgery

Pediatric Cardiac Surgery: A rapidly evolving field

1953-1969
- Early Palliation
- Elective Repair at ± 5 to 6 Years

1970-1982
- Primary Repair in Infancy

1983
- Primary Repair in Neonates - 1st report of Norwood procedure

Neonatal cardiac surgery mortality in 2012
- Transposition 2%
- TGA/VSD 6%
- Interrupted Aortic Arch 4%
- Truncus Arteriosus 8%
- Tetralogy of Fallot 3%
- Norwood procedure 20%

Mortality and Morbidity
- Traditional reporting of outcomes of neonatal cardiac surgery
- In the recent era, maturation of surgical techniques and perioperative care have significantly reduced mortality and morbidity
- These outcomes are easily measured, but not revealing of long-term functional status
“Real world” Outcomes

- Neurodevelopmental
- Psychosocial/behavioral
- Quality of Life

Scope of the problem

- 35,000 infants born each year in the US with significant congenital heart disease
- Nearly one-third require surgical correction or palliation in the first year of life

The deck is stacked: neurologic abnormalities common in CHD

- Babies with CHD higher incidence of structural brain abnormalities
  - MRI studies reveal increased periventricular leukomalacia, absent corpus callosum
  - Microcephaly has been reported in up to a third of CHD patients
- Genetic syndromes associated with developmental delay much more prevalent in CHD (eg. Trisomy 21, 22q11 deletion, Williams, Noonan’s, CHARGE)

Physiologic factors of complex CHD impact preoperative neurologic risk

- Ductal-dependent lesions at risk for profound acidosis, hypoxic-ischemic injury, inadequate tissue perfusion
- Preoperative seizures and intraventricular hemorrhage result from brain hypoperfusion
- Chronic hypoxemia in cyanotic lesions associated with cognitive impairment
- Congestive heart failure impacts postnatal brain development

Intraoperative factors

- Cardiopulmonary bypass
  - Potential for microemboli
  - Activation of inflammatory pathways
  - Anticoagulation can worsen existing IVH
- Deep hypothermic circulatory arrest (DHCA)
  - Cooling to 18C, venous blood drained and circulation is arrested
  - Risks of cognitive impairment when DHCA duration is greater than 50 minutes

Postoperative risks

- Cerebral perfusion and oxygen delivery may be compromised in the postoperative setting
- Seizures are the most common early neurological sequel in the neonate or infant
  - Immature brain has lower seizure threshold
  - Usually occur on 1st to 4th postoperative day
  - Most likely to occur if DHCA extends past 35 minutes
- Strong correlation of postoperative seizures with developmental delay, cerebral palsy and MR
Neurodevelopmental Outcomes: Boston Circulatory Arrest Study

- Prospective randomized study of 171 pts with TGA randomized to DHCA vs CPB
- 1 yr: PDI significantly lower in the DHCA group
- 2.5 yr: postoperative seizures were associated with language delays
- 4 yrs: IQ scores were significantly lower than general population (93 vs. 100)
- 8 yrs: significant deficits in visual-spatial and visual-memory skills, much higher incidence of attention and impulsivity

Neurologic outcomes following Fontan palliation

- Mean IQ in normal range
- > 75% at or above average achievement for school age
- Higher socioeconomic status predicted higher IQ and academic achievement

HLHS Neurologic Outcomes: Longer-term

- A study of 5-7 year olds found median IQ score of 88 (Predicted 100)  
  - Minor neurologic abnormalities in 55%
  - Cerebral Palsy present in 17%
- HLHS pts who had heart transplantation had similar IQ scores (mean 88.5)
- Increased incidence of attention and behavioral disorders
  - Incidence of attention deficit disorders 28%
  - Incidence of anxiety disorders 18%

What are the risks for poor neurologic outcomes in HLHS?

- Preoperative factors were the strongest predictor of poorer neurologic outcome  
  - multiorgan failure
  - preoperative seizures
  - younger gestational age
  - preoperative intubation
  - genetic syndrome
  - correlation between microcephaly and ascending aortic size
- Operative factors: Neurologic injury correlates with longer periods of circulatory arrest
- Postoperative seizures correlate with worse outcome

Functional status

- General health status of 150 8 yr olds following arterial switch for TGA
  - Physical Health Summary scores were equivalent to normal populations
  - Scores were much higher than age matched patients with asthma and JRA

Screening Program Congenital Heart Defects New Jersey Critical 171

Increased incidence of attention and behavioral disorders

NEUROLOGIC OUTCOMES IN HLHS

NEURODEVELOPMENTAL OUTCOMES IN BOSTON ARREST STUDY

NEUROLOGIC OUTCOMES FOLLOWING FONTAN PALLIATION

HLHS NEUROLOGIC OUTCOMES: LONGER-TERM

WHAT ARE THE RISKS FOR POOR NEUROLOGIC OUTCOMES IN HLHS?

FUNCTIONAL STATUS
Skills of adaptive behavior

- Impaired skills of self-care at 24 months following Norwood palliation mostly related to feeding
- Norwood patients had age-appropriate functioning in communication, leisure, preschool academics and self-direction
- Arterial switch patients had near normal functioning in all domains

Midterm quality of life

- 130 4 yr olds who underwent cardiac surgery before 6 weeks of age were given the PedsQL
  - Biventricular repairs had significantly impaired emotional functioning, social functioning and school functioning
  - Single ventricle patients had lower physical functioning than biventricular functioning
  - Older age at repair and lower postop cardiac output associated with lower HRQL scores

Long-term quality of life

- 306 survivors of TGA repair aged 11-15
- QOL scores actually higher in TGA cohort than normative controls (indicating better self-perceived health status)
- ASO children had significantly higher scores in physical functioning, bodily pain, mental health, and general health perceptions
- Use of DHCA associated with worse QOL scores

Summary

- Advances in surgery and perioperative care have dramatically improved the natural history of infants with complex CHD
- Children requiring early cardiac surgery face ongoing neurodevelopmental, psychosocial, and functional challenges

Future directions

- Perioperative care:
  - Regional cerebral oxygen saturation monitoring
  - Current Research: Develop biomarkers of postcardiotomy white and grey matter brain injury
- Operative strategies:
  - Minimizing circulatory arrest by utilizing continuous antegrade cerebral perfusion