Objectives

• Review congenital heart disease (CHD) demographics
• Understand factors that increase risk for developmental delays
• Learn the CHD “neurodevelopmental (ND) phenotype”
• Understand national CHD/ND evaluation and management recommendations
• Review role of PT and OT across continuum of care for this population

CHD Demographics

• CHD affects ~ 1% of live births
• 40,000 births/year in the United States
• 1/3 of infants require surgical or catheterization procedures in the first year of life

CHD Demographics

• Significant decrease in CHD mortality over past 20 years
  – Advances in CHD detection, surgical techniques, and perioperative management
• 85% of patients with CHD now survive into adulthood
• Now finding that significant proportion of CHD survivors have neurodevelopmental delays of varying degrees

High Risk Categories

• Neonates/infants requiring open heart surgery (cyanotic and acyanotic types)
  – Examples: HLHS, IAA, PA/IVS, TA, TAPVC, TGA, TOF, tricuspid atresia
• Children with other cyanotic heart lesions not requiring open heart surgery as neonate/infant
  – Examples: TOF with PA and MAPCAs, TOF with shunt without use of CPB, Ebstein’s anomaly

CHD & NEURODEVELOPMENT

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Considerations Following Open Heart Surgery

- Altered cerebral blood flow in utero and after birth may impact subsequent brain development
  - Increased risk for developmental delay and injury
- In utero brain development is delayed in children with complex CHD (~ 36 weeks)
  - Brain is less mature and more vulnerable at birth than full-term infant without CHD
- Methods of vital organ support during surgery (CPB and DHCA) may result in emboli to the central nervous system or a period of global ischemia
  - These CNS events may contribute to the presence of strokes or thromboses or to increased prevalence of PVL

Children with Cyanotic Lesions NOT Requiring Surgery During Infancy

- May avoid some of the inherent risks associated with early open heart surgery
- May still be at higher risk of developmental delay
  - Chronic hypoxemia caused by underlying CHD
  - Palliative or reparative surgeries later in childhood

Genetic Syndromes Associated with CHD

- Up to 30% of pediatric patients with CHD
  - Trisomy 21
  - Williams Syndrome
  - Noonan syndrome
  - CHARGE
  - VACTERL
  - DiGeorge (22q11.2 Deletion Syndrome)
- Developmental trajectory after surgery tends to be more delayed

Risk Factors for Developmental Delay

- Prematurity (< 37 weeks)
- Developmental delay recognized in infancy
- Suspected genetic abnormality or syndrome associated with developmental delay
- History of mechanical support (ECMO or VAD)
- Heart transplantation
- Cardiopulmonary resuscitation at any point
- Prolonged hospitalization (postoperative length of stay > 2 weeks)
- Perioperative seizures related to CHD surgery
- Significant abnormalities on neuroimaging or microcephaly

CHD “Neurodevelopmental Phenotype”

- Delays and deficits in the following:
  - Cognition (usually mild)
  - Gross and fine motor
  - Communication and pragmatic language skills
  - Visual Motor and visual spatial integration
  - Executive functioning
- Difficulties with:
  - Inattention
  - Hyperactivity
  - Impulsivity
  - Psychosocial adjustment

Developmental Abnormalities

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<th>Age at follow-up (months)</th>
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<th>Language</th>
<th>Social Skills</th>
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Executive Functioning

- Set of behaviors responsible for purposeful, goal directed activity
- Used to organize and direct cognitive activity, emotional responses, and overt behavior.
- It’s not the “know how,” but “how you do it”

- Inhibitory Control, Working Memory, Cognitive Flexibility are the core components, which build into planning, organization, problem solving, and integration of information.

High Rates of Inattention/Executive Dysfunction

- High rates of a lifetime ADHD diagnosis among adolescents with critical CHD
  - 19% for d-TGA and TOF without an identified genetic condition
  - 39% for TOF with an identified genetic condition
  - 53% for kids with single ventricle anatomy

- Executive dysfunction as early as preschool

- Problems across domains: inhibition, working memory, planning, flexibility

NEURODEVELOPMENTAL CARE IN THE HOSPITAL

Developmentally Supportive Care in the Hospital

- Infant Massage
- Skin-to-Skin
- Developmentally supportive positioning
- Cue-Based Feeding
- Cluster Care
- Non-pharmacologic pain management
- Environmental management
- Parent Education and Involvement

Promoting Developmental Care Framework in the PCICU

- Standardize developmental care practices
- Minimize environmental stress
- Create a developmentally supportive environment for critically ill infants
- Minimize ND morbidities associated with ICU environment
- Minimize physiologic stress and altered early experience
- Optimize weight gain
- Serve as an educator and ambassador for families
- Increase parental involvement in care
Role of Physical Therapy – PCICU, intubated and sedated
- Neuromotor assessment
- Complete infant massage training with caregivers
- PROM/AAROM exercise to maintain mobility
- Developmental positioning
- Educate parents and child on sternal precautions
- Maximize early mobility

Role of Physical Therapy – PCICU, VAD and ECMO considerations
- PT and OT will co-treat initially until safe mobility plan is created
- Infants most often sedated and paralyzed but children and teens often awake
- RT/VAD RN always present
- Goal is maximizing mobility opportunities and teaching movement patterns for independence while maintaining line safety

Role of Physical Therapy - Stepdown
- Improve muscle strength, balance, core strength, muscle coordination
- Promote physical activity in daily life
- Assist child in developing or regaining mobility
- Assist child in gaining freedom and independence to participate in recreational activities

Role of Occupational Therapy
- Developmental/Feeding assessment
- Implement developmentally supportive care
- Optimize positioning
- Splinting
- Support infant feeding
- Promote parent engagement and provide parental support
- Identify developmental concerns
- Promote advances in developmental areas/early intervention
  - Fine motor, visual motor, visual perceptual, sensory processing, coordination, cognitive and self-care skills
- Family education to prepare for discharge home

Factors Associated with Receiving Rehabilitation
- Palliative vs. corrective surgeries
- Prematurity
- Genetic syndrome
- Pre-surgical hospital stay of more than 1 day
- Prolonged CPB time
- Adverse events after surgery

WHAT WE ARE DOING AT DUKE
How did we elevate the standard of OT and PT practice?15

- Transition from a reactive (rescue) to proactive (prevention) model of care
- Adopt a holistic approach
- Standardization of care, assessments, and referrals
- Pre-operative neurodevelopmental evaluations
- Promote continuum of care and tracking of developmental progress over time
- Promote evidence-based personalized therapies that optimize adaptive skills
  - Development of individualized, tailored and targeted interventions
  - Identify therapeutic strategies to optimize outcomes and promote resiliency

Parent Education

- Early education packet
  - Role of PT/OT
  - Unique needs of hospitalized neonate/infant
  - What parents can do right now
  - What is to come
- Infant Massage
- Discharge education packet
  - Review of supine, prone, side-lying, and sitting
  - Patient-specific exercises
  - Equipment recommendations
  - Therapy follow-up
  - Sternal precautions
  - CDC month-by-month development

PT Inpatient Workflow

Admission: Automatic PT Consult
Early Education – Assign Primary PT
Surgery/Recovery (on hold)
PT Intervention & I.M Education
Discharge Education

PCICU Initiatives

- Lighting & Noise
- Sleep and Clustered Care
- Policy/Guidelines
- Developmental Care Levels
- Supportive Equipment
- Positioning Guidelines

Cycled Lighting

- Nursing responsibility
- Helps regulate circadian rhythms
- Lights on during day shift
- Lights off during night shift
- Provide dedicated nap times
- Minimize noise level at bedside
- IV pump and phones set at lowest volume
- Respond to all bells promptly
- Use of noise machines and mobiles

Clustered Care

- Clustering several nursing interventions instead of spreading them out
- Allows for longer periods of restorative rest
- Timing nursing assessments with feeding times
- Transition infants to "clustered care" once they are on bolus feeds
Caregiver Communication

- Photo Journals
  - Located in parent mailbox at bedside
  - To be filled out weekly by therapists
  - What am I working on in therapy?
  - What is my progress?
  - What can you help me do this week?
- Blue Therapy Folders
  - Located in therapy file drawer in PCICU and Stepdown
  - Use to consolidate all bedside and caregiver education
- Growth Charts
  - At bedside – nursing staff fills out
- Developmental Care Levels

Developmental Care Levels

- Developmental Care Level to be determined every day in medical rounds
- Visual schematic posted at bedside
- Nursing, therapy, and parents working together to make sure activities are completed consistently and safely

Date:

Developmental Care Levels

FAMILY

- Hold me in your lap or on abdomen.
- Participate in my therapy.
- Hold your phone, books, and toys for stimulation.

CARE TEAM

- Supportive Equipment
  - Chairs
  - Noodle play arches
  - Splinting
    - Wrist and hand splints
    - Thigh binders
    - Figure 8 ankle splints
  - Mirrors, mobiles, vibration boxes, sound machines
  - Positioning equipment
    - Bendy bumpers
    - Snuggle ups
    - Frederick T. Frog
    - Z-flo

Supportive Equipment

**Developmental Follow-Up**

- Pediatric Cardiac Neurodevelopmental Clinic (PCNC)
- Referral to CDSA for early intervention evaluation
- Direct referral to outpatient PT or OT for more acute issues
  - Torticollis
  - Feeding

**PCNC**

- Opened October 2018
  - 2 days/month, increasing to 4 days/month in 2020
- All infants with CHD
- Exceptions: prematurity, other genetic condition with separate developmental clinic at Duke
- Staffed by neonatology, developmental psychologist, PT/OT/ST, social work
  - Neurology and nutrition are on-call
- Follow up intervals are based on risk stratification, developmental needs and current method of feeding
  - Will get formal neurodevelopmental assessments at 1 and 2 years old (Bayley-III)

**Summary**

- Standardization of Care
- Understand periods of risk and cumulative risk burden
- Optimize developmental transitions at key intervals across the lifespan
- Identify high-risk CHD populations
- Neurodevelopmental problems may take years to manifest
- Anticipatory guidance/planning
- Family-centered care model across the care continuum
- Evidence-based practice
  - science-driven profession/apply most up-to-date research to service delivery

**PARENT AND FAMILY IMPACT**
Parental Mental Health

• Clinical levels of psychological distress are apparent in 1/3 of mothers and almost 1/5 of fathers of infants with CHD

• Maternal distress was NOT predicted by disease factors.

• Maternal mental health predicted by:
  – Maternal coping skills
  – Mother’s understanding of the diagnosis
  – Degree of family cohesiveness

Rates of Traumatic Stress in Parents

• Prenatal diagnosis: 39% of mothers

• Hospital discharge: 32% of mothers and 27% of fathers

• One year post-diagnosis: 37% of parents

Potential Family Impact in CHD

• Diagnosis: Prenatal or Postnatal
  – Shock
  – Sense of loss
  – Acceptance
  – Preparation

• Preoperative:
  – Attachment issues
  – Feeding difficulties
  – Medical treatment
  – Financial burden
  – Relationship impact

• Perioperative:
  – Increased anxiety
  – Concern about mortality

• Preschool & School Age
  – Lifestyle adjustment
  – Employment issues
  – Physical activity and sport participation
  – Sibling impact

Family Communication

• Parents appreciate:
  – Support, hope and reassurance to reduce guilt
  – Visual aids to understand diagnosis, trusted websites
  – Typical surgical course and hospitalization
  – Potential outcomes/success rates

• Parents want:
  – More information about peer support
  – Preparation for children not following “typical course”
  – Information on common outcomes and complications (feeding tube)
  – Preparation for long-term challenges that children may face
  – Preparation for impact on family (PTSD, financial, day-to-day)

• Tools and strategies to prevent, detect, reduce problems

Create Opportunities for Parent Empowerment

CHD is a life-long diagnosis

• Support parental adjustment
• Enhancing mothers’ and fathers’ coping strategies
• Promote early involvement and effective interactions related to feeding, caregiving, social and sensory stimulation
• Minimize stress by providing knowledge, coping skills
• Teach parents active, generalizable, problem-solving strategies
  – Feeding problems, positioning, developmental needs

TAKE AWAY THOUGHTS
Take Away – Early Intervention

• Consider parent stress responses and current coping
• Focus on prone positioning, scar massage, identifying/treating torticollis, identifying/treating ROM impairments
• Consider abdominal activation in all positions
• Standardized assessment: AIMS

Take Away – School age

• Consider ND phenotype
  – Assess for difficulties in motor planning, executive function, and attention
• Most kids are not limited in physical activities but may have lower SpO2 limits
• Assess for global strength and endurance
  – Focus on proximal strength, thoracic mobility, flexibility
• Standardized assessment: VMI, BOT-2

Take Away – Preschool

• Focus on supporting motor development
• Consider if future surgeries are planned and monitor activity tolerance
• Monitor for executive dysfunction
• Standardized assessment: Bayley-III, PDMS-2

Take Back - OT

• EI: feeding evaluation, early play skills, sleep
• Preschool: fine and visual motor development, peer interactions, sensory processing
• School age: visual-motor integration and executive function (CHD ND phenotype), proximal strength, fine motor skills, sensory processing,