To Heal a Broken Heart: Long Term Implications of Congenital Heart Disease

Learning Objectives

• Identify long term implications of congenital heart disease (CHD).
• Incorporate interventions targeted for neuro-developmental, psycho-social and developmental outcomes for children and adolescents with CHD.
• Evaluate the PNP and their fellow pediatric-focused APRN’s role in caring for children and adolescents with CHD.
• Apply concepts associated with transition of the adolescent with CHD.

Impact of Congenital Heart Disease (CHD)

• ~ 9 per 1,000 live births of US population
• One of the most common birth defect in the US
• Causes:
  – Multi-factoral
    • Genetics, environment, maternal Diabetes, drug exposure.
  – Not the mother’s fault
• Defect occurs in the first 12 weeks of pregnancy.

Types of Congenital Heart Defects

• A large variety
  – VSD, ASD, PDA, AV canal
  – AS, PS, Coarctation
  – MR, MS, TR, TS
  – Complex lesions
  • TOF, TGA, Truncus, and single ventricle anatomy

Genetics and Defects

• Approximately 30% of pts with CHD have genetic abnormality
• Down’s syndrome
  – 50% will have congenital heart disease
  – 50% of those will have AV canal
• Turner’s syndrome
  – Left heart obstructive lesions
• DiGeorge Syndrome
  – TOF, Truncus, etc.

Disclosures

• No Disclosures
Congenital Heart Surgery

- Began in 1940s.
- Timeline:
  - 1945: First Palliative procedure for TOF-BT shunt (Blalock-Tausig)
  - 1954: First surgical correction of TOF-Lillihei-cross circulation (used parent as oxygenator)
  - 1953: First successful open heart surgery CPB-18 year old female with ASD (Gibbon, Thomas Jefferson Hospital, Philadelphia)

Timeline:
- Prior to 1980's single ventricle lesions considered universally fatal.
- 1980s: Staged repair of Fontan completion
  - Venous return routed to pulmonary circuit and the SV pumping to systemic circulation

Not Just Heart Disease:

- Other organ systems
  - Midline defects: Kidney, Brain abnormalities, Cleft lip and palate, GI and GU abnormalities.
- Heterotaxy syndromes
  - Situs inversus
  - Malrotation
  - Asplenia
  - Polyspleni

From the Beginning

- New areas of research have shown these children have risk from the beginning
  - Abnormal brain structure
  - Alterations in cerebral blood flow
  - Impaired volumetric brain growth
  - Brain maturations at term=35 weeks
  - Increase risk for acquired brain injury
  - Microcephaly
    - As high as 33%
  - Open Operculum
    - Speech and Oral Motor
    - Poor drive/coordination of feedings

From the Beginning

- Operation with Cardiopulmonary bypass (CPB)
  - Incidence of stroke 10%
  - Incidence of periventricular leukomalacia 16% (pre) to 48% (post)
    - White-matter brain injury.
    - Characterized by the necrosis of white matter near the lateral ventricles
    - Infarcts common in the motor tract
From the Beginning

• Cyanosis (critical period of brain development)
  – Prenatally: TGA, SV, and TAPVR
  – Prolonged in SV- up to 3-5 years
• FTT
  – Poor growth parameters
    • Increased caloric needs
    • Poor energy levels to promote intake

Social/Family Dynamics

• Parents asked to be:
  – Mother/Father
  – Nurse
  – Social Worker
  – Developmental Specialist
  – Special Education Teacher
  – Child Advocate

Survival Rates

• Vary by disease complexity:
  – 95% for simple (AS, PS, ASD, VSD)
  – 90% for moderate (coarctation, AVSD, TOF)
  – 80% for complex (single ventricle, truncus arteriosus, TGA)
• 1.0-2.9 Adult Survivors with CHD

Long Term Implications

• As these infants are now living to adulthood there has been a shift in focus (surviving to thriving):
  – Long term complications
  – Neurodevelopmental Outcomes
  – Transition to adulthood

From the Beginning

• ECMO/CPR
  – decreased cerebral perfusion
• Perioperative Seizures
  – Most related to adverse neurodevelopment outcomes
• Prolonged hospitalizations
  – > 2 weeks-increased risk for poor cognitive function

Social/Family Dynamics

• Dealing with Chronic Illness:
  – Increased risk for:
    • Divorce
    • Psychological Disorders
    • Fragile Child
    • Economic Issues
    • Chronic Sorrow
Long Term Complications

• Reoperations:
  – Most congenital heart surgery is palliative not corrective:
    • Single Ventricle: Requires 3 operations by age of 3 and then multiple cath procedures and possible transplant
    • TOF: Requires multiple Pulmonary valve replacements, possible Branch PA interventions and PV reductions.
    • AS: Multiple aortic valve replacements etc.
    • SubAS: can regrow and require multiple reoperations

• Arrythmias
  – Cardiac Surgery
    • Scar Tissue
    • Cuts in Ventricle and atri
    • Underlying disorders
  – Atrial and Ventricular Enlargements
    – May require surgical and interventional Ablation procedures.
    – Lifelong Medications

Long Term Complications

• Single ventricle anatomy
  – Protein Losing enteropathy
  – At risk for ventricular failure/transplant
  – Collateral development
  – Pulmonary AVMs
  – Thromboembolic
  – Clotting problems

Management of Long Term Complications

• Make sure:
  – Have specialist care needed
    • Pediatric Cardiologist
    • Pediatric Cardiothoracic Surgeon
    • Congenital heart disease hospital
    • PT/OT/Speech Therapy
    • Other Specialists to manage co‐morbidities

Management of Long Term Complications

• Medications:
  – Arrhythmia
    • Beta Blockers
    • Digoxin
    • Amiodarone
  – Pulmonary Vascular Resistance
    • Sildenafil
    • Tadalafil
Management of Long Term Complications

• Medications:
  – Function
    • Carvedilol
    • Digoxin
  – Anti-coagulants
    • Aspirin
    • Plavix
    • Coumadin

Neurodevelopmental outcomes

• Pediatric Patients with CHD are at risk for:
  – Developmental Delays as great as 55%
  – Lower IQ
  – Behavioral Problems-40-50% increased (ADHD)
  – Psychosocial Issues-18.6% or higher
  – Impaired Living Skills
  – QOL-reduced health status, exercise, psychosocial impairments

Screening for Developmental Delays

• AHA statement (2012):
  1. Medical Home Model of Care
  2. Risk Stratification of patients with CHD
  3. If High risk:
    - Referral for formal development and medical evaluation
    - Early intervention services
    - Periodic Evaluations for DDs at 12-24mths, 3-5yrs, and 11-12yrs
    - Referral of young adults for higher ed or vocational counseling

Screening and Medical Home

• All children with CHD in a Medical Home
  – Individualized Plans
  – Collaboration with specialists
  – Comprehensive Record
    • Hospital based care
    • Imaging studies
    • Genetic Testing
    • Developmental Evaluations
    • Plan of long term surgical care
    • Transition Record

Risk Stratification

<table>
<thead>
<tr>
<th>High Risk Children with Congenital Heart Disease</th>
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<tbody>
<tr>
<td>1. Neonates or infants requiring open heart surgery</td>
</tr>
<tr>
<td>2. Children with cyanotic lesions not requiring open heart surgery</td>
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<tr>
<td>3. Comorbidities:</td>
</tr>
<tr>
<td>Prematurity (&lt;37 weeks)</td>
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<tr>
<td>DD in infancy</td>
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<tr>
<td>Genetic abnormality</td>
</tr>
<tr>
<td>History of Mechanical support</td>
</tr>
<tr>
<td>Heart transplant</td>
</tr>
<tr>
<td>CPR</td>
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<tr>
<td>Prolonged Hospitalisation (&gt;2 weeks)</td>
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<tr>
<td>Perioperative seizures</td>
</tr>
<tr>
<td>Abnormalities on neuroimaging or microcephaly</td>
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</tbody>
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Screening

• Low risk and High risk:
  – Follow AAP screening guidelines
  – Surveillance at every well child
  – Elicit and attend to parental concerns
  – Developmental history
  – Observations of the child
  – Presence of Risk and Protective Factors
  – Screen for:
    • Screening Tools, Behavioral/Psychosocial Issues, Autism Spectrum Disorder, Fine/Gross Motor Skills
High Risk Evaluation

Screened and referred for formal evaluation
- If identified, early interventions and other medical evaluations

- Screened and referred for formal evaluation
- If identified:
  - Early intervention
  - Genetic Evaluation
  - Structural Brain Imaging
  - Academic and Behavioral Issues
  - Psychosocial Adjustment
  - Self Management

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School Age and Adolescents

- Monitor Academic Difficulties
  - May need learning disabilities testing
  - IEP

- Targeted interventions for:
  - Visual spatial skills
  - Writing skills
  - Longer testing
  - Life skills
  - Coping Adaptive abilities
  - Supporting Networks
  - Empowering experiences

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Transition to Adulthood

- Transition is defined as
  - Process of moving from the pediatric medical system to the adult medical system

- AAP Goal: “Is to maximize lifelong functioning and potential through the provision of high-quality, developmentally appropriate health care services that continue uninterrupted as the individual moves from adolescence to adulthood”

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Complicated in CHD

- Only 45% have access to an adult physician who is familiar with their condition
  - Adult Congenital Heart Disease Clinics
  - Institutions where Congenital Heart Teams work with Adult Teams

- Normal life transitions may be complicated by
  - Developmental Delays
  - Psychosocial Issues
  - Impaired abilities
    - Exercise
    - Work
    - Insurance

Transition Recommendations

Recommendations Regarding Timing and Support of Families
1. Timing based on emotional maturity and developmental level
2. Adolescents and parents engaged in transition plan
3. Adolescents/parents should be asked about understanding of disease, health status, restrictions, and future goals
4. Adolescents and parents encouraged to share concerns about QOL
5. Acknowledge adolescent and parental concerns and fears
6. Pediatric Cardiology should work with adolescent/parents/PCP to create transition binder or passport (including cardiac destination)
7. Providers should begin to direct health discussions toward the adolescent
8. Discuss QOL issues in private with adolescent

Transition Recommendations

Cardiology Follow up Care and Surgical Issues
1. Follow up studies performed by providers with expertise in CHD
2. Regular (~1 year) follow up is required to maintain hemodynamics and prevent complications
3. Non-surgical/surgical interventions need to be individualized and performed in centers with CHD expertise
4. High or mod risk surgeries with complex CHD should be performed in centers with CHD expertise
5. Elective surgery should have preoperative planning with ACHD experts, anesthesiology, and surgical services
6. Intraoperative management should be performed by an anesthesiologist familiar with physiology associated with CHD.
Transition Recommendations (Anticipatory Guidance)

Genetic Counseling and Contraception
1. Should have 3 generation family hx including CHD, pregnancy loss, and DD
2. Genetic evaluation with testing if indicated
3. Recurrence risk and reproductive counseling-prenatal screening or diagnostics for pregnant CHD patients
4. Counseling individualized considering emotional maturity and impact of counseling
5. Begin reproductive counseling early in adolescence (12 years of age)
6. Healthcare provider who are knowledgeable about CHD and reproductive health do the counseling
7. Document interventions to modify high risk behaviors
8. Document prescribed contraception for males and females
9. Comprehensive reproductive services including adolescent gynecology
10. Education materials and resources provide to adolescents and families (stress importance of planning of preg)

Exercise
1. Given general benefits of exercise, some form of routine exercise is advisable for all pts
2. Exercise testing should be performed to establish baseline and repeated for symptoms/concerns
3. Based on results of testing, cardiologist should discuss exercise with family, revise based on clinical status
4. Exercise prescription should include type, HR goals/limits, duration, frequency, and guide to advance if possible (things to avoid and based on guidelines-Bethesda Guidelines)
5. If limited or adverse hemodynamics to exercise, medically supervised cardiac rehab may be beneficial

Education/Employment, Insurance
1. Develop a structured education/teaching plan based on the individual needs (academic abilities, education levels, and developmental maturity)
2. Counsel early in adolescence to identify interests and possible need for education or vocational training
3. View every patient as employable and tailor career/employment counseling to the capabilities
4. Counsel about insurance issues prior to leaving parents policy or loss of eligibility for children’s services
5. Discuss relationship between education/vocational choices and access to insurance should occur early and guide planning.

Conclusion
• Helping Families of CHD
  – Acknowledge feeling of guilt
  – Acknowledge Chronic Sorrow
  – Assist with special education measures
  – Early identification of learning needs
  – Maintain health at maximum and encourage exercise at patient’s level
  – Monitor for and manage complications early

• Long Term Outlook
  – Need for specialists that are prepared to care for these children/adults
  – Adult cardiologists vs Pediatric cardiologists
  – Cross over care
  – Aging population of SV patients with new complications
  – Oldest patients in 40’s
  – Some unknown
  – Advocate for additional research in care and treatment of these complex and precious children
References