Meeting the Needs of the Adult Congenital Heart Disease Patient

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I have no relevant financial relationships to disclose.

Background
- Cardiac surgery for CHD began mid-1950s
  - Initially performed mainly older children and adults
  - Complex defects not approached until mid-1970s
- Contemporary population ACHD patients represent various natural histories
  - Those who survived without surgery
  - Those with palliative or reparative surgeries starting childhood or adulthood
  - Those with CHD addressed as neonate or infant

Background
- Survival to adulthood varies by severity of CHD
  - 98% with mild, 96% with moderate, 56% severe CHD
- Population of adults with CHD has increased dramatically over past 3 decades
  - 2012 estimated >1,000,000
  - >50% with complex CHD
  - Only 1:3 receiving recommended level of care
  - Unaware need for long-term care; lost to follow-up
Late CV and Non-Cardiac Complications

Neurodevelopmental Influences
- Genetics
- Operative strategies
- Neurologic / Circulatory risk factors
- Environmental / Psychosocial impact

Genetics
- Estimated at least 15% CHDs associated with genetic conditions
  - Majority of ACHD have not had testing
  - Subset found to have genetic variation in childhood may not have received counseling
  - Service unavailable or too young to participate
  - Topic resurfaces when patient becomes parent
  - Phenotypic abnormalities suggestive of syndromic CHD may have been missed
  - Physical findings often subtle
  - Late-onset related disorders may only exhibit in adulthood
  - Negative testing in childhood may warrant re-testing as more sensitive tests developed

Operative Strategies
- Operative risk factors influenced by surgical era
  - DHCA
    - Popular 1960s children & adults; 1970s infants
    - By late 1980s concern regarding effect on neuro development
  - CPB
    - Patients cyanotic for years until deemed candidate for bypass
    - Prior 1980s membrane oxygenators and cannulas limited
    - Microemboli linked to chronic cognitive decline
    - Depression, apathy, inattention, dementia
  - Hospitalization - prolonged stays, visitation very limited
Neurologic/Circulatory Risk Factors

- Late Neurologic events
  - Ischemic stroke secondary to
    - venous emboli crossing ASD or PFO
    - atrial fibrillation or flutter
  - Intracranial aneurysms in patients with coarctation
    - hemorrhage even in absence of HTN
  - Chronic cyanosis
    - brain abscess with R→L shunt uncommon but reported
    - secondary erythrocytosis and platelet dysfunction

Environmental/Psychosocial Impact

- Psychological health-anxiety and depression well reported
  - Living with knowledge of significant risk of dying
  - Health declines unexpectedly when perceive themselves as "cured" by childhood interventions
  - Review of congenital history/procedures unsettling
  - Social difficulties due to impaired peer relationships, family overprotection
    - Possible body image issues -scarring, small size, cyanosis
    - Delayed progression to independent adulthood
      - never leaving home or marrying
    - Poor coping or fear resulting from childhood experiences
      - untreated pain, developmentally inappropriate medical care

Case Example - Bill

- Late repair
- Fragmented cardiac and primary care
- ACHD care starting age 51
- Multiple sequelae – cardiac and non-cardiac
- Education / employment challenges
- Medication management and "non-compliance"
- Lacked a social support system
- Psychosocial stressors
- Poor navigation of health care system - "medically naïve"
- Neuropsychology consult age 56

ACHD Patient Needs

- Medication management
- Effective transition to adult-centered health care
- Education and career planning
- Guardianship determination/assessment
- Psychosocial support
Medication Management

- Generic medications and $4 Prescription Plans
  - Limit combination medications
- Pill boxes / medicine organizers
  - Smart phone applications
- Electronic Medication Management Systems
  - E.M.M.A.

Care Transition

- Transition to adult primary care and adult-centered CHD care
  - Who provides best medical home - engage PCP for management non-cardiac health concerns
- Consequences of lapses in care: At time of presentation these patients more likely to:
  - Be symptomatic, have inappropriate medication regimen, require additional diagnostic testing, require urgent cardiac intervention
- Interdisciplinary collaboration is the key
  - draws upon expertise of pediatric and adult providers, surgeons, nurses, allied health professionals is the key
- Multi-site collaboration and sharing
  - Learn from successes of other programs; don’t reinvent the wheel

Education and Career Planning

- Utilize ACHA “Personal Health Passport”
  - If relocating ensure pharmacy, lab, primary care services
- Employment benefits: group insurance > salary
- Individuals with Disabilities Education Act (IDEA) mandates “free appropriate public education” for all children with special needs
  - Not long ago only options 18-21 year olds with intellectual disabilities was to stay in high school in order to receive services under the Act.
  - Now many colleges provide postsecondary programs if student’s IEP team determines needs best met through such a program
- Division Vocational Rehabilitation option for cognitively intact patients with limited physical capacity
  - Every state has a vocational rehabilitation agency to assist individuals with disabilities to prepare for, get, keep, or regain employment.
- Disability determination – resources to guide ACHD patients

Guardianship

- Topic best addressed by PCP or cardiologist
- Consider if cognitive limitations lead to concerns about medical or legal/money decision making
  - begin discussions during adolescence
- Competency evaluation by doctor, psychiatrist or psychologist needed
  - only good for 6 months before court hearing
- Utilize local adult guardianship resources
  - CHW offers guardianship clinic for qualifying adults
  - Wisconsin Guardianship Support Center
Psychological Support

- Screening tools for adults readily available
  - Hospital Anxiety & Depression Scale, Satisfaction with Life Scale
- Treating psychosocial disorders very challenging
  - Few ACHD program with resource of adult psychologist
  - Inpatient services for psychiatry limited; psychology services available but poor transition to outpatient
  - Limited provider pool and financial coverage limiting factor
  - Need to identify best treatment practices for anxiety / depression and target interventions to minimize development
- Chronic pain as contributing factor not be underestimated
  - Referral to chronic pain clinic / specialists

Summary:

- Adults with CHD is a rapidly growing population
- Cognitive, psychosocial and mood disturbances are well-described in adult survivors of CHD
- Neurodevelopmental concerns in these adults may manifest differently than contemporary CHD pediatric population
- Providers must be adaptable and innovative to meet the needs of these individuals
- Expansion of resources for this population needs to be a priority for the congenital cardiology community

Thank You

Contact Information

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