Adult Congenital Heart Disease: The Scope of the Problem

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Congenital Heart Disease

- Recognized for centuries
- Most common of all major birth defects
- 5-9/1,000 live births
  - Higher if BAV and PFO included
- Wide variety of lesions
- Natural and ‘unnatural’ histories
Growth and Impact of CHD Surgery

- 1938 – Ligation of a patent ductus arteriosus (Gross & Hubbard)
- 1944 – First Blalock-Taussig shunt (Baltimore)
- 1945 – Resection of an aortic coarctation (Crafoord and Gross)
- 1953 – Gibbon closes ASD successfully using heart-lung machine
- 1954 – Repair of Tetralogy of Fallot (Lillihei, U Minn)
- 1957, 63 – Senning and Mustard atrial switches for D-TGA
- 1968 – Fontan procedure (Bordeaux)
- 1975 – Arterial switch for D-TGA (Jatene, Brazil)
- 1981 – Norwood procedure for HLHS (Boston)
CHD Pioneers
CHD Surgical Mortality According To Age And Era

Slide Courtesy of Robert Sade, MD
Changing Times

Improving Natural History of Children with CHD = More Adults with CHD

1960s
- Surviving to Adulthood: 35%
- Died in First Year: 15%
- Died within 18 Years: 50%

2010
- Surviving to Adulthood: 85%
- Died in First Year: 10%
- Died within 18 Years: 5%

Age at Death in Patients With CHD

TOF: Palliation and Repair
Adult Congenital Heart Disease (ACHD)

- >39,000 infants born annually with CHD
- 90% survive to adulthood
- 20-25,000 operations for CHD annually
- Currently over 1,500,000 adults in US with CHD
Birth of a (sub) Subspecialty

- In the Beginning...
  - Boston Children’s/ Mass General 1974
  - UCSF and UCLA 1980
  - Mayo 1988
  - Penn State Hershey 1991

- Currently 110 self proclaimed programs in US
- Little formal training available for ACHD physicians
Program Establishment

Cumulative Number of Sites

Year

Davidson, 2006
ACHD Programs in US and Canada, 2017

www.achaheart.org
Adult Congenital Heart Association

- 1998 at Boston Children's
- Now focuses on education, research and advocacy
- ACHA program accreditation
Goals in ACHD

- Sufficient, trained providers
- Sufficient number of Regional Centers
  - Research
  - Clinical care
- Web of referral relationships
- Improved evidence base for practice
Clinical Challenges in ACHD

- Relatively small numbers of patients/lesion
- High percentage of patients ‘lost’ to follow up
- Large burden of complex clinical care
  - History, lesion- and operation-specific
- High demands of imaging - anatomic, physiologic
- Imposition of acquired on congenital heart disease
- Consequences of aging
ACHD: Changing the Conceptual Framework

- Acquired Heart Disease
- CHD
- Congenital Heart Disease (± acquired)
- Lifespan
Societal Challenges in ACHD

- Transition of care
- Location of care
- Working relationship between Medicine and Pediatrics
- Societal recognition (coding, insurance, disability)
- Reluctance to refer
- Insurance access and adequacy
- Neurocognitive issues
Timing of Transition

- Difficult to have a ‘one size fits all’ approach
  - Based on pt’s developmental stage

- Gradual process
  - Emphasize need for autonomy

- Start to educate teens
  - Diagnosis, meds, residual problems, symptoms to be worried about
Location of Care

- Adult Only Hospital?
- Free-standing Children's Hospital?
- “Mixed” hospital?
Residuae and Sequelae of CHD Surgery

- Ventricular Dysfunction
- Vascular Problems
- Valve Disease
- Non-cardiovascular
- Prosthetic materials
- Electrophysiologic
  - Myocardial scars
  - Disorders of impulse formation and conduction
- Endocardial and myocardial effects

CHD Surgery is corrective, not curative!

Perloff, Circ, 1991
Causes of Death in ACHD

- Perioperative: 36 (18%)
- CHF: 41 (21%)
- SD: 51 (26%)
- Other cardiovascular: 36 (18%)
- Non-cardiac: 33 (17%)

Oechslin, Am J Cardiol, 2000
SCD in ACHD

Sudden Death Also Important Cause of CHD Mortality

Death in ACHD

- Avoidable in 21%
  - 341 deaths in 4500 British pts
  - >50% of deaths in Class I or II patients
  - Errors in arrhythmia management
  - Lack of knowledge concerning pt.’s disease
  - Poor operative technique or planning
  - Poor postoperative care
  - Cyanotic and Fontan patients

Somerville, 1995
ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease)

Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

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Recommendations for Care

- Every ACHD patient should be seen by an ACHD specialist at least once.
- Simpler lesions can be treated locally:
  - closed PDA
  - repaired secundum ASD
  - isolated small or repaired VSD
  - bicuspid AV without root involvement
  - mild pulmonic stenosis

32nd Bethesda Conference, 2001
US ACHD Guidelines, 2008
ACCHD Guidelines

ACC/AHA Guidelines for Monitoring ACCHD

<table>
<thead>
<tr>
<th>Recommended visits to an ACCHD center:</th>
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<tbody>
<tr>
<td>Simple</td>
<td>At least once to determine needs for future follow-up</td>
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<tr>
<td>Moderate complexity</td>
<td>Every 12 to 24 months</td>
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<tr>
<td>Great complexity</td>
<td>Every 6 to 12 months</td>
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Who Should Provide Care

- Pediatric Cardiologists??
- Adult Cardiologists??
- Ideally, an ACHD physician!
But the Problem Is...

- Too few ACHD docs
- Too few ACHD fellowship programs
- Lots of current ACHD docs nearing retirement age....
CHC PACC
(Program for Adult Congenital Cardiology)

Interventional Cardiology
Adult EP
CT Surgery
Children’s Heart Center
Hepatology
CICU
Hospitalist Team
Social Work
PICU
Critical Care Team
Endocrinology
Interventional Radiology
Sonographers
Device Clinic
Peds EP
Psychology
MFM and OB/GYN
Children’s Heart Foundation
Where We are Headed...

- ACHA Accredited Comprehensive Care Center
- Fontan FIT Program
- Education Programs
  - Nursing, Inpt. providers,
  - Residents, cardiology fellows
  - PATIENTS!
Thanks