Congenital Heart Disease
Continuum of care

Extent of the problem

5-10 per 1000 live births.
- 1.5 per 1000 have complex CHD
- Untreated - ONLY 15-25% survival
- Treated - 90% survival to adulthood


POFPS 42nd Annual CME Symposium
August 4-6, 2017
The Reason for ACHD

Incidence of CHD: 5-10/1000 live births
Fetal Diagnosis
Improved Surgical Techniques
Lower Perioperative Mortality
Increased Midterm Survival
Increased Early Survival
Early Complete Repair
Advances in NICU Care

ACHD: Incidence

Patients Reaching Adulthood with CHD

Survival rate from Year of Birth (1940-2000) by Complexity of Congenital Heart Disease

<table>
<thead>
<tr>
<th>Year of Birth</th>
<th>Simple</th>
<th>Moderate</th>
<th>Complex</th>
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<tbody>
<tr>
<td>1940 - 1959</td>
<td>90%</td>
<td>55%</td>
<td>10%</td>
</tr>
<tr>
<td>1960 - 1979</td>
<td>95%</td>
<td>65%</td>
<td>50%</td>
</tr>
<tr>
<td>1980 - 1989</td>
<td>95%</td>
<td>90%</td>
<td>80%</td>
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</tbody>
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Care of the Adult with Congenital Heart Disease. 32nd Bethesda Conference. JACC 2001; 37: 1161-98

Devyani Chowdhury, MD, Director
Lancaster, PA
“Congenital Heart Disease”
Devyan Chowdhury, MD

ACHD “survivor” group?
Half million adults with complex CHD**
Only 30,000 receive follow-up care in a ACHD center**

**Webb G. The long road to better ACHD care. Congen Heart Dis 2010; 5: 198–205

Lesions That Make it to Adulthood

- 25% of adults
  - mild form of CHD
  - without need for surgery or interventional catheterization
- Bicuspid Aortic valve
- VSD – small, restrictive
- ASD – secundum
- Pulm. stenosis – mild
- Mitral valve prolapse
- Isolated Congenitally Corrected Transposition of Great Arteries

Adult Congenital Heart Disease (ACHD)

- 15-year survival rate post surgical
  - 80% for complex CHD
  - 95% simple CH
- Currently ACHD is more than Peds CHD
- Approximately 1.5 million adults with CHD worldwide
- 20,000 – 30,000 new CHD patients reach adulthood and add to existing pool of patients
Congenital Heart Disease

Devyani Chowdhury, MD

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INTERSTAGE MORTALITY
Fontan Circulation

- Flow pattern highly dependent on respiration, maximally increased during spontaneous inspiration (sub-atmospheric intra-thoracic pressure)
- Flow attenuation or even reversal seen during the positive-pressure phase of IPPV

The morphologic left ventricle supports a better hemodynamic profile (tricuspid atresia vs. HLHS).

Classical Fontan

Fontan Modifications
“Congenital Heart Disease”
Devyani Chowdhury, MD

Single Ventricle Before and After Fontan

ACHD “survivor” group?
- Multiple physical and/or mental disabilities (musculoskeletal, cerebral palsy, Down’s syndrome, CPB related neurological injury)
- Living with parents or have complex psychosocial needs (multiple scars, lack of self esteem and/or independence)
- Co-morbidities superimposed on already fragile matrix
- Anxious Frequent Flyer patient and/or family!!

Psychosocial issues
30 patients with complex CHD followed for 30yrs
- 90% graduated high school
- 62% graduated from college
- 90% worked regularly
- PTSD (12 pts) Panic disorder (5pts) Major depression (4 pts) – no formal psychiatric treatment!!
- Anger, Anxiety, Apprehension common sentiments


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Presentation of ACHD

- Revision of childhood corrections.
- Newly recognized / untreated CHD
- New rhythm disturbances and/or the need for pacemakers
- Pregnancy issues
- Exercise and training issues
- Adult cardiovascular or other age-related health concerns

ACHD-“survivor” group?

- ACHD symptoms are subtle and not noticed until late!!!
- Noncompliance due to minimal or no symptoms or INSURANCE denial !!!
- Adolescent - invincible and in denial
- Lost to follow-up when they move for education or work-related reasons

Risk Markers in ACHD

High Risk
- Pulmonary hypertension
- Cyanotic CHD
- Severe systemic ventricular dysfunction
- Severe obstructive valvular disease or severe conduit obstruction
- Fontan circulation

Moderate Risk
- Prosthetic valve or conduit
- Systemic-to-pulmonary shunt
- Moderate systemic ventricular dysfunction
- Moderate obstructive valvular disease or moderate conduit obstruction
**Tetralogy of Fallot (ToF)**

- Nonoperative survival
  - 30% at 10 years
  - less than 3% at 40 years
- Complete correction - survival 85% at 36 years.
- Adult survival: (Pink Tet's)
  - Mild Pulm flow limitations
  - aorto-pulmonary collaterals

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**Tetralogy of Fallot (ToF)**

- Pulmonary regurgitation - the main pathology after repair in childhood
- Subsequent worsening RV function – a major determinant for postop morbidity/mortality
- Subsequent reoperations for
  - Pulm Valve replacement (surgical)
  - Transcatheter PV replacement (MELODY valve)

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**Tetralogy of Fallot (ToF)**

- Adult survival: (Pink Tet's)
  - Mild Pulm flow limitations
  - aorto-pulmonary collaterals
Branch PAs

Cardiac MRI

Melody Valve
63 yrs old male repair of Coarctation of Aorta

- Absent to diminished femoral pulses
- brachial-femoral delay
- upper body hypertension
- discrepant BP’s in upper and lower limbs

40 yr old patient with Bicuspid Aortic valve for AVR. Patient has a child who has a Fontan (tattoo) for Hypoplastic Left Heart Syndrome.
Who is managing the store?

- Many times difficult to identify an adult cardiologist who understands the anatomy and physiology.
- Several pediatric cardiologists follow these patients as adults.
- Patients that are older than the physician.
- Does not meet the need.

Stakeholders

- Ongoing care
- Day to day living
- Residual defects
- Arrhythmias
- Pregnancy
- Next generation

Uninterrupted health care: patient centered, age and developmentally appropriate, flexible, and comprehensive.

- Age-appropriate education about medical conditions to promote skills in communication, decision making, self-care, and self-advocacy.
- Greater personal and medical independence and a greater sense of control over health, healthcare decisions, and psychosocial environment.
- Optimal quality of life (QOL), life expectancy, and future productivity of young patients.
The American Academy of Pediatrics states, “The goal of transition in health care for young adults with special health care needs 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.

45% of them lack access to a physician who is familiar with their condition(s).
30% of all young adults 18 to 24 years of age lack a payment source for their health care.

Evaluated by a pediatric cardiologist
Valve is functioning well
Patient gets discharged to adult care
The patient is now 25 years old
Pregnancy
Casually mentions that she has a BAV—can the baby be checked in-utero?

16 year old with BAV
Meanwhile---

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Patient had no knowledge

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Risk to the mother's life
The patient was never educated

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Education

Help a Middle Schooler Succeed

Age of transition

Age of transition
Expectation

- What can the child do?
- Defining an occupation by middle school
- Parents and school to develop their strengths
- Behavior and cognitive challenges

Long term: prognosis

QOL

- Finding a life partner: “baggage”
- Dependence of family unit
- Health insurance
Transposition of great arteries

TGA: arterial switch

TGA: atrial switch

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Health Maintenance Needs

- Usually Handled by Primary Care Provider
  - Vaccinations
  - Cholesterol screening
  - Hypertension screening
  - Cancer screening
  - Assessment of tobacco, alcohol, drug use
  - Nutritional counseling
  - Contraception, sexuality issues
  - Exercise prescription

Education: highest level possible

- Develop a structured educational/teaching plan based on the adolescent's individual academic abilities, educational level, and developmental maturity (Class I; Level of Evidence C).
- Begin counseling early in adolescence to identify interests, as well as the need for additional educational or vocational training and guidance (Class I; Level of Evidence C). Utilize exercise testing as appropriate to assist in determining physical abilities (Class I; Level of Evidence C).
- View every patient as employable, and tailor career/employment counseling to the adolescent's physical capacity, as well as mental and psychological disposition (Class I; Level of Evidence C).

Medical Insurance

- 1. Counseling about health insurance issues before patients with CHD leave their parents' policy or lose their eligibility for children's services, including information regarding the relationship between education/vocational choices and access to insurance benefits, may be beneficial to facilitate continued insurance (Class IIb; Level of Evidence C).
- 2. Discussion of the relationship between education/vocational choices and access to insurance benefits should occur early and guide educational planning (Class I; Level of Evidence C).
1 Comprehensive care that is coordinated and managed through a medical home (usually a primary care provider but in some instances a tertiary care center or a subspecialty practice).
2. Access to healthcare financing.
3. Education of adult providers in managing chronic conditions previously limited to the pediatric population.
4. Ongoing, coordinated communication between patients, families, and pediatric and adult healthcare providers to facilitate transition and transfer.

Disease severity and Stratification of care

- **Group 1 CHD - Simple Heart Disease**
  - community center

- **Group 2 CHD - Moderate Severity**
  - regional congenital heart centers

- **Group 3 CHD - Great Complexity**
  - adult congenital heart disease centers

ACC Task Force 1: 32nd Bethesda Conference, 2001

"And no one shall work for money, and no one shall work for fame, But each for the joy of the working, and each his separate star, Shall draw the Thing as he sees It, for the God of Things as They are!"
- Rudyard Kipling
References:


For Adolescents

1. The timing of transition should be guided by emotional, maturity and developmental level (as opposed to chronological age) for transition planning (Class I; Level of Evidence C).

2. The adolescent should be engaged in transition planning (Class I; Level of Evidence C).

3. The adolescent should be asked about their understanding of their disease in relation to their current health status, restrictions on activities, and future goals (Class I; Level of Evidence C).

4. The adolescent should be encouraged to share concerns about QOL issues (physical restrictions, school, peers, social relationships) (Class I; Level of Evidence C).

5. The adolescent's fears and concerns should be acknowledged in an empathetic, nonjudgmental manner (Class I; Level of Evidence C).

6. The pediatric cardiology provider should initiate and work together with the adolescent on a transition plan using a transition resource binder and/or health "passport" (Class I; Level of Evidence C).
Adolescent

7. Providers should begin to direct health discussions more toward the adolescent than the parent (Class I; Level of Evidence C).

8. QOL issues should be discussed privately with the adolescent (Class I; Level of Evidence C).

9. Be flexible (Class I; Level of Evidence C)

Parents

For Parents

1. The pediatric cardiology provider should initiate discussions on transition planning and partner with parents in the process (Class I; Level of Evidence C).

2. The pediatric cardiology provider should solicit information about parental perceptions of their child’s QOL (Class I; Level of Evidence C).

3. The pediatric cardiology provider should encourage discussion of parental understanding of their child’s disease and concerns in relation to future goals (illness management, education)

4. The pediatric cardiology provider should be nonjudgmental and empathetic when acknowledging fears (Class I; Level of Evidence C).
Recommendations

1. The patient’s “medical home” should reside with the primary care provider, who should provide family and patient-centered care (Class I; Level of Evidence C). The primary care physician should maintain a confidential central record that contains all pertinent information about the patient (Class I; Level of Evidence C).

2. The pediatric cardiologist should prepare a written adolescent transition plan that includes a cardiac destination (Class I; Level of Evidence C).

3. The ACHD expert should prepare and maintain an up-to-date comprehensive cardiac record and transmit that record to the primary care provider (Class I; Level of Evidence C).

Intervention

1. Follow-up studies, both noninvasive and invasive, should be performed by providers with expertise in performance and interpretation of the data specific to CHD (Class I; Level of Evidence C).

2. Regular (at least annual) follow-up is required to ensure appropriate hemodynamics are maintained and to prevent secondary complications (Class I; Level of Evidence C). This will reinforce to the patient that regular follow-up is essential indefinitely.

3. Nonsurgical and surgical interventions must be tailored to the individual patient and should be performed in centers with specific expertise (Class I; Level of Evidence C).

Electrophysiology

- EP and arrhythmias
- Pacemakers and devices