Adult Congenital Heart Disease: What the Primary Care Provider Needs to Know

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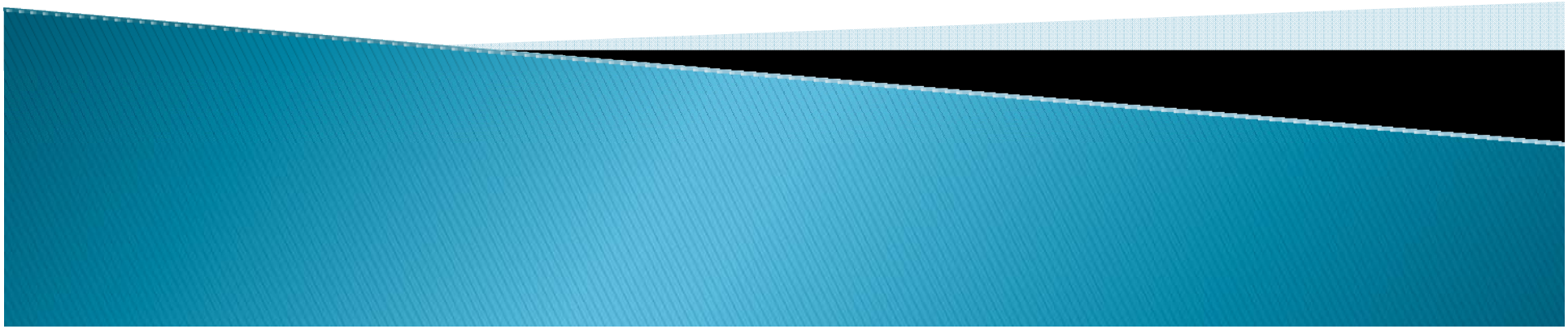
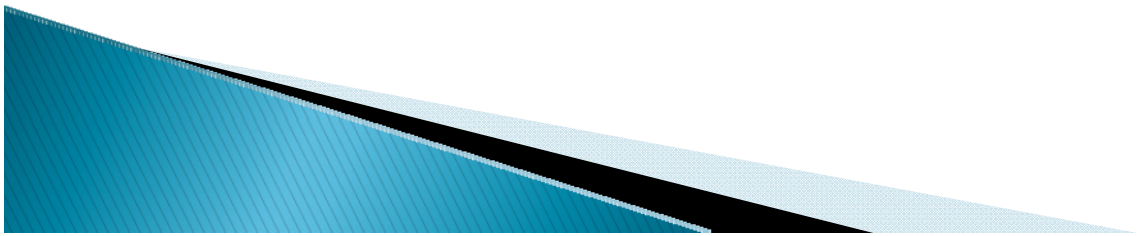


Adult Congenital Heart Disease: Background

- Advances in cardiac surgery have created a new population of adult patients with repaired congenital heart disease.
- Since the advent of neonatal repair of complex lesions in the 1970's, 85% of these patients have survived to adulthood.
  - Current survival rates more like 95%
  - 50 years ago, only 25% of these infants survived beyond the first year of life
- In 2000, 800,000 adults with CHD were alive in US
  - Now, approaching 1 million
- By 2020, 1/150 adults will have some form of CHD



Care of the ACHD Patient: Defining the Problem

- Patients and parents (and physicians) may have a mistaken perception that surgery was curative.
  - Rarely the case
    - Except perhaps in ligated PDA or early repair of isolated ASD or VSD.
  - All other congenital heart surgery should be considered palliative.
  - Late complications are the rule.
  - Lifetime follow-up therefore mandatory.
- Adult cardiology system lacks infrastructure provided by comprehensive pediatric cardiology centers
  - Diagnostic, interventional and surgical expertise as well as APRN's and social workers.
- Transition to adult system occurs when patients (late teens, early twenties) are most likely to ignore their health history and their parent's input.
  - Patients routinely "lost to follow-up" during this period.

Recommendations for Delivery of Care and Ensuring Access (From ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease)

Class I

1. The focus of current healthcare access goals for ACHD patients should include the following:

a. Strengthening organization of and access to transition clinics for adolescents and young adults with CHD, including funding of allied healthcare providers.

b. Organization of outreach and education programs for patients, their families, and caregivers to recapture patients leaving pediatric supervisory care or who are lost to follow-up.

c. Enhanced education of adult cardiovascular specialists and pediatric cardiologists in the pathophysiology and management of ACHD patients.

d. A liaison with regulatory agencies at the local, regional, state, and federal levels to create programs commensurate with the needs of this large cardiovascular population.

2. Health care for ACHD patients should be coordinated by regional ACHD centers of excellence that would serve as a resource for the surrounding medical community, affected individuals, and their families.

3. ACHD patients should carry a complete medical "passport" that outlines specifics of their past and current medical history, as well as contact information for immediate access to data and counsel from local and regional centers of excellence.

ACC/AHA Recommendations of Access to and Delivery of Care (continued)

4. Care of some ACHD patients is complicated by additional special needs, including but not restricted to intellectual incapacities or psychosocial limitations, and designated healthcare guardians should be included in all medical decision making.

5. Every ACHD patient should have a primary care physician. To ensure and improve communication, current clinical records should be on file with the primary care physician and local cardiovascular specialist, as well as at a regional ACHD center; patients should also have copies of relevant records.

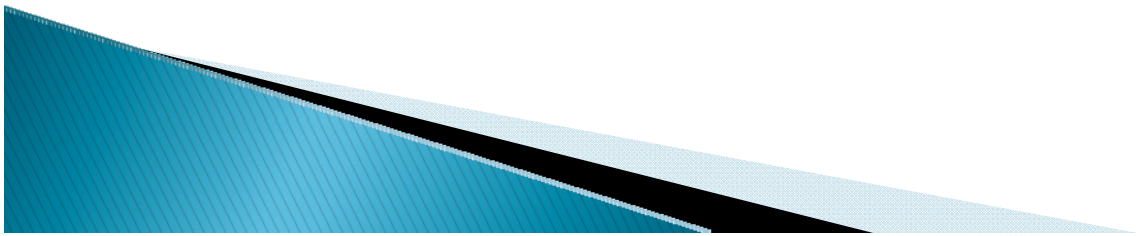
6. Every cardiovascular family caregiver should have a referral relationship with a regional ACHD center so that all patients have geographically accessible care.

Care of the ACHD Patient

- RULE #1:
  - Know the Diagnosis!
  - “I had a hole in my heart” is not a diagnosis.
  - Obtain the primary data from the pediatric cardiologist or the hospital that cared for the child.
  - The surgeon’s original operative note is the gold standard and attempts should always be made to obtain it.
  - If you don’t want to or can’t do this, make sure the cardiologist caring for the patient does.
  - Make sure the patient knows his/her diagnosis. Have him/her carry a “medical passport.”

ACHD Patients At Their Most Vulnerable

- Pregnancy and contraception
- Noncardiac surgery or medical procedures
- Endocarditis prophylaxis



ACHD: Examples of (Not So) Complete Repair

- Tetralogy of Fallot
  - Characterized by VSD and sub-pulmonic stenosis.
  - Repair involves relief of PS but often deforms valve and patient left with significant Pulmonary Valve Insufficiency.
  - PI is hard to hear on exam and see on echo.
  - Long term, this causes RV failure and premature death
  - Many patients require Pulmonary Valve Replacement from age 25-40 years.
- Aortic Coarctation
  - Even with prompt recognition and repair in infancy, there are significant sequelae which limit long term survival.
  - Recoarctation is common.
  - Diffuse aortopathy present with proclivity to aneursym.
  - Concomitant bicuspid aortic valve in 50%.
  - 10% circle of Willis aneurysm.
  - Premature and signficant HTN common and related to poorly defined arteriopathy.

Types of Adult Congenital Heart Disease of Great Complexity*

1. Conduits, valved or nonvalved
2. Cyanotic congenital heart (all forms)
3. Double-outlet ventricle
4. Eisenmenger syndrome
5. Fontan procedure
6. Mitral atresia
7. Single ventricle (also called double inlet or outlet, common, or primitive)
8. Pulmonary atresia (all forms)
9. Pulmonary vascular obstructive disease
10. Transposition of the great arteries
11. Tricuspid atresia
12. Truncus arteriosus/hemitruncus
13. Other abnormalities of atrioventricular or ventriculoarterial connection not included above (ie, crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

*These patients should be seen regularly at adult congenital heart disease centers.

Modified from Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. J Am Coll Cardiol. 2001;37:1170 -5.3

Diagnoses in Adult Patients With Congenital Heart Disease of Moderate Complexity*

1. Aorto-left ventricular fistulas
2. Anomalous pulmonary venous drainage, partial or total
3. Atrioventricular septal defects (partial or complete)
4. Coarctation of the aorta
5. Ebstein's anomaly
6. Infundibular right ventricular outflow obstruction of significance
7. Ostium primum atrial septal defect
8. Patent ductus arteriosus (not closed)
9. Pulmonary valve regurgitation (moderate to severe)
10. Pulmonary valve stenosis (moderate to severe)
11. Sinus of Valsalva fistula/aneurysm
12. Sinus venosus atrial septal defect
13. Subvalvular AS or SupraAS (except HOCM)
14. Tetralogy of Fallot
15. Ventricular septal defect with:
    - Absent valve or valves
    - Aortic regurgitation
    - Coarctation of the aorta
    - Mitral disease
    - Right ventricular outflow tract obstruction
    - Straddling tricuspid/mitral valve
    - Subaortic stenosis

These patients should be seen periodically at regional adult congenital heart disease centers. Modified from Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. J Am Coll Cardiol. 2001;37:1170 –5.3 AS indicates aortic stenosis; HOCM, hypertrophic

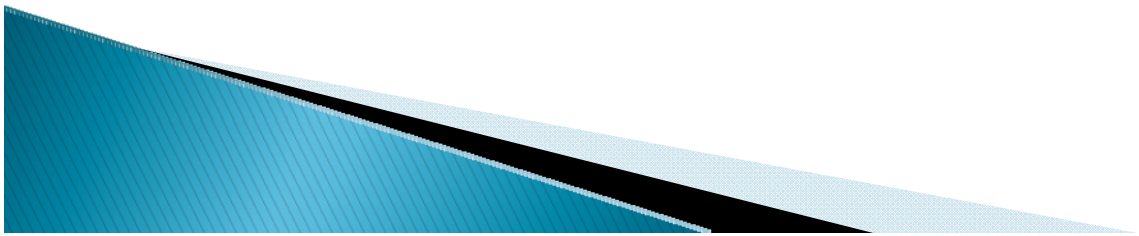
Diagnoses in Adult Patients With Simple Congenital Heart Disease*

1. Native disease
   a. Isolated congenital aortic valve disease
   b. Isolated congenital mitral valve disease (eg, except parachute valve, cleft leaflet)
   c. Small atrial septal defect
   d. Isolated small ventricular septal defect (no associated lesions)
   e. Mild pulmonary stenosis
   f. Small patent ductus arteriosus

2. Repaired conditions
   a. Previously ligated or occluded ductus arteriosus
   b. Repaired secundum or sinus venosus atrial septal defect without residua
   c. Repaired ventricular septal defect without residua

*These patients can usually be cared for in the general medical community.

Modified from Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. J Am Coll Cardiol. 2001;37:1170 –5.3



ACHD: Conclusions

- ACHD patients are a “new population.”
- Their numbers are increasing rapidly.
- Most surgery is palliative, not curative.
- Late complications are very common.
- Follow-up in specialized centers is important for most patients.

- As a primary care provider, key issues are to:
  - Make sure you know the diagnosis.
  - Identify appropriate local and regional specialized caregivers who can follow the patient long term.
  - Recognize times of increased vulnerability (pregnancy, noncardiac surgery) and advocate forcefully for necessary care.