Congenital Heart Disease: Not Just a Pediatric Problem

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- I have no financial disclosures

Case Report #1

- 19 year old male born prematurely (31 weeks) and tetralogy of Fallot
  - Prolonged NICU stay (9 months)
    - Multiple abdominal operation for NEC
    - RVOT patch augmentation to increase pulmonary blood flow
  - Repair of tetralogy of Fallot at 2 years of age with pulmonary homograft
  - Homograft replacement at 6 years of age
  - Implantation of Melody valve in pulmonary position 6 months previously

Case Report #1

- Presented to Sky Ridge with flu-like symptoms for one week
  - Fever to 102º, chills, myalgias, headache, dyspnea
  - Seen by PCP 4 days earlier, no specific therapy
  - Testicular tenderness
  - Treated with Ceftriaxone
- Transferred to RMHC, admitted for viral syndrome and R/O orchitis (2/23/2016)
  - CT angiogram – no pulmonary embolus
  - Echocardiogram – vegetation on Melody valve, RV dilated with elevated pressure, 60 mmHg gradient across pulmonary valve
  - Continued on Ceftriaxone for presumed endocarditis
  - Multiple blood cultures obtained (no growth in all cultures)
  - Acute kidney injury (creatinine 2.9)

Case Report #1

- Cardiac arrest 5 days after admission (2/28/2016)
  - Severe low cardiac output after 30 minutes of CPR
  - Placed on veno-arterial ECMO via femoral vessels
- Surgical procedures
  - Replacement of RV-PA conduit, tricuspid valve replacement (3/3/2016)
    - 34 hour operation
    - 17 units PRBCs, 22 liters Cell Saver
  - Delayed sternal closure (3/7/2016)
  - ECMO decannulation, repair of femoral vessels (3/9/2016)
  - Drainage of pericardial effusion (3/13/2016)
- Hospital course
  - Extubated 4/4/2016
  - Transferred to rehab 4/21/2016
  - Generalized seizure 5/6/2016
  - Discharged 5/12/2016

Scope of the Problem

- Incidence of congenital heart disease
  - 7-8 out of 1000 newborns (perhaps as high as 10 out of 1000 births)
  - Increased in subsequent pregnancies
  - Increased for children of adults with CHD
- Approximately 85% of children with repaired CHD will survive into adulthood (about 8500 per year)
- Estimated 800,000 to 1,000,000 adults with CHD currently in United States

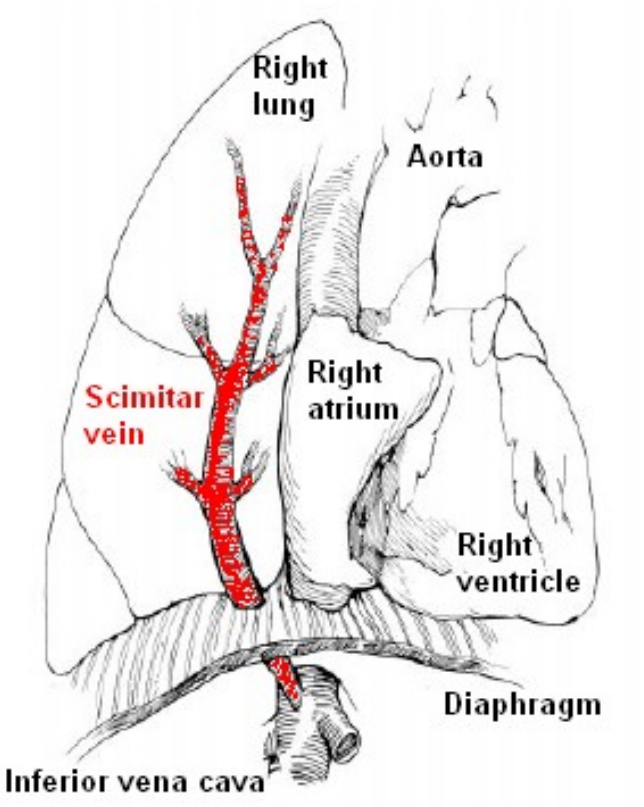
Scope of the Problem

- True incidence may be underestimated
  - At least 10% of cases of CHD will not be diagnosed until adulthood
  - Number of immigrants with unrepaired CHD is increasing
- It is estimated that, within the next decade, 1 out of 150 young adults in the United States will have some form of CHD.

Case Report #2

- 61 year old woman evaluated for abdominal pain caused by diverticulitis
- Referred to adult cardiologist because of an abnormal chest x-ray
- Past medical history
  - Shortness of breath for past 10 years, diagnosed with asthma
  - No complications with seven pregnancies
- Cardiac evaluation (adult cardiologist)
  - Echocardiogram suggestive of partial anomalous pulmonary venous drainage of right lung to inferior vena cava
  - Confirmed with CT angiography
  - Right heart cardiac catheterization: mild pulmonary artery hypertension and Q_p/Q_s=1.5

Case Report #2



Case Report #2

- Referred to adult cardiac surgeon, who thought repair should be best accomplished by congenital cardiac surgeon
- Evaluated by pediatric cardiologist and cardiac catheterization repeated
  - Moderate pulmonary artery hypertension (60/20, mean 39)
  - Elevated PVR (5.8 Wood units, decreasing to 3.0 Wood units with 100% oxygen)
  - Q_p/Q_s=1.8. increasing to 2.4 with 100% oxygen
- Referred for surgical repair, which she underwent without significant complication

Lessons from Case Report #2

- Congenital heart defect unrecognized for 61 years (17 years in United States)
- Incorrect diagnosis of asthma in ~50 year old adult
- Lack of referral to pediatric cardiologist led to
  - Two cardiac catheterizations
  - Unnecessary CT angiogram
  - Initial referral to adult cardiac surgeon who is unfamiliar with anatomy and repair

Guidelines for Managing ACHD Patients

- *ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease*

  JACC 52:e143-e263, 2008

- Task force articles from 32nd Bethesda Conference on “Care of the Adult With Congenital Heart Disease”

  JACC 37:1170-1198, 2001

- *Collaborative Care for Adults with Congenital Heart Disease*

  Circulation105:2318-2323, 2002

Role of Primary Care Provider

- Virtually every PCP will be involved in the care of adults with CHD
- Majority of medical care for adults with CHD will be provided by practitioners who do not have specific training in CHD
- Most medical issues will be managed as if the patient does not have CHD
- Collaborative care with cardiologists and surgeons familiar with CHD is beneficial for most of these patients

Role of Primary Care Provider

- Maintain a low threshold for consulting CHD specialists in patient care
- Maintaining a healthy lifestyle is important for adults with CHD
  - Avoidance of smoking, excessive alcohol, recreational drug use
  - Weight control
  - Exercise
  - Oral hygiene/avoidance of endocarditis

Adults with Simple CHD

- Native disease (unrepaired)
  - Isolated congenital aortic valve disease
  - Isolated congenital mitral valve disease (excluding parachute and cleft mitral valves)
  - Isolated patent foramen ovale or atrial septal defect
  - Isolated small ventricular septal defect
  - Mild pulmonary valve stenosis
- Repaired conditions
  - Ligated or occluded patent ductus arteriosus
  - Repaired atrial septal defect without residua
  - Repaired ventricular septal defect without residua

Adults with Moderate CHD

- Anomalous pulmonary venous drainage, partial or total
- Atrioventricular canal defects, partial or complete
- Coarctation of the aorta
- Ebstein's anomaly
- Right ventricular outflow tract obstruction
- Patent ductus arteriosus (not closed)
- Pulmonary valve stenosis, moderate or severe
- Subvalvar or supravalvar aortic stenosis
- Tetralogy of Fallot
- Ventricular septal defect associated with other lesions

Adults with Severe CHD

- Conduits, valved or nonvalved
- Cyanotic congenital heart disease (all forms)
- Double outlet right ventricle
- Eisenmenger syndrome
- Pulmonary atresia (all forms)
- Pulmonary vascular obstructive disease
- Single ventricle lesions (Fontan procedure)
  - Tricuspid valve atresia
  - Mitral valve atresia
- Truncus arteriosus

Cardiac Concerns for Adults with CHD

- Residual or recurrent hemodynamic problems
  - Valve dysfunction or failure
    - Native pulmonary valve
    - Native aortic valve (including bicuspid aortic valve)
    - Tricuspid valve insufficiency
    - Mitral valve stenosis or insufficiency
    - Prosthetic valve failure
  - Conduit failure
  - Ventricular dysfunction or failure
  - Coarctation of aorta
  - Pulmonary artery stenosis

Cardiac Concerns for Adults with CHD

- Arrhythmias
  - Atrial and ventricular arrhythmias may contribute to morbidity and mortality
  - Possible causes
    - Cardiac incisions, suture lines or patches
    - Injury to or near conduction system
    - Chamber dilation
    - Myocardial injury and fibrosis
    - Anatomic substrate (e.g. *ℓ*-transposition of great arteries)
    - Re-entrant pathways
  - Treatment options
    - Medical therapy
    - Electrophysiologic cardiac catheterization
    - Surgical

Exercise in Adults with CHD

- Exercise tolerance decreased in adults with CHD, repaired or unrepaired
  - Some patients may not perceive decreased exercise tolerance
  - Similar to patients with congestive heart failure
  - May be compounded by
    - Poor chronotropic response to exercise
    - Pulmonary artery hypertension
    - Impaired pulmonary function

Exercise in Adults with CHD

- Few studies have been done to assess exercise tolerance in adults with CHD, so data are limited
  - Decreased exercise tolerance in patients with repaired ASD
  - Not know if exercise and conditioning will reduce symptoms, improve exercise tolerance or improve length/quality of life
- General recommendation is to be as active as possible

Pregnancy in Adult Women with CHD

- Parents should be aware of risk of CHD in offspring
  - 0.8% for general population
  - 3-5% for parents with CHD (mother > father)
- Genetic counseling should be provided
- Risk of pregnancy is generally low in women with good functional class and ventricular function
- High risk in patients with single ventricle (Fontan procedure), *d*-transposition (Senning or Mustard procedure) and *l*-transpisition (systemic RV)

Pregnancy in Adult Women with CHD

- High risk in patients with pulmonary artery hypertension and/or Eisenmenger's complex
- Anticoagulation
  - Conversion from warfarin therapy to heparin
- Fetal echocardiography should be performed to assess cardiac anatomy of fetus
- Careful collaboration among PCP, obstetrician, MFM specialist, cardiologist familiar with CHD, anesthesiologist and neonatologist

Contraception in Women with CHD

- Estrogen-containing oral contraceptive generally not recommended, especially in cyanotic patients or single ventricle (Fontan) patients
- Other oral contraceptives may cause fluid retention and exacerbate CHF
- Barrier methods are preferred, but imperfect
- Tubal ligation may be considered with some women

Cyanotic Adults with CHD

- Should be seen regularly by CHD specialist
- Erythrocytosis
  - Hematocrit may exceed 60%
  - Hyperviscocity of blood
  - Thrombotic complications (stroke)
- Hyperuricemia
  - Arthralgia
  - Gouty arthritis
  - Tophaceous urate deposits

Cyanotic Adults with CHD

- Phlebotomy
  - Rarely used to reduce erythrocytosis
  - Should be performed in center which treats CHD patients
  - Volume replacement with normal saline is recommended
  - Iron deficiency anemia can result with repeated treatment

Cyanotic Adults with CHD

- Surgical considerations
  - Pulmonary artery hypertension/Eisenmenger syndrome significantly increases surgical risk
  - Avoidance of hypovolemia (including preoperative NPO status) is important
  - Meticulous attention to intravenous air is essential
  - Use of invasive vascular catheters should be carefully considered
  - Anticoagulation
    - Preoperative anticoagulation with warfarin
    - Perioperative heparin administration
    - Restarting postoperative warfarin
  - Alternative anesthetic techniques (e.g. epidural)

Psychosocial Issues in Adults with CHD

- Physical appearance
  - Scars, smaller body size, cyanosis, clubbing
- Limitations in achieving educational and vocational goals
  - Learning disabilities
  - Physical limitations
- Achieving independence
  - Transition from adolescence to adulthood
  - Assuming primary role in medical care and decision-making
  - Transition from pediatric care to adult care

Psychosocial Issues in Adults with CHD

- Decreased insurability
  - Health insurance
  - Life insurance
- Avoidance of risky health behaviors
- Family planning
  - Risks of pregnancy and contraception
  - Risk of CHD in offspring
- Coping with chronic illness
- Facing possible early death (patient or spouse)

Summary

- Primary care practitioners will provide the majority of care to adults with CHD
- These patients will have some specific medical needs
  - Surgery (cardiac or non-cardiac)
  - Pregnancy and contraception
- Psychosocial issues may be more prominent in this patient population

Summary

- The primary care practitioner should consult specialists in congenital heart disease (pediatric cardiologist and congenital heart surgeon) in adults who are newly diagnosed
- Patients with moderate or severe congenital heart disease (and some with mild congenital heart disease) should be routinely followed for life by pediatric cardiologist
- Surgical (cardiac or non-cardiac) and obstetric care should be provided in an institution with appropriate facilities, personnel and experience

Dear Dr. Leonard,

Thank you for taking such great care of me and my family while I was in the hospital. Also, I really appreciate you not giving up on me during my surgery. Thanks for doing everything necessary in order to keep me alive. I still find it hard to believe that you stood over me for over 30 hours. I especially appreciate your genuine concern for my twin brother, Zach. Thanks to you, he was able to cope with my ordeal in the hospital. There really aren't words to adequately thank you for making sure that my heart was repaired – and to put me back onto my path to recovery. Finally, thanks so much for making sure my family was safe and helping them stay positive. All in all, thank you for everything you've done and especially for not giving up on me even when it got hard. I will always remember and appreciate you.

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