Introduction to Adult Congenital Heart Disease

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Objectives

• Understand the anatomy and physiology of common congenital heart lesions
• Understand which lesions should be referred to dedicated adult congenital heart programs
• Understand key features in the follow-up of ACHD, as well as resources available to patients and primary care physicians
Scope

- In 2000, there were estimates to be 800,000 adults with congenital heart disease in the United States.
- There are now more adults with congenital heart disease than children.
- 50% of adults with congenital heart disease would have died without intervention.

Historical Repairs

- 1\textsuperscript{st} Coarct Repair
- 1\textsuperscript{st} BT Shunt
- 1\textsuperscript{st} Tetralogy Repair
- 1\textsuperscript{st} Senning/Mustard
- 1\textsuperscript{st} Fontan
- 1\textsuperscript{st} Arterial Switch Procedure
- 1\textsuperscript{st} Norwood Procedure

- 1944
- 1944
- 1954
- 1957 (1960’s), 1963
- 1971
- 1982
- 1979
Implications

- Coarcts – 60’s- 70’s (although many die early)
- Sennings/Mustards -40’s-50’s
- Arterial Switch – Early 20’s
- Norwoods – Late Teens to early 20’s
- Oldest TOF Patients – 70’s
Tetralogy of Fallot

- Over-riding aorta
- Anteriorly deviated septum with narrowing of RVOT
- Large VSD
- Hypertrophied RV
Tetralogy of Fallot

1. Pulmonary stenosis (thickened, narrow pulmonary outflow tract)
2. Thickened right ventricle wall
3. Ventricular septal defect
4. Aorta overrides septal defect

Photo Courtesy of the University of Michigan Congenital Heart Center
Life Expectancy in Unrepaired TOF

66% live to one year
49% to 3 years
24% to 10 years

Surgical Intervention for TOF

- Most Common Cyanotic Congenital Heart Lesion
- First Blalock Taussig Shunt – 1944, Hopkins
- First Complete Repairs(using cross-circulation – 1954 Lillehei, U of MN
Long-term survival in TOF

Issues to Follow in Adult TOF-Sudden Death

• History of Syncope Very Important
• Correlate with QRS duration on resting ECG
Plot of QRS duration in 182 patients

QRS Duration

P<0.0001

QRS of 180ms

Issues to Follow in TOF – Pulmonary Insufficiency

• As a rule, those who underwent an extensive transannular patch will have severe pulmonary insufficiency
• Controversy as to when to replace the pulmonary valve
• Ask about symptoms
• Ask about syncope/arrhythmias
The Systemic Right Ventricle

• Congenital Heart Disease with Systemic Right Ventricle:
  – Congenital Corrected Transposition (L-TGA)
  – D-Transposition of the great arteries following atrial switch procedure
    • Senning Procedure
    • Mustard Procedure
Congenitally Corrected Transposition
D-Transposition of the Great Arteries

PA
Aorta
LV
RV
Atrial Switch Procedures

Intra-atrial baffle (mustard or Senning procedure)

Left Ventricle pumps to Lungs
Right Ventricle pumps to Systemic Circulation
Complications following atrial switch

- Rhythm issues
- Right Ventricular Dysfunction
- Tricuspid insufficiency
- Baffle Obstruction
- Pulmonary Hypertension
  - More common after late repair and with VSD
Survival s/p Atrial Switch

Rhythm problems s/p Atrial Switch

The Fontan Procedure

- Used if only one ventricle or AV valve are present
- Makes use of passive flow from systemic venous through pulmonary bed to pulmonary veins
- First indication: Tricuspid Atresia, now HLHS and other complex single ventricles
- Is it better to have an LV than an RV
The Fontan Procedure

Image Courtesy Dr. Dennis Crowley and Donna Wilkin

Image Courtesy Dr. Adam Dorfman
Complications after Fontan

- Right atrial enlargement, hepatic dysfunction
- Systemic venous collateralization
- Atrial Arrhythmias
- Protein losing enteropathy
- Plastic Bronchitis
- Pulmonary AVM’s
SBE Prophylaxis

• Variable Compliance with New Guidelines
  – Previous Endocarditis
  – Unrepaired complex congenital heart disease
  – Prosthetic or Bioprosthetic Valves
  – Transplant Patients with Valve Disease
  – Residual Lesion which prevents endothelialization (ie VSD patch leak).

Eisenmenger Syndrome

- Eisenmenger physiology develops by 2 years of age with intraventricular connection, but does not occur until adulthood in patients with atrial level shunt.
- Adults with Eisenmenger’s have a better prognosis than primary pulmonary hypertension. (77% at 3 years as opposed to 35% (pre-Flolan))
  - Average age of death 37 years
  - Actuarial survival: 75% at 30 years, 70% at 40 years, 55% at 50 years


Risk Factors

- Syncope
- NYHA Class III or IV
- Complex Congenital Heart Disease
- Sat < 85%
- RV dysfunction
- RVH on ECG
- Down Syndrome
Treatment

- Minimalist Strategy is generally best
- Avoid Phlebotomy
- Look for and Treat Iron Deficiency Anemia
- Need expert care at the time of elective, non-cardiac procedures
- Anticoagulation is controversial
- Pulmonary HTN agents
Pregnancy and Congenital Heart Disease

• Contraindications (Very high risk)
  – Severe Pulmonary HTN (maternal death rate 50%)
  – Severe obstructive Lesions (maternal death rate 17%)
  – Class III or IV CHF
  – Marfan Syndrome with Aortic Root greater than 40mm

High Risk Pregnancy

- Prosthetic Valves
- Obstructive lesions
- Marfan
- Cyanotic CHD
- Pulmonary HTN
- EF < 40%
- Significant Uncorrected Congenital Heart Disease
Indications for C-section

- Obstetric Indications
- Anticoagulation with coumadin at onset of labor
- Pulmonary HTN
- Unstable aortic lesion with risk of dissection
- Severe obstructive lesions
Common Pitfalls

• Know what their baseline O2 sat is. You don’t necessarily have to go for >92% 
  – If they have clubbing and a Hct of 70, their sat is not normal

• Corollary – Not all congenital heart patients are cyanotic 
  • In fact, most are not
Common Pitfalls

- Beware patients with right to left shunts – use venous filters etc.
- Know that patients with previous surgical shunts may not have palpable pulses or BP’s in their arms. Ask the patient.
- When in doubt, don’t phlebotomize unless you check with somebody who really knows what they’re doing.
Common Pitfalls

• You can not always get to the right ventricle via the systemic veins

• All wide complex tachycardia is not V-tach

• All A-flutter does not present with a heart rate of 150
Common Pitfalls

• One can make it to adulthood with undiagnosed congenital heart disease
• Not all pregnant women with CHD need a C-Section
• If a chronically cyanotic patient has a fever, think:
  • Endocarditis
  • Brain abscess
Over the last 50 years, advances in cardiac surgery have allowed many patients with congenital heart disease to survive to adulthood. Patients with complex cardiac anatomy present a challenge to medical providers, and often require access to highly specialized care. This presentation will provide an overview of congenital heart disease in the adult population, with a focus on indications for referral, and potential pitfalls in the care of this complex patient population.

Objectives:

1) Understand the basic anatomy and pathophysiology of congenital heart disease commonly seen in the adult population, including Tetralogy of Fallot, Transposition of the Great Vessels, and single ventricle anatomy.
2) Understand which defects require follow-up in a specialized adult congenital center
3) Avoid classic pitfalls in the management of adult congenital heart disease patients.

The vast majority of congenital heart disease in adults requires regular follow-up in experienced adult congenital heart disease (ACHD) centers. Highly complex patients include those with conduits, Cyanotic disease, double-outlet ventricle, Eisenmenger syndrome, Fontan procedure, mitral atresia, single Ventricle, pulmonary atresia, transposition of the great arteries (TGA), tricuspid atresia, truncus arteriosus, atroventricular or ventricular-arterial discordance,

Additional, somewhat less complex lesions require periodic follow-up at ACHD centers. These include aorto-left ventricular fistulas, anomalous pulmonary and systemic veins, atroventricular septal defect, coarctation of the aorta, Ebstein’s anomaly of the tricuspid valve, right ventricular outflow tract obstruction, primum ASD, patent ductus arteriosus, moderate to severe pulmonary stenosis or insufficiency, sinus of valsalva fistula, aneurysm, sinus venosus septal defect, Sub- or supra-valvar aortic stenosis, tetralogy of Fallot.

Tetralogy of Fallot is the prototype congenital heart lesion, as it is the most common cyanotic congenital heart lesion, and the first complex cyanotic lesion to have both a palliative surgery (Blalock-Taussig Shunt in 1944) and a complete repair (Mid 1950’s). The result of surgical success is that there are currently tetralogy of Fallot patients in their 50’s, 60’s and 70’s. The surgical approach in the early era emphasized aggressive relief of right ventricular outflow tract obstruction with an extensive right ventriculotomy and patch across the pulmonary valve annulus (transannular patch). Therefore, in adults with tetralogy of Fallot, free pulmonary insufficiency and subsequent right ventricular dilatation and dysfunction are extremely common. Patients often develop atrial and ventricular dysrhythmias, and declining exercise capacity. All patients with tetralogy of Fallot require evaluation at an ACHD for formal evaluation. Cardiac MRI has become
the gold standard for measurement of right ventricular size and function, but should be performed at centers with expertise in congenital MRI.

D-Transposition of the great arteries is the most common cyanotic lesion presenting in the neonatal period. Unoperated patients experience near universal mortality in the first months and year of life. Most patients greater than age 20-25 will have undergone an atrial switch procedure (Senning or Mustard procedures). In these procedures, oxygenated blood returning from the left atrium is baffled across the atria to the tricuspid valve and right ventricle, and the deoxygenated blood returning from the systemic veins is baffled to the mitral valve and left ventricle. The result of this procedure is that the patient is no longer cyanotic, but now has a systemic right ventricle and a left ventricle pumping to the lungs. Common late complications include loss of sinus rhythm, chronotropic incompetence, atrial arrhythmias, progressive right ventricular dysfunction, and tricuspid insufficiency. In the mid-1980’s, the arterial switch procedure became widespread. Common complications in this patient population include right ventricular outflow tract obstruction (from the pulmonary valve to the supravalvar region to the branch pulmonary arteries), proximal coronary stenosis, and aortic root dilatation.

The Fontan procedure has allowed long-term survival of patients with only one ventricle. Most older patients have systemic left ventricles, often with tricuspid atresia, or failure of development of the tricuspid valve. In Fontan patients the single ventricle pumps to the systemic circulation. An anastomosis is performed between the systemic veins and the pulmonary arteries. The result is that there is passive flow from the systemic veins through the lungs to the left atrium. Common complications include atrial arrhythmias, systemic ventricular dysfunction, and thromboembolic disease. Protein-losing enteropathy is a serious complication involving diarrhea, and peripheral edema, likely caused by elevated systemic venous pressures. The 5 year survival after diagnosis of protein losing enteropathy is approximately 50%.

11 Classic Pitfalls (Words to the wise)

1. You cannot always get to the right ventricle from the systemic veins
2. Not all congenital heart patients are cyanotic (most are not)
3. Routine, scheduled phlebotomy is not appropriate
4. Many patients do not have blood pressures or pulses in their upper extremities
5. Not all atrial flutter is at 300 with a ventricular rate of 150
6. Right ventricular hypertension does not equal pulmonary hypertension
7. Invasive procedures should be performed at experienced ACHD centers
8. Most women with ACHD do not automatically require a C-section
9. Consider underlying ACHD when starting oral contraceptives
10. All wide-complex tachycardia is not V-Tach
11. A dilated right ventricle must be explained.
References:


