Adult Congenital Heart Disease for the Internist

Saurabh Rajpal, MBBS, MD
Assistant Professor
Department of Internal Medicine
Division of Cardiovascular Medicine
The Ohio State University Wexner Medical Center

Objectives

• To discuss the increasing prevalence of adult congenital heart disease

• To discuss the common congenital heart disease diagnoses encountered in primary care clinics

• To discuss strategies for lifelong care of adult patients with congenital heart disease
Why Should the Internists Care?

Congenital Heart Disease
The Remarkable Journey From the “Post-Mortem Room” to Adult Clinics
Ali J. Marian

EXPLOSION of a “Life-Long” Disease Burden

CHD prevalence 12 / 1000 children
CHD prevalence 6.07 / 1000 adults

55-140,000 new pts/yr
9% increase/yr

### Estimated Survival - Era Effect

![Graph depicting estimated survival probability by age and era effect](Moons et al. Circulation 2010)

### Trends in ACHD

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Specialized ACHD Centers</td>
<td>68</td>
<td>94</td>
<td>126</td>
</tr>
<tr>
<td>Patient Visits</td>
<td>55777</td>
<td>5568</td>
<td>110740</td>
</tr>
<tr>
<td>Publications</td>
<td>4373</td>
<td>6281</td>
<td>6281</td>
</tr>
</tbody>
</table>

## ACHD Patients - Common Issues

<table>
<thead>
<tr>
<th>Quality of life</th>
<th>Pulmonary Hypertension</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transition</td>
<td>Heart Failure</td>
</tr>
<tr>
<td>Birth Control</td>
<td>Arrhythmias</td>
</tr>
<tr>
<td>Pregnancy and CHD</td>
<td>Neurocognitive issues</td>
</tr>
<tr>
<td>Dental Issues</td>
<td>Advance care planning</td>
</tr>
<tr>
<td>Exercise</td>
<td>and advanced directives</td>
</tr>
<tr>
<td>Hep C</td>
<td></td>
</tr>
</tbody>
</table>

### Atrial Septal Defect

https://www.cdc.gov/ncbddd/heartdefects/AtrialSeptalDefect-graphic2.html
Physical Exam – Atrial Septal Defect

- Hyperdynamic precordium
- Loud P2- Pulm HTN
- Signs of RHF rare
- Widely split and fixed S2

Murmurs in ASD

- Soft SEM- LUSB
- Diastolic rumble over LLSB- increased flow TV
- HSM at LLSB- TR

https://www.cdc.gov/ncbddd/heartdefects/AtrialSeptalDefect-graphic2.html

EKG

Secundum ASD
- Incomplete RBBB
- Right Axis Deviation

Primum ASD
- Incomplete RBBB
- Left Axis deviation
ASD – Paradoxical Embolism

Myocardial Infarction and PE after IVDU

ASD and Pulmonary Hypertension
**Associated Anomalies**

- Anomalous pulmonary veins
- VSD
- Mitral regurgitation with primum ASDs due to cleft valve

### RV Dilation

<table>
<thead>
<tr>
<th>Description</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>RVEDV&lt;sub&gt;i&lt;/sub&gt;</td>
<td>145 ml/m&lt;sup&gt;2&lt;/sup&gt;</td>
</tr>
<tr>
<td>RVEF</td>
<td>61%</td>
</tr>
<tr>
<td>LVEDV&lt;sub&gt;i&lt;/sub&gt;</td>
<td>102 ml/m&lt;sup&gt;2&lt;/sup&gt;</td>
</tr>
<tr>
<td>LVEF</td>
<td>56%</td>
</tr>
<tr>
<td>Qp/Qs</td>
<td>1.1</td>
</tr>
<tr>
<td>TR fraction</td>
<td>49%</td>
</tr>
</tbody>
</table>
### Etiologies of RV Dilation

- Tricuspid regurgitation
- Pulmonary regurgitation
- Pulmonary artery hypertension
- Shunt Lesions
- Myocardial abnormalities
  - Uhl’s anomaly
  - ARVC
  - Ventricular dysfunction

### Inferior Sinus Venosus – ASD
### ASD- Indications for Closure

- Significant left-to-right shunt
  - right ventricular volume overload
  - with or without symptoms
  - without pulmonary hypertension*
- Orthodeoxia-platypnea
- Paradoxical embolism
- At the time of another cardiac surgery

### ASD Closure

- Surgical
- Transcatheter
Other Issues with ASDs

- Periodic follow up
- Arrhythmias (also with repaired)
- Pulmonary hypertension
- Scuba diving
- High altitude exposure

Patent Foramen Ovale

Drawn by: Kjetil Lenes
PFO - To close or not to close..

*Patient Factors*
- Hypercoagulable state
- Atrial Fibrillation
- ASCVD Risk Factors
- Presence of devices in the RV

*PFO factors*
- Shunt size
- Atrial Septal Aneurysm

### Ventricular Septal Defect

1. Conoventricular
2. Membranous
3. Inlet
4. Muscular

Source: Centers for Disease Control and Prevention
### Physical Exam and EKG

- Smaller the VSD, louder the murmur
- Holosystolic plateau-shaped murmur at LLSB
- Majority of patients with isolated ventricular septal defect have normal EKG
- EKG signs of LAE and LVH – maybe

### VSD

![Echocardiogram images](image-url)
Indications for Closure of VSD

- Symptoms of heart failure
- Large LV
- Normal PVR

VSD Closure in Adults

- Intervention is rarely required
- Small VSDs do not create a clinically important shunt
- Adult patients with large VSDs and irreversible pulmonary vascular disease – Need expert assessment
Lifelong Follow Up - VSD

- More often repair of the VSD
  - Endocarditis
  - Aortic insufficiency
  - Pulmonary insufficiency
- Surgical repair remains the gold standard for treatment of VSD

Gerbode Defect
**Patent Ductus Arteriosus**

Significant Left to Right Shunt
- Symptoms
- LA and LV enlargement

**Endarteritis**

Source: Patent Ductus Arteriosus (PDA) - American Heart Association

---

**Coarctation of Aorta**

- Familial risk
- Turner syndrome
- Associated anomalies
  - ASD
  - VSD
  - Bicuspid aortic valve
- Hypertension
- Brachial-femoral delay
- Premature CAD, Stroke
- Intracranial aneurysms
- Surgery, Balloon Angioplasty, Stent

Courtesy: Ohio Fetal Medicine Collaborative
Tetralogy Of Fallot (TOF)

Unrepaired

Repaired

EKG in Repaired TOF
Issues in Adults with Repaired TOF

- Pulmonary Regurgitation
- Atrial Arrhythmias
- Ventricular Arrhythmias
- Sudden Cardiac Death
- Residual VSD
- LV Dysfunction and left heart failure
- Right Heart Failure

Tetralogy Of Fallot
Adults with Repaired TOF

- EP Procedures
- Pulmonary Valve Replacement
  - Transcatheter
  - Surgical
- Lifelong follow up with ACHD

Ebstein Anomaly
**Ebstein Anomaly**

- Tricuspid regurgitation
- Right heart failure
- ASD - O2 desaturation
- Surgery-
  - Tricuspid valve replacement
  - Cone procedure
- Atrial arrhythmias
  - Atrial fibrillation
  - Atrial flutter
  - WPW
  - Sinus node dysfunction
  - Sudden cardiac death

**Transposition of Great Arteries**

[Diagram of Transposition of Great Arteries]

Courtesy: Ohio Fetal Medicine Collaborative
Transposition of Great Arteries

42 year old female with D-TGA s/p atrial switch w/syncope
D-TGA Atrial Switch

Complex Congenital Heart Disease

- Eisenmenger Syndrome
- Unrepaired Cyanotic Congenital Heart Disease
- Fontan and Single Ventricle
- Patients Palliated with Systemic to Pulmonary Artery Shunts
**Single Ventricle Anatomy**

<table>
<thead>
<tr>
<th>HLHS</th>
<th>TA</th>
<th>DORV</th>
<th>DILV</th>
<th>Unbalanced AVC</th>
<th>PA</th>
<th>Ebstein</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Surgical Shunts
- BT
- Central
- Waterston
- Potts

Courtesy: Ohio Fetal Medicine Collaborative

PVR >> SVR
Qp:Qs = 0.9:1
Eisenmenger Syndrome

Courtesy: Curt Daniels, MD
Complex Congenital Heart Disease

- 6 monthly to yearly ACHD follow-up
- Know and check O2 saturation regularly
- Check iron stores periodically
- Monitor renal and liver function
- Most have restrictive or other lung disease
- Dental follow up
- High risk of stroke and brain abscess in cyanotic patients

ACHD Patients-Common Issues

- Quality of life
- Transition
- Birth Control
- Pregnancy and CHD
- Dental Issues
- Exercise
- Hep C

- Pulmonary Hypertension
- Heart Failure
- Arrhythmias
- Neurocognitive issues
- Advance care planning and advanced directives
Quality of life

- Quality of a person’s life is related to how satisfied they are with their life overall
- Functional status has to do with a person’s ability to do normal daily activities and perform their roles in life
- Disability paradox
- Response shift
- Sense of coherence

Pregnancy

- High Risk
  - Aortopathies including Marfan syndrome
  - Severe left sided obstructive lesions
  - Fontan
  - Eisenmenger
  - Pulmonary Hypertension
  - Severe LV Dysfunction
## Birth Control

- Hypercoagulable states
- Low dose progestin pills
- IUDs
- Complex congenital heart disease patients should be evaluated in tertiary centers

## Exercise

![Unidentified members exercising](http://digitalcollections.nypl.org/items/025b8e10-63aa-0132-0f5b-58d385a7bb0)

### SBE Prophylaxis

- Prosthetic heart valves
- Prosthetic material used for cardiac valve repair
- Prior history of IE
- Unrepaired cyanotic congenital heart disease
- Repaired congenital heart disease with residual shunts or valvular regurgitation at the site or adjacent to the site of the prosthetic patch or prosthetic device
- Repaired congenital heart defects with catheter-based intervention involving an occlusion device or stent during the first six months after the procedure
- Valve regurgitation due to a structurally abnormal valve in a transplanted heart

### Advance Care Planning and Advance Directives

The place for these difficult conversations should not be in the Intensive Care Unit (ICU)

- 50% of ACHD patients die in the hospital
- Of these, two-thirds die in the intensive care setting and almost a half were on life support
- Only 10% of patients in ACHD care had an end-of-life discussion

*Tobler et al Tobler Am J Cardiol 2012
Tobler et al Palliative Medicine 2012*
Current Concerns Neurodevelopmental Outcomes

- 60 young adults with arterial switch operation re-evaluated at a mean age of 16.9 ± 1.7 years
- Neurologic impairment in 10%
- Periventricular leukomalacia was detected in >50%; its severity correlated with the grade of neurologic impairment
- Magnetic resonance imaging demonstrated moderate or severe structural brain abnormalities in 32% of the patients


New Guidelines

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines

Developed in Collaboration With the American Association for Thoracic Surgery, 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

JACC 2018
COACH
Columbus Ohio Adult Congenital Heart Program