Pediatric Cardiology and Congenital Heart Surgery

The Basics

Amy Hemingway, RN, MSN, CNS, CPNP
Congenital Heart Surgery
Texas Children’s Hospital
Normal Cardiac Anatomy

- Superior Vena Cava
- Aorta
- Pulmonary Artery
- Pulmonary Veins
- Right Atrium
- Pulmonary Valve
- Tricuspid Valve
- Inferior Vena Cava
- Left Atrium
- Aortic Valve
- Left Ventricle
- Mitral Valve
- Right Ventricle
- Descending Aorta
Normal Blood Flow
Normal intracardiac pressures and saturations
Physiology: Things to Remember

Blood will take the path of least resistance
Without flow, distal structures will not develop ("no flow, no grow")
The ventricles are designed very differently
The RV is not designed to be a systemic/high pressure pump
The valve (TV/MV) goes with the ventricle
If not protected, the pulmonary vasculature will start to become obstructive in the first 2 years of life
Bernoulli Equation

\[ 4 \ (V^2) = \text{pressure difference/ gradient} \]

\[ \text{So} \]

If Gradient across AoV is 5m/second

\[ 4 \ (5^2) = 100 \]

\[ \text{So} \]

If SBP is 100 LV pressure is 200!
Single Ventricle Pathway

Blood flows passively to lungs and single pumping chamber pumps to body

Fontan’s 10 commandments

Think of management as a chess match

Hepatic Factor- AVMs form without it

Failing Fontan: plastic bronchitis and PLE
Fontan's 10 Commandments

Age older than 4 yrs
Sinus rhythm
Normal systemic venous return
Normal RA volume
Mean PA pressure <15

PVR < 4 woods units
PA-Ao ratio > 0.75
LVEF > 60%
Competent MV
No PA distortion
Single Ventricle/ Norwood
Bidirectional Glenn SVC to RPA
Fontan
IVC to RPA via GoreTex tube
Classifications of Congenital Heart Disease

Lesions with too much pulmonary blood flow (left to right shunt)
Lesions with not enough pulmonary blood flow (right to left shunt)
Transposition
Obstructive lesions (outflow and inflow)
Regurgitant lesions
Surgical lesions
Too much pulmonary blood flow

- VSD
- PDA
- AP window
- Truncus Arteriosus
- ASD/PFO
- ASD/PAPVR
- CAVC
Too much PBF: Respiratory Symptoms/ FTT

First 3 months, PVR ↓ and SVR ↑

Hypermetabolic state, may need 150kcal/kg/d to gain weight

A rumble on exam means Qp:Qs > 2:1
Treatment

Medical management: diuretics/afterload reduction
device closure
surgical repair vs PA banding
Not enough PBF
Right sided obstructive lesions

Tetralogy of Fallot
Pulmonary Stenosis
Pulmonary Atresia
Treatment for “not enough PBF”

Newborn: if insufficient PBF start PGE to keep PDA open

Establish a reliable form of pulmonary blood flow

Surgery:
- shunt BT or central or Mee
- complete repair

Cardiac cath Lab
- PDA stent
- Balloon valvuloplasty if annulus big enough
Surgery:
BT shunt vs Valvotomy
ToF Repair
Obstructive Lesions-outflow

Left Ventricle
- Aortic stenosis
- Aortic atresia
- Interrupted aortic arch
- Coarctation of the aorta
- Subaortic membrane

Right ventricle
- Pulmonary stenosis
- Pulmonary atresia
- Double chambered RV
Left Sided Obstructive Lesions

Aortic stenosis
Aortic atresia
Interrupted aortic arch
Coarctation of the aorta

LV hypertrophy causes diastolic dysfunction
Coronary perfusion compromised with AS/ AI
Stenosis usually worsens over time
Ross Operation
Move Pulmonary to Aortic position and replace pulmonary with homograft
End to End Coarctation Repair
Aortic Arch Advancement

A

Patent ductus arteriosus

Incision

Phrenic n.

Vagus n.

Descending aorta

B

Phrenic n.

Vagus n.

Patent ductus arteriosus

Descending aorta

C

Descending aorta-to-side aorta anastomosis
Right Sided Obstructive Lesions

May have ductal dependent PBF

Most likely will not worsen over time

Hypoxemia is a result of R→L shunting
Treatment of Outflow Obstructive Lesions

Single ventricle pathway, Balloon dilation or stent implantation

Surgical management: Valvotomy vs valvuloplasty in cath lab
Obstructive AV Valves

Left sided
- pulmonary vein obstruction
- cor triatriatum
- mitral stenosis

Right sided
- tricuspid stenosis
Treatment of Obstructive AV Valves

Balloon dilation (Cardiac Cath lab or operating room if annulus ok)
Surgical relief: valvotomy
Transplantation
Long term medical management
Regurgitant AV valves

**Mitral Regurgitation**
- AV canal defect with ASD & VSD
- Papillary muscle infarct with LCAPA

**Tricuspid Regurgitation**
- Ebstein’s malformation
- Hypoxic
- Big heart
DOL #1

Huge Heart
Desaturated
Ebsteins until proven otherwise
Treatment

Surgical: Starnes, Valvuloplasty, (may or may not work), BT shunt or single ventricle pathway
Valve replacement (Poor options ... lifelong anticoagulation and don't grow)
Long term medical and surgical management
May require transplant vs palliative care
Starnes Operation
Regurgitant lesions - semilunar valves

Right sided

  Pulmonary insufficiency
    Well tolerated (RVEDV 180-200cc/m²)
    Surgical and Catheter based

Left Sided

  Aortic Insufficiency
    Monitor function: EF is deceptive
Pulmonary Insufficiency

(TOF) Absent Pulmonary Valve Syndrome
Post op ToF/ valvotomy
Large PAs compress airways
RV dilation with increased volume load (>180cc/m²)
PAs may need surgical plication
Valve repair vs replace
Aortic Insufficiency

Associated with aortic stenosis
May be caused by prolapse of valve leaflet into a VSD
Will see a fall in diastolic pressure and subsequently decreased coronary perfusion
If isolated, think about Marfan Syndrome
Treat with surgical repair vs replace vs Ross
Transposition of the Great Arteries: TGA

Hypoxic- parallel circuits
Mix via PFO +/- PDA
PGE at birth- DC post
BAS
surgery in first 6 weeks of life if no VSD
Arterial Switch Operation
TAPVR
supracardiac vs infradiaphragmatic

may have early hypoxia-
infradiaphragmatic
Pulmonary edema- ground
glass CXR
They all become
obstructive, eventually
Treatment of TAPVR

Don’t touch the veins!

Pulmonary veins are VERY precarious.

Surgical repair of TAPVR (A surgical emergency if they are obstructed or below the diaphragm. All will become obstructed eventually)

if repeatedly stenotic, stenting may be best option

Heart Lung is only transplant option
The Child With CHD: Things to Remember

Children with heart disease need their immunizations and Flu shots and some need Synagis (transplants do NOT get anything for the 1st year and never get live vaccines).

Digoxin may be given for inotrope.

BNP is an indication of stress/heart failure.

Consider holding diuretics if vomiting or diarrhea occur to avoid dehydration.

The child with a large left to right shunt that doesn’t go into failure is worrisome (PVR)
Everything is done to give them life, so we need to let them live

Thank You

No Borders. No Boundaries
Anomalous Origin of a Coronary

♥ First program of its kind in the world
♥ Intramural vs interarterial
♥ Left more worrisome than right
♥ Unsure if surgery helps
  ♥ Unroofing
  ♥ Translocating
♥ No exercise until cleared 3 months post-op: stress test+ CTA+ Echo
Cardiomyopathy

- Dilated
- LV non-compaction
- Hypertrophic
- Restrictive
# Types of Cardiomyopathy

<table>
<thead>
<tr>
<th>Dilated</th>
<th>LV non-compaction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Idiopathic</td>
<td>Familial</td>
</tr>
<tr>
<td>Familial (~20%)</td>
<td>Idiopathic</td>
</tr>
<tr>
<td>Viral</td>
<td>Hypertrophic</td>
</tr>
<tr>
<td>Metabolic</td>
<td>Familial (~50%)</td>
</tr>
<tr>
<td>Neurologic</td>
<td>Idiopathic</td>
</tr>
<tr>
<td>Autoimmune</td>
<td>Restrictive</td>
</tr>
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<td>Chemotherapy</td>
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Dilated Cardiomyopathy

Most common cardiomyopathy

Myocarditis, idiopathic, CHD

Big dysfunctional ventricle

33% get better

33% need medications

33% need transplant
Hypertrophic Cardiomyopathy

- Thickened left ventricle
- May have obstructed outflow tract
- Hyperdynamic Function
- May have diastolic dysfunction
- Ventricular arrhythmias
- Treat with β-blockers
- Avoid dehydration and tachycardia
- Most common cause of adolescent athlete death
LV Non-Compaction

- Spongy rather than smooth walled
Dilated vs. Hypertrophic
Restrictive Cardiomyopathy

Diastolic dysfunction (poor relaxation)

β-blocker to slow the heart rate and minimize ischemia—usually intravenously as an inpatient

Poor prognosis (<2 years from diagnosis) without transplantation
Ventricular Assist Devices for Dilated and LVNC

Berlin Heart for young children
HeartWare
Cardiac Transplantation

Patients with severely impaired function (systolic or diastolic)

Trade one set of problems for another

Average transplant survives 12 years before needing their second transplant

Transplant coronary artery disease

Post-transplant lymphoproliferative disease (PTLD)
History

Patient History
- Feeding problems
  - Diaphoresis
  - Tachypnea
  - Cyanosis
  - Irritability
- Poor weight gain
- Activity level
- Syncope/Palpitations
- Chest pain

Family History
- Congenital heart disease
- Frequent miscarriages
- Sudden/Early death/ MI under 50 yr. old
- Congenital deafness
- Genetic Syndromes
Who should be referred

Syndrome/ multiple cong. anomalies

(+) FH of CHD, syncope, cardiomyopathy, sudden death

(+ ) ROS: FTT, Cardiorespiratory symptoms

Abnormal vital signs i.e. BP gradient (leg>arm = normal, but arm>leg=refer)

Central cyanosis

Chest wall abnormality i.e. hemihypertrophy

Absent or weak femoral pulses
Who should be referred (cont.)

Active precordium: lifts, taps, thrills, PMI displaced
Abnormal heart sounds i.e.: clicks, fixed split S2, single S2, gallop rhythm
Murmurs that are diastolic or systolic and harsh, long, holosystolic or GIII or greater
Murmurs that persist with postural changes, Valsalva, absence of fever or anemia
The Exam: Things to Remember

Be patient during your exam, take as long as you need to be sure!

Feel 4 extremity pulses and take 4 extremity BPs

Tachypnea in a baby with too much PBF is usually ok as long as they are comfortable

Crackles are one of the very last signs of heart failure in children
Chest Pain
Rarely cardiac etiology: Consider musculo-skeletal, GI or pulmonary causes.

Refer to Cardiology if:

- Family history of MI under 50 years of age
- History of congenital heart disease/arrhythmia
- Occurs with exertion and is associated with dizziness, palpitations, SOB, diaphoresis...is described as tightness NOT sharp & stabbing
- Recent illness with orthopnea and pain when they take a deep breath (consider pericarditis)
- Parent insists
Syncope
Vasodepressor Syncope

Most common
Dizziness, light headedness, LOC
Cough, post-micturition, hair-brushing, standing
Venous pooling in the lower extremities
Decrease in ventricular preload
Compensatory increase in HR and ventricular contractility
Vasovagal reflex: bradycardia, hypertension or both
# Evaluation

## History of event
- Prodrome
- Loss of continence
- Chest pain
- Palpitations
- Vertigo
- Seizure activity
- Loss of consciousness
- Exercise association
- Injuries sustained

## Past Medical History
- CHD
- Syncope
- Seizures
- Medications

## Family History
- Syncope
- Congenital deafness
- Seizures
- Sudden death
Evaluation

Physical Exam
vitals, 4 extremity and orthostatic blood pressures

complete CV exam:
  Precordial activity normal? LV impulse displaced?
  Regular rate? Normal S1, S2? Murmur? Pulses equal? Pink and warm? Cap refill brisk?

neurological exam
Diagnostic Studies

Directed by H & P
CBC, Drug screen, chemistries, b-HCG
EKG to check QTc, ectopic beats, pre-excitation, arrhythmia
Holter monitor or event recorder
Treatment: Vasovagal

Avoidance therapy

Increase fluid intake (\(\downarrow\) Caffeine)

Avoid changing positions quickly

Liberalize salt

Avoid triggers

Thermotabs (450 mg tabs, 2 tabs bid)

Florinef (0.1 mg, 1 tab qd to bid)
Cardiac Syncope - Refer to Cardiology

Occurs while recumbent or during exercise
Little or no prodrome
Prolonged LOC (>5 mins) (+/-) Seizures
Associated with chest pain or palpitations
Family history of sudden death
Injury sustained during fall
Have aortic stenosis, HCM, DCM, RCM
Types of Cardiac Syncope

Primary
- Right or left ventricular outflow tract obstruction
- Coronary artery abnormalities
- Cardiomyopathies

Secondary
- Arrhythmia
Arrhythmias

VT, SVT, junctional tach...
AV block, sinus bradycardia...
Sick sinus syndrome
Long QT syndrome/ Torsade de Pointes
Treatment: Cardiac Causes

- Exercise restriction
- Correct underlying cause
- Medications (β-blocker or ACE inhibitor)
- Radiofrequency ablation
- Pacemaker
- AICD (internal defibrillator)
Kawasaki Disease
Primary Symptoms
Need to have 5 out of 6 unless aneurysms present, then only 4

- Fever >5 days
- Reddening of palms and soles with indurative edema
- Rash-polymorphous exanthema
- Red lips, strawberry tongue
- Bilateral conjunctival congestion
- Cervical lymphadenopathy
Secondary Symptoms

**Cardiac:** new murmur, prolonged PR-QT, abnormal Q wave, low voltage

**GI:** vomiting, diarrhea, abdominal pain, jaundice

**Blood:** Leukocytosis and thrombocytosis

**GU:** Proteinuria

**Resp:** URI symptoms: cough, runny nose

**Muscl-Skeletal:** Joint pain

**CNS:** seizure, facial palsy, paralysis of extremities
Outcome after Kawasaki with aneurysms

May have necrosis of fingers and toes
Aneurysms may resolve
May have rebound stenosis
May be treated with bypass surgery, but rarely an option
May require cardiac transplantation in severe cases
The Pre-participation Sports Physical
Competitive athletes:
High school = 6,000,000
College = 500,000
Professional = 5,000

Sudden death during athletic event is 1:100,000 - 1:300,000 for high school

Death rate is 5 times greater in boys

Death rate is 2 times greater in college vs high school
Sudden Death in Athletes

Italian study (1978-1993) - 200 cases

Cardiac etiology = 163/200 (81.5%)
Cerebral etiology = 15/200 (7.5%)
Unexplained etiology = 12/200 (6%)
Respiratory etiology = 10/200 (5%)

Causes of Sudden Cardiac Death in Athletes
Thank You