Pediatric Cardiology

An introduction to the pediatric patient with heart disease: M-III Lecture
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Introduction
- Pediatric heart disease may be congenital or acquired
- Congenital heart defects occur in about 8 of every 1000 live births
- Successful treatment is available for almost every known defect

Tools of Assessment
- General appearance
- Vital signs (including pulse oximetry)
- Auscultation
- Pulses
- Laboratory data (response to oxygen)

General Appearance
- Alert vs. lethargic
- Thriving vs. wasted
- Pink vs. cyanotic
- Comfortable vs. distressed
- Warm, well-perfused vs. cool, mottled

Vital Signs
- Weight
- Respiratory rate
- Pulse
- Blood pressures
- Systemic saturation (pulse oximetry)

Auscultation
- First heart sound S1
- Second heart sound S2
  - Single, physiologic splitting, fixed split
  - S3, S4, gallop
- Murmurs
- Clicks
- Rubs
Murmurs

- Timing (when in the cardiac cycle)
- Type (ejection vs. regurgitant)
- Location
- Intensity or Grade (how loud is it)
- Quality (blowing, musical, machinery)
- Transmission or Radiation

Pulses

- Pulse rate and regularity
- Right arm, left arm, and lower extremity
- Bounding pulses
- Thready pulses
- Pulsus paradoxus

Basic Categories of Disease

- Dysrhythmias
- Heart Failure
- Structural Abnormalities

Sinus rhythms

- Normal sinus rate for newborn 110-150
- 2yo 85-125; 4yo 75-115; 6yo 60-100
- Transient tachycardia to 180-190 is common, rarely >220 bpm
- Tachycardia often caused by caffeine, sepsis, fever, hypovolemia, anemia

Sinus rhythms (cont.)

- Sinus bradycardia almost never primary heart disease (usually secondary to apnea, CNS injury, or drug exposure)
- Sinus arrhythmia (phasic variation with respiration) is normal in the pediatric population !!!

Premature atrial contractions

- Seen in 35% of term newborns
- Slightly less common in premies
- Can be due to digitalis toxicity
- Will be followed by an incomplete compensatory pause (resets the sinus pacemaker)
- Can be non-conducted
Supraventricular tachycardia
- Heart rate 200-300 with narrow complex (normal QRS)
- WPW responsible for 50% of neonatal cases
- May start and stop abruptly, usually hemodynamically stable
- If possible should obtain 12-lead ECG prior to cardioversion

SVT (continued)
- Convert using IV adenosine 0.1 mg/kg fast IV push (may increase by 0.05 mg/kg increments up to 0.25 mg/kg)
- Treatment of choice in neonates usually digoxin – beware of dosing!
- Treatment in older children usually a beta-blocker (propranolol)
- Parents must know how to count a HR

Premature Ventricular Contractions
- Isolated, unifocal PVCs are normal in the newborn (and common in children)
- Frequent PVCs may be associated with CHD, myocarditis, cardiomyopathy, or caffeine
- Benign PVCs will be suppressed in the setting of sinus tachycardia

Heart Block
- First degree: p wave with every QRS, PR interval long
- Second degree: some but not all p waves followed by QRS (rare in children)

Heart Block (continued)
- Third degree: aka complete heart block
  - P wave completely unrelated to QRS
  - Associated with CHD in 30% of neonatal cases
  - Maternal lupus is by far the most common etiology
  - Pacemakers now small enough for even small neonates

Basic Categories of Disease
- Dysrhythmias
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- Structural Abnormalities
Heart Failure (anatomically normal heart)

- Myocarditis – infection
- Myocardial ischemia – birth asphyxia
- Cardiomyopathy – maternal diabetes or congenital cardiomyopathy
- Dysrhythmias – heart block, SVT
- RV failure due to lung disease
- Other – sepsis, over hydrated, severe anemia

Heart Failure (anatomically abnormal heart)

- At birth: HLHS, severe TR, severe MR
- First week: TGA, premie with large PDA
- 1-4 weeks: critical valve stenosis (AS/PS), coarctation of the aorta
- 4-8 weeks: volume load lesions (large left to right shunt lesions) – so called congestive heart failure

Fetal – Neonatal Circulation

- Basic cardiac structures have formed by 20 days of embryonic life
- Heart is essentially fully developed by 42 days of embryonic life
- Fetal cardiovascular system is dependent on the placenta
- At birth everything must change to adapt from placenta to pulmonary gas exchange

Shunts

- These determine aberrations in BP, SaO2, pulses, etc.
- PDA – patent ductus arteriosus
- PFO – patent foramen ovale
- Intracardiac – VSD
- Intrapulmonary – ventilation/perfusion mismatch

Shunt physiology

- THINK RESISTANCE
- Pediatric cardiology is easy
- Blood flows along the path of least resistance
- If you know the anatomy and you know the relative resistance in the system; you can understand the disease and predict the clinical findings
Pulmonary Vascular Resistance (PVR)

- In utero – very high, blood by-passes the lungs
- Post natal – drops dramatically at birth then gradually over next six months
- In the presence of shunts, PVR is key factor determining if blood shunts R to L or L to R
- Oxygen and nitric oxide decrease PVR

Systemic Vascular Resistance (SVR)

- Must consider SVR in relation to PVR
- This relative resistance determines direction of shunt flow
- SVR normally 10 to 20 times greater than PVR in “adult physiology”
- SVR may be increased with “pressors” dopamine or dobutamine
- SVR can decrease with sepsis, fever, etc.

Obstructive Lesions

- Obstructive lesions may cause R to L shunt or L to R shunt depending on location
- Right sided lesions (TS, PS, branch PS)
- Left sided lesions (MS, AS, Coarctation)

Shunt Physiology

- To understand the physiology and clinical findings in patients with congenital heart disease think “Where is the blood going?” and “Is the blood red, blue, or mixed?”
- Pediatric cardiology is easy!

Patent ductus arteriosus (PDA)

- 45% of infants <1750g, 80% of infants <1200g
- Leads to CHF in 15% pts<1750g, 40% in pts<1500g
- Females > males
- Very important shunt in the setting of congenital heart disease
Ventricular septal defect (VSD)

- 15 to 20% of all CHD
- Commonly present with complex CHD where it may exist as an obligate shunt
- Simple defects often will close spontaneously

Tetralogy of Fallot (TOF)

- 10% of CHD, most common cyanotic lesion
- Classic definition: unrestrictive VSD, pulmonary stenosis, RV hypertrophy, and over-riding aorta
- May be blue or pink depending on degree of PS and relative resistance

Atrial Septal Defect (ASD)

- Primum vs. secundum defect
- Patent foramen ovale vs. ASD
- Generally not significant hemodynamically early on
- If large will eventually cause PVOD (in adulthood) and therefore should be closed
- Can be closed non-surgically
Left Ventricular Outflow tract Obstructions

- Subvalvar AS: muscular (IHSS) or fibrous
- Valvar AS: 3-6% of CHD, M>F, may be critical
- Coarctation of the Aorta: 8-10% of CHD, M>F, Turner’s syndrome
- Hypoplastic Left Heart Syndrome: 1% of CHD, most common cardiac cause of death in first month of life
Right Ventricular Outflow Tract Obstructions

- Pulmonary atresia: if no branch PAs exist, bad disease
- PA with intact ventricular septum: RV severely hypertensive in utero, coronary sinusoids
- Pulmonary stenosis: 8% of CHD
- Peripheral pulmonic stenosis: benign, murmur in axillae

Atrioventricular Valve Abnormalities

- Common AV canal (endocardial cushion defect: Down’s syndrome
- Tricuspid stenosis/atresia: may lead to hypoplastic right heart
- Mitral stenosis/atresia: may be a cause of HLHS
- Tricuspid regurgitation
- Mitral regurgitation
- Ebstein’s anomaly: SVT is common
Complex Others

- Complete Transposition of the Great Arteries (TGA): may need urgent balloon septostomy
- Double Outlet Right Ventricle
- Heterotaxy syndromes
- Ventricular inversion (congenitally corrected TGA)

When to start Prostaglandin

- Ductal dependent systemic circulation
  - Hypoplastic left heart syndrome
  - Critical AS or coarctation
- Ductal dependent pulmonary circulation
  - Severe pulmonary stenosis
  - Cyanotic tetralogy
  - Tricuspid atresia
  - Pulmonary atresia