Cyanotic VS. Acyanotic CHD

Acyanotic heart disease	Cyanotic heart disease
L to R shunt	R to L shunt
obstruction occurring beyond lungs	
blood passes through pulmonic circulation	blood bypasses the lungs
↓	↓
oxygenation takes place	no oxygenation occurs
↓	↓
↓ levels of deoxygenated blood in systemic circulation	↑ levels of deoxygenated blood enters systemic circulation
↓	↓
no cyanosis	cyanosis

Cyanosis: blue mucous membranes, nail beds, & skin secondary to an absolute conc. of deoxygenated Hb of at least 30 g/dL

Description

ACYANOTIC

Description (cont)

L to R shunt lesions

- extra blood displaced through communication frm L to R side of heart → ↑ pulm. blood flow → ↑pulm. pressures
- shunt vol. dependent upon 3 factors:
  1) size of defect
  2) pressure gradient b/w chambers / vessels, and
  3) peripheral outflow resistance
- untreated shunts can result in pulm. vascular disease, L ventricular dilatation & dysfunction, R ventricular HTN & RVH, and ultimately R to L shunts

Obstructive lesions

- present w/ ↓ urine output, pallor, cool extremities & poor pulses, shock, or sudden collapse

CYANOTIC

- syst. venous return re-enters syst. circulation directly
- most prominent feature = cyanosis (O2 sat. <75%)
- hyperoxic test differentiates b/w cardiac & other causes of cyanosis
  --> obtain preductal, R radial ABG in room air, then repeat after child inspires 100% O2
  --> if PaO2 improves to >150 mmHg, cyanosis less likely cardiac in origin
- pre-ductal & post-ductal pulse oximetry
  --> >5% diff. suggests R to L shunt

1. Acyanotic incl.:
- ASD, VSD, PDA
- Coarctation of aorta, Aortic stenosis, Pulm. stenosis

2. Cyanotic incl.:
- Tetralogy of Fallot
- TGA, Total Anomalous Pulm. Venous Return, Truncus Arteriosus, Hypoplastic L Heart Syndrome

Atrial Septal Defect

3 types:`

- *ostium primum*
  -- common in DS
  -- defect located @ mitral / tricuspid valve
- *ostium secundum*
  -- most common type
  -- 50-70%
  -- defect located @ septum b/w L & R atria
- *sinus venosus*
  -- defect located @ entry of SVC into R atrium

Epidemiology

- 6-8% of congenital heart lesions
- common in pts. w/ certain congenital disorders (eg. DS, FAS)

Natural history

- 80-100% spontaneous closure rate if ASD diameter <8mm
- if remains patent, CHF & pulm. HTN can develop in adult life

Clinical features

- hist.: often asymp. in childhood
- phy. exam: grade 2-3/6 pulm. outflow murmur, widely split, & fixed S2
- children w/ large ASDs may hv. signs of heart failure
  --> tachypnea, FTT, hepatomegaly, pulmo. rales/retractions

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Atrial Septal Defect (cont)

Investigations

- ECG: RAD, mild RVH, RBBB (normal ECG doesn't rule out)
- CXR: ↑ pulmo. vasculature, cardiac enlargement (normal ECG doesn't rule out)
- echo: diagnostic

Management

- elective surgical / catheter closure b/w 2-5 yr, though majority req. no surgery
- size <8 mm will likely spontaneously close

Ventricular Septal Defect

most common congenital heart defect

(30-50%)

SMALL VSD (majority)

Clinical Features

- hist.: asymp., normal growth, & development
- phy. exam: early systolic to holosystolic murmur, best heard at LLSB, thrill

Investigations

- echo to confirm diagnosis
  (ECG & CXR are normal)

Management

- most spontaneously close

MODERATE-TO-LARGE VSD

Epidemiology

- CHF by 2 mo
- late secondary pulm. HTN if left untreated

Ventricular Septal Defect (cont)

Clinical features

- hist.: delayed growth, ↓ exercise tolerance, recurrent URTIs or "asthma" episodes
- phy. exam: holosystolic murmur at LLSB,
  mid-diastolic rumble @ apex,
  size of VSD inversely related to intensity of murmur,
  loss of splitting of 2nd heart sound & loud P2 suggests pulm. HTN

Investigations

- ECG: LVH, LAH, RVH (normal ECG doesn't rule it out)
- CXR: ↑ pulm. vasculature, cardiomegaly, CHF (normal CXR doesn't rule out)
  echo: diagnostic

Management

- tx. of CHF & surgical closure by 1 yr, if surgery req.

* Size of VSD inversely related to sound of murmur
  --> loud murmur = smaller hole

Patent Ductus Arteriosus

Patent vessel b/.w descending aorta & L pulm. artery

- normally, func. closure w/i first 15 hr of life
- anatomical closure w/i first days of life

Patent Ductus Arteriosus (cont)

Epidemiology

- 5-10 % of all CHD
- delayed closure of ductus common in premature infants (1/3 of infants <1750 g)
  --> this is diff. frm PDA in term infants

Natural history

- spontaneous closure common in premature infants
- less common in term infants

Clinical features

- hist.: aymp. , or have apneic / bradycardic spells, poor feeding, accessory muscle use, CHF
- phy. exam: tachycardia ✚/━ gallop rhythm,
  bounding pulses,
  hyperactive precordium,
  wide pulse pressure,
  cont. "machinery" murmur best heard @ L infraclavicular area

Investigations

- ECG: may show L atrial enlargement, LVH, RVH
- echo = diagnostic
- CXR: may show normal to mildly enlarged heart, ↑ pulm. vasculature, prominent pulm. artery

Management

- indomethacin (Indocid): antagonizes prostaglandin E2, which maintains ductus arteriosus patency
  --> only effective in premature infants
- catheter or surgical closure if PDA causes resp. compromise, FTT, or persists beyond 3rd mo of life

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Coarctation of aorta

Narrowing of aorta

- almost always at lvl. of ductus arteriosus

Epidemiology

- commonly ass. w/ bicuspid aortic valve (50%) ; Turner synd. (35%)

Clinical features

- hist.: often asymp.
- phy. exam:
  -- BP discrepancy b/w upper & lower extremities (↑ sus. / severity if > 20 mmHg diff.)
  -- diminished / delayed femoral pulses relative to brachial pulses (i.e. brachial-femoral delay)
  -- possible systolic murmur w/ late peak @ apex, L axilla, & L back
  -- if severe, presents w/ shock in neonatal period when ductus arteriosus closes

Investigations

- ECG shows RVH early in infancy, LVH later in childhood
- echo / MRI for diagnosis

Prognosis

- can be complicated by HTN
- if ass. w/ other lesions (e.g. PDA, VSD) can lead to CHF

Coarctation of aorta (cont)

Management

- give prostaglandins to keep ductus arteriosus patent for stabilization & perform surgical correction in neonates
- for older infants & children balloon arterioplasty may be an alternative to surgical correction

Aortic Stenosis

4 types`

- valvular (75%)
- subvalvular (20%)
- supravalvular, & idiopathic hypertrophic subaortic stenosis (5%)

Clinical features

- hist.: often asymp. , b/ may be ass. w/ CHF, exertional chest pain, syncope, or sudden death
- phy. exam: SEM @ RUSB w/ aortic ejection click @ apex (only for valvular stenosis)

Investigations

- echo for diagnosis

Management

- valvular stenosis usually treated w/ balloon valvuloplasty, pts. w/ subvalvular or supravalvular stenosis require surgical repair, exercise restriction req.

Pulmonary Stenosis

3 types

- valvular (90%)
- subvalvular
- or supravalvular

Definition of critical Pulm. S.

Inadequate pulm. blood flow,
Dependent on ductus arteriosus for oxygenation,
Progressive hypoxia & cyanosis

Natural history

- may be part of other congenital heart lesions (e.g. Tetralogy of Fallot)
- or in ass. w/ syndromes (e.g. congenital rubella, Noonan synd.)

Clinical features

- hist.: spectrum frm asymp. to CHF
- phy. exam: wide split S2 on expiration,
  SEM @ LUSB,
  pulmonary ejection click (for valvular lesions)

Investigations

- ECG findings: RVH
- CXR: post-stenotic dilation of main pulm. artery (due to ↑ velocity jest past stenotic valve)
- echo: diagnostic

Management

- surgical repair if critically ill or if symp. in older infants/children

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Tetralogy of Fallot

Epidemiology

- 10% of all CHD
- most common cyanotic heart defect diagnosed beyond infancy w/ peak incidence @ 2-4 mo

Pathophysiology

- embryological defect due to ant. & superior deviation of outlet septum leading to: VSD, RVOTO (i.e. pulm. stenosis ✚/━ subpulm. valve stenosis), overriding aorta, & RVH
  --> infants may initially hv. a L → R shunt (∴ no cyanosis)
  --> however, RVOTO = progressive, leading to ↑ R → L shunting w/ hypoxemia & cyanosis
  --> degree of RVOTO determines direction & degree of shunt, & ∴, extent of clinical cyanosis & degree of RVH

Tetralogy of Fallot (cont)

Clinical features

- hist.: hypoxic "tet" spells
  --> during exertional states (crying, exercise), increasing pulm. vascular resistance & decrease in sys. resistance causes ↑ in R-to-L shunting
  --> clinical features incl.
  paroxysms of rapid & deep breathing, irritability & crying,
  ↑ cyanosis,
  ↓ intensity of murmur {↓ flow across RVOTO),
  pt. squatting for relief (↑ peripheral resistance, ↓ R to L shunting)
  --> if severe, can lead to ↓ lvl. of consciousness, seizures, death

phy. exam:
  --> single loud S2 due to severe pulm. stenosis (i.e. RVOTO)
  --> SEM @ LLSB

Investigations

- ECG: RAD, RVH
- CXR: boot-shaped heart, ↓ pulm. vasculature, R aortic arch (in 25%)
- echo: diagnostic

Management of spells

O2, knee-chest position, fluid bolus, morphine sulfate, propranolol, phenyl-ephrine

Treatment

- surgical repair @ 4-6 mo of age
- earlier if marked cyanosis or "tet" spells

Transposition of the Great Arteries

Epidemiology

- 3-5 % of all congenital cardiac lesions
- most common cyanotic CHD in neonates

Pathophysiology

parallel pulm. & syst. circulations
- Systemic: body → RA → RV → aorta → body
- Pulmonary: lungs → LA → LV → pulm. artery → lungs
- survival dependent on mixing through PDA, ASD, or VSD

Physical exam

- neonates: ductus arteriosus closure causes rapidly progressive severe hypoxemia unresponsive to O2 therapy, acidosis, & death
- VSD present: cyanosis not prominent ; CHF w/i first wks of life
- VSD absent: no murmur

Investigations

- ECG: RAD, RVH, or may be normal
- CXR: egg-shaped heart w/ narrow mediastinum ("egg on a string")
- echo: diagnostic

Management

- symptomatic neonates: prostaglandin E1 infusion to keep ductus open until balloon atrial septostomy
- surgical repair: arterial switch performed in first 2 wk in those w/o VSD while LV muscle still strong

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Total Anomalous Pulmonary Venous Return

Epidemiology

1-2% of CHD

Pathophysiology

- all pulm. veins drain into R-sided circulation (syst. veins, RA)
- no direct oxygenated pulm. venous return to L atrium
- often ass. w/ obstruction @ connection sites
- ASD must be present for oxygenated blood to shunt into LA & syst. circulation

Management

surgical repair in all cases & req. urgently for severe cyanosis

Truncus Arteriosus

Pathophysiology

- single great vessel gives rise to aorta, pulm. & coronary arteries
- truncal valve overlies a large VSD
- potential for coronary ischemia w/ fall in pulm. vascular resistance

Management

surgical repair w/i first 6 wk of life

Hypopplastic Left Heart Syndrome

Epidemiology

- 1-3% of CHD
- most common cause of death frm CHD in first mo of life

Hypopplastic Left Heart Syndrome (cont)

Pathophysiology

- LV hypoplasia may incl.
  --> atretic / stenotic mitral and/or aortic valve
  --> small ascending aorta
  --> coarctation of aorta w/ resultant syst. hypoperfusion

Systemic circulation dependent on ductus patency

Upon closure of ductus, infant presents w/ circulatory shock & metabolic acidosis

Management

- intubate & correct metabolic acidosis
- IV infusion of prostaglandin E1 to keep ductus open
- surgical palliation (overall survival 50% to late childhood) or heart transplant

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