

### 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 from 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 (O<sub>2</sub> 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% O<sub>2</sub>
  - > if PaO<sub>2</sub> 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 S<sub>2</sub>
- children w/ large ASDs may hv. signs of heart failure
  - > tachypnea, FTT, hepatomegaly, pulmo. rales/retractions



### 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.: asymp., 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 just 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  $\pm$  subpulm. valve stenosis), overriding aorta, & RVH
  - > infants may initially hv. a L  $\rightarrow$  R shunt (: no cyanosis)
  - > however, RVOTO = progressive, leading to  $\uparrow$  R  $\rightarrow$  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  $\uparrow$  in R-to-L shunting
    - > clinical features incl. paroxysms of rapid & deep breathing, irritability & crying,
      - $\uparrow$  cyanosis,
      - $\downarrow$  intensity of murmur ( $\downarrow$  flow across RVOTO),
      - pt. squatting for relief ( $\uparrow$  peripheral resistance,  $\downarrow$  R to L shunting)
    - > if severe, can lead to  $\downarrow$  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,  $\downarrow$  pulm. vasculature, R aortic arch (in 25%)
- echo: diagnostic

#### Management of spells

- O2, knee-chest position, fluid bolus, morphine sulfate, propranolol, phenylephrine

#### 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  $\rightarrow$  RA  $\rightarrow$  RV  $\rightarrow$  aorta  $\rightarrow$  body
- Pulmonary: lungs  $\rightarrow$  LA  $\rightarrow$  LV  $\rightarrow$  pulm. artery  $\rightarrow$  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

### Hypoplastic Left Heart Syndrome

#### Epidemiology

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

### Hypoplastic 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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