### CONGENITAL HEART DISEASE

#### Epidemiology
- Incidence 8/1000; >75% survive first few months; 15% have >1 cardiac abnormality
- 75% congenital heart disease is acyanotic

#### Innocent Murmurs
- Venous hum: high pitched continuous murmur over base of heart, neck, SCJ; varies with respiration and position of head; disappears on lying down
- Vibratory murmur: short, buzzing mid-systolic murmur at L sternal edge / apex; louder on lying down, softer on standing
- Pulmonary flow murmur: soft blowing ejection SM over pul area; prominent with exercise, fever, anaemia

#### Guilty Murmurs
- Loud, pan-systolic / diastolic; associated with symptoms or thrill; radiate; not brief

#### Duct Dependent Lesions
- Acyanotic: Coarctation of aorta
  - Critical aortic stenosis
  - Hypoplastic L heart syndrome
- Cyanotic: Transposition of the great arteries
  - Pulmonary atresia / stenosis
  - Hypoplastic R heart syndrome

These depend on flow through PDA to provide systemic / pulmonary flow

They are discovered in 1st - 3rd week of life (ie. Neonatal period) with shock and CV collapse (when closure of ductus arteriosus causes rapid decompensation)

#### Examination: check all BP’s, L and R hand SaO2; metabolic acidosis; maybe cyanosis

#### Investigations: blood gas; ECG and CXR

#### Management:
- **Avoid O2:** O2 is a vasodilator → decreased pulmonary vascular resistance → increased L→R shunt → increased pulmonary blood flow; giving O2 may help if pulmonary HTN or pulmonary vasoconstriction; however it may cause pulmonary overcirculation → steal systemic blood flow if PDA and duct-dependent flow → worsen systemic perfusion
- O2 causes vasoconstriction of PDA → decreases R→L flow across PDA → decompensation
  - Only give O2 if signs and symptoms of inadequate tissue perfusion, or if SaO2 significantly below normal baseline for patient
- **PGE1:** 0.1mcg/kg/min → opens PDA → improvement of systemic flow in mins → titrate down to lowest effective dose (usually 0.05mcg/kg/min)
- **IVF:** 10ml/kg bolus to increase preload and CO
- **NaHCO3:** 1-2mmol/kg for severe metabolic acidosis (pH <7)

#### Others: maybe pressors (eg. Dopamine, dobutamine); give empiric Abx as cannot exclude sepsis

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### Acyanotic Heart Disease = L → R

- 75% congenital heart disease

- Presents at 1-3/12 with CCF (when pulmonary resistance decreases and hence L→R shunt increases → pulmonary overcirculation, hence **acyanotic**)

- **Investigations:** CXR shows cardiomegaly and increased lung markings

- **Aetiology:**
  - ASD (PFO) – 10% congenital heart disease
  - VSD – 25% congenital heart disease (most common)
  - PDA – 10% congenital heart disease; normally closes at 15hrs → closes completely by 3/52 to become ligamentum arteriosum
  - Common AV canal – 3% congenital heart disease; strongly associated with Downs Syndrome
  - Coarctation of aorta – pulmonary HTN develops with closure of PDA → CCF; weak pulses in legs; ash-grey colour
  - Hypoplastic L heart syndrome – decr LV outflow; systemic blood flow based entirely on PDA; CV collapse with closure; single heart sound
  - Aortic stenosis – 6% congenital heart disease; CCF if severe; harsh SM to neck
### Congestive Cardiac Failure In Infants

**Aetiology:** L→R shunt → pulmonary overcirculation → increased preload (eg. VSD, AVSD, truncus arteriosus, PDA; **will present in 1st 3/12**)

1. Acute L heart obstruction (eg. AS, COA à→ increased afterload; **will present in 1st 1/52**)
2. 1Y myocardial → decreased inotropic function (eg. Cardiomyopathy, myocarditis; **can present at any age**)
3. Other (eg. Anaemia, metabolic, toxic; **usually presents on D1**, unlike other causes)

**Symptoms:** SOB after feeding, poor feeding, sweating, tachypnoea, tachycardia, cardiomegaly, gallop rhythm, thrill, enlarged liver / spleen, increased incidence of chest infections, weak pulse, FTT, fatigue; pulmonary rales; increased WOB; no peripheral oedema

**Management:** O2: may causes worsened shunt as above, so aim SaO2 no more than 95%

**Medications:**

1. Frusemide 1-2mg/kg IV → PO frusemide and spironolactone
2. Digoxin (for inotrope): 40mcg/kg if >1/12, 30mcg/kg if <1/12, 20mcg/kg if prem
   - give ½ dose as bolus → ¼ 8-12hrs later → ¼ 8-12hrs later
3. Consider dopamine / dobutamine (5-10mcg/kg/min) / nitroprusside / calcium channel blockers in consultation with cardiologist

**Other:** Elevate head of bed; if give IVF, be careful

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### Cyanotic Heart Disease = R → L

25% congenital heart disease

**Aetiology:** caused by mixing of oxygenated and deoxygenated blood

1. Tetralogy of Fallot = R → L
   - 10% congenital heart disease (most common cyanotic heart disease)
   - 1. Pulmonary stenosis ± hypoplastic pulmonary artery → RV outflow obstruction (à→ESM with thrill in pulmonary area, and at L sternal edge radiating to back)
   - 2. RVH →RV heave
   - 3. Large VSD (subaortic perimembranous); R→L shunt to aorta
   - 4. Over-riding aorta (aortic root overlying septum and VSD → ejection click)

If associated with ASD = Pentalogy of Fallot (occurs in 10%)

**Symptoms:** cyanosis (due to infundibular spasm and decreased pulmonary blood flow; degree of cyanosis depends on extent of RV outflow obstruction and hence R→L shunt); onset of cyanosis in 1st few wks/mths of life; cyanosed after feeding, squatting after exercise (decreased return of desaturated blood from legs, increased aortic pressure, increased afterload → decreased size of R→L shunt)

**Examination:** murmurs as above; clubbing, cyanosis, loud S2; normal pulses, continuous PDA murmur

**Investigations:**

- ECG: ventricular arrhythmias in 40%; AF, flutter common; RAD; RAH; RVH; tall peaked T waves
- CXR: small pulmonary arteries (due to decreased blood flow); boot shaped heart (due to RVH); oligoemic lung fields; no cardiomegaly

**Management:**

- treat by decreasing SVR, HR, agitation, infundibular spasm → decreasing R→L shunt
- Management of Tet spells: aim is to increase pulmonary blood flow by increasing preload, provide pulmonary vasodilation, increase afterload to reverse R→L shunting
  1. O2 100% (usually has little effect though; causes pulmonary vasodilation)
  2. Knees bent posture (increases VR and SVR); rest; abdominal compression; calm child
  3. Morphine 0.1-0.2mg/kg IV/IM (decreases catecholamines and RR)
4. IVF 10-20ml/kg (increases preload, decreases dynamic outflow obstruction; give this before drugs that may cause hypotension)
5. HCO3 1-2mmol/kg IV (corrects acidosis, promotes pulmonary vasodilation) – repeat at 10-15mins
6. Metaraminol 50mcg/kg IV over 10-15mins → 0.25-1mcg/kg/min infusion (increases afterload → decreases R→L shunt)
7. Esmolol 500mcg/kg over 1min → 50mcg/kg/min infusion (decreases RV outflow obstruction via decreased infundibular spasm, increases pulmonary outflow)
8. Ketamine, RSI

Operative Management: corrective surgery (when very young, <1% surgical mortality in children; 90% 30yr survival with good levels of function); early palliative OT if severely ill (pulmonary valvuloplasty to increase pulmonary arterial blood flow)

Complications: Tet spells, polycythaemia (→ thrombosis), consumptive coagulopathy, endocarditis

Transposition of the Great Vessels = R → L
5-8% congenital heart disease

Epidemiology: most common cyanotic lesion manifesting in newborn period
Pathophysiology: only compatible with life if mixing of R and L circulations – VSD (in 20-40%), ASD, PDA (3)
Symptoms: onset of severe cyanosis within hours (ie. PDA dependent) unresponsive to O2
Examination: loud S2; no audible murmurs (systolic murmur if VSD)
Investigations: CXR (cardiomegaly; egg shaped heart; increased pulmonary vasculature); ECG (RAD, RVH)
Management: urgent balloon atrial septoplasty will provide rapid improvement; arterial-switch operation (low mortality, excellent long-term outcome)

Ebstein’s Anomaly = R → L
Pathophysiology: leaflets of tricuspid valve displaced into right ventricle, portion of right ventricle located in right atrium → tricuspid regurgitation (sometimes stenosis); ASD in 80%
Symptoms: vary from very mild to very severe; cyanosis worsens with increased R→L shunt through ASD
Examination: wide split S1 and S2; S3 and S4; systolic murmur at lower L sternal edge (due to TR); hepatomegaly
Investigations: ECG (P pulmonale, RBBB, 1st degree heart block); CXR (cardiomegaly, R atrial enlargement, decreased pulmonary vascular markings)
Management: temporary arterial shunt from L→R if severe to increase pulmonary blood flow; repair

Persistent Truncus Arteriosus = R → L
<1% congenital heart disease
Pathophysiology: single artery arises from heart then branches into pulmonary artery and aorta; pulmonary blood flow may be increased (→ CCF), normal or decreased (→ cyanosis)
Symptoms: mild cyanosis, CCF in newborn
Investigations: CXR (cardiomegaly, pulmonary plethora); ECG (LVH and RVH)

Tricuspid Atresia = R → L
1-2% congenital heart disease
Pathophysiology: no tricuspid valve; development of R ventricle and pulmonary artery wrong → decreased pulmonary blood flow; needs mixing to survive (ASD, VSD, PDA) as R atrium needs R→L shunt in order to empty
Investigations: CXR: slight cardiomegaly, pulmonary oligaemia
ECG: RAH, LAH, LVH, superior axis
**Eisenmenger Syndrome**  
L → R becomes R → L

**Pathophysiology:** L→R shunt → increased pulmonary blood flow → pulmonary hypertension (increased arteriolar muscles) → becomes R→L shunt; occurs with VSD, ostium primum defect, transposition of great vessels with large shunt

**Symptoms:** haemoptysis (20%), CVA (10%), paradoxical embolism, brain abscess (5%); symptoms occur in adolescents / young adults

**Examination:** clubbing, cyanosis; normal / high JVP; small volume pulse; RV heave; loud P2; R sided S4; murmur of original defect disappears when Eisenmenger syndrome develops; decrescendo diastolic murmur (due to pulmonary regurgitation), pansystolic murmur (due to tricuspid regurgitation)

**Investigations:** ECG: AF, flutter; RAD; RVH; P pulmonale  
CXR: cardiomegaly; large pulmonary art

**Management:** no surgical treatment available; maintain intravascular volume; avoid hypoxia and vasodilation

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**CXR Summary**

| Cardiomegaly | TOF, TGV, Ebstein’s; Eisenmenger’s; ASD; VSD |
| Pulmonary plethora | L→R shunt; TGV; patent truncus arteriosus |
| Pulmonary oligaemia | TOF, Ebstein’s |
| Pulmonary venous congestion | L heart obstruction |
1st breath → increased PaO₂ in lungs → decreased PVR → increased pulmonary blood flow → closure of ductus venosus and PDA (at 15hrs) → increased blood flow through LA → LAp > RAp → closure of PFO (at 3 months)