PAEDIATRIC CARDIOLOGY

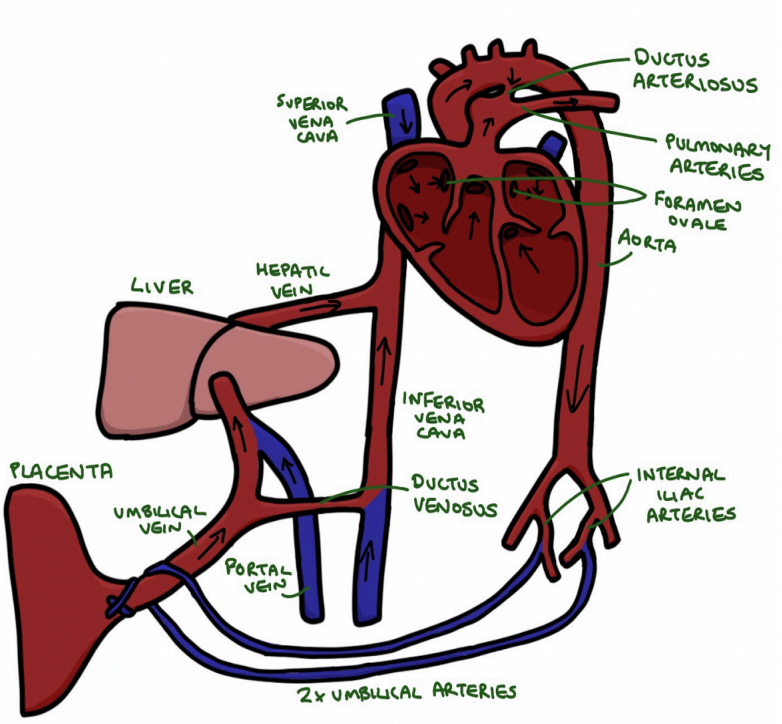
NORMAL FOETAL CIRCULATION:

STEP 1: Cord clamping
1) immediate vasoconstriction (areterial vasospasm) due to cold exposure
2) ↑ systemic vascular resistance

STEP 2: Baby Cries / 1st breath (secs)
1) Alveoli expands = amniotic fluid pushes into pulmonary vessels
2) ↓ **pulm. vascular resistance**
3) Reduced RA pressure and high LA pressure squashes atrail septum
4) Mechanical pressure closure of foramen ovale

STEP 3: Ductus venosus closes (mins)
1) absent flow through umbilical veins
2) closure occurs days later to become ligamentum venosum

STEP 4: Ductus Arteriosus closes (>10 hrs)
1) increased blood O2 causes prostaglandin drop
2) Keep patent using **prostin (PGE analog)** for cyanotic HD (e.g. trcicuspid atresia) - enable L --> R shunting
3) **NSAIDs** for closure



HEART EMBYROLOGY

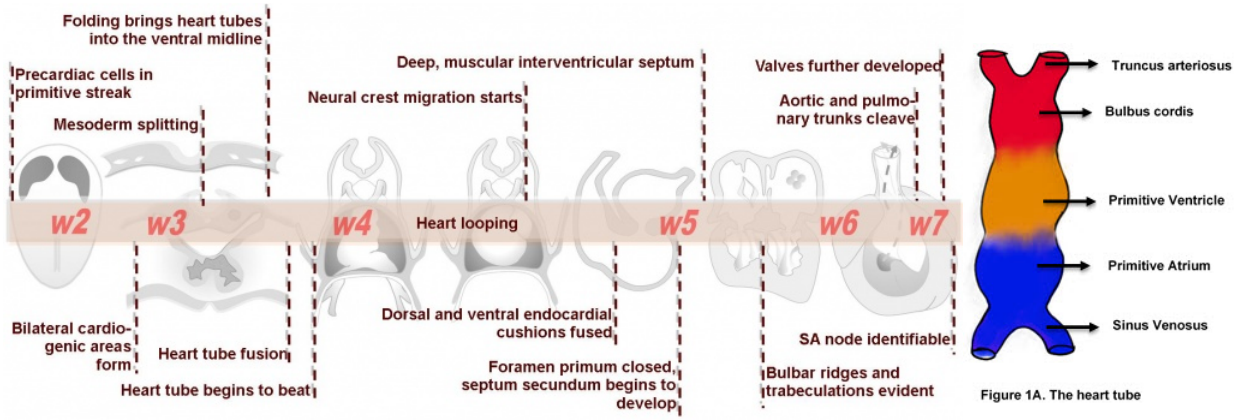
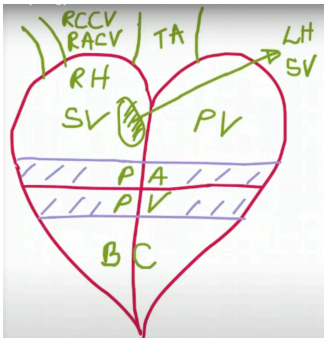
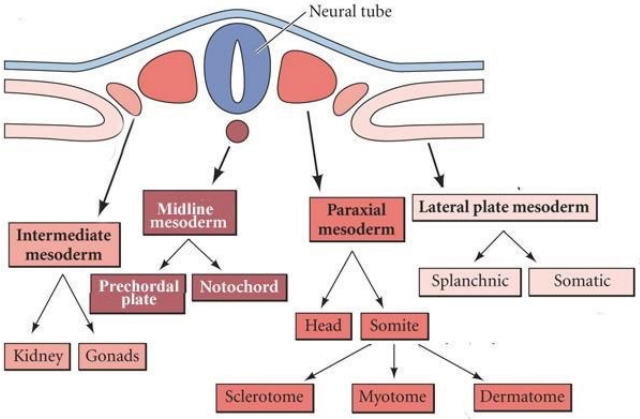


Figure 1A. The heart tube

Foetal structure	Adult Derivative
1. **sinus venosus**	forms **SVC** and part of the **right atrium**
2. **primordial atrium**	forms the anterior parts of the **right and left atria**
3. **primordial ventricle**	forms most of the **left ventricle**
4. **bulbus cordis**	forms most of the **right ventricle** and **parts of the outflow tract**s for the aorta and pulmonary trunk
5. **truncus arteriosus**	form the **pulmonary trunk and aorta**





CONGENITAL HEART DEFECTS

	Aortic Stenosis	Pulmonary stenosis	ASD	VSD	PDA	Eisenmenger syndrome	TOF	TGA	Coarctation of aorta	Ebstein's anomaly
Type	Cyanotic	Cyanotic	Acyanotic (L → R)	Acyanotic	Acyanotic (L → R)	Cyanotic (R → L)	Cyanotic (R → L)	Cyanotic (R → L)	Cyanotic (R → L)	Cyanotic (R → L)
RF		• TOF • William • Noonan • Cong. rubella	• Pre-term • Females	• Most common type • Pre-term • Turner's • Down's	• Pre-term	Acyanotic HD (ASD, VSD, PDA)	• Cong. Rubella • Advanced age • EtOH • GDM	Most common	Turner's	
PP	**Narrowed aortic valve – restrict blood flow from LV to aorta** ➢ Pts may have one, two, three or four leaflets	Abnormal development of 3 leaflets – thickened or fused	• Defect in atrial septum due to incomplete fusion of endocardial cushions (septum primum and secondum) ***Different types*** • PFO • Ostium secondum • Ostium primum → leads to AVSD	Defect in ventricular septum due to membranous septum that has not been formed	**Failure of ductus arteriosus closure** • Higher pressure in aorta than pulmonary vessels (creates L → R shunt) • **Non-congenital**	• When blood flows from R) side of heart to left across structural heart lesion bypassing lung • secondary to PHTN where pulmonary pressure > systemic pressure leading to R → L shunt)	• VSD, • RVH, • PS, • Overriding aorta – entrance to aorta placed further R) than normal	When bulbar ridges do not go 180 deg spiral *If VSD or PDA– this is good – as some mixing between arterial and venous systems	Narrowing of aortic arch reduces pressure distal and increases pressure proximally	Congenital tricuspid valve lower in R) side of heart ➔ bigger RA and smaller RV
Murmur	Ejection systolic in R) 2ND IC space rad to carotids DDx: HOCM	Ejection systolic L) 2nd IC space DDx: HOCM	Mid-systolic crescendo-decrescendo - upper LSE	Pan-systolic – lower LSE + systolic thrill - 3rd, 4th IC spaces (DDx:, MR, TR)	Continuous crescendo-decrescendo murmur "machine-like" murmur	**DEPENDS**	Ejection systolic	**NONE**	Systolic L) infraclavicular area	**NONE**
Signs	Asymptomatic • Palpable thrill and slow rising narrow PP • SOBOE	Asymptomatic • Palpable thrill • RVH – R) heave • Raised JVP (giant A waves)	Fixed split S2 sound (no change w/ inspiration or expiration) • Child sx = SOB, FTT, LRTi • Adult sx = SOB, HF, stroke	Asymptomatic → • **Child sx** = SOB, FTT, poor feeding, ↑RR • **Adult sx** = SOB, HF, stroke	• 2nd heart sound • Wide PP • Child sx = SOB, FTT, LRTi • Adult sx = SOB, HF, stroke • **Sudden cyanosis +cardiac failure**	• Loud P2, raised JVP, peripheral oedema, R ventricular heave • Cyanosis, SOB, clubbing, facial plethora (polycythaemia)	• Cyanosis, clubbing, • poor feed, FTT • **"Tet" spells** – acute worsening R → L shunt after exertion, crying, waking as PVR increases and SVR decreases (XS CO_2 – vasodilation of SVR)	If decompensated ➢ RDS ➢ Tachycardia ➢ Poor feeding ➢ Poor wt gain • Sweating	• **L) ventricular heave** • **Underdeveloped L) arm** (reduced L subclavian artery) • **↑WoB, Poor feeding, grey, floppy baby**	• **Gallop rhythm (3rd + 4th HS)** • **HF signs (oedema)** • **Cyanosis, SOB,** • **Poor feeding** • **Syncope**
Comp.	• L) vent. Outflow tract obstruction • Heart failure (early) • V. arrythmia • Bacterial endocarditis • Sudden death		• Stroke (VTE) • AF or flutter • Migraine w/ aura (if PFO) • PHTN → RVH (RHF) → LVH → Eisenmenger	• IE risk • PHTN → RVH → LVH → Eisenmenger	• Asymptomatic (if small) • PHTN → RVH → LVH → Eisenmenger	Reduced LE by 20 years → due to HF, infection, VTE, haemorrhage	Severe TET spells ➢ Irritable ➢ Cyanotic ➢ LOC ➢ Seizures ➢ Death	• VSD • PS • Coarctation of aorta	• Heart failure → death (especially newborn)	• Cardiac arrest • WPW • ASD
Ix	**ECHO (gold-standard)**	**ECHO (gold-standard)**	ECG, CXR, ECHO (assess size of L → R shunt)	ECG, CXR, ECHO (assess size of L → R shunt)	ECG, CXR, ECHO (assess size of L → R shunt)		**Ante-natal scan or newborn check** ECG, CXR, ECHO ➢ "boot-shaped heart" due to R) vent. Thickening	**Ante-natal scan or newborn check** ECG, CXR, ECHO "egg-on string"	**4-limb BP** – high BP in limbs supplied before coarctation and low BP distal to coarctation **CT angiography**	**ECHO (gold-standard)**
Mx	*Regular f/u with paed. Cardiologist* • ECHO • ECG • Exercise testing *monitor progression + watch and wait *Rx for significant stenosis:* • Valve replacement (via femoral vein) • Percutaneous balloon aortic valvuloplasty • Surgical aortic valvuloplasty		Referral to paediatric cardiologist • Asymptomatic in childhood • **Elective closure** at 3-5yo via transcatheter surgical closure or open heart surgery • **Anti-coags** – reduce risk of clots and strokes	Referral to paediatric cardiologist • Detected via newborn check or antenatal scans • Trans-catheter closure before 18/12 old • ABx prophylaxis to reduce risk of IE esp. post-op	• ECHO until 1 yo *If failure to close or symptomatic* • NSAIDs – indomethacin • Trans-catheter via femoral vein • Surgical closure / ligation	*correct underlying defect* 1) FiO2 – manage Sx only 2) PHTN → sildanefil 3) Rx arrythmias 4) Polycythaemia-venesection 5) Thrombosis = prophylactic anticoags 6) IE = prophylactic ABx *Definitive Mx:* ➢ Heart-lung transplant	"*TET spells*" Squat or knees to chest to increase SVR → increase blood into pulm. Vessels ➢ *FiO2* ➢ *BB* ➢ *IVF* ➢ *Morphine* ➢ *Phenylephrine (increase SVR)* *General Mx* Prostin infusion to keep PDA patent until total surgical repair (90% live into adulthood if successful)	*General Mx* Prostin infusion to keep PDA patent until total surgical repair **1) Balloon septostomy** (insert into foramen ovale to create large ASD) **2) Open heart surgery** (definitive Mx) -> cardiopulm. Bypass machine to perform "arterial switch" procedure within 1st few days of birth	*Asymptomatic* ➢ Watch and wait *Severe coarctation* • Prostin – to keep PDA open while waiting surgery • Surgery to correct coarctation and ligate ductus arteriosus	*Medical Mx for* • Arrythmias • HF • Prophylactic ABx for IE *Definitive Mx* ➢ Surgically correct defect

Nb:

- **innocent (flow) murmurs** = soft, short, systolic (**grade 2**), symptomless, situational dependent (heard if unwell or feverish)
- **acyanotic** – ASD, VSD, PDA, PFO
- **Cyanotic** – **TOF, TGA**, **Truncus arteriosus**, **Total anomalous venous return** , **Tricuspid atresia** (obstructed L) heart), **Hypoplastic L heart**