❤️ Cardiovascular Pathology

Congenital Heart Diseases

🗺️ Right-to-Left Shunts (Cyanotic)

Key idea: Deoxygenated blood bypasses lungs → enters systemic circulation → early cyanosis ("blue babies") 🫐

Often present at birth and may require urgent intervention + PDA maintenance (Prostaglandin E_1).

🔑 The "5 T's"

Condition	Key Clue

Truncus arteriosus	1 vessel
Transposition of great arteries (TGA)	2 vessels switched
Tricuspid atresia	3 = Tri
Tetralogy of Fallot (TOF)	4 defects
TAPVR	5 letters

1️⃣ Truncus Arteriosus

📌 Core Defect

Failure of aorticopulmonary septum formation → single vessel leaving heart

🔬 Pathophysiology Flowchart

Neural crest cell migration defect → Failure of truncus to divide → One common outflow tract → Mixing of oxygenated + deoxygenated blood → Cyanosis early in life

🧠 Key Points

- Almost always associated with VSD
- Blood fully mixes → less severe cyanosis than TGA initially

⚠️ Exam Trap

If you see single great vessel + VSD → think truncus arteriosus

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2️⃣ D-Transposition of Great Arteries (TGA)

📌 Core Defect

- Aorta from RV (anterior)

- Pulmonary artery from LV (posterior)
- Parallel circulations (no mixing!)

🔬 Pathophysiology Flowchart

Failure of aorticopulmonary septum to spiral → Great vessels reversed → Systemic circulation: deoxygenated blood recirculates → Pulmonary circulation: oxygenated blood recirculates → No oxygen delivery to body → Life incompatible unless mixing present

🧠 Survival Depends On:

- VSD
- PDA
- Patent foramen ovale

📸 Classic Finding

- "Egg-on-a-string" appearance on CXR 🥚

⚠️ Exam Tips

- Most die within months without surgery
- Prostaglandin E_1 used to keep PDA open

3️⃣ Tricuspid Atresia

📌 Core Defect

- Absent tricuspid valve
  - No RA → RV flow
  - Hypoplastic RV

🔬 Pathophysiology Flowchart

No tricuspid valve → Blood cannot enter RV → RA pressure ↑ → Blood shunts RA → LA via ASD → Requires VSD to reach lungs → Survival depends on mixing

🧠 Key Requirements for Life

- ASD (mandatory)
- VSD (mandatory)

⚠️ Exam Tip

If no tricuspid valve + hypoplastic RV + required shunts

→ Tricuspid atresia

4️⃣ Tetralogy of Fallot ⭐

📌 Cause

Anterosuperior displacement of infundibular septum

🧩 The 4 Components (PROVe mnemonic)

Feature	Explanation

Pulmonary stenosis	Most important determinant 🔥
RVH	Due to pressure overload
Overriding aorta	Receives blood from both ventricles
VSD	Allows R→L shunting

🔬 Pathophysiology Flowchart

Pulmonary stenosis → RV pressure ↑ → Blood shunts R → L via VSD → Deoxygenated blood enters systemic circulation → Cyanosis 🫐

😤 "Tet Spells"

Triggered by:

- Crying 😭
- Fever 🌡️
- Exercise 🏃

Mechanism:

↑ RV outflow obstruction → ↑ R→L shunt → Sudden cyanosis

🧠 Squatting Mechanism

Squatting → ↑ SVR → ↓ Right-to-left shunt → ↑ Blood to lungs → Improves cyanosis ✅

📸 Classic Finding

- Boot-shaped heart on CXR 👢

🧬 Association

- 22q11 deletion (DiGeorge syndrome)

🇸 TAPVR (Total Anomalous Pulmonary Venous Return)

📌 Core Defect

Pulmonary veins drain into right heart instead of left atrium

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🔬 Pathophysiology Flowchart

Pulmonary veins connect to SVC/coronary sinus → Oxygenated blood returns to right heart → Mixing occurs in RA → Requires ASD (± PDA) → Some oxygenated blood reaches systemic circulation

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🧠 Key Point

- ASD is essential for survival

⭐ Extra: Ebstein Anomaly (Not a classic "5 T", but important)

📌 Core Defect

Downward displacement of tricuspid valve → RV becomes partially "atrialized"

🔬 Pathophysiology Flowchart

Tricuspid valve displaced → RV size/function reduced → Tricuspid regurgitation → RA enlargement → Right-sided heart failure

🧠 Associations

- Accessory conduction pathways (⚡ arrhythmias)
- Often linked to maternal lithium use

🧠 Quick Comparison Table

Condition	Key Defect	Survival Depends On	Classic Clue
Truncus arteriosus	Single vessel	Mixing already present	VSD + 1 vessel
TGA	Parallel circulation	VSD/PDA/PFO	Egg-on-string
Tricuspid atresia	No tricuspid valve	ASD + VSD	Hypoplastic RV

TOF	4 defects	Severity of PS	Boot-shaped heart
TAPVR	Pulmonary → right heart	ASD	All blood to right side

⚠️ High-Yield Exam Traps

- No mixing = death → think TGA
- Cyanosis relieved by squatting = TOF
- Single vessel = truncus arteriosus
- Missing tricuspid valve = tricuspid atresia
- All pulmonary veins wrong connection = TAPVR

❤️ Left-to-Right Shunts

(Acyanotic → Late Cyanosis)

🔑 Core Concept

- Blood flows from left (high pressure) → right (low pressure)
  - ↑ Pulmonary blood flow 🌊
  - No early cyanosis (acyanotic at birth)
  - Cyanosis develops later if reversal occurs (Eisenmenger) 🫐

📊 Frequency

VSD > ASD > PDA

⚖️ Right vs Left Shunts

Feature	Right → Left	Left → Right

Cyanosis	Early 🫐	Late 🕑
Mechanism	Deoxygenated → systemic	Oxygenated recirculates to lungs
Example	TOF, TGA	VSD, ASD, PDA

1️⃣ Ventricular Septal Defect (VSD) ⭐

📌 Core Defect

Opening in interventricular septum → LV to RV shunt

🔬 Pathophysiology Flowchart

VSD present → Blood flows LV to RV → ↑ Pulmonary blood flow → ↑ Return to left heart → LV volume overload → Heart failure (if large defect) ❤️‍🩹

🧠 Key Features

- Often asymptomatic at birth
- Symptoms appear weeks later (↓ pulmonary resistance after birth)
- Small defects → self-resolve
- Large defects → HF + pulmonary HTN

🧪 Oxygen Saturation Changes

- ↑ O_2 in RV + pulmonary artery

⚠️ Exam Tips

- Most common congenital heart defect
- If untreated → Eisenmenger syndrome

2️⃣ Atrial Septal Defect (ASD)

📌 Core Defect

Opening in interatrial septum → LA to RA shunt

🧠 Types

Type	Features
Ostium secundum	Most common, isolated
Ostium primum	Associated with other defects (e.g., Down syndrome)

🔬 Pathophysiology Flowchart

ASD present → Blood flows LA to RA → ↑ Right heart volume → ↑ Pulmonary blood flow → RV dilation → Possible HF later

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🩺 Classic Finding

- Wide, fixed split S2 🔊

🧪 Oxygen Saturation Changes

- ↑ O_2 in RA, RV, pulmonary artery

⚠️ Exam Gold

- Paradoxical embolism 🧠

  Venous clot → crosses ASD → enters systemic circulation → stroke

🧠 Key Distinction

- ASD ≠ Patent Foramen Ovale (PFO)
  - ASD = true defect
  - PFO = failed fusion

3️⃣ Patent Ductus Arteriosus (PDA)

📌 Core Concept

- Fetal connection persists between aorta & pulmonary artery

🔬 Normal vs Pathologic Flow

Fetal life:

Right → Left (normal)

After birth:

↓ Pulmonary resistance

→ Left → Right shunt

🔬 Pathophysiology Flowchart

PDA remains open → Blood flows aorta → pulmonary artery → ↑ Pulmonary blood flow 🌊 → ↑ Left heart volume → LV overload → HF

🩺 Classic Finding

- Continuous "machine-like" murmur 🤖

🧠 Regulation

Factor	Effect
PGE (Prostaglandins)	Keeps PDA open
↑ O_2	Closes PDA

⚠️ High-Yield Clinical Point

Uncorrected PDA → Pulmonary HTN → Shunt reversal (R → L) → Differential cyanosis 🫐 (Lower limbs cyanotic, upper normal)

🔥 Eisenmenger Syndrome (VERY IMPORTANT)

📌 Definition

Reversal of long-standing left to right shunt → right to left shunt

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🔬 Pathophysiology Flowchart

Chronic L to R shunt → ↑ Pulmonary blood flow → Pulmonary vascular remodeling → Pulmonary hypertension → RV pressure ↑ → RV > LV pressure → Shunt reverses (R → L) → Cyanosis + clubbing + polycythemia

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🧠 Key Features

- Late cyanosis
- Clubbing of fingers ✋
- Polycythemia (↑ RBCs due to chronic hypoxia)

⚠️ Exam Traps

- Any long-standing VSD, ASD, PDA → Eisenmenger
- Once reversal occurs → surgery usually contraindicated

🧠 Quick Comparison Table

Condition	Shunt	Key Feature	Complication
VSD	LV → RV	Most common	HF, Eisenmenger
ASD	LA → RA	Fixed split S2	Paradoxical emboli
PDA	Aorta → PA	Machine murmur	Differential cyanosis

Eisenmenger	R → L (reversal)	Late cyanosis	Clubbing, polycythemia

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🎯 Final High-Yield Summary

- Left → Right = Acyanotic early
- Right → Left = Cyanotic early
- Eisenmenger = the turning point ⚠️
- Always think:
  - Volume overload → HF
  - Pulmonary HTN → reversal → cyanosis

❤️ Coarctation of the Aorta (CoA)

📌 Core Defect

Narrowing of the aorta near the ductus arteriosus insertion (juxtaductal region)

🔬 Pathophysiology Flowchart

Aortic narrowing (juxtaductal) → ↑ Resistance to blood flow distal to narrowing → ↑ Pressure proximal (upper body) → ↓ Perfusion distal (lower body) → Collateral circulation develops (intercostal arteries) → Rib erosion → notching on CXR

🧠 Key Clinical Features

Feature	Explanation
Upper extremity HTN 💪	Blood accumulates before narrowing

Weak/delayed femoral pulses 🦵	↓ Flow to lower limbs
Brachiofemoral delay	Radial pulse before femoral
Cold lower extremities ❄️	Poor perfusion

📸 Classic Imaging Finding

- Rib notching due to enlarged intercostal arteries 🦴

🧬 Associations

- Bicuspid aortic valve
- Turner syndrome
- Other congenital heart defects

⚠️ Complications

- Heart failure ❤️‍🩹
- Berry aneurysm → risk of cerebral hemorrhage
- Aortic rupture
- Infective endocarditis

👶 Persistent Pulmonary Hypertension of the Newborn (PPHN)

📌 Core Problem

Failure of pulmonary vascular resistance (PVR) to decrease after birth

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🔬 Pathophysiology Flowchart

High PVR persists after birth → Blood avoids lungs →

Right-to-left shunting via:

- Foramen ovale
- Ductus arteriosus

→ ↓ Oxygenation → Cyanosis 🫐 + respiratory distress

🧠 Clinical Presentation

- Tachypnea 😮‍💨
- Cyanosis 🫐
- Signs of respiratory distress

🧪 Key Diagnostic Clue

Preductal O_2 saturation > Postductal O_2 saturation

⚠️ Important Distinction

- Pulses are equal (unlike coarctation ❗)

🧬 Risk Factors

- Meconium aspiration
- Neonatal pneumonia
- Abnormal pulmonary vascular development

🧬 Congenital Cardiac Defect Associations

Condition / Exposure	Associated Cardiac Defects
Prenatal alcohol (Fetal alcohol syndrome) 🍷	VSD, ASD, PDA, Tetralogy of Fallot

Congenital rubella 🤰	PDA, pulmonary artery stenosis, septal defects
Down syndrome	AV septal defect (endocardial cushion defect), ASD, VSD
Maternal diabetes 🍬	TGA, truncus arteriosus, tricuspid atresia, VSD
Marfan syndrome 🕸️	MVP, aortic regurgitation, thoracic aortic aneurysm/dissection
Lithium exposure (prenatal) 💊	Ebstein anomaly
Turner syndrome (45,XO)	Bicuspid aortic valve, coarctation of aorta
Williams syndrome	Supravalvular aortic stenosis
22q11 deletion (DiGeorge)	Truncus arteriosus, Tetralogy of Fallot

🧠 Pattern Recognition

Think like this in MCQs:

Upper limb HTN + weak femoral pulse → Coarctation of aorta

Cyanotic newborn + preductal > postductal O_2 → PPHN

Maternal lithium history → Ebstein anomaly

Down syndrome + heart defect → AV septal defect

⚠️ High-Yield Traps

- Coarctation ≠ equal pulses (there is delay!)
- PPHN ≠ structural defect (it's functional vascular problem)
- Rib notching = chronic collateral flow (CoA)

- Preductal vs postductal O_2 difference = shunting clue

🎯 Final Takeaway

- Coarctation = obstruction problem 🚧
- PPHN = failure of neonatal transition 🌬️
- Associations = memorization goldmine 🧠🔥

-> The End <-