

Cyanotic Congenital Heart Anomalies:

Congenital Heart Anomaly	Pathophysiology	Diagnosis (echo)	Heart Murmurs	Signs and Symptoms	Management
Ventricular Septal Defect (VSD) <i>VCP3A3</i>	Hole in the ventricular septum (membranous or muscular part) L→R shunt, biventricular hypertrophy→ pulm HTN	Echocardiogram, Catheterization, CXR- enlarged proximal pulmonary arteries	Pansystolic/Holosystolic murmur in lower left sternal border, loud S2 (pulm HTN)	Fatigue Shunt reversal R→L /Eisenmenger effect- cyanosis bulging sternum, systolic thrill	Small-Observation CHF- digoxin + diuretics, Pulm HTN/volume overload/supracristal VSD-surgery
Atrial Septal Defect (ASD)	Hole in the atrial septum. L→R High in septum: sinus venosus Mid: ostium secundum (m/c) Low: ostium primum	Echocardiogram septum moves in paradoxical way , Catheterization	split S2, systolic murmur	Secundum-asympt, Primium- fatigue, recurrent PNA, MR	Bact endocarditis prophylaxis , definitive is Surgery
Coarctation of the Aorta	Narrowing of the aorta at origin of left subclavian artery near ligamentum arteriosum, associated with turner syndrome in women	BP, Echocardiogram, CXR notching of ribs & figure 3 sign	Systolic murmur may be heard over the back or abdomen	HTN in arms but ↓BP & pulse in lower extremities, radio-femoral delay	Surgical decompression, or percutaneous balloon arthroplasty >2years
Patent Ductus Arteriosus (PDA)	Persistence of ductus arteriosus after birth- hole b/w pulm artery and aorta . L→R, Common in LBW, rubella infected babies, females	Echocardiogram, CXR cardiomegaly, ↑pulm vascular markings	Continuous machinery-like murmur, bounding with wide pulse pressure	Breathlessness and sweating while feeding,	In Preterm babies- indomethacin Surgery- ligation and division of ductus
Patent foramen ovale	Hole in atrial septum that closes at birth normally	Bubble echo	murmur	Closes spontaneously	Observe, surgery closure if symptoms occur
Aortic Stenosis	Narrowing of the aortic valve or aorta. Most common form- valvular	Echocardiogram, Catheterization	Mid systolic crescendo decrescendo murmur , soft S2	Chest pain, Fainting, Fatigue	Medications, Balloon valvuloplasty , Surgical
Pulmonary Stenosis	Narrowing of the pulmonary valve or artery	Echocardiogram, Catheterization	Mid systolic crescendo decrescendo murmur	Cyanosis, Fatigue, Shortness of breath	Balloon valvuloplasty , Surgical
Atrioventricular Canal Defect	Incomplete fusion of atrial and ventricular septa and malformation of AV valves	Echocardiogram, Catheterization	Holosystolic murmur	Poor weight gain, Respiratory infections	Surgical repair

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TSE <i>Tetralogy of Fallot</i>	Combination of four heart defects (PVOR)-pulmonary stenosis-VSD-over riding of aorta over ventricular septum-right ventricular hypertrophy	Echocardiogram, Chest X-ray: boot shaped heart + diminished pulmonary vascular markings ' oligemic lung fields '	Systolic ejection (pulmonic stenosis), holosystolic murmur (VSD)	Cyanotic attacks/ blue spells usually in morning after crying, Dyspnea	Squatting (↓left-right shunt by ↑SVR), iron supplements, endocarditis prophylaxis , Palliative surgery- Blalock-taussig, waterston, potts shunt Surgical repair (total correction)

<i>Transposition of the Great Arteries (TGA)</i>	Switched positions of the pulmonary artery and aorta , two parallel circuits m/c infants of diabetic mothers	Echocardiogram, Arterial blood gases, CXR; egg placed on its side	Single loud second heart sound (S2), systolic murmur	Cyanosis tachypnea within 1hr of birth , large babies (TGA w/ VSD), Poor feeding	PGE IV after birth, digoxin, diuretics, Surgery after 2 wks age but in the meantime -Rashkind/BAS, mustard procedure
<i>Truncus Arteriosus</i>	Single large vessel arises from the heart , instead of separate aorta and pulmonary arteries	Echocardiogram, Catheterization , CXR; waterfall/hilar comma sign	Single loud heart sound, Continuous murmur	Cyanosis, Poor feeding, diaphoresis	Surgical repair
<i>Total Anomalous Pulmonary Venous Drainage (TAPVD)</i>	Pulmonary veins don't connect normally to the left atrium	Echocardiogram, Cardiac catheterization	Loud S2	Cyanosis, Rapid breathing, feeding intolerance, pulmonary edema	Surgical repair (reattaching pulmonary veins to the left atrium)
<i>Tricuspid Atresia</i>	Absence of tricuspid valve	Echocardiogram, Catheterization	Single loud S2, maybe S3	Cyanosis, trouble feeding	Symptomatic (digitalis diuresis for congestion) surgery blalock-taussig shunt
<i>Ebstein Anomaly</i>	Malformation of the tricuspid valve, displacement towards the apex, backflow into right atrium Also with ASD	Echocardiogram, right atrial enlargement (tall broad p waves)	Systolic murmur (Tricuspid regurgitation), S3,S4	Cyanosis, Heart failure	Medications, Surgery Da Silva's cone repair