

Cardiac Evaluation of the Infant

Three Typical Presentations Depending on Lesion:

Cyanosis

Cardiovascular Collapse

Congestive Heart Failure/Overcirculation of the Pulmonary Circuit

Cyanosis:

Cardiac or Pulmonary Cause?

Pulmonary:

No murmur

Increase respiratory distress (tachypnic, retractions)

Changes in PCO₂

Less cyanosis with agitation

CXR-small heart/parenchymal changes

Quiet precordium

Cardiac:

Murmur

Less s/s respiratory distress

Minimum changes in PCO₂

More cyanosis with agitation

CXR-Large heart or normal/pulmonary edema

Palpation of precordium + RV impulse/heave.

LESIONS PRESENTING WITH CYANOSIS:

5 T's:

Transposition

Tetralogy of Fallot with pulmonary atresia/stenosis

Tricuspid atresia

Truncus arteriosus

Total anomalous pulmonary venous return (obstructed)

TRANSPOSITION OF THE GREAT ARTERIES:

Most common cyanotic lesion in newborn period; 5% of all congenital heart defects.

90% present in the first day of life.

Aorta arises from the Right Ventricle; Pulmonary Artery arises from the Left Ventricle.

Parallel circulation instead of series.

Must have some mixing at the atrial, ventricular or ductus level.

CXR- Oval or egg shaped heart due to anterior Aorta and large Right Ventricle.

PGE may be used to maintain ductal patency.

Atrial septostomy is life saving.

Definitive repair- Arterial switch +/- Ventricular Septal Defect closure if present.

TETRALOGY OF FALLOT:

Most common form of cyanotic heart disease; 6-10% of all congenital heart disease.

Constellation of- Large Ventricle Septal Defect, Right Ventricle Outlet Track obstruction (pulmonic stenosis), Aorta override, Right Ventricle Hypertrophy.

Degree of cyanosis dependent on degree of Right Ventricle Outlet Tract obstruction.

Significant RVOT obstruction presents early with cyanosis.

Mild RVOT obstruction presents later without cyanosis (pink TET) with murmur.

CXR- Boot shaped heart due to RVH.

Severe TET with Pulmonary Atresia.

Severe stenosis requires PGE to maintain pulmonary blood flow.

Initial palliation with systemic to pulmonary shunt in infant with small Pulmonary atresia/stenosis (early BT Shunt).

Primary repair in infant with well-formed pulmonary arteries (3-5 months)

Asymptomatic (pink) repaired later during the first year.

TRICUSPID ATRESIA:

Third most common form of cyanotic heart disease; 1% of all congenital heart disease.

No direct connection of the Right Atrium to Right Ventricle.

Systemic blood enters left heart via Atrial Septal Defect; the Right Ventricle is underdeveloped.

The cyanotic neonate is stabilized with PGE.

Palliation with BT shunt; later BDG (hemi-fontan) then eventually Fontan.

TRUNCUS:

Less than 1% of all congenital heart disease.

Single great artery arises from the heart supplying systemic, pulmonary and coronary circulation.

Degree of cyanosis depends on amount of pulmonary blood flow (generally increased).

**Although classically grouped with cyanotic lesions, has increased pulmonary flow and some degree of congestive heart failure.

Tachypnea, cardiomegaly, + murmur, bounding pulses (run-off to pulmonary circuit)

OR cyanotic shunting to systemic circulation secondary to respiratory illness (parallel circulation)

Early surgical intervention with Right Ventricle to Pulmonary Artery conduit.

TOTAL ANOMALOUS PULMONARY VENOUS RETURN (Obstructed):

Pulmonary veins drain into systemic venous system.

Supracardiac, cardiac, infracardiac (Superior Vena Cava, Right Atrium, portal circulation)

Systemic output is maintained by R to L shunt at the atrial level (Atrial Septal Defect)

Significant obstruction most common seen with infradiaphragmatic connection.

Presents with cyanosis and pulmonary edema.

Unobstructed veins will present later, generally first two months with tachypnea, +/- cardiomegaly and milder cyanosis (wheezing!)

Surgical repair.

CARDIOVASCULAR COLLAPSE:

The newborn with cardiovascular collapse usually presents in the first two weeks of life. This constitutes a medical emergency. Infants

discharged from the nursery prior to constriction of the ductus, without obvious signs, as perfusion and pulses were maintained. The infant is brought back as the ductus closes. History of poor feeding, tachypnea and poor color. May be in florid shock, hypotensive with absent or diminished pulses. Lethargy and decreasing level of consciousness is also present. "Ductal Dependent" PGE IS LIFE SAVING AND NECESSARY.

LESIONS PRESENTING WITH CARDIOVASCULAR COLLAPSE:

Hypoplastic Left-Heart Syndrome
Critical Aortic Stenosis
Coarctation of the Aorta and Interrupted Aortic Arch

HYPOPLASTIC LEFT HEART SYNDROME:

Most common cause of death from heart disease during the first week of life.

Diminutive Left Ventricle, critical Aortic Stenosis or Atresia, Mitral Stenosis or Atresia, hypoplastic proximal aorta.

Following ductal constriction, shock ensues rapidly due to decrease systemic flow.

Stabilization with PGE is warranted early.

Three Options: Norwood, cardiac transplant or nothing.

CRITICAL AORTIC STENOSIS:

Presents in shock secondary to compromised systemic flow, again with constriction of ductus. PGE is life saving, usually need airway support, treatment of acidosis and inotropic support. If Left Ventricle is small may need Norwood palliation. If Left Ventricle adequate, intraoperative valvotomy or balloon valvuloplasty in the catheterization lab.

COARCTATION OF THE AORTA AND INTERRUPTED AORTIC ARCH:

Coarctation accounts for 5-8% of congenital heart disease; Interrupted Aortic Arch < 1%.

Following ductus closure, severe Coarc and IAA present in shock.

Recognition of pulse discrepancy is critical to the clinical diagnosis.

Acidosis, poor perfusion is present, may have renal failure.

PGE, treat acidosis, airway management, inotropic support for severe early presentation.

Mild Coarc may present later with milder but significant shock.

CONGESTIVE HEART FAILURE:

Presents more gradually and beyond the neonatal period. Symptoms may be vague and difficult to recognize. Difficulty feeding, tachypnea, excessive sweating, irritability, pallor and poor weight gain. Cardiac murmurs are usually appreciated.

Two Main Groups:

Ventricular Failure (less common)

Excessive Pulmonary Circulation From Left-To-Right Shunting. (most common)

Lesions Presenting with Ventricular Failure:

Myocarditis

Dilated Cardiomyopathy

Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery
(seen as Pulmonary Vascular Resistance falls)

LEFT VENTRICLE FAILURE:

Typical presentation- poor feeding, pallor, tachypnea, sweating but more acute.

Cool and pale (sympathetic stimulation/compromise); may be in shock.

Liver palpable.

CXR- cardiomegaly, increased pulmonary vascular markings

Severe cases- airway management, inotropic support and afterload reduction

Milder cases- inotropic support, afterload reduction and diuresis.

Lesions Presenting with Pulmonary Overcirculation:

Moderate to Large Ventricular Septal Defect.

Atrioventricular Septal Defect

Large Patent Ductus Arteriosus

VENTRICLE SEPTAL DEFECT:

Presents with murmur and/or signs of congestive heart failure.

Natural decline in pulmonary vascular resistance may be delayed, especially large defects.

Infants with low resistance have significant L to R shunt and present in Congestive Heart Failure.

Growth failure may be present.

Prominent holosystolic murmur, diastolic rumble and hepatomegaly.

Medical management with balancing of circulation, diuresis, inotrope.

Surgical repair.

ATRIOVENTRICLE SEPTAL DEFECT:

Consists of a common Atrioventricular Valve, primum Atrial Septal Defect and Ventricle Septal Defect.

L to R shunt and Atrioventricle Valve regurgitation leads to significant Congestive Heart Failure.

Those without signs of Congestive Heart Failure will have increased Pulmonary Vascular Resistance.

40-50% of Down's Syndrome will have congenital heart disease; 40% of these will have some Atrioventricle Septal Defect accounting for nearly 85% of infants with complete Atrioventricle Septal Defect.

Medical management with balancing circulation, inotrope, diuresis.

Surgical repair.

PATENT DUCTUS ARTERIOSUS:

Present with large murmur, more subtle signs of Congestive Heart Failure.

Bounding pulses and a widened pulse pressure due to pulmonary artery runoff.

Medical management with balancing circulation, supportive care.

Surgical ligation.