CONGENITAL HEART DISEASE

CLASSIFICATION:

<table>
<thead>
<tr>
<th>CYANOTIC</th>
<th>Pulmonary plethora</th>
<th>1. TGA</th>
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<tbody>
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<td></td>
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<td>2. Truncus Arteriosus</td>
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<td>3. TAPVR</td>
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<td>4. single ventricle</td>
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<td>5. double outlet right ventricle</td>
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<td>6. hypoplastic left heart syndrome</td>
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<td>Pulmonary oligemia</td>
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<td>1. TOF</td>
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<td>2. TA</td>
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<td>3. Ebstein's anomaly</td>
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<td>4. Pulmonary atresia</td>
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<table>
<thead>
<tr>
<th>ACYANOTIC</th>
<th>Pulmonary plethora</th>
<th>L &gt; R shunts, ie</th>
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<tbody>
<tr>
<td></td>
<td></td>
<td>1. VSD</td>
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<td>2. ASD</td>
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<td>3. PDA</td>
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<td>4. AV Canal defect</td>
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<tr>
<td>Normal lung vascularity</td>
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<td>1. Coarctation</td>
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<td>2. Congenital aortic stenosis</td>
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<td>3. Pulmonary artery stenosis</td>
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ACYANOTIC CHD WITH PULMONARY PLETHORA:

- VSD
- ASD
- PDA
- AV canal defect

VSD:

- commonest intracardiac lesion in children.
- Isolated defect in about 20% of pts with CHD.
- 5% associated with other lesions particularly chromosomal abnormalities such as trisomy 13, 18, 21.
- Many small VSD's close spontaneously - anatomic classification important re expectant vs surgical management

Clinically:
- Blowing PSM at lower sternal border
- CHF, recurrent RTI, FTT
- Present after 1st month of life because pulmonary vascular resistance falls with subsequent development of interstitial oedema.
- In the older child if pulmonary vascular resistance remains high → pulm. HPT
- Rarely, pulmonary vascular obstructive disease occurs - reversal of blood flow L→R shunt occurs (Eisenmenger reaction)

**CLASSIFICATION**

**MEMBRANOUS:**
- Anterior
- Posterior
- Supracristal
- Gebode LV - RA shunt

**MUSCULAR:**
- 1/more defects
- Swiss cheese

**CXR:**
- pulm:systolic blood flow >2:1
- Biventricular enlargement
• Increased pulmonary vascularity
• Dilated pulmonary trunk
• LA enlargement with posteromedial displacement of the left mainstem bronchus
• CHF signs may appear after the 1st month of life

CARDIOMEGALY, PLETHORA, ENLARGED PULM. TRUNK

ECHO:
• LA hypertrophy
• Colour flow Doppler demonstrates flow across the defect.
PERIMEMBRANOUS DEFECT IN THE PARASTERNAL LONGAXIS PLANE

MRI:

5 YR OLD BOY: MUSCULAR VSD

ASD

• 2nd commonest cardiac anomaly
- 10% of all CHD
- Is the commonest intracardiac shunt that persists into adulthood.

**CLASSIFICATION**

OSTIUM SECUNDUM: 80-90%
Midseptum (fossa ovalis)
Usually isolated >1cm

SINUS VENOSUS: 5%
Defect in posterior wall close to SVC entry
law PAPVD

PATENT FORAMEN OVALE
Defective approximation of ostium secundum and primum

ENDOCARDIAL CUSION DEFECTS 5-10%
Low in atrial septum +/or aberrant PV, PS, Eisenmenger rx,

** Clinically:**

Pxs usually asymptomatic
Present in adolescence with:
- Mild dyspnoea
- Asymptomatic cardiac murmur
- Harsh systolic murmur along upper LSB
- Loud S2 with fixed splitting
Ostium primum ASD: MR

**CXR:**

- Normal in infancy, changes noted 1st in childhood
- RV dilation
- Pulmonary trunk enlargement
- Plethoric lung fields
- Mild rotation of the heart and great vessels to the left.
- Absent SVC contour d/t rotation over the spine
ECHO: (transthoracic with transoesophageal is diagnostic)

4 chamber subcostal view
OSTIUM PRIMUM ASD
SVC

Sinus Venosus defect with overriding SVC - rotated mediastinum, prominent RPA, absent SVC shadow
AV CANAL (ENDOCARDIAL CUSHION) DEFECTS

- 4% of all CHD
- D/t abnormal development of endocardial cushion tissue.
- Spectrum: common AV valves -> AV canal defect
- Complete AV canal defect incl. ASD, VSD which occur less frequently with common AV valves.
- Association with asplenia/ polysplenia
- Partial AV canal defect - ASD and cleft mitral septal defect.
Clinically:

- FTT
- Dyspnoea
- Fatigue
- Pulm HPT:

- more commonly in Down’s syndrome (40-50% of Down’s)

CXR:

- Indistinguishable from ASD.
- Complete defect - RA < RV enlargement

PDA

- Persistant communication between descending aorta and LPA.
- 8-10% of CHD (Increased incidence in prem babies)
- 1 in 3000 term infants.
- F:M = 2:1
- Anatomic closure in 95% of infants by the 3rd month.
- Remains open d/t:
  - low pO2 in arterial blood
  - increased fetal PG levels

- HMD in a prem baby cxs the ductus to close

Clinically:

- Usually asymptomatic.
- Range of symptoms from a machinery murmur in neonate - frank CF in infancy.

- Rx: sx clip
  indomethacin

CXR: (may be normal)

Neonate:
  - pulmonary plethora
  - LA enlargement

Infant / young child:
  - pulmonary plethora
- enlarged aortic knuckle
- LA and LV enlargement
- filling of AP window

Partial closure: ductus bump or diverticulum
Closed ductus: ligamentous arteriosum calcification

**ECHO:**
Connection between DA and LPA directly demonstrated

**OBSTRUCTIVE LESIONS:**
(acyanotic with normal lung vasculature):

- Coarctation of the aorta
- Congenital aortic stenosis
- Pulmonary artery stenosis

**ECHO**
- modality of choice and is diagnostic
- functional assessment also possible

**ANGIOCARDIOGRAPHY**
- rarely performed

**CARDIAC MRI**
• being used more often and allows excellent detailed visualisation of cardiac anomalies

**CXR:**

• Value varies tremendously.
• Sometimes possible to make a diagnosis on CXR eg. aortic coarctation in an adolescent/ adult but not in a child.
• Diagnostic value is also dependent on the type of lesion.
• Type of abnormality and haemodynamic consequences may be evident on CXR eg. PDA complicated by Eisenmenger reaction.

**Diagnostic features on CXR:**

• lung fields - normal/ increased or decreased vascularity
• size and position of the heart
• shape of the heart
• position, size and shape of the ascending aorta, aortic arch and MPA
• assoc. skeletal anomalies
• position of the viscera and main bronchi

**CYANOSIS AND Oligaemia:**

• Tetralogy of Fallot
• Tricuspid atresia
• Ebstein's anomaly

**TETRALOGY OF FALLOT**

• Commonest (12%) of cyanotic CHD.

• 4 components:
  → infundibular PS
  → RVH
  → high VSD
  → overriding of the aorta

• Is the consequence of eccentric separation of the truncus → hypoplasia of the PV, MPA, RPA and LPA
• Thought to be d/t aberrant development of the infundibulum.
If mild PS and large VSD present → pink TOF as there is sufficient pulmonary flow for oxygenation.
At the other extreme: pulmonary atresia and VSD → blue TOF.
Most infants are in the middle of the spectrum.

**Clinically:**
- Cyanosis after 3-6 months.
- Exertional dyspnoea.
- Hypoxic spells relieved by squatting.
- Squatting increases venous systemic return.
- Associated with trisomy 21, VACTERL abn., TOF

**CXR:**
- BOOT shaped heart (RVH with rotated heart and upturned apex)
- Oligaemia
- Absence of the pulmonary segment
- Reticular interstitial pattern caused by collateral flow in the upper lung fields medially
- Enlarged AA
- RT sided aortic arch in 25% of pxs
- Pink Fallot: plethora, hollow pulmonary bay
ECHO:
- Ant displacement of infundibular septum
- excl. 2nd VSD on colour doppler
- confirms large gradient over PS

SUBCOSTAL LONG AXIS VIEW:
STENOSED RVOT BY THICKENED INFUNDIBULAR SEPTUM
HIGH VSD & AORTA

MRI:
High VSD with overriding aorta
TRICUSPID ATRESIA

- 1.5% of CHD
- Infants cyanotic at birth and there is an obligatory ASD/VSD or PDA (uncommon) for survival.
- Transposition of the great vessels in 30%.
- PS in 50%.
- May be assoc with RT sided aortic arch or TAPVR
- The obligatory R → L shunt at atrial level leads to LVH.

CXR:
- Normal to diminished pulmonary blood flow.
- LVH (rounded apex)
- prominent RA contour.
- Concave pulmonary artery segment.
- May have RT sided aorta.

EBSTEIN'S MALFORMATION

- <1% of CHD
- Tricuspid valve tx “displaced” into RV → severe incompetence
- RA enlarged
- RV partially atrialised
- ASD always present
CXR Findings:

- Box shaped heart
- Oligaemia
- RA enlargement with rounding of SVC/ RA junction

Clinically:

- Palpitations, cyanosis
- Usually present in 1st month of life.
- Poorly fxning RV → flow backup → increased cyanosis because of R to L shunting via ASD.
- Severe pulmonary HPT
CYANOSIS AND PLETHORA:

- TGA
- CONGEN. CORRECTED TGA
- PERSISTENT TA
- ANOMALOUS PULM VENOUS RETURN
- DOUBLE OUTLET RV
- SINGLE VENTRICLE

TRANSPOSITION OF THE GREAT ARTERIES

- 5% OF CHD
- Aorta arises from RV and MPA from LV
- Aorta comes to lie anterior to the pulmonary artery.
- Deoxygenated blood circulates to the body and oxygenated blood circulates to the lungs.
- Incompatible with life if no ASD/ VSD/ PDA

Clinically:

- Intensely cyanotic at birth.
- Unresponsive to 100% O2 if no VSD
- Degree of pulmonary blood flow determines the degree of cyanosis
- Admixture of oxygenated and non oxygenated blood - less cyanosis
CXR:
At birth normal cardiac size, mild cardiomegaly later
- Hyperinflated and plethoric lung fields
- Lack of normal thymic outline due to stress
- Egg on the side/ apple on a string shaped heart

- Rx: J antene procedure or palliative if large VSD
CONGENITALLY CORRECTED TGA

- Occurs when there is inversion of the ventricles with normal atrial relationship.
- Aorta lies to the left of the MPA
- Coexisting lesions: VSD, single ventricle, PS, TR
- Clinical findings depend on the severity of intracardiac lesions.

CXR:
- Often normal
- Varying pulmonary vascularity but usually normal/ decreased
- Abn. bulge in left upper cardiac border and mediastinum d/t ascending aorta arising from the left side and being border forming.
PERSISTENT TRUNCUS ARTERIOSUS

- Due to developmental failure of septation of the primitive truncus arteriosus into the aorta and MPA.
- <2% of CHD; 1:10,000 live births
- There is an obligatory shunt at the ventricular level.

Collett Edward’s Classification (based on origin of MPA)

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<tr>
<th>I</th>
<th>Originate from aortic trunk</th>
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<tbody>
<tr>
<td>II</td>
<td>Origin of PA from ascending aorta</td>
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<tr>
<td>III</td>
<td>Separate orifices</td>
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<tr>
<td>IV</td>
<td>MPA originates from the descending aorta</td>
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Clinically:

- Present in early infancy with cyanosis, FTT, dyspnoea, CHF
- High association with Di George’s syndrome with TA occurring in 10% of these pts and coarctation of the aorta in 30%.

CxR:

- Mild oval cardiomegaly
- Plethora which may be asymmetrical
- 35% RT side aortic arch
- Pulmonary artery waist d/t thymic stress
Rx:
- Ratselli procedure- valved conduit between RV and PA, VSD closed.

**ANOMALOUS PULMONARY VENOUS RETURN**
- Is the return of pulmonary venous blood not to the LA.
- Total/ Partial

**TAPVR:**
- 1% of CHD
- 2X commoner in males

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<tr>
<th>Location</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Supracardiac</td>
<td>50%</td>
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<tr>
<td>Cardiac</td>
<td>30%</td>
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<tr>
<td>Infracardiac</td>
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<tr>
<td>Mixed</td>
<td>5%</td>
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Obstruction to venous return occurs

Clinically:
- **Present in the first week of life.**
- High pulmonary flow and good admixture - mild cyanosis
- More severe pulmonary venous obstruction - severe cyanosis
- A third of pts have other abnormalities eg. truncus

CXR:

With obstruction:
Congested pulmonary vessels and a normal cardiac silhouette in obstruction.

Without obstruction:
- Cardiomegaly (RV and RA enlargement)
- Enlarged pulmonary artery segment
- Increased pulmonary blood flow.
- In older pts- classic snowman/ figure 8

**TAPVR:** SNOWMAN/ FIG 8/ COTTAGE LOAF HEART-
CONVEXITY OF SUP. MED., RVH

**PAPVR**
- Only part of venous drainage is anomalous.
- Assoc with sinus venosus ASD.
- Most drain to SVC or left sided SVC.
- Best example is the scimitar syndrome

**SINGLE VENTRICLE**
- Present with early cyanosis and CHF.
- Aorta and pulmonary trunk are transposed.
- Invariably associated with asplenic syndrome.
- CXR: cardiomegaly and CHF

**DOUBLE OUTLET RV**
- Aorta and pulmonary trunk arise from RV.
- VSD almost always persists
- Great arteries may be transposed.
- PS may be present.

**HYPOPLASTIC LEFT HEART SYNDROME**
- Commonest cause of CF in the neonate.
- Presents at birth.
- Hypoplasia/ atresia of aortic and mitral valves.
• Obligatory L → R shunt at atrial level most common
• CXR: globular heart, pulmonary congestion in 24 hours

EISENMENGER REACTION
• Occurs in L → R shunts (ASD, VSD, PDA)
• Development of obliterative pulmonary arteriosclerosis
  → pulmonary vascular occlusive dx
  → progressive increase in pulmonary resistance and decrease in the L→R
  shunt until pulmonary resistance exceeds systemic resistance
  → preferential RV flow into systemic circulation
  → shunt reversal of R → L