Cyanotic Congenital HD

+ visible cyanosis
  Hb ≈ 5gms/dl circulates unbound to O_2
  + PO_2 < 85%

+ mechanism of cyanosis
  ① decrease or insufficient Blood flow to the lung
  ② Deoxygenates or desaturated Blood is pumped to the body.

+ Eisenmenger's complex → Cyanotic presentation not (cyanotic disease) Seconday to R-L shunt.

+ Classification according to pulmonary Flow

- always oligemic ↓PBF
  - critical pulmonary stenosis
  - Ebsteins anomaly
  - ToF, TA, pA — mainly
- always plethoric ↑PBF
  - TGA
  - TAPVR
  - single ventricle without PS
  - Double outlet RV, truncus arteriosus — in case of NO PS or large VSD could presented as plethoric
    → mainly plethoric unless there is PS

+ Complecation :
  ① polycythemia with anemia
  ② clotting abnormalites (hypercoagulable state)
  ③ tet spells (Hypercyanotic episode)
  ④ CNS injury (abscess, embolism)
  ⑤ increase risk of → endocarditis.
  → pulmonary HTN.

+ Cardiac Finding : (Cyanosis ... Cardial)
  ① Tachypnea without other signs of RDS
  usually Full term, normal delivery baby
  ② Good air entey (No crackles, No abnormal sounds
  ③ murmur → for example: continuous murmur of PDA
  Note → may be absent in serious Forms of cyanotic HD.

④ chest X-ray & look For:
  - heart, cardiomegaly, other sign.
  - lung: plethoric / oligomic (+ or ↑ pulmonary vascular markings)

⑤ little or No increase PO_2 with Oxygen administration.
RS ... CVS ... Cardiac cause

↳ Hyperoxic test 100% oxygen then ↓ paO_2
+ more than 150 → think about respiratory Causes, or CNS
+ less than 150 → Cardiac causes
+ less than 50 → parallel circulation like TGA
  - Mixing lesion with reduce pulmony Flow.

+ difference Between preductal (R hand) post ductal (Feet) : O_2 sat
  > 3% → abnormal
  > 10% → pathologic.

Note
- periphral cyanosis in childnen usualy normal unless, if there is sever localized with Coldness → this indicate ischemic events.
- central cyanosis may be came with periphral cyanosis

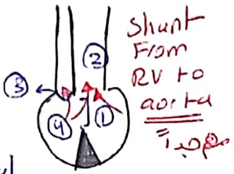
+ causes of central cyanosis :
  ① Respiratory
  ② cardiac
  ③ metabolic (acidosis)
  ④ Neurological

※ If cyanotic defect is suspected based on labrotory test, prostaglandin E_1 Sould be started or made available. → ductus arteriosus ... pulmonary circulation.

Tetralogy of Fallot (TOF)

→ mechanism (2)

- most common
- **Features :**
  1. VSD
  2. overriding aorta
  3. PS mainly infundibular (subpulmonary)
  4. RVH → secondary to obstruction



- **clinical presentation :**
  1. recurrent hypercyanotic attack (Tet's spells) → exacerbating by crying, Feeding, exertion.
  2. in First few month's of life (3-4) months of age.
  3. may be associated with DiGeorge Syndrome → so dysmorphic baby
  4. Failure to thrive → وزنه مابزيد
  5. 
     1. single S2
     2. ejection systolic murmur ← due to PS
        لما ح نسمع غير pangstolic لأن RV and LV almost equal pressure.

murmur → Flow through stenotic valve
→ shunting between 2 different gradient.

- **Diagnostic test :**
  - CXR → Boot-shape, oligemic lung.
  - ECG → RAD → lead I -ve, lead 3 +ve (تخيلها)
  - Eco → VSD, RVH

- **Complication :**
  1. cerebral thrombosis due to polycythemia / dehydration
  2. Brain abscess (N/V/ convulsion headache)
  3. Bacterial endo carditis
  4. Paroxysmal Hypercyanotic attacks → hypoxic spells
     - 1st 2 yrs of life
     - self-limiting غالبا
     - If it prolonged may lead to :
       1. LOC
       2. hemi-paresis

1. patch to main pulmonary A.
2. transannular patch → valve
3. remove some of Heart muscle → below the valve

- **managment**
  1. managment of Hypercyanotic spell
  2. surgical correction of TOF

- **Hypercyanotic spells** → usually self limiting But If prolonged need Rx → وغالباً لا يجي المريض على الطوارئ رح يكون prolonged حتى لو فيه قريب أمك استشفيه ١٠ دقايق

1. Knee-chest position
   ↓ R-L shunt, ↑ pulmonary Flow due to ↑ SVR
   also ↑ venous return to right side so ↑ RV volume so ↑ pulmonary Flow.
2. oxygen → لأنه الطفل رح يكون صار له فترة hypoxemia ويكون صار عنده acidosis
3. Bolus of Normal Saline (IV) to increase RV volume so ↑ pulmonary Flow.
4. Morphin (SC or IM) 0.1 mg / kg
   suppress RC and hyperpnea, ↓ irritability
5. Sodium bicarbonate ($NaHCO_3$)
   - 1 mEq/kg IV → can repeat in 10-15 mint
   - For Acidosis (↓ RC - stimulating effect of acidosis)
6. propranolol or esmolol (IV push)
   ↓ spasm (Relaxation of Right venticular outlet tract)
7. phenylephrine + Ketamine
   later on If patient not respond to increase ↑ systemic after load, ↑ SVR

- **surgical correction of TOF**
  1. اذا كان الرضيع Symptomatic من الولادة بعطي PGE1 ومكن نعمل وبعدين shunt (Temporary repair) لا repair عملياً لحد ما يكبر
  2. أما اذا كان الرضيع typical Form يعني انه ال (good Flow) هوه ممكن نسكت cyanosis وبعدين صار عنده ١٠٠ لحد (4-6 شهور) وزمن وقت العملية عادي I

Total surgical correction : (4-6 month of age)
1. closure of VSD with a patch
2. reconstruction of the Right venticular outflow tract (RVOT reconstruction)

Tricuspid atresia (1)

- 3rd most common
- features:-
  1. Absent TC valve
  2. RV hypoplastic
  3. VSD
  4. ASD or patent foramen ovale → must be present

* لازم يكون عنده Newborn الـ Ductus Arteriosus مفتوح عشان الـ PBF

* ممكن يكون معاه PS أو TGA in 30% of cases (high)

• Clinical presentation:

1. cyanosis → From birth, always present
2. S1 S2
   1. single S2
   2. pansystolic murmur VSD ← لو ممكن يجي ejection systolic لو كان فيه PS
   3. continuous murmur of PDA

• Diagnostic test:

CXR → ↓ pulmonary vascularity (PBF)↓ - oligemic lung

* ممكن شكل القلب تكون واضحة زي ما نشوفها normal وفيه بعض الأحيان ممكن تجي TOF زي boot shape

ECG → LAD, RA dilation — lead I, lead III

• Management:

1. PGE1
2. Surgical - single ventricular stages correction:

1- Shunt or pulmonary bandage
   ↳ in ↓ pulmonary flow
   ↳ in case of ↑ pulmonary flow

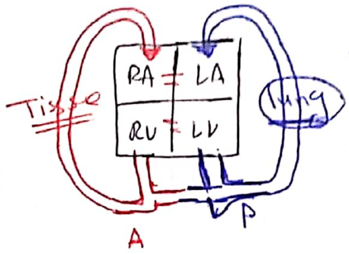
2- Bidirectional Glenn shunt
   SVC ⇌ pulmonary arteries

3- modified Fontan operation.
   RA ⇌ PA OR IVC ⇌ PA

※ Rashkind procedure (Balloon atrial septostomy)

Dextro-Transposition of Great arteries (TGA) (3)

- most common defect at birth



① الـ aorta طالعة من anterior RV والـ pulmonary artery طالع من الـ LV (2 separate circulation)

- degree of saturation will depend on the degree of mixing of the 2 parallel circuits.
- site of mixing: ASD, PDA, VSD
  ← هذولهم مبدأيًا يكونوا موجودين

هذا سلاح ذو حدين من ناحية إنه ممكن يعملنا more mixing وممكن نستغني عن الـ ASD لو كان صغير بحيث يجيبنا الـ (Rashkind procedure) مع إنها هي أفضل من الـ VSD بحيث لو كان الـ VSD كبير راح نحط Patch ممكن مع الـ complication + weakness يصير فيه ← (myocardial contractility) stenosis later on OR Residual VSD

يعني هو شي كويس يجينا مع VSD ممكن فيه بعض الحالات بساعد.

• Clinical Presentation:

1. cyanosis From birth (always present)
2. S1 S2
   - No murmur (if No VSD, small PDA)
   - Loud, single S2 (anterior aorta) ← ما يطلع الـ pulmonary
3. If CHF, hepatomegaly and dyspnea develop.

• Diagnostic test:

CXR → plethoric lung, egg-on a string, narrow mediastinum.

هذي رح تكون not oligemic وبعد ما نعطيهم PGE1 ويصير فيه PDA ← ↑ PBF وبالتالي يصير فيه (pulmonary edema)

ECG → (RVH)

Eco → anterior aorta, posterior pulmonary artery

• Laboratory studies:

1. severe arterial hypoxemia, ± acidosis
   hyperoxic test $PO_2 < 50$
2. Hypoglycemia, hypocalcaemia.

• managment :

① PGE1 infusion, oxygen, correct metabolic acidosis, hypoglycemia hypocalcemia. digoxin + diuretics in case of CHF.

② cardiac - catheterization + Balloon atrial septostomy
→ If atrial shunt is not sufficient

③ surgical : Arterial switch procedure.

arterial trunks are transected and switched to restore normal anatomy.

Coronary arteries are ~~fest~~ resected and re-implanted

→ as early as possible in First 2 weeks of life.
"anatomical + physiological correction"
(لو اتأخرنا اكتر هيكون مجرد بس physiological correction)
(left ventricle الـ Risk ان ماذا لو اتأخرت كده failure)
عضلته هتضعف وترجع aorta مش هيقدر يعمل pumping ← against high resistant
هيفشل HF

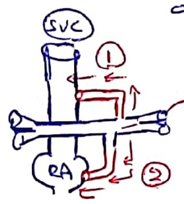
Total Anomalous pulmonary Venous Return (TAPVR)

• No direct communication exists between the pulmonary vein and the LA, instead, they drain anomalously into the systemic venous tributaries or into the RA.

• types

①- supracardiac : pulmonary veins drain into SVC

②- Cardiac : drain into right atrium direct or coronary sinus.



pulmonary vein empty in Right atrium via connective veins.

③ infra cardiac (sub diaphragmatic) → pulmonary vein drain into IVC (to portal vein, ductus venosus) hepatic vein.

④ mixed type : (combination of the other types)

* ASD or PFO necessary for survival.
• left side of heart relatively small.

• clinical precentation
• cyanosis (mild from birth)
• recurrent pneumonia, recurrent chest infection in infancy.
• sign of CHF (tachypnea, dyspnea tachycardia, hepatomegaly)
• ejection systolic murmur
• wide Fixed ~~splett~~ splitting S2
• mid-diastolic rumble.

أصلاً بتكون الـ ASD كبيرة عشان هيك بتكون الـ Systemic circulation بس بنقص الوقت نحميس (extra-Blood in RV) عشان high pulmonary Flow والـ

quadruple rhythm is present

* ملاحظة: النوع الـ infracardiac بصير منطقة فيها (Congested Venous return) بصير وبالتالي high resistant بصير الـ pulmonary edema
→ due to obstruction and congested venous return → not due to volume over load like in type 1+2
* عشان هيك هاد بسمى (Obstructive type)

وريكون منافيه pulmonary HTN ياخذ وعلى
early close to the valve so ⇒
- single S_2 or narrow splitting
- marked cyanosis + RDS in neonatal period with failure to thrive.
- Loud + gallop rhythm are present
- No murmur → ليش مرحلة الـ pulmonary HTN severe تعيق وصول عكس الـ Flow
- pulmonary crackels + hepatomegaly.

- **Diagnostic test**

CXR → Right side dilation, plethoric lung (mild in infracardiac type)
(Snow man only in supracardiac type. early before age of 4 months)

ECG → without obstruction (RBBB) RVH → rsR in V_1
→ with obstruction RVH tall R waves in Right precordial leads is present.

- **managment** → medical ← O_2, loop Diuretics For pulmonary edema, $NAHCO_3$ / Acidosis, anti-failure therapy with diuretics or/and digoxin
→ surgical

obstructive الـ يعتبر (Top emergency) بدك تعمل عملية فورية

Ebstein anomaly

- **feature**:

① downward displacement of the septal and posterior leaflets of tricuspid valve into the RV cavity ("atrialized RU")

② TR

③ ASD, PFO

④ RA dilation.

- **clinical presentation**

① cyanosis → severe case with CHF of first few days of life
- 50% mortality rate
- RV very small, need to shunt
- cyanosis due to ① PBF↓ ② TR → increase RAP increase R→L shunt

② milder case → dyspnea, Fatigue, mild cyanosis, palpitation

③ wide split S_2

④ Hepatomegaly due to RHF وممكن تكون LL edema الـ ascitis

⑤ If Right side dilatation → triple or quadruple rhythm.

- **Diagnostic test**

CXR → mild case → normal
Severe → Box-shaped or Balloon-shaped Heart
↓ pulmonary vascular marking

ECG → - RBBB / RA dilation
- First degree AV block
- WPW syndrome (20%)
- SVT episode

- **managment**:

بعطيه PGE2 لحد ما أقلل الـ PVR ويتحسن بعدها بيرجعه ويصير يعمل

Follow up → medical anti-Failure
→ Surgical

* if cyanosis at birth give PGE_1 في الحالات الـ severe

Truncus Arteriosus (rare)

Features →

① Single arterial trunk with a truncal valve leaves the heart and give rise to the pulmonary, systemic, and coronary circulation.

② large VSD

③ DiGeorge syndrome with hypocalcemia 33%.

Clinical presentation

- variable presentation according to pulmonary artery anatomy
  - ↳ if small pulmonary artery → oligemic lung with cyanosis
  - ↳ If good size pulmonary artery → plethoric lung with sign of CHF with tachypnea + dyspnea.

- Single S_2
- wide pulse pressure.
- Truncal valve regurgitation
- continous murmur →

  diastolic , systolic يعني
  murmur

- **Diagnostic test**

  CXR → variable depend on Type TA
  Cardiomegaly in ↑ pulmonary vascularity

  ECG → Biventricular hypertrophy BVH

- **management** :

  in ↑ PBF → anti-failure (Lasix, loop diuretic)

  in ↓ PBF → PGE_1

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other cyanotic disease

- Hypoplastic left heart syndrome
- DORV, single ventricle PA, critical pulmonary stenosis.