Clinical congenital heart disease

Fb/Nurse-Info

Some clinical aspects

"paediatric and adolescent accent"

- Classification (modified for simplicity)

- **cyanotic** - with ↑ pulm blood flow
  - with ↓ pulm blood flow
  - unclassifyiable – *ebsteins/ TGA IVS*
- **acyanotic** – largely shunt lesions
- **stenotic** - *outflow & arterial obstructions*

Cyanosis

caused by > 5gm/dl reduced Hb

- Clinical detection depends on
  - % arterial blood that is desaturated
  - Hb Concentration !!

- If art O2 satn is 60%,
  cyanosis is detectable if Hb > 12.5gm/dl !
  but not if Hb < 10 gm/dl !
  ie 4gm/dl insufficient for detection of cyanosis !

Detection of cyanosis

- Astute physician/ paed cardiologist
  detects when reduced Hb 3 gm/dl
  Others detect at 5gm/dl

- *Better to overdiagnose than underdiagnose* !

- Clinical diagnosis of cyanosis is inaccurate

M Tynan in Andersons Paediatric cardiology 2007




Cyanosis -some aspects

- Some CCHD with Rt to Lt shunt and ↑ P B flow

  UO TAPVR/ Truncus/ TGA-VSD/ Single ventr Physiol etc

  - may have low saturations
  - but undetectable cyanosis clinically

  i.e. 88-92% !!

- Polycythemic patients appear cyanosed
- Methhaemoglobinaemia !!

Hyperoxic test
cyanosed or not

- Pulse oximeter - not always reliable

  "a random number generator"

- Rt radial ABG ***in air*** and ***after 5-10 min O2***

  paO2 > 250mmHg -excludes CCHD

  paO2 > 160 -CCHD unlikely

  (UO TAPVR False negative !)

  paO2 < 100 -CCHD likely *(usually lower)*

  (severe Lung disease *(high paCo2)*, PPHN/PFC)

- "Radial ABG more useful than ECG or CXR in detection of cyanotic heart disease"

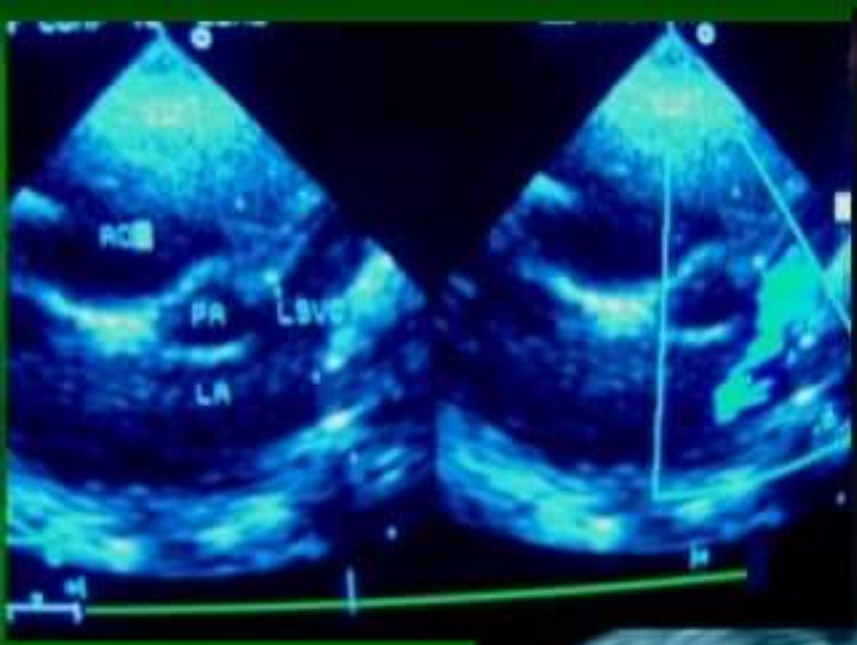
Warburton 1981

C C H D
in 3 major circumstances

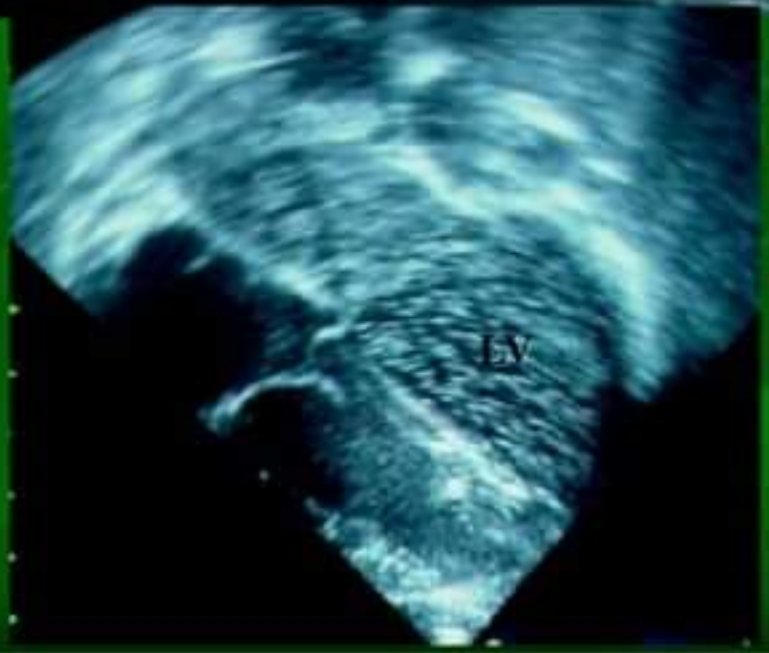
- Pulmonary obstruction with avenue for right to left shunting
- Discordant AV connection *i.e transpositions*
- Common mixing situations *i.e common atrium single ventricle etc*

Unusual causes of cyanosis ***without murmurs !*** surviving to adolesc./ adult life

- Left SVC to LA
- IVC to LA
- Rt. SVC to LA
- Pulm. AV Fistulae *(Ostler Rendu Weber syndr)*


PA
LA


AO
LV
LAVV
LA
CS


LV

Cyanosis – which category?

- Symptomatology
- Clinical examination
- Chest X ray

Fallot physiology

- Systemic venous return unable to reach lungs
- Shunted right to left away from pulm circulation
- ASD/VSD essential for this to occur;
- Or a common chamber !

PLUS

Fallot physiology

- ***Obstruction at***

  - RA outlet - *i.e Tric atresia*

  - infund/valvar Pulm stenosis

  - rarely branch PA stenosis/ DCRV

  - High PVR – *Eisenmenger !*

    obstructed pulm arterioles !!

CCHD with ↓ pulm blood flow

pulmonary oligaemia on CXR

- Symptomatology
- Inspection findings
- Auscultatory findings
- Chest Skiagram

CCHD with ↓ PBF - symptoms

- Exertional dyspnoea
- Cyanosis, spells, seizures
- CNS complications

- No recurrent RTI/ no diaphoresis
- No breathlessness at rest

except in extremes / anaemia

CCHD - ↓ PBF
- inspection /palpatory findings

- Cyanosis & clubbing
- polycythemia
- Quiet precordium to inspection & palpation
- No Harrisons sulcus or precordial bulge
- Apex well within limits if visible

- No palpable sounds or thrills

CCHD with ↓ PB Flow
auscultatory findings

- Normal first heart sound
- Single second heart sound
- Pulm component inaudible

- Stenotic pulmonary murmur
  - slightly after S1
  - stops short of S2
- Other murmurs – ductal/ MAPCA/ AR

Ejection murmur in Fallot physiology

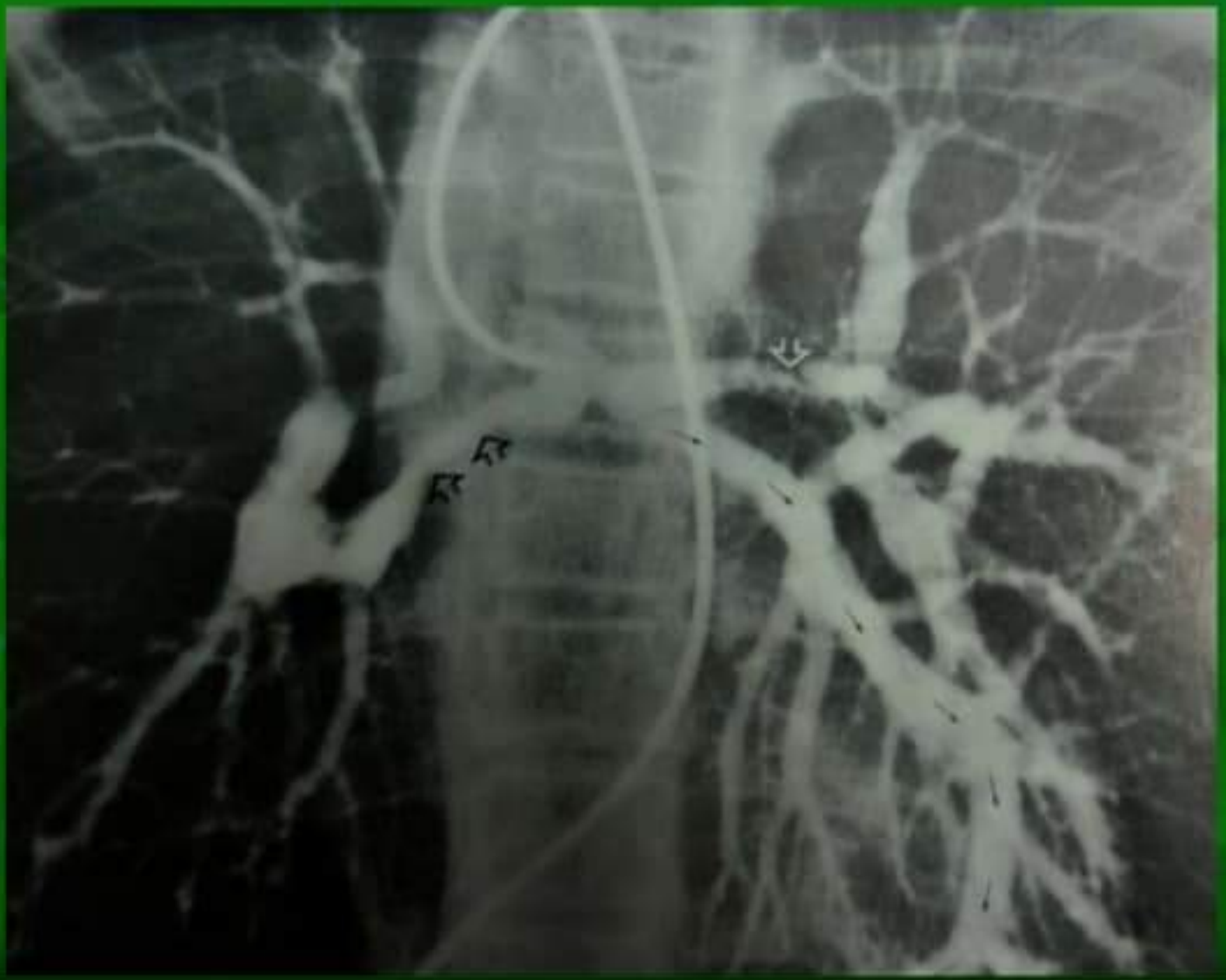
- *Length & loudness inversely proportional to severity of stenosis*

  In isolated PVS – the opposite !

- Absent murmur – *acquired pulm atresia*
  - *MAPCA murmur over back*
  - *soft ductal murmur (tortuous)*

- **To & Fro** – *Aortic regurg / Abs PV syndrome*

MAPCAS



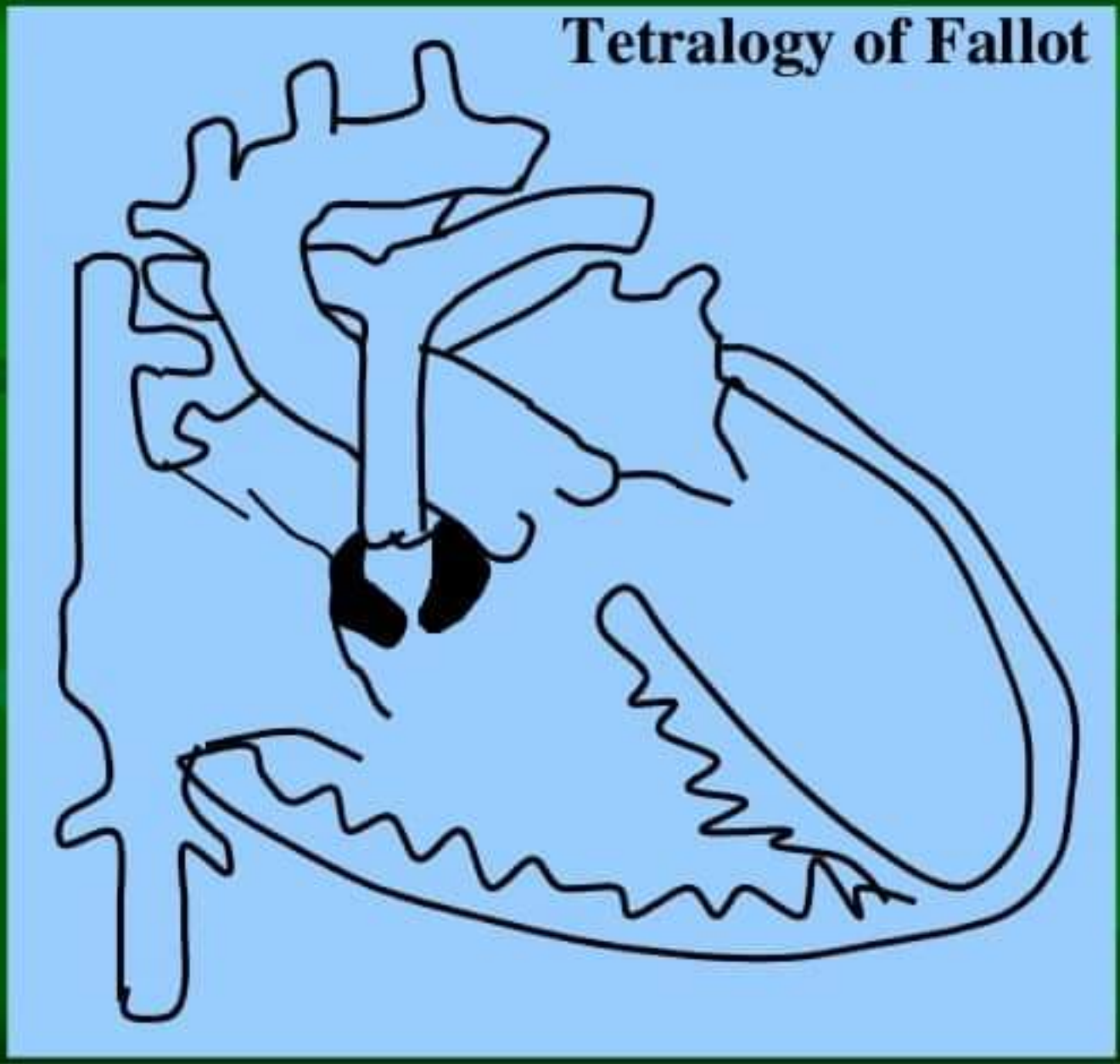
CCHD with ↓ Pulm.blood flow

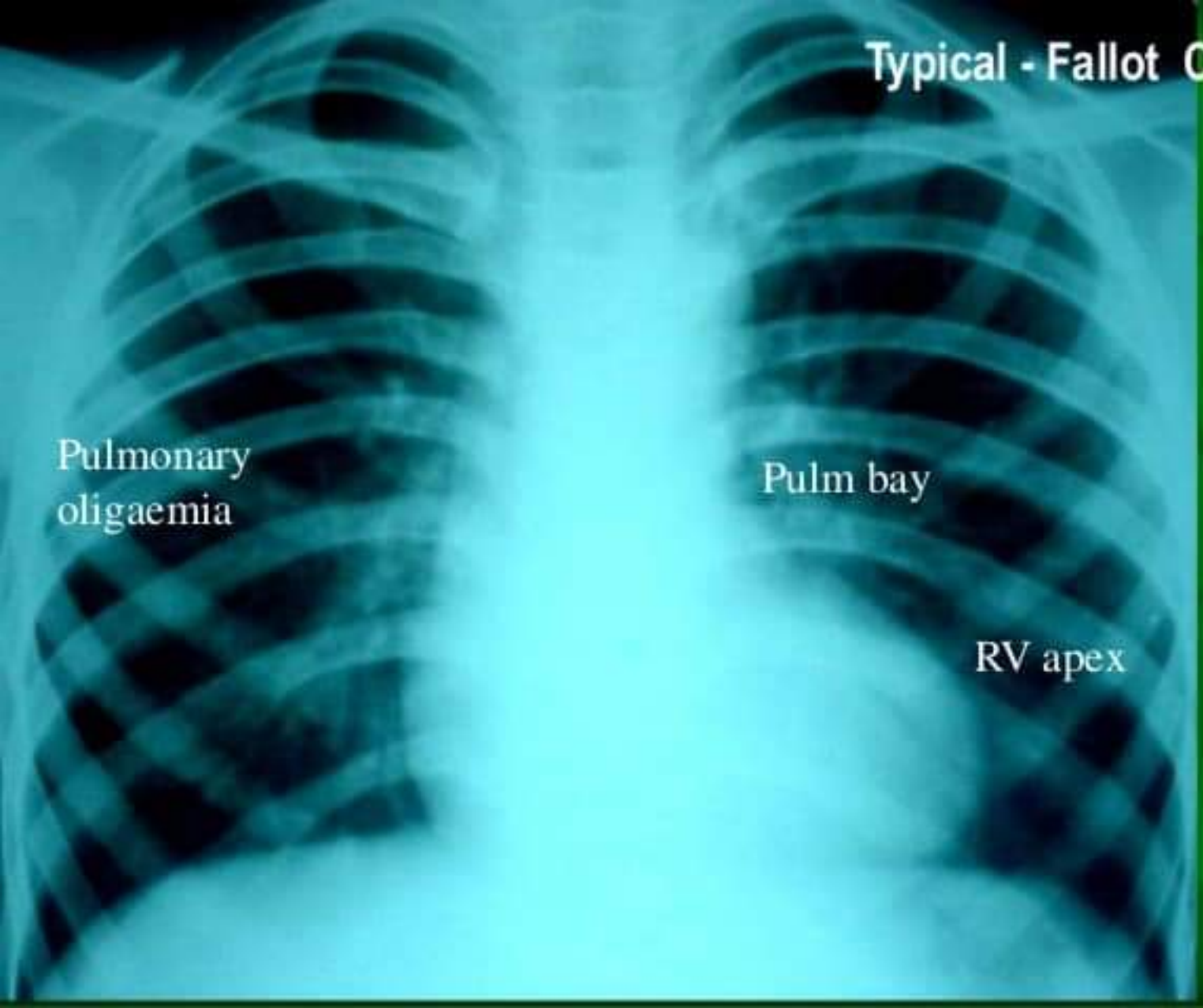
- Tetralogy of Fallot
- VSD - PS
- DORV – VSD – PS
- Tricusp. atresia - PS
- Single ventricle - PS
- TGA with VSD – PS
- Corr.transp.-VSD-PS
- ASD - PS

Chest skiagram in CCHD with ↓ PBF

- Small heart
- Pulmonary bay
- Pulmonary oligaemia
- Right aortic arch/ RA enlargement/ differential vascularity/ narrow pedicle in various defects

Tetralogy of Fallot




Typical - Fallot CXR
Pulmonary oligaemia
Pulm bay
RV apex

CNS complications of CCHD with ↓ PBF

- Paradoxic embolus
- Cerebral thrombosis
- Cerebral abcess
- Seizures
- Hypoxic damage
- Endocarditis & vegetations
- Postoperative strokes

CCHD with ↑ pulm blood flow

- **Transpositions** with VSD/Duct/ASD

- **Common mixing** situations

  Mixing at
  - atrial level – *TAPVR/Comm Atr*
  - ventric level – *DORV/Single ventric*
  - arterial level – *comm art trunk*

- *Mild cyanosis, CCF, resp symptoms, ex dyspnoea*

CCHD with ↑ Pulm blood flow

- Seldom survive to adolescence/ adulthood
- UO TAPVR/ comm atrium- the exceptions

- Most have Eisenmenger by then
  and those features dominate

CCHD ↑ P B Flow
easy diagnosis – rare

- **Clinical differentiation not always possible**

(Tynan M, Andersons paed cardiology 2007)

- Brisk pulses, ej click, to& fro murmur – Truncus
- Sm. pulses, RV impulse, wide split S2,TV MDM – TAPVR
- AV regurg murmur, wide split, TV MDM – comm. atrium
- Sing S2, cont murmur over back – p atr / MAPCAS

CCHD with ↑ P B Flow - **symptoms**

- Respiratory symptoms predominate
- Growth retarded – weight & height
- Scrawny, sick, dyspnoeic patient
- Recurrent LRTI/Pneumonias
- Chronic lung disease- bronchiectasis etc
- Diaphoresis/ breathlessness at rest
- Exertional dyspnoea, limited activity.

CCHD with ↑ P B Flow
inspection findings

- Sickly underweight individual
- Cyanosis & clubbing -mild to moderate
- Severe PHT, Eisenmenger – *modifies findings*
- Harrisson's sulcus, precordial bulge
  Active precordium, RV, LV, PA pulsations
  Obvious cardiomegaly

CCHD with ↑ P B Flow
palpatory findings

- Active precordium
- RV impulse – DORV, TAPVR, TGA VSD PS
- LV Impulse – Single ventricle, AVSD-AV regurg
- Palpable second sound / Thrills rare

CCHD with ↑ P B Flow
auscultatory findings

- Single second heart sound
- Loud pulm component, if heard
- Ejection click – pulmonary/ truncal

CCHD with ↑ P B Flow
auscultatory findings -2

- Pulm flow – ejection murmur
- MD murmur - *if no severe PHT/ Eisenmenger*
- PR/ TR murmurs may dominate
- To & fro murmurs in- *Truncus/ abs PV syndr.*
- MR murmur *in complex AVSD /comm Atrium*

CCHD with ↑ P B Flow
radiographic findings

- Cardiomegaly *(unless sev. PHT/Eisenmenger)*
- Dilated PA
- Pulmonary plethora
- Atrial enlargement
- RV/LV/ Biventric. -*Depends on anatomy/age*

Keys to clinical diagnosis

- Work in order
- Pulses,pulses, pulses
- Colour ie. Cyanosis, pallor, polycythemia
- Inspect – for chest form, pulsations
- Palpate to determine – which ventricle ?
- *Forget the murmur !!*
- Listen first to S1, and then to S2
- Can you split the second sound ??
- Then concentrate on the components
- *Finally the murmurs* – systolic – ejection or pansyst.
- Is there a diastolic murmur

The second heart sound the key to diagnosis of CHD

- Single
- Normal split
- Wide variable split
- Wide fixed split
- Reverse split
- Loud A2
- Loud P2