Septal defects with severe PHT: When is it too late to close them?

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Septal defects with severe PHT

• Rare in the developed world

 Not an uncommon problem in the developing countries

0.8% of all the out patients – Eisenmenger's syndrome

Severe PHT: A spectrum

• With normal or near normal PVR

• With elevated PVR which is reversible

• With elevated PVR which is irreversible

Two basic things

- PHT is not synonymous with PVR
 - 6 month old with a large PMVSD and systemic PA pressure
 - 16 year old with a large PMVSD and systemic PA pressure
- High PVR is not necessarily irreversible

When is it too late?

• Severe PHT with irreversibly high PVR

• Closing the defect – COUNTERPRODUCTIVE

• Severe PHT with normal/reversibly high PVR

 Closing the defect – Significant benefit in terms of QOL as well as longivity

Million Dollar question

• How to clinically or otherwise recognize

• Irrevrsible PVR

• Normal/reversible PVR

Holistic Approach

- Clinical
- ECG
- CxR
- 2DE/CD
- Cardiac catheterization
 - Basal
 - After O2 and pulmonary vasodilators
 - Temporary (balloon) occlusion of the defect

Hemodynamic point of view

- Pretricuspid shunts: Predominantly diastolic
 - Shunt reversal may not occur despite high PVR till such time RV diastolic function is normal
 - Shunt reversal with normal or mildly elevated PVR due to associated TR

 Post tricuspid shunts: Predominantly systolic or continuous

Clinical Evaluation

- Symptoms: Characterized by fixed output state and hypoxia
 - SOB
 - Easy fatigability
 - Syncope
 - Claudication
 - Hemoptysis

Clinical Evaluation

- Signs:
 - Cyanosis \pm clubbing
 - Small heart (except valve regurgitation)
 - RV apex
 - Parasternal heave
 - EC
 - Narrow splitting of A2P2 with loud P2
 - Short/absent murmurs
 - No flow murmurs

Post exercise SaO2/PO2

- UL for ASD/VSD
- LL for PDA
- Avoid in the presence of H/O post exertional syncope

Clinical Evaluation

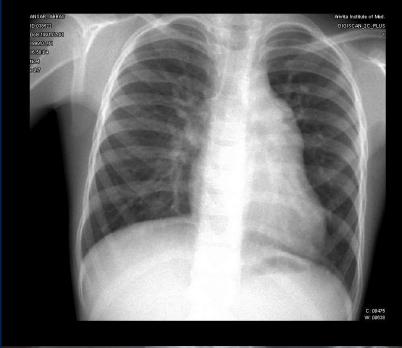
- Age
- Presence of Down's syndrome
- Born at high altitude
- Unilateral pulmonary artery stenosis/absence



X-ray chest

- Normal sized heart
- Large central PAs
- Presence of peripheral pruning







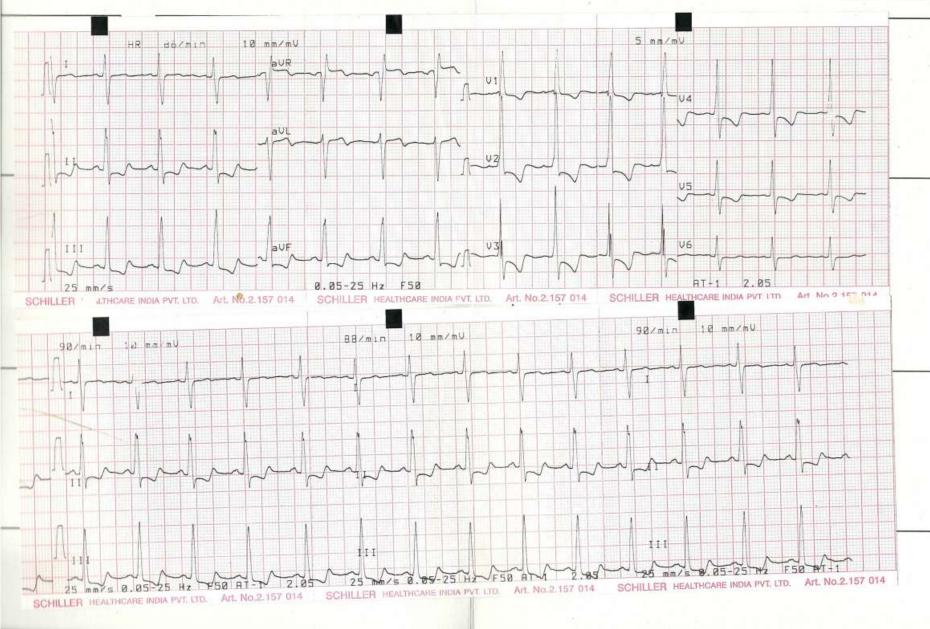
ECG

- Right axis deviation
- RVH
- Absence of LV forces

Rashme Parmar

04/08/10

"Please obtain pho copy of this ECG record fades with



2DE/CD

- RA and RV are pressure overloaded
- PA pressure is systemic or more
- Flow becomes bidirectional
- TR and PR develop
- Estimates of PA systolic and mean pressure





Recommendation for Cardiac Cath

CLASS I

 Cardiac catheterization to assess the operability of adults with intra/extracardiac shunt and PAH (*Level of Evidence: C*)

Cardiac Catheterization – Is it a Gold Standard?

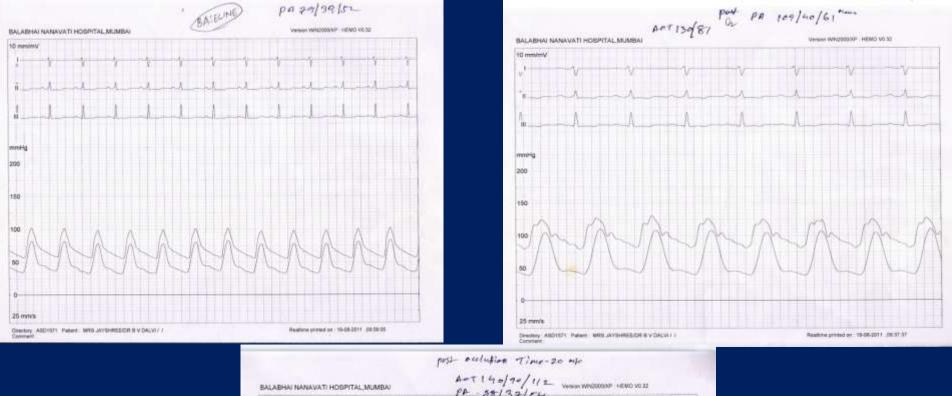
- O2 consumption: **ASSUMED**
- AV Oxygen difference: Dissolved oxygen is **ASSUMED** to be zero. Not true for patient breathing O2
- Mixed venous blood is ASSUMED to be in various combinations of SVC and IVC
- Poiseuille-Hagen equation for PVR **ASSUMES**
 - Steady state fluid
 - Neutonian fluid
 - Rigid system

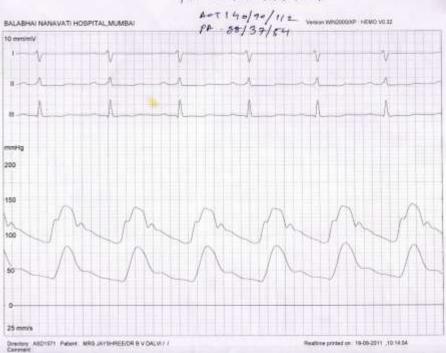
Sources of error

- Recording pressures: Balancing, zeroing, transducer frequency response
- PO2 used to calculate SaO2 rather than direct measurement of SaO2

Cath Numbers

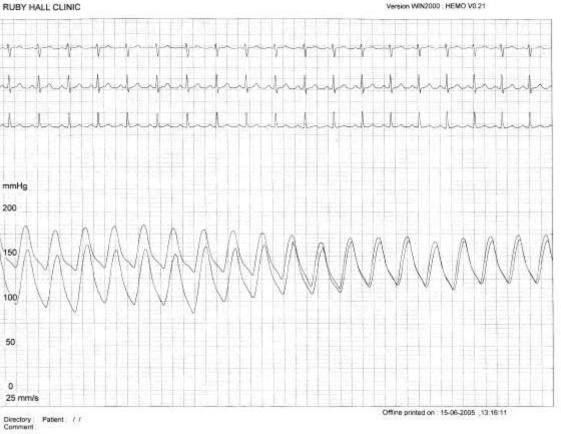
- PVR estimation on room air, with 100% O2 and with NO
 - PVRI > 8 Wood units. m²
 - PVR/SVR > 0.66
- Balloon occlusion on catherterization table : < 25% drop in PAP (ASD)
 No fall or increase (PDA)







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Conclusion

- It is difficult to judge with one single parameter whether it is too late
- One needs to take a holistic view and multidimensional approach
- Counseling the family appropriately is very essential

In Jurisprudence

"Justice delayed is

justice denied"

"Treatment delayed is treatment denied"