Hemodynamic assessment prior to closure of heart defects in presence of pulmonary hypertension

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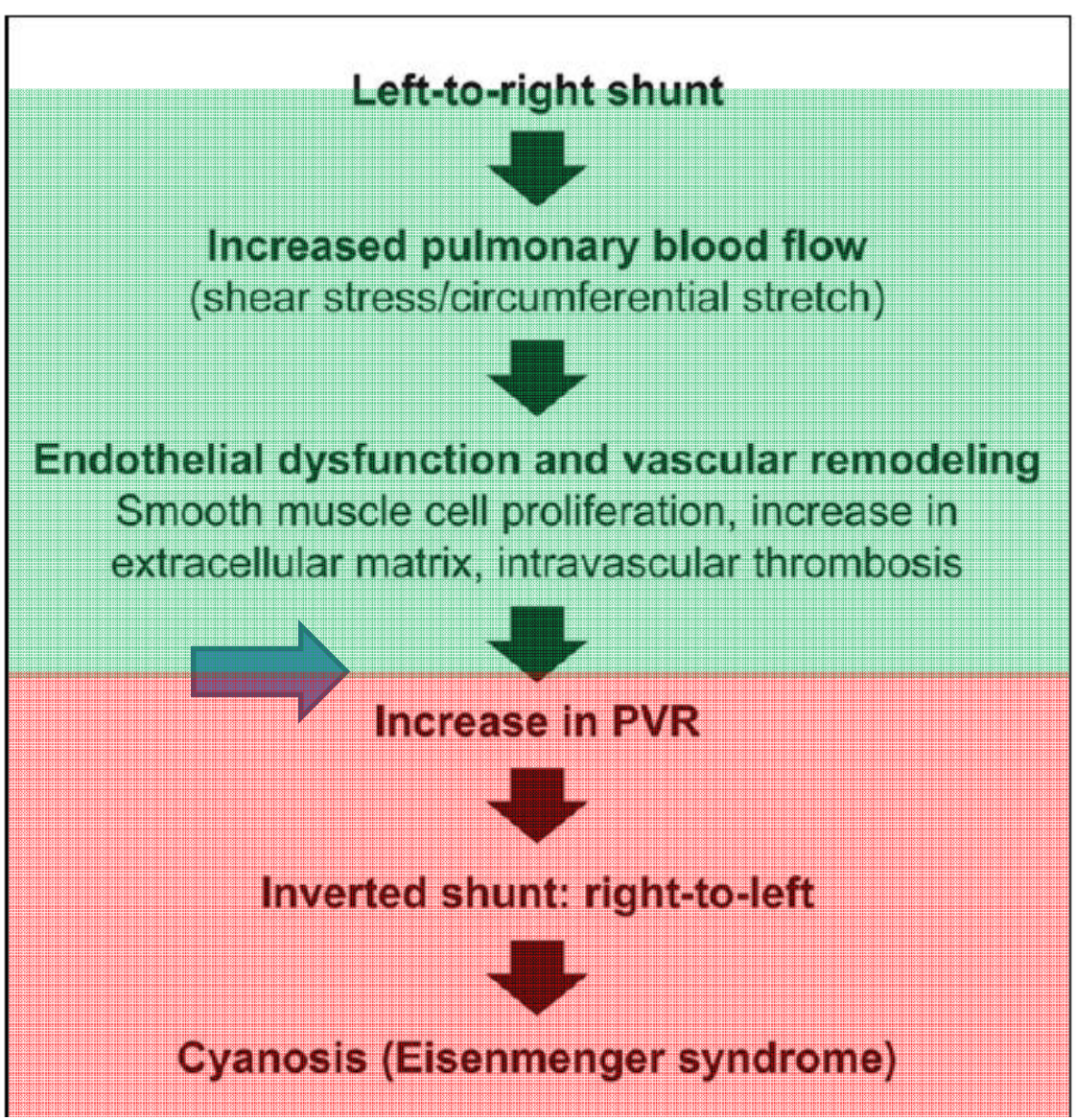
Hemodynamic assessment in heart defects with pulmonary hypertension?

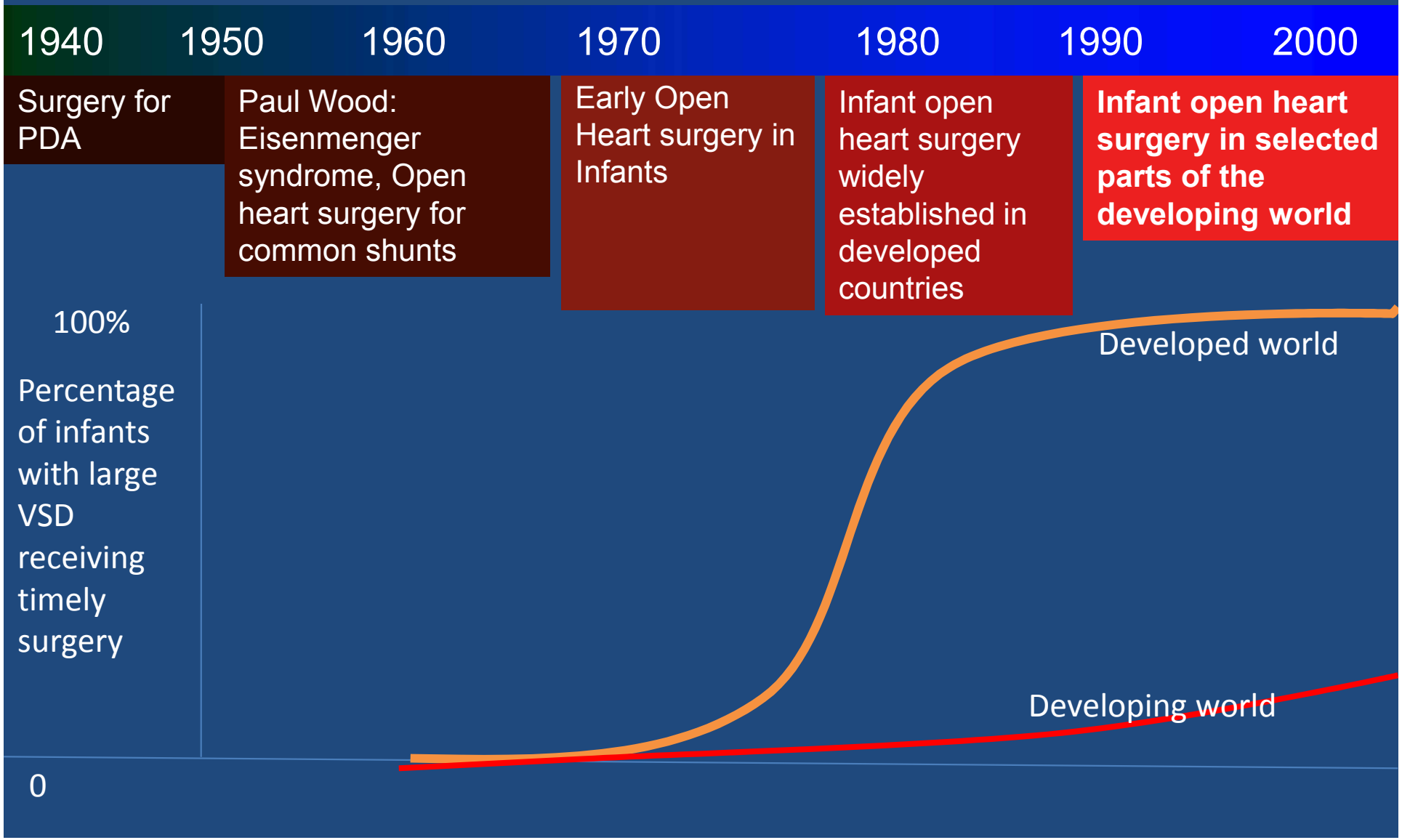
1. Why worry about pulmonary hypertension?
2. How do we decide on surgical closure of heart defects in presence of pulmonary hypertension?
3. Should we do anything different for catheter closure of heart defects?
4. Should we do anything different after the availability of newer therapy for pulmonary hypertension?

Congenital Heart Disease (L-R shunts) and Pulmonary Hypertension



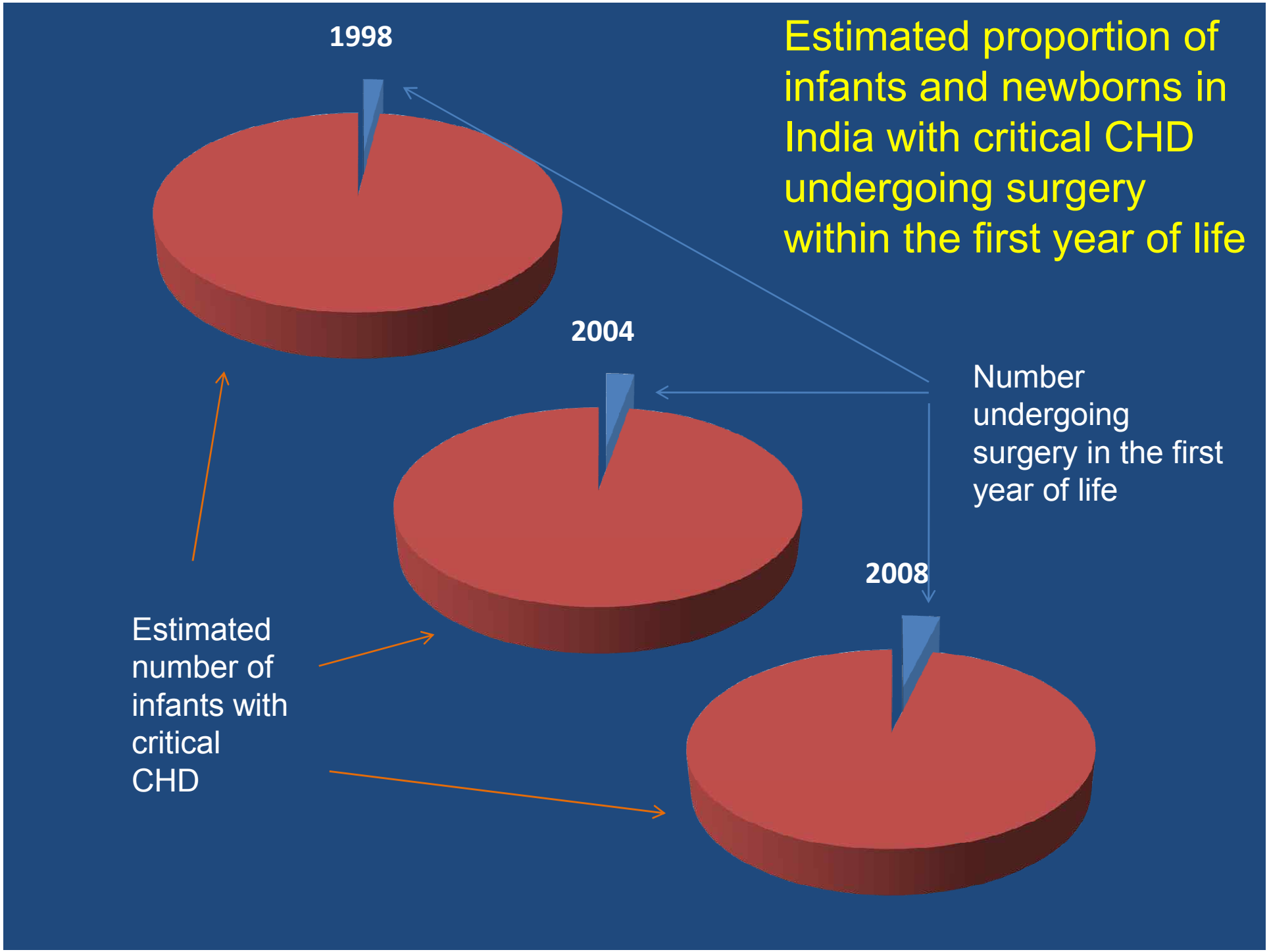
Maurice Beghetti, and Nazzareno Gali, *J. Am. Coll. Cardiol. 2009;53;733-740*

Congenital Heart Disease



Operations for CHD (Global Situation): Data from Year 2000

- Total Open Heart Procedures: 1,211,624
- Total Congenital operations: 159,482
- Adult Congenital: 25,556
- Pediatric Congenital: 81,506
- Infant/Newborn: 52,419


Estimated proportion of infants and newborns in India with critical CHD undergoing surgery within the first year of life
1998
2004
2008
Number undergoing surgery in the first year of life
Estimated number of infants with critical CHD

Parts of The World Where the Average Child in the Region has Access to Congenital Heart Surgery

Late Diagnosis of CHD: Consequences

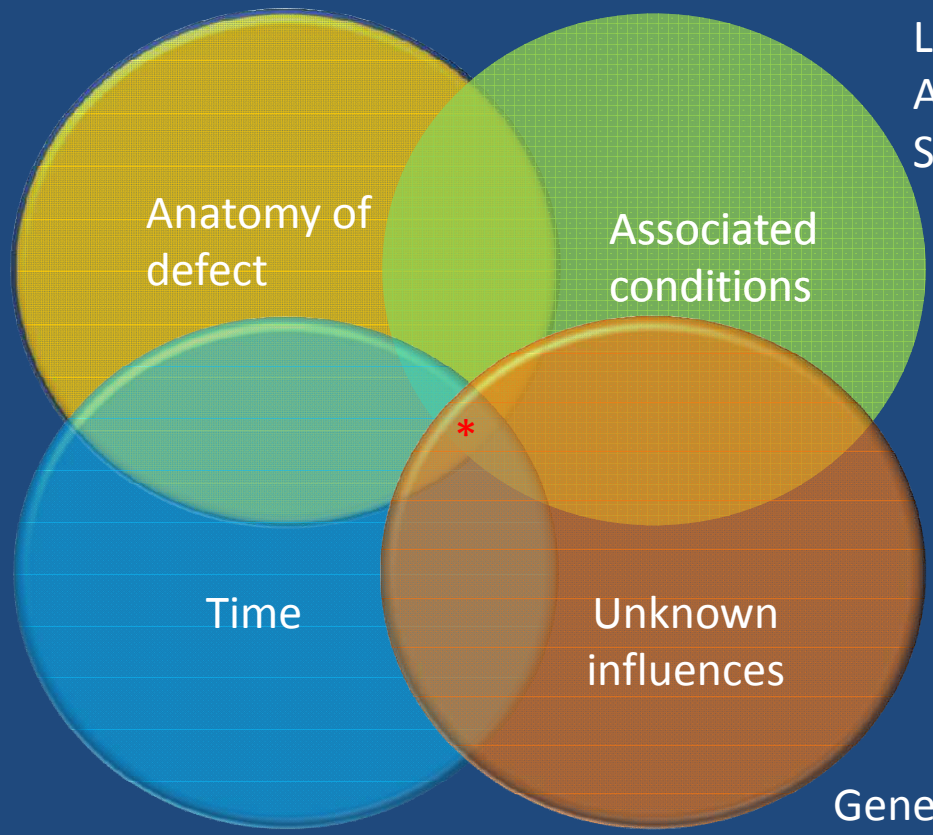
- Survivors selected by natural history
- Shunt lesions: Most children with large shunts with ↑ Qp would not survive beyond 2 years age
- Relatively large proportion of ↑ PVR among survivors

Why worry about pulmonary hypertension in CHD?

- Most of the world has limited access to congenital heart surgery
- ASD can present late anywhere
- An occasional VSD or PDA escape detection may present later in childhood
- Insights into mechanisms of development and progression of pulmonary hypertension

What determines the development of pulmonary vascular obstructive disease (PVOD)?

Pre vs. post tricuspid
Size
Associated lesions: pulmonary venous hypertension



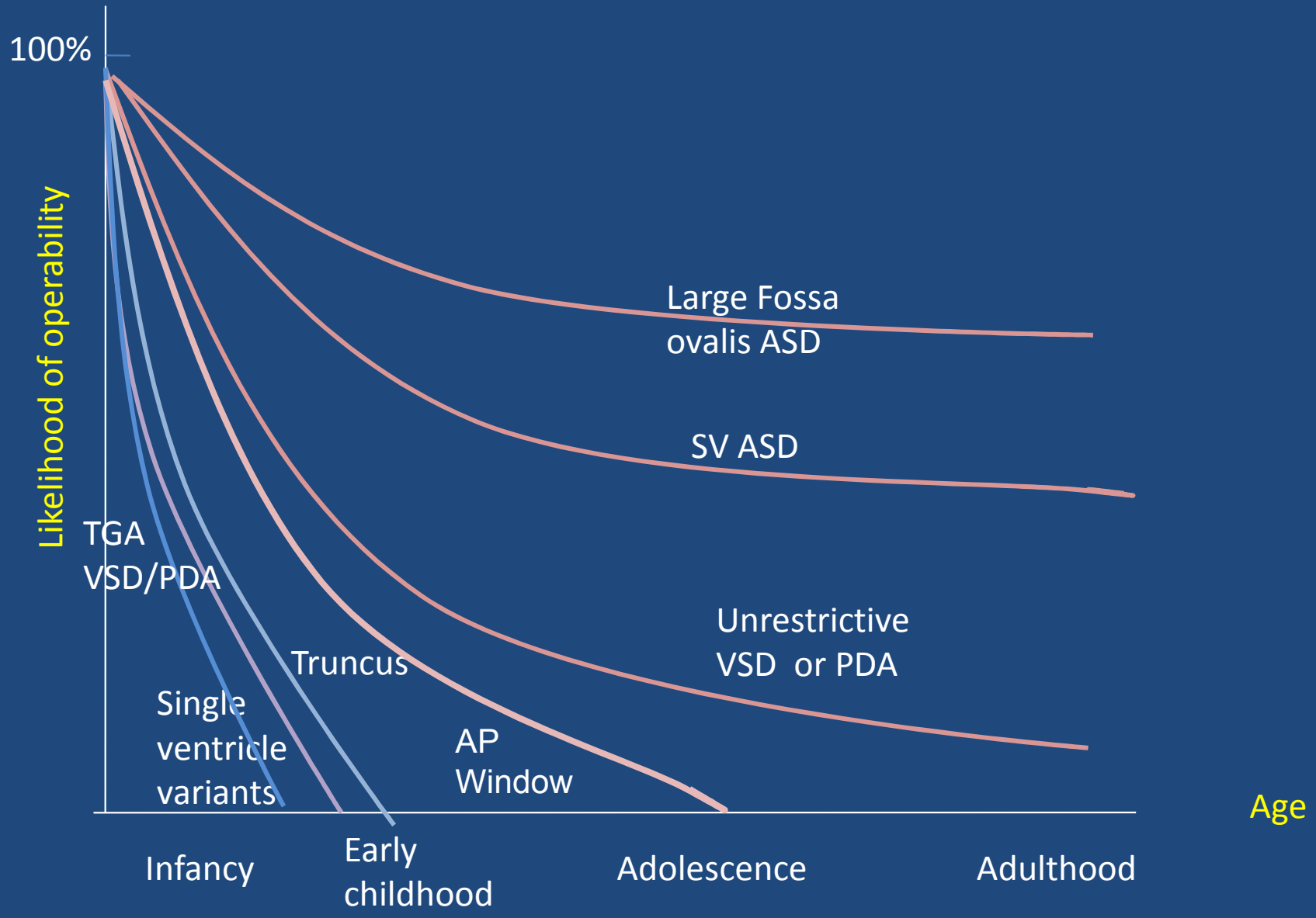
Lungs and airways
Altitude
Syndromes: Tri-21

Genetic???

Determinants of development of PVOD in L-R shunts

- Pre-tricuspid shunts:
  - gradual ↑in Qp as RV accommodates and enlarges
  - ASD, PAPVC
- Post tricuspid shunts: Direct transmission of pressure head:
  - VSD (systolic),
  - PDA, AP-Window (systolic and diastolic)

Concepts: Defect vs. PVOD Risk



Risk of development of PVOD: Other (unknown) influences

Remarkable individual variability

- ASD with PAH in an infant
- VSD with shunt reversal in an infant
- Operable AP window in a teenager
- Operable large VSD in an adult

Prediction for an individual patient is sometimes quite challenging

Case example

- 5 month old, first seen in 2002, large fossa ovalis ASD, pulmonary hypertension, RV systolic pressure 54 + RA pressure, L-R shunt (vigorous)
- Symptomatic, tachypnea, failure to thrive, heart failure
- Catheterized to “understand the hemodynamics better”

Case example….

HEMODYNAMIC DATA			
Condition 1 :	☒ Breathing Room air	☐ O2 Inhalation (FIO2 =)	☐ Ventilation, (FIO2 =)

A) Oxymetry and pressure data were recorded for SVC, RA, PA, LV, AO, PV, in

Basal state	and	**After administration of O2** at 10L/min for 10 minutes which revealed
• Large L->R Shunt		Large L->R Shunt
• QP/QS::3.1>1		QP/QS:>4
• PVRI:2.3		PVRI:0.29

B) Elevated right heart filling pressure and Moderate PAH.

C) Angios:

LUPV Injection PA view normal drainage to LA

PA angiogram PA view showed confluent PA anatomy,RPA:6.8MM,LPA:5.7MM,normal arborization,no evidence of pulmonary stenosis.

LV angio LAO,Cran. View showed normal LV,normal coronaries,and arch vessels.

PLAN

Atrial Septal Defect Closure

Operated in the same admission with uneventful post-operative course

Seven years later…

Case Example #2

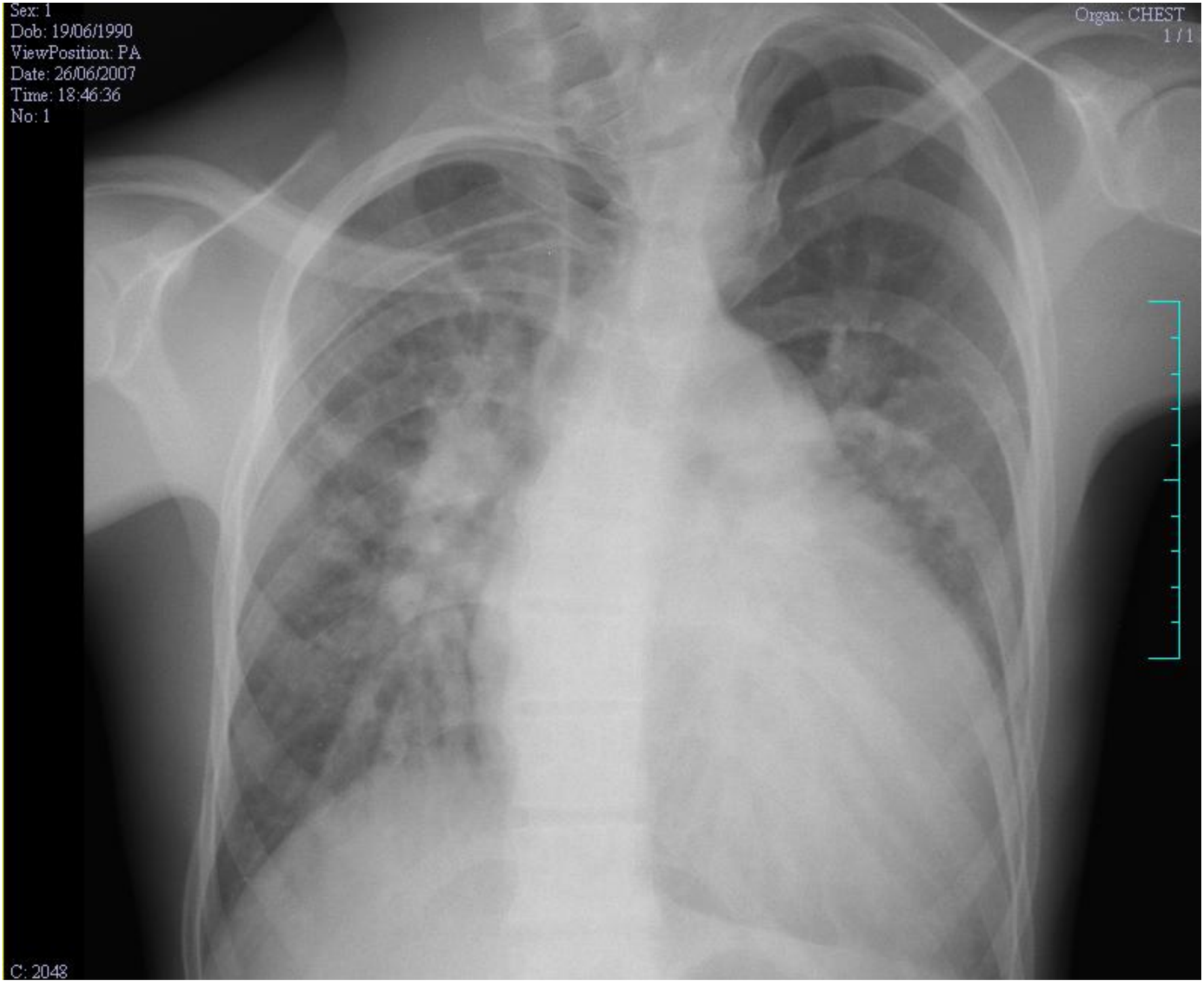
- 17 year old
- Detected to have CHD in the early infancy. Cardiac cath planned but could not be accomplished.
- Relatively “asymptomatic”
- Occasional episodes of respiratory infection+.
- AV malformation of calf, heart disease needed to be fixed before vascular surgery.

Case #2: Physical Exam

- HR: 110/min, RR: 32/min.
- SpO2 on room air 97%.
- CVS: S1 normal, S2 normally split with loud P2, grade 5/6 Pansystolic Murmur at present at LLSB with a clear mid diastolic murmur at left 4th ICS.

Case #2: Echo

- Large inlet VSD (2 cm), gradient of 35 mm Hg
- Predominant L-R flows
- Grade I straddling of tricuspid valve
- LA and LV enlargement
- Flow acceleration in the pulmonary valve (gradient of 30 mm Hg)


Sex: 1
Dob: 19/06/1990
ViewPosition: PA
Date: 26/06/2007
Time: 18:46:36
No: 1
Organ: CHEST
1 / 1
C: 2048

Case #2: PFT

Parameter	UM	Description	Pred.	SD	TEST#1	%Pred.
Best FVC	l(btps)	Best Forced Vital Capacity	4.59	0.56	2.18	47.4
Best FEV1	l(btps)	Best Forced Exp Volume in 1 sec	3.95	0.47	1.85	46.7
FVC	l(btps)	Forced Vital Capacity	4.59	0.56	2.18	47.4
FEV1	l(btps)	Forced Exp Volume in 1 sec	3.95	0.47	1.85	46.7
PEF	l/sec	Peak Expiratory Flow	9.03	1.11	7.02	77.7
PIF	l/sec	Peak Inspiratory Flow			3.40	
FEV1/FVC%	%	FEV1 as % of FVC	84.0	7.2	84.9	101.1
FEF25-75%	l/sec	Forced mid-expiratory flow	4.81	0.96	2.64	55.0
MEF75%	l/sec	Max Exp Flow @ 25% FVC	7.63	1.57	6.61	86.7
MEF50%	l/sec	Max Exp Flow @ 50% FVC	5.09	1.21	3.14	61.7
MEF25%	l/sec	Max Exp Flow @ 75% FVC	2.42	0.72	1.27	52.7
FET100%	sec	Forced Expiratory Time			3.3	
IC	l(btps)	Inspiratory Capacity			1.13	

Restrictive physiology

Case #2: Hemodynamic data

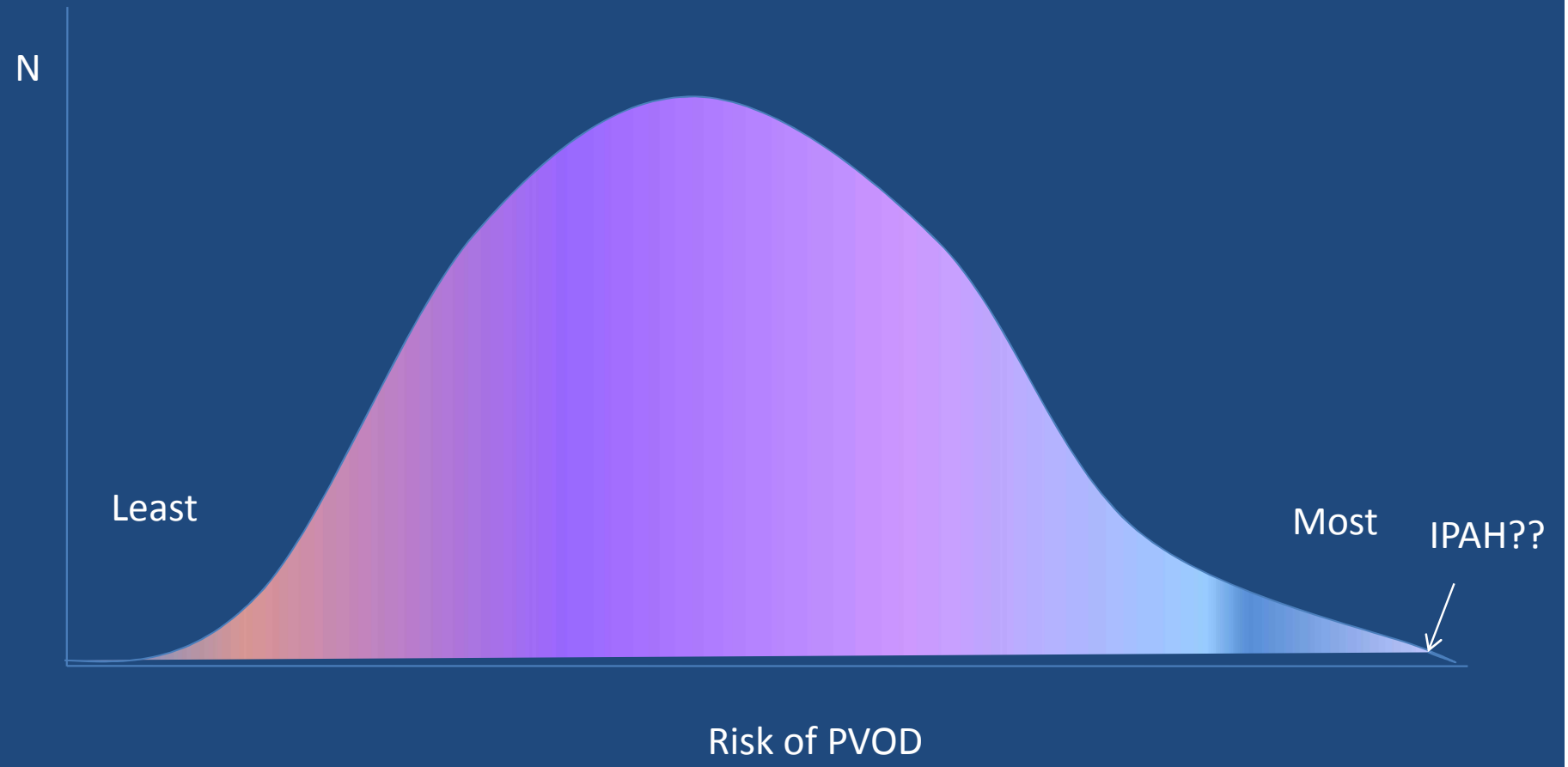
Chamber	Sys(v)	Dias(a)	Mean	O2 %
SVC				66.3%
RA	11	12	8	
PA	85	29	51	87.8%
PA wedge				
PV				98 (a)
RV	75	ED= 10		
Ao	110	60	88	97.5%

Case #2: Hemodynamic data

Parameter	Condition 1 (room air)	Condition 2 (FiO2 100)	Condition 3 (NO 40PPM + O2)
O2 Consumption	135	143	143
Qp	7.54	20.22	19.13
Qs	2.47	3.54	5.39
Qp/Qs	3.06	5.72	3.55
PVRI	5.43	1.98	1.46
SVRI	29.2	16.7	17.26

VSD closure done; Normalization of PA pressures in the post op. period

Other (?Genetic) Influences



Operability in L-R shunts

- How do we decide on operability of L-R shunts today?
  - What tools do we have?
  - What are their limitations?

Deciding operability of L-R shunts

- Clinical evaluation
- Chest X-ray and ECG
- Measurement of oxygen saturation
- Echocardiography
- Resting and post exercise ABG (PO2)
- Cardiac catheterization
- ?MRI

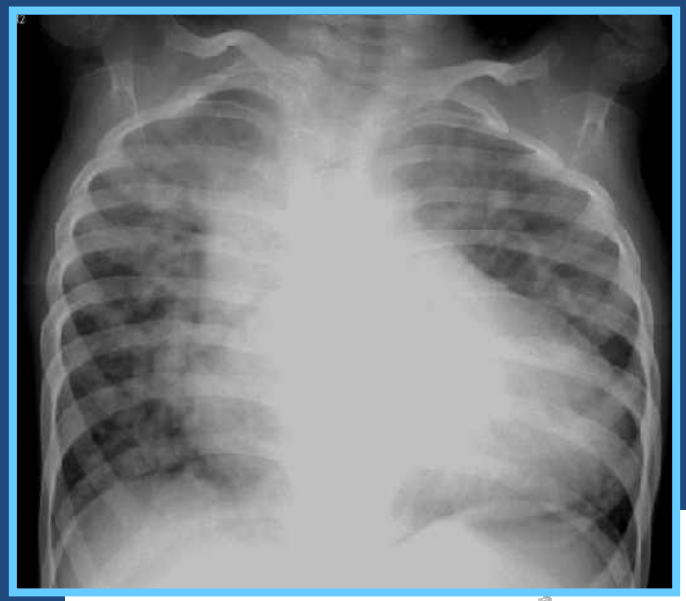
Viswanathan S, Kumar RK, Assessment of operability in congenital cardiac shunts with increased pulmonary vascular resistance, Cathet Cardiovasc Interv. 2008; 71:665-70

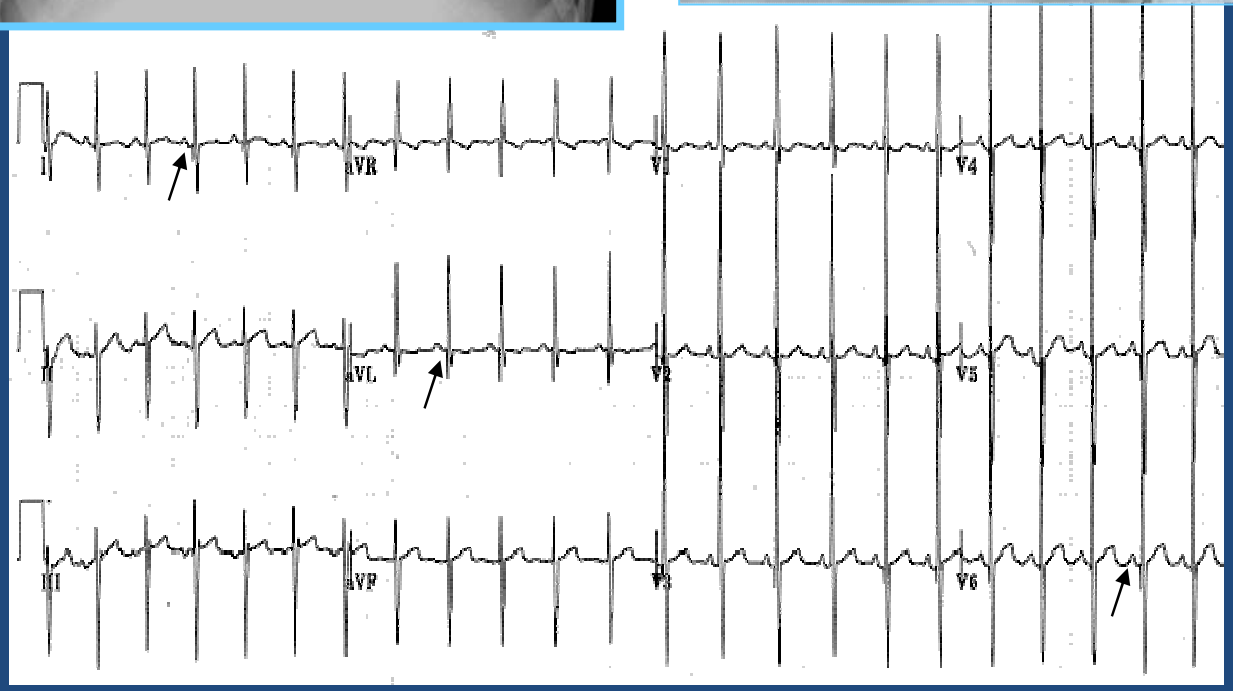
What principles govern decision on operability?

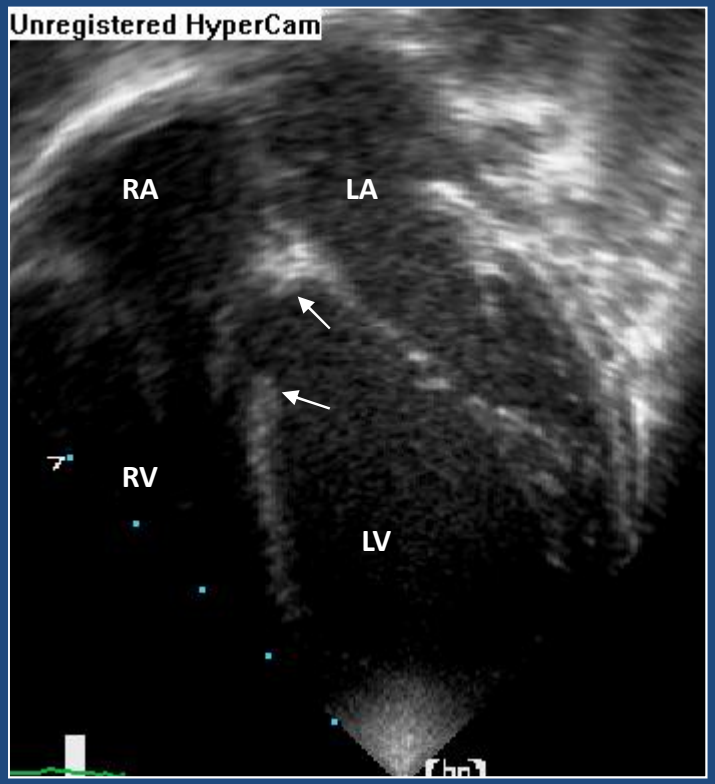
- Post tricuspid shunts:
  - Operable if significant shunt in the basal state irrespective of PA pressure
- Pre-tricuspid shunts:
  - Pulmonary hypertension (anything more than mild) warrants concern especially if basal shunt is not obvious

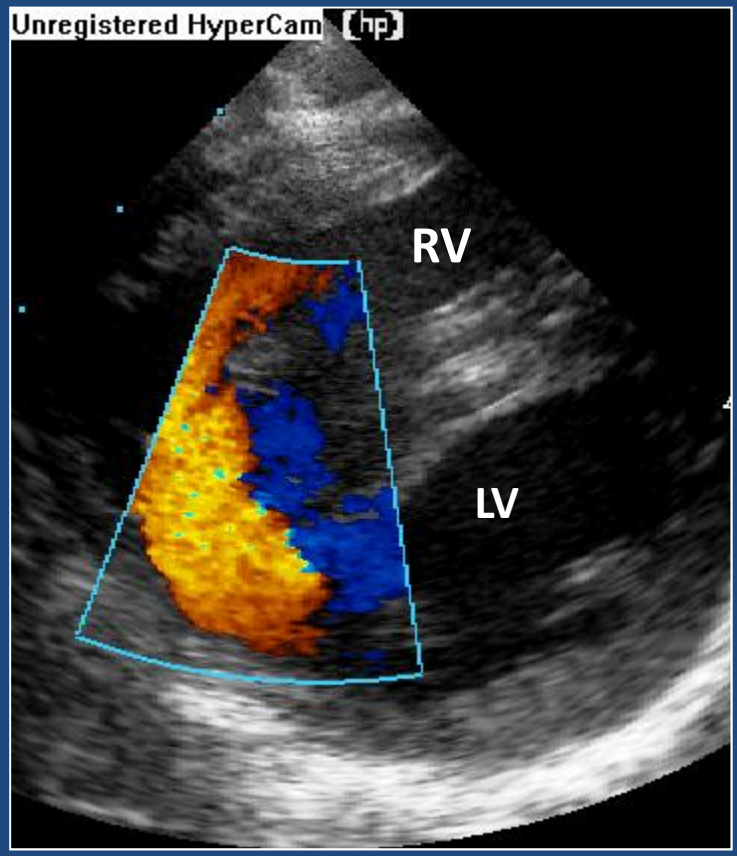
Deciding operability: Principles

- Age: benefit of doubt to younger patients.
- Lung, airway and ventilation issues can elevate PVR and confound assessment
- Pulmonary venous hypertension can result in reversible elevations in PVR

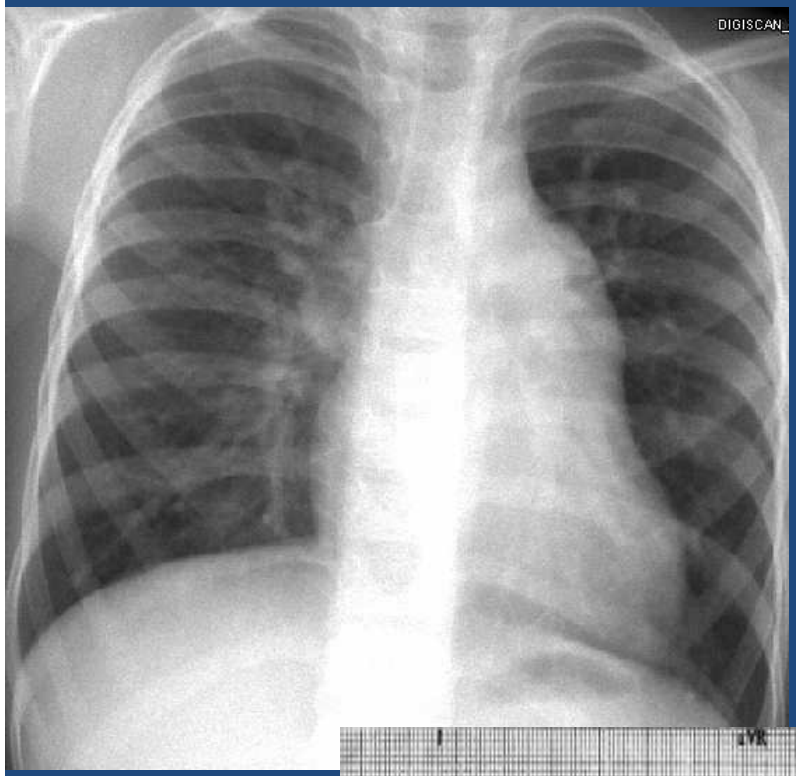


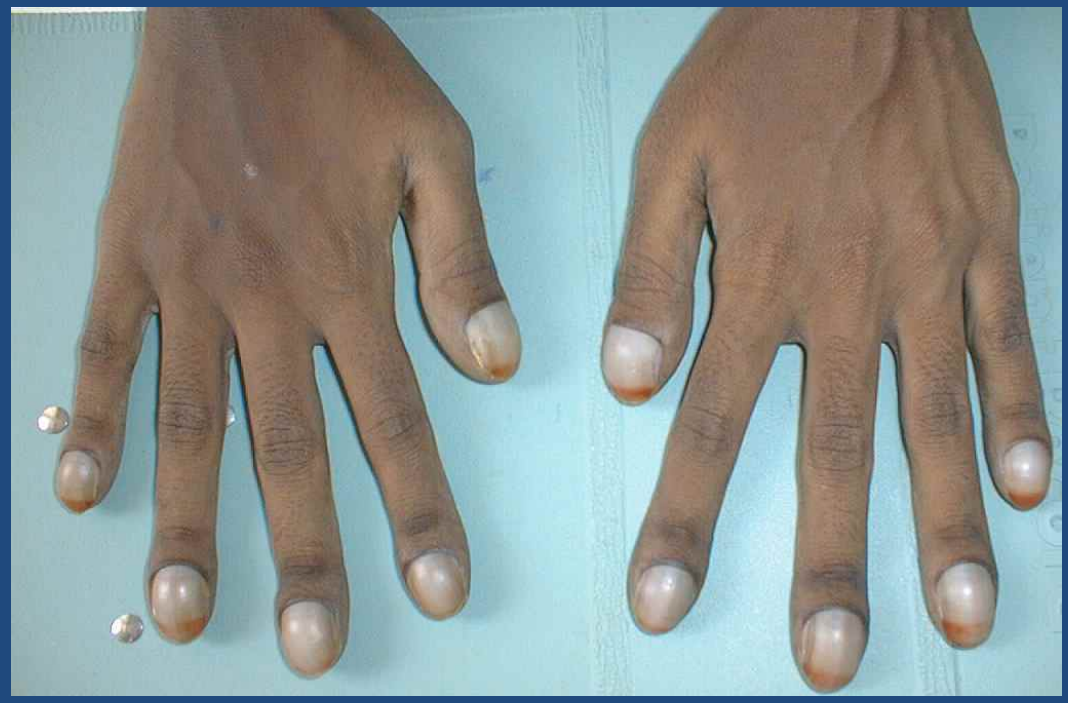

I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6

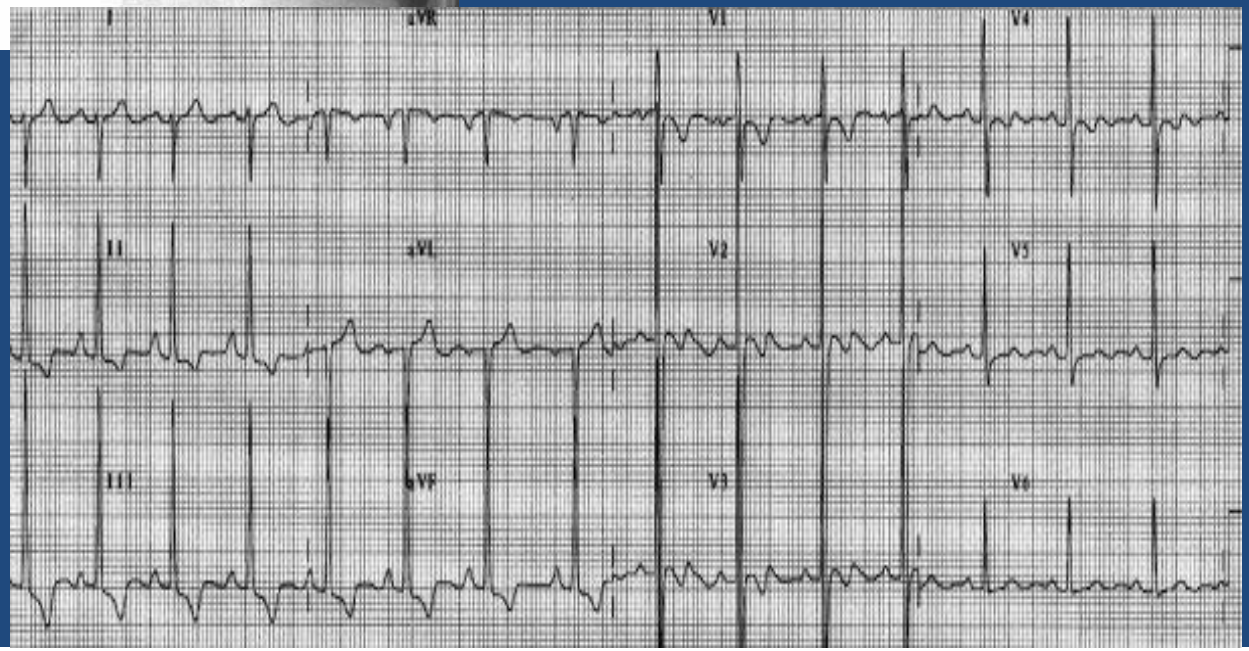




Clearly Operable: Cath not required



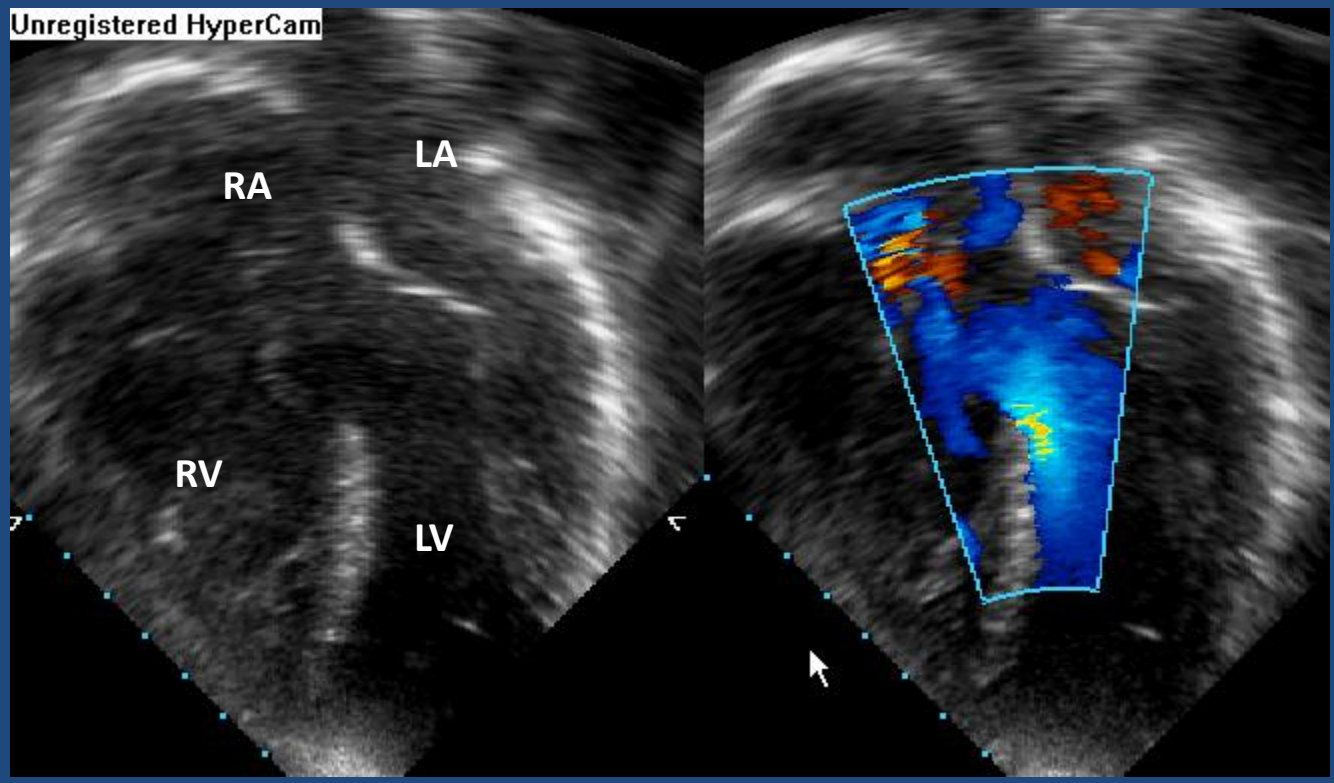




26 year old

Blue

Single loud S2



Clearly Inoperable: Catheterization unnecessary

Clinical spectrum of post-tricuspid shunts with PAH

Operable

Clear clinical /noninvasive evidence of a large left – right shunt

- Failure to thrive, ↑ precordial activity, mid diastolic murmur at apex,
- Cardiac enlargement, ↑pulmonary blood flow
- Q in lateral leads on ECG, good LV forces
- LA/LV enlargement, exclusively L-R flows across the defect

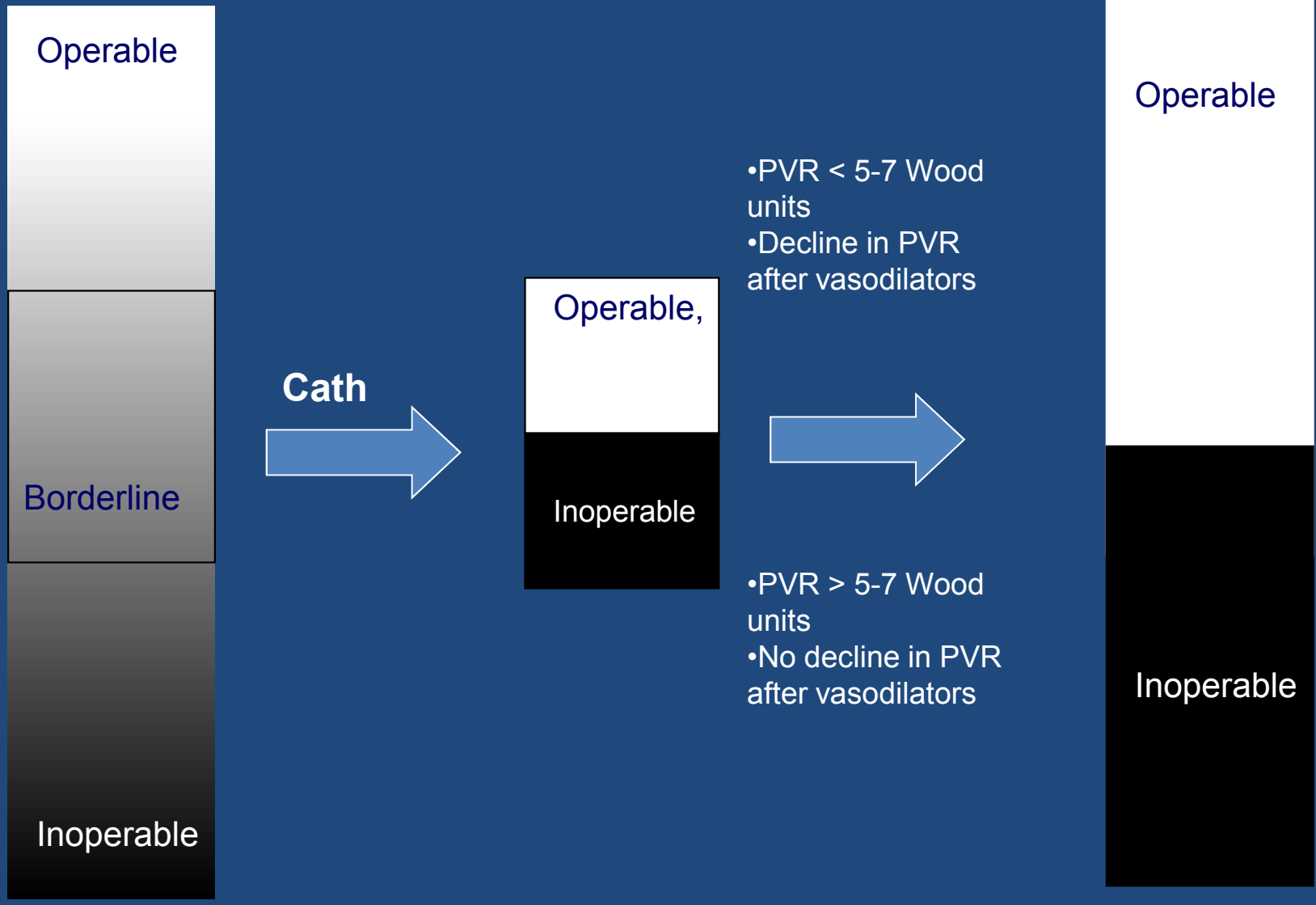
Borderline clinical non-invasive data: uncertain operability

Clear evidence of shunt reversal resulting from high PVR.

- Cyanosis, quiet precordium, no MDM
- Normal heart size, peripheral pruning
- No Q in lateral leads, predominent RV forces
- No LA LV enlargement, significant R-L flows across the defect

Inoperable

Ideally…..


Operable
Borderline
Inoperable
Cath
Operable,
Inoperable
•PVR < 5-7 Wood units
•Decline in PVR after vasodilators
•PVR > 5-7 Wood units
•No decline in PVR after vasodilators
Operable
Inoperable

PVR Estimation by Cardiac Catheterization

Pulmonary artery mean pressure − Pulmonary venous mean pressure

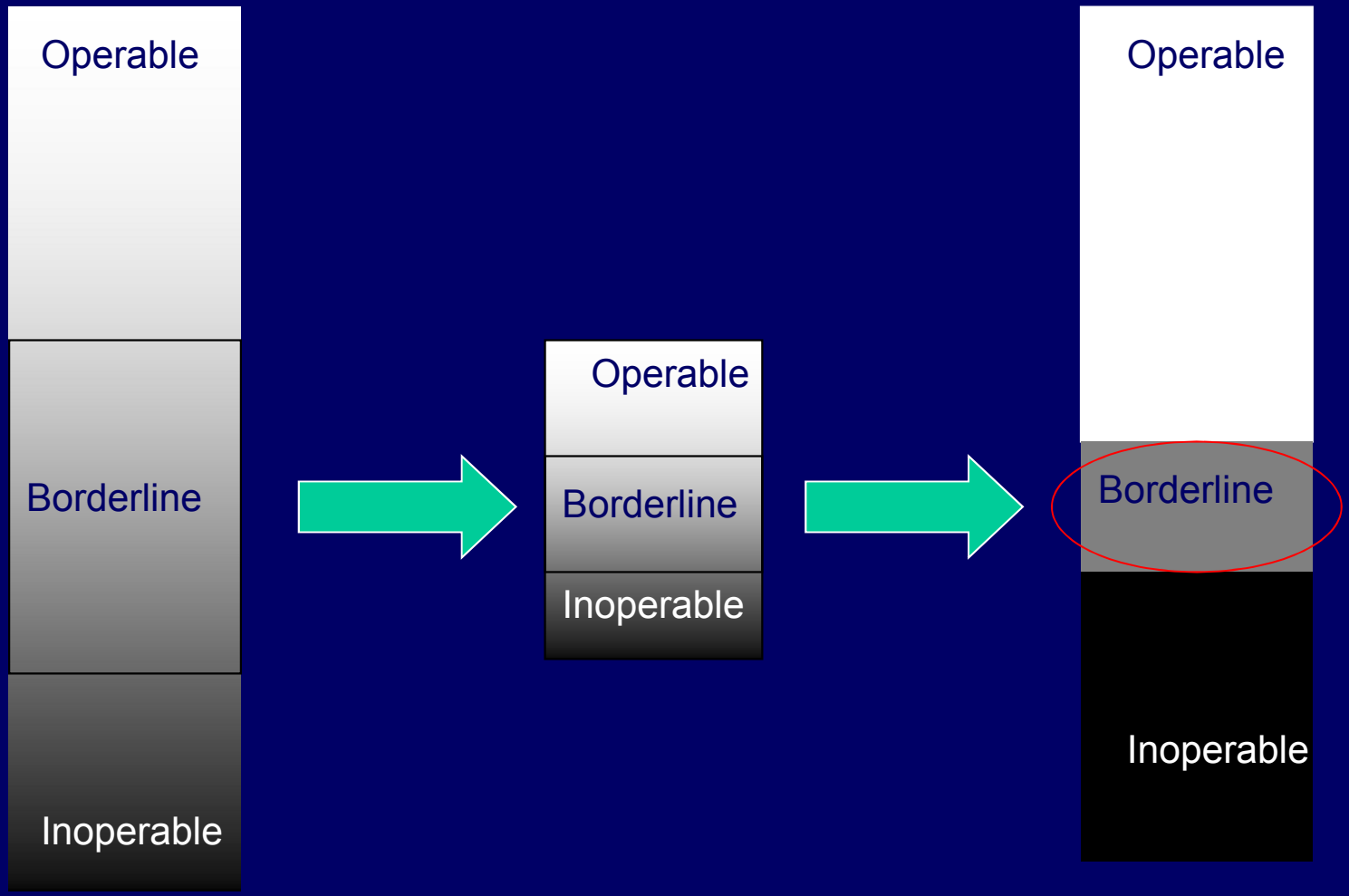
$$PVR = \frac{\text{Trans-pulmonary gradient}}{\text{Pulmonary blood flow}}$$

$$\frac{\text{Oxygen consumption}}{\text{PVO2 content} - \text{PA O2 Content}}$$

Sources of Error / Limitations in Catheterization Data

- Assumed oxygen saturations
- Assumed pulmonary vein saturation
- “Non-physiologic” state
- Calculated PVRI (basal and post-pulmonary vasodilator) has not been adequately standardized against the gold standard “surgical outcome”

In the Real World……



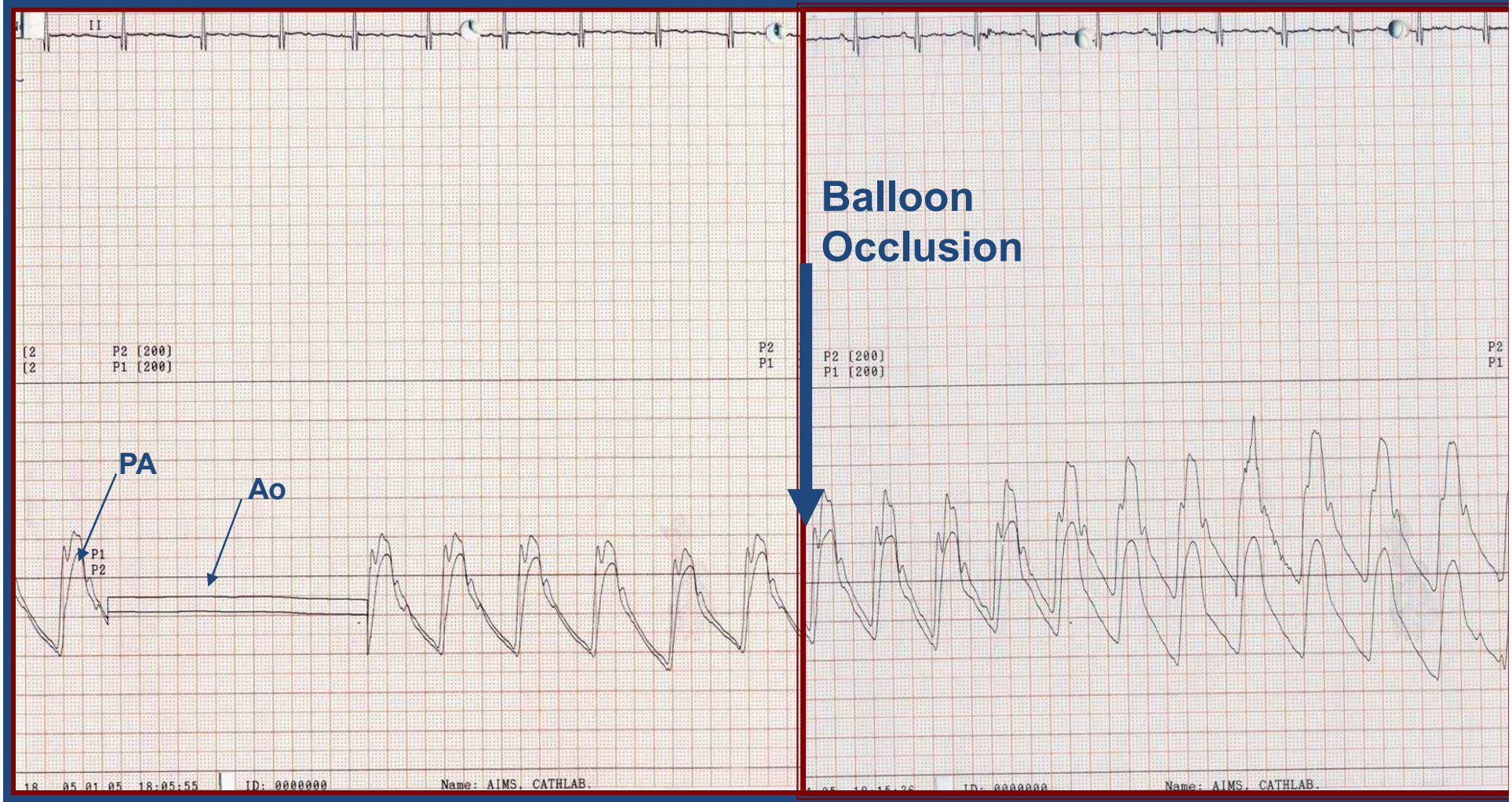
What else can be done in the cath lab?

- Test occlusion of the defects:
  - ASD
  - PDA
- Little validation with long term data
- Immediate reduction of PA pressure may not translate into long term benefits

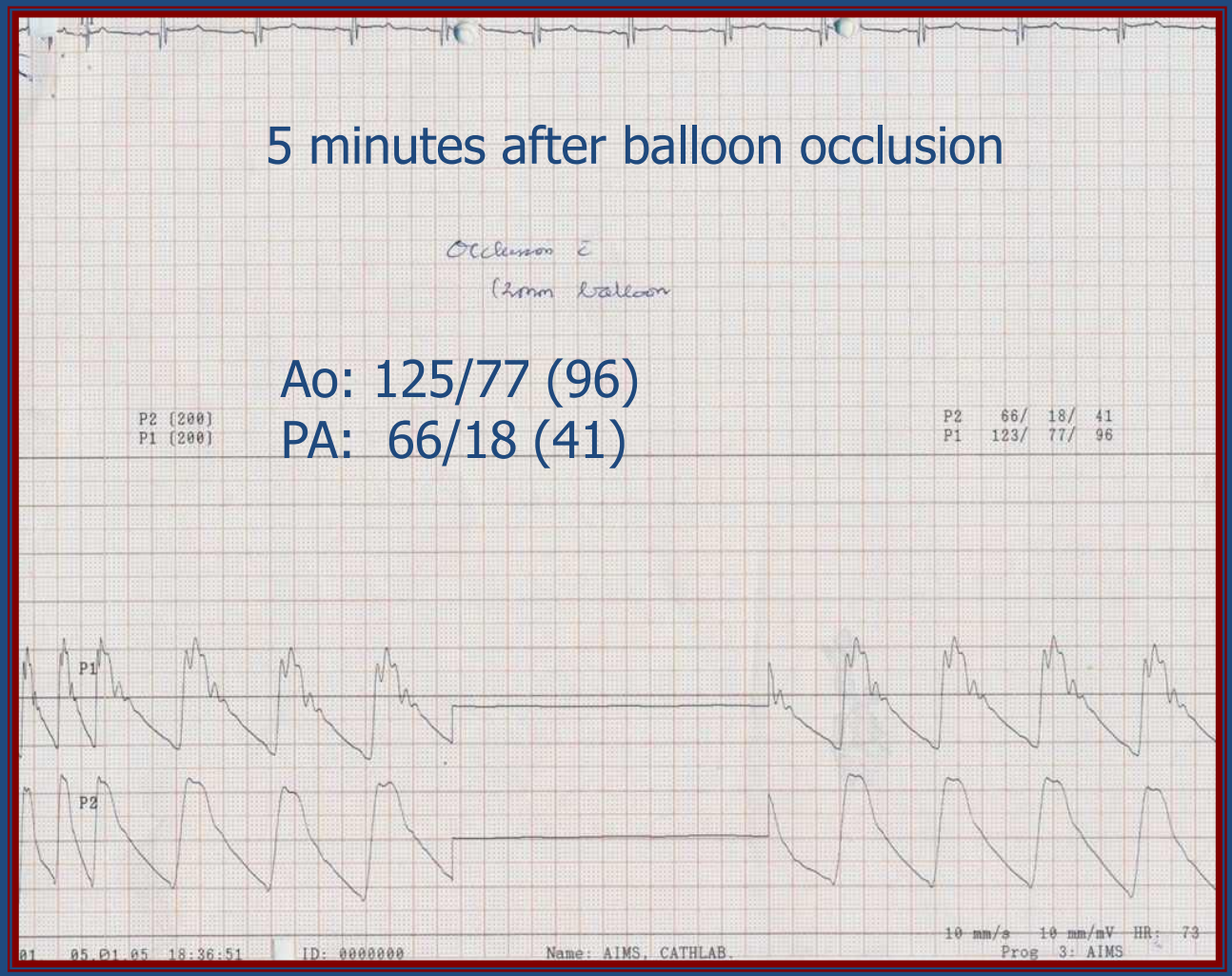
Illustrative Example

- 16 year old boy, 9.4 mm duct
- Nearly systemic PA pressures (Ao 120/60, mean: 90; PA 110/60: mean: 80)
- LL O2 Saturation: 96%
- Qp/Qs: 1.15:1 (Qp 3.8; Qs: 3.3)
- Basal PVRI: 18.75 Wood Units; PVRI/SVRI ratio: 0.66

Illustrative Example



Illustrative Example



Pulmonary Hypertension in ASD

- Clinical clues: limited utility
- Clear evidence of flow reversal (sats < 90%) suggests ↑↑ PVR
- Echo evidence of elevated PA pressure (RVSP > 50-60 mm Hg: Cause for concern, need for careful assessment of hemodynamics
- A decline in PO2 on exercise (>mm Hg 10 mm Hg) suggests high PVR (>7 wood units)

Should we have different thresholds for closing defects in the cath lab (vs. surgery)

- Answer outside the cath lab: No
- Practice inside the cath lab: ??
- Since it is easier: Lower thresholds?
- Think long-term
- Fenestrated ASD device

Should we change our practice because we now have some drugs for PAH?

- Lifelong requirement?
- Outcomes when compared to natural history?
- Expense and availability

The Way Forward……

We need data

- Need for good prospective studies
- Cut-offs
- Basal PVR, O2, NO, O2+NO
- Test-occlusion in the cath lab
- Meticulous follow-up after closure
- SCAMPS??

Standardized clinical assessment and management plans (SCAMP)

1. All patients assessed and managed the same way unless the data center is notified in writing
2. Precise entry criteria
3. Prescribed and enforced follow-up
4. Deviations from protocol are allowed but must be justified
5. *Not randomized, not research, no IRB*
6. Lends itself to multicenter study

Conclusions

- We need better data on our patients.
- Until then: Comprehensive approach and look at all data available (clinical, non-invasive, hemodynamic)
- Guidelines based on combined experience and wisdom
- Holistic and not “hole-istic”