Differential perfusion and regurgitation of pulmonary artery ; does it matter?

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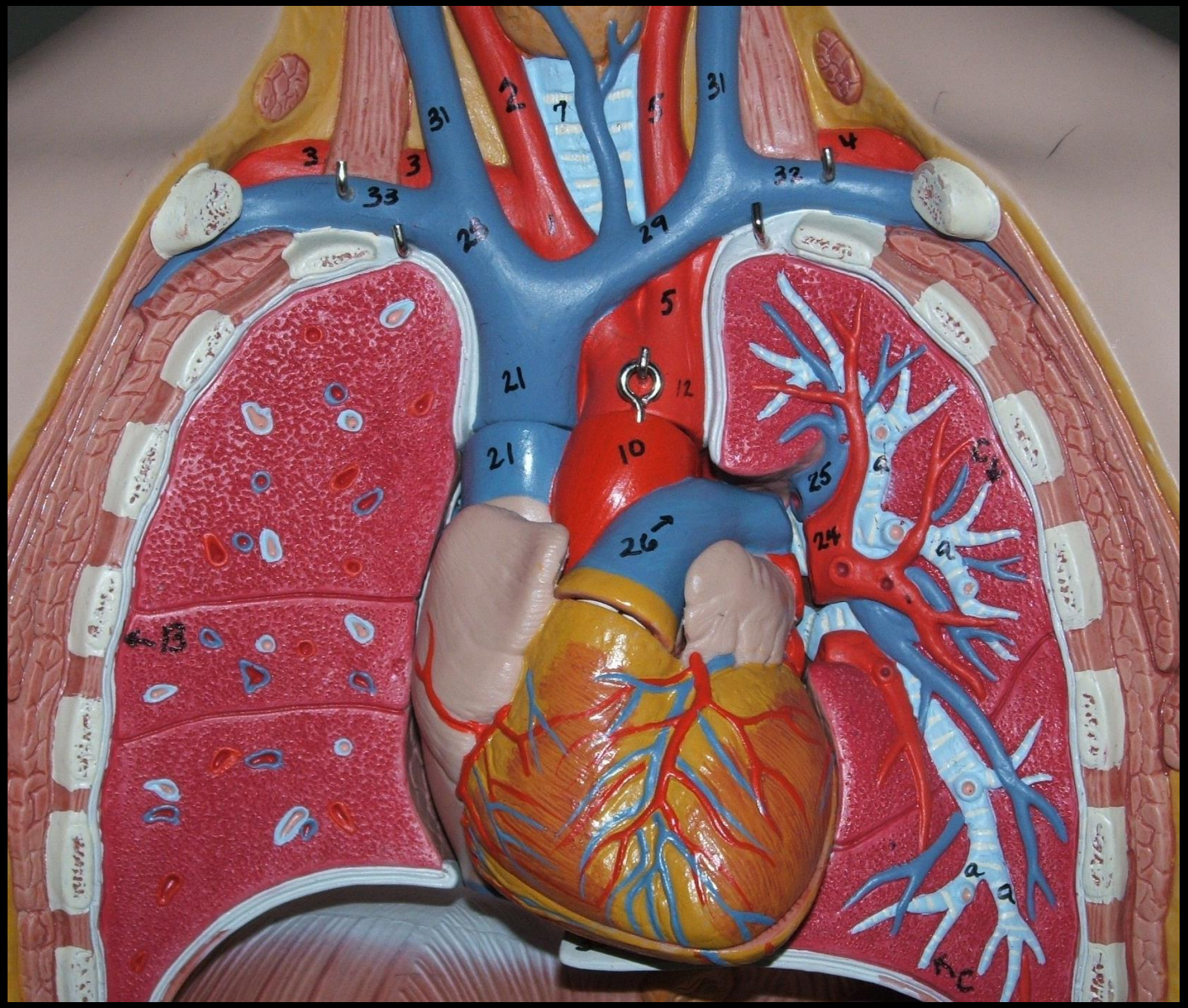
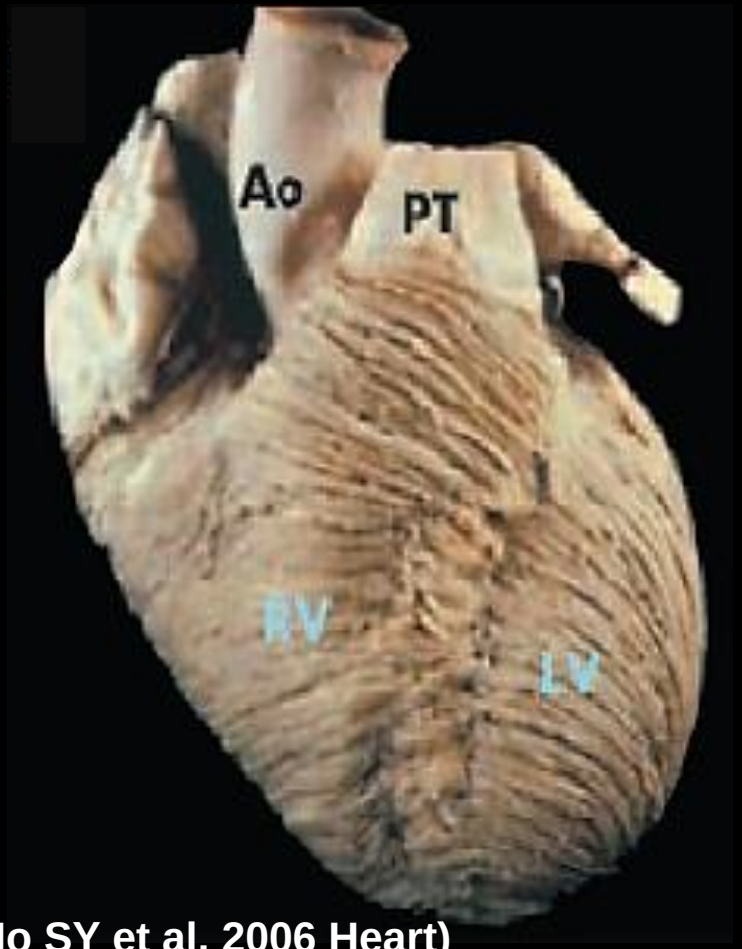
Heart and Lung



Relationships among two ventricles and two great arteries

The heart is a helix that contains an apex

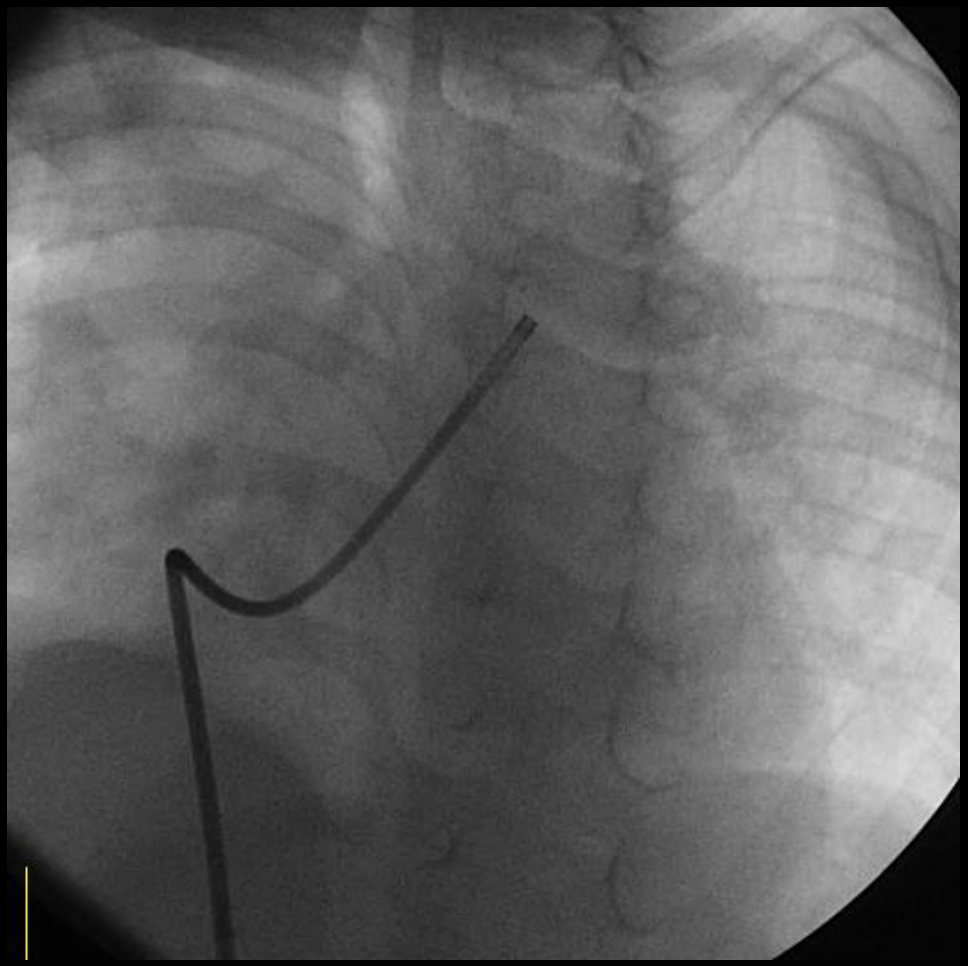


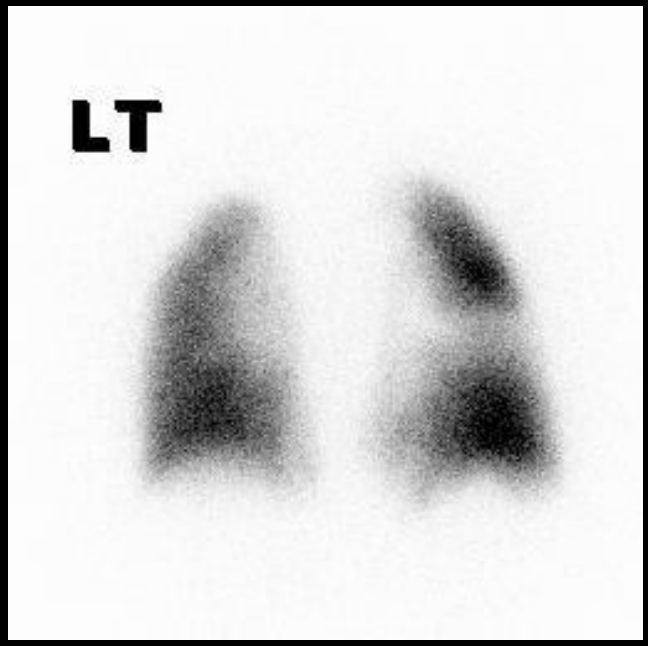
(Ho SY et al. 2006 Heart)

Continuity of superficial muscle among RV, LV and great arteries

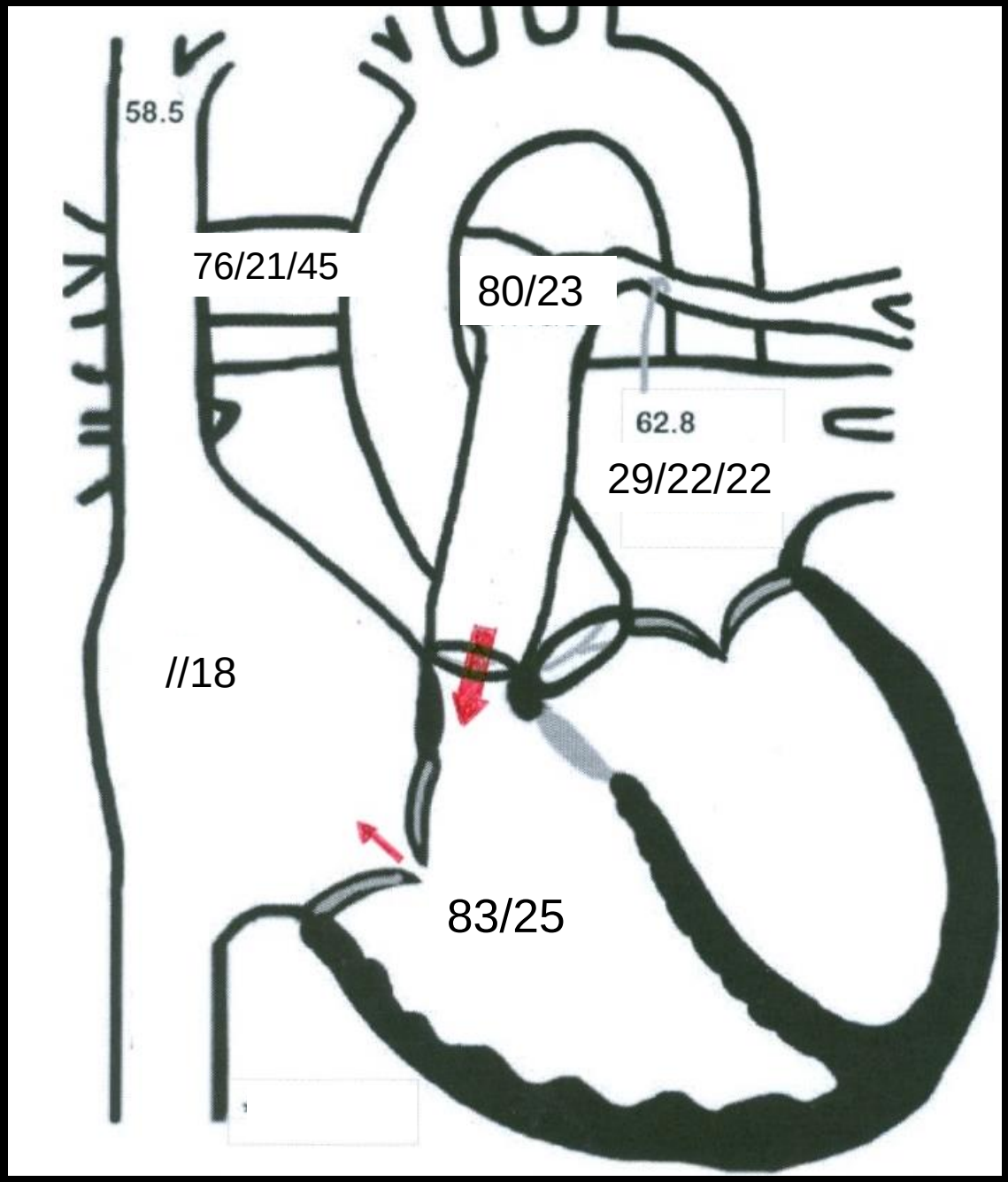
Lung perfusion in this patient?

s/p DORV with subaortic VSD, M/13 month-old





	LT	RT
Upper	7.0	13.4
Middle	18.7	20.0
Low	15.5	25.3
Total	41.3	58.7



$$\text{Flow} = \frac{\text{Pressure}}{\text{Resistance}}$$

	LT	RT
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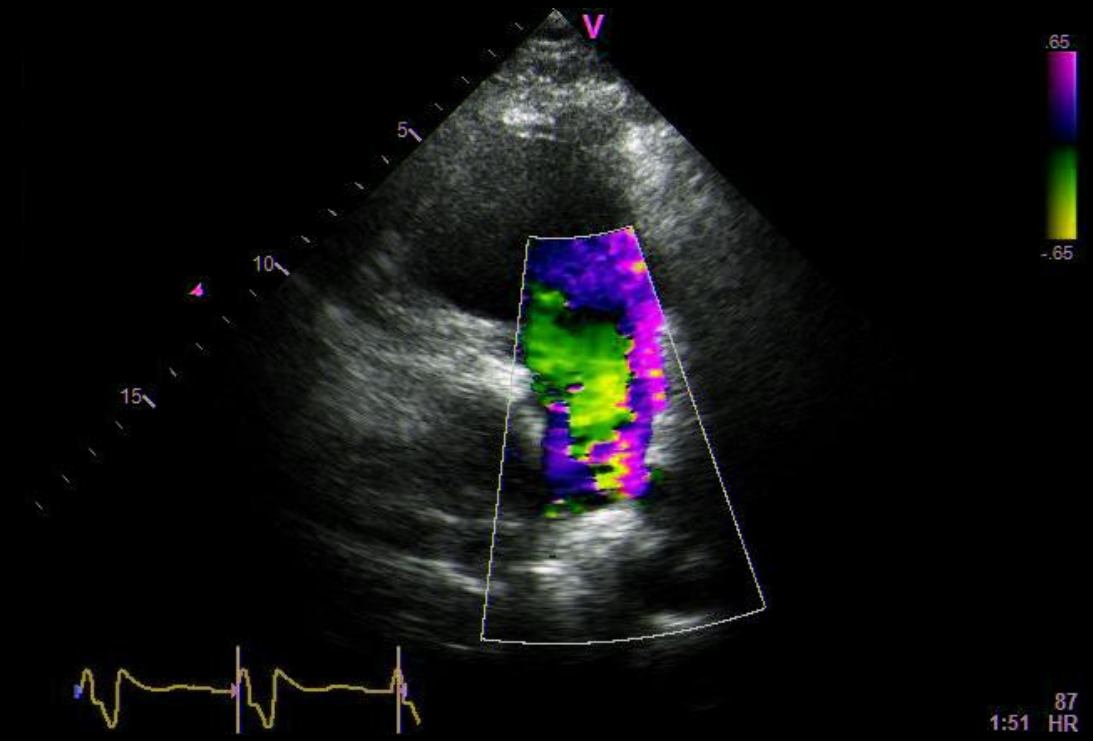
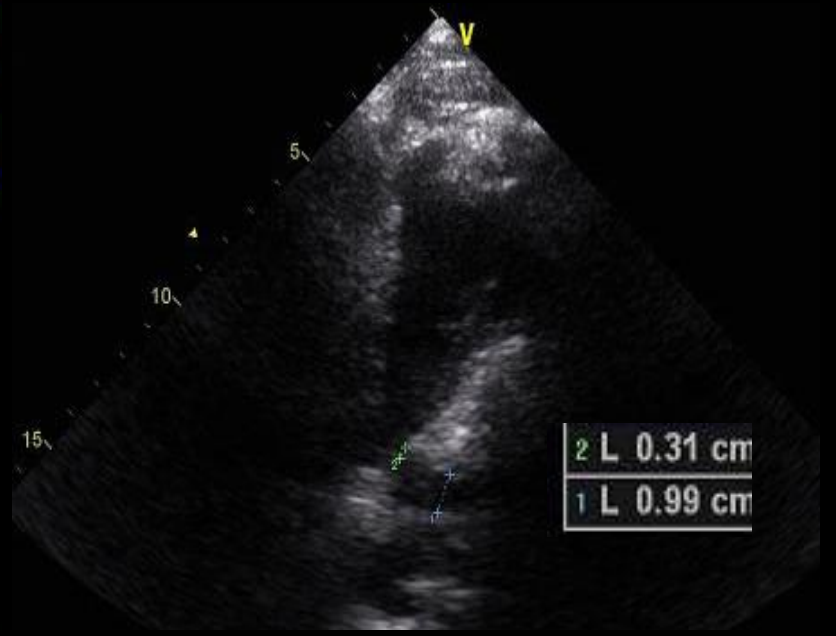
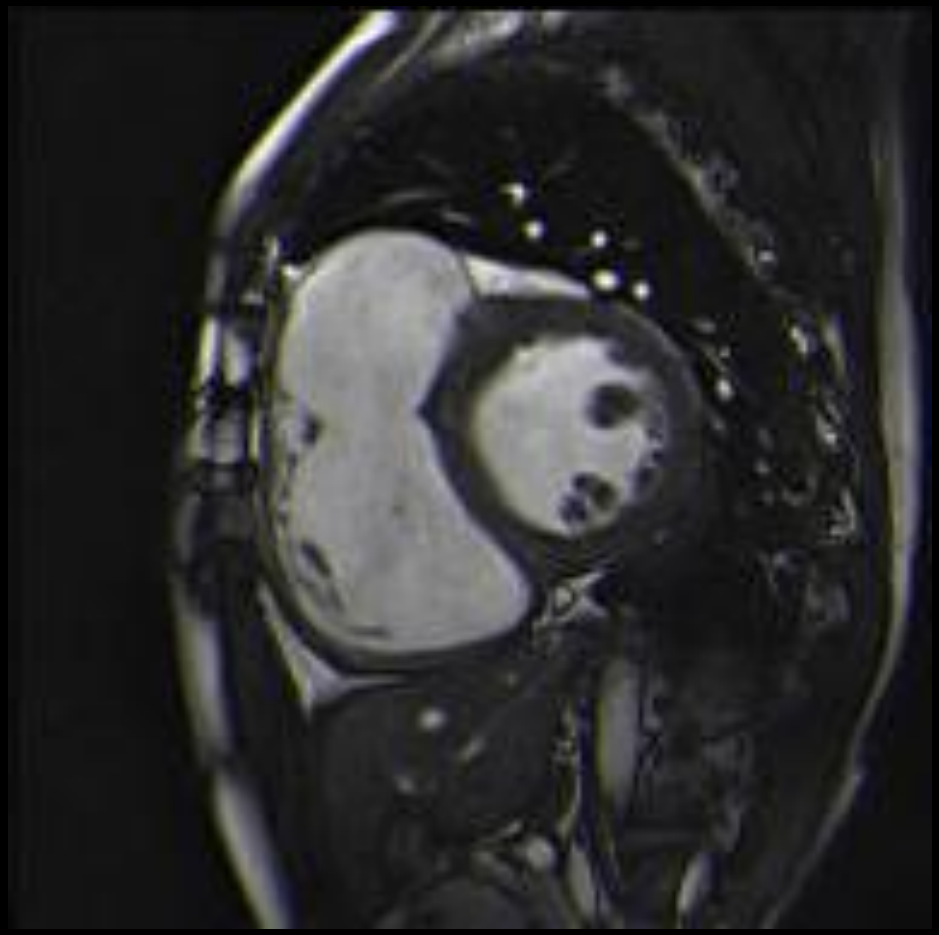
s/p TOF, 39/ Male



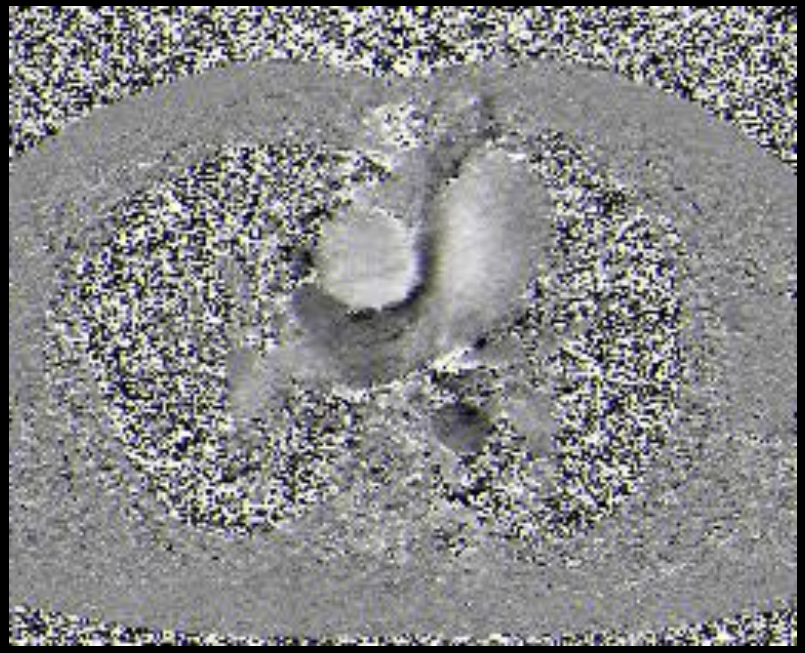


Cardiac MRI, s/p TOF, 39/ Male



- 1. Severe PR
  - PR fraction = 62.3%
  2. RV severe dilatation
  - RV indexed EDV
- = 403.8 ml/m^2
  3. RVOT aneurysmal change

Cardiac MRI, s/p TOF, 39/ Male



1. LPA focal tight stenosis
2. Lung perfusion

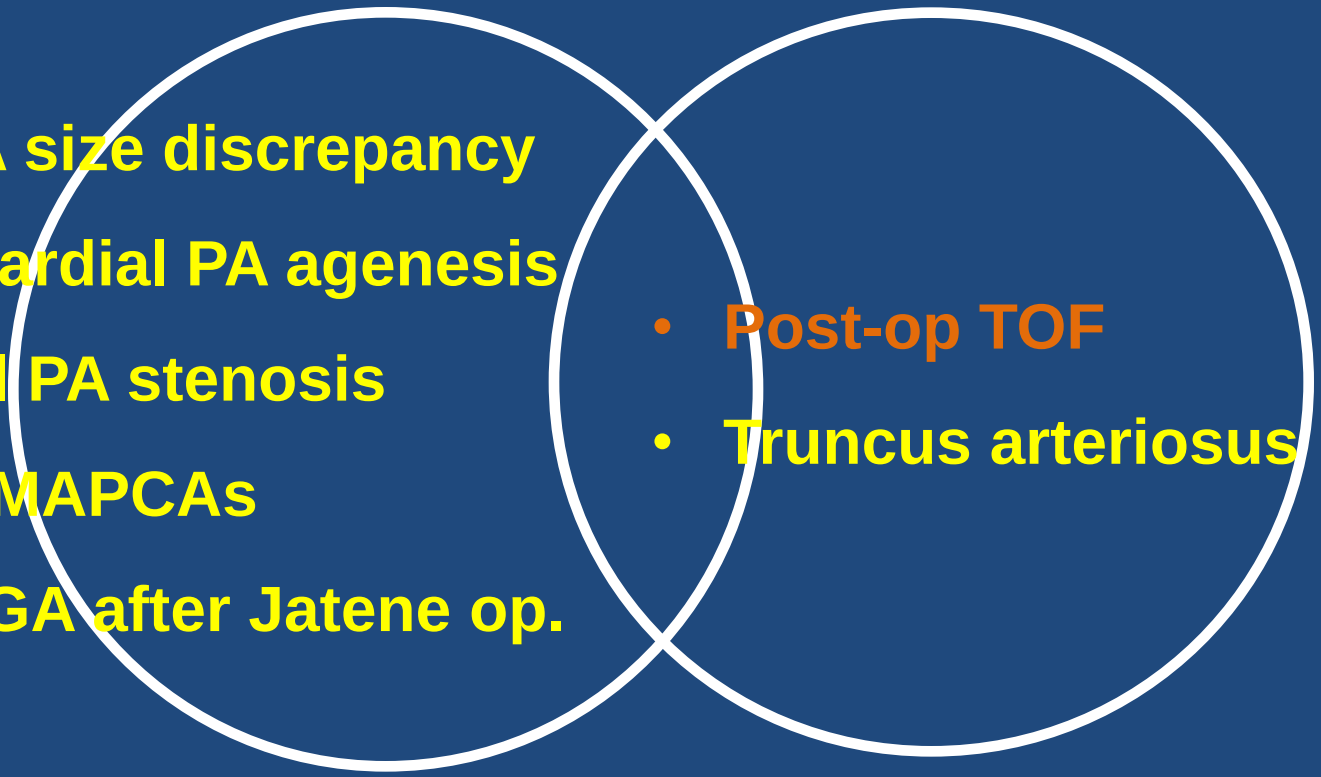
   - Right 85.6% : Left 14.4%

	Forward Volume	Reverse Volume	Net Forward Volume	Regurgitation Fraction, %
RPA	139.5	88.9	**50.7**	63.6
LPA	17.7	9.0	**8.5**	51.7
RPA+LPA	157.2	97.9	**59.2**	**62.3**
MPA	153.0	136.8	16.3	89.4

Affected Disease Category

Differential perfusion

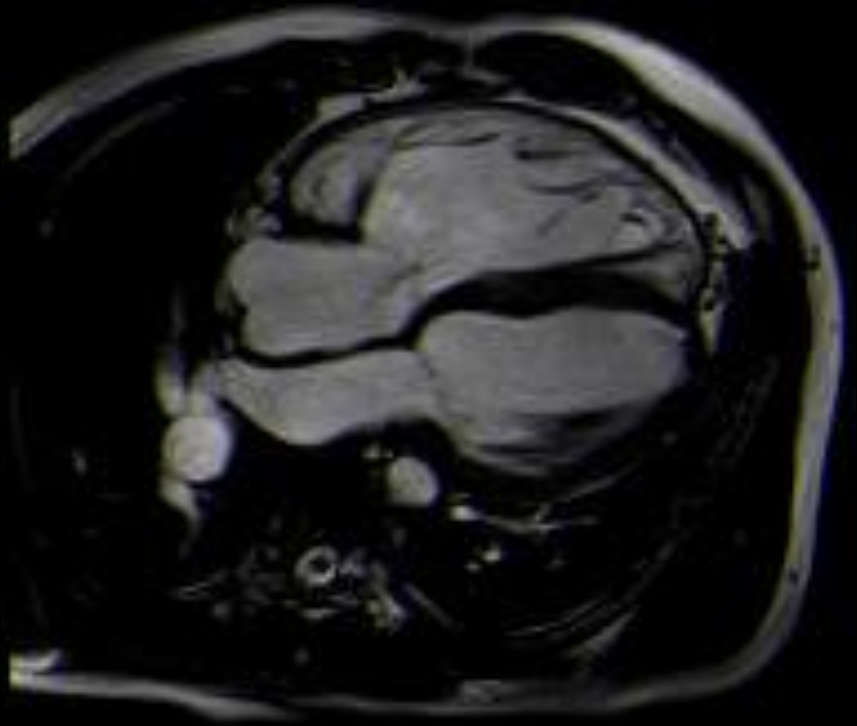
Differential regurgitation

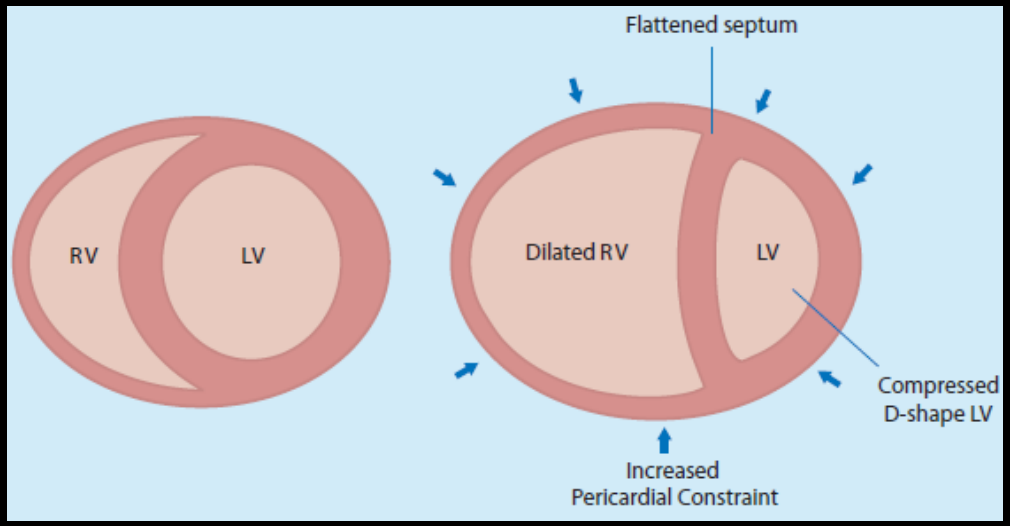


- **Branch PA size discrepancy**
  - **Intrapericardial PA agenesis**
- **Segmental PA stenosis**
- **CHD with MAPCAs**
- **Post-op TGA after Jatene op.**

- **Post-op TOF**
- **Truncus arteriosus**

The **effect** of **chronic PR** on **RV**

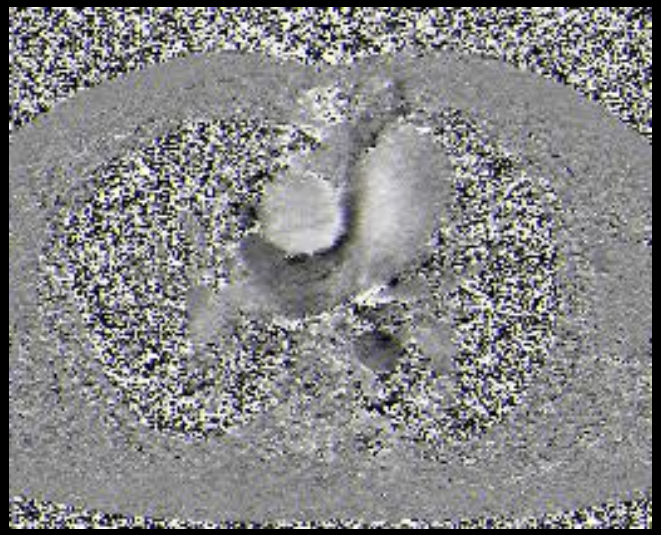


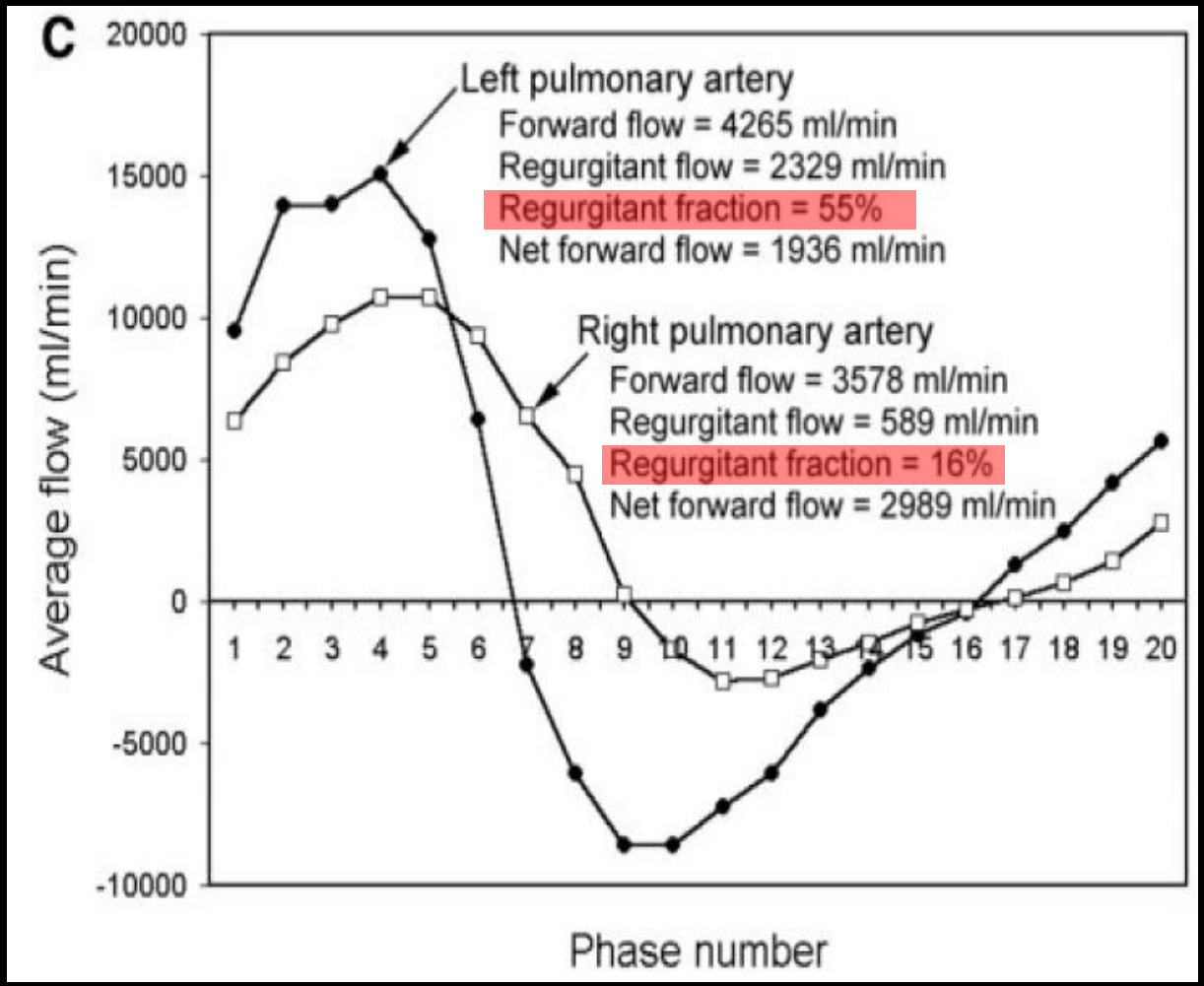


(Hadhad F et al. 2008 Circulation)

Differential regurgitation of pulmonary artery

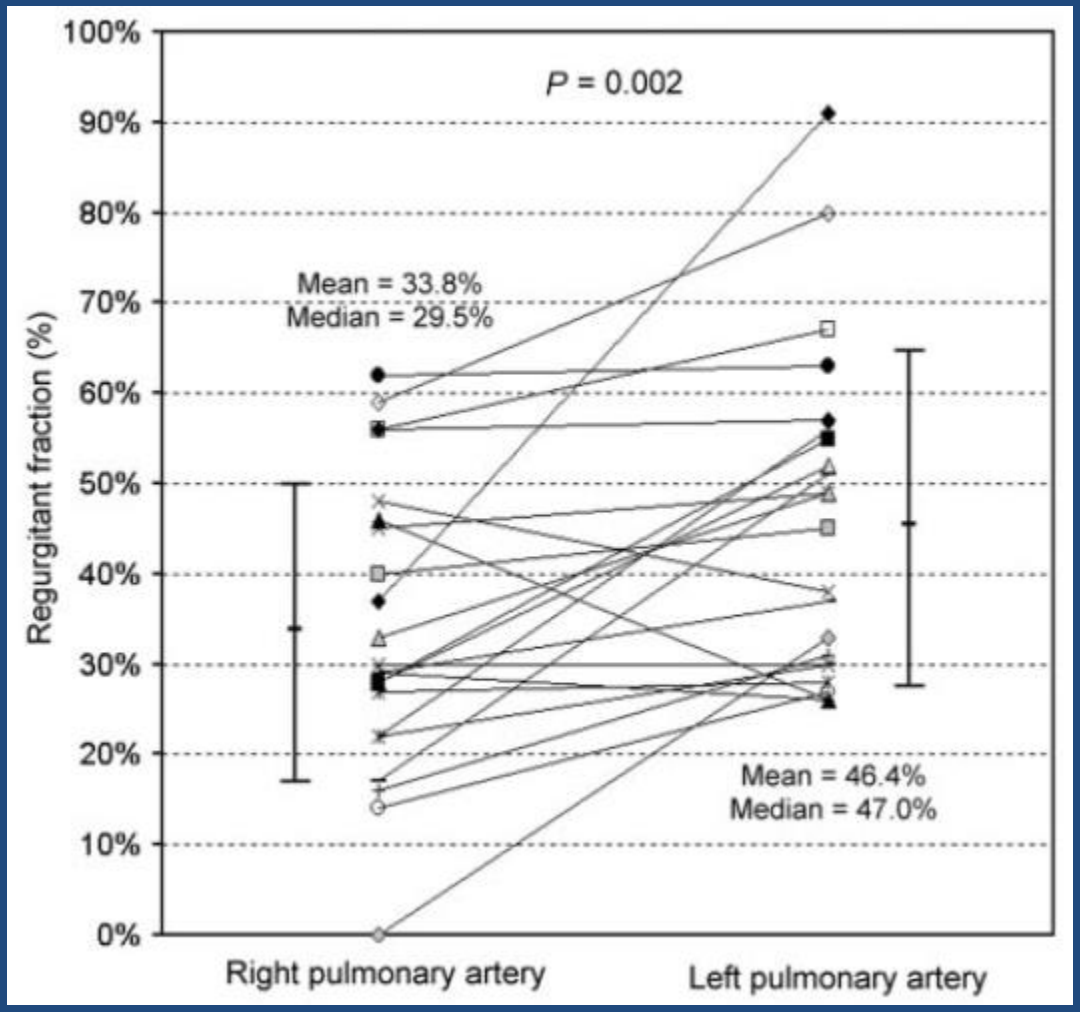
- Toronto Children's Hospital
- N = 22
- Age: (3.5~17.2) years
- TOF: 19, PA•VSD: 3





(Kang IS et al. 2003 Circulation)

Differential regurgitation of pulmonary artery



Size Difference	Net Blood Flow Distribution: RPA>LPA (n=9)	RPA=LPA (n=11)	RPA<LPA (n=2)
RPA>LPA (n=7)	6	1	0
RPA=LPA (n=12)	3	8	1
RPA<LPA (n=3)	0	2	1

(Kang IS et al. 2003 Circulation)

Differential regurgitation of pulmonary artery

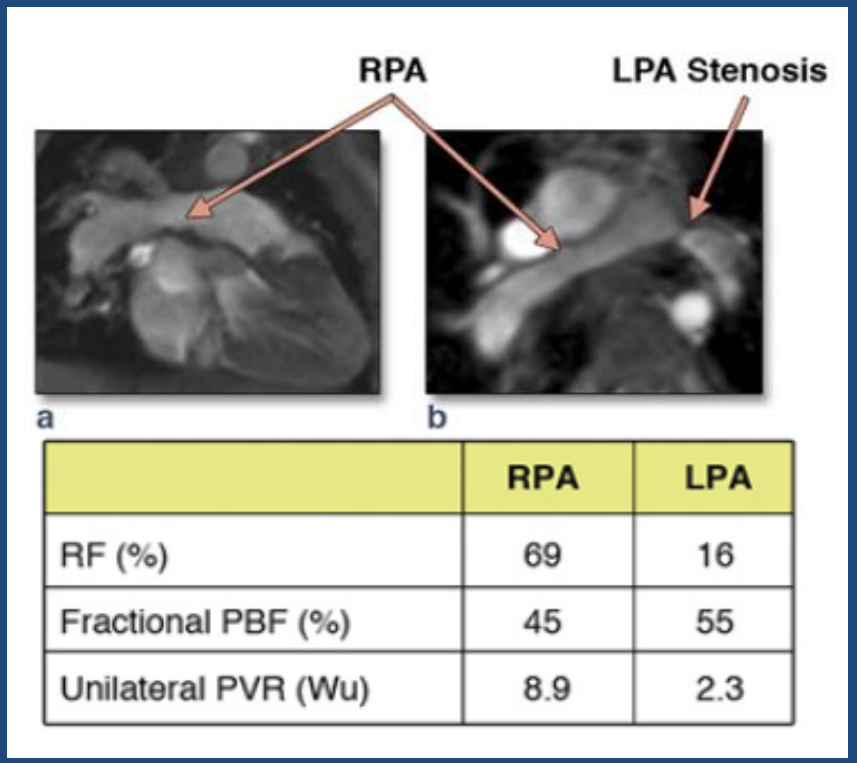
Conclusions

: **PR after repair of TOF** is commonly associated with differential regurgitation in the branch pulmonary arteries, which is **usually greater in the LPA**. Although the cause of this disparity requires further investigation, those patients with a significant unilateral contribution to total PR may be amenable to **localized techniques to reduce regurgitation**.

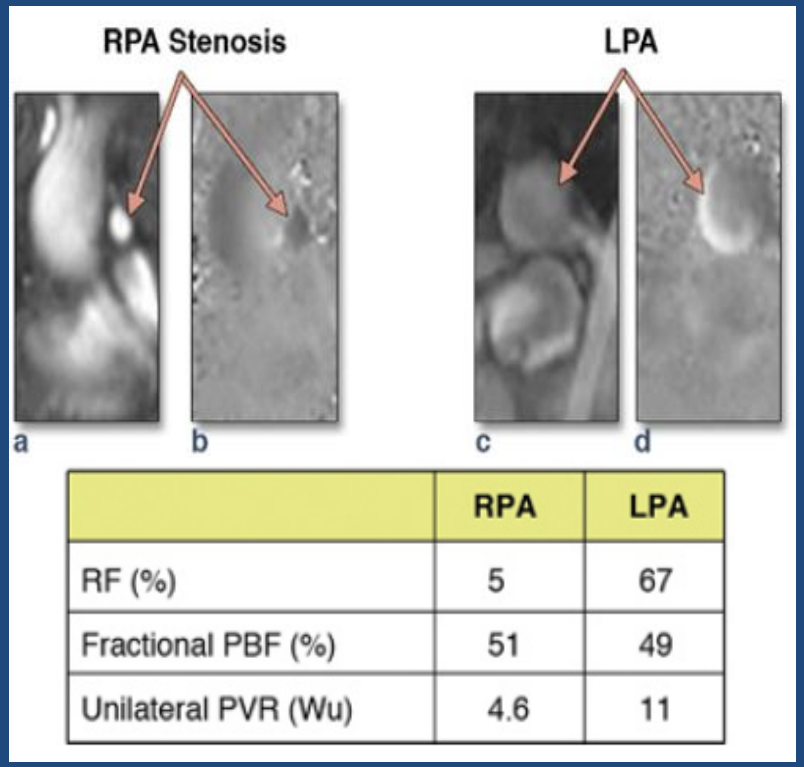
(Kang IS et al. 2003 Circulation)

Differential branch PA regurgitation vs PA anatomy, pul. vascular resistance

- Children's Hospital of Philadelphia
- 76 patients (2003-2006), mean age: 12.6±6.9 (0.1~35.5) years
- retrospective : cardiac MRI and cardiac cath. data



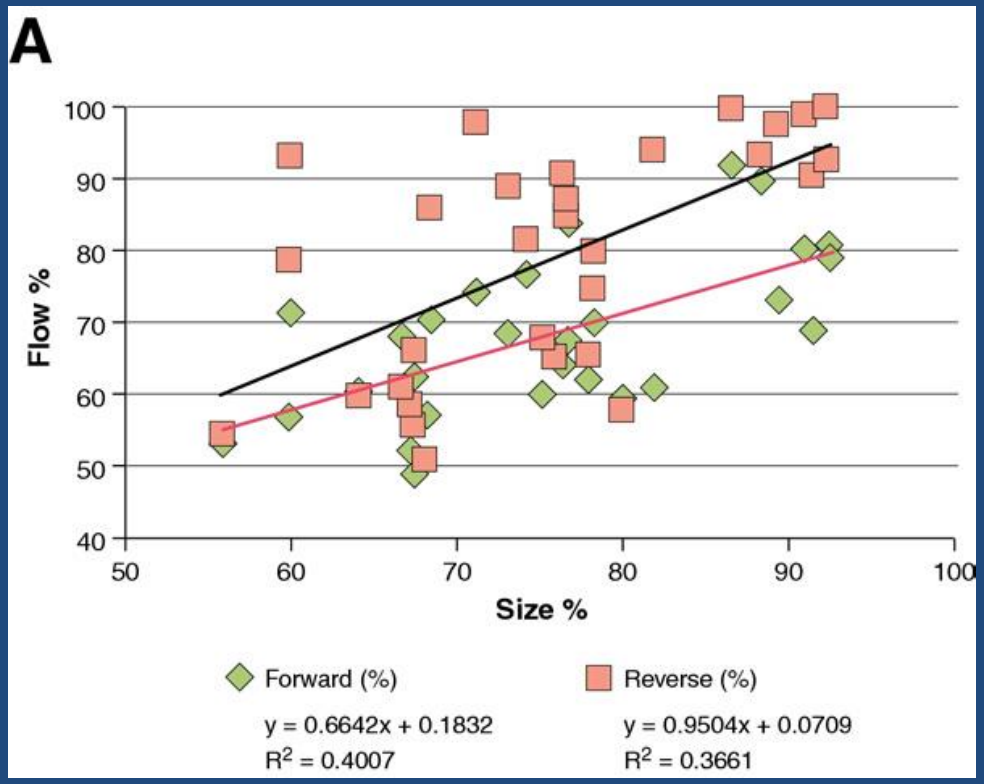
	RPA	LPA
RF (%)	69	16
Fractional PBF (%)	45	55
Unilateral PVR (Wu)	8.9	2.3



	RPA	LPA
RF (%)	5	67
Fractional PBF (%)	51	49
Unilateral PVR (Wu)	4.6	11

(Harris MA et al. 2011 J Am Coll Cardiol Img)

Differential branch PA regurgitation vs PA anatomy, pul. vascular resistance

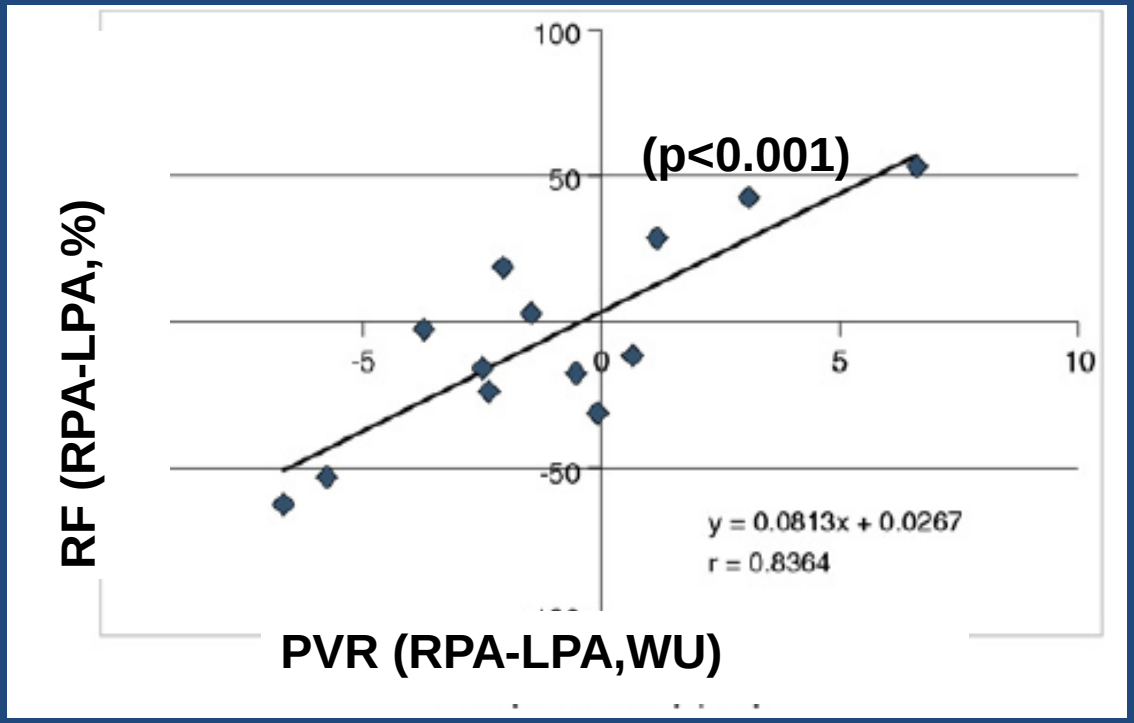


Increased PR fraction of larger versus smaller branch PA

(39% vs. 21%, p<0.001)

(Harris MA et al. 2011 J Am Coll Cardiol Img)

Differential branch PA regurgitation vs PA anatomy, pul. vascular resistance



$$\text{Flow} = \frac{\text{Pressure}}{\text{Resistance}}$$

Differential Branch PA RF strongly correlated with

differential branch PA resistance

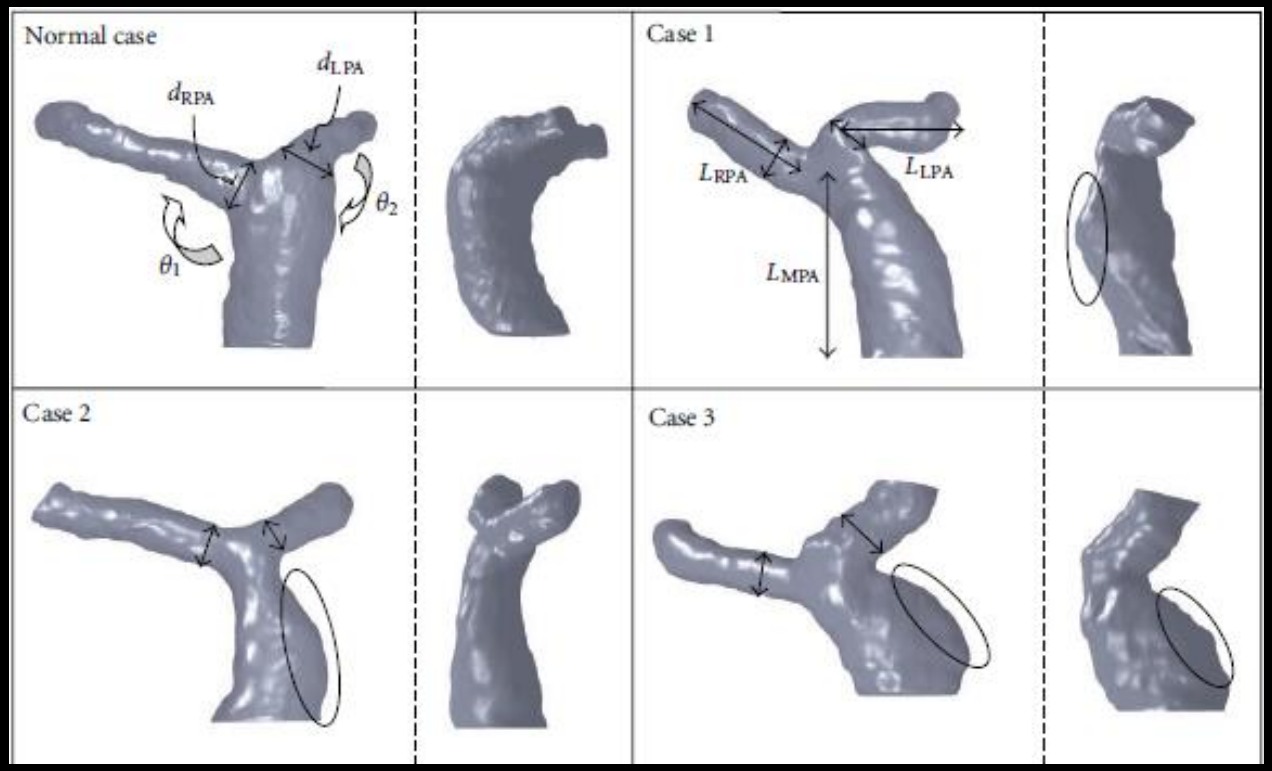
(Harris MA et al. 2011 J Am Coll Cardiol Img)

Differential branch PA regurgitation vs PA anatomy, pul. vascular resistance

Conclusions: **BPA RF is a function of the relative PVR and the presence of BPA stenosis or size discrepancy**. Contrary to prior reports, the LPA RF was only elevated in patients with relatively equal-sized BPAs. In the setting of BPA stenosis or size discrepancy, net flows will not identify unilateral increases in PVR. Therefore, **measuring the differential RF** is an important tool for screening patients for unilateral increases in PVR, which can affect the **indication and timing for repair of BPA stenosis.**

(Harris MA et al. 2011 J Am Coll Cardiol Img)

Differential branch PA regurgitation vs PA anatomy

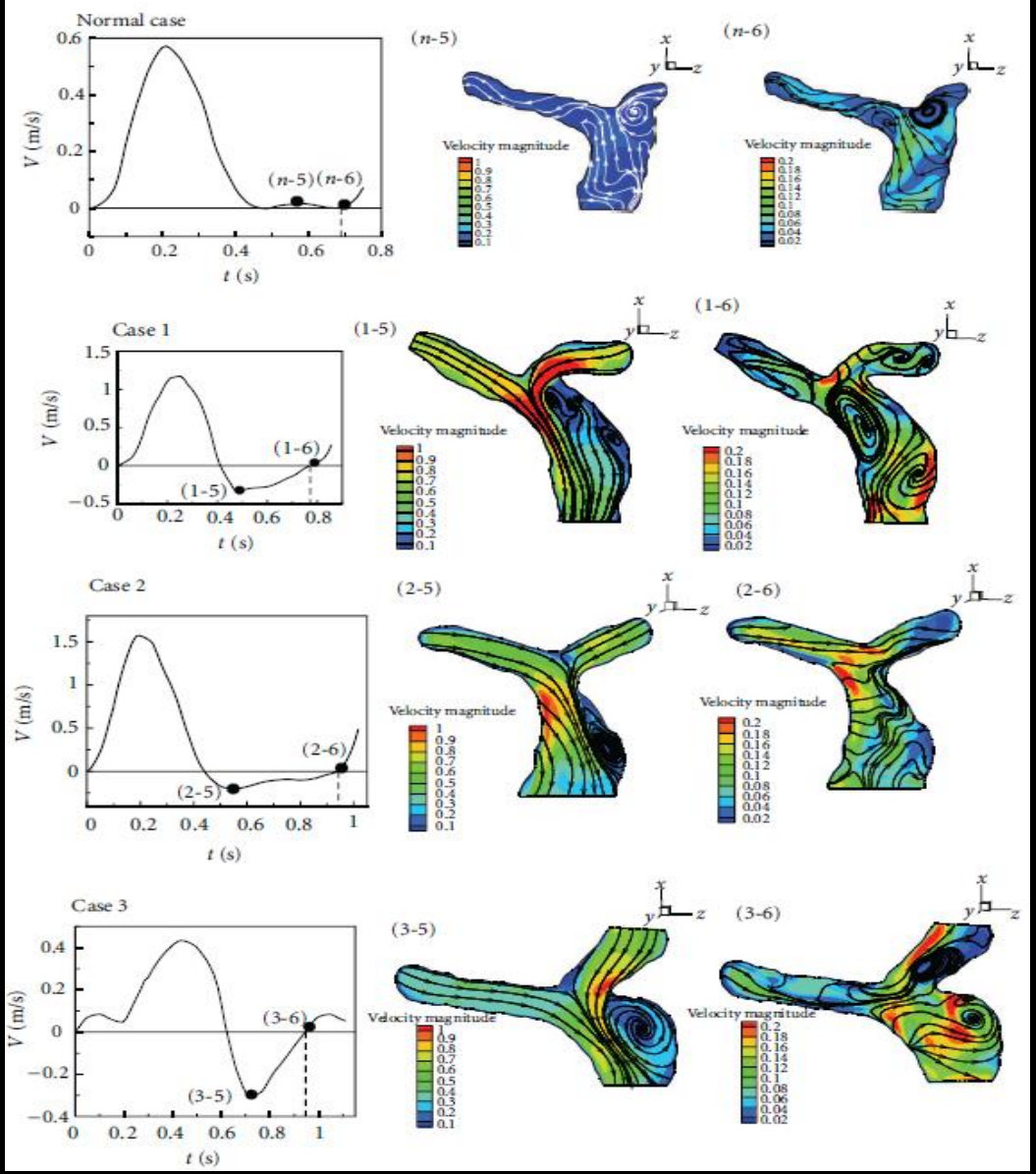


Regurgitant Fraction (%)

Case	MPA	LPA	RPA
1	33.7	44.9	25.2
2	16.4	18.6	14.9
3	28.8	30.4	26

(Chern MJ et al. 2012 Comput Math Methods Med)

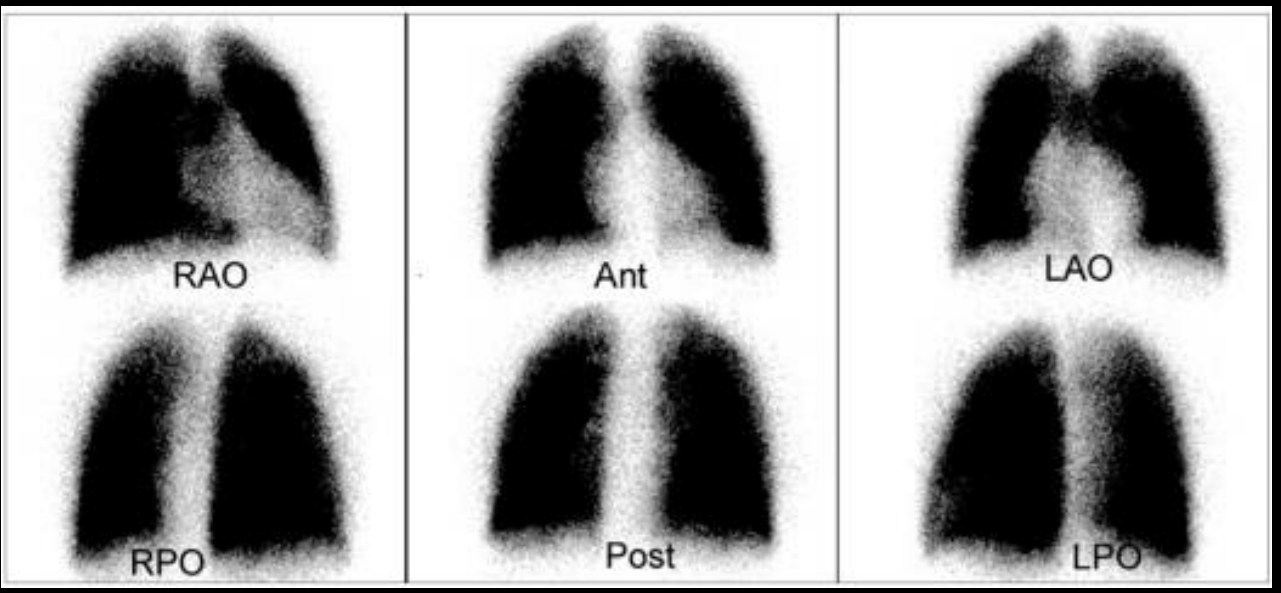
Numerical Study for Blood Flow in Pulmonary Arteries after Repair of Tetralogy of Fallot



- The blood flow is influenced by
  - bifurcation angles
  - geometry of PA
- The regurgitation
  - happens first in LPA due to the small angle between LPA and MPA

(Chern MJ et al. 2012 Comput Math Methods Med)

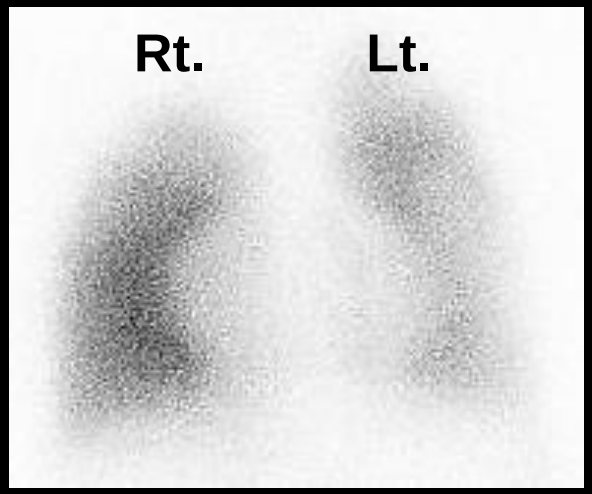
Differential perfusion of pulmonary artery



정상

Rt. : Lt. = 55 : 45

(Fathlal A. 2010 Heart views)



LUNG	LT	RT
Upper	11.0	6.8
Middle	14.9	33.3
Low	8.7	25.3
Total	34.6	65.4

The effect of differential pulmonary perfusion on lung

- **Pulmonary hypertension** in contralateral PA
  - right ventricular hypertrophy

- **Hemoptysis** from systemic artery collaterals to the hypo-perfused lung

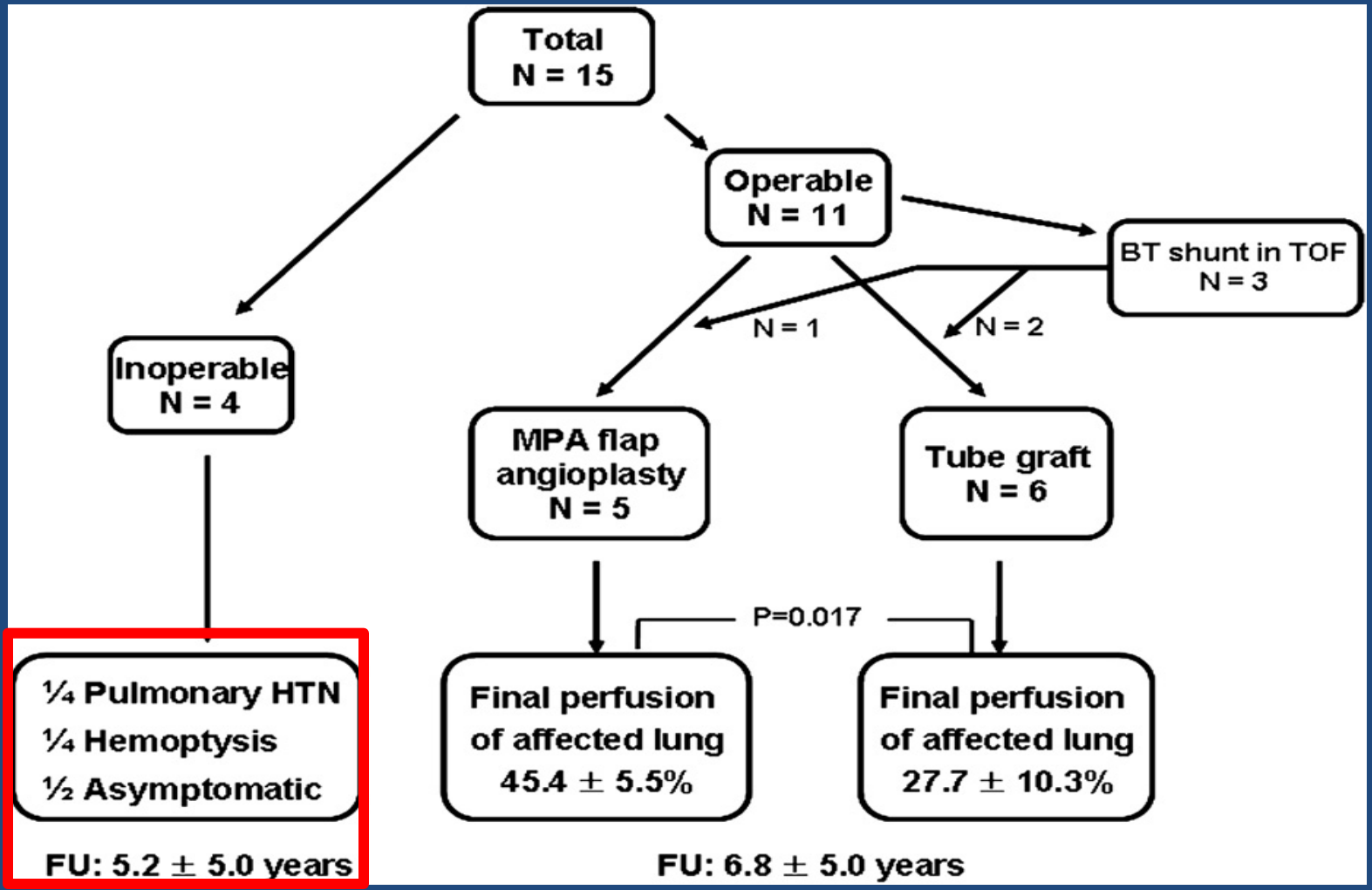
- **Poor development** of hypo-perfused **lung and chest**

The effect of differential pulmonary perfusion on lung

- Left intrapericardial PA agenesis
- Male/33 month-old

Chronic effects of differential pulmonary perfusion on lung

Intrapericardial unilateral PA agenesis (1991-2008, SNUCH)

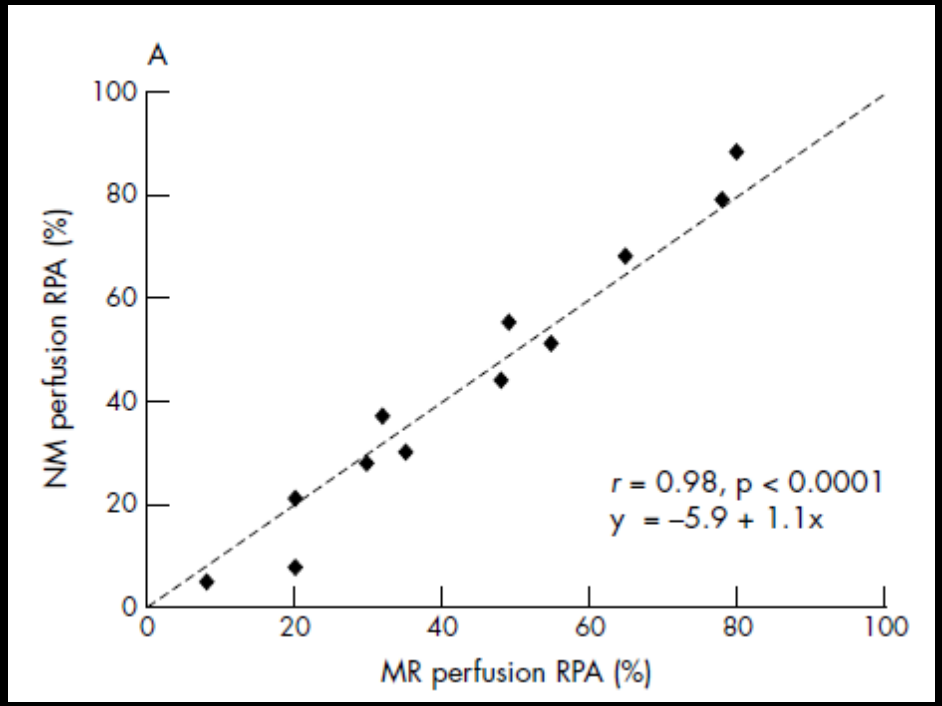


(Kim GB et al. 2011 J Thorac Cardiovasc Surg)

How to measure differential pulmonary perfusion?

Lung perfusion scan or cardiac MRI for quantitative measurements!

- **PA segmental stenosis**
  - **CHD with MAPCAs**
  - **Williams syndrome**
  - **Alagille syndrome**
- **Branch PA size discrepancy**
  - **unilateral PA agenesis**
- **Post-op TGA after Jatene op.**

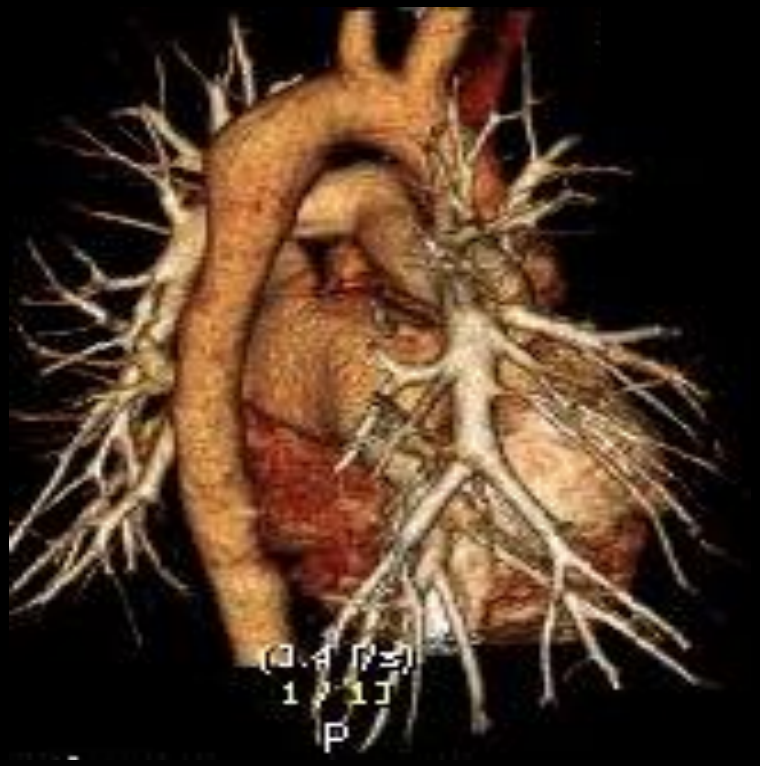


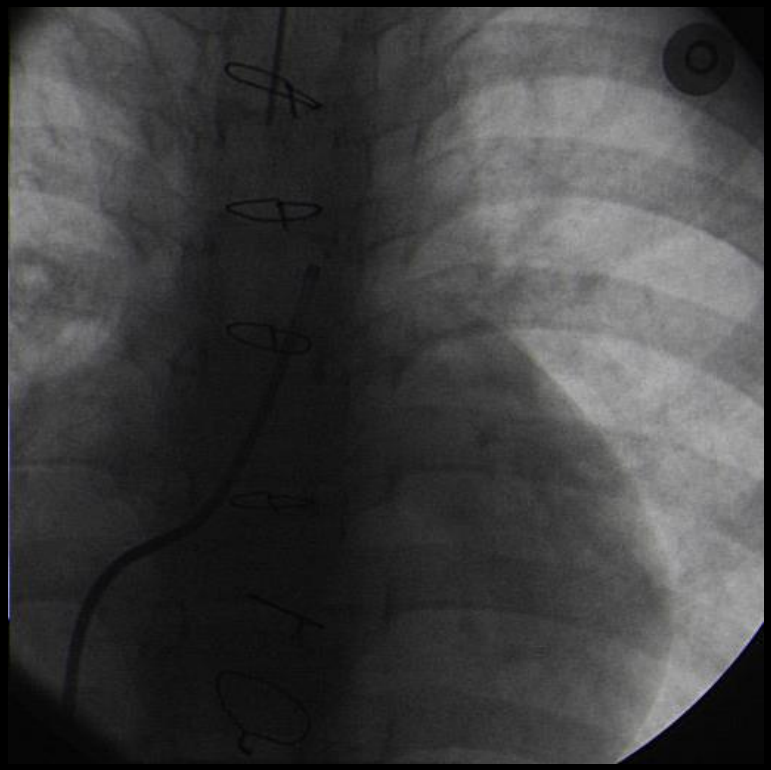
(Sridharna S et al 2006 Heart)

Segmental branch level : **Lung perfusion scan** >> MRI

How to measure differential pulmonary perfusion?

Cardiac CT, MRI or catheter-based angiography for anatomic evaluation





Segmental branch level : **Catheter-based angiography** >> CT, MRI

Differential perfusion and regurgitation of pulmonary artery ; does it matter?

- **It does matter !!**
  - Relationships between 2 ventricles and 2 great arteries !
  - Over-perfusion versus under-perfusion of branch PA

    ➔ poor development of hypo-perfused lung and chest

    pulmonary HTN, hemoptysis, etc
  - Excessive regurgitation from branch PA

    ➔ RV volume overloading ➔ RV failure

    ➔ LV failure, arrhythmias, sudden death, etc
- **Differential perfusion and regurgitation after TOF repair**
  - Depend on PA anatomy, pulmonary vascular resistance
  - Need multi-modality diagnostic tests : echocardiography, lung perfusion scan, CT, MRI, cardiac cath. etc.


Thank you for attention !