Significant PR without Significant Symptoms & Signs in repaired ToF case

: *Early intervention ?*

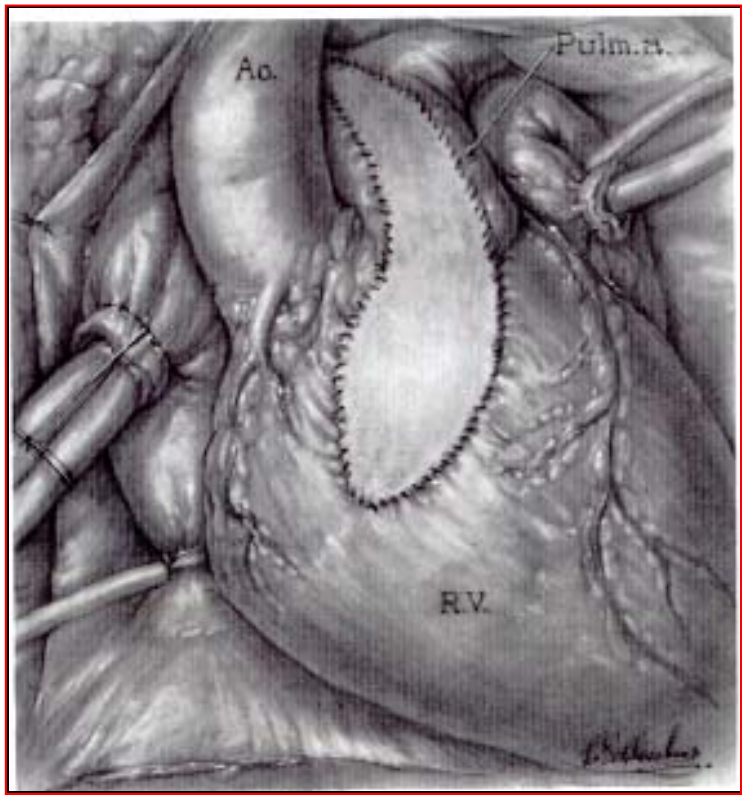
Pusan National University Yangsan Hospital
Siho Kim

Patient presenting with

- S/P ToF, 16 year-old
- RV volume 150ml/m2, EF 55%, PR fraction 40% on MRI
- no TR
- Non-sustained VT and/or PVCs on Holter monitor
- QRS duration on surface 12 leads ECG:150 ms
- NYHA class II, normal Ex CP test (one unproven episode of dizziness)

- ⇒ Severe PR without significant symptoms and signs
- Suggestion : Early PVR

ToF – Tale with a very long tail



- Residual lesions
- Sequellae : mainly, RV failure
  - Ventricular Dysfunction & Impairment Of Exercise Tolerance
  - Arrhythmia and Sudden Cardiac Death
  - **Pulmonary Regurgitation**

Indication of PVR

Indications for pulmonary valve replacement late after tetralogy of Fallot repair and associated surgical procedures in some of the most representative studies in the field

Study	Indication for PVR	Additional procedures
Bove et al. [17]	Conduit stenosis (n=3), symptoms (n=2), progressive cardiomegaly (n=3) and new onset of tricuspid insufficiency (n=3).	Not available
Warner et al. [24]	Diminished exercise tolerance and echocardiographic evidence of progressive PR with severe RV dilatation.	Pulmonary artery augmentation (n=6), closure of residual left to right shunts (n=3) and subendocardial resection for monomorphic ventricular tachycardia (n=1). Six patients had either preoperative or postoperative balloon dilations of pulmonary artery stenoses.
Yemets et al. [58]	Exercise intolerance, progressive RV dilatation with evidence of new onset TR or symptomatic ventricular tachycardia.	66% of the patients: VSD closure (n=13), cryoablation for sustained monomorphic VT (n=13), branch pulmonary angioplasty (n=30), miscellaneous (n=25).
d'Udekem et al. [57]	Exertional dyspnea, fatigue and syncope (six patients, four of which had sustained ventricular tachycardia)	Patch repair of a hypoplastic left pulmonary artery (n=1)
Conte et al. [47]	Cardiomegaly, significant to severe RV dilatation and dysfunction, fatigue, reduced exercise tolerance and ventricular tachyarrhythmia (10 patients).	VSD (n=10), ASD (n=2), PAPVD (n=1), branch pulmonary arterioplasty (n=4), tricuspid valve plasty (n=2), aortic valve plasty (n=1) and pacemaker insertion (n=1).
Eyskens et al. [59]	Severe pulmonary regurgitation with progressive signs of RV volume overload, which was assessed by increasing cardiomegaly on chest X-ray, progressive RV dilatation or worsening RV function as seen on echocardiography or angiocardiography, progressive tricuspid valve regurgitation, decreasing exercise tolerance or ventricular arrhythmias.	Not reported
Therrien et al. [62]	Significant pulmonary regurgitation with either (1) exercise intolerance, (2) progressive RV dilation (identified from serial transthoracic echocardiographic studies) or (3) clinical arrhythmia, with most patients (56%) fulfilling two or three criteria.	Aneurysm resection 40%, relief of RVOT obstruction 40%, tricuspid annuloplasty 20%, cryoablation for VT 12% and other miscellaneous.
Discigil et al. [64]	Decreased exercise tolerance in 58%, right heart failure in 21%, arrhythmia in 14%, syncope in 10% and progressive isolated right ventricular dilatation in 7%.	Tricuspid valve repair/replacement (n=18), residual VSD (n=12), ASD (n=4), pulmonary artery patch angioplasty (n=17) and RVOT enlargement (n=13).
Hazecamp et al. [65]	Symptomatic deterioration (fatigue and dyspnea at exercise), increase in right ventricular dimensions as measured with echo or MRI studies and/or progression of ventricular arrhythmias	Small residual VSDs 8, de Vega tricuspid annuloplasty 10, reduction of a dilated ascending aorta 2, resection of subaortic stenosis 2, atrial septal defect closure 1 and closure of a coronary artery fistula 1.
Vliegen et al. [66]	Objective evidence of important right ventricular dilatation, with or without deterioration of NYHA or the presence of tricuspid regurgitation (TR). Co-indications included the presence of (supra)ventricular arrhythmias, prolonged QRS duration (>180 ms) and a residual VSD.	Resection of infundibulum 8%, tricuspid valve repair 15%, closure of VSD 12% and closure of atrial septal defect (type II) 4%. Calcified outflow tract patch material was resected as much as possible.

- Uncertainty regarding
  - Indication
  - Timing for PVR
- Lack of reliable tools & guidelines

Indication of PVR

1. Patients with serious ventricular arrhythmias, when associated with severe PR and RV dilatation, with or without RV dysfunction.
2. Symptomatic patients with long-standing severe PR and RV dilatation with or without RV dysfunction.
3. Asymptomatic or symptomatic patients with moderate to severe PR and haemodynamically significant associated lesions that need surgical management.
4. ***Asymptomatic*** *patients with* ***severe PR*** *and evidence of* ***progressive RV dilatation and dysfunction*** *(ECHO,RNA,CMR) and/or* ***progressively diminishing exercise tolerance.***

Indication of PVR

4. ***Asymptomatic*** *patients with* ***severe PR*** *and evidence of* ***progressive RV dilatation and dysfunction*** *and/or* ***progressively diminishing exerc ise tolerance.***

- Risk in asymptomatic patients

  Heart 2000;84:416-420, Semi thorac cardio surg2005;17:155-9

  - RV end-diastolic volume ≥ 170 (*or 120*) mL/m^2

    Am J Cardiol 2005;95:779-782

  - QRS duration ≥ 180 msec — *Am J Cardiol 1997; 80:160-163*
  - Increased rate of change in QRS duration (≥ 3.5 msec/yr)

    Lancet 2000;356:975-981

  - RV end-systolic volume (ESV) ≥ 85 (*or 95*) mL/m^2

    Am J Cardiol 2005;95:779-782

  - NYHA II or higher — *Circulation 2002;106:1703-1707*

Indication of PVR

- **Risk** in asymptomatic patients
  - RV end-diastolic volume ≥ 170 (*or 120*) mL/m^2
  - QRS duration ≥ 180 msec
  - Increased rate of change in QRS duration (≥ 3.5 msec/yr*)*
  - RV end-systolic volume ≥ 85 (*or 95*) mL/m^2
  - NYHA II or higher

Should do something, BEFORE ~

- RV end-diastolic volume = 170mL/m^2
- QRS duration =180 msec
- Increased rate of change in QRS duration (>3.5 msec/yr*)*
- RV end-systolic volume mea sured **reaches** 85 mL/m^2

Tail of Sequelae

- **Ventricular Dysfunction & Impairment Of Exercise Tolerance**
  - Exercise tolerance was improved after PVR
  - Evaluation of symptoms alone does not reflect the functional RV derangement *Circulation 1995;91:1775-81*
  - Potential for RV recovery after PVR might be compromised in adult patients *JACC 2000;36:1670-5, ATS 2002;73:1794–800*
  - Waiting for symptoms to appear may allow irreversible RV dysfun ction to occur and result in minimal benefits from PVR *Int J Cardiol 2004;97 Suppl 1:91-101*
- Plus, The later PVR, the greater chance of RVOT procedure that contributes to poor RV recovery *Int J Cardiol 2004;97 Suppl 1:91-101* *Circulation. 2002;106:1703-1707*

Tail of Sequelae

by Cardiac MR study

In adult patients with PR and RV dilatation,
We advocate a less restrictive management concerning PVR in these patients.

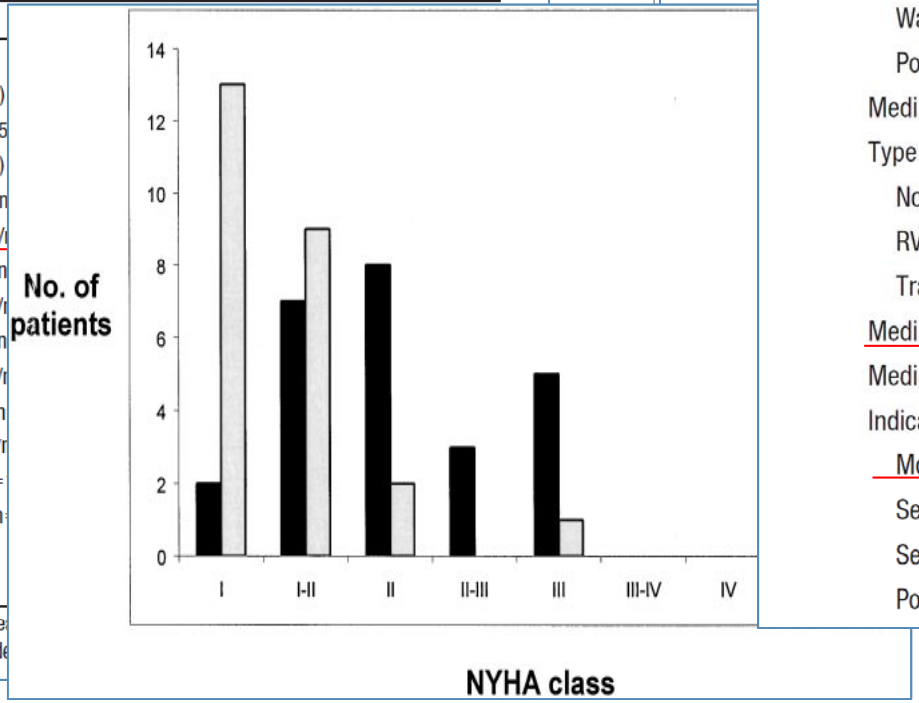
TABLE 1. Demographic and Surgical Characteristics: Total Cohort of Patients

Variable	(n=26)
Male sex	15 (58)
Previous palliative shunt	
Blalock Taussig	7 (27)
Waterston	3 (12)
Potts	1 (4)
Median age at initial repair, y	5.0±4.2
Type of repair	
No patch	10 (38)
RV patch	6 (23)
Transannular path	10 (38)
Median age at PVR, y	29.2±9.0
Median duration of follow-up, mo	7.4±2.4
Indications for PVR	
Moderate PR (20% to 40%)	11 (42)
Severe PR (>40%)	15 (58)
Severe RV dilatation (RV/LV>2)	13 (50)
Poor validity	10 (38)

TABLE 2. MRI and Clinical Data Before and After PVR

Variables	
MRI	
PR, % (n=25)	
PR, mL (n=25	
TR, % (n=22)	
RV-EDV, mL (n	
RV-EDV-I, mL/	
RV-ESV, mL (n	
RV-ESV-I, mL/	
LV-EDV, mL (n	
LV-EDV-I, mL/	
LV-ESV, mL (n	
LV-ESV-I, mL/	
RV-EF, % (n=	
$RV\text{-}EF_{cor}$, % (n	
Clinical	
NYHA (n=26)	

Values are me
I indicates inde



Tail of Sequelae

- **Arrhythmia and Sudden Cardiac Death**
  - Holter monitoring is not helpful for identification of patients with risk *JACC 1994;23:1151-5 (12yr f/u), Lancet 2000;356:975-981*
  - **QRS duration stabilized** (it continued to increase in a control group with similar follow-up duration), **but failed to decrease** as one would have hoped. It is likely that patients *would benefit more if valve replacement were performed before the onset of symptoms.* *Semi thorac cardio surg 2005;17:155-9*

Tail of Sequelae

- **Pulmonary Regurgi**
  - Pulmonary regurgita
    lesion for patients w
  - None of the patients
    ns *(PVR)*, implying a
  - **At least moderate P**
    enosis, was the culpr
    ped SVT. Furthermor
    dden death.

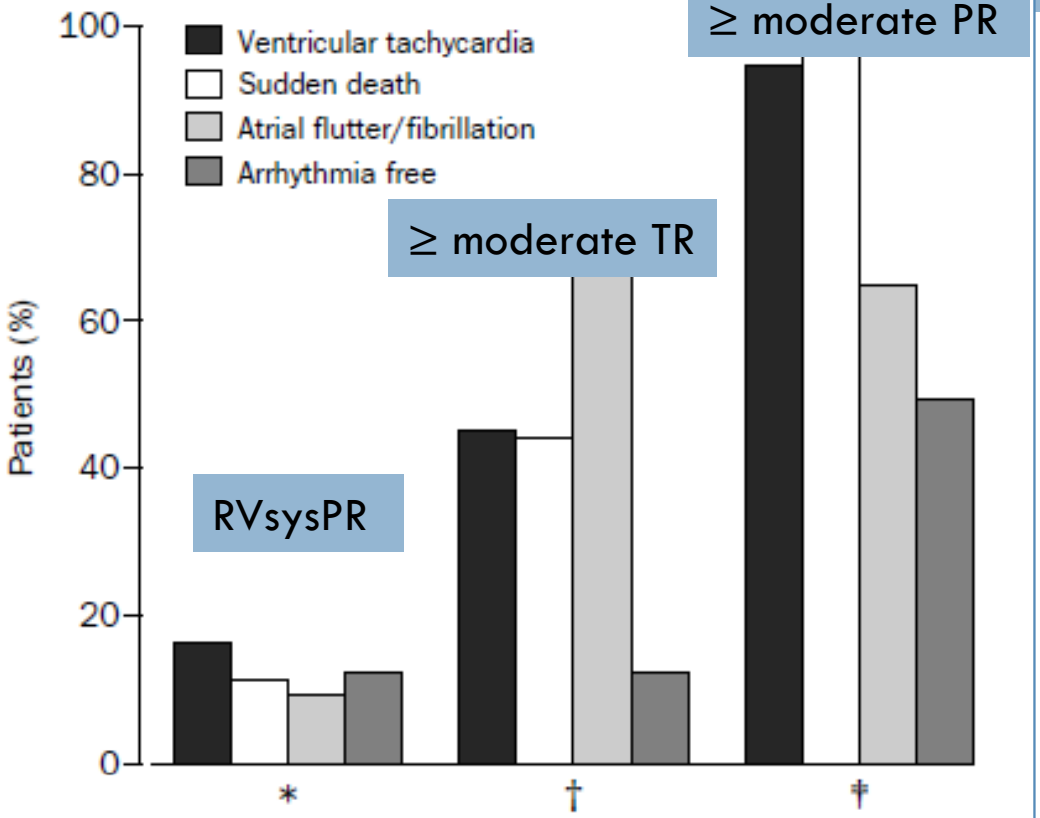


Figure 3: **Echocardiographic data from 456 patients late after repair of tetralogy of Fallot**

Data obtained during preceding 12 months from final patient assessment in 1995, or before occurrence of arrhythmia and sudden death.

*Right-ventricular systolic pressure (>60 mm Hg). †Tricuspid regurgitation ⩾moderate). ‡Pulmonary regurgitation (⩾moderate)

Inconstant value of Tx tool over time

- *Ex) ASD device closure*
  - Advance in Technology
  - Support from Society or System (Healthcare System)
  - Patient's preference – Life style or Occupation

Indication of PVR

- On How Bad ?
- On When Bad ? : *Timing*
  - *Tail of tale : propensity, not the cross-sectional results*

On When Bad ? : *Timing*

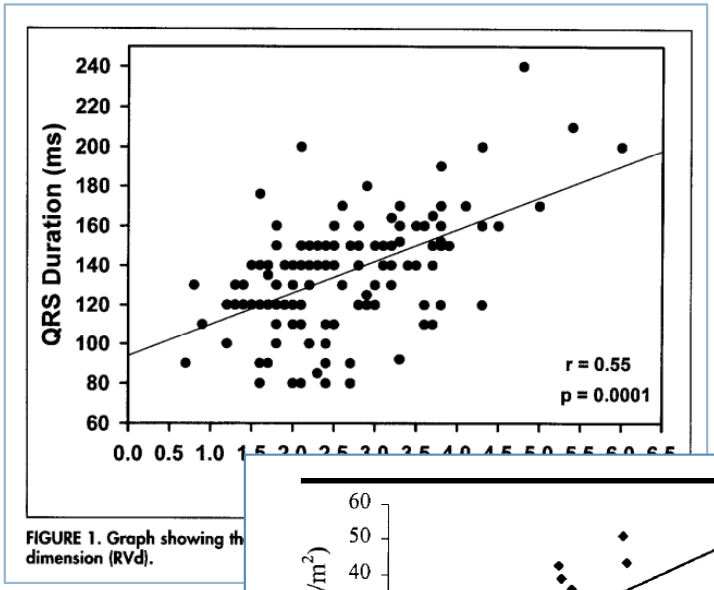


FIGURE 1. Graph showing th
dimension (RVd).

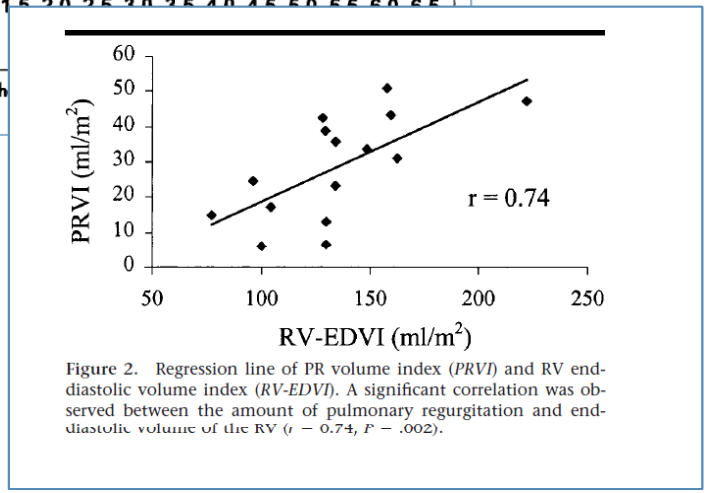
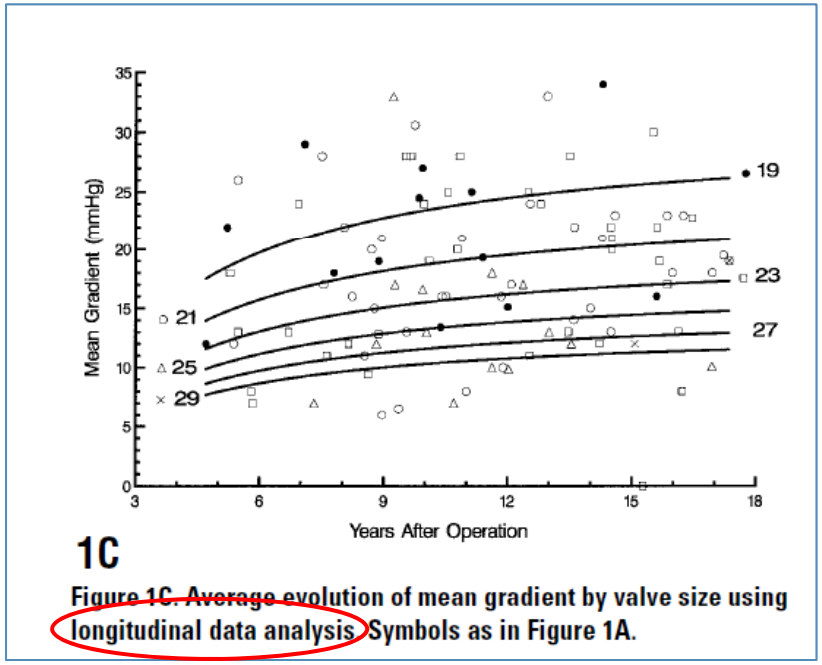


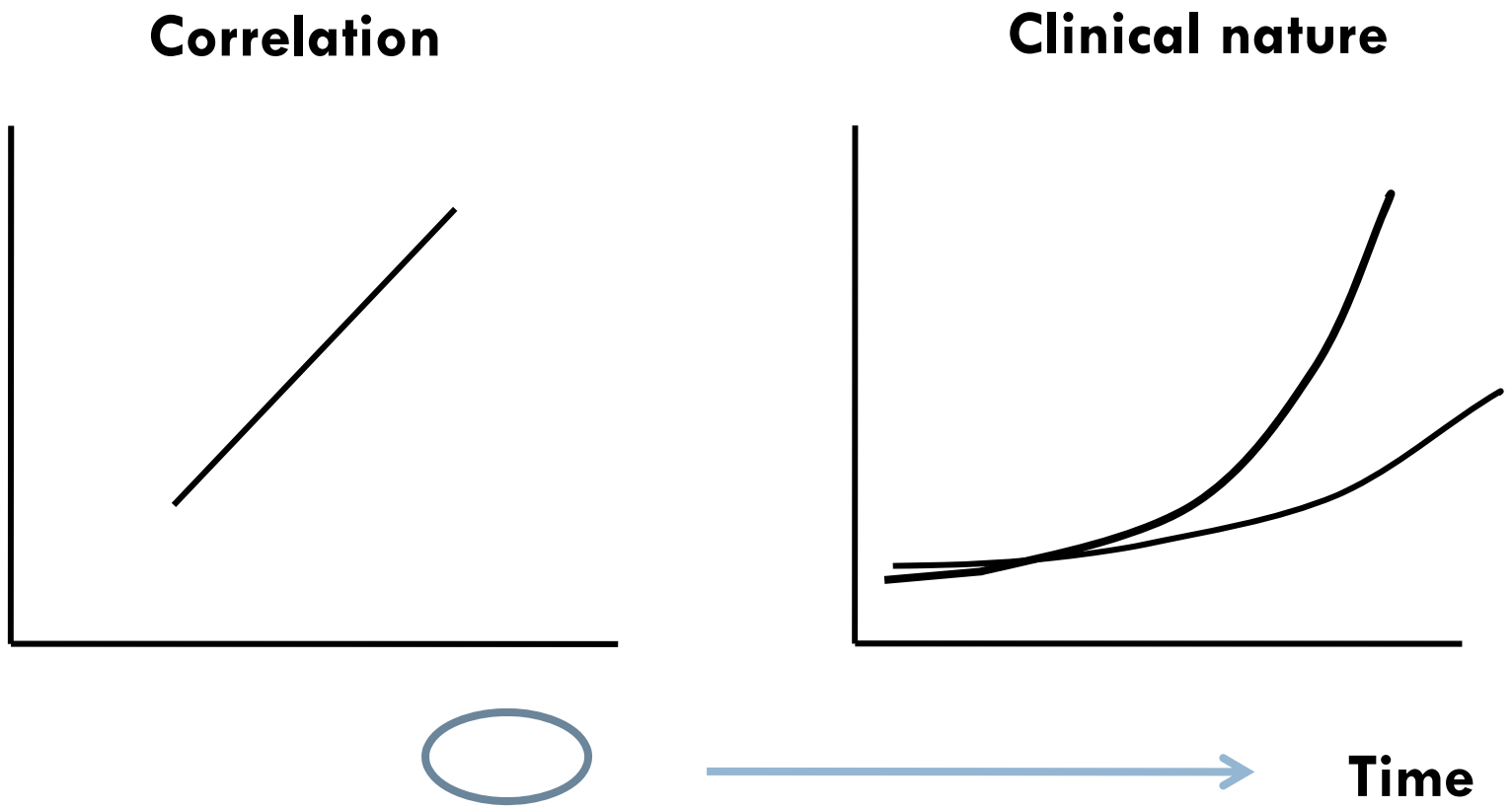
Figure 2. Regression line of PR volume index (*PRVI*) and RV end-diastolic volume index (*RV-EDVI*). A significant correlation was observed between the amount of pulmonary regurgitation and end-diastolic volume of the RV ($r = 0.74$, $P = .002$).



1C

Figure 1C. Average evolution of mean gradient by valve size using longitudinal data analysis. Symbols as in Figure 1A.

On When Bad ? : *Timing*



More clues for making a decision

- **Serial trend on clinical information**
- **OP date and Days from the operation**
- **OP tech – Annular preservation or Transannular patch ?**
- **Other pathologic lesion in RV**
- **Tissue Doppler**
- **MRI :**
  - Volume/mass ratio
  - RV end-systolic & end-diastolic volume
- **EKG :**
  - Rate of change of QRS duration
  - Inhomogeneity of repolarisation
  - Microvolt T wave alternans
- **Gene expression**
- **Occupation or wishful life style**

Suggestion : *Early PVR*

- **More clinical reports** were required
  - Well-designed, cumulative data
  - Cardiac MR
- Don't get the degree of hemodynamic deterioration to **progress beyond the capacity** for recovery after intervention.
  - Vigilant F/U on asymptomatic or minimally symptomatic patients
- **More liberal approach with earlier timing of PVR** before symptoms associated with the hemodynamic substrate ensue, which is depends on
  - The quality of healthcare system of each society
  - How vigilant the patient follow-up programs are.
- **?** : simply **reducing RV** size is "good enough" vs. **reaching a normal RV** size on long-term morbidity and mortality