



TURIN  
October  
24<sup>th</sup>-26<sup>th</sup>  
2019

## 31 GIORNATE CARDIOLOGICHE TORINESI

*Everything you always  
wanted to know about*  
Cardiovascular Medicine

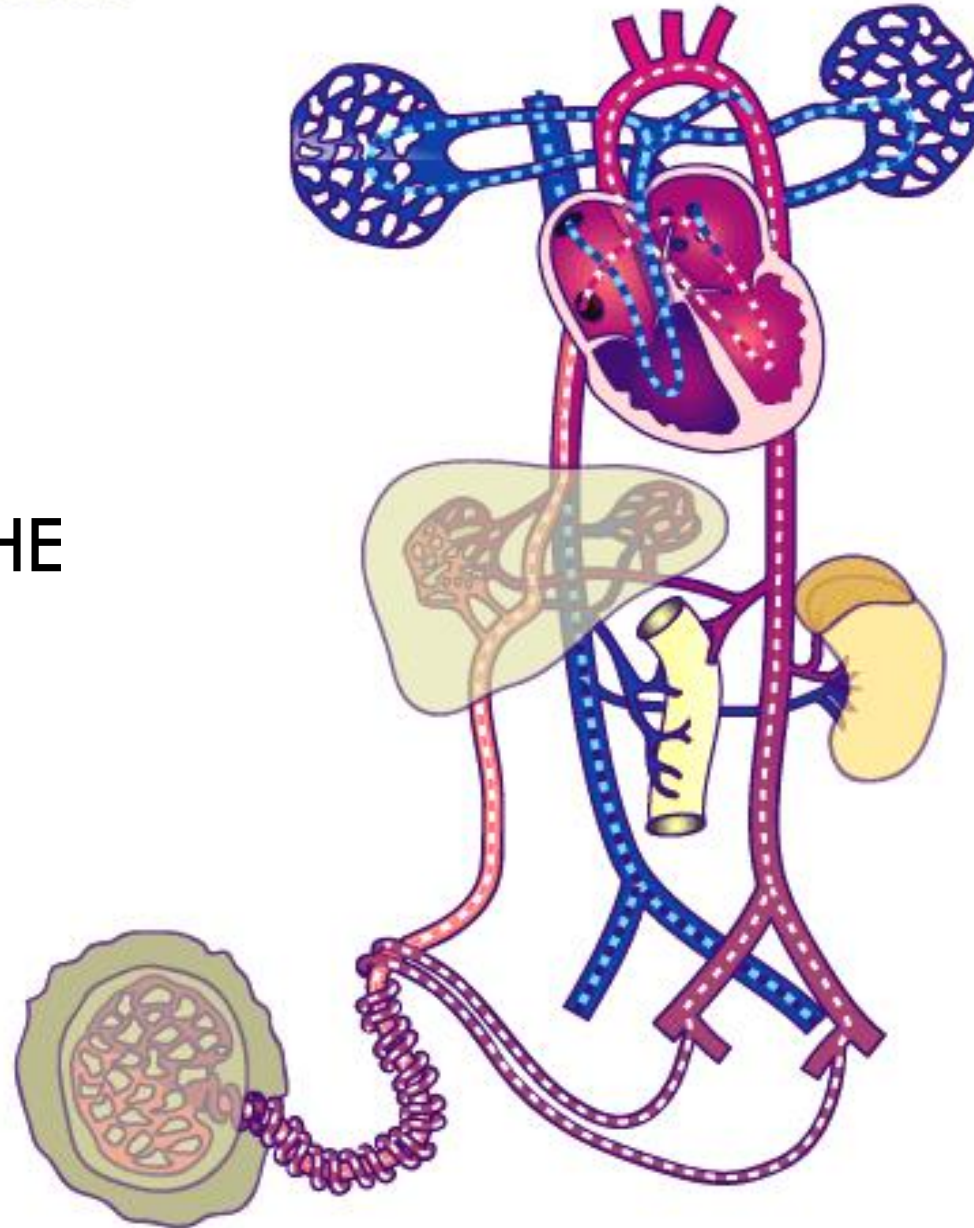


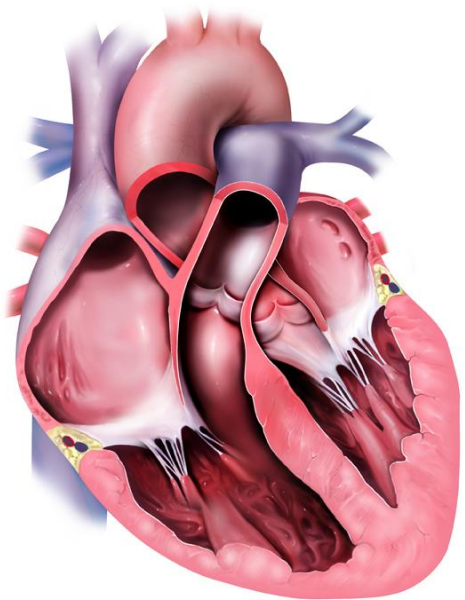
# SPECTRUM OF CARDIOVASCULAR AND PULMONARY INVOLVEMENT IN GROWN UP CONGENITAL HEART PATIENTS

G AGNOLETTI

before birth

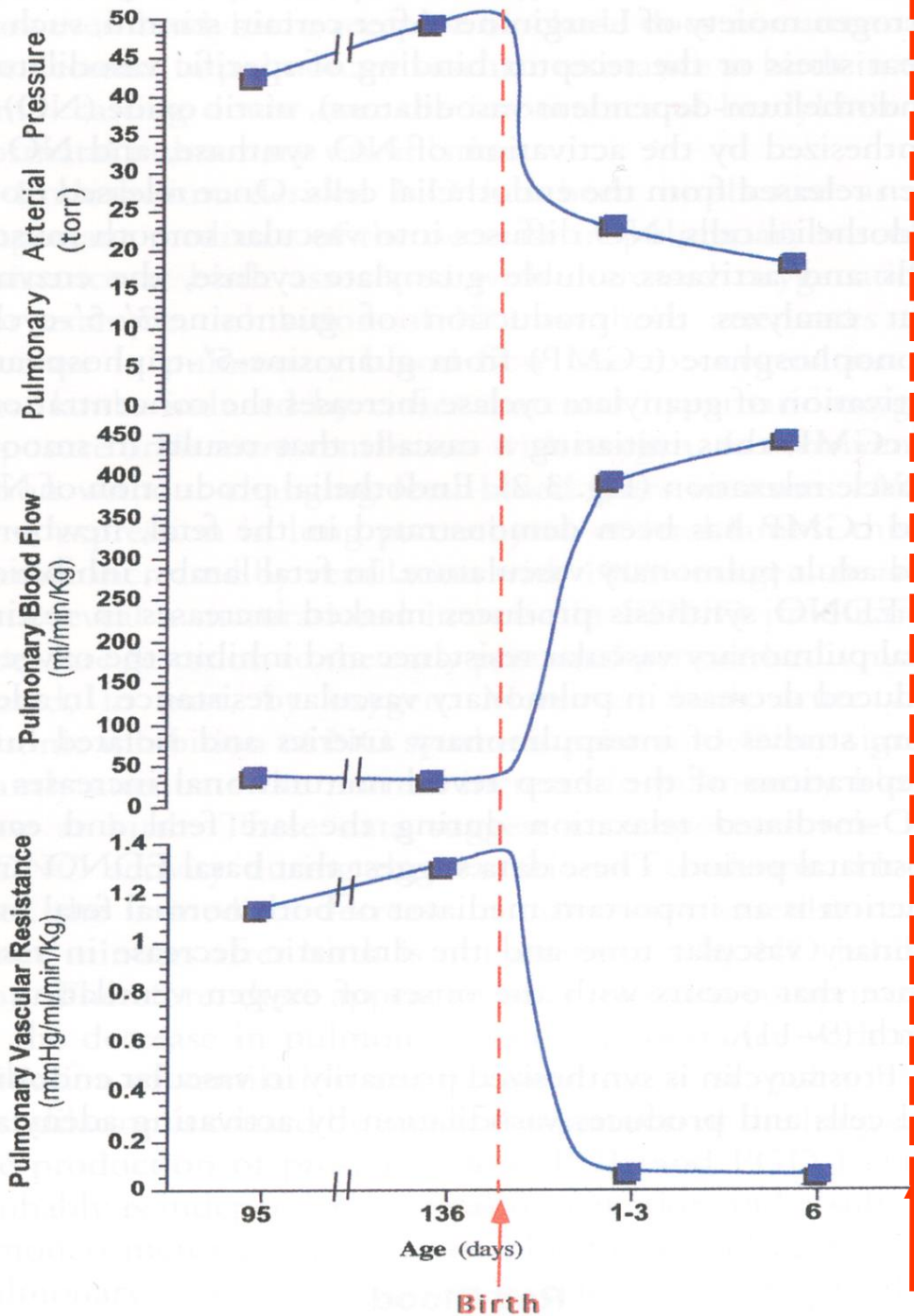
JULE VERNE  
LET'S START FROM THE  
BEGINNING...





Normal Heart

AT BIRTH

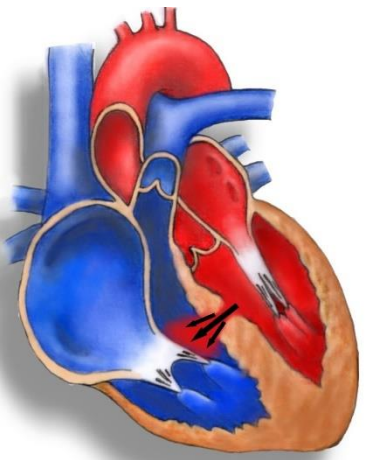


PRESSURE

FLOW

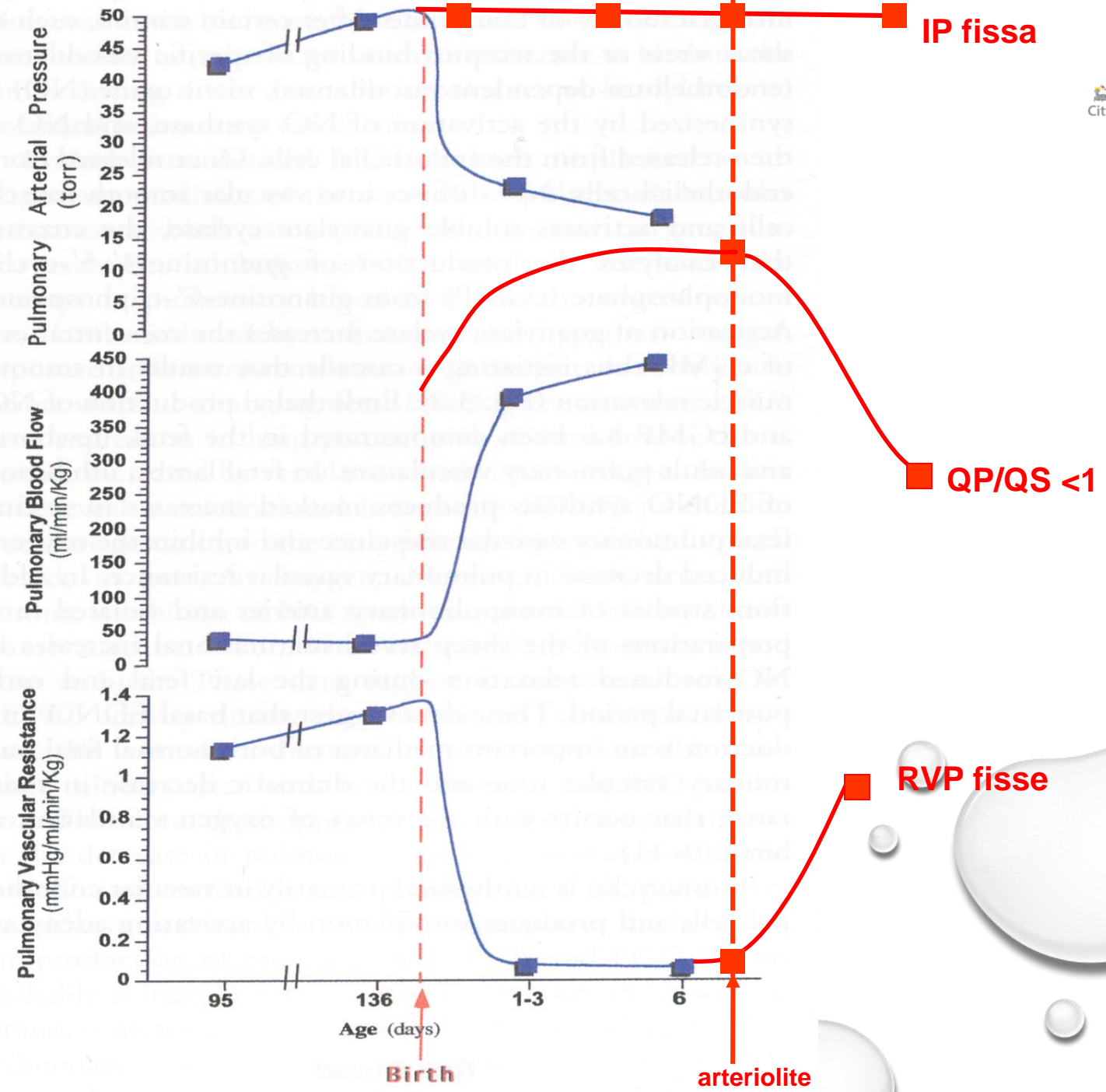
PVR

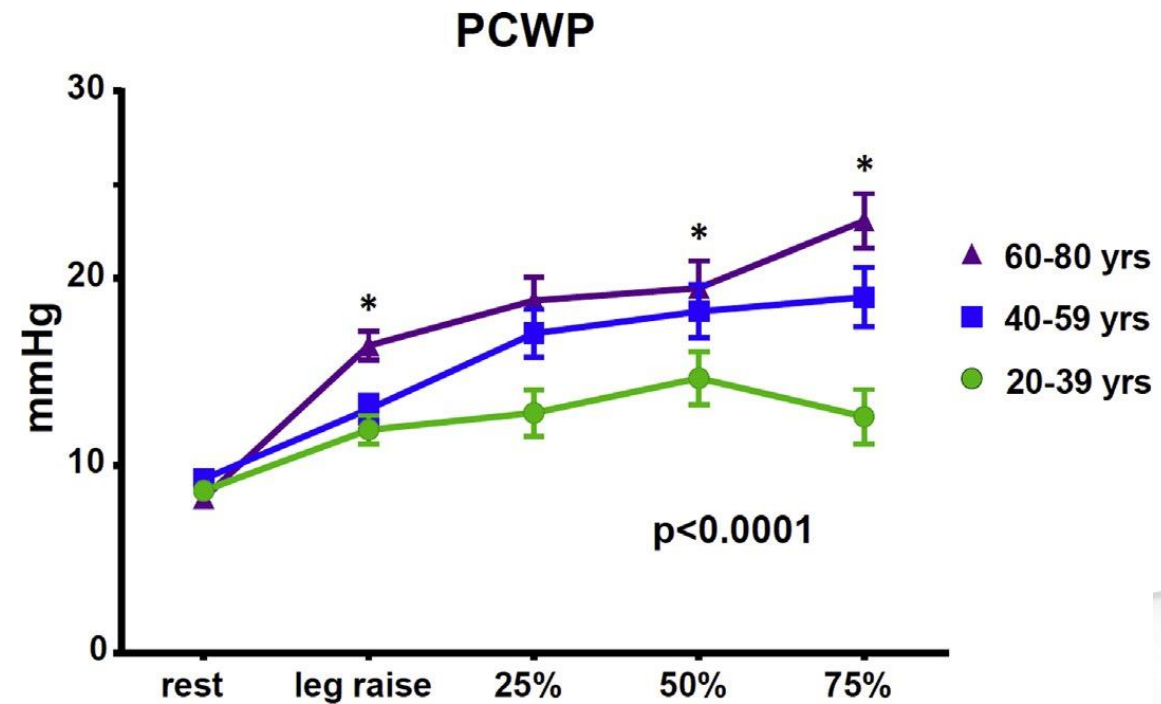
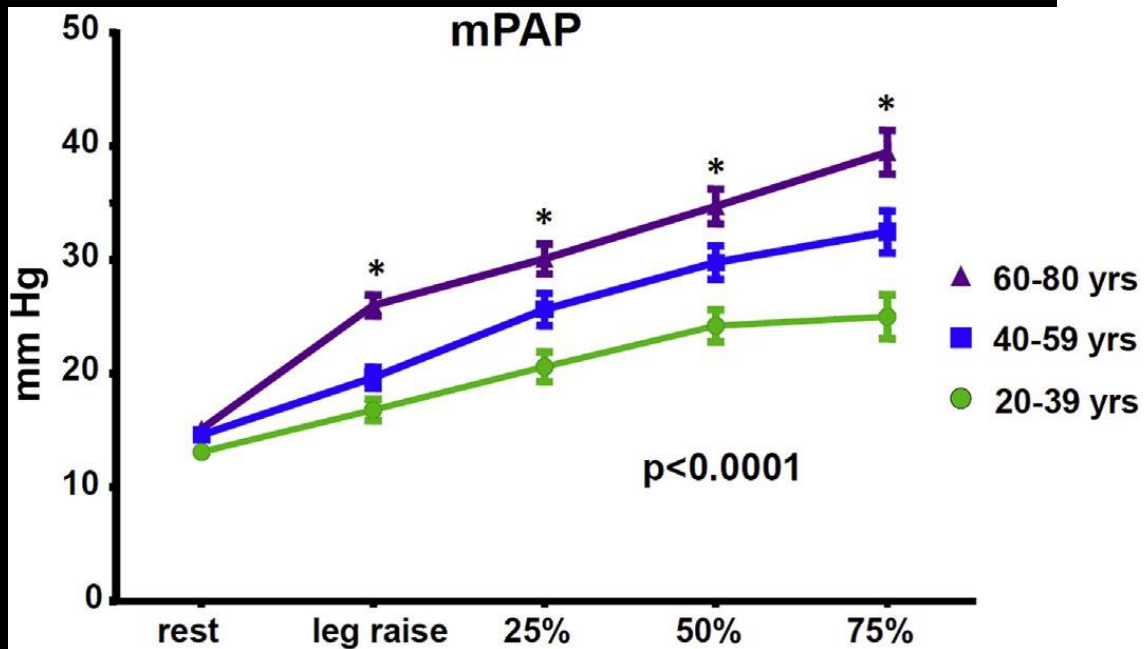
Shunts



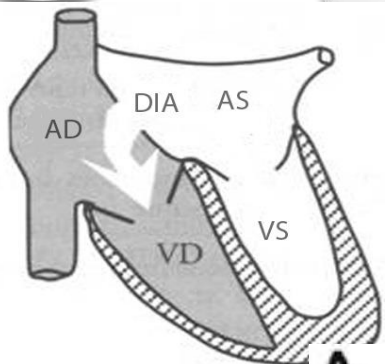
Normal Heart

AT BIRTH

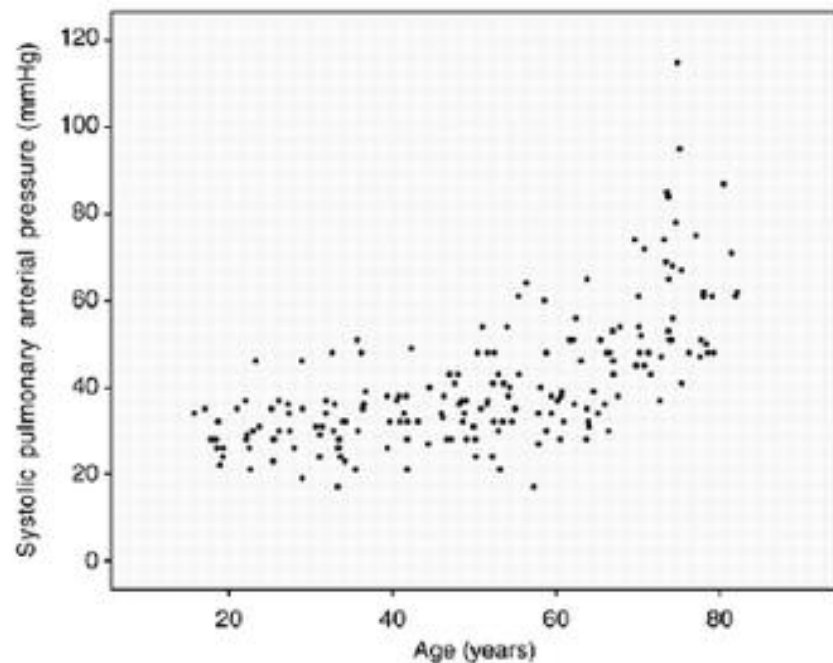
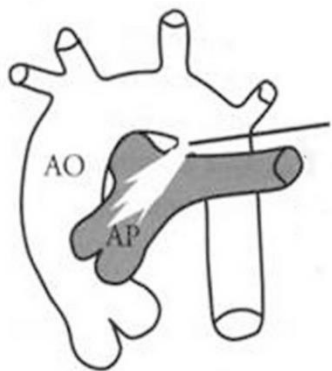
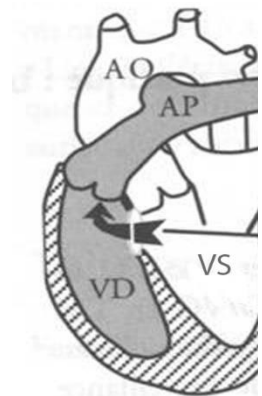




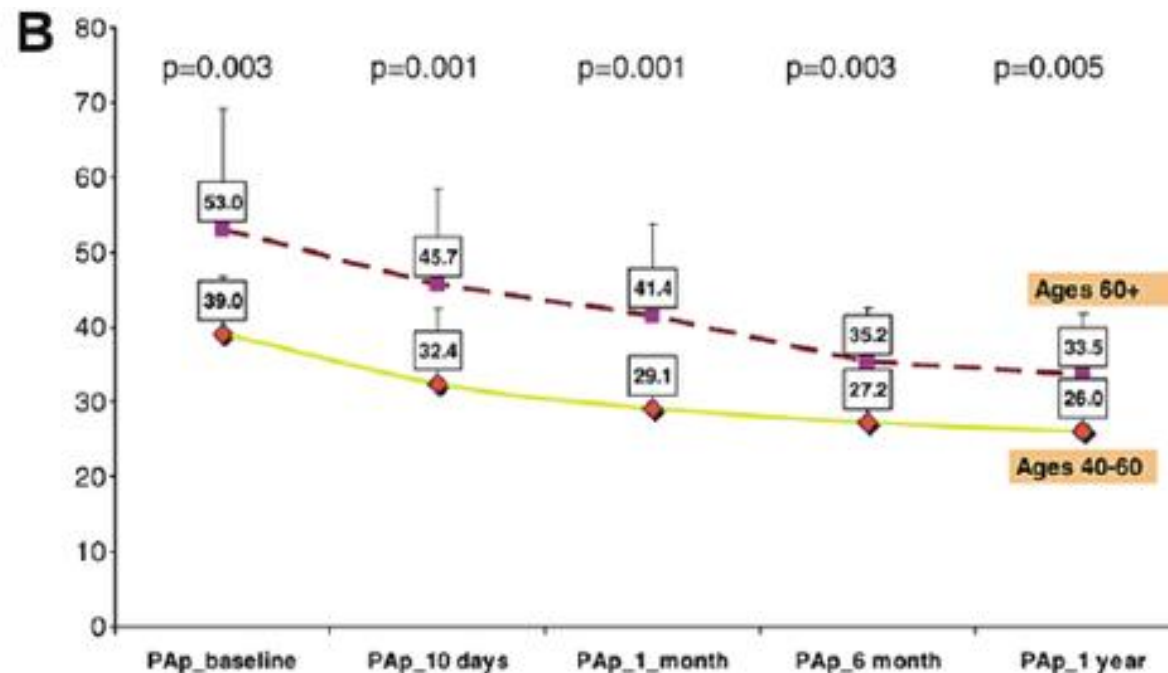
Cardiac AND LUNG aging was  
 progressive without sex differences  
 in healthy participants



**A**



**After closure**

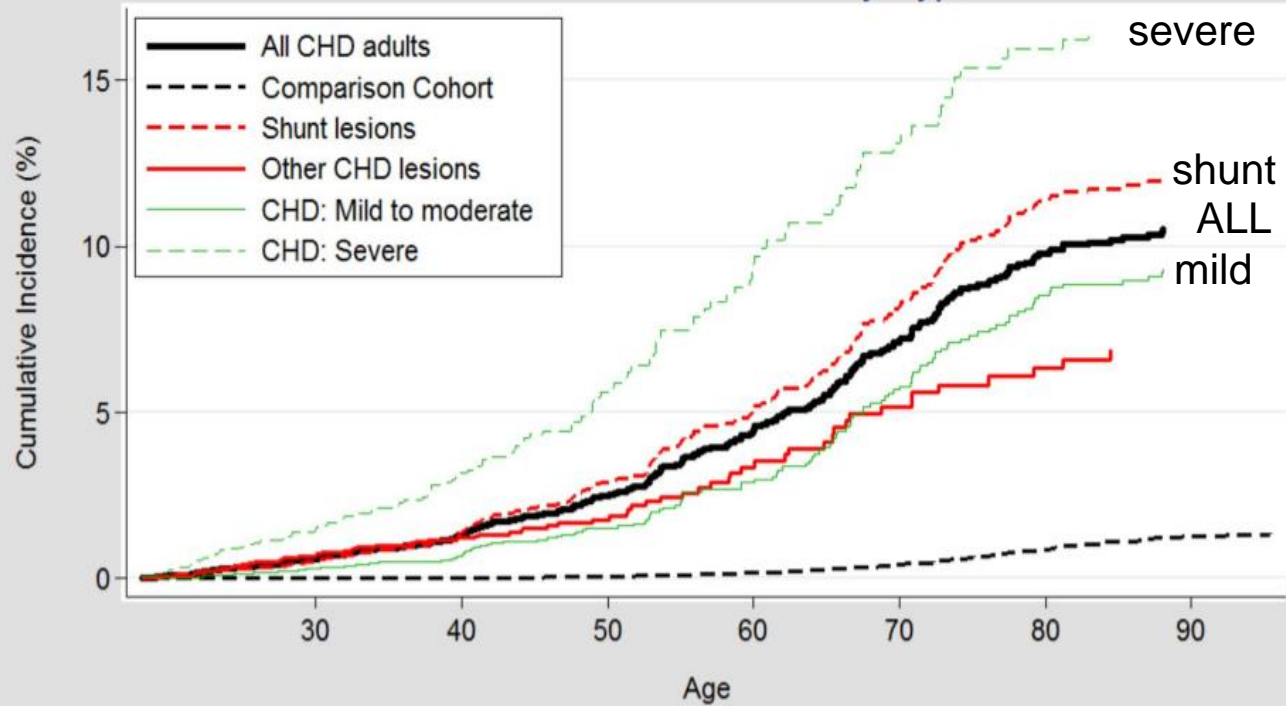


Relationship between pulmonary artery pressure and age in patients with **secundum ASD**

# PAH-CHD

## Incidence

Cumulative incidence of Pulmonary Hypertension



## Mortality

Mortality Rate (Kaplan-Meier)

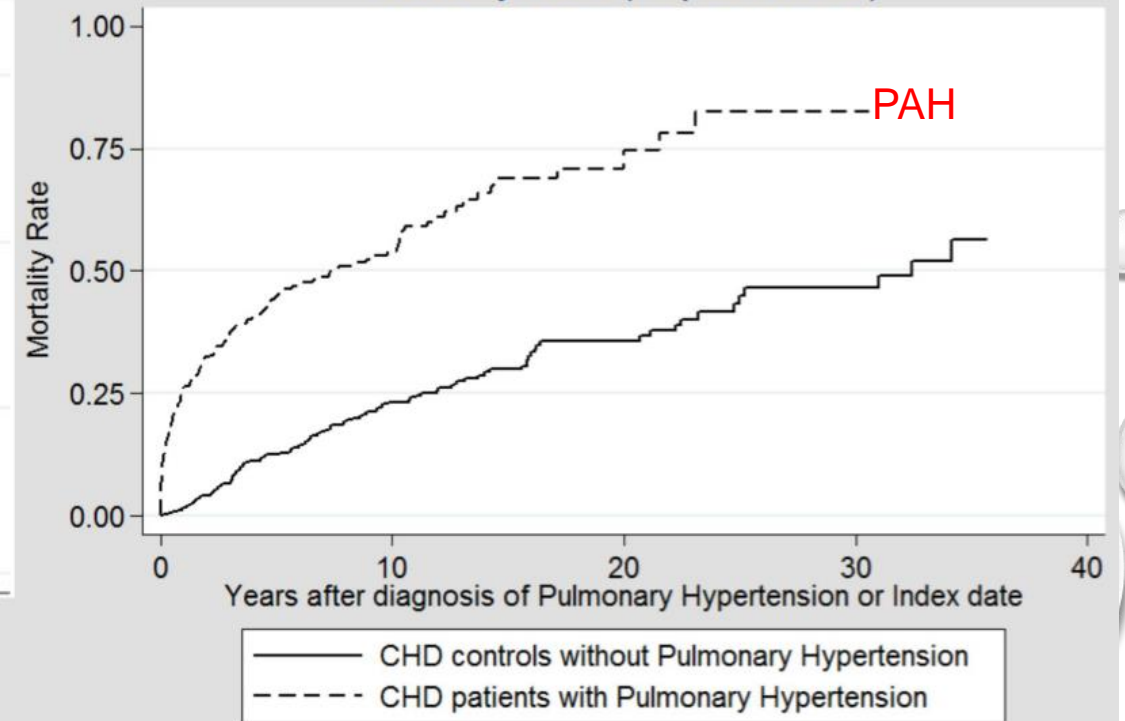
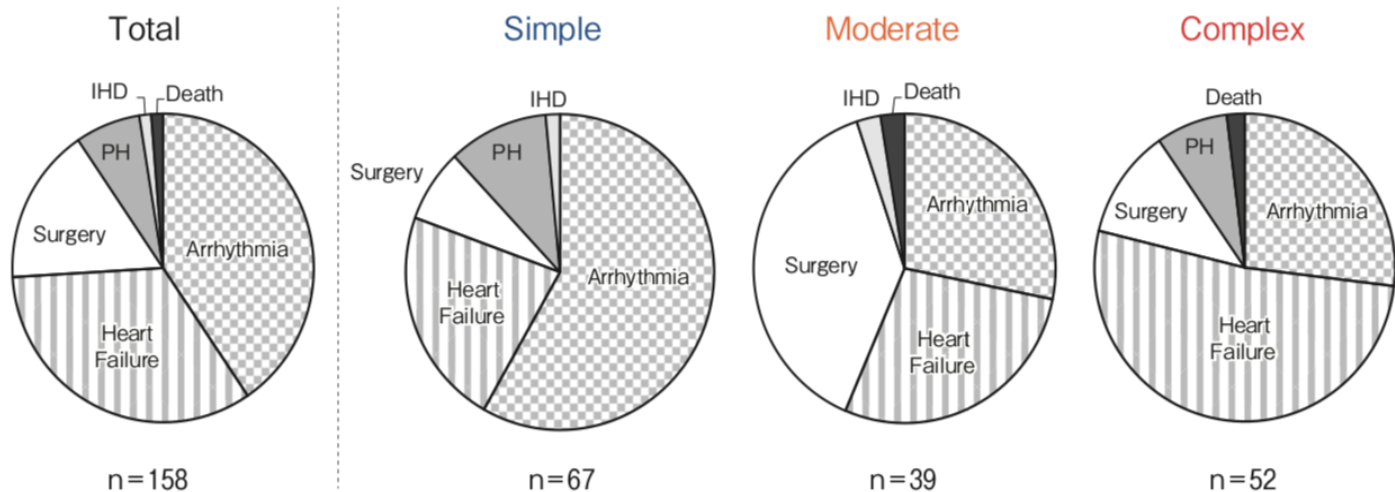


Figure 1 Cumulative Incidence of Pulmonary Hypertension.tiff

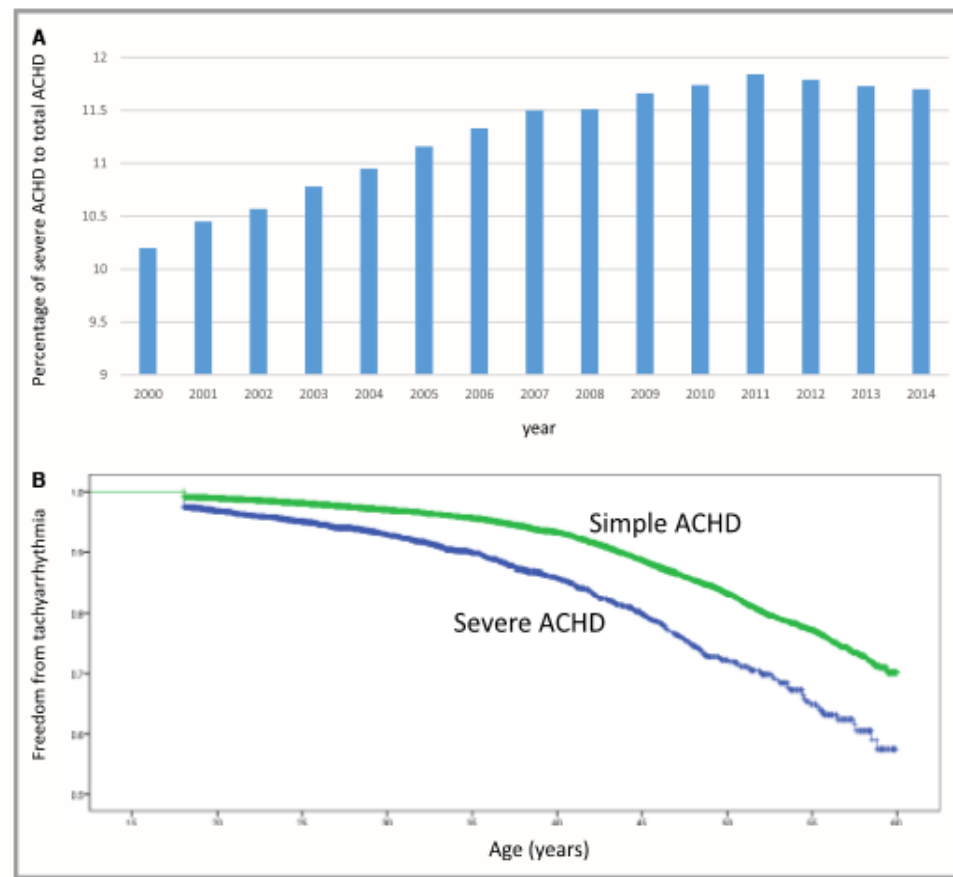
# Increase of patients with complex CHD

Adult CHD: Clinical Spectrum and the Trend Wu et al



**Fig. 2** Causes of hospitalization. The causes of hospitalization depending on ACHD complexity are shown. Transcatheter intervention was the most common cause in the Simple group, with surgery the most common cause in the Moderate group, and heart failure in the Complex group. IHD, ischemic heart disease; PH, pulmonary hypertension.

Freedom from tachyarrhythmia



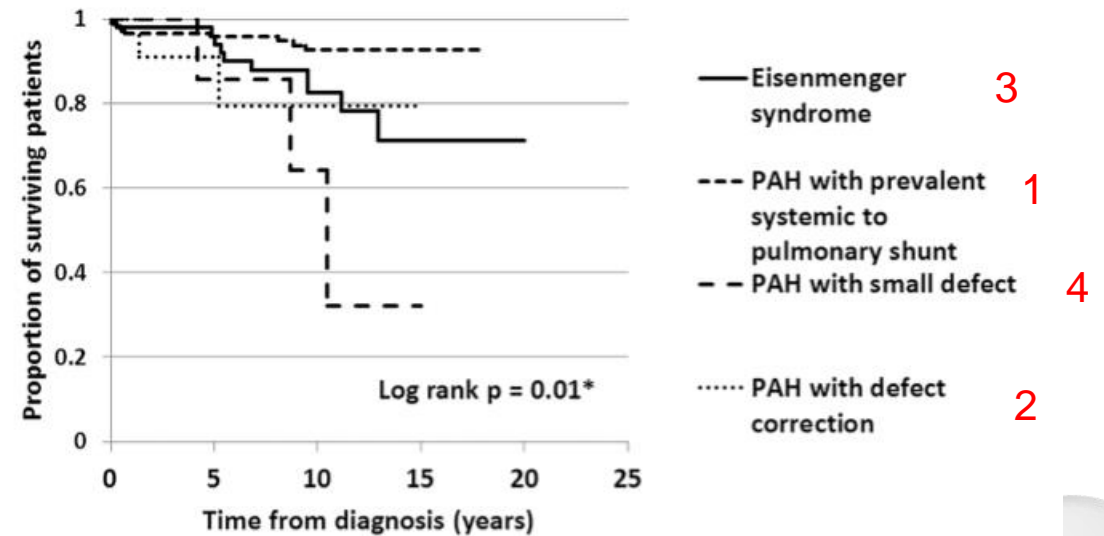
**Figure 1.** A, Percentage of severe adult congenital heart disease in the whole adult congenital heart disease cohort from 2000 to 2014. B, Freedom from tachyarrhythmia in simple adult congenital heart disease (simple ACHD) and severe adult congenital heart disease (severe ACHD) cohorts.



# PAH-CHD

## Survival

○ PAH-CHD: 70-90% at 25 yrs



Number at risk				
98	49	29	4	Eisenmenger syndrome
244	138	75	10	PAH with prevalent systemic to pulmonary shunt
11	6	3	0	PAH with small defect
13	8	3	0	PAH with defect correction

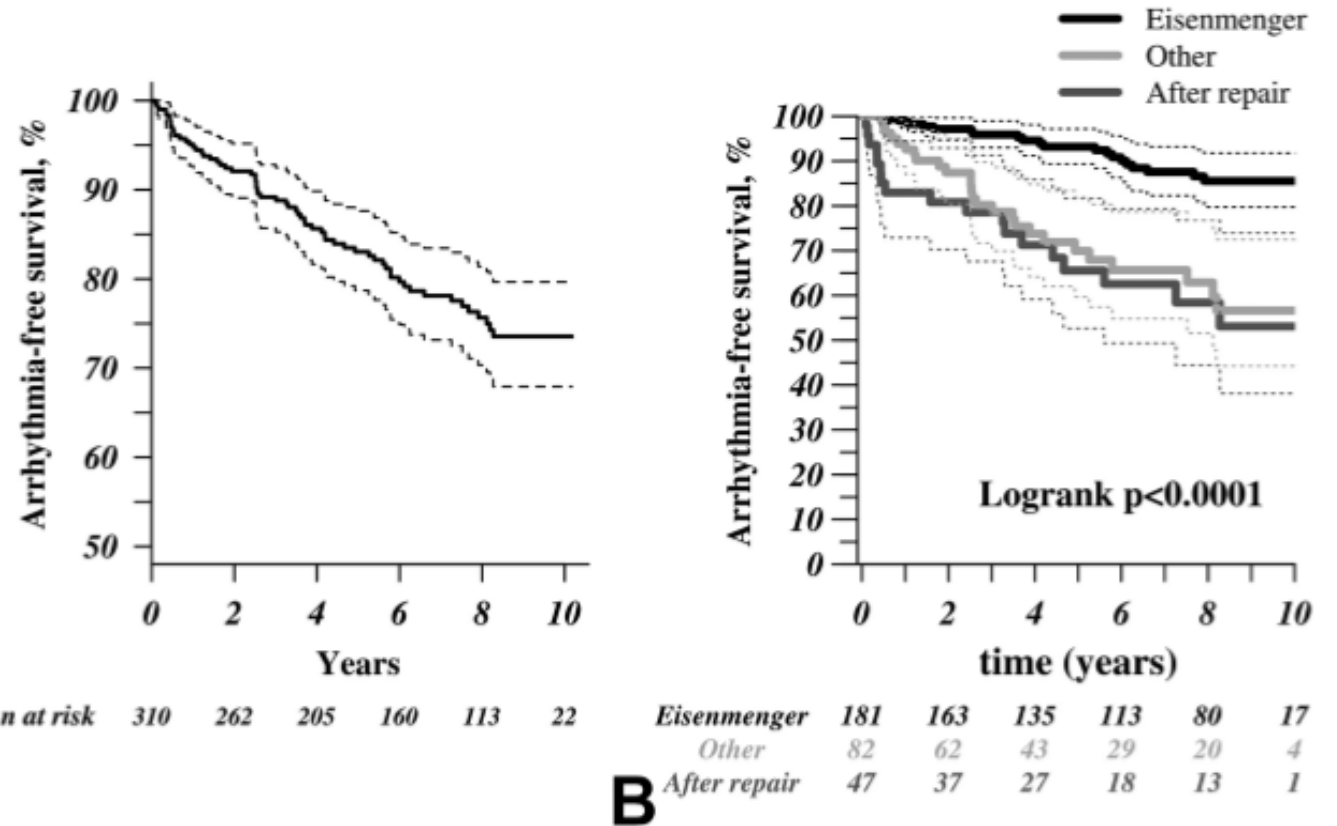
Fig 3. Cumulative survival of all PAH-CHD patients with different clinical classifications: 1) Eisenmenger syndrome (solid line), 2) PAH with prevalent systemic to pulmonary shunt (---), 3) PAH with small defect (-.-) and 4) PAH after defect correction (...).

# NOT ONLY PAH

- PAH / arrhythmia
- PAH / heart failure
- PAH / iron - PLT
- PAH / polypharmacy ...

Hospitalisations for heart failure predict mortality in pulmonary hypertension related to congenital heart disease

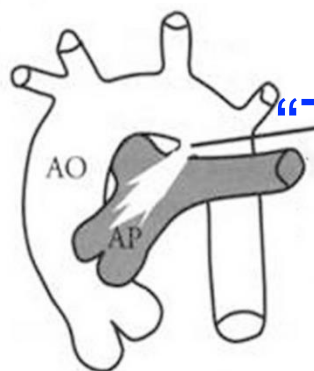
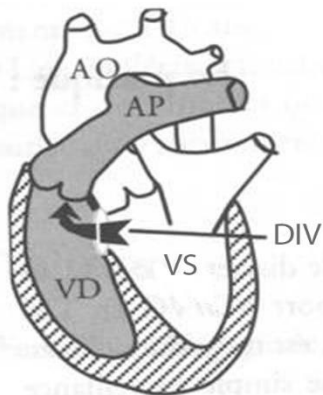
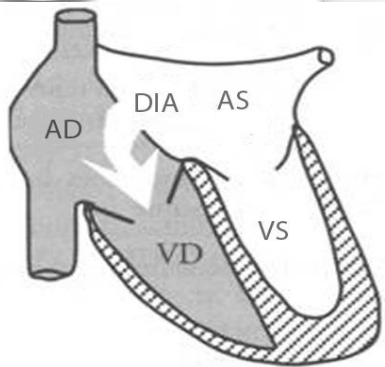
Despoina Ntiloudi,<sup>1</sup> Sotiria Apostolopoulou,<sup>2</sup> Konstantinos Vasiliadis,<sup>3</sup>  
 Alexandra Frogoudaki,<sup>4</sup> Aphrodite Tzifa,<sup>5</sup> Christos Ntellos,<sup>6</sup> Styliani Brili,<sup>7</sup>  
 Athanasios Manginas,<sup>8</sup> Antonios Pitsis,<sup>9</sup> Marios Kolios,<sup>10</sup> Haralambos Karvounis,<sup>1</sup>  
 Costas Tsioufis,<sup>7</sup> John Goudevenos,<sup>10</sup> Spyridon Rammos,<sup>2</sup> George Giannakoulas,<sup>1</sup> on  
 behalf of the CHALLENGE investigators



**Figure 2** The incidence of arrhythmia in the overall pulmonary arterial hypertension with congenital heart disease (PAH-CHD) population (A) and PAH-CHD subgroups (B). The 'other' group contains patients with PAH and left-right shunts as well as patients with PAH-CHD and small (coincidental) defects.

## Beneficial effects of percutaneous ASD closure in adults.

Definite effects of ASD closure	Possible Effects of ASD closure
RV size ↓	Atrial arrhythmias ↓
LV size ↑	
PA pressure ↓	
RA size ↓	
Exercise capacity ↑	
NYHA class ↓	



« Pulmonary artery pressure improves after ASD closure regardless of patient age ».

“Not too old to be closed.....”

“The benefit of atrial septal defect closure in elderly patients”

Congenit Heart Dis. 2009 Am J Cardiovasc Dis. 2012, Neth Heart J. 2010.....

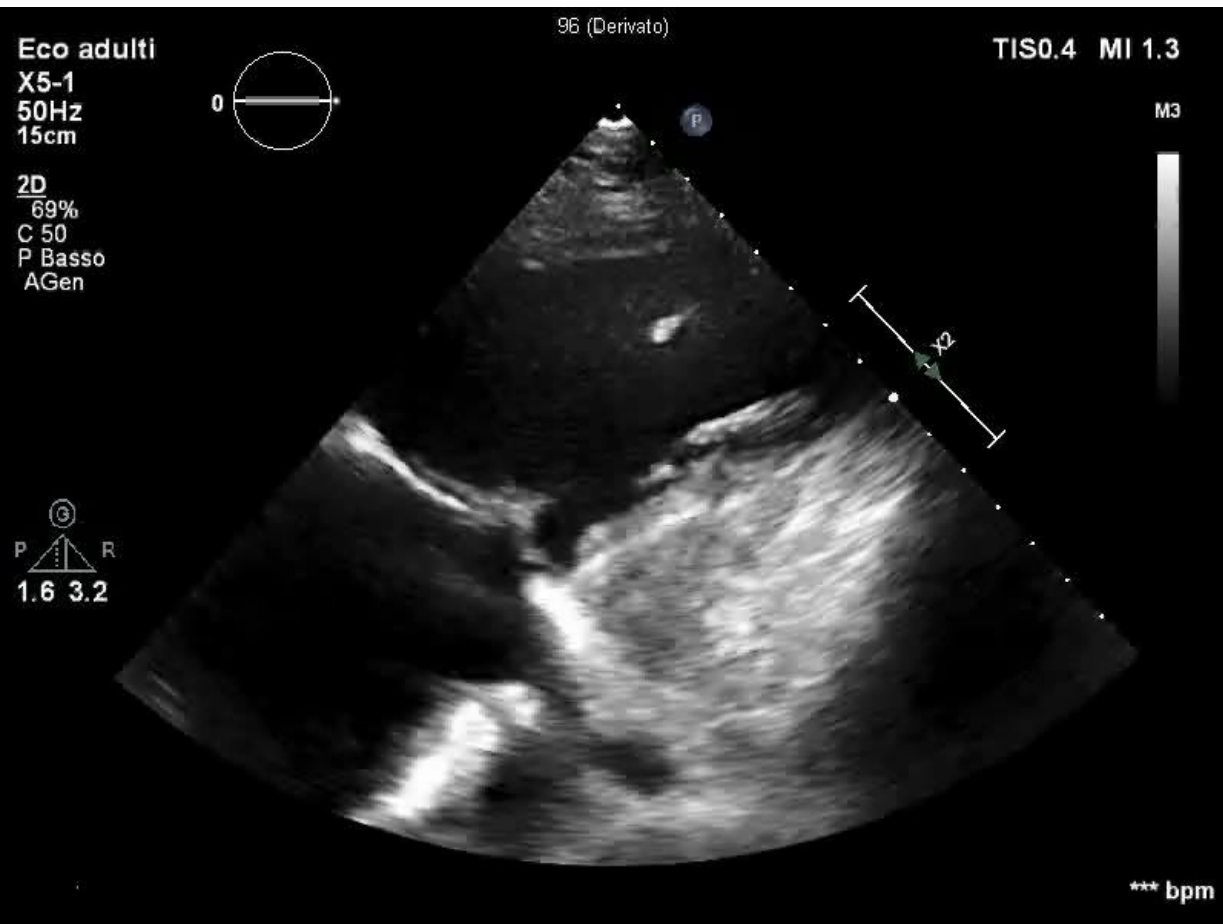


- Giuseppe aa 73
- 2003 (67aa) chiusura percutanea ASD (per evitare l'insorgenza di FA)
- 2019: FA, iperteso, displasia polmonare con steno-insufficienza, VD dilatato +++

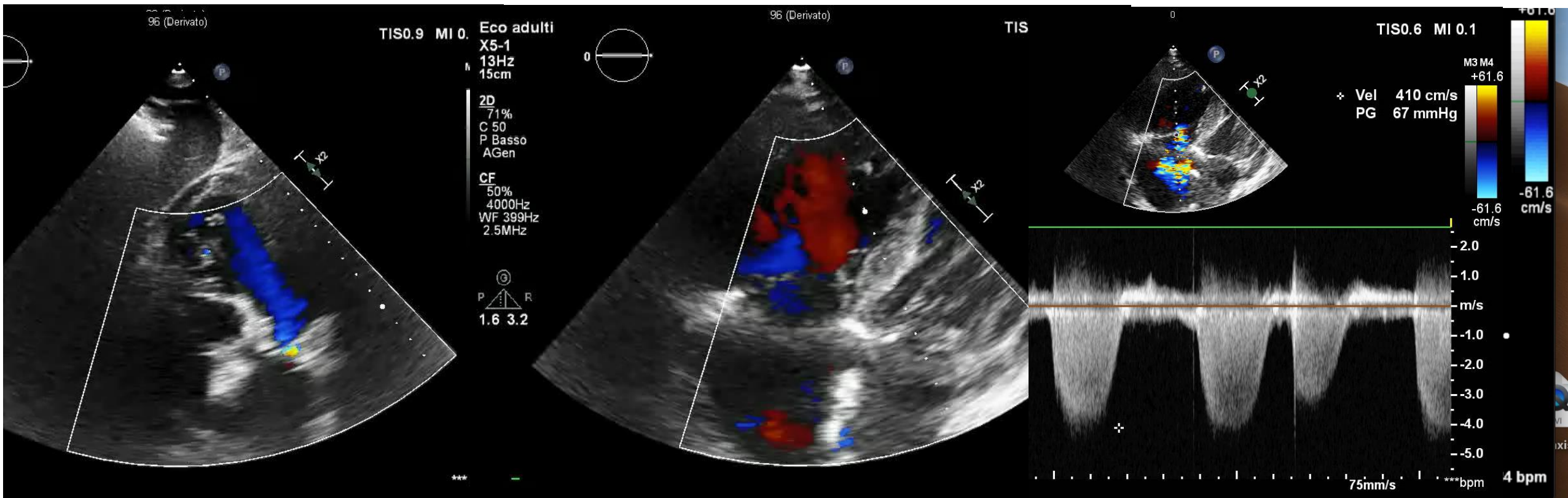
**INDICAZIONE A  
VALVOLAZIONE  
POLMONARE**

# ECO PRE VALVOLAZIONE

## II VD

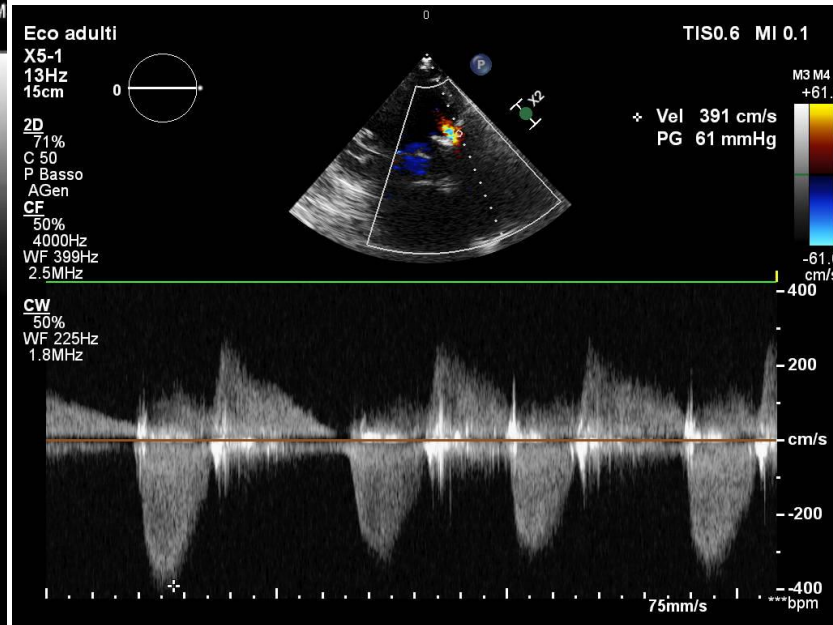


# ECO PRE VALVOLAZIONE IP - IT

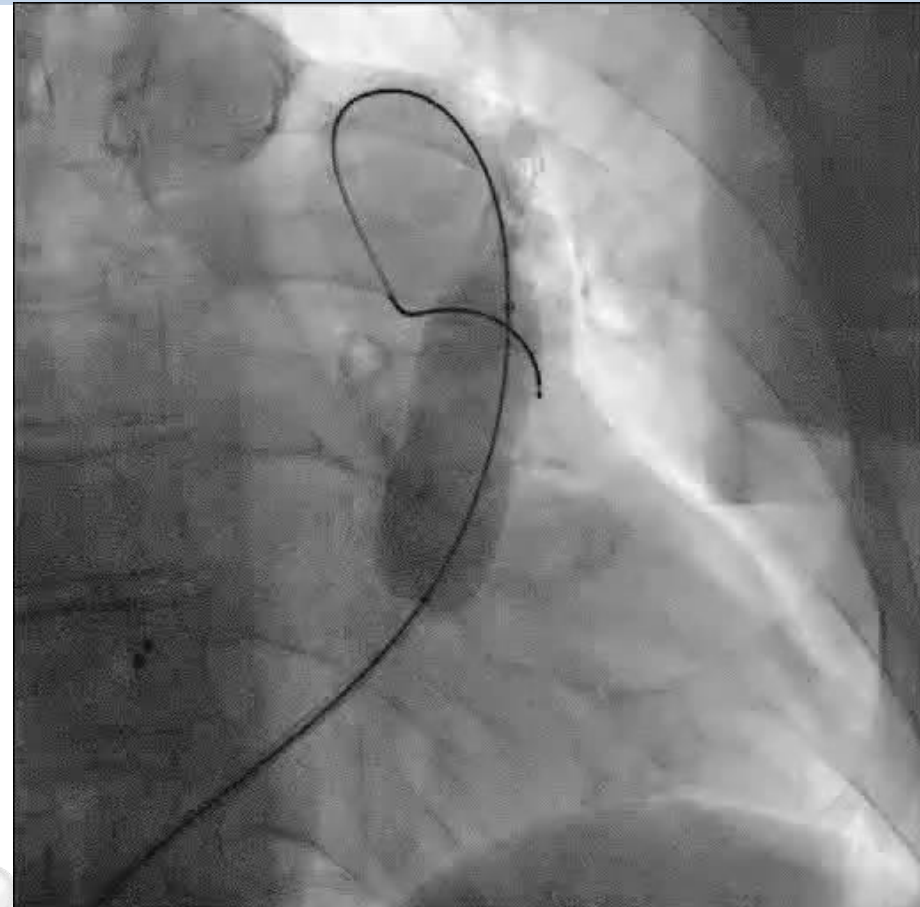
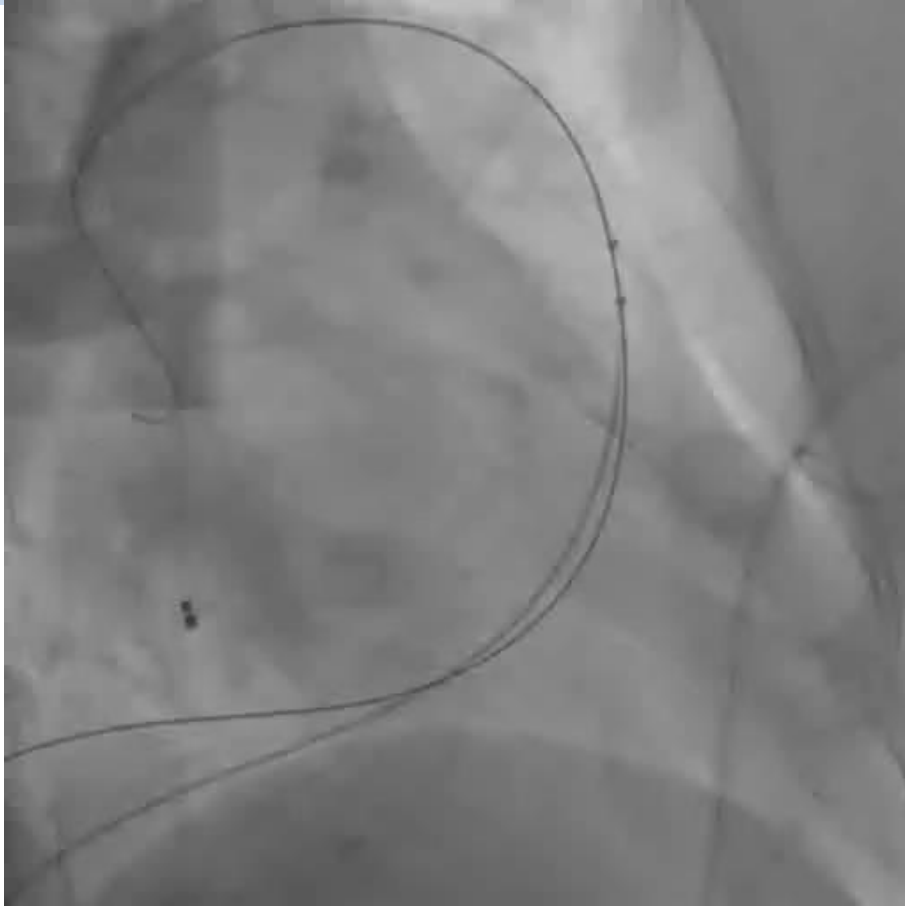


# ECO PRE VALVOLAZIONE

## SVP



# VALVOLAZIONE Calibrazione



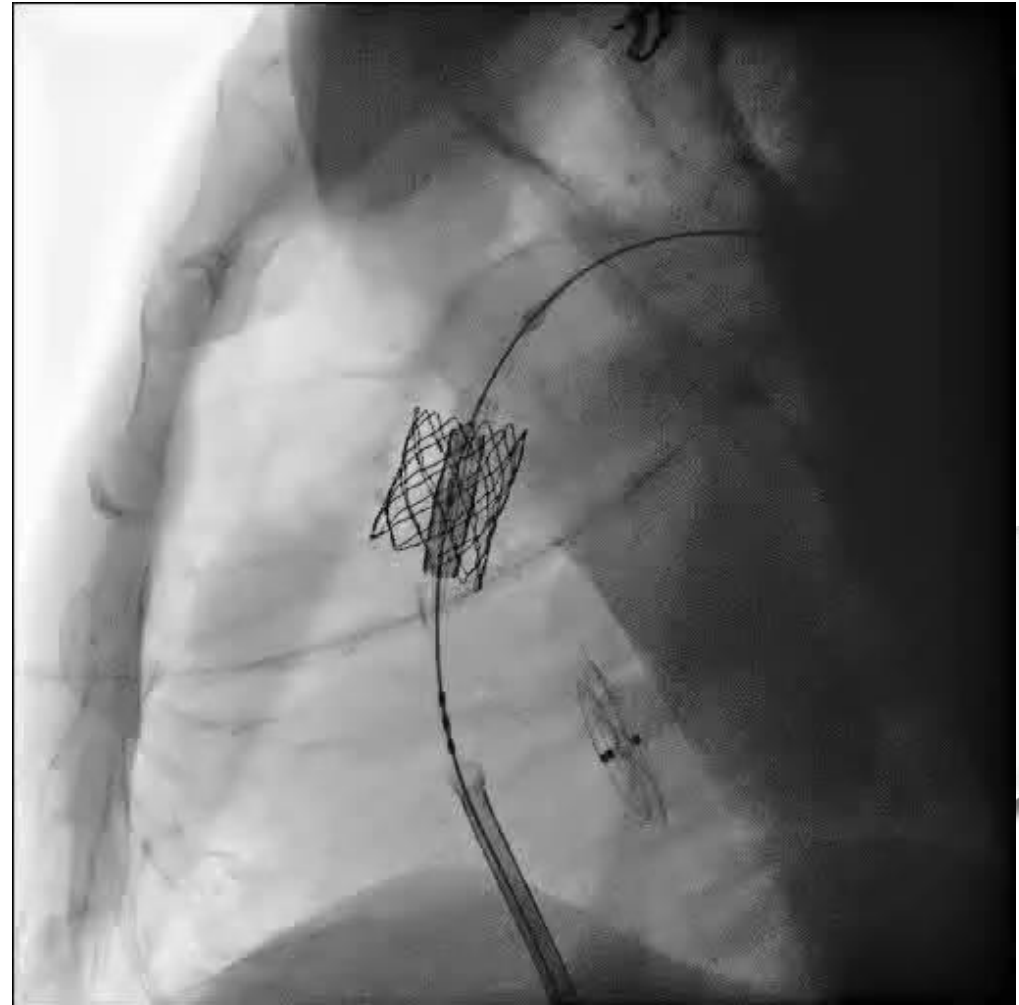
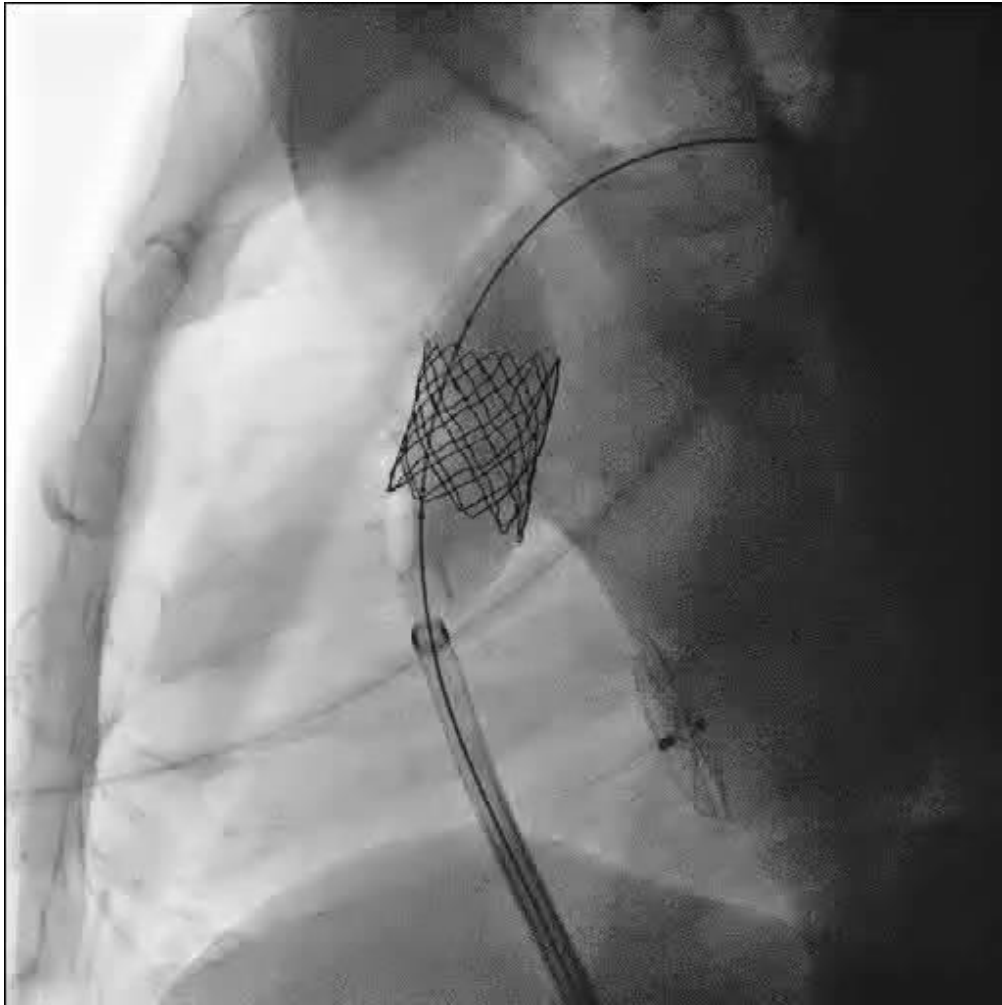
?

**Pressioni basali: Ao:220/110/ mm Hg !!!!! PA 94/29/55 mmHg.....**



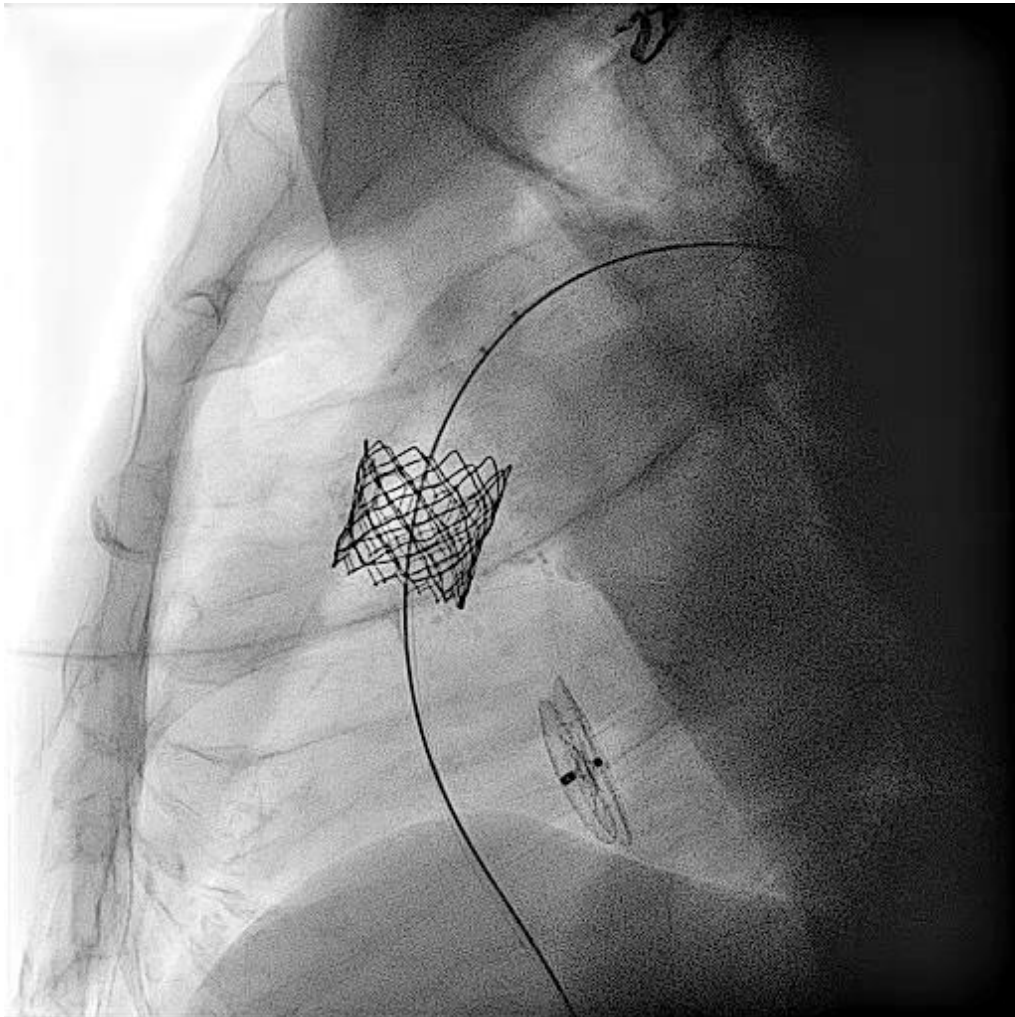
# VALVOLAZIONE

## Prestenting e valvolazione



# VALVOLAZIONE

## Sapien 29 mm

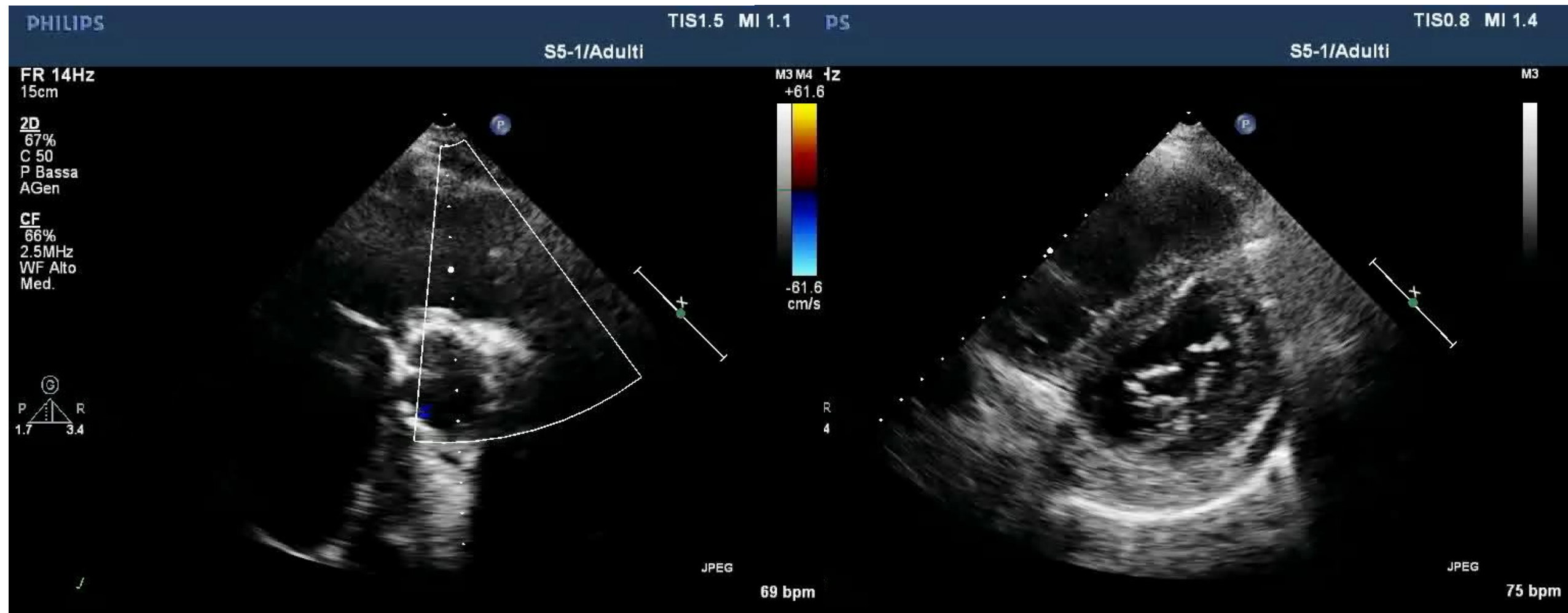


### PRESSIONI FINALI

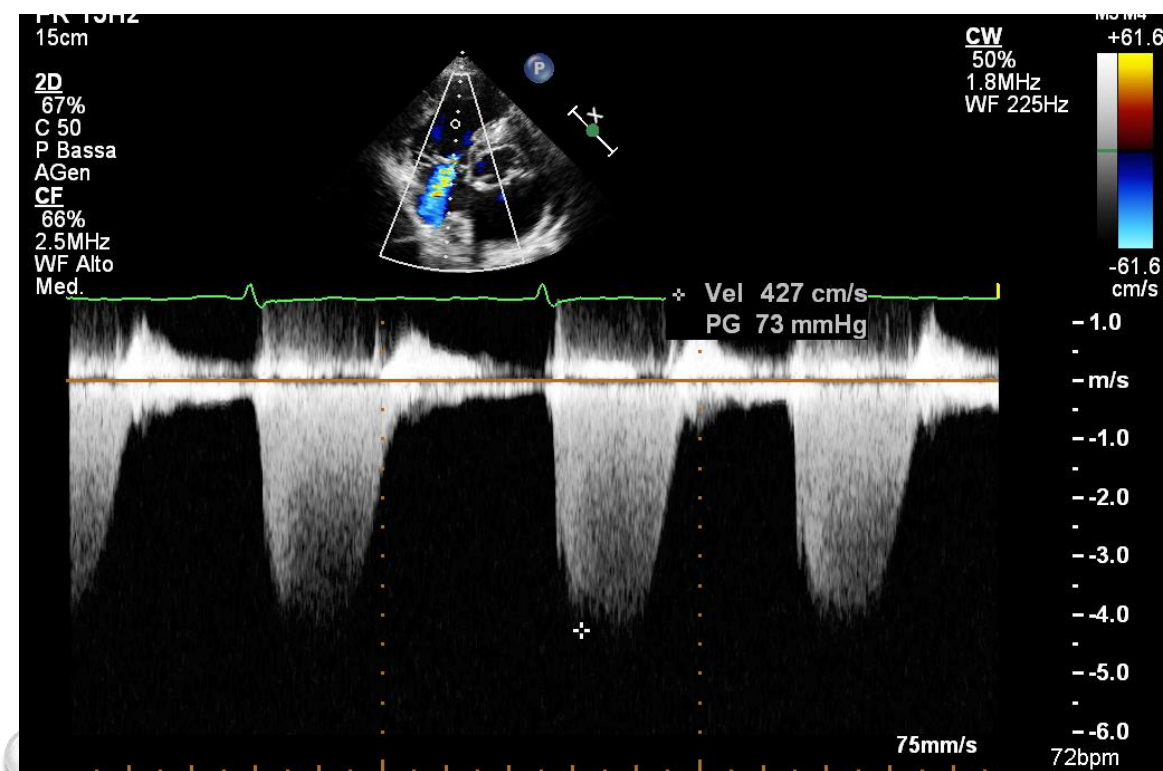
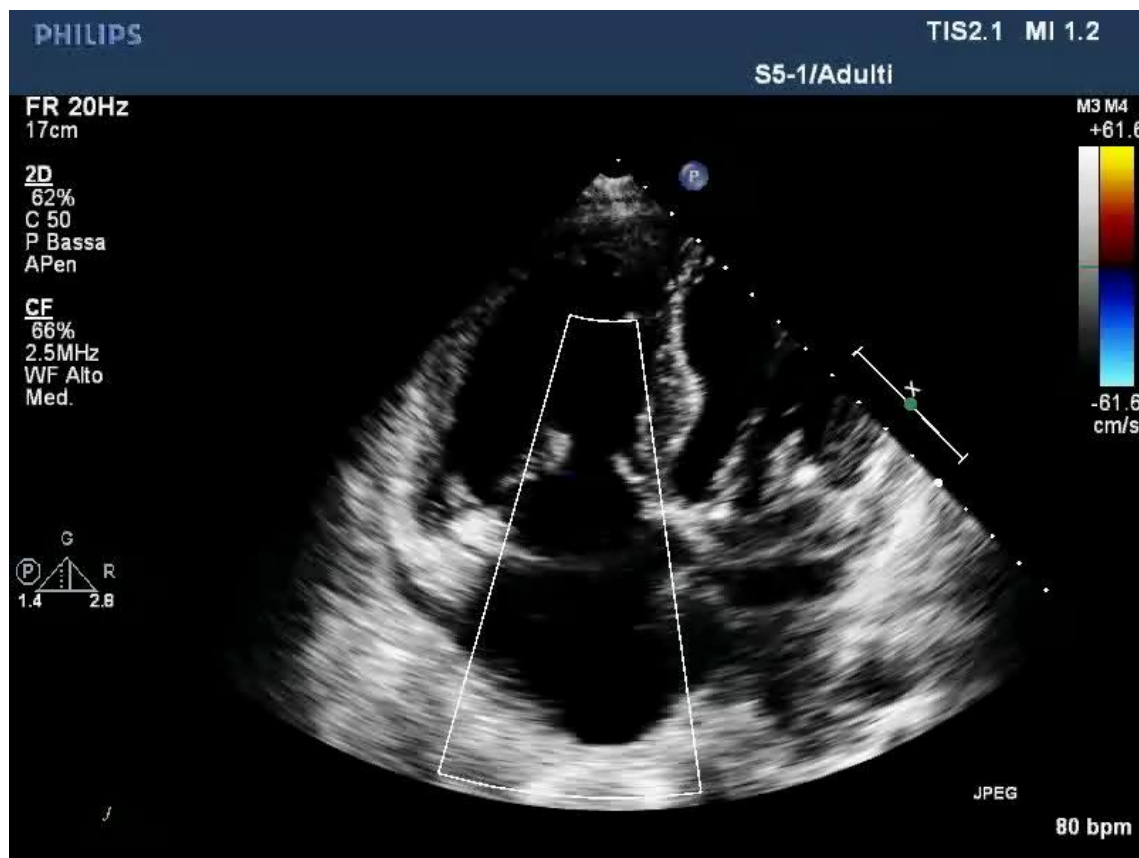
- Aorta 170/78/105 mmHg
- Polmonare 92/46/62 mmHg



# ECO POST VALVOLAZIONE



# ECO POST VALVOLAZIONE



## In hindsight.....

- Valutazione ambulatorio IP
- Terapia con anti-ipertensori polmonari
- Programmato restudio per RVP e reattività

Vivi come se dovessi  
morire domani.  
Impara come se  
dovessi vivere per  
sempre.

M. Gandhi



# In hindsight.....

Col "senno di poi" si  
comprende sempre tutto. Ci  
vorrebbe un "senno di prima"  
per farsi meno male.

