

# Eisenmenger Update

Queenstown 2017  
Prof David Celermajer  
University of Sydney

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**THE EISENMENGER SYNDROME  
OR PULMONARY HYPERTENSION WITH REVERSED CENTRAL SHUNT\***

BY

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# ES 2017

1. Novel insights into  
outcomes

2. Therapies- what  
works?



# PAH – Now a Treatable Illness?

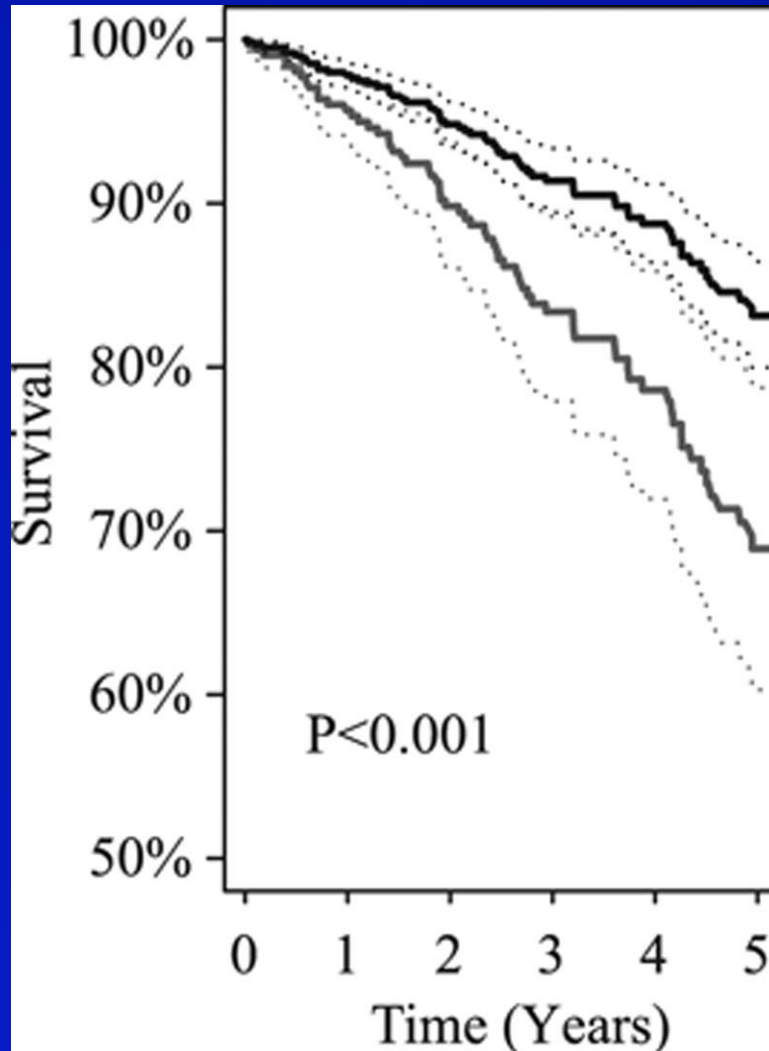


*Sometimes improvement  
is obvious...*

The stories of KC, DT and DY.....



# ES – Predictors of Survival



n = 1098

13 countries, 38 yrs

Main predictor of  
POOR outcome –  
pre-tricuspid shunt

Other predictors

Older age

Lower O<sub>2</sub> saturation

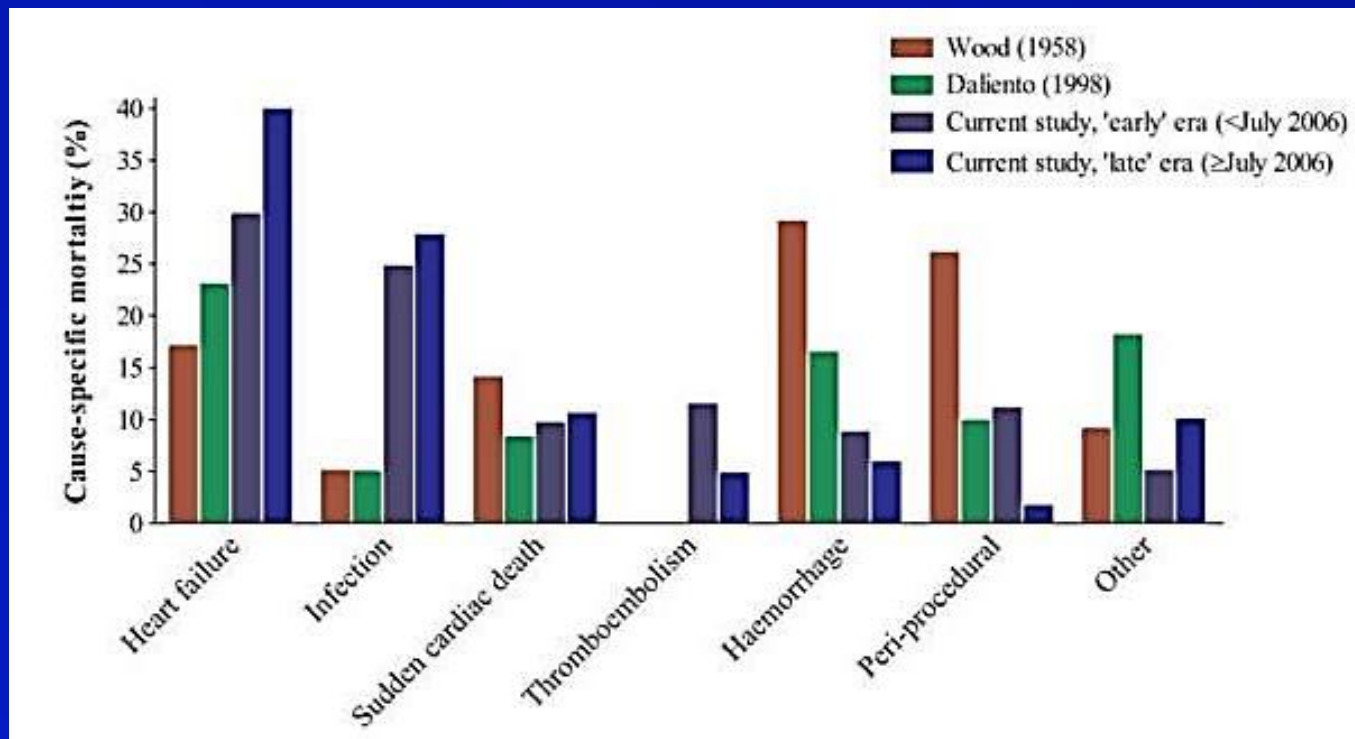
Loss of sinus rhythm

Kempny et al., *Circulation*, 2017

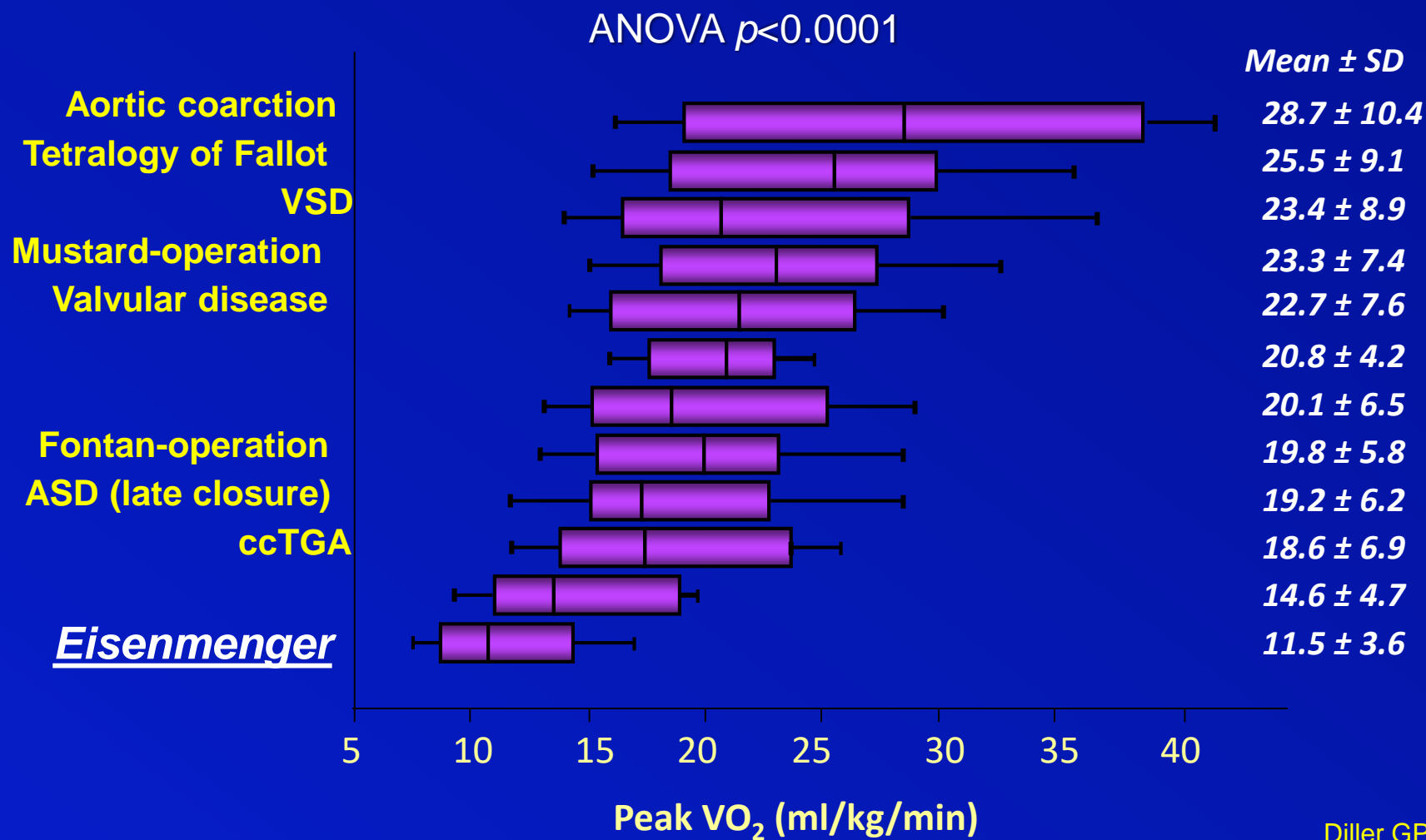
# Changing pattern of ES mortality

n = 1546

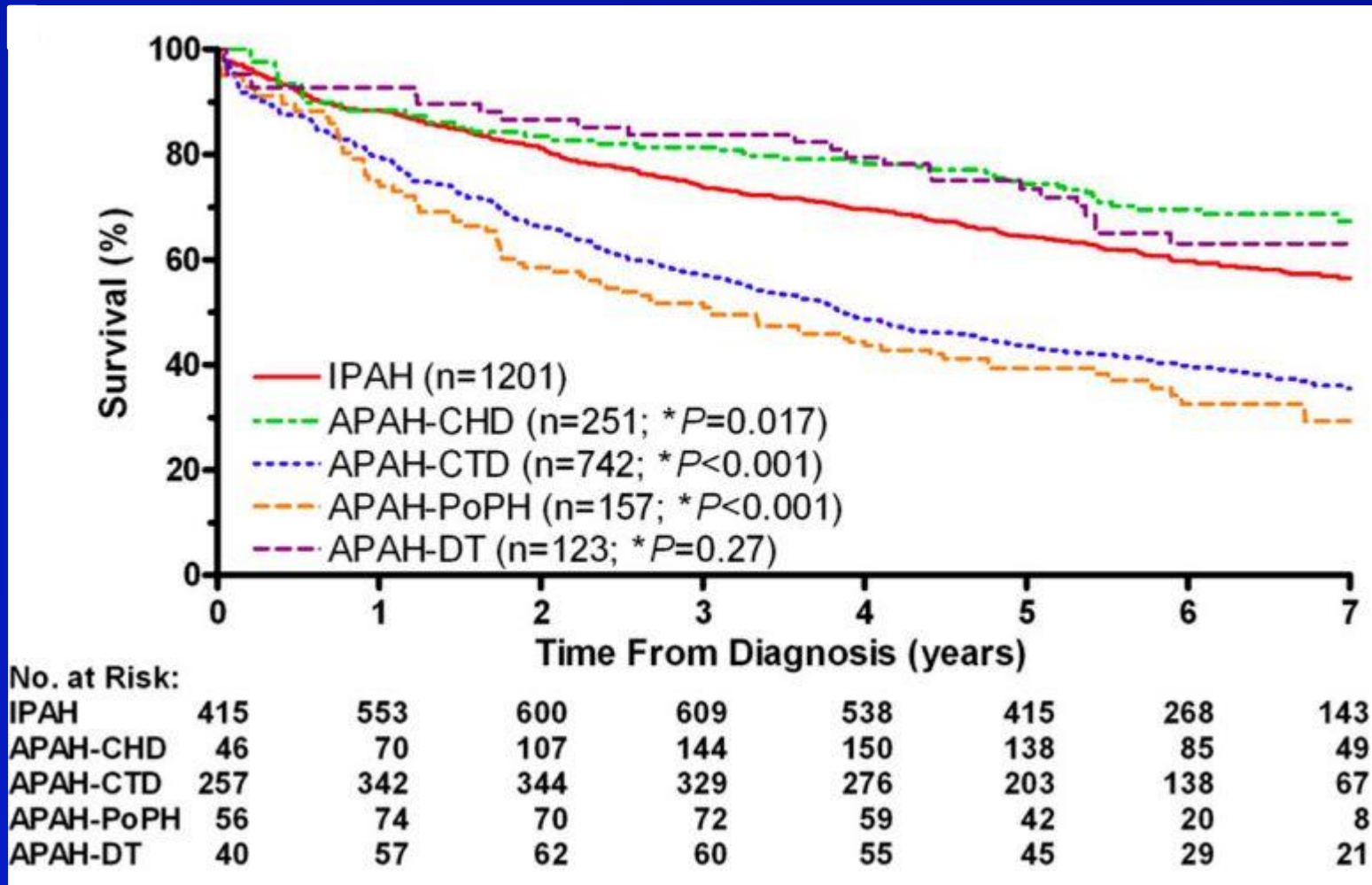
558 deaths



# Exercise capacity in adults with CHD - by underlying diagnosis



# REVEAL REGISTRY; ES Does Better??

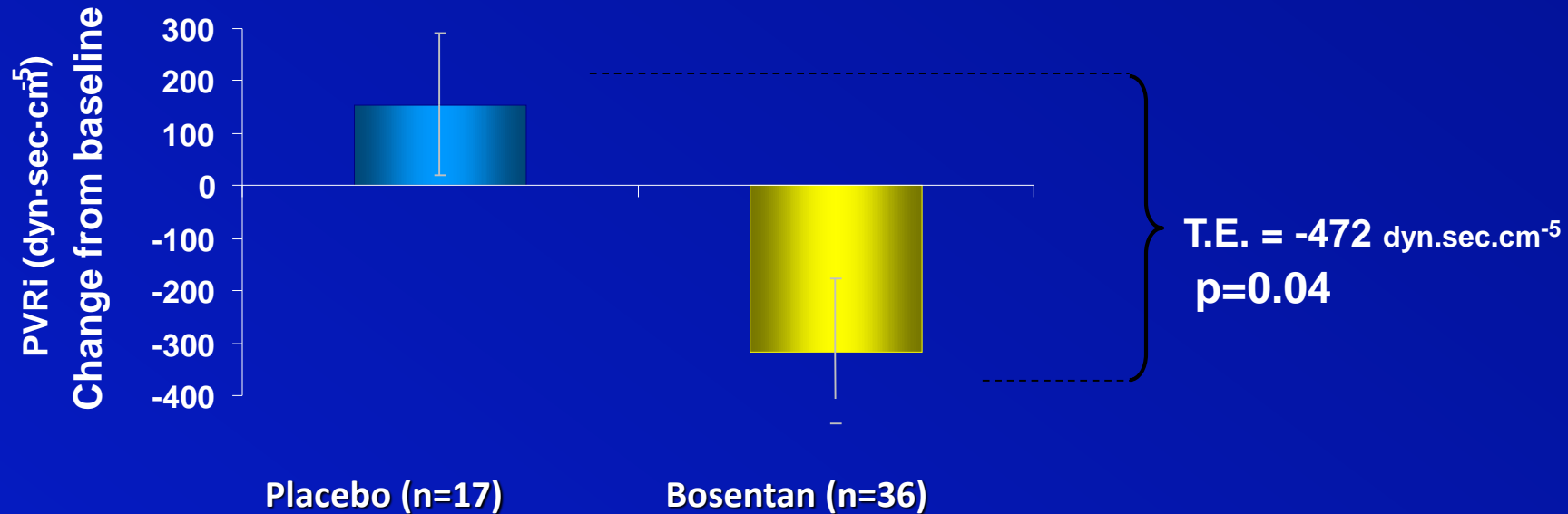


# Supportive Care for ES

- Avoid dehydration; pregnancy
- Care with surgical procedures; anaesthesia
- Digoxin, diuretics, antiarrhythmics, anticoagulants;  
NONE modify survival!
  - diuretics and hyperviscosity
  - anticoagulants and haemoptysis/haemorrhage
- Phlebotomy ONLY if symptoms of ↑ viscosity
- Transplantation
- Specific pulmonary vasodilators?

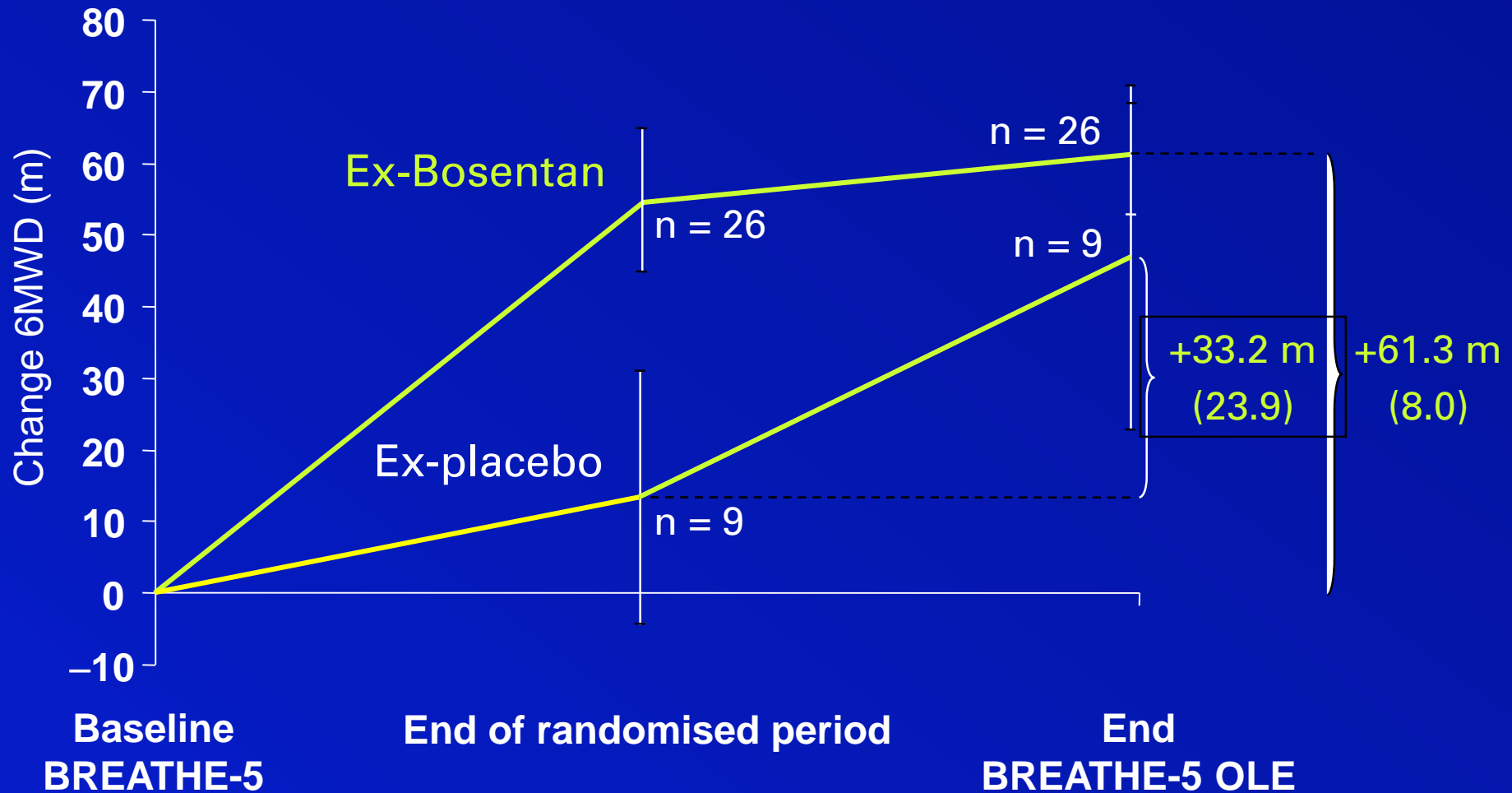


# BREATHE-5; Bosentan Reduces Pulmonary Vascular Resistance



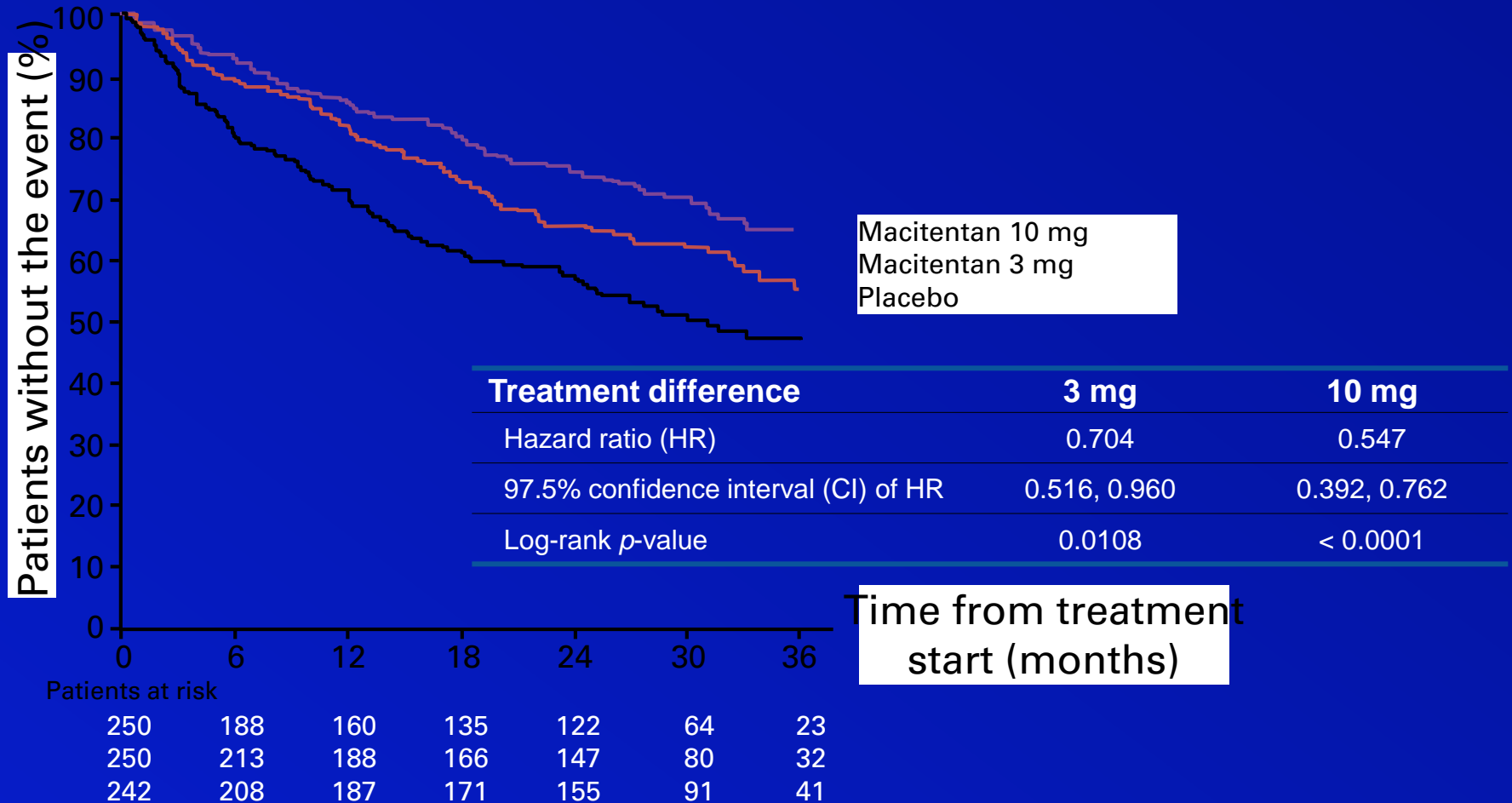
# Treatment of ES

## BREATHE 5 - Bosentan



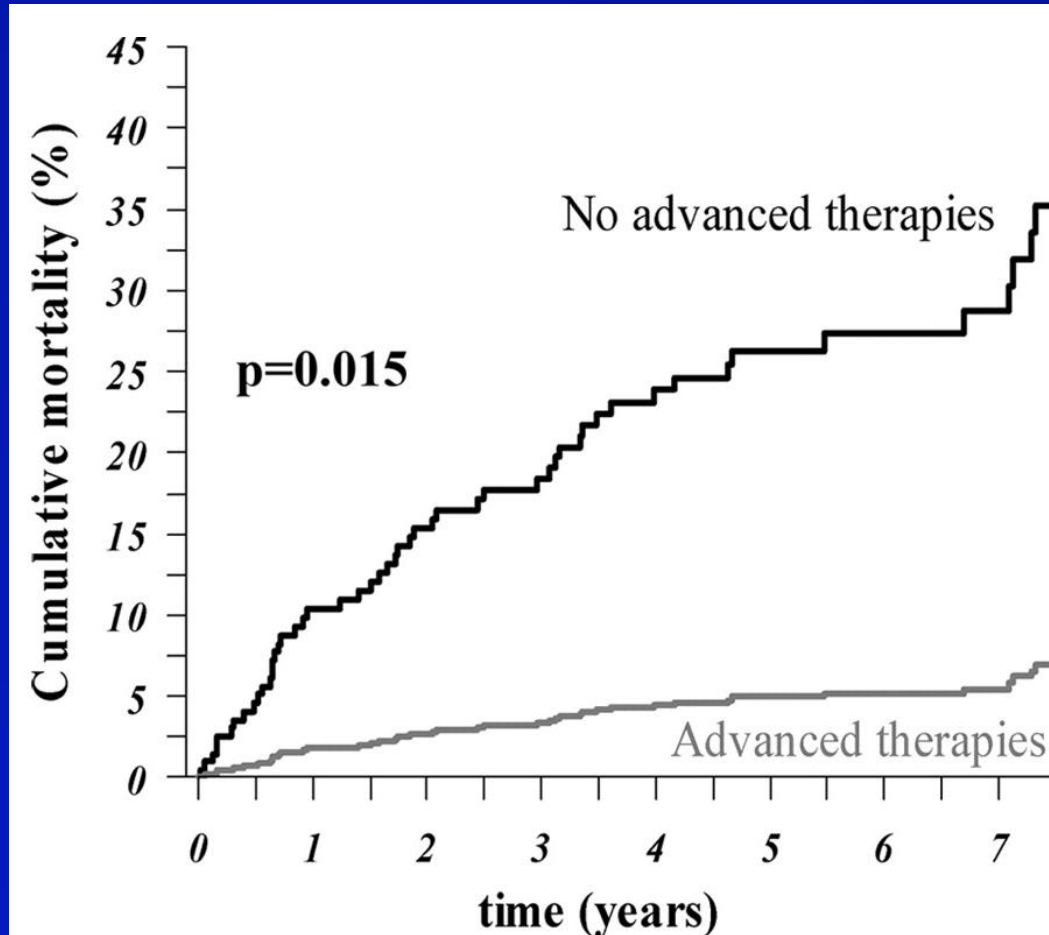
# SERAPHIN – Macitentan in PAH

(n = 762; 61 with CHD – PAH)(NB - 51 in BREATHE-5)



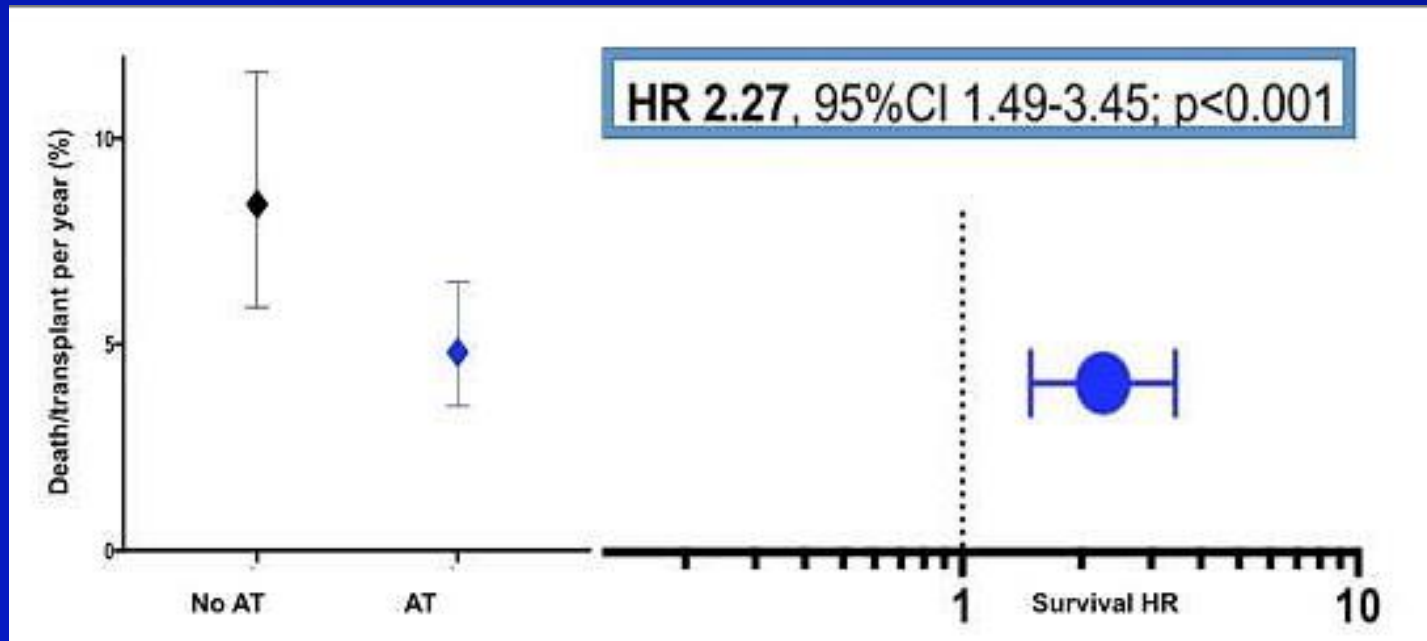
**What about MAESTRO???**

# Advanced Therapies in ES?

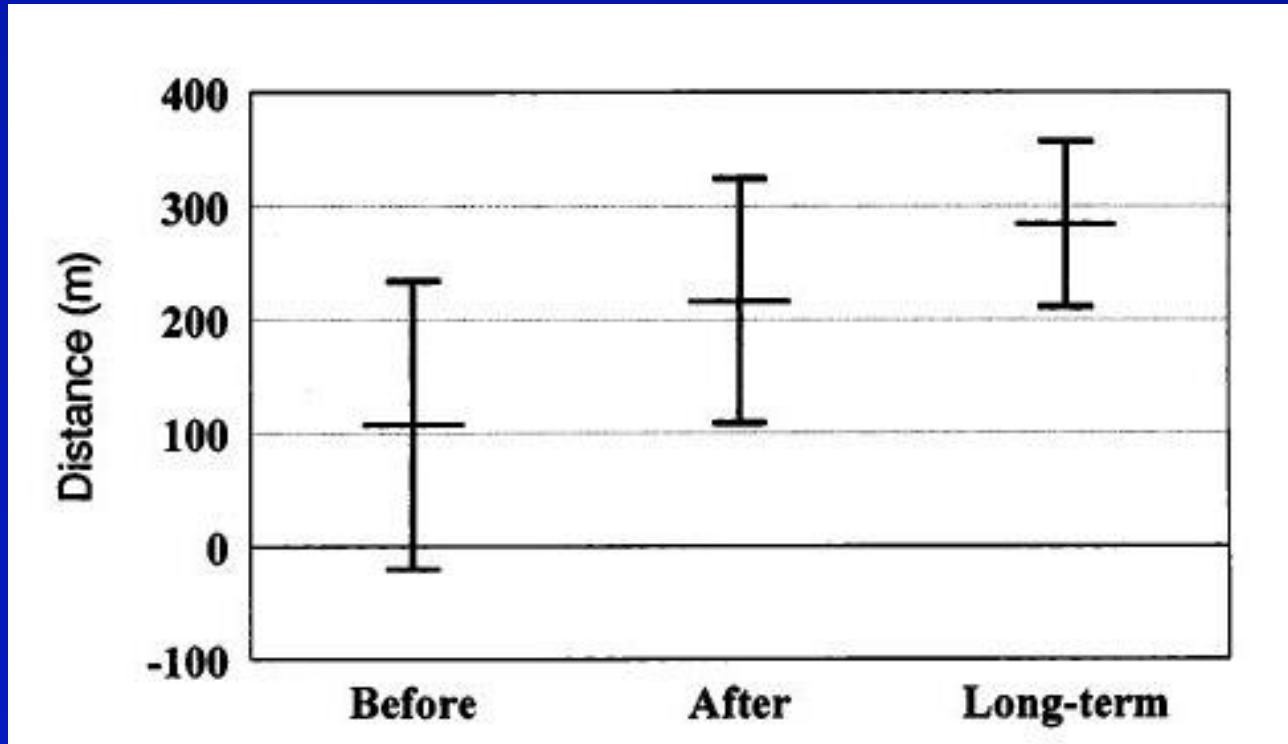


# Advanced Therapies in ES - ANZ

- n = 253
- 12 ANZ centres
- AT in 72% - (sicker)



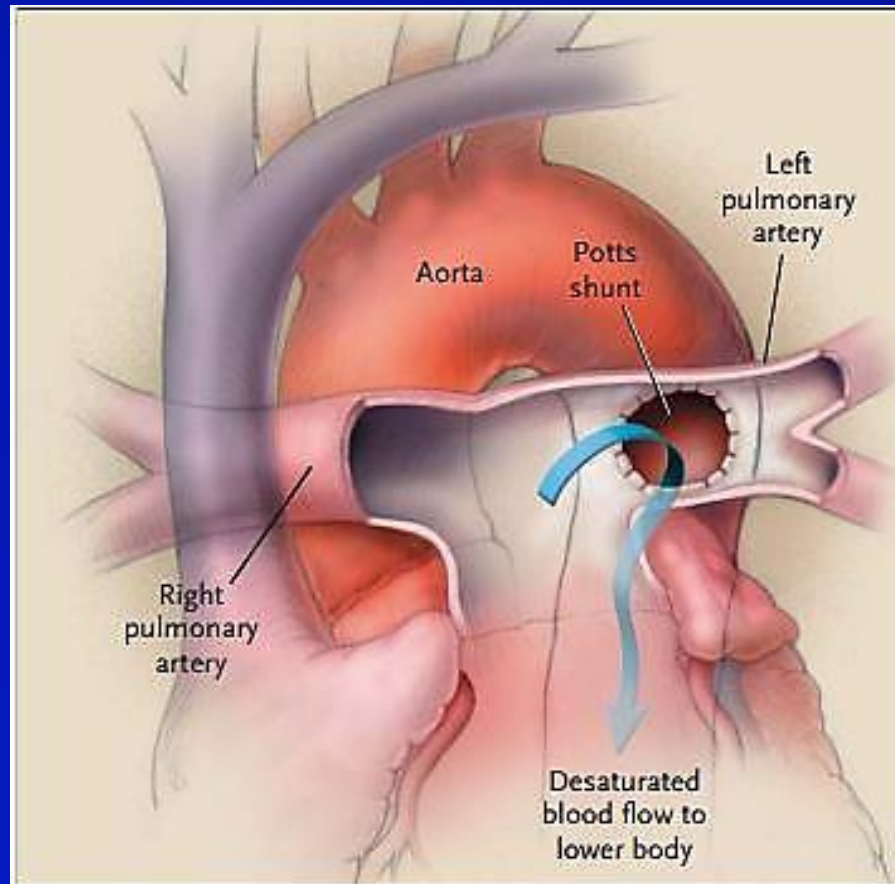
# Converting IPAH to ES – I – The Effect of BAS in iPAH



Sandoval et al., *JACC* (1998)

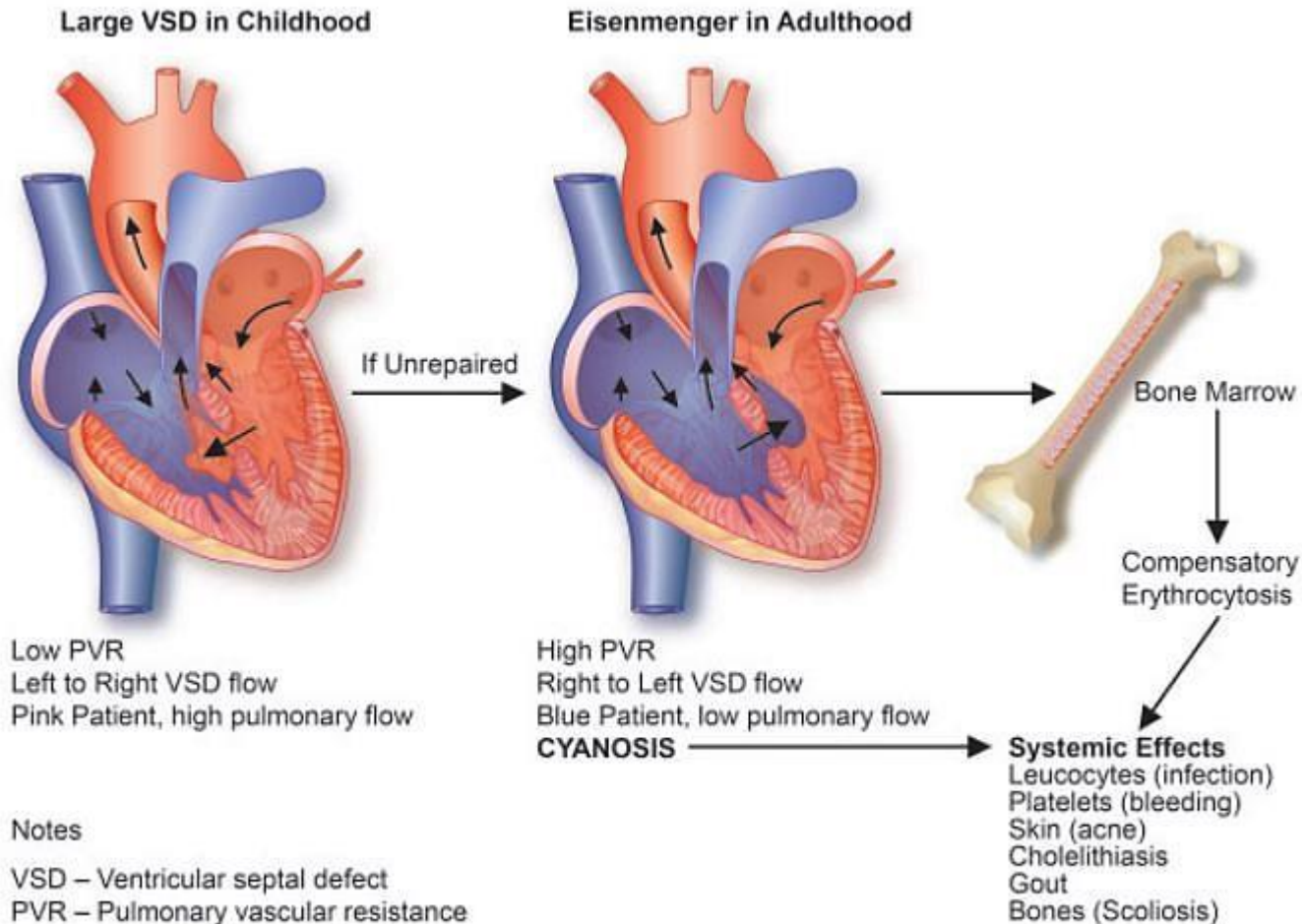
# Converting iPAH to ES II

## Potts' Shunt





# ES Effects



# ES 2017 - Conclusions

## **1. Continues to fascinate!**

- Perpetually blue – what does that effect?
- Why better than iPAH and what clues are there?

## **2. Changing patterns of prognosis and mode of demise**

## **3. Medications help – but remember supportive care!**

