Pulmonary hypertension and congenital heart disease

The treat and repair approach - does it make sense?

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Congenital heart defects and pulmonary hypertension

- **Types of Eisenmenger syndrome**

- **High flow PHTA**
  - Preventive surgery

- **Eisenmenger syndrome**
  - Surgical ways
Classical causes for ES

Pre-tricuspid forms

Post-tricuspid forms
Ventricular development

Hyperplasia

Hypertrophy
Potential for hyperplastic dvp

Hyperplasia

Hypertrophy
Evolution of P Hypertension in post-tricuspid valve hypertension
Large « non-restrictive » VSD

PVR

Qp/Qs

Years

2 m 5 10 20
Golden rule

Non-restrictive pressure shunts should be corrected within 6 months
Unrestrictive VSD

$RV/LV < 0.5$

3 months  3 – 4 hours
6 months  3 – 4 days
Chronic forms

Functional Endothelial dysfunction

Morphological Endothelial / medial scarification
Chronic VSD

Pulm vascular resistance < 6 Woods
P diastolic pressure < 50% systemic
Reactivity ++

Closure of the VSD
Carefulness to the tricuspid valve
Avoid infundibulotomy

Beware the tricuspid valve
Chronic VSD, pre-terminal PVD

- Pulm vascular resistance \[6 - 8\] Woods
- P/S diastolic pressure \(< 80\%\)
- Reactivity \(+\)

- L to R or bidirectional shunt at rest
- R to L shunt at exercise
Chronic VSD, pre-terminal PVD

- Closure and NO therapy only (?)
- Valved patch technique (?)
- Banding of the PA ?
VSD closure with a valved patch
Banding of the PA

Remodeling of PVD
Chronic VSD, terminal PVD

- Pulm vascular resistance: 8 - 10 Woods
- P/S diastolic pressure: > 90 %
- Reactivity: (-)

Exclusive R to L shunt
Never close the communications !!!
Chronic VSD, terminal PVD

Create a communication
Pulmonary-aortic connection
Ultimate treatment
Transplantation
**Tx in Eisenmenger syndrome**

Prognosis Eisenmenger. Saha. Int J Cardiol, 1994;45
Tx in Eisenmenger. Waddell. J Heart Lung Tx, 2002;21
Indications for transplantation

• Progressive signs of heart failure
• Quality of life
• Uni/bilateral lung transplantation with cardiac repair
• Heart-lung transplantation
Tx in Eisenmenger syndrome

- Simple defect ASD / VSD with good RV/LV function
- Uni/bilateral lung Tx + Cardiac defect repair
Tx in Eisenmenger syndrome

• More complex defects (VSD, A-V canal, Truncus arteriosus)

• Reduced RV/LV function

• Heart-Lung transplantation
Conclusion

Low / median PVR
Closure of communication

Median-high PVR
Valved closure

High-fixed PVR
Create a communication

H-L or Lung Tx
Heart failure
Quality of life