Pediatric PAH after successful neonatal arterial switch operation for transposition of the great arteries

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Declaration of interest

- I have nothing to disclose
Transposition of the great arteries

- Most common cyanotic CHD (5% of all CHD)

Van der Linde et al, JACC, 2011
https://concor.net/nl/transpositie-van-de-grote-vaten.html
Drawing: dr. J.P.M. Hamer
PAH in TGA

• Uncorrected $\Rightarrow$ rapid development of PVD/PAH

• Atrial redirection procedure:
  • Performed $\geq 6$ months after birth
  • PAH reported in 7% of the cases

Haworth, Br Heart J, 1984
Ebenroth et al, Am J Cardiol, 2000
Arterial switch operation

• Anatomic repair within weeks after birth

• Pulmonary hemodynamics virtually normalized

• PAH not to be expected, but clinically recognized

• Clinical characterization lacking
Aims of the study

• To gain more insight in this clinical entity by:
  • Presenting an international cohort of children with PAH after neonatal ASO for TGA
  • Describing its epidemiology and clinical course
Methods

• Retrospective, international, multicenter study

• Including children who:
  • Underwent ASO for simple TGA (± VSD)
  • At neonatal age (<6 weeks after birth)
  • Between 1989 and 2014
  • No residual defects
Participating centers

Total: 25 children
Methods

• Data collection:
  • Patient characteristics
  • ASO and peri-operative phase
  • PAH (diagnosis, treatment)
  • Endpoint or last follow-up

• PAH confirmed by cardiac catheterization
• In 1 child → echocardiographic diagnosis
Methods

• Treatment intensity:
  • Calcium channel blocker (CCB) therapy
  • Mono, dual, triple PAH-targeted therapy

• Primary endpoint:
  • Potts shunt
  • Lung transplantation (LTx)
  • Death
  • If not, censored at last follow-up
Patient characteristics

- Gestational age > 36 weeks
- 76% males
- 13% PPHN
- 24% concomitant VSD
- 84% Rashkind procedure
- Median age ASO 8 days (IQR 6, 10)
Age first PAH detection (months)
Hemodynamics

mPAP, mean pulmonary artery pressure; PVRi, indexed pulmonary vascular resistance
PAH therapy

- Triple therapy (40%)
- Dual therapy (32%)
- Mono therapy (24%)
- CCB therapy (4%)

CCB = calcium channel blocker
Survival

Median follow-up 5.1 years (IQR 2.9, 12.1)

- 2 Potts shunts
- 4 LTx
- 8 deaths
Survival after ASO

P = 0.025

Children at risk:
- <1yr after ASO:
  - 16
  - 5
  - 2
  - 0
  - 0
- >1yr after ASO:
  - 9
  - 8
  - 7
  - 3
  - 1
Survival after first PAH detection

P=0.257

Children at risk:
- <1yr after ASO: 16, 5, 2, 0
- >1yr after ASO: 9, 4, 3, 3

(%)
Discussion

• First study clinically describing this entity

• Review of literature →
  reported incidence 0.5-0.9%

Losay et al, Circulation, 1990
Rivenes et al, Tex Heart Inst J, 1998
Hutter et al, J Thorac Cardiovasc Surg, 2002
Cordina et al, Pediatr Cardiol, 2010
Clinical implications

• 15 years after ASO for TGA:
  • Survival ~90%
  • (Almost) no deaths after 5 years

• PAH important factor in prognosis

• Early- or late-onset PAH
Take home messages

1. Children with neonatal ASO for TGA should have lifelong clinical follow-up including screening for PAH

2. Two phenotypes (early- and late-onset PAH) with similar fatal course from PAH detection