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

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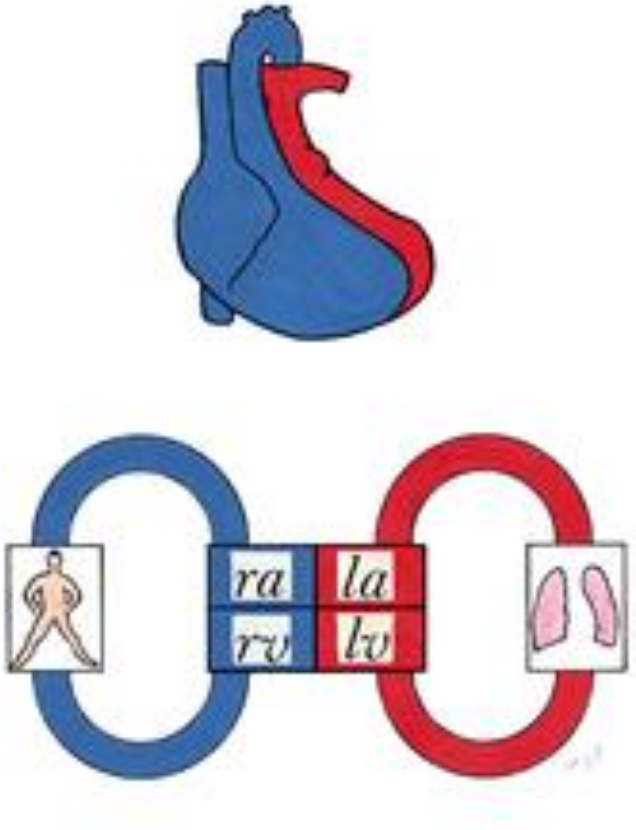
Declaration of interest

- 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 → rapid development of PVD/PAH
- Atrial redirection procedure:
  - Performed ≥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

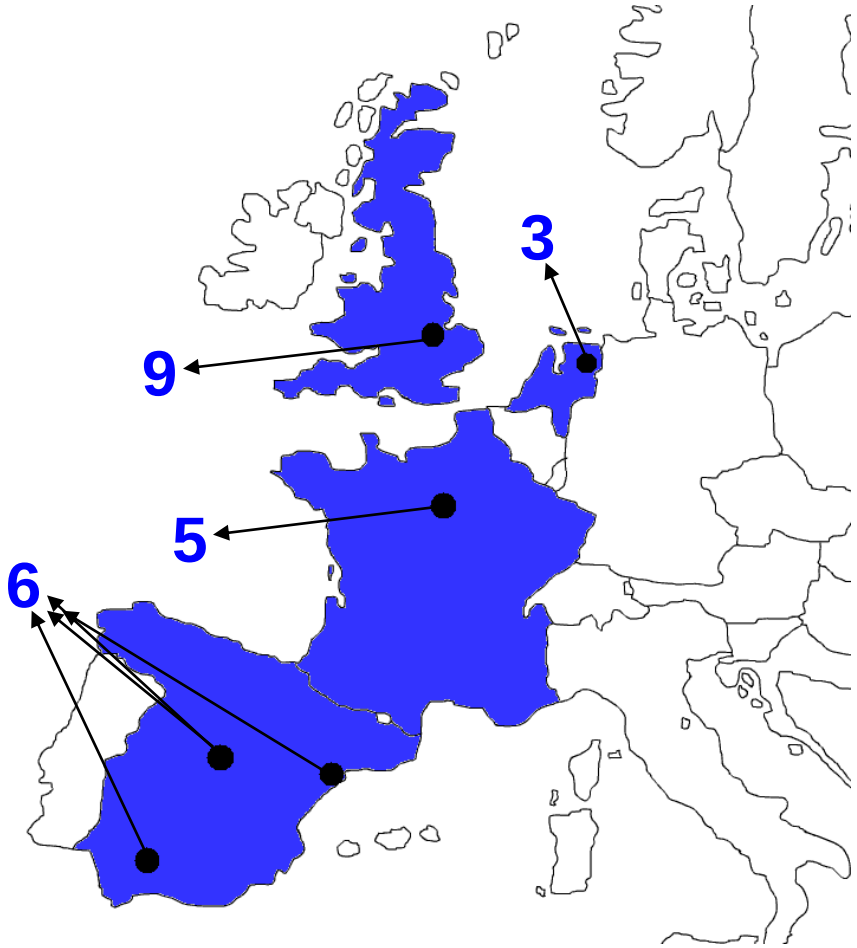
Aims of the study

- 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

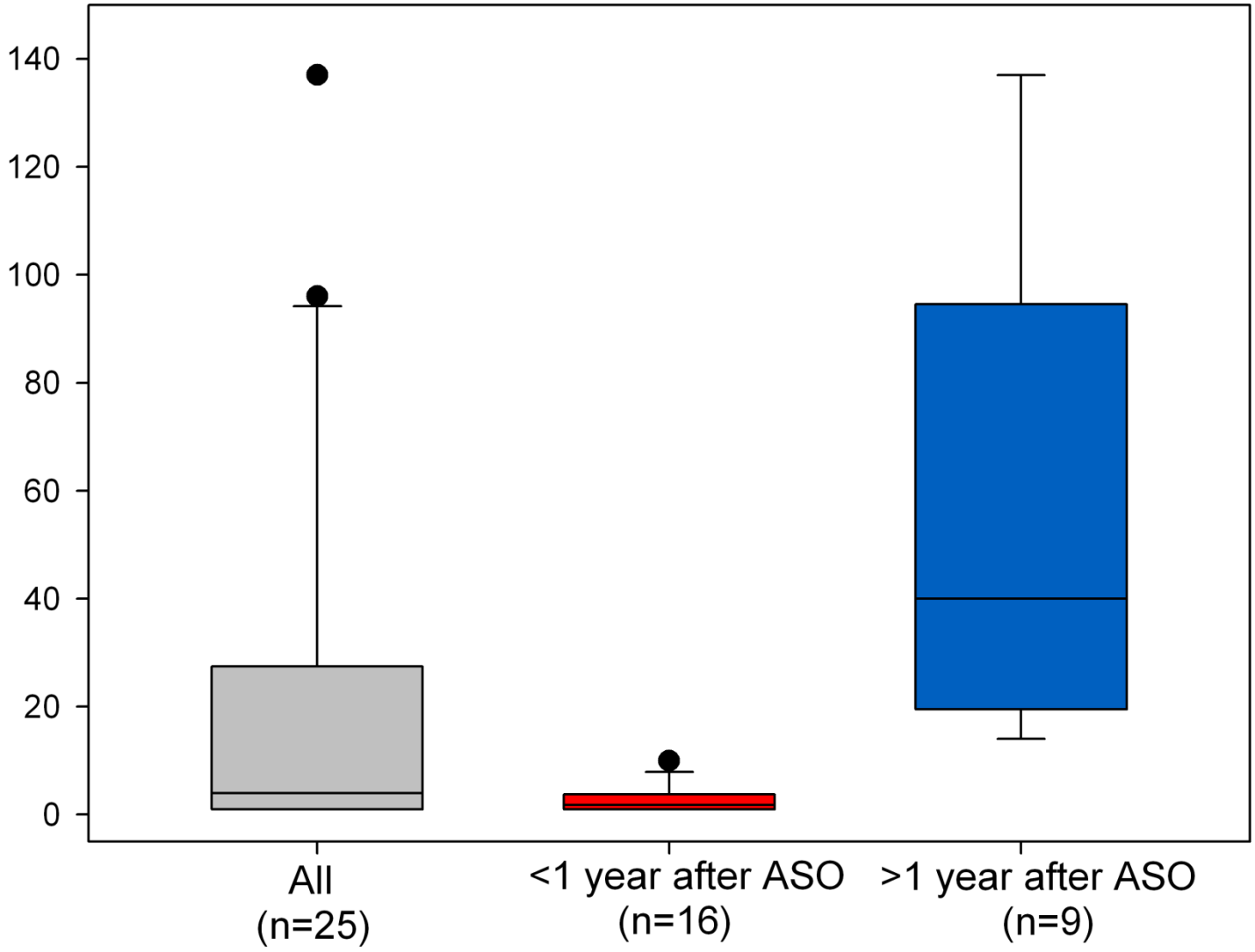
- Treatment intensity:
  - Calcium channel blocker (CCB) therapy
  - Mono, dual, triple PAH-targeted therapy
- Primary endpoint:
  - Potts shunt
  - Lung transplantation (LTx)
  - Death
  - If not, censored at last follow-up

Patient characteristics

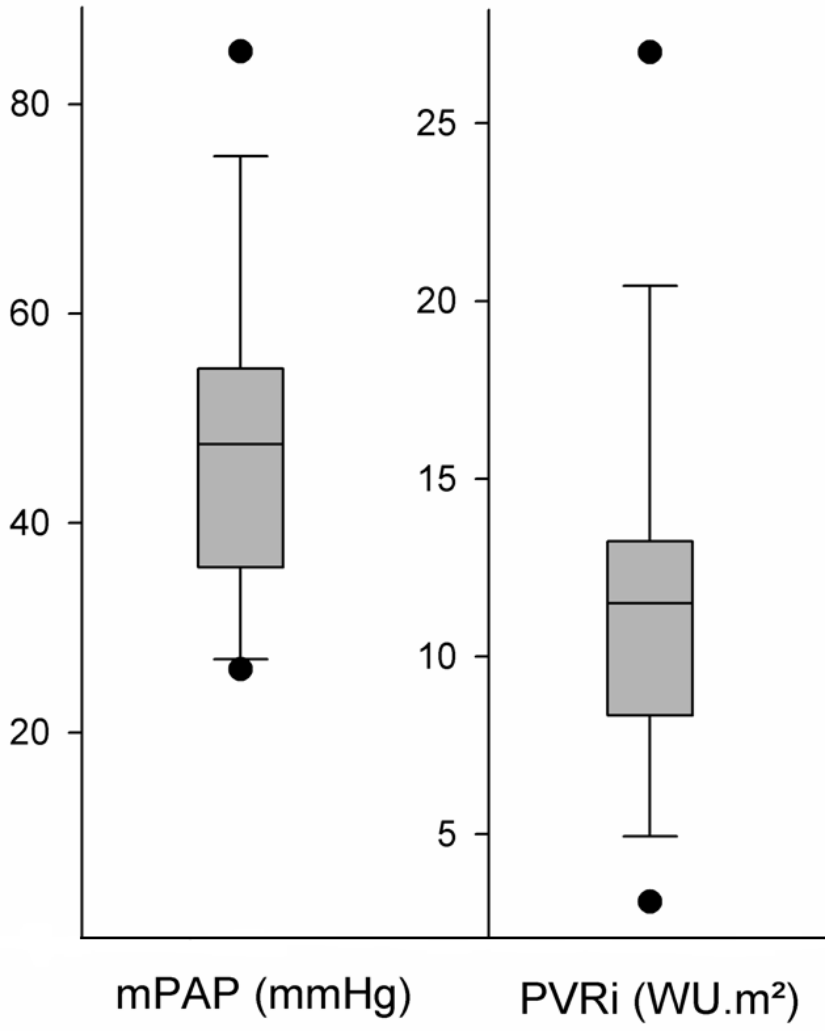
- Gestational age > 36 weeks
- 76% males
- 13% PPHN
- 24% concomitant VSD

- 84% Rashkind procedure
- Median age ASO 8 days (IQR 6, 10)

Age first PAH detection (months)

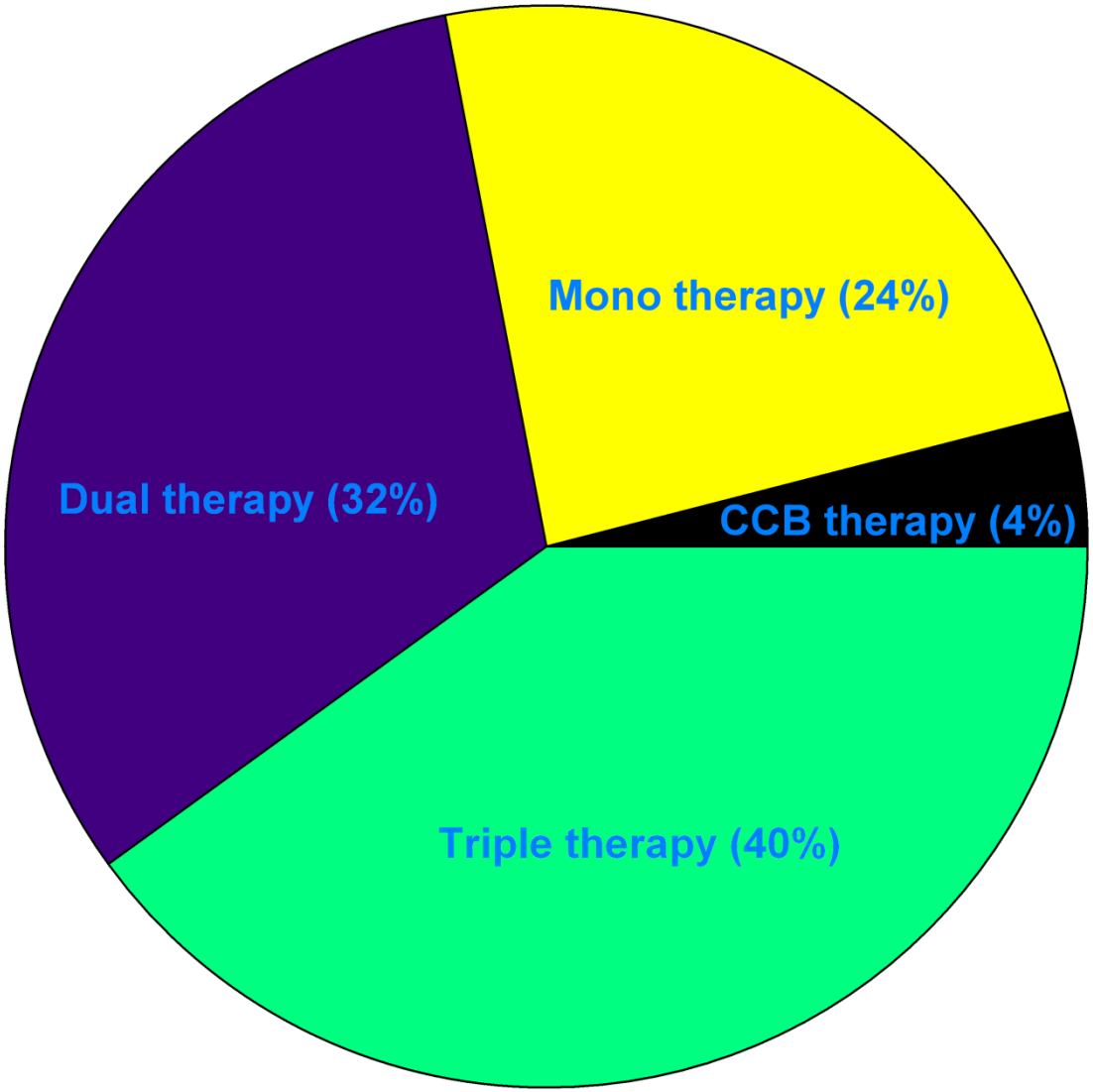


Hemodynamics



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

PAH therapy

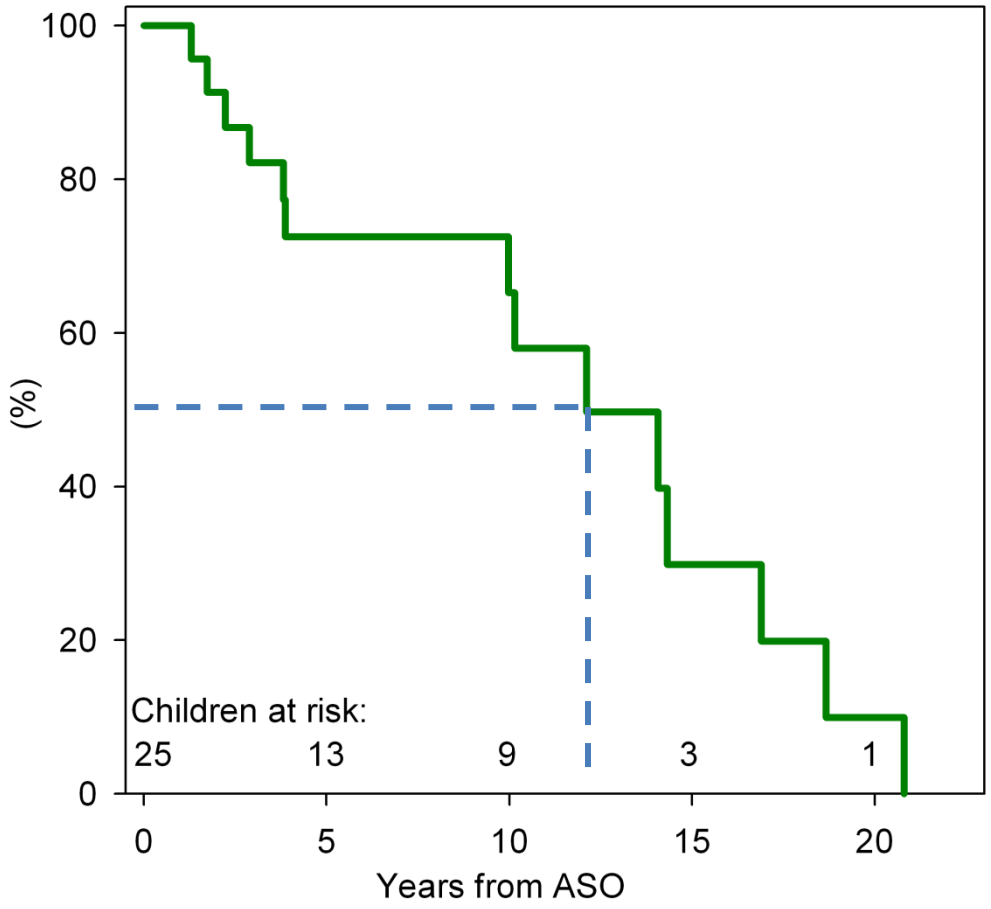


CCB = calcium channel blocker

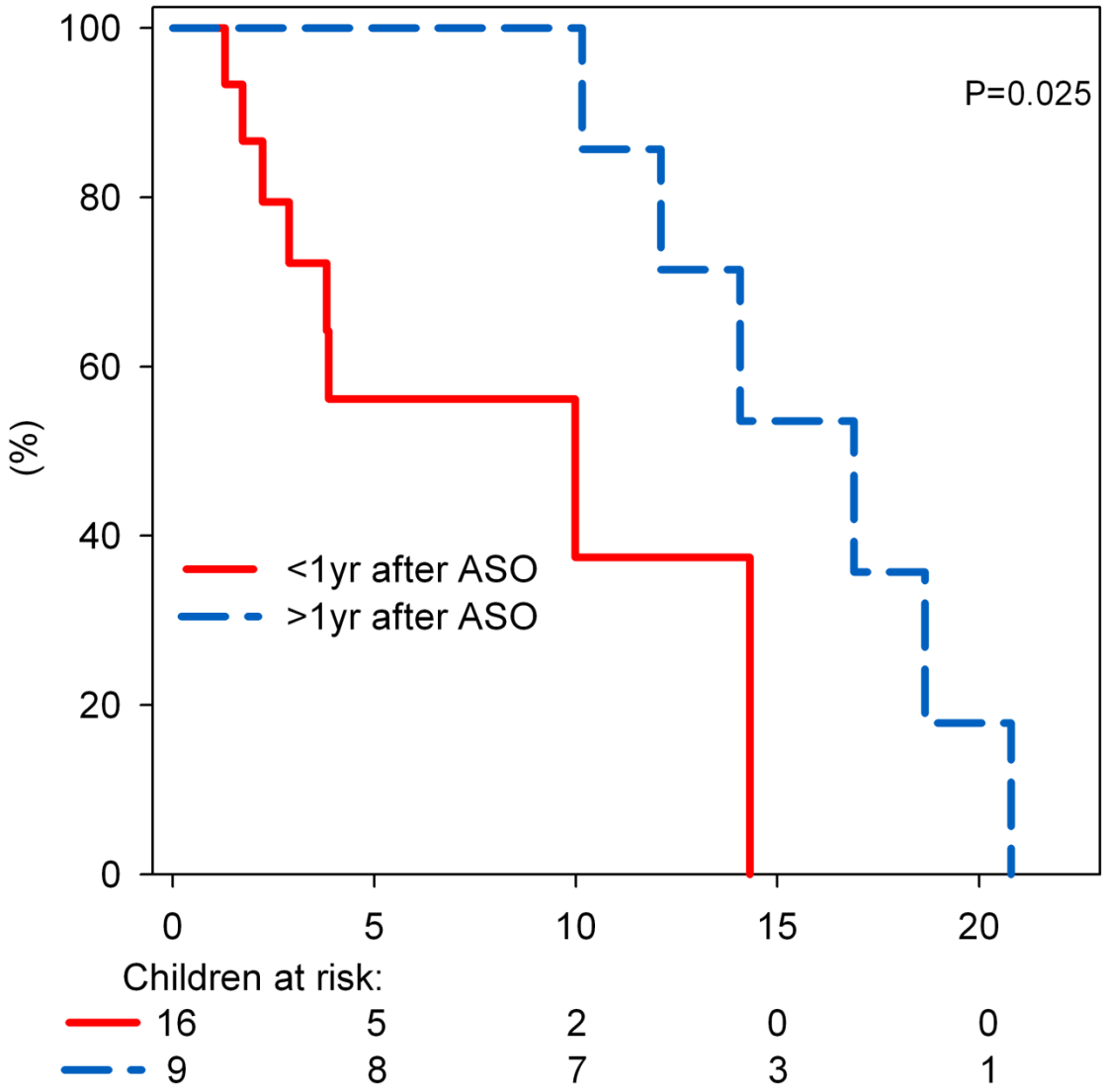
Survival

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

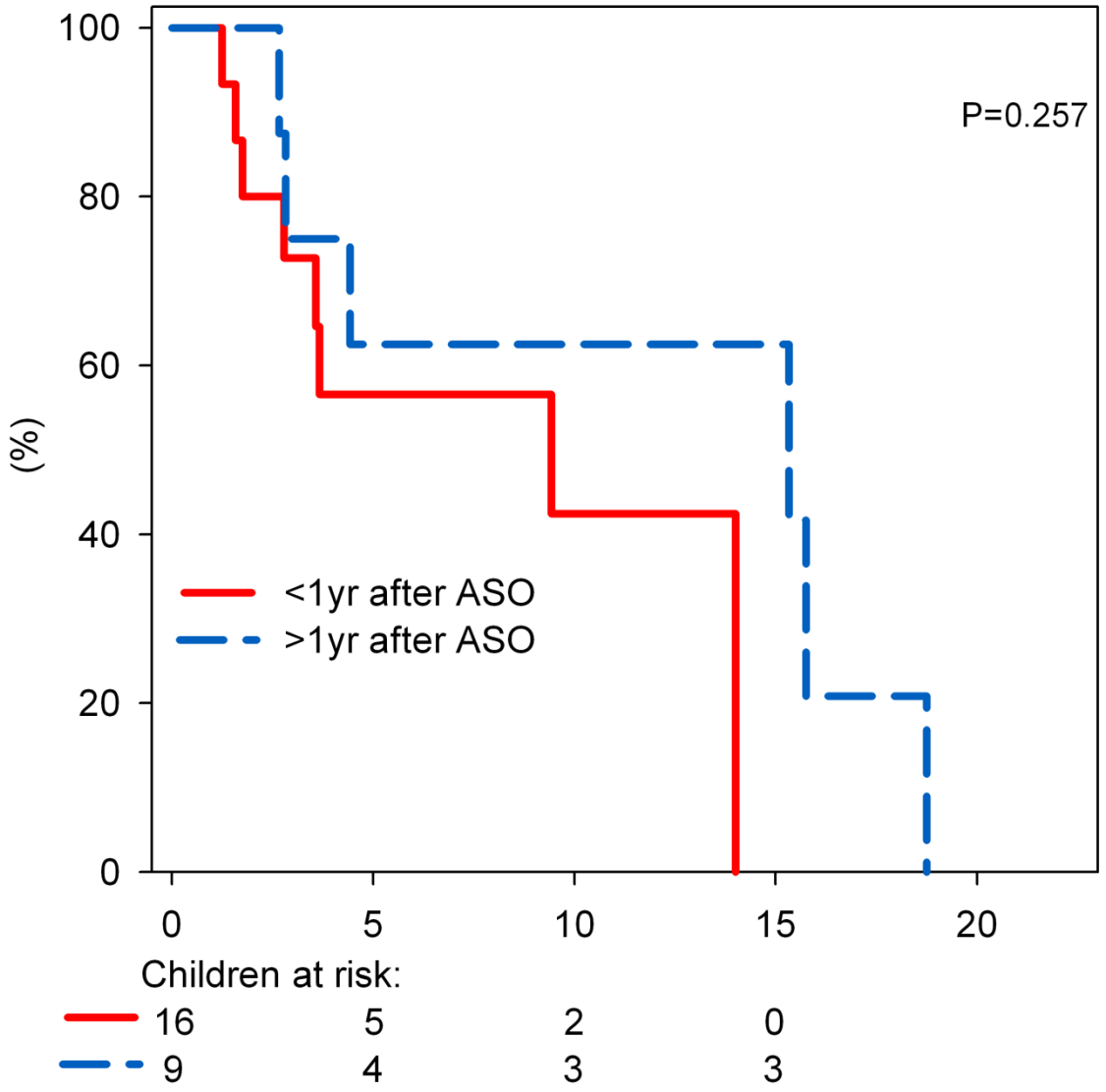
- 2 Potts shunts
- 4 LTx
- 8 deaths



Survival after ASO



Survival after first PAH detection



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
Roofthooft et al, Ann Thorac Surg, 2007
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

Losay et al, Circulation, 2001

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