

Pulmonary arterial hypertension in children with transposition of the great arteries after successful neonatal arterial switch operation

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Introduction– Pulmonary arterial hypertension (PAH) has been sporadically described in children after neonatal arterial switch operation (ASO) for transposition of the great arteries (TGA). To describe epidemiology and clinical course of this particular concurrence, we present a series of children with PAH after neonatal ASO for TGA.

Methods– In nine dedicated pediatric pulmonary hypertension centers in Europe and North-America data were collected of children diagnosed with PAH after neonatal ASO (≤ 6 weeks after birth) for TGA between 1989 and 2014. Children with significant residual shunt-defects were not included.

Results– Twenty-five children were identified (median age of ASO 8 days). Six children (24%) had a concomitant ventricular septal defect. In 14 children (56%), PAH was diagnosed within six months after ASO. The remaining children were diagnosed after a median of 30 months (interquartile range: 14 – 93 months). Twenty-three children (92%) received PAH-targeted therapies during their disease course. During a median follow-up after ASO of 5.1 years, 2 children received a Potts shunt, 4 children underwent lung transplantation and 8 children died. One-, 5-, 10- and 15-year Potts shunt- and lung transplantation-free survival after ASO was 100%, 73%, 65% and 30%, respectively. Of the survivors without lung transplantation or Potts shunt (median follow-up after ASO 4.7 years), all but one child were in World Health Organization Functional Class I-II at last follow-up.

Conclusions– PAH after successful neonatal ASO for TGA represents a specific disease entity with a putative incidence of 0.5-0.9% of children that undergo ASO for TGA. Onset of PAH varies from directly after ASO to first detection only in adolescence. Its clinical course varies from rapid deterioration and death to a prolonged course in good clinical condition. Since prolonged exposure to unfavourable hemodynamics is not present in these children, specific prenatal pulmonary hemodynamics in TGA or genetic make-up may play a role in the typical development of PAH in these children.