Causes and outcomes of pulmonary hypertensions confirmed with right heart cath in infants younger than 6 months

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Background
The epidemiology of pulmonary hypertension in infants younger than 6 months is poorly known as they are either excluded of registries or do not have right heart catheterization to characterize their hemodynamic.

Objectives
We retrospectively reviewed all RHC performed over a period of 8 years in infants less than 6 months of age with the purpose to evaluate pulmonary hemodynamic. We define PH as mean PAP > 25 mmHg and PVRi > 3 WU.m2 for those with biventricular physiology with or without shunt. Any value of pulmonary wedge pressure was accepted. For segmental PH, only mPAP was considered. Patients were classified according to the Nice classification.

Results
210 infants < 6 months were included into the study. 184 had PH confirmed. Causes of PH and/or indications for RHC were: group 1 (Pulmonary arterial hypertension) - evaluation of left-to-right shunts for operability or pulmonary blood flow contribution to clinical condition in 69 (37.5%), idiopathic PAH in 3, PVOD in 1, associated PAH-other in 5 (total 42.4%); group 2 (Postcapillary PH) - diagnosis of pulmonary veins anomalies in 29 (15.7%), and PH in obstructive left heart diseases in 26 (14.1%); group 5 (Multifactorial) - scimitar syndrome in 10; PH after the ASO for TGA in 6; complex congenital heart diseases and segmental PH in 14 (total 16.3%); group 3 (developmental lung diseases) isolated in 5 and with coincidental CHD in 16 (total 11.4%). There was one RHC related death due to sepsis. One-year mortality in the total population at one year was 35/184 (19%) and mainly related to persisting PH. 76% of survivors had normal pulmonary pressure at one year of age either after surgical correction of CHD or improvement of their lung disease. 37 patients (24% of survivors) had PH at one year of age.

Conclusion
PH in young infants is mainly associated with congenital heart diseases (CHD) with all groups being represented. Idiopathic PAH and developmental lung disease are rare in this age group. The transitory aspect of PH is frequent in all PH-CHD groups. Persisting PH at one year is present in a fourth of the population.