

## MP1-10

### **Pulmonary hypertension in the preterm infant with bronchodysplasia can be caused by pulmonary vein stenosis: a must-know entity**

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**Background:** Pulmonary vein stenosis is a rare cardiac defect. It has been described associated to other congenital heart defects and to prematurity. Haemodynamically it typically causes postcapillary pulmonary hypertension but a precapillary component can be observed.

**Objective:** Retrospective analysis from 1998 till 2012 in two French pediatric congenital cardiac centers of all premature infants with pulmonary vein stenosis. Diagnostic modalities, haemodynamics, pulmonary vein anatomy and outcome are described.

**Results:** 15 premature infants <37 weeks of gestation with the diagnosis of pulmonary vein stenosis were identified. Median gestational age at birth was 28+5 weeks (range: 25+5–35 weeks). Median birth weight was 790 g (range: 585-1500g). Nearly all the infants (86%) had bronchodysplasia. 27% (n=4) had associated cardiac defects other than persistent arterial duct or secundum atrial septal defect. Six infants (40%) had a first cardiac catheterization for exploration of pulmonary hypertension without visualization of pulmonary vein stenosis. In 73% of infants the diagnosis of pulmonary vein stenosis was suspected by echocardiography during follow-up. The remaining infants were diagnosed during cardiac catheterization, by pulmonary computer tomography and one during cardiac surgery. Median age at diagnosis was 6.8 months (range: 1.5-71 months). The majority of infants (60%) had initially unilateral pulmonary vein stenosis affecting in 89% one of the left pulmonary veins. Median initial mean pulmonary artery pressure (PAP) at diagnosis was 40 mmHg (range: 24-70 mmHg). Treatment modalities included: surgical intervention (pulmonary venoplasty, sutureless, lobectomy, heart and lung transplantation) for 6 patients, decision of non-intervention in 6 patients, interventional cardiac catheter (percutaneous pulmonary vein dilatation) in 2 patients and additional medical treatment for pulmonary hypertension in three patients. Overall mortality was high with only 46% of patients still alive at latest follow-up (median: 6 years; range: 1.2-10.9). Median follow-up until death was 7.2 months (range: 3.6-12.1 months).

**Conclusion:** Pulmonary vein stenosis is an unusual cause of pulmonary hypertension in the premature infant with bronchodysplasia. Diagnosis can be difficult since initial echocardiography can be normal and the disease progressive. The diagnostic method of choice is cardiac catheterization. Treatment options are numerous by surgical or interventional means but prognosis remains extremely poor.