Pulmonary hypertension can be caused by pulmonary vein stenosis: a must-know entity

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Pulmonary hypertension (PH) is a common problem in the extremely premature infant. Chronic pulmonary disease (CLD) is mostly the reason. Pulmonary vein (PV) stenosis on the contrary is a rare cardiac defect which has been reported in association with prematurity and other heart defects. Haemodynamically it typically causes postcapillary PH but a precapillary component can be observed.

Characteristics of the study population
- Median age at birth: 28+5 WG (25+5-35)
- Median birth weight: 790 g (585-1500)
- Bronchodysplasia: 86% n=13
- Associated cardiac defects: 93% n=14
  - ASD: 60% n=9
  - PDA: 40% n=6
  - VSD: 27% n=4
  - Coarctation, hypoplastic arch: 13% n=2
- Median age at diagnosis: 6.8 months (1.5-71)
- Median PAPm: 40 mmHg (24-70)

Results: 15 premature infants with PH due to PV stenosis were identified. The majority (86%) had moderately or severe CLD due to prematurity. 73% of infants had initially a normal echocardiography and the diagnosis of PV stenosis was suspected during follow-up. 27% had a first cardiac catheterization for causal exploration of PH without visualization of PV stenosis.

Modalities of diagnosis of PV stenosis
- 1st cardiac catheterization: n=2
- 2nd cardiac catheterization: n=1
- Cardiac catheterization + CT: n=8
- Surgery: n=3

PV stenosis at diagnosis
- One or more LPV: 13% n=2
- LPV + one RPV: 13% n=2
- All PV: 33% n=5
- One or more RPV: 40% n=6

Treatment strategy after diagnosis
- No intervention: 7% n=1
- Interventional cardiac catheterisation: 7% n=1
- Surgical intervention: 13% n=2
- Only medical treatment (PAH): 33% n=5
- Transplantation: 13% n=2

Methods: Retrospective analysis from 1998 – 2012 in two French Cardiac centers. Focus was on diagnostic mode, haemodynamics and outcome.

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PV stenosis is an unusual cause of PH 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. In case of non intervention the disease mostly leads to death due to RV failure because of severe PH. Surgical and interventional treatments exist but prognosis remains compromised with a high mortality in the first months after diagnosis. Decision to intervene has to take in consideration associated co-morbidities in this setting.