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

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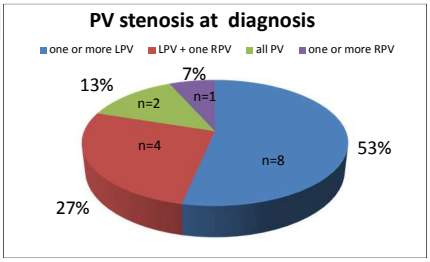
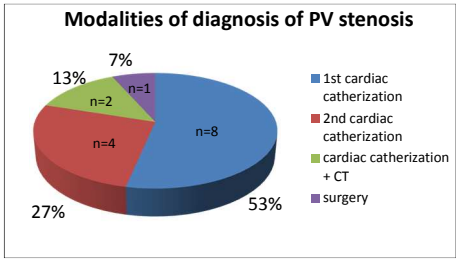
Pulmonary hypertension (PH) is a common problem in the extremely premature infant. Chronic pulmonary disease (CLD) is mostly the reason. Pulmonary vein (PV) stenosis on the contrary is a rare cardiac defect which has been reported in association with prematurity and other heart defects. Haemodynamically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

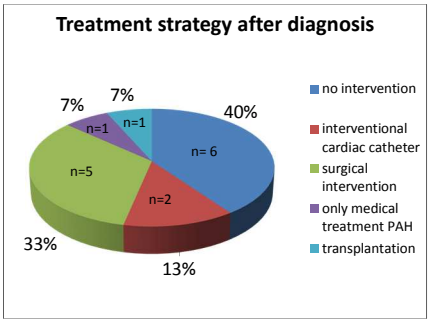
Methods: Retrospective analysis from 1998 – 2012 in two French Cardiac centers. Focus was on diagnostic mode, haemodynamics and outcome.

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.

Caracteristics 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)

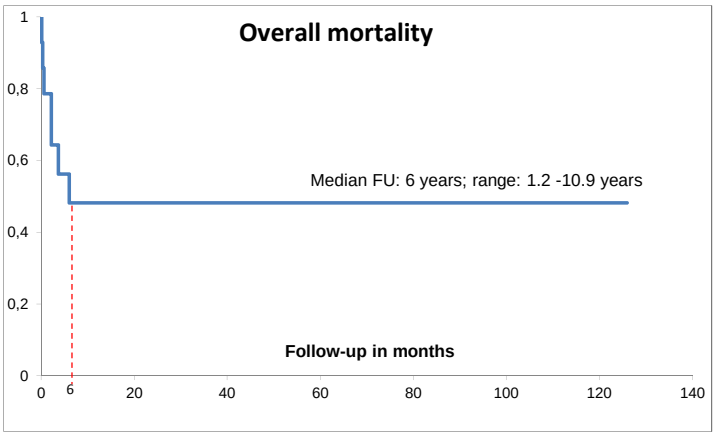


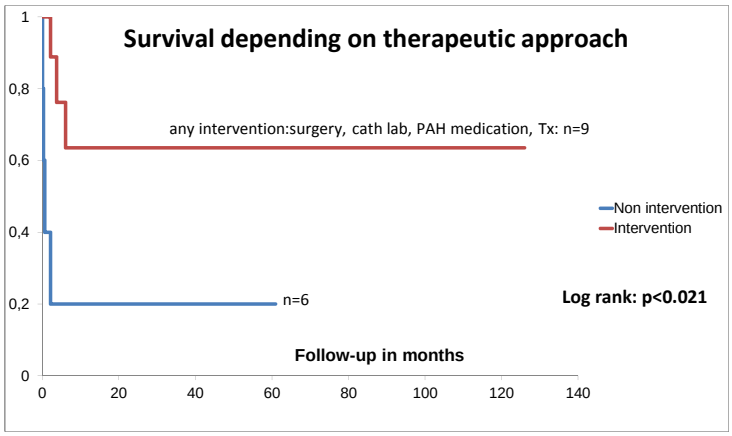




Case N°	Birthweight	Localisation	Treatment strategy	Outcome
1	1420 g	LPV, SRPV	no intervention	died
2	585 g	SLPV	no intervention	died
3	910 g	LPV,RPV	no intervention	lost to FU
4	900 g	LIPV	surgical plasty of 4 PV	alive
5	610 g	LPV, IRPV	surgical plasty LPV, right bilobectomy	died
6	1500 g	ILPV	percutaneous dilatation of ILPV twice	died
7	760 g	SRPV	percutaneous dilatation of SRPV medical treatment PAH	alive
8	1040 g	ILPV	left inferior lobectomy medical treatment PAH	alive
9	780 g	LPV	no intervention	alive
10	790 g	LPV,RPV	no intervention	died
11	970 g	SLPV	1st Op: plasty LSPV; 2nd OP: sutureless	died
12	660 g	LPV, SRPV	no intervention	died
13	680 g	LPV, SRPV	stent PV ->heart lung transplantation	alive
14	955 g	LPV	medical treatment PAH	alive
15	680 g	LPV	sutureless LPV	alive

Survival was significantly better in the intervention group, but even then global mortality remained high **(37%),** especially in the first six months after diagnosis. Medical decision for non intervention was taken after parental consent based on importance of PV stenosis and disease progression, severity of prematurity, importance of associated CLD and cardiac and extracardiac co-morbidities.





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.