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Title: Acute vasodilator response in different forms of pre-capillary pulmonary hypertension

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Body: Background: acute vasodilator testing (AVT) is well established in idiopathic pulmonary arterial hypertension (IPAH). Few data exist about its use in other forms of pulmonary arterial hypertension (PAH) or chronic thromboembolic pulmonary hypertension (CTEPH). Objetives: Analyze results of AVT in non-IPAH and CTEPH; and its predictive value on patient status over 1yr of calcium channel blockers (CCB), compared with IPAH. Methods: clinical and haemodynamic parameters were reviewed in 232 consecutive patients with pre-capillary PH (53±16 yrs; 66% female; mPAP 46±12 mmHg; CI 2.53±0.86 l.min.m²; PVR 829±473 din.s.cm). Patients were classified in IPAH (n=58, 25%), PAH associated with connective tissue disease (PAH-CTD) (n=40, 17.2%), HIV (PAH-HIV) (n=31, 13.4%), portal hypertension (PoPH) (n=53, 22.8%), congenital heart disease (PAH-CHD) (n=10, 4.3%) and CTEPH (n=40, 17.2%). Venice criteria defined a positive response. Long-term responders to CCB (LTR-CCB) were defined as achieving functional class I-II, 6 min walk distance ≥450m and no addition of targeted therapy. Results: shown in table.

Acute vasodilator testing and long-term response to CCB

	IPAH	PAH-CTD	PAH-HIV	PoPH	PAH-CHD	CTPH
Positive AVT, n (%)	10 (17.9)	5 (13.5)	0*	5 (10.6)	0	4 (11.4)
LTR-CCB, n	7	0	--	3	--	2
% LTR-CCB among Positive AVT	70	0*	--	60	--	50*
% LTR-CCB among overall	12	0	--	5.6	--	5

*p<0.05 compared with IPAH

Conclusion: As in IPAH, a proportion of patients with PAH-CTD, PoPH and CTEPH show positive AVT. Whereas at least 50% of acute responders with PoPH and CTEPH show long-term response, no patient

with PAH-CTD present long-term response. AVT and vasodilator treatment is justified in PoPH and CTEPH, although with close clinical control.