

## CLINICAL PRESENTATION OF PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION AND PULMONARY ARTERIAL HYPERTENSION: A DATABASE PRELIMINARY STUDY.

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**BACKGROUNDS:** Chronic thromboembolic pulmonary hypertension (CTEPH) and pulmonary arterial hypertension (PAH) are two rare diseases of different etiologies resulting in an elevation in mean pulmonary artery pressure (PAPm). Data comparing the clinical presentation of both diseases are lacking.

**METHODS:** We performed a retrospective database review on 860 CTEPH and 506 PAH patients between 2000 and 2011 and analysed clinical presentation and exercise capacity at the time of diagnosis +/- 3 months.

**RESULTS:** table 1.

		<b>CTEPH</b>	<b>PAH</b>	<b>P</b>
	n	860	506	/
	sex(M), %	52	28	<b>&lt;0.0001</b>
CAMPHOR	n	244	266	/
	S	12.02 ± 5.81	12.52 ± 6.63	0.3632
	A	11.22 ± 5.94	12.78 ± 7.57	<b>0.0105</b>
	QOL	10.55 ± 6.46	10.64 ± 7.16	0.8750
6MWT	n	597	434	/
	6MWD, m	290 ± 124	262 ± 122	<b>0.0004</b>
	SaO <sub>2</sub> , %	88 ± 6	85 ± 10	<b>&lt;0.0001</b>
TLCO	n	381	245	/
	TLCO, %pred	32 ± 7	27 ± 11	<b>&lt;0.0001</b>
BNP	n	149	122	/
	NT-proBNP,pg/mL	2231 ± 2575	2689 ± 3305	0.2125
AGE	n	425	371	/
	AGE,y	58 ± 15	57 ± 17	0.2448
WHO	I,%	0.7	1.1	0.5777
	II,%	16.3	16.2	0.9676
	III,%	69.1	62.2	<b>0.0350</b>
	IV,%	11.8	18.1	<b>0.0129</b>
RHC	PAPm,mmHg	42.1 ± 13.9	49.7 ± 15.6	<b>&lt;0.0001</b>
	RAPm, mmHg	9.3 ± 6.3	9.1 ± 5.9	0.5835
	PCWP, mmHg	11.5 ± 5.9	11.9 ± 7.1	0.3519
	CO, L/min	3.94 ± 1.16	3.65 ± 1.27	<b>0.0012</b>
	CI, L/min.m <sup>2</sup>	2.09 ± 0.56	2.12 ± 0.71	0.4001
	PVR, woods	8.9 ± 5.8	11.0 ± 6.1	<b>&lt;0.0001</b>

CAMPHOR: quality of life evaluated by the camphor questionnaire (S:symptoms, A:activity, QOL quality of life), 6MWT:6minute walk test, SaO<sub>2</sub>: saturation of blood in oxygen at the end of the 6MWT, TLCO: total lung diffusion capacity for carbon monoxide, NT-proBNP: N-terminal fragment of pro- brain natriuretic peptide, WHO: world health organisation functional class, RHC: right heart catheterisation, RAPm: right auricular pressure, PCWP : precapillary wedge pressure, CO: cardiac output, CI: cardiac index, PVR: pulmonary vascular resistance.

When scleroderma patients are excluded from the PAH population the 6MWdistance (274±120 m) and TLCO%pred (30±11%) are similar to that of CTEPH group. (P=0.0573 and P = 0.15 respectively).

CONCLUSIONS: CTEPH patients present at earlier state of the disease, more often in WHO class III and with lower PAPm and PVR. CTEPH patients tend to have longer 6MWdistance and higher TLCO.