

**LONG TERM QUALITY OF LIFE (QoL) IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH): A COMPARISON OF LONGITUDINAL OBJECTIVE AND PATIENT REPORTED OUTCOMES FOLLOWING COMMENCEMENT PULMONARY ARTERIAL HYPERTENSION TARGETED MONOTHERAPY**

N. Doughty,<sup>1</sup> J Twiss<sup>2</sup> S., P. McKenna,<sup>2</sup> J. Pepke-Zaba<sup>1</sup>

1. Papworth Hospital Pulmonary Vascular Disease Unit, (PVDU) Cambridge, UK.

2. Galen Research, Manchester UK

**Objective:** To determine long term changes in QoL measured by Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR) , 6MWT and NT-Pro BNP in patients with distal or residual CTEPH .

**Methods:** Data obtained retrospectively from database at Papworth PVDU .Patients with diagnosis of CTEPH not suitable for (distal distribution of disease), and with significant residual Pulmonary Hypertension (PH) at right heart catheterization (RHC) 3-12 months post Pulmonary Endarterectomy (PEA). We included patients treated with mono targeted therapy at baseline for which data were available with at least one follow up within first year.

**Results:** n=72. (45f) Mean age 61yrs in WHO class III/IV (70/2). 38 discontinued due to clinical worsening requiring additional intervention. 37=sildenafil, 30=bosentan, 5= prostanoids. Significant changes were observed: 1) Initial improvement of CAMPHOR scores at 4 and 12 months were not maintained at 2 years; Symptom and Activity scores worsened between 12 and 24 months. 2) The 6 minute walk test (6MWT) improved and retained statistical significance for two years.3) Reduced levels of NT-Pro-BNP up to two years were observed. Data was analyzed with Microsoft excel and presented as Mean (SD). Change scores significance analysed using Wilcoxon Signed Rank test. (p<0.05)

**Table 1:**

	6 minute walk test	CAMPHOR symptoms	CAMPHOR activity	CAMPHOR QoL	NTProBNP
<b>Baseline to 4 months</b>					
n	55	59	58	59	40
Change score mean (SD)	35.9 (77.3) (increase)	1.8 (4.9)	0.87 (4.0)	2 (4.9)	892.8 (1868.3) (decrease)
p	0.0008*	0.011*	0.132	0.002*	0.001*
<b>Baseline to 12 months</b>					
n	55	59	58	59	40
Change score mean (SD)	35.9 (77.3) (increase)	1.8 (4.9)	0.87 (4.0)	2 (4.9)	892.8 (1868.3) (decrease)
p	0.0008*	0.011*	0.132	0.002*	0.001*
<b>Baseline to 24 months</b>					
n	39	45	45	45	30
Change score mean (SD)	29.3 (78.3) (increase)	0.8 (4.6)	0.1 (4.4.) (increase)	1.3 (5.4)	733.5 (1893.7) (decrease)
p	0.029*	0.2	0.69	0.06	0.01*
<b>4 months to 12 months</b>					
n	53	52	53	53	38
Change score mean (SD)	13 (72.2) (increase)	0.3 (3.7)	0.4 (4.0) (increase)	0.54 (3.6)	319.5 (949) (increase)
p	0.35	0.83	0.43	0.4	0.007*
<b>12 months to 24 months</b>					
n	36	42	41	42	28
Change score mean (SD)	316 (115.3) (decrease)	1.4 (4.6) (increase)	1 (3.0) (increase)	0.8 (4.5) (increase)	267.3 (726.7) (increase)
p	0.067	0.05*	0.02*	0.32	0.36

**Conclusion:** In response to PAH targeted monotherapy, patients with distal or residual CTEPH show significant improvement up to 12 months in objective and patient self-reported outcomes. Between 12 and 24 months all measures showed deterioration, though only the CAMPHOR showed a statistically significant difference between 12 and 24 months.