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Long-Term Outcome Of Patients With Chronic Thromboembolic Pulmonary Hypertension: Results Of An International Prospective Registry Comparing Operated Versus Non Operated Patients

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Abstract Body

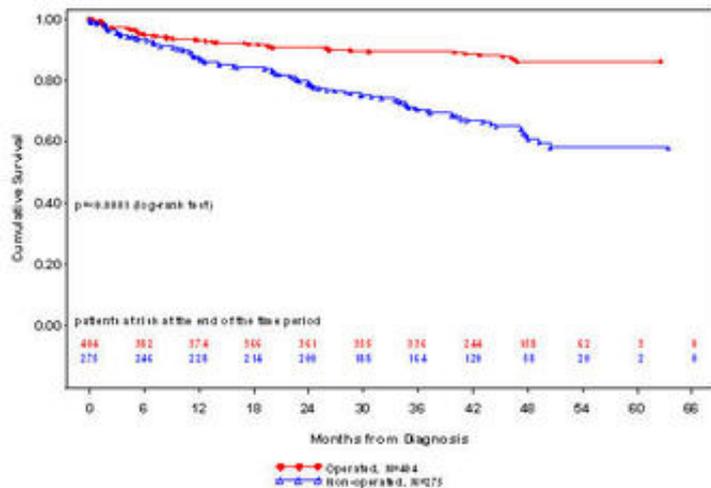
< **Introduction:** The only potentially curative treatment of chronic thromboembolic pulmonary hypertension (CTEPH) is surgery (pulmonary endarterectomy). The immediate postoperative mortality rate of pulmonary endarterectomy (PEA) has been decreased to less than 5%. After PEA the majority of patients has a dramatic improvement in hemodynamics, functional status and long-term survival. However a substantial number of patients with CTEPH are not operated; in this population lifelong oral anticoagulation is strongly recommended, whereas the role of pulmonary arterial hypertension (PAH) targeted therapy is uncertain.

Methods: A total of 679 incident patients with CTEPH were included in an international prospective registry over a 24 months period. Patients were on anticoagulation for at least 3 months prior to diagnosis, were older than 18 years at diagnosis and not treated with PH targeted medication before diagnosis.

Results: 404 (60 %) patients were operated. 275 (40%) patients were not operated and received at least one specific PAH therapy during the follow up in 61 % of cases. The main reasons for not being operated were: inaccessible occlusion (112), too high PVR (28), co-morbidity (37), refusal (38), others (60). For all patients the minimum follow up was 3 years with a maximum of 5 years. The non-operated population was older, predominantly female, had a lower FEV1, a lower walking distance, less history of acute venous thromboembolism and a more distal disease on angiography; in contrast, NYHA functional class, RAP and PVR were similar (Table).

Table: Patient Characteristics			
	Operated n=404	Non-operated n= 275	p-value
Age (years)	60 (18-84)	67 (22-86)	<0.0001
Gender (% male)	55	43	0.0038
6MWD (m)	340 (20-700)	315 (11-677)	0.0045
NYHA class I-II/III/IV (%)	19/69/12	18/69/13	ns
mRAP (mmHg)	9 (0-38)	8 (0-40)	ns
PVR (dynes.s.cm-5)	728 (97-2880)	676 (165-2800)	ns
FEV1 (L in 1 sec)	2.5 (0.9-5.0)	2.2 (0.7-4.7)	<0.0001
History of acute VTE (%)	81	72	0.0040
COPD (%)	8	13	0.0594
Angiographic obstruction:			
Main PA(%)	26	11	<0.0001
Lobar PA(%)	62	48	0.0050
Segmentan PA(%)	12	41	<0.0001
6MWD: 6-minute walk distance; NYHA; New York Heart Association; mRAP: mean right atrial pressure; FEV1: Forced Expiratory Volume; VTE: Venous thromboembolism; COPD: Chronic Obstructive Pulmonary Disease; PA: Pulmonary Artery			

Operated patients had a much better survival than non operated patients with a 89.3 % and 70.5 % survival rate at 3 years respectively (Figure); this difference was maintained after adjustment for age, gender, NYHA functional class, mean RAP and history of cancer. Both in operated and non-operated patients, NYHA functional class at diagnosis was the main prognostic factor.



Conclusion: in this incident cohort of patients with CTEPH referred to experienced centres, CTEPH is predominantly treated with surgery: 60 % of patients underwent pulmonary endarterectomy. Operated patients had a significantly better long term survival than non-operated patients. These data support the guidelines indicating that all patients with CTEPH have to be referred to an experienced surgical centre for evaluation of operability. In patients non eligible for surgery, the effect of medical therapy on long term outcome needs to be properly evaluated.

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