

The Europe, Middle East and Africa (EMEA) chronic thromboembolic pulmonary hypertension (CTEPH) registry: study design and baseline demographics

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The EMEA CTEPH registry is an ongoing, international, prospective, Phase IV observational study investigating the epidemiology, diagnosis, and treatment of CTEPH patients in EMEA countries.

Key inclusion criteria were ≥ 18 years, and CTEPH diagnosed by RHC. Patients will be monitored for 36 months post-enrollment. Primary assessments are change from initial to final visit in routine clinical parameters (i.e. 6MWD, WHO functional class, and hemodynamics). Secondary assessments include patient demographics, CTEPH subtype, diagnostic and treatment patterns, PEA eligibility and outcomes, and long-term outcomes (clinical parameters, mortality, complications, hospitalizations, healthcare provider visits).

Recruitment closed with 231 patients enrolled. Data presented are from the July 2018 data cut (n=205). The table shows patient baseline characteristics. Most patients were prevalent (173 [84%]; diagnosed >6 months prior to initial visit and received PH-targeted therapy); 32 patients (16%) were incident (diagnosed <6 months prior to initial visit and received no PH-targeted therapy). The commonest symptoms were shortness of breath (94%), fatigue (43%), edema (36%), and chest discomfort (30%). The commonest techniques used at

diagnosis were echocardiography (85%), ECG (72%), computer tomography angiography (67%), and RHC (66%). At initial visit, 42% of patients underwent RHC, and 94 patients (46%) were considered operable. Of the 64 patients (31%) who underwent PEA before initial visit, 50% experienced recurrent/residual PH, and 50% were not yet assessed. For the 109 inoperable patients, the reasons were non-patient related (23%), comorbidities (42%), and distal disease (35%). At initial visit, 34% of patients were not receiving PH-targeted therapy. In those 136 patients receiving therapy, the commonest were PDE5i (49%), riociguat (40%), and ERA (21%); 71% were receiving monotherapy and 25% were receiving combination therapy.

This registry is the first of its kind in EMEA countries and is expected to provide insight into diagnosis and treatment of CTEPH in these regions.

Baseline demographics		
Female, n (%)		134 (65)
Age, years		
	At diagnosis	53 (15)
	At registry inclusion	55 (15)
Race, n (%)		
	White	179 (87)
	Black/African American	1 (<1)
	Asian	27 (13)
	Not reported	1 (<1)
Body mass index, kg/m ²		31 (7)
History of venous thromboembolism, n (%)		140 (68)
	Length of hospitalization	21 (51)
Time between symptom onset and diagnosis, years		2.3 (3.1)
Distance of residence from specialist center, km, n (%)		
	<100	95 (46)
	≥100–<300	13 (6)
	≥300	72 (35)
	Undeterminable	25 (12)

Data are mean (standard deviation), unless otherwise stated