Purpose
A U.S. CTEPH Registry was organized to characterize the demographics, evaluation, and clinical course of CTEPH in the United States and to assess short and long term outcomes of surgical and non-surgical therapy of CTEPH in the U.S. health care system.

Background
Chronic thromboembolic pulmonary hypertension (CTEPH) is a rare, life-threatening condition that represents the most severe long-term complication of acute pulmonary embolism. Yet the symptoms of CTEPH, which include exercise intolerance, fatigue, and dyspnea, are nonspecific. As such, CTEPH may be underdiagnosed in patients presenting with such symptoms or may be misdiagnosed as pulmonary arterial hypertension due to a lack of disease awareness. CTEPH is the only form of PH that is potentially curable, and only with pulmonary thromboendarterectomy (PTE), so it is critical that it be appropriately diagnosed. The epidemiology and demographics of CTEPH have yet to be adequately described in the U.S. population.

Methods
Multi-center, prospective observational registry of newly diagnosed CTEPH patients. First subjects were enrolled in April 2015 with plans for a total enrollment of 750 subjects over a 3-year period at 30 U.S. sites.

Results
As of May 2017, 588 subjects were submitted for radiologic adjudication and 548 subjects were enrolled in the Registry (6.7% did not meet adjudication criteria). Median age = 56 yrs (range 16-87 yrs), 50.4% male. Caucasians comprise 72% of the population, Blacks/AA 21%, Asians 1.4%. Hispanic or Latino = 6%. WHO Functional Class: I (2%), II (20%), III (64%), IV (11%). History of DVT reported in 49% and acute PE in 87%. Lupus anticoagulant was the most common thrombophilia identified (13%). Most subjects were anticoagulated with warfarin (50%), with 35% treated with NOACS. Prior to PTE surgery, 54% of subjects were on pulmonary arterial hypertension target medical therapy (PAH therapy), the most common being sGCS (54% of those on PAH therapy, 28% of all subjects) followed by PDE-5 inhibitors (23% on PAH therapy, 12% all subjects), parenteral prostanoids (8% on PAH therapy, 4% all subjects), or ERAs (8% on PAH therapy, 4% all subjects) with 7% of subjects on more than one PAH therapy. 45% of subjects were on supplemental O2 at enrollment, 57% on diuretics.

Enrollment RHC mean values: RA 10 mmHg, mPAP 45 mmHg, PCWP 12 mmHg, C.O. 4.8 L/min, C.I. 2.5, TPR 10.3 WU. Operability assessments performed on 456 subjects by enrolling centers yielded 87% subjects operable, 10% inoperable and 3% unsure. Of the 440 subjects referred to surgical centers, 92% were deemed operable and 98% of those were referred for surgery. Early post-operative hemodynamics are available from 334 subjects: RA 8.6 mmHg, mPAP 26.3 mmHg, C.O. 5.8 L/min, TPR 4.7 WU. Fifteen subjects have undergone balloon pulmonary angioplasty.

Conclusion
Demographic characteristics of initial patients in the U.S. CTEPH Registry appear to be similar to those previously reported. Despite the vast majority of subjects being deemed operable, medical therapies for CTEPH/PAH are commonly being used. Preliminary data also confirms significant early hemodynamic improvement following PTE surgery.

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Abstract Title
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