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Title: Plasma high fibrinogen and low plasminogen levels predict poor prognosis in patients with inoperable chronic thromboembolic pulmonary hypertension

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Body: Background: Chronic thromboembolic pulmonary hypertension (CTEPH) comprises organizing thrombotic obstructions in the pulmonary arteries, and progressive pulmonary hypertension. However, there are few reports about the abnormality of coagulation and fibrinolysis system in CTEPH. Purpose: The aim of this study was to investigate correlation between coagulation and fibrinolysis system, and severity and prognosis in patients with CTEPH. Methods and results: We studied 91 patients (18 male, 55±14 [SD] years, mean pulmonary arterial pressure 45±12 mmHg) diagnosed as having inoperable CTEPH at Chiba University Hospital between 1986 and 2011. We retrospectively investigated the relationship between plasma fibrinogen and plasminogen levels, severity and prognosis. Eighteen patients who had both high fibrinogen level (≥291 mg/dl, median) and low plasminogen level (<100.5 ng/ml, median) showed significantly lower cardiac index (2.27±0.66 vs. 2.70±0.58 L/min/m², p=0.0079), higher pulmonary vascular resistance (PVR) (1030±602 vs. 746±340 dyne*sec*cm⁻⁵, p=0.0090), and poor survival (5-year survival 35.4 vs. 88.6%, p<0.001) compared with other 73 patients. Multivariate analysis revealed that plasma fibrinogen, plasminogen, and PVR were independent predictors for survival. Conclusion: Plasma high fibrinogen and low plasminogen levels are poor prognostic factors for patients with inoperable CTEPH.