

**LONG TERM OUTCOMES IN PATIENTS WITH SYMPTOMATIC LIVER  
HEREDITARY HEMORRHAGIC TELANGIECTASIA**

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*Introduction:* Symptomatic liver involvement by HHT is unusual and the natural history of patients with this manifestation is poorly understood.

*Methods:* Clinical chart review and analysis of patients evaluated at a single HHT center over a 12-year period.

*Results:* Of 2500 patients evaluated in this center, 51 presented with or developed symptoms related to liver HHT. Median age at initial evaluation was 64 years (range 17-73) years and 41/51 (80%) were women. Patients were initially classified into Type 1, symptomatic high cardiac output state (39/51 patients, 76.5%), Type 2, portal hypertension (9/51 patients, 17.5%), and Type 3, biliary abnormalities (3/51 patients, 6%). In the 39 Type 1 patients, mean cardiac index (CI) was 5.9 per minute per square meter (range 2.9-8.5), mean pulmonary capillary wedge pressure was 15 mm Hg (range 4-32), and mean pulmonary artery pressure was 28 mm Hg (range 9-61). Type 1 and 2 patients were treated with diuretics, correction of anemia and anti-arrhythmics to control dyspnea, edema and prevent recurrent atrial fibrillation. Type 3 patients with evidence of infection were treated aggressively with antibiotics and those with biliary pain were given frequent small meals and bile salts. During the mean follow-up of 4.8 years, 17/51 (33%) of patients died. In Type 1 patients, 10/39 died at a median of 4 years after initial evaluation due to spontaneous biliary duct necrosis and/or liver hemorrhage (4), right heart failure with superimposed complications (4), liver necrosis post-embolization (1) and unknown (1). In Type 2 patients, 6/8 died at a median of 4 years due to esophageal varices (1), intractable bleeding from gastrointestinal telangiectases (2), presumed pulmonary emboli (1), pancreatic cancer (1), and lung cancer (1). One of 3 Type 3 patients died after failed liver transplantation.

*Conclusions:* Aggressive symptomatic treatment helps to control heart failure symptoms in Type 1 HHT, preventing overt pulmonary edema. Right heart failure may contribute to death, but Type I patients also “cross-over” and may die of biliary necrosis or liver hemorrhage. Strategies to treat liver AVMs with transplantation need to be carefully formulated.