OUTCOMES OF SYMPTOMATIC LIVER VASCULAR MALFORMATIONS IN HEREDITARY HEMORRHAGIC TELANGIECTASIA

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HHT patients with symptomatic liver AVMs can present with high cardiac output heart failure (type 1), portal hypertension (type 2) or biliary ischemia (type 3), however, their natural history remains poorly understood. The clinical course and factors related to mortality were evaluated at a single HHT center over a 12-year period. Of 2500 HHT patients, 48 had symptomatic liver AVMs, including 37 with type 1, 8 with type 2, and 3 with type 3 manifestations. Age at initial evaluation was 58±12 years and 38 were women. Of the type 1 patients, 4 subsequently developed biliary ischemia and were classified as type 3 cross-over patients. Over 5.6±3.5 year follow-up, 16/48 (33%) patients died, including 6/33 type 1 (18%, age 74±5), 5/8 type 2 (63%, age 68±6), 1/3 type 3 (33%, age 74), and 4/4 type 3 cross-over (100%, age 52±15). Kaplan-Meier survival was best in type 1 patients (p=0.03). In bivariate analysis, type at last visit (p=0.004), refractory GI bleeding (p=0.02) and ascites (p=0.04) were associated with mortality. In type 1, cross-over to type 3 (p=0.003) and refractory GI bleed (p=0.01) were associated with mortality. Thus, in symptomatic liver HHT, type of presentation has important prognostic implications. Type 1 has a favorable prognosis unless biliary ischemia or severe GI bleeding develops.