HEART FAILURE AND PULMONARY HYPERTENSION IN PATIENTS WITH HEPATIC AVM AND HHT

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Background: Manifestations of hepatic AVMs in HHT syndrome include biliary disease, portal hypertension, and heart failure (HF).

Methods: We analyzed HHT patients with liver AVMs (n=37) to determine whether those with HF (n=26) had evidence of left ventricular (LV) systolic dysfunction, pulmonary or portal hypertension.

Results: The cardiac index was elevated in all patients, but was higher in those with vs. without HF (5.7 (0.2) vs. 4.3 (0.4) l/min/m^2, mean (SE), P<0.01). Mean PCWP (16 (1) vs. 12 (1.5) mmHg; P<0.05) and pulmonary artery pressures were also higher in patients with HF (29 (2) vs. 23 (3) mmHg, P=0.09). Elevated pulmonary pressures (mean $\geq$25 mmHg) were more common in those with vs. without HF (15/26 vs. 2/11; P<0.05), but the mean pulmonary vascular resistance index was normal in both groups (2.4 (0.3) and 3 (1) U? m^2; PNS). Severe pulmonary hypertension (systolic >60 mmHg) was present in 4/26 patients with HF and 2/11 without HF. The mean hepatic venous pressure gradient was lower in those with vs. without HF (3 (1) vs. 12 (3) mm Hg; P<0.05). Only 2 patients with HF had portal hypertension. Echocardiograms in symptomatic HF patients demonstrated normal LV ejection fractions (>50%), normal end-diastolic and end-systolic LV diameters (5.2 (0.2) and 3.1 (0.2) cm, respectively), but a tendency to left atrial enlargement (4.3 (0.2) cm).

Conclusions: Symptomatic HF occurs prior to the development of significant LV enlargement or systolic dysfunction in HHT patients with hepatic AVMs. In patients with HF, mild pulmonary hypertension is common, but is usually associated with normal pulmonary vascular resistance, while portal hypertension is typically absent. Severe pulmonary hypertension occurs infrequently in patients with liver HHT and is independent of the presence of HF.