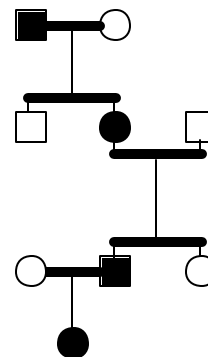


# IVth HHT Scientific Conference

Tenerife, Canary Islands, April 2001



## LIVER INVOLVEMENT IN HEREDITARY HEMORRHAGIC TELANGIECTASIA (HHT) – AN UPDATE ON OUTCOME

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**Background:** Liver involvement in HHT has been recently characterized in 19 symptomatic patients (New Engl J Med 2000;343:931-6). The aim of this study is to describe the evolution of 31 patients with liver HHT (19 from the original description plus 12 additional patients) followed for a median period of 24 months (range 0-72). All patients have angiographic or CT evidence of hepatic involvement by HHT and absence of concomitant liver disease.

**Results:** Of 31 patients, 17 (55%) belong to the high output heart failure (HF) group, 8 (26%) to the portal hypertension (PHT) type and 6 (19%) to the biliary (BIL) type. **1) HF** (n=17; one newly diagnosed, median follow-up 24 months): symptoms are stable in 10, improved in 4 patients on medical therapy and worsened in 2. One patient died after hepatic artery embolization for incapacitating HF. Another patient with stable HF died of biliary sepsis. **2) PHT** (n=8; median follow-up 20 months): 3 patients have improved on medical therapy, one patient has developed recurrent variceal hemorrhage, four patients have died (three from intractable gastrointestinal bleeding from telangiectases). One of these developed biliary sepsis prior to death. **3) BIL** (n=6; one newly diagnosed, median follow-up 31 months): liver enzymes and symptoms have improved spontaneously in three patients. One patient developed HF and another developed ascites, liver failure and death.

**Conclusions:** Liver HHT is an entity characterized by exacerbations and remissions as well as transitions from one type to another. The HF type can be managed medically in most patients, the PHT type carries the worst prognosis while the BIL type can be accompanied by spontaneous remissions.

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**Structured Scientific Programme:** Friday 20th - Monday 23<sup>rd</sup> April 2001  
**Informal Collaborative Workshops:** Tuesday 24th - Thursday 26<sup>th</sup> April 2001  
**Conference Venue:** Hotel Semiramis Puerto de la Cruz, Canary Islands

Conference Chairman: Claire Shovlin  
Scientific sub-committee: Michelle Letarte and Carmelo Bernabeu.  
Clinical sub-committee: Bob White, Kees Westermann, Henri Plauchu, and Poul Vase