

Liver Transplantation for High Output Heart Failure Secondary to HHT: A Case Report and Review of the Literature

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4 Months later	21	57/24(35)	25	10.7/4.9	12%
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Table 1: Summary of hemodynamic evaluation results through the course of treatment. Abbreviations: RA: Right Atrium, PA: Pulmonary pressure, PB nary

Liver transplantation is a treatment option in patients with HHT [4,15]. Indications for transplant include biliary necrosis and/or heart failure not responding to conventional therapy [4]. In the United States, patients with HHT may be eligible for MELD exception points if they have evidence intractable heart failure or if they develop bile duct necrosis [16].

The outcomes after liver transplant are good with the actuarial 1-, 5-, and 10-year patient and graft survival rates being 82.5% in the largest case series, 40 patients, from the European Liver Transplant Registry [15]. In a study by Boillot et al, the hyperdynamic circulation disappeared after liver transplantation in all patients studied. At time of last follow-up, all patients were asymptomatic and had normal computed tomography of the liver and right heart catheterization 6 months after transplant [17].

The disease can recur after transplant. In the series of Lerut et al, two first cousin females, transplanted for mixed cardiac and biliary HHT disease, presented with focal vascular dilatations throughout the allograft at 156 and 84 months post-liver transplant [15].

Our patient initially presented with heart failure that failed to respond to conventional medical therapy. She also failed to obtain a sustained response to AVM embolization and bevacizumab, leaving transplantation as the only viable option. Liver transplantation resulted in an improvement in the symptoms of heart failure as early as two months after transplant.

Patients with HHT benefit from a multidisciplinary approach to their care. Physicians treating patients with HHT should consider referring patients with HHT and intractable heart failure for a liver transplant evaluation.

References

1. Garcia-Tsao G, Korzenik JR, Young L, Henderson KJ, Jain D, et al. (2000) Liver disease in patients with hereditary hemorrhagic telangiectasia. *N Engl J Med* 343: 931-936.
2. McDonald J, Wooderchak-Donahue W, VanSant Webb C, Whitehead K, Stevenson DA5, et al. (2015) Hereditary hemorrhagic telangiectasia: genetics and molecular diagnostics in a new era. *Front Genet* 6: 1.
3. Brenard R, Chapaux X, Deltenre P (2010) Large spectrum of liver vascular lesions including high prevalence of focal nodular hyperplasia in patients with hereditary haemorrhagic telangiectasia: the Belgian Registry based on 30 patients. *Eur J Gastroenterol Hepatol* 22: 1253-1259.

4. Falgout M, Palda VA, Garcia-Tsao G, Reistoff U, McDonald J, et al. (2011) International guidelines for the diagnosis and management of hereditary haemorrhagic telangiectasia. *J Med Genet* 48: 73-87.
5. Sabbà C, Pompili M (2008) Review article: the hepatic manifestations of hereditary haemorrhagic telangiectasia. *Aliment Pharmacol Ther* 28: 523-533.
6. Ianora AA, Memeo M, Sabba C, Cirulli A, Rotondo A, et al. (2004) Hereditary hemorrhagic telangiectasia: multi-detector row helical CT assessment of hepatic involvement. *Radiology* 230: 250-259.
7. Dupuis-Girod S, Chesnais AL, Ginon I (2010) Long-term outcome of patients with hereditary hemorrhagic telangiectasia and severe hepatic involvement after orthotopic liver transplantation: a single-center study. *Liver Transpl* 16: 340-347.
8. Garcia-Tsao G (2007) Liver involvement in hereditary hemorrhagic telangiectasia (HHT). *J Hepatol* 46: 499-507.
9. Bauer T, Britton P, Lomas D, Wight DG, Friend PJ, et al. (1995) Liver transplantation for hepatic arteriovenous malformation. *Am J Surg* 170: 100-103.