Eur J Vasc Endovasc Surg (2016) 51, 594–601

Clinical Outcomes of Patients with Severe Hepatic Hereditary Hemorrhagic Telangiectasia After Banding of the Hepatic Artery and Banding/Ligation of Branches of the Hepatic Artery

Z.-C. Liu [a], X.-F. Lu [a,b], H. Yang [a], H.-D. Liu [a], X. Song [c], S.-L. Ning [a], Y.-F. Xu [a], Y.-X. Chen [a,*]

[a] Department of Hepatobiliary Surgery, Qilu Hospital of Shandong University, Jinan 250012, Shandong Province, China
[b] Department of General Surgery, Jinan Central Hospital, Jinan 250013, Shandong Province, China
[c] Department of General Surgery, Shandong Provincial Hospital, Jinan 250021, Shandong Province, China

WHAT THIS PAPER ADDS

The liver is one of the most commonly affected organs in the context of hereditary hemorrhagic telangiectasia (HHT). If left untreated, liver complications and flow related heart failure with serious outcome are seen. This paper evaluates the technical and clinical outcomes of double hepatic artery banding/ligation in patients with symptomatic hepatic HHT and reports excellent results, suggesting that double hepatic artery banding/ligation is technically feasible and effective in selected patients with hepatic HHT.

Objective/background: To evaluate the effectiveness of double banding/ligation of hepatic arteries in treating patients with hepatic hereditary hemorrhagic telangiectasia (HHHT).

Methods: From January 2004 to December 2013, 35 patients were diagnosed with HHHT, among whom 11 woman and two men with a mean ± SD age of 44 ± 9 years were treated by double hepatic artery banding/ligation for cardiac insufficiency and/or portal hypertension. The outcomes were evaluated prospectively by measuring clinical manifestations, imaging features, liver and cardiac function, pulmonary arterial systolic pressure, and post-operative complications. Quality of life was evaluated with the Short Form Health Survey questionnaire.

Results: For each patient, the common hepatic artery and one branch of the left and/or right hepatic artery were banded, and other significantly dilated hepatic artery branches were ligated. No patient died after surgery. Clinical symptoms were improved in all patients, although ischemic cholangitis was observed in two patients and treated conservatively. Cardiac function, classified per the New York Heart Association (NYHA) cardiac functional grading, improved (NYHA III–IV vs. NYHA I–II); pulmonary arterial systolic pressure significantly decreased in all patients (48 ± 8 mmHg vs. 24 ± 4 mmHg; $P < .001$) and remained in the normal range (26 ± 3 mmHg) at the end of follow up. The levels of γ-glutamyl transpeptidase and alkaline phosphatase decreased in 11 patients (144 ± 94 U/L vs. 71 ± 34 U/L; $P = .003$) and 10 patients (207 ± 71 U/L vs. 105 ± 32 U/L; $P = .001$), respectively. Patients were followed up for 50 ± 28 months (range 6–113 months); one death resulted from causes unrelated to surgery and all dimensions of quality of life improved in all surviving patients.

Conclusions: This study helps to establish double hepatic artery banding/ligation as an effective therapy for selected patients with HHHT.

Article history: Received 22 April 2015, Accepted 12 December 2015, Available online 1 February 2016

Keywords: Arteriovenous malformations, AVM, Cardiac insufficiency, Hepatic artery, Liver, Portal hypertension

INTRODUCTION

Hereditary hemorrhagic telangiectasia (HHT), also termed Rendu–Osler–Weber disease, is a multisystemic vascular dysplastic disorder inherited in an autosomal dominant manner. It is characterized by mucocutaneous telangiectases with hemorrhagic tendency and arteriovenous malformations (AVMs) in visceral organs such as liver, lung, brain, and gastrointestinal tract.[1] Its prevalence has been estimated to range between 1 in 5,000 and 1 in 8,000 in the general population.[2] Liver involvement has recently been reported in 41–78% of patients with HHT.[3–5] Although < 10% of patients are symptomatic,[3] patients may develop severe complications such as high output cardiac failure (HOCF), portal hypertension, biliary ischemia, or encephalopathy, which mainly result from high flow

* Corresponding author.
E-mail addresses: surgeonchenyuxin@163.com; yxchen@sdu.edu.cn (Y.-X. Chen).

http://dx.doi.org/10.1016/j.ejvs.2015.12.020

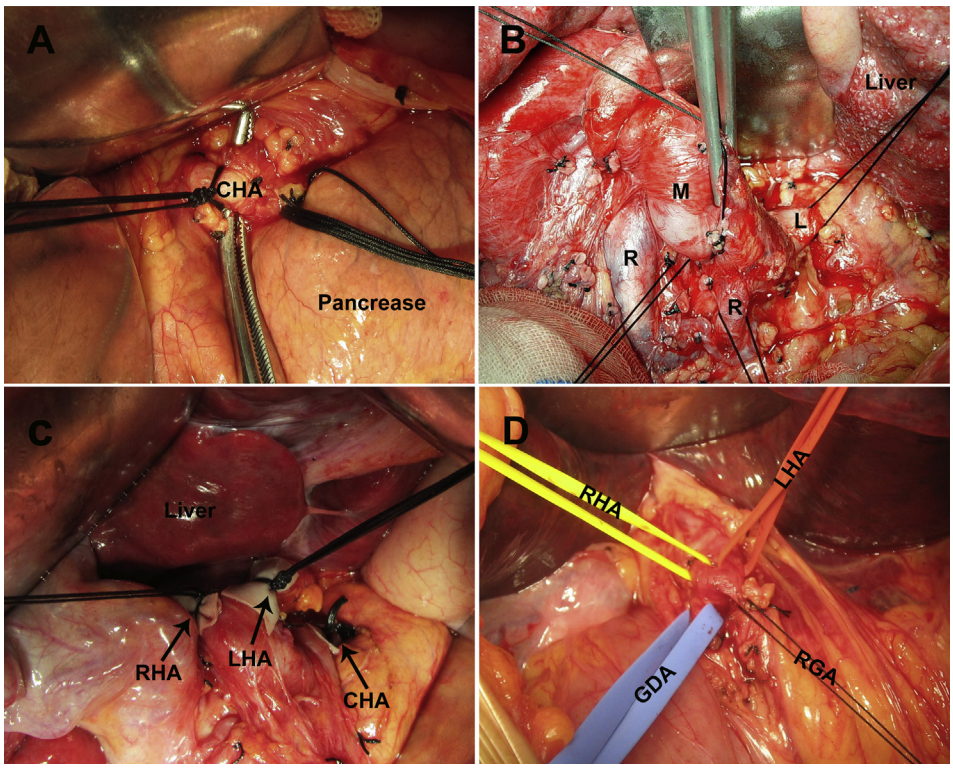

Figure 1. Intra-operative photographs. (A) The origin of common hepatic artery was exposed and banded. (B) Enlarged hepatic arteries were tortuous with a cavernous distribution and were dissected carefully. (C) Banding of the common hepatic artery (CHA) and left (L) hepatic artery (LHA), and ligation of right (R) hepatic artery (RHA). Hepatic arteries were encased by a pericardial patch. (D) Branches of CHA and collateral vessels. Note. M = middle hepatic artery; GDA = gastroduodenal artery.

hepatic vascular anomalies with arteriovenous shunting (arteriovenous, arterioportal, portovenous, or a combination).[6,7]

It is generally accepted that symptomatic patients with hepatic HHT (HHHT) should be treated.[6–9] For patients with complicated liver AVMs failing to respond to intensive medical treatment, several surgical approaches, including arterial embolization and liver transplantation,[10–14] have been proposed.[15] The feeding artery to a liver with AVMs is often a large caliber vessel leading to hyperperfusion of the liver.[9] Furthermore, AVMs grow in an expanding pattern due to the high blood flow between the arteries, and veins and cardiac output are significantly correlated with the diameter of the common hepatic artery in these patients.[16,17] Thus, ligation or banding of these dilated arteries can be a potentially effective treatment, which has been used in limited cases with undefined long-term results.[18–23]

In this study, the clinical data of patients with HHHT treated by double hepatic artery banding/ligation (both at the origin of the common hepatic artery and where its branches empty into the liver) were retrospectively analyzed in order to determine long-term outcomes.

MATERIALS AND METHODS

Protocol

From January 2004 to December 2013, 35 consecutive patients were diagnosed with HHHT at Qilu Hospital of Shandong University, China. Diagnosis depended on the Curacao criteria: epistaxis, telangiectasia, visceral AVM, and family history (a first degree relative with HHT). A definite diagnosis was made if three of the four criteria were met.[24] Twenty-two patients were not treated by double hepatic artery banding/ligation because they were either asymptomatic, had extensive biliary disease, significant portovenous shunting with symptoms suggestive of portosystemic encephalopathy, cirrhosis with impaired liver function, or irreversible cardiopulmonary changes. Symptomatic treatment for these patients included correction of anemia and arrhythmias, pain control with analgesics, and systemic antibiotics when indicated. Complications of portal hypertension were treated as recommended in cirrhotic patients.[25] A total of 13 patients with HHHT treated with double hepatic artery banding/ligation were enrolled in this study. The study protocol was reviewed and approved by the ethics committee of Qilu Hospital of Shandong University, and written informed consent was obtained from all patients.

Clinical presentations were assessed pre-operatively. Quality of life was evaluated using the Short Form-36 Health Survey (SF-36) questionnaire before operation. These patients were evaluated pre-operatively by computed tomography (CT) and CT angiography (CTA) to confirm the existence of AVMs in the liver or other organs such as brain and lung. All the patients were classified according to the New York Heart Association (NYHA) cardiac functional grading and Child-Pugh liver functional grading. Pulmonary