

CASE REPORT

Laparoscopic Double Hepatic Artery Banding/Ligation for Patients With Hepatic Hereditary Haemorrhagic Telangiectasia (HHHT)

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Introduction: Hepatic hereditary haemorrhagic telangiectasia (HHHT) is a rare autosomal dominant genetic disease. Some patients may develop cardiac failure, portal hypertension, and biliary ischaemia. To date, there is no standard surgical treatment for HHHT. The present authors propose a move from open to laparoscopic surgery; however, laparoscopic surgery has not been reported previously as a surgical treatment for HHHT.

Report: Two women were admitted with histories of exertional dyspnoea and upper abdominal pain, respectively. Combined with recurrent epistaxis and their positive family history, a diagnosis of clinical HHHT was made based on Curacao criteria after comprehensive evaluation of imaging features. Next generation sequencing (NGS) results also confirmed typical gene mutations responsible for HHT. Both patients underwent laparoscopic double hepatic artery banding and or ligation successfully and were discharged four to six days after operation without severe complications. The symptoms of cardiac insufficiency including exertional dyspnoea and shortness of breath of the first patient improved six months after the operation. The second patient, with epigastric pain, remained pain free without medication three months after the operation.

Discussion: Laparoscopic surgery for HHHT is technically challenging. Clinical data and follow up information showed that laparoscopic double hepatic artery banding and or ligation was a technically feasible surgical approach for HHHT patients with simple hepatic artery dilation.

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INTRODUCTION

Hereditary haemorrhagic telangiectasia (HHT), also termed Rendu-Osler-Weber disease, is a rare autosomal dominant genetic disease and may cause high output cardiac failure, portal hypertension, and biliary lesions.¹ Although several surgical strategies including hepatic artery banding, arterial embolisation, and liver transplantation have been proposed for HHHT, there is no standard surgical treatment.² The authors reported the effectiveness and feasibility of double hepatic artery banding and or ligation for the treatment of HHHT in 2016.³ With advances in laparoscopic surgery, most abdominal operations can be completed laparoscopically and the minimally invasive approach can greatly reduce peri-operative pain, however, the laparoscopic approach has never been reported for HHHT treatment. Recently, the present authors succeeded in performing laparoscopic double hepatic artery banding and or ligation, based on open experience, in two

female HHHT patients. The surgical procedures including clinical data are described. To the present authors' knowledge, this is the first description of laparoscopic double hepatic artery band and or ligation for HHHT treatment.

REPORT

Case 1

A 48 year old woman was referred with a two year history of exertional dyspnoea. Physical examination found multiple mucocutaneous telangiectasia on the lips and tip of the tongue (Fig. 1A). Abdominal contrast computed tomography (CT) revealed a dilated common hepatic artery with a diameter of approximately 12 mm, arteriovenous fistulae, diffuse intrahepatic vascular malformations, splenomegaly, and many hyperplastic nodules (Fig. 1B and C). Doppler echocardiography estimated that pulmonary artery systolic pressure was approximately 124 mmHg and right heart catheterisation (RHC) confirmed notable pre-capillary pulmonary hypertension and higher cardiac output (7.2 L/min).

Case 2

A 30 year old woman was admitted with a two month history of upper abdominal pain. Abdominal contrast CT

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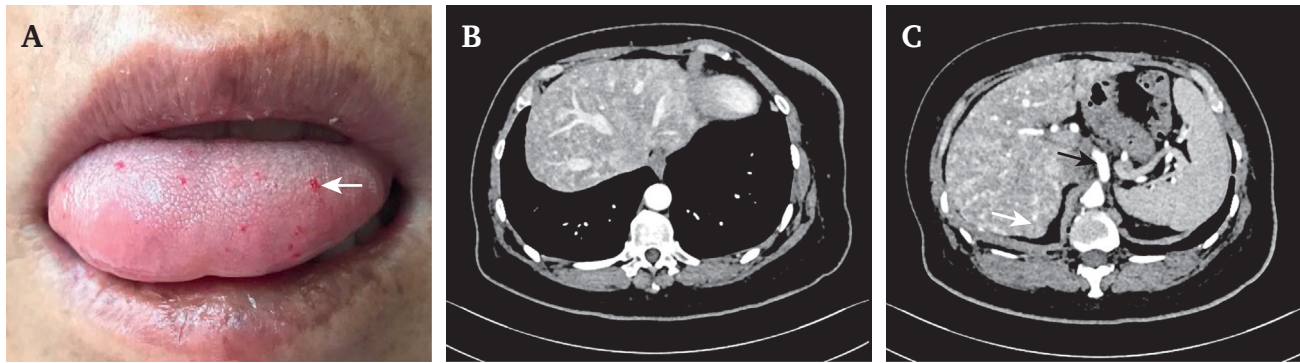


Figure 1. (A) Multiple telangiectasias were located on the tongue mucosa (white arrow). (B) Contrast computed tomography (CT) showed that the hepatic vein was enhanced earlier during the arterial phase. (C) CT showed dilated and tortuous hepatic arteries (black arrow), splenomegaly and multiple hyperplastic nodules (white arrow), indicating the existence of portal hypertension.

showed dilation of the coeliac trunk and common hepatic artery with a diameter of almost 10 mm (Fig. 2A), and diffuse intrahepatic vascular malformations. In addition, some hyperplastic nodules were found in the parenchyma of the liver (Fig. 2B). Cardiac function was normal and Doppler echocardiography estimated that the pulmonary artery systolic pressure was approximately 26 mmHg, which is within the normal range.

Operation procedure and results

Patients were placed in the supine position with their legs separated. The observation trocar was placed in the inferior margin of the umbilicus. Four operation ports were made in the midclavicular and anterior axillary lines symmetrically. Diffuse telangiectasias were found on the liver surfaces of both patients (Fig. 3A and B). Surgical management was focused on the origin of the common hepatic artery, arterial branches flowing into the liver, and collateral circulation.

The first step was to dissect the origin of the common hepatic artery and band it until the desired blood flow reached 200–300 mL/min while monitoring via intra-operative laparoscopic ultrasound (Fig. 3C), which was consistent with the authors' previous experience from open operations. Thus, the hyperperfusion of the liver could be

controlled, but the blood supply was kept sufficient for the biliary system. A non-absorbable number 0 silk suture is recommended for a longer effective time and for yielding granulomata.

After the hepatic artery hyperdynamic flow was alleviated, it was safer to manage the tortuous branches of the hepatic artery that supplied the liver. The branches that feed the liver lobes that were predominantly involved with arteriovenous malformations (AVMs) were ligated completely, and the other branches received banding. The gastroduodenal artery and other innominate collateral branches arising from the aorta, superior mesenteric artery, and left gastric artery were ligated or clipped for all patients to avoid recurrence of liver hyperperfusion. In addition, the patient in case 1 received cholecystectomy for cholelithiasis. Finally, liver biopsy was carried out for further pathological diagnosis with the consent of the two patients.

The follow up durations were 32 and 18 months, respectively. Neither patient suffered severe complications, and both were discharged four to six days after the operation. The symptoms of cardiac insufficiency of the patient in case 1, including exertional dyspnoea, shortage of breath, and oedema of the lower extremities, improved six months after the operation. The heart function changed to

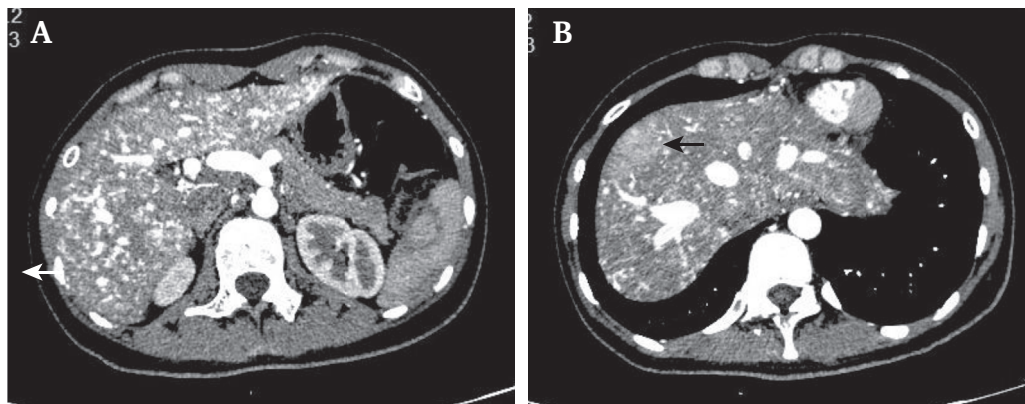


Figure 2. (A) Computed tomography (CT) showed a dilated coeliac trunk and common hepatic artery. (B) Contrast CT showed hyperplastic nodules (black arrow) and hepatic vein during the arterial phase, indicating the existence of hepatic arteriovenous fistulae.



Figure 3. (A), (B) Diffuse telangiectasia were seen on the liver surface of both patients. (C) The origin of the common hepatic artery was dissected and banded with non-resorbable suture under intra-operative laparoscopic ultrasound control.

NYHA (I – II) from NYHA (III) after reevaluation. The pulmonary artery systolic pressure of the patient decreased from 124 mmHg to 26 mmHg after the operation, although the patient had drug targeted pulmonary hypertension, including ambrisentan and tadalafil throughout. The patient with epigastric pain has remained pain free without medication three months after the operation. Although there was a transient increase in aminotransferases after the operation, the elevated alkaline phosphatase (ALP) and γ glutamyl transpeptidase (GGT) levels of patient 2 finally decreased to normal levels, which demonstrated that the biliary system was improved after the operation. The SF-36 questionnaire revealed that the quality of life of the two patients was improved after the laparoscopic surgery.

DISCUSSION

This report describes the surgical procedures of double hepatic artery band and or ligation for the treatment of HHHT. The longest follow up time for this procedure as described previously was 22 years, indicating that the procedure is not simply a transient and temporising measure.³ The present authors have been performing open operations and have accumulated HHHT patient data over a number of years. Developments in laparoscopic surgery have prompted the authors to consider how to relieve the trauma and pain associated with HHHT, and whether or not it is possible to complete double hepatic artery band and or ligation using a laparoscopic approach. This led to successful laparoscopic surgery in two patients as described here. To the present authors' knowledge, this is the first description of laparoscopic double hepatic artery band and or ligation for HHHT treatment, with improvement in symptoms of cardiac insufficiency and epigastric pain. A possible future direction would be to consider robot assisted laparoscopic surgery for HHHT treatment.

Laparoscopic double hepatic artery banding and or ligation is technically more demanding because the arteries are generally tortuous and have poor elasticity secondary to abnormal development.⁴ Firstly, banding and or ligation was performed both at the origin of the common hepatic artery and where its branches flowed into the liver. As is known, anatomical variation of vessels in HHHT patients is notable and clinicians should make an individualised surgical plan

for each patient before the operation to maximise effectiveness and safety.⁵ Consistent with previous surgical experience from open operations, the present authors routinely made use of intra-operative laparoscopic ultrasound to achieve the desired hepatic artery flow of 200–300 mL/min.^{3,6} The peri-hepatic ligaments should be intact to preserve the possible collateral circulation from the diaphragm to the liver and biliary tract. Decreased hyperdynamic circulation after banding will facilitate subsequent hepatic artery branch dissection. To prevent recurrence of hepatic AVM, gastroduodenal artery (GDA) ligation was carried out in both patients because the blood flow from the superior mesenteric artery (SMA) system through the inferior pancreaticoduodenal artery can reduce the effectiveness of the operation. In addition, other collateral vessels supplying blood to the liver, as recognised on CT imaging, should also be ligated during the operation for better results.

Generally, patients with cardiac failure symptoms and serious portal hypertension who do not respond to conservative therapy are candidates for double hepatic artery banding and or ligation operations. In the present authors' opinion, patients with simple hepatic artery dilation without serious portal hypertension or portal cavernous transformation are potential candidates for a laparoscopic operation. The authors recommend that HHHT patients with cardiac insufficiency receive RHC to evaluate pulmonary artery systolic pressure and cardiac output more objectively before surgery.⁷ Embolisation of the hepatic artery and orthotopic liver transplantation are alternative options for those who are not suitable for the procedure.⁸ However, high recurrence rates and shortage of donors are unavoidable problems.⁹

In conclusion, laparoscopic double hepatic artery banding and or ligation is a technically feasible and effective surgical approach for selected HHHT patients with simple hepatic artery dilation. The minimally invasive surgery could improve the quality of life of HHHT patients, although more patients and large scale prospective controlled studies are needed to confirm the long term therapeutic outcome.

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CONFLICT OF INTEREST

None.

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