Laparoscopic ligation of congenital extrahepatic portosystemic shunt in children with hyperammonemia: a single-institution experience

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Abstract

Purpose A congenital extrahepatic portosystemic shunt (CEPS) that is associated with hyperammonemia requires occlusion of the shunt vessels. We sought to evaluate the effectiveness and safety of laparoscopic ligation of CEPS in children with hyperammonemia.

Methods Laparoscopic ligation of CEPS was performed in 7 children with hyperammonemia. The median patient age was 5.2 years (range, 1–16 years). Before the laparoscopic procedure, a catheter inserted through the femoral vein was placed in the portal vein via the shunt vessel. The shunt vessel was dissected and taped laparoscopically. After measuring the portal vein pressure under temporal occlusion, the shunt vessels were eventually ligated.

Results The types of shunts according to location were patent ductus venosus (n = 2), splenorenal shunt (n = 2), gastrorenal shunt (n = 2), and superior mesenteric vein–inferior vena cava shunt (n = 1). Laparoscopic ligation of the shunt vessel was completed without complications in all patients. The median portal vein pressure was 19 mmHg after ligation. The median preoperative blood ammonia level was 94 μg/dL (range, 71–259 μg/dL), which decreased after ligation in all patients. No postoperative liver failure occurred.

Conclusion Laparoscopic ligation of CEPS is safe and effective in children with hyperammonemia.

Abbreviations

CPSS: congenital portosystemic shunt

CEPS: congenital extrahepatic portosystemic shunt

Introduction

Congenital portosystemic shunt (CPSS) is a rare malformation that leads to hyperammonemia, hypermanganesemia, hepatic encephalopathy, pulmonary hypertension, hepatopulmonary syndrome, and liver tumor[1]. CPSSs are divided into intrahepatic- and extrahepatic shunts[2]. Moreover, congenital extrahepatic portosystemic shunt (CEPS) is also classified into 2 types: type I, in which the liver is not perfused with portal blood, and type II, in which the liver is perfused with portal blood [2].

Portosystemic shunt with symptoms requires occlusion of the shunt vessels to improve or prevent other symptoms[1, 3, 4]. Type II CEPS can be closed surgically or through an interventional radiological procedure[1, 3, 5]. We prefer surgical closure because of the risk of migration of embolic materials. Especially at our institution, laparoscopic surgical ligation is the first choice for occlusion of type II CEPS because of its less invasiveness and reliability. The aim of this study is to evaluate the usefulness, safety, and effectiveness for laparoscopic ligation of type II CEPS.

Methods

We retrospectively assessed seven cases of laparoscopic ligation of type II CEPS performed at our institution from January 2003 to December 2016. Two cases (case 3 & 5) were reported previously in 2004 [6]. The study protocol was approved by the institutional review board (protocol no. 15191). We reviewed the patients' age at diagnosis, sex, course of laboratory data, shunt location, imaging findings, operative findings, portal vein pressure, treatments, complications related to treatment, and prognosis from the patients' medical records. Blood ammonia levels were assessed before the surgery, within one month and at 6 months to 1-year after surgery. Portal vein pressure was assessed before the surgery and at the time of surgery before and after the ligation. Surgery was indicated when patients had hyperammonemia. Before the laparoscopic procedure, a catheter inserted through the femoral vein was placed in the portal vein via the shunt vessel. The portal vein pressure was measured using this catheter. The shunt vessel was dissected

and taped laparoscopically (Figure 1). After measuring the portal vein pressure under 15 minutes of temporal shunt occlusion, the shunt vessels were eventually ligated. The criteria for indicating primary ligation at our institution were as follows: (i) portal vein pressure <25 mmHg after ligation, (ii) pressure gradient <5 mmHg at ligation, and (iii) absence of findings of intestinal wall edema and redness such as would accompany intestinal congestion.

Results

We performed laparoscopic ligation of type 2 CEPS in 7 patients during the study period. Their demographic characteristics are shown in Table 1. The patients comprised 5 boys and 2 girls. The median age at diagnosis was 1.5 years (range, 0–14 years). The median age at surgery was 5.2 years (range, 1–16 years). The triggers for diagnosis were hypergalactosemia (n = 4), hyperammonemia (n = 2), and liver function disorder (n = 1). Two patients were being treated for cardiopulmonary disease. The other patients had no relevant medical history and complications. The types of shunt according to location were patent ductus venosus shunt (n = 2), splenorenal shunt (n = 2), gastrorenal shunt (n = 2), and superior mesenteric vein–inferior vena cava shunt (n = 1). The median preoperative blood ammonia level was 94 μg/dL (range, 71–259 μg/dL; normal blood ammonia level, 5–43 μg/dL). There were no cases of hepatopulmonary syndrome or pulmonary hypertension. Liver tumor was detected in 2 patients, which was diagnosed as focal nodular hyperplasia (FNH) in both cases. The size of FNH was reduced two years after surgery in case 6, and was not reduced 6 months after surgery in case 7. There was no obvious case of hepatic encephalopathy and intracerebral disorder due to CEPS. Although 1 patient (case 6) had pervasive developmental disorder, parkinsonism, and hallucination, it was not obvious whether these conditions were related to hyperammonemia.

We performed primary ligation in all patients in accordance with the institutional criteria mentioned above. The surgical findings are shown in Table 2. The median operative time was 157 min (range, 126–182 min).

Laparoscopic ligation of the shunt vessel was completed without complications. The median portal vein pressure was 18 mmHg (range, 13–20 mmHg) before ligation and 19 mmHg (range, 13–23 mmHg) after ligation. The median pressure gradient was 2 mmHg (range, 0–3 mmHg). Blood ammonia levels decreased after ligation in all patients and normalized in 5 patients (Figure 2). The 2 patients with cardiovascular disease (cases 6 and 7) continued to take oral lactulose because of mild elevation in blood ammonia level. The median follow-up time was 6.7 years (range, 1.4–14.7 years). Recurrence of hyperammonemia and postoperative liver failure were not observed.

Discussion

CPSS is classified into various types according to the location of the shunt and presence of intrahepatic portal flow[1-4, 7]. In this study, we treated type II CEPSs in which the shunt vessels existed in the extrahepatic lesion and an intrahepatic portal flow was perfused with portal blood.

Patients with CPSS present with a wide spectrum of symptoms and complications that may develop during their lifetime. Pediatric patients are usually diagnosed as having hyperammonemia or hypergalactosemia[1, 4]. Neonates may be diagnosed on routine screening as having galactosemia without enzyme deficiency[1, 4, 8]. Hepatic encephalopathy, hepatopulmonary syndrome, and pulmonary hypertension are the most prominent manifestations caused by long-term portosystemic shunting and are more often observed in children[1, 3, 5]. Although optimal timing of treatment for children with CEPS has not been defined, some review papers suggested that all patients with extrahepatic shunts should receive early intervention regardless of the symptoms, as early treatment has been shown to prevent hepatopulmonary syndrome and other pulmonary complications, and may allow the progress of intellectual and psychosocial development [1, 3, 4]. The likelihood of encephalopathy increases with age and is related to the shunt flow [1, 5]. Children whose condition is undiagnosed or who are placed under long-term surveillance due to mild symptoms and remain treatment-free may develop clinical symptoms at a later age [1, 9, 10]. Unusual findings such as

parkinsonism and spastic paraparesis have also been described in association with hyperammonemia [1, 9]. Considering the above points, early intervention for hyperammonemia and CEPS may be important to prevent encephalopathy, hepatopulmonary syndrome, and intellectual and psychosocial disorders. Therefore, surgery is now indicated at our institution when patients have hyperammonemia that remains uncontrolled by non-surgical treatment.

A high portal vein pressure after ligation may induce postoperative liver failure[1, 8, 11, 12]. We routinely measure the intraoperative portal vein pressure before and after temporal ligation in order to decide whether the primary ligation is possible according to our institutional criteria. The criteria for indicating primary ligation are controversial and have not established yet. The criteria at our institution mentioned in the Methods section are established based on our preliminary experiences and the previous reports[3, 4, 6, 13, 14]. No postoperative liver failure occurred in this study, therefore our criteria was thought to be adequate. In type 2 CEPS, shunt occlusion is usually performed using a surgical or an interventional radiological approach[1, 3-7, 11-16]. Although interventional radiology is a less invasive method, the choice between the radiological or surgical approach depends on local expertise, shunt anatomy and size, and the patient's fitness[1, 3, 7, 16]. If the shunt is wide and short or in the case of failed embolization, the surgical approach is preferable[1, 13]. At our institution, surgical intervention, especially laparoscopic intervention, is the first choice in the treatment of patients with type 2 CEPS[6, 14]. The laparoscopic approach is as minimally invasive as the interventional radiological approach. With the laparoscopic approach, it is possible to not only measure the portal vein pressure but also check, under direct view, for intestinal congestion and the liver appearance at the time of ligation, which is impossible with the interventional radiological approach. In our study, laparoscopic ligation of type 2 CEPS was possible in various types of shunts in children aged 1 to 16 years, without complications and recurrence of hyperammonemia. The limitation of laparoscopic shunt ligation is that it is not suitable for patients with intrahepatic CPSS. Moreover, the limitations of this study are its small number of patients, single-institution design, and short follow-up duration.

Conclusion

We performed laparoscopic ligation of type II CEPS at various locations in children aged 1–16 years. Our ligation criteria are useful for preventing postoperative liver failure. Laparoscopic ligation of type II CEPS has no risk of inadvertent migration and pulmonary embolization of embolic agents, as in interventional radiology. Furthermore, it is a safe and effective method for reducing serum ammonia levels and preventing hepatopulmonary syndrome, pulmonary complication and intellectual or developmental disorder.

Conflict of interest statement: Yuichi Takama and other co-authors have no conflict of interest.

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