ORIGINAL

Successful preemptive surgical division of type 2-congenital extrahepatic portosystemic shunt in children

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Abstract : Purpose : A congenital extrahepatic portosystemic shunt (CEPS) is a rare abnormality. The shunts are classified into 2 types. Of these, a type 2-shunt is a side-to-side one, which may be treated by a simple shunt division. The aim of this retrospective study was to clarify the effects of a surgical shunt division on 4 children with type 2-CEPS. Patients : Between June 2002 and June 2008, 4 children with type 2-CEPS underwent a surgical shunt division. Various clinical factors of each patient, including shunt types, shut ratios evaluated by portal scintigraphy using ^{123}I-iodoamphetamine, serum levels of ammonia and total bile acids before and after surgery were evaluated. Findings : Two children had a conventional open surgery and the other two had a laparoscopic surgery. The serum levels of ammonia as well as total bile acids of these children decreased significantly to the normal levels within a month after the surgical shunt divisions. All the children had a better clinical course. Conclusions : A shunt division, especially by laparoscopic surgery, is an effective therapy for type 2-CEPS. To the best of our knowledge by reviewing literatures, our cases are the youngest ones treated by laparoscopic shunt division. J. Med. Invest. 56 : 49-54, February, 2009

Keywords **:** *congenital extrahepatic portosystemic shunt, laparoscopic surgery, shunt closure*

INTRODUCTION

A congenital extrahepatic portosystemic shunt (CEPS) is an uncommon abnormality. One of the earliest descriptions about this abnormality was made by Abernethy in 1793 (1). Murray, *et al.* demonstrated there were only 61 reported patients with CEPS until 2003 (2). Gitzelmann, *et al.* screened 145,000 newborns for hypergalactosemia and discovered 4 babies with type 2-CEPS (3). With the advancement of diagnostic modalities, there have been an increasing number of reported patients with CEPS recently (4). However, as of yet, the precise incidence was unknown. These shunts are classified into 2 types. A type 1-shunt is an end-to-side one with which the portal vein terminates in the IVC, whereas a type 2-shunt is a side-to-side one with which there is a venous communication between a portal and a systemic circulation (5). A patient with type 1-shunt cannot be treated by a shunt division because of the lack of the portal vein into the liver. On the other hand, a patient with type 2-shunt may be treated by a simple shunt division because a portal venous circulation into the liver exists. So far, there have been only several case reports on the results of shunt occlusion for such patients (6-14). Because of its rarity, it is difficult to assess the true effectiveness of a shunt ligation on patients with type 2-CEPS.

Here, we present 4 children with type 2-CEPS

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treated by a simple surgical shunt division in our hospital, especially focusing on laparoscopic surgery, and the effects of such treatment on these children.

PATIENTS AND METHODS

Patients

Between June 2002 and June 2008, four children with type 2-CEPS were admitted to our hospital to undergo surgery for this abnormality. All the four children were male. The mean age and the mean body weight on admission were 18.3 months (range ; 2-31) and 10.6 kg (range ; 4.6-14.4), respectively (Table 1). They were found to have hypergalactosemia by neonatal mass screening, but the activities of galactokinase, galactose-1-phosphate uridyltransferase and uridine diphosphate galactose 4'-epimerase were within normal. Further examinations including 3 dimensional-CT and angiography revealed the children had type 2-CEPS. Two children had another abnormality. One had hydronephrosis and the other had pulmonary arterial stenosis.

The shunts existed between the portal vein and the left renal vein in Child 1, between the splenic vein and the left renal vein in Child 2, between the portal vein and the inferior vena cava in Child 3, and between the superior mesenteric vein and the left renal vein in Child 4. The representative angiography of Child 2 is shown in Fig. 1A. Shunt ratios were determined by portal scintigraphy using ^{123}I-iodoamphetamine per rectum (7). The ratios were calculated as follows : counts obtained in the lung divided by total counts obtained in the liver and lung.

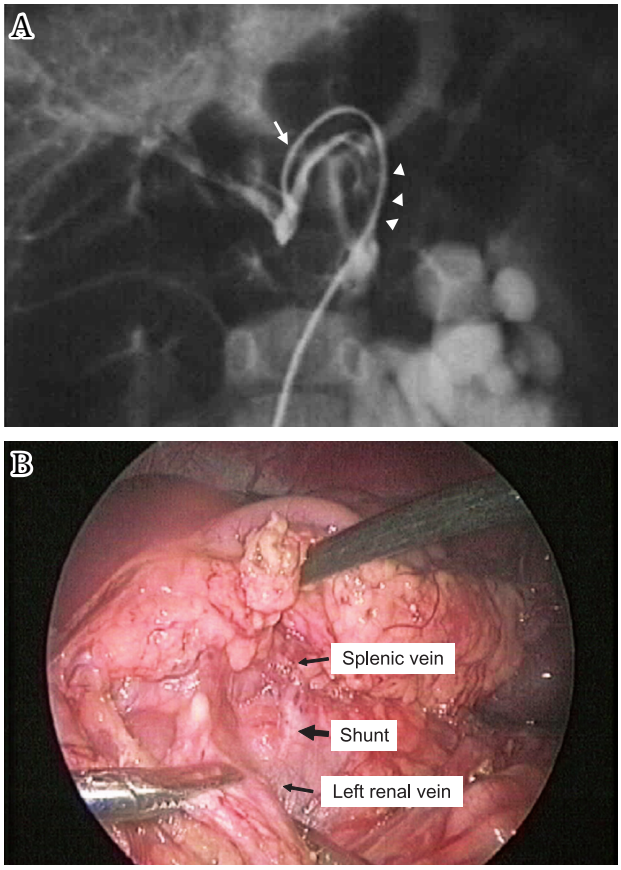

Fig. 1. Representative angiography (A) and operative image (B) of a type 2-congenital extrahepatic portosystemic shunt. (A) A shunt between the splenic vein and the left renal vein in Child 2. A catheter from the right femoral vein was introduced into the splenic vein via the shunt. White arrow indicates the splenic vein. Arrow heads indicate the portosystemic venous shunt between the splenic vein and the left renal vein. (B) An operative image of the shunt through a laparoscope. The shunt between the splenic vein and the left renal vein are visualized.

The mean shunt ratio was 71% (range ; 53.5-78.1, normal< 5) (Table 2).

Table 1. Children's characteristics on admission

Child	Age	Body weight (kg)	Hepatic encephalopathy	Developmental delay	Other abnormalities
1	2y7m	14.4	None	None	None
2	1y11m	13.9	None	None	Hydronephrosis
3	1y7m	9.5	None	None	Pulmonary artery stenosis
4	2m	4.6	None	None	None

Table 2. Preoperative data

Child	Shunt anatomy	Shunt ratio (%)	Serum ammonia (normal range ; 12-66 μg/dl)	Serum total bile acids (normal range ; 2.9-11.0 μmol/L)
1	PV-LRV	75.0	77	65.7
2	SPV-LRV	53.5	69	45.2
3	PV-IVC	77.7	93	87.0
4	SMV-LRV	78.1	92	98.1

PV, portal vein ; LRV, left renal vein ; SPV, splenic vein ; IVC, inferior vena cava ; SMV, superior mesenteric vein

All the parents of these children were given a written informed consent preoperatively. We previously reported on Child 3 as a case report (8).

Statistical analysis

Data are expressed as a mean±SD. Statistical significance was defined as $P<0.05$. Two dependent variables were compared using paired t test. Statistical analyses were performed using Stat View 5.0J software (Abacus Concepts Inc., Berkeley, CA).

RESULTS

Operation results

Child 1 and 2 underwent a laparoscopic shunt division. A representative image through a laparoscope in the operation of Child 2 is shown in Fig. 1B. Child 3 had an open laparotomy because the patent portal vein was not visualized on preoperative evaluation and we thought direct inspection of the portal system was inevitable. By direct inspection of the abdominal cavity and monitoring the portal pressure, we could safely divide the shunt. The laparoscopic surgery on Child 4 was converted to open surgery, which was due to uncertainty about the patent portal flow because the spleen was markedly swollen after shunt ligation. Child 2 had a prolonged hospital stay after operation because another treatment for hydronephrosis was needed which was performed in the urologic department. The results of each operation are summarized in Table 3. There were no major complications after surgery and the children had a good clinical course during the follow-up periods (range ; 6-78 months).

Improvement of serum parameters

Serum levels of ammonia (normal range ; 12-66 μg/dL) and total bile acids (normal range ; 2.9-11.0 μmol/L) significantly decreased to the normal levels within a month after surgery as compared to preoperative levels (82.6±11.7 vs. 30.3±10.4 μg/dL, 74.0±23.4 vs. 5.2±2.7 μmol/L, respectively) (Fig. 2A, B).

Table 3. Operation results

Child	Operation type	Operation time (min)	Blood loss (ml)	Hospital stay after operation (day)	Complication
1	Laparoscopic	330	< 5	9	None
2	Laparoscopic	310	< 5	33	None
3	Open	320	16	11	None
4	Laparoscopic to Open	457	20	14	None

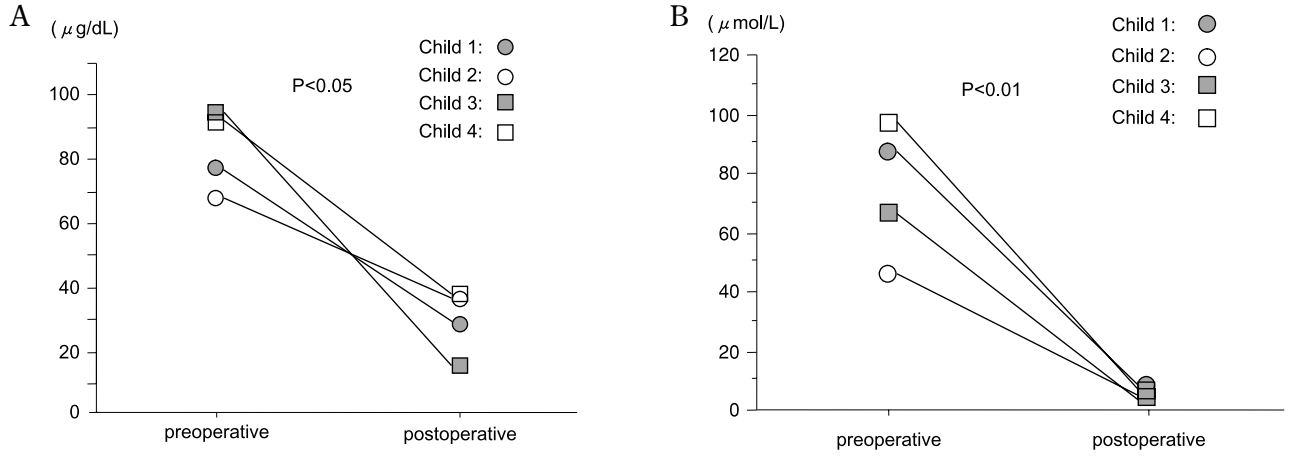

Fig. 2. Changes of serum levels of ammonia (A) and total bile acids (B).
The serum levels of ammonia and total bile acids significantly decreased to the normal levels within a month after surgery as compared to the preoperative levels.

DISCUSSION

We have shown a simple shunt division could be performed on 4 children with type 2-CEPS without any major complications and resultant normalization of serum parameters.

With the current advancement of diagnostic modalities and mass screening of newborns, especially in Japan, there are an increasing number of reported cases of CEPS (4). Because CEPSs may not be as rare as thought in the past, it should be included in the differential diagnoses of encephalopathy in children with hyperammonemia or hypegalactosemia. Finding a shunt presents a potentially curable cause for brain dysfunction.

Because patients with a type 1-CEPS lack the patent portal vein, shunt occlusion is contraindicated. In such patients, liver transplantation is the only potentially curative treatment (6, 15, 16). On the other hand, in type 2 patients, there is a possibility that a patient can enjoy a favorable life without any consequences by a simple shunt occlusion. Uchino, *et al.* identified 51 cases of congenital portosystemic shunt in Japan ; 34 of the intrahepatic type and 17 of the extrahepatic type, of which spontaneous closure was occurred only in the intrahepatic type, but not in the extrahepatic type (17). If patients with CEPS are left untreated, they will sustain various consequences such as repeated encephalopathy (7, 18), neurocognitive dysfunction (14), pulmonary hypertension (19), hepatopulmonary syndrome (9), cataract (3), liver atrophy (16), liver tumors (10, 14) and so on. Eroglu, *et al.* reported damage incurred during development in childhood could reduce the functional capacity of the adult (14). These shunts should be preemptively occluded by either surgical or radiological intervention after ruling out severe associated anomalies such as cardiac defects or maldevelopment of the intrahepatic portal as well as hepatic veins (4, 6, 7).

The surgical shunt divisions are performed by either conventional open surgery (6-10) or laparoscopic surgery (11), whereas the nonsurgical interventional shunt occlusions are performed using various devices (7, 12-14). There are the pros and cons of each intervention. Nonsurgical radiological intervention for type 2-CEPS is very attractive because of less invasiveness than other approaches. Besides, it makes staged occlusive procedures possible to permit the portal system to adjust to its new hemodynamic status. However, there are a few weak points in this approach. Dislodgement of coils into the systemic circulation is a possible risk and occluded shunts may be recanalized. Also, we cannot confirm whether there is any macroscopic appearance of serious blood congestion in the intestine. Open surgery is considered the most secure method for shunt closure in spite of its invasiveness. We can directly inspect those shunts and make sure whether there is any serious blood congestion in the intestine after temporary shunt closure. Shunt division can be achieved very easily and recanalization of the shunt cannot be possible. Most patients with type 2-CEPS, however, are so young that we should minimize the invasiveness of this treatment. Currently, the application of laparoscopic surgery to various diseases has been dramatically expanding even in pediatric surgery. Although technically demanding, this approach is far less invasive than open surgery. We can visualize those shunts and confirm gross appearance of the intestine, the liver and the spleen through a monitor. With the recent development of instruments and devices in the field of laparoscopic surgery, most type 2-CEPSs are considered amenable to this approach. We attempted this approach on three cases, two of which were completed laparoscopically and the other was converted to open surgery because of remarkable congestion of the spleen and uncertainty of the patent portal flow. To the best of our knowledge by reviewing literatures (Table 4), our cases are the youngest ones treated by laparoscopic surgery. It is needless to say that the children who underwent laparoscopic surgery recovered faster after surgery.

Table 4. Reported laparoscopic division of type 2-CEPS

Author	Year	Age (year-old)	Gender
Kimura, *et al.* (11)	2004	8	Male
		3	Female
Yamaguchi, *et al.* (20)	2007	54	Female
Seaman, *et al.* (21)	2008	48	Male
Nii, *et al.* (present report)	2009	2	Male
		1	Male

Ikeda, *et al.* reported one child with type 2-CEPS whose hepatic veins later revealed to have been poorly developed. After ligation of this child's shunt, severe liver congestion and ascites appeared, which necessitated releasing the ligation (7). Before permanent shunt closure, a test clamp should be applied to a shunt to confirm there is no steep increase

of the portal pressure. If the intrahepatic portal system is considerably hypoplastic, portal hypertension may result from abrupt changes in portal circulation. Severe portal hypertension may cause new collateral vessel formation as seen in cirrhotic patient (10). We should not sever those shunts so that occluded shunts can be released in case of fatal blood congestion in the portal circulation.

In conclusion, we strongly advocate an occlusion of type 2-congenital extrahepatic portosystemic shunts even if the patients are asymptomatic, because children with such shunt will suffer various consequences by leaving this abnormality untreated. Laparoscopic shunt division will become a standard treatment for such abnormality.

REFERENCES

1. Abernethy J : Account of two instances of the uncommon formation in the viscera of the human body. Phil Trans B83 : 59-66, 1793
2. Murray CP, Yoo SJ, Babyn PS : Congenital extrahepatic portosystemic shunts. Pediatr Radiol 33 : 614-620, 2003
3. Gitzelmann R, Forster I, Willi UV : Hypergalactosaemia in a newborn : self-limiting intrahepatic portosystemic venous shunt. Eur J Pediatr 156 : 719-722, 1997
4. Stringer MD : The clinical anatomy of congenital portosystemic venous shunts. Clin Anat 21 : 147-157, 2008
5. Morgan G, Superina R : Congenital absence of the portal vein : two cases and a proposed classification system for portasystemic vascular anomalies. J Pediatr Surg 29 : 1239-1241, 1994
6. Howard ER, Davenport M : Congenital extrahepatic portocaval shunts-the Abernethy malformation. J Pediatr Surg 32 : 494-497, 1997
7. Ikeda S, Sera Y, Ohshiro H, Uchino S, Uchino T, Endo F : Surgical indications for patients with hyperammonemia. J Pediatr Surg 34 : 1012-1015, 1999
8. Takehara Y, Mori K, Edagawa T, Sugimoto M, Takehara H, Ito M, Kuroda Y : Presumed hypoplastic intrahepatic portal system due to patent ducts venous : Importance of direct occlusion test of ductus venosus under open laparotomy. Pediatrics International 46 : 484-486, 2004
9. Morikawa N, Honna T, Kuroda T, Kitano Y, Fuchimoto Y, Kawashima N, Kawasaki K : Resolution of hepatopulmonary syndrome after ligation of a portosystemic shunt in a pediatric patient with an Abernethy malformation. J Pediatr Surg 43 : E35-E38, 2008
10. Kanamori Y, Hashizume K, Kitano Y, Sugiyama M, Motoi T, Tange T : Congenital extrahepatic portocaval shunt (Abernethy type 2), huge liver mass, and patent ductus arteriosus-a case report of its rare clinical presentation in a young girl. J Pediatr Surg 38 : E15-E20, 2003
11. Kimura T, Soh H, Hasegawa T, Sasaki T, Kuroda S, Yuri E, Tomoda K, Fukuzawa M : Laparoscopic correction of congenital portosystemic shunt in children. Surg Laparosc Endosc Percutan Tech 14 : 285-288, 2004
12. Chiu SN, Chien YH, Wu MH, Wang JK, Chen SJ : Transcatheter closure of portal-systemic shunt combining congenital double extrahepatic inferior vena cava with vascular plug. J Pediatr 153 : 723, 2008
13. Yamagami T, Yoshimatsu R, Matsumoto T, Terayama K, Nishimura A, Maeda Y, Nishimura T : Successful embolization using interlocking detachable coils for a congenital extrahepatic portosystemic venous shunt in a child. J Pediatr Surg 42 : 1949-1952, 2007
14. Eroglu Y, Donaldson J, Sorensen LG, Vogelzang RL, Melin-Aldana H, Andersen J, Whitington PF : Improved neurocognitive function after radiologic closure of congenital portosystemic shunts. J Pediatr Gastroenterol Nutr 39 : 410-417, 2004
15. Charre L, Roggen F, Lemaire J, Mathijs J, Goffette P, Danse E, Lerut J : Hematochezia and congenital extrahepatic portocaval shunt with absent portal vein : successful treatment by liver transplantation. Transplantation 78 : 1404-1405, 2004
16. Shinkai M, Ohhama Y, Nishi T, Yamamoto H, Fujita S, Take H, Adachi M, Tachibana K, Aida N, Kato K, Tanaka Y, Takemiya S : Congenital absence of the portal vein and role of liver transplantation in children. J Pediatr Surg 36 : 1026-1031, 2001
17. Uchino T, Matsuda I, Endo F : The long-term prognosis of congenital portosystemic venous shunt. J Pediatr 135 : 254-256, 1999
18. Raskin NH, Bredesen D, Ehrenfeld WK, Kerlan RK : Periodic confusion caused by congenital extrahepatic portacaval shunt. Neurology 34 : 666-669, 1984
19. Ohno T, Muneuchi J, Ihara K, Yuge T, Kanaya

Y, Yamaki S, Hara T : Pulmonary hypertension in patients with congenital portosystemic venous shunt : a previously unrecognized association. Pediatrics 121 : e892-e899, 2008
20. Yamaguchi S, Kawanaka H, Konishi K, Anegawa G, Yoshida D, Kinjo N, Tomilkawa M, Hashizume M, Maehara Y : Laparoscopic disconnection of a huge paraumbilical vein shunt for portosystemic encephalopathy. Surg Laparosc Endosc Percutan Tech 17 : 212-214, 2007
21. Seman M, Scatton O, Zalinski S, Chrissostalis A, Legman P, Soubrane O : Laparoscopic division of a portosystemic shunt to treat chronic hepatic encephalopathy. HPB (Oxford) 10 : 211-213, 2008