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Brief Report

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An unusual interventional approach to treat Type 2 Abernethy malformation in children: two case reports

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Introduction

Abernethy malformation is a rare congenital vascular abnormality characterized by portal venous blood draining into systemic veins through an abnormal shunt vein [1]. There is a consensus that symptomatic Abernethy malformation should be treated. The proposed therapeutic option for Type 1 Abernethy malformation is liver transplantation, while Type 2 can be treated with shunt closure [1–3]. Many studies have shown that the symptoms of Type 2 can be alleviated after shunt closure, with no significant difference in effectiveness between surgical ligation and interventional closure [1, 3–6]. Interventional closure has become increasingly important due to its advantages in diagnosis and treatment, and it is minimally invasive [1]. A transjugular or transfemoral approach is the conventional interventional approach to the shunt vein. However, in some cases, the above approaches may be difficult, such as in the recently reported adult case, which is considered as the first to be treated using percutaneous transhepatic interventional closure [7].

Here, we present two pediatric patients with Type 2 Abernethy malformation successfully treated by using ultrasound-guided percutaneous transhepatic interventional shunt closure to highlight the conditions needed for this approach.

Case report

Case 1

A 2-year-old girl presented with bloody stools for 6 months. The laboratory examination revealed mild anemia (hemoglobin, 91 g/L), normal blood ammonia before meals (ammonia, 42 μmol/L), and hyperammonemia after meals (ammonia, 84 μmol/L). The diagnosis of Type 2 Abernethy malformation was confirmed by using abdominal computed tomography (CT), which revealed an almost normal portal vein with a dilated inferior mesenteric vein connection between the splenic vein and the right internal iliac vein, and obvious rectal varices in the pelvis (Figure 1A). Brain magnetic resonance imaging (MRI) showed a hyperintense signal on T1-weighted imaging in the globus pallidus, and colonoscopy showed a diffuse vascular network in the mucosa of the sigmoid colon and a perianal vascular mass. Since the shunt vein was tortuous and it was difficult to place the sheath retrograde through the transfemoral approach, interventional closure via a transhepatic approach was chosen after multidisciplinary consultation. Ultrasound-guided percutaneous transhepatic interventional shunt closure was successful (Figure 1B). Hyperammonemia was not detected immediately after closure. Abdominal ultrasounds were performed daily to monitor for thrombosis and showed thrombosis on the first day after closure, gradually extending to the inferior mesenteric vein. Anticoagulation therapy using low-molecular-weight heparin calcium was initiated on the fourth day after closure and continued for 10 days. Abdominal CT, 2 weeks after closure, showed no thrombosis in the splenic, superior mesenteric, or portal vein and showed that the rectal varices in the pelvis were significantly reduced. In addition, the patient's hematochezia improved considerably, with no anemia. Colonoscopy 6 months after closure also indicated a regression of the vascular network in the rectum and anal canal.

Case 2

A 14-year-old girl presented with a headache for 2 months. The patient had been treated several times in other hospitals due to intermittent hematochezia. The laboratory examination was unremarkable, with normal blood ammonia before and after meals. Brain MRI showed a hyperintense signal on T1-weighted imaging in the globus pallidus. The diagnosis of Type 2 Abernethy malformation was confirmed by using abdominal CT, which revealed an almost normal portal vein with a dilated inferior mesenteric vein communicating with the left internal iliac vein (Figure 1C). Interventional closure via a transhepatic approach was chosen after multidisciplinary consultation and the procedures were successful (Figure 1D). After closure, abdominal ultrasounds were performed daily to monitor for thrombosis, which gradually occurred, extending to the inferior mesenteric vein on the fourth day. Anticoagulation therapy using low-molecular-weight heparin calcium was initiated on the fifth day. However, thrombosis occurred in the superior mesenteric and portal veins on the sixth day. The

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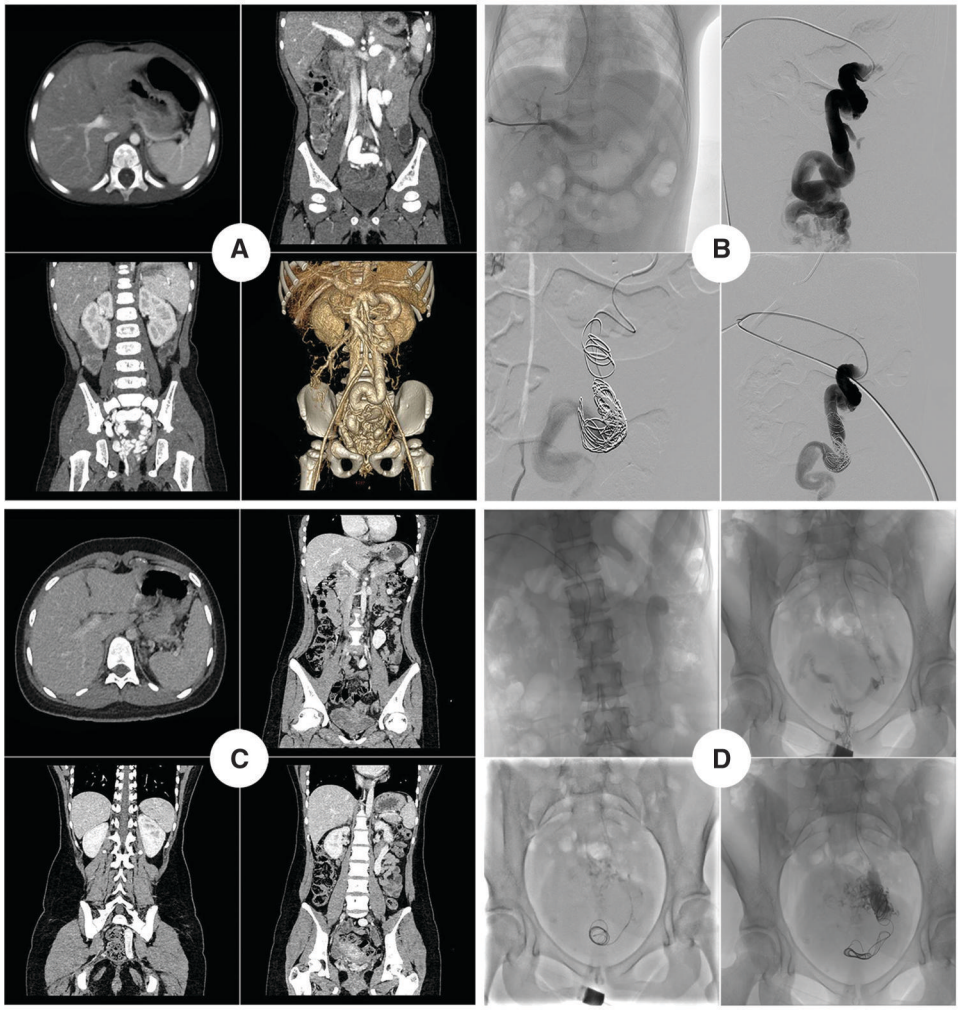

Figure 1. The images of contrast-enhanced abdominal computed tomography (CT) and operation procedures of ultrasound-guided percutaneous transhepatic interventional closure. (A) Contrast-enhanced abdominal CT with three-dimensional reconstruction in Case 1 shows an almost normal portal vein with a dilated inferior mesenteric vein connection between the splenic vein and the right internal iliac vein and obvious rectal varices in the pelvis. (B) Operation procedures in Case 1. Percutaneous transhepatic portal vein puncture was performed under ultrasound guidance, followed by insertion of a 4 French sheath. Portal pressure was measured at ~11 mmHg. After angiography confirmed shunt blood flow to the right internal iliac vein, four interlocking coils were inserted anterogradely into the inferior mesenteric vein to embolize the shunt. Repeat angiography showed that the shunt blood flow velocity was significantly slowed. Portal pressure was measured again at ~15 mmHg and recovered to 12 mmHg after 20 minutes. (C) Contrast-enhanced abdominal CT in Case 2 shows an almost normal portal vein with a dilated inferior mesenteric vein communicating with the left internal iliac vein. (D) Operation procedures in Case 2. The patient underwent anterograde angiography of the inferior mesenteric vein after percutaneous transhepatic puncture of the portal vein under ultrasound guidance, which was confirmed to be connected to the bilateral internal iliac vein; thus, the diagnosis of Type 2 Abernethy malformation was considered. Then, three interlocking coils were inserted anterogradely along the portal vein to the distal end of the inferior mesenteric vein to embolize the shunt. Repeat angiography showed that the shunt blood flow velocity was significantly slowed. The portal pressure was measured at ~24 mmHg and recovered to 21 mmHg after 20 minutes.

patient received one dose of alteplase (25 mg) in addition to heparin anticoagulation on the 10th day and was discharged on the 24th day when abdominal ultrasound showed partial dissolution of the thrombus in the superior mesenteric and portal veins. The headache of the patient disappeared after closure and the hematochezia also improved. Abdominal ultrasound 3 months after closure indicated complete dissolution of the thrombus in the superior mesenteric vein and portal vein.

Discussion

Selecting a reasonable therapeutic method for Abernethy malformation depends on the type of malformation, complications, and associated abnormalities [8]. According to the classification based on the portosystemic shunt system proposed by Kobayashi *et al.* [9], the two patients in our study were also Type C patients. Unlike the Type C patients reported in previous studies, the shunt vein between the inferior mesenteric vein and the iliac vein in our two patients was extremely tortuous and complicated, making it difficult to reach the shunt vein through the conventional interventional approach [8, 9]. In addition, the two patients had relatively normal main portal veins. Therefore, percutaneous transhepatic interventional closure was chosen. To our knowledge, our study reports the second and third cases of Abernethy malformation treated by using ultrasound-guided percutaneous transhepatic interventional closure, which is the first time this approach has been applied to Type C patients.

After closure, the shunt vein should be closely monitored to prevent thrombosis involving the splenic, superior mesenteric, and portal veins. In practice, monitoring thrombosis is difficult because the thrombus can form and grow very quickly. Zhang *et al.* [3] suggested that heparin anticoagulation could be

routinely applied after shunt closure. In Case 1, heparin anticoagulation was used when thrombosis was detected on the fourth day after shunt closure and the thrombosis eventually did not spread to the portal vein. However, in Case 2, the thrombosis spread to the portal vein with the same anticoagulation regimen. Developing a reasonable anticoagulation therapy is challenging and further large-sample studies are needed.

Although ours is a small case series of two children, based on our observations, we believe that patients with Type 2 Abernethy malformation who can be treated using percutaneous transhepatic interventional closure should have suitable vascular conditions, including an almost normal portal vein that cannot be reached easily from the systemic veins due to the presence of tortuous and complicated shunt veins.

Authors' Contributions

J.Yan, D.Z., W.P., and Y.C. conceived and designed the project. C. L. and J.Yin completed the procedures. J.Yan, C.L., and D.Z. collected the data. J.Yan, C.L., D.Z., and J.Yin analysed and interpreted the data. J.Yan, C.L., and D.Z. drafted the manuscript. All authors read and approved the final version of the manuscript.

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Conflict of Interest

None declared.

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