A case report on the treatment of diaphragmatic eventration-induced Budd-Chiari syndrome: the first innovative application of diaphragm plication

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Introduction: Budd-Chiari syndrome (BCS) is a condition in which the hepatic outflow tract is obstructed. In some rare situations, abnormal diaphragmatic eventration (DE) is the main cause of BCS; furthermore, severe cases of DE-induced BCS can lead to liver congestion, functional impairment, and cirrhosis. Only three cases of DE-induced BCS have been reported in the literature to date, and no specific treatment modalities have been described. Herein, we present the first case in which minimally invasive surgery was used to treat the DE-induced BCS.

Case report: A 72 years-old woman presented with hepatic encephalopathy as the first symptom and a diagnosis of cirrhosis. Contrast-enhanced computed tomography (CT) and abdominal ultrasound imaging revealed obstruction of the hepatic outflow tract, stenosis of the hepatic vein (HV) at the opening of the inferior vena cava (IVC) and dilatation of the lower segment of the IVC. These findings were consistent with a diagnosis of BCS. After three interventional balloon dilatations, the stenosis could not be resolved. Chest radiographs revealed a marked elevation of the right diaphragm (by approximately three vertebral levels) compared with the left diaphragm. During the thoracic surgery consultation, DE was diagnosed as a possible underlying cause of BCS, and diaphragmatic plication (DP) was proposed as a surgical treatment. The patient then underwent DP at our center and was discharged on postoperative day 4 without complications. The follow-up examinations revealed that the patient's blood ammonia level and internal diameter of the IVC had returned to normal.

Conclusion: Diaphragmatic eventration is an exceedingly rare cause of BCS, and DP offers novel therapeutic insights for such condition. Etiology-driven strategies and appropriate multidisciplinary modalities are extremely necessary for diagnose and treat BCS.

KEYWORDS

Budd-Chiari syndrome, diaphragmatic eventration, diagnosis, diaphragmatic plication, case report

Introduction

Budd-Chiari syndrome (BCS) is caused by the obstruction of the hepatic outflow tracts (1). In some cases, this condition can cause recurrent abdominal pain and bloating of the right upper abdomen, ascites, jaundice, liver bruising and enlargement. Furthermore, severe BCS can progress to cirrhosis and eventually to liver failure (2). The etiology of the disease can be broadly classified as primary, lesions in the hepatic vein (HV) or inferior vena cava (IVC) associated with structural narrowing of the veins, thrombosis due to abnormal coagulation processes, and thromboembolic obstruction due to malignancy (3). External compression of the IVC, such as compression by solid tumors and abdominal organs through hernia formation, is the secondary cause of BCS (3). Diaphragmatic hernia (DH), which is relatively rare, can also contribute to this condition (4). Some of these contents, including the colon, the omentum or even part of the liver lobe, may herniate due to congenital weaknesses in the development of the diaphragm (5). A small percentage of diaphragmatic injuries are caused by trauma, which can compress the outflow tract by hernia formation and reduce the diameter of the IVC due to diaphragmatic scar healing and fibrotic tissue contracture (4).

Diaphragmatic eventration (DE) is a congenital developmental defect in the muscular part of the diaphragm that only remains attached to the sternum, ribs and dorsal lumbar vertebrae (6). According to embryological theory, structural defects in the diaphragm are caused by abnormal or delayed migration of myofibroblasts in the upper cervical segment (7, 8). DE is commonly induced by diaphragmatic paralysis that is caused by tumor- or trauma-mediated damage to the phrenic nerve (9, 10). Dyspnea is the most prominent clinical manifestation in patients with DE, and ventilation/perfusion mismatch and loss of chest wall compliance are among the factors that contribute to dyspnea. Some patients develop mild hypoxemia and attempt to compensate by hyperventilation, which can result in mild respiratory alkalosis (6). Atypical gastrointestinal symptoms may also occur in a subset of patients (especially those with left hemidiaphragm eventration), such as epigastric pain, bloating, heartburn, regurgitation, belching, nausea, constipation, and inability to gain weight (8, 11). However, DE-induced BCS is much rarer. Only three cases have been reported in the literature to date by Saujani et al. (12), Doğan et al. (13). However, there is no description of the treatment for this disease. Here, we report the first case of DE-induced BCS that was successfully resolved by diaphragmatic plication (DP), a minimally invasive surgery. This case was reported in accordance with the CARE Guidelines (14).

Case report

The patient was a 72 years-old Chinese woman who was admitted with chronic abdominal pain lasting more than 30 years and recent reduced directionality lasting 2 weeks. A total of 15 days prior to presentation, she experienced blurred consciousness upon waking up in the morning. Cranial computed tomography (CT) imaging and magnetic resonance imaging (MRI) were performed at another hospital to exclude cerebrovascular disease and other abnormalities, and a scale-based assessment suggested disorientation. The patient's blood ammonia level reached 90.2 μmol/L, while her liver and kidney function were normal. After treatment with oral lactulose and mentholated ornithine preparations, improvements in orientation and consciousness were observed. The patient had a history of type II diabetes mellitus and was taking metformin and glimepiride regularly, achieving good glycemic control. No other significant findings were noted in terms of family history, psychosocial history, or lifestyle history. On physical examination, the vital signs were normal, the skin and sclera were not yellowed, no mass was palpable in the upper abdomen, no tenderness was found in the abdomen, and Murphy's sign was negative. After consulting our hospital, the following blood laboratory indices were measured: white blood cell (WBC) count, 3.69×10^9/L; hemoglobin (HBG), 105 g/L; alanine transaminase (ALT), 24.6 U/L; aspartate aminotransferase (AST), 43.9 U/L; and total bilirubin (TBIL), 33.7 mmol/L. Contrast-enhanced CT imaging revealed narrowing of the IVC above the HV confluence, cirrhosis, dilation of the main portal trunk to 13 mm, a tortuous splenic vein and esophagogastric fundic varices (Figure 1). Abdominal ultrasound revealed a constriction of the IVC (at the opening of the HV) approximately 0.33 cm in width, an increase in flow velocity to 134 cm per second (Figure 2C) and a dilatation of the IVC at its lower segment to 2.8 cm (Figure 2A). Combined with the patient's CT and ultrasound assessment of IVC morphology, it was evident that the hepatic outflow tract was obstructed, thus confirming the diagnosis of BCS.

However, the patient's treatment process was not very smooth, and the patient experienced twists and turns. The treatment of BCS began with an interventional approach using a balloon to dilate and shape the stenotic IVC. The patency of the IVC was restored after balloon inflation (Figure 3A), but the original stenosis returned immediately after balloon withdrawal, and there was no change in the internal diameter of the stenosis (Figure 3B). There was no benefit from three rounds of balloon inflation. Postoperative chest radiographs showed a marked elevation of the right diaphragm in contrast to the left diaphragm, which was elevated by approximately three vertebral levels, consistent with a diagnosis of DE (Figure 4A). Subsequently, ultrasound examination of diaphragmatic mobility revealed that during calm breathing, the right diaphragm amplitude was 0.20–0.47 cm and did not change significantly; the left diaphragm amplitude was 2.4 cm; which was consistent with the diagnosis of right DE (Figure 5). Tumors of thoracic and mediastinal origin and those caused by inflammation were ruled out by positron emission tomography/computed tomography (PET/CT). Based on these findings, the CT images were further reviewed. It was suggested that the elevation of the diaphragm led to liver transposition and torsion of the HV at the confluence of the IVC. This approach helped to clarify the distortion of the IVC despite several interventional balloon dilatations due to the external force exerted by liver traction. To prevent further elevation of the diaphragm from worsening the obstruction, DP was recommended by thoracic surgeons. However, the efficacy of this procedure for treating BCS is inconclusive and has not been reported in the literature. Nonetheless, DP minimally invasive, and even if it is ineffective, it does not increase the difficulty of open surgery.
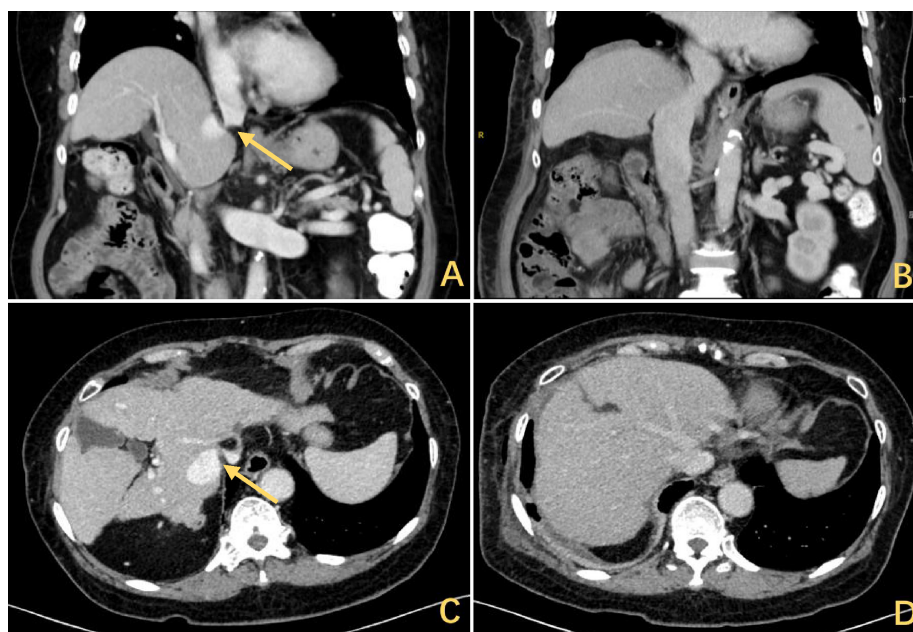
FIGURE 1
The CT scan showed a DE with hepatic elevation leading to a reduction in the angle of confluence between the HV and the IVC and obstruction of the hepatic outflow tract. The arrows highlighted in yellow indicate the stenosis, **(A)** coronal and **(C)** axial. Release of outflow tract obstruction after DP, **(B)** coronal and **(D)** axial. CT, computed tomography; DE, diaphragmatic eventration; HV, hepatic vein; IVC, inferior vena cava; DP, diaphragmatic plication.
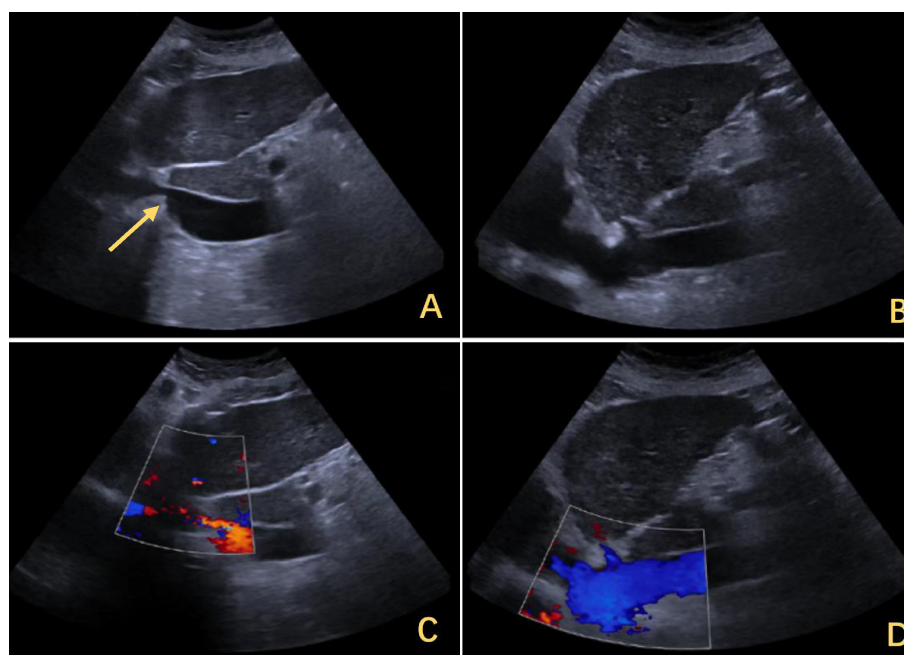

FIGURE 2
Ultrasound findings of IVC stenosis with outflow obstruction. **(A)** The IVC was approximately 0.33 cm wide, with its lower segment expanding to 2.8 cm. The arrows highlighted in yellow indicate stenosis. **(C)** The flow increased to 0.134 m per second. **(B)** After the release of the outflow tract obstruction after DP, the diameter of the IVC increased to 1.2 cm. **(D)** The flow rate returned to normal. IVC, inferior vena cava; DP, diaphragmatic plication.

A 3 cm incision was made at the right mid-axillary line, intercostal space five, and the pleural cavity was entered by dissecting through the layers. Intraoperatively, thoracoscopy was performed for exploration, revealing no pleural effusion or pleural nodules, and no pleural adhesions were observed. Electrical stimulation of the phrenic nerves resulted in a weak diaphragmatic
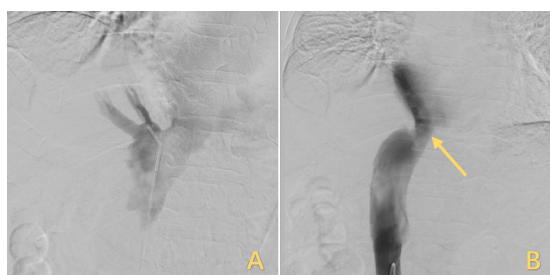
FIGURE 3
Treatment for IVC stenosis with interventional balloon dilatation.
(A) The inflated balloon was used to open the narrowed IVC.
(B) After the balloon was withdrawn, the IVC returned to its original
shape, but the stenosis remained unrelieved, and the arrows
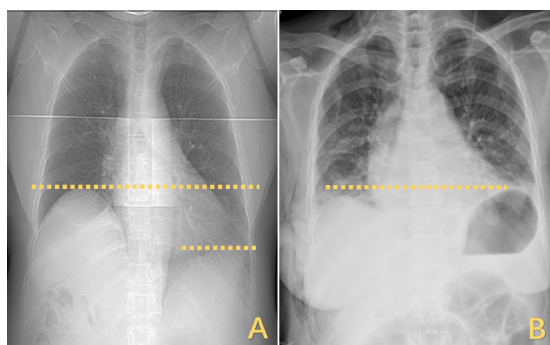highlighted in yellow indicate the stenosis. IVC, inferior vena cava.

FIGURE 4
Changes in the horizontal position of the right diaphragm before
and after DP. **(A)** The right diaphragm was significantly raised
compared to the left diaphragm, which was elevated by
approximately three vertebral levels. **(B)** Postoperatively, both
diaphragms returned to the same level. DP, diaphragmatic plication.

contraction response. Pressing down on the diaphragm was
attempted, followed by esophageal ultrasound, which revealed that
the width of the IVC increased to 14 mm. The tendon plate area of
the diaphragm was lifted with oval forceps (Figure 6A), the root was
sutured, and the free discarded diaphragm was fixed (Figure 6B).
A thoracic close drainage was preplaced, and after adequate lung
reinflation and air evacuation, the drainage was removed.

The right diaphragm dropped to almost the same level as
the left diaphragm on the third day after DP (Figure 4B). The
patency of the IVC stenosis was restored with by increasing
the internal diameter to 1.2 cm (Figure 2B). And the flow rate
returned to normal (Figure 2D). The patient's blood ammonia
level decreased to 66.4 μmol/L. She had a thoracic close drainage
removed on the third postoperative day, had no postoperative
complications, and was discharged on the fourth postoperative day.
At 1 year postoperative follow-up, ultrasonographic evaluation of
IVC revealed a maximum diameter of 1.46 cm, with a mean flow
velocity of 0.5 m per second and peak velocity of approximately
1 m per second (Figure 7). Serial blood ammonia monitoring
demonstrated persistently normal levels throughout the follow-up
period (Figure 8).

Discussion

Only three cases of DE-induced BCS have been reported in
the literature. The diagnosis of these patients is challenging and
can only be deduced by fully combining the patients' imaging
data and clinical symptoms. The case described in this article
was diagnosed only after a review of judgment, following several
failed interventions. Therefore, this disease is rare and often
misdiagnosed, leading to delayed or ineffective treatment.

The key to resolving secondary BCS is to remove the cause
of the obstruction, e.g., the removal of the solid tumor and the
tumor thrombus and the closure of the hernia ring to repair the
diaphragmatic hernia. In contrast, the etiology of BCS does not lend
itself to conventional surgical solutions. Diaphragmatic paralysis
resulting in DE causes a change in the anatomical position of
the liver, resulting in a corresponding increase of 2–3 vertebral
levels. Additionally, the angle at which the HE converges into
the IVC changes from an obtuse to an acute angle (15). This
process is accompanied by a compensatory increase in hepatic
outflow through short veins that originate from the central and
caudate lobes of the liver and connect directly to the IVC (16). The
central hepatic lobe becomes hypertrophied, and the lobe in which
outflow tracts drain through the obstructing hepatic vein becomes
atrophied (17). This results in changes in the center of gravity of the
liver that contribute to its axial displacement, which is centered on
the IVC. This displacement further reduces the angle of HV-IVC
convergence and exacerbates outflow tract obstruction (15).

In conventional surgery, it is assumed that resecting part of the
liver and the abnormal diaphragm, and simultaneously repairing
the diaphragmatic defect with a artificial patch, will restore the
anatomical position and thereby resolve the confluence obstruction
(18). However, this approach is highly invasive and involves
permanent implantation of artificial materials, which have a limited
lifespan. Importantly, no effective treatments were available for
the three previously reported cases (12, 13). In our case, the
treatment was innovative, and minimally invasive thoracoscopic
surgery can achieve the same efficacy as conventional treatments
by lowering the plane of the liver by folding the diaphragm of the
inflated segment. Based on accurate diagnostic reasoning, a suitable
minimally invasive treatment was chosen, resulting in a satisfactory
therapeutic outcome.

Congenital weakness of the diaphragm is the most common
cause of DE (6). The diaphragm is attached to the thoracic ribs
and vertebrae. The prevalence of DE in the population is less
than 0.05%, and it is more common on the left side than on
the right (19). The first report of using DP to treat DE was
published in 1923 by Morrison et al. (20). Since then, the safety
and efficacy of DP have been recognized, and thus, it has been
widely practiced. Beshay et al. (21) reviewed the postoperative
lung function of 134 patients who underwent DP and found that
the procedure was effective in reducing debilitating dyspnea and
improving lung function in patients suffering from diaphragm
eventration. Groth et al. (22) followed up with their patients after
DP by collecting questionnaire responses and reported significant
short-term and midterm improvements in respiratory quality of
life and pulmonary function test results. A retrospective study by
Kocher et al. (23) reported that performing minimally invasive
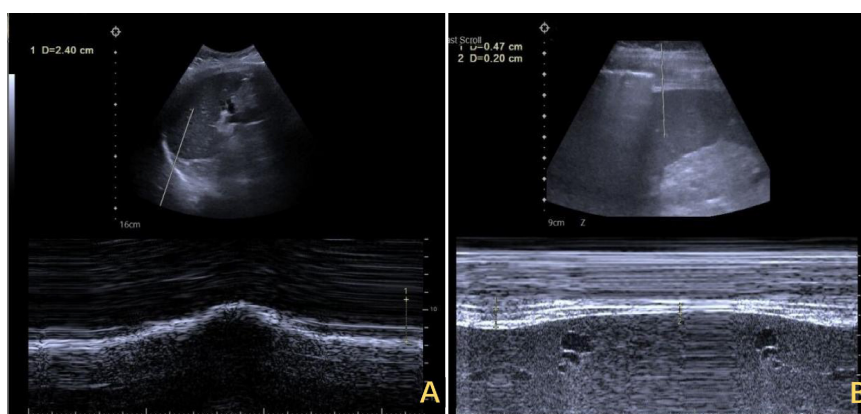DP not only significantly improves respiratory symptoms but

FIGURE 5
Ultrasound assessment of bilateral diaphragm amplitude. **(A)** The left diaphragm showed an amplitude of 2.4 cm at end of inspiration. **(B)** The right diaphragm exhibited minimal amplitude throughout the respiratory cycle, ranging from 0.20 to 0.47 cm.
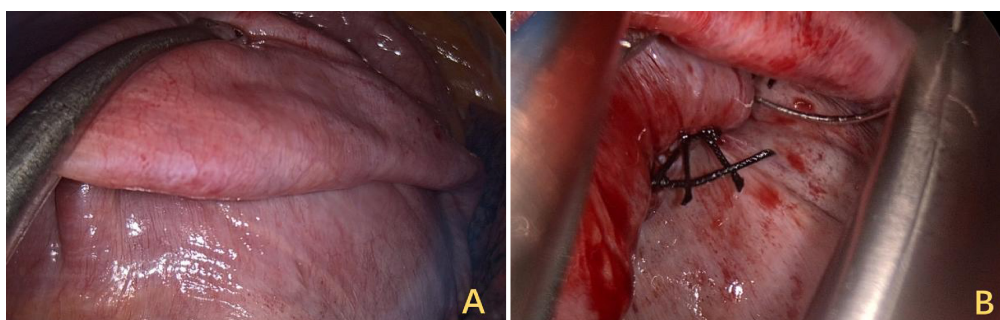

FIGURE 6
Surgical field view of trans-thoracoscopic DP. **(A)** Restoring tension by folding the diaphragm onto a flat surface. **(B)** At the base of the folded segment, sutures were used for continuous suturing and fixation. DP, diaphragmatic plication.
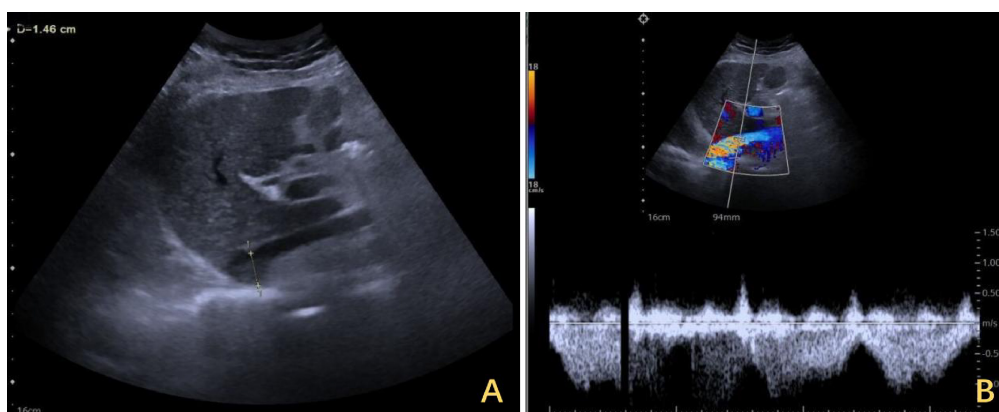

FIGURE 7
One-year follow-up ultrasound of the IVC after DP. **(A)** The maximum diameter of the IVC was 1.46 cm at its widest segment. **(B)** The IVC exhibited a mean flow velocity of 0.5 m per second, with a peak velocity of approximately 1 m per second. IVC, inferior vena cava; DP, diaphragmatic plication.

also simplifies the procedure, increases efficiency, and ensures perioperative safety. The diaphragm can be approached from the thorax or the abdomen. Either approach may be performed with open or minimally invasive techniques. The only goal of diaphragm plication is to treat dyspnea; hence, operative intervention is indicated exclusively for patients who are symptomatic (19). Studies suggest that the procedure leads to an improvement in vital capacity, forced expiratory volume, and overall lung function, particularly in patients with diaphragmatic paralysis due to phrenic nerve injury or other causes of diaphragm dysfunction (24–27).
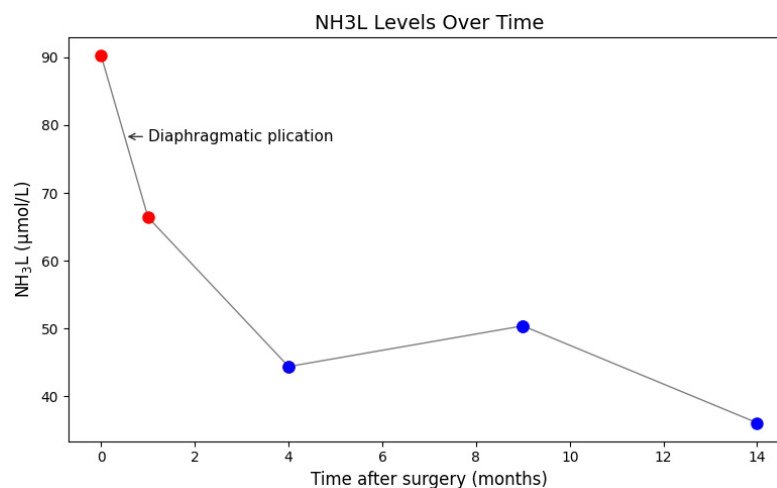
FIGURE 8
Serial blood ammonia monitoring following DP. Blood ammonia levels decreased immediately postoperatively and remained within normal range. DP, diaphragmatic plication.

While the recurrence of diaphragmatic dysfunction is uncommon, it can occur in some cases, particularly if the underlying cause is not fully addressed (e.g., ongoing phrenic nerve damage). Long-term failure rates are generally low, but it remains a potential concern for a subset of patients (28).

In this case study, DP reversed the liver dysfunction caused by BCS and even alleviated the need for liver transplantation. Surgical indications for DP have expanded, thus providing a new option for treating such patients.

Conclusion

A subset of BCS patients require CT, MRI and ultrasound to determine IVC morphology and alignment after primary etiology has been ruled out. DH and DE are extremely rare causes of BCS. DP can be an effective and minimally invasive treatment for BCS if it is clearly diagnosed. Future multicenter studies involving larger patient cohorts will be essential to confirm these findings and establish stronger evidence-based medical support.

Data availability statement

The original contributions presented in this study are included in this article/supplementary material, further inquiries can be directed to the corresponding author.

Ethics statement

The studies involving humans were approved by the Ethics Committee of Tsinghua Changgung Hospital. The studies were conducted in accordance with the local legislation and institutional requirements. The participants provided their written informed consent to participate in this study. Written informed consent

was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

JM: Conceptualization, Data curation, Investigation, Writing – original draft. BT: Data curation, Investigation, Writing – review and editing. XW: Validation, Writing – review and editing. LY: Data curation, Investigation, Writing – review and editing, Conceptualization. YL: Data curation, Investigation, Writing – review and editing, Formal Analysis. YW: Data curation, Investigation, Writing – review and editing. PW: Writing – review and editing.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Correction note

This article has been corrected with minor changes. These changes do not impact the scientific content of the article.

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