



# Decompression surgery of the celiac trunc—a single center experience

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**Background:** Abdominal pain from extrinsic compression of the celiac trunk, by a fibrous diaphragmatic arcuatum ligament is commonly referred to as median arcuate ligament syndrome (MALS). This rare condition requires a detailed preoperative workup and a customized minimally invasive treatment strategy. Celiac trunc decompression, furthermore plays a key role in patients undergoing liver transplantation.

**Methods:** We reviewed our institutional database for patients with MALS as well as liver transplant candidates which also required a celiac trunc decompression during the transplant procedure.

**Results:** Between 2016 and 2019, fourteen patients were diagnosed with MALS. Mean patient age was 51±19 years, and MALS was predominantly present in women (n=9, 64%). Two patients received laparoscopic decompression surgery; six patients qualified for a conservative follow up due to mild symptoms and another 6 patients refused a surgical intervention. Mean operative time was 50.6. No abdominal drains were placed. MALS associated symptoms resolved in all treated patients. Morbidity was 0%. Patients were discharged on postoperative day 3. In 2019, n=32 liver transplants were performed at our institution. Four patients (12.5%) required celiac trunc decompression during the transplant procedure to reestablish sufficient blood flow (233±25 mL/min) through the hepatic artery. Decompression surgery related morbidity was 0%. Overall morbidity was 18.75% (n=6) and 60-day mortality was 3.2% (n=1).

**Conclusions:** Decompression surgery for the celiac trunc is necessary in patients with MALS and asymptomatic patients undergoing liver transplantation. Both minimally invasive and open decompression strategies are feasible and safe. General diagnostic and therapeutic guidelines are in need.

**Keywords:** Laparoscopic celiac trunc decompression surgery; median arcuate ligament syndrome (MALS); laparoscopic surgery; liver transplantation

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## Introduction

Median arcuate ligament syndrome (MALS), first described in 1963 by Harjola *et al.* (1), results from extrinsic compression of the celiac artery by the median arcuate ligament. Clinical symptoms comprise nausea, vomiting chronic abdominal pain, postprandial pain and unintentional weight loss. The location of the pain is variable, but most commonly spotted in the epigastrium. MALS is a rare condition, which predominantly prevails in mid aged women who have a lean body habitus (2).

Hypotheses for the etiology of MALS are diverse (3) and range from visceral ischemia (4), neuropathic compression (5) to vascular steal phenomena, caused by collateral vessels (6). Incidental findings also demonstrated that celiac trunc compression or stenosis may furthermore be present in a relatively high number (7,3%) of asymptomatic patients (7).

In case of celiac trunc stenosis or even occlusion, an increased blood flow through the pancreaticoduodenal arcade from the superior mesenteric artery (SMA) may be sufficient to compensate an otherwise insufficient arterial blood flow to the liver and stomach. However, subsequent arterial hyper-perfusion of the pancreaticoduodenal arcade, may foster the development of visceral artery aneurysms within this circulatory system (8). Decompression of the celiac trunc and reestablishment of sufficient arterial blood flow through the celiac route is hence mandatory before any endovascular intervention (e.g., embolization of the aneurysm) on the pancreaticoduodenal route may be considered, since occlusion of the entire arcade may result in acute ischemia induced liver injury. It is furthermore critical to recognize celiac trunc compression in patients requiring a liver transplant, since the arterial reconstruction during transplant comprises dissection of the hepatic artery and frequent division of the collateral circulation including the gastroduodenal artery (9).

The current literature predominantly provides case reports or small case series which describe both open and laparoscopic techniques for celiac trunc decompression (3). Currently, universally valid diagnostic criteria do not exist, however doppler ultrasound, CT angiograph and angiographic evaluation of possible changes in celiac velocities which are altered by inspiration and expiration, are commonly applied.

It is hence our aim to raise awareness of this rare disease, provide an insight in to our patient collectives, namely those with MALS as well as a group of liver transplant recipients

who required celiac trunc decompression at the time of transplant.

## Methods

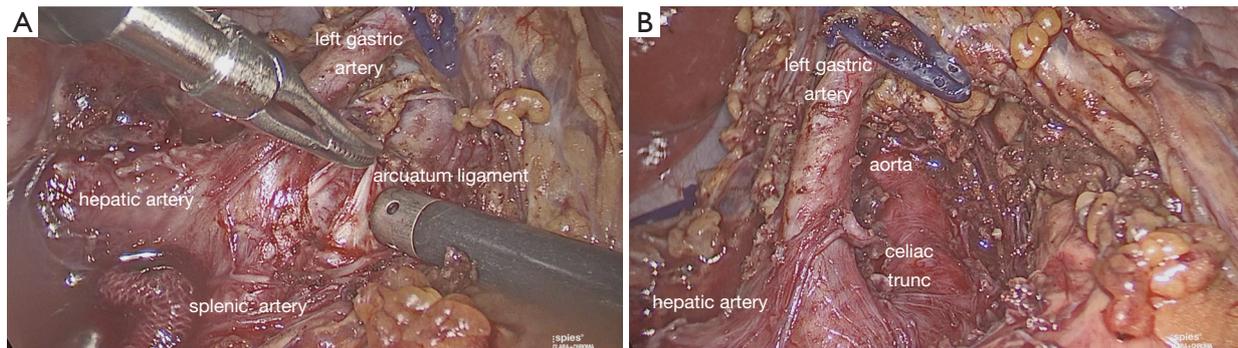
Medical datasets of patients diagnosed with celiac artery compression syndrome were evaluated retrospectively for the years 2016 to 2019. For this survey, the diagnosis code I77.4 was used to recover patient data stored in our institutional data base. Additionally, patients undergoing liver transplantation in 2019 were reviewed for coeliac trunc decompression procedures during transplantation. Anonymized patient details, clinical histories, operative details and outcomes were retrospectively reviewed. SPSS and Excel was used for statistical analysis. This retrospective study was approved by the institutional review board.

### *Minimally invasive operative technique for median arcuate ligament decompression surgery*

The detailed laparoscopic surgical procedure for median arcuate ligament decompression, is provided in *Video 1*. As an example, we demonstrate the case of a 31-year-old female patient (*Figure 1*), who reported of nausea, vomiting and postprandial pain for the last 6 months. Doppler ultrasound examination and CT angiography revealed a compression of the coeliac trunc, caused by the median arcuate ligament (*Figure 2*). The patient was scheduled for minimally invasive surgery, following informed consent.

In short, surgery was performed under general anesthesia and in French position. Following a team timeout, and under sterile conditions a 12 mm camera trocar was inserted into the abdomen through the umbilicus. A capno-peritoneum of 12 mm Hg was maintained constantly through the entire operation. After explorative laparoscopy using a 30° optic which revealed no contraindications for the planned procedure, a 10 mm and a 5 mm trocar were placed into the right and left upper abdomen, respectively. Another 5 mm trocar for suction and retraction could additionally be placed into the left subcostal region. For liver retraction a Nathanson liver retractor was inserted through a sub xiphoidal incision.

In a first step, the minor omentum was transected and the left gastric artery was dissected free from the accompanying vein. The coeliac trunc trifurcation, comprising the left gastric artery, the hepatic artery and the splenic artery should be identified and dissected up to the perivascular



**Figure 1** Intraoperative view during laparoscopic celiac trunk decompression surgery. The celiac trunk trifurcation comprises the left gastric artery, splenic artery and hepatic artery. Compression results from a low riding median arcuate ligament MALS (A). The entire ligament must be dissected up to the anterior aorta (B) to provide sufficient decompression and prevent perhaps recurrence. MALS, median arcuate ligament syndrome.

sheet (Figure 1).

In a next step, first ramifications of the arcuatum ligament located on the celiac trunk were transected using laparoscopic scissors and bipolar forceps. Another valuable instrument widely applicable for this type of operation are ultrasonic sheers, which might facilitate delicate and fast preparation along the coeliac trunk, up to the front of the abdominal aorta. It is of importance, that all diaphragmatic muscle fibers anterior to the aorta are dissected free at a length of 4–5 cm to prevent perhaps a recurrence of celiac trunk compression. Finally, all trocars were removed and the incisions were closed using PDS 3-0 interrupted sutures. Skin closure was performed by intracutaneous running sutures. Postoperative doppler ultrasound examinations were performed on postoperative day three, before discharge of the patient.

#### ***Median arcuate ligament decompression during liver transplantation***

In liver transplant candidates with celiac trunk compression, adequate arterial blood flow has to be established prior hepatectomy and subsequent liver transplantation. Screening for celiac trunk compression is performed by preoperative CT angiography. Arterial blood flow was intraoperatively measured by doppler ultrasound and the Medistim MiraQ Flow measurement system. In case of celiac decompression, a subsequent arterial blood flow of more than 200 mL/second, including the expiratory phase, was rated sufficient for transplant and reperfusion.

## **Results**

### ***Minimally invasive decompression of the median arcuate ligament***

Twelve patients were diagnosed with (I77.4) coeliac trunk compression syndrome between 01/2016 and 12/2019. Patient demographics are listed in *Table 1*. Mean patient age was  $51 \pm 19$  years and MALS had a higher incidence in women (women:  $n=9$ , 64%; men  $n=5$ , 36%). To date  $n=2$  (16,7%) were treated by minimally invasive decompression surgery. One patient presented with a classical MALS (Figure 2), and one patient additionally presented with an aneurysm of the pancreaticoduodenal arcade which was coiled three days after coeliac trunk decompression surgery (Figure 3). Mean operative time was  $50 \pm 6$  minutes. No abdominal drainages were used.

The remaining  $n=10$  (83.3%) patients either presented with mild clinical symptoms ( $n=6$ , 60%) and therefore did not qualify for surgery, or refused surgery and preferred a clinical follow up strategy instead ( $n=4$ , 40%). With regard to clinical surveillance group, two patients refused further monitoring and were hence lost in follow-up.

Our diagnosis treatment algorithm for patients with MALS comprised the following: After (I) clinical examination and survey of the medical history, (II) duplex sonography, (III) CT angiography and (IV) upper GI endoscopy is performed. In case of severe clinical symptoms due to stenosis of the coeliac trunk a minimally invasive release of the median arcuate ligament is recommended and the patient is scheduled for surgery after informed consent.

**Table 1** Table of patients diagnosed with MALS

Nr.	Gender	Age	Diagnostic procedures	Treatment
1	F	46	Doppler US; CT angiography, upper GI endoscopy, angiography	No treatment, mild symptoms
2	M	80	Doppler US; CT angiography, upper GI endoscopy,	No treatment, higher age, mild symptoms
3	M	57	Doppler US; CT angiography, upper GI endoscopy, angiography	Indication for laparoscopic decompression, patient in follow up
4	F	47	Doppler US; CT angiography, upper GI endoscopy, angiography	Indication for laparoscopic decompression, patient lost in follow up
5	F	70	Doppler US; CT angiography, upper GI endoscopy	Indication for laparoscopic decompression, patient in follow up
6	F	53	Doppler US; CT angiography, upper GI endoscopy	Indication for laparoscopic decompression, patient lost in follow up
7	F	40	Doppler US; CT angiography, upper GI endoscopy	Mild stenosis, no treatment, mild symptoms
8	M	61	Doppler US; CT angiography, upper GI endoscopy	Indication for laparoscopic decompression, patient lost in follow up
9	F	79	Doppler US; CT angiography, upper GI endoscopy	No treatment, higher age, mild symptoms
10	F	20	Doppler US; CT angiography, upper GI endoscopy	Indication for laparoscopic decompression, patient in follow up
11	M	30	doppler US; CT angiography, upper GI endoscopy	Indication for laparoscopic decompression, patient in follow up
12	F	31	doppler US; CT angiography, upper GI endoscopy	Laparoscopic decompression
13	F	44	doppler US; CT angiography, upper GI endoscopy aneurysm of pancreaticoduodenal arcade	Laparoscopic decompression, angiographic coiling of aneurysm
14	M	20	doppler US; CT angiography, upper GI endoscopy	Mild stenosis, no treatment, mild symptoms

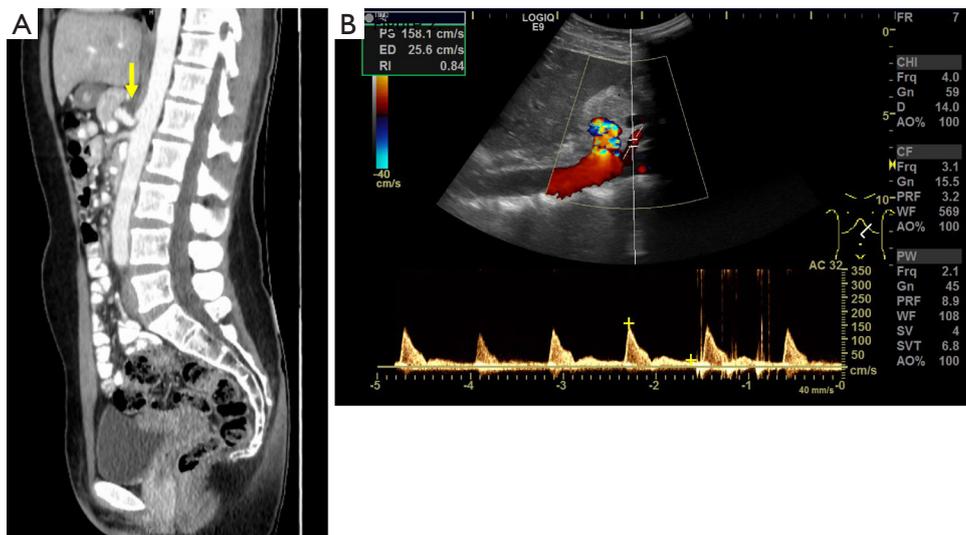
To date 2 patients (nr 13 and nr 14) received successful laparoscopic decompression surgery. Six patients presented with mild symptoms and hence did not qualify for surgery. Another six patients refused a surgical procedure and preferred a short interval surveillance strategy. Two patients did not meet their follow up appointments and were hence lost in follow-up. MALS, median arcuate ligament syndrome.

Postoperative duplex sonography is generally performed on postoperative day three, which is the regular day of discharge. If clinical symptoms are resolved, which was the case in our first two patients, treatment is completed. In case of persistent symptoms, angiography and pressure gradient examination is performed and the patient is evaluated for further angioplasty, stent placement or surgical vascular reconstruction.

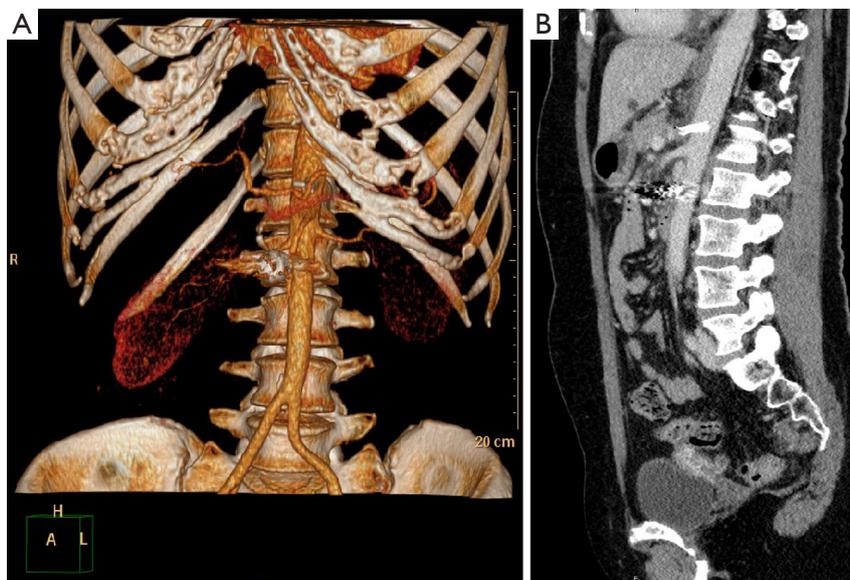
#### ***Decompression of the median arcuate ligament during liver transplantation***

In 2019 a total number of n=32 orthotopic liver transplants were performed at our institution. Demographic patient data and indications for liver transplantation are listed in *Table 2* and *Figure 4*, respectively. The average waiting

time for liver transplant recipients was  $6.4 \pm 6.4$  month and the average Model for End Stage Liver Disease (MELD) at time of enrolment and transplant was  $18 \pm 8$  and  $20 \pm 9$  respectively. Leading indications for transplant were alcohol induced liver cirrhosis (ASH, 44%) as well as cirrhosis from non-alcoholic steatohepatitis (NASH, 16%) and primary sclerosing cholangitis (PSC, 13%). Based on preoperative CT screening, four patients (12.5%) were identified with a ligamentous compression of the celiac trunc with otherwise normal arterial anatomy (Michels Type 1). In these cases, decompression of the celiac artery was performed as a first step of the transplant procedure. It is of importance to facilitate adequate arterial blood flow prior hepatectomy, since this potential time-consuming maneuver should not be performed during the an-hepatic phase of liver transplantation. All celiac



**Figure 2** Patient with MALS. CT angiography (A) displays a stenosis of the celiac trunk as a result of a low riding median arcuate ligament. A post-stenotic dilation of the celiac artery is visible. Doppler ultrasound (B) commonly displays reduced arterial flow which is even more pronounced in steep expiration. Since the expiratory phase lasts two thirds of the respiratory cycle, a significant reduction in overall celiac arterial blood flow may exist in patients with MALS. MALS, median arcuate ligament syndrome.



**Figure 3** Patient with MALS (#) and aneurysm (X) of the pancreaticoduodenal arcade. 3D reconstruction (A) and sagittal CT scans (B) show successful aneurysm coiling (X) and celiac artery stenting (#) which was performed two days after laparoscopic median arcuate ligament release.

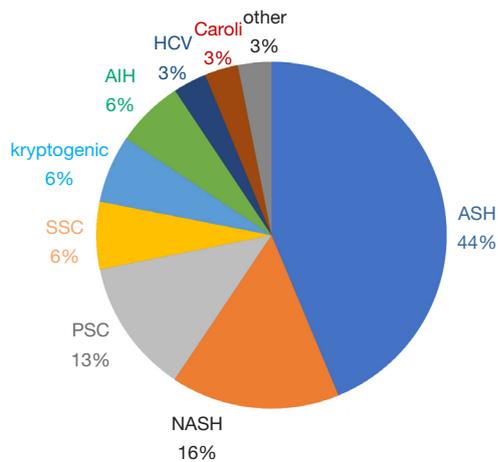
trunc decompressions were successful. Graft reperfusion was performed as “artery first” and subsequent portal vein reperfusion. Arterial flow was controlled by intraoperative doppler ultrasound and MiraQ Flow measurement

( $233 \pm 25$  mL/min). Overall morbidity, defined as Clavian Dindo grade 3b or higher, was 18,75% (n=6). Sixty-day mortality was 3.2% (n=1). Morbidity attributed to celiac trunc decompression was 0%.

**Table 2** Demographic table of patients who received a liver transplant in 2019 at our institution

Number of LTX	n=32
Number of celiac trunc decompression	n=4 (12.5%)
Age	60±7
Gender	f=22 (31.3%); m=10 (68.7%)
Waiting time (month)	6.4±6.4
MELD at listing	18±8
MELD at transplant	20±9

MELD, Model For End-Stage Liver Disease.



**Figure 4** Indications for liver transplants performed at our institution in 2019. Cirrhosis from ASH, NASH, PSC, secondary sclerosing cholangitis (SSC), cryptogenic and AIH account for the majority of all transplants. Cirrhosis from chronic hepatitis C virus infection (HCV) and Caroli disease only account for a minor number of liver transplants performed at our center. ASH, alcoholic steatohepatitis; NASH, non-alcoholic steatohepatitis; PSC, primary sclerosing cholangitis; AIH, autoimmune hepatitis.

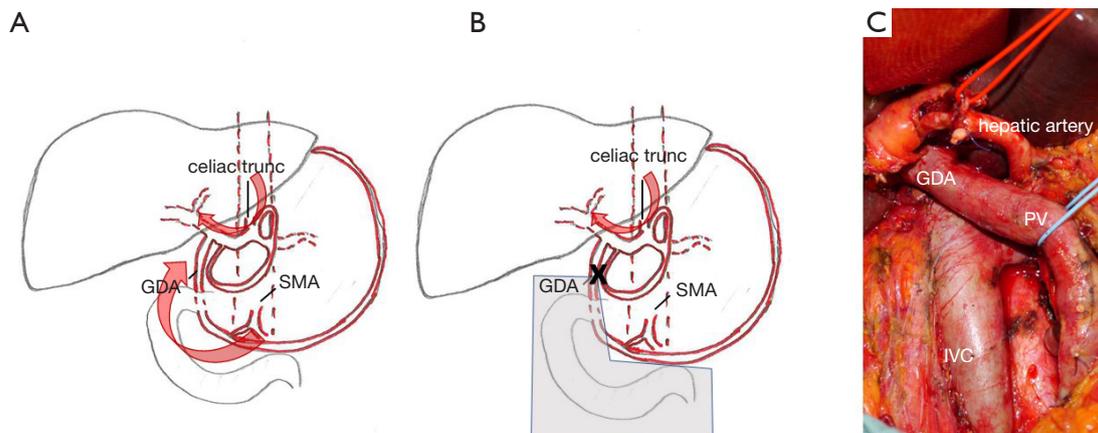
## Discussion

Minimally invasive decompression of the celiac trunc has been increasingly accepted as the gold standard treatment for MALS. First reported in 1995 by Loffeld *et al.* (10), the laparoscopic approach bears several patient benefits, including smaller incisions and decreased postoperative morbidity, and in parallel provides the surgeon with an improved surgical view of the operation field (11). More recently, the application of robotic assisted techniques (12)

have been introduced to the minimally invasive surgical world (13), which potentially are capable to even outperform the laparoscopic techniques in terms of technical (enhanced 3-D vision and intraabdominal maneuverability) and operator based (tremor elimination, operators comfort) criteria (14). In the shade of these modern surgical techniques, endoscopic retroperitoneal and open surgical approaches are gradually tapered from the daily surgical business (15). This is especially true for open surgical techniques which are mainly reserved for cases which require more than just ligamental decompression.

The diagnosis of MAL syndrome, because of its resemblance to other abdominal disorders, commonly is a diagnosis of exclusion. Currently no standardized diagnostic criteria exist, however Kim *et al.* recently provided a well-arranged algorithm for diagnosis and management of MALS, which hence has also been applied to our patients (14). This comprises a clinical patients history workup, followed by ultrasound CT scan and upper GI endoscopy. In case of normal findings diagnostic strategies are accompanied by a more detailed CT or MRT angiography, as well as a CT angiography of the celiac trunc with detailed pressure gradient measurements. In case of celiac stenosis, a minimally invasive decompression is aspired. If symptoms are resolved after surgery patients are followed up by doppler ultrasonography. In case of symptoms` persistence an alternative approach comprising angioplasty and stenting is taken into account. The high reticence in regard to decompression surgery, furthermore highlights the fact that commonly patients are torn between a high degree of physical and psychological affliction and fear of the surgical intervention.

The liver is unique in that it is supplied by dual blood inflow. While the portal vein provides the major part of hepatocyte`s blood consumption, the bile ducts solely depend on arterial blood flow from the hepatic artery (9). Flawless revascularization during transplant is key to successful postoperative graft function and necessary to prevent ischemic type bile-duct lesions (ITBL). Arterial reconstruction depends on the anatomical conditions and commonly involves the anastomosis of the donor hepatic artery to the recipient`s hepatic artery just at the origin of the gastroduodenal artery. “Artery first” reperfusion, performed as a standard in our institution, has been demonstrated to be beneficial (16,17) since biliary epithelial cells are more susceptible to warm ischemia than hepatocytes (18). It is hence crucial that proper surgical measures are taken, which in some circumstances include



**Figure 5** Schematic drawing of the celiac trunk and SMA. In case of celiac trunk compression (A) arterial perfusion from the SMA, pancreaticoduodenal arcade, and GDA may be sufficient for adequate arterial liver perfusion; in case of pancreaticoduodenectomy (B,C) ligation and transection of the GDA may only be performed once sufficient perfusion via the celiac trunk and subsequent hepatic artery is guaranteed. In case of ligamentous celiac trunk stenosis, decompression surgery may be performed before ligation of the GDA. SMA, superior mesenteric artery; GDA, gastroduodenal artery.

the decompression of the celiac trunk, to facilitate adequate arterial blood supply at time of reperfusion.

With regard to celiac trunk compression or occlusion, one more group of individuals require special attention. Patients scheduled for operations which comprise the division of the (GDA) gastroduodenal artery (e.g., pancreatoduodenectomies, visceral aneurysm repair etc.), must be screened carefully, since an intraoperative division of the GDA may stop an otherwise vital retrograde arterial blood flow to the liver (Figure 5) and ultimately result in ischemic liver failure.

In the era of minimally invasive surgery different surgical disciplines benefit from each other (19). Our laparoscopic operations were performed by a team of both hepatobiliary surgeons and vascular surgeons. We strongly believe that fundamental laparoscopic skills and a detailed knowledge of the vascular anatomy and a faithful preoperative diagnostic workup (20) is necessary for successful treatment of celiac compression syndromes.

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## Footnote

*Conflicts of Interest:* All authors have completed the ICMJE uniform disclosure form (available at <http://dx.doi.org/10.21037/ls.2020.03.08>).

RS serves as an unpaid editorial board member of *Laparoscopic Surgery* from Oct 2019 to Sep 2021. The other authors have no conflicts of interest to declare.

*Ethical Statement:* The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). This retrospective study was approved by the Institutional Review Board of University Clinic Leipzig and individual consent for this retrospective analysis was waived.

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