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Case Report

Trans-Esophageal Echocardiography of the Descending Aorta and Celiac Trunk as an Intraoperative Monitoring for Median Arcuate Ligament Syndrome (MALS) Treatment: Technique Proposal and Two Case Reports

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Abstract: Dunbar syndrome is caused by the compression of the celiac artery by the median arcuate ligament (MAL) and the diaphragmatic crura, presenting with abdominal pain. The treatment is surgical, with the aim of dissecting and separating the diaphragmatic crura from the celiac axis, restoring blood flow in the celiac trunk. We propose the use of transesophageal echocardiography to measure the peak systolic velocity in the celiac trunk and to evaluate, in real time, the efficacy of the surgery during MAL resection. Two clinical cases with practical implications are reported. In one of these, the ultrasound exam was useful in providing an intraoperative confirmation of the outcome of the resection.

Keywords: Dunbar syndrome; median arcuate ligament syndrome (MALS); echocardiography; abdominal pain

Citation: Lubian, M.; Nisi, F.; Giustiniano, E.; Ripani, U.; Piccioni, F.; Cecconi, M. Trans-Esophageal Echocardiography of the Descending Aorta and Celiac Trunk as an Intraoperative Monitoring for Median Arcuate Ligament Syndrome (MALS) Treatment: Technique Proposal and Two Case Reports. *Surgeries* **2023**, *4*, 17–25. https://doi.org/10.3390/surgeries4010003

Academic Editor: Cornelis F.M. Sier

Received: 21 October 2022
Revised: 1 January 2023
Accepted: 4 January 2023
Published: 8 January 2023

1. Introduction

Dunbar syndrome, also known as median arcuate ligament syndrome (MALS), is caused by the compression of the celiac artery by the median arcuate ligament (MAL) and the diaphragmatic crura. The MAL is a band of fibrous tissue anteriorly connecting the diaphragmatic crura surrounding the aortic hiatus; the position of the MAL may vary, leading to extrinsic compression of the celiac artery, while an abnormal origin of the celiac artery is less common [1,2]. MALS is characterized by symptoms of foregut ischemia, such as abdominal postprandial or exercise-induced pain, often located in the epigastrium, and unexplained weight loss; other signs and symptoms are nausea and abdominal bruits. The pathophysiologic mechanism of the syndrome is unclear [3]. The most probable hypothesis is that during digestion, an increased demand for intestinal blood flow cannot be supplied by a compressed artery, causing transient intestinal ischemia. Moreover, the chronic compressing of the celiac ganglion MAL may lead to the irritation of sympathetic pain fibers, causing the development of neuropathic pain [1,2]. However, most of the patients with compression of the artery have no symptoms, probably due to the development of collateral circulation which compensates for the insufficient blood flow in the celiac trunk [2,4].

The grade of compression varies with respiration, with a reduction in compression during inspiration, as the MAL moves caudally, and a greater constriction during expiration. Moreover, the compression of the artery may lead to hyperplastic intimal changes and the

development of poststenotic dilation and aneurysms, mostly in the pancreaticoduodenal arcade [4].

During physical examination, epigastric tenderness or bruits may be revealed, especially during expiration; however, neither of these symptoms are specific for the diagnosis of MALS [1]. Diagnosis is confirmed by Doppler ultrasound (US), multi-slice computed tomography angiography (MSCTA), magnetic resonance (MR) angiography, or invasive selective angiography.

The treatment is surgical, both open and laparoscopic, with the aim of dissecting and separating the diaphragmatic crura from the celiac axis, thus restoring safe blood flow in the celiac trunk [2,4–6]; endovascular intervention is not suitable because it does not solve the extrinsic compression, and it has thus been proven ineffective [1]. Successful treatment has been confirmed by Doppler ultrasound follow-up, showing the presence of a normal flow in the celiac artery after laparoscopy [7].

With these two case reports, we describe a new approach using intraoperative transesophageal Doppler to guide the surgeon during the procedure and confirm successful decompression of the celiac artery after the surgery.

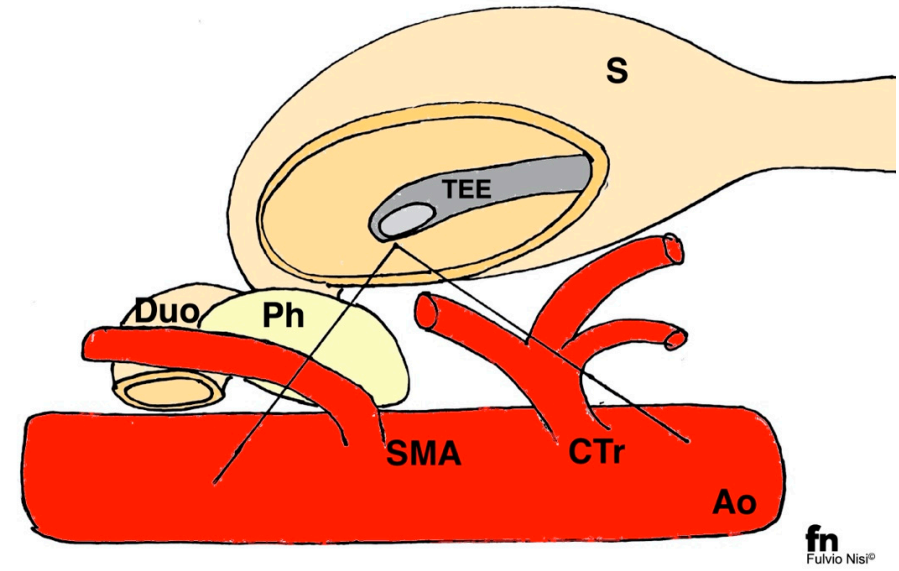
2. Materials and Methods

The evaluation of the descending aorta is included in the imaging plans for TEE recommended by the American Society of Echocardiography. However, the celiac artery (CTr), superior mesenteric artery (SMA), or other visceral branches [8] are not evaluated by any of these imaging planes. Nevertheless, the use of TEE to assess the proximal third of the abdominal aorta and its branches is reported in some preliminary studies [9–11]. According to a previous report from Orihashi et al., CTr and SMA visualization with TEE can be easily learned and achieved, as well as the visualization of both renal arteries, even if only in a smaller percentage of cases [9].

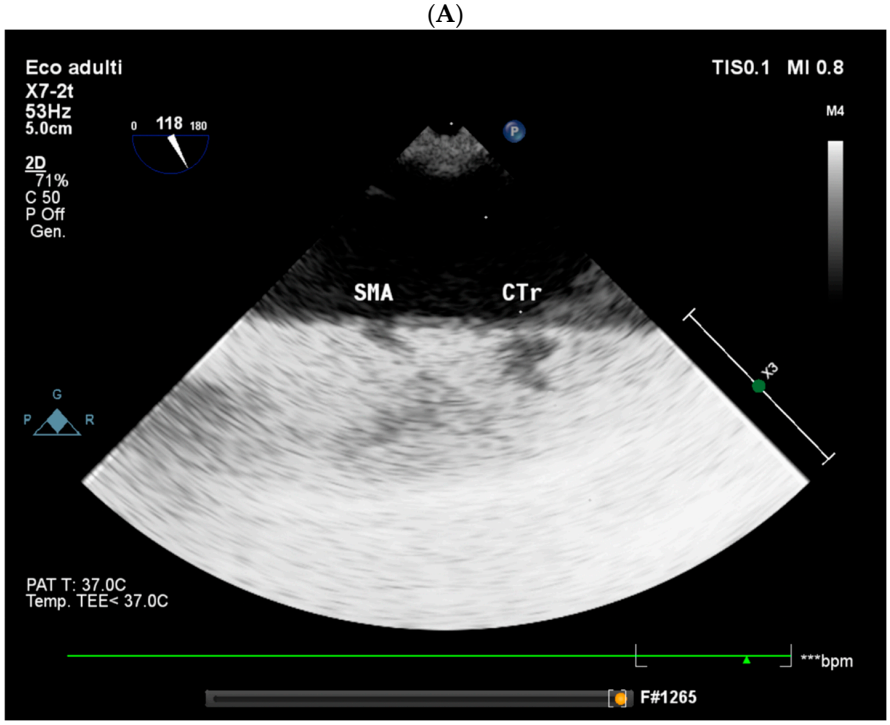
2.1. Technique Description

The device used in this study was a Philips Affiniti 70c ultrasound machine with an X7-2t transducer. After advancing the probe for routine transgastric views to approximately 40–45 cm depth, it was rotated 180° and flexed posteriorly, with the aim of locating the descending thoracic aorta at the horizontal image plane (upward flexion of the probe, short-axis view) and maintaining it on the screen. The probe was advanced farther until CTr and SMA serially appeared at the 1 to 3 o'clock position of the aorta. The former soon divided into branches and moved away from the aorta, whereas the latter stayed adjacent to the aorta. In the longitudinal scan, the long-axis view of the aorta, along with the origin of both the CTr and the SMA, was depicted (Figure 1). Its distal bifurcation is the distinguishing hallmark of the CTr.

In order to achieve a clear view of the vessel and blood flow signal, the color Doppler signal velocity scale is lowered. The peak systolic velocity (PSV) and end-diastolic velocity flow evaluation of both vessels are assessed. The CTr and SMA show different blood flow patterns. The CTr artery supplies the low-resistance vascular beds of the liver and spleen; for this reason, its waveforms show a low-resistance pattern, with high end-diastolic velocity. On the contrary, the SMA supplies the vascular beds of the small intestine and colon, which have high resistance; hence, its waveforms have a high-impedance flow pattern with low end-diastolic velocity. An accurate depiction of the technique, exploited in aortic dissections, can be found in the work of Moral et al. [12].



(**A**)



(**B**)

Figure 1. (**A**): Scanning planes and images at the level of the celiac trunk (CTr) and superior mesenteric artery (SMA) in the longitudinal scan of the aorta (Ao). The stomach (S), duodenum (Duo), and pancreatic head (Ph) are displayed. (**B**): The TEE probe (TEE) can be seen through a virtual section of the stomach wall. The orientation of ultrasound beam is shown by means of two continuous lines (longitudinal to the aortic major axis). The illustration was created by one of the authors (F.N.).

2.2. Case Report 1

An 18-year-old woman was referred to the vascular surgery department of our hospital. She has been suffering from post-prandial abdominal pain, vomiting, dyspepsia, and weight loss for one year, and needed total parenteral nutrition. She had no previous abnormal medical history or other medical condition, and her laboratory blood tests were normal. During a previous hospitalization, she was tested and empirically treated for inflammatory bowel disease and Helicobacter pylori gastritis, but both conditions were ruled out since the treatment was not beneficial.

During a subsequent hospitalization in a different hospital, abdominal US was performed and, since it showed a CTr narrowing and a dynamic change in flow associated with breathing, the diagnosis of MALS was confirmed. Hence, she underwent a laparoscopic surgical procedure for dissection of the MAL, but six months later, she presented at our hospital because a recurrence of the symptoms began a few weeks after surgery; a new diagnostic work-up began.

The first examination performed was a Color Doppler US, which showed a narrowing of the celiac artery > 50%; tortuosity of the celiac trunk and a dynamic change in the arterial diameter during deep inspiration and expiration was shown by abdominal MRI angiography. Therefore, the patient was scheduled for a new surgical procedure in our hospital for dissection of the MAL.

After the induction of general anesthesia, the trans-esophageal echographic transducer was placed, and the peak systolic velocity (PSV) was measured, according to the method previously described. The mean value of three measurements realized during expiration was 133 cm/s. During the surgery, the TEE probe was left in place in standby mode, in order not to overheat and damage the surrounding tissues. At the end of the procedure, PSV was measured again, with the same hemodynamic conditions, ventilation setting, and pneumoperitoneum pressure value used in the first measurement. It showed a reduction in velocity, now oscillating with respiratory variations, between 76 and 98 cm/s (Figure 2).

(**A**)

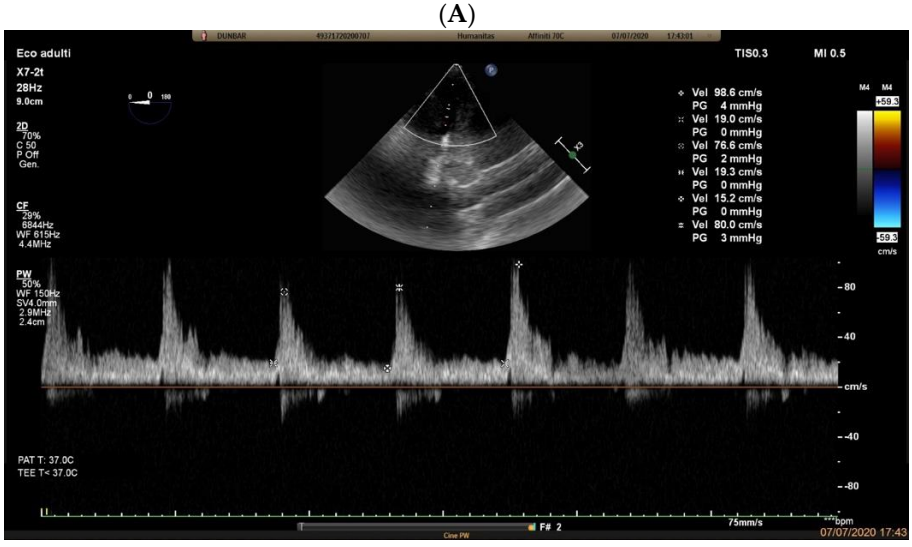


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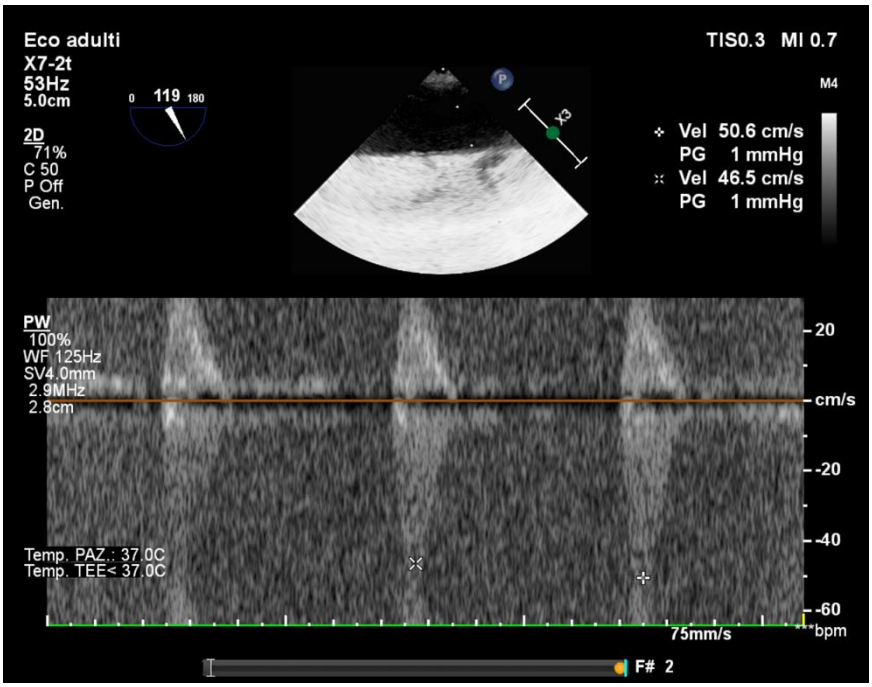
Figure 2. *Cont.*



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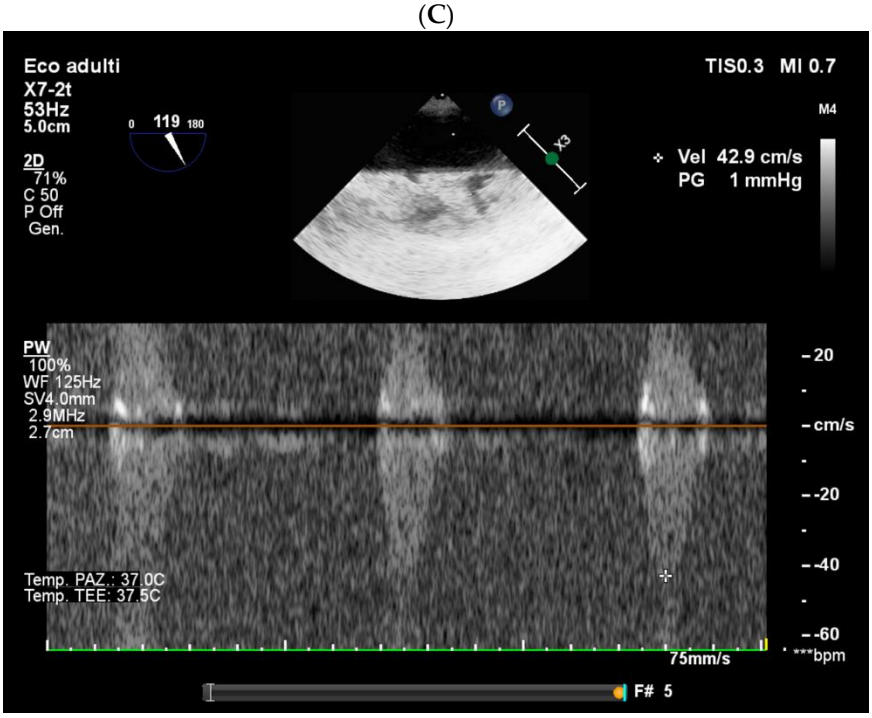

(**D**)

Figure 2. (**A**) Trans-esophageal pulse Doppler evalutaion of the celiac trunk after induction of general anesthesia in patient one. (**B**) Trans-esophageal pulsed wave Doppler evaluation of the celiac trunk at the end of the laparoscopic section of the arcuate ligament in patient one. Notice the variation in the PSV with the mechanical respiratory cycle. (**C**) Trans-esophageal pulse Doppler assessment of the celiac trunk after induction of general anesthesia in patient two. (**D**) Trans-esophageal pulse Doppler evaluation of the celiac trunk at the end of the procedure in patient two.

Once the procedure had been completed, the patient was awakened from general anesthesia and was observed in the post anesthesia care unit; finally, she was transferred to the ward for postoperative monitoring. During the hospitalization, she experienced no adverse events and no symptoms recurrence with food intake, which started on day 1

after surgery; on postoperative day 3, the patient was discharged from the hospital and no events were reported at one month follow-up [13].

2.3. Case Report 2

A 42-year-old man was referred to the Emergency Department of our hospital, suffering from recurrent pancreatitis for five years, with a known condition of pancreas divisum. He had been previously hospitalized several times in different hospitals and one year before, had undergone endoscopic retrograde cholangiopancreatography (ERCP) with sphincterotomy. However, he did not show any improvement and experienced new episodes of pancreatitis.

In our hospital, an abdominal CT scan was performed, which showed the presence of hemoperitoneum and a rounded hemorrhagic area in the head of pancreas, suggesting active bleeding; hence, the patient underwent an endoscopic procedure to embolize the bleeding pancreatic branch.

The arteriography revealed the lack of opacification of the common hepatic artery, and the hepatic branches were dulled by the pancreaticoduodenal arcade, with inverted flow. Moreover, both the pancreaticoduodenal artery and other small pancreatic arteries showed irregular courses and aneurysmal dilations.

In a subsequent multi-slice CT-angiography (MSTCA) follow-up, the compression of celiac artery was evident and, although the patient had no symptoms, given the suspicion that the episodes of pancreatitis were caused by the alteration of the splanchnic arterial vascularization due to MALS, the patient was scheduled for a laparoscopic surgical MAL dissection.

After the induction of general anesthesia, the trans-esophageal transducer was placed, and the PSV was measured. The average PSV value of three successive expiratory measurements was 47.5 cm/s. Due to the scars from previous pancreatitis' episodes, the surgical procedure was technically challenging, and the surgeon himself admitted difficulties in identifying the MAL and resecting it. During the surgery, the TEE probe was left in place in standby mode, in order not to overheat and damage the surrounding tissues. At the end of the procedure, the PSV was measured again, with the hemodynamics, ventilation setting, and pneumoperitoneum pressure unchanged. Still, the average value was 46 cm/s, not showing any significant change or reduction (Figure 2). The patient was then awakened and, after a brief observation in the post anesthesia care unit, he was transferred to the ward for post-operative observation. During the hospitalization, there were no new events, and the patient had no symptoms of MALS. Written informed consent and ethical approval for clinical case publication were obtained from both patients 1 and 2.

3. Discussion

MALS is a rare condition, more frequent in young people from 18 to 40 years old, especially women (4:1 ratio) and thin patients; the syndrome has a incidence of 2:100,000 [14], but the real prevalence of celiac artery compression is unknown, since it is often an incidental finding during examinations performed for other reasons [1,2]. Diagnostic imaging by Doppler US, MSCTA, MR angiography, or invasive selective angiography usually portrays a narrowing of the celiac artery, with a characteristic hooked appearance, which is different from atherosclerotic narrowing. Importantly, the examination should be performed during inspiration and expiration to identify dynamic variations in artery compression [4]. The same imaging methods have been used during follow-up visits to postoperatively confirm the success of treatment [7,15]. Indeed, successful treatment was confirmed by the presence of a normal flow in the celiac artery as noted by a Doppler ultrasound examination. The main limitation of this approach is related to performing the surgical procedure without any immediate assurance of its effectiveness. Indeed, both clinicians and patients have to wait postoperatively for clinical signs to appear and/or ultrasound follow-up to be carried out to know if the treatment has been effective. Although intraoperative pre- and post-decompression flow velocity studies could be performed to assess the success of the

procedure [16], previously used methods, such as surface ultrasonography or intravascular ultrasonography, either necessitate an exposure of the abdominal aorta or are invasive and necessitate another system and operator in the operative field [9].

In these two cases, we proposed the use of intraoperative TEE color-Doppler to evaluate in real-time the efficacy of the procedure. Compared to the previously mentioned technique, intraoperative TEE is easy to perform for this purpose, and is also faster and less costly. TEE is widely used to evaluate the heart and the thoracic aorta, but its usefulness to visualize the upper abdominal aorta and its branches has not yet been validated. However, in many cases TEE has been used to evaluate abdominal aortic vessels, especially in the context of aortic dissection [9,11,12,15,17]. Indeed, TEE may be useful to evaluate CTr and SMA involvement in abdominal aortic dissection and can distinguish between static and dynamic obstruction mechanisms [12]. In detail, the celiac artery has been proved to be that easy to identify in 97% of patients, and it has been proposed as an anatomical landmark to be routinely used during procedures involving the descending aorta [18].

In the first case, this new method was useful in assessing the outcome of the surgical procedure, showing, in real-time, a reduction in the PSV associated with the establishment of a normal blood flow in the celiac artery. Postoperative improvement in symptoms confirmed the clinical efficacy of the surgery. Indeed, the patient had already undergone surgery to resect the MAL without clinical results, since she experienced new symptoms. Similar outcomes have been described before [19], and we believe that an intraoperative monitoring of the efficacy of the procedure is particularly suitable when dealing with relapses of MALS.

In the second case, the result is controversial. Considering that the patient did not have any symptoms, and that the baseline PSV value, compared with the first case, was much lower, it is conceivable that the stenosis was not clinically significant from the beginning, and that the surgical resection was either futile or ineffective, since even post-procedure, the PSV did not change at all. In addition, the resection may not have been effective due to the procedural impediment reported by the surgeon caused by previous pancreatitis.

In addition, it is important to emphasize that laparoscopic surgery, which is currently the most commonly used procedure in these patients, has some advantages, but also some limitations. Specifically, it allows for performing small surgical incisions, carries a low risk of complications, and ensures a higher comfort level for the patients. On the other hand, the main disadvantage is that the resection is technically challenging, sometimes causing an incomplete release of the celiac trunk [1,4]. For this reason, it would be mandatory to define a method that can be used for the intraoperative monitoring of the success of the procedure.

It is worth noting that values predictive of the stenotic flow in the CTr and SMA are currently unknown. Using abdominal ultrasonography, a stenosis > 70% is identified by a PSV > 200 cm/s at the CTr level [20], with 87% sensitivity, 80% specificity, a positive predictive value of 63%, and a negative predictive value of 94%, but this has not yet been validated for TEE [18]. Still, we can expect similar values, even with TEE, although some doubts remain about values suggestive of stenosis when the obstruction of the celiac trunk is an incidental finding, or in patients with mild symptoms.

Finally, although the ability to visualize only the proximal third of the abdominal aorta and its branches is enough to evaluate the flow in the celiac trunk during MAL resection [12], the lack of standardization and validation in TEE regarding the abdominal aorta limits the general applicability of this approach. Moreover, although unlikely, using TEE as "transgastric echoangiography" may cause mucosal damage, and to date, further investigation is mandatory to establish the safety of this method.

4. Conclusions

Trans-esophageal echocardiography has been proven to be a simple, inexpensive, and easily reproducible way to measure the flow and peak systolic velocity in the celiac artery. Using TEE as an intraoperative monitoring during MAL resections may be useful to evalu-

ate the success of the surgical procedure, allowing the surgeon to obtain an intraoperative confirmation of the outcome of the resection, without the need to wait for postoperative follow-up evaluation. Intraoperative transesophageal US complements preoperative US assessment and postoperative follow-up aimed at increasing the quality of care in patients affected by MALS, especially in complex cases.

Author Contributions: Conceptualization, F.N. and E.G.; methodology, E.G.; investigation, F.N. and E.G.; writing—original draft preparation, M.L.; writing—review and editing, F.N.; supervision, E.G., F.P. and M.C.; project administration, U.R. All authors have read and agreed to the published version of the manuscript.

Funding: This research received no external funding.

Institutional Review Board Statement: The study was conducted in accordance with the Declaration of Helsinki and was approved by the Institutional or Ethics Committee of Humanitas Research Hospital.

Informed Consent Statement: Informed consent was obtained from all subjects involved in the study, and written informed consent has been obtained from the patients to publish this paper.

Data Availability Statement: Not applicable.

Conflicts of Interest: The authors declare no conflict of interest.

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