Abstract
Median arcuate ligament syndrome (MALS) is a rare cause of abdominal pain and weight loss, which is likely caused by compression of either the celiac artery (CA) or plexus by the median arcuate ligament. A case of MALS in a 25-year-old female with severe postprandial pain and weight loss is herein described. An imaging study demonstrated the abnormal "stealing" of the blood flow from the superior mesenteric artery (SMA) circulation through the pancreaticoduodenal arcade to the hepatic circulation, which was corrected by laparoscopic dissection of the MAL followed by percutaneous transluminal angioplasty (PTA) of the CA.

Key words: median-arcuate-ligament-syndrome, laparoscopy, additional PTA

Introduction
MALS is a rare disease. A review of the Japanese clinical database revealed only eighty case reports in Japan, since 1985, and laparoscopic surgery performed in only one case. English publications show over four hundred case reports since 1965. Forty of those cases were treated by laparoscopic surgery.

The median arcuate ligament (MAL) is a fibrous band formed of crossing the right and left diaphragmatic crura at the level of the CA. There are several patterns as the locations of the MAL. Concerning the anatomical variations of the MAL, it lies on the basal of the CA in the common cases. However, there is a report that about 20% of individuals have anatomical variables to the MAL, are located anterior and superior to the CA. For these anatomical reasons and the presence of structures like ganglions, the CA is compressed, narrowed. This leads abdominal pain and weight loss, the syndrome due to the aforementioned reasons is called MALS.

Case Report
A 25-year-old female was admitted to a nearby clinic because of repeated postprandial epigastric pain. She had 10 years history of upper abdominal discomfort, which was enhanced by eating, and had lost 3kg during the last 3 months. Upper GI endoscopy had shown negative findings. Enhanced computed tomography revealed the stenosis and poststenotic dilatation of the CA, accompanied by a dilated pancreaticoduodenal arcade, which was suggestive of increased collateral flow through the superior mesenteric artery. Based on these findings, median arcuate ligament syndrome was suspected, and the patient was referred to our hospital.

On admission, the patient’s height was 171cm, and her weight was 48kg. Her blood pressure was 111/68mmHg, and heart rate was 85/min. The laboratory findings revealed no undernutritional sign, the total protein level was 7.3g/dl and the albumen level was 4.6g/dl. Her abdomen was soft and flat, and no bruit was detected on auscultation. When a patient was first seen, we performed duplex abdominal ultrasound. The image was that the origin of the CA was narrowed and we found the blood flow of the CA was decreased. Detailed contrast en-
Enhanced three-dimensional CT revealed a hook-shaped stenosis of the CA with post-stenotic dilatation. It was conspicuous with full exhalation, suggestive of extrinsic compression by the MAL. Digital-subtraction angiography was performed to access the dynamic blood supply to GI tract. The selective angiography of the CA revealed no blood flow into the gastroduodenal artery, and the selective angiography of the SMA demonstrated a contrast enhancement of the proper hepatic artery through a dilated pancreaticoduodenal arcade. These findings suggested that the SMA was supplying collateral blood flow to the ischemic proper hepatic artery.

The patient underwent laparoscopic division of the MAL and celiac ganglionectomy. The surgical procedure was as follows: The patient was placed under general anesthesia in the supine position, with her legs spread apart. An optical 12mm Trocar was used to enter the abdomen midway on the umbilicus and four laparoscopic ports were used at the two hypochondriums and the bilateral regions. A liver retractor was placed to allow for the visualization of the surgical field by holding up the liver. The avascular region of the gastrohepatic omentum was divided by using a harmonic, and the right crus was isolated inferiorly to the cardia. The left gastric artery was identified, looped, and then was retracted with vascular tape, to expose the anterior aspect of the CA. The MAL was identified as a musculoaponeurotic band crossing over the anterior wall of the CA. The MAL and surrounding celiac ganglia were dissected using bipolar scissors, and the anterior surface of the CA and the aorta were exposed (Fig. 1). A surface imprint was recognized on the anterior aspect of the CA (Fig. 2), thus clearly showing the presence of both extrinsic compression and localized stenosis.

Intraoperative duplex scan and angiography were not routinely used. A drain was then placed near the CA, and the wounds were then closed in a routine fashion.
Although the patient’s postoperative course was uneventful, she had only a limited improvement of the symptoms. CT angiography at 1 week after the laparoscopic dissection of the MAL suggested that the CA was free from the extrinsic compression, but that localized stenosis remained at the origin of the CA. Four weeks after the operation, PTA was performed for the stenotic lesion of the CA. The initial selective angiography of the SMA showed a persistent “stealing” of blood flow through the pancreato-duodenal arcade to the proper hepatic artery (Fig. 3).

The stenotic lesion of the CA was confirmed with lateral aortography, and dilated with a PTA balloon of 4mm in diameter. After dilation, the “stealing” of blood flow from the SMA to the proper hepatic artery disappeared (Fig. 4). After the PTA was successfully performed, complete relief of the symptoms was achieved immediately. Three months after the PTA, the patient was doing well without any relapse of the symptoms, and she had also gained 3kg in weight.

Discussion
MALS or celiac artery compression syndrome (CACS) is a condition characterized by abdominal pain attributed to compression of the CA, and possibly the celiac ganglia, by the MAL. It was first described in a case report by Harjola et al in 1963. Subsequently, Dunbar et al. reported a series of successful treatments for this entity by surgical decompression of the CA in 1965. Controversies exist regarding the clinical features, mechanism and pathophysiology of this condition, and the observation of CA compression in asymptomatic patients had led to questions about the true existence of the syndrome. The clinical presentation in MALS varies, and both the diagnosis and the treatment can be challenging. Most patients are young females, with a thin body habitus. The pain is located in the upper abdomen and is often postprandial. Relief may be obtained in the knee chest position. The mechanism responsible for the pain is not completely understood. Different theories exist, but the most accepted one is that the increased blood demand through a compressed CA leads to foregut ischemia and subsequent pain, especially after meals. Some criticize this theory because altered CA circulation can be compensated by collateral blood flow from the SMA. In fact, ligation of the CA in cases of trauma, cancer resection, or thoracoabdominal aneurysm repair is tolerated well if the remaining mesenteric arteries are normal. Another theory, in which MALS cause symptoms due to mesenteric ischemia as a consequence of a "stealing" syndrome, where blood flow is diverted away from the mesenteric circulation to compensate for impaired blood flow.
flow through a stenosed CA, has also been proposed. This finding can be demonstrated with selective angiography of the SMA, where blood is shunted through the pancreaticoduodenal arcade to the hepatic circulation, and this finding often disappears after the successful release of the CA compression and stenosis. Our case clearly demonstrated that the mesenteric blood distribution was normalized after the successful treatment of the CA stenosis (Fig. 2, 3). Altered CA circulation may also cause an aneurysm to form at the pancreaticoduodenal artery or gastroduodenal artery, presumably as a result of the increased collateral flow in these arterial beds. In the presence of CA stenosis, the supply to the liver and spleen may be derived from the SMA via the inferior pancreaticoduodenal artery. The resultant chronic high flow through the pancreaticoduodenal arcade may lead to the formation of aneurysms. Other, less accepted theories suggest that the pain comes from direct compression of the celiac ganglia. The treatment for MALS includes the surgical release of the CA compression with dissection of the celiac ganglia. Initial attempts to treat MALS by endovascular angioplasty or stenting are not recommended, because the extrinsic compression may cause recoil restenosis, dissection, and fracture of the stent. On the other hand, chronic compression of the CA may lead to luminal stenosis by intimal hyperplasia, medial fibrodysplasia, and disorganization of the adventitia. This explains why surgical release of the CA compression by the MAL does not always relieve symptoms, and in some cases, further interventions like angioplasty or splanchnic revascularization may be needed. Our case required PTA after surgical release of the CA compression in order to achieve symptom relief. We thought that the operative indication for MALS was that the clinical symptoms include postprandial abdominal pain, nausea, vomiting, and unintentional weight loss were not improved by conservative treatment. Recently, laparoscopy has been considered to be a novel approach for the management of MALS. Both laparoscopic and open MAL lysis and celiac ganglionectomy can be safely performed with minimal patient morbidity and mortality. Late but milder recurrence of symptoms is frequently seen after both approaches. The laparoscopic approach thus make it possible to avoid laparotomy and it may be associated with shorter inpatient hospitalization and a decreased time until the start of oral food intake. We believe that laparoscopy is therefore a useful tool for the treatment of MALS, once an adequate diagnosis is established.

References