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POINT OF VIEW

The celiac axis compression syndrome (CACS): critical review in the laparoscopic era

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ABSTRACT

The celiac axis compression syndrome (CACS) due to median arcuate ligament (MAL) was first described by Harjola in 1963; originating postpandrial abdominal pain, weight loss, epigastric bruit and celiac axis stenosis > 75% in angiographic studies. This clinical condition has been the origin of controversies about its pathogenesis, diagnosis and its long term clinical results.

Advances in diagnostic imaging as 64 multidetector–row CT (MDCT), 3-D reconstruction, magnetic resonance (MR) and color duplex ultrasonography, provide better understanding of the syndrome and allow to identify the best candidates for surgical division of MAL fibers.

Since the introduction of laparoscopic approach, and also endovascular procedures, in 2000, a new perspective has established in this challenging syndrome. With the occasion of our own experience, a critical review of the syndrome is presented.

Key words: Celiac axis compression. Median arcuate ligament. Laparoscopic treatment.

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INTRODUCTION

The celiac axis compression syndrome (CACS), also known as median arcuate ligament (MAL), was first described by Harjola in 1963. In 1965 Durban reported the first cases serie with surgery (1,2).

The syndrome is clinically characterized by the triade: postpandrial abdominal pain, presence of an epigastric bruit which increases in expiration and the assessment of extrinsic compression (> 50%) of the celiac axis by the arcuate ligament by vascular imaging (angiography, CT-angiography, etc.).

Since the first description, the syndrome has been submitted to several controversies based on the variabilities in the results and also different physiopathological interpretations. Some authors have considered the CACS as the most frequent cause of chronic mesenteric ischemia, meanwhile its existence has been challenged by other authors (3-8). The introduction of CT-angiography with three-dimensional reconstruction, duplex abdominal ultrasound (US) and laparoscopic surgery has promoted a renewed interest in the syndrome. We present a critical review of the pathogenic mechanisms, the therapeutic options and results.

ANATOMIC BASES

The arcuate ligament is formed by tendinous fibers between the two diaphragmatic cruras, which form the ventral arch of the aortic hiatus of which four variants have been reported. An abnormal fibrous thickening of MAL obstructing from 70 to 100% of the celiac trunk lumen has been previously reported in necropsy findings and surgical descriptions as well (9-12).

Another cause of the syndrome has been related with the celiac trunk (CT) origin in the aorta. The CT origines between the 11 thoracic and first lumbar vertebra. The mentioned origin can be located craneally within the thorax, causing a compression by the MAL's fibres (1,9,12). A craneally origin of the CT has been described in patients with astenic habitus. Some of the patients describe

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a pain relief with the knee-chest position, due to the relaxing of the vessels impingement by the MAL.

Charette, Balaban and other authors (13,14), related the symptons with a thickening of the celiac plexus sustaining an extrinsic compression of the CT with flow impairment (15-19).

The syndrome is more frequent in young females with a thin body habitus, who many of them have previous history of weight loss as consequence of the intense postpandrial pain. As matter of fact, one of the characteristics of good response to surgical release was the mentioned above according to a seminal paper of Reilly, published in 1985, who reported the long-term follow-up (median 9 years, range 1-18) of 51 patients surgically treated (20).

A simultaneous stenosis of the celiac trunk with superior mesenteric artery and renal arteries compression originating chronic intestinal ischemia and renovascular hypertension have described (21-23).

PHISIOPATHOLOGY

Several hipothesis have been proposed regarding the origin of the syndrome, although is a topic of academic debate. The most common theory relates the symptons with the visceral ischemia originated by the extrinsic compression of the CT. In support of this there are the radiologic findings (CT-angiography, MR-angiography), the surgical descriptions and the symptons release (close to 80%) after surgical division of the MAL and release of the CT (4-6,18,23-25).

By contrast, another facts question this mechanism. The most mentioned is the experimental and clinical evidence that the complete occlusion of two of three main major abdominal arteries are required to provoque abdominal angina, as have been shown in animals or in oncologic and thoracoabdominal aneurism cases (7,11,26).

In 1969, Bron et al. reported 12.5% of asymptomatic patients with "radiologic findings" of CT compression. In 1972 Colapinto found signs of compression and stenosis on the 31% of asymptomatic patients (27,28). These data and the symptoms recurrence following the surgical division of the MAL would support the caution in attributing the patients' symptoms to the arterial occlusion and the existence of the syndrome (4,5,26).

The second pathogenic theory is known as "steal phenomenon" by which blood from superior mesenteric artery territory is diverted through collateral vessels concurrent with a high demand of oxygen by the gut. Kalapatapu has recently reproduced the symptoms inducing a selective splachnic vasodilatation in the superior mesenteric artery (31). In the experience of this author, the induction of symptoms is a clear sign for selecting surgical candidates, obtaining a 75% of success with MAL release.

In favour of this theory would be the reported consequences in young athletes after intense exercise in liver transplantation and following pancreatoduodenectomies in patients who had an unnoticed celiac trunk compression (29,30,32-35).

CLINICAL FEATURES AND DIAGNOSIS

The intense postpandrial epigastric pain (mean duration 20-30 minutes), nausea and vomiting are the most common symptons. The postpandrial pain induces a "food fear" and avoid eating, being cause of weight loss (5-10 kg). The pain may be relieved in the knee-chest position, and is more frequent in young females (3/1) about 40-50 years; although it has been reported in children and adults. The mean duration of symptoms range between three months and more than 10 years (5,7,11,20,23,26,27, 36).

Because of the difficulty in diagnosis of the syndrome, many patients have been previously diagnosed of functional disorders and had undergone an extensive gastrointestinal evaluation including endoscopy, motility studies, abdominal CT or sugical intervention as exploratory laparoscopy/laparotomy, without establishing a diagnosis. In some series, 10-15% of the patients had psiquiatric antecedents (5,7,11,36).

On physical examination, patients usually have a thin and leptosomatic habitus and an epigastric bruit –which increases in deep expiration– is present in 83% (20,24,37); although abdominal bruits occur in approximately 30% of healthy adults (18,38).

Imaging studies

The radiologic confirmation of CT compression and severe stenosis or occlusion is mandatory for the diagnosis. The gold standard of CTC syndrome was aortic-arteriography with lateral aortogram until the introduction of CT-angiography, 3D reconstruction and magnetic resonance (MR)-angiography.

In the lateral projections of aortography and selective mesenteric studies, an image of "hook" –characteristic of the MAL effect upon celiac axis- is assessed (Fig. 1). Postestenotic dilatation, collateral circulation and pancreaticoduodenal artery aneurisms have been reported (7,11,20,28). The patency of superior mesenteric artery and renal arteries must be evaluated. Duplex ultrasound have shown to be useful in assesing changes in CT orientation and increase in the peak systolic velocity (> 1,8 m/sec) in inspiration *vs*. expiration (39).

Currently the thin-section multidetector CT scanners along with 3D software in sagital plane is optimal for assesing the proximal portion of the celiac trunk, ruling out other causes of celiac artery occlusion or narrowing as atherosclerotic disease (40-42) (Fig. 2).

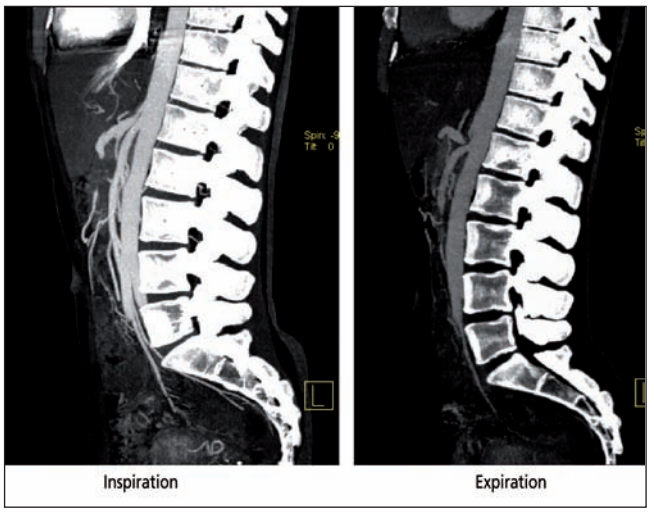

Fig. 1. Sagital contrast entrance volume-rendered technique showing hooked appearance and CT stenosis in a patient with postpandrial pain.
Estenosis tronco celiaco. Imagen sagital. Imagen 3D de una paciente con dolor agudo postpandrial y que muestra una angulación y estenosis del tronco celiaco.

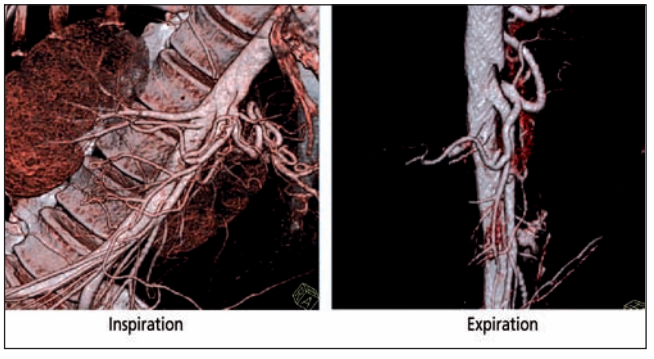

Fig. 2. Sagital 3-D volume-rendered image showing the CT stenosis in expiration.
Estenosis tronco celiaco. Imagen sagital y reconstrucción 3D con contraste mostrando la estenosis marcada en espiración.

Some authors have developed more specific studies in order to asses the CT occlusion and stablish a more precise surgical indication. Mensik et al. studied 320 patients with symptons of chronic intestinal ischemia, with selective angiography and gastric tonometry, yelded the CT syndrome in 43 patients (13.4%), indicating the surgical treatment exclusively in those who presented signs of gastric ischemia (gradient intraluminal gastric and arterial PCO_2), (43), obtaining a 83% of symptoms relieve in these patients.

We have previously described how Kalapatapu y cols. reproduced the gastric ischemic signs by inducing in four patients a selective vasodilatation in the splachnic bed, with 75% of abdominal pain resolution in the surgically treated. These findings confirm the ischemic theory of the syndrome and establish an objective criteria for surgical selection (31).

TREATMENT

Several endovascular procedures as angioplasty with or without stenting have been carried out with poor results. The sustained ant tight compression of the diaphragmatic fibers led to permanent changes on the vessel wall, fragmenting the stents and making this procedure unsuccesfull (44).

The other surgical techniques are the simple surgical division of the MAL and CT release with blood flow restoration assessing by Doppler-US and other complex procedures including vascular reconstruction with patch angioplasty of the celiac artery, aortoceliac bypass with saphenous vein or Dacron graft and reimplantation of the celiac trunk in the aorta (20,44,45-47).

The most common procedure is the surgical division of the constrictive fibers of the celiac plexus and release of the celiac trunk up its origin the aorta (Table I). Since the introduction of the laparoscopic approach in 2000, excelent results –with the benefits of this procedure, less hospital stay, faster recovering, less postoperative pain, etc. (48-55)– have been reported. The technique is depicted in figure 3.

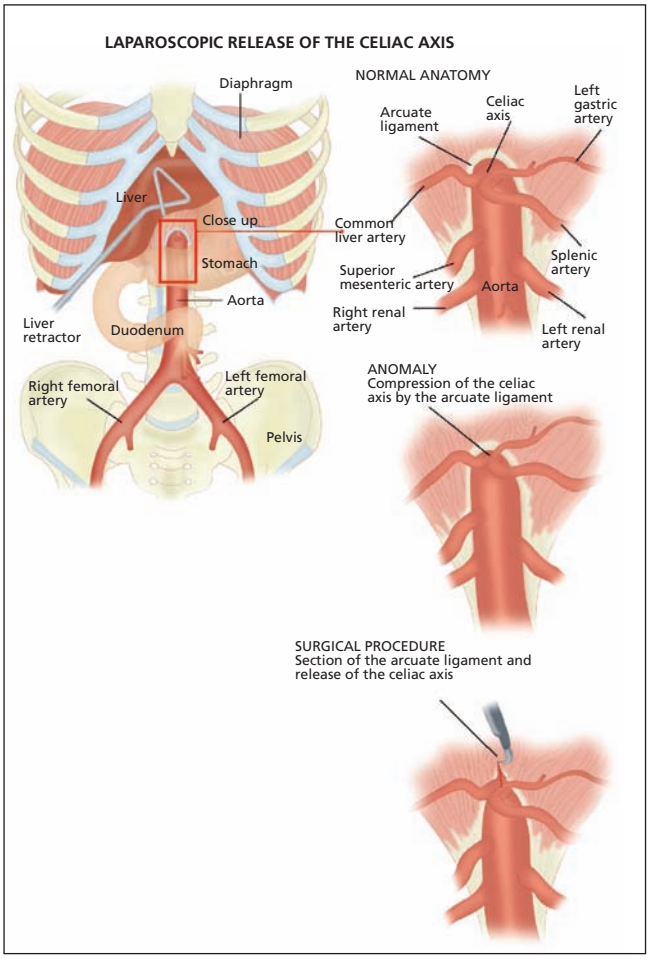

Fig. 3. A diaphragmatic representation of laparoscopic MAL division and CT release.
Representación esquemática de la técnica laparoscópica de la sección del ligamento arcuato y liberación del tronco celiaco.

Table I. Results obtained by the simple division of the MAL in the most numerous series

Author (ref.)	*Year*	*# cases*	*Release MAL**	*Improvement %*	*Follow-up*
Dunbar (2)	1965	13	13	69%	9-15 months
Szilagyi**(4)	1972	165	129	82%	1 month-4 years
Evans (5)	1974	47	47	83%	6 months-11 years
Watson (36)	1977	20	20	80%	6 months-4 years
Reilly (20)	1985	51	33	65%	1 year-18 years

*MAL: median arcuate ligament; **Literature review 1963-1971.

Table II. Experience with laparoscopic approach in the treatment of CTC syndrome

Author (ref.)	*Year*	*# cases*	*Surgery*		*Hospital stay*	*Follow-up*	*Improvement*
			Lenght	*Blood loss*			
Roayaie (48)	2002	1	135 min	55 cc	15 hours	3 months	Yes
Dordoni (49)	2002	1	125 min	65 cc	48 hours	6 months	Yes
Carbonell (50)	2005	1			72 hours	7 months	Yes
Baldassarre (51)	2007	1	130 min	-	No data	3 months	Yes
Jaik** (52)	2008	1	160 min	50 cc	48 hours	6 months	Yes
Jarry (53)	2008	1	168 min	-	24 hours	1.5 months	Yes
Duffy (55)	2008	1	218 min	-	24 hours	10 months	Yes
Vaziri (54)	2009	3	$\bar{X}$: 151 min (range: 114-217)	< 50 cc	24 hours	6 months	Yes
Rotellar*	2009	7	$\bar{X}$: 111 min (range: 90-120)	< 50 cc	65 hours (range 48-96)	6 months-8 years	Yes 2/3 Average 1/3

*Authors' results in 2009; **Surgery using Da Vinci Surgical System (Intuitive Surgical, Sunnyvale, California).

The cases reported in these series were well selectionated: young females with intense pain and weight loss. In our experience of 7 cases underwent surgery by laparoscopic technique, similar results were obtained (Table II).

As mentioned before, the results have been variable. Perhaps the main reason of that is the short-term follow-up and the diversity of abdominal pain medical profiles diagnosed as CTC syndrome.

The seminal paper paper published by Reilly in 1985 describing the long-term follow-up of 51 patiens treated surgically was crucial for explaining these divergences. In this report, 80% of young patients (40-60 years) with postpandrial pain, weight loss and postestenotic dilatation in the celiac angiogram, releaved the symptoms. Similar results were published by other authors and the same prognostic factors were found in regression studies: epigastric pain related with ingesta as more relevant parameter (20,46,56,57).

The accumulated experience points out the importance of a detalied anamnesis and the selective imaging techniques as Doppler-US, CT-angiography; avoiding unnecessary explorations.

The laparoscopic approach and minimally invasive techniques: MAL division, with ulterior endovascular techniques when necessary –angioplasty, Stent-, open a new "era" in the diagnosis and treatment of the syndrome described in 1963.

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