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GUIDELINES

EDITOR'S CHOICE

International Union of Angiology consensus document on vascular compression syndromes

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ABSTRACT

Vascular compression syndromes (VCS) are rare diseases, but they may cause significant symptoms interfering with the quality of life (QoL) of patients who are often in their younger age. Given their infrequent occurrence, multiform clinical and anatomical presentation, and absence of dedicated guidelines from scientific societies, further knowledge of these conditions is required to investigate and treat them using modern imaging and

surgical (open or endovascular) techniques. This consensus document will focus on known VCS, affecting the arterial and venous system. The position paper, written by members of International Union of Angiology (IUA) Youth Committee and senior experts, will show an overview of pathophysiology, diagnostic, and therapeutical approaches for patients with VCS. Furthermore, this document will provide also unresolved issues that require more research that need to be addressed in the future.

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Key words: Consensus; Peripheral vascular diseases; Quality of life.

1. Introduction and scope of the document

Most modern vascular practice encompasses (atherosclerotic and non-atherosclerotic) arterial disease and venous insufficiency. External compression of otherwise normal arterial and venous vessels represents a different and infrequent pathology. Indeed, vascular compression syndromes (VCS) are rare diseases, but they may cause significant symptoms interfering with the quality of life (QoL) of patients who are often in their younger age. These syndromes can cause ischemia (arterial entrapment) or stasis (venous entrapment) (Figure 1). Given their infrequent occurrence, multiform clinical and anatomical presentation, and absence of dedicated guidelines from scientific societies, further knowledge of these conditions is required to investigate and treat them using modern imaging and surgical (open or endovascular) techniques.

The approach to diagnosis and management of VCS should be tailored to each patient based upon several factors, including presence and degree of symptoms and their impact on QoL, vascular anatomy, availability of vascular conduits for revascularization, as well as comorbidities and age of patients. Indeed, conclusive evidence has yet to be published in literature, which is heterogeneous, and mainly coming from retrospective single-center series; however, some standards of practice may be established.

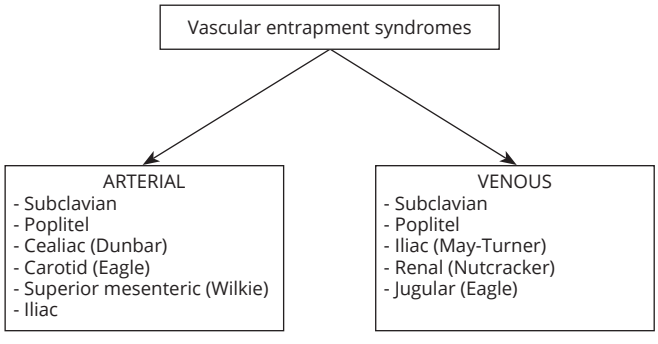

Figure 1.—Classification of vascular entrapment syndromes (VES).

This consensus document will focus on known VCS, affecting the arterial and venous system. The position paper, written by members of International Union of Angiology (IUA) Youth Committee and senior experts, will show an overview of pathophysiology, diagnostic, and therapeutical approaches for patients with VCS. Furthermore, this document will provide also unresolved issues that require more research that need to be addressed in the future.

2. General considerations on diagnosis

Vascular compression syndromes diagnosis depends on the type of vessel involved, the affected anatomical site and the underlying cause producing it. Doppler ultrasonography, computed tomography angiography or magnetic resonance angiography, are all essential tools for proper diagnosis, therapeutic planning, and follow-up (Table I).[1]

2.1. Doppler ultrasonography (DU)

It is considered the first line examinations for the diagnosis of vascular compression. It is notable for being a quick, inexpensive, non-invasive and dynamic examination. Enables clinicians to highlight the morphological changes and, also, to measure the degree of vascular stenosis. It is possible to identify the site of compression and indirect signs, such as dilatations (prestenotic and poststenotic tract). Also, it is possible to determine the increase flow in stenotic area and the peak systolic velocity (PSV) reduction in the prestenotic tract. This allows us to quantify

Table I.—*Imaging examinations for diagnosis of vascular compression/entrapment syndromes.*

Test	Advantages	Disadvantages	Indications
Radiography	• Cheap • Easily accessible • Non-invasive • Good for seeing the bones	• Radiation • Bidimensional • Bad for soft tissues	Thoracic outlet syndrome
Duplex ultrasound	• Quick • Readily accessible • Non expensive • Non-invasive • No radiation • Hemodynamic information (flow, stenosis degree, etc.) • Functional/provocation tests	• Bad visualization if overlying bones/air • Patient-, anatomy- and explorer-dependent	• Thoracic outlet syndrome • Popliteal entrapment • Visceral entrapment • Iliac artery endofibrosis • Femoro-popliteal vein entrapment • Iliac vein entrapment syndrome (May Turner)
IVUS	• Almost every vascular territory • Does not have X-ray proyection limitations. • Identify and describe lesions in the vessel walls • Very accurate and real time diameter measurements (for balloon/stent selection) • Can be simultaneously combined with arteriography/phlebography	• Invasive • Expensive • Requires some training	• Thoracic outlet syndrome • Popliteal entrapment • Visceral entrapment • Iliac artery endofibrosis • Femoro-popliteal vein entrapment • Iliac vein entrapment Syndrome • Nutcracker
Computed Tomography	• Contrast enhanced • Acurate definition of vessels/lessions • Anatomical structure relations • Bony and soft structures • 3D/multiplanar reconstructions • Non-invasive • Relatively available and quick	• Radiation • Contrast-induced nephropathy	• Thoracic outlet syndrome • Popliteal entrapment • Visceral entrapment • Iliac artery endofibrosis • Femoro-popliteal vein entrapment • Iliac vein entrapment syndrome • Nutcracker syndrome
Magnetic Resonance	• Good for shoft tissue • Non-invasive • Can provide hemodynamic information/flow direction • Dynamic studies • Non-ionizing radiation • Can visualize different structures depending on potentiation, avoiding contrast • 3D	• Nephrogenic sclerosis (gadolinium) • Time-consuming • Not easily available • Vascular image protocols difficult to establish	• Popliteal entrampment • Visceral entrapment • Femoro-popliteal vein entrapment • Iliac vein entrapment • Nutcracker/pelvic congestion
Angiography	• Hemodynamic information • Can confirm diagnostics • Can associate endovascular treatments • Allow dynamic imaging/provocation tests • Allow functional tests (intravascular pressure)	• Ionizing radiation • Contrast induced nephropathy • Invasive • Access complications	• Thoracic outlet syndrome • Popliteal entrapment • Visceral entrapment • Iliac artery endofibrosis • Femoro-popliteal vein entrapment • Iliac vein entrapment syndrome • Nutcracker syndrome

the degree of stenosis, which is achieved with the flow ratio.[2] Nevertheless, anatomical and patient factors may ultimately obscure vessels visualization and diagnosis of compression syndrome, for example difficult assessment of iliac veins within the abdomen or the subclavian vein below the clavicle. Usually, to reach a conclusive diagnosis a second-line method is needed to identify the compression, to exclude other causes of compression or also to highlight complications.[1-3]

2.2. Intravascular ultrasound imaging (IVUS)

IVUS allows direct evaluation of endoluminal content, which has recently become the reference method for arterial and venous pathologies. This method is particularly useful for the diagnosis and management of numerous vascular diseases, including aneurysms, dissections, thrombosis, and stenosis, and it has also been used in some vascular compression syndromes. IVUS adds maximum efficiency and information before deciding to implant any device. Although, it is still an expensive tool.[1-3]

2.3. X-ray

Radiographs of the chest and cervical spine are considered first-line in the imaging evaluation of all forms of thoracic outlet syndromes to evaluate for potential bony abnormalities.

2.4. Computed tomography (CT)

Contrast-enhanced CT is the most frequently recommended imaging method in clinically suspected syndromes. CT provides accurate detection of vascular structures and their relationship with adjacent organs. It provides a rapid, accurate, and reliable assessment of suspected compression syndrome, overcoming many of the limitations of US. Moreover, isotropic CT datasets allow robust post-processing capabilities. Multiplanar reconstruction facilitates image creation, with two-dimensional plane the best for the patient's anatomy visualization, while three-dimensional volume rendering provides an overall perspective. In addition, maximum intensity projection techniques are ideally suited to highlighting vascular structures.[4] The predisposing anatomic abnormalities may be incidentally discovered even in asymptomatic patients on abdominal CT imaging examinations performed for unrelated indications. Contrast-enhanced CT may be performed with different protocols according to the specific syndrome to maximize the visibility of involved vessels and allow the identification of typical imaging features and possible complications (for example with provocative maneuver in thoracic outlet syndrome). So that, dynamic imaging may be performed to evaluate patients with inconstant symptoms manifesting in a specific position.[5] Disadvantages of CT include risks associated with ionizing radiation exposure, which are of greater significance in the population most likely to be imaged for compression syndromes compression (young patients, premenopausal women).[1, 4]

2.5. Magnetic resonance angiography (MRA)

MRA has emerged as a leading imaging modality in the assessment of venous compression syndromes. MRA may be preferred in children or young patients due to the absence of ionizing radiation. It takes more exam time to be performed than CT but the potential risks of ionizing radiation exposure are mitigated. With the new MRA optimal venous contrast timing could be achieved. Non contrast MRI are also possible in patients with renal insufficiency. Like CT, MRA datasets allow 2D and 3D postprocessing techniques. MR flow-sensitive sequences also allow assessment of venous flow directionality, helping to predict the hemodynamic significance of compressive lesions.[5, 6]

2.6. Angiography

Venography or arteriography are still considered the gold standard, but they are an invasives procedures. Are useful for the direct measurement of pressure gradients and are sometimes needed in certain cases to establish the definitive diagnosis. These invasive modalities may be limited or reserved for situations in which intervention is planned.[5-7]

In conclusion, imaging studies are fundamental tools for the diagnosis therapeutic planning and monitoring of vascular compression syndromes. The combination of noninvasive imaging modalities, which allow the precise evaluation of anatomical structures, and invasive techniques, that are useful for the direct measurement of pressure gradients, is helpful to establish the definitive diagnosis. However, the invasive procedures should be limited to the most doubtful cases or when the endovascular treatment needs to be performed.[5-7]

3. General considerations on treatment and follow-up

3.1. Conservative treatment of vascular compression syndromes

The treatment of vascular compression syndromes (VCS) can be more challenging than other vascular injuries because of their psychological implications. Typically, correcting VCS involves a team effort from medical professionals specializing in several different fields. Physicians must work together to determine the underlying cause of their patient's symptoms and then create a treatment plan to address those causes.[8]

Treatment of VCS usually consists of surgery to correct the problem. In some cases, though, conservative methods are preferred. These include lifestyle changes, stress reduction and exercise. Arterial insufficiency is one of the most treatable causes of chronic pain syndromes in patients with VCS and many patients experience anxiety and depression after sustaining an injury to their limbs. This can make it difficult for physicians to determine whether the patient's symptoms are improving or worsening over time.[9] For people with VCS, for instance with popliteal entrapment syndrome (PES), exercise like walking is an important part of a treatment plan. Preferably, exercise should be done at least three times a week for at least twenty minutes at a time. Maintaining regular exercise habits will help reduce symptoms and promote overall health.[10] Moreover, in patients suffering from thoracic outlet syndrome (TOS) weight loss, and disease-specific physical therapy (PT) should be considered. PT for TOS includes periscapular muscle reinforcement, tendon and nerve gliding procedures, and postural exercises to reduce the compression on the involved neurovascular structures.[11]

Common VCS also include deep vein thrombosis (DVT), and acute compartment syndrome. These disorders cause severe pain, swelling and disability in the affected

limb. Depending on the severity of the disorder, physical therapists may help treat these conditions. Physical therapy can help patients with VCS reduce their pain levels through stretching and strengthening exercises. Historically, patients with acute DVT were confined to bed rest to prevent the clot from dissolving and causing a pulmonary embolism. Recent studies suggest that early ambulation and compression stockings are treatment options for acute VTE.[12] In addition, physical therapy can also help patients improve their mobility so that they can perform necessary tasks every day without straining their affected limb. Patients with vascular entrapment and DVT may also need to wear a compression stocking to reduce swelling and pain. Therapeutic options include compression sleeves that fit over the foot, ankle or knee, or compression tights for the calf muscles only.

3.2. Surgical and endovascular treatment of vascular compression sydromes

The pathophysiology of vascular compressive syndromes is typically related to the compression of a major blood vessel by an adjacent structure, such as a muscle, ligament, or bone. This compression can lead to a reduction in blood flow and subsequent ischemia, or a reduced venous return and subsequent deep venous thrombosis. VCS are rarely life-threatening and might require prompt surgical intervention to alleviate the pressure and restore proper blood flow and nerve function. Both endovascular and open surgical approaches are in current use, and the choice of surgical approach depends on various factors, such as the location and severity of the compression, the presence of associated complications, and the patient's overall health status.

Endovascular approaches are becoming increasingly popular, as they offer all the classical advantages over traditional open surgery. However, it has been very successful in treating stable compressions, mainly without any joint involvement. The endovascular approach has revolutionized the treatment of patients with May Thurner syndrome, a condition that was previously mostly left treated with lifelong compression, with pattencies rising to 73.4-98% in 12 months.[13] Additionally, the endovascular approach has provided a new lease of life to patients with previously forgotten pathologies like nutcracker syndrome and pelvic congestion syndrome.[14, 15] On the other side, still lacks proper evidence to prove its efficacy in some cases, and head-to-head comparisons with open approach.

Traditional open-surgical approaches have been the standard of care for treating vascular compressive syndromes for many years, still dominating in flexure regions. Usually, involve making a large incision to access the affected blood vessel.

Open surgery has been overwhelmingly successful in treating arterial entrapment syndromes, and it is hard for endovascular approach to surpass open surgery results in the next few years. In the popliteal entrapment syndrome, the artery is compressed by surrounding structures, and open surgery is still the preferred approach.[16, 17] Many centers report patency close to 100%.[18] In venous disease, it is rare to use a venous conduit, with only a few reports of procedures like the Palma and May-Husni procedures or even trauma, restricted to some specialized centers.[19]

The hybrid approach, which involves a combination of open surgery and endovascular techniques, is another treatment option for vascular compressive syndromes. The treatment of Paget-Schroetter syndrome (PSS) has been successful with a hybrid approach, which involves the removal of extrinsic compression using open surgery, as well as recanalization of the vein with endovascular techniques. Robotic surgery is also a treatment option that has been described in some cases of PSS.[20] Also, the use of thrombolysis as the first step in managing PSS combined with anticoagulation, followed by first rib resection and venous percutaneous transluminal angioplasty (VenoPTA) has resulted in improved final vein patency. This approach showed an odds ratio of 17 [4-199] ($P<0.001$) and lead to improved functional outcomes.

3.3. Antithrombotic therapy and special situations in vascular compression syndromes

Although surgery and endovascular interventions are mainstays of the management of VES, the pharmacological therapy, anticoagulation in particular, is cornerstone treatment, especially in these types of VES, where thromboembolism plays an important role. Among them, those in which the veins are compressed, deserve special attention, as veins are more susceptible to thrombosis, than arteries.

Management of May-Thurner syndrome (MTS) in pregnant patients is a matter of the utmost importance. The anatomic changes throughout gestation, including decreased venous return secondary to caval compression of the gravid uterus, increased venous stasis, and an increased hepatic production of coagulation factors (VII, VIII, X and fibrinogen) all contribute to the increased incidence of venous thromboembolism (VTE) in pregnancy, which is exacerbated in those with MTS.[21]

Anticoagulation in pregnant patients is accomplished with either low molecular weight heparin (LMWH) or unfractionated heparin. LMWH can be continued until de-

livery and held with signs of labor or 12-24 hours prior to scheduled delivery, depending upon the dose utilized. The risk of thrombosis is greatest in the weeks following delivery; therefore, more aggressive therapy with intermediate or adjusted dose anticoagulation could be considered for patients with additional risk factors. Nevertheless, systemic anticoagulation as monotherapy is insufficient treatment for patients with MTS, given that it does not address the underlying compressive etiology. Post-thrombotic syndrome (PTS) occurs in as many as 73% of patients with MTS treated with either anticoagulation alone or in combination with thrombectomy. Therefore, anticoagulation should be used as an adjuvant treatment to a more aggressive approach in these patients.[21]

Recent findings suggest that pregnancy is not contraindicated after endovenous stenting for MTS, supporting the concept that women of child-bearing age with symptomatic MTS can be considered for treatment.[22] However, patients undergoing iliac vein stenting with MTS and VTE risk factors require prophylactic anticoagulation during pregnancy.[22]

Paget-Schroetter syndrome (PSS) is another type of VES that deserves special attention. In a recent meta-analysis, the proportion of PTS was higher in patients with PSS, than secondary upper extremity deep vein thrombosis (UEDVT).[23]

While initial management for all UEDVT patients consists of anticoagulant therapy, in patients with PSS, the underlying venous thoracic outlet syndrome necessitates a more aggressive management strategy.[24] Several therapeutic options exist, apart from surgical decompression through first rib resection, including catheter-directed thrombolysis and percutaneous transluminal angioplasty of the vein. However, some controversies exist regarding their indication and timing.[24] Nevertheless, recent ESVS guidelines recommend to consider thrombolysis within the first two weeks in selected young and active patients with UEDVT with severe symptoms.[25] Although no randomized controlled data support this recommendation, very recent analysis strongly suggests higher rates of thrombus and symptoms resolution with thrombolysis, followed by first rib resection, in PSS patients.[26]

4. Superior mesenteric artery and celiac artery compression syndromes

4.1. Superior mesenteric artery compression syndrome

4.1.1. Definition and pathophysiology

Wilkie syndrome is a rare (0.013-0.3%) compressive syndrome that results in upper small bowel obstruction due to entrapment of the third part of the duodenum between the superior mesenteric artery ventrally and the pulsation of the anterior wall of the aorta dorsally at the lower part of second lumbar vertebra or upper part of third lumbar vertebrae.[27] This syndrome is also known as superior mesenteric artery syndrome (SMA syndrome), cast syndrome, aortomesenteric syndrome, mesenteric root syndrome, chronic duodenal ileus and vascular duodenal compression.

David Wilkie gave the first comprehensive description in 1927 as a chronic duodenal obstruction; however, it was first mentioned in 1861 by Carl Von Rokitsansky.[28] It is more common amongst females in the second and third decades. Wilkie syndrome is attributed to the narrowing of the aortomesenteric angle and aortomesenteric distance of SMA to Aortic wall with loss of mesenteric or perivascular fat.[27, 29] Peri-mesenteric fat acts as a natural cushion to maintain the aortomeseteric angle to prevent extrinsic compression.

The normal aortomesenteric angle is 45°, but it varies from 20° to 70°, and the anortomesenteric distance range from 10-28 mm (Figure 2A, B). Any decrease in the angle below 16° and the distance less than 7-8 mm is diagnostic of Wilkie Syndrome (Figure 3A, B).[27] This aortomesenteric angle contains the left renal vein, the pancreatic uncinate process and the third part of the duodenum. The duodenum is held in that angle by the ligament of Trietz and is cushioned by the retroperitoneal fat and lymphatics. The main contributing factor is severe weight loss, resulting in diminution of visceral fat and narrowing of the angle.

Wilkie syndrome could have congenital or acquired causes (Table II).[27] Congenital causes include abnormal high insertion of the ligament of Treitz with high duodenal

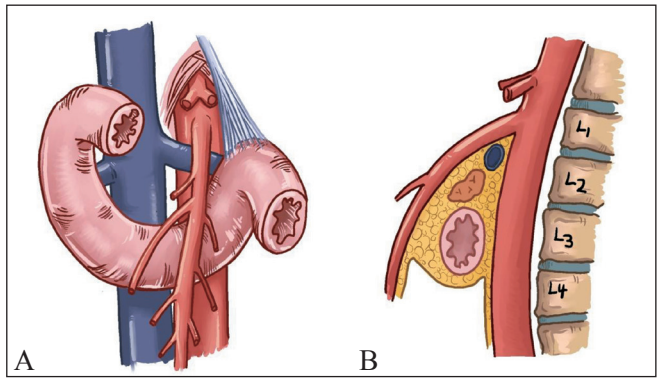

Figure 2.—Schematic diagram showing: A) normal anatomical relationship between superior mesenteric artery (SMA), duodenum, uncinate process, and left renal vein, where duodenum, Uncinate process and left renal vein pass underneath the superior mesenteric artery; B) normal aortomesenteric angle and aortomesenteric distance.

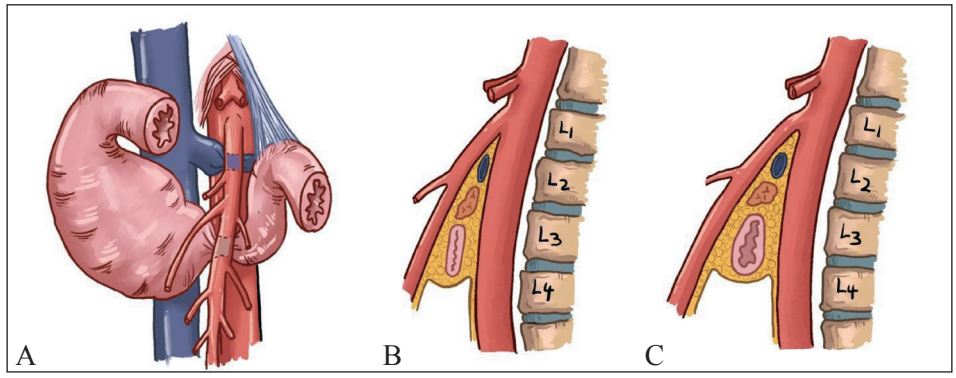

Figure 3.—Schematic diagram showing: A) Compression of the duodenum (Wilkie or superior mesenteric artery syndrome) and left renal vein (nutcracker syndrome) by superior mesenteric artery with dilation of the proximal duodenum; B) reduced aortomesenteric angle and distance with compression of the duodenum (Wilkie or superior mesenteric artery syndrome) and left renal vein (nutcracker syndrome) by the proximal superior mesenteric artery; C) post total parenteral nutrition (TPN), with the restoration of the mesenteric fat and increase in aortomesenteric angle and aortomesenteric distance.

TABLE II.—*Core features of Wilkie Syndrome: symptoms, diagnosis and management.*

SMA syndrome or Wilkie Syndrome			
Causes	Symptoms	Diagnosis	Management
(a) Congenital: • Abnormal insertion or abnormally high ligament of Treitz • Hypertrophy of the ligament • Duodenal malrotation to a cranial position • Short intestinal mesentery • Anomalous or low origin of the SMA • High duodenal fixation • Increased lumbar lordosis • Visceral ptosis • Peritoneal adhesions (b) Acquired: • severe weight loss (tumours, burn, malabsorption syndrome, anorexia nervosa, malignant cachexia, AIDS, prolonged bed rest, poly-trauma, hyper-catabolic state and drug abuse) • postoperative (spinal surgery, body casting, open aortic aneurysm or dissecting aortic aneurysm repair)	• Weight loss • Post-prandial abdominal pain • Early satiety • Bloating • Vomiting	(a) Initial: upper gastrointestinal series (b) Confirmative: • Computed tomography • Magnetic resonance imaging	(a) Conservative: • Nasogastric tube • Enteral feeding • Total parenteral nutrition (b) Surgical: • Strong procedure • Gastrojejunostomy • Duodeno-jejunostomy ± distal duodenum resection

fixation, hypertrophy of the ligament, duodenal malrotation to a cranial position, short intestinal mesentery, the anomalous or low origin of the SMA, increased lumbar lordosis, visceral ptosis, and peritoneal adhesions. Similarly, acquired causes are due to hyper-catabolic state with weight loss due to tumours, burn, malabsorption syndrome, anorexia nervosa, malignant cachexia, AIDS, prolonged bed rest, poly-trauma, and drug abuse.

Post-operative reasons are attributed to corrective spinal surgery, body casting, and open aortic aneurysm or dissecting aortic aneurysm repair.[27, 29] A complex Wilkie Syndrome is usually associated with renal 'Nutcracker syndrome' where the left renal vein is wedged between SMA and the aorta (Figure 3A-C).[30]

4.1.2. Diagnosis

A high degree of suspicion is required for diagnosis due to the vague and non-specific presentation and symptoms onset to diagnosis time vary from 1-5 years.[31] Symptoms include post-prandial abdominal pain, early satiety, bloating and vomiting in the background of unexplained weight loss. These symptoms are aggravated by lying supine after eating and are relieved by assuming the left lateral decubitus, prone or knee-chest position. Physical examination is always inconclusive except for the distended abdomen. The symptoms are also relieved by elevating the root of the mesentery by Haynes' manoeuvre, where pressure is exerted below the umbilicus in the cephalad and dorsal direction.[32]

Upper gastrointestinal series of barium swallow shows proximal dilatation with distal narrowing of the duodenum and pinpoint the diagnosis in 95% of the cases.[32-34]

Haynes radiological criteria characteristic of Wilkie's Syndrome:[27, 32-34]

• dilatation of first and second parts of the duodenum with or without gastric dilatation;

• abrupt vertical cut-off over the third part of the duodenum;

• transient hold-up of the contrast in the gastroduodenal segment with to and fro motion;
• quick distal flow of the contrast on assuming the left lateral position.

These criteria are essential to pinpoint the duodenal outflow obstruction.

However, contrast computed tomography is becoming the diagnostic modality of choice as it provides crucial information about the aortomesenteric angle/distance, perivisceral fat content, and site of obstruction to rule out other pathologies.[27, 35, 36] We learned through our experience that a narrow SMA angle alone is not diagnostic, as it is not uncommon to see a narrow aortomesenteric angle in paediatric populations and those with low Body Mass Index (BMI).

4.1.3. Management and follow-up

Initial conservative management consists of decompression of the duodenum with a nasogastric tube followed by correction of the electrolyte imbalance and nutritional support.[27, 35, 36] It is essential to promote weight gain and restore the perivisceral fat in order to obliterate the narrow aortomesenteric angle. Weight Gain can be achieved by tube feeding or total parenteral nutrition (TPN). Most patients respond to conservative management (Figure 3C); however, some will require surgical intervention.[35]

Surgical interventions include Strong's procedure, gastrojejunostomy, and duodenojejunostomy.[27, 31, 35, 36] Strong's procedure, especially in paediatric patients, consists of the division of the Treitz ligament; however, it is not recommended due to its high failure rate. Similarly, gastrojejunostomy is associated with increased postoperative complications of blind loop syndrome, biliary reflux, peptic stomal ulceration, and failure to decompress.

The presence of marked gastroduodenal dilatation requires duodenojejunostomy between the dilated second part of the duodenum and a loop of the jejunum just distal to the duodenojejunal flexure, and it is increasingly the surgical treatment of choice due to a higher success rate with rare recurrence.[27]

4.1.4. Unresolved issues and goals for future research

Wilkie Syndrome lacks consensus on diagnosis and optimal management strategy, including duration of conservative treatment. Although conservative management has a high success rate (80-86%), around 16% show relapse.[27, 31, 35, 36] Surgical treatment has failure rate of 8-21%, and patient selection is the key.[27, 31, 35, 36] Endo-luminal or laparoscopic surgery is a viable option,[37] but there are limited studies with uncertainty about its efficacy compared to open surgical intervention in the long run. Finally, as most studies are either case-based or retrospective, it is important that we establish an international registry to conduct more multicentre prospective or controlled studies to further understand the pathophysiology and optimal management of this rare disease.

4.2. Celiac artery compression syndrome

4.2.1. Definition and pathophysiology

Celiac artery entrapment syndrome, more commonly referred to as median arcuate ligament syndrome (MALS), is the clinical syndrome resulting from compression of the median arcuate ligament (MAL) on the celiac axis. The celiac artery branches from the aorta below the MAL at approximately the T11-L1 vertebral level. Surrounding the celiac artery origin, the celiac plexus is the largest autonomic plexus and includes a network of nerves which relay visceral sensory information from the foregut and midgut, including nociceptive impulses (Figure 4A). MALS occurs due to variations in the position of the celiac origin or the ligament which ultimately render an individual more prone to compression.

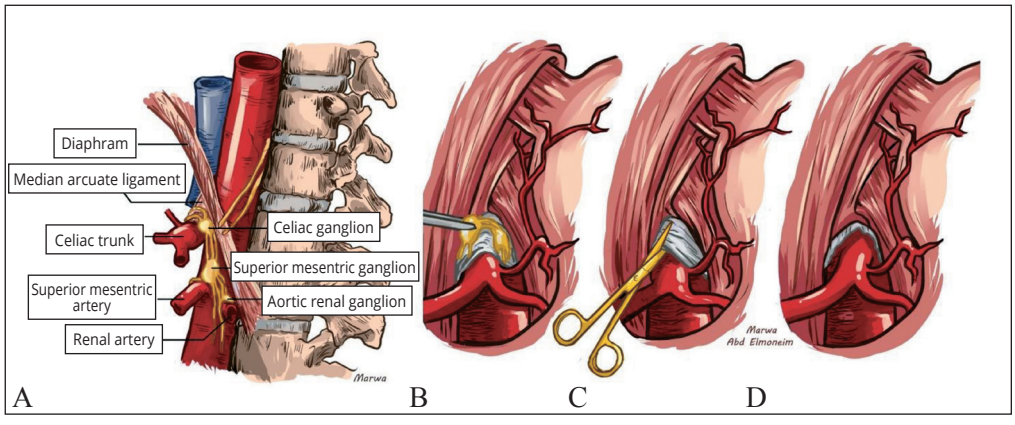

Figure 4.—Schematic diagram showing: A) Anatomical relationship between celiac trunk, median arcuate ligament and celiac ganglion; B-D) decompression with celiac sympathectomy in patients with median arcuate ligament syndrome. The whole celiac axis is dissected free from the tough paraaortic dense autonomic nerve tissue of the CA plexus. Freeing the whole CA to its third division is mandatory by removing any tissue overlying the anterior and medial-lateral aspects of the CA, including large nerve complexes and lymphatics.

Although up to 25% of the population may have MAL compression, it is estimated that approximately 1% of those develop symptoms.[38] While previously considered to be a primary vascular disease, it is now appreciated that MALS is also a neuropathic condition.[39, 40] Leading theories on the aetiology of pain in MALS center on the compression of the celiac axis and plexus with resulting intermittent foregut ischemia and overstimulation of pain fibres, respectively.[39-41] However, there is no widespread consensus on the pathophysiology of MALS, making thoughtful patient selection critical to diagnosis and management.

4.2.2. Diagnosis

Patients with MALS are often found have non-specific gastrointestinal complaints, such as epigastric pain, nausea, vomiting, or weight loss (Table III). Patients also often are referred followed gastroenterology work-up which is important given the non-specific clinical presentation.

A full blood count, liver enzymes, albumin, ferritin, thyroid function tests, glycated haemoglobin (HbA_{1c}), homocysteine, abdominal pelvic ultrasound, OGD, colonoscopy, DUS of celiac and SMA before and after drinking milk and in inspiration and deep expiration, and CTA aorta are required for differential diagnosis. A practical therapeutic algorithm was developed to guide physician (Figure 5).

Mesenteric duplex with peak systolic velocity (PSV) measurements during inspiration and expiration are performed. Turbulent celiac artery flow with PSV >200 cm/s or no flow during expiration, which decreases during inspiration or rest, are suggestive of MALS.

If positive duplex findings advocate celiac stenosis in the context of symptoms consistent with MALS, the patient is considered for CTA to delineate the CA entry and folding angle. CTA will demonstrate a focal celiac narrowing with post-stenotic dilation.

If positive, we commence total parenteral nutrition (TPN) for one to two weeks, aiming for albumin above 30 g/dL to improve the patient's general ability to tolerate surgery.

Those presenting with an atypical pattern of symptoms or who display equivocal duplex findings undergo more imaging with a HIDA liver scan to rule any dysfunctional biliary secretion and are consequently discussed with a gastroenterologist with a particular interest in MALS for further workup for alternative diagnoses. Gastric exercise tonometry is used as an adjunctive diagnostic test, to measure arterial and gastric CO_2 levels before, during, and after exercise. A positive test consists of gastric-arterial CO_2 level difference >0.8 kPa, an increase in gastric CO_2 level above baseline, or an arterial lactate <8 mmol/L.

4.2.3. Management and follow-up

Treatment is indicated in patients with confirmed celiac artery compression with respiratory manoeuvres on duplex. Celiac plexus block is utilised in some centres for temporary relief of MALS symptoms and a possible prognostic modality that supports its neurogenic pathophysiology.[39-42]

DeCarlo *et al.* found that the absence of symptom improvement after celiac sympathetic block is an independent risk factor for treatment failure after surgery.[43] In the largest MALS cohort including 516 patients enrolled in the Vascular Low Frequency Disease Consortium, investigators found that patients who did not have symptomatic relief after celiac plexus block had a 2.18-fold increased likelihood of surgical treatment failure.

Surgical decompression *via* MAL release remains the mainstay of treatment. Laparoscopic, open, and robotic-assisted approaches have been described.[43, 44] In addition to MAL release, concomitant celiac neurolysis is also often performed to address the neuropathic component of the disease (Figure 4B-D).[39, 40] Open and laparoscopic MAL release appear to have similar rates of long-term treatment success, although open release is associated with a higher rate of perioperative complications such as ileus and increased length of stay, moreover laparoscopic repair may result in serious bleeding from the friable coeliac axix.[40, 43, 45]

After MAL release, confirmation of decompression is performed intraoperatively either under direct visualization, intraoperative duplex ultrasound, or aortography in cases where a hybrid room is available.

Postoperatively, patients are followed with surveillance

TABLE III.—*Core features of MALS symptoms, diagnosis and management.*

Syndrome	Symptoms	Diagnosis	Management
Celiac artery compression syndrome	• Epigastric pain that may be worsened by eating, exercise, or forward flexion • Unintentional weight loss • Nausea or vomiting	• Duplex ultrasound with respiratory manoeuvres • CT angiography • Endoscopy and gastroenterology work-up to rule out other aetiologies	• Celiac plexus block for physiologic testing and prognostication • Surgical MAL release • Revascularization for residual or recurrent celiac artery stenosis

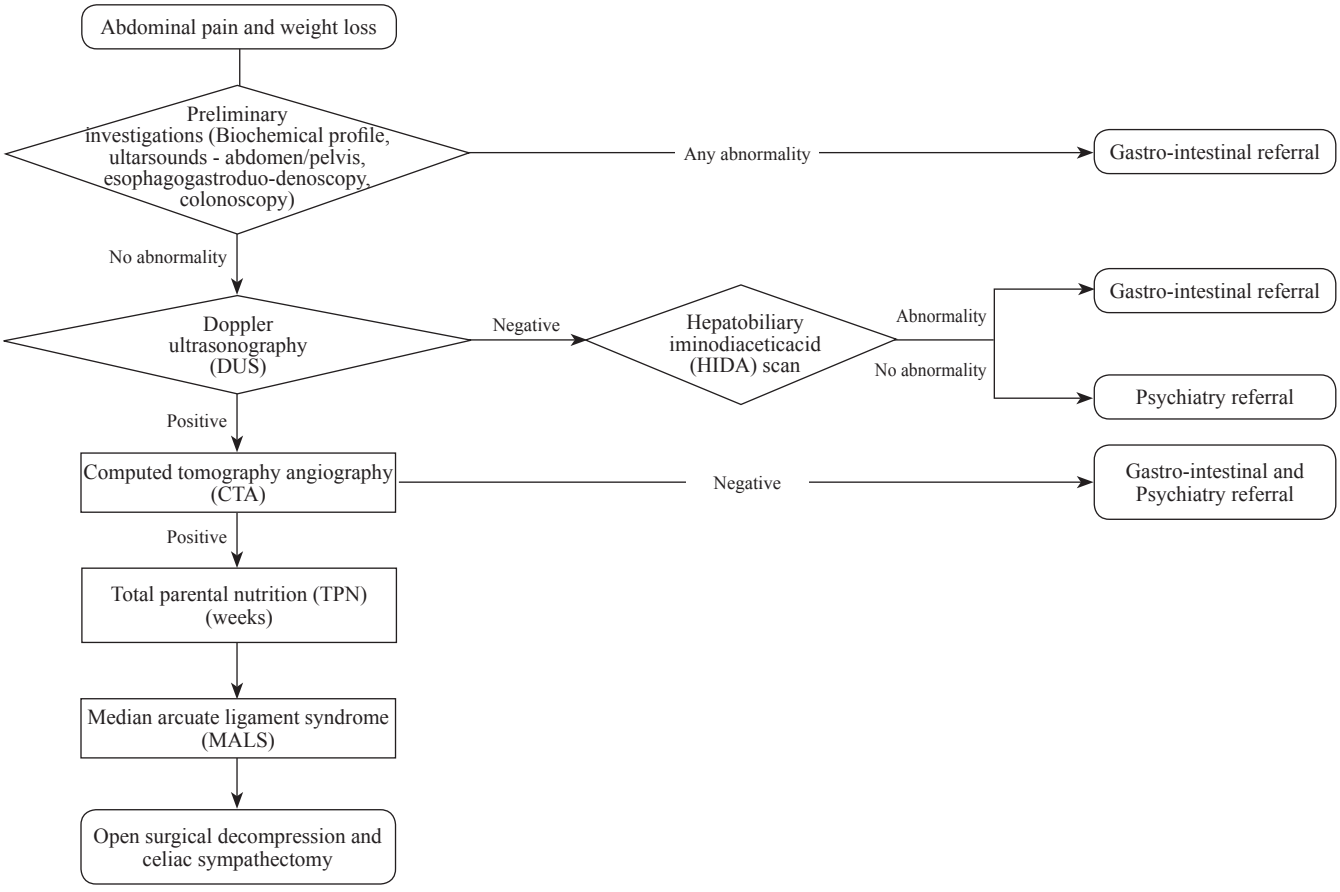

Figure 5.—Practical therapeutic algorithm employed in patients with abdominal pain and weight loss.

duplexes. In the case of unresolved symptoms and residual or recurrent stenosis, revascularization is recommended. Both angioplasty, stenting, and mesenteric bypass have been reported, but no technique has been reported as superior.[46] Lastly, in comparison with open and laparoscopic approaches, robotic release has been associated with inferior results, with approximately 65% of patients reporting no symptom relief at one year.[40, 43]

4.2.4. Unresolved issues and goals for future research

MALS remains a challenging entity to diagnose and treat. After an extensive pathway of work-up and subsequent surgical release, the overall rate of treatment failure is high, inspite early pain relief, as 35% of patients report symptom recurrence at one year, followed by 50% at three years.[39, 40, 43, 44]

Currently, although celiac plexus block is a proposed modality for prediction of treatment success after surgical decompression, more studies are needed to determine optimal patient selection. Laparoscopic release appears to be associated with greater reinterventions rates, driven by endovascular celiac artery angioplasty or stenting.[43] Given this, it has been suggested that an open-first approach is reasonable in those who are expected to undergo concomitant revascularization.[39, 40] However, there are currently no known radiographic parameters to guide need for revascularization. Knowledge of specific pre-operative symptoms, comorbidities, or imaging findings to guide patient selection would be welcome knowledge and would help advance the management of this often debilitated patient population.

5. Iliac artery endofibrosis

5.1. Definition and pathophysiology

Iliac artery endofibrosis (IAE) is a rare condition that predominantly affects young, male, endurance athletes, particularly cyclists. It is defined as pathological subintimal

fibrosis that results in impaired blood flow and claudication, with over 90% of patients affected in the external iliac artery, often with a fibrotic segment measuring up to 6cm in length.[47] IAE commonly affects the left leg, and sometimes, bilateral disease is present in patients with unilateral symptoms.[48]

The true incidence of IAE is unknown but a study in elite cyclists suggest the prevalence may be as high as 10-20%,[49] and its incidence is expected to increase due to the growing participation in cycling and other endurance sports that involve repetitive hip flexion, such as speed skating, endurance running, and triathlons.

Unlike atherosclerosis, the risk factors for IAE are not fully understood, and familial predisposition to cardiovascular disease does not appear to represent a significant risk factor. The pathophysiology of IAE remains unclear, with several theories proposed in the literature. The most commonly suggested explanation is that repetitive stretching of the EIA during high-intensity cycling leads to direct mechanical stress, causing the development of endofibrosis and subsequent stenosis. Although arterial kinking and vasospasms have also been described, controversy exists as to whether these are responsible for limiting blood flow during exercise.[50, 51]

The most common symptom of IAE is exertional discomfort, particularly in the thigh, which is typically relieved within five minutes of ceasing exercise. Patients may also experience leg weakness and numbness in the thigh or calf. Without treatment, IAE may progress in those who continue athletic activity, with some reports of EIA occlusion.[52]

5.2. Diagnosis

The diagnosis of iliac artery endofibrosis is often delayed due to the non-specific nature of its symptoms as well as normal physical examination at rest. In high-intensity athletes, muscle pain and fatigue are expected, further complicating the diagnosis, as symptoms are often attributed to musculoskeletal or neurological causes. Additionally, young, healthy athletes are not typically associated with arterial disease, contributing to the diagnostic delay.

Non-invasive imaging and testing are essential in the diagnosis of IAE. The first line investigation includes Ankle-Brachial Pressure Index (ABPI) and Doppler US, which are conducted before and immediately after exercise to confirm or exclude IAE. The length of exercise testing varies depending on the time it takes for patients to experience symptoms.

Patients with IAE would usually have a normal pre-exercise ABPI that decreases after exercise. There is currently no consensus regarding the absolute value of post-exercise ABPI drop, although a drop between 21-40 mmHg was considered to be diagnostic.[53]

Doppler US findings of monophasic post-exercise waveforms throughout the iliac arteries, with a peak systolic velocity (PSV) of >350 cm/s and end diastolic velocity of >150 cm/s are also suggestive of severe disease.[54]

Cross-sectional imaging such as CT and MRI and invasive modalities like angiograms offer little value in the initial diagnosis of IAE due to the high sensitivity of exercise ABPI and Doppler ultrasound. However, they may be used in the event of diagnostic uncertainty to highlight findings in the later stages of IAE.

5.3. Management and follow-up

The management of IAE involves educating patients on available treatment options and attempting conservative measures such as reducing exercise intensity or discontinuing cycling. Non-operative interventions should be the mainstay of treatment. Cyclists should be advised to reduce the time spent cycling and make adjustments, such as raising handlebars or adjusting saddle position forward to reduce hip flexion. Unfortunately, these measures often fail to relieve symptoms and are not always realistic for professional athletes. Endovascular approaches such as angioplasty and endoluminal stenting appear to be ineffective in treating IAE.

In the absence of specific medical therapy and reliable endovascular treatment for IAE, open surgery remains the preferred treatment when conservative approaches fail, and quality of life is severely affected. The surgical options vary depending on the investigation findings (Figure 6). Patients with abnormal vessel fixation and arterial kinking will benefit from an arterial release of fibrous tissue, whereas patients with longer EIA may benefit from vessel shortening with endarterectomy and vein patch repair. In cases of occluded external iliac artery, bypass surgery should be considered. Other alternatives to treat a proven intravascular lesion with normal vessel length include complete resection of the stenosed segment and replacement with an interposition graft. Arterial release should not be performed in isolation but rather as an adjunct for patients with arterial kinking and significant endoluminal narrowing.

In the post-operative period, patients should refrain from exercising for at least 6-8 weeks. Follow-up appointments should include annual clinical assessments with non-invasive Doppler ultrasound, when available, to detect potential complications such as pseudo-aneurysms.

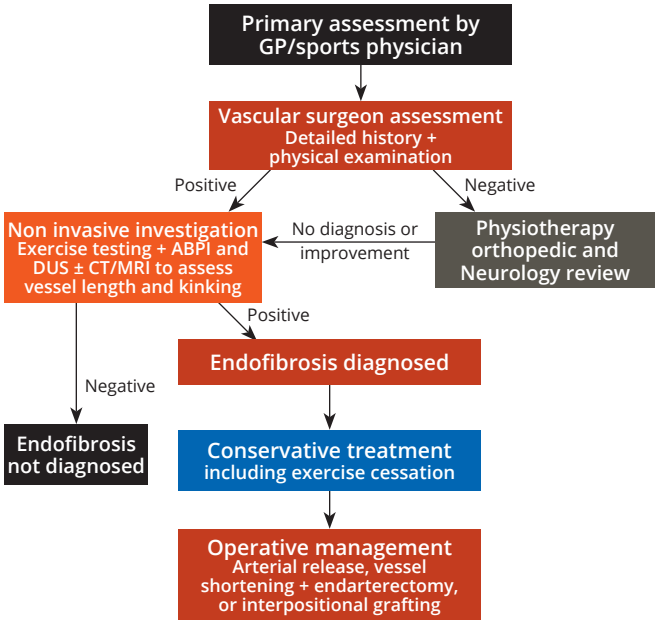

Figure 6.—Flow diagram of proposed evaluation and management plans for suspected or confirmed iliac artery endofibrosis.

5.4. Unresolved issues and goals for future research

Currently, there is a lack of standardized care recommendations for the assessment and management of IAE, except for a management consensus issued by the INSITE Group.[53] To improve patient outcomes and prevent harm, vascular surgeons worldwide should collaborate to develop a disease registry for IAE patients undergoing surgery. This can enhance understanding and promote clinical research on a larger scale.[55]

Finally, it is crucial to collaborate with governing bodies to advance research in this field, including implementing an exercise screening program to identify individuals at risk of developing IAE. Such initiative can lead to earlier diagnosis of IAE in high-endurance athletes and ultimately prevent further progression of the condition.

6. Femoral and popliteal compression syndrome

6.1. Definitions and pathophysiology

First described in 1879 by Stuart, popliteal entrapment syndrome (PES) is a uncommon vascular pathology affecting approximately 0.17-3.5% of the population resulting from an embryologic mal-development of the muscle heads of the lower limb and the femoral artery.[56, 57] In the course of preparing a dissection of a patient who had succumbed to gangrene, Anderson Stuart noted an aberrant course of the medial head of the gastrocnemius in relation to the popliteal artery, with the medial head of the gastrocnemius (MHG) arising much higher than normal and the popliteal artery does not pass through the popliteal fossa.[56] While the sciatic artery and femoral artery converge to become the popliteal artery within the popliteal fossa and the sciatic artery regresses and gives off the tibial arteries, the myofascial planes of the popliteal fossa develop in an improper relationship to each other.[57, 58]

This pathologic process leads to extrinsic vessel compression and can induce progressive injury to the popliteal vessels leading to thrombosis, embolization, or aneurysmal degeneration.[58]

More specifically, PES can either be a congenital abnormality or a functional abnormality.

Congenital/anatomical PES is broken down into five different abnormalities.[57]

- type I: Aa aberrant medial course of the popliteal artery around a normally positioned MHG;
- type II: MHG attaches abnormally and more laterally on the femur causing the popliteal artery to pass medially and inferiorly;
- type III: abnormal fibrous band or accessory muscle arising from the medial or lateral condyle encircling the popliteal artery;
- type IV: popliteal artery lying in its primitive deep or axial position within the fossa, becoming compromised by the popliteus muscle or fibrous bands;
- type V: the entrapment of both the popliteal artery and vein due to any of the causes mentioned above.

Functional PES has no anatomic abnormality that leads to symptoms:[57]

- type VI: the muscular hypertrophy, resulting in a functional compression of both the popliteal artery and vein.

There exists another classification scheme known as the Heidelberg Classification which simplifies the pathology into three main categories:[59]

- type 1: the problem lies in the course of the popliteal artery;
- type 2: the muscular insertion is atypical;
- type 3: both of the abovementioned conditions are present.

6.2. Diagnosis

Clinical suspicion is paramount in the diagnosis of this rare pathology. Understanding the epidemiology will alert the practicing clinician to the possibility of the diagnosis which

should then lead to a thorough history and physical examination. Key epidemiologic facts that clinicians should think about are that approximately 85% of affected individuals are males with no significant cardiac comorbidities in addition to the onset of symptoms arising in the second or third decade of life in the majority of cases.[57, 60, 61] Furthermore, the syndrome is typically seen in athletic individuals, with approximately 30% of patients presenting with bilateral complaints.[57, 60, 61]

Presenting symptoms that should steer the evaluating practitioner towards this diagnosis include leg and foot exertional pain that can be quite severe and disabling and are relieved by rest. As the entire neurovascular bundle can be compromised, the patient might likewise notice lower extremity numbness, swelling, paresthesia, discoloration, pallor, and coolness depending on the affected structures.[57, 62-64] Moreover, the physical examination usually reveals hypertrophy of the calf muscles. Provocative maneuvers during physical exam such as dorsiflexion/plantar flexion of the foot may elicit diminished, unequal, or even absent pulses, and as the disease progresses, the ABI will also decrease.[58, 65]

Once clinical suspicion is raised, imaging is the next step in the workup of this patient population. Imaging can be difficult given the lack of true pathology other than aberrancy of muscle groups and trajectory of vascular structures. As such, magnetic resonance imaging (MRI) has been touted as one of the mainstays of imaging, however it should be combined with a flow-based assessment using duplex.[57, 58, 66] Functional testing has also been described with active and passive movements of the foot during imaging, whether it be non-invasive studies such as duplex or during active angiography.[57] Imaging signs to be looking for include medial deviation of the popliteal artery, popliteal artery stenosis or occlusion, or possible post-stenotic dilatation of the popliteal artery.[57]

6.3. Management and follow-up

Symptomatic PES typically requires surgical intervention. The types I-V more commonly require open surgery as the artery is extrinsically compressed and requires muscle/tendon release (if popliteal artery is normal) and/ or bypass (if popliteal artery is compromised). In type VI, or functional PES (FPES), the need for treatment depends upon the presence of symptoms. Asymptomatic type VI is usually observed. The clinical presentation and severity will influence the procedure as earlier intervention may simply require a release of the muscle while chronicity and damage to that segment of the artery, *i.e.*, fibrosis, stenosis, or thrombosis, requires reconstruction. In Type 5, involvement of the vein may warrant its decompression. Success using endovascular approaches is limited as this does not address the extrinsic compression. Treatment of FPES with ultrasound-guided Botox injection has been described, but further study is warranted to determine efficacy and durability.[67] This would not address any damage to the artery that may be present.

Imaging pre-procedurally may demonstrate arterial occlusion and the surgeon can be prepared for a reconstruction with either vein (short saphenous or greater saphenous) or prosthetic. One should be prepared to reconstruct as the damage to a patent artery may not be appreciated until this vessel is exposed and palpated. Use of intraoperative duplex US has been reported to help determine need for bypass.[68]

6.4. Surgical approach

The posterior approach is through an S-shaped configuration over the popliteal fossa. This approach has been described and includes vertical incision of the deep fascia with care to preserve the underlying medial sural nerve. Dissection is carried down directly over the popliteal artery.[69] The tibial nerve should be avoided which lies lateral to the artery and vein. The peroneal nerve should also be identified and carefully preserved. The heads of the gastrocnemius muscles are split to expose the distal aspect of the popliteal artery. Debulking of muscle or fibrous bands can then be performed and assessment of the artery with or without arterial reconstruction. Advantages of the posterior approach include better visualization of the muscle to be divided. Types III and IV can more easily be addressed this way dividing fibrous bands or the popliteus muscle (type IV).

A medial approach may be preferred depending upon the need for access to the more proximal and distal targets during reconstruction. A successful medial approach treating type I, II, or III entrapment has been described by dividing the medial head of gastrocnemius muscle, both muscular and tendinous portions, in addition to the division of any abnormal muscle slips, heads, and tendinous bands. In limbs with FPES, authors used a medial approach as well dividing the entire muscular portion of the medial head of the gastrocnemius muscle at the level of the tibial plateau, followed by the careful mobilization and lysis of the distal half of the popliteal artery. Only the tendon of the medial head of the gastrocnemius muscle was left intact.[70]

6.5. Outcomes

Successful outcomes of PES surgery have been described with shorter term results and minimal complica-

tions.[58, 64, 71] A larger study described the experience with PES in 88 limbs.[70] Of 18 limbs with occlusion of the popliteal artery, 15 underwent bypass grafting with reversed saphenous vein grafts, all of which remained patent during the follow-up period (median follow-up, 4.2 years; range, 1 to 10 years). One popliteal artery occlusion treated with thrombectomy and vein patching occluded within 6 months and necessitated subsequent vein grafting. Two limbs with inoperable occluded popliteal arteries were not subjected to reconstruction (one necessitated amputation because of advanced ischemia, and the second had extensive thrombosis of the distal run-off). In four limbs, moderate presenting symptoms abated without surgery after the discontinuation of an extreme exercise program. The remaining limbs underwent surgical decompression (all popliteal arteries remained patent, with a median follow-up of 3.9 years)

Popliteal vein entrapment surgical decompression has been described with good results.[72] Anomalies of the medial head of the gastrocnemius muscle caused entrapment in 60% of the patients; anatomic course venous anomalies were infrequent (7% of the patients). Significant relief of pain and swelling occurred in the patients who had surgery.

Functional popliteal artery entrapment syndrome surgical management was evaluated in 36 athletes who had a total of 56 limbs treated. Of the patients, 27 (75%) had bilateral symptoms and evidence of entrapment. Nine percent of limbs underwent a bypass along with debulking owing to arterial occlusion. Postoperatively, there were no nerve or vascular complications noted, two patients had wound/seroma complications (6%). At initial follow-up, all patients reported mild symptom improvement, but at midterm follow-up (mean follow-up time, 16 months), six (17%) reported mild to moderate recurrence of symptoms.[73]

Postoperative surveillance with color flow duplex ultrasound at 1, 3, 6, 12, and 18 months can be performed.[74]

6.6. Unresolved issues and goals for future research

Earlier detection with a heightened awareness can help avoiding misdiagnosis of PES.[60] Careful workup and understanding of the anatomic variants are important in obtaining good outcomes.

7. Thoracic outlet syndrome

7.1. Definition

Thoracic outlet syndrome (TOS) consists of a group of distinct pathologies and symptoms arising as a result of compression of structures at the thoracic outlet. TOS is recognized to encompass three conditions:

- neurogenic TOS (nTOS), caused by compression of the brachial plexus nerve roots within the scalene triangle, subcoracoid space, or both;
- venous TOS (vTOS), caused by compression of the subclavian vein and leading to the thrombosis;
- arterial TOS (aTOS), caused by compression of the subclavian artery and leading to arterial stenosis, aneurysm formation, and thromboembolism.

nTOS is the most common presentation in more than 95% of the cases.[75] Management is usually conservative, employing physiotherapy and postural exercises, however pain or muscle wasting may be indications for surgery, but it is out of the scope of this chapter.

7.2. Anatomy

Successful surgical treatment for all three types of TOS depends on a sound understanding of the relationships between musculoskeletal and neurovascular structures in the region of the thoracic outlet, as well as the many anatomic variations likely to be encountered. The scalene triangle is bounded by the anterior scalene muscle, the middle scalene muscle, and the first rib. The brachial plexus and subclavian artery pass through this space and over the first rib, whereas the subclavian vein passes over the first rib immediately in front of the scalene triangle. The costoclavicular space lies between the clavicle and the first rib and is bordered superiorly by the subclavius muscle, medially by the costoclavicular ligament, and posteriorly by the insertion of the anterior scalene muscle tendon on the first rib. The brachial plexus and subclavian artery pass over the first rib behind the costoclavicular space, whereas the subclavian vein passes over the first rib through the front part of the costoclavicular space. The subcoracoid space lies inferior to the clavicle and underneath the pectoralis minor muscle tendon, just below its insertion on the coracoid process. All of the structures of the neurovascular bundle pass through this space before reaching the axilla.

7.3. Pathophysiology and clinical presentation

In comparison to nTOS and vTOS, aTOS is the least common presentation of TOS, accounting for 1-2% of all cases.[76] During extreme shoulder abduction, the thoracic outlet is physiologically narrowed. As such, those who perform repetitive overhead arm movements such as swimming or throwing, are at risk of developing aTOS. Nearly half of patients presenting with aTOS have cervical ribs and around a third have soft tissue anomalies (Figure 7).[76]

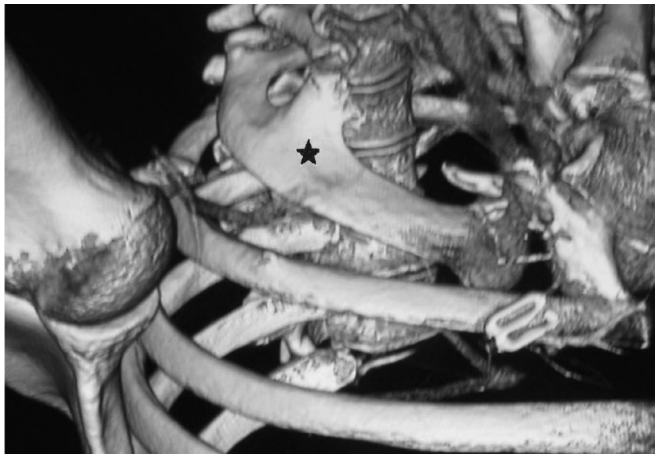

Figure 7.—Computed tomography scan with three-dimensional reconstruction demonstrates a right anomalous first rib (marked with asterisk).

The etiology is chronic repetitive injury to the subclavian artery as it exits the thoracic outlet. This injury may cause subclavian artery stenosis, intimal ulceration, poststenotic dilatation and true aneurysm. In asymptomatic patients a pulsatile mass or supraclavicular bruit can be detected on physical examination. Arm claudication is caused by areas of stenosis, which may be fixed, because of longstanding injury, or dynamic and occur with the arm abducted or extended only. Critical or acute limb ischemia is because of emboli of fibrin-enriched platelet aggregates that originate from an ulcerated mural thrombus in the aneurysmal segment.[75]

In vTOS most patients are sporting individuals, musicians or manual workers undertaking repetitive movements. Hypertrophy of the subclavius or scalenus anterior muscles, impingement at the costochondral junction by the costoclavicular ligament (particularly if the ligament inserts more laterally than normal on the first rib), or by calus formation from clavicule fracture, may cause extrinsic compression of the vein. Repetitive extrinsic compression during upper limb movement may eventually lead to fibrosis and scarring in the vein wall resulting in local thrombosis of the axillo-subclavian vein (Paget-Schroetter) syndrome.[77] Symptoms are precipitated by working with the arms elevated and are relieved by dependency, a pathognomonic feature of TOS. Acute presentation reveals an aching, swollen, tense, blueish-purple arm due to venous engorgement.

7.4. Diagnostic workup

Chest X-ray is used for the detection of bony abnormalities. DUS is very useful in detecting the occlusions or aneurysmal changes of subclavian artery in aTOS, and lack of flow on augmentation or clot in vTOS. Provocation manoeuvres may be very useful when both vein and artery compressed and incompletely blocked. CTA or MRA are the choices of investigation. Various image post processing (*e.g.* volume/surface rendering, maximal intensity projection) provide an unrivalled view of the vessels and the corresponding bony anatomy, excellent for planning surgery.

7.5. Management and follow-up

7.5.1. Arterial TOS

The main treatment goals of arterial TOS include a removal of the structures compressing subclavian artery, removal of the source of distal embolisation, and restoration of distal arterial perfusion.[78] All these procedures can be performed using a supraclavicular approach (Figure 8). The first step of this approach is the transverse skin incision two fingerbreadths above the clavicle, beginning at the lateral border of the sternocleidomastoid muscle. After mobilising the fat pad and phrenic nerve, anterior scalene muscle is divided and lifted superiorly to detach it from the additional structures underneath, including the pleural apex, the subclavian artery, and the brachial plexus nerve roots. The next step is a division of the middle scalene muscle. After complete scalenectomy, the procedure is continued with additional decompression that can include an excision and removal of the cervical rib, the first rib, or both of them. A removal of the cervical rib should be performed as close to cervical spine as possible. Supraclavic-

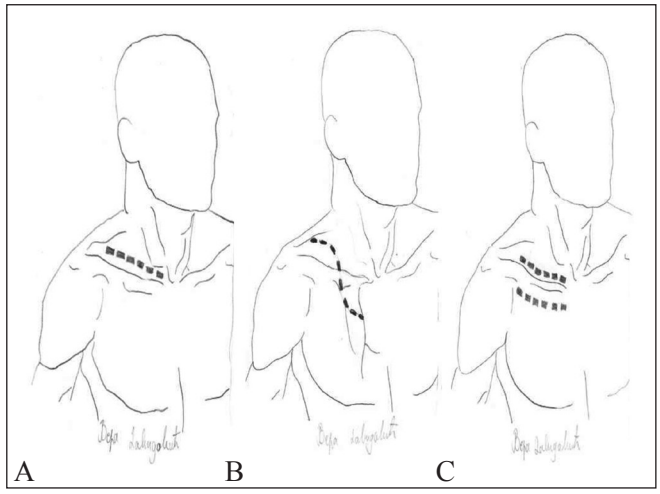

Figure 8.—A) Supraclavicular approach; B) supraclavicular approach with infraclavicular extension; C) combined supra- and infraclavicular approach.

ular approach enables also, a repair and replacement of the mid portion of the subclavian artery. Complete removal of the first rib requires further additional exposure. There are two options. The first one is obtained through sigmoid incision that has its supra- and infraclavicular segments (Figure 8). The clavicle should be left intact. However, in cases with more distal extension of SA caused by thoracic outlet compression, this sigmoid incision cannot provide complete vascular reconstruction. In such cases, additional exposure is obtained through separate, transverse infraclavicular incision (Figure 8). It provides excellent exposure to the proximal axillary artery where the distal anastomosis can be performed after SA excision, by using end-to-end interposition grafting technique.[79]

7.5.2. Venous TOS

Younger patients who do not want to accept the significant chance of a swollen limb interfering with activities should be offered intervention with thrombolysis and surgery.[80] Patients with late presentation, beyond a timeframe conductive to thrombolysis (around 14 days) should be treated initially with anticoagulation and reviewed at 3 months for evidence of dissolution of clot, at which time surgery may be offered if there is evidence of persistent vein impingement. Catheter-directed thrombolysis in acute presentation at this young age group is safe. When the clot is lysed, treatment with low molecular weight Heparin (LMWH) or direct oral anticoagulants (DOACs) is continued until surgery. The goals of surgery are to achieve complete decompression of the subclavian vein with subsequent definitive management of residual vein stenosis or occlusion. The surgical decompression can be undertaken *via* transaxillary approach, excising the first rib from beneath the vein.[81] Another option is to use the infraclavicular or combined supra- and infraclavicular approach to extend resection of the posterior part of the first rib (Figure 8). This combined approach may allow vein patch angioplasty or vein resection and graft interposition of a fibrosed, resistant stenosis within the subclavian vein; although this is rarely necessary. Balloon angioplasty and stenting is not advised in this region due to high incidence of rethrombosis.

7.6. Follow-up

The patient is seen usually one month after surgery for examination and duplex ultrasound imaging. The patient is reassessed at 6 months and re-evaluated at 1 year and yearly thereafter. Patients with aTOS who have undergone a vascular reconstruction, as well as decompression, warrant single antiplatelet therapy regardless of the conduit type used (vein or prosthetic). Patients with vTOS are maintained on oral anticoagulation for 3 months, and if they remain asymptomatic, anticoagulation may be safely discontinued.

7.7. Unresolved issues and goals for future research

It remains unknown whether patients presenting with the functional compression on subclavian artery, noted on CTA or conventional angiography, should be offered surgical decompression or close monitoring. While in the presence of arterial structural damage the indication for treatment is clear, the evidence regarding the surgical treatment of functional aTOS in asymptomatic patients is still very scarce, and there is no current consensus among experts.

As for vTOS the main question remain regarding the treatment indication. The evidence of whether to repair the scarred or compressed subclavian vein remains also vague. The interval of surgical decompression after thrombosis range from couple of days after thrombolysis up to several months after initial treatment.

8. Renal vein compression syndrome

8.1. Definition

Compression of the left renal vein (LRV) between the superior mesenteric artery (SMA) and the abdominal aorta is a relatively frequent incidental finding (nutcracker phenomenon) that may also cause symptoms of venous congestion of the left flank, kidney or pelvis (nutcracker syndrome). The condition is more common in females, and it can present at any age.

8.2. Mechanism of renal vein compression

The most typical mechanism is anterior compression of the LRV between the aorta and the SMA (Figure 9). In some cases, the third portion of the duodenum also lies between the aorta and the SMA, in front of the LRV. Therefore, anterior nutcracker may rarely occur with superior mesenteric artery syndrome (compression of the duodenum by the SMA, *i.e.* Wilkie's Syndrome). A retro-aortic or circum-aortic renal vein compressed between the aorta and the vertebral body can cause a posterior nutcracker syndrome. Obstruction to flow due to narrowing and stretching of the LRV over the aorta can also be caused by other mechanisms such as renal ptosis, an asthenic body habitus with minimal or no retroperitoneal fat, prominent lumbar lordosis or an abnormally high course of the LRV

or abnormal SMA branching. Other mechanisms may include excess fibrolymphatic tissue between the SMA and the aorta, compression from a testicular artery, para-aortic lymphadenopathy, pancreatic or retroperitoneal tumors or a gravid uterus.[83]

8.3. Clinical presentation

Asymptomatic Nutcracker phenomenon is frequent and in one review, >50% narrowing of the LRV was present in 51% to 72% of computed tomography (CT) angiograms.[84] The most frequent symptoms, when present, include abdominal and left flank pain (43.4-65.2%), macroscopic hematuria (39.1-69.5%), microscopic hematuria (8.6-21.7%), proteinuria (4.3-26.1%), and varicocele (8.7-21.7%).[85] Increased venous pressure leads to development of renal pelvic varices that rupture into the renal pelvic collecting system. Flank or abdominal pain can be exacerbated by positional changes (sitting, standing, travelling in a vehicle that shakes) with possible radiation to the posteromedial thigh and buttock. Symptoms of pelvic vein congestion are present due to high reflux in the left gonadal veins.[86] Left sided varicocele in males, pelvic congestion syndrome, dyspareunia, dysuria, dysmenorrhea, increased frequency of polycystic ovaries in females, orthostatic proteinuria, orthostatic intolerance, anemia, chronic fatigue in children and adolescents, other compressive syndromes like iliac vein compression (May-Thurner Syndrome) may also be present.

8.4. Diagnostic evaluation

Diagnosis is challenging and it is frequently a diagnosis of exclusion of all other possible diseases that cause hematuria, flank pain or chronic pelvic venous congestion. Physical examination and clinical history are critical. Eliciting a history of the onset of symptoms, precipitating, aggravating, and relieving factors, progression over time are important. The extent and nature of lifestyle limitations should be assessed. If hematuria is present, a complete urologic evaluation with urinalysis, urine culture, cytology, urethrocystoscopy, and in some patients renal biopsy is indicated to rule out other causes and localize the source to the left ureteric orifice.

8.4.1. Duplex ultrasonography

A renal venous duplex ultrasound is the first screening study we use for the diagnosis of left renal vein compression.[82, 87, 88] Duplex images are obtained both in supine and in the left lateral decubitus position (Figure 10).[89] Major ultrasound features include peak velocity ratio of ≥5 between the narrowed and distended portions of the left renal vein (PVN/D ratio), or an anteroposterior diameter ratio of ≥5 between the distended and narrowed portions of the left renal vein (APD/N diameter).[85] The sensitivity and specificity of Duplex ultrasound is reported at 78% and 100% respectively.[90, 91] However, there is wide variation, due to small study size as well as operator dependence and need of a dedicated ultrasonographer.[84]

8.4.2. Computed tomography venography

CT venography can demonstrate the nature and degree of LRV narrowing, extrinsic compression, retroperitoneal and renal hilar collaterals. It is essential to have a dedicated imaging protocol with timed pre-hydration to capture the venous phase with maximal distention of the LRV. Dehydration may lead to false positive compression of the LRV. The CT features of Nutcracker syndrome include a narrow aorto-mesenteric angle (6-16°, normal around 40°), narrowing of the LRV (beak sign, triangular shape of LRV

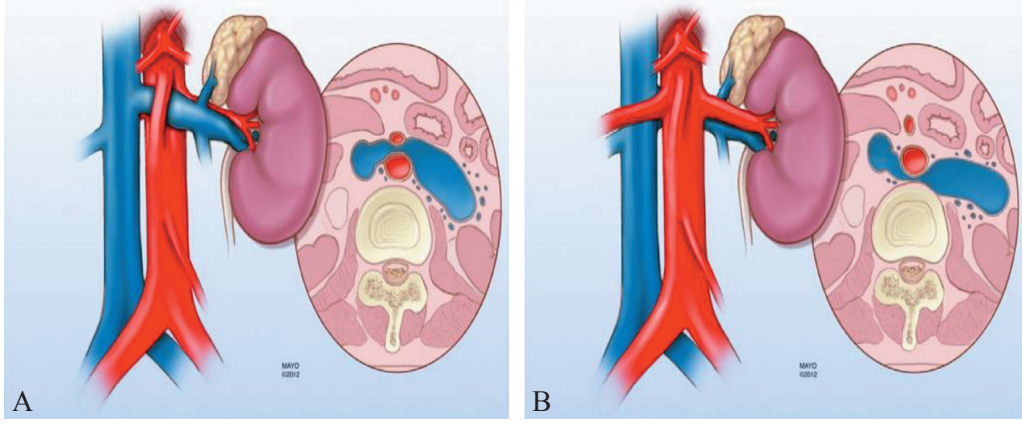

Figure 9.—Diagrammatic representation of the nutcracker syndrome (NS): A) anterior with extrinsic compression of the left renal vein (LRV) between the aorta and the superior mesenteric artery (SMA); B) exstrinsic compression of the left renal vein (LRV) between the aorta and the spine (from Said *et al*.).[82]

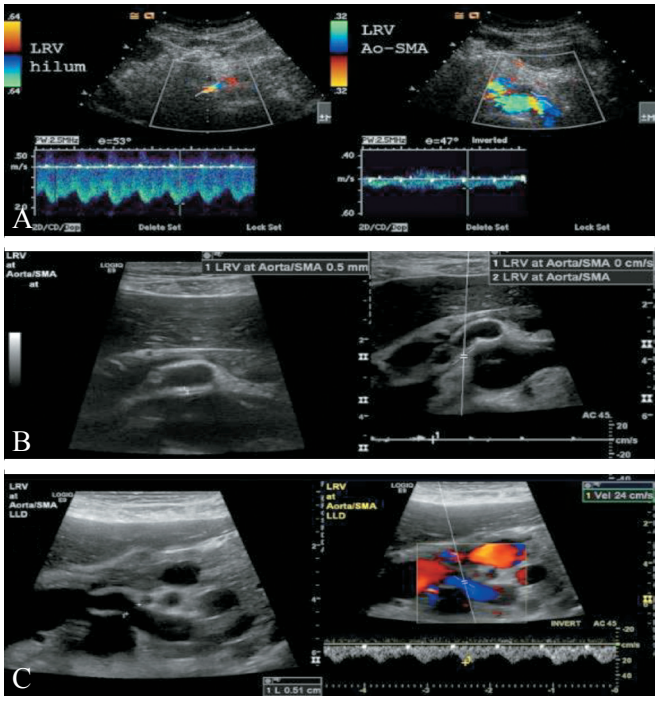

Figure 10.—A) Duplex ultrasound images showing the Peak systolic velocity at the hilar segment of the left renal vein (left) and in the aorto-mesenteric angle (right); PSV ratio 11.3; B) Duplex ultrasound images in supine position in a patient with suspected NCS showing significant compression of the LRV on B mode image (left) and no flow through the vein on color flow (right); C) Imaging in the same patient in left lateral decubitus (LLD) position showing a 10-fold increase in the diameter of the LRV (left) and normal flow and peak systolic velocity (right).

due to severe aortomesenteric narrowing, the beak angle <32-41°), a LRV diameter ratio (hilar to aorto-mesenteric) ≥5 and prominent collaterals in the retroperitoneum and renal hilum, distended left gonadal vein.[82, 92]

8.4.3. Contrast venography

Considered to be the gold standard for diagnosis. Unfortunately, there can be great variability with patient position, hydration and the degree of collateralization, and these can lead to both false positive and false negative results. Thus, treatment is not based on imaging parameters alone. The venographic criteria for Nutcracker syndrome include a venous pressure gradient of 2-14 mm Hg between the hilar LRV and the IVC (normal 0-1 mm Hg), >50% narrowing of the LRV, collaterals and gonadal venous reflux

8.4.4. Intravascular ultrasound

Intravascular ultrasound has not been validated for the diagnosis of NCS but can measure the degree of venous stenosis.[93]

8.5. Management of renal vein compression syndromes

The first therapeutic approach for the patient with nutcracker syndrome (NTS) should be conservative. In pediatric patients, this pathology usually resolves spontaneously without surgical treatment.[94] In adult patients, conservative approach consists of gaining weight and performing exercises for back muscles. There is evidence for the use of hemorheological drugs or antithrombotic therapy.

8.6. Endovascular approach

When conservative treatment has failed, we consider an invasive treatment. Endovascular first approach is not the gold standard of NTS. After the first period of enthusiasm related to the technique of stent implantation and ovarian vein embolization, serious complications started to occur.[95, 96]

There are several reasons why primary stenting should not be the first line approach for the treatment of NTS:

- there is no dedicated stent model developed for the purpose of NTS treatment;
- it is an area of great mobility, where an implant may have a risk of migration and serious consequences, ranging up to 7%;[95, 96]
- the LRV is conical in its shape. It measures between 8-10 mm near the renal hilum, and between 14-16 mm in the area of the inferior vena cava (IVC) confluence. As there are no conical shaped stents, an excessive stent oversizing can expand the vein at the level of hilum, causing chronic pain;[97]
- the LRV has inverted C shape and a thin wall. A straight metallic stent can cause chronic perforation and associated pain if its end remains in the region where vein passes over the aorta and changes direction towards the posterior and caudal aspect;[97]
- the LRV follows an oblique and ascending course from kidney to IVC. The implantation of stent straightens the course of the vein and can cause a new functional stenosis. This is because the vein goes vertically towards renal hilum at the level of stent distal insertion. This kinking is known as the curtain effect and could cause poor kidney drainage.[97]

The question remains in what circumstances we should perform LRV stenting in the presence of NTS. Endovascular treatment in these cases is currently restricted only to cases of a very hostile abdomen, or as a rescue option of surgical complications (Figure 11). There may be patients who reject open redo procedure. In these cases, the vein no longer follows a horizontal or ascending course but its angle is surgically modified and the risk of stent

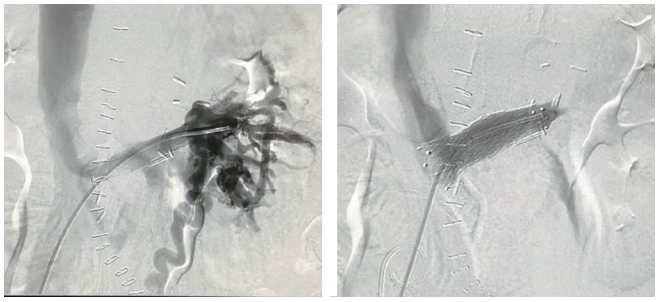

Figure 11.—Endovascular salvage after failed open surgery.

migration is almost impossible. However, chronic pain may appear due to the impact of the end of the stent on the curve of the vein before its angulation towards the renal hilum.

In posterior NTS vein has downward course, making stent migration almost impossible.[98] But the pressure between the aorta and vertebra can cause stent fracture on the long-term scale. Even so, this would be a possible indication because in case of failure, open surgery could always be performed by renocaval bypass.

8.7. Surgical treatment of nutcracker syndrome

Classic open surgery is complex and invasive, but still nowadays is the treatment of choice. Both a right subcostal incision extending to the left side and midline laparotomy are used as choices of surgical approach. Different interventions[82, 87, 99, 100] have been described for the treatment of NTS, but most commonly used techniques are:

- renocaval prosthetic bypass;
- vein derivation procedures:
  - transposition of the LRV to the IVC;
  - transposition of the gonadal vein to the IVC;
  - transposition of the distal left gonadal vein to the left common iliac vein;
  - bypass from LRV to IVC with reversed saphenous vein;
- renal autotransplantation;
- external stent (shield technique);
- radical nephrectomy.

One of the most commonly used open surgical techniques is the renocaval prosthetic bypass (Figure 12). One very important surgical detail to take into account is that the prosthesis should be completely covered with a layer of omentum, thus preventing the occurrence of enteroprosthetic fistula.

LRV transposition has been considered previously as the first line treatment option. However, it is associated with the high rates of restenosis. In order to avoid this hybrid technique has been developed with simultaneous stent implantation.

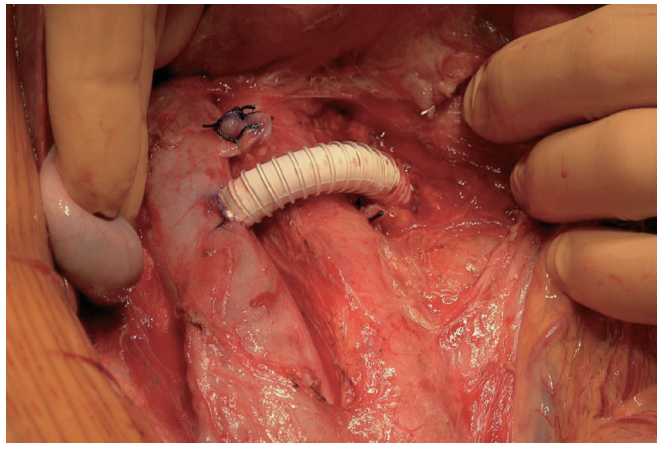

Figure 12.—Renocaval bypass using prosthetic polytetrafluoroethylene (PTFE) graft.

8.8. Follow-up

In general, concern about a possible thrombosis may prompt intense anticoagulation therapy, and in the first 24 hours the most important postoperative complication is hemorrhage, rather than thrombosis. After the initial postoperative period patients are usually anticoagulated for 6 months using direct oral anticoagulants. Patients are scheduled for follow-up visit at 3 and 6 months and then annually after. Doppler ultrasound is used for follow-up imaging control.

8.9. Unresolved issues and goals for future research

There are no dedicated stents for the treatment of NTS. Having this in mind, stents should be used in cases after failed surgery, in Posterior NTS or when facing hostile abdomen. Taking in mind the complexity of open repair, patients should be transferred to the reference center.

9. Iliac vein compresion syndrome

9.1. Definition

Extrinsic compression of iliac vein by the arterial system against bony structures in iliocaval area is commonly known as May-Thurner Syndrome (MTS). The most frequent condition for anatomic reasons is the left common iliac vein compression (LCIV) between the spine and the right common iliac artery. This condition may represent a

cause of obstructive venous disease due to increased pressure in proximal venous system and unfortunately is rarely considered and diagnosed. Evolution of chronic hypertension may lead to severe venous disorders or deep vein thrombosis (DVT). The exact incidence and prevalence of MTS remains unknown and in fact underestimated since most of the cases are asymptomatic and require no treatment. May-Thurner syndrome turned out to be the etiology in 2-5% of patients who came up with symptomatic lower extremity venous disorder.

9.2. Pathophysiology

Venous compression can be permissive and asymptomatic, but three factors may lead development of symptoms:

- chronic inflammation and fibrotic response due to persistent extrinsic trauma may cause the formation of spurs, webs, channels and diaphragms inside the vessel;
- external inflammation may lead to increased vessel rigidity;
- flow alteration secondary to lower limb reflux and loss of volume during backflow to the heart may lead to vein collapse and gradually to thrombosis.

Iliac vein entrapment syndrome can occur due to compression in several anatomic points. Currently, the following patterns of compression are recognized:

- left common iliac vein (LCIV) compressed by right common iliac artery (RCIA);
- right common iliac vein (RCIV) compressed by right common iliac artery (RCIA); left common iliac artery (LCIA); right internal iliac artery (RIIA); right external iliac artery (REIA);
- left external iliac vein (LEIV) compressed by LEIA;
- right external iliac vein compressed by REIA;
- RCIV compressed by aortic bifurcation;
- inferior vena cava compressed by RCIA.

Patients with MTS can present extreme heterogeneity of symptoms, ranging from leg swelling with persistent oedema to venous claudication, symptomatic varicose veins or phlebitis. Most rarely, in case of primary DVT presentation, phlegmasia cerulea dolens can occur.

All patients with left sided chronic venous disorders need to be investigated for iliac vein entrapment syndrome, particularly those with history of isolated or recurrent DVT. All causes of extrinsic compression need to be excluded during differential diagnosis, including pelvic masses, iliac artery aneurysms, bladder distension, spondylolisthesis or lumbar discopathy, lymphadenopathy and tumors.

While the exact natural history of MTS is not clear, some authors believe it evolves in three clinical stages:[101]

- stage 1: asymptomatic LCIV compression;
- stage 2: formation of intraluminal spurs;
- stage 3: occurrence of left iliac deep vein thrombosis.

9.3. Diagnostic evaluation

Persistent narrowing of vein compressed with stenosis more than 50% is considered as adequate suspect of MTS; other indicators are represented by venous collaterals, identification of intraluminal changes, diameter of proximal vessels and flow patterns.

9.3.1. Duplex ultrasound (DUS)

Although it provides high resolution, sensitivity and specificity, the abdominal ultrasound scan may present some limitations in compressive syndromes. The deep location of veins and standard prone position of patients can limit the correct evaluation and may lead to the over diagnosis of compressions. To overcome this limitation, this examination needs to be performed in supine and semi-sitting position. Following DUS in standing position focused on reflux, the patient is invited to lay in supine position and femoral vein evaluation is performed using a 4-7 MHz linear array transducer. In non-occlusive conditions such as MTS, flow patterns can be absolutely normal with regular phasicity of common femoral vein, but other cases can reveal reduced phasicity of flow, reduction in amplitude and limited response to compressive or Valsalva maneuver. Iliac and caval vessels need to be investigated with 2-3 MHz probe in B mode in order to directly evaluate diameters and morphologic compression. Peak vein velocity (PVV) is measured in the post-stenotic and compared to the pre-stenotic segment, which is often dilated. If the PVV gradient is more than 2.5, the findings are significant. Reflux on ipsilateral internal iliac vein is associated with proximal compression due to compensatory reversal flow.

9.3.2. Plethysmography

It is a volumetric tool to evaluate venous reflux and proximal obstruction. Rapid filling and low elevation drainage are indicative of global venous incompetence and obstruction, respectively. This technique can be used to assess severity of venous symptoms, particularly in severe reflux, but is not currently recognized as a diagnostic tool for MTS.

9.3.3. Computerized tomography/magnetic resonance (CT/MR) venography

MR venography and CT venography are second level diagnostic tools in case of suspicion of iliac vein entrap-

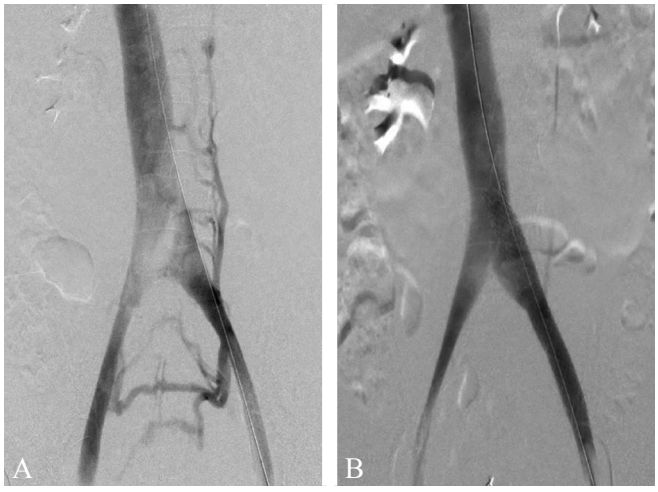

Figure 13.—Intraoperative phlebography before (A) and after the stenting of left common et external iliac vein (B).

ment syndrome, and provide a complete examination of the deep venous system and surrounding structures, that can be enhanced by 3-D reconstruction. Magnetic resonance venography is the most versatile imaging, since its dynamic sequences can provide information regarding velocity and volume while providing high-quality morphologic images of the compressed vein, such as fibrotic scarring, post-thrombotic fibrosis as well as collaterals and varicose veins. Computed tomography has some disadvantages related to radiation exposure and iodinate contrast but it is well recognized that provides high quality studies and complete evaluation of the venous anatomy.

9.3.4. Venography

Nowadays it is often used in combination with devices such as intravascular ultrasound, allowing for simultaneous diagnosis and intervention. When used in iliac vein entrapment syndromes, phlebography can provide information about the patency of vessels, anatomical variations and associated reflux. It is often possible to observe the classical arterial "shadow" associated with the "pancake effect" typically present in anteroposterior view and secondary to vein enlargement below the compression (Figure 13A). Furthermore, phlebography can show hypertrophic collateral circulation, in particular internal iliac vein branches and ascending lumbar vein.

9.3.5. Intravascular ultrasound

Performed immediately after venography, IVUS is recognized as the method of choice for the diagnosis of MTS, helping to identify the compressed area and intraluminal changes of diseased vessel. IVUS can provide information about vein diameter and distance between the two healthy segments that need to be identified as landing zones for stenting. After deployment, it's mandatory to perform a final control to ensure stent expansion and correct resolution of compression disease.

9.4. Treatment

Treatment of asymptomatic patients, even with high grade stenosis or occlusion, is not supported by any robust evidence that suggests this reduces the risk of subsequent DVT. In case of C3-C6 CEAP classification classes, endovascular interventions by means of venoplasty and stenting need to be considered. Furthermore, venous claudication (heaviness and pain during exercise) with persistent edema represent a possible indication in young patients if associated to debilitating quality of life.

Conservative measures, such as compression stockings and anticoagulation therapy, are the first-line treatment, followed by endovascular approach which is focused to restore patency of compressed/occluded vessel by endoluminal stent implantation. Several devices are available on the market with different features. The goal of stenting MTS is to solve compression achieving correct venous return (Figure 13B).

It becomes crucial to deploy stent with "high radial force" to resist to focal load of extrinsic compression but simultaneously with good "flexibility" to accommodate in the venous "curves" anchoring the proximal side to external iliac vein. According to common practice, venous stents need to be positioned under IVUS-guidance, therefore avoiding excessive jailing in the IVC confluence that may lead to contralateral iliac DVT. Referral center are provided by different stent configurations and physician can adequately decide which stent to implant regarding the IVC confluence anatomy. Several trials are ongoing with satisfactory results for MTS treatment and long-term results with secondary patency near 100%. No difference between dedicated venous stents and non-dedicated venous stent was reported in current literature.

One of the most crucial and debated issue is stent migration. This event may occur in case of undersizing or in case of positioning of short devices. IVUS guidance led to right choice of sizing and length and allows for correct identification of landing zones in external iliac vein and in IVC confluence.

After treatment, immediate anticoagulation with low-molecular weight heparin is started and intermittent pneumatic compression is administered for the first 12 hours,

followed by active mobilization thereafter. There is significant discussion on the best post-procedural pharmacological strategies to employ. In 2018, the Imperial College of London conducted an International Delphi Consensus, enrolling 106 experts from 78 venous stenting centres in 28 countries, and aiming to investigate antithrombotic practices after venous stent.[102] It was consensual that LMWH is the anticoagulant of choice in the first 2-6 weeks after venous stenting, and after this period, oral anticoagulation should be initiated, with most studies using vitamin K antagonist as the ideal anticoagulation regimen. The duration of the oral anticoagulation depends on the clinical setting, so that:

• following iliac vein stenting due to non-thrombotic compressive iliac vein lesions, anticoagulation should be instituted in the first 6-12 months, and can be discontinued after this period if the appearance on ultrasound is satisfactory;

• following DVT thrombolysis and iliac vein stenting, anticoagulation can be discontinued between 6-12 months if thrombophilia screen is negative and it is the first DVT and US appearances of the stent are satisfactory;

• in patients with multiple DVT's and iliac vein stenting, anticoagulation should be lifelong.

Finally, the role of antiplatelet therapy after venous stenting is unclear, and the available evidence is conflicting. Although antiplatelets have been shown to be beneficial in preventing restenosis of arterial stents, these effects cannot necessarily be extrapolated to venous stents. As such, no consensus among experts was obtained.

9.5. Follow-up and re-intervention

Regular follow-up with imaging studies is essential in the management of MTS, regardless of the type of intervention used, and DUS is usually recommended as the first line treatment option given its non-invasive nature. However, there are no standardized ultrasound follow-up surveillance guidelines for iliac vein stenting, with literature mostly reporting institutional protocols, that often differ among each other.[103-105]

The dificulties that surround DUS-based follow-up strategies after venous stenting extend beyound the timings, since there are no established ultrasound surveillance criteria to predict impeding stent failure in the venous system. As such, while the presence of thrombus, loss of phasicity and lack of augmentation with provocative maneuvers are often seen as predictors of failure, CT or MRI are often necessary to better caractherize patency and stent behaviour in such settings.

Finally, there are no universally acepted criteria for re-intervention. Most high-volume centers opt for a symptom-based re-intervention strategy, championing the idea that re-intervention should occur when patients become symptomatic, even without obvious diagnostic ultrasound changes.[106] Although intuitive, this strategy requires strict clinical follow-up, as well as patient education towards its condition and alarming signs, which is sometimes difficult in certain settings. For now, most centers provide re-intervention in a case-by-case basis, taking into account the individual patient's symptoms, comorbidities, and risk factors.

9.6. Unresolved issues and goals for future research

Despite the availability of various treatment options for MTS, there are still unresolved issues and controversies in the management of this condition. The following are some of the areas of uncertainty and goals for future research in MTS.

• optimal timing and duration of anticoagulation therapy;

• optimal imaging modality for surveillance: while DUS is the recommended imaging modality for surveillance of MTS, there is a need to investigate the role of CT and MRI in the detection and management of restenosis and complications;

• optimal criteria for re-intervention: the criteria for re-intervention in MTS are not universally accepted, and there is a need for further research to establish evidence-based guidelines for the management of restenosis and complications;

• long-term outcomes of endovascular intervention: while endovascular intervention is the preferred treatment option for MTS, data on the long-term patency and structural behavior of stents is still lacking.

10. Vascular compression syndromes of the neck

10.1. Definition and pathophysiology

Arterial compression of the neck is a rare clinical picture, whereas venous entrapments are more frequent. The classic Eagle syndrome is characterized by an elongated styloid process compressing the surrounding nerves which causes pain, dysphagia and otalgia, often exacerbated by yawning and swallowing. Depending on the angle variation of the styloid elongation the tip of the styloid may impact on the internal carotid artery. The eventual penetration into the wall may cause transient ischemic attacks and\or stroke.[107]

10.2. Pathophysiology

The internal jugular vein (IJV) is the main bilateral pathway of the cerebral venous system (CVS), which functions are mainly represented by blood and catabolites drainage from the brain, refilling of the right atrium, and maintenance of thermic homeostasis. IJV compression alters of course the function above and creates a condition of chronic cerebral venous insufficiency. According to the recent theory of the glymphatic system (the peculiar lymphatic system of the brain), the interstitial space full of solutes and waste products is cleaned by a flow converging into the paravenous cerebro-spinal fluid (CSF) space to be discharged. Fluid exchanges between venular lumen and paravascular space depend primarily on transmural pressure (TMP).[108] TMP can be influenced by IJV compression according to Poiseuille's law ($Q=\Delta P/R$, where Q is the flow, ΔP is the pressure gradient, and R is the resistance to flow). Indeed, the increased R of IJV flow due to obstructions increases the TMP. In addition, IJV compression modifies the physiological drop of the venous pressure between cortical veins and dural sinuses. This fundamental mechanism allows the CSF to leave the craniospinal space with the same speed with which it is produced.[108]

Based on such rationale, several neurologic diseases such as idiopathic intracranial hypertension (IIH), inner ear functions, headache, multiple sclerosis (MS), Parkinson and Alzheimer's disease, have already been reported to be associated with internal jugular vein (IJV) stenosis.[109-113] Indeed, a CVS obstruction at any level would lead to an increased TMP, a decreased paravascular interstitial fluid flushing and an increased collection of solutes and waste products in the interstitial space. These products could increase the neuroinflammation and lead a different kind of nervous structures damages.

10.2.1. C1-styloid jugular nutcracker (C1JN)

Recently, one more vascular Eagle variant has been described. In this case, the IJV is compressed by a bone nutcracker between the styloid process and the arch atlas (Figure 14A).[107] Often the styloid process is elongated, but in some cases, it can present normal dimensions with a different angle (Figure 14B). Clinical presentation is characterized by headache, tinnitus, insomnia, visual disturbances, hearing impairment.[107]

10.2.2. C1 jugular nutcracker and perimesencephalic subarachnoid hemorrhage

This is a particular kind of cerebral hemorrhage not correlated to a clear intracranial vascular malformation. The etiopathogenesis is still discussed, but most of the possible causes are related to CVS hypertension. Scerrati *et al.* showed a possible association of this hemorrhage to the presence of C1JN, compressing the IJV at the passage through the styloid process and the transverse process of C1. Again, an extracranial CVS hampered outflow would increase venular pressure, thus predisposing wall rupture and bleeding.[114]

10.2.3. Omohyoid muscular entrapment (OME)

Idiopathic intracranial pressure is usually associated to the female sex and an elevated body mass index. Cerebral ventricles dimensions are usually normal or decreased. In 2019 De Bonis *et al.* described the JEDI (jugular entrapment, dilated ventricles, intracranial hypertension) syndrome in a woman suffering from bad headaches, visual loss (Frisen grade 4 papilledema) and pulsating tinnitus. In this case, ventricles were dilated and a FDG PET showed a diffuse hypometabolic cerebral state. The patient also suffered from a bilateral jugular vein entrapment between the upper and the lower bell of the omohyoid muscles (OME) (Figure 14C). The compression of the IJV determined a complete absence of IJV flow with scarce collateral circulation. Surgery was performed with bilateral section of omohyoid muscles. Headache and tinnitus disappeared after surgery and papilledema progressively improved with visual acuity restoration and hydrocephalus regression.[115] Finally, mono-lateral OME can be associated with IJV intraluminal obstacles such as the presence of malformed valves or septa, frequently seen in multiple sclerosis patients. In a multicenter randomized double blinded trial, 19% of patients showed at venography OME, on which balloon angioplasty was ineffective.[113] Quite the contrary, open omohyoid resection fixed the brain outflow with consistent and significant improvement either of cerebral ventricles volume or brain perfusion.[116]

10.3. Diagnosis

A jugular stenosis is defined, according to Jayaraman, as a caliber reduction >80% on axial cuts compared with the normal vein proximal to the stenosis. Common radiological findings include the failure of vein appearance due to the occlusion, venous congestion with the enlargement of typically diminutive veins from collateral drainage and reversal of venous flow.[117]

Color Doppler ultrasound (US) is mandatory to evaluate both in the transverse and longitudinal plane the IJV morphology and hemodynamics. Particularly, the detection of blocked IJV flow at J3 segment, just above the carotid bi-

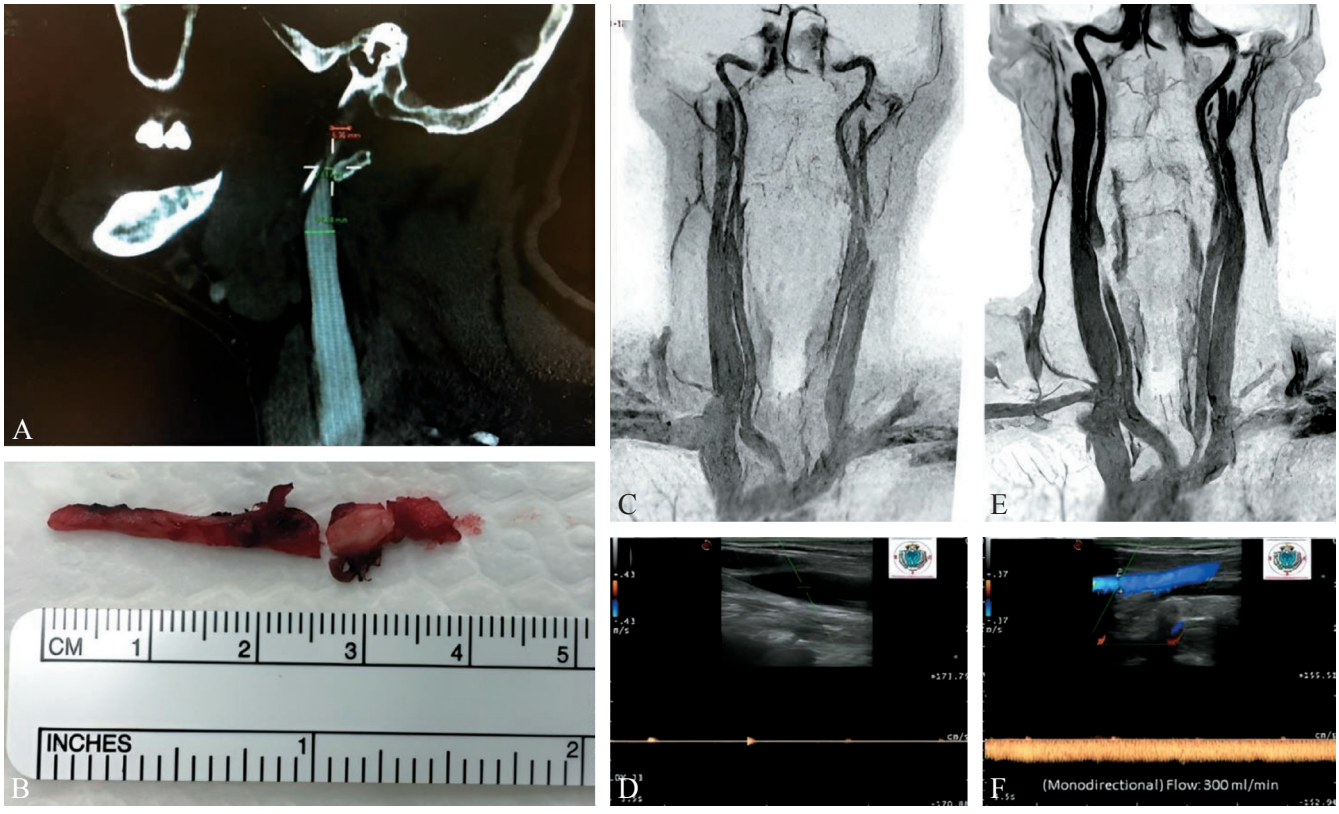

Figure 14.—A) CTA reconstruction of C1-styloid nutcracker of the IJV; B) specimen of an elongated styloid process (normal length is less than 2 cm), removed to treat an Eagle vascular syndrome; C) MRV clearly showing the disappearance of the J3-J2 segments of the left IJV due to omohyoid muscle entrapment; D) the Duplex ultrasound of the same case showing the IJV obstruction with above vein dilation and absence of Doppler flow; E) MRV at 1-year follow-up of the same case with restoration of the IJV continuity; F) Duplex ultrasound of the same case showing the flow restoration in the IJV.
CTA: computerized tomography angiography; IJV: internal jugular vein; MRV: magnetic resonance venography.

furcation, could be the consequence of C1JN and needs to be further investigated.[107, 108, 114] Moreover, just below J2 the OME can be detected and considered significant in case of IJV absence of flow (Figure 14D, F).

Cervical plethysmography on a tilt chair allows to assess either the jugular flow rate and the flow resistance, respectively decreased and increased in case of significant IJV compression.[118]

CT or MRI (TOF sequences) venography are useful for the classification of the severity and the anatomical location of the stenosis (Figure 14A, C).[107, 109, 110, 112]

10.4. Management and follow-up

First line treatment is the use of prophylactic anticoagulants/antiplatelet drugs to prevent thrombus formation. Styloidectomy is the most frequently performed surgical procedure for the carotid variant of the Eagle syndrome, with eventual repair of the artery with different techniques (Figure 14B). Surgery (styloidectomy or C1 anterior arch removal) is used for C1JN, while open muscular section is currently the best option for OME. Both procedures are proposed to patients with severe persistent symptoms, especially related to the possible IIH, where also jugular stenting has been proposed.[111]

Follow-up is based on clinical, ultrasound and radiological monitoring.

10.5. Unresolved issues and goals for future research

IJV obstruction has been associated to several neurologic diseases. However, the exact pathogenesis is still not completely clear. Although the discovery of the glymphatic system permits to explain several issues, still solid basis is lacking. For instance, Zhou *et al.*[112] showed the IJV obstruction on behalf of a kinking of the innominate artery linked with several neuroimaging and clinical abnormalities. Further studies are required. Finally, from a technical

point of view, OME treatment could benefit of the development of innovative venous stents.

11. Final remarks and conclusions

Vascular compression syndromes (VCSs) are rare and little-known entities that represent a very important subset of vascular pathology.

The recognition of VCSs is difficult when they are still asymptomatic due to the non-specificity of their symptoms. They are often accidentally identified during routine clinical examination. Only when the compression is severe and patient becomes symptomatic it is possible to identify important characteristic and signs that help clinicans in making a differential diagnosis.

The diagnosis may be difficult due to clinicians' lack of knowledge and the rarity of this condition. Knowledge of these signs and typical symptoms is essential for their recognition. When patients have minimal asymptomatic vascular compressions, in most cases the diagnosis of VCS is overlooked, exposing them to serious risks of potential complications such as thrombosis, pulmonary embolism, cerebral embolism, etc.

Recognizing VCS promptly and evaluating its severity is essential for proper therapeutic planning. DU is the first-line examination for the diagnosis of VCSs. It enables clinicians to highlight the morphological changes and above all to measure the degree of vascular stenosis.

In some cases, ultrasonography is not sufficent, and to make a definite diagnosis a second-line diagnostic modality is necessary to identify the type of compression and highlight the complications, as well as to exclude other causes of patient signs and symptoms.

IVUS, which has recently become the reference diagnostic method for arterial and venous pathologies, allows direct evaluation of endoluminal content.

Digital subtraction angiography has been the mainstay of imaging-based diagnosis for most vascular compression syndromes, but other methods such as CTA, and MRA are used with increasing frequency for initial diagnostic evaluation.

Because vascular compression syndromes are caused by the external compression of vessels, endoluminal treatment alone is rarely adequate and surgical decompression is likely to be required for optimal and durable clinical benefit.

References

1. Farina R, Foti PV, Pennisi I, Vasile T, Clemenza M, Rosa G, *et al.* Vascular compression syndromes: a pictorial review. Ultrasonography 2022;41:444–61.

2. Farina R, Foti PV, Conti A, Iannace FA, Pennisi I, Fanzone L, *et al.* The role of ultrasound imaging in vascular compression syndromes. Ultrasound J 2021;13:4.

3. Rangel Villalobos E, Busquier Cerdán T, Cortés Sañudo X, Avilés Vázquez I, Estellés López R, Pérez Ramírez C. Vascular compression syndromes: the value of Doppler ultrasonography. Radiologia (Engl Ed) 2022;64:17–25. [Engl Ed]

4. Gozzo C, Giambelluca D, Cannella R, Caruana G, Jukna A, Picone D, *et al.* CT imaging findings of abdominopelvic vascular compression syndromes: what the radiologist needs to know. Insights Imaging 2020;11:48.

5. Zucker EJ, Ganguli S, Ghoshhajra BB, Gupta R, Prabhakar AM, Picone D, *et al.* Imaging of venous compression syndromes. Cardiovasc Diagn Ther 2016;6:519–32.

6. Czihal M, Banafsche R, Hoffmann U, Koeppel T. Vascular compression syndromes. Vasa 2015;44:419–34.

7. Noorani A, Walsh SR, Cooper DG, Varty K. Entrapment syndromes. Eur J Vasc Endovasc Surg 2009;37:213–20.

8. Gupta PC, Atturu G. A Multidisciplinary Approach to Neurogenic Thoracic Outlet Syndrome: Sinking Deeper or Stepping Out of a Chaotic Quagmire Onto Terra Firma? Eur J Vasc Endovasc Surg 2021;61:1025.

9. Balderman J, Holzem K, Field BJ, Bottros MM, Abuirqeba AA, Vemuri C, *et al.* Associations between clinical diagnostic criteria and pretreatment patient-reported outcomes measures in a prospective observational cohort of patients with neurogenic thoracic outlet syndrome. J Vasc Surg 2017;66:533–544.e2.

10. Miller TL, Backs R, Vaccaro PS. Popliteal Artery Entrapment Syndrome: A Diagnostic and Treatment Enigma for Orthopaedic Surgeons. J Am Acad Orthop Surg 2021;29:e834–45.

11. Panther EJ, Reintgen CD, Cueto RJ, Hao KA, Chim H, King JJ. Thoracic outlet syndrome: a review. J Shoulder Elbow Surg 2022;31:e545–61.

12. Andò G, Trio O, Manganaro R, Manganaro A. To promote endothelial function: the elusive link between physical therapy of venous thromboembolism and improved outcomes? Int J Cardiol 2016;214:31–2.

13. Bashar K, Shalan A, Sharafat Ali S, Tang T, Tiwari A. Endovascular versus medical treatment of venous compression syndrome of the iliac vein - a systematic review. Vasa 2021;50:22–9.

14. Senechal Q, Echegut P, Bravetti M, Florin M, Jarboui L, Bouaboua M, *et al.* Endovascular Treatment of Pelvic Congestion Syndrome: Visual Analog Scale Follow-Up. Front Cardiovasc Med 2021;8:751178.

15. Scultetus AH, Villavicencio JL, Gillespie DL. The nutcracker syndrome: its role in the pelvic venous disorders. J Vasc Surg 2001;34:812–9.

16. Kim SY, Min SK, Ahn S, Min SI, Ha J, Kim SJ. Long-term outcomes after revascularization for advanced popliteal artery entrapment syndrome with segmental arterial occlusion. J Vasc Surg 2012;55:90–7.

17. Meier TO, Schneider E, Amann-Vesti B. Long-term follow-up of patients with popliteal artery entrapment syndrome treated by endoluminal revascularization. Vasa 2010;39:189–95.

18. Yamamoto S, Hoshina K, Hosaka A, Shigematsu K, Watanabe T. Long-term outcomes of surgical treatment in patients with popliteal artery entrapment syndrome. Vascular 2015;23:449–54.

19. Kosasih S, Moore H, Lane TR, Davies AH. Deep Venous Reconstruction: A Case Series. Cureus 2017;9:e1518.

20. Gharagozloo F, Meyer M, Tempesta B, Gruessner S. Robotic transthoracic first-rib resection for Paget-Schroetter syndrome. Eur J Cardiothorac Surg 2019;55:434–9.

21. Schrufer-Poland TL, Florio K, Grodzinsky A, Borsa JJ, Schmidt L. Management of May Thurner Syndrome in Pregnant Patients. J Cardiovasc Dev Dis 2022;9:410.

22. Speranza G, Sadek M, Jacobowitz G. Common iliac vein stenting for May-Thurner syndrome and subsequent pregnancy. J Vasc Surg Venous Lymphat Disord 2022;10:348–52.

23. Thiyagarajah K, Ellingwood L, Endres K, Hegazi A, Radford J, Iansavitchene A, *et al.* Post-thrombotic syndrome and recurrent thrombo-

embolism in patients with upper extremity deep vein thrombosis: A systematic review and meta-analysis. Thromb Res 2019;174:34–9.

24. van den Houten MM, van Grinsven R, Pouwels S, Yo LS, van Sambeek MR, Teijink JA. Treatment of upper-extremity outflow thrombosis. Phlebology 2016;31(Suppl):28–33.

25. Kakkos SK, Gohel M, Baekgaard N, Bauersachs R, Bellmunt-Montoya S, Black SA, *et al.*; Esvs Guidelines Committee. Editor's Choice - European Society for Vascular Surgery (ESVS) 2021 Clinical Practice Guidelines on the Management of Venous Thrombosis. Eur J Vasc Endovasc Surg 2021;61:9–82.

26. Karaolanis G, Antonopoulos CN, Koutsias SG, Giosdekos A, Metaxas EK, Tzimas P, *et al.* A systematic review and meta-analysis for the management of Paget-Schroetter syndrome. J Vasc Surg Venous Lymphat Disord 2021;9:801–810.e5.

27. Welsch T, Büchler MW, Kienle P. Recalling superior mesenteric artery syndrome. Dig Surg 2007;24:149–56.

28. Yale SH, Tekiner H, Yale ES. Historical terminology and superior mesenteric artery syndrome. Int J Surg Case Rep 2020;67:282–3.

29. Edwards KC, Katzen BT. Superior mesenteric artery syndrome due to large dissecting abdominal aortic aneurysm. Am J Gastroenterol 1984;79:72–4.

30. Shi Y, Shi G, Li Z, Chen Y, Tang S, Huang W. Superior mesenteric artery syndrome coexists with Nutcracker syndrome in a female: a case report. BMC Gastroenterol 2019;19:15.

31. Awaludin R, Ab Rahim H, Syazana Arivai D, Refaie Elkeleny M. Superior Mesenteric Artery Syndrome: A Worldwide Descriptive Study with Literature Review. Int J Med Stud. 2016;4:50–4.

32. Karrer FM, Jones SA. Superior Mesenteric Artery (SMA) Syndrome Treatment & Management; 2018 [Internet]. Available from: https://emedicine.medscape.com/article/932220-overview [cited 2023, Jun 30].

33. Niaz Z, Chaudhary FA, Gardazi JR. Superior mesenteric artery syndrome: an uncommon cause of upper intestinal obstruction. J Coll Physicians Surg Pak 2006;16:666–8.

34. Unal B, Aktaş A, Kemal G, Bilgili Y, Güliter S, Daphan C, *et al.* Superior mesenteric artery syndrome: CT and ultrasonography findings. Diagn Interv Radiol 2005;11:90–5.

35. Zaraket V, Deeb L. Wilkie's Syndrome or Superior Mesenteric Artery Syndrome: fact or Fantasy? Case Rep Gastroenterol 2015;9:194–9.

36. Lee TH, Lee JS, Jo Y, Park KS, Cheon JH, Kim YS, *et al.* Superior mesenteric artery syndrome: where do we stand today? J Gastrointest Surg 2012;16:2203–11.

37. Pottorf BJ, Husain FA, Hollis HW Jr, Lin E. Laparoscopic management of duodenal obstruction resulting from superior mesenteric artery syndrome. JAMA Surg 2014;149:1319–22.

38. Horton KM, Talamini MA, Fishman EK. Median arcuate ligament syndrome: evaluation with CT angiography. Radiographics 2005;25:1177–82.

39. Sultan SA, Acharya Y, Mustafa M, Hynes N. Two Decades of Experience With Chronic Mesenteric Ischaemia and Median Arcuate Ligament Syndrome in a Tertiary Referral Centre: A Parallel Longitudinal Comparative Study. Cureus 2021;13:e20726.

40. Sultan S, Hynes N, Elsafty N, Tawfick W. Eight years experience in the management of median arcuate ligament syndrome by decompression, celiac ganglion sympathectomy, and selective revascularization. Vasc Endovascular Surg 2013;47:614–9.

41. Goodall R, Langridge B, Onida S, Ellis M, Lane T, Davies AH. Median arcuate ligament syndrome. J Vasc Surg 2020;71:2170–6.

42. Barbon DA, Hsu R, Noga J, Lazzara B, Miller T, Stainken BF. Clinical Response to Celiac Plexus Block Confirms the Neurogenic Etiology of Median Arcuate Ligament Syndrome. J Vasc Interv Radiol 2021;32:1081–7.

43. DeCarlo C, Woo K, van Petersen AS, Geelkerken RH, Chen AJ, Yeh SL, *et al.* Factors associated with successful median arcuate ligament release in an international, multi-institutional cohort. J Vasc Surg 2023;77:567–577.e2.

44. Columbo JA, Trus T, Nolan B, Goodney P, Rzucidlo E, Powell R, *et al.* Contemporary management of median arcuate ligament syndrome provides early symptom improvement. J Vasc Surg 2015;62:151–6.

45. Romero-Velez G, Barajas-Gamboa JS, Pantoja JP, Corcelles R, Rodriguez J, Navarrete S, *et al.* A nationwide analysis of median arcuate ligament release between 2010 and 2020: a NSQIP Study. Surg Endosc 2023;37:140–7.

46. Jimenez JC, Harlander-Locke M, Dutson EP. Open and laparoscopic treatment of median arcuate ligament syndrome. J Vasc Surg 2012;56:869–73.

47. Rousselet MC, Saint-Andre JP, L'Hoste P, Enon B, Megret A, Chevalier JM. Stenotic intimal thickening of the external iliac artery in competition cyclists. Hum Pathol 1990;21:524–9.

48. Peake LK, D'Abate F, Farrah J, Morgan M, Hinchliffe RJ. The Investigation and Management of Iliac Artery Endofibrosis: Lessons Learned from a Case Series. Eur J Vasc Endovasc Surg 2018;55:577–83.

49. Schep G, Bender MH, van de Tempel G, Wijn PF, de Vries WR, Eikelboom BC. Detection and treatment of claudication due to functional iliac obstruction in top endurance athletes: a prospective study. Lancet 2002;359:466–73.

50. Peach G, Schep G, Palfreeman R, Beard JD, Thompson MM, Hinchliffe RJ. Endofibrosis and kinking of the iliac arteries in athletes: a systematic review. Eur J Vasc Endovasc Surg 2012;43:208–17.

51. Fisher AT, Tran K, Dossabhoy SS, Sorondo S, Fereydooni A, Lee JT. Anatomic Factors Contributing to External Iliac Artery Endofibrosis in High-Performance Athletes. Ann Vasc Surg 2022;87:181–7.

52. O'Ceallaigh P, Burns P, McLaughlin R, Leader M, Bouchier-Hayes D. Complete external iliac artery occlusion in a 34-year-old cyclist. Eur J Vasc Endovasc Surg 2002;23:376–7.

53. INSITE Collaborators (INternational Study group for Identification and Treatment of Endofibrosis). Diagnosis and Management of Iliac Artery Endofibrosis: Results of a Delphi Consensus Study. Eur J Vasc Endovasc Surg 2016;52:90–8.

54. D'Abate F, Paraskevas KI, Oates C, Palfreeman R, Hinchliffe RJ. Color Doppler ultrasound imaging in the assessment of iliac endofibrosis. Angiology 2017;68:225–32.

55. Hinchliffe RJ. Iliac artery endofibrosis. Eur J Vasc Endovasc Surg 2016;52:1–2.

56. Stuart TP. Note on a Variation in the Course of the Popliteal Artery. J Anat Physiol 1879;13:162.

57. Davis PS. Popliteal Artery Entrapment Syndrome. Vasc Endovascular Surg 1982;16:239–42.

58. Deveze E, Bruneau A, Hersant J, Ammi M, Abraham P, Picquet J. Popliteal Entrapment Syndrome: Diagnostic, Surgical Management, and Short-Term Results of a Ten-Year Experience. Ann Vasc Surg 2023;88:139–44.

59. Hoelting T, Schuermann G, Allenberg JR. Entrapment of the popliteal artery and its surgical management in a 20-year period. Br J Surg 1997;84:338–41.

60. Wady H, Badar Z, Farooq Z, Shaw P, Kobayashi K. Avoiding the Trap of Misdiagnosis: Valuable Teaching Points Derived from a Case of Longstanding Popliteal Artery Entrapment Syndrome. Case Rep Med 2018;2018:3214561.

61. Mustapha JA, Sarkar R, Rastogi U. Awareness and Early Diagnosis of Popliteal Artery Entrapment Syndrome Is Needed. JACC Case Rep 2022;4:429–32.

62. Kumar R, Warren P, Mannava K. Popliteal Artery Entrapment Syndrome Presenting with Critical Limb Ischemia in an Adolescent. J Pediatr 2020;217:215–215.e1.

63. Pandya YK, Lowenkamp MN, Chapman SC. Functional popliteal artery entrapment syndrome: A review of diagnostic and management approaches. Vasc Med 2019;24:455–60.

64. Ghaffarian AA, Nkansah R, Quiroga E, Tran N, Starnes BW, Singh N. Clinical Outcomes of a Diagnostic and Management Protocol for Pop-

liteal Artery Entrapment Syndrome at a Large Referral Center. Ann Vasc Surg 2022;87:140–6.

65. Shahi N, Arosemena M, Kwon J, Abai B, Salvatore D, DiMuzio P. Functional Popliteal Artery Entrapment Syndrome: A Review of Diagnosis and Management. Ann Vasc Surg 2019;59:259–67.

66. Alsaadi MJ, Aljabri B, Sulieman A, Mahmoud MZ. The reliability of duplex ultrasound in diagnosing popliteal artery entrapment syndrome: an observational pilot study. J Radiat Res Appl Sci. 2022;15:100472.

67. Wadhwani A, Nutley M, Bakshi D, Mirakhur A. Treatment of Functional Popliteal Artery Entrapment Syndrome with Ultrasound-Guided Botox Injection. J Vasc Interv Radiol 2018;29:1780–2.

68. White JM, Golarz SR, White PW, Craig RM, Whittaker DR. Intraoperative duplex ultrasound criteria for performing interposition bypass in the treatment of popliteal artery entrapment syndrome. Ann Vasc Surg 2015;29:124.e7–12.

69. Beseth BD, Moore WS. The posterior approach for repair of popliteal artery aneurysms. J Vasc Surg 2006;43:940–4, discussion 944–5.

70. Levien LJ, Veller MG. Popliteal artery entrapment syndrome: more common than previously recognized. J Vasc Surg 1999;30:587–98.

71. Fujimura N, Hosokawa K, Obara H, Igari K, Akamatsu D, Matsumoto H, *et al.* Incidence, diagnosis and treatment of popliteal artery entrapment syndrome in current vascular practice in Japan. Cardiovasc Interv Ther 2021;36:506–13.

72. Raju S, Neglen P. Popliteal vein entrapment: a benign venographic feature or a pathologic entity? J Vasc Surg 2000;31:631–41.

73. Lavingia KS, Dua A, Rothenberg KA, Fredericson M, Lee JT. Surgical management of functional popliteal entrapment syndrome in athletes. J Vasc Surg 2019;70:1555–62.

74. Carter A, Murphy MO, Halka AT, Turner NJ, Kirton JP, Murray D, *et al.* The natural history of stenoses within lower limb arterial bypass grafts using a graft surveillance program. Ann Vasc Surg 2007;21:695–703.

75. Sanders RJ, Hammond SL, Rao NM. Diagnosis of thoracic outlet syndrome. J Vasc Surg 2007;46:601–4.

76. Peek J, Vos CG, Ünlü Ç, van de Pavoordt HD, van den Akker PJ, de Vries JP. Outcome of surgical treatment for thoracic outlet syndrome: systematic review and meta-analysis. Ann Vasc Surg 2017;40:303–26.

77. Illig KA, Doyle AJ. A comprehensive review of Paget-Schroetter syndrome. J Vasc Surg 2010;51:1538–47.

78. Davidovic LB, Zlatanovic P, Dragas M, Koncar I, Micic M, Matejevic D. Arterial thoracic outlet syndrome: a 30-year experience in a high-volume referral center. J Cardiovasc Surg (Torino) 2022;63:687–94.

79. Desai SS, Toliyat M, Dua A, Charlton-Ouw KM, Hossain M, Estrera AL, *et al.* Outcomes of surgical paraclavicular thoracic outlet decompression. Ann Vasc Surg 2014;28:457–64.

80. Persson LM, Arnhjort T, Lärfars G, Rosfors S. Hemodynamic and morphologic evaluation of sequelae of primary upper extremity deep venous thromboses treated with anticoagulation. J Vasc Surg 2006;43:1230–5, discussion 1235.

81. Stilo F, Montelione N, Benedetto F, Spinelli D, Vigliotti RC, Spinelli F. Thirty-year experience of transaxillary resection of first rib for thoracic outlet syndrome. Int Angiol 2020;39:82–8.

82. Said SM, Gloviczki P, Kalra M, Oderich GS, Duncan AA, D Fleming M, *et al.* Renal nutcracker syndrome: surgical options. Semin Vasc Surg 2013;26:35–42.

83. Radisic MV, Feldman D, Diaz C, Froment RO. Unexplained hematuria during pregnancy: right-sided nutcracker phenomenon. Int Urol Nephrol 2007;39:709–11.

84. Kim SH. Doppler US and CT Diagnosis of Nutcracker Syndrome. Korean J Radiol 2019;20:1627–37.

85. Velasquez CA, Saeyeldin A, Zafar MA, Brownstein AJ, Erben Y. A systematic review on management of nutcracker syndrome. J Vasc Surg Venous Lymphat Disord 2018;6:271–8.

86. Meissner MH, Khilnani NM, Labropoulos N, Gasparis AP, Gibson K, Greiner M, *et al.* The Symptoms-Varices-Pathophysiology classification of pelvic venous disorders: A report of the American Vein & Lymphatic Society International Working Group on Pelvic Venous Disorders. Phlebology 2021;36:342–60.

87. Reed NR, Kalra M, Bower TC, Vrtiska TJ, Ricotta JJ 2nd, Gloviczki P. Left renal vein transposition for nutcracker syndrome. J Vasc Surg 2009;49:386–93, discussion 393–4.

88. Erben Y, Gloviczki P, Kalra M, Bjarnason H, Reed N, Duncan A, *et al.* Treatment of Nutcracker Syndrome with Open and Endovascular Interventions. J Vasc Surg Venous Lymphat Disord 2014;2:116.

89. Chait J, Sen I, Kalra M. Nutcracker Syndrome: How to Diagnose It and When/How Should It Be Treated in the Pelvic Venous Disease Population. Tech Vasc Interv Radiol 2021;24:100734.

90. Takebayashi S, Ueki T, Ikeda N, Fujikawa A. Diagnosis of the nutcracker syndrome with color Doppler sonography: correlation with flow patterns on retrograde left renal venography. AJR Am J Roentgenol 1999;172:39–43.

91. Kim KW, Cho JY, Kim SH, Yoon JH, Kim DS, Chung JW, *et al.* Diagnostic value of computed tomographic findings of nutcracker syndrome: correlation with renal venography and renocaval pressure gradients. Eur J Radiol 2011;80:648–54.

92. Ananthan K, Onida S, Davies AH. Nutcracker Syndrome: An Update on Current Diagnostic Criteria and Management Guidelines. Eur J Vasc Endovasc Surg 2017;53:886–94.

93. Barnes RW, Fleisher HL 3rd, Redman JF, Smith JW, Harshfield DL, Ferris EJ. Mesoaortic compression of the left renal vein (the so-called nutcracker syndrome): repair by a new stenting procedure. J Vasc Surg 1988;8:415–21.

94. Miró I, Serrano A, Pérez-Ardavín J, March JA, Polo A, Conca MÁ, *et al.* Eighteen years of experience with pediatric nutcracker syndrome: the importance of the conservative approach. J Pediatr Urol 2020;16:218.e1–6.

95. Sebastian T, Erdoes G, Bratu VA, Baumgartner I, Kucher N. Endovascular extraction of a migrated large self-expanding laser-cut renal venous stent from the right ventricle. J Vasc Surg Cases Innov Tech 2017;3:79–82.

96. Hartung O, Grisoli D, Boufi M, Marani I, Hakam Z, Barthelemy P, *et al.* Endovascular stenting in the treatment of pelvic vein congestion caused by nutcracker syndrome: lessons learned from the first five cases. J Vasc Surg 2005;42:275–80.

97. Rodríguez Morata A, González-Fajardo JA. Controversias en el síndrome de congestion pélvica. Una perspectiva hemodinámica. Angiologia 2020;72:215–78.

98. Rodríguez-Morata A, Robles-Martín ML, Reyes-Ortega JP. Endovascular treatment of posterior nutcracker syndrome with a new autoexpandable stent. J Vasc Surg Venous Lymphat Disord 2019;7:118–21.

99. Decaestecker K, Van Parys B, Van Besien J, Doumerc N, Desender L, Randon C, *et al.* Robot-assisted Kidney Autotransplantation: A Minimally Invasive Way to Salvage Kidneys. Eur Urol Focus 2018;4:198–205.

100. Fuentes-Perez A, Bush RL, Kalra M, Shortell C, Gloviczki P, Brigham TJ, *et al.* Systematic review of endovascular versus laparoscopic extravascular stenting for treatment of nutcracker syndrome. J Vasc Surg Venous Lymphat Disord 2023;11:433–41.

101. Ibrahim W, Al Safran Z, Hasan H, Zeid WA. Endovascular management of may-thurner syndrome. Ann Vasc Dis 2012;5:217–21.

102. Milinis K, Thapar A, Shalhoub J, Davies AH. Antithrombotic Therapy Following Venous Stenting: international Delphi Consensus. Eur J Vasc Endovasc Surg 2018;55:537–44.

103. Abdul-Haqq R, Novak Z, Pearce BJ, Matthews TC, Patterson MA, Jordan WD Jr, *et al.* Routine extended follow-up surveillance of iliac vein stents for iliocaval venous obstruction may not be warranted. J Vasc Surg Venous Lymphat Disord 2017;5:500–5.

104. Neglén P, Hollis KC, Olivier J, Raju S. Stenting of the venous outflow in chronic venous disease: long-term stent-related outcome, clinical, and hemodynamic result. J Vasc Surg 2007;46:979–90.

105. Liu Z, Gao N, Shen L, Yang J, Zhu Y, Li Z, *et al.* Endovascular treatment for symptomatic iliac vein compression syndrome: a prospective consecutive series of 48 patients. Ann Vasc Surg 2014;28:695–704.

106. Black S, Janicek A, Knuttinen MG. Re-intervention for occluded iliac vein stents. Cardiovasc Diagn Ther 2017;7(Suppl 3):S258–66.

107. Zamboni P, Scerrati A, Menegatti E, Galeotti R, Lapparelli M, Traina L, *et al.* The eagle jugular syndrome. BMC Neurol 2019;19:333.

108. Jessen NA, Munk AS, Lundgaard I, Nedergaard M. The Glymphatic System: A Beginner's Guide. Neurochem Res 2015;40:2583–99.

109. Liu M, Xu H, Wang Y, Zhong Y, Xia S, Utriainen D, *et al.* Patterns of chronic venous insufficiency in the dural sinuses and extracranial draining veins and their relationship with white matter hyperintensities for patients with Parkinson's disease. J Vasc Surg 2015;61:1511–20.e1.

110. Beggs C, Chung CP, Bergsland N, Wang PN, Shepherd S, Cheng CY, *et al.* Jugular venous reflux and brain parenchyma volumes in elderly patients with mild cognitive impairment and Alzheimer's disease. BMC Neurol 2013;13:157.

111. Zhou D, Meng R, Zhang X, Guo L, Li S, Wu W, *et al.* Intracranial hypertension induced by internal jugular vein stenosis can be resolved by stenting. Eur J Neurol 2018;25:365–e13.

112. Zhou D, Ding J, Asmaro K, Pan L, Ya J, Yang Q, *et al.* Clinical Characteristics and Neuroimaging Findings in Internal Jugular Venous Outflow Disturbance. Thromb Haemost 2019;119:308–18.

113. Zamboni P, Galeotti R, Salvi F, Giaquinta A, Setacci C, Alborino S, *et al.*; Brave Dreams Research Group. Effects of Venous Angioplasty on Cerebral Lesions in Multiple Sclerosis: Expanded Analysis of the Brave Dreams Double-Blind, Sham-Controlled Randomized Trial. J Endovasc Ther 2020;27:1526602819890110.

114. Scerrati A, De Bonis P, Zamboni P, Dones F, Fontanella M, Cenzato M, *et al.* A New Insight in Nonaneurysmal Subarachnoid Hemorrhage: The Potential Role of the Internal Jugular Veins. J Neurol Surg A Cent Eur Neurosurg 2022;83:344–50.

115. De Bonis P, Menegatti E, Cavallo MA, Sisini F, Trapella G, Scerrati A, *et al.* JEDI (jugular entrapment, dilated ventricles, intracranial hypertension) syndrome: a new clinical entity? A case report. Acta Neurochir (Wien) 2019;161:1367–70.

116. Zamboni P, Menegatti E, Cittanti C, Sisini F, Gianesini S, Salvi F, *et al.* Fixing the jugular flow reduces ventricle volume and improves brain perfusion. J Vasc Surg Venous Lymphat Disord 2016;4:434–45.

117. Jayaraman MV, Boxerman JL, Davis LM, Haas RA, Rogg JM. Incidence of extrinsic compression of the internal jugular vein in unselected patients undergoing CT angiography. AJNR Am J Neuroradiol 2012;33:1247–50.

118. Zamboni P, Menegatti E, Conforti P, Shepherd S, Tessari M, Beggs C. Assessment of cerebral venous return by a novel plethysmography method. J Vasc Surg 2012;56:677–85.e1.

Conflicts of interest
The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

Authors' contributions
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