

Vascular compression syndromes

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Summary: Dealing with vascular compression syndromes is one of the most challenging tasks in Vascular Medicine practice. This heterogeneous group of disorders is characterised by external compression of primarily healthy arteries and/or veins as well as accompanying neural structures, carrying the risk of subsequent structural vessel wall and nerve damage. Vascular compression syndromes may severely impair health-related quality of life in affected individuals who are typically young and otherwise healthy. The diagnostic approach has not been standardised for any of the vascular compression syndromes. Moreover, some degree of positional external compression of blood vessels such as the subclavian and popliteal vessels or the celiac trunk can be found in a significant proportion of healthy

individuals. This implies important difficulties in differentiating physiological from pathological findings of clinical examination and diagnostic imaging with provocative manoeuvres. The level of evidence on which treatment decisions regarding surgical decompression with or without revascularisation can be relied on is generally poor, mostly coming from retrospective single centre studies. Proper patient selection is critical in order to avoid over-treatment in patients without a clear association between vascular compression and clinical symptoms. With a focus on the thoracic outlet-syndrome, the median arcuate ligament syndrome and the popliteal entrapment syndrome, the present article gives a selective literature review on compression syndromes from an interdisciplinary vascular point of view.

Key words: Vascular compression syndrome, thoracic outlet syndrome, median arcuate ligament syndrome, Dunbar's syndrome, popliteal entrapment syndrome

Introduction

Vascular compression syndromes, characterised by clinical symptoms arising from the external compression of normal, non-diseased blood vessels, belong to the most controversially discussed disorders in Clinical Medicine. Being relatively infrequent, reliable diagnostic criteria have not yet been established. Moreover, positional extrinsic compression of vascular structures can be found in a substantial proportion of healthy individuals, resulting in serious diagnostic uncertainties. Finally, the level of evidence regarding the treatment of vascular compression syndromes is generally poor. The present review article aims to critically review the available literature to provide a scientific basis for clinical decision making. The main focus of this paper lies on the thoracic outlet syndrome (TOS), the median arcuate ligament syndrome (MALS, also called Dunbar's syndrome) and

the popliteal entrapment syndrome (PES). May-Thurner-Syndrome and the Hypothenar-Hammer-Syndrome have been recently reviewed in this journal [1, 2] and thus were not included in this work. More rare vascular compression syndromes are briefly discussed.

Thoracic outlet syndrome

Definition and epidemiology

The term "TOS" was introduced by Peet et al. in 1956 [3] and is used today to describe a heterogeneous disease complex characterised by upper extremity symptoms resulting from (positional) compression of the neurovascular bundle at the upper thoracic outlet [4, 5]. The scarce epidemiological data available should be interpreted with caution. Reported incidences range from 3 to 80 cases per 1,000 population, and women seem to be more frequently affected

than men. The onset of symptoms is typically observed between 20 and 50 years of age [6].

Pathoanatomy

The upper thoracic outlet consists of three compartments, namely the interscalene triangle, the costoclavicular space and the retropectoralis minor space [7]. Within this container, the brachial plexus, the subclavian artery, the subclavian vein and also sympathetic nerve fibres can be compressed by various anatomical abnormalities, either congenital or developmental (Tab. I, Fig. 1) [3–7]. The most common location of compression is the costoclavicular space. Of note, the subclavian vein does not cross the interscalene triangle and instead runs in front of the anterior scalene muscle. Thus, hypertrophied scalene muscles or cervical ribs usually do not cause subclavian vein compression [8]. In addition to anatomical factors, it is assumed that

static factors may play an aetiological role, e.g. postural abnormalities of the spine and the shoulder girdle, or hypertrophy of the scalene muscles due to body building [9]. Moreover, local trauma to the shoulder or neck

(e.g. whiplash injury) may contribute to the development of TOS via post-traumatic scarring and shortening of the affected musculature [4, 5].

Classification and clinical presentation

The term TOS neither specifies the components of the neurovascular bundle being compressed nor the compressing anatomical structures [5]. Therefore, according to the clinical symptoms, TOS should be discriminated in arterial TOS (aTOS), venous TOS (vTOS, also referred to as “thoracic inlet syndrome”) and neurogenic TOS (nTOS) [4–7]. Based on the presence or absence of objective diagnostic findings, nTOS can be further categorised into true and disputed nTOS [6]. While aTOS, vTOS and true nTOS are well defined and broadly accepted, disputed nTOS is subject of a highly controversial interdisciplinary debate. Sceptics even argue that if this disease entity really exists it is quite rare [10]. On the contrary, proponents consider it to be a common disorder and in many

surgical series patients with disputed nTOS constituted up to 90 % of the study population.

The incidence of vTOS was much lower in these series (3–5 % of all TOS cases), with primary upper extremity deep venous thrombosis (also entitled “Paget-Schroetter-Syndrome” or “effort thrombosis”) being the first manifestation of vTOS in most affected subjects. It typically occurs in the dominant arm after strenuous, unusual or repetitive physical activity and presents with sudden onset oedematous arm swelling and/or upper extremity discomfort or pain [8]. vTOS presenting with intermittent arm swelling and cyanosis due to positional subclavian vein obstruction (McCleery-Syndrome) is quite rare [11].

Less than 1 % of TOS cases are arterial. In these cases, repetitive arterial compression has led to vessel wall damage, resulting in scarred stenosis or the development of ectasia/aneurysm (Fig. 2). Structural vessel wall damage predisposes to mural thrombus formation, being a hazardous source of arterial embolism into the arm and digital arteries (Fig. 3) [5]. Acute or subacute digital ischemia is the leading clinical manifestation of aTOS and the clinical spectrum ranges from secondary Raynaud’s phenomenon to finger necrosis due to critical ischemia. Arm claudication is a less frequent manifestation of aTOS. Symptoms of nTOS are non-radicular in nature and typically influenced by the position of the upper limb and/or neck. The most frequent symptoms are pain and paresthesia of the arm, hand and fingers (with paresthesia restricted to the ulnar aspect of the forearm and hand in most cases due to predominant affection of the lower plexus) [4–7]. Further symptoms include pain of the shoulder, neck and supraclavicular fossa as well as occipital headache. In advanced disease, patients may experience a loss of

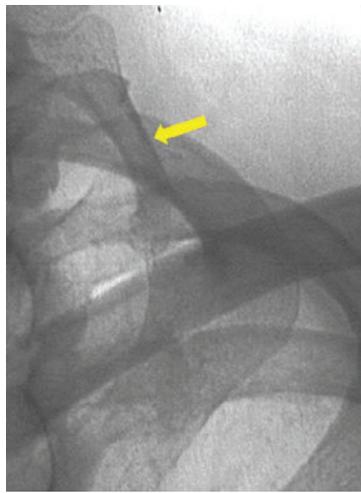


Figure 1: Left-sided cervical rib in a patient with TOS (arrow), (TOS = thoracic outlet syndrome).

Table I: Aetiology of the thoracic outlet syndrome

Anatomical abnormalities

Bony structures

Complete or incomplete cervical rib

Elongated C7 transversal processus

Abnormal first rib (e.g. exostosis)

Abnormal clavicle (e.g. callus or malunion after fracture)

Soft tissue abnormalities

Scalene muscle hypertrophy

Minor scalene minor muscle

Pectoralis minor muscle

Subclavian muscle

Various ligamentous abnormalities (e.g. costoclavicular ligament)

Postural abnormalities

Hypotension of the shoulder girdle musculature

Trauma

Neck trauma (e.g. whiplash injury resulting in muscle scarring and shortening)

Repetitive upper extremity activities related to work or athletics

strength and impairment of fine motor skills of the hand [6]. Raynaud's phenomenon secondary to affection of sympathetic fibres can also be a clinical feature of nTOS and can hamper the discrimination of nTOS and aTOS [5]. Among the multitude of disorders to be considered in the differential diagnosis of nTOS, some of the most important are cervical radiculopathy, ulnar or median nerve entrapment and shoulder pathologies [6].

Noteworthy, bilateral symptoms (particularly in nTOS and vTOS) as well as combined vascular and neurologic symptoms are not uncommon [12, 13].

Diagnostic workup

Diagnosis of TOS is challenging. This is underlined by the results from a cohort study reporting on patients who underwent surgery for established TOS. Patients reported a mean of 3.1 ± 1.1 physician-consultations prior to diagnosis and a mean latency between symptom onset and diagnosis of 46.0 ± 83.5 months [14]. A detailed medical history and thorough physical examination are of utmost importance in the clinical evaluation of suspected TOS. Visual inspection may reveal predisposing postural variants such as drooping shoulders [7] as well as sequelae of the syndrome (e.g., hand muscle atrophy in nTOS, digital necrosis in aTOS, prominent subcutaneous collateral veins or livid discoloration and oedema of the upper limb in vTOS) [4, 8]. Bilateral pulse palpation, arterial auscultation and Allen's Test are essential for detection of peripheral arterial complications in aTOS. Neurological assessment has been described elsewhere in detail [6, 7]. Clinical provocative manoeuvres are the key elements in the diagnosis of TOS. Various tests aiming to uncover compression at different compartments of the thoracic outlet have been

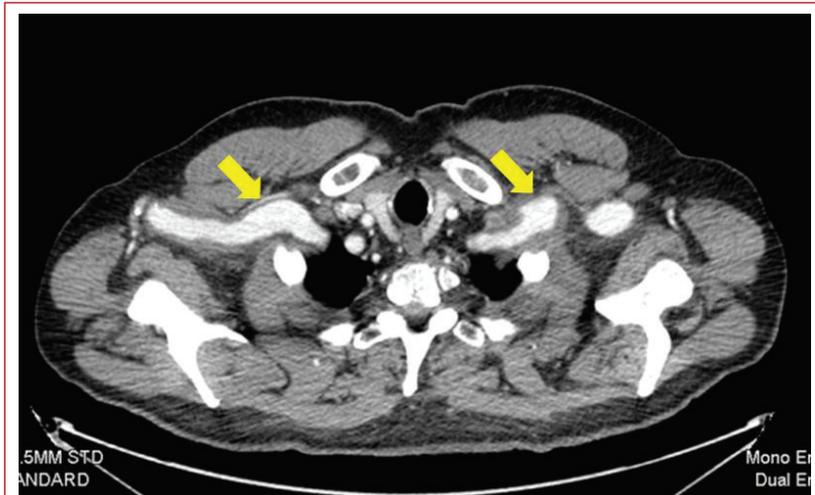


Figure 2: Bilateral subclavian artery aneurysms (arrows) with mural thrombus in a patient with aTOS. (aTOS = arterial thoracic outlet syndrome).

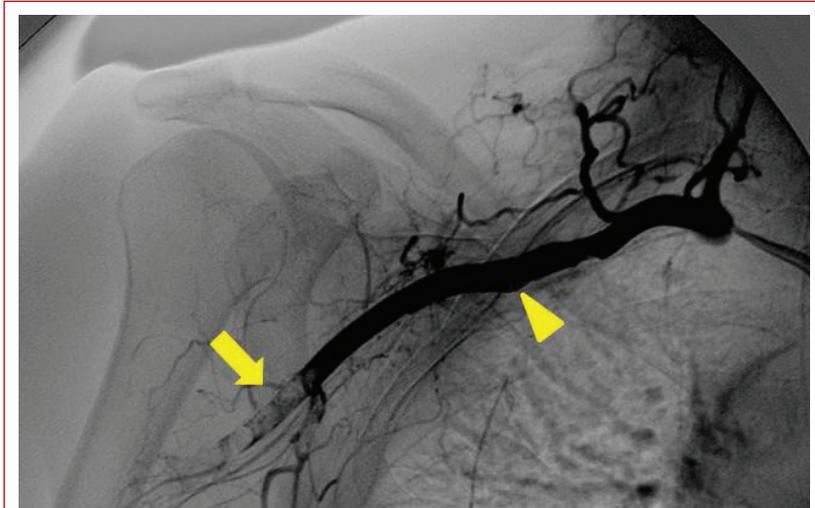


Figure 3: Embolic axillary artery occlusion (arrow) originating from subclavian artery ectasia (arrowhead) in a patient with aTOS. (aTOS = arterial thoracic outlet syndrome).

described in the literature (Tab. II). Tests are considered to be positive when the patient's symptoms can be reproduced and/or arterial pulse loss or arterial bruits appear. Unfortunately there is (a) no consensus on standardised testing and (b) the diagnostic accuracy of the tests is

unclear as a diagnostic gold standard is missing. The main problem is the substantial rate of positive test results in healthy individuals, implying the risk of over-diagnosis [9, 15–17]. A diagnostic approach with a combination of several tests (arm abduction, elevation and external rotation as well

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Table II: Clinical provocative manoeuvres for the diagnosis of TOS (modified according to [5])

Test	Proposed mechanism	Procedure*	Diagnostic** accuracy
Adson's test	Compression at the interscalene triangle	The patient breathes in deeply, then extends and rotates the neck towards the side being tested. <u>Positive test:</u> loss of radial pulse, supraclavicular bruit and/or pain/paresthesia	Sensitivity 79 % Specificity 74–100 %
Costoclavicular manoeuvre (Military exercise-test)	Costoclavicular compression	The patient breathes in and retracts both shoulders, while the examiner draws the patient's arm down. <u>Positive test:</u> loss of radial pulse, subclavian bruit and/or pain/paresthesia	Sensitivity: unknown Specificity 53–100 %
Wright's test	Costopectoral compression	Passive retraction of the hyperabducted, externally rotated arm <u>Positive test:</u> loss of radial pulse, axillary bruit and/or pain/paresthesia	Sensitivity 70–90 % Specificity 29–53 %
Elevated arm stress test (Roos test)	Not specified	90° abduction and full external rotation of both arms with the head in neutral position. While holding this position, the Patient opens and closes the hands repeatedly for 3 minutes. <u>Positive test:</u> pain and/or paresthesia, early discontinuation of the test for relief of symptoms	Sensitivity 52–84 % Specificity 30–100 %
Modified upper limb tension test (Elvey test)	Compression of the brachial plexus in nTOS	(1) Active arm abduction with extended elbows (2) Dorsiflexion of both wrists (3) Tilting the head to each shoulder <u>Positive test:</u> Pain down the arm or paresthesia (ipsilateral in position 1 and 2; contralateral to the side the head is tilted in position 3)	Sensitivity: 98 % Specificity: unknown (limited)

*All tests are performed with the patients in a seated position and the radial pulse being palpated by the examiner.
**Diagnostic accuracy according to the final clinical diagnosis.



Figure 4: Intra-arterial angiography showing focal luminal narrowing (arrow) of the left subclavian artery in neutral position (A) and complete occlusion (arrowhead) with arm abduction in a patient with TOS (B). (TOS = thoracic outlet syndrome).

as shoulder retraction and head rotation) seems to increase the diagnostic accuracy and is thus recommended for clinical practice [18].

Non-invasive vascular laboratory testing is the next step in objectifying TOS with vascular compression. The diagnostic approach should include acral photoplethysmography for detection of digital artery obstructions and colour duplex sonography for detection of morphological abnormalities of the subclavian arteries (mural thrombus, aneurysm) and subclavian veins (post-thrombotic sequelae). Complete colour duplex sonography of the upper extremity arteries also enables the investigator to detect important alternative reasons for digital artery occlusions such as the hypothenar hammer syndrome [2]. Dynamic testing with provoca-

tive manoeuvres performed in a sitting position is helpful in confirming positional arterial compression. With this regard, pulse volume recording, continuous wave Doppler sonography and colour duplex sonography can be applied. However, it should be noted that the sensitivity of colour duplex sonography is probably not superior to clinical provocative testing with combined manoeuvres [18]. Confirmation of arterial compression may also contribute to the diagnosis of vTOS, and a study applying colour duplex sonography revealed arterial compression in every second patient with nTOS [19]. It must be pointed out that the absence of arterial compression does not allow ruling out nTOS [5]. Thus, in suspected nTOS, further testing (nerve conduction studies and electromyography) is required. A more detailed description of electrodiagnostic tests in suspected nTOS is provided elsewhere [4,6]. Conventional X-ray imaging of the chest and neck easily allows the detection of cervical ribs (Fig. 1) [5]. Computed tomography (CT) and magnetic resonance imaging (MRI) including CT- and MR-angiography in neutral position and under provocation are helpful for uncovering the site of vascular compression and for characterisation of the compressing structure when surgical decompression is intended [20–24]. Intra-arterial angiography still has a limited role in the diagnostic workup of patients with suspected aTOS, mainly in assessing the digital arteries for embolic occlusions, in investigating the subclavian arteries in neutral and provocative position in cases of suspected aTOS remaining ambiguous after non-invasive diagnostics, and for planning arterial reconstruction (Fig. 4) [5]. Similarly, contrast venography with postural manoeuvres can be a useful diagnostic tool in suspected vTOS [8]. However, venography results should be interpreted with caution, as partial venous

compression at the thoracic outlet is universally found in normal subjects with arm abduction/external rotation [24]. When interpreting the results of vascular imaging, the examiner should take into account that passive arm abduction in a supine position may fail to replicate the true pathophysiology of TOS. Cornelis et al. found that arteriography revealed a significantly lower rate of severe positional arterial stenosis when performed in a supine position (31 %) compared to the investigation done in a sitting position (87 %) in patients with established TOS [25].

It is of great importance to recognise that the detection of postural vessel compression by vascular imaging in an asymptomatic patient does not justify the diagnosis of TOS.

Treatment principles

Treatment of TOS has two goals: relief of the patient's symptoms and prevention of complications. Depending on the type and severity of TOS, conservative and invasive surgical approaches can be applied. The level of evidence on which treatment decisions can be based is still very low, with the majority of the published reports being retrospective single centre experiences [9, 26]. Some factors particularly impede the interpretation of the currently available literature: heterogeneous diagnostic criteria, lack of characterisation of the TOS-type in many studies, difficulties in reporting changes in subjective symptoms and differences in the outcome definitions. As of today, only two small prospective, randomised trials dealing with the treatment of TOS have been published, but there is no randomised trial comparing surgical vs. conservative treatment [27, 28]. How to treat the condition is therefore one of the most controversial points in the scientific debate surrounding TOS.

Conservative treatment includes patient education to avoid provocative arm positions (e.g., overhead use of the upper extremities), pain control, and individually tailored physical therapy aimed to strengthen the muscles of the pectoral girdle and to restore a normal posture [6, 29]. With conservative treatment, rates of clinical improvement between 50 and 90 % have been reported [6, 9]. Duration of symptoms and additional psychoemotional disturbances may negatively impact the treatment result [9]. The principles and controversies of conservative TOS-treatment are outlined in detail in the reviews of Hooper and Vanti et al. [9, 29]. Surgical decompression of the neurovascular bundle of the thoracic outlet can include resection of bony structures (first rib, cervical rib, callus resulting from a clavicle fracture), scalenotomy, neurolysis of the brachial plexus and resection of fibrous bands. In retrospective studies, rates of significant clinical improvement were between 45 and 92 % [4]. The rate of severe complications (particularly nerve and vessel injury) has been reported to be very low in experienced centres [13, 14, 30, 31]. Of note, recurrent symptoms of TOS seem to be common in the long-term follow-up and were the reason for second surgical procedures in about 30 % of patients in the large series published by Urschel et al. [30]. Thus, when referring a patient for surgical treatment of TOS, one should be aware that (a) the procedure carries a minor risk for significant neurovascular complications and (b) that a substantial percentage of patients will have no benefit from the surgical procedure.

Decompressive surgery can be performed via a transaxillary or an anterior supraclavicular approach. Introduced by Roos in 1966 [32], the transaxillary approach has become the most popular procedure in the surgical treatment of TOS [6, 13,

14, 31]. It provides a good exposure of almost the entire first rib. As the incision is hidden in the axilla, the cosmetic result is usually excellent. A shortcoming of this approach lies in its limited exposure of the neural structures. The supraclavicular approach offers a good exposure of the dorsal part of the first rib and of cervical ribs when present. It allows wide exposure of the brachial plexus but, on the contrary, carries a considerable risk of nerve injury (phrenic nerve, brachial plexus, cervical sympathetic chain). When indirectly comparing the results of cohort studies applying either the transaxillary or the supraclavicular approach, clinical success rates appear quite similar [4]. In a small prospective randomised trial comprising 55 patients with nTOS, Sheth et al. compared neuroplasty (without first rib resection) through a supraclavicular approach with transaxillary resection of the first rib. Vascular TOS and the presence of a cervical rib were exclusion criteria. During a mean follow-up of 37 months, significantly more patients in the group randomised to transaxillary first rib resection reported clinical improvement (75 % vs. 48 %) [27]. The decision on which approach to use in an individual patient should take into account the pathoanatomy, the presence of structural arterial damage and how familiar the surgeon is with the specific procedure. Endoscopic and computer-assisted (Da-Vinci-system) procedures for transaxillary first rib resection have been proposed as alternatives to traditional open surgery [33, 34]. Whether these techniques bear the potential to reduce local complications needs to be clarified in future studies.

Treatment approach stratified according to the type of TOS

In vTOS presenting with primary upper extremity deep venous thrombosis (Paget-Schroetter-Syndrome), the

treatment goals are prevention of (a) the post-thrombotic syndrome and (b) of recurrent thrombosis. With this regard it is noteworthy that severe post-thrombotic syndrome is virtually unknown after primary upper extremity deep venous thrombosis [35]. Moreover, there seems to be no clear correlation between morphological sequelae (i.e. residual venous stenosis and occlusion) and the presence and severity of the post-thrombotic syndrome [36]. Some authors advocate decompressive surgery for the treatment of Paget-Schroetter-syndrome, usually embedded into a multimodal approach with catheter-directed thrombolysis and angioplasty with or without stent placement to restore venous patency [37]. Although an abundance of mainly retrospective single centre studies reported impressive results of thrombolysis alone or in combination with surgical decompression, high-quality evidence proving the concept that invasive treatment reduces the frequency of the post-thrombotic syndrome and of recurrent thrombosis is lacking. The current ACCP-guidelines recommend anticoagulation treatment for a minimum duration of 3 months for all patients. Catheter-directed thrombolysis should be considered only for a subgroup of patients with extensive thrombosis suffering from severe symptoms for less than 14 days and having a very low bleeding risk [38]. According to the guidelines, surgical decompression should be confined to exceptional circumstances [38]. On the contrary, first rib resection and venolysis appear to be reasonable treatment options in patients suffering from McCleery-Syndrome [11]. Despite a lack of systematic data, attributable to the rarity of this disease variant, surgical decompression is generally accepted in aTOS to prevent (recurrent) ischemic events [6, 29]. Surgery should be performed early after restoration of distal blood flow

when required (e.g., catheter directed thrombolysis in upper limb ischemia secondary to arterio-arterial embolism). Following decompression, reconstruction of the subclavian artery must be performed in case of morphological alterations (particularly aneurysms). This frequently requires a combined transaxillary and supraclavicular approach.

Most patients with nTOS should primarily receive conservative therapy for a few months [6, 29]. With this regard, a double-blind, randomised, controlled trial failed to prove a positive effect of botulinum toxin injections vs. placebo injections to the scalene muscles on pain in subjects with nTOS [28]. When conservative treatment does not result in an improvement of patient's symptoms and related disability, surgical decompression of the brachial plexus can be considered. Noteworthy, limited data from cohort studies comparing conservative and surgical treatment yielded contradictory results but raised concerns about the efficacy of surgical treatment in nTOS [39–41]. An observational study indicated that a highly selective algorithm with initial TOS-specific physical therapy in all nTOS-patients and surgical decompression performed only in a subset of patients depending mainly on the success of conservative treatment may improve the clinical success rate of surgery [42]. Only in patients with true nTOS suffering from severe muscle weakness or atrophy a surgery first-strategy may be warranted [6, 29]. In a study focusing on long-term quality of life (QOL), Rochlin et al. identified several factors that predicted the clinical outcome of decompression surgery in nTOS patients. Significantly poorer scores on validated QOL-assessment-instruments were associated with comorbid chronic pain syndromes, opioid use, smoking, age ≥ 40 years, neck and/or

shoulder disease and surgical complications [12].

Median arcuate ligament syndrome (Dunbar's syndrome)

Definition and epidemiology

MALS is probably the most controversial vascular compression syndrome. MALS was broadly recognised as a clinical syndrome after publication of a series comprising 15 patients treated with surgical decompression by Dunbar et al. in 1965 [43]. Epidemiologic data are not available. The female to male ratio is estimated at 3:1 and the typical patients are women with an asthenic habitus aged between 20 and 40 years. However, MALS has been reported in patients of any age group [44].

Pathoanatomy

The median arcuate ligament is a musculofibrous arch bridging the crura of the diaphragm to form the aortic hiatus. It traverses the aorta above the origin of the celiac trunk, and the celiac ganglion is adjacent to the ligament. Imaging studies showed some degree of external compression of the celiac trunk resulting from the median arcuate ligament in about one of five asymptomatic individuals [45]. Low insertion of the diaphragm and a high origin of the celiac trunk are considered to increase the risk of celiac artery compression [46]. Compression is mostly pronounced in expiration and can affect also the superior mesenteric artery and rarely the renal arteries.

In MALS, positional compression of the celiac artery is associated with clinical symptoms. However, the actual pathomechanism behind the clinical syndrome still remains to be elucidated. Given the abundant splanchnic collateral circulation,

most authors argue that obstruction of the celiac trunk secondary to external compression is able to cause symptomatic ischemia of the upper gastrointestinal tract. Interestingly, a more recent study applying gastric exercise tonometry challenged this theory as it documented a gradient between gastric and arterial blood carbon dioxide level after exercise, indicating gastric ischemia secondary to blood redistribution in patients considered as having typical MALS [48]. A postprandial steal phenomenon via collaterals to the superior mesenteric artery may contribute to gastric ischemia. Another disputed mechanism is compression and irritation of the celiac plexus resulting in alteration of gastric myoelectrical activity [49].

Fixed stenosis with post-stenotic dilatation of the celiac trunk is frequently seen in MALS and a result of structural vessel wall alterations secondary to chronic compression. Some cases with aneurysms of the pancreaticoduodenal artery in association to MALS have been described, possibly related to the increased collateral blood flow via this artery [50].

Clinical presentation

MALS may present with a variety of symptoms, with the classical triad consisting of postprandial abdominal pain, weight loss and nausea/vomiting. However, presentation with unspecific complaints such as chronic, continuous or intermittent, vague pain epigastric pain, sometimes radiating to the flanks or back, seems to be common. A more characteristic presentation is exercise-induced abdominal pain in young athletes who may experience symptom relief when leaning forward [46]. In case of compression of the renal arteries by the diaphragmatic crura, the leading clinical consequence is renovascular hypertension [47].

Diagnostic workup

The optimal diagnostic workup is highly disputed. First of all, MALS is a diagnosis of exclusion and should be considered in cases with abdominal symptoms remaining unclear after thorough gastroenterological diagnostic testing including endoscopy of the upper and lower gastrointestinal tract. Physical examination may reveal epigastric tenderness or an abdominal bruit. The most reasonable first step in diagnostic imaging is functional colour duplex sonography with respiratory manoeuvres. In MALS, a significant increase in PSV with deep expiration can be observed. A case-control study limited by its very small sample size suggested a peak systolic expiratory flow velocity of more than 350 cm/s together with a celiac trunk deflection angle during expiration of more than 50° as a diagnostic criterion for MALS (sensitivity 83%, specificity 100%) [51]. Computed tomography and magnetic resonance tomography allow excellent visualization of external compression of the celiac trunk, but intra-arterial angiography, performed with respiratory manoeuvres remains the reference standard of diagnostic imaging in MALS. A hook-shaped appearance of the celiac trunk observed in the sagittal plane during expiration is the typical angiographic finding in MALS (Fig. 5). Provocation of a steal phenomenon by direct vasodilator injection into the superior mesenteric artery has been suggested as a diagnostic criterion for MALS [52]. Intra-arterial pressure measurement and intravascular ultrasound may further elucidate the dynamic character of celiac artery stenosis secondary to external compression [53]. Gastric exercise tonometry may substantially add to the diagnosis of MALS. According to a protocol published by Otte et al., arterial blood gas analysis and gastric pCO₂-measurement via a nasogastric tube



Figure 5: Selective intra-arterial angiography of the celiac trunk in a female patient suffering from median arcuate ligament syndrome (MALS): Fixed stenosis of the celiac trunk origin visible during inspiration (arrow) (A). Further luminal narrowing resulting in subtotal stenosis after expiration (arrow) (B). Courtesy of Prof. Gunnar Tepe, Rosenheim, Germany.

are performed at rest and after 10 minutes of submaximal exercise on a cycle ergometer. Effective gastric acid suppression prior the test with proton pump inhibitors is mandatory. The main criterion for a positive (pathological test) is a post-exercise gradient between gastric and arterial blood carbon dioxide of >6 mmHg (0.8 kPa) [54]. The combination of colour duplex sonography and gastric exercise tonometry has been shown to have excellent diagnostic accuracy in the diagnosis of chronic gastrointestinal ischemia and hence may be a reasonable first line diagnostic approach in suspected MALS

[55]. Independent validation of these promising results by other working groups is pending.

Treatment

When the diagnosis of MALS is considered to be very likely, surgical decompression is the treatment of choice. Both open surgical decompression, performed via a short upper midline laparotomy, and laparoscopic decompression comprise transection of the median arcuate ligament and the crus of the diaphragm proximal to the celiac artery as well as transection of the ganglionic tissue over the aorta. Revascularisation of the celiac artery

is frequently required due to fibrous stenosis, occlusion or even aneurysmal degeneration. Open surgery allows direct reconstruction (patch or aorto-coeliac bypass), whereas laparoscopic treatment requires an additional endovascular approach with stenting of the celiac artery. Robot-assisted laparoscopic surgery has been performed successfully in a limited number of MALS-patients [56].

In a systematic review of 21 retrospective studies on 400 patients, complete postoperative symptom relief was achieved in 96 % and 78 % after laparoscopic and open surgery, respectively [57]. Additional endovascular procedures were required in 12 % of patients who underwent laparoscopic surgery. The frequency of late symptom recurrence did not differ between the treatment modalities (6–7 %). Thus, the individual patient needs to be informed explicitly that there remains a small chance of symptom persistence or recurrence despite proper surgical treatment. In 9.1 % of subjects who underwent laparoscopic treatment, bleeding complications forced conversion to open surgery [57]. Recently, Klimas et al. showed similar rates of immediate postoperative symptom relief (100 %) and recurrence of symptoms (6.9 %) in a large series of children and adolescents who underwent laparoscopic decompression of the celiac trunk for MALS [58].

Table III: Popliteal Vascular Entrapment Forum classification for popliteal entrapment syndrome [60, 61]

Classification	Pathomechanism
Type I	Popliteal artery displaced medially by the medial head of the gastrocnemius muscle
Type II	Medial head of the gastrocnemius head attached laterally to the popliteal artery
Type III	Accessory muscle strings/fibrous bands arising from the medial head of the gastrocnemius muscle
Type IV	Popliteal artery compressed by muscle strings/fibrous bands arising from the popliteal muscle
Type V	Entrapment of the popliteal vein
Type VI	Other variants
Type F	Functional entrapment

Endovascular treatment by means of celiac artery stenting without prior release of external compression is not recommended due to the risk of stent fracture/crush [46].

Popliteal entrapment syndrome

Definition and epidemiology

PES describes a group of conditions characterised by clinical symptoms arising from compression of the popliteal artery, the popliteal vein, and/or the tibial nerve in the popliteal fossa by surrounding musculoskeletal structures [59–62]. Although anatomic abnormalities obviating the course of the popliteal vessels have been reported much earlier, the term PES was first introduced in 1965 [63]. The prevalence of PES in the general population is basically unknown. According to a systematic review the mean age at the time of diagnosis was 32 (range 20.7–41) years, with a median percentage of 83% male patients [62].

Pathoanatomy

A considerable heterogeneity in the use of different classification systems in published studies on PES must be acknowledged [62]. Nowadays broadly accepted is the classification system proposed by the popliteal vascular entrapment forum in 1998 (Tab. III) [60, 61].

Types I to IV refer to anatomical entrapment of the popliteal artery. The basis for PES types I–III is an abnormal medial migration of the medial head of the gastrocnemius muscle during embryogenesis, resulting in medial deviation and/or compression of the popliteal artery (Fig. 6,7). Type IV (popliteal artery entrapment by the popliteal muscle) can be explained by the persistence of a segment of the primitive axial artery during embry-

nal development. A detailed description of the embryological fundamentals of PES is provided elsewhere [60,

61]. Repetitive compression may lead to intimal damage, mural thrombus formation, stenosis or aneurysm

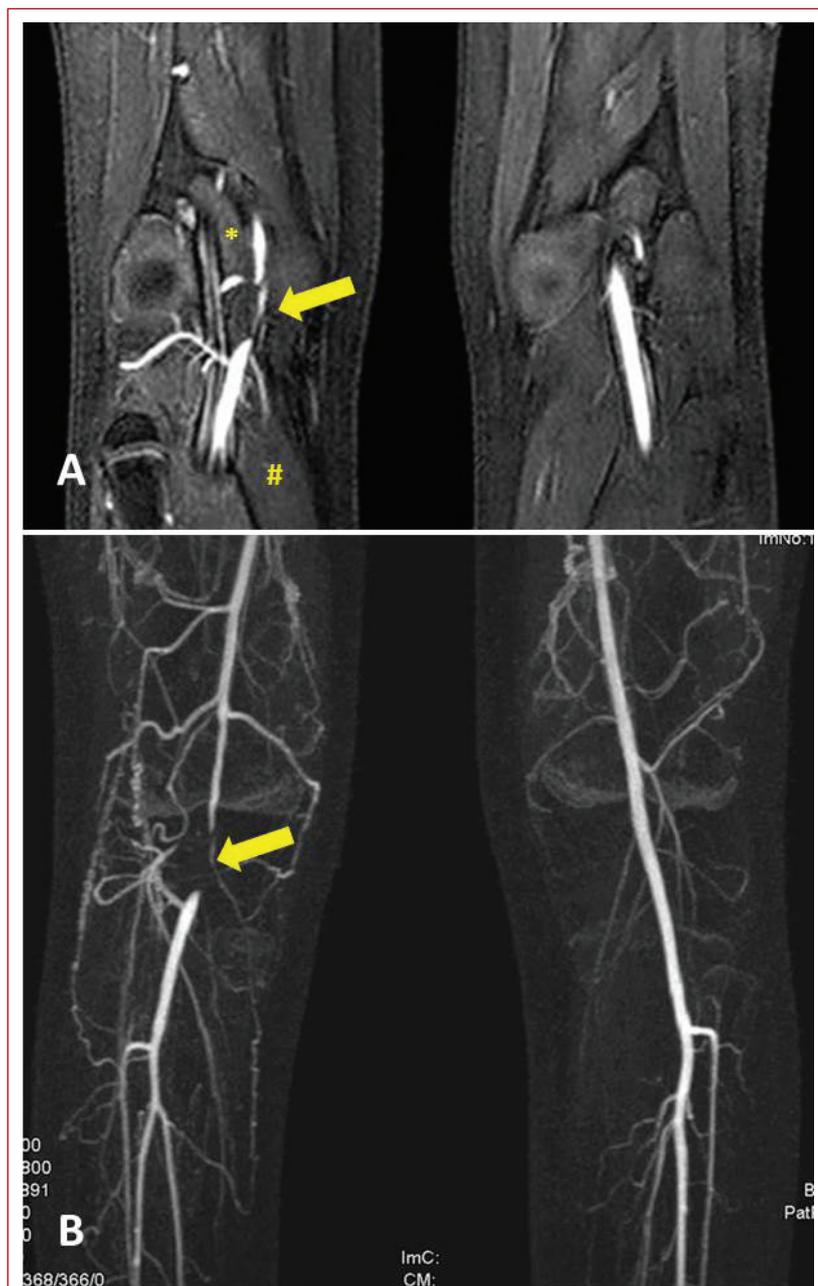


Figure 6: Eleven-year old girl presenting with acute occlusion of the right popliteal artery. Magnetic resonance -imaging documents medial deviation of the occluded artery and compression between the medial head of the gastrocnemius muscle (#) and the semimembranosus muscle (*), corresponding to popliteal entrapment syndrome (PES) type I. The left popliteal artery has a regular anatomical course.

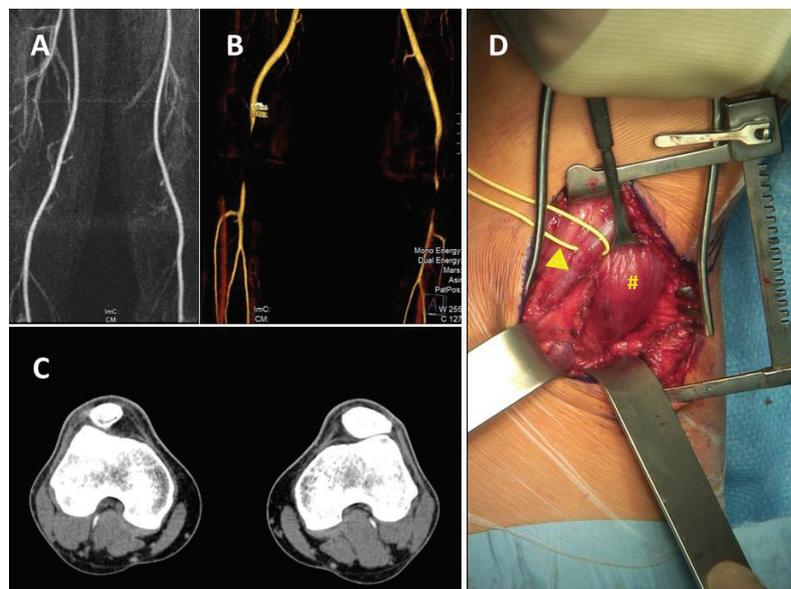


Figure 7: Young male patient complaining of bilateral calf claudication. MR-angiography without provocative manoeuvres reveals no pathological findings (A). Computed tomography-angiography performed during plantar flexion shows filiform narrowing of the popliteal artery on both sides (B,C). Intraoperatively, an abnormal lateral insertion of the hypertrophied medial head of the gastrocnemius muscle (#) is confirmed, corresponding to popliteal entrapment syndrome (PES) type II (D).

formation. Structural arterial damage, as seen in approximately 10 % of patients, carries the inherent risk of local thrombotic occlusion and distal embolisation.

Functional entrapment of the popliteal artery occurs in the absence of anatomical abnormalities and is found in about one out of four PES-cases [62, 64]. Hypertrophy of the gastrocnemius muscles, as frequently seen in competitive athletes and persons abusing anabolic steroids, has been attributed to be causative for positional compression [59]. However, Hoffmann et al. observed no differences in the rate of significant popliteal artery compression during plantar flexion in normally vs. highly trained individuals [65]. In this and other studies, some degree of popliteal artery compression with maximal plantar flexion was found in 53

to 88 % of asymptomatic individuals, clearly indicating that the phenomenon per se does not imply a clinically relevant disease [65, 66]. If the popliteal vein is additionally involved, the pathology is classified as type V. The true rate of popliteal vein entrapment, with or without concomitant arterial involvement, is basically unknown, as is the rate of tibial nerve compression in the popliteal fossa.

Clinical presentation

Among other disorders such as large vessel vasculitis, persistence of the sciatic artery, adventitial cystic disease and iliac artery endofibrosis, PES is an important differential diagnosis of symptomatic lower leg ischemia in younger patients. The absence of cardiovascular risk factors and an athletic habitus should heighten the index of suspicion. Most patients complain

of intermittent claudication during walking or running, but acute limb ischemia secondary to local thrombosis or distal embolism occurs in 11 % of symptomatic limbs [62]. If the diagnosis is initially missed, affected limbs are at risk of developing critical limb ischemia due to recurrent embolism. The frequency of bilateral symptomatic PES varies considerably between studies (0–100 %, median 38 %) [62]. Diagnosis of unilateral symptomatic PES should necessarily prompt investigation of the asymptomatic contralateral leg.

The main symptom of venous entrapment is calf swelling, whereas tibial nerve entrapment typically goes along with pain in the popliteal fossa and paresthesia/numbness of the foot.

In a systematic review, the median duration of symptoms until the correct diagnosis could be established was 12 months. Noteworthy, a correlation between duration of symptoms and the presence of irreversible structural vascular damage could not be established [62].

The neuromuscular symptoms (calf cramps, plantar paresthesia) of functional PES may resemble those of chronic recurrent exertional compartment syndrome (CRECS), a disorder also typically occurring in young physically active adults [64]. By contrast to classical claudication, lower limb pain in CRECS usually persists over hours and days after exercise [67].

Diagnostic approach

As in other vascular compression syndromes, the diagnostic approach has not been standardised. The first diagnostic step is non-invasive vascular laboratory testing, including systolic ankle pressure measurement, pulse volume recording, and colour duplex sonography. When performed with adequate provocation manoeuvres by means of forced active plantar

flexion, sensitivities between 90 and 100 % have been reported for each of the aforementioned techniques in a limited number of studies which used surgical confirmation as the reference standard [62]. Colour duplex sonography offers the opportunity to depict both morphological changes of the vascular structures (focal wall thickening, stenosis with post-stenotic aneurysm, thrombotic occlusion) and haemodynamic changes provoked by forced active plantar flexion. In some cases, the underlying anatomic abnormality may also be detected, for instance a muscular slip between the popliteal artery and vein [68]. Treadmill testing is useful for further characterisation of the functional relevance of the disorder when presenting with intermittent claudication.

In recent years, cross-sectional imaging techniques (MR- and CT-angiography) have been increasingly using in the diagnostic workup of suspected TOS. These techniques allow excellent visualisation of the popliteal fossa's anatomy and detection of abnormal musculotendinous structures (Fig. 6, 7). In neutral position, structural abnormalities of the popliteal vessels and, in PES type I, a medial deviation of the artery can be found. Repeated imaging during forced plantar flexion has been shown to have excellent sensitivity for the diagnosis of PES [62]. MRI has the advantage of being free of radiation exposure, but movement artefacts during image acquisition in provocation position may limit its diagnostic accuracy [59].

Traditionally, PES has been diagnosed by conventional digital subtractive arterio- or venography with provocation of vascular compression (forced active plantar flexion). Although still considered the reference imaging method, conventional angiography do not provide anatomical information on the perivascular structures responsible for compression, and thus

should be reserved for cases remaining ambiguous despite previous cross sectional imaging studies or when endovascular treatment of acute leg ischemia is intended.

Popliteal artery occlusion, found in a median of 24 % of patients at the time of diagnosis [62], may hamper the diagnostic efforts to uncover a vascular compression syndrome. In these cases, evidence of popliteal artery compression on the contralateral leg may further underline the suspected diagnosis of PES.

Treatment

It is generally accepted that anatomical PES requires surgical treatment, including decompression of the popliteal vessels and, if structural vessel damage has occurred, arterial or venous revascularisation [59–62]. This recommendation, however, is mainly based on evidence coming from retrospective cohort studies. Interpretation of these studies is limited by several factors, including differences in cohort characteristics and the lack of standardised outcome measures. Indeed, half of the studies included in a systematic review did not clearly describe symptom resolution after surgery [62].

For surgical treatment, either a dorsal or a medial approach can be used. The choice between both techniques is primarily guided by the presence or absence of structural vessel damage. In the absence of arterial injury, the posterior access is more favourable as it allows excellent visualisation of the muscular/ligamentous structures being causative for entrapment [61]. There is no consensus on whether or not to perform reattachment of the medial head of the gastrocnemius muscle to the medial femur condyle after transection [62].

When arterial damage has occurred, standard techniques of reconstruction must be employed in addition to surgical decompression. In this

situation, a medial approach is preferable, facilitating harvesting of the great saphenous vein and also offering quicker return to physical activity [61]. In case of popliteal artery wall lesions without thrombotic occlusion or distal embolic occlusions, interposition of a short popliteo-popliteal saphenous vein graft is the method of choice. When acute thromboembolic occlusions of the below the knee arteries are present, consideration should be given to catheter-directed thrombolysis in order to improve the arterial outflow. Subsequent surgical revascularization and decompression of the popliteal artery are mandatory, as otherwise recurrent ischemia is very likely. In case of popliteocrural obstructions not approachable by endovascular techniques, femorocrural vein bypass surgery is required. In the young, otherwise healthy patients, the saphenous veins are usually eligible as graft material. Patency rates have been assumed to be generally better than in arteriosclerotic peripheral arterial disease, but Sinha et al. observed a median failure rate of 27 % (range 0–88 %) in their systematic review [62].

Treatment success as indicated by postoperative resolution of symptoms was reported in a median of 77 % (range 70–100 %) of patients with symptomatic arterial compression and in 53 % (range 48–57 %) of patients with symptomatic venous compression [62]. Minor or major amputations had to be performed only in exceptional cases. Data on the long-term outcome of revascularisation operations in the treatment of PES are scarce. In the currently largest retrospective cohort study, comprising 88 limbs of 48 patients, the primary patency rate of 15 reversed saphenous vein grafts was 100 % after a median follow-up of 4.2 years (range 1–10 years), as was the primary patency rate of the popliteal arteries in all limbs managed

Table IV: Rare vascular compression syndromes [73-80]

Syndrome	Pathoanatomy	Symptoms	Diagnosis	Treatment
Eagle syndrome	Internal carotid artery compression by an abnormally elongated styloid process	Neck pain, dysphagia, cranial nerve palsies, Horner's syndrome. Rare: transient ischemic attack and stroke in the anterior cerebral circulation	Intra-arterial angiography with provocative manoeuvres; spiral CT of the head and neck with 3D-reconstruction	Resection of the abnormal structure(s); Anticoagulation with or without surgical revascularization in case of structural arterial damage.
Rotational vertebral artery ischemia (Bow hunter's syndrome)	Cervical spine pathologies, e.g. osteophytes and disc prolapse, leading to positional vertebral artery compression	Dizziness or Syncope occurring with head rotation. Rare: posterior circulation stroke secondary to vertebral artery dissection/occlusion ("Bow hunter's stroke")		
Langer's axillary arch	Accessory muscular band crossing the axilla, found in up to 8% of the general population. Axillary vein obstruction and or thrombosis is a very uncommon complication.	Symptoms of intermittent upper extremity venous congestion; Upper extremity deep venous thrombosis	Colour duplex sonography; venography; magnetic resonance imaging	Resection of the aberrant muscle bundle
Axillary artery / posterior circumflex humeral artery (PCHA) injury	Repetitive compression of the third portion of the axillary artery or the PCHA against the head of the humerus	Typically occurring in overhead athletes (baseball pitchers, volleyball players). Arm claudication secondary to axillary artery occlusion; acral ischemia due to peripheral embolism from posterior circumflex humeral artery aneurysms	History of professional overhead sports activities; Intra-arterial angiography	Staged revascularization including thrombectomy or thrombolysis, exclusion of PHCA aneurysm and/or partial axillary artery resection with vein graft interposition
Brachial artery compression syndrome (Popeye syndrome)	Compression of the brachial artery in the cubital fossa due to hypertrophy of forearm muscles or the lacertus fibrosus	Intermittent claudication of the forearm; hand and finger ischemia following local arterial thrombosis with or without peripheral embolism	Muscular men with hypertrophied forearm muscles; pain and disappearance of the radial pulse with forearm pronation and resisted elbow flexion; colour duplex sonography and intra-arterial angiography with provocative manoeuvres	Surgical release of the lacertus fibrosus
External iliac artery endofibrosis	Multifactorial compression and tethering of the external iliac artery (main factors: repetitive hip hyperflexion, psoas muscle hypertrophy), resulting in vessel tortuosity, kinking and endofibrosis.	Typically occurring in competitive cyclists. Thigh pain, weakness or cramps at (sub)maximum exercise or during hip hyperflexion, predominantly left leg.	Competitive athletes, particularly professional or top amateur cyclists. Iliac artery bruit; ankle pressure drop after cycle-ergometer based exercise (standard treadmill testing usually without abnormal results); colour duplex sonography; magnetic resonance imaging with provocative manoeuvre (hip hyperflexion)	Adjustment of cycling posture, reduction of cycling intensity. Surgical or endovascular treatment should be avoided if possible. Surgical release of the iliac artery is the only invasive procedure supported by low level evidence.

Syndrome	Pathoanatomy	Symptoms	Diagnosis	Treatment
Adductor canal compression syndrome	Intimal damage with subsequent arterial thrombosis at the outlet of the adductor canal where the neurovascular bundle crosses the vastoadductor membrane	Unilateral superficial femoral artery obstructions occurring in younger male patients without obvious cardiovascular risk profile, particularly competitive athletes	Difficult with diagnostic imaging; evidence for external compression during surgical revascularization	Surgical decompression during femoropopliteal graft interposition
Dorsalis pedis artery entrapment syndrome	Compression of the dorsalis pedis artery by the extensor hallucis brevis tendon	Foot claudication, blue toe syndrome	Cessation of the dorsalis pedis artery Doppler signal during dorsiflexion of the foot	Transection of the extensor hallucis brevis tendon and transposition to extensor hallucis longus tendon
Left renal vein entrapment syndrome (Nutcracker syndrome)	Compression of the renal vein between the aorta and the steep-angled origin of the superior mesenteric artery (anterior nutcracker syndrome) or between the aorta and the vertebral column (posterior nutcracker syndrome in case of retroartical course of the left renal vein)	Left flank and abdominal pain; haematuria; clinical manifestations attributable to left renal-to-gonadal vein reflux, i.e. symptoms of pelvic congestion syndrome, varicocele, lower limb varices.	Urological investigations to confirm unilateral haematuria and rule out other sources of haematuria; colour duplex sonography; cross sectional imaging; retrograde venography with assessment of the reno-caval pressure gradient	Dependent on the severity of symptoms and patient's age: Surveillance vs. surgical or endovascular revascularization

by surgical decompression only (median follow-up 3.9 years) [69]. More recently, study groups from Japan and Korea published follow-up data after vein graft interposition/bypass surgery, demonstrating primary graft patency rates at 5 years between 74 and 100 % [70–72]. The patency rate was negatively influenced by outflow obstruction and increasing length of the bypass in the study by Kim et al. [70]. Of note, bypass occlusions in the very long-term (12 and 23 years after surgery) have been reported [71]. Aneurysmatic degeneration of venous interponates is another important complication in the long-term. In this case, persistent compression should be ruled out. These findings underscore the need for regular long-term surveillance at least of patients who underwent revascularisation procedures. Very limited data suggest that a conservative approach with cessation of intense sports activities may be an appropriate treatment approach in PES with functional entrapment [69].

No clear recommendations can be made concerning primarily asymptomatic patients with arterial compression. It seems reasonable to perform preventive decompressive surgery of the asymptomatic contralateral leg in patients with symptomatic PES to avoid arterial ischemic complications. Reflecting current clinical practice, this approach was applied in almost all studies reporting surgical treatment in PES [62].

Other vascular compression syndromes

As mentioned above, May-Thurner Syndrome and Hypothenar-Hammer Syndrome were subjected to dedicated reviews in recent issues of this journal [1, 2]. Therefore, we did not include either condition in the present work. A variety of other vascular compression syndromes has been reported. Related to the rarity of these disorders, evidence for diagnostic and therapeutic decision making at best

comes from retrospective case series [73–80]. This is particularly true for compression syndromes of the extremities, with less than 20 cases each reported in the literature. A concise overview is provided in Table IV. Recently, a contribution of external compression between the sternum and the aortic arch to the development of left innominate vein stenosis in haemodialysis patients has been suggested [81]. In addition to classic compression syndromes, resulting from entrapment of blood vessels by adjacent anatomical structures, abnormalities in the course or morphology of large and medium sized arteries may result in the compression of adjacent organs. Superior mesenteric artery syndrome (Wilkie's syndrome), for instance, is characterised by compression of the third portion of the duodenum by the narrow-angled superior mesenteric artery. Other examples are left main bronchial obstruction by large thoracic aortic aneurysms, and a right aberrant subclavian artery (arteria lusoria) leading to oesopha-

geal obstruction. Although primarily caused by a vascular disease process, the clinical manifestations implicate that patients with such disorders primarily present to other medical subspecialties.

Conclusions

Vascular compression syndromes pose a challenge to Vascular Medicine specialists for several reasons. Affected patients, typically young and middle-aged persons without severe chronic comorbidities, commonly experience severe impairment of quality of life. Overtreatment of individuals with positional vascular compression but no clearly established causal relation between clinical symptoms and vascular compression or vascular damage must be avoided. On the contrary, patients in whom the causality is obvious need proper and timely treatment to release subjective symptoms and to avoid complications, particularly limb ischemia in aTOS and PES. The complex nature of vascular compression syndromes is what makes the management of subjects suspected to suffer from or diagnosed with these disorders so challenging.

Conflicts of interest

There are no conflicts of interest existing.

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