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REVIEW

The Investigation and Management of the Abdominopelvic Vascular Compression Syndromes in Patients with Ehlers–Danlos Syndrome and Hypermobility Spectrum Disorder

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Objective: Abdominopelvic Vascular Compression Syndrome(s) (VCS) are rare disorders with diverse symptoms that appear to occur more frequently in patients with Ehlers–Danlos Syndrome (EDS) and Hypermobility Spectrum Disorder (HSD). The reported associations between EDS/HSD, Postural Orthostatic Tachycardia Syndrome (POTS), and Mast Cell Activation Syndrome (MCAS) further complicate the diagnosis and management of the VCS in this specific patient population. This review summarises the established literature on this complex topic, highlighting these relationships, with the aim to propose a framework for recognising and managing VCS among patients with EDS/HSD. Given the limited body of literature on this topic, we also aim to underscore the need for further research within this specific patient population.

Methods: A PRISMA-guided systematic review was conducted using PubMed and Ovid/Medline databases. VCS included Median Arcuate Ligament Syndrome (MALS), Superior Mesenteric Artery Syndrome (SMAS), Nutcracker Syndrome (NCS), and May–Thurner Syndrome (MTS). Given the limited number of studies, small cohort studies and case reports/series were also reviewed.

Results: Of 183 screened studies, 62 met the inclusion criteria. Only six studies directly addressed the VCS in EDS/HSD. Five discussed an EDS–POTS association, two described links between MCAS, POTS, and EDS, and five associated POTS with VCS. Only one study explored all four conditions.

Conclusion: Evidence suggests an association between EDS/HSD, VCS, POTS, and MCAS but remains limited. Underdiagnosis and delayed treatment are common and underscore the need for multi-disciplinary care. Invasive imaging and interventions appear generally safe in EDS/HSD, excluding vascular EDS, yet robust safety and outcome data and tailored diagnostic or treatment algorithms are lacking and require further investigation.

Keywords: Ehlers–Danlos syndrome, EDS, hypermobility spectrum disorder, HSD, median arcuate ligament syndrome, MALS, mast cell activation syndrome, MCAS, May–Thurner syndrome

Introduction

Ehlers–Danlos Syndrome (EDS) and Hypermobility Spectrum Disorder (HSD) are characterised by joint hypermobility, skin hyperextensibility, and tissue fragility.[1] EDS is a hereditary connective tissue disorder, and HSD is diagnosed when a patient meets some, but not all, criteria for EDS. The overall prevalence of EDS is 1 in 5000, with 13 subtypes. Hypermobile EDS (hEDS) is the most common (1 in 320), followed by classical EDS (1 in 20,000–40,000) and vascular EDS (vEDS) (1 in 100,000 or 4% of all EDS cases).[2,3] Diagnosis is mainly based on clinical criteria, with genetic testing available for 12 of the 13 subtypes (hEDS has no available genetic test). There are no genetic tests for HSD, which is thought to be polygenic similar to hEDS.

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Depending on the subtype, the inheritance pattern can be autosomal dominant, autosomal recessive, or de novo. For vEDS (characterised by even greater tissue fragility), genetic testing is required to confirm the diagnosis (autosomal dominant COL3A1 mutation).[4] However, certain clinical characteristics can indicate a diagnosis (eg translucent skin, thin vermilion of the lips, narrow nose, micrognathia, prominent eyes).[5]

Whilst not formally established, there is increasing recognition of an association between EDS/HSD, the Abdominopelvic Vascular Compression Syndrome(s) (VCS), Postural Orthostatic Tachycardia Syndrome (POTS) and Mast Cell Activation Syndrome (MCAS).

This association appears to arise from a shared pathophysiological basis: the connective tissue abnormalities of EDS/HSD result in increased tissue laxity and diminished structural support of the surrounding matrix, potentially making these patients more at risk of developing a VCS. Additionally, heightened vascular distensibility promotes venous pooling in the extremities and may precipitate a maladaptive autonomic adrenergic response with associated dizziness and palpitations, as seen in POTS.

POTS (a form of dysautonomia) has been described in >50% of patients with EDS/HSD.[6] Additionally, it has been found that 1 in 3 patients with MCAS have hEDS, and up to 30% of patients with POTS have MCAS.[3,7] While numerous diagnostic criteria exist, it is notable that 17 of the 48 recognised symptoms of MCAS overlap with 33 symptoms associated with POTS.[7] Furthermore, symptoms of POTS overlap significantly with those of the VCS. This substantial concurrence lends support to the observed comorbidity between these conditions but also underscores the potential for attribution bias and diagnostic overshadowing.[7] It has been postulated that excessive mast cell activation and their release of vasodilatory substances such as histamine and tryptase may exacerbate POTS and the symptoms of the VCS by intensifying venous pooling.

Pelvic Congestion Syndrome (PCS), which contributes to 30–40% of patients with chronic pelvic pain of unknown cause can arise as a result of VCS, especially NCS and MTS.[8,9]

There appears to be a complex interplay between these conditions, each potentially exacerbating the other. Significant symptom overlap makes differentiating between them challenging, resulting in diagnostic uncertainty and delayed diagnosis.[7] While hypotheses about the underlying pathophysiological link between these conditions exist, robust scientific evidence of their association is lacking. Emerging genomic studies are beginning to reveal the molecular foundations underlying hypermobility, indicating that these syndromes likely represent a heterogenous group of connective-tissue disorders with variants across numerous structural and regulatory genes, holding promise that molecular stratification may ultimately clarify these relationships.[10]

This review underscores the importance of clarifying the interplay between hypermobility, VCS, MCAS, POTS and PCS, and the importance of having a framework for the diagnosis and management of the VCS in EDS/HSD patients by also recognising and treating these co-existing conditions.

Methods

Associations between VCS, EDS/HSD, MCAS, and POTS were screened using the PubMed and Ovid MEDLINE medical databases, including keywords and Medical Subject Headings (MeSH). This screen included imaging studies and interventions used for the VCS in this patient population. We searched for studies in Australia and internationally.

"Renal vein compression" was used as an alternative to NCS. "Joint instability" and "hypermobility" were used as alternatives to HSD. For MTS, other terms used in the search were "iliocaval venous compression syndrome", "Cockett syndrome", and "venous spur".[11] For MALS, "coeliac artery compression syndrome", "Dunbar Syndrome", and "coeliac axis syndrome" were used as alternative terms. For SMAS, "Cast Syndrome", "Wilkie Syndrome", "arterio-mesenteric syndrome" and "chronic duodenal ileus" were used as alternative terms.[12]

Given the limited published research on this topic, we included case studies/series, small observational, and retrospective cohort reports in our review, which made up the majority of the search results. No systematic reviews or meta-analyses were found on this topic. We included articles that commented on the symptomatology, diagnosis, imaging, and treatment of VCS with regard to POTS, MCAS, and PCS and any articles that discussed the association and underlying pathophysiology of these conditions and EDS/HSD (Figure 1). Excluded articles were abstract only and non-English articles, and those that focused on other vascular manifestations of tissue laxity such as aneurysmal change.

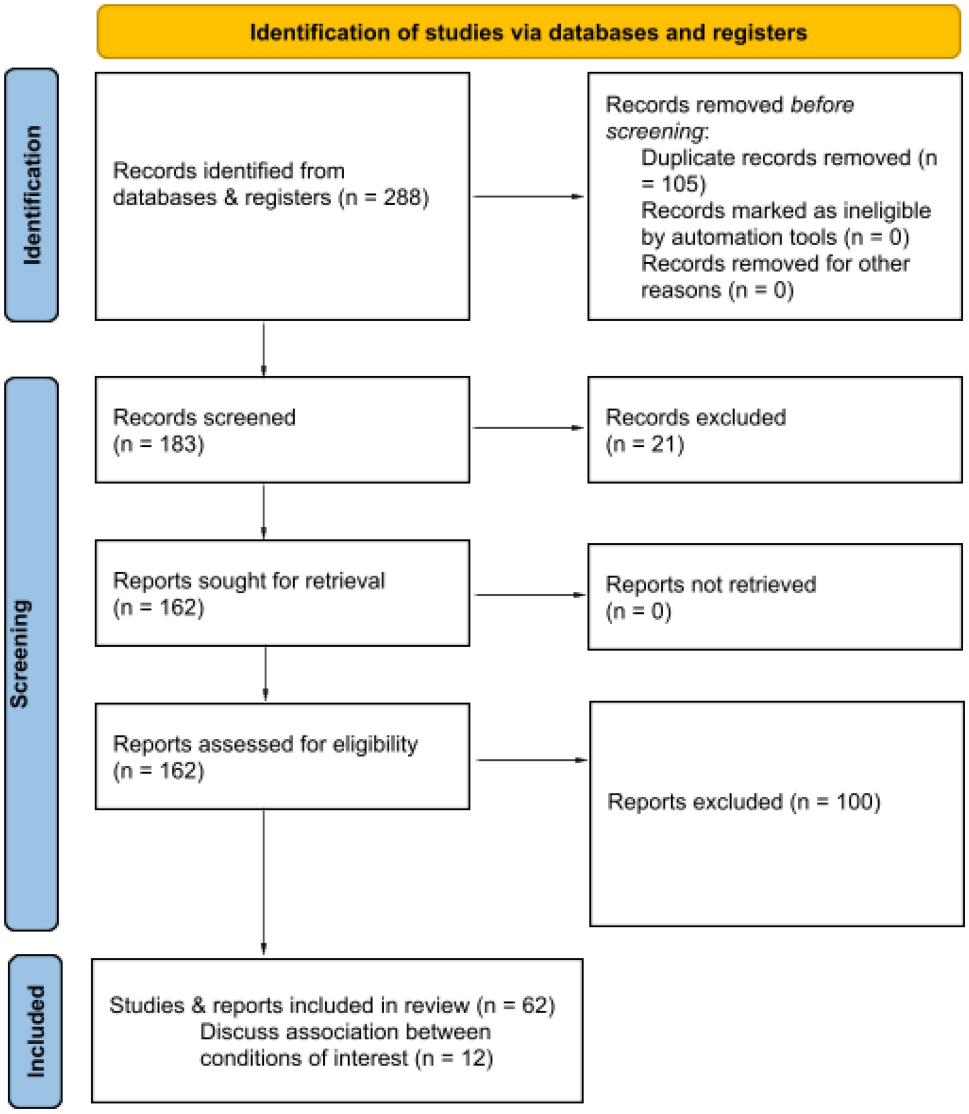

Figure 1 Systematic Review of the Literature on Vascular Compression and EDS/HSD.

Titles and abstracts were independently screened by two reviewers using the Covidence-Screening Tool. Full-text articles were assessed for their eligibility. Data were independently extracted by two reviewers using a standardised form.

Results

Association Between EDS/HSD, the VCS, and POTS, MCAS and PCS

None of the studies from the search strategy revealed the exact prevalence of VCS, POTS, and MCAS in the EDS/HSD population. However, our findings indicated a strong association between them which is considered to be greater than that in the general population. The most obvious link lies in their overlapping or shared symptoms. However, tissue laxity, dysregulation of autonomic nerves, and venous pooling appear to be the relevant pathological processes that may contribute to and exacerbate each condition.

The VCS are a group of rare disorders in which a blood vessel compresses or is itself compressed by an adjacent structure. Frequently, VCS result from a diminished angle between the vessel and an adjacent structure, secondary to the loss of abdominal fat and/or supportive connective tissue between these structures. Increased tissue laxity and fragility caused by alterations in collagen are likely to underlie the association between vascular compression syndromes and EDS/HSD, causing hypermobility of the abdominopelvic viscera and altering anatomical associations between intra-abdominal structures.[13]

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The spectrum of clinical presentations and non-specific symptoms mean that VCS are frequently under-recognised or seen as diagnoses of exclusion, causing delays in treatment.[14–16] Table 1 outlines the aetiology, risk factors, and clinical features of common VCS.

POTS, defined by a heart rate increase of ≥ 30 beats per minute within 10 minutes of standing without orthostatic hypotension,[11] has been found in up to 50% of patients with EDS/HSD.[11,16,29] Collagen abnormalities in EDS/HSD can affect nerves as well as the blood supply to nerves which can result in POTS. Symptoms of POTS include palpitations, dizziness, "brain fog", syncope, dyspnoea and gastrointestinal upset (eg gastroparesis with associated nausea and vomiting).[11]

The symptoms of POTS can overlap with those of VCS, making the diagnosis challenging. Dysautonomia in EDS can affect vascular tone, which may exacerbate the effect of compression on the blood vessels in the VCS (Figure 2).[8] VCS can lead to a reduction in venous return and aggravate POTS (Figure 2).[2] This is indicated by patients in some studies with complete resolution of POTS after surgical intervention for underlying VCS.[6,12,17,30]

NCS, MALS, and SMAS can worsen orthostatic intolerance postprandially, likely through reduced venous return and irritation of autonomic nerves such as the coeliac plexus.[9] In POTS, gastrointestinal symptoms may be explained by active autoimmune muscarinic antibodies which cause an autonomic imbalance.[18] A cycle of aggravated symptoms can arise when POTS and VCS co-occur with worsening chronic fatigue syndrome, dizziness, and gastro-intestinal symptoms.[8]

MCAS occurs due to abnormalities in mast cells or abnormal release of their mediators. Patients may experience muscle pain, fatigue, headache, itching, urticaria, reflux, dyspnoea, chills, paraesthesia, presyncope/syncope, eye irritation and nausea.[18] Gastrointestinal symptoms are common in MCAS, POTS, and the VCS.[18] The gastrointestinal symptoms of MCAS have been postulated to be due to inappropriate release of mast cell mediators.[18]

MCAS can exacerbate pre-existing inflammation around compressed blood vessels in the VCS, which can result in worsening of VCS symptoms (Figure 2).[8] MCAS can also worsen the symptoms of POTS owing to vasodilation induced by mast cell mediators (Figure 2).[18] Given the overlapping symptoms of POTS, MCAS and VCS clinicians should search for (and manage) each of these conditions when caring for patients with EDS/HSD. As the symptoms of these conditions overlap, there are delays in the diagnosis of each condition.[18]

PCS is an underdiagnosed and poorly understood condition with a sparsity of high-quality literature to guide treatment.[19] Symptoms include irritable bowel syndrome, vulvar varicosities, dyspareunia, pelvic pain worsening upon standing and lower-limb venous pathology.[19–21] Essentially, there is pelvic venous insufficiency with reflux or obstruction of the gonadal or internal iliac veins.[19]

The venous pooling that occurs in POTS, may result in or exacerbate PCS however it is unknown what percentage of patients with PCS have POTS.[4] According to a case report by Wilhelmstoetter et al[22] of a patient with EDS, POTS, NCS, and MTS, who was found to have PCS on a venogram with intravenous ultrasound (IVUS), the authors stated, that

> There is a significant lack of literature describing guidelines on how to manage this special patient population. To date, there are no reports of managing patients with EDS who are diagnosed with PCS and NCS.[22]

An observational study of 169 patients with hEDS or HSD found that 72.8% presented with multiple types of VCS.[23] Due to abnormalities in collagen and the resulting increased vessel elasticity in EDS/HSD, blood vessels are at higher risk of compression by surrounding structures; therefore, these patients are more likely to develop multiple VCS.[8,13] Stubberud et al described a unique case in which a patient with hEDS and POTS had NCS, MTS, MALS and SMAS.[17] This makes it important for all four VCS to be on the differential list for EDS/HSD and/or POTS patients presenting with the clinical features outlined in Table 1.

A further study found that 69% of patients with POTS had significant (>50%) left common iliac vein compression (ie May–Thurner syndrome).[24] Another study of 73 patients with POTS at a single healthcare centre found that 77% of them had pelvic venous compression (iliac vein, renal vein or both).[25] A survey of 242 patients with MALS found that 53% had dysautonomia.[26] Concomitant NCS has been reported in up to 18% of patients with PCS.[27] Another study found that 18% of patients with PCS had "loose skin or lax joints", 6% had EDS, and 9% had POTS.[15] The prevalence of other VCS in POTS is unknown but is postulated to be greater than that in the general population, given that POTS is highly associated with EDS/HSD.[11]

Table 1 The Aetiology, Risk Factors and Clinical Features of the VCS

VCS	Aetiology	Risk Factors	Clinical Features
NCS	**Anterior**: Left renal vein compression between SMA and aorta;[17] <90 degree angle between structures[17] **Posterior**: Left renal vein compression between aorta and vertebral body[18]	• Correction spinal surgery for scoliosis[19] • Renal ptosis[19] • Major weight loss[19] • Tumour compression[19]	• Proximal bowel obstruction[19] • Chronic abdominal/flank pain[17,19] • Food intolerances[19] • Weight loss; malnutrition[19] • Varicose veins (VV)[19] • Varicocele (usually left sided)[19] • PCS[17,19] • Orthostatic proteinuria[19] • Orthostatic haematuria (micro/macroscopic)[20] • Orthostatic intolerance including POTS[19] • Headache[1,19,21] • Syringomyelia/myelopathy[22]
MTS	Left common iliac vein compression by right common iliac artery[23]	• Female gender[11] • Age: 20–40[11] • Multiparity[11] • Postpartum[11] • Oral contraceptive use[11] • Scoliosis[11] • Lumbar vertebral degeneration[11] • Vertebral disc protrusion[11] • Spondylolisthesis[11] • -Osteophytes[11]	• DVT[11,24] • PE[23] • Chronic venous insufficiency (VV, ulceration, venous claudication)[23] • Recurrent superficial vein thrombosis[23] • PCS[23] • Phlegmasia cerulea/alba dolens (rare)[11]
MALS	Coeliac artery and/or coeliac plexus compression by the MAL[25]	• Females (4:1 ratio)[26] • Age: 40–60[26] • Low BMI[26]	• Chronic post-prandial and/or exercise induced epigastric pain[26] • Nausea/vomiting[23,25] • Weight loss; malnutrition[23] • Relief with leaning forward or knee-chest position[26] • 35% of cases have an epigastric bruit that is louder in expiration[26]

(*Continued*)

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Table 1 (Continued).

VCS	Aetiology	Risk Factors	Clinical Features
SMAS	3rd part of duodenum is compressed between SMA and abdominal aorta[18] Can co-exist with anterior NCS	• Congenital anatomically low SMA origin or high attachment of angle of Treitz[27] • Reduced aortomesenteric angle[18] • Severe weight loss (loss of fatty tissue around the surrounding structures)[18] • Acquired: burns, eating disorders, spinal trauma, corrective surgery for scoliosis, malignancy, malabsorption disorders[12,18]	• Concurrent MALS[28] • Proximal bowel obstructive symptoms[27] • Post-prandial epigastric pain • Early satiety • Abdominal distension • Nausea; vomiting • Weight loss; malnutrition[27] • GORD[27] • Symptom relief in prone, knee-chest or lateral decubitus position[27] • High-pitched bowel sounds; succession splash[27] • Gastric perforation, gastric pneumatosis, obstructing duodenal bezoar (rare)[27]

Abbreviations: VCS, Vascular Compression Syndrome; NCS, Nutcracker Syndrome; MTS, May–Thurner syndrome; MALS, Median Arcuate Ligament Syndrome; SMAS, Superior Mesenteric Artery Syndrome; SMA, Superior Mesenteric Artery; EDS, Ehlers–Danlos Syndrome; HSD, Hypermobile Spectrum Disorder; PCS, Pelvic Congestion Syndrome; POTS, Postural Orthostatic Tachycardia Syndrome; MAL, Median Arcuate Ligament; VV, varicose vein.

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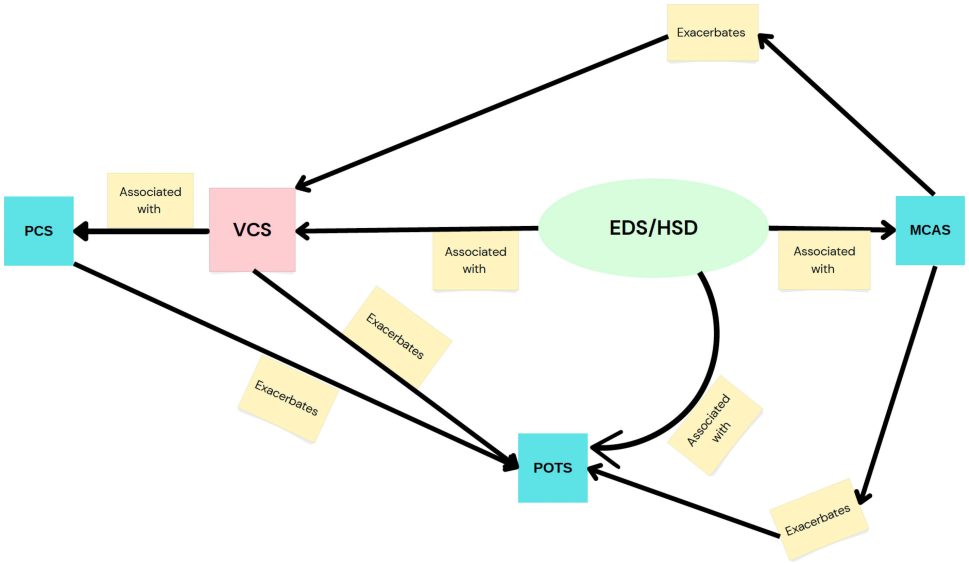

Figure 2 VCS and its Association with EDS/HSD, POTS, MCAS and PCS.
Abbreviations: EDS, Ehlers–Danlos Syndrome; HSD, Hypermobility Spectrum Disorder; MCAS, Mast Cell Activation Syndrome; POTS, Postural Orthostatic Tachycardia Syndrome; VCS, Vascular Compression Syndrome; PCS, Pelvic Congestion Syndrome.

Regarding MCAS, there is a lack of studies examining the prevalence of VCS in this condition. One study found that 23% of patients with MALS had MCAS.[26] Given the strong association of MCAS with EDS/HSD and POTS it is likely that the prevalence of the VCS in patients with MCAS is also greater than the general population.[3,7] In a limited case series of 12 patients with symptomatic and radiological MALS, 50% had POTS, 42% had MCAS, hEDS was found in 42% and other VCS were seen in 33%.[13]

Despite the lack of robust large-scale population studies in this field, the literature highlights common underlying pathological processes and reported symptoms of POTS, MCAS, PCS, and VCS. Therefore, it is crucial to consider each of these conditions in patients with EDS/HSD with this constellation of symptoms. In particular, vascular EDS can lead to more severe VCS symptoms owing to the heightened blood vessel fragility observed in this subtype.[8]

Imaging for VCS in EDS/HSD Patients

Table 2 indicates the first- and second-line imaging modalities for the detection of VCS in the general population, which did not differ between the EDS/HSD groups. Despite vascular fragility, invasive imaging for the EDS and HSD groups is generally considered safe; however, there are no large cohort studies on long-term safety outcomes or adverse effects.[3]

Table 2 Imaging Modalities for the Detection of VCS in the EDS/HDS Population

VCS	First Line Imaging Modality	Second Line/Further Imaging Modalities
NCS	Doppler ultrasound of renal vein flow velocity or CT/MR abdominal contrast angiogram/venogram[13,28]	Definitive diagnosis is with direct contrast venography and/or intravascular ultrasound (IVUS)*[20]
MTS	Venous Doppler ultrasound	CT/MR venography* Catheter-based venography* IVUS with venography*
MALS	CT or MR contrast angiogram of the abdomen*	CT/MR angiography with three-dimensional reconstruction combined with duplex inspiratory and expiratory views* Catheter-based arteriography with inspiratory and expiratory views*

(*Continued*)

Table 2 (Continued).

VCS	First Line Imaging Modality	Second Line/Further Imaging Modalities
SMAS	Diagnosis of exclusion: if high suspicion, then CT/MR contrast angiogram of the abdomen*	Upper gastrointestinal series with barium contrast Trans-abdominal ultrasound

Notes: *Catheter-based diagnostic procedures with injected contrast in patients with vEDS are generally discouraged because of the risk of arterial tears or dissection. Even injection pressure from angiogram can cause arterial aneurysms in these patients. Invasive imaging is only used to identify life-threatening bleeding sites in patients with vEDS before surgery.

Catheter-based diagnostic procedures with injected contrast in patients with vEDS are generally discouraged (because of the risk of arterial tears or dissection).[1,28] The injection pressure can cause arterial aneurysms in these patients.[7] Invasive imaging is only used to identify life-threatening bleeding sites in patients with vEDS before surgery.[1,28] Even routine elective surgery of any kind, including procedures such as colonoscopy or endoscopy, is discouraged in vEDS patients.[7]

NCS

Doppler ultrasound is often the first imaging modality used.[28,31] The ability of ultrasound to show left renal vein compression is dependent on the Doppler angle and sample volume of the image (ie operator dependent).[32] The diagnostic classification varies, but it is generally accepted that an angle between the aorta and SMA of less than 35° in the sagittal plane is sufficient for diagnosis.[33] The sensitivity of ultrasound is 69–90% with a specificity of 89–100%.[32]

CT can show pelvic varices or gonadal vein dilatation, provide more information on the relationship between the involved anatomical structures, but does not allow the measurement of flow direction and velocity and exposes patients to radiation.[34] Although MR-based imaging modalities offer a radiation-free alternative, they can be expensive; for example there is a limited rebate in Australia, and this imaging modality is usually performed at private centres.

Intravascular ultrasound (IVUS) is another diagnostic modality that can concurrently assist in the management of NCS by confirming stent placement and patency. With contrast venography and/or IVUS, renocaval pressure gradient measurements can be performed.[28,31,35] Pelvic varicosities and reflux through the adrenal or gonadal veins have also been reported.[28,31]

MTS

Various imaging modalities have been used to diagnose May–Thurner syndrome, including Doppler ultrasonography, CT and MR angiography.

Venous Doppler ultrasound is mainly used to investigate DVT, including iliocaval DVT, but can reveal iliac vein stenosis and post-stenotic turbulence if the correct angle of insonation is used.[16,36] Doppler ultrasound, although operator-dependent, has high sensitivity and specificity for detecting proximal DVT, but difficulties can occur with obesity or overlying gas.[6] If there is no DVT but a high index of suspicion for MTS, reflux duplex ultrasonography can be used.[6]

CT/MR venography have a >95% specificity and sensitivity.[6,20] These modalities may have a superior ability to detect other causes of extrinsic venous compression. CT angiography allows the definition of the extent of thrombus extension, as well as visualisation of collateral circulation; however, suboptimal contrast opacification may limit the diagnostic value.[6] MR venography provides better images of spinal and pelvic structures and identifies lumbar vertebral degeneration, disc protrusion, spondylolisthesis, and osteophytes, which can be associated with MTS (Table 1).[6]

Catheter-based venography with invasive haemodynamic pressure measurements remains the gold standard for diagnosis and allows for optimal identification and classification of stenotic lesions in MTS.[6,37] Repetitive pressure injury to the venous endothelium in MTS from the compressing right iliac artery results in intimal hypertrophy, endovenous fibrous band formation, and venous spur formation.[6,37]

An adjunctive imaging modality to further define the intraluminal effects of MTS is intravascular ultrasonography (IVUS).[37] IVUS can show the precise morphology of a venous "spur", the distribution and severity of pathology, and the spatial relationships of anatomical structures.[6] IVUS has a sensitivity and specificity of > 98% for MTS.[6] It can be used during treatment to assist with accurate stent placement and can be helpful for follow-up to identify the severity of potential in-stent restenosis.[6,7]

MALS

CT or MR contrast angiography of the abdomen is usually the first-line imaging modality used to investigate coeliac artery stenosis.[26] Splanchnic artery aneurysms or post-stenotic dilatation may also be found.[26] Figure 3 shows MALS, as observed on a sagittal CT angiogram (arterial phase). There is a tight stenosis at the origin of the coeliac trunk with evidence of compression from the MAL above. Distal flow in the coeliac artery is maintained.

If CT/MR angiogram is negative but there remains a high index of suspicion, three-dimensional reconstruction combined with duplex inspiratory and expiratory views can be used.[26] During expiration, compression of the coeliac artery increases, corresponding to increased systolic velocities in the coeliac artery on duplex ultrasonography.[26,38] This is an advanced imaging modality which may be difficult to access.

Provocative investigations can also be performed, such as mesenteric angiograms with papaverine injection (to reduce symptoms of mesenteric ischaemia) or coeliac plexus blockade (to determine if abdominal pain improves.[25,39] A positive response to provocative investigations can be used to screen or select patients who are more likely to have successful outcomes and symptom reduction following MALS interventions.[13,25,39]

A last-line diagnostic imaging modality is catheter-based arteriography with inspiratory and expiratory views, which allows simultaneous venous angioplasty or stenting, as discussed further below.[26]

SMAS

Plain abdominal X-rays are useful for investigating acute causes of abdominal pain but have little merit in the work-up of EDS/HSD patients with chronic abdominal pain and gastrointestinal symptoms.[40]

Functional scans, such as the upper gastrointestinal series with barium contrast or transabdominal ultrasound with positional manoeuvres, are higher-yield modalities for identifying SMAS. Upper gastrointestinal series with barium contrast demonstrate contrast passage delay from the duodenum into the small bowel, sometimes with obstruction at the third portion of the duodenum.[40] The stomach and duodenum may be dilated and show retention of contrast.[40]

The most sensitive measure for diagnosis is an aortomesenteric artery angle ≤25°on transabdominal ultrasound, especially if the aortomesenteric distance is ≤8 mm.[40] Positional manoeuvres, such as the lateral decubitus or standing position, may reveal abnormalities.[40]

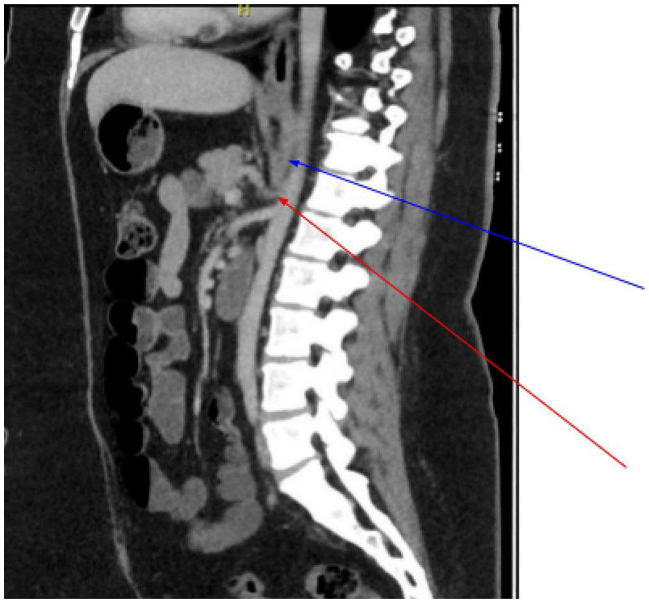

Figure 3 Median Arcuate Ligament Syndrome (MALS) on CT Angiogram in a patient with EDS. The blue arrow points to the median arcuate ligament. The red arrow shows the tight stenosis of the coeliac trunk as it passes under the MAL.

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CT/MR contrast angiogram of the abdomen also shows a reduction in the aortomesenteric angle, and efforts should be made to assess the amount of retroperitoneal and intra-abdominal fat, the presence of high attachment of the ligament of Treitz on the duodenum, and/or a low origin of the SMA, and other abnormalities of the SMA.[40]

Management of VCS in EDS/HSD Patients

The management of VCS in EDS/HSD patients is the same as that in the general population, with the exception of vEDS, where invasive procedures are not performed given the increased vessel/organ fragility.

NCS

Treatment of NCS is non-operative if there are minimal symptoms.[34] If there is significant weight loss, dietetic input is needed to aid in weight gain to increase intra-abdominal fat, which acts as a cushion to reduce compression of the left renal vein, and this may be sufficient to reduce symptoms.[32,41] Some patients require total parenteral nutrition or nasogastric tube feeding.

In highly symptomatic patients or those in whom medical management has failed, invasive management options may be considered after appropriate preoperative optimisation. Endovascular stenting is usually considered as the first-line treatment.[5,16,42] In some cases, open surgery may be required to attach the renal vein to the inferior vena cava 2–5 cm below the normal attachment site to reduce compression.[43,44] Open gonadal vein transposition, coiling, or ligation of pelvic varicosities with or without anterior nephropexy can also be performed.[28,31,45]

There is some mention in the literature of spleno-renal and gonado-caval venous bypass surgery for NCS however these are limited to a few isolated case series.[28,46] In case of recurrence, an endovascular approach with plasty or stenting is favoured.[44]

No large cohort studies have been found studying the safety and efficacy of these procedures in the EDS/HSD population however case reports have shown good outcomes with open surgical and endovascular treatments.[14,23,25,29]

MTS

In the general population, iliac vein DVT is normally treated non-operatively with therapeutic anticoagulation.[47] The compressive element that predisposes patients to DVT formation in MTS necessitates that pharmacological management is almost always in conjunction with minimally invasive or open surgical intervention. Catheter-directed thrombolysis or open thrombectomy may be used with or without balloon angioplasty and stenting of the iliac veins to reduce compression by the iliac artery.[16,36,47] In some cases, venous bypass, or vascular transposition is performed.[37,43]

There is significant heterogeneity in the literature regarding the duration of postoperative anticoagulation, choice of agent, and the effects of concurrent antiplatelet use. Patients are mostly anticoagulated for a minimum of 6–12 months for proximal DVT and are administered lifelong aspirin monotherapy if a stent is inserted. Lifelong anticoagulation therapy is not uncommon; however, decisions need to be guided by individual patient risk factors. Percutaneous stent placement may be implemented.[48] No large cohort study has described the safety of thrombolysis/thrombectomy or venous angioplasty/stenting for MTS in the EDS/HSD population.

If no DVT is found, MTS is present on imaging, and the patient is highly symptomatic, patients can undergo catheter-based venography with venous angioplasty and stenting. It is important to balance recurrent thrombosis with the risk of bleeding, particularly in patients with EDS/HSD who are at a higher risk of bleeding.

Pelvic congestion is more common in MTS and NCS. Smith et al[3] conducted a retrospective, non-blinded study examining the efficacy of pelvic vein insufficiency treatment in 52 female patients with EDS/HSD between 2018–2021. The authors found that embolisation and sclerotherapy for gonadal vein reflux, pelvic varices, and/or iliac vein stenting significantly improved symptoms of chronic pelvic pain, interstitial cystitis and dysautonomia.[3] There were no significant rates of major complications associated with these procedures.[3] IVUS was also performed without complications in these patients.[3]

MALS

In asymptomatic MALS with evidence of collateral flow on imaging, no intervention is needed; however, patients should be followed up as they can develop symptoms.[26] For symptomatic MALS, coeliac artery decompression success depends on appropriate patient selection (favourable factors are symptomatic patients aged between 40–60 years, with weight loss

of approximately 9 kg, or those who have had a positive response to provocative investigation).[26,35,42] Decompression with median arcuate ligament release was performed laparoscopically or using an open approach.[26] Neurolysis or coeliac ganglionectomy can be performed concurrently to reduce associated neuropathic pain.[26] Bypass grafting may be reserved in cases of failure of less invasive treatments.

In recent years, laparoscopic decompression has become the preferred approach owing to the lower rate of all-cause perioperative complications; however, some studies have found a higher rate of reintervention.[4,35,49]

Concurrent revascularisation (coeliac artery stenting or angioplasty) has been shown to improve long-term symptomatic relief in both open and laparoscopic interventions.[26]

Long-term success and symptom resolution have been variably reported in the literature. Columbo et al[50] reported a good response (75% of patients were symptom-free at 6 months in a study of 21 patients in the general population), whereas Bagley et al[51] found only a fair degree of long-term symptom relief in their patients. Late symptomatic recurrence occurs in ~10% of cases post-procedure (laparoscopic or open).[26,35] A large-scale systematic review by Metz et al[52] suggested overall positive outcomes in terms of symptom relief (up to 70%) in the treatment of MALS.

SMAS

For SMAS, nasogastric tube placement may be required to decompress the dilated stomach and proximal duodenum if present and to assist with feeding to increase intra-abdominal fat if there is significant weight loss.[40] This may be sufficient to resolve symptoms from the perspective of increasing the aortomesenteric angle by fat padding.[40]

If this fails, the options include laparoscopic duodenojejunal bypass or open surgery (Strong's procedure, gastrojejunostomy or duodenojejunostomy).[40] In Strong's procedure, the ligament of Treitz is divided to mobilise the duodenum. The duodenum is then positioned on the right side of the SMA to avoid it being present in the space between the SMA and aorta.[40] Failure occurs in approximately 25% of Strong's procedures as per Scovell et al[40] Duodenojejunostomy has greater success rates than Strong's procedure or gastrojejunostomy and is the preferred surgical procedure.[40]

Sandmann et al[23] conducted an observational study (between 2010–2020) of 169 patients (122 with hEDS/HSD) and found that the surgical and endovascular techniques for MALS and SMAS cannot produce a durable result given the hyper-elastic tissue which creates instability in the operative area. The authors stated that these patients required "adaptive operative techniques" to stabilise their operative sites.[2] To treat MALS (and MALS in combination with another VCS), the MAL was partially resected.[2] Approximately 26% of 169 patients were readmitted for symptomatic re-compression post procedure (it is unclear from the study what percentage were hEDS/HSD patients).[2] Only 4 out of the total patients were asymptomatic at 3 months.[2] Superior mesenteric artery transposition was performed in patients with isolated SMAS ("Strong procedure").

In this series, approximately 90.5% of the patients reported being symptom-free at 6 months, which is a much higher success rate than that reported by Scovell et al[40] The procedure was safe and there were no significant rates of major complications.

Discussion

EDS/HSD, VCS, POTS, and MCAS form a complex web of associations and overlapping symptoms (Figure 2). VCS has been shown to directly exacerbate the symptoms of POTS through venous pooling, or in the case of MALS, irritation of nearby autonomic nerves, as well as indirectly when they provoked PCS.

MCAS can also worsen the symptoms of POTS and VCS, postulated to occur via its effect on vascular tone and permeability secondary to the release of substances such as histamine and pro-inflammatory cytokines.

All of these conditions were observed at a higher frequency in EDS/HSD patients. Therefore, it is important to look for VCS, POTS, and MCAS in patients with EDS/HSD to treat these conditions, which will also aid in reducing symptom burden.

In numerous case reports, VCS treatment has been shown to relieve the symptoms of PCS and POTS. For instance, left renal vein compression, as in NCS, may result in pelvic congestion with a subsequent raised dural sac and intracranial pressure, causing a persistent, debilitating headache.[17] Stubberud et al described a case in which left renal vein transposition and gastrojejunostomy resulted in resolution of medication-resistant generalised headache in a young woman with hEDS, POTS, severe NCS, MTS, MALS, and SMAS.[17]

A case series by Rozen et al[53] documented four female patients (ages 19–59 years) with generalised hypermobility and chronic headache which worsened in the Trendelenburg position.[54] Novel time-resolved contrast-enhanced magnetic resonance (MR) angiography of the abdomen revealed regional epidural venous plexus congestion and retrograde left lumbar vein flow.[54] The patients underwent embolisation of the left second lumbar vein using microcoils, and each reported a subsequent improvement in headaches post-treatment.[54]

In a study by Frantz et al[55] of 33 non-EDS/HSD patients, ligation of the renospinal trunk helped reduce the symptoms of myelopathy and syringomyelia which were postulated to have occurred due to NCS.

The interplay between EDS/HSD, POTS, MCAS, PCS, and VCS emphasises the need for a multi-disciplinary approach to managing these conditions, including cardiology/clinical pharmacology (POTS), immunologists/allergists (MCAS), as well as input from gastroenterologists, rheumatologists, neurologists, neurosurgeons, vascular surgeons, interventional radiologists, and clinical geneticists.

Apart from the vEDS subtype, invasive testing to identify the VCS is deemed safe in patients with EDS and is being utilised in centres across Australia. Catheter-based procedures are generally discouraged in vEDS patients (reasons outlined in "Results").[1,28,56] For patients with vEDS, non-invasive ultrasound or CT/MR angiography are safer options than invasive imaging.[56] If these tests are negative, but there is a high degree of suspicion for one of the VCS, it is often too high risk to perform invasive testing in the vEDS population, and therefore formulating a diagnosis can be difficult in this group. It is important that genetic testing for suspected vEDS is performed, as once there is a diagnosis of vEDS, clinicians can approach the imaging and management of the VCS in these patients in a safer manner.

Many imaging modalities are expensive and are often performed in private centres. Access to imaging and specialist appointments (including long waiting lists) can be an issue for patients who are severely disabled by EDS/HSD/POTS, who may be already facing financial difficulties, or who live regionally/rurally.

Despite positive results in the literature, vascular intervention may not fully resolve all symptoms of VCS, and surgical intervention should be in conjunction with non-operative management and allied health involvement. The role of dietetics for nutritional optimisation and weight gain, especially in NCS, MALS, and SMAS, cannot be underestimated. Delayed wound healing, which predisposes patients to wound infections, is expected in EDS patients given tissue fragility. This may be exacerbated by malnutrition due to significant weight loss from the VCS.

In patients with the vEDS subtype, endovascular or open surgery to treat VCS is typically not performed, unless there is a life-threatening situation. Some clinicians may prefer to await genetic testing to rule out vEDS before attempting a procedure. However, waiting times to see a clinical geneticist and return of genetic testing can take several months. No large cohort studies have focused specifically on the risks of invasive imaging or endovascular/open vascular interventions in the vEDS group and obtaining clinical ethics approval for this would be difficult.

Another important point is that the prevalence of the four VCS tends to be higher in females.[5,9,16–19,34,36] EDS is also more common in females (7:3),[57] including the vEDS subtype.[58] Currently, there are no studies on the management of VCS in pregnant women with EDS/HSD, identifying a large research gap.

Studies with larger sample sizes of patients with EDS/HSD are needed to definitively establish the safety and efficacy of endovascular/invasive procedures for correcting VCS. These studies should follow up the patient for several years to determine if the symptoms remain resolved and if any complications from the procedures arise. These studies should also describe whether POTS/PCS symptoms are significantly reduced after VCS correction. The treatment of MCAS with correction of the VCS should be investigated, as no studies have been conducted on this topic. Studies should help to establish formal and internationally recognised imaging and treatment algorithms for VCS in patients with EDS/HSD.

Conclusion

The prevalence of VCS in the EDS/HSD population is likely higher than that in the general population given the abnormalities in vascular collagen in these patients. It is known that there are delays in the diagnosis of patients with VCS, EDS/HSD, POTS, PCS and/or MCAS.[15,16,57] This could be due to clinicians' inexperience in this area, the lack of published studies in this area, and the constellation of symptoms (sometimes overlapping), which may make diagnosis difficult.

Patients with an EDS/HSD commonly have more than one VCS. Correcting one VCS without correcting the other(s) in a single patient leads to worse outcomes.[49] Furthermore, the conditions associated with VCS (POTS, MCAS, PCS)

also need to be treated concurrently, as they can exacerbate each other and the VCS. This indicates the need for clinicians to have a high degree of suspicion of VCS and their associated conditions.

Invasive imaging and procedures for correcting VCS in patients with EDS/HSD are generally considered safe. For vEDS, the increased fragility of blood vessels can lead to more severe VCS symptoms[8] and contraindicates invasive imaging and invasive treatment procedures, leading to difficulties in managing VCS symptoms in these patients.

No large cohort study has examined the safety and long-term efficacy of invasive imaging and procedures (or treatment algorithms) for VCS in the EDS/HSD population. Further research in these areas can greatly improve patient care and increase clinician awareness and understanding of these conditions and their associations.

The diagnostic and management guideline for VCS in EDS/HSD proposed in this review may serve as a foundation for further development as additional studies in this area emerge.

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