TECHNICKÁ UNIVERZITA V KOŠICIACH
Strojnícka fakulta
Katedra Biomedicínskeho Inžinierstva a Merania

ABDOMINO–PELVIC VASCULAR COMPRESSION SYNDROMES

&

SYMPTOMATIC JOINT HYPERMOBILITY SYNDROMES

(connective tissue disorders)

Mgr. Silvia Šišková

06 / 11 / 2024

Odborný školiteľ:

Dr. h. c. mult. prof. Ing. Jozef Živčák, Dr.Sc., MPH

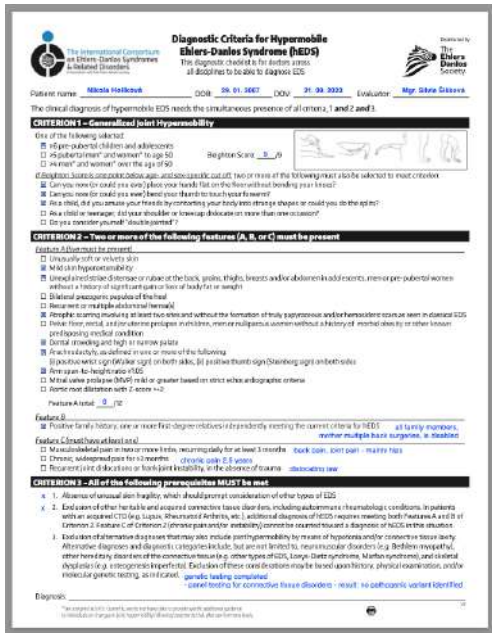
PATIENT ORGANIZATION

AVKS – Asociácia Vaskulárnych Kompresívnych Syndrómov a Ehlers-Danlos (SK & CZ), o.z.

- 1.5 year
- 297 members

Wilkie, Dunbar, Nutcracker, May-Thurner & EDS (Ehlers-Danlos) / SK & CZ ›

- 71 cases - complex dg. management



II VENOUS SUMMIT
CCEV 2023
Endovascular Surgery Chapter
MÁLAGA 14 -15 ABRIL

CERTIFICADO DE ASISTENCIA

A favor de

SILVIA ŠIŠKOVÁ

por su asistencia al **II Venous Summit**, organizado por el Capítulo de Cirugía Endovascular de la Sociedad Española de Angiología y Cirugía Vascular celebrado en Málaga, el 14 y 15 de abril de 2023.

Y para que conste donde convenga se expide el presente certificado.

Dr. Alejandro Rodríguez Morata
Presidente del Capítulo de Cirugía Endovascular

Dra. María Isolina de Haro Sanz
Secretaria del Capítulo de Cirugía Endovascular

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The Interconnected Relationships between Abdomino-Pelvic Vascular Compression Syndromes and Symptomatic Joint Hypermobility Syndromes, Their Co-existing Conditions and Comorbidities, and Their Significance in Patient Diagnostic and Treatment Management

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15 November 2023

INTRODUCTION

Abdomino-pelvic Vascular Compression Syndromes are currently defined as rare conditions, where vessels in the abdomen and pelvis are compressed by other vessels or other structures and organs, or where vessels compress other structures and organs. These include Median Arcuate Ligament Syndrome, Left Renal Vein Compression Syndrome, Superior Mesenteric Artery Syndrome, Iliac Vein Compression Syndrome, Pelvic Venous Insufficiency, and Inferior Vena Cava Syndrome.[2,5,7,8,10]

The described syndromes present with a wide variety of clinical symptoms as well as anatomic variations. Moreover, the symptomatology frequently also overlaps with many other conditions. Thus, the diagnostic process is often very demanding and unclear, as well as the patient treatment management.[4,7,8]

Most affected seem to be patients of young age and these conditions have a significant impact on their quality of life.

Recently, multiple studies have pointed out the link between Abdomino-Pelvic Vascular Compression Syndromes and Symptomatic Joint Hypermobility Syndromes and their co-existing conditions.[7-10] However, a more in-depth research is lacking. The objective of this study is to perform such reseach in order to allow for a better understanding of the interplay of these conditions.

METHODS

A combination of retrospective data collection and a prospective diagnostic study is being utilized in order to carry out this research study. The studied patient population consists of 2 main groups, including international patient population and local Slovak-Czech patient population. These two populations differ quite extensively in terms of the scope of differential diagnostic process so far completed. Whereas a distinctive portion of the international patient group has already been evaluated on a broader and more in-depth scale in terms of all Abdomino-Pelvic Vascular Compression Syndromes, Symptomatic Joint Hypermobility syndromes and their co-existing conditions, the local Slovak-Czech patient group has only been evaluated for 1 or 2 compression syndromes. Evaluation of Symptomatic Joint Hypermobility Syndromes and their co-existing conditions is completely absent in this population.

Therefore, this research is pursuing a thorough investigation of data obtained from the above mentioned foreign patients, while at the same time is initializing and performing a similar diagnostic process with the local Slovak-Czech patient population. In the end, a comparative analysis and evaluation of the combined results of these 2 populations will allow for a more in-depth understanding of the interconnected relationships of these conditions.

The estimated number of patients in the international population is approximately 20 000, while approximately 100 in the local Slovak-Czech population. However, patient numbers are rising continuously and rapidly.

DISCUSSION & CONCLUSIONS

With the increasing amount of recent evidence suggesting the correlation between Abdomino-Pelvic Vascular Compression Syndromes, Symptomatic Joint Hypermobility Syndromes and their co-existing comorbid conditions, a strong need for more research and understanding of these interconnected relationships arises.[7-10,14]

A better and a more in-depth understanding of all these conditions and their interplay is necessary in order to allow for advancements in the diagnostic and treatment patient management. Early and correct diagnosis and treatment, in particular, represent direct predictors of a better patient outcome, thus higher patient quality of life.

REFERENCES

Abdomino-Pelvic Vascular Compression Syndromes (APVCS):

Median Arcuate Ligament Syndrome (MALS) / Dunbar Syndrome
Left Renal Vein Compression Syndrome (NCS) / Nutcracker Syndrome
Superior Mesenteric Artery Syndrome (SMAS) / Wilkie's Syndrome
Iliac Vein Compression Syndrome (MTS) / May-Thurner Syndrome
Pelvic Venous Insufficiency (PVI) / Pelvic Congestion Syndrome (PCS)
Inferior Vena Cava Syndrome (IVCS)

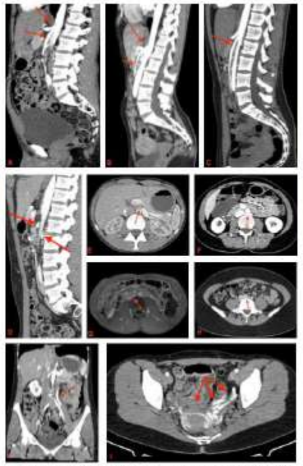

Figure 1. CT images of vascular compression syndromes

Median Arcuate Ligament Syndrome is the compression of celiac artery by the median arcuate ligament. Sometimes, the ligament compesses not only the celiac artery but also the the adjacent nerves. Hence, we differentiate between 2 types of MALS, vascular MALS and neurogenic MALS, or also a combination of these two.[5,8]

In Left Renal Vein Compression Syndrome, the left renal vein is compressed in an acute angle created between the aorta and the superior mesenteric artery. However, other surrounding organs and structures may contribute to the left renal vein compression as well. There are also other variations of NCS, such as posterior NCS, where the left renal vein is compressed between the spine and the aorta, or a circumaortic NCS, which is a combination of anterior and posterior NCS due to left renal vein congenital branching.[5,8]

Superior Mesenteric Artery Syndrome is defined as compression of the third part of the duodenum in the acute angle between aorta and superior mesenteric artery.[1] Thus, NCS and SMAS may often co-exist together.

In Iliac Vein Compression Syndrome, most often the right iliac artery compresses the left iliac vein. However, compressions may occur in multiple other spots as well and simultaneously.[2,3,5,8]

Pelvic Venous Insufficiency or Pelvic Congestion Syndrome is usually the result of the above described NCS and MTS, or a combination of these.[3,5,7-10]

It is important to stress that, APVCS many times coexist together and multiple APVCS are often found simultaneously in one patient, especially patients with Symptomatic Joint Hypermobility Syndromes, such as Ehlers-Danlos Syndrome or Hypermobile Spectrum Disorders.[7-10]

Multisystemic symptomatologic manifestations of APVCS:

headache, vertigo, dizziness, syncope or pre-syncope, nausea, chest pain, restricted breathing ability, palpitations, shortness of breath, abdominal pain, early satiety, loss of appetite, feeling full, sensation of food getting stuck in GI tract, extreme belching and bloating, inability to eat enough, unintentional severe weight loss, diarrhea, constipation, back pain, flank pain, pelvic pain, vaginal pain, dyspareunia, painful menstruation, bladder pain, dysuria, bladder dysfunction, burning sensations, swelling, heaviness, leg pain, leg numbness, blood pooling, thrombosis[1-3,5,7-10]

This list is not exhaustive of all the possible symptoms. It is also important to note that the severity and combination of symptoms may vary in each individual.

Symptomatic Joint Hypermobility Syndromes

Symptomatic Joint Hypermobility Syndromes are a group of conditions, which present with joint laxity, hypermobility and tissue fragility. They are associated with a vast number of comorbidities, specific to the different subtypes. Among Symptomatic Join Hypermobility Syndromes belong the 14 types of Ehlers-Danlos Syndromes, Hypermobility Spectrum Disorders, Marfan Syndrome, Osteogenesis Imperfecta, Loeys-Dietz Syndrome, and Stickler Syndrome.[4,6,13]

Given that connective tissue is presented throughout the whole human body, multiple organs and body systems may be affected. It is also important to note that the combination of manifestations as well as their severities may vary distinctively from individual to individual. In addition, many of these comorbid conditions share similar clinical symptomatology, thus making the differential diagnosis often challenging.

Figure 2. The Beighton Scoring System. Diagnostic criteria for measuring joint hypermobility.[11]

Symptomatic Joint Hypermobility Syndromes (SJHS):[4,6]

- Ehlers-Danlos Syndromes (EDS)
- Hypermobility Spectrum Disorders
- Marfan Syndrome
- Osteogenesis Imperfecta (Brittle Bone Disease)
- Loeys-Dietz Syndrome
- Stickler Syndrome

HSD & hEDS - related comorbidities:[4,6,11,12]

- musculoskeletal
- immunological
- gastrointestinal
- cardiovascular and autonomic
- neurological
- urogynecologic
- cutaneous/dermatological
- ocular, oral, mandibular
- sleep, fatigue, pain and psychological impact

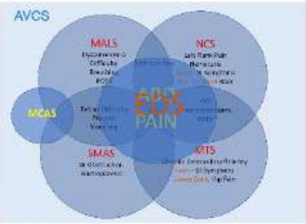

Figure 3. The overlapping relationships and manifestations of vascular compression syndromes, EDS and co-existing conditions.[1]

The figure above represents the overlapping manifestations of Abdomino-Pelvic Vascular Compression Syndromes as well as Symptomatic Joint Hypermobility Syndromes and their co-existing conditions.[7,8] Thus, at the same time, it emphasises the need for a multidisciplinary diagnostic and treatment approach. Such guidelines should therefore comprise of an in-depth analysis of all Abdomino-Pelvic Vascular Compression Syndromes, Symptomatic Joint Hypermobility Syndromes and their co-existing comorbidities. Only aforementioned approach is able to provide the best possible opportunity for a correct and successful patient treatment outcome.

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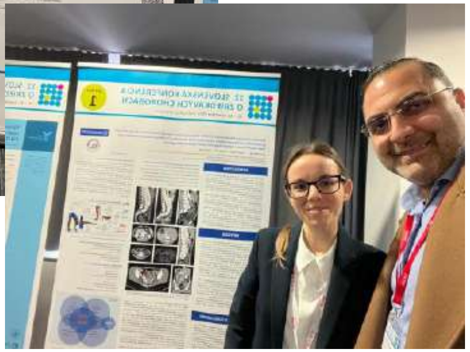
Mgr. Silvia Šišková

Disclosures:
Human subjects: All CT images used in this publication were obtained and used with the patients' permissions.

12. SLOVENSKÁ KONFERENCIA O ZRIEDKAVÝCH CHOROBÁCH

15. – 16. november 2023, Hotel Saffron Bratislava



www.ehlers-danlosuv-syndrom.org www.tarlovovacysta.org www.avks.sk

Ehlers-Danlosův syndrom a syndrom hypermobility

Eliška Kopicová [1] | Silvia Šišková [2] | Jozef Živčák [3]

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[2] Technická Univerzita v Košiciach, Strojnícka Fakulta, Katedra biomedicínskeho inžinierstva a merania, Košice, Slovensko; Asociácia Vaskulárnych Kompresívnych Syndrómov a Ehlers-Danlos, o.z., Bratislava, Slovensko
[3] Technická Univerzita v Košiciach, Strojnícka Fakulta, Katedra biomedicínskeho inžinierstva a merania, Košice, Slovensko

Ehlers-Danlosovy syndromy (Q79.6) jsou skupinou 13(+) vrozených poruch pojivové tkáně, které jsou spojovány s mutacemi více než 20 různých genů. Každý z typů EDS má trochu jiné příznaky (a každý pacient i v rámci jednoho typu a jedné rodiny může mít trochu jinou kombinaci a intenzitu potíží), ale obecně tyto poruchy charakterizuje kloubní hypermobilita a křehkost tkání.[1]

Syndrom hypermobility (M35.7) je porucha pojivové tkáně vyznačující se symptomatickou hypermobilitou (například hypermobilitou v kombinaci s chronickou bolestí a dalšími (nejen) muskuloskeletálními obtížemi).[1]

Podobnými diagnózami je Marfanův syndrom, Loeys-Dietzův syndrom, cutis laxa, Sticklerův syndrom a Osteogenesis imperfecta (OI).[1]

Některé z možných symptomů:

Hypermobilita, subluxace a/nebo dislokace kloubů, chronická bolest, chronická únava, gastrointestinální potíže, skolióza a kyfóza, svalová hypotonie, hyperextenzibilní a/nebo jemná, "těstovitá" či křehká kůže, pomalé hojení, snadná tvorba modřin, atrofické či jinak abnormální jizvy, aneurysma, hernie, prolapsy, degenerativní onemocnění páteře a kloubů, poranění šlach a vazů - a další.[1-3]

Pacienti s Ehlers-Danlosovými syndromy mohou zažívat **neurologické a/nebo muskulární potíže** jako je: dysautonomie (především syndrom posturální ortostatické tachykardie - POTS), idiopatická intrakraniální hypertenze (IIH), Chiariho malformace 1. typu, únik mozkomíšního moku, kraniocervikální nestabilita (CCI) a atlantoaxiální nestabilita (AAI), skolióza, syndrom fixované míchy (v důsledku abnormalit filum terminale), Tarlovovy cysty, neuropatie, bolesti hlavy, svalová hypotonie, svalová slabost, myalgie.[1-3]

Mezi další možné komorbidity se řadí například neurovývojové poruchy (např. poruchy autistického spektra (PAS) a ADHD), syndromy vaskulární komprese, poruchy aktivace žírných buněk (např. syndrom aktivace žírných buněk (MCAS)), gastroparéza - a další.[1-3]

Diagnostika a management symptomů:

EDS se obecně diagnostikuje genetickým testováním, avšak genové mutace nejčastějšího typu (hypermobilní EDS - hEDS) nejsou tak úplně známy (diagnostikuje se tedy dle klinických znaků a symptomů). Syndrom hypermobility může diagnostikovat ortoped, revmatolog, neurolog či rehabilitační lékař. Ve světě se diskutuje o tom, jaký je opravdový rozdíl mezi syndromem hypermobility a hypermobilním typem EDS (hEDS).[1-3]

Ač tyto poruchy nejsou léčitelné a pouze se řeší jednotlivé symptomy, správná diagnóza pomáhá. Řešení a management potíží závisí na individuálních potížích pacienta (často je však doporučována fyzioterapie či ergoterapie).

Zdroje:

1. Gensemer C, Burks R, Kautz S, Judge DP, Lavallee M, Norris RA. **Hypermobile Ehlers-Danlos syndromes: Complex phenotypes, challenging diagnoses, and poorly understood causes.** *Developmental Dynamics*. 2021; 250:318–344.

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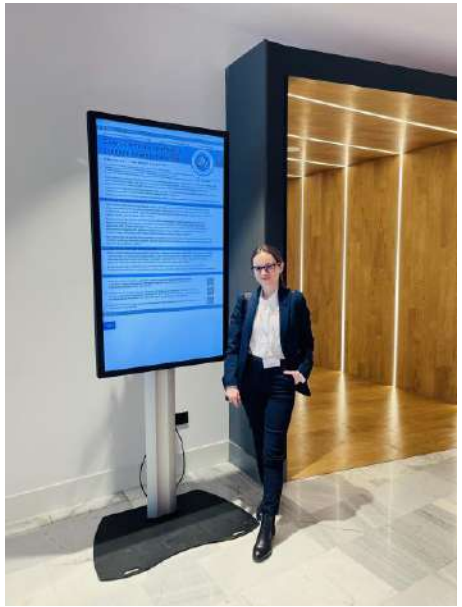
3. Castori M, Voermans NC. **Neurological manifestations of Ehlers-Danlos syndrome(s): A review.** Iran J Neurol 2014; 13(2): 190-208.



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Strojnícka fakulta

The Biomechanisms and Links between Abdomino-Pelvic Vascular Compression Syndromes and Symptomatic Joint Hypermobility Syndromes

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[3] Dr. Ricardo Gil Sánchez, Internal medicine. Rare disease speciality, Calle del Doctor Zamenhof 29 Bajo, 46008 Valencia, Spain; Hospital La Fe, Valencia, consulta@dricardogil.com
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Abstract: The link between Abdomino-Pelvic Vascular Compression Syndromes (APVCS) and Symptomatic Joint Hypermobility Syndromes (SJHS) highlights the need for a better understanding of the biomechanisms involved in the pathogenic processes of these disorders. This paper discusses 41 patient cases from Slovakia and Czech Republic primarily diagnosed with APVCS, but not evaluated further. Within the investigation process, these patients were clinically assessed for SJHS, in particular the hypermobile type of Ehlers Danlos Syndrome and Hypermobility Spectrum Disorder. The results point out the need to acknowledge the link between these two conditions and call for further research to better understand the biomechanisms involved in these pathogenic processes.

Keywords: Abdomino-Pelvic Vascular Compression Syndromes (APVCS); Median Arcuate Ligament Syndrome (MALS, Dunbar Syndrome); Left Renal Vein Compression Syndrome (NCS, Nutcracker Syndrome); Superior Mesenteric Artery Syndrome (SMAS, Wilkie Syndrome); Iliac Vein Compression Syndrome (MTS, May-Thurner Syndrome); Pelvic Venous Insufficiency (PVI); Pelvic Congestion Syndrome (PCS); Symptomatic Joint Hypermobility Syndromes (SJHS); Ehlers-Danlos Syndrome (EDS); Hypermobility Spectrum Disorder (HSD)

1. Introduction

Abdomino-Pelvic Vascular Compression Syndromes are currently defined as rare conditions, where vessels in the abdomen and pelvis are compressed by other vessels or other structures and organs, or where vessels compress other structures and organs. These include Median Arcuate Ligament Syndrome, Left Renal Vein Compression Syndrome, Superior Mesenteric Artery Syndrome, Iliac Vein Compression Syndrome, Pelvic Venous Insufficiency, and Inferior Vena Cava Syndrome [1-3,5,8-11,16]. These described syndromes present with a wide variety of clinical symptoms as well as anatomic variations. Moreover, the symptomatology frequently also overlaps with many other conditions. Thus, the diagnostic process is often very demanding and unclear, as well as the patient treatment management [5,8,9,16]. Most affected seem to be patients of young age and these conditions have a significant impact on their quality of life.

Recently, a few studies have pointed out the link between Abdomino-Pelvic Vascular Compression Syndromes and Symptomatic Joint Hypermobility Syndromes and their co-

SCIENTIFIC COOPERATIONS - SVK

NÚSCH BA

MEDIREX

ULBGaKG LF UK a UNB

AGEL Šaca

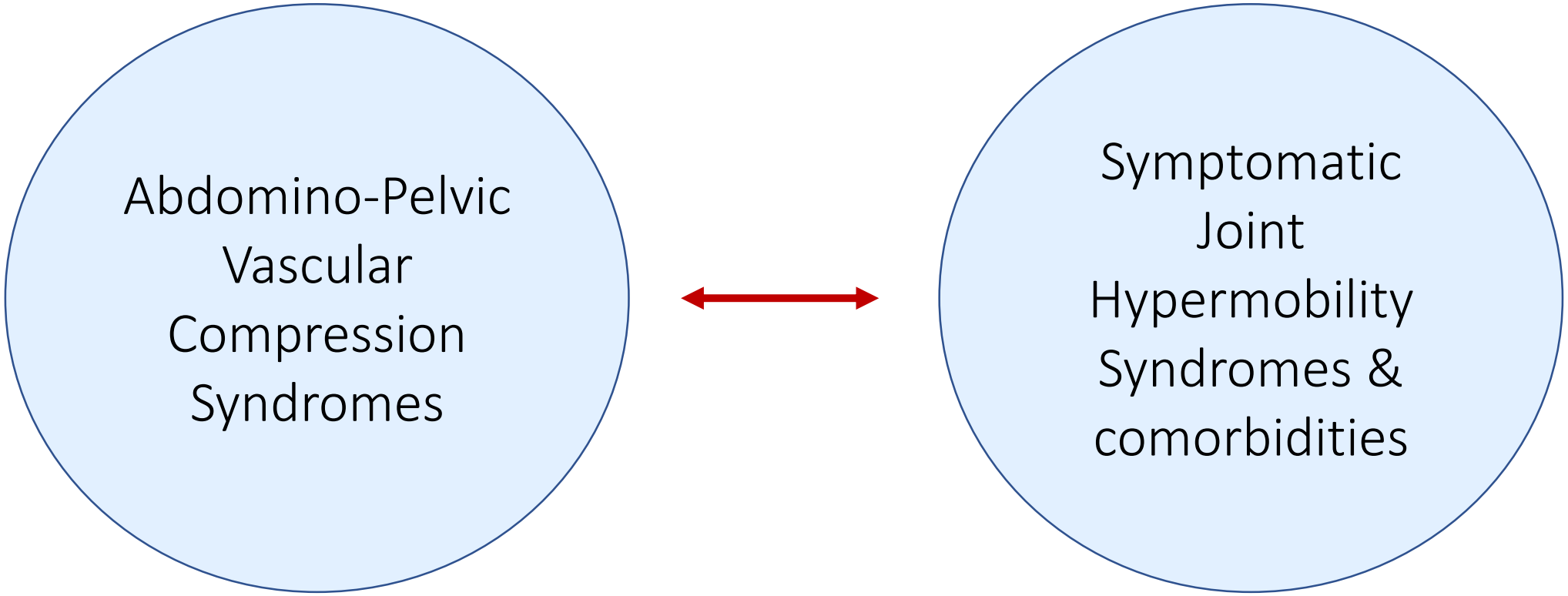
VÚSCH KE

Penta Hospitals Bory BA

FN Motol Praha

ASSIDUO BA

STUDY TOPIC


Abdomino-Pelvic
Vascular
Compression
Syndromes
Symptomatic
Joint
Hypermobility
Syndromes &
comorbidities

SYMPTOMATIC JOINT HYPERMOBILITY SYNDROMES

Ehlers-Danlos Syndromes *(14 types)*

EDS

Hypermobility Spectrum Disorders

HSD

Marfan Syndrome

group of heritable connective tissue disorders

Other:

Stickler Syndrome
Loeys-Dietz Syndrome
Osteogenesis Imperfecta
Cutis Laxa

EDS TYPES

Name of EDS Subtype	*IP**	*Genetic Basis*	*Protein Involved*
Classical EDS (cEDS)	AD	Major: *COL5A1, COL5A2*	Type V collagen
		Rare: *COL1A1* c.934C>T, p.(Arg312Cys)	Type I collagen
Classical-like EDS (clEDS)	AR	*TNXB*	Tenascin XB
Cardiac-valvular EDS (cvEDS)	AR	*COL1A2* (biallelic mutations that lead to *COL1A2* NMD and absence of pro α2(I) collagen chains)	Type I collagen
Vascular EDS (vEDS)	AD	Major: *COL3A1*	Type III collagen
		Rare: *COL1A1* c.934C>T, p.(Arg312Cys) c.1720C>T, p.(Arg574Cys) c.3277C>T, p.(Arg1093Cys)	Type I collagen
Hypermobile EDS (hEDS)	AD	Unknown	Unknown
Arthrochalasia EDS (aEDS)	AD	*COL1A1, COL1A2*	Type I collagen
Dermatosparaxis EDS (dEDS)	AR	*ADAMTS2*	ADAMTS-2
Kyphoscoliotic EDS (kEDS)	AR	*PLOD1*	LH1
		FKBP14	FKBP22
Brittle cornea syndrome (BCS)	AR	*ZNF469*	ZNF469
		PRDM5	PRDM5
Spondylodysplastic EDS (spEDS)	AR	*B4GALT7*	ß4GalT7
		B3GALT6	ß3GalT6
		SLC39A13	ZIP13
Musculocontractural EDS (mcEDS)	AR	*CHST14*	D4ST1
		DSE	DSE
Myopathic EDS (mEDS)	AD or AR	*COL12A1*	Type XII collagen
Periodontal EDS (pEDS)	AD	*C1R*	C1r

* Inheritance Pattern: AD = autosomal dominant; AR = autosomal recessive

SJHS & COMORBIDITIES

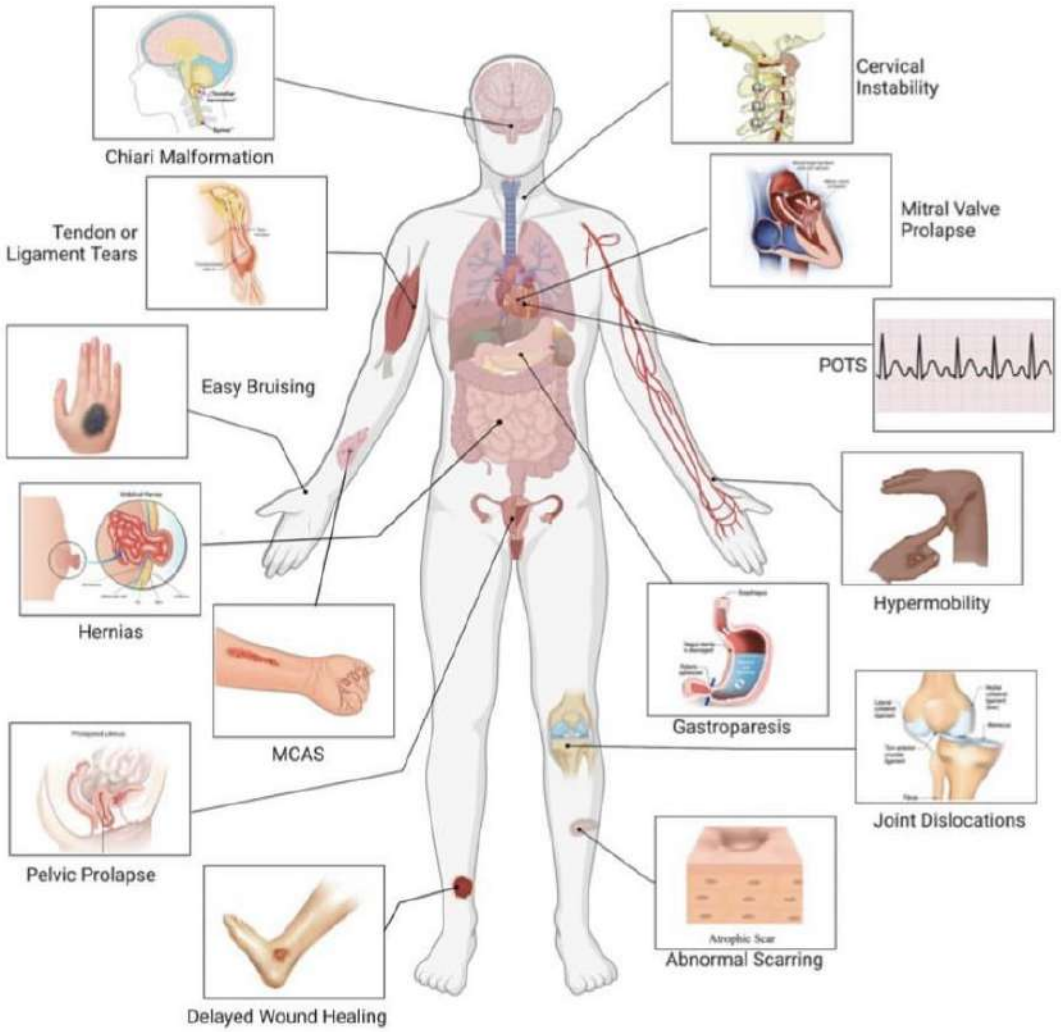
Musculoskeletal

Cutaneous/ dermatological

Cardiovascular & autonomic

Gastrointestinal

Reproductive & urinary



Ocular

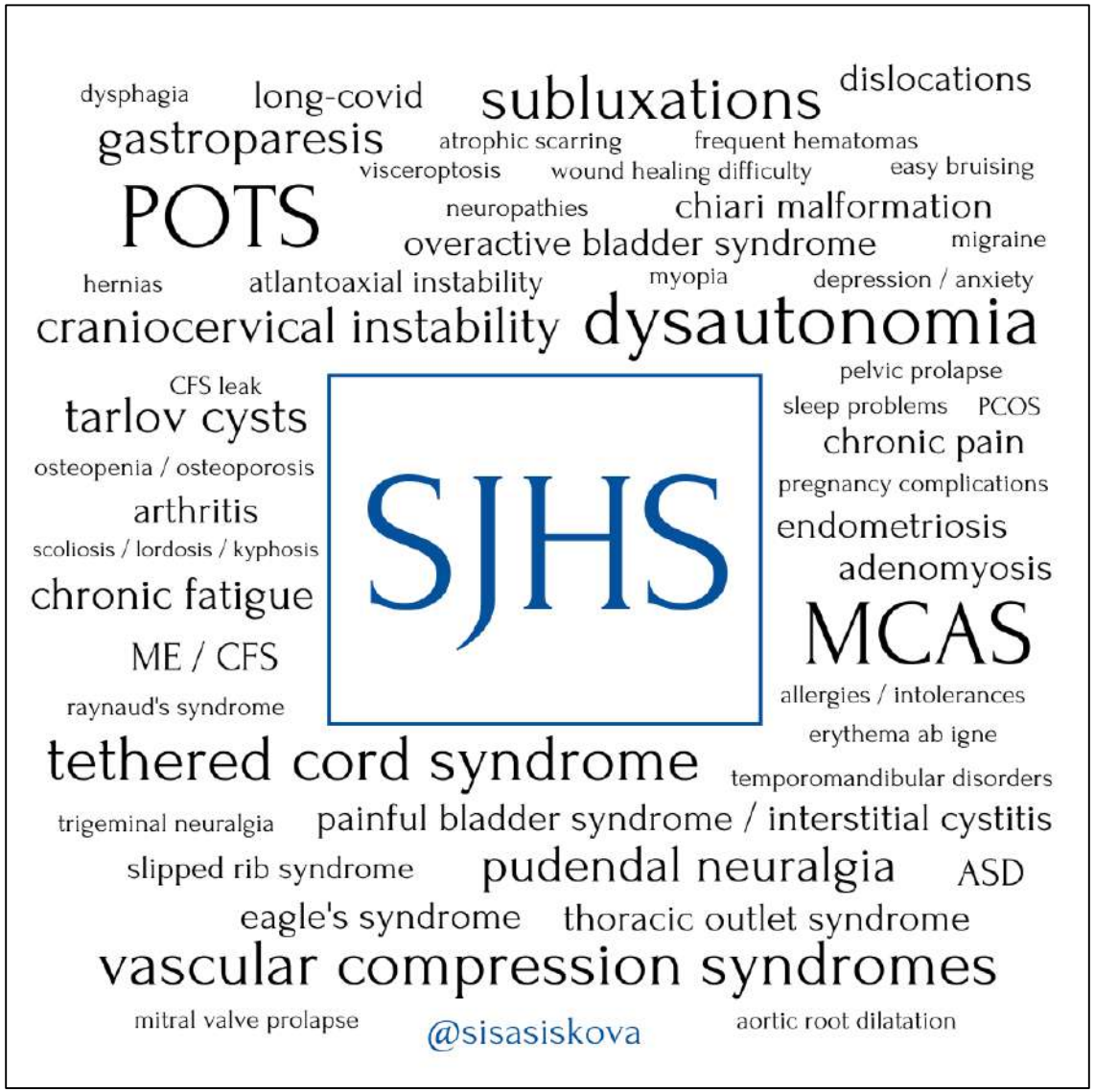
Oral, mandibular, voice

Immunological

Neurological

Sleep, fatigue, pain, mental

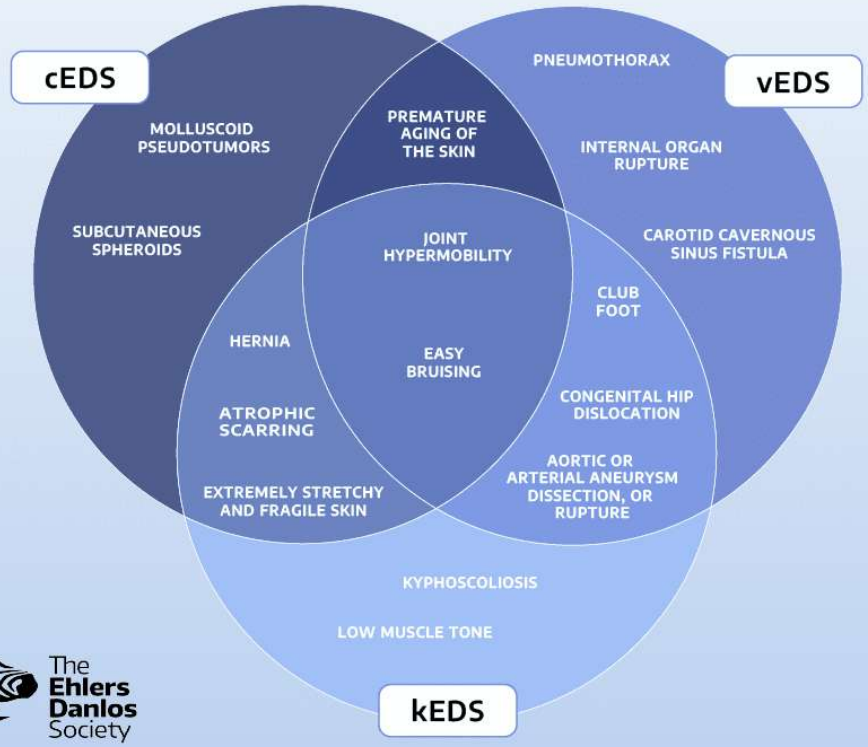
COMORBIDITIES



EDS SPECTRUM

THE EDS SPECTRUM

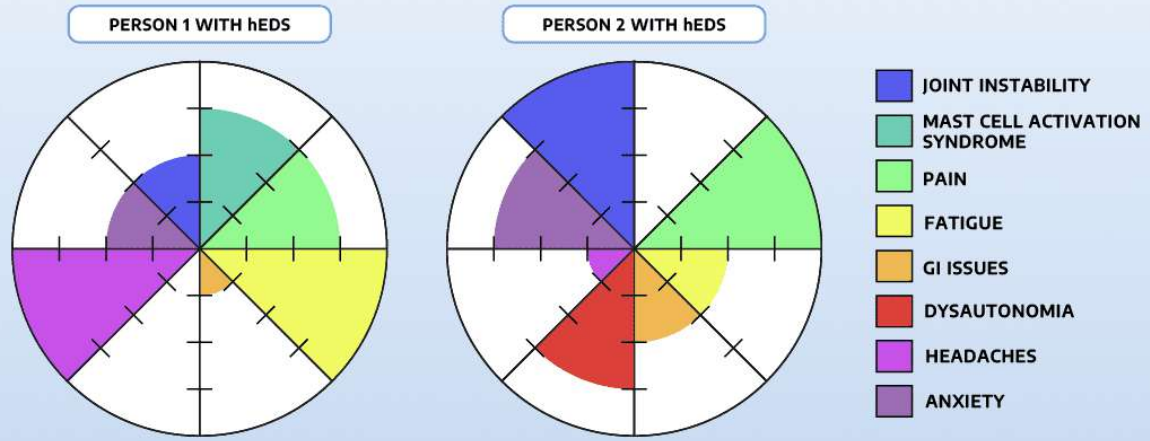
EACH TYPE OF EDS HAS ITS OWN COMBINATION OF SIGNS AND SYMPTOMS. SOME SIGNS AND SYMPTOMS ARE COMMON IN MULTIPLE TYPES OF EDS, WHILE OTHERS ARE ONLY ASSOCIATED WITH ONE SPECIFIC TYPE.



The Ehlers Danlos Society

THE EDS SPECTRUM

PEOPLE WITH THE SAME TYPE OF EDS MAY EXPERIENCE DIFFERENT SIGNS AND SYMPTOMS

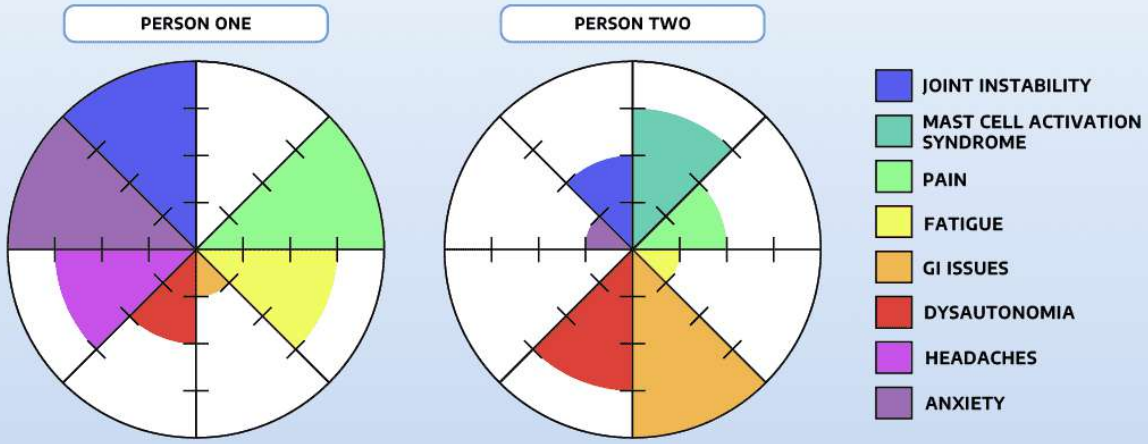


The Ehlers Danlos Society

HSD SPECTRUM

THE HSD SPECTRUM

TWO DIFFERENT PEOPLE WITH HSD MAY EXPERIENCE VERY DIFFERENT SYMPTOMS



The Ehlers Danlos Society

THE HSD SPECTRUM

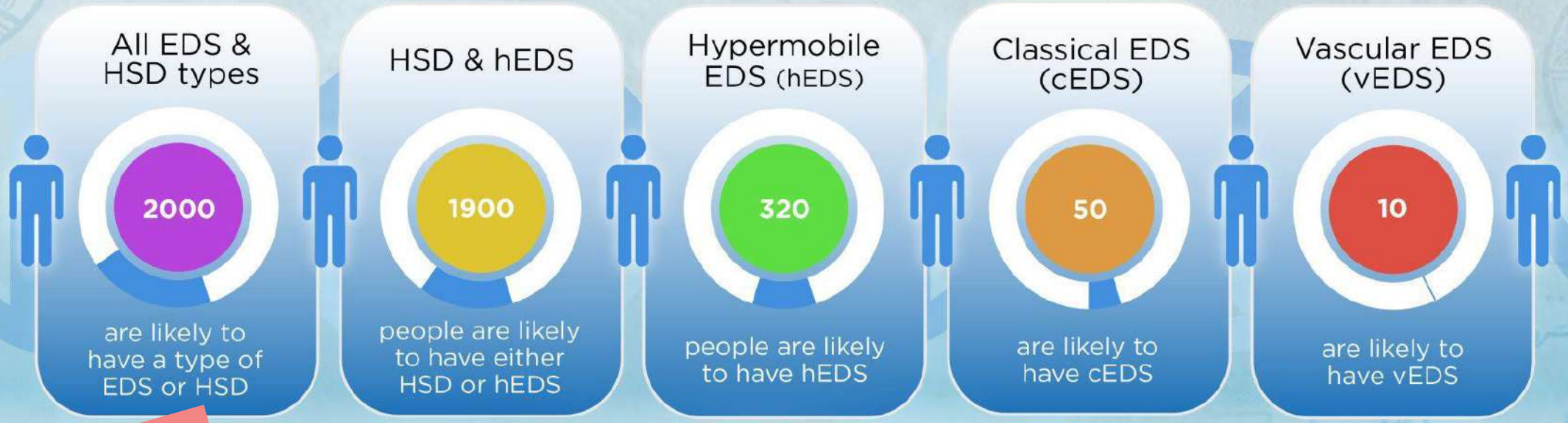
THE HYPERMOBILITY SPECTRUM DISORDERS DO NOT EXIST ON A LINEAR SPECTRUM LIKE THIS:

EACH PERSON'S EXPERIENCE IS A COMBINATION OF THE SPECIFIC SYMPTOMS THAT AFFECT THEM. THE SPECTRUM LOOKS MORE LIKE THIS

The Ehlers Danlos Society

HOW PREVALENT ARE THE EHLERS-DANLOS SYNDROMES (EDS) & HYPERMOBILITY SPECTRUM DISORDERS (HSD)?

PER MILLION IN THE POPULATION



1 in 500

LESS THAN ONE IN A MILLION

Arthrochalasia EDS (aEDS)
Brittle Cornea Syndrome (BCS)
Cardiac-Valvular EDS (cvEDS)
Classical-Like EDS (clEDS)
Dermatosparaxis EDS (dEDS)

The Ehlers Danlos Society
EDS & HSD AWARENESS MONTH 2022

Kyphoscoliotic EDS (kEDS)
Musculocontractural EDS (mcEDS)
Myopathic EDS (mEDS)
Periodontal EDS (pEDS)
Spondylodysplastic EDS (spEDS)

The Ehlers-Danlos Society

The International Consortium on Ehlers-Danlos Syndromes & Related Disorders
In Association with The Ehlers-Danlos Society

Diagnostic Criteria for Hypermobile Ehlers-Danlos Syndrome (hEDS)

This diagnostic checklist is for doctors across all disciplines to be able to diagnose EDS

Distributed by The Ehlers Danlos Society.

Patient name: _____ DOB: _____ DOV: _____ Evaluator: _____

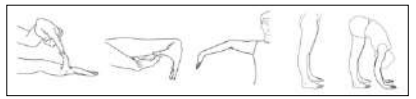
The clinical diagnosis of hypermobile EDS needs the simultaneous presence of all criteria, 1 **and** 2 **and** 3.

CRITERION 1 – Generalized Joint Hypermobility

One of the following selected:

- ☐ ≥6 pre-pubertal children and adolescents
- ☐ ≥5 pubertal men and woman to age 50
- ☐ ≥4 men and women over the age of 50

Beighton Score: _____/9



If Beighton Score is one point below age- and sex-specific cut off, two or more of the following must also be selected to meet criterion:

- ☐ Can you now (or could you ever) place your hands flat on the floor without bending your knees?
- ☐ Can you now (or could you ever) bend your thumb to touch your forearm?
- ☐ As a child, did you amuse your friends by contorting your body into strange shapes or could you do the splits?
- ☐ As a child or teenager, did your shoulder or kneecap dislocate on more than one occasion?
- ☐ Do you consider yourself "double jointed"?

CRITERION 2 – Two or more of the following features (A, B, or C) must be present

Feature A (five must be present)

- ☐ Unusually soft or velvety skin
- ☐ Mild skin hyperextensibility
- ☐ Unexplained striae distensae or rubae at the back, groins, thighs, breasts and/or abdomen in adolescents, men or pre-pubertal women without a history of significant gain or loss of body fat or weight
- ☐ Bilateral piezogenic papules of the heel
- ☐ Recurrent or multiple abdominal hernia(s)
- ☐ Atrophic scarring involving at least two sites and without the formation of truly papyraceous and/or hemosideric scars as seen in classical EDS
- ☐ Pelvic floor, rectal, and/or uterine prolapse in children, men or nulliparous women without a history of morbid obesity or other known predisposing medical condition
- ☐ Dental crowding and high or narrow palate
- ☐ Arachnodactyly, as defined in one or more of the following:
  (i) positive wrist sign (Walker sign) on both sides, (ii) positive thumb sign (Steinberg sign) on both sides
- ☐ Arm span-to-height ratio ≥1.05
- ☐ Mitral valve prolapse (MVP) mild or greater based on strict echocardiographic criteria
- ☐ Aortic root dilatation with Z-score >+2

Feature A total: _____/12

Feature B

- ☐ Positive family history; one or more first-degree relatives independently meeting the current criteria for hEDS

Feature C (must have at least one)

- ☐ Musculoskeletal pain in two or more limbs, recurring daily for at least 3 months
- ☐ Chronic, widespread pain for ≥3 months
- ☐ Recurrent joint dislocations or frank joint instability, in the absence of trauma

CRITERION 3 - All of the following prerequisites MUST be met

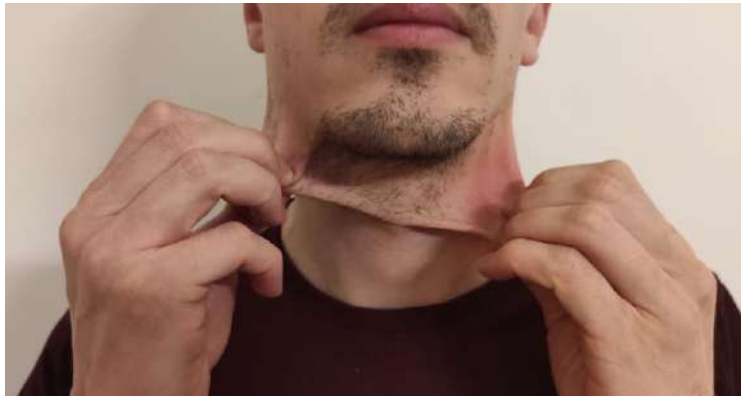
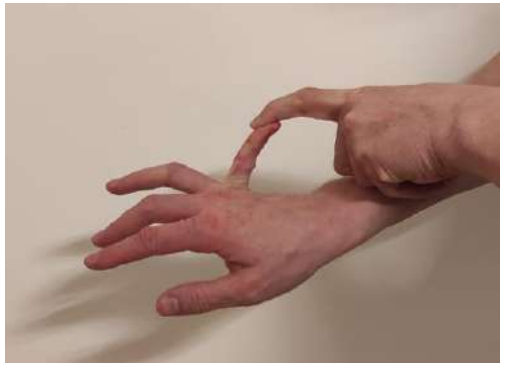
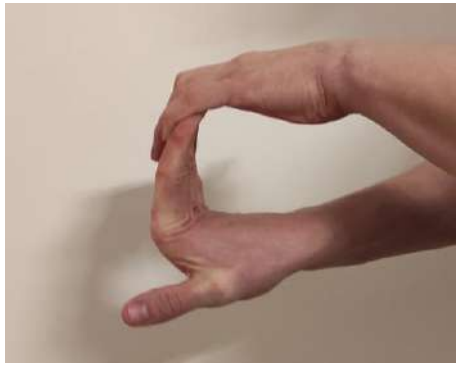
1. Absence of unusual skin fragility, which should prompt consideration of other types of EDS
2. Exclusion of other heritable and acquired connective tissue disorders, including autoimmune rheumatologic conditions. In patients with an acquired CTD (e.g. Lupus, Rheumatoid Arthritis, etc.), additional diagnosis of hEDS requires meeting both Features A and B of Criterion 2. Feature C of Criterion 2 (chronic pain and/or instability) cannot be counted toward a diagnosis of hEDS in this situation.
3. Exclusion of alternative diagnoses that may also include joint hypermobility by means of hypotonia and/or connective tissue laxity. Alternative diagnoses and diagnostic categories include, but are not limited to, neuromuscular disorders (e.g. Bethlem myopathy), other hereditary disorders of the connective tissue (e.g. other types of EDS, Loeys-Dietz syndrome, Marfan syndrome), and skeletal dysplasias (e.g. osteogenesis imperfecta). Exclusion of these considerations may be based upon history, physical examination, and/or molecular genetic testing, as indicated.

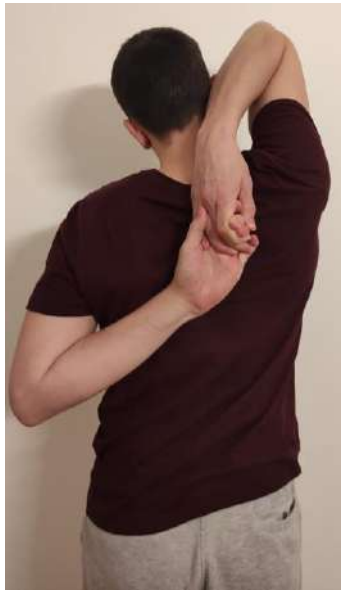
Diagnosis: _____

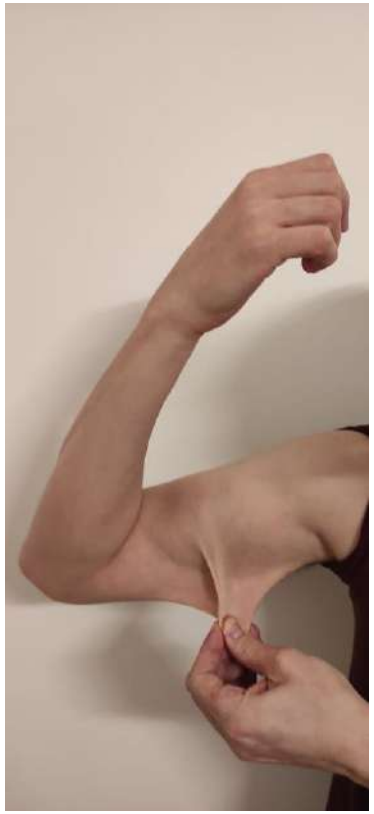
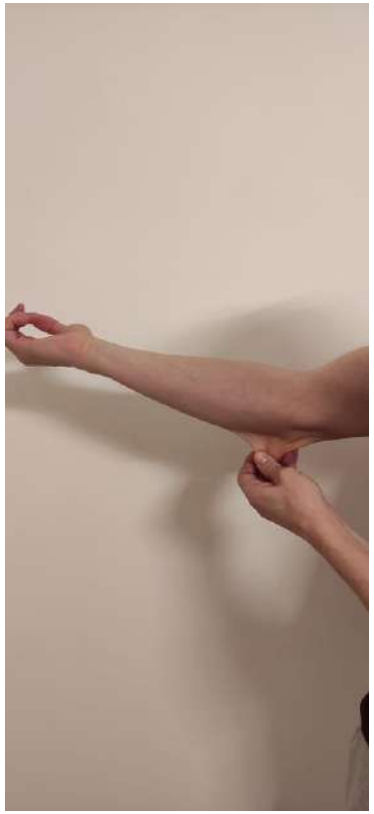
v9

THE 2017 INTERNATIONAL CLASSIFICATION OF THE EHLERS-DANLOS SYNDROMES

The Ehlers-Danlos Society

SOME RELEVANT PUBLICATIONS

American Journal of Medical Genetics Part C (Seminars in Medical Genetics) 175C:8–26 (2017)

ARTICLE

The 2017 International Classification of the Ehlers–Danlos Syndromes

FRANSISKA MALFAIT,* CLAIR FRANCOMANO, PETER BYERS, JOHN BELMONT, BRITTA BERGLUND, JAMES BLACK, LARA BLOOM, JESSICA M. BOWEN, ANGELA F. BRADY, NIGEL P. BURROWS, MARCO CASTORI, HELEN COHEN, MARINA COLOMBI, SERWET DEMIRDAS, JULIE DE BACKER, ANNE DE PAEPE, SYLVIE FOURNEL-GIGLEUX, MICHAEL FRANK, NEETI GHALI, CECILIA GIUNTA, RODNEY GRAHAME, ALAN HAKIM, XAVIER JEUNEMAITRE, DIANA JOHNSON, BIRGIT JUUL-KRISTENSEN, INES KAPFERER-SEEBACHER, HANADI KAZKAZ, TOMOKI KOSHO, MARK E. LAVALLEE, HOWARD LEVY, ROBERTO MENDOZA-LONDONO, MELANIE PEPIN, F. MICHAEL POPE, EYAL REINSTEIN, LEEMA ROBERT, MARIANNE ROHRBACH, LYNN SANDERS, GLENDA J. SOBEY, TIM VAN DAMME, ANTHONY VANDERSTEEN, CAROLINE VAN MOURIK, NICOL VOERMANS, NIGEL WHEELDON, JOHANNES ZSCHOCKE, AND BRAD TINKLE

American Journal of Medical Genetics Part C (Seminars in Medical Genetics) 175C:48–69 (2017)

ARTICLE

Hypermobile Ehlers–Danlos Syndrome (a.k.a. Ehlers–Danlos Syndrome Type III and Ehlers–Danlos Syndrome Hypermobility Type): Clinical Description and Natural History

BRAD TINKLE,* MARCO CASTORI, BRITTA BERGLUND, HELEN COHEN, RODNEY GRAHAME, HANADI KAZKAZ, AND HOWARD LEVY

American Journal of Medical Genetics Part C (Seminars in Medical Genetics) 175C:148

ARTICLE

A Framework for the Classification of Joint Hypermobility and Related Conditions

MARCO CASTORI,* BRAD TINKLE, HOWARD LEVY, RODNEY GRAHAME, FRANSISKA MALFAIT, AND ALAN HAKIM

Clinical Reviews in Allergy & Immunology (2020) 58:273–297
https://doi.org/10.1007/s12016-019-08755-8

The Relationship Between Hypermobile Ehlers-Danlos Syndrome (hEDS), Postural Orthostatic Tachycardia Syndrome (POTS), and Mast Cell Activation Syndrome (MCAS)

Alison Kohn[1] · Christopher Chang[1,2,3,4]

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DOI: 10.1002/dvdy.220

REVIEW

Developmental Dynamics WILEY

Hypermobile Ehlers-Danlos syndromes: Complex phenotypes, challenging diagnoses, and poorly understood causes

Cortney Gensemer[1] | Randall Burks[1] | Steven Kautz[2] | Daniel P. Judge[3] | Mark Lavallee[4] | Russell A. Norris[1]

Topics in PAIN MANAGEMENT
Vol. 38, No. 8 Current Concepts and Treatment Strategies March 2023

CONTINUING EDUCATION ACTIVITY

Hope for Hypermobility: Part 1—An Integrative Approach to Treating Symptomatic Joint Hypermobility

Victoria Daylor, BFA, Cortney Gensemer, PhD, Russell A. Norris, PhD, and Linda Bluestein, MD

Topics in PAIN MANAGEMENT
Vol. 38, No. 9 Current Concepts and Treatment Strategies April 2023

CONTINUING EDUCATION ACTIVITY

Hope for Hypermobility: Part 2—An Integrative Approach to Treating Symptomatic Joint Hypermobility

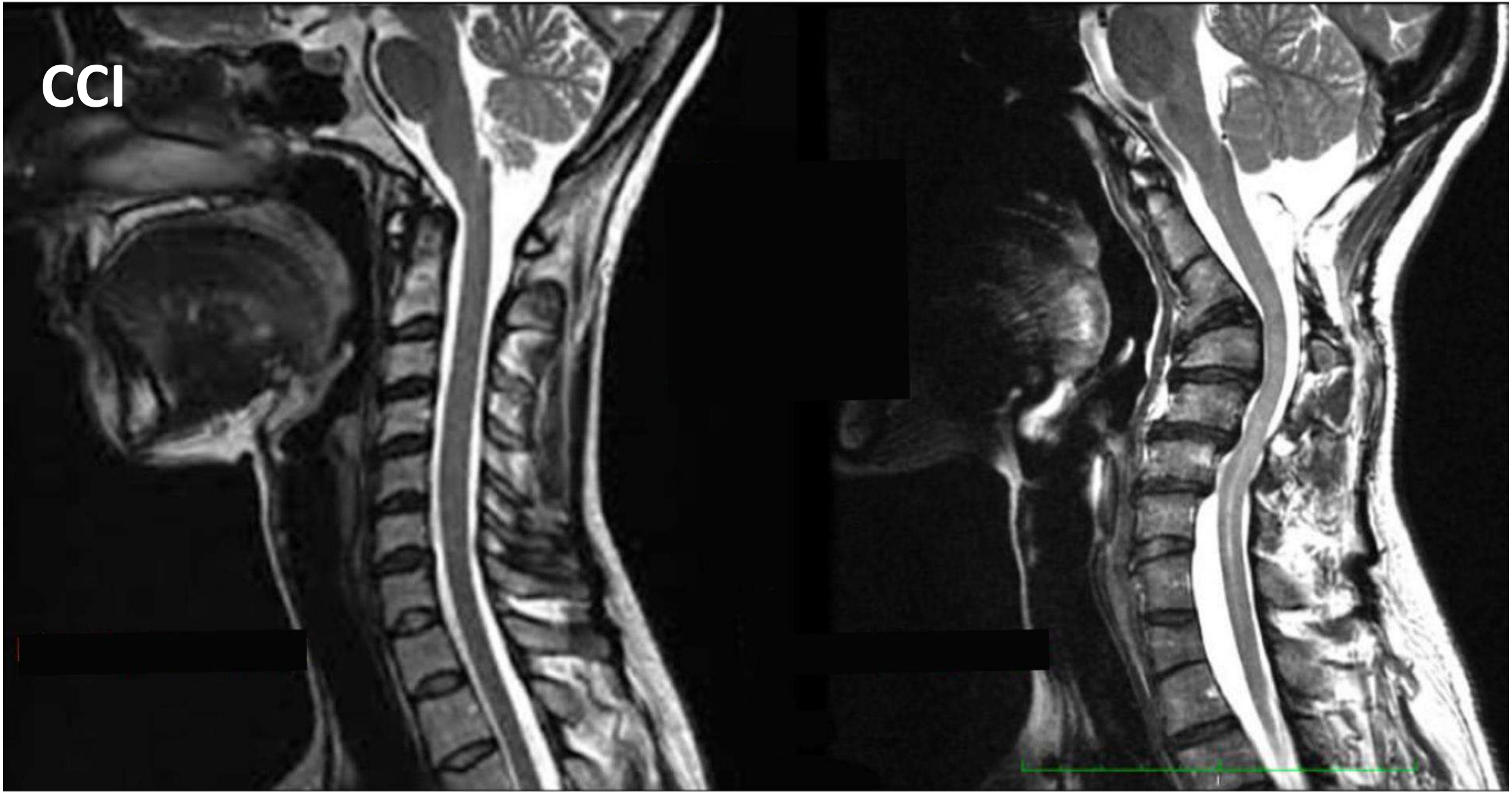
Victoria Daylor, BFA, Cortney Gensemer, PhD, Russell A. Norris, PhD, and Linda Bluestein, MD

American Journal of Medical Genetics Part C (Seminars in Medical Genetics) 175C:226–236 (2017)

RESEARCH REVIEW

Mast Cell Disorders in Ehlers–Danlos Syndrome

SURANJITH L. SENEVIRATNE, ANNE MAITLAND,* AND LAWRENCE AFRIN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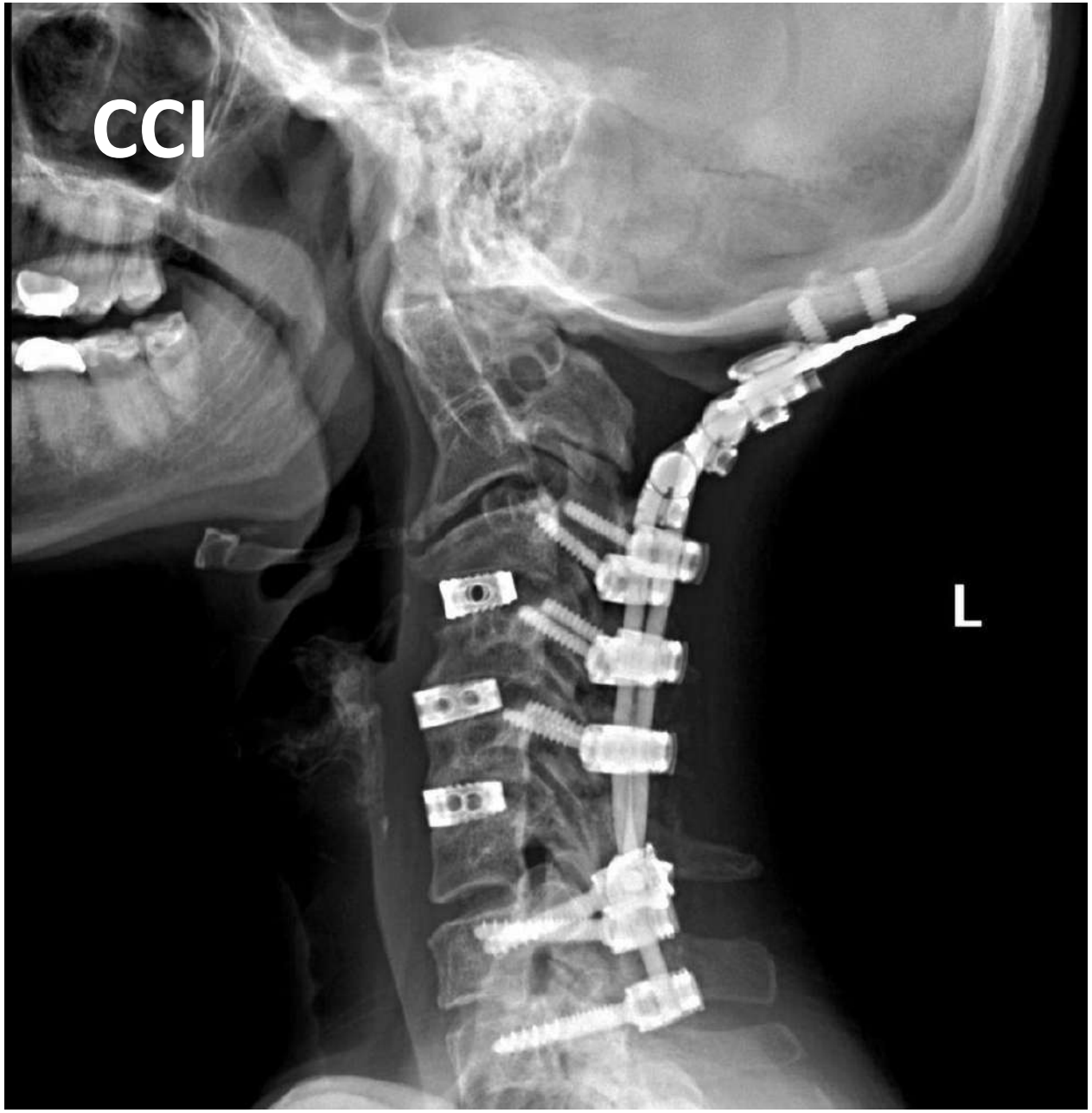
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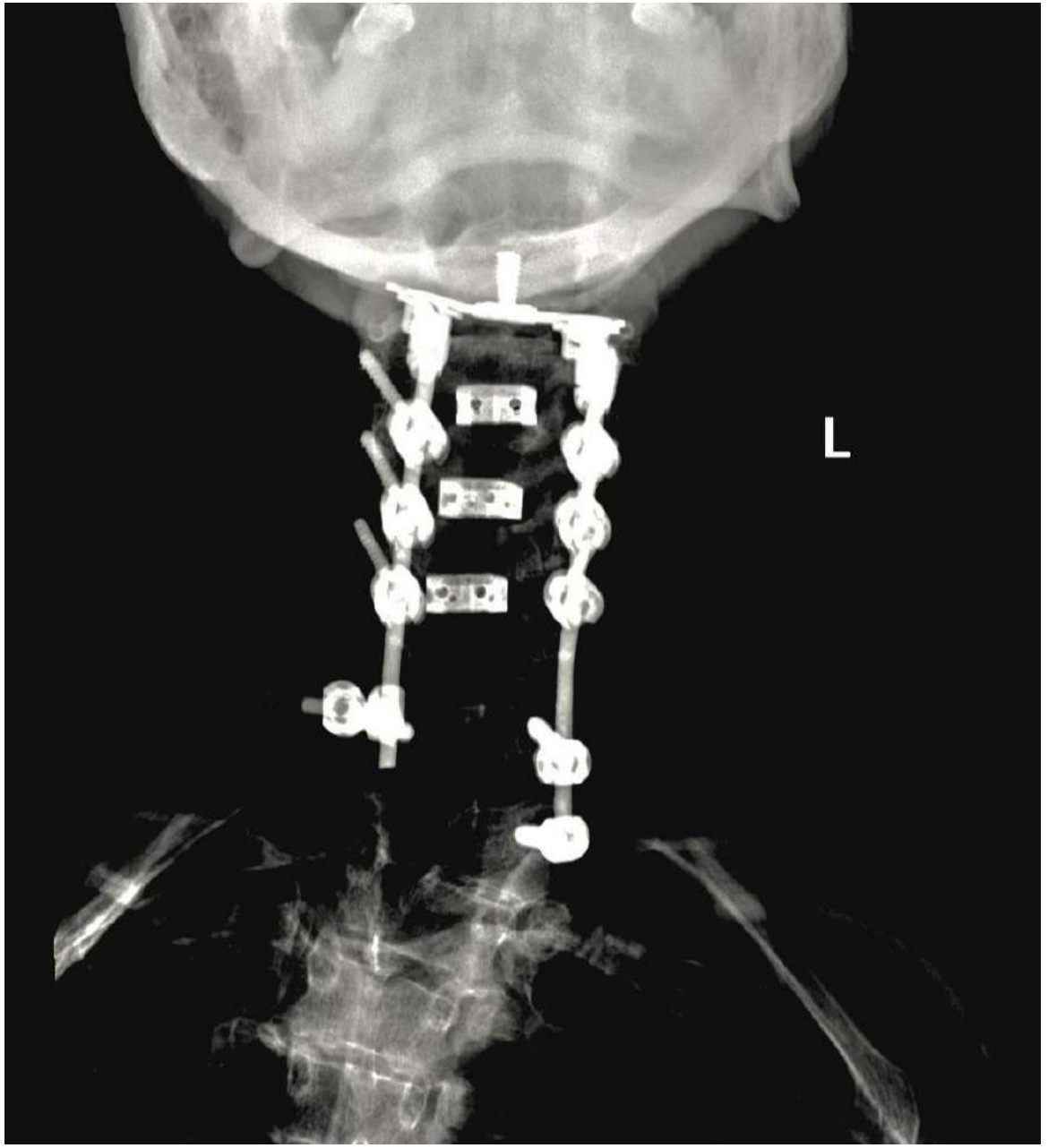
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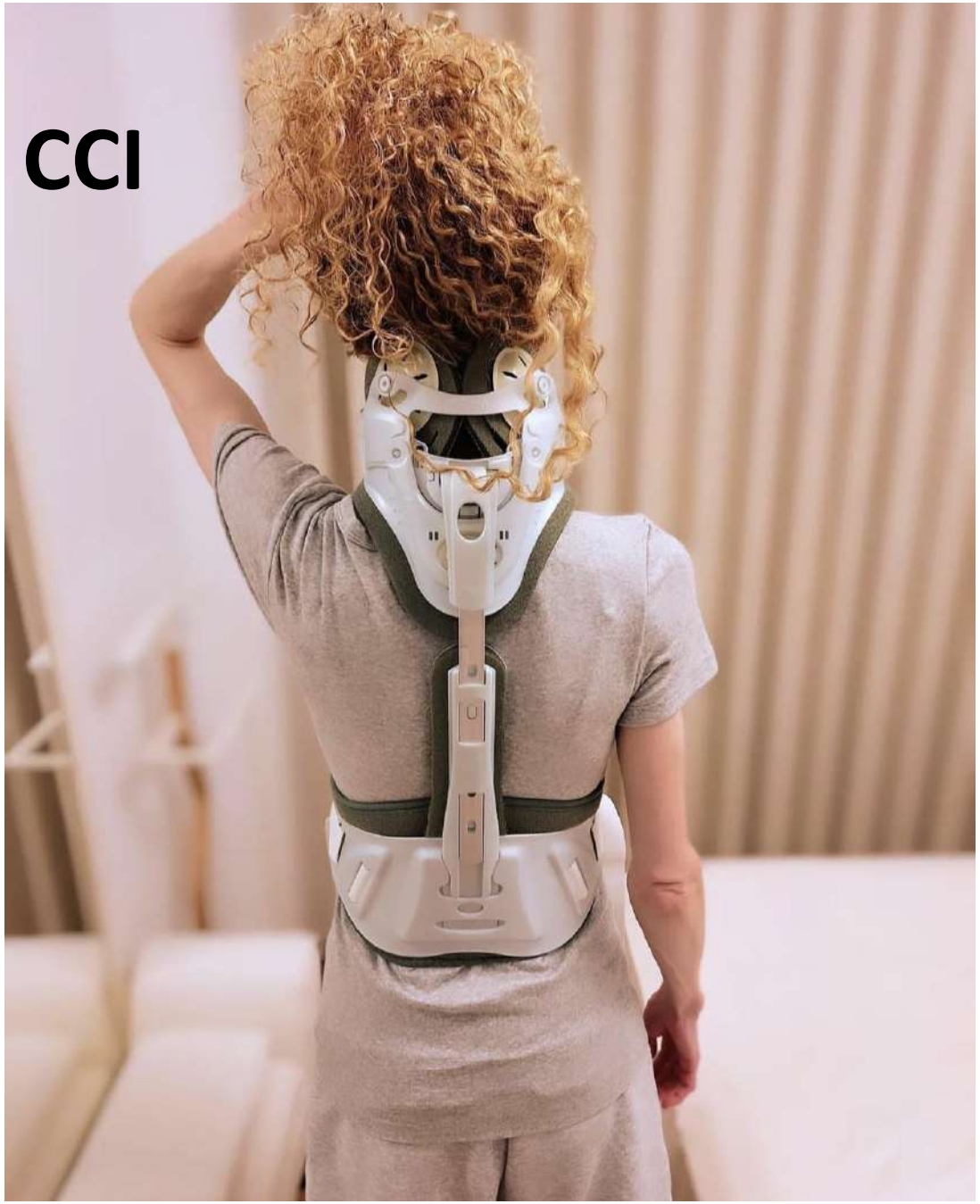
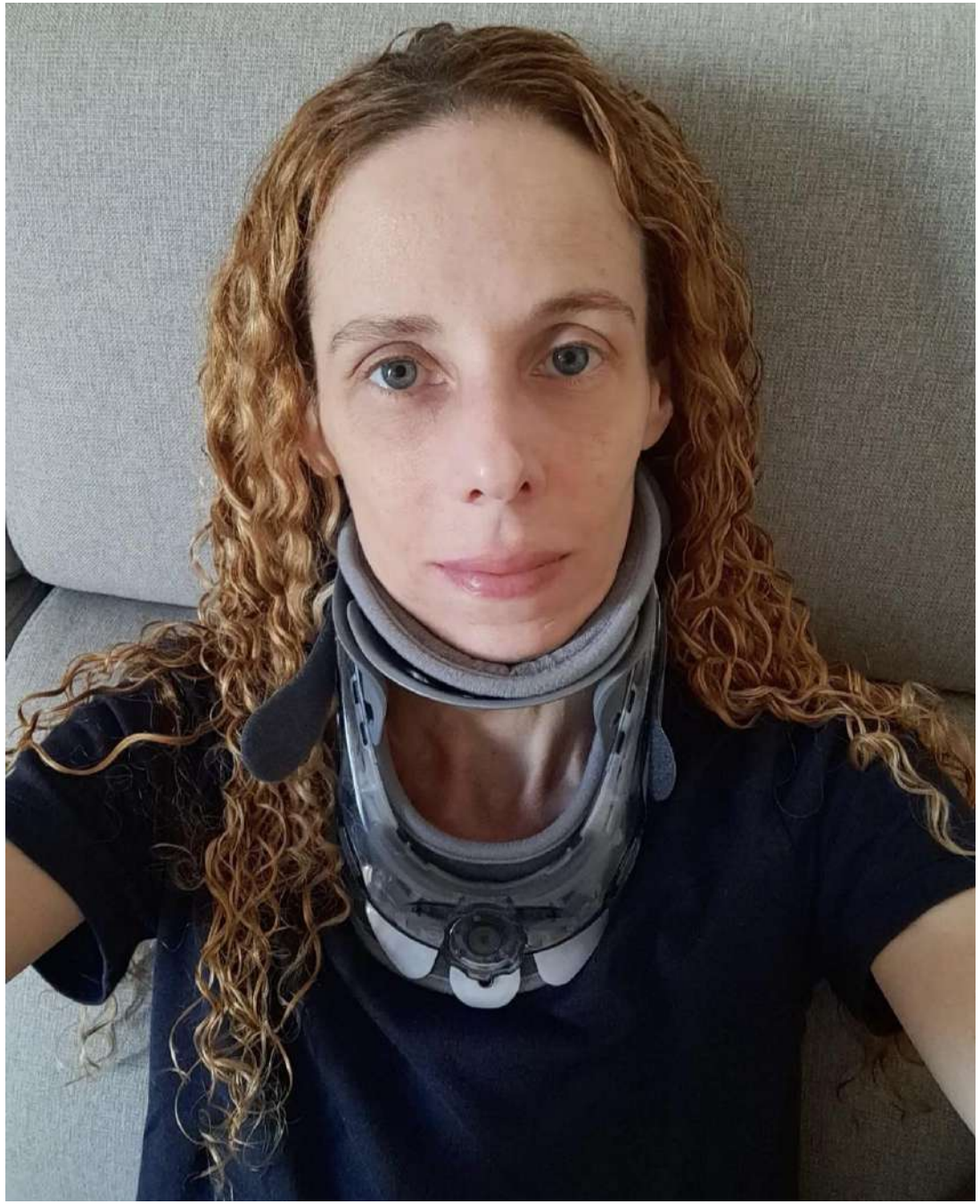
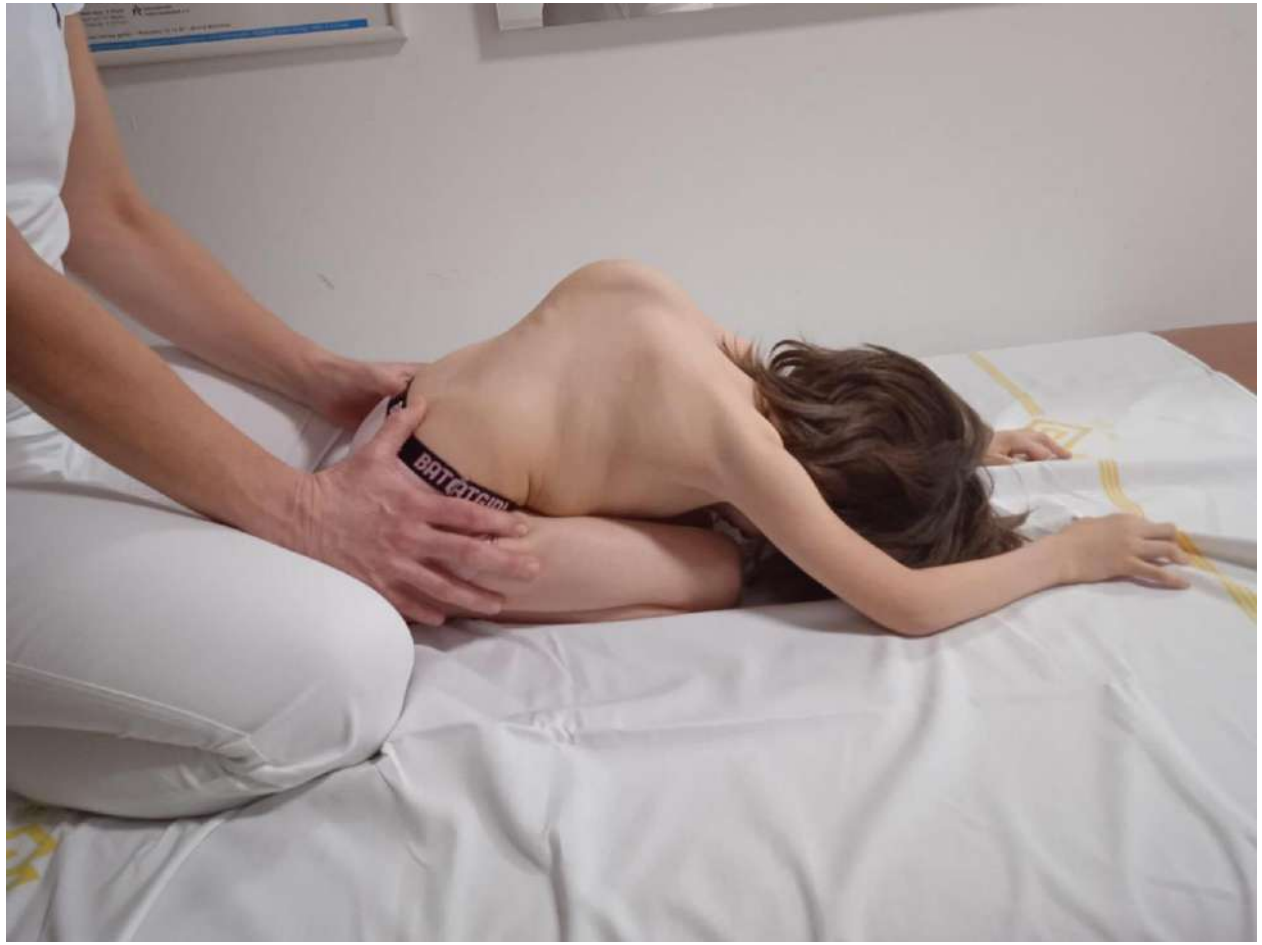


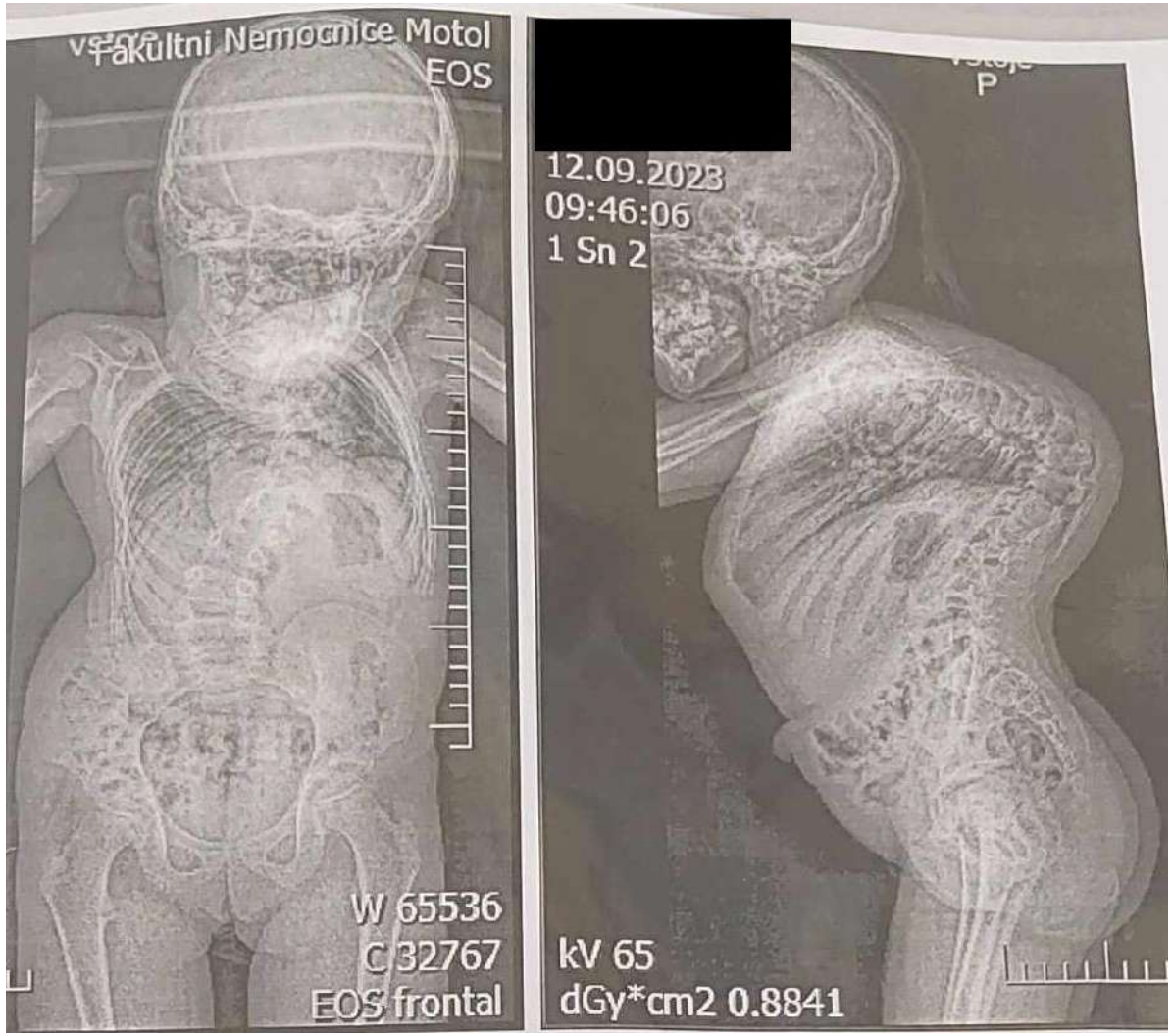


severe kyphoscoliosis in kEDS patient

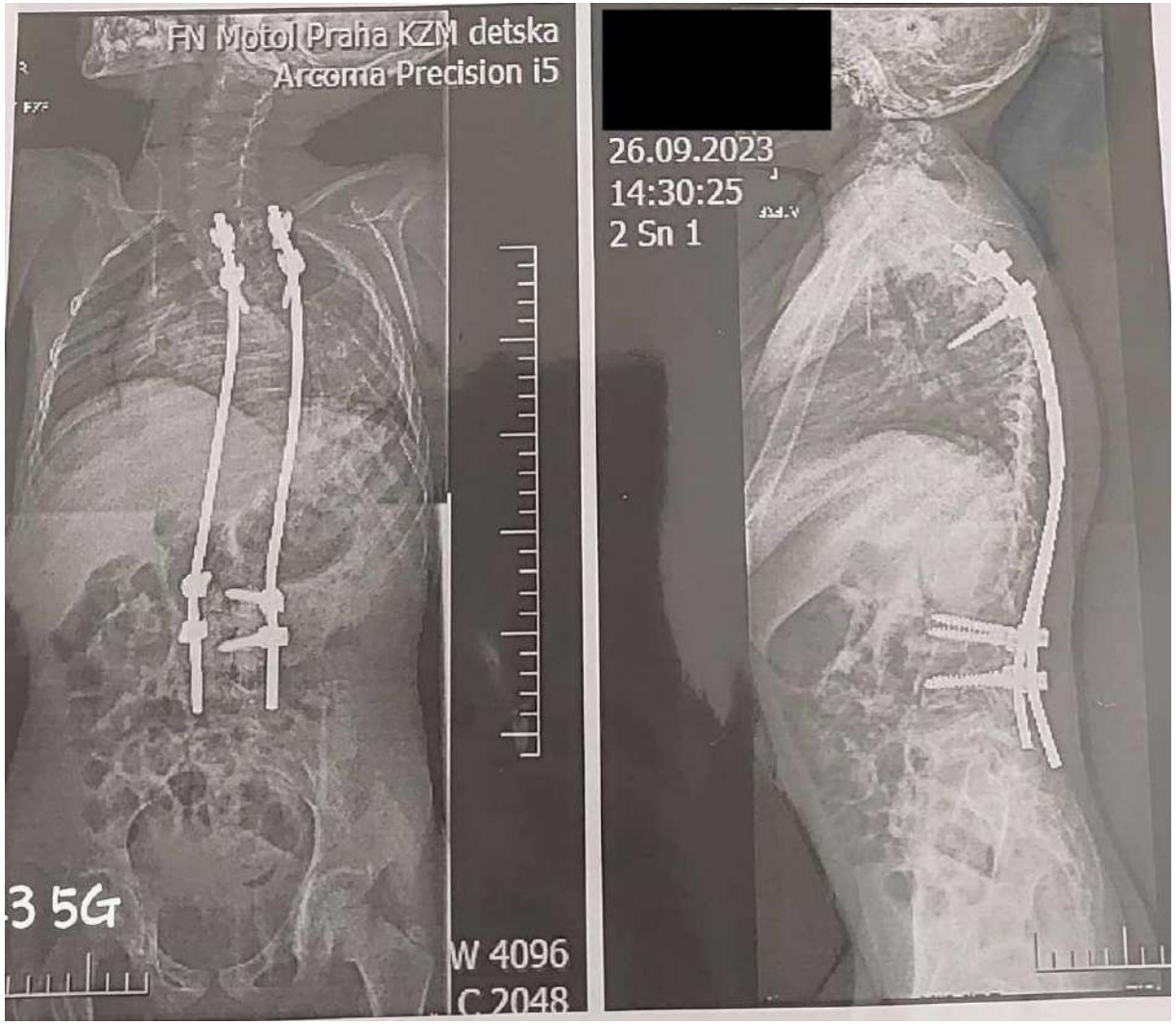


severe kyphoscoliosis in kEDS patient

Pre surgery

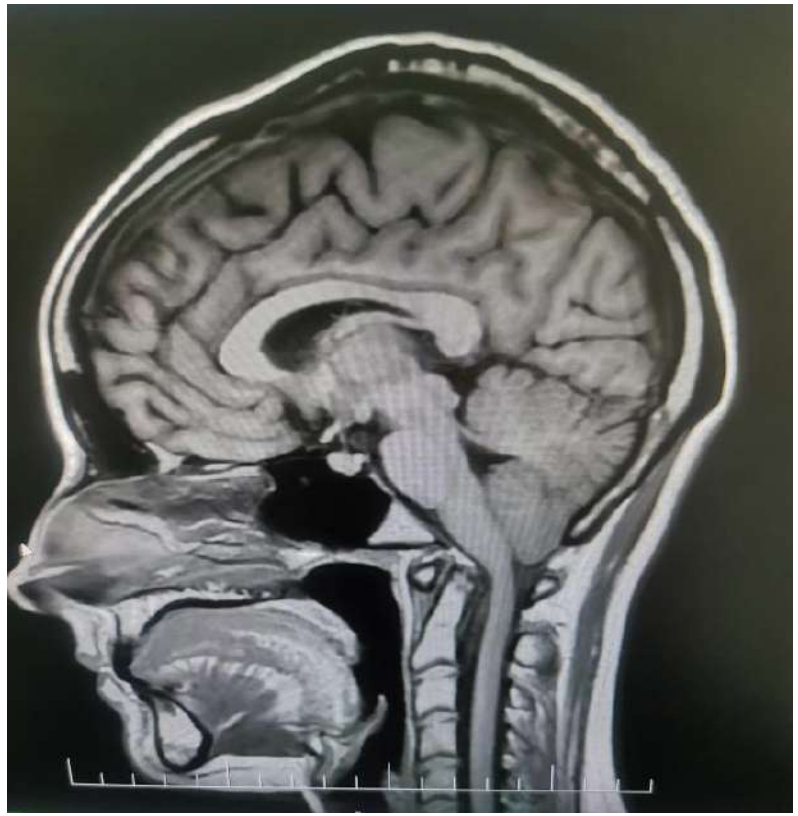


Post surgery

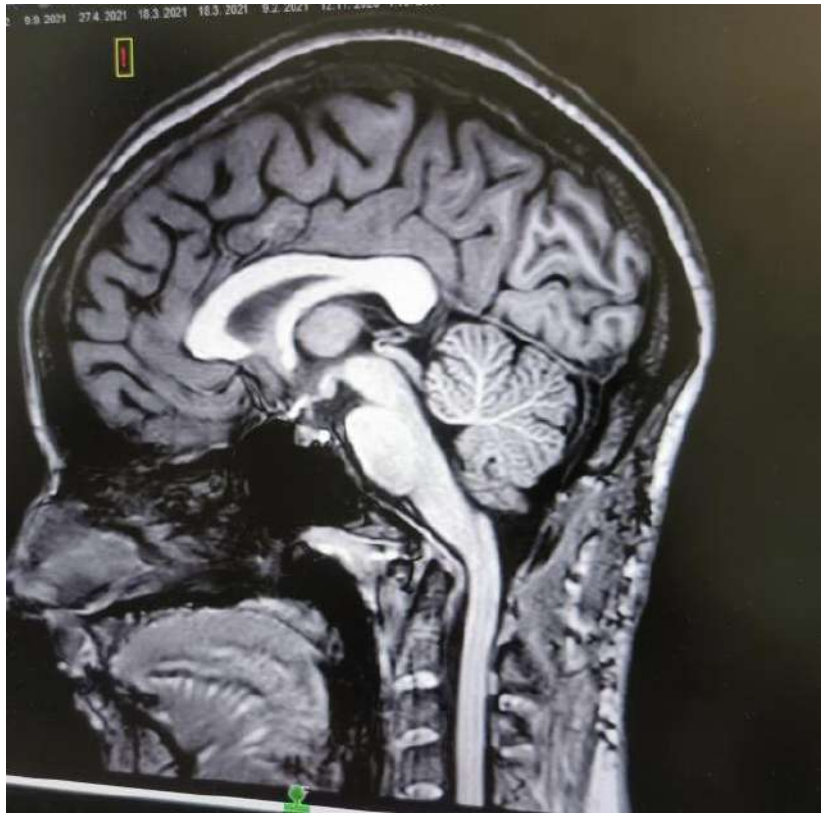


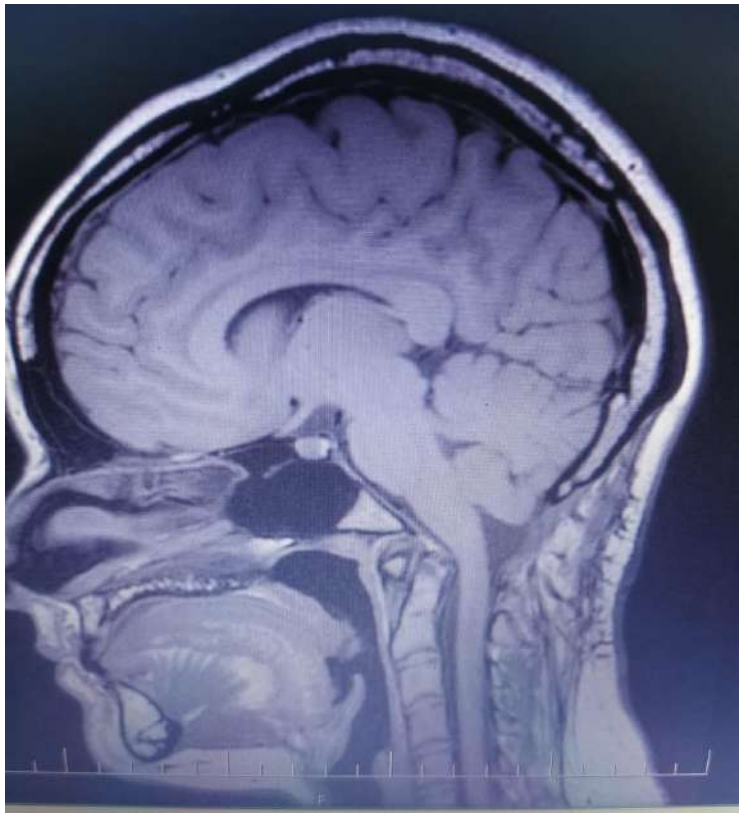
Chiari Malformation

Pre surgery



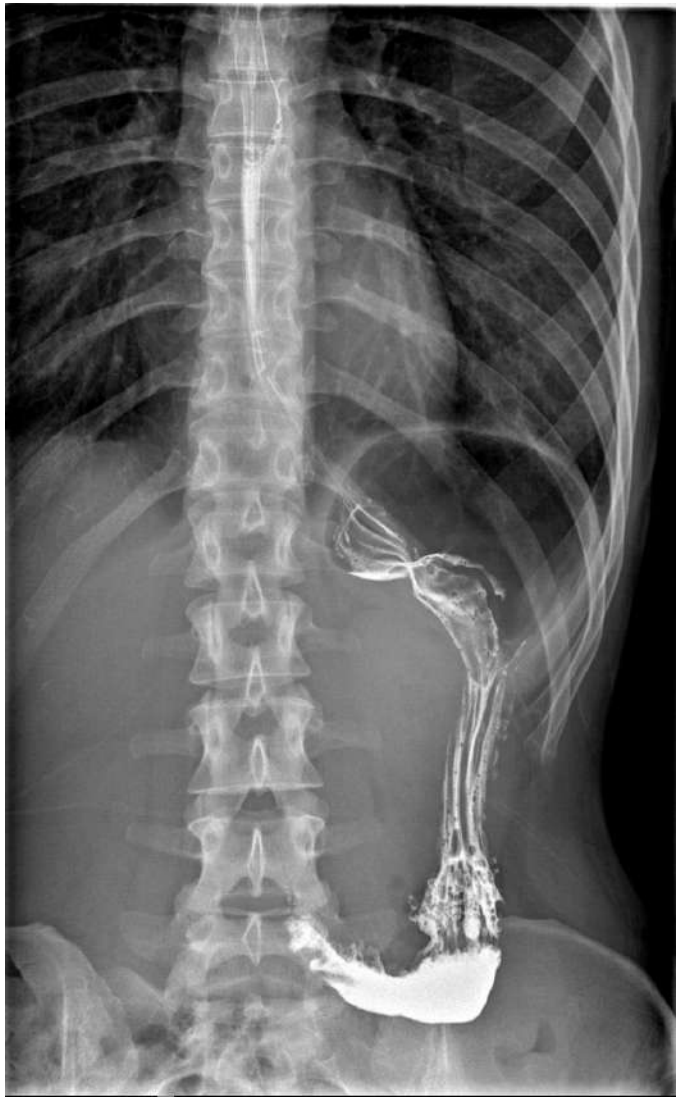
Post surgery

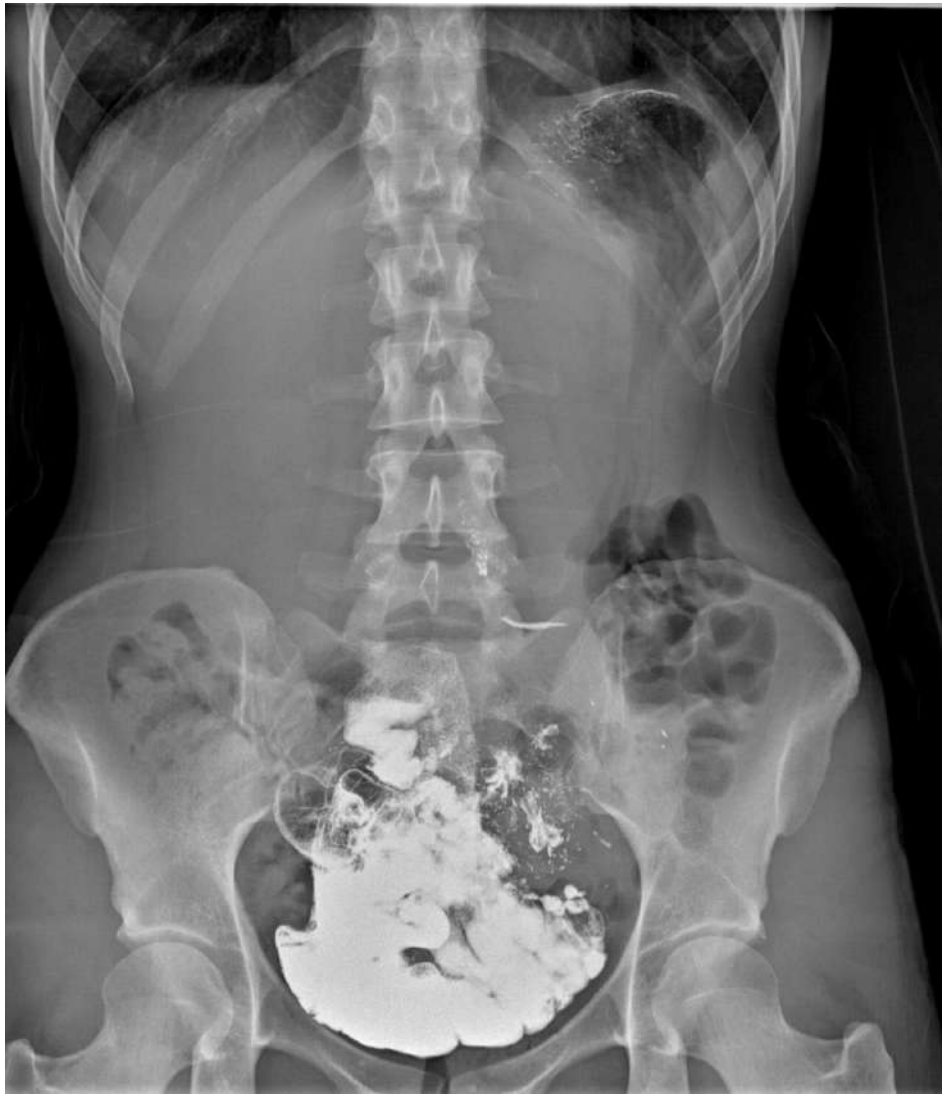


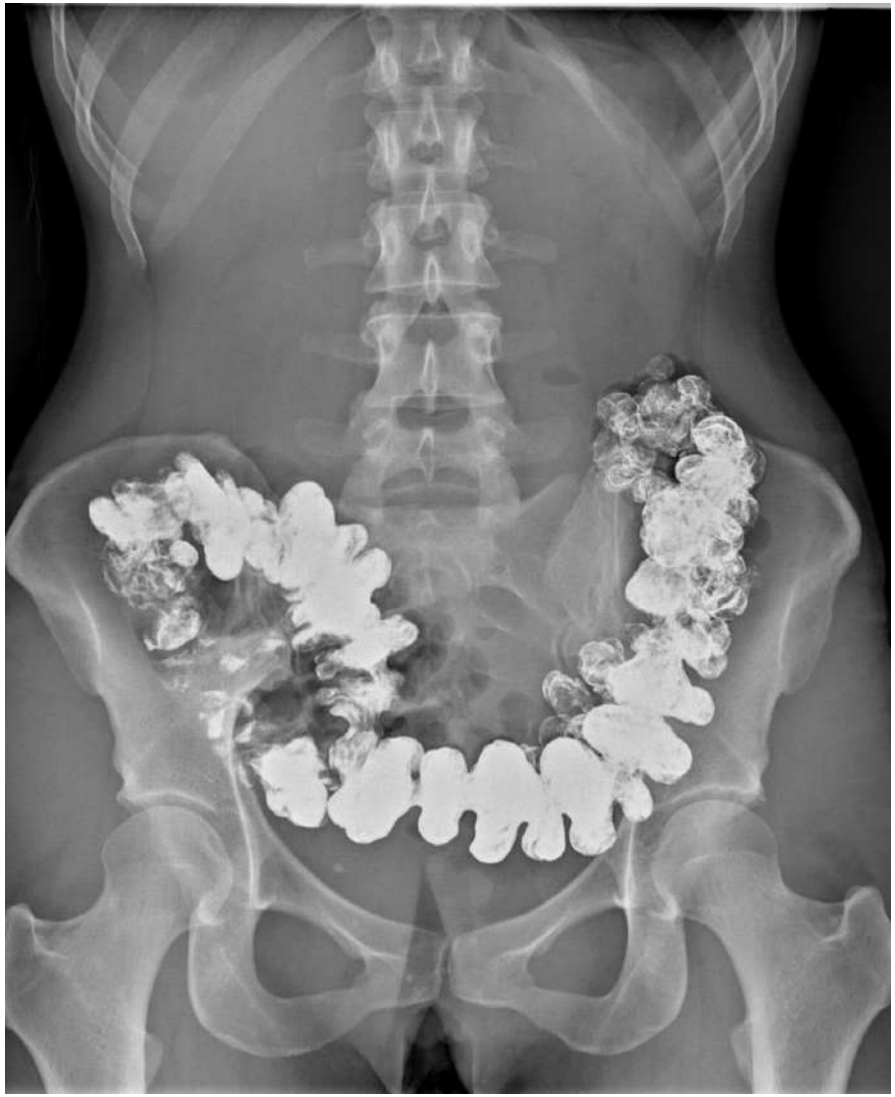


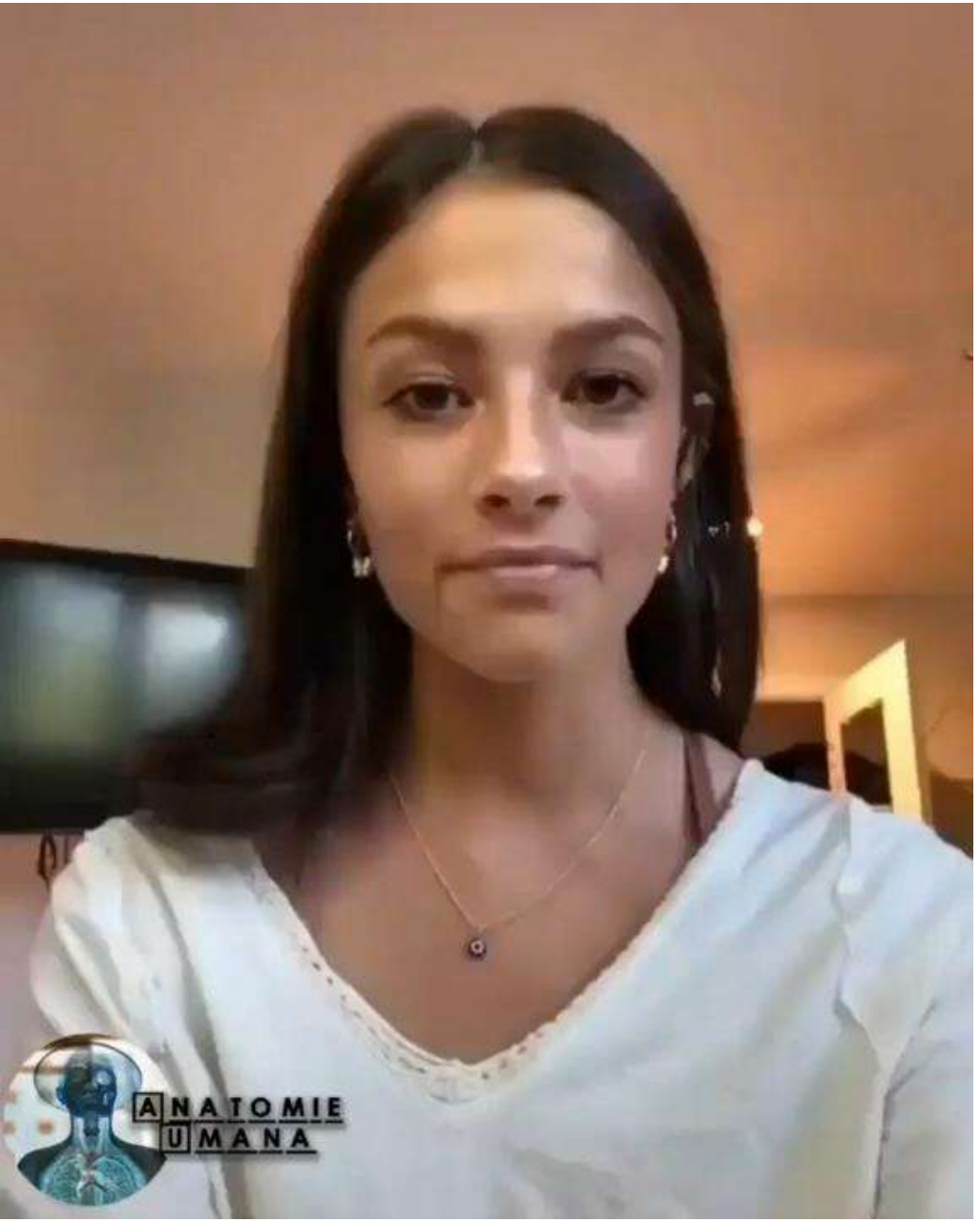
decompressive craniectomy, tonsillectomy, duroplasty, C1 laminectomy

Prolapses

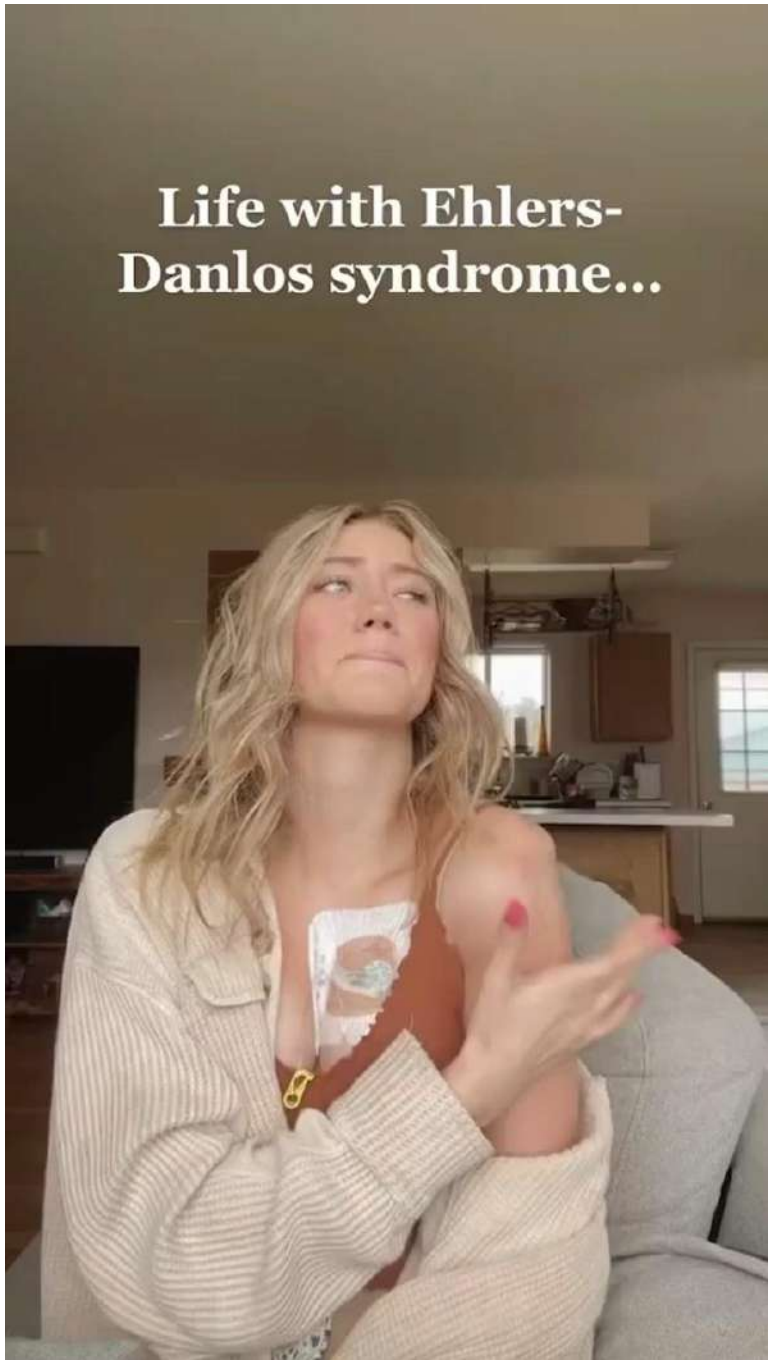







ANATOMIE
UMANA


Doctor React: A man with Ehlers Danlos Syndrome


Life with Ehlers-
Danlos syndrome...

UROGYNAECOLOGIC COMORBIDITIES

- Visceral and pelvic organ prolapse (POP) and pelvic floor dysfunction
  - bladder dysfunction, Pudendal neuralgia
- Pregnancy and menstrual cycle complications
  - infertility, spontaneous abortions, prelabor membrane rupture, preterm labor, failure to progress in labor, etc.
- Vascular Compression Syndromes (NCS & MTS -> PCS MALS, SMAS)
- Autonomic Dysfunction (POTS)
  - bladder dysfunction
- Mast Cell Activation Syndrome (MCAS)
  - Interstitial cystitis / Painful bladder syndrome
- Spinal instabilities (CCI, AAI)
- Slipped Rib Syndrome (SRS)
- Endometriosis, Adenomyosis, PCOS

4.3 | Gynecological manifestations

Gynecological manifestation of hEDS can range from pelvic organ prolapse (POP), to pregnancy and menstrual cycle complications. In a small sample of patients with unspecified EDS subtypes, patients experienced both urinary incontinence and history of POP.[115] POP has also been found to be more common in patients with benign joint hypermobility syndrome.[115,116] In a patient-reported survey, infertility issues have been reported in 44% of patients with EDS, hEDS was the most affected type of EDS compared to 10% of general population.[117] Spontaneous abortions have been reported in 28% of hEDS patients and 57% of EDS patients while impacting only 15% of the general population.[117,118] Despite some evidence of pregnancy complications, including prelabor membrane rupture, preterm labor and failure to progress in labor,[119] other published data indicates that hEDS/JHS are associated with a normal risk of serious adverse pregnancy outcomes.[120]

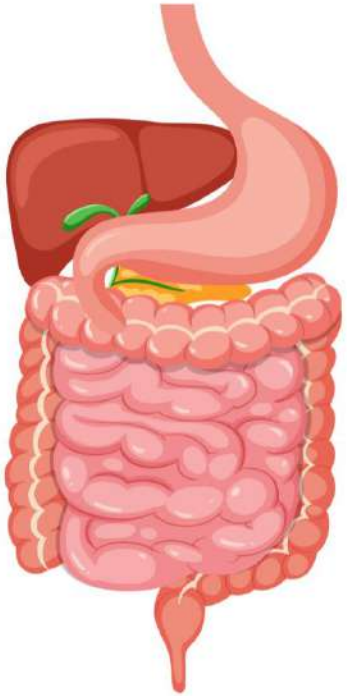
An increase in dislocations and symptoms at puberty, during pregnancy, postpartum and during the perimenstrual period have both been reported along with an improvement after menopause.[117,118] In the general population, ligament laxity has been shown to be influenced by estrogen, progesterone, relaxin and testosterone and has been best evaluated in the context of anterior cruciate ligament (ACL) injury in females. Knee ligament laxity and risk of ACL injury occurs more frequently during preovulatory phase and ovulatory phase of the menstrual cycle, when estrogen exceeds progesterone.[121-123] Hormonal contraceptives have been found to have a possible protective role in ACL tears.[122,124,125] The influence of hormones on ligament laxity, combined with patient-reported fluctuations in symptoms that coincide with hormonal shifts, indicate that more research is needed to establish the role of hormones in hEDS.

GI COMORBIDITIES

- Vascular Compression Syndromes (MALS, NCS, SMAS, MTS)
- Autonomic Dysfunction (POTS – Postural Orthostatic Tachycardia Syndrome)
- Mast Cell Activation Syndrome (MCAS)
- Functional gut dysmotility (gastroparesis, etc.)
- Prolapses - Visceroptosis (gastroptosis, enteroptosis, coloptosis, nephroptosis, rectal and pelvic prolapse, rectocele, etc.)
- Hernias (hiatus hernia, etc.)
- Spinal instabilities (CCI, AAI)
- Slipped Rib Syndrome (SRS)
- Esophageal disorders (GERD, Eosinophilic esophagitis)
- Temporomandibular disorders

GI COMORBIDITIES

Hypermobile EDS diagnostic criteria is consistently limited to skin fragility or elasticity and hypermobile joints.[2] Due to the emphasis on specific joint and skin elasticity, diagnostic criteria for hEDS frequently neglects gastrointestinal manifestations, despite their high prevalence.[170] The frequency of gastrointestinal symptoms is higher than previously assessed among hEDS patients. While GI symptoms experienced by affected individuals are primarily functional and nonlife threatening in nature, their impact upon the patient's quality of life is significant. Clinical assessment of gastrointestinal symptoms associated with hEDS should be constructed to address diagnosed and under-treated gastrointestinal complaints among hEDS patients.[170] Gastrointestinal complaints are common in EDS and generalized joint hypermobility.[88,171-173] Abdominal pain, bloating, nausea, reflux symptoms, vomiting, constipation and diarrhea are commonly experienced GI symptoms.[171] In a widespread survey inquiring about GI symptoms among hEDS populations, 79.3% of participants reported gastroesophageal disease (GERD), 48% reported symptoms congruent with irritable bowel syndrome, and 36% reported motility issues, specifically functional constipation.[170] Dysmotility and delayed gastric emptying (gastroparesis) was highly reported, which may be attributed to the high prevalence of dysautonomia among hEDS patients.[171,174]



Gastrointestinal physiological studies by Mayo Clinic surveyed 36 EDS patients of various subtypes, a majority of whom presented with type III (hypermobile type): 28% of patients who underwent colonic transit studies had abnormal results, with either slow or fast transit.[172] There is currently no standardized clinical assessment nor care guidelines for the management of hEDS-related gastrointestinal symptoms.[174] Anatomical abnormalities among hEDS patients may be attributed to structural changes in collagen located in the smooth muscle of gastrointestinal pathology, presenting as diverticulosis, rectoceles, and prolapse. Celiac disease is also reported to be more prevalent in hEDS.[175] Recurrent abdominal pain, chronic gastritis and constipation/diarrhea was reported by hEDS patients.[176]

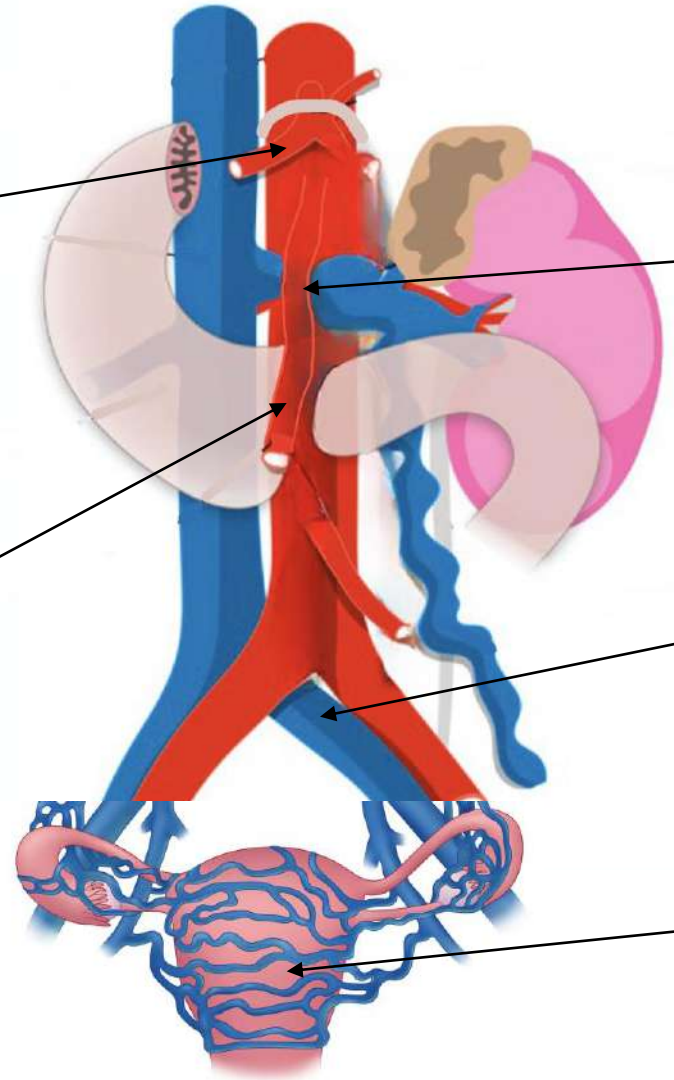
DIAGNOSTIC TOOLS

Vascular Compression Syndromes	CTAG – oral & IV contrast, doppler ultrasound, venography, IVUS, + tests below
Autonomic Dysfunction *(Neuro/Cardiovascular)*	CARTs – cardiovascular autonomic reflex tests: HUTT / NASA Lean Test - orthostatic test , Valsalva maneuver, deep breathing, QSART – quantitative sudomotor axon reflex test, skin biopsy for SFN
Mast Cell Activation Syndrome *(Allergoimmuno)*	CD117 staining, tryptase in blood, leukotrienes in urine (IMD lab Berlin), Colontransit-time study
Functional GI Dysmotility *(gastroparesis, dysphagia, etc.)*	Scintigraphy / Gammagraphy RTG barium study (SMAS), Smart Pill, Colontransit-time study, Manometry
Prolapses / Visceroptosis	RTG barium study, CT with oral & IV contrast, MRI
Hernias, Spinal instabilities, Slipped Rib Syndrome, etc.	MRI, physical examination, etc.

ABDOMINO-PELVIC VASCULAR COMPRESSION SYNDROMES

Median Arcuate Ligament Syndrome
MALS
(Dunbar S.)

Superior Mesenteric Artery Syndrome
SMAS
(Wilkie S.)

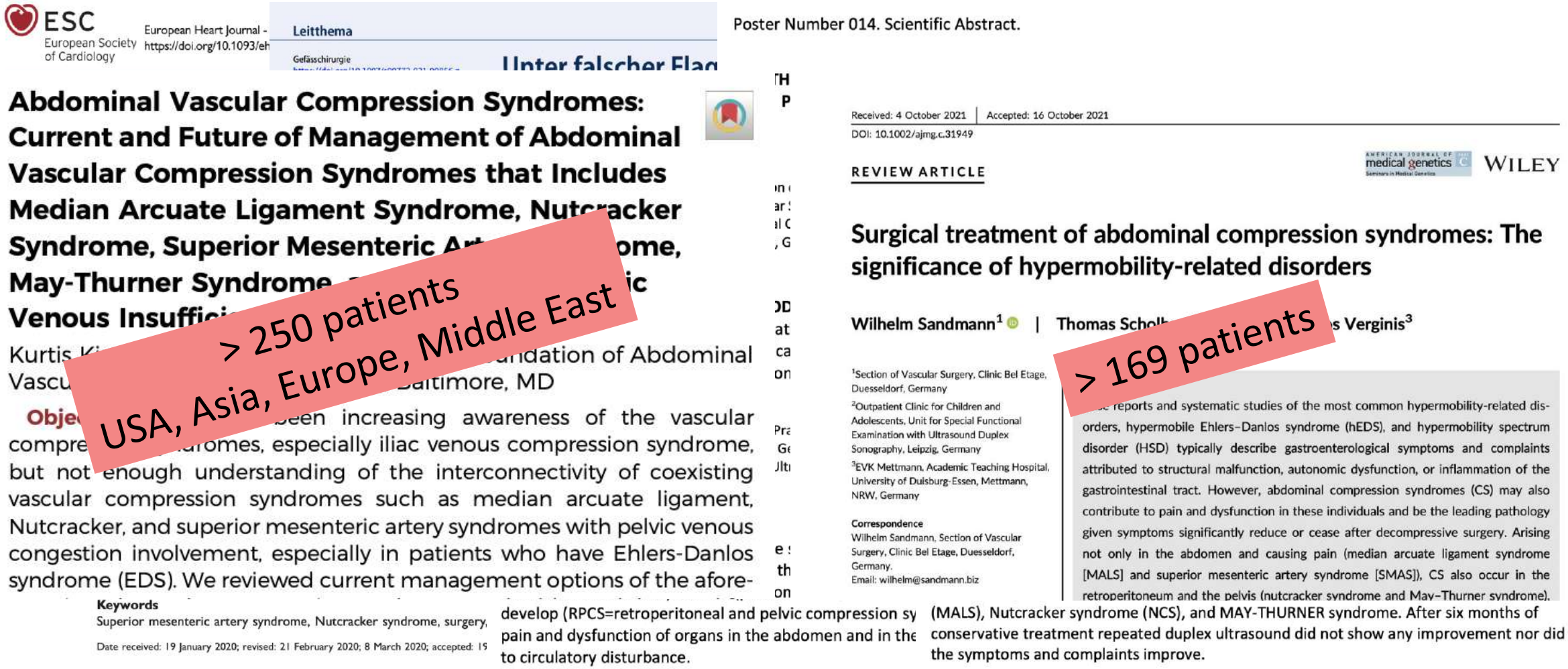


Left Renal Vein Compression Syndrome
NCS
(Nutcracker S.)

Iliac Vein Compression Syndrome
MTS
(May Thurner Syndrome)

Pelvic Congestion Syndrome
PCS /
Pelvic Venous Insufficiency
PVI

PUBLISHED RESEARCH



ESC European Society of Cardiology

European Heart Journal - https://doi.org/10.1093/eh

Leitthema

Gefässchirurgie

Unter falscher Flag

Poster Number 014. Scientific Abstract.

Abstract

Abdominal Vascular Compression Syndromes: Current and Future of Management of Abdominal Vascular Compression Syndromes that Includes Median Arcuate Ligament Syndrome, Nutcracker Syndrome, Superior Mesenteric Ar ome, May-Thurner Syndrome ic Venous Insuffici

> 250 patients

USA, Asia, Europe, Middle East

Kurtis Ki ndation of Abdominal Vascu Baltimore, MD

Obje been increasing awareness of the vascular compre romes, especially iliac venous compression syndrome, but not enough understanding of the interconnectivity of coexisting vascular compression syndromes such as median arcuate ligament, Nutcracker, and superior mesenteric artery syndromes with pelvic venous congestion involvement, especially in patients who have Ehlers-Danlos syndrome (EDS). We reviewed current management options of the afore-

develop (RPCS=retroperitoneal and pelvic compression sy pain and dysfunction of organs in the abdomen and in the to circulatory disturbance.

Keywords

Superior mesenteric artery syndrome, Nutcracker syndrome, surgery,

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REVIEW ARTICLE

AMERICAN JOURNAL OF medical genetics C Seminars in Medical Genetics

WILEY

Surgical treatment of abdominal compression syndromes: The significance of hypermobility-related disorders

> 169 patients

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[2]Outpatient Clinic for Children and Adolescents, Unit for Special Functional Examination with Ultrasound Duplex Sonography, Leipzig, Germany

[3]EVK Mettmann, Academic Teaching Hospital, University of Duisburg-Essen, Mettmann, NRW, Germany

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reports and systematic studies of the most common hypermobility-related disorders, hypermobile Ehlers–Danlos syndrome (hEDS), and hypermobility spectrum disorder (HSD) typically describe gastroenterological symptoms and complaints attributed to structural malfunction, autonomic dysfunction, or inflammation of the gastrointestinal tract. However, abdominal compression syndromes (CS) may also contribute to pain and dysfunction in these individuals and be the leading pathology given symptoms significantly reduce or cease after decompressive surgery. Arising not only in the abdomen and causing pain (median arcuate ligament syndrome [MALS] and superior mesenteric artery syndrome [SMAS]), CS also occur in the retroperitoneum and the pelvis (nutcracker syndrome and May–Thurner syndrome). (MALS), Nutcracker syndrome (NCS), and MAY-THURNER syndrome. After six months of conservative treatment repeated duplex ultrasound did not show any improvement nor did the symptoms and complaints improve.

SK

STUDY POPULATION

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Surgical Treatment of Wilkie's Syndrome by Vascular Transposition

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Abstract

Introduction

Superior mesenteric artery syndrome (SMAS), also called mesenteric duodenal compression syndrome, Wilkie's syndrome, chronic duodenal ileus or cast syndrome, is a rare clinical condition defined as a compression of the third portion of the duodenum in between the SMA and abdominal aorta (AA), due to narrowing of the space between them. SMAS is primarily attributed to loss of the intervening mesenteric fat pad, leading to partial or complete duodenal obstruction. Its manifestations are complex and non-specific, including postprandial epigastric pain, nausea, vomiting, early satiety, weight loss and anorexia. SMAS may present as an acute syndrome, or it may have an insidious onset with chronic symptoms. SMAS mainly affects females between 10 and 40 years of age. This study aims to discuss the safety and efficacy of vascular decompression of the duodenum by infrarenal transposition of SMA.

90 patients

Research Article

International Journal of Probiotics and Dietetics

Management of Wilkie's Syndrome in Vascular surgery

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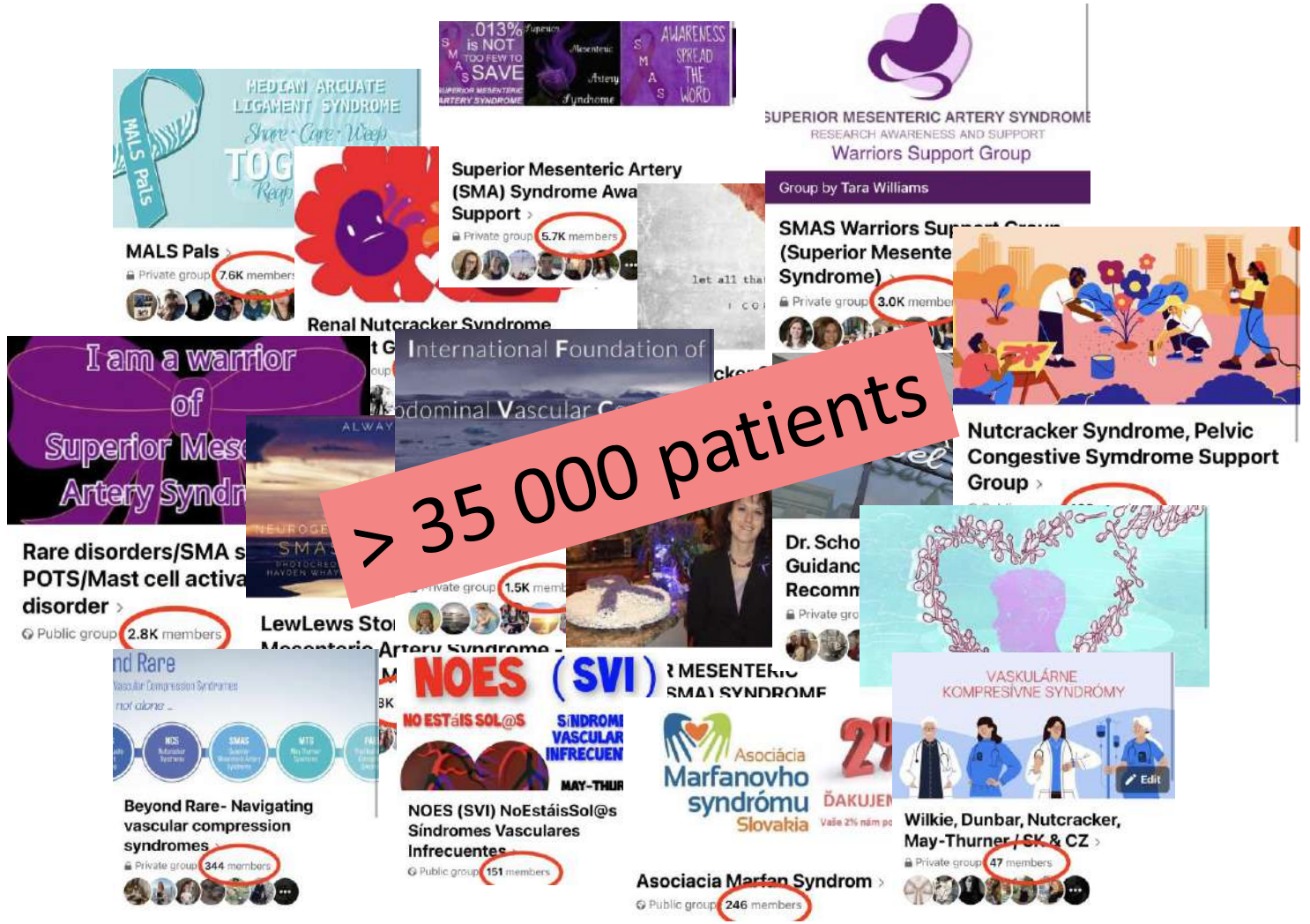

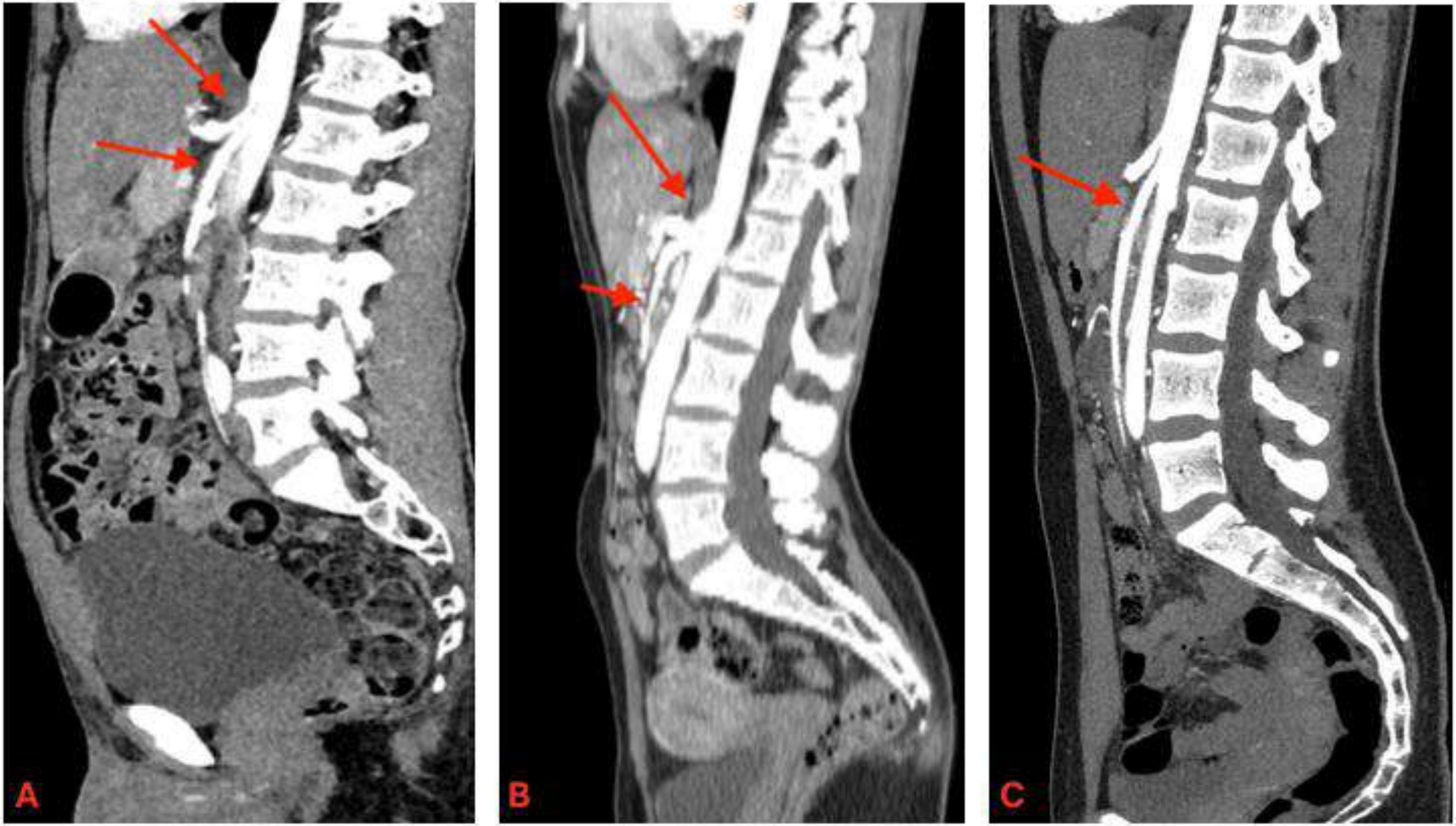
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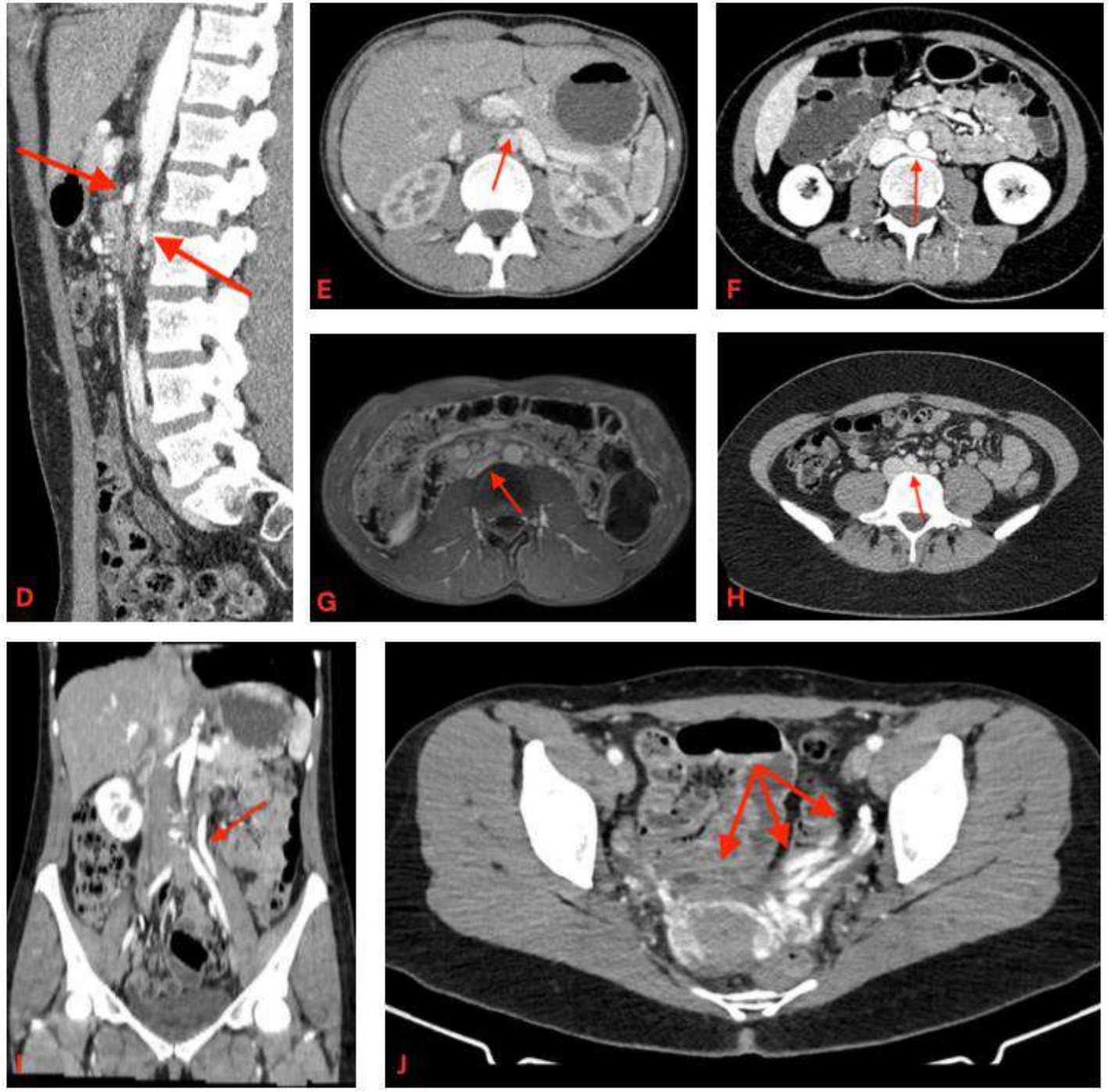
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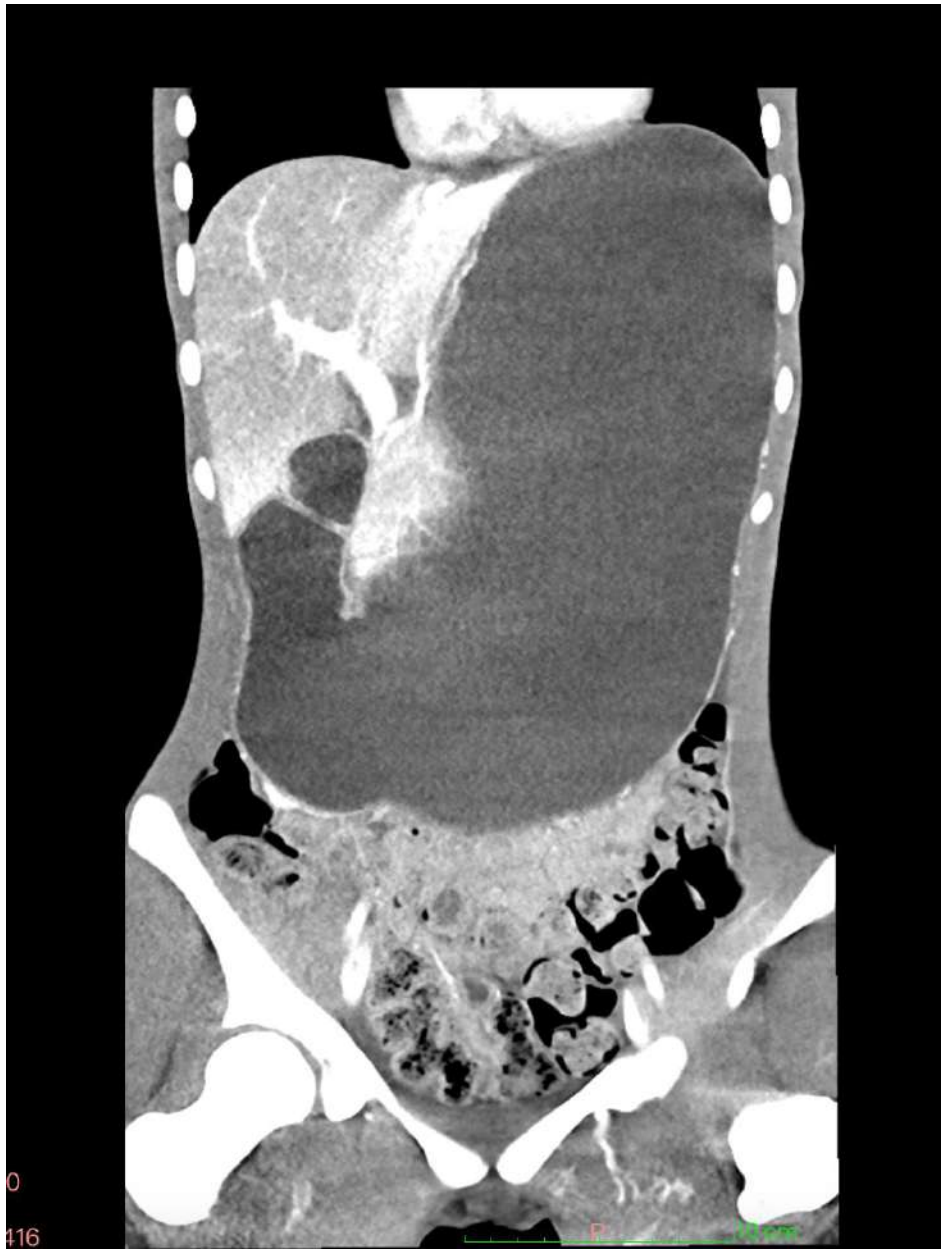
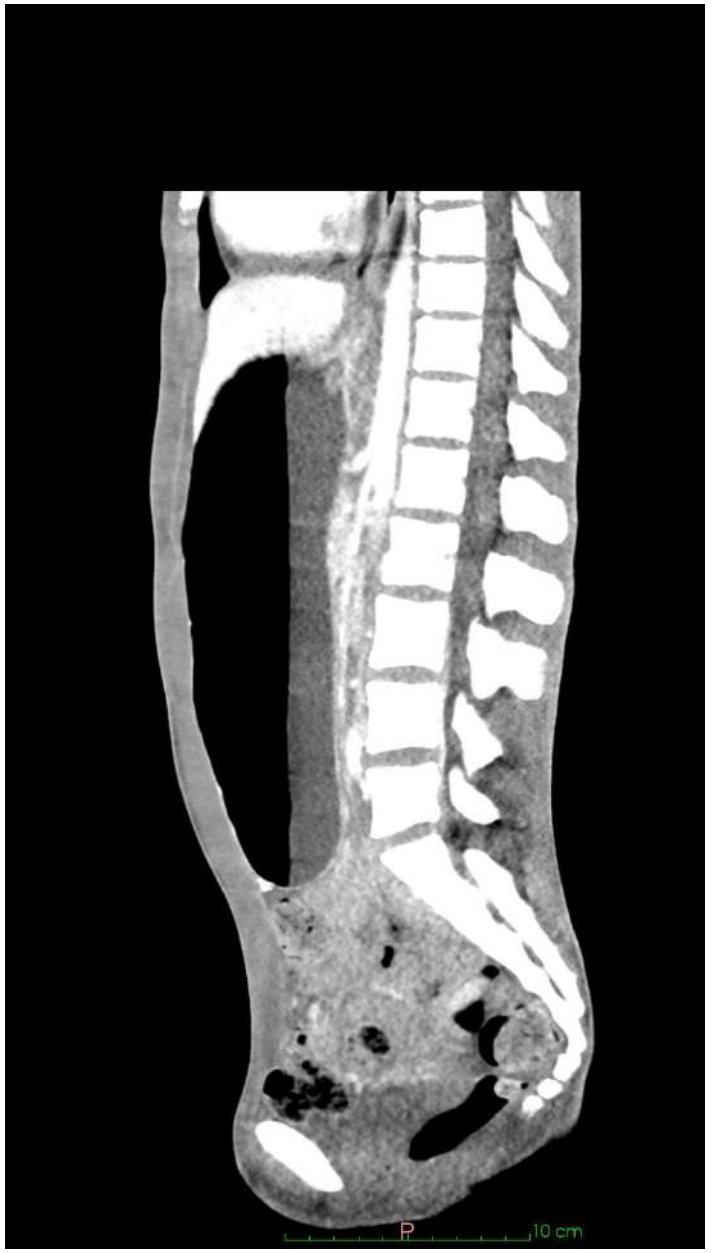
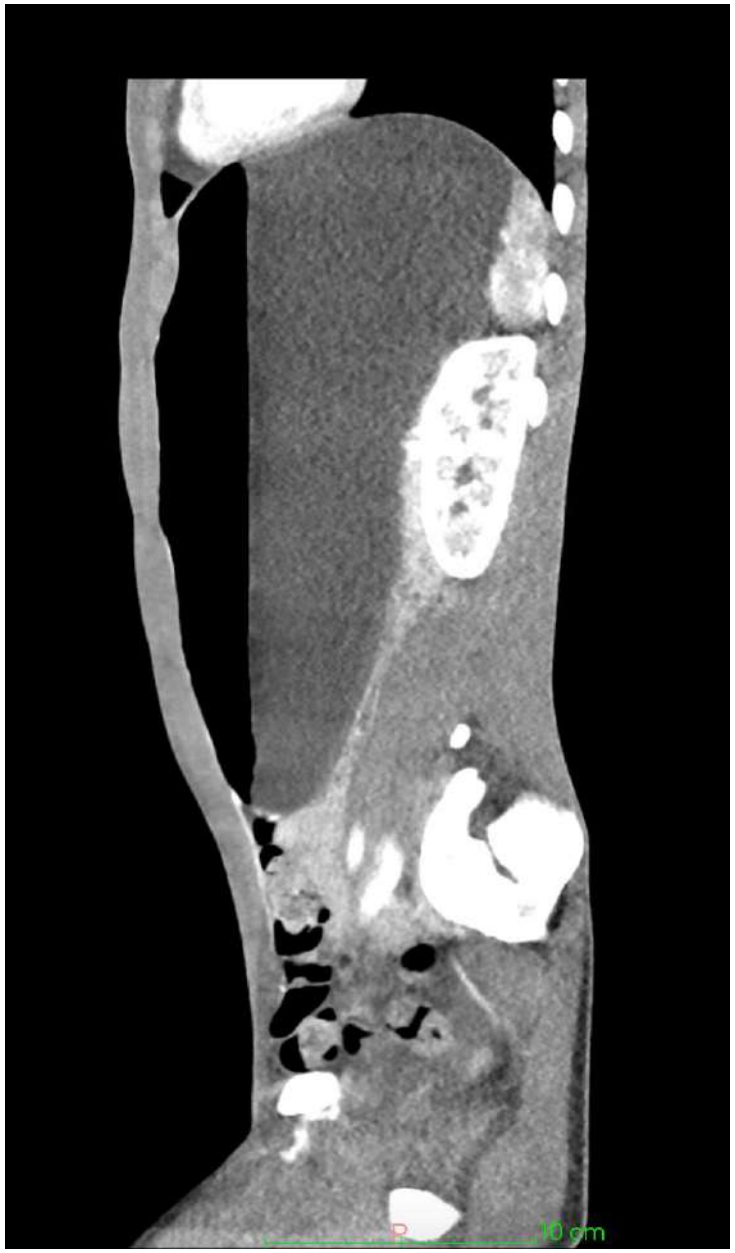




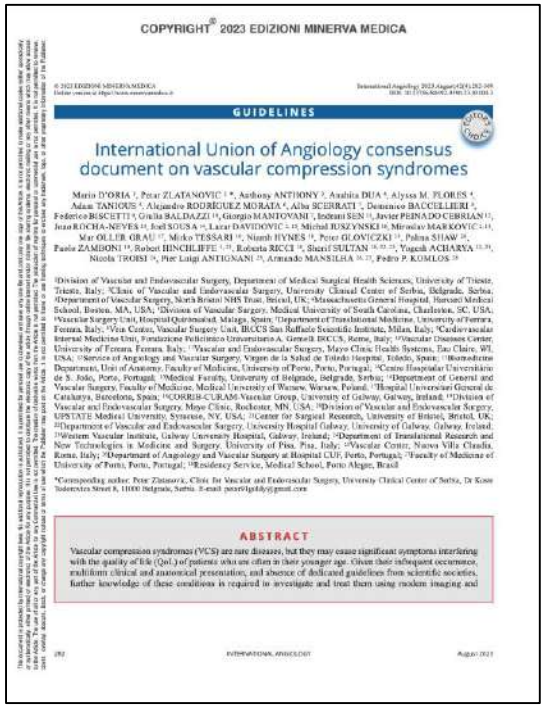


… also possible IAH and ACS

DIAGNOSTIC IMAGING

DIAGNOSTING IMAGING

Angiología y Cirugía Vascular
Dr. Rodríguez Morata



GUIDELINES

International Union of Angiology consensus document on vascular compression syndromes

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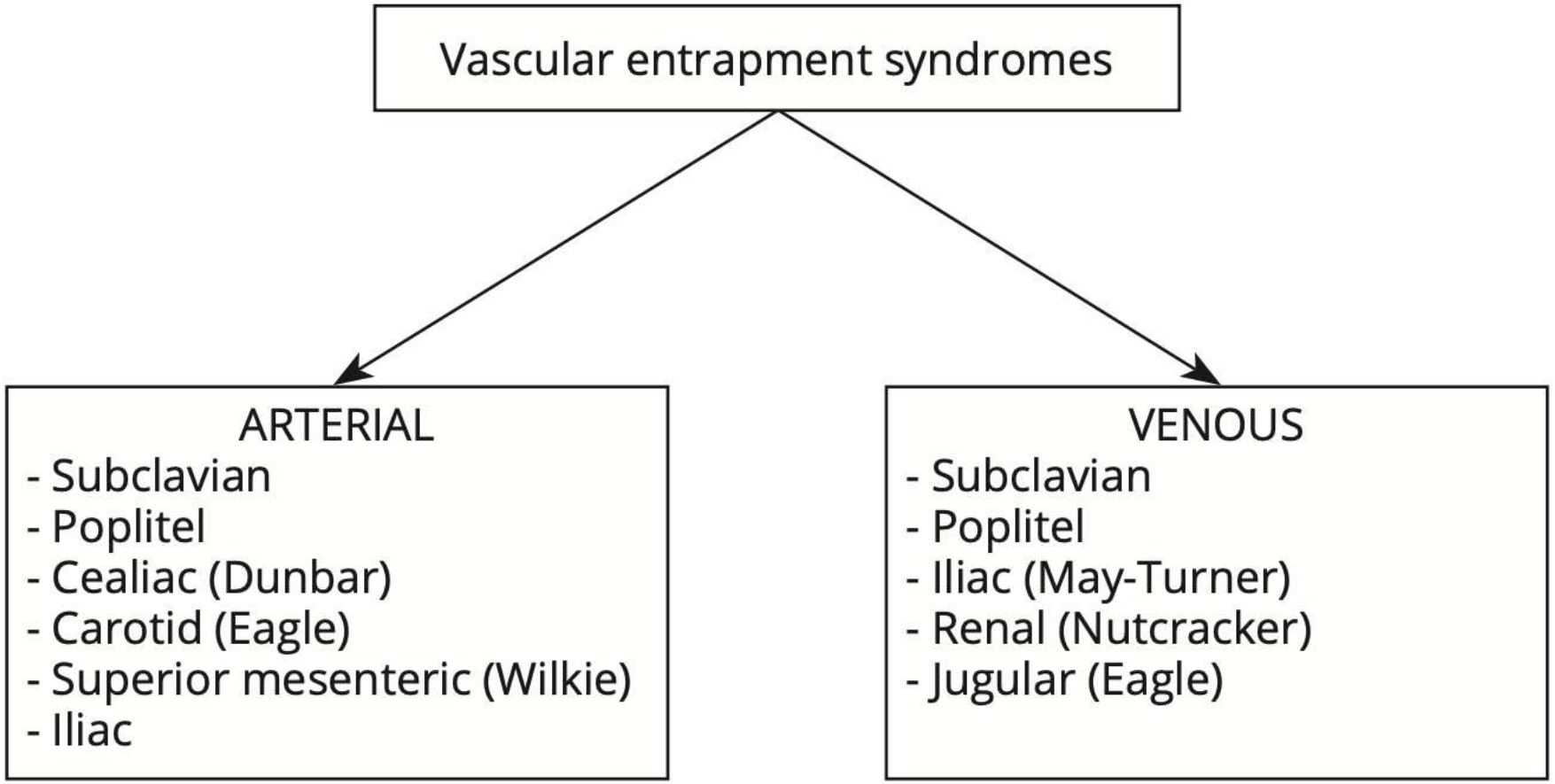
ABSTRACT

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

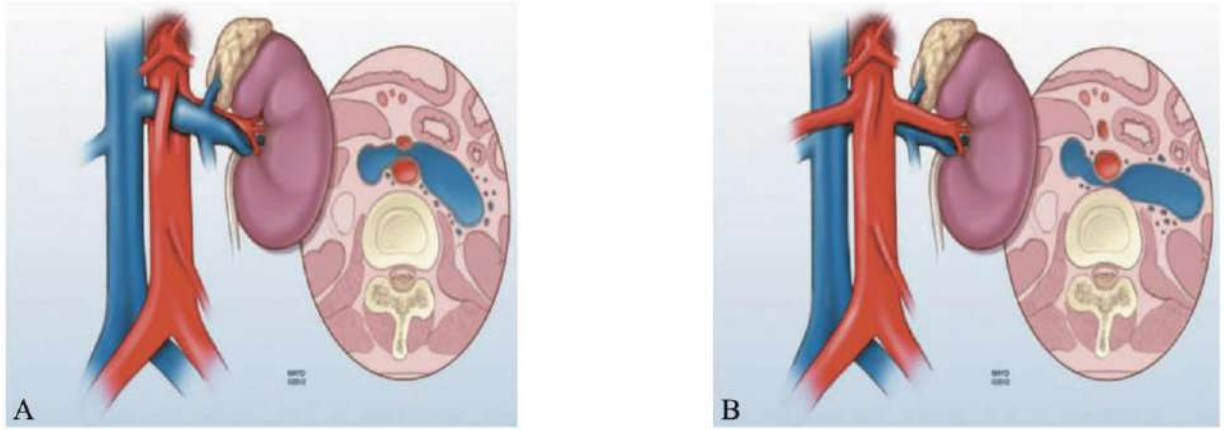
TABLE I.—*Imaging examinations for diagnosis of vascular compression/entrapment syndromes.*

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

D I A G N O S T I N G I M A G I N G

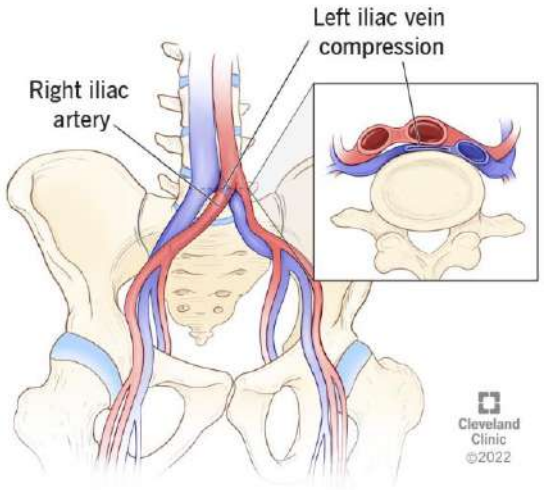


N C S – Nutcracker's Phenomenon / Left Renal Vein Compression S.



Nutcracker Syndrome			
Mechanisms	Symptoms	Diagnosis	Management
• frequent incidental finding • anterior, posterior / retro-aortic, circum-aortic • renal ptosis • astheic body habitus, min. retroperitoneal fat • lumbar lordosis • abnormally high LRV course, abnormal SMA branching • excess fibrolymphatic tissue • other compressions (testicular artery, para-aortic	• asymptomatic NCS > 50% narrowing LRV - 51-72% CT cases • abdominal, left-flank pain • hematuria, proteinuria • varicocele, pelvic congestion syndrome • dyspareunia, dysuria, dysmenorrhea • polycystic ovaries, orthostatic intolerance • anemia, chronic fatigue • other compression s. - May-Thurner s.	• duplex ultrasonography • computed tomography venography • contrast venography • IVUS	• conservative (weight gain, physiotherapy - back, antithrombotic therapy • endovascular approach: stenting • open surgery: renocaval prosthetic bypass, LRV, LOV, SMA transposition, renal autotransplantation, PTFE external stent, radical nephrectomy • no safe and efficient technique • no dedicated stent

M T S – May Thurner Syndrome / Iliac Vein Compression S.



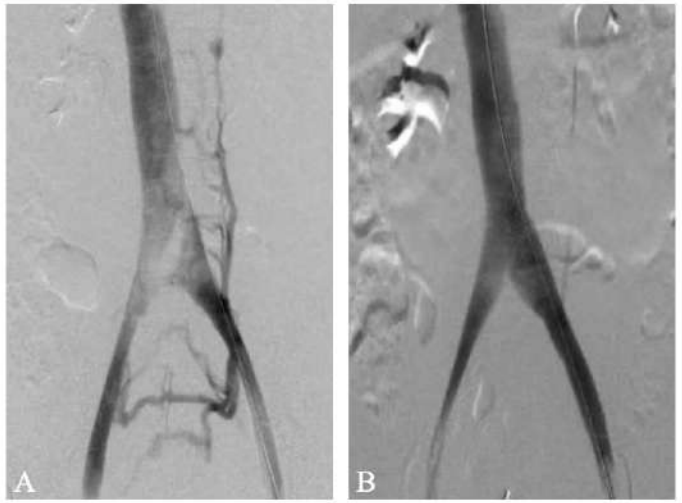
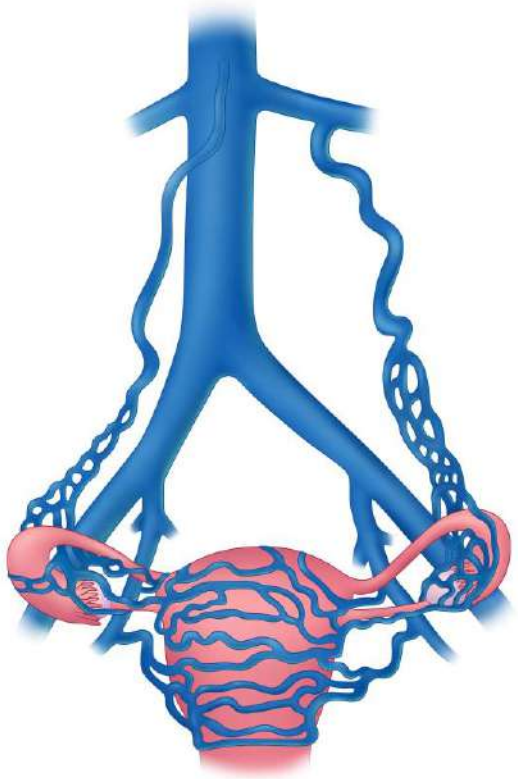


Figure 13.—Intraoperative phlebography before (A) and after the stenting of left common et external iliac vein (B).

May Thurner Syndrome			
Mechanisms	Symptoms	Diagnosis	Management
• LCIV compressed by RCIA • RCIV compressed by RCIA / LCIA / RIIA / REIA • LEIV compressed by LEIA • REIV compressed by REIA • IVC compressed by RCIA • Stage 1: asymptomatic LCIV compression • Stage 2: formation of intraluminal spurs • Stage 3: occurrence of left iliac deep vein thrombosis	• leg swelling, oedema, venuous claudication, symptomatic varicoe vein, phlebitis, DVT • orthostatic intolerance, pelvic congestion syndrome, functional GI motility issues, urinary issues	• duplex ultrasonography • plethysmography • computerized tomography / magnetic resonance • venography • IVUS	• conservative (compression stockings, anticoagulation therapy) • endovascular approach • open surgery: transposition, PTFE external stent

P C S - Pelvic Congestion S. / P V I – Pelvic Venous Insufficiency

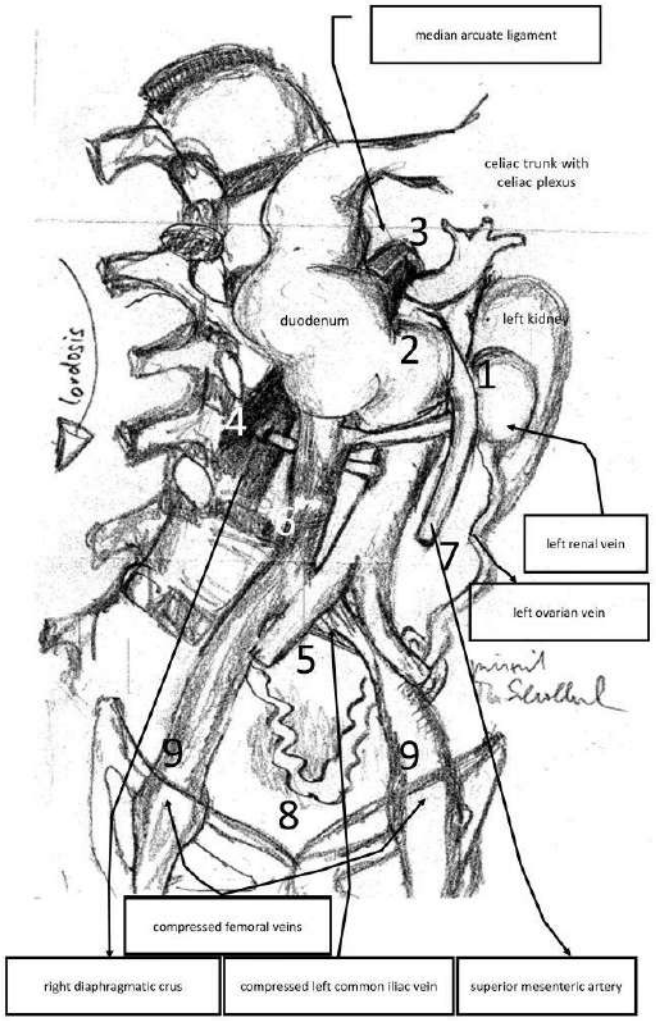


NCS – LRV Compression S.

MTS – IV Compression S.

PCS / PVI

Lordogenetic Midline Congestion Syndrome



Prof. Dr. med. habil.
THOMAS
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LORDOSIS – one of the decisive factors of all APVCS

M A L S – Median Arcuate Ligament Syndrome (Dunbar S.)

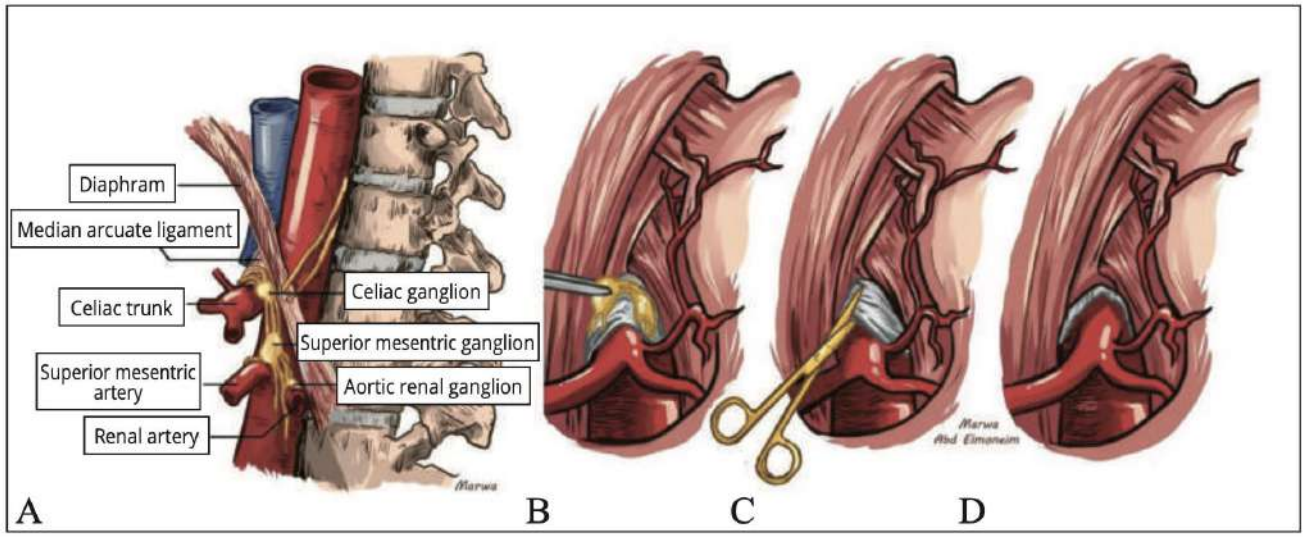
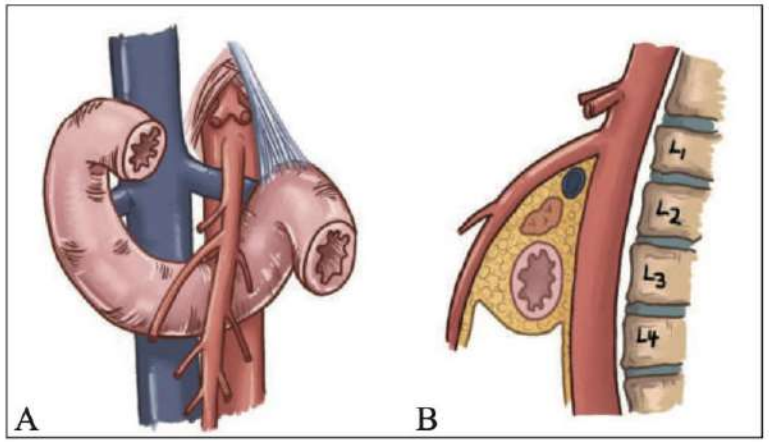
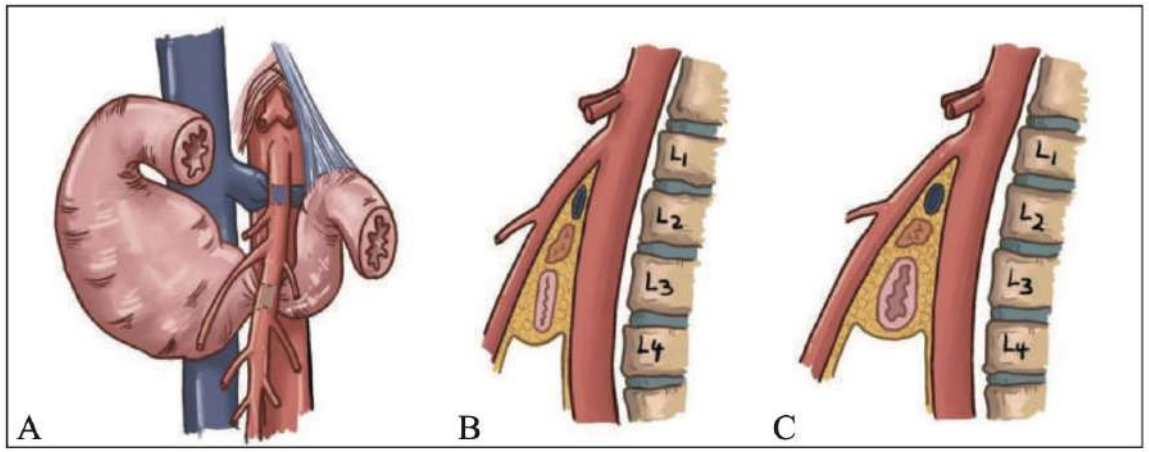


TABLE III.—*Core features of MALS symptoms, diagnosis and management.*

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

S M A S – Superior Mesenteric Artery Syndrome (Wilkie S.)





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

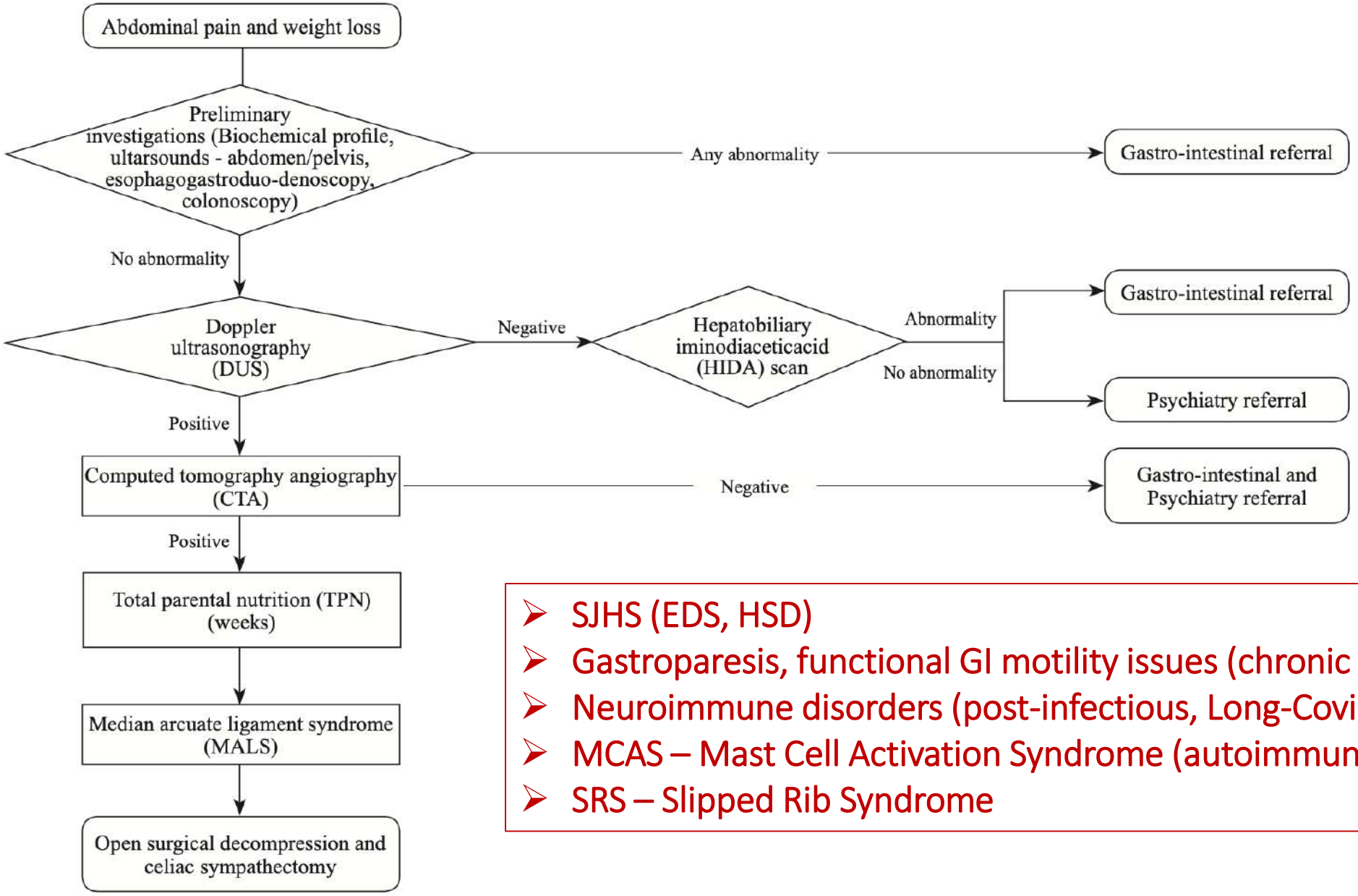


Figure 5.—Practical therapeutic algorithm employed in patients with abdominal pain and weight loss.

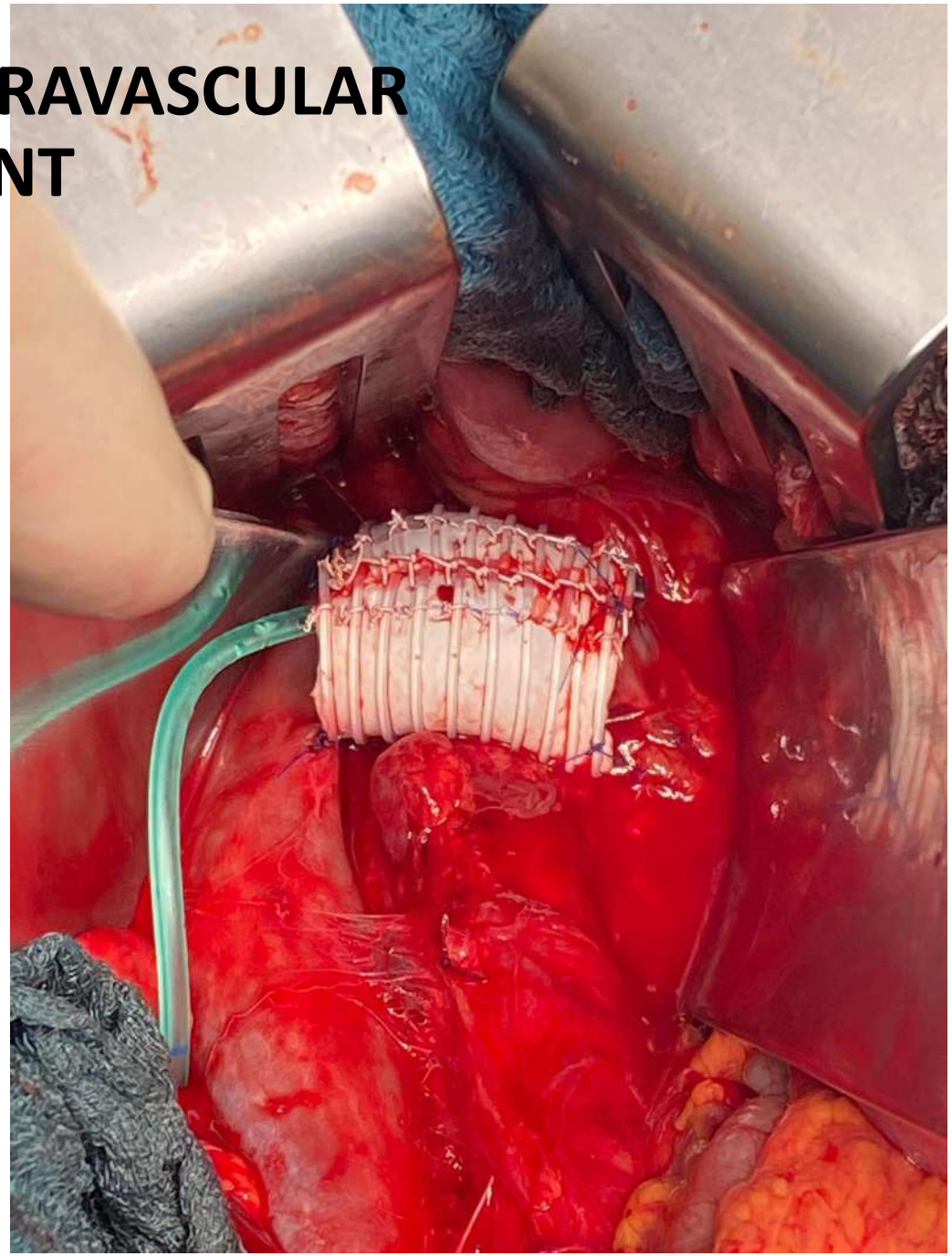
- SJHS (EDS, HSD)
- Gastroparesis, functional GI motility issues (chronic SIBO)
- Neuroimmune disorders (post-infectious, Long-Covid, etc.)
- MCAS – Mast Cell Activation Syndrome (autoimmune)
- SRS – Slipped Rib Syndrome

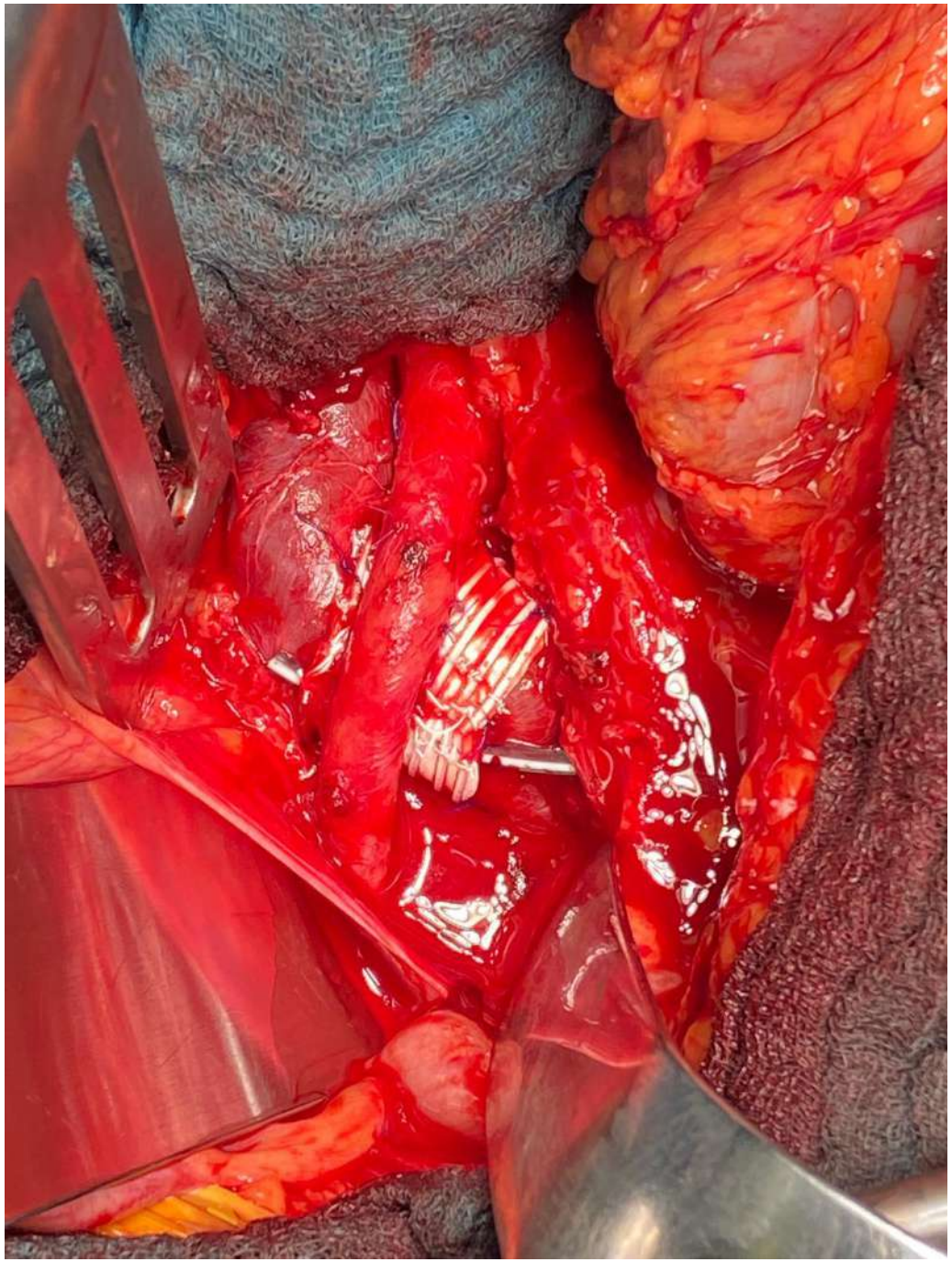
VASCULAR COMPRESSION SYNDROMES

SURGICAL APPROACHES

MALS	NCS	SMAS	MTS	PCS
• MAL release through resection, with / without neurectomy • Laparoscopic / open approach • Celiac plexus block	• Transposition of LRV or LOV into VC, with / without an extension patch • Infrarenal transposition of SMA • Endovascular / extravascular PTFE stents • Renocaval prosthetic PTFE bypass • Hybrid technique: endovascular stent & open surgical sewing of the stent in LRV • Kidney autotransplantation • Radical nephrectomy	• Gastric bypasses, Roux-en-Y: gastrojejunostomy / duodenojejunostomy • Duodenal derotation, modified LADD's procedure • Infrarenal transposition of SMA • PTFE extravascular stents	• Endovascular stents • Iliac vein transpositions	• Depending on primary / secondary PCS • embolization of varicose veins • Treating NCS and/or MTS

EXTRAVASCULAR STENT





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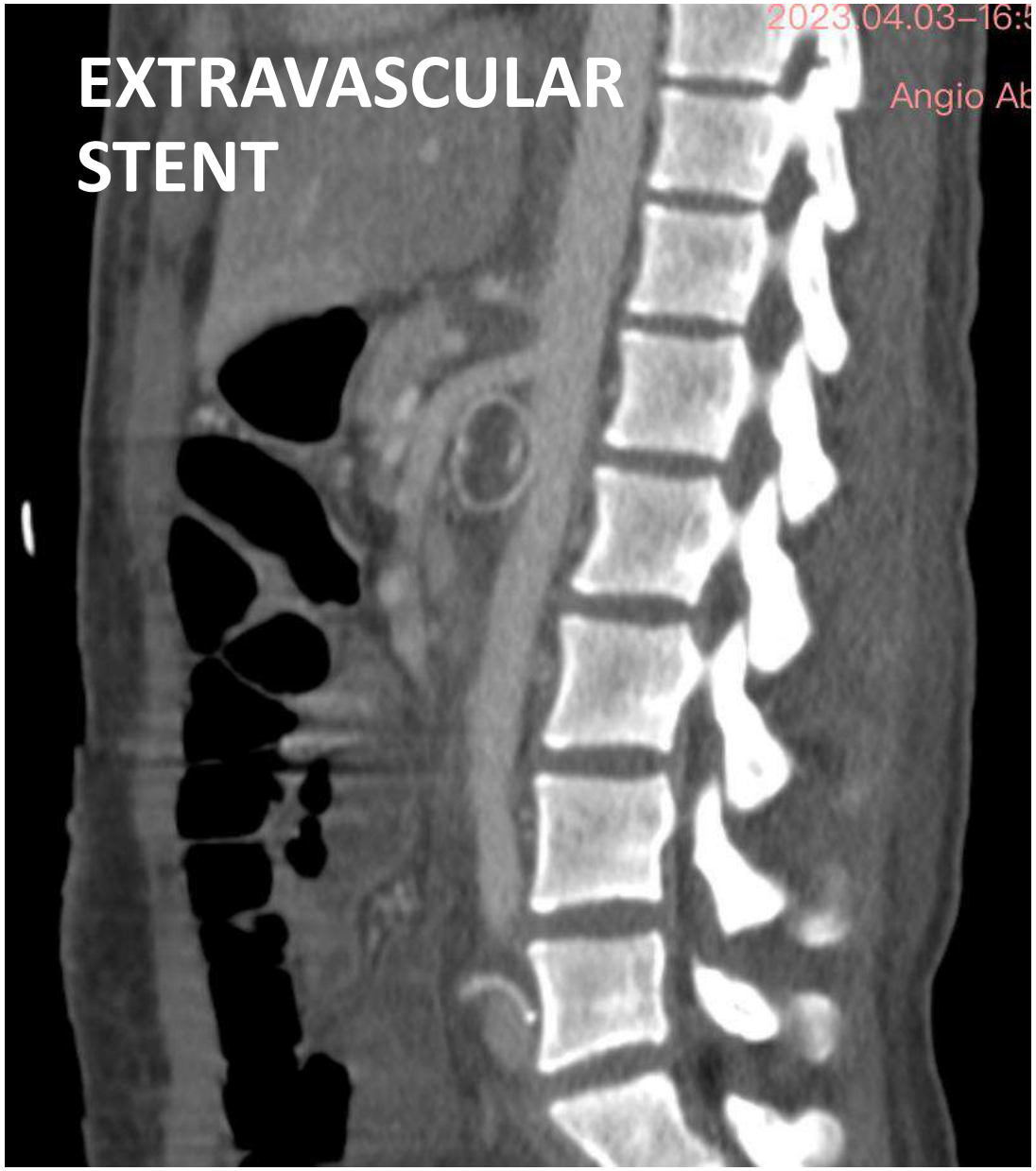

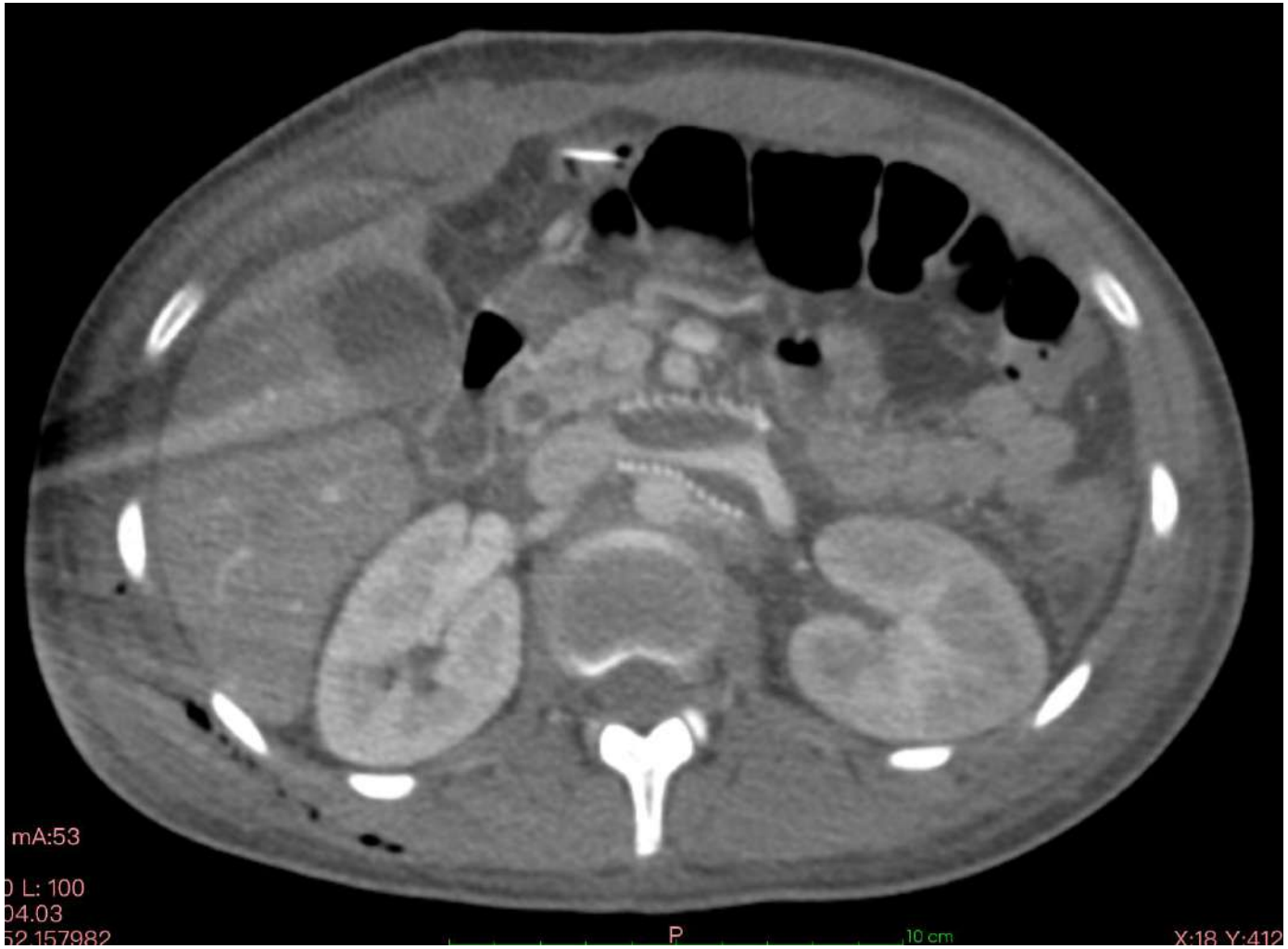

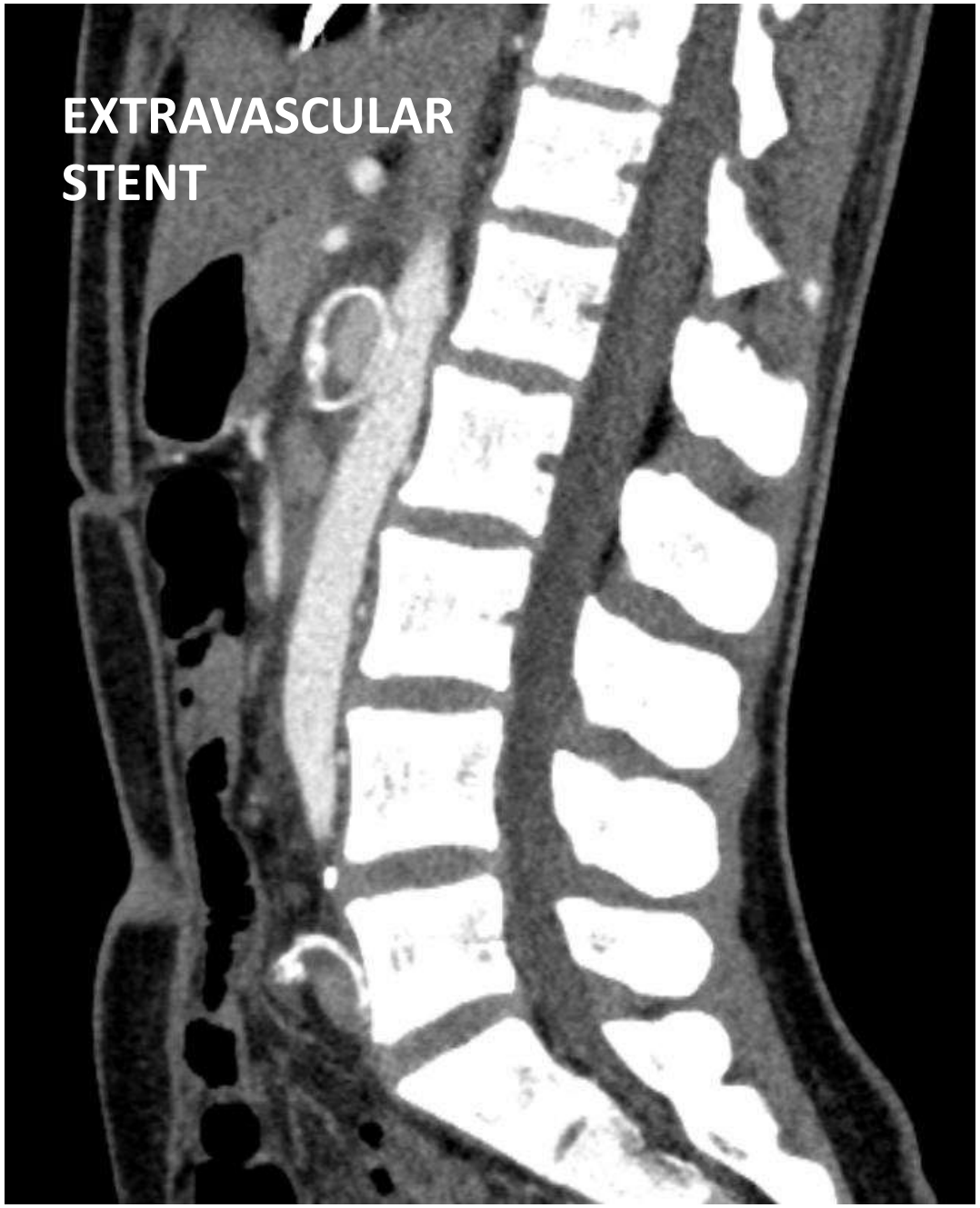
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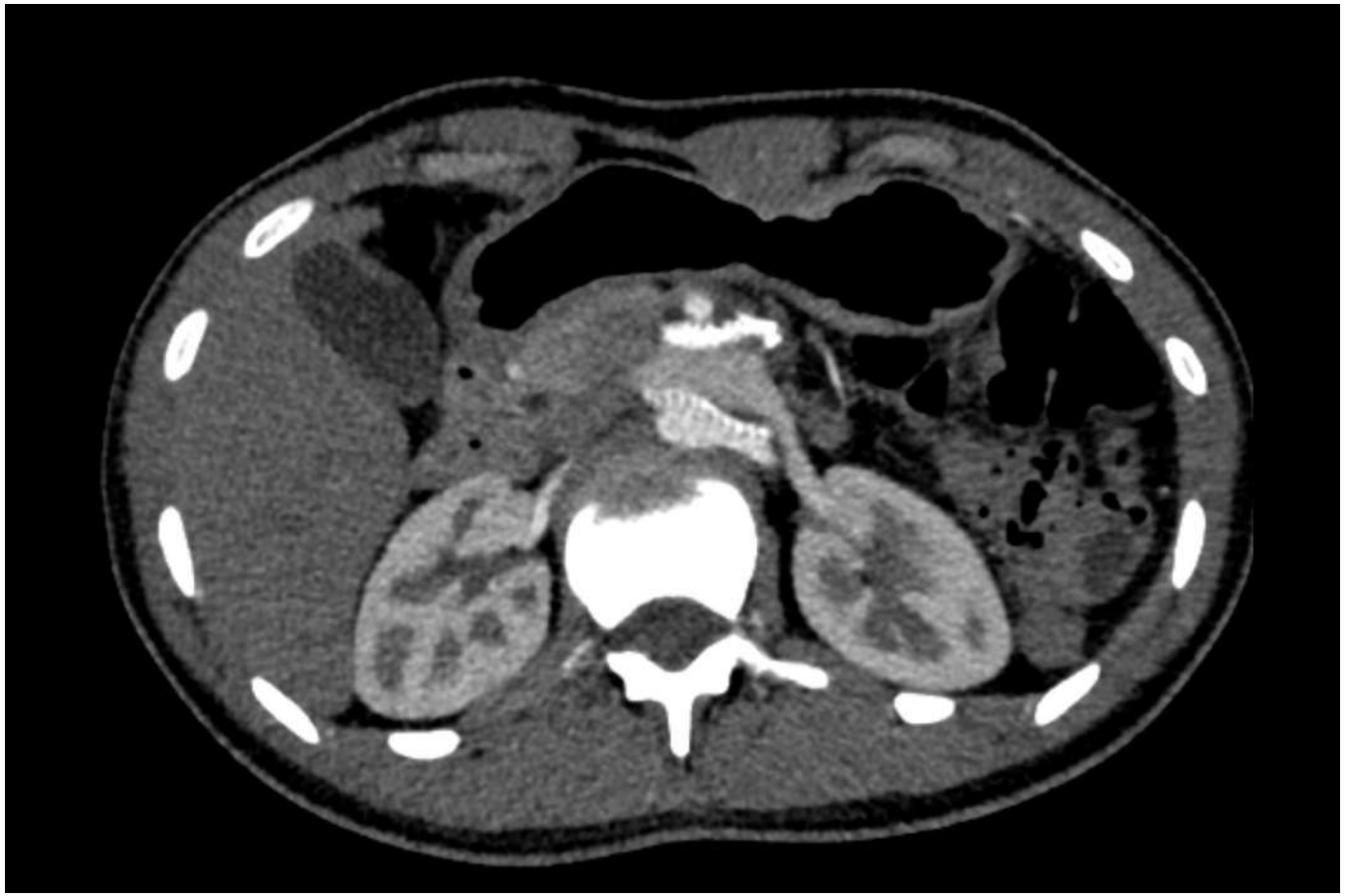

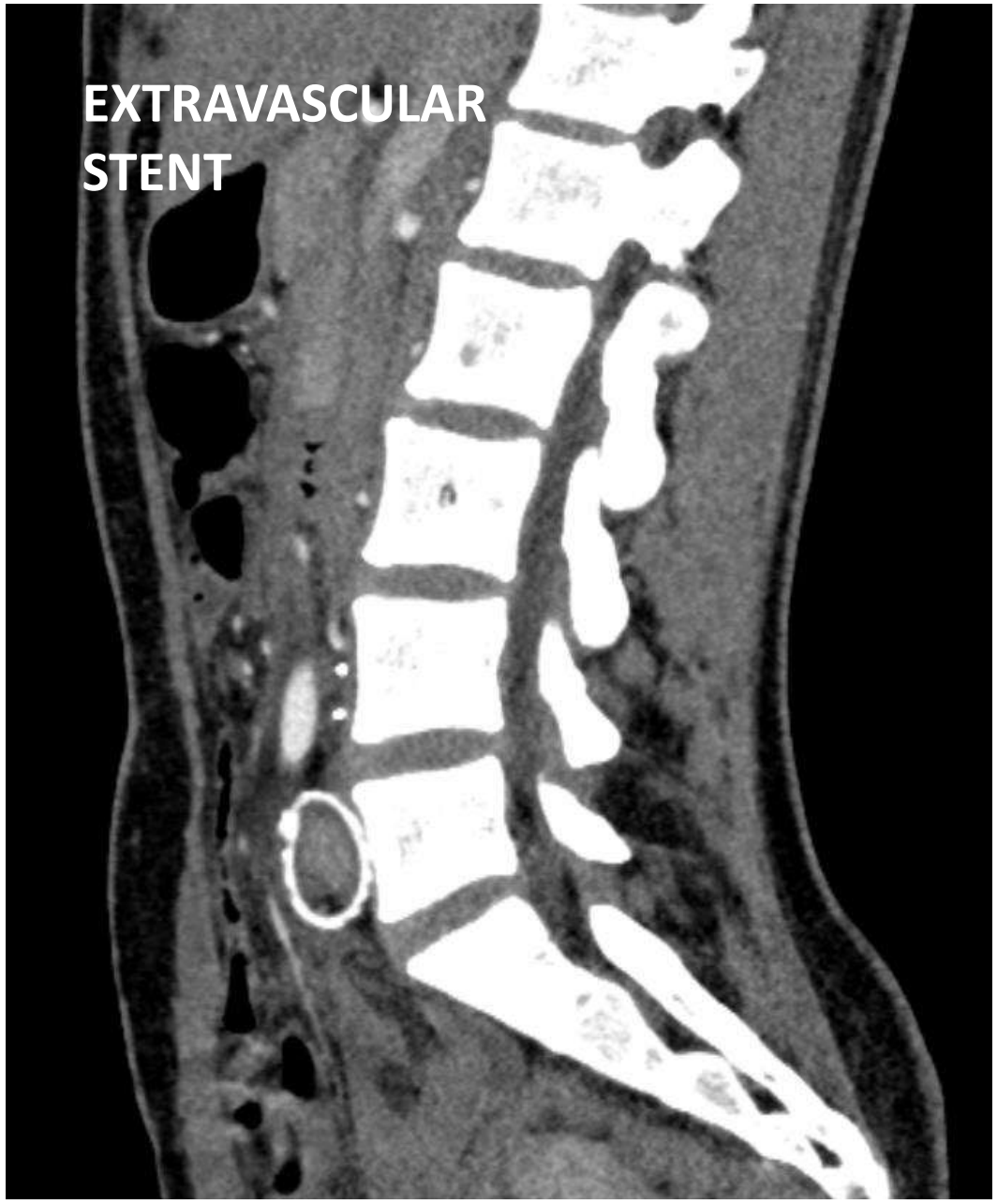
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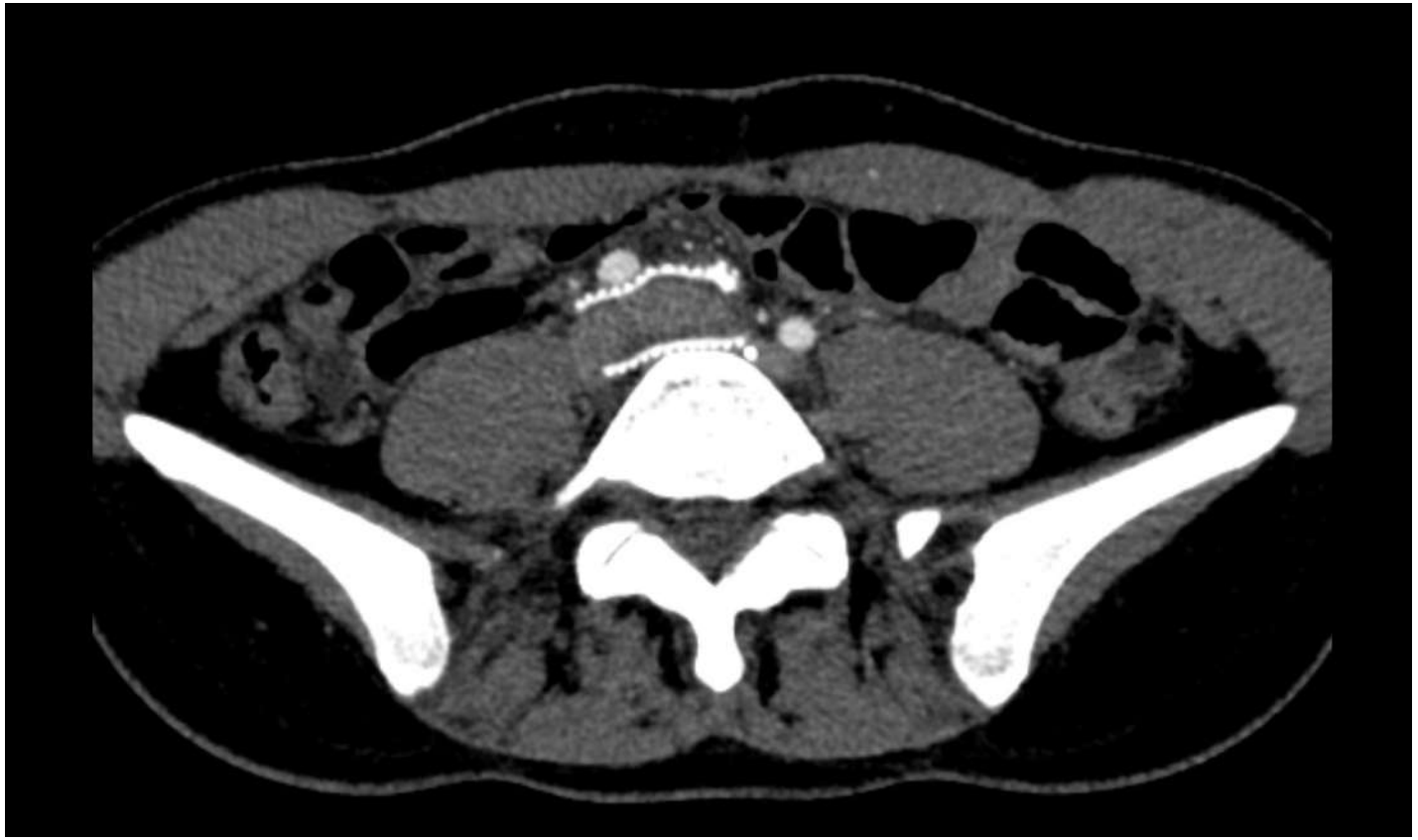
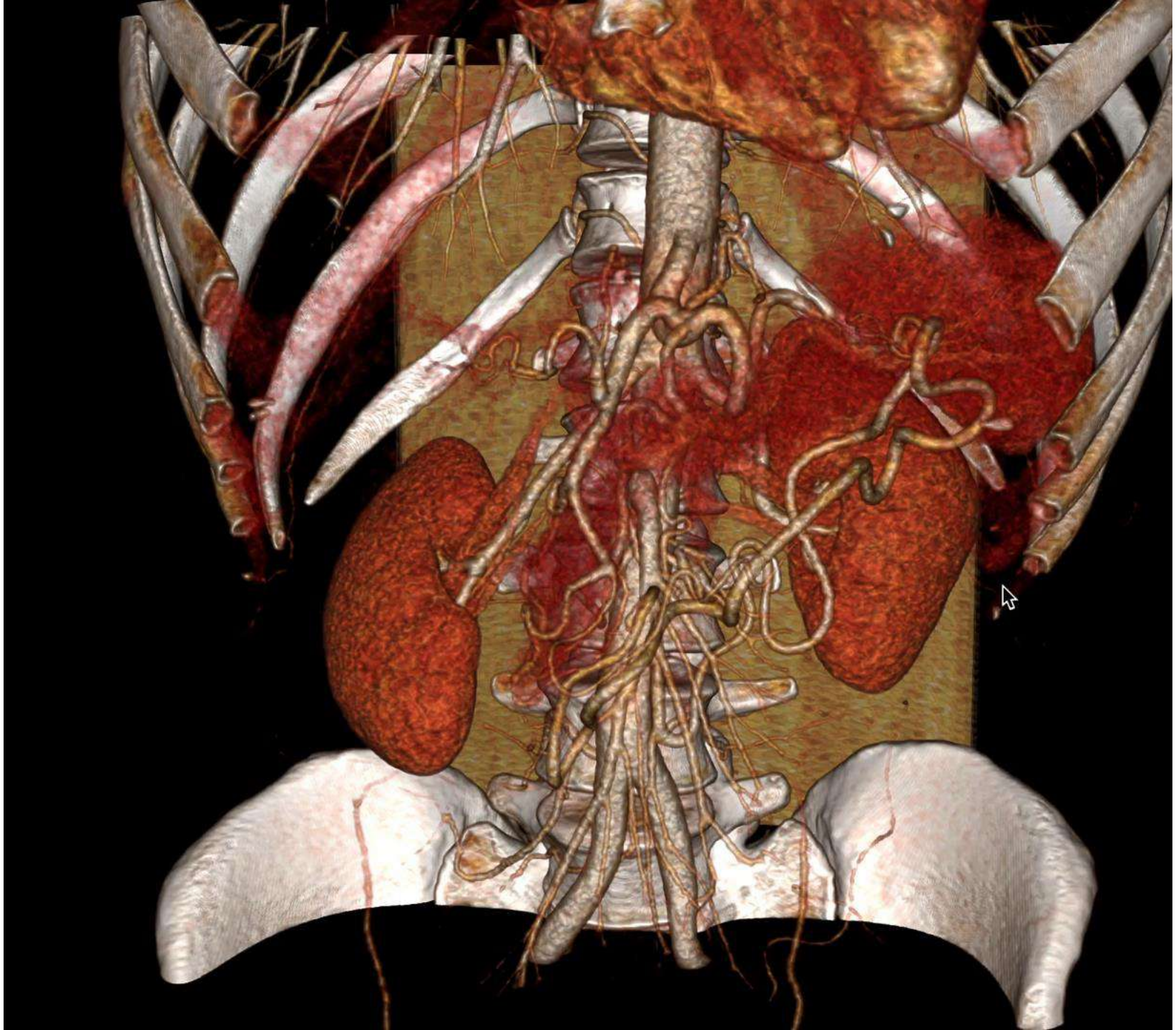
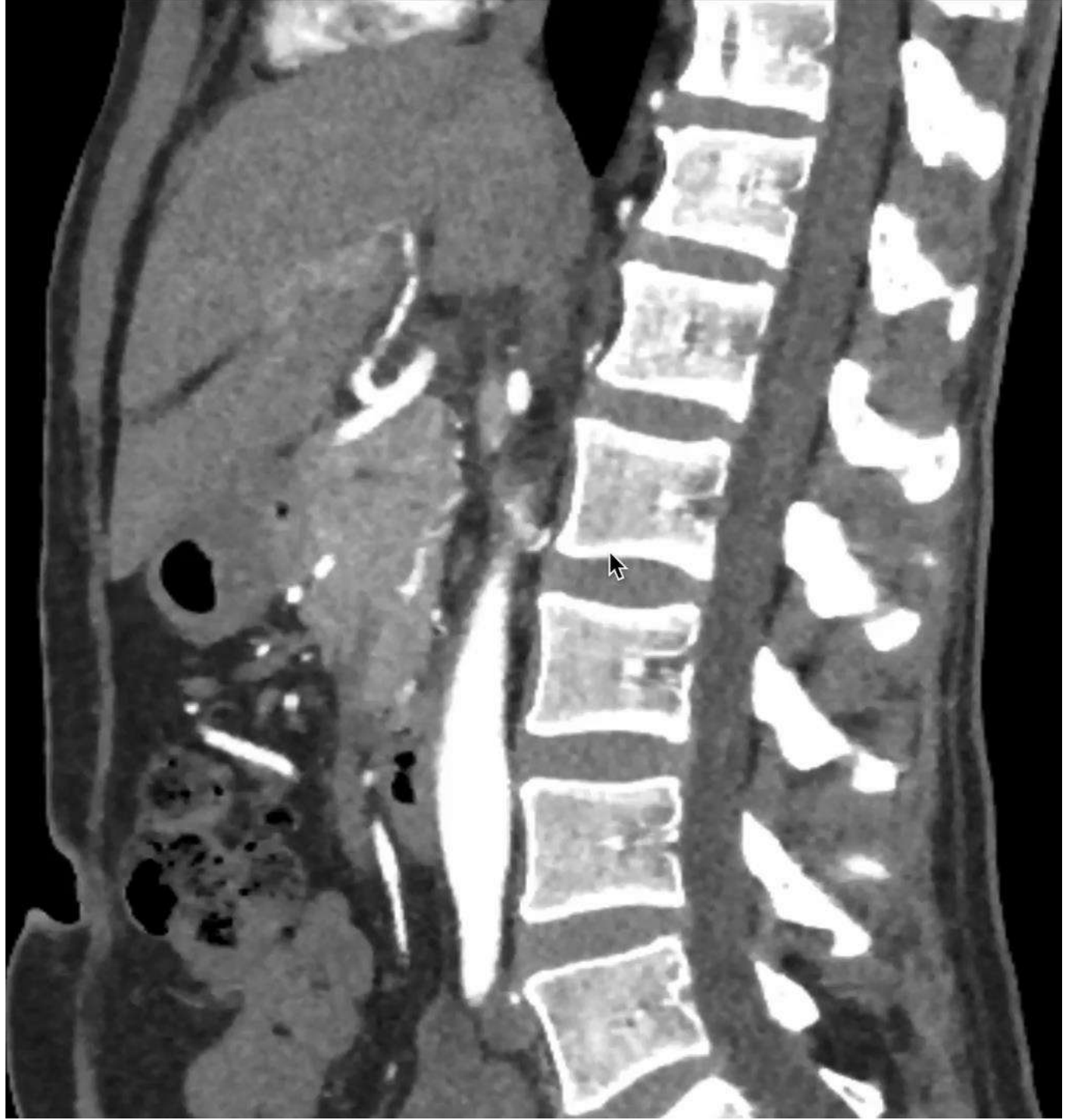


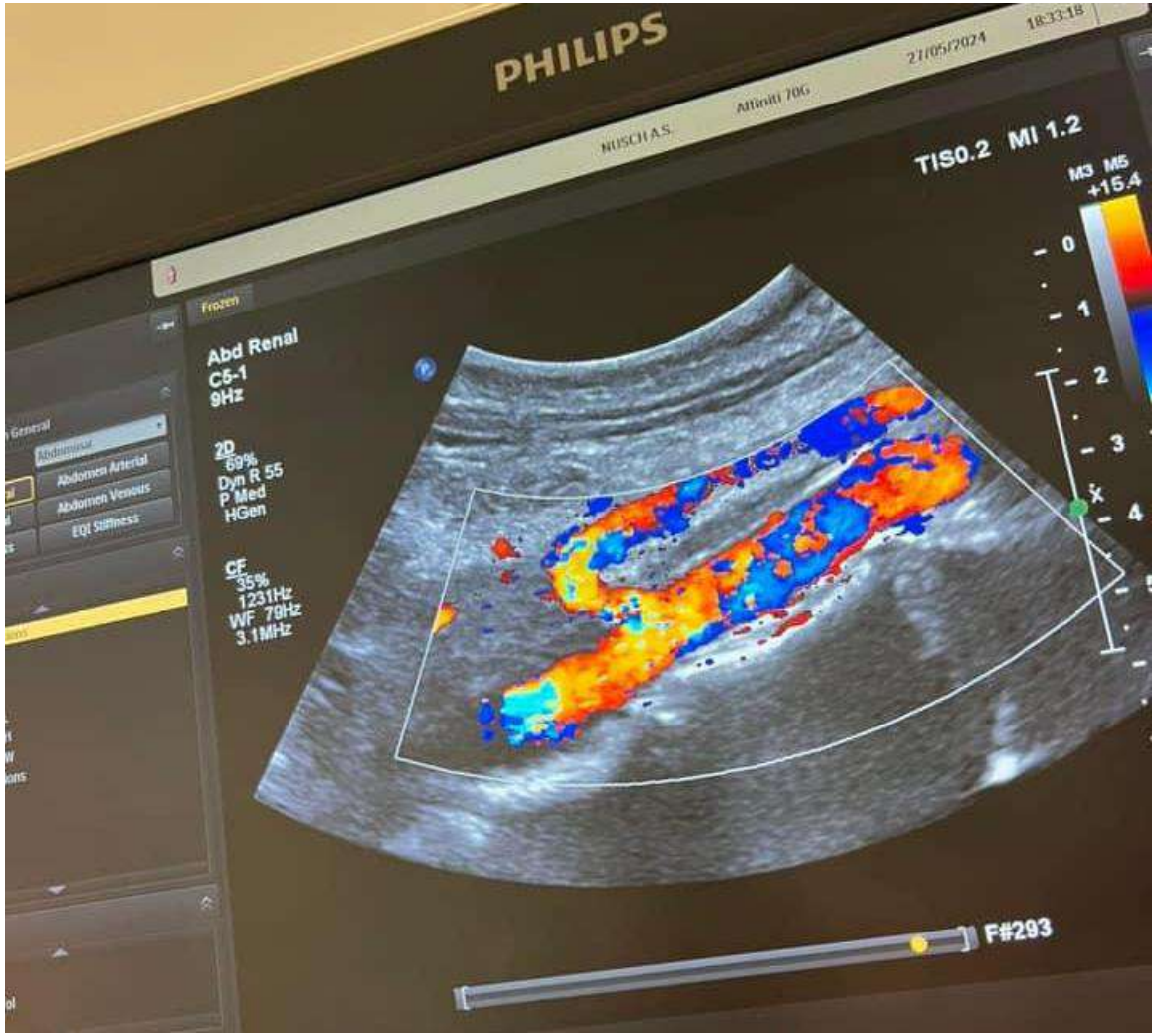
SMA TRANSPOSITION

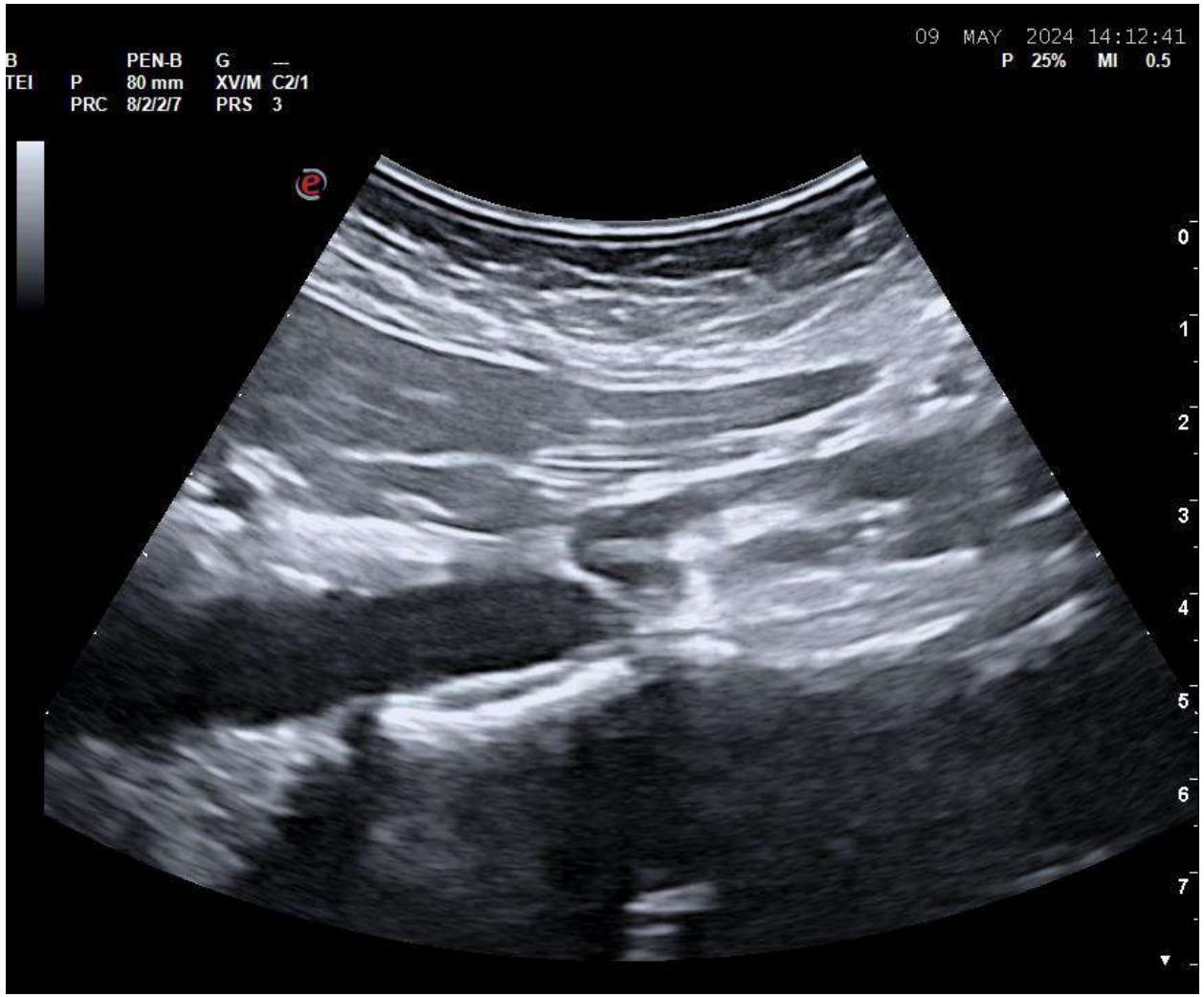


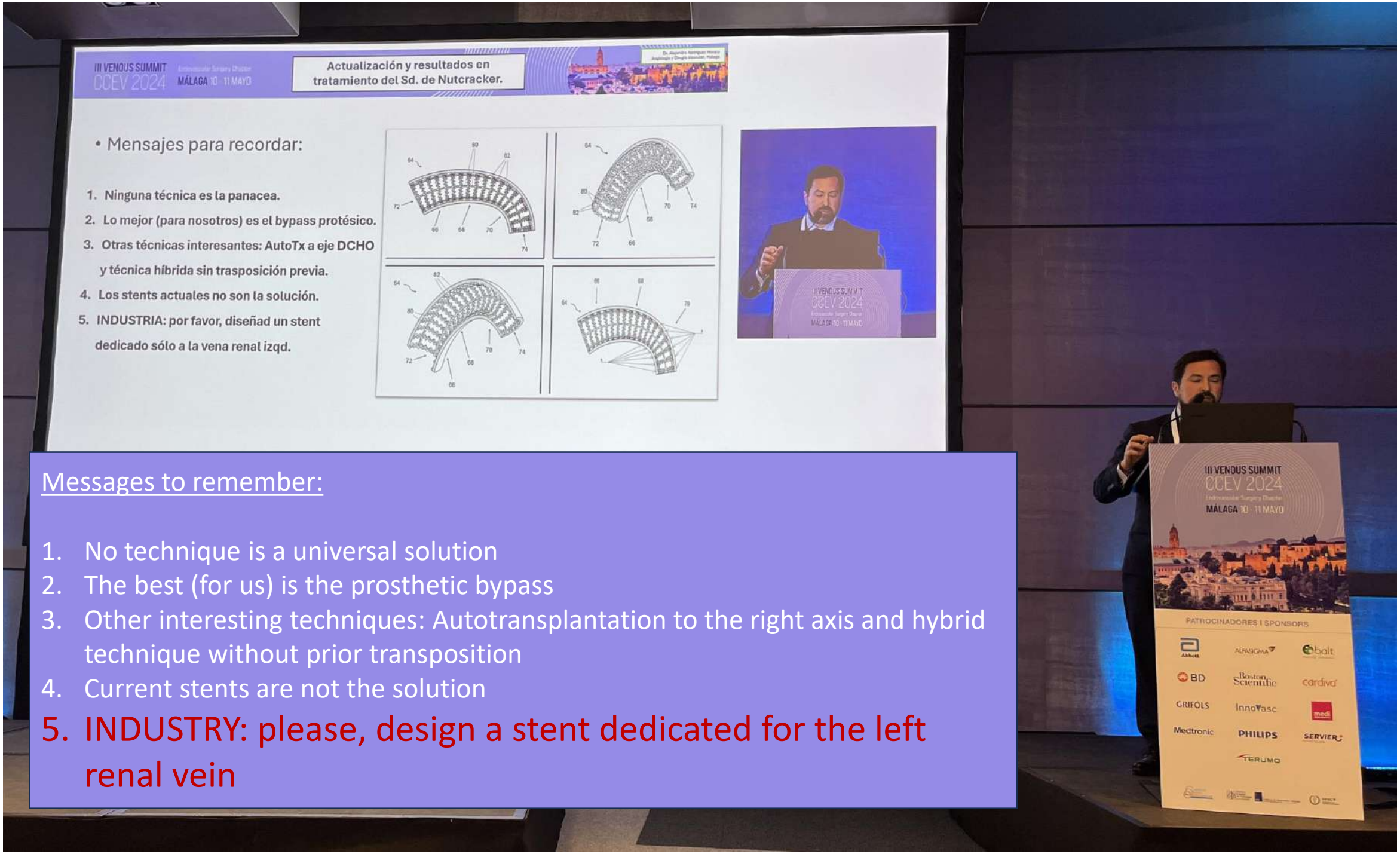
SMA TRANSPOSITION



SMA TRANSPOSITION







Messages to remember:

1. No technique is a universal solution
2. The best (for us) is the prosthetic bypass
3. Other interesting techniques: Autotransplantation to the right axis and hybrid technique without prior transposition
4. Current stents are not the solution
5. INDUSTRY: please, design a stent dedicated for the left renal vein

capitulo_cirugia_endovascular

capitulo_cirugia_endovascular Closing out the first day of the III Venous Summit our president, Dr. @rodriguezmorata, with an extensive lecture on the current approach to Nutcracker Syndrome. It highlights prosthetic bypass as the current best option and calls for the industry to develop a specific left renal vein stent.

NEED FOR THE DEVELOPMENT OF A NEW STENT

CONCLUSION

APVCS - rare diseases???, young patients, significant symptoms, ↓ QoL

multiform clinical & anatomical presentation, absence of dedicated guidelines from scientific societies

↓

further knowledge required to investigate & treat

↓

modern imaging and surgical (open or endovascular) techniques

? rare ?

hEDS / HSD
1 in 500

? unknown & misdiagnosed ?

CURRENT ISSUES

rare disease

≈ 5 years

lack of understanding

↓

incorrect / no diagnosis

↓

long time to diagnosis / treatment

↓

no treatment

↓

deterioration of patient condition & early deaths

↓

diagnosis – no treatment / treatment unavailability locally

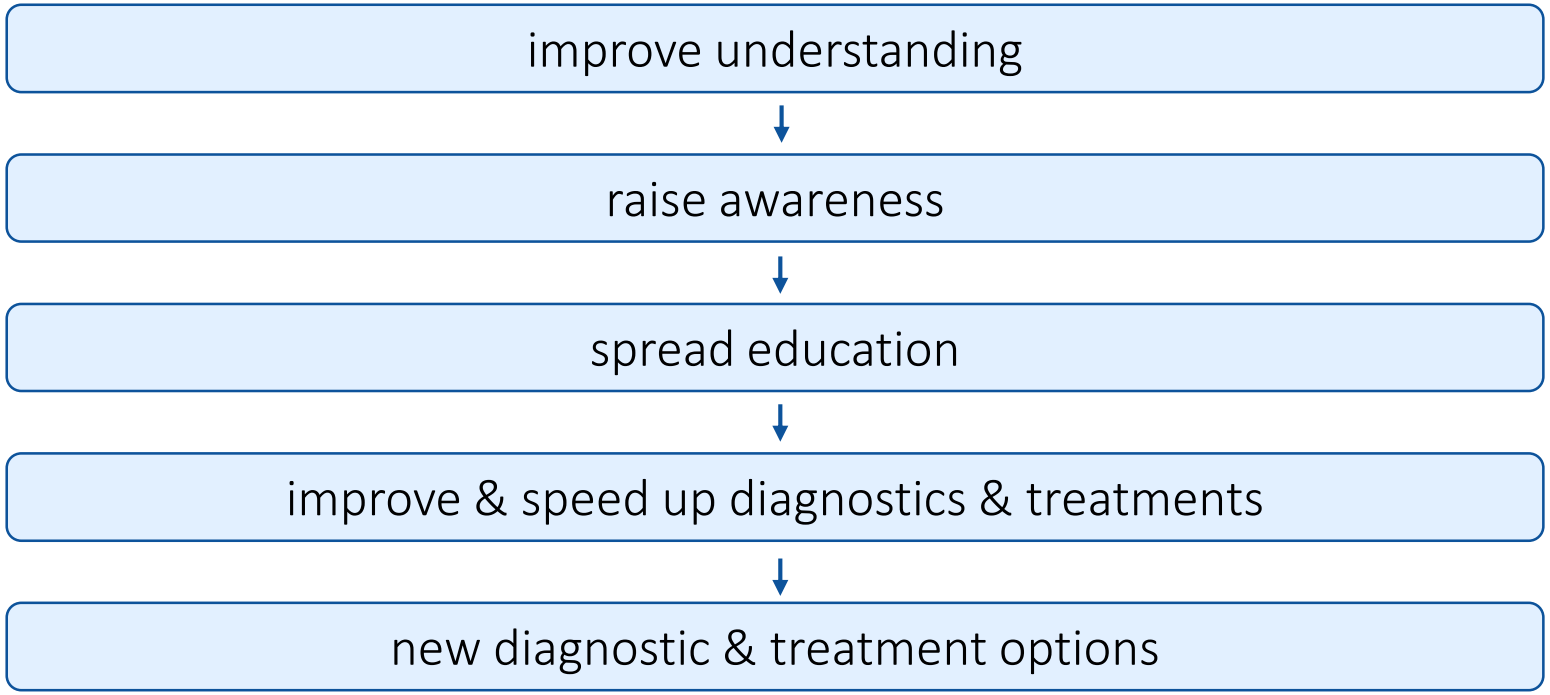
↓

burden to health care, economic and social systems

EDS

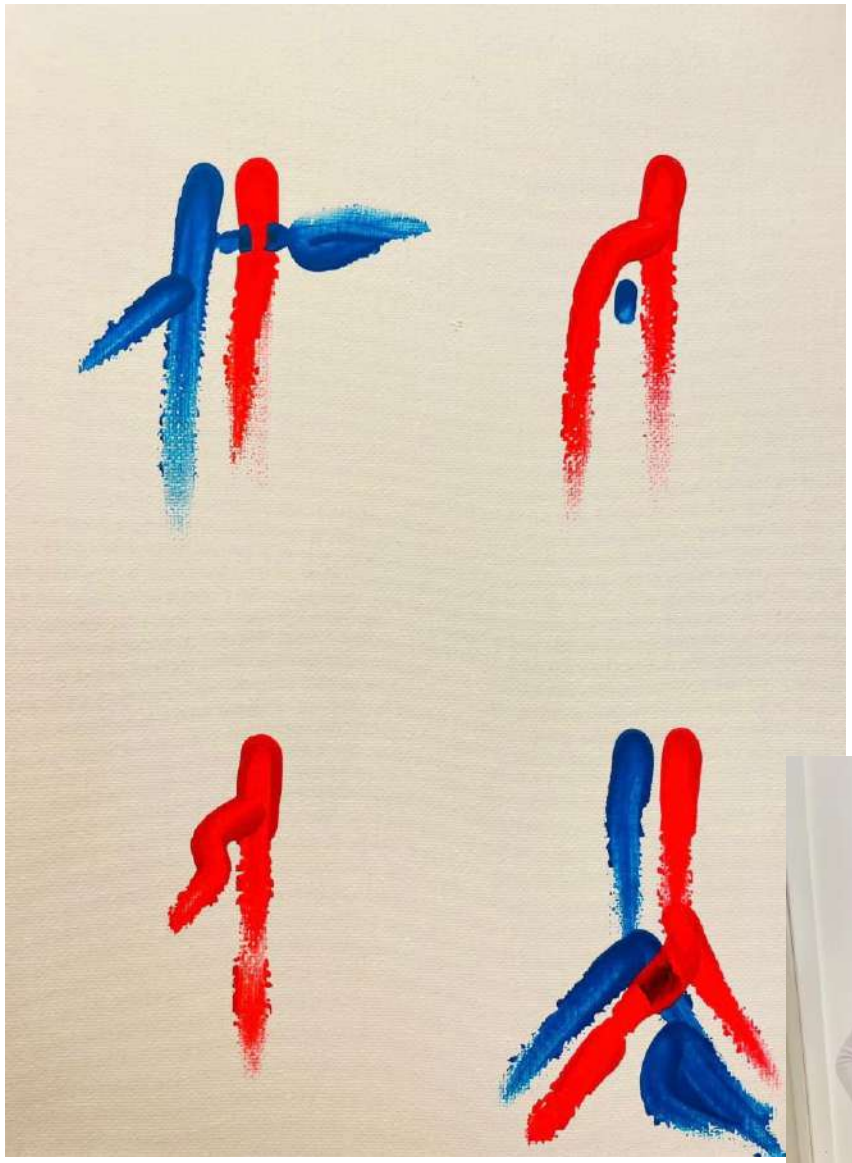
≈ 12 years

STUDY OBJECTIVE



“All things excellent are as difficult as they are rare.”

- Baruch Spinoza



Thank you very much for

Your attention!

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