

VISCERAL ARTERIAL COMPRESSION SYNDROMES and THEIR RELATIONSHIP to Genetic DISORDERS: CURRENT OPTIMAL DIAGNOSIS and TREATMENT
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Nothing to disclose



CLINIC Bel Etage
The upper level of medical care.

HEINRICH HEINE UNIVERSITÄT DÜSSELDORF

“You are operating something, which does not exist !”



Thomas SCHMITZ-RIXEN
President. GSVS 1967



Wilhelm Sandmann



We Organized a symposium in regard to compression syndromes.

What and WHICH ARE VISCERAL ARTERIAL COMPRESSIONS

MALS: Compression of the Celiac Artery
 (First Surgery by P.T. HARIOLA 1963,D. DUNBAR 1965)
Compressed structure: Celiac artery (trunc)
Compressing structure: Tendon of the Diaphragm: MAL

SMAS: Compression of the Duodenum III (WILKIE 1921, 1927)
Compressing structure: Superior mesenteric artery
Compressed structure: duodenum III part

ETIOLOGY:
 HYPERMOBILITY SYNDROME DISORDER (HSD); HYPERMOBILE EHLERS-DANLOS SYNDROME (hEDS)

Compression syndromes are very often related to hypermobility disorder syndromes

???? UNDERLYING ETIOLOGY ????

Beighton score 1-9





CS Female 83%
 Affected by hEDS/ HSD 70%

EHLERS-DANLOS-Syndrome

The Challenge for Diagnostik Work Up

- MALS and SMAS in HDS/ hEDS Patients present rarely as a Monocompression
- If abdominal Compression Syndrome(s) is/are also present, the patient is categorized psychotic (80% female)

„You don't have pain in Your abdomen – you have problems in Your head!“

Hypothesis
„Hypermobility Disorders represent „Instability“ due to Hyperelasticity of the fibrous tissue and force the Body to develop „Stability“, which results in „Compressions“

Surgical treatment of abdominal compression syndromes: The significance of hypermobility-related disorders¹
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American Journal of Medical Genetics
Visceral compressions present rarely as a monocompression syndrome

Abstract
 Case reports and systematic studies of the most common hypermobility-related disorders, hypermobile Ehlers-Danlos syndrome (hEDS), and hypermobility spectrum disorder (HSD) typically describe gastrointestinal symptoms and complaints attributed to structural malfunctions, 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 pathologic genes symptoms significantly reduce or cease after decompressive surgery. Aiming not only at 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 (inferior vena cava and Neri-Thurner syndromes), these latter conditions causing chronic pelvic congestion syndrome (PCS). Here, we report primarily on our experience of the assessment and management of MALS and SMAS in a cohort of cases with a surprising prevalence of hEDS and HSD. To our knowledge, this is the first cohort report of its kind in hEDS, HSD, and CS. We recommend that CS are considered in hEDS and HSD individuals with gastrointestinal and other organ complaints within the ‘‘belt’’ area. These CS can be identified using functional ultrasound duplex examination in experienced hands, and in appropriate cases stabilizing surgery can substantially improve quality of life.

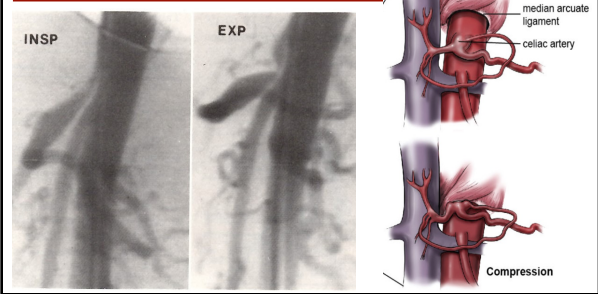
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Routine Diagnostic Algorithm for Patients with abdominal Pains

- PALPATION , AUSCULTATION
- Ultrasound of the Abdomen-Peristalsis
- Gastroscopy
- **Gastroduodenoscopy : air insufflation with pressure !!!**
- Barium contrast passage
- Scintigraphic documentation of remaining food in the stomach
CTA / MRA

Dunbar-Syndrome (MALS)



Treatment Modalities for visceral arterial compression syndromes

MALS

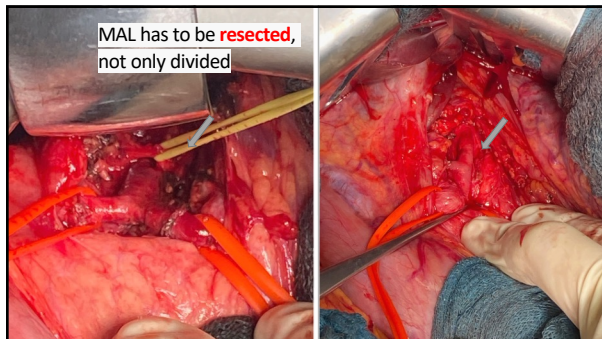
- Reimplantation of splenic artery
- Stenting celiac artery
- Resection of MAL: open, laparoscopic, Da Vinci

SMAS

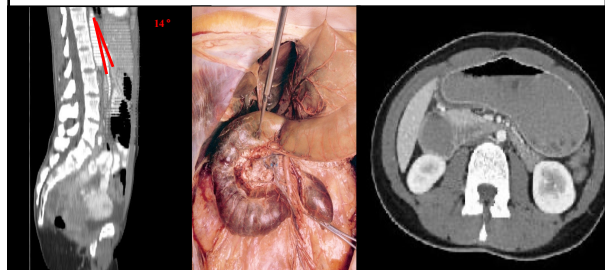
- Duodeno-jejunostomy
- Percutaneous entero- jejunostomy (PEJ)
- ROUX-Y -Anastomosis
- A.mesenterica superior transposition
- Decompression of left renal vein by ringenforced PTFE tube graft

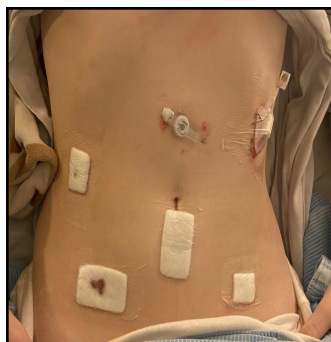
Methods to treat the DUNBAR Syndrome (MALS)

- 1) Pokrovsky, A.V. 1962, Decompression via thoracoabdominal approach
- 2) Transection of MAL (HARJOLA 1963 , fibrous-altered ganglionic plexus
- 3) Resection of MAL (DUNBAR 1965, 27 cases , ligamentum arcuatum syndrome
- 4) Marable et al. 1966 „combined compression „MAL and celiac plexus“
- 5) Sandmann et al , 1977,Transection of the splenic artery at the spleen and implantation into the aorta
- 6) Grotemeyer et. al ,2009,
- 7) Multicenter Study: Transection of MAL
a) open b) laparoscopic c) Da Vinci JVS 2021 Best results : Open surgery
- 8) Predominant etiology: hEDS, HSD: Sandmann et al: 2021 (Am.J.Med.Gen.)



(SMAS) - WILKIE-Syndrome PATHOLOGY

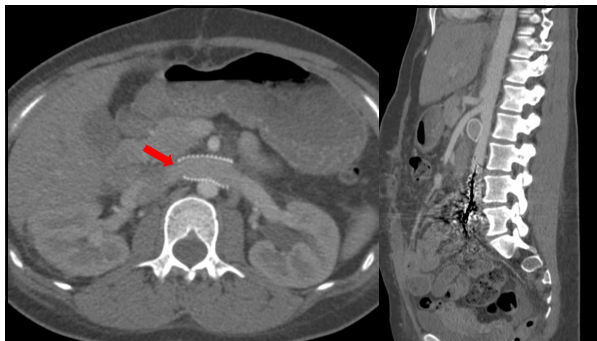
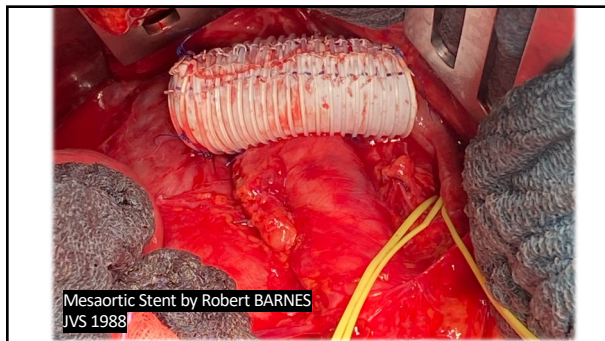
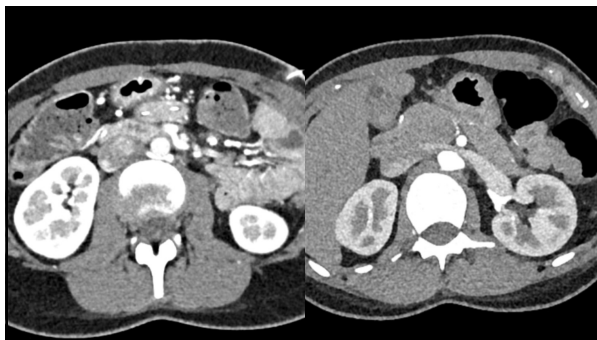
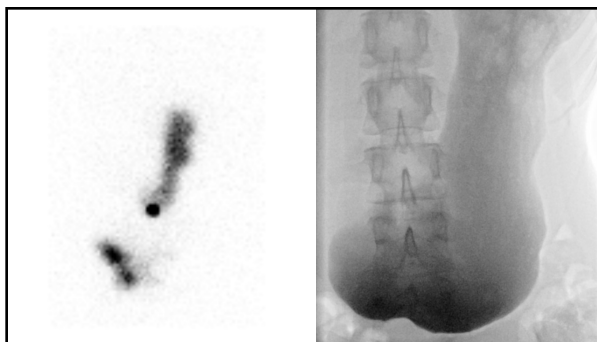




„You don't have pain in Your abdomen,
You have problems in Your head! “

„YOU can eat!!!! “
Dr. H.R. pediatrician,confronted
and offended The Patient
Miss Lo..... Th..... 14y

16 years old girl with NCS and SMAS
(WILKIE) just after implantation of PEJ
to prevent further loss of weight
(Negative Intelligence)



Patients	n =	%	EDS n =	%
female	228	84%	164	72%
male	44	16%	27	61%
Patients total	272	100%	191	100%
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No. compression syndromes	n =	%	EDS n =	%
One	43	16%	17	40%
Two	98	36%	68	69%
Three	80	29%	64	80%
Four	51	19%	42	82%
	272	100%	191	70%

Compression syndromes	n =	% (based on 272 patients)	EDS n =	EDS %
MALS alone	27	10%	6	22%
SMAS alone	2	1%	0	0%
NCS alone	10	4%	9	90%
MTS alone	4	1%	2	50%
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Leading syndrome				
MALS multi	159	58%	117	74%
SMAS multi	86	32%	68	79%
NCS multi	216	79%	174	81%
MTS multi	179	66%	148	83%
MALS, SMAS	3	1%	1	(33%)
NCS, MTS	47	17%	39	83%
NCS, SMAS, MTS, MALS	51	19%	42	82%

