Poster Number 014. Scientific Abstract.

THE SURPRISING FREQUENCY OF HYPERMOBILE EHLERS-DANLOS (hEDS) AMONG PATIENTS WITH ABDOMINAL AND PELVIC COMPRESSION SYNDROMES (APCS)

W. Sandmann[1], K. Verginis[2], S Jacobi[1], T. Scholbach[3]

[1] Section of Vascular Surgery, Clinic Bel Etage Pradus Medical Center, Düsseldorf, Germany, [2] Department of Vascular Surgery, EVK Mettmann, Mettmann, Germany, [1] Section of Vascular Surgery, Clinic Bel Etage Pradus Medical Center, [3] Private Practice for Special Ultrasound Duplex Diagnostic Studies and Pedriatics, 04229 Leipzig, Germany.

INTRODUCTION

EDS patients complain often about GI symptoms, which remain mostly unexplained. Identical symptoms and complaints are typical for abdominal and pelvic compression syndromes (APCS).

OBJECTIVES

During follow-up of our patients operated for APCS (MALS= median arcuatum ligament syndrome, SMAS= superior mesenteric artery syndrome, NCS= nutcracker syndrome, MTS= MAY-THURNER syndrome) we realised, that many patients showed signs and behaviour of hyper-elasticity. These are also typical for hEDS. In contrary the prevalence of EDS in the normal population is reported to be 1:5000. So, the question arose as to whether EDS may be an etiological factor for the development of APCS.

METHODS

Over a period of 18 years 142 primary operations were performed for APCS by the senior author (W. S.) by open median laparotomy (female 80%, mean age 29,9 Y). One hundred and six patients (Feb 2017 to Dec 2019) were treated at one single institution: 79 cases primary with a first procedure, and 27 secondary MALS (n=12), SMAS (n=5), and NCS/ MTS n=10) after unsuccessful treatment elsewhere.
All patients had undergone duplex ultrasound studies besides previous gastroenterological workup with numerous and repetitive gastro-duodeno- and colonoscopies and imaging methods (CTA 90%).
AT least 1/3 presented with typical signs of hEDS, and two presented with the vascular type after genetic testing.
We could not differentiate at the time whether one type of APCS was more prone to EDS, but the prevalence of hEDS among those undergoing recurrent surgery was highly significant. Before realizing that the coincidence of APCS and hEDS was very high surgery for decompression consisted of:
1) Resection of MAL (not only dividing) for MALS,
2) Transposing the SMA into the infrarenal aorta for SMAS and sometimes for NCS, and also
3) vein patchplasty to construct a delta mouth at the left renal vein (LRV),
4) Resection of the venous spur (if present) and scar at the iliac cava junction, vein patchplasty, and
5) Elongation of the right iliac artery with a short segment of autologous vein or PTFE graft.

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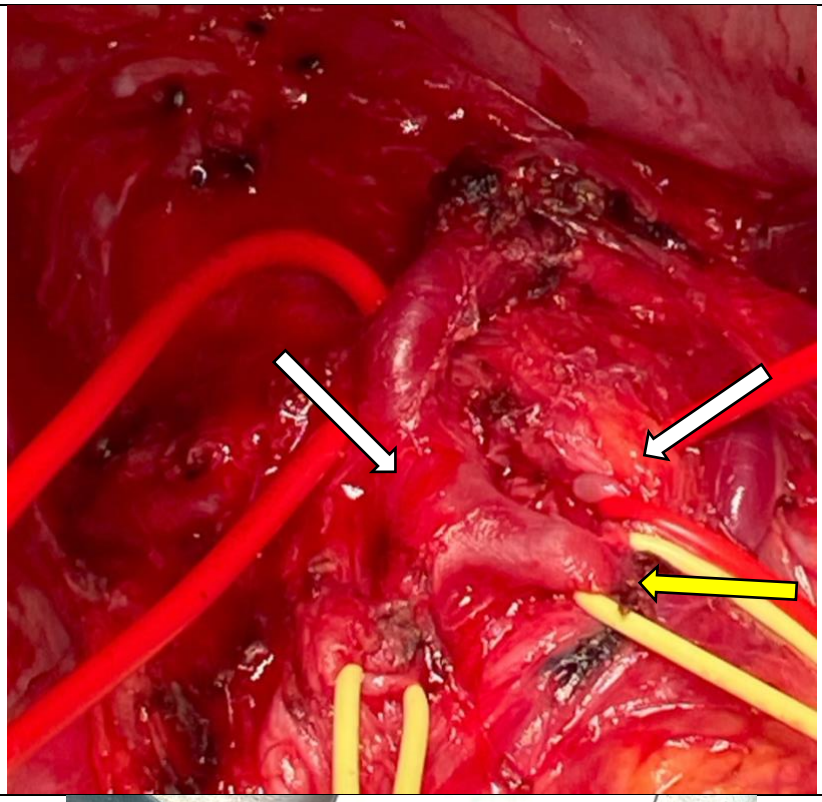
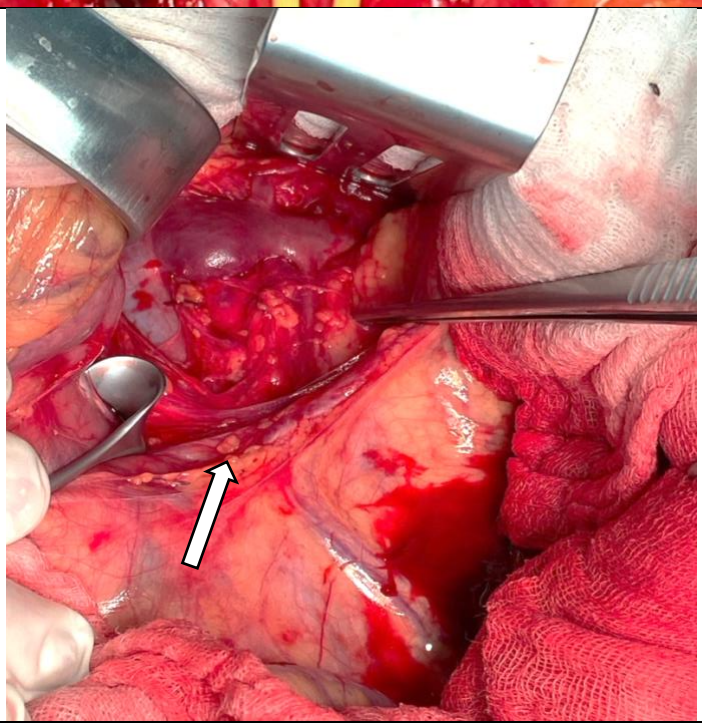
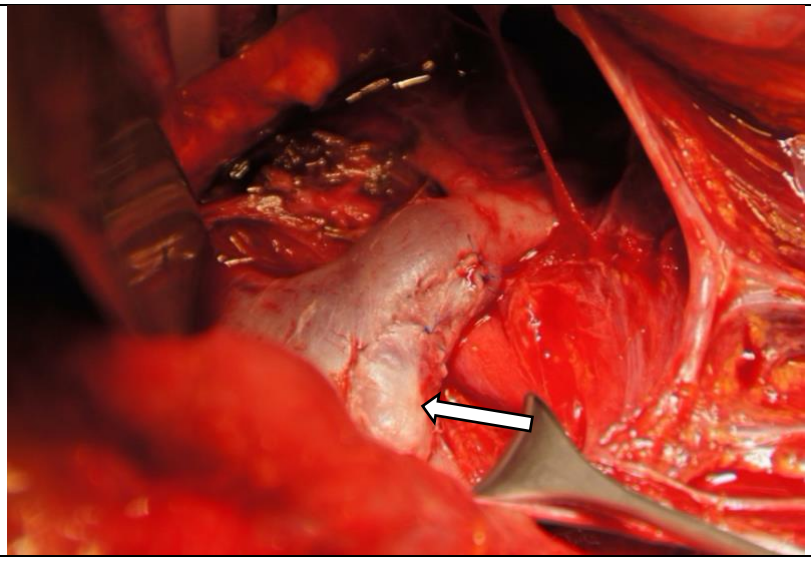
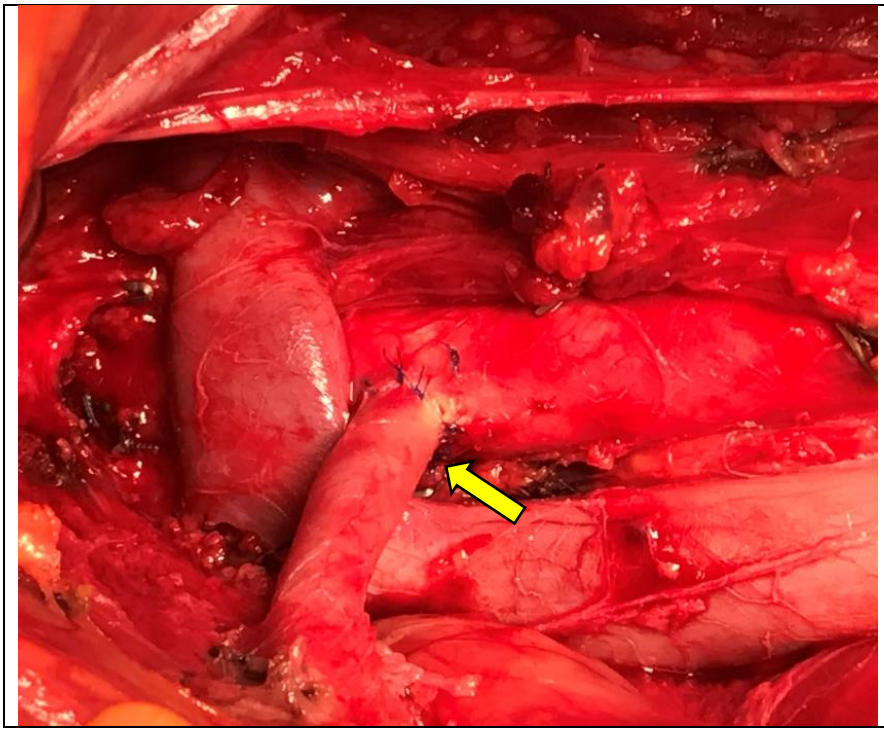
	Median arcuatum ligament (white arrows) compressing the celiac trunk (yellow arrow) after previous laparoscopic surgery for MALS
	Left renal vein, decompressed. The previously compressed proximal segment became dilated, its venous wall is thinner than the distal part. (white arrow suprarenal vein)



Enlargement of the mouth of the left renal vein with a venous patch (white arrow) harvested from the left groin. This method avoids complete transposition of the left renal vein and is very useful in patients without hEDS, because the method compensates later shrinking of the venous suture.



Transposed superior mesenteric artery (yellow arrow) in a case with WILKIE's (SMAS) syndrome. The left renal vein gets also space.

RESULTS

Decompression for MALS was successful in all but two cases in which the celiac trunk had to be replaced.

SMA transposition was successful in all but two cases in which an aneurysm at the SMA – aortic anastomosis developed. However, all patients had gained substantial weight and could eat again.

After we learned that the fibrous tissue problem (hEDS) was involved, we changed the operative technique at LRV and left iliac vein (LIV) and stabilised the veins in the last 25 cases using the "external stent" technique published by R. Barnes, which was highly effective and successful except in two NCS-cases where the tube possibly was not sufficiently fixated.

CONCLUSION

Although we have cared for patients with APCS for almost 20 years, it is only lately that we have understood that, unexpectantly, hEDS may be an underlying weakness, which reduces the stability of the body constitution and allows the development of compression syndromes.

In regard to this new insight decompression techniques should include stabilisation of compressible structures.

REFERNECES

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DECLARATIONS OF INTEREST

None

NOTE TO READER

Also see associated abstract numbers 004 and 005