

## **EDS COMPLICATED BY SEVERE SCOLIOSIS AND THREE RETROPERITONEAL AND PELVIC COMPRESSION SYNDROMES (RPCS)**

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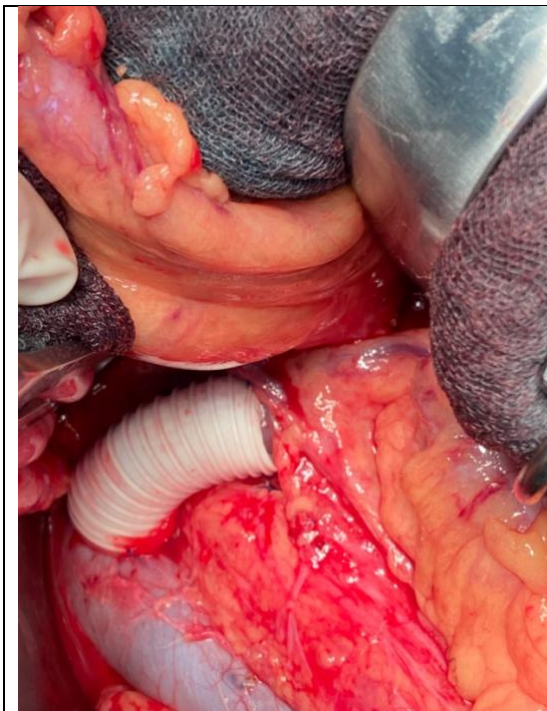
### **CASE PRESENTATION AND DIAGNOSES**

A 29-year old, slim line female (weight 62Kg / height 177cm) had lost 7kg in weight due to heavy pain from eating. She suffered from Postural Orthostatic Tachycardia Syndrome (POTS), nausea, heavy flank and pelvic pain, numbness and weakness of both legs, and required the use of a wheelchair.

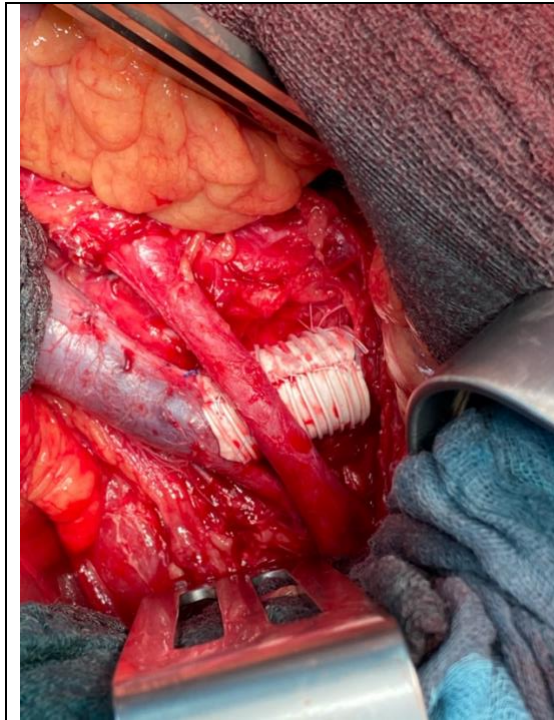
She was diagnosed in 2016 to have three RCPS, namely, Median Arcuatum Lig. syndrome (MALS), Nutcracker syndrome (NCS), and MAY-THURNER syndrome. After six months of conservative treatment repeated duplex ultrasound did not show any improvement nor did the symptoms and complaints improve.

Decompressive surgery was performed in April 2017. The procedure entailed resection of MAL, minimal resection of the solar plexus, enlarging the mouth of the left renal vein (LRV) and the left common iliac vein (LCIV), elongation of the right common iliac artery by a PTFE graft and temporary adjunctive arteriovenous fistula in the groin which was closed three months later by suture strangulation.

A second surgery was undertaken in May 2019. At this time the patient was still wheelchair dependent. The surgery involved resection of all scar tissue around the LRV and the LCIV; and, placing "External stents" made from 20 mm diameter ring enforced PTFE tube grafts around the LRV and LCIV with fixation accordingly.



Example of external stent (modification of the original external stents technique for treatment of nutcracker syndrome by R.Barnes) The tube is drained by a vacuum drainage, which cannot be seen in this photo, and is fixed with stay sutures to the inferior vena cava. It is also hold in place by the suprarenal and the ovarian vein, for which separate wholes were cut into the tube.



External stent protecting the left common iliac vein against compression between the promontorium and the overriding right common iliac artery in a patient with primary MAY-THURNER syndrome. The adventitia of the artery is fixed by sutures to the stent and the stent is fixed to the paravertebral fascia.

## OUTCOMES

After the first surgery the patient was able to eat without pain, gain weight, and other symptomatology markedly improved for up to 6-12 months before recurrence of NCS and MTS with more severe symptomatology. After the second surgery the patient gained further weight, had no nausea, could work. Her POTS was less severe; she could walk without pain, and dance without any limits.

## CONCLUSIONS

In EDS patients, surgical reconstitution of compressed veins as in NCS and MTS is not durable. The “external stent” technique is effective and to our mind more efficient and durable than any catheter supported inner stent technique, because the veins are protected against further compression and any injury to the endothelium of the veins is avoided, which might be ideal in young individuals with otherwise disease-free veins in the future perspective.

## REFERENCES

Barnes, Robert et al. (1988): JVS, Volume 8, Number 4, 415-421; Mesoaortic compression of the left renal vein (the so-called nutcracker syndrome): Repair by a new stenting procedure.

## DECLARATIONS OF INTEREST:

None

**NOTE TO READER:** See also abstract numbers 005 and 014