Single-center initial experience with minimally invasive extravascular stent placement for nutcracker syndrome in adolescent patients

Salvatore Fabio Chiarenza,[1] Giulia Brooks,[1] Paolo Magagna,[2] Cosimo Bleve,[1] Jacopo Dall'Acqua,[3] Stefania Marconi,[4] Elisa Zolpi[1]

[1]Department of Pediatric Surgery, Pediatric Minimally Invasive Surgery and New Technologies, San Bortolo Hospital, Vicenza; [2]Department of Cardiac Surgery, San Bortolo Hospital, Vicenza; [3]Diagnostic and Interventional Angiography, Department of Radiology, San Bortolo Hospital, Vicenza; [4]Department of Civil Engineering and Architecture, University of Pavia, Pavia, Italy

Abstract

Nutcracker Syndrome (NCS) is characterized by impeded blood flow from the Left Renal Vein (LRV) into the inferior vena cava due to an abnormally narrow angle between the abdominal aorta and superior mesenteric artery. This syndrome is usually associated with hematuria, abdominal pain and orthostatic proteinuria. We described three patients diagnosed with NCS (mean age 16.3 years). The aortomesenteric angle, LRV diameter ratio and peak velocity ratio were assessed through doppler ultrasonography and CT angiography. A kidney model was printed out for surgical planning. An extravascular stent was designed based on the LRV's structure using computer software and printed in three dimensions with a precision setting of 20µm. Patients underwent laparoscopic placement of the extravascular stent. The mean duration of surgery was 180 minutes. There were no intraoperative complications. CT examinations revealed that pre- and postoperative AMA ranged from 18.7°±4.3° to 55.0°±4.4, respectively. No side effects were observed in the follow-up period (range 12-24 months). At present, treatment guidelines for NCS are unclear, and the different therapeutic principles need to be applied in a patient-specific manner. Our results confirm previous reports in literature concerning the efficacy and safety of ES in pediatric patients.

Correspondence: Giulia Brooks, Department of Pediatric Surgery, Pediatric Minimally Invasive Surgery and New Technologies, San Bortolo Hospital, via Ferdinando Rodolfi 37, 36100, Vicenza, Italy.
Tel.: +390444752875.
E-mail: giulia.brooks@aulss8.veneto.it

Key words: 3D-printing, extravascular stent, minimally invasive surgery, nutcracker syndrome.

Contributions: SFC, conceptualization (lead), writing - review and editing (lead); GB, writing - original draft preparation; PM, resources (equal), software (equal); CB, formal analysis, data curation; JDA, investigation, resources (equal); SM, resources (equal), software (equal); EZ, conceptualization, writing - review and editing, data curation (lead).

Conflict of interest: there are no competing interests to declare.

Ethics approval: the Ethics Committee of Ulss 8 Berica approved this study (08/21 from 25/06/2021). The study is conformed with the Helsinki Declaration of 1964, as revised in 2013, concerning human and animal rights.

Informed consent: all patients participating in this study signed a written informed consent form for participating in this study.

Patient consent for publication: written informed consent was obtained from a legally authorized representative(s) for anonymized patient information to be published in this article.

Availability of data and materials: all data generated or analyzed during this study are included in this published article.

Received: 7 October 2024.
Accepted: 3 October 2025.

Licensee PAGEPress, Italy
La Pediatria Medica e Chirurgica 2025; 47:353
doi:10.4081/pmc.2025.353

Introduction

Nutcracker phenomenon is an anatomic anomaly characterized by an extrinsic compression of the Left Renal Vein (LRV), resulting in alteration of the blood flow to the inferior vena cava. The LRV is most commonly entrapped between the Superior Mesenteric Artery (SMA) and the Abdominal Aorta (AA) (this is referred to as nutcracker phenomenon). Nutcracker Syndrome (NCS) is the association of this condition with signs and symptoms caused by impaired venous flow, the most common being hematuria, proteinuria and abdominal pain.[1] NCS is one of the abdominal vascular compression syndromes along with the SMA syndrome (in which there is a duodenal compression between the SMA and AA). Among treatment options, extravascular stenting is a mini-invasive, safe and efficacious technique.

We hereby report our experience with extravascular stenting for NCS in three pediatric patients.

Materials and Methods

Patients' characteristics are summarized in Table 1.

Patient 1

A 16-year-old boy presented a 2.5-year history of chronic abdominal pain, localized in the epi-mesogastrium with left flank irradiation, associated with weight loss. The patient had experienced a significant height growth (50 centimeters in one year). His Body

Mass Index (BMI) was 16 kg/m². Blood tests and renal scan were normal. Doppler ultrasound was used to calculate the LRV hilar-to-aortomesenteric Diameter Ratio (DR) and the LRV hilar-to-aortomesenteric Peak Velocity Ratio (PVR), which were 8 and 8.6 respectively (Figure 1). The CT angiography showed an aorto-mesenteric angle (AMA) of 23°, a DR of 4 and LRV flux acceleration at the compression site.

Patient 2

A 17-year-old boy was referred to our center for a 2-year history of chronic left flank pain, associated with weight loss (5 kilograms in 18 months) and macrohematuria. He also presented an important height growth (30 centimeters) in one year. His BMI was 19.3 kg/m². Blood tests and renal scan were normal. The sonographic parameters were DR=12.5 and PVR=9, while CT angiography demonstrated an AMA of 22° and mild duodenal compression by the SMA.

Patient 3

A 16-year-old girl presented a 1-year history of chronic left flank pain, associated with macrohematuria, proteinuria, a 6-kilogram weight loss in one year and a 2-year history of amenorrhea. Her BMI was 16.4 kg/m². Blood tests and renal scan showed no pathologic alterations. The ultrasound parameters were DR 11.3 and PVR 9, while CT-angiography showed an AMA of 14.4° with a width between the SMA and AA of 4 millimeters.

All patients were offered nutritional and psychological support for at least 1-2 years. Due to little response to conservative treatment, we decided to place an extravascular stent through a mini-invasive approach. We obtained the ethical committee's approval for all patients. Using a 64-detector CT scanner (GE LightSpeed VCT, Boston, MA, USA), we obtained images with a slice thickness of 1.0 mm for all patients. The CT image output in the DICOM format was exported to Mimics (Version 19.0, Materialise NV, Belgium). 3D-DOCTOR software (Version 4.0, Able Software Co., Lexington, MA, USA) was adopted for kidney modeling. These programs are used for image segmentation, converting CT 2D images into optimized 3D anatomic models. The data were transferred to a Fused Deposition Modeling (FDM) 3D printer (TD-III, 3D Printing Research Center of the Fourth Military Medical University, China) in the standard tessellation language format (Figure 2).

3D models were used to create customized titanium stents, tailored to each patient's vessel anatomy (Figure 3).

Results

We chose a transperitoneal laparoscopic approach to place the extravascular stents. The patient was positioned in right lateral decubitus with left flank elevation. A urinary catheter was placed preoperatively. A 12-millimeter balloon camera port was inserted through the umbilicus, along with four operative 5-millimeter bladeless trocars: two in the left flank (along the emiclavear and median axillary

Table 1. Patients' key characteristics.

Patient	Age (y)	Sex	BMI (kg/m²)	Symptoms/signs	Onset before surgery (y)	AMA (°)	LRV DR	LRV PVR before surgery	LRV PVR after surgery
1	16	M	16	Abdominal pain, weight loss	2.5 y	23	8	8.6	1.6
2	17	M	19.3	Abdominal pain, weight loss, macrohematuria	2 y	22	12.5	9	2.7
3	16	F	16.4	Abdominal pain, weight loss, macrohematuria, proteinuria	1 y	14.4	11.3	9	1.8

y, years; M, male; F, female; AMA, aorto-mesenteric angle; LRV DR, left renal vein diameter ratio (hilar-to-aortomesenteric); LRV PVR, left renal vein peak velocity ratio (hilar-to-aortomesenteric).

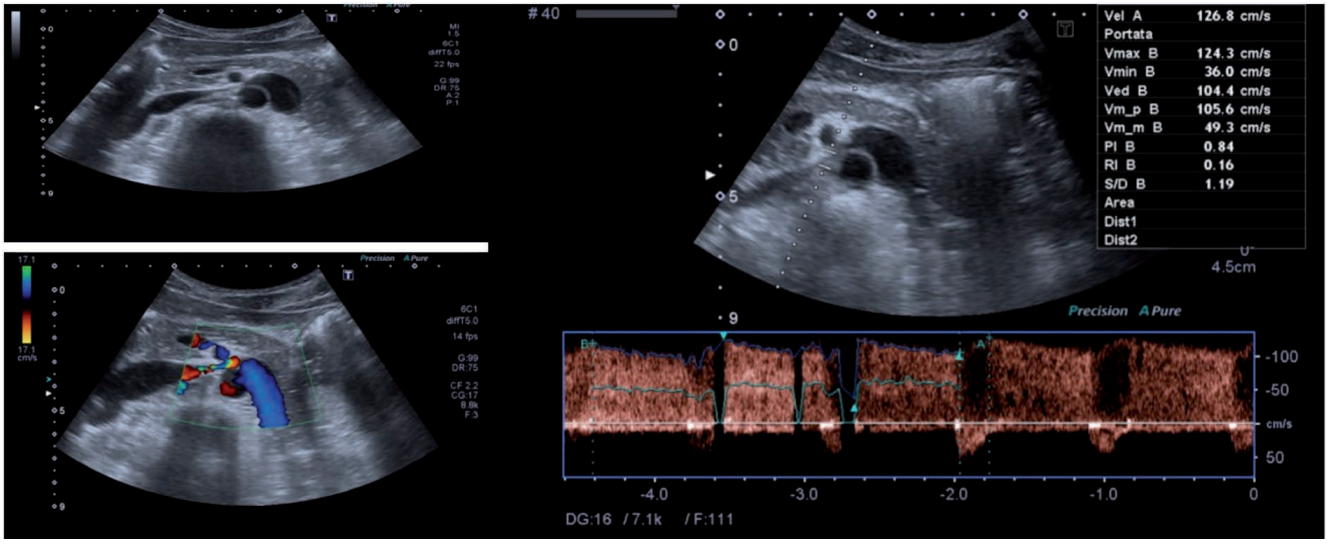

Figure 1. Doppler ultrasound images of patient 1 showing the LRV stenosis due to compression by the SMA and peak velocities.

lines) and two along the supra-umbilical linea alba (in the epigastrium and mesogastrium), as shown in Figure 4.

A 30° 10-millimeter camera was used. Access to the retroperitoneum was obtained through incision of the left paracolic gutter and medial mobilization of the transverse and left colon. To gain exposure of the aortomesenteric angle, we carried on an accurate dissection of the LRV from the hilar region to its caval insertion, ligating the periarterial lymphatic vessels. The titanium stent was inserted

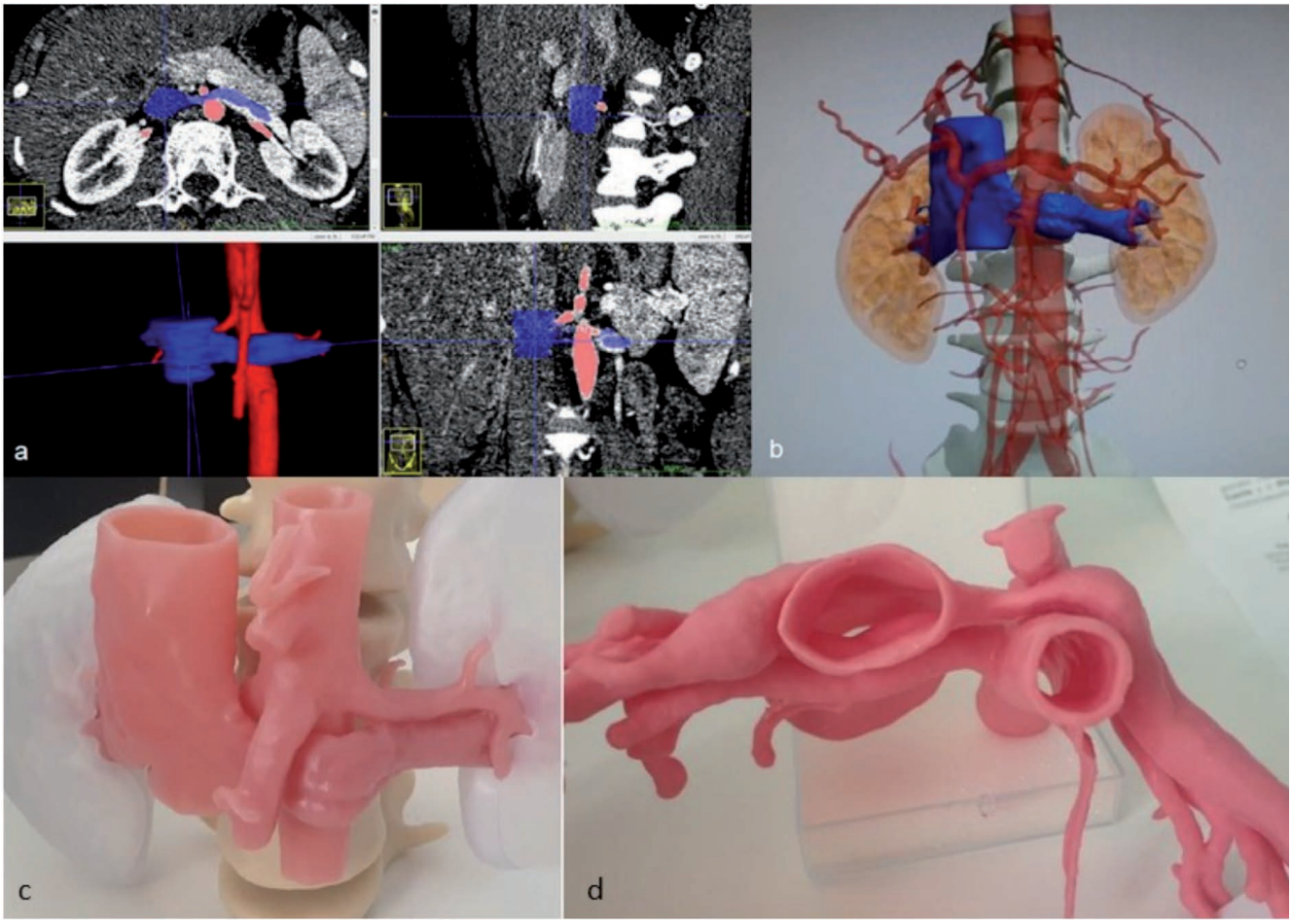

Figure 2. a) CT images (LRV and inferior vena cava in blue, SMA and AA in red); b) 3D model after image conversion; c-d) printed model of the patient's vessels obtained from 3D reconstruction of the CT images.

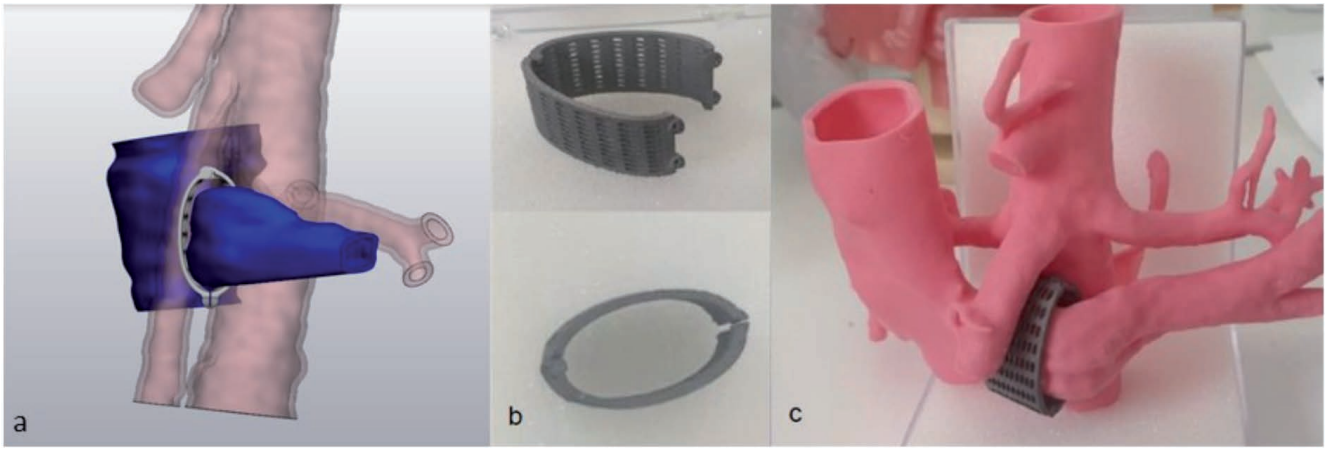

Figure 3. a) individualized extravascular stent project using the 3D software; b) stent model and final titanium device; c) expected result after stent positioning.

through the umbilical port and placed at the site of venous compression. The device was closed around the vessel and fixed to the fibrous periaortic tissue using silk sutures. Restoration of venous flow and absence of vessel kinking were verified. Finally, a laminar drain was placed near the stent. The mean duration of surgery was 180 ± 10 minutes. The hospital stay ranged from 4 to 7 days. All patients were discharged without complications.

The postoperative follow-up ranges from 12 to 24 months. Clinical evaluations are scheduled once a month during the first six months, then upon a trimestral basis. So far, all patients are free of symptoms and have gained their previous weight within six months from surgery. Renal doppler ultrasound is performed every three months during the first year after surgery; homogenous LRV caliber and blood flow have been observed in all patients (Figure 5).

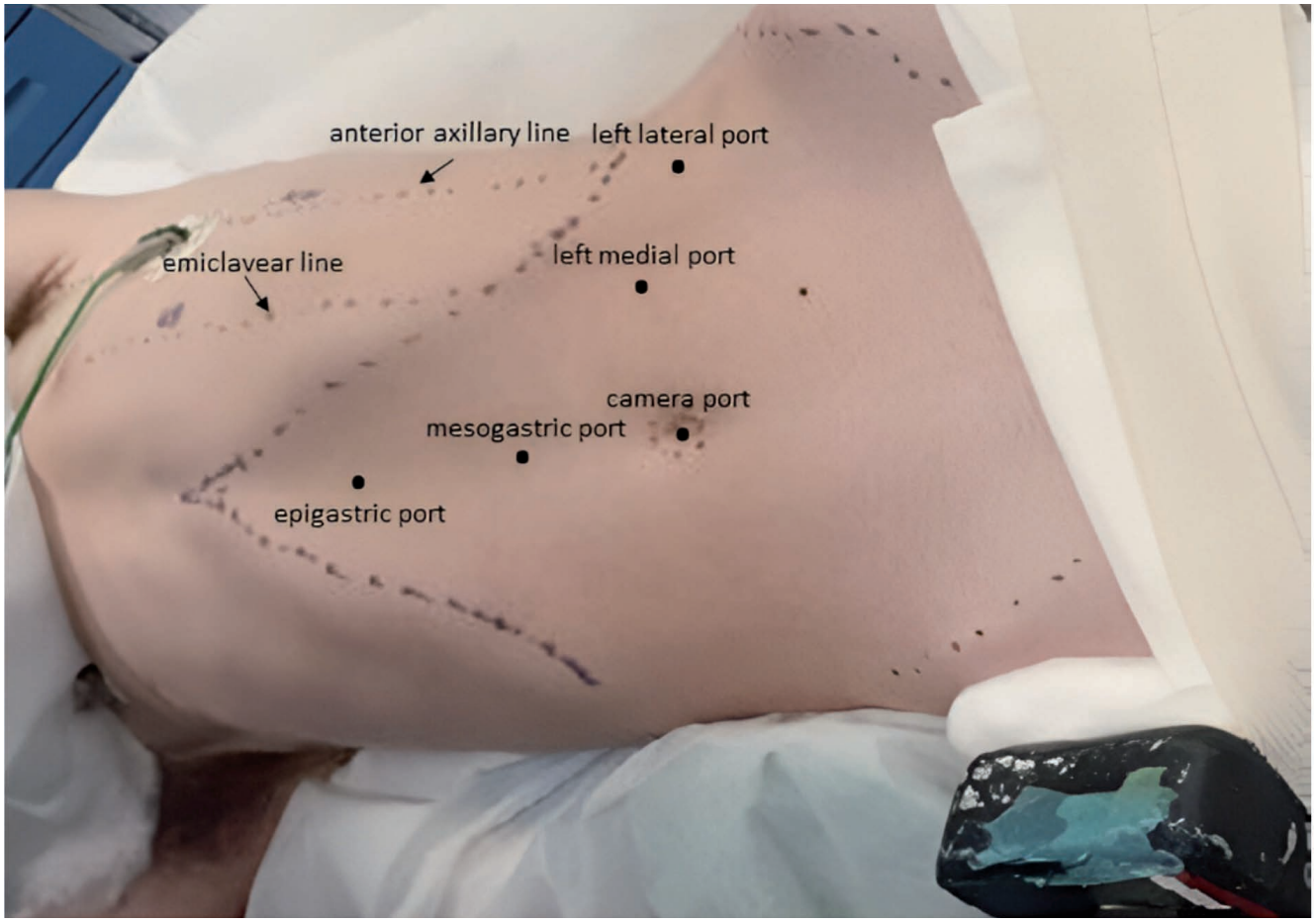

Figure 4. Trocar disposition. The patient is placed in right lateral decubitus with left flank elevation.

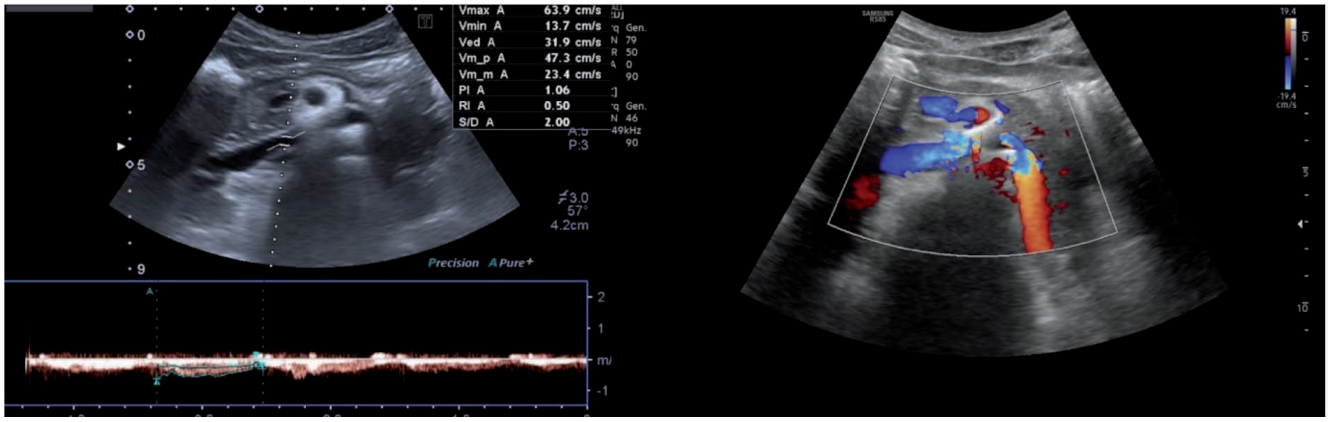

Figure 5. Patient 1: post-operative doppler ultrasound showing an unimpeded blood flow at the stent level.

Specifically, the stent did not impede normal blood flow and PVR reduction was demonstrated one month after surgery (Table 1). This reduction has been confirmed at the following ultrasounds performed three and six months after surgery.

All patients underwent a post-operative CT scan three months after surgery (Figure 6) that demonstrated AMA increased to 55.0°±4.4 (preoperative value 18.7°±4.3°). No stent migration was observed.

Discussion

The incidence of the nutcracker phenomenon in the general population is unknown, as many cases remain asymptomatic. Considering NCS, a bimodal peak incidence is described, with most cases diagnosed in adolescence/young adulthood and in the third/fourth decade of life. The incidence appears higher in females and in individuals with low BMI.[2] Correlation between low weight and nutcracker phenomenon/NCS can be explained by a reduction in retroperitoneal adipose tissue, which decreases the AMA and can facilitate renal ptosis and LRV kinking. NCS manifestations are a consequence of obstructed LRV blood flow. The most reported signs and symptoms are hematuria, abdominal pain and orthostatic proteinuria.[1] Venous congestion can also manifest as left varicocele in men or pelvic congestion syndrome in women.[3] When suspecting NCS, the most important diagnostic tools are doppler ultrasound and CT angiography. Sonographic signs suggestive for NCS include flow velocity at the stenosis site >100 cm/s and LRV DR (hilar-to-aortomesenteric diameter ratio) >3 in supine position or >5 after 15 minutes in orthostatic position.[4] The most important CT findings are stenosis of the LRV between the SMA and AA ("beak sign"), AMA angle below 32° (normal values range between 38° and 65°) and a LRV DR > 4.9.[2] Retrograde venography directly measures the pressure gradient between the LRV and inferior vena cava; a gradient > 3 mmHg is considered diagnostic for NCS.2 de Los Reyes *et al.*[5] proposed a diagnostic algorithm that can be adopted when encountering patients with symptoms suggestive of NCS to guide further management. In patients <18 years of age, conservative treatment is generally recommended as the first-line option as growth of adipose tissue may alleviate venous compression, while surgery should be considered if symptoms persist after two years of follow-up.[6] Open surgery interventions include LRV, SMA or left gonadal vein transposition; reno-caval bypass; left kidney autotransplantation; nephropexy and nephrectomy. Nowadays, these techniques are rarely used due to the associated high morbidity compared to mini-invasive ones. In recent years, endovascular surgery has gained popularity due to its efficacy and safety. However, it is not void of complications, most frequently stent migration in the venous system (7.3% of cases as reported in literature)[3] or venous occlusion. Furthermore, the lack of long term follow-up does not permit evaluating its safety in young patients, in which the LRV lumen may increase with growth. Therefore, in recent years, mini-invasive extravascular stent placement has been proposed as a safe and effective technique, associated with low morbidity and complication rates.[6] Extravascular stents can also be considered as a second line treatment in case of endovascular stent migration.[7]

In our center, conservative treatment is the first option for NCS. Nutritional support is needed to increase the retroperitoneal adipose tissue and the AMA, while psychological consultations are fundamental in helping patients to cope with chronic pain. Surgical treatment is a second-line option when conservative therapy fails. After reviewing the literature, we chose to place an extravascular stent, tailored for each patient thanks to anatomic reconstruction and 3D printing software. These tools effectively guide the choice of the surgical approach (open vs. mini-invasive, transperitoneal vs. retroperitoneal) and allow the creation of a patient-specific stent, whose length, width and convexity can be adapted to the AMA and the vessels' length and caliber. The optimization of the stent's dimensions and size renders its insertion and positioning easier and reduces the risk of dislocation or vessel damage. We chose titanium due to its biocompatibility, compressive strength, lightweight and optimal quality/cost ratio.[8]

Our series, along with other reports in literature,[8,9] confirms that extravascular stent is an effective and safe treatment option also in the pediatric population. The transperitoneal mini-invasive approach is safe and feasible in expert hands and no patient necessitated open conversion. Based on current literature, there are no studies providing long-term follow-up in pediatric patients. In our series, we chose a stent size that could adapt to the vessel's diameter increase, basing ourselves on the 3D-models. More studies are needed in order to evaluate how the stent may adapt to patients' growth.

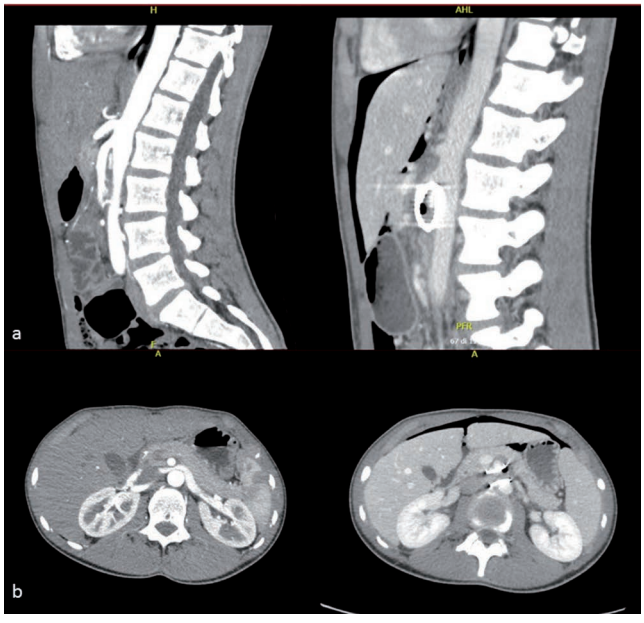

Figure 6. Patient 1: pre and post-operative CT scan (a. sagittal and b. transverse sections) showing correct ES position and resolution of LRV stenosis.

References

1. Agarwal A, Litra F, Barr LL. A rare cause of abdominal and flank pain in children: nutcracker syndrome. Cureus 2021;13:e16422.
2. Granata A, Distefano G, Sturiale A, et al. From nutcracker phenomenon to nutcracker syndrome: a pictorial review. Diagnostics 2021;11:101.
3. de Macedo GL, dos Santos MA, Sarris AB, Zanetti Gomes R. Diagnosis and treatment of the Nutcracker syndrome: a review of the last 10 years. J Vasc Bras 2018;17:220-8.
4. Zhang H, Li M, Jin W, et al. The left renal entrapment syndrome: diagnosis and treatment. Ann Vasc Surg 2007;21:198-203.
5. de Los Reyes T, Keefe DT, Rickard M, Lorenzo AJ. Diagnosis and therapeutic strategies for nutcracker syndrome. Curr Opin Urol 2021;31:135–7.

6. Chen FM, Chen XL, Jiang XN, et al. A new technique: laparoscopic resection of fibrous ring and placing extravascular stent in patients with nutcracker syndrome: a report of 5 cases. Urology 2019;126:110-5.
7. Tian L, Chen S, Zhang G, et al. Extravascular stent management for migration of left renal vein endovascular stent in nutcracker syndrome. BMC Urol 2015;15:73.
8. Wang H, Guo YT, Jiao Y, et al. A minimally invasive alternative for the treatment of nutcracker syndrome using individualized three-dimensional printed extravascular titanium stents. Chin Med J (Engl) 2019;132:1454-60.
9. He D, Wang H, Jiao Y et al. Successful resolution of nutcracker syndrome with 3D printed PEEK extravascular stent in an adolescent boy. Acta Clin Croat 2022;61:138-44.