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# A Case of Nutcracker Syndrome Combined with Wilkie Syndrome with Unusual Clinical Presentation

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Data Collection B  
Statistical Analysis C  
Data Interpretation D  
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**Conflict of interest:** None declared

**Patient:** Male, 27-year-old  
**Final Diagnosis:** Nutcracker syndrome  
**Symptoms:** Vomiting  
**Medication:** —  
**Clinical Procedure:** Endovascular stent grafting  
**Specialty:** Gastroenterology and Hepatology • Radiology

**Objective:** Rare disease

**Background:** Nutcracker syndrome and Wilkie's syndrome are rare vascular diseases due to the abnormal course of the superior mesenteric artery originating from the abdominal aorta with reduced angle ( $<22^\circ$ ) and consequent compression of the left renal vein (nutcracker) and duodenum (Wilkie). Here, we report the case of a patient with a rare combination of these 2 syndromes and with unusual clinical manifestation of post-prandial pain.

**Case Report:** We describe the case of a young male patient with rapid weight loss, coupled with post-prandial abdominal pain, with sub-acute onset, not associated with other symptoms. The ultrasound examination found an aorto-mesenteric angle of  $18^\circ$  and compression of the left renal vein and left varicocele. A CT study was performed to exclude oncological diseases and/or other pathologies responsible for the pain and weight loss, which confirmed the ultrasound findings and showed compression of the third part of the duodenum. The patient underwent endovascular treatment, with stent placement in the left renal vein, which resolved the vascular compression and of the duodenum, with regression of symptoms.

**Conclusions:** The ultrasound scan promptly highlighted the reduction of the aorto-mesenteric angle and the signs of venous congestion of the left renal vein. Based on this experience, in patients with weight loss and post-prandial pain, in our opinion, diagnostic investigations should also be extended to the study of the aorto-mesenteric angle to confirm or exclude any vascular and/or duodenal compression.

**MeSH Keywords:** Mesenteric Artery, Superior • Renal Nutcracker Syndrome • Superior Mesenteric Artery Syndrome • Ultrasonography, Doppler, Color

**Abbreviations:** CT – computed tomography; MR – magnetic resonance

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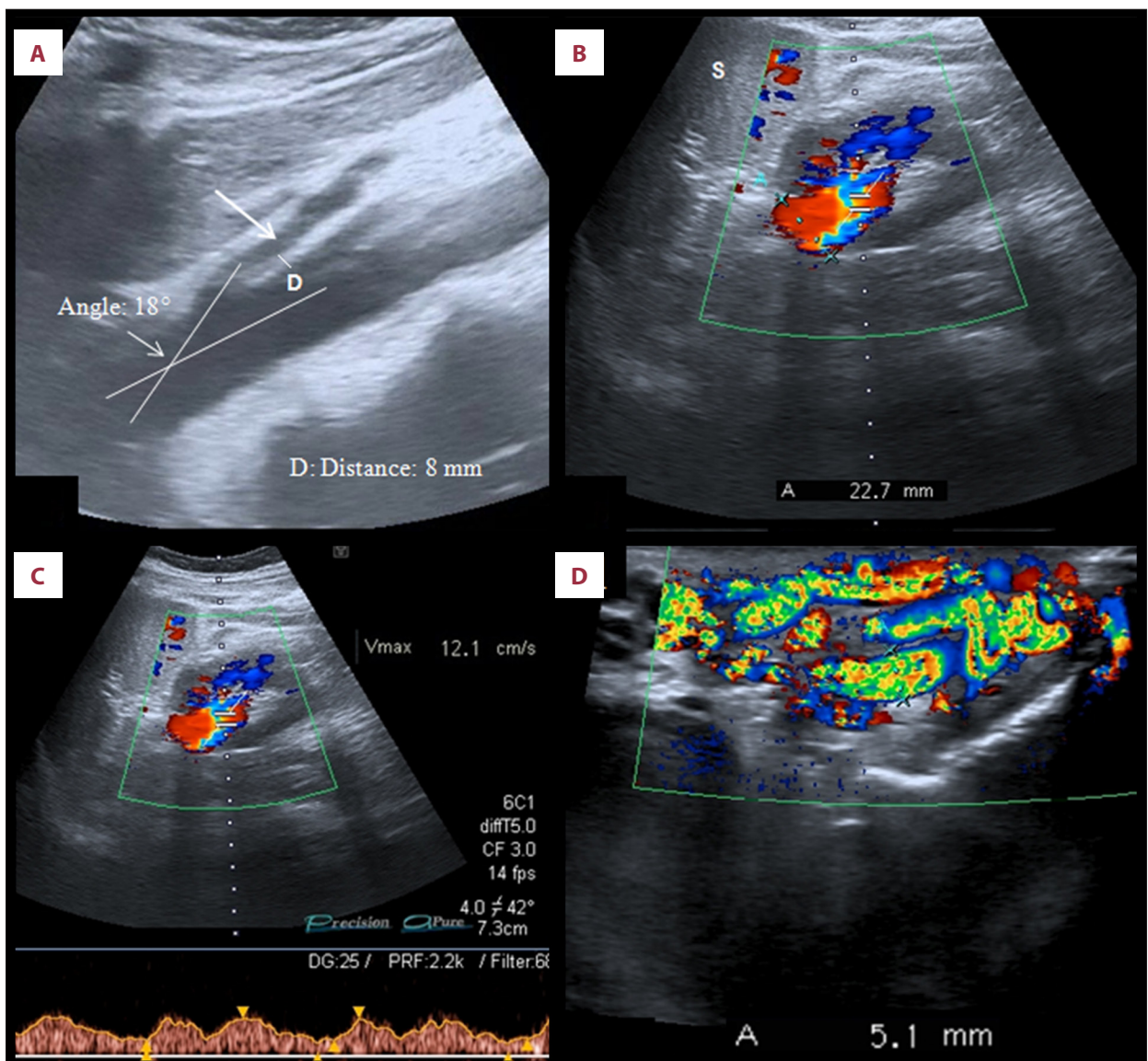
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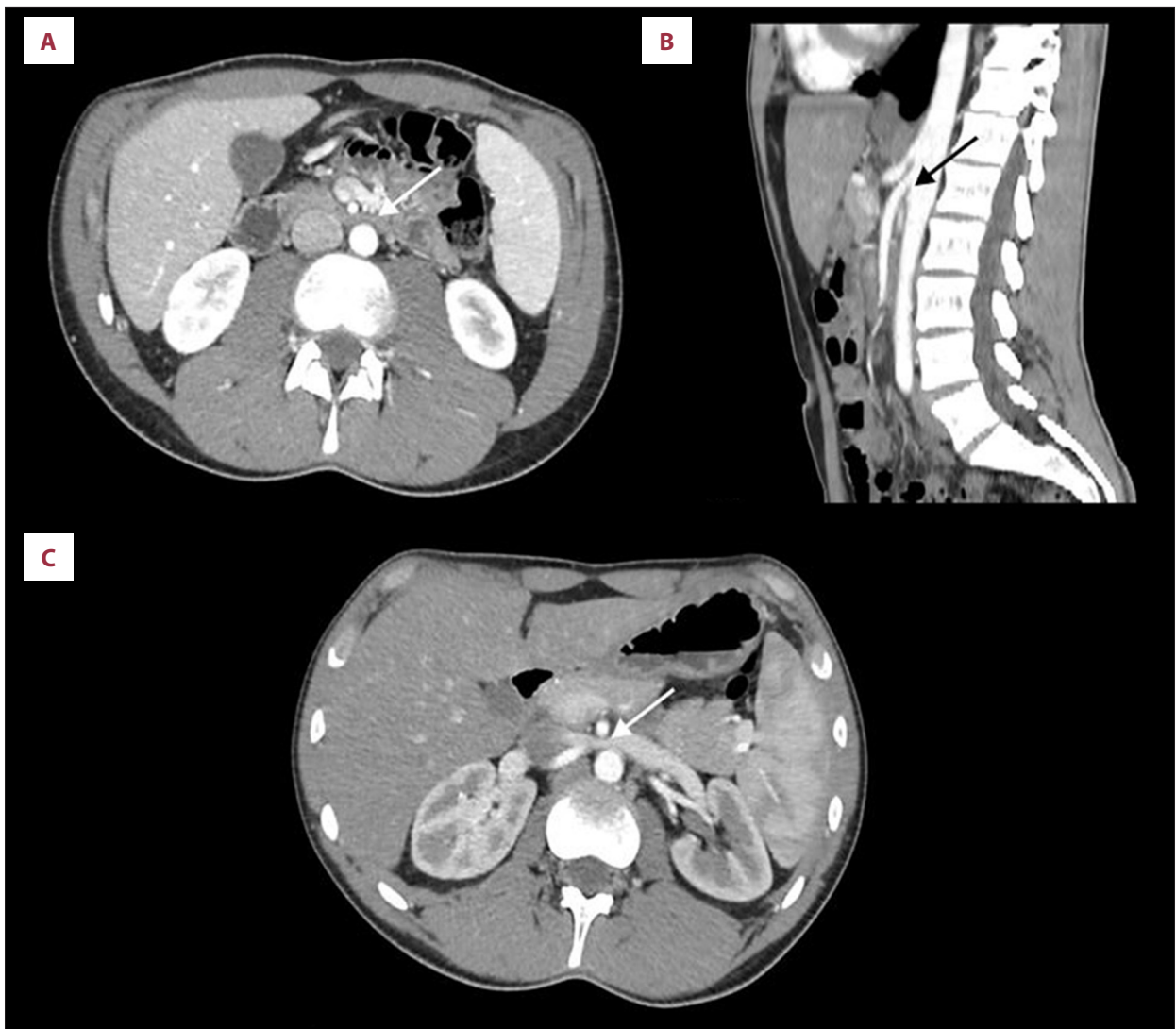
## Background

The Wilkie syndrome [1] and nutcracker syndrome [2] are rare vascular pathologies caused by an anomalous course of the superior mesenteric artery which, at the origin, presents angulations with the abdominal aorta less than 22 degrees and a distance between the superior mesenteric artery and aorta less than 8 mm. The combination of these 2 syndromes is very rare [3]. In nutcracker syndrome, reduction of the aorto-mesenteric angle causes compression of the left renal vein with

consequent congestion of the venous outflow from left kidney and consequent varicocele; while in Wilkie's syndrome, the reduced aorto-mesenteric angle and reduced aorto-mesenteric distance causes duodenal compression resulting in sub-occlusive crisis, vomiting, and post-prandial pain. This syndrome can be congenital or acquired. The congenital form is less frequent and symptoms can already manifest in developmental age. The most frequent is the form, in which the reduction of the aorto-mesenteric angle is due to a reduction of the fat surrounding the abdominal aorta and superior mesenteric artery,



**Figure 1.** B-mode Ultrasound: longitudinal sub-xyphoid scan of the abdominal aorta performed in supine decubitus position and scans of the left kidney in right lateral decubitus performed before treatment. (A) The aorto-mesenteric angle (short arrow) and the aorto-mesenteric distance (long arrow) appears reduced, at 18° and 7 mm, respectively. (B) The caliber of the left renal vein at the hilum is 22.7 mm. Spleen (S). (C) The duplex Doppler shows a flow reduction in left renal vein (maximum speed of about 12.1 cm/s) compared to the contralateral vein. (D) The scans with color Doppler, performed in the pampiniform plexus, show the presence of left varicocele (vein diameter 5.1 mm) (IV degree of Sarteschi).



**Figure 2.** CT scan of the abdomen. (A) The reconstruction on an axial plane shows the narrowing of the aorto-mesenteric angle and the compression of the duodenum (white arrow). (B) The reconstruction according to a sagittal plane show the characteristic vascular pattern of Wilkie's syndrome with aorta and superior mesenteric artery taking on the beak-like appearance (black arrow). (C) This scan shows the compression of the left renal vein (white arrow).

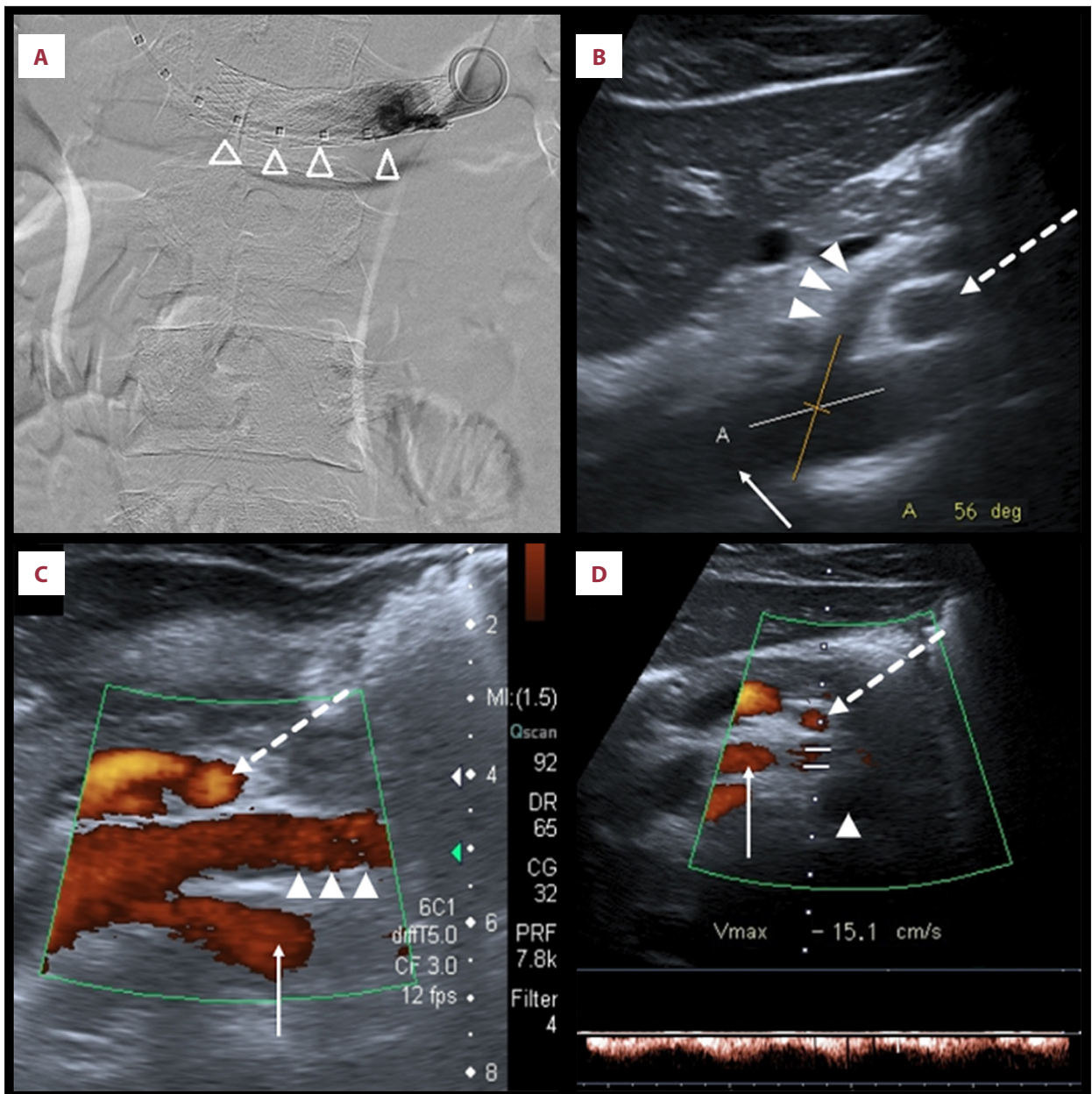
mainly caused by rapid weight loss, which is very common in anorexic patients [4].

The symptomatology is non-specific and is common to many other abdominal pathologies; the most frequent signs they are belching, abdominal fullness and post-prandial pain, biliary reflux, and biliary vomiting and nausea. Depending on the degree of obstruction of the duodenum, patients can only have post-prandial pain if the obstruction is mild and may increase if the obstruction becomes severe with weight loss, nausea, and bilious vomiting [5].

The preferred treatment when symptoms are mild consists of a high-calorie diet to induce the restoration of normal layer

of peri-vascular adipose tissue and normal angulation of the aorto-mesenteric angle [6]. The other 2 therapeutic approaches are surgical treatment [7] and endovascular stenting [8]. The elective surgical treatment consists of resection of the first duodenal loop and retro-vascular duodenum and in the packaging of the anastomosis between the duodenum and the second duodenal loop, which are carried anteriorly. Endovascular stenting consists of positioning a stent in the left renal vein to restore the correct aorto-mesenteric angle and regular flow. Ultrasonography is the first-line exam for the diagnosis of nutcracker syndrome; it allows clinicians, with high sensitivity, to measure the angle and aorto-mesenteric distance and to assess the congestion of the left renal vein and the presence of the left varicocele. In patients with elevated intestinal meteorism,



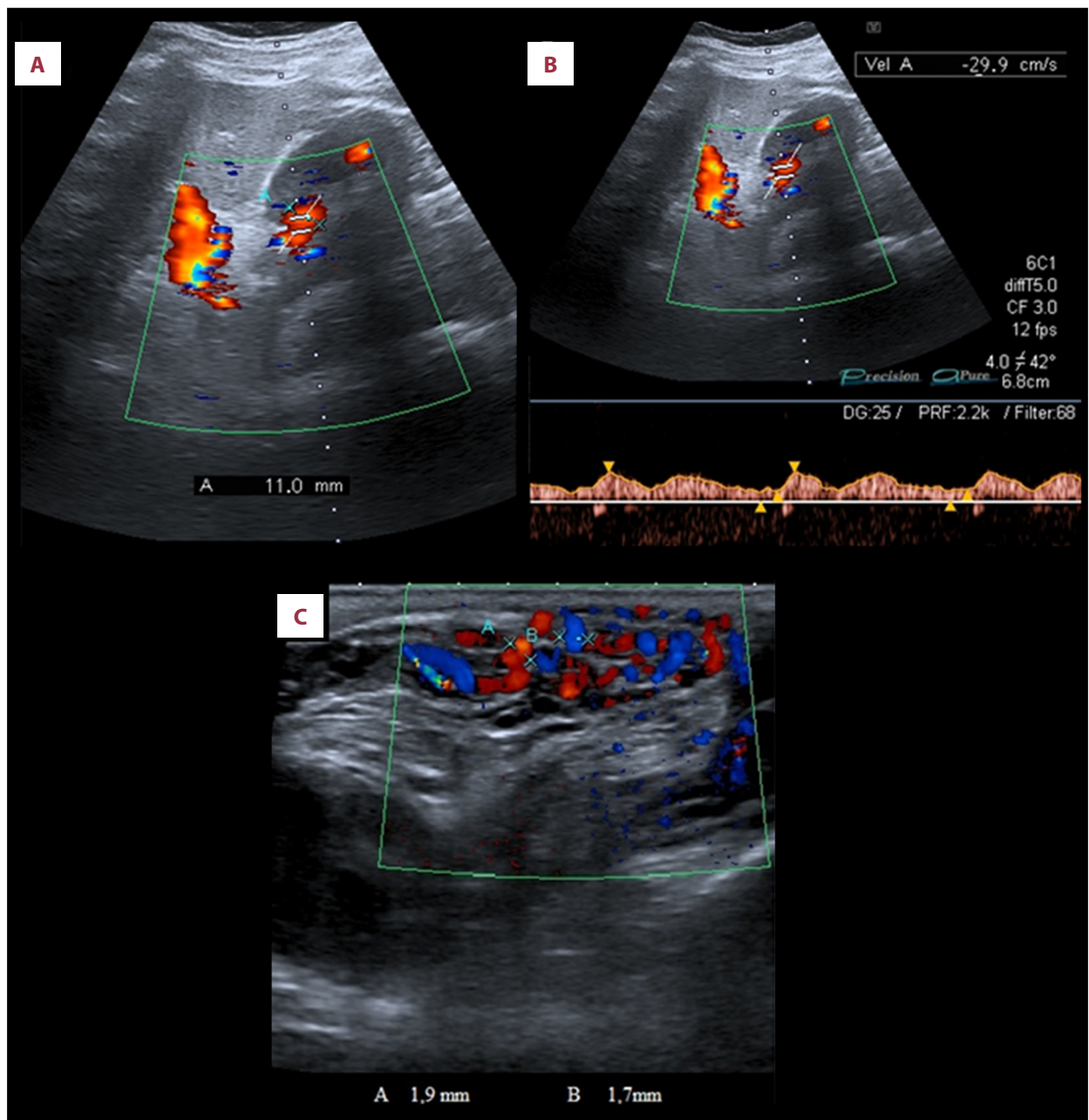


**Figure 3.** (A) This angiographic image shows the stent after positioning in the left renal vein (arrow heads). (B) The B-mode exam shows the presence of the endovascular stent in the left renal vein (dotted arrow). Upper mesenteric artery (arrow heads). Abdominal aorta (short arrow). Aorto-mesenteric angle. (C) The power Doppler scans demonstrates the presence of flow within the vascular stent (arrow heads). Upper mesenteric artery (dotted arrow). Abdominal aorta (short arrow). (D) The duplex Doppler test confirms the patency of the vascular stent (short arrow) showing a flow with a peak velocity of about 15.1 cm/s. Abdominal aorta (arrow head). Upper mesenteric artery (dotted arrow).

which hinders ultrasound examination, a CT scan may be indicated, which allows assessing both vascular alterations and duodenal compression [9,10]. We describe below a rare case of combined Wilkie's syndrome and nutcracker syndrome associated with an unusual clinical manifestation of post-prandial pain not associated with other symptoms.

### Case Report

Our patient was a 27-year-old man, height 1.75 m, weight 63 kg, who was underweight (body mass index 17.37) and had experienced rapid weight loss (loss of 10 kg in 3 months), with painful post-prandial crises at the sub-acute onset, located at the epigastrium.



**Figure 4.** Color Doppler scans of the left renal vein and pampiniform plexus after the positioning of the endovascular stent. (A) The scans performed in right lateral decubitus, after endovascular procedure, show a caliber reduction of the left renal vein (diameters of 11 mm). (B) Increased flow of the left renal vein (29.9 cm/s). (C) Color Doppler scans, performed in supine decubitus position, in the pampiniform plexus, demonstrate regression of the left varicocele (maximum vein diameter <1.9 mm).

The patient underwent ultrasound examination with B-mode, color, power, and duplex Doppler of the abdomen. An Aplio XG (Toshiba) ultrasound device, with a 3.5 MHz convex and a 7.5 MHz linear probe, was used. The ultrasound scans showed a reduction of the aorto-mesenteric angle (18°), measured at the origin, and aorto-mesenteric distance (7 mm) measured at 2 cm from the origin; a compression of left renal vein in the

tract of passage between the superior mesenteric artery and aorta (diameter <3 mm) (Figure 1A). The scan of the left renal vein, performed at the renal hilum, showed a diameter of about 11 mm (Figure 1B) and a maximum speed of about 12.1 cm/s (Figure 1C). Ultrasound scans performed in the pampiniform plexus showed the presence of left varicocele (Figure 1D).

The patient did not show changes in the laboratory analysis related to varicocele, such as hematuria, proteinuria, or clinical symptoms, with no groin, lumbar, or testicular pain, and no testicular swelling.

The patient was subsequently subjected to an CT examination that excluded side pathologies. The CT confirmed compression of the duodenum (Figure 2A, 2B) and compression of the left renal vein (Figure 2C). After placement of an endovascular stent in the left renal vein (Figure 3A), the ultrasound showed increased aorto-mesenteric angle ( $56^\circ$ ) (Figure 3B), patency of the stent (Figure 3C) with a maximum flow of 15.1 cm/s (Figure 3D), caliber reduction (11 mm) (Figure 4A), and increased flow (29.9 cm/s) of the left renal vein (Figure 4B), as well as varicocele regression (Figure 4C). In the following days, he had a gradual regression of post-prandial pain.

## Discussion

The measurements to be performed for a correct diagnosis are standardized and widely described in the literature [11]. In our case, the ultrasound finding of an aorto-mesenteric angle reduction ( $<22^\circ$ ) and an aorto-mesenteric distance reduction ( $<8$  mm), associated with flow congestion in the left renal vein with consequent left varicocele, allowed the diagnosis of nutcracker syndrome, while stenosis of third distal duodenal revealed by CT examination allowed diagnosis of Wilkie's syndrome. The singularity of this case is also due to the symptomatology reported by the patient; in fact, the sole presence of intense post-prandial pain crisis not associated with vomiting represents an unusual clinical manifestation of the syndrome, which almost always presents with an emetic crisis. In this regard, other studies describe similar symptomatologic manifestations, but with associated dyspepsia [12,13]; in our case, this expanded the range of diagnostic hypotheses (e.g., pancreatitis, gastritis, gastric ulcer, duodenal ulcer, cholelithiasis, hiatal

hernia, diverticulitis, and gastro-oesophageal reflux), making diagnosis more complicated. Ultrasound does not allow a complete diagnostic assessment since the study of duodenal compression diagnostic integration with other methods such as RM-enterography [14], fluoroscopy [15], and endoscopy [16] is necessary. The best treatment to achieve restoration of the correct aorto-mesenteric angle, after the failure of a high-calorie diet, is, in our opinion, the interventional procedure, which is less invasive, less expensive, and involves shorter hospital stay compared to surgery. However, it must be pointed out that, due to the complications of endovascular stenting, especially the migration and occlusion of the stent, controversies still exist regarding the optimal treatment.

## Conclusions

The importance of this case lies in the combination between 2 rare syndromes and the unusual symptomatology. Thanks to the ultrasound examination, it was possible to reach the diagnosis of nutcracker syndrome. Ultrasound has many advantages, including the absence of ionizing radiations, repeatability, and low cost. In our opinion, based on this experience, in patients with weight loss and post-prandial pain, diagnostic investigations should also be extended to the study of the aorto-mesenteric angle to confirm or exclude any vascular compression.

## Acknowledgements

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## Conflict of interest

None.

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