



## Case Report | Olgu Sunumu

### RARE CAUSE OF SMALL BOWEL OBSTRUCTION: WILKIE'S SYNDROME, A CASE REPORT

#### İNCE BARSAK OBSTRÜKSİYONUNDA NADİR BİR ETYOLOJİ: WİLKİE SENDROMU, OLGU SUNUMU

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#### ABSTRACT

Mechanical blockage of the bowel frequently causes abdominal pain, nausea, vomiting, and distention. Post-surgical adhesions are the most common etiology. Wilkie syndrome develops as a result of compression of the third part of the duodenum between the aorta and the superior mesenteric artery, and it is extremely rare. The patients are mostly cachectic due to inadequate nutrition. Surgical intervention, especially duodenojejunostomy, is a preferred method in the treatment of the disease. A multidisciplinary approach is important in the diagnosis, treatment, and management of the disease.

**Keywords:** Wilkie syndrome, superior mesenteric artery syndrome, duodenojejunostomy, duodenal obstruction

#### ÖZ

Mekanik barsak obstrüksiyonunda sıklıkla karın ağrısı, mide bulantısı, kusma ve distansiyon görülür. Sıklıkla operasyon sonrası yapışıklıklara sekonder olarak görülür. Wilkie sendromu duodenum üçüncü kısmının aort ve superior mezenterik arter arasında sıkışması sonucunda oluşan, ender görülen, mekanik barsak obstrüksiyon sebeplerinden biridir. Hastalar bulantı, kusma, yetersiz beslenme nedeniyle çoğunlukla kaşektiktir. Cerrahi girişim, özellikle duodenojejunostomi, hastalığın tedavisinde tercih edilen bir yöntemdir. Multidisipliner yaklaşım hastalığın tanı, tedavi ve yönetiminde önemlidir.

**Anahtar Kelimeler:** Wilkie Sendromu, süperior mezenter arter sendromu, duodenojejunostomi, duodenal obstrüksiyon

## Introduction

The signs and symptoms of Wilkie's syndrome (WS), an extremely rare condition, include nausea, bilious vomiting, postprandial epigastric discomfort, anorexia, and weight loss.<sup>1</sup> It is also referred to in the literature as "cast syndrome", "arterio-mesenteric duodenal compression syndrome", "superior mesenteric artery syndrome", and "chronic duodenal ileus".<sup>1,2</sup> WS incidence as determined by radiological investigations It ranges from 0.2% to 0.78%.<sup>3</sup> The incidence in the general population has been estimated to range from 0.0024% to 0.34%.<sup>1-3</sup> The prevalence of having WS is between 0.0024% and 0.3%.<sup>3</sup> Although it can occur at any age, it more frequently affects children and teenagers. Most individuals receive their first diagnosis between their first and fourth decades. The female-to-male ratio by gender is 3:2. No racial propensity is mentioned. Patients who have undergone surgical correction for scoliosis, congenitally short or hypertrophic Treitz ligament, duodenal malrotation, or an aortic aneurysm usually experience it. Numerous studies in the literature have reported a familial predisposition in the literature.<sup>4</sup> The pathogenesis is due to compression of the third segment of the duodenum between the superior mesenteric artery and the abdominal aorta. The aorta mesenteric angle narrowing, which is 20° (normal: 38-65°), the decrease in aortomesenteric distance, which is 10 mm (normal: 10-28 mm), and proximal duodenal dilatation are significant in the diagnosis of WS.<sup>1-4</sup> Surgery is recommended when conservative treatment fails. Although laparoscopic duodenojejunostomies are currently performed, open duodenojejunostomies may be preferred. In this article, we present a 54-year-old

male patient with WS that we treated with a duodenojejunostomy.

## Case Report

A 54-year-old man who had been experiencing postprandial epigastric pain, nausea, bilious vomiting, loss of appetite, and gradual weight loss for six months was admitted to our outpatient clinic. It was found that the patient. Who had a history of diabetes mellitus, did not have his endocrinology outpatient controls and had irregular drug use. He was cachectic, and his body mass index was 15.9 kg/m<sup>2</sup>. Abdominal examination showed abdominal distension and mild tenderness in the epigastric area. In laboratory examinations glucose: 237 (70-105 mg/dL), creatinin: 2.91(0.7-1.3 mg/dL), total protein: 49.9 (63-86 g/L), albumin: 28.5 (35-55 g/L), amylase: 159 (25-125 U/L), lipase: 123 (13-60 U/L), c reactive protein: 34.5 (0.1-5 mg/L), WBC: 12.5 (4-10.5 10<sup>3</sup>/μL). A nasogastric catheter was inserted into the patient, and 4.5 liters of bile content were drained. X-rays of the abdomen and lungs revealed that the stomach was ptotic to the pelvis (Figure 1). In the gastroscopy, alkaline reflux, erythematous pangastritis, and an enlarged stomach were observed. While the duodenal bulb was normal, enlargement was observed in the second and third parts of the duodenum. No stricture or obstructive pathology was observed (Fujinon EG 530, Japan) (Figure 2). An intravenous and oral contrast-enhanced abdominal computerized tomography showed gastric ptozsis, dilatation of the second part of the duodenum, and compression of the third part (Figure 3).

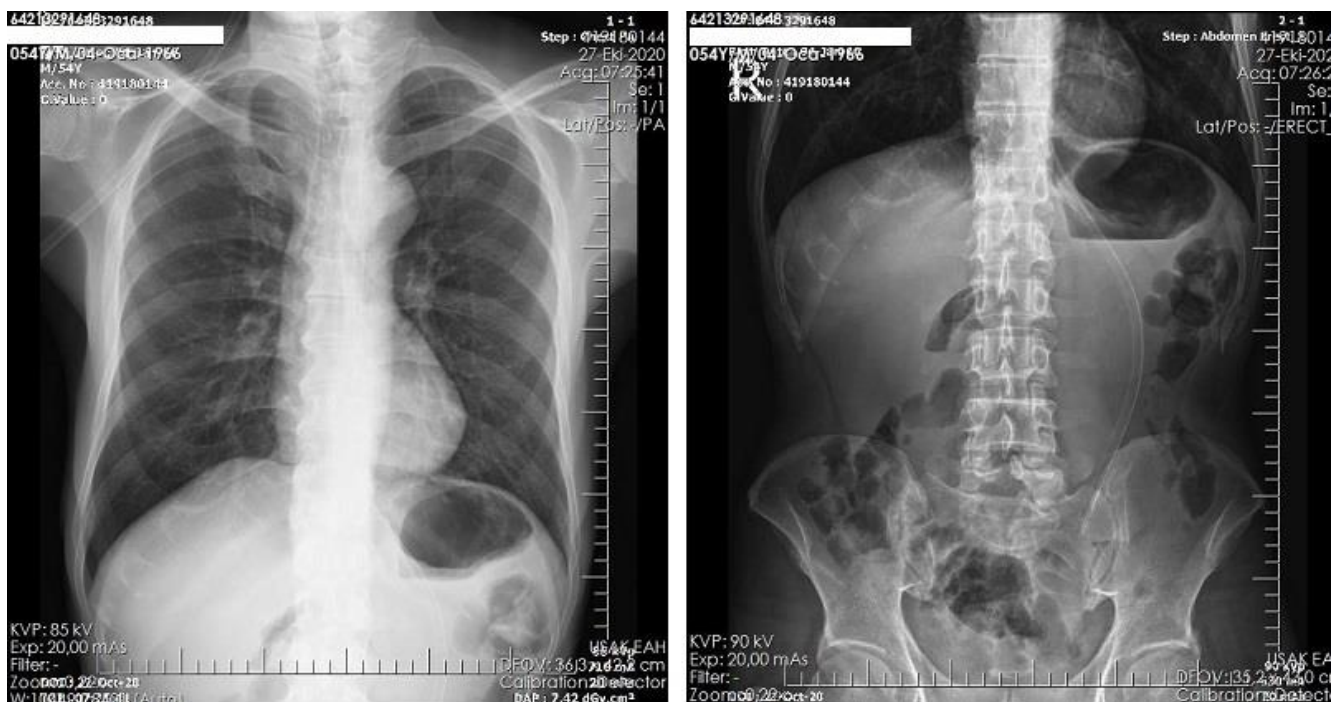


Figure 1. Abdominal and lung x-rays

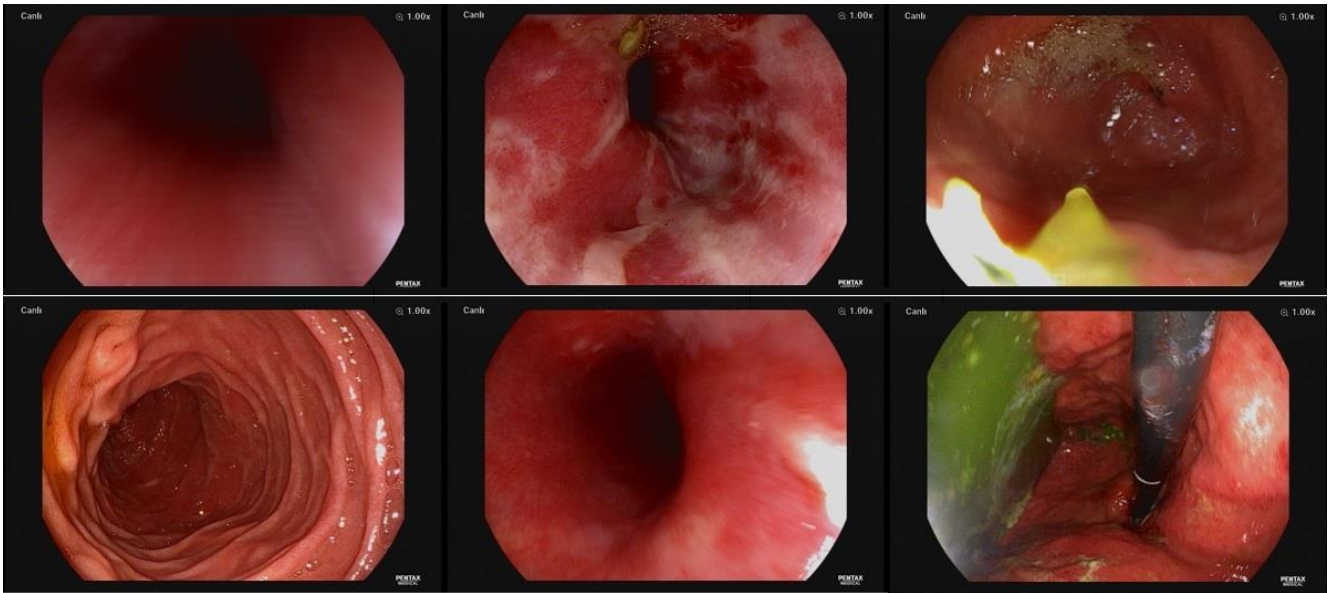


Figure 2. Upper gastrointestinal system endoscopy findings

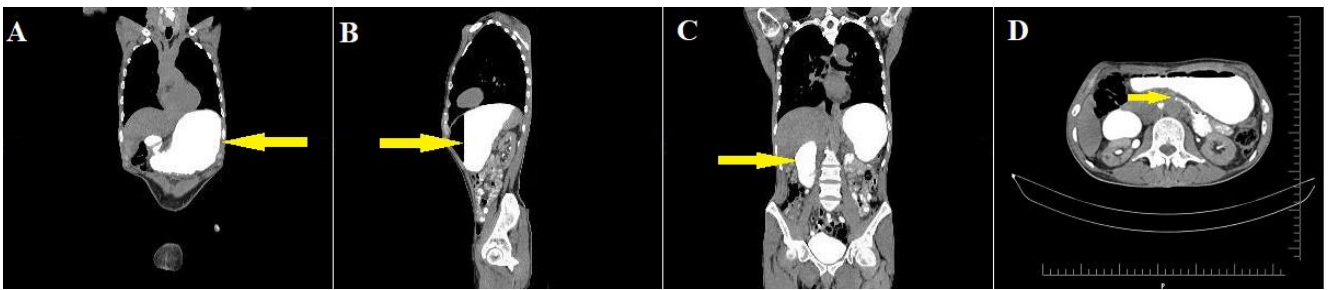


Figure 3. Oral contrast-enhanced abdominal computerized tomography

Aorto-mesenteric angle of  $19.5^\circ$  and the aorto-mesenteric distance of  $< 1$  cm (0.53) (Figure 4). Wilkie's syndrome was diagnosed. After two weeks of total parenteral nutrition therapy, the patient was taken to conservative management therapy. He was discharged home with nasojejunal feeding, given a prescription for erythematous pangastritis. After three weeks of observation, the patient refused to continue her nasojejunal nutrition. The patient was prepared for surgery. During the operation, it was observed that the stomach was ptotic. Duodenum was exposed with Kocher maneuver. It was observed that the duodenum

was dilated from the third part towards the proximal (Figure 5). Retrocolic side to side stapled duodenojejunostomy (60 mm) was done between the 3rd part of the duodenum and jejunum at 40 cm from Treitz (Figure 6). On the 5th postoperative day, gastrograhin passage radiography was taken. No leak was observed and passage from duodenum to jejunum was detected (Figure 7). The patient recovered smoothly and was discharged home on the 14th postoperative day. In the sixth month control, it was detected that the patient received fifteen kilograms.

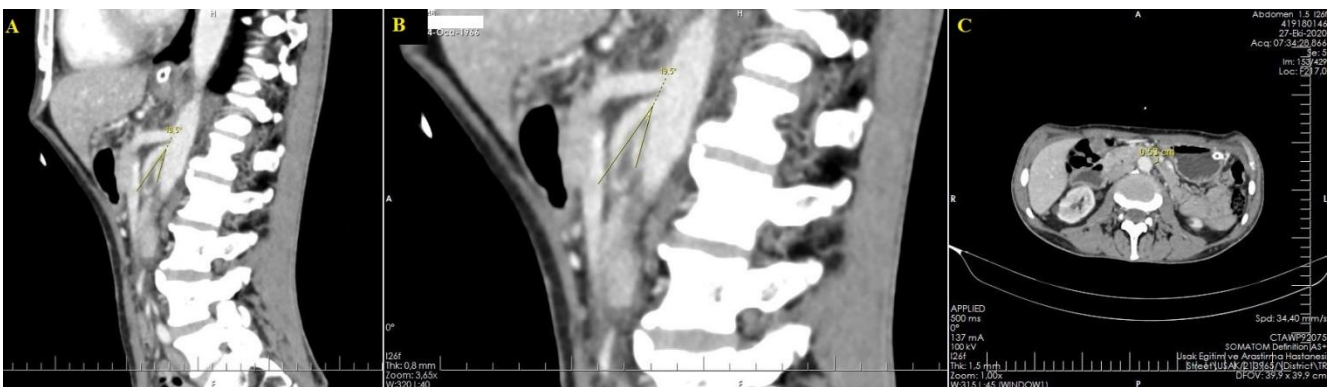


Figure 4. Aorto-mesenteric angle and the aorto-mesenteric distance



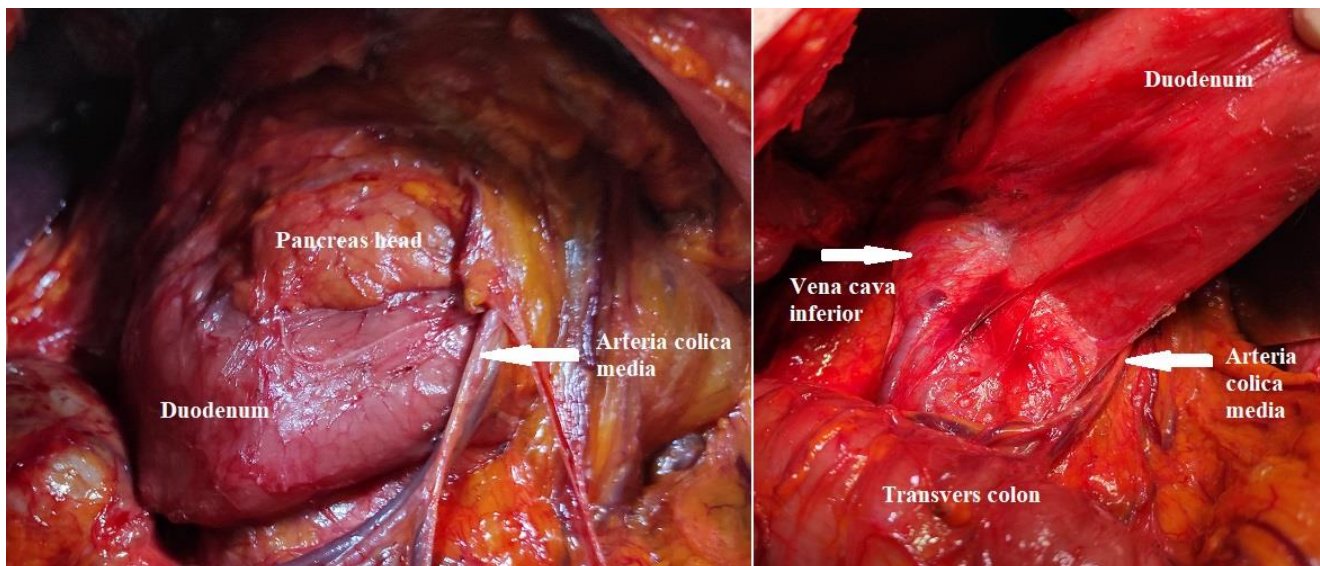


Figure 5. Operation findings

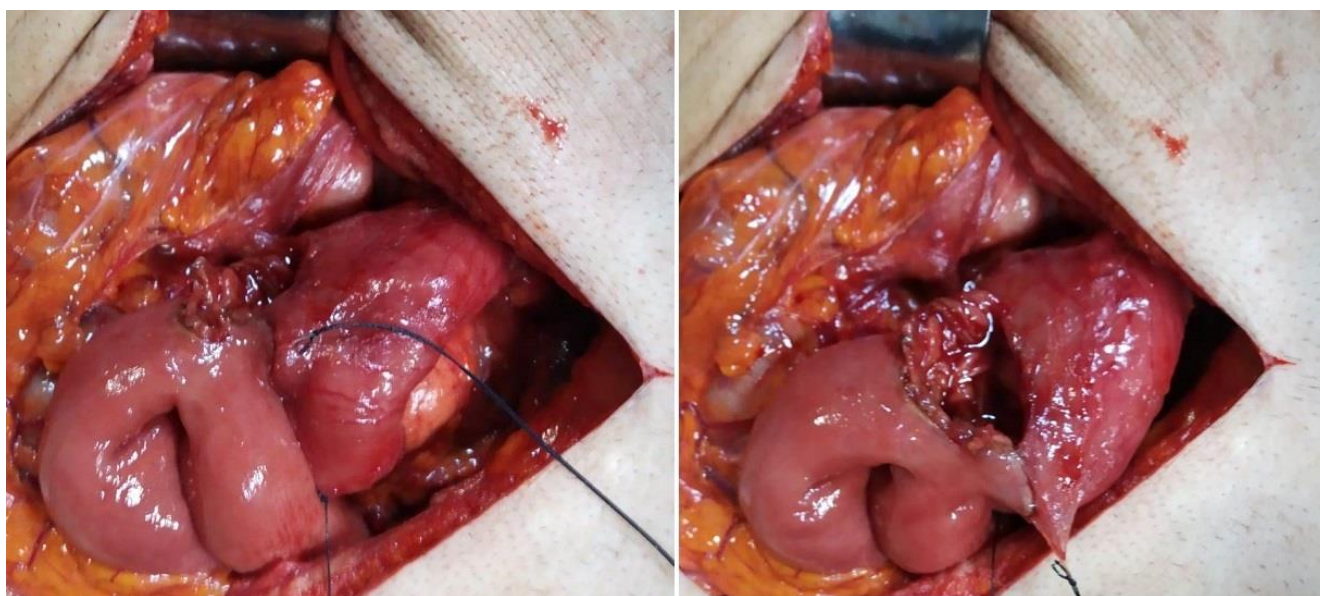


Figure 6. Retrocolic side to side stapled duodenojejunostomy



Figure 7. Passage graphy of duodenojejunostomy

### Discussion

The small bowel mechanical obstruction symptoms in WS are present. Clinical, endoscopic, and radiologic findings are crucial for diagnosis. In the differential diagnosis, it is important to consider pancreatic neoplasm, duodenal or small intestine tumors, gastric antral neoplasm, and duodenal stricture brought on by inflammatory bowel disease. In patients with Wilkie's syndrome, abdominal computed tomography with water-soluble contrast scans of the upper gastrointestinal tractus reveal dilated stomach, proximal duodenum, and visibly compressed third section of duodenum.<sup>3</sup> The normal aorto-mesenteric angle (AMA) and distance are 25°-60° and 10-20 mm, respectively. We measure AMA 19.5° and distance < 1 cm (0.53). WS can be treated medically or surgically. Medical treatment such as bowel rest, fluid replacement, parenteral nutrition, restoration of electrolyte imbalance, and nasojejun feeding may be effective in acute situations.<sup>5</sup> In our case, we started the treatment conservatively, but the operation decision was

made because the patient could not tolerate and refused to be fed with a nasojejun tube. There are three options for the surgical treatment of WS: Strong procedure, gastrojejunostomy, and duodenojejunostomy. In the Strong procedure, it is aimed to widen the distance between the duodenum and aorta by dividing the Treitz ligament without disrupting the intestinal integrity.<sup>6</sup> Gastric decompression is provided by gastrojejunostomy; however, it may not be adequate to relieve duodenal obstruction, necessitating a subsequent procedure.<sup>7</sup> The most popular surgical technique, with a success rate of up to 80-90%, is an open or laparoscopic duodenojejunostomy.<sup>8,9</sup> For the patient, open surgery was preferable. Gastroparesis and gastric atony are significant issues in phytotic stomachs following surgery. To ensure postoperative motility in our situation, we maintained administering metoclopramide intravenously for ten days and neotigmine methylsulfate for two days.

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### Compliance with Ethical Standards

Written informed consent was obtained from the patient for the publication of this case report.

### Conflict of Interest

All author declared no potential conflict of interest with respect to research of this article.

### Author Contribution

All authors contributed equally to this article.

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