

# Rapunzel Syndrome Causing Gastric Outlet Obstruction in A 12-Year-Old Girl: Report of A Case and Review of the Literature

## 12 Yaşındaki Bir Kız Çocukta Mide Çıkım Tıkanıklığına Neden Olan Rapunzel Sendromu: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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### Abstract

Rapunzel syndrome (RS) is a rare condition presenting with hair accumulation in the stomach that extends to small intestine. It may cause gastric outlet and intestinal obstruction. The most common clinical signs are abdominal pain, palpable mobile mass and vomiting. A 12-year-old girl with a history of trichotillomania, trichophagia, attention deficit and hyperactivity disorder presented with gastric outlet obstruction. A bezoar completely filling the stomach and reaching to third part of the duodenum was detected in abdominal ultrasound. Removal of the trichobezoar via gastrotomy was the treatment of choice. Herein we report a 12-year-old girl with RS to discuss the clinical features and treatment options in this rare presentation.

**Key Words:** Bezoar, trichobezoar, Rapunzel Syndrome, child

### Öz

Rapunzel sendromu (RS) midede biriken saçların ince barsakta obstrüksiyona neden olabilecek şekilde uzandığı nadir bir durumdur. Mide çıkımında ve barsakta tıkanıklığına neden olabilir. En sık görülen klinik belirtiler karın ağrısı, ele gelen mobil kitle ve kusmadır. Trikotilomani, trikofaji, dikkat eksikliği ve hiperaktivite bozukluğu öyküsü olan 12 yaşında kız hasta, mide çıkım tıkanıklığı nedeniyle başvurdu. Batın ultrasonunda mideyi tamamen dolduran ve duodenumun üçüncü kısmına uzanan bezoar tespit edildi. Trikobezoarın gastrotomi yoluyla çıkarılması tercih edildi. Burada, bu nadir görülen tablonun klinik özelliklerini ve tedavi seçeneklerini tartışmak amacıyla 12 yaşında RS'li bir kız çocuğunu sunuyoruz.

**Anahtar Kelimeler:** Bezoar, trikobezoar, Rapunzel Sendromu, çocuk

### Introduction

Bezoars are concretions that occur as a result of the accumulation of non-absorbable fibers such as hair and plant fibers in the gastrointestinal tract. The most common type of bezoar is the trichobezoar, which is mostly made of hair. Trichobezoar is a rare condition almost exclusively seen in young females (1). Rapunzel syndrome (RS) is an unusual and rare form of trichobezoar extending into the small intestine. The name "Rapunzel" syndrome comes from the Grimm Brothers' fairytale and this syndrome was first described by Vaughan et al. in 1968 (2).

The most common clinical findings in trichobezoar cases are abdominal pain, palpable firm and mobile masses. However, more serious complications can also be seen in trichobezoars that extend to the small intestines, such as intestinal obstruction, perforation, intussusception, cholangitis and pancreatitis (3,4). A 12-year-old girl diagnosed with RS, which causes gastric outlet obstruction and has trichobezoars extending along the duodenum, is presented to discuss the difficulties experienced in the diagnosis and treatment of trichobezoars with acute gastric obstruction.

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## Case Presentation

A 12-year-old female, admitted to hospital with epigastric mass and non-bilious vomiting. Heart rate was 90/min, respiratory rate was 16/min, blood pressure was 120/80 mmHg and body temperature was 37.5 °C. In her medical history that she has been following up for trichotillomania, trichophagia and attention deficit and hyperactivity disorder. She has been using methylphenidate (20 mg, Ritalin, USA) and risperidone (0.5 mg, Risperdal, Turkey) treatment. Laboratory findings were all within normal limits, including complete blood cell counts, serum electrolyte levels, liver and renal function tests. She complained of weight loss in last ten days. Abdominal examination revealed a non-tender palpable mass in the epigastric region. Abdominal ultrasonography (USG) revealed large foreign body completely filling the stomach. Abdominal computed tomography (CT) scan confirmed a large bezoar completely filling the stomach and extending to the third part of the duodenum (Figure 1). Upper gastrointestinal endoscopy revealed that her entire stomach was filled with trichobezoar and it extended from the gastric cardia to the pylorus. It was not possible to remove the bezoar with forceps and surgical exploration was planned in the same session. Laparotomy was performed with midline incision above umbilicus and the bezoar was removed by gastrotomy on the anterior aspect of the stomach (Figure 2).

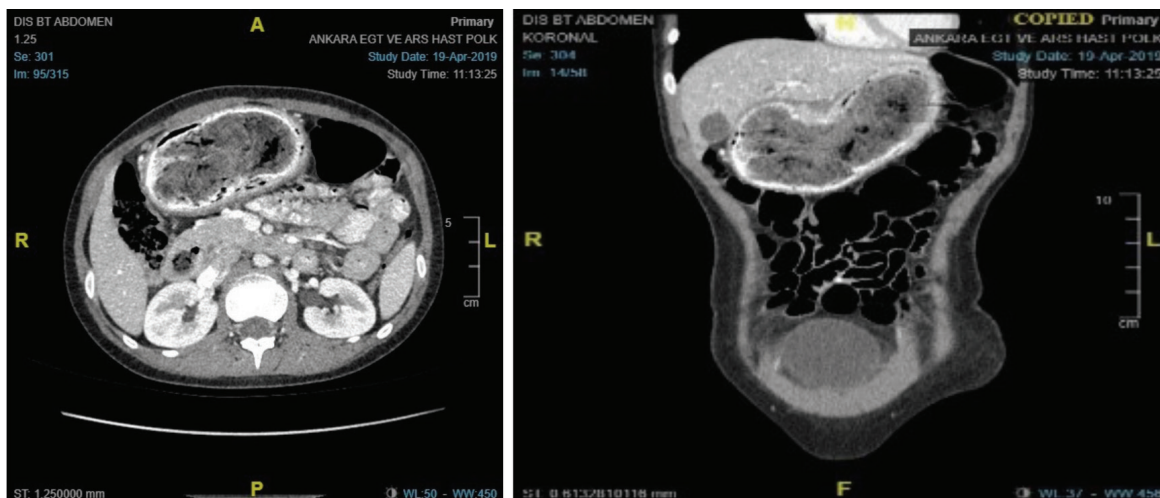
She had an eventful postoperative recovery and fed orally on the 3<sup>rd</sup> postoperative. She was discharged after detailed psychiatric counselling on the 6<sup>th</sup> postoperative day. Two years later, she was admitted to our department with abdominal pain. She had stopped her psychiatric medical treatment for the last 6 months and started trichophagia. The USG showed no gastric outlet obstruction. She was consulted to pediatric psychiatry department again and has been following up with no symptoms.

A detailed PUBMED searched was performed by using the keywords "child, bezoar, obstruction, Rapunzel". Table 1 summarizes these cases with RS with gastric outlet obstruction.

## Discussion

The word "bezoar" comes from the Arabic word "bedzehr" or the Persian word "padzhar," meaning "protecting against a poison" (3). The first reference to a bezoar in a human was in 1779 during an autopsy of a patient who died from gastric perforation and peritonitis (4). Bezoars are named as phytobezoar (fruit, vegetable fibres), lactobezoar (milk residues) and trichobezoar (hair, nylon) according to their content. Trichobezoars are due to the accumulation of indigestible materials such as hair, nylon or wool in the stomach associated with trichotillomania and trichophagia. Human hair is resistant to digestion and peristalsis. As the hair continues to be swallowed, it becomes a mass by accumulating in the stomach folds along with the mucus and nutrients. Although it is more common in girls, mainly because they have long hair, a boy eating his sister's hair has also been reported (5).

RS is a rare form of trichobezoar and trichobezoars extend into the duodenum and small intestine. Affected patients can remain asymptomatic for a long time. The bezoar continues to grow and symptoms appear in the late stages. The most common findings in children are epigastric mass, epigastric pain, nausea, vomiting, weight loss, hematemesis, diarrhea or constipation (6). Besides causing gastrointestinal obstruction, they can also cause stomach ulcers, obstructive jaundice and acute pancreatitis (3,4). Protein-losing enteropathy, iron deficiency anemia and megaloblastic anemia are other complications related malabsorption.



**Figure 1:** Axial and coronal CT scan showing extension of the trichobezoar  
CT: Computed tomography



**Figure 2:** Whole trichobezoar showing stomach cast, duodenal cap and long tail

The bezoar sometimes breaks off from the large mass and causes obstruction in the small intestines and may cause emergency surgical operation (7). In addition to obstruction, they can cause intussusception in different segments of the intestine. USG may be the first choice in diagnosis because the bezoar may cause intussusceptions (10). The acute surgical treatment in RS mostly correlates with obstructions and complications. Trichobezoars that usually cause partial gastric outlet obstruction and complete obstruction is extremely rare (3,11). We found 21 cases reporting RS that cause complete or partial gastric outlet obstruction with small intestine extension (Table 1). The most common clinical symptom was abdominal pain, palpable mass and vomiting. Most of the cases present with non-bilious vomiting. Our patient was diagnosed after the non-bilious vomiting and clinical findings of gastric outlet obstruction. One should be aware of whether

**Table 1: Complete or partial gastric outlet obstruction with Rapunzel syndrome**

Number	Study	Sex	Age of patient	Presenting features	Diagnostic methods	Management
1	Flaherty et al. (17)	Female	1.5	Vomiting Early satiety	CT	Laparotomy
2	Kumar et al. (18)	Male	3	Vomiting Constipation PICA Abdominal pain Abdominal mass	USG/CT	Endoscopy, laparotomy
3	Kim and Nam (19)	Female	8	Vomiting Epigastric pain Abdominal mass	CT	Endoscopy, laparotomy
4	Gonuguntla and Joshi (3)	Female	5	Vomiting Abdominal pain Abdominal mass	CT	Laparotomy
5	Khanna et al. (20)	Female	6	Vomiting Abdominal pain	Abdominal radiograph	Laparotomy
6	Lalith et al. (21)	Female	12	Vomiting Abdominal pain Constipation Loss of appetite	Abdominal radiograph USG CT	Laparotomy
7	Mazzei et al. (22)	Female	14	Vomiting Weight loss Epigastric pain Abdominal mass	USG, CT	Endoscopy, laparotomy
8	Kyin et al. (23)	Female	11	Abdominal pain Vomiting Constipation Weight loss	CT	Laparotomy
9	Czerwińska et al. (24)	Female	16	Vomiting Abdominal mass	USG, CT	Laparotomy
10	Raikar et al. (25)	Female	12	Vomiting Abdominal pain	CT	Endoscopy, laparotomy
11	Winsett et al. (26)	Female	13	Vomiting Diarrhea Epigastric pain	CT	Laparotomy
12	Rogers et al. (27)	Female	2	Vomiting Diarrhea	Endoscopy	Laparotomy

Table 1: Continued						
Number	Study	Sex	Age of patient	Presenting features	Diagnostic methods	Management
13	Al Wadan et al. (28)	Female	7	Vomiting Abdominal pain Distention	Endoscopy	Laparotomy
14	Chogle et al. (29)	Female	3	Vomiting Pain Weight loss	USG, CT, MRI, ERCP	Laparotomy
15	Nwankwo et al. (30)	Female	7	Abdominal pain Vomiting Nausea	Abdominal radiograph	Laparotomy
16	Singla et al. (31)	Female	9	Abdominal pain Vomiting Loss of appetite	USG	Laparotomy
17	Quraishi et al. (32)	Female	5	Vomiting Abdominal mass	Barium contrast study CT	Endoscopy laparotomy
18	Senapati et al. (33)	Female	8	Fever Vomiting Abdominal pain Hematochezia	Abdominal radiograph USG Barium contrast study	Laparotomy
19	Sharma et al. (34)	Female	12	Vomiting Hematemesis	Abdominal radiograph Endoscopy	Laparotomy
20	Hirugade et al. (5)	Male	6	Abdominal pain Vomiting Loss of appetite Weight loss	Abdominal radiograph Barium contrast study	Laparotomy
21	Belsky et al. (35)	Female	6	Abdominal pain Vomiting Constipation	Abdominal radiograph Oral contrast study	Laparotomy
22	Our case	Female	12	Vomiting Abdominal mass Weight loss	USG CT	Endoscopy laparotomy

trichotillomania is in the history or alopecia is on physical examination when bezoar is clinically suspected. The history of these patients may include other psychological disorders that cause bezoar, such as pica, obsessive compulsive disorder, depression, anorexia nervosa and substance abuse. In addition to psychiatric problems, social causes such as family quarrels, school changes, loss of parents can also create a predisposition (8). Our patient had a history of trichotillomania, trichophagia and psychological reasons such as the separation of the mother and father. However, the history of trichotillomania is suggestive for diagnosis, all patients with bezoars should be consulted to psychological evaluation.

Trichobezoars can be detected as a palpable mobile, well-circumscribed mass in physical examination. Erect abdominal X-ray may be useful for diagnosis and evaluation of complications. However, despite the large ileal perforation due to trichobezoar, a case without pneumoperitonium on erect abdominal X-ray was reported, which was associated with the trichobezoar completely covering the perforated area (9). Trichobezoars can be diagnosed a firm epigastric

mass in 88% of cases and USG is often the first imaging study performed in the clinical setting of a lump in the epigastrium (10). USG could be used to support diagnosis but few other causes of upper abdominal masses are showed intense sonic shadow like trichobezoar in USG (11). CT is another radiological option after USG to show the size and configuration of the bezoar and to differentiate it from gastrointestinal tumors such as gastrointestinal stromal tumor extending into the gastric lumen, thus it helps diagnose and demonstrate mechanical bowel complications (12). Although CT and magnetic resonance imaging (MRI) examination are used to exclude other mass causes in the differential diagnosis, upper gastrointestinal system endoscopy is also considered the gold standard for diagnosis. In this case, it was shown that the bezoar extended to the small intestine in the USG and CT performed before the endoscopy. In our case, the stomach was completely filled with hair and the attempts for endoscopic removal was failed. Although most of the patients underwent endoscopy, only one of the patients had successful endoscopic removal (13). Patients with partial gastric obstruction were more eligible for endoscopic treatment.



If endoscopic removal is not possible, removal of bezoars with gastrotomy via laparotomy is recommended. The first successful surgical intervention was made by Schonborn in 1885 (14). The first attempt at laparoscopy was made by Nirasawa et al. (15). In addition to surgical treatment, various other modalities such as extracorporeal shock wave lithotripsy (ESWL), enzymatic therapy, metoclopramide and acetylcysteine have been reported as successful treatment options (15). In our case, surgical treatment was the only choice because of very large trichobezoar completely filling the stomach. Patients with small intestinal extension should also undergo detailed exploration of the entire duodenum and jejunum.

Recurrence of trichobezoar is not uncommon. But psychological treatment is one of the most important steps in these patient's treatment and it is also regular part to prevent recurrence. The most important evidence of this is that the case who had gastrotomy because of bezoar nine years ago, was operated again for bezoar at the age of twenty-five, because she continued her psychiatric treatment irregularly (16). In some cases, as in our patient, although the diagnosis is known, advanced tests and treatments may be required in cases who do not use their medications regularly and do not comply to appropriate treatment. Therefore, preventive measures should be taken into consideration in patients who are incompliant to their psychiatric treatment.

## Conclusion

RS is a rare form of trichobezoars that can cause gastric outlet obstruction and require emergency surgical treatment. When endoscopic methods are insufficient in large masses, detailed imaging methods should be used to evaluate mass size and extension before the surgery. Psychiatric evaluation should be performed in all cases to prevent recurrence.

## Ethics

**Informed Consent:** Informed consent was obtained from the patient's family.

## Authorship Contributions

**Conflict of Interest:** All authors declare no conflict of interest.

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