HK J Paediatr (new series) 2023;**28**:173-177

*Original Article*

# Surgical Management of Rapunzel Syndrome: A Retrospective Report from Two Children's Medical Centres

C ***Wang***, S ***Feng***, J ***Zhang***, Y ***Tian***, Y ***Wu***, W ***Wu***

**Abstract** ***Background:*** Rapunzel syndrome is a stomach trichobezoar with a long hairy tail that extends into the duodenum and small bowel. Endoscopy, often followed by laparotomy, and psychological intervention were all part of traditional treatment. The purpose of this study was to discuss our experience with seven cases of trichobezoars in the gastrointestinal tract. ***Materials and methods:*** A retrospective review of all cases of trichobezoar at two children's hospitals from 2010 to 2020 was performed. Demographic data, presenting complaints, imaging, surgical treatment, and subsequent management were collected. ***Results:*** All 7 patients were female, ages 4 to 13 years (mean, 9.3 years). Although multiple imaging modalities were necessary for preoperative diagnosis, most patients were accurately diagnosed without endoscopic evaluation (71%). All patients required an exploratory laparotomy for definitive treatment. At laparotomy, 3 patients were found to have post-pyloric extension of the trichobezoar (43%). No surgical complications or recurrences were discovered. ***Conclusions:*** Rapunzel syndrome should be diagnosed with a high degree of clinical suspicion, which can be established with a comprehensive history and radiographs, and laparotomy should be indicated. To avoid additional trichophagia and trichobezoars, psychiatric evaluation and management are essential.

**Key words** *Acute abdomen; Intussusception; Rapunzel syndrome; Trichobezoar; Trichophagia*

## Introduction

Rapunzel syndrome is a type of trichobezoar that is extremely rare. It was called after a lovely story about a young woman penned by the brothers Grimm in 1812.[1] It mainly affects young women under the age of 20 and causes abdominal pain, nausea, vomiting, bloating, early satiety, weight loss, diarrhoea, and constipation. In delayed and neglected cases, complications such as anaemia, haematemesis, gastric ulcers, intestinal obstruction, perforation, and peritonitis were frequently reported.[2-4]

Despite the fact that a history of hair eating can aid with diagnosis, patients frequently present to the ER with abdominal pain, making preoperative diagnosis challenging. Abdominal X-rays reveal a nonspecific mass, while computed tomography scans reveal heterogeneous masses containing trapped air.[5] Trichobezoars can be distinguished from other probable epigastric mass aetiologies (such as pseudocyst, duplicated cyst, or tumour) with a computed tomographic (CT) scan.[6] For diagnosis, endoscopy is useful. Endoscopic removal of the bezoar, on the other hand, is rarely a permanent solution. Furthermore, endoscopic bezoar fragmentation can result

**Department of Pediatric Surgery, Hangzhou Children's Hospital, Hangzhou, China**
C **Wang** (汪晨) *MD*
S **Feng** (馮召廣) *MD*
J **Zhang** (章捷) *MD*
Y **Tian** (田由) *MD*
Y **Wu** (吳曄明) *MD*

**Department of Pediatric Surgery, Xinhua Hospital, Shanghai Jiao Tong University School of Medicine. Shanghai, China**
W **Wu** (鄔文傑) *MD*

Correspondence to: Dr W **Wu**
*Email: wuwenjie0416@163.com*

*Received November 19, 2021*

in small bowel blockage. Despite several publications describing laparoscopic techniques to treatment, traditional laparotomy has remained the mainstay of treatment for bezoars that do not flow into the peritoneal cavity.[7,8]

The purpose of this study was to summarise the clinical manifestations, physical examinations, radiological examination, and surgical outcomes of seven cases of Rapunzel syndrome and discuss the merits of current practice patterns.

## Materials and Methods

From January 2010 to December 2020 in Shanghai Xinhua Hospital and Hangzhou Children's Hospital, seven consecutive cases of gastrointestinal tract (GIT) trichobezoars were reviewed after taking permission from the Hospital Institutional Review Board. Demographic data, elements of presentation, imaging, surgical treatment, and subsequent management were reviewed.

## Results

There were seven female patients and no male patients identified. The median age at the time of presentation was 11 years old (range 4-13 years old). Table 1 summarises demographic information and characteristics. Stomach pain, abdominal distension, and vomiting were all common presenting symptoms in our study (Table 1). Five patients (71%) presented with acute symptoms as a result of complications (intestinal obstruction in 4 patients and intussusception in 1). A history of trichophagia was mentioned in 5 (71%) of the patients. A palpable abdominal mass was the most prevalent physical examination finding. Only two individuals reported with bilious vomiting and intestinal blockage on radiographs. An abdominal scan revealed that one of the patients had intussusception.

When the patients presented to the emergency department, the workup for the etiology of their problems was different. Table 2 summarises the results of the preoperative evaluation. For the most part, multiple imaging modalities were necessary to accurately diagnose the bezoar. Despite the lack of solid confirmation via endoscopy, the majority of patients had a correct preoperative diagnosis of a trichobezoar.

**Table 1** Patient clinical data

| | **Patient population (n = 7)** |
|---|---|
| Female | 7 (100%) |
| Mean age | 9.3 y |
| Clinical manifestations | |
| Abdominal pain and vomiting | 5 (71%) |
| Abdominal distension | 2 (29%) |
| Chronic GI symptoms | 5 (71%) |
| Small bowel obstruction | 2(29%) |
| Peritonitis | 3 (43%) |
| Palpable abdominal mass | 3 (43%) |
| Trichophagia | 6 (86%) |
| Trichotillomania | 3 (43%) |

**Table 2** Preoperative evaluation

| | **Patient population (n = 7)** |
|---|---|
| Preoperative diagnosis of trichobezoar | 5 (71%) |
| Abdominal radiograph | 3 (43%) |
| Abdominal ultrasound | 3 (43%) |
| Abdominal computed tomography scan | 2 (29%) |
| Upper GIT endoscopy | 4 (57%) |

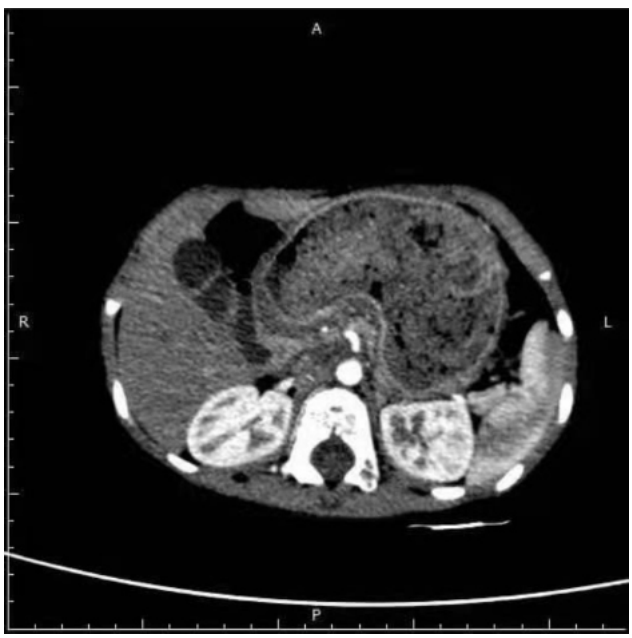


**Figure 1** Abdominal CT scan abdomen showing intragastric bezoar.

Following is a summary of the patients' perioperative care. A diagnostic endoscopy was performed on one patient who had been diagnosed with small intestinal obstruction in order to ascertain the cause. After detecting two hard lumps in the jejunum that looked like trichobezoars, the treatment was changed to a laparotomy. History, examinations, or endoscopy were used to make a preoperative diagnosis of trichobezoar in 5 individuals (71%). The patients who remained were diagnosed during surgery. After clinical optimisation, all of the patients received open surgery, with the exception of six who were operated on as an emergency. Endoscopic retrieval was attempted in three individuals, however none of them were successful. During the operation, one patient developed jejuno-jejunal intussusception while swallowing magnetic beads (Figures 3). Our operational findings, surgical techniques, and patient outcomes are detailed in Table 3. All of the patients had no complications. The average length of stay was 12 days, with a range of 10 to 18 days. Following surgery, the patients were discharged and referred to the psychiatric department. Five patients who were recently managed are being followed up on, with the rest of the patients being lost to follow-up.

## Discussion

Rapunzel syndrome is an uncommon form of trichobezoar in which long hair loops from the stomach protrude into the small intestine.[1] It is especially common in young females who have trichotillomania, a concurrent psychological condition that is typically untreated.

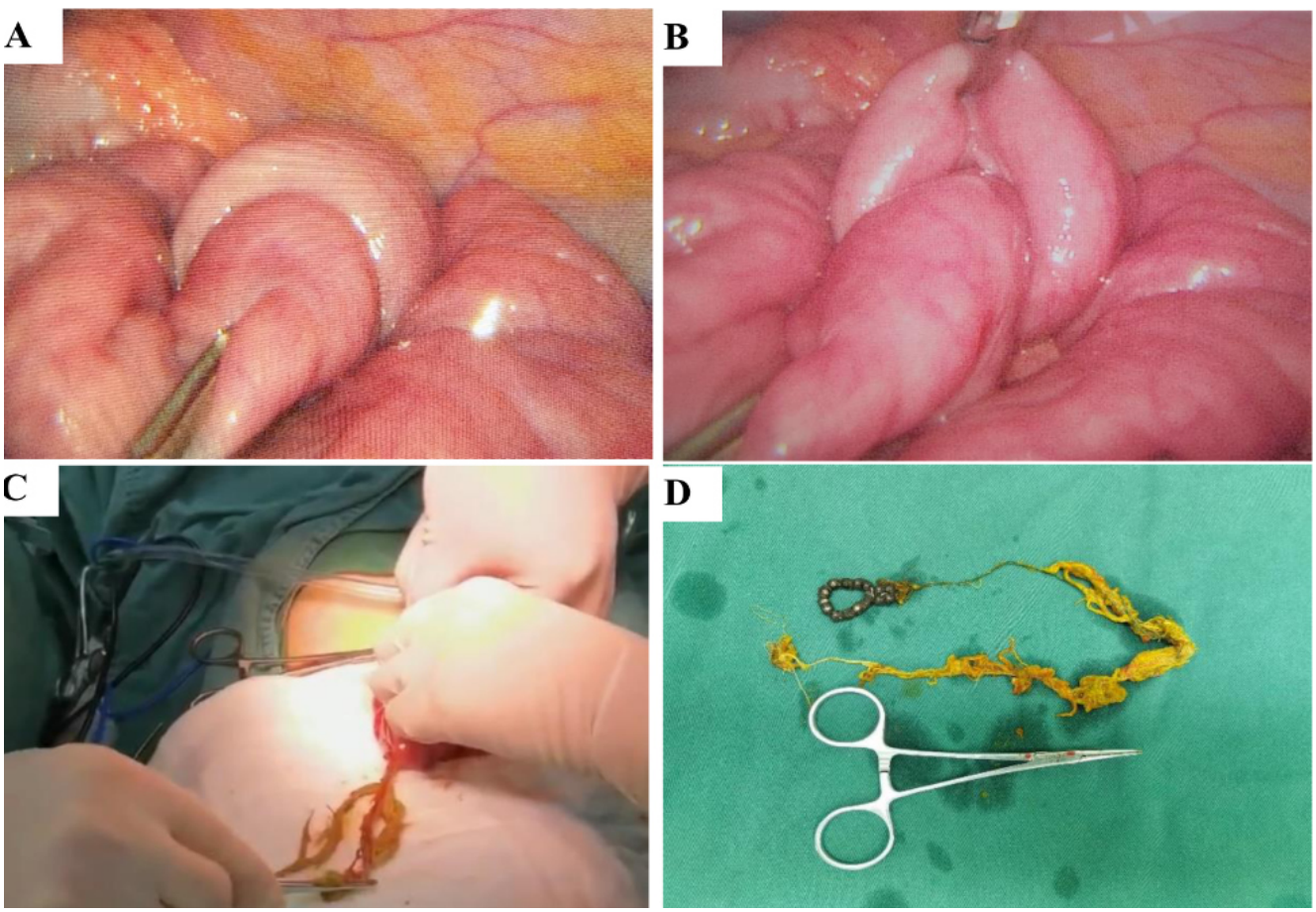


**Figure 2** (A, B) Laparoscopy finding of a jejuno-jejunal intussusception, with attempted laparoscopic removal failed. (C) Enterotomy performed for removal of the intestinal trichobezoar concretion after reduction of jejuno-jejunal intussusception. (D) Complete trichobezoars and magnetic beads were removed, consisted of: (1) sixteen magnetic beads were found in the proximal jejunum, (2) a small intestinal trichobezoar.

Stomach trichobezoar can cause bleeding, perforation, and obstruction of the gastric outlet. Rapunzel syndrome commonly manifests later in life, after years of absorption of enormous volumes of hairs. Trichobezoar is most commonly found in the stomach, although it can also spread to the duodenum, resulting in the Rapunzel syndrome.

Rapunzel syndrome is frequently diagnosed late due to a low threshold of suspicion and the fact that individuals may go years without symptoms until the bezoar has grown large enough to cause intestinal obstruction. Depending on the degree of obstruction, the patient may have a palpable lump in the belly, abdominal discomfort, nausea, vomiting, weakness, and constipation. In the gastrointestinal tract, the bezoar can create an obstruction or a gastric ulcer. The telescoping of the proximal jejunum into the distal jejunum, which leads to intussusception, may have started with the extension and migration of the trichobezoar tail into the jejunum.[9] However, the lack of comorbidities and a complete medical history may make a preoperative diagnosis impossible. Furthermore, the majority of parents did not mention a history of hair consumption. Radiological examinations may confirm the preoperative diagnosis. In the context of a patient with trichophagia and a palpable mass, plain radiographs frequently show a stomach shaped partly opacified area. CT scan is a superior examination because it not only detects but also defines the extent of a heterogeneous bezoar.[10,11] Upper GI endoscopy is the preferred diagnostic method, and it can also be used to treat minor bezoars.[12]

Endoscopic retrieval was attempted in four children in our study, but it was unsuccessful. To yet, the success rate of endoscopic retrieval is not encouraging. Laparotomy was used to successfully treat all of our patients. In a recent analysis of all case reports involving bezoars, 100 (92.5%) of 108 patients were treated with a laparotomy, with a

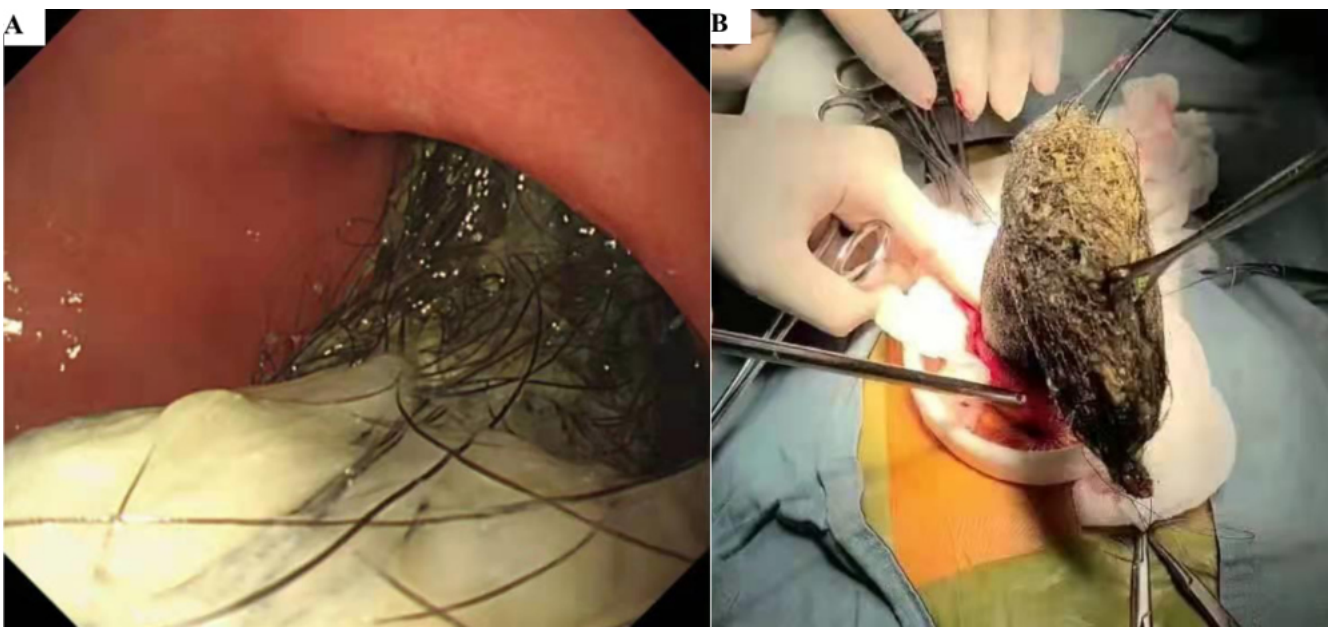


**Figure 3** (A) Endoscopic finding of a trichobezoar extended beyond the pylorus, with attempted endoscopic removal failed. (B) A large gastric bezoar with duodenal extension measuring 10.5 × 10.5 × 8.5 cm as it is delivered through the gastrotomy.

**Table 3** Operative findings, surgical procedure and outcome

| Patients | Surgery performed | Dimensions in bezoar | Outcome |
|---|---|---|---|
| Case 1 | Gastrotomy and removal | 15*8*6 cm | Alive |
| Case 2 | Gastrotomy and removal | NR | Alive |
| Case 3 | Enterotomy, Gastrotomy and removal | 8*4*4 cm | Alive |
| Case 4 | Gastrotomy and removal | 17*13*10 cm | Alive |
| Case 5 | Gastrotomy and removal | 10*6*6 cm | Alive |
| Case 6 | Enterotomy, Gastrotomy and removal | 16*8*6 cm | Alive |
| Case 7 | Enterotomy and removal | 5*4*4 cm | Alive |

success rate of 99 percent and a complication rate of 12 percent.[13] Our findings are comparable, with a success rate of 100 percent and a complication rate of 0 percent. The average length of stay in the hospital was around one week, and all of the patients had good outcomes. One patient with a preoperative diagnosis of small bowel blockage was treated with laparoscopy at first, but when the jejuno-jejunal intussusception was discovered, the surgery was changed to an open procedure. Some authors were against the laparoscopic method because of the increased risk of bezoar contents spilling into the abdomen and the high conversion rates.[13]

Because the impulsive behaviour associated with trichophagia in these patients is difficult to manage, and the risk of recurrence is considerable, long-term mental treatment is essential, as well as follow-up endoscopic or contrast testing.[14] Psychiatric care was sought by 71% of our patients. Every patient should get a psychiatric evaluation as part of their therapy.[13] A randomised control trial investigated the efficacy of behavioural therapy compared with minimal attention control on patients with trichotillomania found that behavioural therapy was a superior treatment strategy for both management of symptoms and durability of treatment gains.[15] Furthermore, A recent randomised control trial that compared habit reversal training (HRT) with treatment-as-usual (TAU) revealed that trichotillomania was largely improved with HRT compared to TAU.[16] No one in our study experienced a recurrence. Recurrence that necessitated reoperation was uncommon, and it was largely owing to a lack of competent psychiatric follow-up.[17]

In conclusion, the diagnosis of Rapunzel syndrome requires a high index of clinical suspicion, which can be established with a thorough history combined with radiography. Early diagnosis and prompt surgical intervention after rapid clinical optimisation help ensure a good prognosis. Psychiatric evaluation and management are key to prevent further trichophagia and trichobezoars.

## Statement of Ethics

All procedures performed in studies involving human participants were in accordance with the ethical standards of the Hangzhou Children's Hospital ethical review committee and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient's parents for publication of this article and any accompanying images.

## Disclosure Statement

The authors declare no conflict of interests.

## Funding Sources

The study was supported by grant from the Medical health Science and Technology project of Zhejiang Province (2022492938).

## References

1. Phillips MR, Zaheer S, Drugas GT. Gastric trichobezoar: case report and literature review. Mayo Clin Proc 1998;73:653-6.
2. Zent RM, Cothren CC, Moore EE. Gastric trichobezoar and rapunzal syndrome. J Am Coll Surg 2004;199:990.
3. Wadlington WB, Rose M, Holocomb GW Jr. Complications of trichobezoars: a 30-year experience. South Med J 1992;85:1020-2.
4. Klipfel AA, Kessler E, Schein M. Rapunzal syndrome causing gastric emphysema and small bowel obstruction. Surgery 2003;133:120-1.
5. Hoover K, Piotrowski J. St Pierre K, Katz A, Goldstein AM. Simultaneous gastric and small intestinal trichobezoars–a hairy problem. J Pediatr Surg 2006;41:1495-7.
6. Alsafwah S, Alzein M. Small bowel obstruction due to trichobezoar: role of upper endoscopy in diagnosis. Gastrointest Endosc 2000;52:784-6.
7. Dorn HF, Gillick JL, Stringel G. Laparoscopic intragastric removal of giant trichobezoar. JSLS 2010;14:259-62.
8. Fraser JD, Leys CM. St Peter SD. Laparoscopic removal of a gastric trichobezoar in a pediatric patient. J Laparoendosc Adv Surg Tech A 2009;19:835-7.
9. Naik S, Gupta V, Naik S, et al. Rapunzel syndrome reviewed and redefined. Dig Surg 2007;24:157-61.
10. El Hajjam M, Lakhloufi A, Bouzidi A, Kadiri R. CT features of a voluminous gastric trichobezoar. Eur J Pediatr Surg 2001;11:131-2.
11. Naran AD, Naran D, Haller JO. CT findings of small bowel trichobezoar. Emerg Radiol 2002;9:93-5.
12. Benatta MA. Endoscopic retrieval of gastric trichobezoar after fragmentation with electrocautery using polypectomy snare and argon plasma coagulation in a pediatric patient. Gastroenterol Rep (Oxf) 2015;41:1371-5.
13. Gorter RR, Kneepkens CMF, Mattens ECJL, Aronson DC, Heij HA. Management of trichobezoar: case report and literature review. Pediatr Surg Int 2010;26:457-63.
14. Tiwary SK, Kumar S, Khanna R, Khanna AK. Recurrent Rapunzel Syndrome. Singap Med J 2011;52:128-30.
15. Franklin ME, Edson AL, Ledley DA, Cahill SP. Behavior therapy for pediatric trichotillomania: a randomized controlled trial. J Am Acad Child Adolesc Psychiatry 2011;50:763-71.
16. Rahman O, McGuire J, Storch EA, Lewin AB. Preliminary randomized controlled trial of habit reversal training for treatment of hair pulling in youth. J Child Adolesc Psychopharmacol 2017;27:132-9.
17. Pegues JN, Lucas ED, Morris M. Recurrent Rapunzel Syndrome in an Adolescent Female. Am Surg 2023;31348231173942.