

# Rapunzel Syndrome Revealed by an Umbilical Hernia: A Case Report and Review of the Literature

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## ABSTRACT

**Introduction:** Rapunzel syndrome is a rare entity defined by the presence of a trichobezoar that can sit throughout the digestive tract. It results in psychiatric disorders, trichotillomania and trichophagia. Hernial strangulation is an exceptional mode of revelation of this pathology.

We report a case of Rapunzel syndrome revealed by a recurrent strangulated umbilical hernia.

**Case report:** This was a 22-year-old female patient with a history of umbilical hernia repair 6 months ago, admitted to the emergency department of Idrissa Pouye General Hospital for painful umbilical swelling. Examination on admission revealed a strangulated umbilical hernia with firm contents and an inferior umbilical scar. Biological work-up was unremarkable. Surgical exploration via an inferior umbilical approach revealed a strangulated umbilical hernia with a 3 cm neck, containing a viable small intestine, and the presence of a 50 cm long inert foreign body in the terminal ileum, enclosed at the ileocaecal angle. We performed a 1 cm enterotomy to extract the foreign body, which was made into a trichobezoar, then closed the enterotomy, treated the sac and cured the hernia using aponeurography. Postoperative recovery was straightforward. She was discharged on the third postoperative day. She was referred to the psychiatric ward for psychiatric and psychological follow-up.

**Conclusion:** Rapunzel syndrome is a rare condition. Its mode of revelation by a strangulated umbilical hernia is very rare, even exceptional. The evolution can be marked by serious mechanical complications such as intestinal occlusion.

## Keywords:

Rapunzel syndrome, Strangulated umbilical hernia, Surgery, Trichobezoar, Pseudo-occlusive syndrome.

## Introduction

Rapunzel syndrome is a rare entity defined by the presence of a trichobezoar that can sit all over the digestive tract. It results from psychiatric disorders, trichotillomania and trichophagia [1]. It was first described in 1968 by Vaughan. It takes its name from a Grimm Brothers fairy tale [2]. It occurs most often in young girls with psychiatric disorders [1,3]. Hernial strangulation is an exceptional mode of revelation.

The aim of this study was to report a case of Rapunzel syndrome revealed by a recurrent strangulated umbilical hernia.

## Observation

A 22-year-old female patient with a history of umbilical hernia repair was admitted to the emergency department of Dakar's Idrissa Pouye General Hospital with painful umbilical swelling. Questioning revealed a history of trichophagia. General examination revealed good general condition, with coloured, anicteric mucous membranes. There was no dehydration or malnutrition. Her blood pressure was 120/80mmHg, temperature

37.4°C, heart rate 83 beats/min, respiratory rate 23 cycles/min and saturation 95% on room air. Physical examination revealed an umbilical scar and the presence of a painful, irreducible umbilical swelling suggestive of recurrence of a strangulated umbilical hernia with firm contents. The other hernial orifices were free, and the rectal exam was normal. The extended examination of other devices and systems revealed no abnormalities. The blood count showed a natraemia of 132mg/l, a kalaemia of 3.5mmol/l and a chloraemia of 105mmol/l. Hemoglobin was 11g/dl, creatinine 12mg/dl. Surgical exploration was indicated. The patient underwent emergency surgery under general anaesthesia via an arciform incision in the lower umbilicus, using the old scar.

Surgical exploration revealed a 3 cm snare. The hernia sac contained a viable small loop with an inert foreign body lying in the terminal ileum for 50 cm, enclosed at the ileocaecal angle. We performed the following procedures:

- A 2 cm-long enterotomy on the anti-mesenteric border opposite the mass (Figure 1), to extract the foreign body made of a mass of hair (Figure 2).
- The enterotomy was closed with 4 simple stitches of absorbable suture, Polyglactine 910 USP 3/0 (vicryl® 3/0).

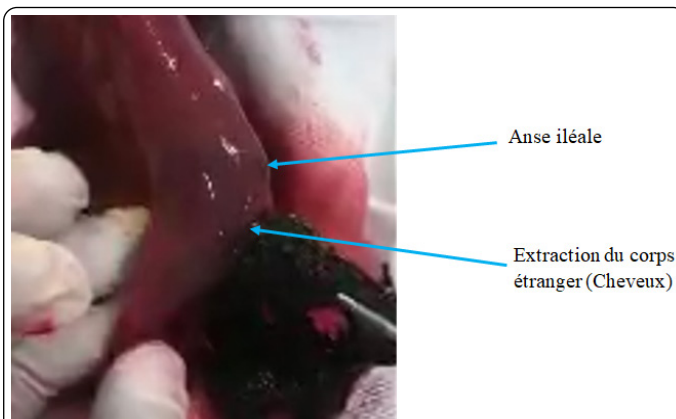


Figure 1: Surgical view of trichobezoar extraction after enterotomy.



Figure 2: Trichobezoard after extraction.

- Fasciotomy hernia repair with "X" stitches using non-absorbable Polyester USP 2 (Mersuture® 2) thread.

Postoperative recovery was straightforward, and the patient was discharged on the 3rd postoperative day. She was referred to the psychiatric department for psychiatric and psychological follow-up.

After 12 months, the patient had no recurrence of umbilical hernia or trichophagia after cognitive-behavioural therapy.

## Discussion

A bezoar is an agglomeration of food or foreign matter that accumulates and remains in the gastrointestinal tract. Its frequency is estimated at 0.15% of gastrointestinal foreign bodies [1]. There are 6 types of bezoar: trichobezoar, phytobezoar, lactobezoar, lithobezoar, pharmacobezoar and plasticobezoar

[5]. It is a pathology of young girls, with an average age of under 20. However, two cases have been reported in males, and one in a 37-year-old woman [1,6,7]. It presents as a pseudo-occlusive syndrome [8]. Clinical examination may reveal alopecia and an epigastric abdominal mass [9]. Biological tests may show anemia and hypoalbuminemia [10]. Our patient was admitted with a strangulated umbilical hernia and her biology was unremarkable. As the bezoar increases in size and progresses, its evolution can be marked by mainly mechanical complications, including intermittent small bowel obstruction, duodeno-jejunal fissuring, pancreatico-biliary obstruction, peritonitis or even pancreatitis [11,12].

There are 6 types of bezoar: trichobezoar, phytobezoar, lactobezoar, lithobezoar, pharmacobezoar and plasticobezoar [5]. It is a pathology of young girls, with an average age of under 20. However, two cases have been reported in males, and one in a 37-year-old woman [1,6,7]. It presents as a pseudo-occlusive syndrome [8]. Clinical examination may reveal alopecia and an epigastric abdominal mass. Biological [9] tests may show anemia and hypoalbuminemia [10]. Our patient's case appears to be the first description of a Rapunzel syndrome revealed by hernial strangulation in our practice setting. Recurrence of the umbilical hernia and strangulation could be explained by hyperperistalsis secondary to obstruction of the ileocolic valve by the trichobezoar. Computed tomography with injection of the contractile product is the reference examination for diagnosis. It shows a well-defined oval intra-luminal mass with air bubbles trapped in the interstice, or a homogeneous marbled appearance in the region of the stomach or intestine. Endoscopy allows direct visualization of the bezoar in the stomach [13].

In the context of strangulated umbilical hernia, we have not performed a morphological examination. The indication for emergency surgery was immediately established. Treatment has a dual objective: extraction of the bezoar and prevention of recurrence. The method of extraction depends on the size and location of the mass [14]. Endoscopy allows extraction of small masses, laparoscopic surgery for medium-sized masses and open surgery for large masses, notably in Rapunzel syndrome [15,16]. Other therapeutic means are currently being evaluated, including enzymatic dissolution, the Nd-YaG laser and extracorporeal wave fragmentation [14]. We performed an enterotomy extraction following laparotomy. In the largest review of trichobezoar management, 100 of 108 patients (92.5%) were treated by laparotomy, with only 12% complications, making it the treatment of choice for trichobezoar, particularly in Rapunzel syndrome [17]. Prevention relies on psychiatric follow-up with psychotherapy, cognitive-behavioural therapy and selective serotonin reuptake inhibitors [18]. We referred the patient to a psychiatric service for follow-up. Our patient's trichophagia has stopped, with a satisfactory outcome.

## Conclusion

Rapunzel syndrome is a rare disorder affecting children and young girls. Its evolution can be marked by serious mechanical complications such as intestinal obstruction. Its mode of manifestation in the form of a strangulated umbilical hernia is very rare, if not exceptional. Hence the interest of this case report. Prevention of Rapunzel syndrome and its complications requires psychiatric and psychological follow-up.

## Conflict of Interest

None.

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