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Case Report

# Rapunzel Syndrome: An Unusual Onset Case Report

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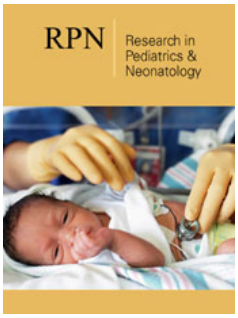


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## Abstract

Bezoar is an accumulation of organic substances in the gastrointestinal tract. This condition is extremely rare, their incidence in the pediatric population is unknown. Bezoars usually occur in females, adolescents, and children with psychiatric or neurological disorders. Trichobezoar refers to the accumulation of hair usually in the stomach and even beyond it (Rapunzel syndrome). We present the clinical case of a school-age female patient who started with neurological symptoms (headache, syncope) and then developed gastrointestinal symptoms. Only later was clarified a history of bullying at school and consequent anxious behaviors (trichotillomania and trichophagia). It's very important a thorough medical history, especially in patients with Trichobezoar, to highlight psychological disorders that can cause trichotillomania and trichophagia.

**Keywords:** Trichobezoar; Trichotillomania; Trichofagia; Laparoscopy

## Introduction

Bezoar is an accumulation of exogenous matter in the stomach or intestine. Rapunzel syndrome refers to a trichobezoar that extends from the stomach into the entire small bowel [1]. Bezoars are classified on the basis of their composition [2]. Trichobezoars are composed of the patient's own hair. Phytobezoars are composed of a combination of plant and animal material. Trichophytobezoars are mix composition. It's defined Orthobezoar when mineral salt are deposited on a nucleous of animal or vegetable. Lactobezoars were previously found most often in premature infants and may be attributed to the high casein or calcium content of some premature formulas. Shallowed chewing gum may occasionally lead to bezoars. This condition is extremely rare, their incidence in the pediatric population is unknown [3]. Most bezoars have been found in females with underlying personality problems or neurologically impaired individuals. About 70% of patients with trichobezoar intestinal obstructive syndrome are women under age 20 years [4]. Bezoars can be fatal because they can cause gastric and bowel perforation, peritonitis, bleeding, or ischemic to necrotic changes of the gut [5]. Patients diagnosed with a bezoar usually are asymptomatic for many years [6]. Some bezoars are found incidentally with imaging, where they appear as masses or filling defects on abdominal radiographs and computed tomography (CT) scans. Therefore, a thorough medical history is important, especially in patients with Trichobezoar, to highlight psychological disorders that can cause trichotillomania and trichophagia. In fact, about 6% of patients with Tricotillomania develop a trichobezoar [5]. Diagnosis can be established by an upper endoscopy with extraction sampling to determine the composition or extract the whole bezoar in fragments if it is only in the stomach and/or proximal bowel. Although endoscopy is the preferred diagnostic study to differentiate bezoars, but only a few cases in the literature describe the complete endoscopic removal of a trichobezoar after mechanical or chemical fragmentation, and these cases refer exclusively to pediatric patients in whom presumably the total foreign body mass was easier to handle [7]. Conventional laparotomy is the treatment of choice for trichobezoar with satellite fragments beyond the stomach or extending into the small bowel as in Rapunzel syndrome [1-5].

## Case Report

We present the case of a trichobezoar intestinal obstructive syndrome in a 12-year-old girl. The girl was hospitalized for recurrent syncopal episodes, which started a few months ago, some of which with doubtful loss of consciousness. In addition, frequent headache episodes were reported for about 3 years, well responsive to paracetamol. The medical history did not highlight family pathologies, while the pathological history showed the presence of hepatic hemartoma, diagnosed at the age of 2, for which periodic ultrasound examinations are performed, and an episode of Schonlein-Henoch purpura at the age 5 years old.

Physical examination revealed a palpable tumor within the epigastrium, initially interpreted as referable to hepatic hemartoma, for which we planned a control abdominal ultrasound. During the hospitalization, several tests were performed including blood biochemical examination, routine electrocardiogram, dynamic electrocardiogram, echocardiography, vestibular examination, eye examination, electroencephalogram, head magnetic resonance imaging (RMI), excluding the organic disease of nervous system and circulatory system. All tests were substantially normal. Subsequently, in light of all this, psychological consultation was also performed suspecting psychological involvement, which detected a condition of psychological distress due to anxiety disorder, for which child neuropsychiatric consultation was planned.

In the meantime, a few days after admission, another abdomen ultrasound was performed which showed a mass in the stomach that was not well interpretable, for which a CT of the abdomen was performed which showed the presence of solid tissue in the epigastric area not well defined, extending for about 6 cm, with no evident cleavage plane towards the head of the pancreas, the gastric antrum (apparently infiltrated) and the duodenal C. For further study, MRI of the abdomen was performed which interpreted the abdominal lesion as likely pancreatic nature (annular pancreas?). In the meantime, the symptoms had changed as the girl began to present abdominal pain and vomiting and appeared particularly dejected. Apart from hypochromic and microcytic anemia, laboratory test results were unremarkable. Therefore a new ultrasound evaluation was carried out which this time highlighted, in the context of the gastric lumen, the presence of a solid mass, extended for about 13cm from the gastric body to the duodenum. The subsequent barium abdominal X-ray raised the suspicion of trichobezoar, later confirmed by upper gastrointestinal tract endoscopy which showed a large trichobezoar occluding the stomach from the fundus to the antrum with extension into the duodenum and a peptic ulcers was detected in the area of Vater's papilla underneath the attachment base of the hair mass as a result of pressure necrosis (Figure 1). Repeated attempts at fragmentation and extraction of the mass have been made, but without success, for which a gastrostomy and removal of the trichobezoar were used (Figures 2 & 3). The post-surgery course was regular, without complications. Subsequently, as planned, child neuropsychiatric consultation was performed, from which a condition of trichotillomania emerged for about 2 years, probably linked to bullying at school and an anxious state. Currently, the girl is in good health in the absence of symptoms referable to relapse and is followed up by the neuropsychiatrist.

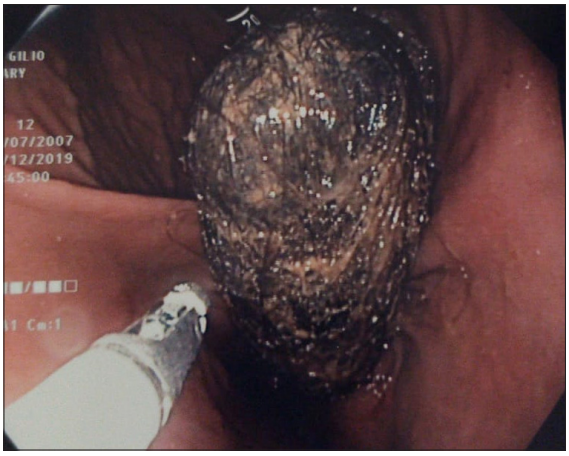

**Figure 1:** Endoscopic view of the trichobezoar.

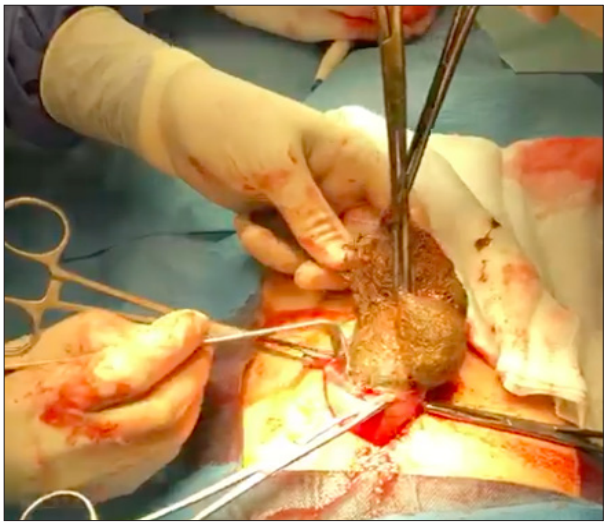

**Figure 2:** Surgical breach with the trichobezoar.

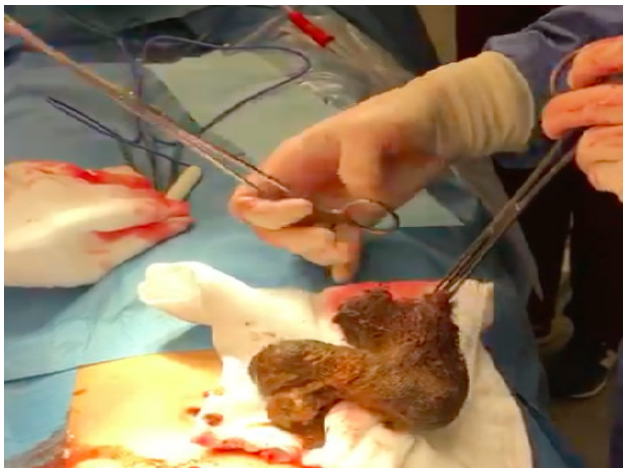

**Figure 3:** Intraoperative view of the trichobezoar.

## Discussion

Bezoars are masses of partially or totally indigestible material, that is, unassailable by gastrointestinal secretions, which generally gather in the stomach and can cause various dyspeptic disorders up to gastrointestinal occlusion. Trichobezoars are clusters of undigested hair, mucus and fats. They are responsible for Rapunzel Syndrome, named after the character created by the brothers Grimm's imagination, a rare medical condition in which the hair, which people ingest, forms a tangled mass that remains trapped in the stomach and extends to the small intestine. A trichobezoar occurs mainly in association with a psychiatric disorder affecting usually young women, having the tendency of pulling out their own hair (trichotillomania) and eating it (trichophagia). Affected people are often asymptomatic. Symptoms and signs are related to the mass that progressively occupies the space of the gastric lumen and its obstacle to gastric function. Most common symptoms are abdominal pain, vomiting, and gastrointestinal bleeding from asymptomatic anemia to hematemesis [8]. Moreover, patients can complain early satiety, obstruction, peritonitis, intussusception and weight loss [9]. A few cases present with obstructive jaundice [10], pancreatitis [11,12], appendicitis [13] and gut perforation [14,15]. Abdominal examination typically reveals an upper abdominal mass (Lamerton's sign) [9].

The Gold standard of diagnosis of trichobezoars is upper gastrointestinal endoscopy [16]. Others modalities are of additional help. The upper abdomen CT is reported to be more effective than ultrasound and barium meal in revealing concomitant gastric and intestinal bezoars, and it also has proven accuracy in determining the level and degree of intestinal obstruction [17]. However, upper endoscopy is the most sensitive examination for diagnosis and also plays a fundamental therapeutic role. Treatment of trichobezoars aims to completely remove the mass and prevent recurrence. Laparotomy is still the treatment of choice for large trichobezoars because of its advantages which include the simplicity of the procedure, less time required and feasibility to examination all the bowel for satellite lesions [18]. However, laparoscopic intervention seems to be gaining ground over open surgical operation and there are several cases reporting the laparoscopic removal of extremely long and large trichobezoars [19]. Recurrence has been reported owing to improperly treated psychiatric conditions and missed follow-up [20].

## Conclusion

The diagnosis of trichobezoar is easy after detailed enquiry of medical history, paying close attention to the risk factors (trichotillomania, trichophagia, alopecia). In our case report the initial diagnosis was oriented towards a neurological pathology (headache, syncope) and only later did neuropsychiatric disorders emerge. Therefore, it is very important to pay attention to the psychological aspects, to avoid diagnostic delays or misdiagnoses. There are no standardized techniques for treatment. In the case of small sized trichobezoars, endoscopic treatment may be useful, on the contrary in the case of large, non-fragmentable or molded trichibezoars, the surgical approach is the treatment of choice.

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