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# Upper gastrointestinal bleeding, an unusual presentation of Rapunzel syndrome

Carlos Torres-Salinas[1], David Anccasi-Hermoza[2]

*Gastrointestinal bezoars in the pediatric age group have an unusual presentation, and their clinical manifestation is mainly associated with obstructive conditions of the gastrointestinal tract, and are generally made up of hair and food debris. In a smaller number, it has been seen that from its usual location, which is the stomach, bezoar structures could extend into the duodenum, jejunum, ileum, and even colon, giving the appearance of a "Rapunzel tail".*

*We present the case of a 7-year-old girl with an atypical initial presentation of Rapunzel syndrome, secondary to a trichobezoar. The patient did not have previous psychiatric history, nor reports of any disturbance at school. She was admitted to the emergency room due to upper gastrointestinal bleeding and required a subsequent transfusion of blood products. During her hospitalization, an upper digestive endoscopy was performed, with the diagnosis of Rapunzel syndrome, in addition to the presence of two Forrest II-C, III gastric ulcers and severe erythematous gastritis in the region of the antrum and body. Medical treatment was started without success, for which the bezoar had to be surgically removed.*

*Therefore, we want to point out the importance of deepening certain aspects such as the medical history and background in this type of patient, since it is a disorder linked to behavioral and mood alterations; likewise, remember the less frequent clinical symptoms such as upper gastrointestinal bleeding, but which in turn can be lethal if we don't act promptly. In the same way, the comprehensive approach with the support of the family and the intervention of psychology and/or psychiatry is vital to prevent recurrences.*

**Key words:** BEZOARS; GASTROINTESTINAL HEMORRHAGE; ANXIETY

## INTRODUCTION

Gastrointestinal bezoars are indigestible aggregates that were previously ingested, the trichobezoar is within the classification of these, its composition is human hair and food remains. Sometimes it can be made up of synthetic doll hair, these being very dangerous due to their potential flammable reaction with electrocautery and argon plasma coagulation from endoscopes. They are most often located in the stomach and rarely extend to the duodenum, jejunum, or ileum. and even the colon, a condition then called Rapunzel syndrome (1, 2).

It is a rare entity, with incidences ranging from 0.068% - 0.43% (3), and it is usually associated with such as diffuse abdominal pain, nausea, vomiting, poor weight gain, obstructive jaundice, protein-losing enteropathy, abdominal mass, and usually associated psychiatric symptoms (4, 5). We present the case of a 7-year-old patient with upper gastrointestinal bleeding as disease onset, secondary to trichobezoar.

## CASE REPORT

Female patient of 7 years 8 months with weight: 22.5 kg, Body Mass Index/Age: 0.72 Standard Deviation. Family

[1] School of Medicine, Continental University, Peru
[2] Departament of Pediatrics, Ramiro Prialé Prialé Hospital, Peru

**Correspondence to:**
Carlos Torres-Salinas; Departament of Pediatrics; Ramiro Prialé Prialé Hospital; Av. Independence N° 266; Postal address No. 12006

FIGURE 1. **Trichobezoar with "Rapunzel tail" going towards the duodenum, in addition to Forrest II-C, III gastric ulcers, and severe antrocorporal erythematous gastritis.**

member reports that the patient pulls out her hair and compulsively ingests it for two years, denies prior hospitalizations and there is no history of psychiatric evaluation. The mother denied that her daughter had presented previous symptoms of abdominal pain, early satiety, or apparent weight loss.

She entered the emergency room with a 7-hour illness, characterized by diffuse colicky abdominal pain, intensity 7/10. Initially, it was accompanied by two vomits with traces of blood, followed by four episodes of hematemesis with an approximate volume of 150 ml in total; and did not show symptoms of intestinal obstruction, heart rate: 105 bpm, respiratory rate: 30 bpm, blood pressure: 100/70 mmHg. At physical examination there was a moderate abdominal distention with a palpable upper abdominal mass. In the auxiliary tests, hemoglobin stands out at 10.3 g/dl (Normal value: 12-14 g/dl), with normal corpuscular constants.

She was hospitalized with a diagnosis of upper gastrointestinal bleeding and probable acute gastropathy of etiology to be determined. During hospitalization, she received a transfusion of an immune globular package on two occasions and did not present any event of hematemesis. As part of the work plan, an upper digestive endoscopy procedure is scheduled for diagnostic-therapeutic purposes.

Thus, during the endoscopic procedure, a mass made up of hairs that occupy 70% of the gastric chamber (Figure 1) is visualized, which emits a "Rapunzel tail" prolongation towards the duodenum. Similarly, 2 Forrest II-C and III gastric ulcers, respectively, and severe intracorporal erythematous gastritis are visualized.

FIGURE 2. **Surgical approach by gastrotomy with trichobezoar extraction. The yellowish tuft is the classic "Rapunzel tail" image and indicates the end of the trichobezoar.**

Endoscopic removal of trichobezoar was attempted without success, and for this reason, it was decided to start medical management with proton pump inhibitors and instillation by gastroschisis of Coca-Cola ® during the following 24 hours without improvement of the condition, on the contrary, the patient presented abdominal pain type colic and nausea, deciding to suspend the instillation.

She was managed with proton pump inhibitors, a wide liquid diet alternated with a soft diet, and on the third day of hospitalization, a medical meeting was held, to decide on surgical management (gastrotomy and removal of trichobezoar) (Figure 2).

After surgery, the patient progressed favorably, being discharged 17 days after surgery, with outpatient follow-up by pediatrician, pediatric surgeon, and psychiatrist who evaluated the patient, concluding: behavioral emotional disorder and mild intellectual disability.

## DISCUSSION

The denomination comes from the Arabic "badzehr" or from the Persian "panzehr" which means antidote, the same one that within the history of medicine was used for this purpose, but without success. Currently, the term "bezoar" is not used to refer to a failed antidote, but to a potentially serious medical problem that requires timely diagnosis and appropriate therapy (5).

For a trichobezoar to form, the habit of pulling out hair (trichotillomania) is first required, 30% of them ingest the hair (trichotillophagia), and if gastric peristalsis is poor, a greater accumulation of hair in the stomach will be appreciated. The same will generate symptoms such as abdominal pain, vomiting, weight loss, halitosis, obstructive jaundice,

enteropathy due to protein loss, and in some cases a palpable abdominal mass (6, 7).

The trichobezoar belongs together with the phytobezoar, pharmacobezoar, lactobezoar, and other rarer ones, to the bezoars described in humans and they are differentiated according to their composition; specifically, the trichobezoar, due to its enzyme-resistant properties, its smooth and slippery surface cannot be digested and can remain stagnant in the gastrointestinal system. In some cases, the hairball extends from the stomach to the small intestine and colon, being called Rapunzel syndrome, as in the present case (8).

Concerning its association with upper gastrointestinal bleeding and considering its notable infrequency, the literature does not show clear explanations in this regard. For this reason, we postulate that this may be due to a secondary effect derived from the proteolytic enzymes produced in the stomach, the same ones that in their eagerness to degrade the trichobezoar end up damaging the gastric mucosa and in their evolution would cause gastric ulcerations. And in cases of giant trichobezoars, a valid hypothesis could be compromised gastric perfusion. However, this would not explain the focal lesions typical of gastric ulcers.

On the other hand, this entity is closely related to psychiatric disorders such as anxiety, depression, and obsessive-compulsive disorder. A remission of said behavior has been seen with both pharmacological therapy and behavioral therapy; the latter being the one with the best results (9). Likewise, when expanding the medical history, it was evidenced that there were previously conduct disorders, anxiety, and poor school performance.

Within the complementary examinations, upper digestive endoscopy is recommended, since it contributes to the diagnosis and removal of the bezoar can even be attempted, the use of computed tomography has been set aside given the irradiation of the pediatric patient and its limitation in terms of treatment (10, 11).

In the case presented, we resorted to the use of upper gastrointestinal endoscopy for another purpose, since the unusual presentation of the picture with upper gastrointestinal bleeding forced us to consider possible gastric ulcers as the basic cause. Thus, the presence of 2 Forrest II-C and Forrest III gastric ulcers was confirmed, as well as the presence of a mass of hairs that covered most of the gastric chamber and projected into the duodenum.

Regarding management, the use of Coca-Cola® is described, especially in phytobezoars, with a success rate of 50% if used alone, or up to 91.3% in combination with endoscopic management. However, its response to Coca Cola ® is poor against diopyrobezoars (60.6% vs. 23%) (12).

Likewise, the literature describes that endoscopic extraction of trichobezoar associated with Rapunzel syndrome have low success rates that reach up to 5%; Therefore, in these specific cases, it is recommended that they be removed surgically since they have a good response that ranges from 92.5% - 99% of cases resolved (13).

In our patient, we were able to observe that there was no response to medical management with the instillation of Coca-Cola, nor to the endoscopic one; having to resort to surgical management, which showed favorable evolution. The patient was followed up on an outpatient basis in the pediatric surgery, pediatrics, psychiatry, and nutrition clinics.

## CONCLUSION

Trichobezoar in the pediatric age shows a low incidence, it is worth noting its association with psychiatric disorders. For this reason, it is important to make an assessment not only of the medical condition but also of the social context and the family nucleus of the patient.

Its clinical presentation generally includes gastrointestinal symptoms that are sometimes non-specific and it is extremely unusual for them to start with upper gastrointestinal bleeding, as in the present case; but it is important to consider it as a diagnostic possibility if the child associates symptoms of trichotillomania and trichotillophagia.

There is no consensus on the best approach in terms of its management, especially in pediatric ages. However, we consider it prudent to assess the cases individually, and if necessary, opt to start with non-invasive therapy and if the condition does not resolve, resort to surgical management.

Likewise, once the urgency or emergency is resolved, as the case may be, it is essential to work together with the family and the support of other areas such as psychiatry and/or psychology.

Although there are few reports on the recurrence of this pathology, it is logical to think that having an altered behavioral basis, these patients should be followed up to treat the primary causes and thus avoid future episodes.

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SAŽETAK

# Gornje gastrointestinalno krvarenje, neuobičajena prezentacija Zlatokosinog sindroma

Carlos Torres-Salinas, David Anccasi-Hermoza

*U djece, gastrointestinalni bezoari su općenito sačinjeni od kose i ostataka hrane i imaju neuobičajenu kliničku prezentaciju sa simptomima uglavnom kao posljedica opstrukcije gornjeg gastrointestinalnog trakta. U manjem broju pacijenata bezoari se mogu protezati u dvanaesnik, jejunum, ileum pa čak i u debelo crijevo , dajući izgled „Zlatokosinog repa".*

*Prikazujemo slučaj 7-godišnje djevojčice s atipičnom inicijalnom prezentacijom Zlatokosinog sindroma, kao posljedica trihobezoara. Pacijentica nije imala anamnezu pozitivnu na psihijatrijske bolesti, niti prijave bilo kakvih smetnji u školi. Primljena je na hitnu pomoć zbog krvarenja iz gornjeg dijela probavnog sustava te joj je bila potrebna naknadna transfuzija krvnih pripravaka. Tijekom hospitalizacije učinjena je endoskopija gornjeg probavnog trakta s dijagnozom Zlatokosinog sindroma, uz postojanje dva Forrest II-C, III želučana ulkusa i teškog eritematoznog gastritisa u predjelu antruma i tijela. Započeto je konzervativno liječenje, koje je bilo bezuspješno, zbog čega je bezoar morao biti kirurški odstranjen. Stoga želimo ukazati na važnost produbljivanja određenih aspekata kao što su anamneza s naglaskom na psihijatrijsku pozadinu kod ove vrste pacijenata, budući da se radi o poremećaju povezanom s promjenama u ponašanju i raspoloženju. Isto tako, važno je imati na umu rjeđe simptoma kao što je krvarenje iz gornjeg dijela probavnog sustava, ali koji pak može biti smrtonosno ako ne djelujemo odmah. Na isti je način sveobuhvatan pristup uz podršku obitelji i intervenciju psihologa i/ili psihijatra od vitalnog značaja za sprječavanje recidiva.*

**Ključne riječi:** BEZOARI; GASTROINTESTINALNO KRVARENJE; ANKSIOZNOST