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**Case report**

# Rapunzel Syndrome in a Colombian Female Adolescent: A Case Study and Literature Review

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R E S U M E N

This article presents a case of Rapunzel syndrome (gastric trichobezoar with extension into the small intestine) in a female adolescent patient with mixed anxiety, depression, and obsessive-compulsive disorder. She suffered from trichotillomania secondary to family dysfunction and school bullying. At her arrival to the emergency department, she was experiencing abdominal pain, though no abdominal mass was palpated or seen on ultrasound, and her amylase and lipase levels were elevated. She developed an acute pancreatitis which required laparotomy. In addition to the case report, a review of the available literature on the subject is presented.

## Síndrome de Rapunzel en una adolescente colombiana: estudio de caso y revisión de tema

A B S T R A C T

En este artículo se presenta un caso de síndrome de Rapunzel (tricobezoar gástrico con extensión al intestino delgado) en una adolescente con trastorno mixto de ansiedad, depresión y trastorno obsesivo-compulsivo. La paciente sufría de tricotilomanía asociada a disfunción familiar y matoneo escolar. A su llegada al departamento de urgencias, la paciente experimentaba dolor abdominal, aunque no se palpó ni se visualizó por ecografía ninguna masa abdominal, y sus niveles de amilasa y lipasa eran elevados. Debutó con pancreatitis aguda, por lo que requirió laparotomía. En adición al reporte de caso, se presenta una revisión de la literatura disponible sobre el tema.

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## Introduction

In *Epidemia I*, Hippocrates already described the habit of pulling out hair as a disease. Later on, in the 18th century, Baudaman first described a trichobezoar in a 16-year-old male, but it was not until 1889 that Hallopeau introduced the term trichotillomania (TTM). Today, it is defined in the DSM-V (*Diagnostic and Statistical Manual of Mental Disorders. Fifth Edition.*) as recurrent hair pulling resulting in functional deterioration, without any other medical condition or mental disorder that can better explain it, and it has an estimated prevalence of one to two percent of the word population (1). Rapunzel syndrome is a form of trichobezoar extending into the intestine, formed from the ingestion of hair and associated with trichotillomania (1 y 2).

### *Case*

A healthy 14-year-old female consulted for a 16-day history of abdominal pain, vomiting and constipation. She had already consulted two weeks before to the emergency department. Back then surgical pathology was ruled out and she was discharged home. However, she was brought again to the emergency department for hyporexia and collapse. The patient also presented dehydration and soft abdomen with pain on epigastric palpation and no palpable abdominal mass or signs of peritoneal irritation.

She came from a single-parent family (dissolved by separation), with a history of school bullying and low academic performance, social isolation, selective appetite for liquids, sleep disturbances, and self-mutilation by wrist cutting. On admission, she had neutrophilia and thrombocytosis. A total abdominal ultrasound showed normal results, but amylase and lipases levels were elevated more than three times above the normal limit (Table 1).

An abdominal computerized tomography (CT) showed a trichobezoar extending into the small intestine (see Image 1). At the physical examination, she didn´t present alopecia, but she did have signs of onychophagia. Her eyebrows and eyelashes were normal.

The trichobezoar was extracted via laparotomy, and its extension into the duodenum and part of the jejunum was detached in its extraction (see Image 2). No gastric ulcer was seen.

On psychiatric examination, she was found to have normal attention and activity levels. She was laconic, with logical, coherent thinking, and little concern regarding her current illness. No body dysmorphic disorder was reported. She was egodystonic, without delusional thinking, ideas of self-aggression or aggression towards others, or active death or suicidal ideation. She manifested sadness with an anxious tone, no sensory alterations, a preserved sense of reality, partial introspection, adequate developing prospection, and no tics or stereotypical movements. She was diagnosed with mixed anxiety, depression, and obsessive-compulsive disorder (OCD), with TTM related to family dysfunction. There was no family history of obsessive-compulsive disorder or other psychiatric illnesses, nor any record of aggression or trichotillomania.

She began treatment with a Selective Serotonin Reuptake Inhibitor (SSRI), with persistent family conflicts and depression. She consulted again two months after discharge for exogenous intoxication with non-tricyclic antidepressants, metoclopramide, acetaminophen and diphenhydramine, requiring hospitalization on a mental health unit. The patient was discharged with outpatient pediatric follow-up and continued her schooling without further gestures or suicide attempts, receiving family support and psychotherapy.

**Table 1. Labs performed during hospitalization.**

| Labs | 05 May | 19 May | 21 May | 25 May | 27 May | 30 May | 01 Jun | 23 Jul | 25 Jul |
|---|---|---|---|---|---|---|---|---|---|
| Leu | 20.2 | 22.8 | 22.3 | 8.3 | 5.14 | 5.28 | | | 8.23 |
| Hgb | 15 | 15 | 13.5 | 13 | 12.5 | 11.3 | | | 13.3 |
| Hct | 43 | 43 | 38.7 | 37.8 | 36 | 33.7 | | | 40 |
| Plt | 466 | 509 | 473 | 411 | 271 | 201 | | | 371 |
| Amylase | 78 | 1384 | | 117 | | | | | |
| Lipase | | 7484 | | | | | | | |
| BUN | | 5.9 | 6 | 11.9 | | 9.5 | 9.1 | 10.2 | 6.5 |
| Albumin | | 4.3 | | | | | | | |
| Creatinine | | | 0.53 | 0.47 | | 0.6 | 0.5 | 0.47 | 0.61 |
| AST | | 14 | 17 | 18 | | | | 15 | 14 |
| ALT | | 15 | 12 | 20 | | | | 16 | 14 |
| GGT | | | | | | | | | 24 |

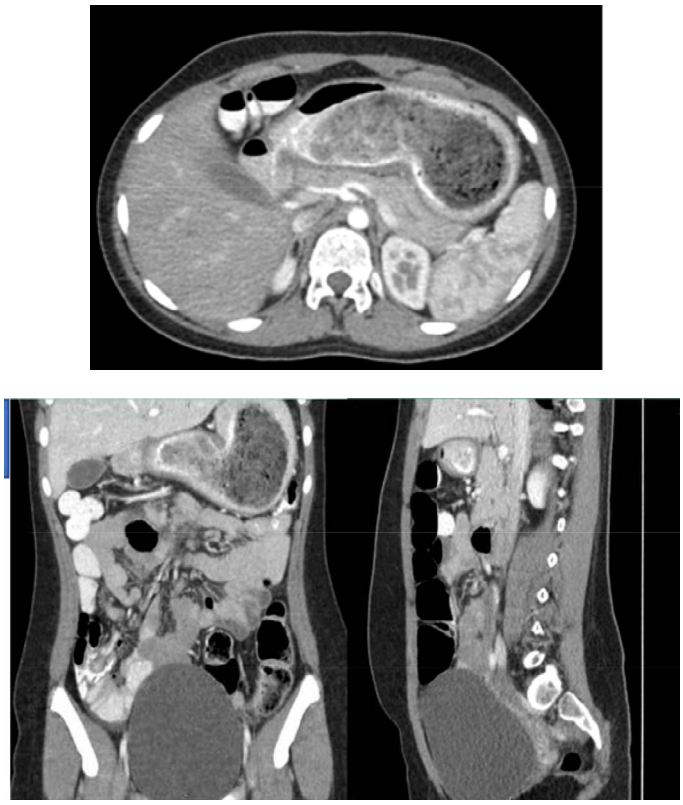

**Image 1.** Simple computerized axial tomography. A. Axial cut: gastric chamber occupied by an isodense, non-homogenous material, from the pylorus within the duodenal bulb, with filiform passage of contrast. B. Coronal cut: gastric and duodenal trichobezoar and filiform passage of contrast which highlights the tail of the Rapunzel syndrome. C. Sagittal cut: Duodenal bulb with oral radio-opaque contrast and an isodense, corresponding to the tail of a Rapunzel syndrome.

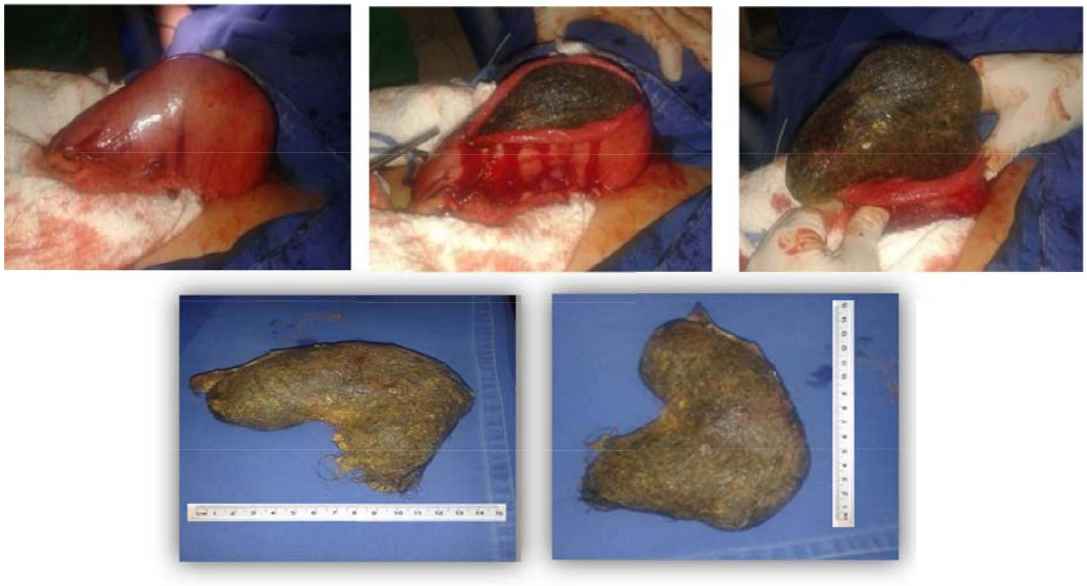

**Image 2**. Gastric and duodenal trichobezoar made up of hair and intestinal digestive juices. Note the detachment of the trichobezoar tail on the lower end. Weight: 350 grams.

## Discussion

The patient with whom this case report is concerned consulted the emrgency department for abdominal pain, without any abdominal mass palpable or visible on ultrasound at the admission. Neither did she have alopecia.

More than 120 cases of Rapunzel syndrome have been reported (3-86), with a mean age of presentation in children of 6.5 years (2.5-18 years, SD 0.7 years), and a female:male ratio of 10:1. The most common symptom for consultation was abdominal pain with 63% of the cases. Likewise, in 38% of the reported cases no abdominal mass was palpated.

In our case, pancreatitis was suspected because of the elevated amylase and lipase levels, which were more than three times above the normal level. In the available literature on the topic, we only found three cases of elevated amylase and lipase levels (48, 49, 64), such as ocurred in our case study, that according to the standards proposed by the INSPPIRE Consortium (The International Study Group of Pediatric Pancreatitis: In Search for a Cure) (87) fulfilled two criteria for acute pancreatitis, which later was ruled out since the serum levels of amylase and lipase returned to normal after surgery. It is interesting to note that only 33% of the cases reported ultrasound as the diagnostic tool that permitted visualization of the trichobezoar. The most frequently described complications were ulcer, perforation, anemia, recurrence (which were ruled out in the our case), and digestive tract obstruction due to mass effect.

The initial assessment of each child by the healthcare team (nurses, psychologists, general physicians, physicians in training such as students, interns and residents, as well as specialists in different areas of healthcare), should look for subtle signs, such as hairs in different growth stages, as well as affected eyebrows, eyelashes and/or nails (which were not identified in this case), since early documentation of these lesions allows quick intervention and avoidance of the progression to chronicity and greater global involvement of the child´s function. In school or rural settings, attention should be paid to teacher reports of conducts such as pulling or biting hair and/or nails, which could indicate TTM or onychophagia (88).

The diagnosis is clinical, and is associated with OCD, anxiety and/or depression affecting the patients' functioning (89). A greater severity of TTM in adolescents with family dysfunction (90), such as was identified in this case, has also been documented. However, various genetic etiological hypotheses have been proposed, such as neurobiological pathway involvement of neurotransmitters like serotonin (5HT2A, SLC6A4) and dopamine (DRD4, DRD1) (still under investigation), since studies show that only the 5HT2A t103c variant differs when patients with TTM and controls are compared (91). Another hypothesis about its causes is the dysregulation of the glutamate metabolism, which alters impulse control by increasing neuronal oxidative stress in specific central nervous system zones (92). Tomography is useful, because it allows more detailed visualization of the progression in the digestive tract. 95% of the cases found in the existing literature required laparotomy, just as in the present case. The rest were treated with laparoscopy, which is technically difficult.

Mansueto et al. have proposed a therapuetic intervention in four phases. The first consists of functional evaluation and analysis of the history, behaviors and functional consequences of the trichotillomania. The information obtained from this functional analysis is used in the second phase to identify its cognitive, sensory, affective, motor, or environmental modalities. This identification allows the implementation of therapeutic strategies by the interdisciplinary team in the third phase; and in the fourth phase, the strategies are evaluated and modified according to the clinical results (93).

The treatment and psychological follow-up diminishes recurrence, as does behavioral therapy (94), which is not statistically significantly different to SSRIs in relation to trichotillomania. Likewise, no study of SSRIs versus placebos has shown strong evidence of improvement in trichotillomania or behavior (95). Other pharmacological alternatives, such as N-acetylcysteine (a glutamate modulator), olanzapine and chlorpromazine, only have preliminary evidence in small studies, and therefore are not recommended for routine treatment (96).

There are psychological treatments aimed at guaranteeing behavior control in adolescents with TTM before adulthood and before functional impact in childhood (97). Among these non-pharmacological interventions, cognitive-behavioral therapy and habit reversal training are highlighted (98), since this disorder has a social and economic impact as it affects emotional expressions and social relationships through avoidance, increases medical consults, and decreases productive school and work time (99). The interdisciplinary management focus in this case study was determinant, since decreased recurrence and complications depend on the short and long-term follow-up of the patient and his/her family, as was proposed in 2016 by Gawłowska-Sawosz, et al. (100).

## Conclusion

Rapunzel syndrome is rare. It should be considered in cases of chronic, recurrent abdominal pain, associated with hyporexia and intestinal obstruction, in female adolescents with psychiatric disorders. The biopsychosocial environment should be assessed and support be given through interdisciplinary follow-up of the patient and the family, avoiding a functional impact on the child's social, educational, emotional, cognitive and motor abilities, which can affect his or her growth and development.

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## Conflict of interests

The authors declare no conflict of interests.

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