# A Rare Case Report of Rapunzel Syndrome with Multiple Small Gut Intusseceptions

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## ABSTRACT

Rapunzel syndrome (RS) is a condition defined as the presence of trichobezoar in the form of hair. It is found in patients with psychiatric disorders called trichotillomania (the habit of pulling hair) and trichophagia (repeated ingestion of hair). Children with pica eat non-digestible things which include long threads, hair, grass, plastic toys, *etc*. in order to cope with conditions such as anxiety, stress, depression, fear, or abuse. The cause of eating indigestible things in children is not known. In this case report, the authors present a case of a 13-year female with a history of weight loss and abdominal pain on and off. On investigations, a large trichobezoar was detected in the form of hair and threads along with multiple jejunal intussusceptions. Surgical removal of trichobezoar was done by opening the stomach and intussusceptions were pulled back from the distal intestine. The patient was discharged home in stable condition.

**Key Words:** *Rapunzel, Syndrome, Hair, Intussusceptions, Surgery*.

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## INTRODUCTION

Rapunzel syndrome (RS) is a condition defined as the presence of trichobezoar in the stomach or small bowel. The word Rapunzel is taken from a female cartoon character who had long hair.[1] Pica is defined as a condition of eating disorder in which a person eats things that are not considered as food e.g., toys, grass, and threads.[2] It is present in patients with psychiatric disorders called trichotillomania (the habit of pulling hair) and trichophagia (repeated ingestion of hair).[3] Intussusception is the telescoping of one part of the gut into another and in most of the cases, the aetiology remains unclear. This condition is most common in children less than five years of age but can present later in life with colicky abdominal pain.[4]

Here, the authors report a case of a 13-year girl who presented in the outdoor department with a history of abdominal pain on and off. Later, on investigation, it turned out to be a case of RS; stomach and proximal small bowel containing a huge trichobezoar with multiple small bowel intussusceptions.

## CASE REPORT

A 13-year girl presented to the surgical outdoor department of a teaching hospital, with complaints of abdominal pain and distention on and off.

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She was accompanied by her mother, who gave a history of decreased appetite of her daughter with vomiting after some meals. These symptoms have continued for the last three years. Initially, these symptoms occurred monthly, but now, with an increase in frequency, they started happening weekly.

Physical examination showed a cachexic female patient with a body mass index (BMI) of 15 kg/m$^2$ and normal vital signs. The abdominal examination showed a distended abdomen without any tenderness or signs of peritonitis or palpable abdominal mass. The chest x-ray was normal. Abdominal x-ray showed a distended stomach with displaced small gut loops. The abdominal ultrasound report showed a distended stomach with collapsed distal gut loops. Computer tomography (CT) revealed intestinal obstruction at the level of the ileum with distended proximal stomach and duodenum and suspicion of bezoar and jejunoileal intussusception (Figure 1). The diagnosis of intestinal obstruction due to bezoar and intussusception was made. A plan of exploratory laparotomy under general anaesthesia after taking informed and written consent from the family of the patient was finalised.

On exploration, the whole of the stomach and duodenum was grossly distended with the telescoping of jejunum into jejunum and ileum into jejunum was noted. A firm mass was palpable in the whole of the stomach and duodenum, which could not be milked distally, so a gastrostomy was performed through the anterior surface of the stomach. The stomach lumen contained a bezoar of about 12 × 15 cm, composed of long hair along with threads of different sizes and diameters extending into the duodenum and jejunum (Figure 2). This mass was evacuated, and the gastrostomy was closed. Intussusception needed no intervention, and proximal loops were pulled back easily with no

serosal tear. Surgery remained uneventful, and the patient was shifted to the ward, where she stayed for six days. She was discharged home on the 7th postoperative day and referred to a child psychiatrist for counselling and treatment to prevent future incidents of eating hair and threads.

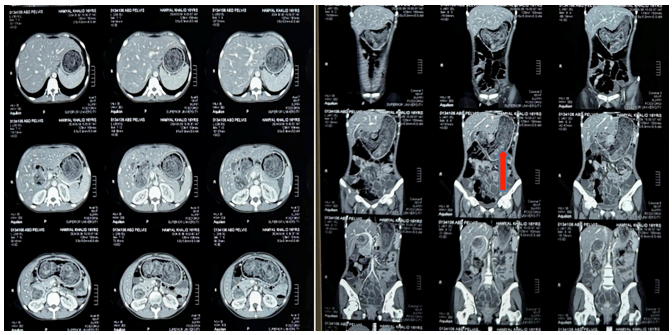

**Figure 1: Axial and coronal views of CT scan of the abdomen showing a mass completely filling the stomach and extending into duodenum and jejunum (red arrows).**

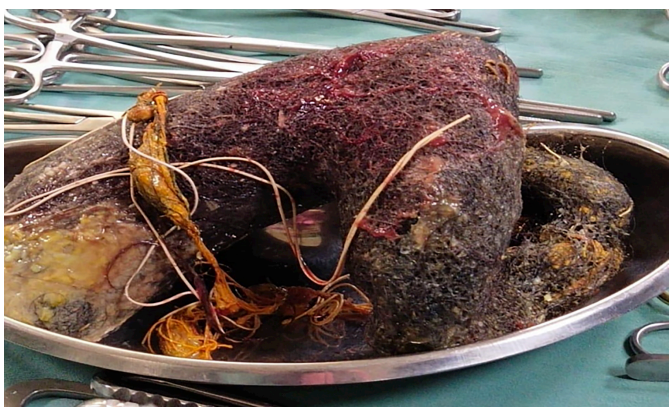

**Figure 2: Huge mass composed of hair and fibres after removing from stomach extending into duodenum and jejunum.**

## DISCUSSION

Pica or compulsive eating disorders and trichobezoars are rare conditions mostly found in children with psychiatric disorders. The word pica is derived from a bird species, the Eurasian magpie, which has a reputation for eating unusual objects. The children with pica eat non-digestible things which include long threads, hair, grass, plastic toys, *etc*., in order to cope with conditions such as anxiety, stress, depression, fear, or abuse.[5] The cause of eating indigestible things in children is not known. One reason is the developmental pattern in which infants tend to put everything in their mouth, although this condition of pica goes away as the child grows up.[6] The term RS is derived from a cartoon character named Rapunzel, which was first described in a fairy tale by the Grimm brothers in which a princess Rapunzel takes advantage of her long hair to escape her lover. When trichobezoar of the stomach extends into the duodenum and jejunum, this condition is called RS and was first described by Vaughan *et al*. in 1962.[7] The diagnosis of RS is tricky, and it is highly suspected in female patients with alopecia and low BMI, along with a distended abdomen and palpable mass in the epigastrium.

Usually, the patient develops symptoms such as abdominal pain, vomiting, and loss of appetite. With the passage of time, upper gastrointestinal bleeding, acute obstruction, nutritional deficiencies, protein-losing enteropathies, and anaemia develop, which indirectly affect the growth of the patient, affecting their height and weight as compared to normal healthy individuals.[2,8] In this case, the patient presented in the outdoor department with symptoms of vomiting and abdominal pain on and off. This is an unusual presentation as the patient had a loss of appetite and severe weight loss, but somehow she managed to hide it from her parents and survived with such a huge trichobezoar.

In literature, very few cases are reported showing RS with intussusception. In most of these cases, the intussusception is ileo-ileal, while in our case, the intussusception was jejuno-jejejunal and jejuno-ileal. Intussusception is present in almost all reported cases of RS, and the association of these two pathologies is not clearly understood. It is considered that the tail of trichobezoar or the free-floating threads interfere with the peristalsis of the gut, hence providing a nidus to develop intussusception.[3] The lowest age of child reported in literature having RS with an intussusception is 2.5 years, which was managed by enterotomy.[9]

The definite diagnosis of RS is confirmed by a CT scan of the abdomen and pelvis. Endoscopy is a useful tool for diagnosing as well as for removal of small trichobezoars but it is not suitable for large solid masses which are densely adherent to the gastric folds.[2] For trichobezoars larger in size and too hard for dissolution or retrieval or if there is intestinal perforation, open surgery is usually considered for complete evacuation of the mass. Surgical procedures used for the removal of trichobezoars include gastrostomy, enterostomy, and resection and anastomosis of bowel, depending upon the situation.[5] Postoperative psychiatric counselling of patients as well as parents is very important. Removing the stress factors from the child's daily life and keeping a friendly relationship with the child will help engage the child from stressful thoughts and ultimately prevent such incidents in the future.[10]

## CONCLUSION

RS is a rare type of trichobezoar which is mostly associated with small gut intussusception. The suspicion of RS should be kept in mind in diagnosing an unexplained weight loss with anorexia, especially in female children. Psychiatric counselling is very important to prevent these cases from happening in the future.

### PATIENT'S CONSENT:

Informed consent was taken from the parents of the patient to share the relevant data only for the purpose of publication while maintaining the confidentiality of the patient's identity.

### COMPETING INTEREST:

The authors declared no conflict of interest.

### AUTHORS' CONTRIBUTION:

ASA, FU, MI, MS, MSI: Conceptualisation, design, drafting, and critical revision of the manuscript for important intellectual content.
All authors approved the final version of the manuscript to be published.

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