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*Case Report:*

# Rapunzel Syndrome: Trichobezoar in a 13 Years Old Girl.

**Authors**
**Krishnanand,** Associate Professor,
**Roshan Chanchalani,** Assistant Professor,
**Kamlesh Dhruv,** Associate Professor,
**Chirayu Medical College and Hospital, Bhopal-Indore Highway, Near Bairagarh, Bhopal-462030, Madhya Pradesh, India.**

**Address for Correspondence**
**Dr Krishnanand,**
Associate Professor,
Department of Surgery,
Chirayu Medical College and Hospital,
Bhopal-Indore Highway, Near Bairagarh,
Bhopal - 462030, Madhya Pradesh, India.
**E-mail:** drkrishnanand@yahoo.in

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**Abstract: Background:** Rapunzel syndrome is a rare type of trichobezoar with an extension of the hair into the small bowel. Clinical presentation is deceptive and vague ranging from abdominal mass to gastrointestinal symptoms. **Case Presentation:** We present a 13 years old girl with Rapunzel syndrome, where the trichobezoar was not suspected at all especially with negative history of trichophagia. In majority of the cases the diagnosis was made very late in the history of the disease, at a stage where surgery is the only cure for this syndrome. **Conclusion:** A trichobezoar represents a serious surgical condition. It is important to consider such diagnosis in face of suggestive symptoms, even if signs of trichotillomania are not present. The discrepancies between the prevalence of trichotillomania and trichobezoars due to trichophagia may be due to issues related to self-selection of patients and symptom severity. Such issues may also be important in the study of impulsive-compulsive spectrum models and to their relevance to impulse control disorders.
**Key Words:** Rapunzel syndrome; Trichobezoar

**Introduction:**
This syndrome is named after the girl with the long tresses in the fairy tale written by the Grimm Brothers in 1812. The Rapunzel syndrome was first reported in the literature by Vaughan et al. in 1968.[1] For centuries bezoars have been known to occur in the form of undigested masses found in the stomach of animals and humans. Nevertheless, this finding has become more common in humans as a result of more frequent manipulation of the gastrointestinal tract.[2] From the increase in the incidence of this finding and the current ease with which knowledge is disseminated today, it is clear that the Rapunzel syndrome, two cases of which were reported by Vaughan et al., remains rare, a finding of phytobezoars being more common.[2] This fact was recently confirmed in a literature review performed in 2007 that identified 27 cases of Rapunzel syndrome described between 1968 and 2006.[3] A few other reports on this syndrome were published in the medical literature after this date.[4-6]
There are several different forms of presentation of this syndrome; however, in general, it involves the presence of a gastric trichobezoar with a long tail extending beyond the duodenum, as found in the case reported here. The factor responsible for this syndrome is the compulsion of patients to pull out their own hair and swallow it, processes referred to as trichotillomania and trichophagia, disorders that affect young girls with or without known psychiatric disorders.[3-4]

**Case Presentation:**
A 13 year old female came to surgery OPD with the history of abdominal mass associated with epigastric pain, sense of fullness, vomiting after meals for the last 6 months. On examination she was found mildly dehydrated, abdominal palpation (Fig.01) revealed a well defined mass occupying the upper 1/3rd of the abdomen, the mass was not tender and was firm in consistency measuring approx 8cm X 10 cm. On further evaluation, a microcytic anaemia was detected, while all the laboratory parameters were within the normal range. An abdominal ultrasound revealed a superficially located broad band of high amplitude echoes along the anterior wall of mass with sharp, clean post acoustic shadowing and a hyperechoic curvilinear dense strip with acoustic shadowing and no through transmission. An upper gastrointestinal contrast study was eventually performed which showed a grossly distended stomach with irregular filling defects extending into the proximal gut (Fig.02), mottled filling defect and a plain abdominal CT reported a mobile intragastric mass consisting of compressed concentric rings,

with a high density pattern due to the presence of entrapped air and food debris (Fig.03).

The patient was referred to Surgery, and through upper mid line incision the stomach was opened (Gastrotomy fig.04) between two Vicryl stay suture. A huge Trichobezoar was identified which took the shape of the stomach (Figure 05). There was a long tail of hair extending through the pylorus into the small bowel (fig.06). By this feature the diagnosis was clear of a Rapunzel syndrome. Hairball was removed intact and stomach was closed in double layers using continuous vicryl and silk sutures. The patient had an uneventful postoperative course and was discharged after six days. The parents were advised to visit pediatric psychiatry for follow up.

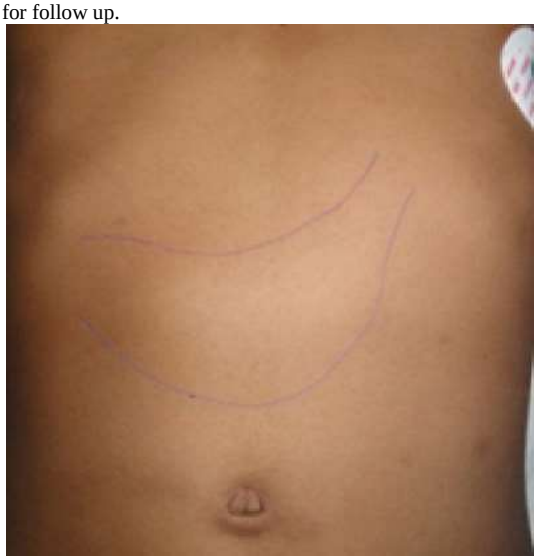

**Fig. 1: Palpable abdominal mass**

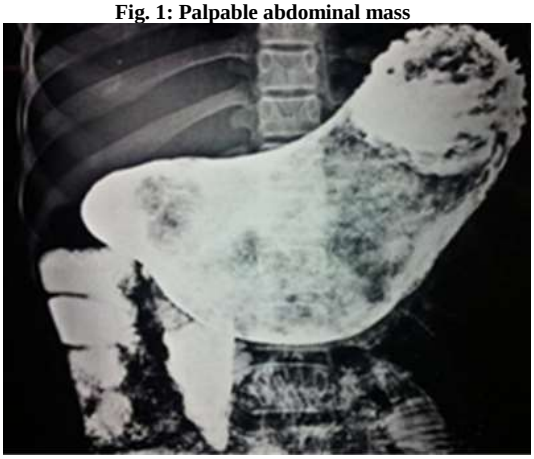

**Fig. 2: GI Contrast study**

**Discussion**:

The common presentation of trichobezoar is in young females usually with an underlying psychiatric disorder. In our case the presentation is in a very young age with hair extending down to the small bowel, causing symptoms, which could mimic gastrointestinal infections and infestation especially in endemic areas. The commonly accepted definition of Rapunzel syndrome is that of a gastric trichobezoar with a tail extending to the jejunum, ileum or the ileocecal junction.

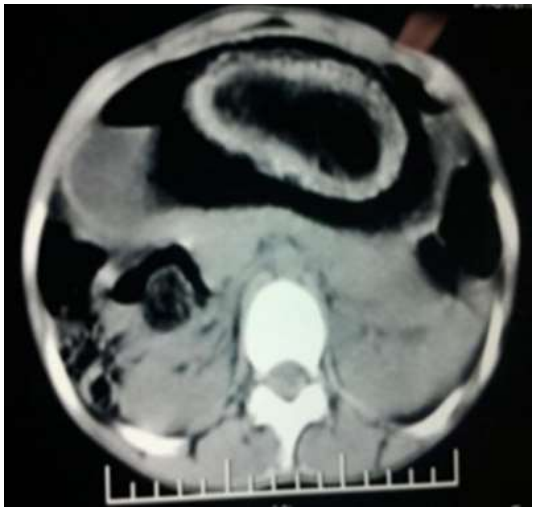

**Fig. 3: CT image**

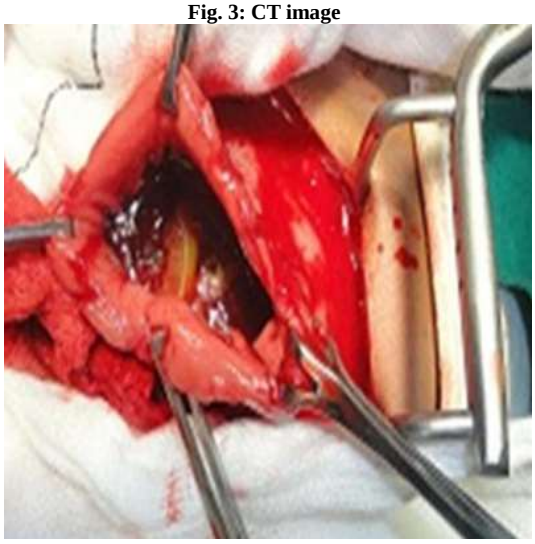

**Fig. 4: Gastrostomy**

**Fig. 5: Trichbezoar removed**

**Fig. 6: Rapunzel**

Majority of cases of trichobezoar present late, due to the low index of suspicion by the physician. Of 131 collected cases of trichobezoar, a palpable abdominal mass was present in (87.7%), abdominal pain (70.2%), nausea and vomiling (64.9%), weakness and weight loss (38.1%), constipation or diarrhoea (32%) and haematemesis (6.1%). The laboratory investigations revealed low haemogiobin in about 62% (average).[7]

Patient with Trichotillomania (a psychological condition that involves strong urges to pull hair), around 30% will engage in trichophagia, and of these, only 1% will go on to eat their hair to the extent requiring surgical removal.[8]Less than half of the patients give a history of trichophagia. There has been few cases of recurrence following successful surgery.[9]

The early detection of trichophagia and trichobezoar depends on an effective screening for trichotillomania and related behaviours, in order to prevent a possibly life-threatening condition with important medical and surgical morbidity. Such effort must include a better collaboration between medical and surgical specialties, dealing with particular aspects of therapeutic relationship regarding shame and guilt as well as considering that trichophagia may be more often present than the majority of clinicians, psychiatrists in particular, would expect.

**Conclusion:**

A trichobezoar represents a serious surgical condition. It is important to consider such diagnosis in face of suggestive symptoms, even if signs of trichotillomania are not present. The discrepancies between the prevalence of trichotillomania and trichobezoars due to trichophagia may be due to issues related to self-selection of patients and symptom severity. Such issues may also be important in the study of impulsive-compulsive spectrum models and to their relevance to impulse control disorders.

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