

The Role of Bio-Psychological Factors in Understanding Trichophagia Disorder (Rapunzel Syndrome)

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ABSTRACT

Trichophagia is a somatic symptom disorder associated with trichotillomania (hair pulling) that results in the eating of hair. This can lead to serious medical consequences and can even be fatal. People who eat hair for a long time may experience abdominal pain, nausea, vomiting, diarrhea, or constipation due to the accumulation of hair that forms hairballs, medically known as trichobiosis. Trichophagia is a psychiatric somatic symptom disorder that is more common, though not exclusively, in women. Also known as Rapunzel syndrome, trichophagia can lead to serious, sometimes life-threatening medical problems that may require gastrointestinal surgery. It is likely caused by the growth of a hairball that accumulates in the small intestine. When the small intestine runs out of space, the hair invades surrounding areas. Symptoms do not appear until the digestive tract (esophagus to rectum) becomes blocked with hair. This syndrome is often undiagnosed or untreated and is often misdiagnosed as a digestive problem.

Keywords: Trichophagia, Rapunzel Syndrome, Trichotillomania, Psychology Disorder.

Introduction to Trichophagia and Trichotillomania

Trichotillomania is a somatic symptom disorder characterized by hair pulling. Hair can be pulled from anywhere it grows (e.g., eyebrows, eyelashes, pubic hair, etc.). Hair pulling from the head often leaves noticeable patches, often leading the person to wear hats or wigs to try to hide or camouflage the bald spots. People may avoid going to work or socializing. Approximately 20% of people with trichotillomania eat their hair after pulling it out [1].

Trichotillomania and trichophagia are body-focused repetitive behaviors (BFRBs) that are most common in women who begin eating and pulling out their hair during childhood or adolescence. In men, the disorder often begins at an earlier age, in early to mid-childhood. Children may eat their own hair, the hair of their dolls, found hair, or hair from paintbrushes [1].

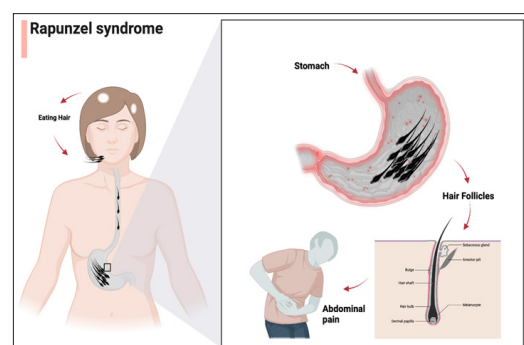


Figure 1: Schematic of symptoms associated with Rapunzel syndrome in the stomach [1].

Trichophagia (hair eating) can be the final stage in a series of ritualistic hair-pulling behaviors. Some people roll the pulled hair into a ball and swallow it whole, which is especially dangerous if they have long hair. However, not everyone eats their hair, and not all people who eat hair pull out their hair. Related to trichotillomania, trichophagia is considered a body-

focused repetitive behavior diagnosis. Other repetitive behaviors serve the same soothing purpose as hair pulling and eating. Other behaviors that often occur with trichotillomania and their associated frequencies include:

Hair pulling (51%)
Nail biting (30%)
Lip/cheek biting (26%)
Nose picking (12%)

Signs and Symptoms of Trichophagia

Unfortunately, the signs and symptoms of Rapunzel syndrome usually do not appear until the physical symptoms of hairballs have accumulated over years, if not decades. People with trichophagia may seek medical help due to symptoms such as abdominal pain or swelling, nausea, and vomiting. Many patients deny any history of trichotillomania or trichophagia, even if specifically asked [2].

The eight symptoms of trichophagia are:

Abdominal pain or swelling
A lump in the upper abdomen
Halitosis (bad breath)
Peritonitis (inflammation of the abdominal wall)
Nausea
Vomiting
Feeling full after eating small meals
Weight loss

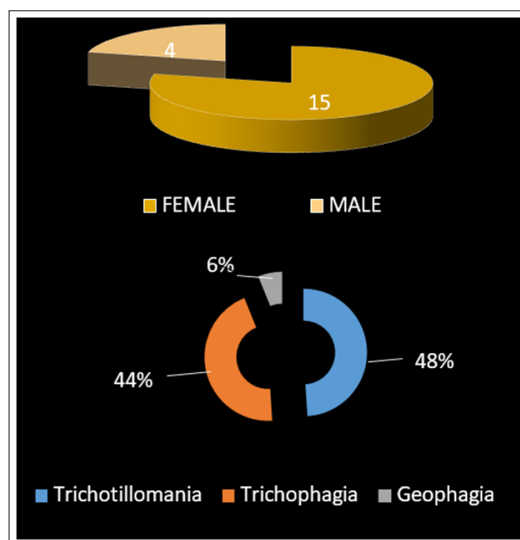


Figure 2: Schematic of a pie chart of the frequency of males and females with Rapunzel syndrome [2].

Rapunzel Syndrome Health Risks

Eating hair has several health implications. Human hair is indigestible due to its enzyme-resistant nature and smooth, slippery surface, meaning it stagnates in the digestive system. As a result, the hair gets trapped between the folds of the stomach lining, leading to the formation of hairballs. On average, only 1 percent of patients with trichophagia develop hairballs [2]. Trichobezoars are seen in 24 percent of women with trichotillomania and may be more complicated in women and men with long hair [2].

Potential health consequences of hairballs include:
Ulcers that perforate the stomach and spread bacteria
Infected or inflamed abdominal cavity (peritonitis)
Gastrointestinal bleeding due to mucosal erosion
Intestinal obstruction

It should be noted that Rapunzel syndrome can be fatal. While the prevalence of hairballs in humans is low, the mortality rate can reach 30% if left untreated [2].

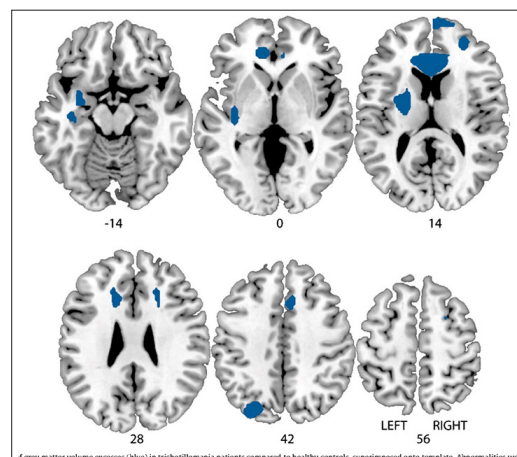


Figure 3: 3D schematic of brain tissue disorder in Rapunzel syndrome [3].

Etiology and Triggers of Rapunzel Syndrome

There is no definitive cause for Rapunzel syndrome, but there appears to be a neurobiological predisposition in people who engage in these behaviors. Conditions such as intellectual disabilities, anemia, and iron deficiency have been found in people with trichophagia, but it is not clear what role they play in causing it [3].

Emotional States Associated with Trichophagia

The behaviors of trichotillomania and trichophagia appear to have a calming effect. These behaviors are used as a way to regulate emotion and cope with stress, boredom, negative or upsetting feelings, anxiety, tension, loneliness, fatigue, or hopelessness. Other emotional factors that contribute to these behaviors include parental discord, grief, or other family problems. People often feel guilty, ashamed, and embarrassed about pulling or eating their hair. This can lead to social isolation in fear of being judged negatively by others. Social isolation often leads to depression and loss of self-esteem, resulting in a desire to self-soothe by pulling or eating hair. Tension often builds up, which triggers the urge to eat hair that has been pulled from the body or from other sources (e.g., loose hair from others). The urges can be done consciously as part of a routine or unconsciously, similar to a nervous habit. Longer periods can lead to feelings of depersonalization, trance-like states, and loss of time [4].

Diagnosis of Rapunzel Syndrome

As mentioned, the diagnosis of trichophagia is often made only after a visit to the doctor for gastrointestinal pain or other related symptoms, most likely due to a hairball. Tests are performed by gastrointestinal endoscopy, ultrasound, CT scan, or X-ray of the abdomen [5].

Treatment of Rapunzel Syndrome

Psychological treatment is also recommended. It is important to seek help from a therapist who specializes in the treatment of trichotillomania and offers habit change training. Along with behavioral therapy, this type of training identifies triggers for behaviors and teaches skills that address emotional factors and stress management [5].

In habit reversal therapy, patients learn to become aware of internal and external cues, as well as predictable and unpredictable situations that lead to hair pulling. They practice competitive behaviors and movements, such as clapping hands, sitting on hands, or keeping hands busy by engaging in activities such as knitting, crocheting, needlework, coloring, or other tasks that involve the hands [5].

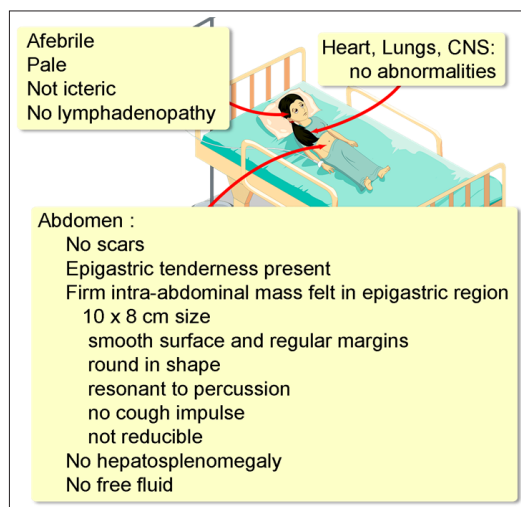


Figure 4: Symptoms associated with Rapunzel syndrome [4].

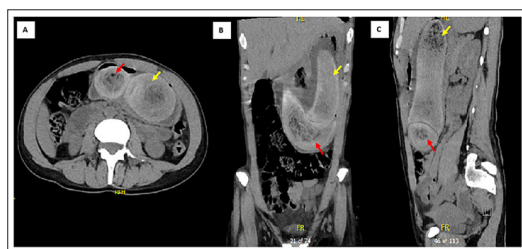


Figure 5: Radiological images of a stomach containing a hair mass in a person with Rapunzel syndrome [5].

Autogenic training is another behavioral therapy option designed to reduce stress and improve body relaxation. It teaches relaxation techniques designed to lower blood pressure, heart rate, and body temperature. Other cognitive behavioral therapy (CBT) strategies may include writing, contingency planning for action based on desires, self-monitoring of symptomatic behaviors, monitoring thoughts that occur before and after episodes, and restructuring cognitive distortions and negative automatic thoughts [5].

Surgery: Emergency surgery may be performed depending on the severity of the case. Surgical options include:

Laparotomy (open surgery)

Laparoscopy (a local abdominal surgery that breaks up and

removes the hairball)

Endoscopy (a nonsurgical procedure that inserts a tube down the throat into the esophagus)

Medication

Psychiatric medications have also been prescribed to treat the disorder, including N-acetylcysteine, naltrexone, topiramate, neuroleptics, and selective serotonin reuptake inhibitors [5].

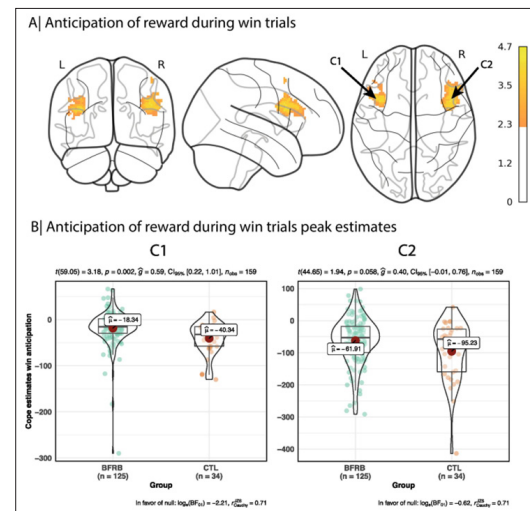


Figure 6: Schematic of brain tissue disruption in the reward system in Rapunzel syndrome [5].

Trichophagia is a serious mental illness that can be successfully treated with CBT and medication. It is important to seek help as soon as you recognize the problem to prevent hairballs from becoming surgically invasive or health-threatening. It is important for people to know that even if they keep their hair eating a secret, they are not alone [5].

Discussion

Rapunzel syndrome is a rare intestinal disease that occurs in humans affected by trichotillomania and caused by eating hair (Trichophagia). In this condition, the trichobezoar (a mass of hair) extends from the stomach to the cecum and is named after the cartoon character Rapunzel, the long-haired girl. The indigestibility and slippery nature of the hair strands lead to the hair getting trapped in the folds of the gastric mucosa and preventing the hair strands from being expelled. Over time, the hair strands accumulate in the stomach and the mass thickens and expands to form a solid body in the shape of a stomach, which can sometimes even enclose the entire stomach space. Since this mass is too large to pass through the stomach, it can lead to gastric paralysis (atony).

Alopecia areata, pulling out hair from the roots and eating it, playing with pulled hair or rubbing it on the lips or face, pulling out eyebrows, eyelids, and other body hair, and pulling hair from pets, dolls, or blankets can be signs of a predisposition to this syndrome. The cause of hair-pulling disorder is still unknown, but like many complex disorders, it is likely the result of a combination of genetic and environmental factors. Family history plays an important role in the development of this condition, and it usually appears in the second decade of life and

is a lifelong problem that can be caused by anxiety or obsessive-compulsive disorder (OCD). People with Rapunzel syndrome have a tendency to pull out and eat their own hair, and sometimes other people's hair or wig hair, due to mental disorders or mental retardation. Alopecia areata and halitosis (bad breath) can be clues on physical examination. The prevalence of this syndrome in young people with emotional and genetic disorders in the second decade of life is especially common in girls with long hair, of course, this condition can also occur in men due to eating other people's hair, of course, one case of Rapunzel syndrome has been reported in an 18-year-old boy due to eating cotton.

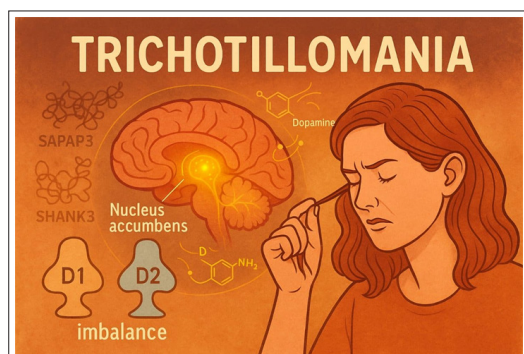


Figure 7: Schematic of the role of the hormone dopamine in hair pulling [5].

To diagnose the syndrome, an upper gastrointestinal endoscopy must be performed to allow direct visualization, and this method also allows for sampling and therapeutic intervention, but a CT scan of the abdomen is required to accurately determine the size and extent of trichobezoar lesions. The most common symptoms of trichobezoar are abdominal pain, nausea and vomiting, anorexia and weight loss, and less common side effects include hematoma and muscle cramps, peritonitis, gastric ulcers, megaloblastic anemia, acute pancreatitis, gastric emphysema, enteropathy and even death. Management and treatment of trichobezoar requires complete removal of the mass and primary prevention of recurrence due to physical, emotional, or genetic causes. Depending on the size and location of the mass, removal may be done endoscopically or surgically. In case of early detection and small size of the mass, lithotripsy or intramuscular injection of enzymes as well as medication (metoclopramide and acetylcysteine) can be effective, but in case of large mass, surgery is performed [1-5].

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