



## **Desmosis Coli**

Constipation is a very serious condition in children. Medical manageable conditions associated with chronic constipation can be classified into normal transit constipation, slow transit constipation and disorders of defecation with retention in the rectum. The most common, slow transit constipation, can be caused by genetic links, dysfunctional enteric nerves, decrease levels of substance P, reduced numbers of interstitial cells of Cajal and disorders of connective tissue synthesis. Desmosis coli is a disturbance of the intramural connective tissue mesh network of the colonic wall which leads to a hypoperistalsis syndrome with chronic constipation in the absence of any anomaly of the vegetative gut innervation. The peristaltic movement of the gut is a function of the alternating contraction and relaxation of circular and longitudinal muscles. This movement is induced by a tendon-like connective-tissue net in the circular and longitudinal muscles, which are both rooted in a connective-tissue plexus layer. This connective tissue layer is not developed in children with desmosis coli. Clinically, the child develops hypoperistalsis or aperistalsis with massive elongation and dilatation of the colon. Hirschsprung's disease can coexist with desmosis coli as a familiar trait. The enteric nervous system is normal or near-normal in the affected areas of desmosis coli. Two major subtypes of desmosis can be distinguished: the rare congenital (primary) aplastic desmosis of childhood, and the more common atrophic (secondary) desmosis of adulthood typically incomplete and associated with a hypoperistaltic syndrome. Seromuscular biopsies are required to prove desmosis in gastrointestinal dysmotility disorders. Management of desmosis coli is controversial.

### **References:**

- 1- Meier-Ruge WA, Holschneider AM, Scharli: New pathogenetic aspects of gut dysmotility in aplastic and hypoplastic desmosis of early childhood. *Pediatr Surg Int.* 17(2-3):140-3, 2001
- 2- Meier-Ruge WA: Desmosis of the colon: a working hypothesis of primary chronic constipation. *Eur J Pediatr Surg.* 8(5):299-303, 1998
- 3- Marshall DG, Meier-Ruge WA, Chakravarti A, Langer JC: Chronic constipation due to Hirschsprung's disease and desmosis coli in a family. *Pediatr Surg Int.* 18(2-3):110-4, 2002
- 4- Habner U, Meier-Ruge W, Halsband H: Four cases of desmosis coli: severe chronic constipation, massive dilatation of the colon, and hypoperistalsis due to changes in the colonic connective-tissue net. *Pediatr Surg Int.* 18(2-3):198-203, 2002
- 5- Meier-Ruge WA, Bruder E: [The morphological characteristics of aplastic and atrophic desmosis of the intestine]. *Pathologe.* 28(2):149-54, 2007
- 6- Bruhin-Feichter S, Meier-Ruge W, Martucciello G, Bruder E: Connective tissue in gut development: a key player in motility and in intestinal desmosis. *Eur J Pediatr Surg.* 22(6):445-59, 2012

## **Transfer Burn Center**

Burns are a major source of injury in children that must be managed within the confines of specialized centers whenever possible to obtain best results in morbidity, mortality and rehabilitation. Burns are classified as First degree (partial thickness, superficial, red, sometimes painful), Second degree (partial thickness, skin may be red, blistered, swollen, very painful), or Third degree (full-thickness, whitish, charred, translucent, no pin prick sensation in burned area). The American Burn Association has established strict criteria for admission or transfer of infants and children to such regional burn centers. They include: 1- Children with less than 10 years of age with burns compromising more than 10% of total body surface area. 2- Children with age above the 10 years with more than 20% total body surface area burned. 3- Children of all ages with full-thickness burn above the 5% of total body surface area. 4- Children with evidence of inhalation injury. 5- Burn to face, eyes, ears, genitalia or joints. 6- Any burn associated with a major fracture or significant traumatic injury. 7- Third degree burns in any age group. 8- Chemical burns. 9- Electrical burns. 10- Burn injury in patients with preexisting medical disorders that could complicate management, prolong recovery, or affect mortality. 10- Burned children in hospitals without qualified personnel or equipment for the care of children. 11- Burn injury in patients who will require special social, emotional, or rehabilitative intervention.

### **References:**

1- [www.ameriburn.org](http://www.ameriburn.org)

## **Rapunzel Syndrome**

Bezoars are concretion of human or vegetable fibers that accumulate in the gastrointestinal tract. The most common type of bezoar is the trichobezoar which is mostly made of hair or hairlike fibers. Trichobezoars most commonly present during adolescent years and during the second decade of life. Most cases of trichobezoars occur in female patients. Eating hair denotes an underlying psychiatry disorder. Most trichobezoars lodge within the body of the stomach. When the trichobezoar extends beyond the pyloric muscle and beyond as a tail is called Rapunzel syndrome. The distal end of the bezoar may be in the jejunum, ileum or the colon. Most children with trichobezoars suffer from psychiatry disorders including trichotillomania (pulling out of their own hair) and trichophagia (eating of hair). Hair is retained in the folds of the gastric mucosa, and as it accumulates the peristalsis creates an enmeshed ball. The stomach dilates significantly. Decomposition and fermentation of fats give the bezoar and the child breath a putrid smell. The black color of the bezoar is due to the effect of acid on hair protein denaturalization. Most cases are asymptomatic for a long period of time. Most common presenting signs are abdominal pain, nausea, vomiting, obstruction and peritonitis. Other times children present with anorexia, weight loss, hematemesis or intussusception. Bezoars can cause gastric ulceration, obstructive jaundice, acute pancreatitis, and gastric emphysema. Imaging (UGIS, Ultrasound and CT-Scan) shows the bezoar as a mass or filling defect and are diagnostic. Endoscopic retrieval of the bezoar is rarely a definitive treatment. Management of trichobezoar includes removal by

gastrotomy and/or enterotomy and behavioral therapy to avoid recurrence.

**References:**

- 1- Ventura DE, Herbella FA, Schettini ST, Delmonte C: Rapunzel syndrome with a fatal outcome in a neglected child. *J Pediatr Surg.* 40(10):1665-7, 2005
- 2- Naik S, Gupta V, Naik S, Rangole A, Chaudhary AK, Jain P, Sharma AK: Rapunzel syndrome reviewed and redefined. *Dig Surg.* 24(3):157-61, 2007
- 3- Gonuguntla V, Joshi DD: Rapunzel syndrome: a comprehensive review of an unusual case of trichobezoar. *Clin Med Res.* 7(3):99-102, 2009
- 4- Crawley AJ, Guillerman RP: Rapunzel syndrome. *Pediatr Radiol.* 40 Suppl 1:S100, 2010
- 5- Kohler JE, Millie M, Neuger E. Trichobezoar causing pancreatitis: first reported case of Rapunzel syndrome in a boy in North America. *J Pediatr Surg.* 47(3):e17-9, 2012
- 6- Fallon SC, Slater BJ, Larimer EL, Brandt ML, Lopez ME. The surgical management of Rapunzel syndrome: a case series and literature review. *J Pediatr Surg.* 48(4):830-4, 2013

---

\* Edited by: Humberto Lugo-Vicente, MD, FACS, FAAP  
Professor of Pediatric Surgery, University of Puerto Rico - School of Medicine,  
Rio Piedras, Puerto Rico. Director - Pediatric Surgery, San Jorge Childrens Hospital.  
Address: P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico USA 00922-0426.  
Tel (787)-786-3495 Fax (787)-720-6103 E-mail: [titolugo@coqui.net](mailto:titolugo@coqui.net)  
Internet: <http://home.coqui.net/titolugo>

©PSU 1993-2013  
ISSN 1089-7739