ESOPHOGEAL STRICTURES

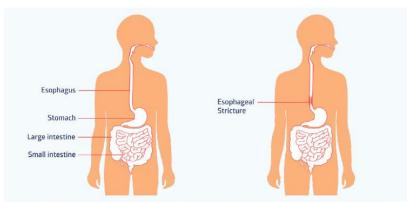
Esophageal stricture (ES) is a narrowing in the esophagus – the muscular tube that carries food and liquids from the mouth to the stomach.

- Most common in recessive dystrophic and junctional EB.
- Narrowed esophagus makes it difficult to swallow food and sometimes liquid.
- Major cause of poor nutrition in recessive dystrophic and junctional EB.
- Not only affects the intake of nutrients but also limits food choice often times the patients favorite foods are removed from the diet affecting enjoyment of eating and quality of life.

How does an ES form?



- Esophagus in individuals with dystrophic and junctional EB has extremely fragile surface lining and makes it easy for it to blister in response to even the most minor trauma.
- Blistering can lead to the formation of scar tissue in the wall of the esophagus and can cause it to narrow or even get blocked.
- Can begin in childhood and risk increases as the patient gets older.



The lining of the esophagus is very fragile in individuals with dystrophic or junctional EB. Friction from rough foods can cause scarring and thickening of the esophageal wall, forming an esophageal stricture.

Symptoms

- Difficulty swallowing (dysphagia)
- Pain with swallowing
- Weight loss or difficulty gaining weight and poor growth
- Regurgitation of food, when food comes back into the mouth from above the stricture
- Food gets stuck in the esophagus (food impaction)
- Frequent burping or hiccups
- Heartburn (burning sensation behind the breast plate bone)

Tests



Barium swallow test: For this test the patient swallows liquid barium, which coats and fills the esophagus, so that it shows up on X-ray images. X-ray pictures are then taken and the radiologist can see if there is a narrowing in the esophagus. Barium is nontoxic and is often flavored to improve the taste.

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Upper gastrointestinal endoscopy: The doctor will place an endoscope (a flexible tube with a light and video camera attached) through the mouth and into the esophagus. It allows the doctor to examine the esophagus and upper intestinal tract.

Treatment



- Esophageal dilatation is a procedure where a dilating device, often a balloon, is placed across the stricture and then inflated to stretch the stricture open and widen the esophagus at that point. The dilatation can be done through the mouth by placing the balloon under X-ray guidance or through an endoscope under the guidance of a camera. For comfort, this procedure may be performed under sedation, including a general anesthetic. A local anesthetic spray may also be applied to the back of the throat. Repeat dilatations are often required to adequately stretch the esophagus. Because EB is a lifetime condition, multiple procedures are often required because of recurrence.
- Despite esophageal dilation, some patients present with significant failure to thrive and require the placement of a gastrostomy tube (also called G-tube) is a tube inserted through the abdomen in the stomach. The tube delivers nutrition directly to the stomach.
- Sometimes, when esophageal strictures are worsened by acid reflux from the stomach into the esophagus, medications such as acid blocking drugs (proton pump inhibitors or H2 blockers) may be added to prevent the stricture from returning. Corticosteroids may also be added and can be given in a form that coats the esophagus (topical) or that is absorbed in the stomach (prednisone pill/liquid).
- Sometimes, extra calories are given by the intravenous route to supplement your child's nutrition. If there is a G-tube (gastrostomy) in place, the dietitian may increase the calories given via this route.

Useful Websites/links:

- www.swrwoundcareprogram.ca
- EB Resources, including EB handbook: https://www.debracanada.org

Disclaimer: Unless stated otherwise, information contained in this document is taken from Epidermolysis Bullosa: A Handbook for EB Patients and Families, Developed by the Section of Dermatology at the Hospital for Sick Children with the Support of DEBRA Canada and Sick Kids Hospital Department of Dermatology EB Expert Team - all content used with permission. This document was created by SWRWCP (August, 2019)



