

Post-fundoplication high-resolution esophageal manometry in a patient with Ehlers-Danlos syndrome

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A 10-year-old boy with developmental delay and Ehlers-Danlos syndrome (EDS) presented with gastroesophageal reflux disease refractory to acid suppression therapy. Esophagogastroduodenoscopy revealed esophagitis, while multichannel pH-impedance monitoring confirmed elevated esophageal acid exposure. Nissen fundoplication was subsequently performed. Three weeks postoperatively, the patient reported worsening dysphagia for both solids and liquids. A contrast esophagogram showed intact fundoplication, but delay of contrast passing through a markedly narrow channel. Endoscopic balloon dilatation of the esophagogastric junction (EGJ) to 15 mm was completed, but dysphagia symptoms persisted. High-resolution esophageal manometry (HREM) confirmed the presence of persistent EGJ outflow obstruction despite prior dilation (Fig. 1), as reflected by an elevated mean integrated relaxation pressure (IRP) of 30.6 mmHg and elevated intrabolus pressure of 15.4 mmHg. Esophageal body peristalsis and upper esophageal sphincter motility were normal. Repeat balloon dilatation to 20 mm was performed and resulted in improvement in dysphagia.

In adults with persistent dysphagia after Nissen fundoplication despite normal endoscopic findings, HREM has been used to show an association between outflow resistance (measured by a longer length of distal esophageal high-pressure zone and increased IRP) and dysphagia compared to asymptomatic patients [1]. EDS patients commonly suffer from gastrointestinal disorders, such as gastroesophageal reflux disease, that require Nissen fundoplication. However, these

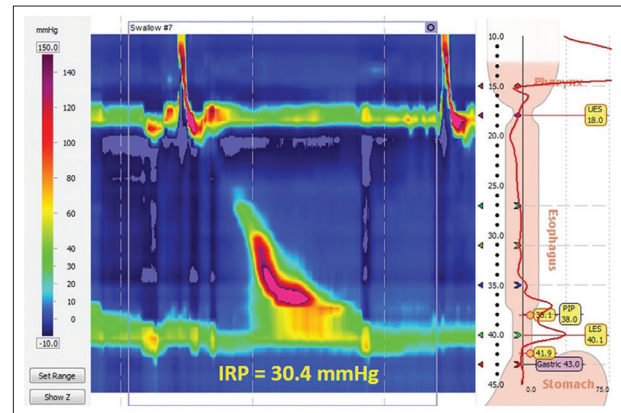


Figure 1 Representative example of a swallow with functional esophagogastric junction obstruction. Note the elevated integrated relaxation pressure at the esophagogastric junction and the slight compartmentalized pressurization in the distal esophagus during peristalsis

patients are at higher risk of endoscopy-related complications, including perforation from excessive dilatation [2]. Thus, HREM can be helpful to confirm EGJ obstruction post-fundoplication, especially when considering more aggressive dilation in high-risk patients [3]. The role of HREM in evaluating the status of children post fundoplication has not previously been reported.

References

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