



Peroral endoscopic myotomy in a child with Triple A syndrome (Allgrove syndrome)

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Triple A syndrome (AAA) or Allgrove syndrome is a rare, autosomal recessive disorder that usually manifests with 3 main cardinal symptoms: achalasia, alacrimation, and adrenal deficiency.¹ A mutation on chromosome 12q13 for the ALADIN protein must be confirmed for the diagnosis.²

Although pneumatic balloon dilatation and Heller myotomy have been reported as possible treatments for esophageal achalasia in Allgrove syndrome, limited experience with peroral endoscopic myotomy (POEM) in children with AAA has been reported.^{3,4} For POEM in children weighing >10 kg, no particular tools or settings are required; nevertheless, particular attention to insufflation-related adverse events is recommended

because their severity can be greater than in the adult population.

CASE REPORT

We report a case of a 7-year-old boy with 1-year history of persistent vomiting and failure to thrive who was referred to our tertiary center. His weight was 11 kg. High-resolution manometry was performed and revealed a typical pattern of type 1 achalasia (Fig. 1). An anesophagogram confirmed the bird-beak appearance of the esophagealgastric junction and contrast level (Fig. 2). Associated alacrimation and severe malnutrition were

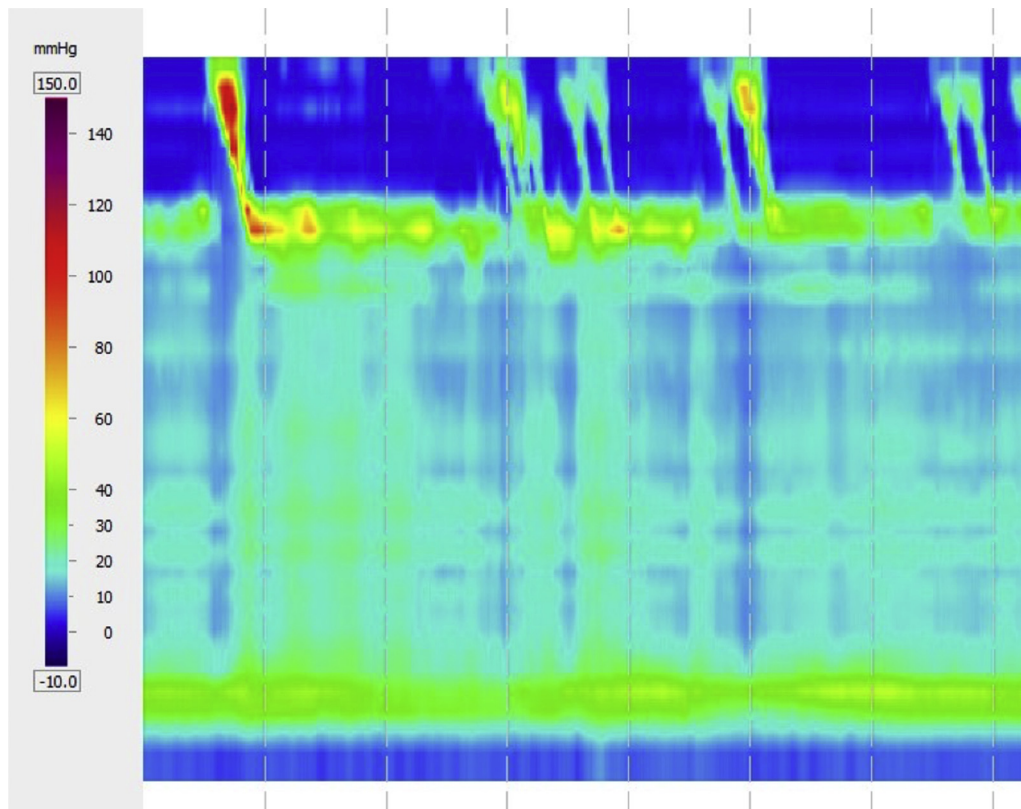


Figure 1. High-resolution manometry.

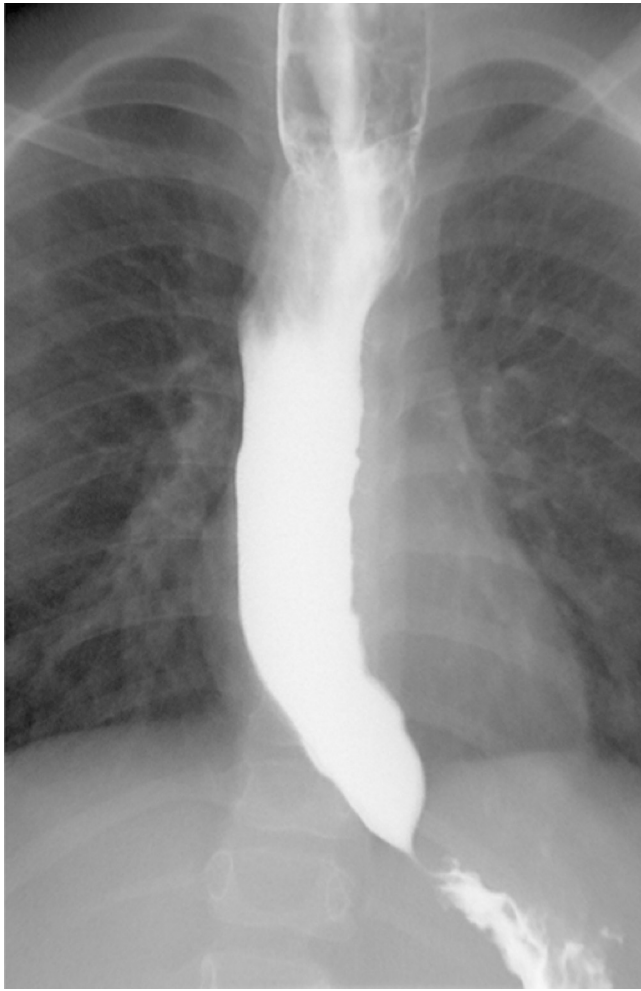


Figure 2. Esophagram before peroral endoscopic myotomy.

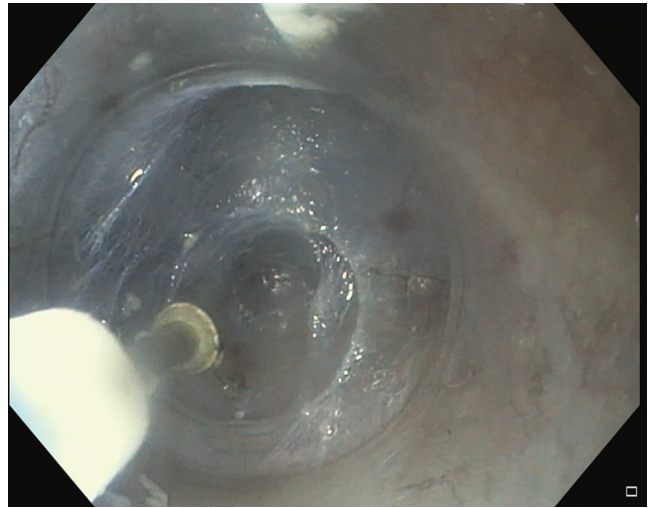


Figure 4. Peroral endoscopic myotomy procedure: tunnel creation.

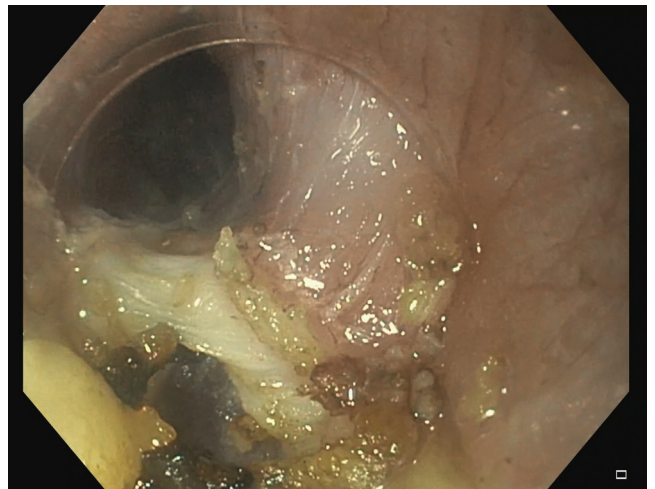


Figure 5. Peroral endoscopic myotomy procedure: myotomy.

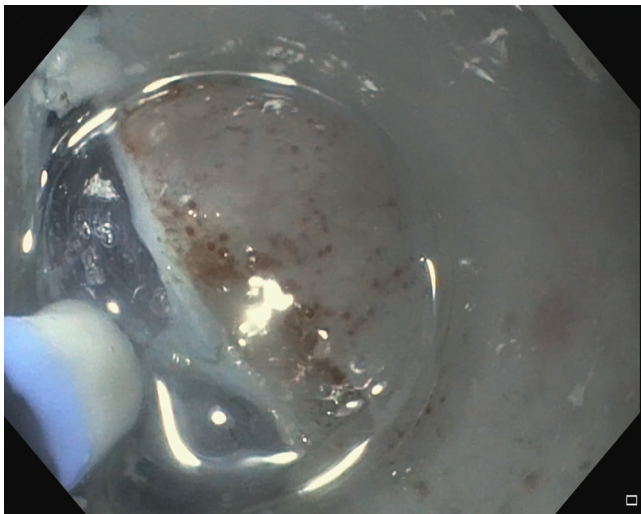


Figure 3. Peroral endoscopic myotomy procedure: mucosal incision.

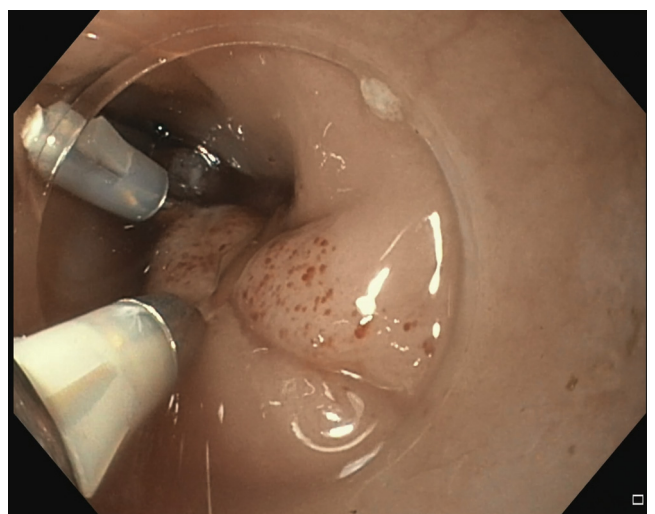


Figure 6. Peroral endoscopic myotomy procedure: closure with endoclips.



Figure 7. Esophagram after peroral endoscopic myotomy.

also noted, and an adrenocorticotrophic hormone stimulation test showed a low cortisol response. AAA was suspected, and genetic testing confirmed the diagnosis.

Periprocedural enteral nutrition was started, and POEM was performed by using a hydrodissection knife (HybridKnife T; Erbe Elektromedizin, Tuebingen) (Figs. 3-6, Video 1, available online at www.VideoGIE.org).

The procedure was uneventful, and the esophagram in the first postoperative follow-up confirmed the absence of esophageal leakage (Fig. 7). Progressive refeeding was started, and the patient was discharged without symptoms and with a nasogastric tube for supportive enteral nutrition. The tube was removed after 15 days.

Three months after the procedure, the patient had good tolerance for feeding and was experiencing recovery of the growth curve (6 kg weight increase), without any late adverse events (Eckardt's score = 0/12).

CONCLUSIONS

POEM was shown to be safe and effective in a child with AAA, a disorder that is reported to be sometimes refractory to conventional treatment.^{3,4} This procedure represents a definitive, minimally invasive treatment for esophageal achalasia.^{5,6} A high level of suspicion and adequate experience is recommended for AAA diagnosis, and long-term follow-up studies are awaited.

DISCLOSURE

All authors disclosed no financial relationships relevant to this publication.

Abbreviations: AAA, Triple A syndrome; POEM, peroral endoscopic myotomy.

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<https://doi.org/10.1016/j.vgie.2020.02.007>