# Rare Cause of Dysphagia in Children

Hussein Ahmed, MRCSEd, EBPS, Arab Board (Ped Surgery)*
Zafer Skef, American Board (Ped Surgery)** Martin T. Corbally, FRCS, FRCSI, FRCSEd***

**Dysphagia in children is generally caused by strictures secondary to gastroesophageal reflux disease or post-repair of trachea-esophageal atresia/fistula.**

**A four-year-old female presented with a history of dysphagia for solid food and vomiting of undigested food. Barium swallow showed esophageal duplication cyst, which was confirmed by esophagoscopy. Surgical resection was curative. Causes other than stricture or dysmotility should be considered in children with unrelenting dysphagia.**

***Bahrain Med Bull 2017; 39(3): 172 - 174***

Alimentary tract duplications are rare anomalies found in one out of every 4500 autopsies[1]. Usually, the duplication has variable symptoms, location and size. The origin of such anomaly remains unknown[2]. Fitz recognized the condition as intestinal duplication in 1884. Ladd in 1930 described the three essential features of esophageal duplication as follows: internal lining similar to alimentary tract; having smooth muscle layer; and proximity to a part of the alimentary tract[3]. The duplications are usually located on the mesenteric border[1].

The small intestine is frequently involved in gastrointestinal duplication; gastric, duodenal, rectal and thoracoabdominal presentations are uncommon[4]. Thoracic and thoracoabdominal duplications constitute about 10-20% of all duplication cysts.

The aim of this presentation is to report a four-year-old female with an esophageal duplication cyst.

## THE CASE

A four-year-old female presented to the Pediatric Surgery outpatient clinic complaining of dysphagia to solid food and non-bilious vomiting containing undigested food particles after every meal. There was no history of other gastrointestinal or upper respiratory tract symptoms.

Although the patient's weight was on the tenth percentile, she looked clinically well. A contrast esophagogram revealed a complex esophageal duplication with communication to the normal esophagus, see figure 1.

A flexible esophagoscopy with a trial of esophageal dilatation was performed; however, the dilator could not be passed through the connecting ostium and biopsy revealed stratified squamous epithelium with mild active inflammation, see figure 2.

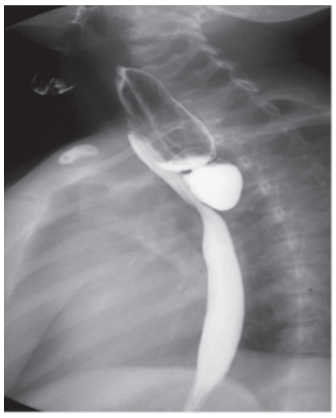

**Figure 1: Contrast Esophagogram Revealed a Blind Ending Esophageal Pouch Connecting to another Tubular Structure Anterior to the Pouch Which Extended Inferiorly to the Stomach**

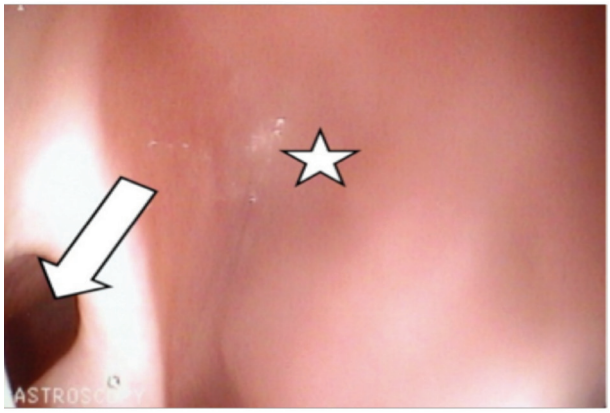

**Figure 2: Flexible Esophagoscopy Revealed Blind Ended Pouch (Star) and Side Communication with the Tubular Structure (Arrow)**

* Senior Registrar
Department of Pediatric Surgery
King Hamad University Hospital
** Consultant and Head of Department
Department of Pediatric Surgery
Security Forces Hospital
Riyadh, Saudi Arabia
*** Consultant and Chief of Medical Staff
Department of Pediatric Surgery
King Hamad University Hospital
The Kingdom of Bahrain
E-mail: martin.corbally@khuh.org.bh

A diagnosis of complex tubular esophageal duplication was made and a right extrapleural thoracotomy performed with resection of the duplication and end to end anastomosis, see figure 3.

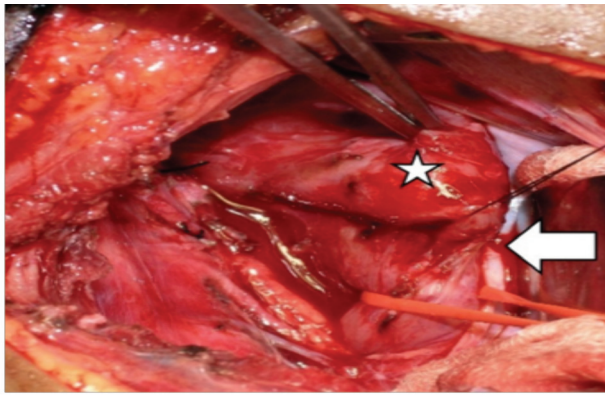

**Figure 3: Intra-Operative, the Blind Pouch (Star), Anterior Esophagus (Vessel Loop) and Communication between Both (Arrow)**

The patient had an uneventful postoperative course, and a contrast esophagogram performed seven days postoperatively revealed no leak or stricture, see figure 4.

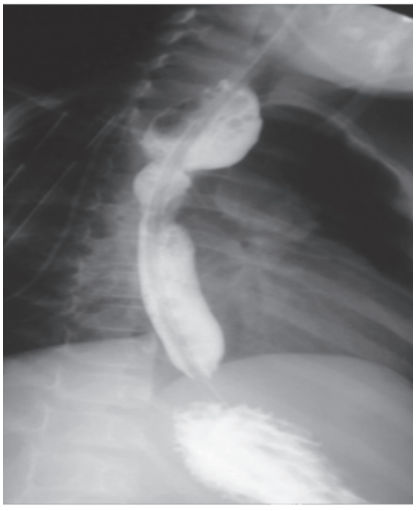

**Figure 4: Contrast Study Performed Seven Days Postoperative Revealed No Leak or Stricture**

Follow-up at four months revealed no dysphagia and significant weight gain to the 25th percentile. A degree of gastro-esophageal reflux was confirmed by a nuclear medicine study and responded to ranitidine.

## DISCUSSION

The incidence of esophageal duplications is approximately 1 in 8,200 live births, and it is approximately 10% to 15% of all GIT duplications[5,6].

Two types of esophageal duplication were recognized, cystic and tubular. The tubular form accounts for 5% to 10% of all cases and is frequently reported in the middle and lower esophagus and it tends to present in late childhood or adulthood[6,7]. The presentation may be asymptomatic or with complications, such as dysphagia, digestive hemorrhage, retrosternal pain, respiratory symptoms (recurrent pneumonia, stridor, respiratory distress)[5,8].

Esophageal duplications could be an isolated anomaly or associated with other GIT duplications, vertebral anomalies and esophageal atresia[5,8,9]. The diagnosis of esophageal duplication may be difficult. Contrast esophagogram is recommended for children with dysphagia or suspected esophageal anomalies. CT and MRI could be used for anatomical details and to look for associated vertebral anomalies[5,6,7]. Esophagoscopy could be used to visualize the communication between the esophagus and the duplication; in addition, it is used to obtain tissue for histopathology[6,7]. Malignant transformation is rare; adenocarcinoma was reported commonly; however, squamous carcinoma has also been reported[8,10].

Surgical resection has been recommended for symptomatic patients or where there is a risk of malignancy[6]. Careful consideration must be given to surgical intervention in asymptomatic patients given the nature of a thoracotomy and its inherent risks.

## CONCLUSION

**Esophageal duplication is a rare congenital anomaly. The cystic tube is the most common and few cases were reported with tubular duplication.**

**The diagnosis can be established with an esophagogram as well as CT with oral contrast. Some authors advise excision to avoid the possible risk of malignant transformation although rare and others preserve operative management only for complications. Howerver, surgery is needed when the condition is symptomatic.**

---

**Author Contribution:** All authors share equal effort contribution towards (1) substantial contribution to conception and design, acquisition, analysis and interpretation of data; (2) drafting the article and revising it critically for important intellectual content; and (3) final approval of manuscript version to be published. Yes.

**Potential Conflicts of Interest:** None.

**Competing Interest:** None.

**Sponsorship:** None.

**Acceptance Date:** 20 June 2017.

**Ethical Approval:** Approved by the Research and Ethics Committee, King Hamad University Hospital, Bahrain.

## REFERENCES

1. Puligandla PS, Nguyen LT, St-Vil D, et al. Gastrointestinal Duplications. J Pediatr Surg 2003; 38(5):740-4.
2. Bremer JL. Diverticula and duplications of the intestinal tract. Arch Pathol 1944; 38: 132-40.

3. Ladd WE, Gross RE. Abdominal Surgery of Infancy and Childhood. Philadelphia: W. B. Saunders Co, 1941; 289–290.
4. Iyer CP, Mahour GH. Duplications of the Alimentary Tract in Infants and Children. J Pediatr Surg 1995; 30(9):1267-70.
5. Chiengkriwate P, Patrapinyokul S, Rergkliang C, et al. Incomplete Duplication of the Esophagus: A Case Report. J Med Assoc Thai 2005; 88(8): 1123-7.
6. Estefan Ventura D, Reibscheid S, Colleoni R, et al. Surgical Images: Soft Tissue Tubular Duplication of the Esophagus. Can J Surg 2008; 51(3):205-6.
7. Khan N, Mansoor C, ul-Haq A, et al. Tubular Duplication of Esophagus: Rare Congenital Malformations Require Individualized and Innovative Procedures. Pak Med J Sci 2006; 22(4):465-7.
8. Tanaka K, Takeo N, Takuya I, et al. Excision of Esophageal Duplication Cyst in Children. Jikeikai Med J 2013; 60:73-6.
9. Ettayebi F. Association of Small Bowel Duplication and Esophageal Duplication: Treatment by Video Surgery. Pediatric Endosurgery & Innovative Techniques. 2002, 6(4): 281-4.
10. Jacob R, Hawkes ND, Dallimore N, et al. Case Report: Squamous Carcinoma in an Oesophageal Foregut Cyst. Br J Radiol. 2003; 76(905):343-6.