Case Report

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# Achalasia Cardia in Infant: A Rare Case Report

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**Abstract**

Oesophageal achalasia is a idiopathic disorder characterized by abnormal motility and failure of relaxation of lower esophageal sphincter, with its usual presentation in young adults. It is an uncommon disease in childhood and extremely rare under the age of one year. We are reporting a case of a 1 year female who presented with persistant vomitting and was diagnosed of having with Achalasia Cardia with barium swallow and was treated with Heller's Modified Cardiomyotomy.

**Keywords:** Achalasia Cardia; Heller's Cardiomyotomy.

## Introduction

Achalasia cardia is a disorder of normal mobilization of esophagus. It is characterized by aperistalsis of the body of the esophagus. The lower esophageal spincter fails to relax appropriately at the time of swallowing the food [1]. Achalasia cardia was first described by Sir Thomas Willis in 1674. He described esophageal dilation with whalebone in a patient who had difficulty in swallowing and a dilated esophagus. The term achalasia was first used by Hurst in the year of 1927, which implies absence of relaxation, i.e inadequate LES relaxation in the absence of distal mechanical obstruction [2]. The condition is very unusual in pediatric age group below 14 years of age and more particularly in infancy. the incidence of achalasia cardia has been estimated to be 0.5–1.0 in 100,000 persons per year, and the world wide prevalence has been estimated to be around 8 per 100,000. About 5% of all cases occur in children and fewer still being reported in infants [3].

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## Case report

A 1 year female first product of nonconsanguineous parents presented to us with regurgitation of feeds and each feed since 6 months. She used to regurgitate unaltered feeds immediately after ingestion. The vomitting was non projectile, nonbilious, not blood stained, containing food particles. Shehas past history of hospitalization two months ago for pneumonia. The antenatal, natal and post natal history was uneventful. The family histiry was insignificant. On general examination, the infant was underweight and anaemic with a weight of 6 kg (below the 5$^{th}$ percentile) and a length of 68 cm (below the 50$^{th}$ percentile). The systemic examination was normal.

Routine laboratory investigations were within normal limits. Barium swallow showed abrupt narrowing of cardio-oesophageal junction suggestive of Achalasia Cardia. Motility study could not be done because of non-availability of esophageal manometry facilities. Oesophagoscopy was done, which ruled out a stricture.

As the symptoms were persistant the patient was operated by Heller's Modified Cardiomyotomy by laproscopy approach. The Nasogastric tuube was

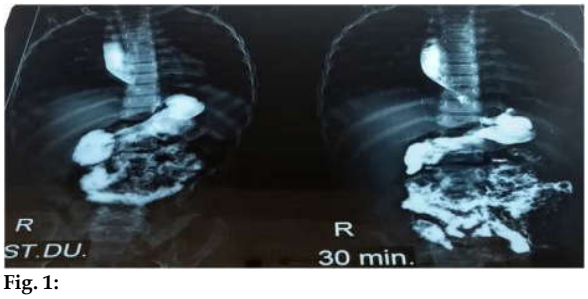


**Fig. 1:**

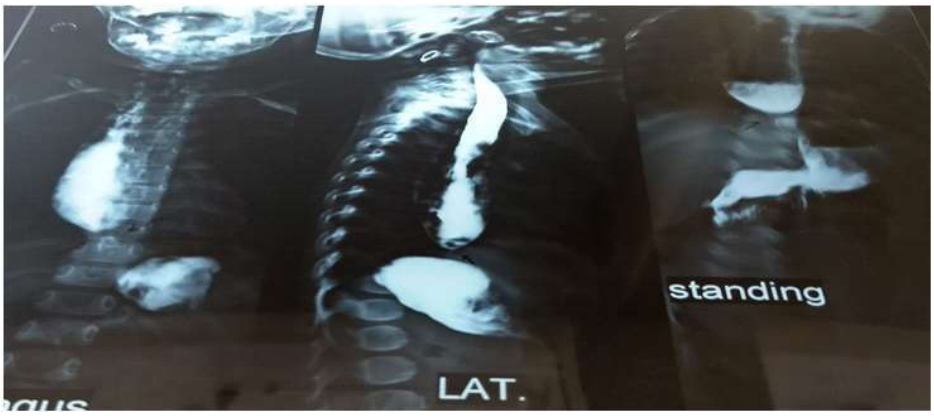


**Fig. 2:** Barium Swallow showing abrupt narrowing of Oesophageus.

removed on 10th post of day and oral feeding was resumed. The girl became asymptomatic in the post-operative period, and she started to gain weight with improvement of her general condition. (Fig. 1, 2).

## Discussion

Oesophageal achalasia is a rare motility disorder seen very less in children. Thomas Willis was the first scientist described achalasia cardia in year 1674. Incidence of achalasia has been estimated as 1 in 10,000 persondworld wide. Only 4-5% of patients with achalasia are diagnosed prior to 15 years of age [4].

Achalasia cardia is a motility disorder. It is idiopathic in nature. Relaxation failure of lower esophageal spinchter is the main pathophysiology of achalasia cardia. Absence of peristaltic wave leads to dilatation of the lower end of osophagus. This ultimately results in functional obstruction at the gastro-esophageal junction. The disorder can be classified on the basis of etiology. There is primary neurogenic abnormality associated with failure of inhibitory nerves supplying spincter causing progressive deration of ganglionic cells. And secondary to gastroesophageal reflux disease, Chagas disease or viral infections leading to deficiency of myentric plexus ganglionic cells. Allgrove and colleagues described a rare condition (Allgrove Syndrome) where achalasia cardia was associated with isolated glucocorticoid deficiency with alacrimiai.e absence of tears [5,6].

The clinical features of achalasia cardia are dysphagia, regurgitation of feeds, recurrent vomitting, growth failure, failure to thrive, retrosternal burning sensation, chest pain and recurrent lower respiratory tract infections. Vomitting of uncurdled milk after every feed is commonest classical feature of achalasia cardia. This is also a feature of GERD. During evaluation of children with persistent vomiting or failure to thrive the diagnosis of Achalasia should be kept in mind [7]. Plain radiograph of chest and abdomen may demonstrate the absence of the fundic air bubble gas due to absence of gas in stomach. As a diagnostic modality the barium swallow is done. It shows the features like.

- Dilated esophagus,
- Body aperistalsis with smooth narrowing of the distal esophagus and
- Esophago-gastric junction also described as “bird-beak” sign.

Endoscopic assessment is essential to rule out other causes of esophageal obstruction, e.g. congenital membrane and acquired stricture, webs, ringsetc [8].

The treatment is aimed to relieving the functional obstruction in distal esophagus anfdesophago-gastric junction. This helps in passage of food easily.

The gold standard for the management of achalasia cardia is a modified Heller’s oesophago cardiomyotomy with fundoplication by open or laparoscopic surgery. In this surgery, the muscular layer of lower end of esophagus is dilated.

Other modalities of treatment described in older children and adults with varying success rate are pharmacological and mechanical pneumatic dilatation of oesophageus. The pharmacological management includes drugs like smooth muscle relaxants like isosorbide dinitrate or calcium channel blockers (nifedipine). Injection botulinum toxin for smooth muscle relaxation of smooth muscles can also be used as a treatment modality [9].

We conclude our case report with aobseravation that cases of vomitting after each feed and dyaphagia

should be further evalauted for achalasia cardia.In the patients of older age group, though GERD is a more common condition than achalasia cardiaolder age, the latter condition should be kept in mind in the differential diagnosis in infants with symptoms of recurrent regurgitation, persistent vomiting, dysphagia and failure to thrive and investigated accodingly. An prompt recognition and prompt surgical intervention are necessary to accomplish a successful outcome.

## References


1. Patti MG, Fisichella PM, Perretta S, et al. Impact of minimally invasive surgery on the treatment of esophageal achalasia: a decade of change. J Am Coll Surg. 2003;196:698–703.
2. Reynolds JC, Parkman HP. Achalasia Gastroenterol Clin North Am. 1989;18:223–55.
3. Hamza AF, Awad HA, Hussein O. Cardiac Achalasia in Children. Dilatation or Surgery? Eur J Pediatr Surg. 1999;9:299–302.
4. Fisichella PM, Raz D, Palazzo F, et al. Clinical, radiological and manometric profile in 145 patients with untreated achalasia. World Journal of Surgery. 2008;32:1974–9.
5. Allgrove J, Clayden GS, Grant DB. Familial glucocorticoid deficiency with achalasia of the cardia and deficient tear production. Lancet. 1978;1:1284–6.
6. Wang L, Li YM, Li L. Meta-analysis of randomized and controlled treatment trials for achalasia. Dig Dis Sci. 2009;54:2303–11.
7. Thomas RJ, Sen S, Zachariah N, et al. Achalasia cardia in infancy and childhood: an Indian experience. J R CollSurgEdinb. 1998;43:103–4.
8. Thomas R J, Sen S, Zachariah N et al. Achalasia cardia in infancy and childhood: an Indian experience. J R CollSurgEdin 2008;43(2):103-4.
9. Carl-Christian A Jackson and Donald C Liu. Achalasia. In: Stefano Guandalini eds. Textbook of pediatric gastroenterology and nutrition. London: Taylor and Francis, 2004.pp.61-72.