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Achalasia Cardia in an Infant

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Esophageal achalasia is a neuromuscular disorder of unknown etiology characterized by abnormal motility and failure of relaxation of lower esophageal sphincter, with its usual presentation in fourth and fifth decades of life. It is an unusual lesion in childhood and extremely rare under the age of one year. We report a 7-month-old child with esophageal achalasia treated with modified Heller's esophagomyotomy.

Case Report

A 7-month-old male infant was admitted with complaints of regurgitation of feeds for three months, and respiratory distress off and on since then. The baby would

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effortlessly regurgitate every feed even in upright position within minutes of ingestion. He was earlier admitted a month ago and was treated for bronchopneumonia. The regurgitation was attributed to a heightened gastroesophageal reflux and he was discharged without any contrast or motility studies. The patient had no family history of the same symptoms. On examination, the infant was malnourished and anemic with a weight of 4.7 Kg (below the 5th percentile) and a length of 67 cm (below the 50th percentile). Examination of other systems was unremarkable.

Routine laboratory investigations were within normal limits. Barium swallow with cine-esophagogram revealed irregular and abnormal peristalsis, occasional skipping of peristaltic waves and inhibition of normal cardio-esophageal relaxation to the passage of the barium bolus *(Fig.* 2). Motility study could not be done because of non-availability of esophageal manometry facilities at our institution. An esophagoscopy was done, which ruled out a stricture. As symptoms persisted, the infant underwent a modified Heller's anterior esophagocardiomyotomy through abdominal route with short-wrap Nissen's fundoplication. The nasogastric tube was removed on fourth post operative day and the child could resume normal feeding from the next day. Barium swallow studies done on seventh post operative day showed a free passage of barium into the stomach; no gastroesophageal reflux could be illustrated

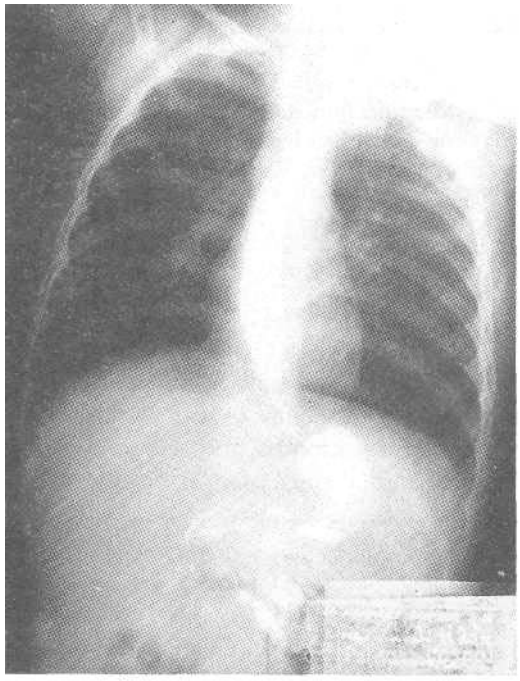

Fig. 1. Preoperative barium swallow revealing dilation of the esophagus with characteristic 'beaking' of the esophagogastric junction. There was marked delay of passage of barium into the stomach.

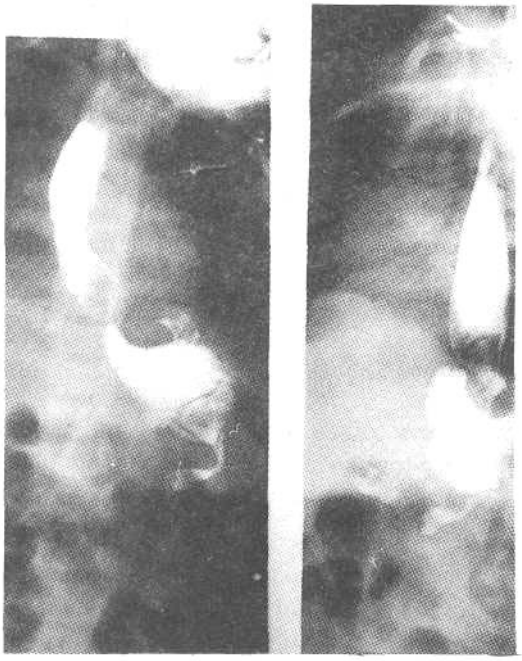

Fig. 2. Barium swallow performed a week after Heller's esophagomyotomy. Note ballooning of esophageal mucosa and definite change in the contour of the terminal part of the esophagus and prompt passage of barium into the stomach.

even in head-down position *(Fig. 2)*. His follow-up examination has revealed dramatic relief of symptoms and satisfactory weight gain.

Discussion

Esophageal achalasia is a relatively rare problem in children with less than 250 cases reported in the world literature till 1988(1). A search of English literature revealed only 30 cases of achalasia in infancy reported till date(2-ll). The first case was probably reported by King in 1953(2). This child was diagnosed at 6 months of age. After failure of therapy with anticholinergic medication, a Heller esophagomyotomy was performed at age of 9 months with excellent symptomatic improvement.

In view of the rarity of achalasia in infants, no one physician or institution can have an extensive experience with the disease; in fact the two largest institutional series ever reported comprised only 4 patients each(9,10). This results in delayed diagnosis in many instances. A recent global survey of esophageal achalasia in childhood revealed that although 18% of children had symptom onset during infancy, only 6% of these patients were identified as having achalasia during infancy(11).

This calls for a high index of suspicion and an awareness that this entity may affect the infants as well. The usual symptomatology include dysphagia, regurgitation, pulmonary infection and weight loss. Retardation of growth and development as well as the severity of pulmonary symptoms have been noted to be much more profound in this age group.

In most infants, a barium swallow with cine-esophagogram will reliably provide the diagnosis. Although motility studies are desirable for an absolute evidence for the diagnosis, even a very recent worldwide survey revealed that such studies were performed in less than 30% of patients in the best of centers(11). These studies should ideally be performed in all cases if sophisticated means of evaluating esophageal motility are available.

Medical therapy has not provided long-term relief in these patients. Esophageal dilation with pneumatic dilators has been used in older children, but the disadvantages and risks of the procedure do not permit its used in infants(3).

The treatment of choice is modified Heller esophagomytomy; it is safe and has excellent long term results. A trans-abdominal myotomy along with anti-reflux procedure has been reported to give the best results(11), although some surgeons advise a transthoracic route(8). Others feel that antireflux procedures need not be added in children because postoperative reflux esophagitis is not a significant problem in this age group(3). The operation has invariably resulted in dramatic relief of symptoms, satisfactory weight gain and disappearance of pulmonary symptoms.

In conclusion, achalasia cardia should be kept in mind in the differential diagnosis of intractable regurgitation and dysphagia in infants. An early diagnosis would result in prompt surgical relief.

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