

CHAPTER 50

ACHALASIA

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Introduction

Achalasia is an uncommon oesophageal problem in children and consequently can be slow to diagnose. It is a condition of unknown aetiology characterised by poor or absent motility of the body of the oesophagus and the failure of the lower oesophageal sphincter to relax. Infants may present with failure to thrive, pulmonary symptoms, vomiting, dysphagia, and growth retardation, but due to the rarity of the condition, many children present late with any of the above symptoms as well as with significant nutritional compromise. In the absence of an identifiable cause, treatment is directed at symptoms.

Demographics

The prevalence of this condition is unclear; a worldwide survey of paediatric surgeons with experience in achalasia in childhood, however, documented information concerning 175 children.¹ The condition appears to be more common in boys, and familial cases do exist but are rare. Regurgitation of food and dysphagia are common. Profound weight loss is a significant feature and is the most frequent symptom, with 18% of patients presenting in infancy but only 6% diagnosed during that time frame. Fewer than 5% of the cases are diagnosed in childhood. The mean age for diagnosis in adult life is 45 years, and the incidence is approximately five new cases per million population per year.

Aetiology/Pathophysiology

Achalasia is a motor disorder of unknown aetiology characterised by the failure of relaxation of the lower oesophageal sphincter along with poor peristalsis of the oesophagus. The three manometric requirements of the diagnosis of achalasia are: (1) hypertension of the lower oesophageal sphincter, (2) incomplete or absent relaxation of the lower oesophageal sphincter, and (3) weak or absent peristalsic contractions in the body of the oesophagus after swallowing. Transient achalasia can be due to corrosive ingestion and ganglion cell damage, as seen in *Trypanosoma cruzi*, or Chagas disease.²

Poor clearance of foodstuffs and saliva produces stagnation with a consequent structural change in the calibre of the oesophagus and a change in the mucosal integrity with resultant oesophagitis. Overflow and aspiration are responsible for the respiratory symptoms associated with this condition. The condition is associated with a late incidence of oesophageal carcinoma.³

History

Multiple, quite disparate presentations make achalasia a difficult diagnosis in some children. The diversity of symptomatology, ranging from foregut symptoms to advanced pulmonary sepsis as a consequence of aspiration, can distract from the diagnosis. The relative rarity of the condition compounds the situation. Nevertheless, typical features are pain on swallowing, dysphagia, vomiting, failure to thrive, and respiratory symptoms including chronic lung sepsis with wheezing. A family history of achalasia is present on rare occasions.

Physical Examination

Similarly, physical examination covers a range of findings from mild weight loss to an advanced pulmonary sepsis and severe malnutrition. Achalasia can be associated with adrenocorticotrophic hormone (ACTH)-resistant adrenal insufficiency and alacrima (an absence of tears).⁴ Hence, presentation may have the features of achalasia, addisonianism, and alacrima (triple A, or Allgrove syndrome). All other physical findings are nonspecific.

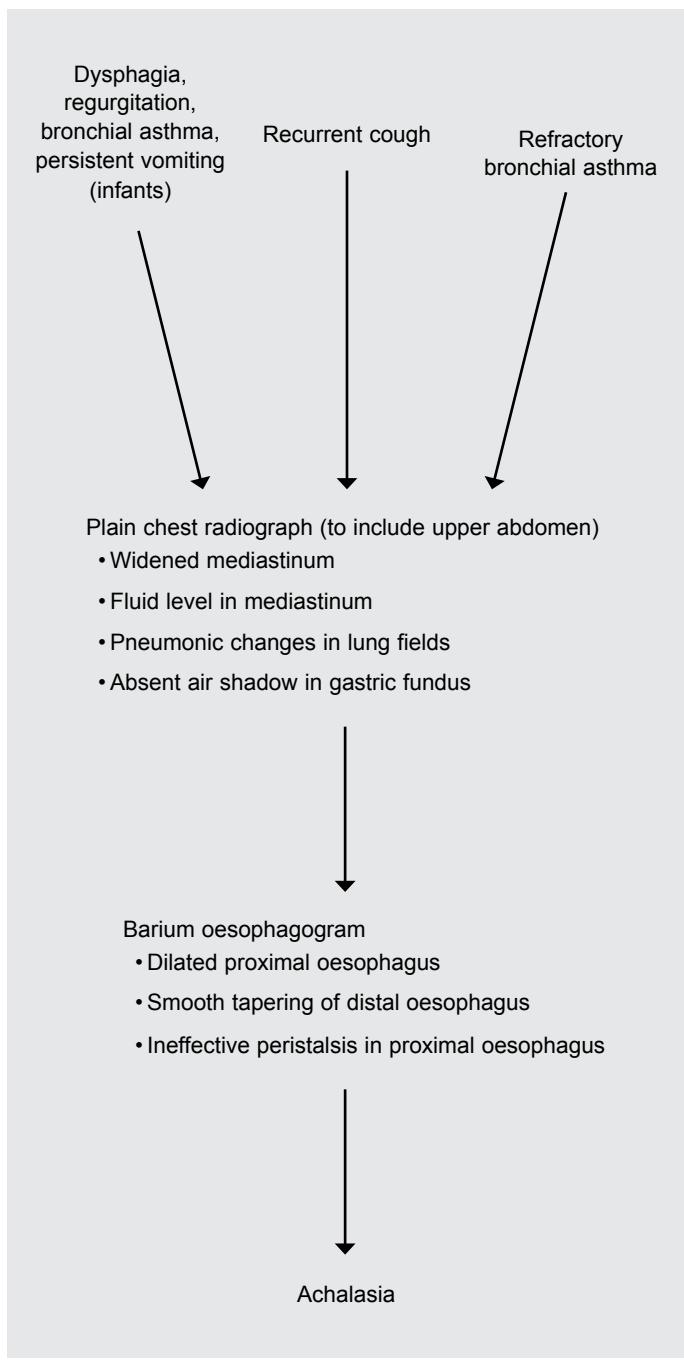
Investigations

Chest x-ray can demonstrate an air-fluid level in the oesophagus with a characteristic absence of the gastric air bubble on an erect film. In the absence of standard facilities (as is the case in most centres in Africa and other developing countries), a high index of suspicion as well as appropriate use and interpretation of a barium swallow are necessary for prompt diagnosis of achalasia. The barium swallow typically demonstrates a dilated oesophagus above a narrowing “rats tail” or “bird’s beak” (see Figure 50.1).

Manometry is the confirmatory investigation with a resting lower oesophageal pressure in excess of 15 to 20 mm Hg and a failure of



Figure 50.1: Typical barium swallow in an oesophageal achalasia patient.



Source: Adapted from Chirdan LB, et al., Childhood achalasia in Zaria Nigeria. *E Afr Med J* 2001; 78:497–499.

Figure 50.2: Algorithm for the diagnosis of oesophageal achalasia in children in countries with limited diagnostic facilities.

the lower oesophageal sphincter to relax on swallowing, along with an absence of peristalsis being the confirmatory features. Endoscopy excludes other causes of organic obstruction or infection in the distal oesophagus.

An algorithm for the diagnosis of achalasia in children in the developing countries and countries with limited diagnostic facilities, seen in Figure 50.2, has been suggested by Chirdan et al.⁵

Management

The three therapeutic modalities of achalasia management are pharmacological treatment, pneumatic dilatation, and esophageal myotomy. An overriding consideration is the nutritional status of the child at outset, and due to the unpredictability of the efficacy of any of the procedures, restoration of good nutritional status is paramount. That can be effectively achieved with nasogastric feeding.

Pharmacological Treatment

Per-endoscopic four quadrant injection of botulinum toxin (100 unit) has been shown to substantially reduce the lower oesophageal sphincter pressures (preinjection, 44 mm Hg; postinjection, 16 mm Hg) with substantial sphincter relaxation. The clear disadvantage of this technique is the need for its repeated application.⁶

The calcium channel blocker nifedipine has been used with good effect in adults, but its use has been limited to those adults unfit for any other form of intervention.⁷

Pneumatic Dilatation in Children

The aim of pneumatic dilatation in children is to relieve obstruction by a gentle disruption of the lower oesophageal sphincter. This procedure is carried out under anaesthesia using radiological and manometric control. An upper gastrointestinal contrast study is performed following the procedure to exclude oesophageal perforation. The number and periodicity of repeat dilatations is patient-specific. It is the recommended initial therapeutic method of choice in older children, but if initial attempts fail to provide satisfactory relief, surgical treatment is then indicated. Only 25% of children show significant improvement with pneumatic dilatations alone, and few children under the age of 9 years have responded to this treatment.⁸

Oesophageal Myotomy

The operative approaches include an abdominal approach, an open thoracic approach, or—more recently—an endoscopic approach through the chest or abdomen.⁹ These approaches share the same objective of performing a modified Heller's procedure, which includes a myotomy of the lower oesophagus, preserving the integrity of the oesophageal mucosa. (Heller initially performed his procedure on both sides of the oesophagus; that is, a double oesophagomyotomy.) This is often accompanied by a subsequent fundoplication to prevent gastro-oesophageal reflux frequently produced by the Heller's procedure, and some authorities restrict the myotomy to 0.5 cm on the cardia, thereby reducing the need for antireflux procedures. In those patients for whom transabdominal or a laparoscopic oesophagomyotomy has been performed, this is very frequently accompanied by an antireflux procedure,⁹ although some recent authors evaluating the long-term results have suggested that symptomatic improvement in the longer term can be equally effectively obtained without an antireflux procedure.¹⁰

Surgical Procedure

Oesophageal myotomy through the abdominal route is described in detail in this section. An antireflux procedure is usually not necessary because reflux is rare among African children.

Position and Anaesthesia

General anaesthetic with endotracheal intubation is used, with precaution taken to avoid aspiration of oesophageal contents, especially during induction.¹¹ The child is placed in the supine position. In centres where paediatric endoscopes are available, preoperative oesophago-

goscopy is used to ensure complete evacuation of retained food and secretions from the oesophagus. A nasogastric tube of appropriate size is passed into the stomach.

Incision and Exposure

An upper midline abdominal incision extending from the xiphisternum to the umbilicus is used. The incision could be extended to just below the umbilicus to give more access. Once the peritoneum is opened, adequate exposure of the abdominal oesophagus is gained by retracting the left lobe of the liver with a wide liver retractor anterosuperiorly. Alternatively, the left triangular ligament is divided in its avascular plane, and the left lobe of the liver is retracted towards the midline.

Mobilisation

The next stage of the operation is the exposure of the oesophageal hiatus and mobilisation of the distal oesophagus. For those who want to add a floppy Nissen fundoplication, the fundus of the stomach is freed by ligating and dividing the short gastric vessels in the gastosplenic ligament. The phrenicoesophageal membrane is then stretched by applying downward traction on the stomach and retracting the diaphragm upwards. The avascular membrane is incised with a scissors, exposing the muscularis of the oesophagus, and the anterior vagus nerve is seen on the oesophagus. The exposed distal oesophagus is now encircled by using a combination of blunt and sharp dissection, taking care not to injure the posterior vagus nerve. A rubber sling or nylon tape is then placed around the distal oesophagus, and 5–8 cm of the oesophagus is exposed by using blunt dissection.

Myotomy

The myotomy is done on the anterior oesophagus extending to 1 cm of the fundus of the stomach. An incision is made in the distal oesophagus; the divided muscle is then parted with a blunt haemostat, exposing the mucosa. The muscle is separated from the underlying mucosa by pledget dissection. This is continued to about at least half the circumference, freeing the oesophagus from the constricting muscle. This is extended to 1 cm of the fundus. The stomach and oesophagus are then distended with air from the nasogastric tube, and the exposed mucosa is carefully inspected for perforation. In the event of a perforation, the mucosal defect is closed with polyglycolic acid sutures. Finally, the hiatus is narrowed posteriorly by placing deep sutures through the diaphragmatic crura, leaving sufficient space along the oesophagus that can admit the tip of the finger.

Wound Closure

The abdominal wound is closed in layers with nylon sutures. The skin is closed with subcuticular suturing.

Laparoscopic Approach

When a laparoscope is available, the pneumoperitoneum is established through standard procedure and a 5–10 mm camera port is placed through the infraumbilical skin crease. Three to four additional 5-mm ports are placed to permit retraction and dissection of the oesophageal hiatus. The same principles are adhered to as in the open operation, with mobilisation of the distal oesophagus under direct vision. Following circumferential encircling of the oesophagus with the umbilical tape and with caudal retraction on this tape, the distal oesophagus is dissected and

exposed for several centimetres. A myotomy is performed on the distal oesophagus vertically with extension just distal to the cardio-oesophageal junction. The muscle is elevated on either side of the myotomy with graspers anchoring the edge of the muscle and with the dissection of the muscle from the mucosa performed with endoscopic scissors.

Postoperative Care

Intravenous fluid is stopped after 3–4 days, and the nasogastric tube is removed by day 4.

The most worrying postoperative complication is leakage from an undetected perforation of the oesophageal mucosa, making a subsequent contrast study a requirement of the postoperative period.

Prognosis and Outcomes

The vast majority of patients have an immediate and lasting benefit from their surgery. Failure of the initial operation should be managed by an attempt at a second myotomy (on the contralateral side of the oesophagus), but more than 80% of patients have long-term relief of symptoms and resumption of appropriate growth following the first operation.

Evidence-Based Research

Due to the rarity of achalasia in children, no prospective studies exist that compare the various modalities of treatment and their long-term outcome in Africa. Guidance is derived mainly from retrospective experiences. Table 5.1 presents evidence-based research using Heller's procedure.

Table 5.1: Evidence-based research.

Title	Evaluating long term results of modified Heller limited esophagomyotomy in children with esophageal achalasia
Authors	Vaos G, Demetriou L, Velaoras C, et al.
Institution	Second Department of Pediatric Surgery, P and A Kyriakou Children's Hospital, Athens, Greece; First Department of Pediatric Surgery, P and A Kyriakou Children's Hospital, Athens, Greece; Department of Pediatric Surgery, Penteli General Children's Hospital, Athens, Greece
Reference	J Pediatr Surg 2008; 43:1262–1269
Problem	The role of modified transabdominal Heller's myotomy in the long-term outcome of children with achalasia.
Intervention	Heller's limited oesophagomyotomy.
Comparison	To evaluate long-term symptom relief after intervention using subjective outcome, Barium esophagogram, esophageal pH, and oesophageal manometry.
Outcome/effect	Excellent to good results observed in 93.3% of patients, late Barium esophagogram showed a significant decrease in oesophageal diameter compared to preoperative values ($p < 0.01$), and the late oesophageal manometry showed a significant decrease of lower oesophageal sphincter pressure ($p < 0.05$).
Historical significance/Comments	This report, although retrospective and in a small population of children, showed that the long-term outcome of children treated with modified Heller's myotomy can be quite satisfactory.

Key Summary Points

1. Achalasia is often diagnosed late in childhood.
2. An awareness of the condition as it affects children is key to diagnosis.
3. A combination of failure to thrive, respiratory symptoms, and food aversion should prompt investigation of upper gastrointestinal (GI) tract.
4. Heller's myotomy (open or laparoscopic) should follow correction of nutritional deficit, with good results being expected.
5. Strict attention must be paid to ensuring mucosal integrity.

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