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ABSTRACT

Objectives: **To study the presentation, management and outcome of achalasia of the oesophagus in children.**
Design: **A retrospective case study.**
Setting: **Ahmadu Bello University Teaching Hospital, Zaria, Nigeria.**
Subjects: **Seven children managed for achalasia of the oesophagus in a period of 19 years.**
Results: **There were six boys and one girl. The median age at presentation was ten years (range three months to fifteen years). The median duration of symptoms before presentation was 20 months (range: 2 - 24 months). Three children were treated on several occasions for pneumonia before the diagnosis was made. Dysphagia, vomiting and regurgitation were the main presenting symptoms. Diagnosis was by barium oesophagogram. Six had modified Heller's myotomy via celiotomy and a Thal fundoplication in a three month old. Transthoracic Heller's myotomy was done in the seventh child. There was complete resolution of symptoms in six children. One patient had post-operative retrosternal pain, which was controlled by oral nifedipine.**
Conclusion: **Achalasia of the oesophagus should be excluded in children with recurrent chest infection. Modified Heller's myotomy without antireflux procedure via the abdominal route is effective in relieving symptoms of achalasia in children.**

INTRODUCTION

In achalasia of the oesophagus there is failure of relaxation of the lower oesophageal sphincter during swallowing, leading to stasis and progressive oesophageal dilatation. The aetiology is unknown(1). It is a common benign oesophageal condition in adults in Zaria, Nigeria(2) and probably the commonest cause of dysphagia in eastern Nigeria(3). It is, however, uncommon in children accounting for less than five per cent of reported cases(4). While much has been written on achalasia in children from developed countries, reports focussing on children from tropical Africa are scanty. This is a retrospective review of childhood achalasia seen over a 19-year period at the Ahmadu Bello University Hospital, Zaria, northern Nigeria.

MATERIALS AND METHODS

Seven children with achalasia were treated at our hospital between January 1981 and December 1999. Their case records, operation notes and discharge summaries have been retrospectively reviewed. Three of the children were part of earlier reports on achalasia generally in Zaria(2,5).

RESULTS

There were six boys and one girl. The median age at presentation was ten years (range: three months to fifteen years). The median duration of symptoms before presentation was 20 months (range two to twenty four months). Only one child presented as an infant. The other six children presented after five years of age. One child had presented at six years of age but the diagnosis was not made until he was 21 years.

Symptomatology: The symptoms and signs at presentation are shown in Table 1. Dysphagia was present in all children. Regurgitation and/or vomiting was seen in five children. The three-month old infant had excessive vomiting with dehydration on presentation which was corrected by intravenous fluid therapy before operation. Three children had received treatment on several occasions for pneumonia (two children) or asthma (one child) before referral. Three children had bilateral parotid swelling which regressed after treatment.

Evaluation: Plain chest roentgenography showed widened mediastinum in six children and evidence of chest infection in addition in three children. Barium swallow was diagnostic in all children showing a dilated oesophagus with tertiary peristaltic waves, absent primary peristalsis in the body of the oesophagus and smooth narrowing of the cardio-oesophageal junction. Oesophagoscopy in four children revealed dilated oesophagus and tight oesophago-gastric junction. There was no evidence of oesophagitis in any patient. One patient had bronchoscopy in addition which showed purulent discharge from the trachea. All had preliminary blood count, urea and electrolytes and haemoglobin level. In two children, the haemoglobin was <10gm/dl, necessitating blood transfusion. One of them had hookworm infestation for which he received antihelminthics.

Table 1

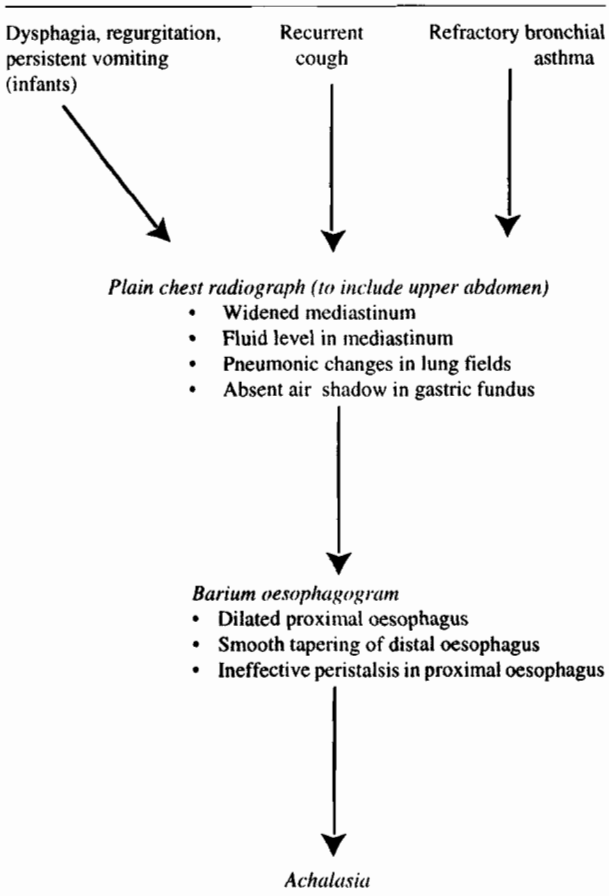
Features of achalasia of the oesophagus in seven children

S/No	Age (years)	Sex	Duration of symptoms (months)	Presentation	Treatment	Duration of follow up (months)	Outcome
1	13	M	22	Dysphagia, regurgitation, parotic swelling and weight loss.	Transthoractic Heller's myotomy	18	Excellent
2	10	M	24	Dysphagia, vomiting, pneumonia, weight loss	Heller's myotomy (trans-abdominal)	20	Good
3	6	M	20	Recurrent chest infection, dysphagia regurgitation(diagnosis made at 21 years of age)	Heller's myotomy / (trans-abdominal)	6	Excellent
4	10	M	20	Dysphagia, respiratory symptoms, parotid swelling, regurgitation	Dilatation initially Heller's myotomy (trans-abdominal)	15	Good
5	15	M	15	Dysphagia, vomiting, retrosternal pain	Heller's myotomy (trans-abdominal) postoperative, nifedipine 10mg before meals	180	Moderate
6	10	M	8	Dysphagia, weight loss parotid swelling, anaemia, hookworm infestation	Heller's myotomy (trans-abdominal)	6	Excellent
7	3 mths	F	2	Vomiting, dysphagia, failure to thrive, anaemia	Heller's myotomy+ Thal fundoplication(trans-abdominal)	3	Excellent

mths = months

Figure 1

Algorithm for the diagnosis of achalasia of the oesophagus in children in a developing country



Histopathology: Two oesophageal muscle biopsy specimens were examined histologically; in one, there were no ganglion cells, while ganglion cells were present in that from a three month old child.

Treatment: One child had oesophageal dilatation as initial treatment. He had Heller's myotomy six months later as symptoms persisted. All seven children had modified Heller's myotomy. Six were through a celiotomy with a Thal fundoplication in addition in a three month old girl and one via the thoracic route. The myotomy extended 3-8 cm on the anterior surface of the distal oesophagus and 1-2 cm onto the anterior wall of the stomach. There was iatrogenic oesophageal mucosal perforation in three children which were repaired. All the three children with chest infection were treated with antibiotics.

Outcome: Complications of surgery included mucosal perforation in three children treated by primary suturing at operation. Vomiting occurred in two children lasting 48 hours after operation and was treated conservatively. One patient had retrosternal pain, a month after discharge from the hospital. Barium studies showed no oesophageal stenosis or gastro-oesophageal reflux. This symptom was relieved by a short course of nifedipinde. The children were followed up for a median duration of 15 months (range six months to five years). The outcome based on the Vantrappen and Hellemann classification(6) was excellent/ good in six children and moderate in one child.

DISCUSSION

In this report, the patients were predominantly boys as in other reports(7,8). The median age at presentation,

onset of symptoms and duration of symptoms before presentation are also similar to reports from other parts of the world(7,9). In one report on achalasia in 175 children from twelve countries, regurgitation was the predominant symptom in 83%(8). The predominant symptoms in our patients were dysphagia, regurgitation and vomiting. Three patients in this report had been treated on several occasions for pneumonia or bronchial asthma. Parotid enlargement, which is a common feature of achalasia in adults in tropical Africa(2,3,5) was present in two patients and may be due to chronic undernutrition. Barium oesophagography was the main diagnostic investigation in this report. Though oesophageal manometry is the current standard diagnostic procedure in achalasia, we have continued to rely on barium oesophagography due to lack of facilities for manometry. In the absence of standard facilities, a high index of suspicion and appropriate use and interpretation of barium oesophagography is necessary for prompt diagnosis (Figure 1). The aim of treatment in achalasia is relief of the obstruction at the oesophagogastric junction. Medical treatment with long acting nitrates and the calcium channel blocker, nifedipine have not given long term relief of symptoms but nifedipine may be useful as initial treatment while definitive treatment is planned(10,11). In one of our patients, nifedipine was useful in relieving post Heller's myotomy retrosternal pain. Oesophageal dilatation gives good results in adults(6) but the results in children is poor(12). One report, however, found pneumatic dilatation to be as effective as surgery in children(13). Dilatation was not effective in one of our patients who had it as initial treatment. Modified Heller's myotomy remains the most widely used definitive treatment in children. Though some reports have recommended antireflux procedures to prevent gastro-oesophageal reflux after modified Heller's myotomy(14,15), six of our patients did not have antireflux procedure but none developed gastro-oesophageal reflux. A Thal fundoplication was added in the infant due to excessive vomiting at presentation. It would appear that antireflux procedures might not be necessary in all children.

Childhood achalasia is uncommon in our environment as in other parts of the world. The features are similar but diagnosis is often delayed due to lack of suspicion and absence of standard diagnostic facilities. Use of the proposed algorithm should lead to earlier recognition and treatment in environments similar to ours.

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