Oesophageal achalasia in children: report on 19 cases from three different institutions

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ABSTRACT

Nineteen cases of oesophageal achalasia in children have been collected from three different pediatric institutions. Main clinical, diagnostic and therapeutic aspects of this disease are analyzed and reported. All patients underwent operation through an abdominal approach and oesophagomyotomy was always associated to an antireflux procedure. This latter was realized by different techniques without significative difference in long term follow-up. Good results were obtained in 17 patients while in one case a light dysphagia and in the other case a light GER resulted.

RIASSUNTO

Vengono presentati diciannove casi di acalasia esofagea in età pediatrica trattati in tre differenti istituti di chirurgia pediatrica. I principali aspetti clinici, diagnostici e terapeutici vengono presi in esame. Tutti i piccoli pazienti sono stati sottoposti ad intervento chirurgico per via addominale e l'esofagomiotomia è sempre stata associata ad una plastica antireflusso. Quest'ultima è stata realizzata con tecniche diverse senza differenze significative al follow-up. In 17 casi si sono raggiunti buoni risultati mentre in un caso era ancora presente una lieve disfagia ed in un altro caso un lieve reflusso gastro-esofageo.

KEY WORDS

Achalasia - Oesophageal myotomy

Achalasia is a functional disorder characterized by a motility disturbance of the distal oesophagus and failure of the lower sphincter to relax in response to swallowing. The etiology is still unknown. Theories have included degeneration or decrease of parasympathetic postganglionic neurons of the intramural plexus, involutional process secondary to a lesion affecting the extrinsic vagal fibers or the vagal nuclei (1). In pediatric age, particularly in early onset cases, the hereditary factors should be considered (1, 2). Data obtained from the literature show that achalasia is an uncommon disease in pediatric age; moreover it is symptomatic only in 5% of cases under 14 years of age (3-5).

The clinical onset of achalasia is generally obscure and symptoms are not always pathognostic. In the attempt to diagnose early cases, this affection has always to be taken into account in the differential diagnosis of recurrent vomiting and regurgitation in presence of weight loss, dysphagia and, sometimes, bronchopulmonary infections which cannot be easily interpreted. Moreover, this affection can present in the newborn with asphyxia, pallor, tachycardia and "ab ingestis" cyanosis. In children over two years of age, dysphagia is a prevailing symptom and presents as "paradoxical", i.e., it becomes manifest during ingestion of liquids whereas solid meals are allowed to pass into the stomach, because they stimulate the sensitive endings to cause reflex opening of the cardias sphyncter (6).

Some authors (7, 8) classify symptoms by dividing them into three main stages. The first one is characterized by painful dysphagia and postprandial regurgitation without any change in oesophageal caliber; in the second, so called "compensation" stage, a mitigation of symptoms are referred, whereas an increase in the oesophageal size occurs. In the third, the "decompensation" stage, a recurrence of dysphagia and a remarkable dilatation of the oesophagus are observed. Postprandial regurgitation is generally encountered and sometimes this is voluntary, in order to relieve retrosternal pain.

In the present paper the experience of three different institutions on a total of 19 cases of oesophageal achalasia in pediatric age is reviewed.

PATIENTS AND METHODS

From 1975 to 1989 nineteen children with achalasia have been treated in three different institutions and respectively: 8 cases at the Gaslini Children Hospital of Genoa, 6 at the Division of Pediatric Surgery of the 2nd School of Medicine of Naples and 5 at the Division of Pediatric Surgery of the Pescara Hospital. Ten were male and 9 female. The patient's age ranged from 2 to 14 years (mean: 8 years). Preoperative diagnostic investigations included: in all cases a contrast oesophagogram, manometry in 10 cases, endoscopy in 7 cases and 24 hours pH-monitoring in 3 cases. No patient underwent forceful pneumatic dilatation of the lower oesophageal sphincter and the primary treatment in each case consisted of an oesophagomyotomy associated in 18 cases to an antireflux procedure (another case underwent an Allison plasty when was reoperated for relapse of dysphagia due to insufficient myotomy). This latter was realized by different techniques: Allison (three), Nissen (four), Thal (three), Lortat Jacob (four), Pellerin (five). In 6 instancies the extent of the myotomy as well as the antireflux procedure were guided by intraoperative manometry.

RESULTS

The main clinical data concerning the 19 patients are summarized in table 1. The delay between the onset of symptoms and definitive diagnosis ranged from 1 to 36 months (mean: 13

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months). The symptoms were in all 19 cases initially digestive (dysphagia, vomiting, frequent regurgitation, retrosternal pain, etc.) and in 7 cases (37%) successively also respiratory (recurrent infections). In 13 patients (68%) an important weight loss (> 20%) occurred.

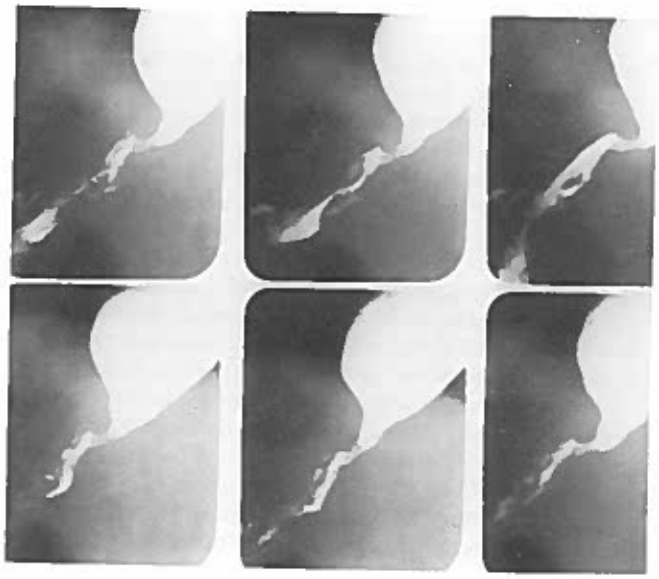

Figure 1: Seried oesophagogram: it is evident the difficult and narrowed passage of the contrast through the distal oesophagus.

In one case an associated asymptomatic double renal system was present. The most important diagnostic investigation was the contrast oesophagogram which resulted diagnostic in all cases (Fig. 1, 2, 3). Manometry invariably demonstrated absence of peristalsis, uncoordinated tertiary contractions and high pressure non relaxing lower oesophageal sphincter. pH-monitoring, carried out only in 3 cases, showed a pathologic reflux in 2, correlated endoscopically with a mild oesophagitis, and it was negative in the other case. All patients were operated on with the oesophagomyotomy through an abdominal approach and the length of the myotomy varied between 4 and 10 cms including a tract onto the stomach. All patients received also an antireflux procedure as previously described (Tab. 1).

The follow up of this series ranges between 8 months and 14 years (mean: 5.5 years) and 17 (89%) patients are actually doing well completely free of correlated symptoms (Fig. 2, 3).

Relapse of dysphagia occurred in 2 cases (10%) and the cause attributed to insufficient myotomy, One of them underwent a new Heller procedure, this time associated to an Allison plasty and his problem resolved completely (follow up 4 years).

The other patient presented only a residual light dysphagia and was controlled periodically without presenting major problems. Only a patient (5%) presented a light GER postoperatively even if he underwent an antireflux procedure (Lortat Jacob) at time of the myotomy. This problem has been controlled by medical therapy alone. There are not significant differences, in term of postoperative complications, among the various antireflux techniques employed. Finally, one child (5%) underwent a relaparotomy after two years for intestinal adhesions.

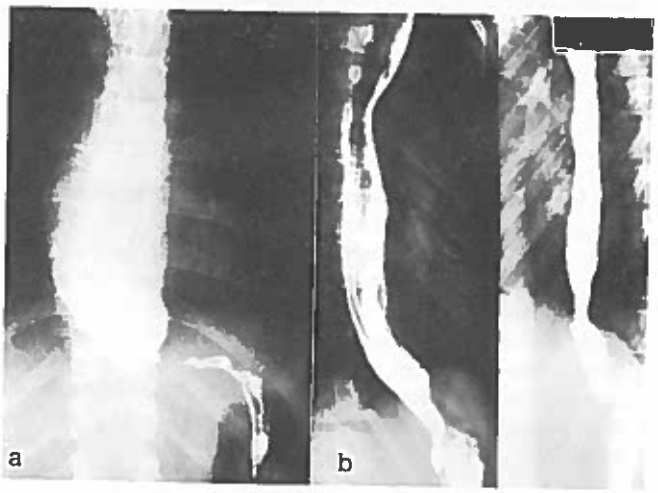

Figure 2: a) preoperative oesophagogram and b) 6 months after myotomy.

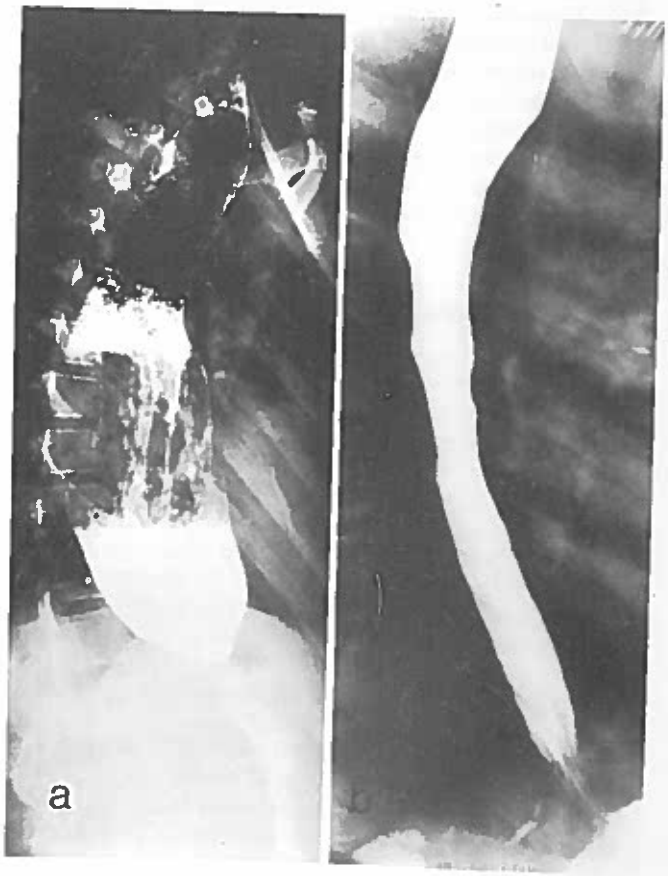

Figure 3: a) preoperative enormous dilatation of the oesophagus and b) control oesophagogram 1 year after myotomy shows an almost normal caliber of the oesophagus.

N°	NAME	AGE	SEX	SYMPTOMS DURATION	DIAGNOSTIC INVESTIGATIONS	TREATMENT	RESULTS FOLLOW-UP ( )
1	A.B.	10 y	f	12 m	oesophagogram	OM + Allison Pl.	doing well (5y)
2	M.G.	11 y	m	15 m	oesophagogram	OM	relapse of symptoms after 2 y - New OM + Allison Pl. doing well (4y)
3	G.R.	4 y	f	12 m	oesophagogram	OM + Allison Pl.	doing well (10y)
4	M.T.	7 y	m	8 m	oesophagogram pH-monitoring endoscopy	OM + Nissen Pl.	doing well (6y)
5	N.M.	9 y	m	12 m	oesophagogram endoscopy	OM + Nissen Pl.	doing well (4y)
6	M.S.	4 y	m	1 m	oesophagogram pH-monitoring endoscopy	OM + Thal Pl.	doing well (2y)
7	L.O.	2 y	f	24 m	oesophagogram pH-monitoring endoscopy manometry	OM + Thal Pl.	doing well (2y)
8	G.L.F.	11 y	m	5 m	oesophagogram endoscopy manometry	OM + Pellerin Pl.	doing well (8m)
9	D.B.	6 y	m	6 m	oesophagogram	OM + Lortat Jacob Pl.	residual light dysphagia (7y)
10	R.R.	14 y	f	18 m	oesophagogram manometry	OM + Lortat Jacob Pl. I.O. manometry	doing well (6y)
11	M.C.	12 y	m	36 m	oesophagogram manometry	OM + Lortat Jacob Pl. I.O. manometry	residual light GER - medical treatment - fair condition (5y)
12	F.M.	7 y	m	7 m	oesophagogram manometry	OM + Nissen Pl. I.O. manometry	doing well (5y)
13	F.D.P.	11 y	f	12 m	oesophagogram endoscopy manometry	OM + Nissen Pl. I.O. manometry	doing well (7y)
14	S.F.	11 y	f	4 m	oesophagogram	OM + Lortat Jacob Pl.	doing well (14 y)
15	E.E.	9 y	m	6 m	oesophagogram manometry	OM + Pellerin Pl.	doing well (11y)
16	I.E.	2 y	f	15 m	oesophagogram endoscopy manometry	OM + Thal Pl.	relaparotomy for intestinal adhesions after 2 y - doing well (8y)
17	E.M.	7 y	f	36 m	oesophagogram	OM + Pellerin Pl.	doing well (4y)
18	A.M.	11 y	f	7 m	oesophagogram manometry	OM + Pellerin Pl. I.O. manometry	doing well (3y)
19	T.C.	11 y	m	9 m	oesophagogram manometry	OM + Pellerin Pl. I.O. manometry	doing well (2y)

TABLE 1 : Main clinical data concerning the 19 cases of oesophageal achalasia observed at the Gaslini Hospital (1-8), Pediatric Surgery of the Pescara Hospital (9-13) and Pediatric Surgery of the 2nd school of Medicine of Naples (14-19). OM= OesophagoMyotomy; I.O.= IntraOperative.

DISCUSSION

Several therapeutic approaches to oesophageal achalasia have been reported in literature. The medical therapy has been employed in the early stage of the disease, either by stimulating the oesophageal peristalsis (domperidone, metoclopramide) or by opposing the action of gastric juice on the oesophageal mucosa with antiacid and anti-H2 medications. However, the management of oesophageal motility disorders using these pharmacological measures has been disappointing. The administration of nifedipine, a calcium entry blocker, has been recently proposed but its usefulness in this disease remains to be proven (9).

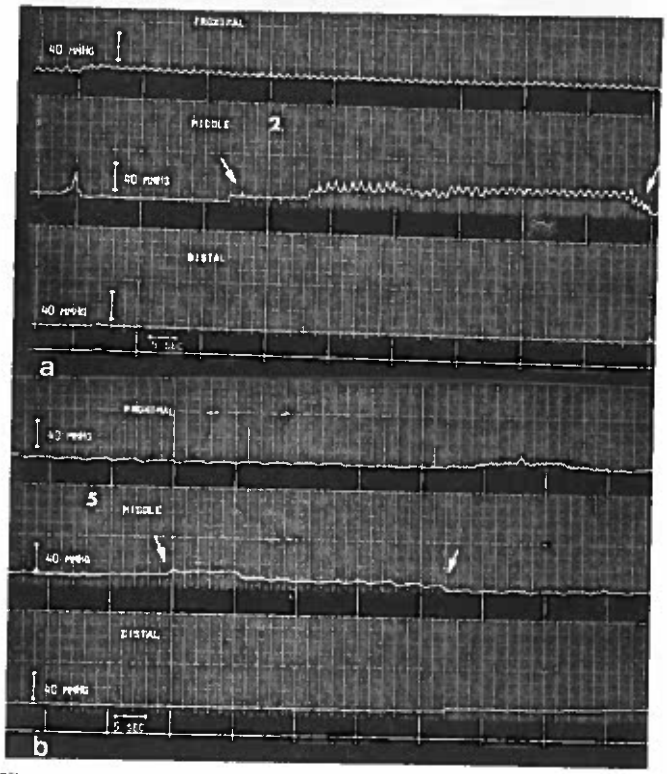

Figure 4: Intraoperative manometry: a) superior channel (proximal) measures the pressure of the oesophageal body; medium channel (middle) shows the high pressure of the LES and lower channel (distal) measures the basal pressure of the gastric fundus; b) between the arrows the "pressure fall" of the LES after myotomy.

Forceful pneumatic or hydrostatic dilatation has been advocated as the treatment of choice in adults. We have not experience with this technique, but revision of the literature on this subject prompted us to prefer surgical myotomy to forceful disruption of oesophageal fibers that in children may be harmful and variable in results (10-16).

Oesophagomyotomy must be extended for 7-10 cms or insufficient relaxation of the lower sphincter may result. More accurate myotomy may be obtained under manometric guidance (Fig. 4). Because the major long term complication after myotomy is gastroesophageal reflux (3-50%) an antireflux procedure must be associated in the same time. Several different techniques have been reported in the literature and in our series 5 different plasties were utilized without significant difference.

Recently Taylor and Myers reported on their experience in 20 pediatric patients with oesophageal achalasia, of which 19 underwent myotomy through a thoracic approach (17). This route is preferred by the Authors because they "...believe that as long as myotomy is not extended onto the stomach more than few millimetres and the oesophageal hiatus is not disrupted, an antireflux procedure is not needed". Even if we have not experience with the thoracic approach, we believe that the abdominal route is preferable because it allows a complete and extended myotomy on the oesophago-gastric junction without postoperative minimal residual dysphagia as reported by the Australian Authors.

From the diagnostic point of view the oesophagogram together with manometry are the most reliable and sensitive methods of diagnosing achalasia. Endoscopy and pH-monitoring of the oesophagus are not essential for diagnosis and should be performed only to rule out other oesophageal and gastro-duodenal conditions (11, 18).

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