Diagnosis and Surgical Management of Children with Oesophageal Achalasia: A 10-Year Single-Centre Experience in Morocco

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Abstract

Introduction: Achalasia is a primary oesophageal motility disorder with unknown aetiology. The aim of this study was to evaluate our 10-year experience in the diagnostic process and surgical management of oesophageal achalasia (OA) in children. Methods: A retrospective review of all children (age: 0–15 years) treated for achalasia at the department of paediatric surgery from 2007 to 2016 was conducted. The demographics, presenting symptoms, associated diseases, diagnostic process, outcomes and complications were analysed. Results: Fourteen patients were identified, with a mean age of 5.2 years. There were eight female and six male patients. The most common symptom was chronic vomiting, in all patients (100%), followed by weight loss in 10 (71.4%), cough in 6 (42.9%), dysphagia in 5 (35.7%) and chest pain in 3 (21.4%). The mean duration of symptoms until diagnosis was 36.3 ± 29.1 months. Three patients underwent an open Heller myotomy (HM) and 11 laparoscopic HM (LHM) including three conversions. The reasons for conversion were mucosal perforation in two cases and liver bleeding in one patient. The mean operating time and the average length of postoperative stay in the patients of LHM group were, respectively, 2.0 ± 0.7 h and 4 ± 1.5 days. The mean follow-up was 43.2 months. Conclusion: The diagnosis of OA in children is based on clinical arguments and especially on the barium oesophagram findings. Laparoscopic myotomy is the most effective surgical approach in children.

Keywords: Children, diagnosis, Heller myotomy, laparoscopy, oesophageal achalasia

INTRODUCTION

Achalasia is a primary oesophageal motility disorder with unknown aetiology, characterised by the absence of oesophageal peristalsis and increased or normal resting pressure of the lower oesophageal sphincter (LOS) which fails to relax completely in response to swallowing.\(^1\) The incidence of achalasia in childhood is 0.18/100,000 children/year.\(^2\) Symptoms such as poorly digested food, weight loss and chronic cough are said to be more common presenting features in children.\(^3\) Achalasia is diagnosed with a barium swallow study and may be confirmed with oesophageal manometry. An upper endoscopy and biopsy are reasonable to rule out oesophagitis.\(^4\) The treatment of OA can be either medical or surgical, and its aim is the reduction of LOS pressure, with consequent suppression of outflow obstruction and symptomatic relief.\(^5\) Pharmacological management of achalasia in children is almost always unsuccessful, and pneumatic dilatation is rarely carried out in the paediatric age group.\(^6\) Amongst the multiple treatments for achalasia, LHM seems to be the procedure of choice.\(^6\) Recently, the per-oral endoscopic myotomy has been establishing itself as a valid alternative in the treatment algorithm of achalasia.\(^8\) In low-income countries, specifically in Morocco, few studies have been performed about the surgical management of OA in children. We, therefore, undertook this study to evaluate our 10-year experience in the diagnostic process and the surgical management of OA in children.

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How to cite this article: Idrissa S, Oumarou A, Mahmoudi A, Elmadi A, Khattala K, Bouabdallah Y. Diagnosis and surgical management of children with oesophageal achalasia: A 10-year single-centre experience in Morocco. Afr J Paediatr Surg 2021;18:155-9.
METHODS

Data collection

A retrospective review was conducted on medical records of children treated for OA at the Department of Pediatric Surgery of Hassan II university Hospital through January 2007 to December 2016 using hospital information systems (Hosix) and the patients register. The diagnosis of OA suspected clinically (chronic vomiting, dysphagia, weight loss, and so on) was confirmed by a combination of barium oesophagram, upper endoscopy and oesophageal manometry. All patients who had Heller myotomy (HM) for achalasia in our institution during the study were included in the study. Patients were excluded from the study if they had not received intervention for achalasia in our institution or had insufficient medical records for data collection. Information collected for each patient included age, sex, associated diseases, presenting symptoms and duration of the symptoms. Early and late complications, operating times, length of postoperative stay and length of follow-up were also analysed. The University of Sidi Mohamed Ben Abdellah Research Ethics Committee approved the project, and informed consent was obtained from all patients.

Surgical technique

Our approach to the HM has undergone a progressive evolution from open to our current laparoscopic approach, which we will detail here. The patient is placed in the supine position and four trocars (one transumbilical 10 mm trocar and 3 trocars of 5 mm) are inserted into the abdomen under direct visualisation [Figure 1a]. We perform the LHM after opening the pars flaccida of the lesser omentum to allow for dissection of the right and left crura from the oesophagus and transection of the phrenoesophageal ligament [Figure 1b]. This sentence could be replaced by this one: The myotomy begins on the lower part of the lower esophageal sphincter and continues to 4 cm above on the lower third of the esophagus, and then extends to the stomach for 2 cm [Figure 1c]. To exclude oesophageal perforations, the methylene blue is injected through a nasogastric tube, and an oesophageal perforation is detected when the methylene blue is seen. We finish the operation by adding a Dor fundoplication [Figure 1d]. The surgical technique is summarised in Figure 1.

Statistical analysis

Statistical data were analysed by Database were analysed with Microsoft (Redmond, Redmond, Washington, uni stade) Office Excel(Version 2010). Categorical variables were expressed as proportions (as well as in percentages) and continuous variables as mean ± standard deviation and range (minimum to maximum).

RESULTS

Demographics, presenting symptoms and associated diseases

Fourteen patients were identified with a mean age at surgery of 5.2 years ± 3.0 years (range: 0.9 year to 9 years). There were eight female (57.1%) and six male (42.9%) patients. Consanguinity was found in nine (64.3%) patients. Four patients (28.6%) had Allgrove syndrome (or 3A syndrome). Two patients (14.3%) had achalasia and alacrima without ACTH insensitivity. One of them had a psychomotor retardation. The most common symptom was chronic vomiting in 14 (100%), followed by weight loss in 10 (71.4%), cough in 6 (42.9%), dysphagia in 5 (35.7%) and chest pain in 3 (21.4%). The mean duration of symptoms until diagnosis was 36.3 ± 29.1 months (range, 8–90 months). These results were summarised in Table 1.

Diagnosis process

The barium oesophagram showed oesophageal dilation in all patients. The “bird-beak” sign was found in seven patients (50%). The upper endoscopy revealed a dilated oesophagus with increased resistance at the gastro-oesophageal junction in all cases and highlighted an oesophagitis in four (28.6%) patients. The manometry performed in only two patients (14.3%) found, an absence of oesophageal peristalsis with increased intra-oesophageal pressure and incomplete relaxation of the LOS on deglutition. Pulmonary X-rays were of great diagnostic utility in five (35.7%) of our patients, showing an air–fluid level. These results were summarised in Table 1.

Surgical treatment

Amongst 14 patients, three underwent an open HM (OHM) and 11 had LHM (including three conversions). In the eight patients who underwent LHM alone, an additional fundoplication was performed in seven patients (Dor n = 6, Toupet n = 1, as per surgeon preference). The reasons for the conversions were mucosal perforation in two cases (18%) and liver bleeding in one patient (9%). The mean operating times and the average length of postoperative stay in the patients of LHM group were, respectively, 2.0 ± 0.7 h (range 1, 20–3, 20 h) and 4 ± 1.5 days (range: 2–6 days). Amongst the six patients who underwent OHM (three OHM alone and three LHM conversions into OHM), a Nissen fundoplication was...
performed in five patients. These results were summarised in Table 1.

**Follow-up and outcome**

The mean follow-up was 43.2 months (range: 6–110 months). All our patients had a good outcome with a symptom relief and good weight gain. There was no mortality during the follow-up. There were no repeated operations. No patient presented symptoms of gastro-oesophageal reflux disease. In two patients, dysphagia and vomiting persisted and required oesophageal balloon dilation with complete resolution of their symptoms after 2 for one and 3 months for the other. Dysphagia persisted in one patient despite balloon dilation, and a repeat surgery option was considered. These results were summarised in Table 1.

**Discussion**

Achalasia is a rare disease, particularly regarding the paediatric population since <5% of patients have symptoms when they are below 15 years of age.\(^1\) Therefore, the cases of OA in children population are limited. To the best of our knowledge, the largest study included 42 children and was performed by Meyer et al.\(^9\). Described in 1674 by Sir Thomas Willis, the aetiology of OA remains unknown, and in the majority, it is idiopathic. However, it has been established that a spasm or a failure to relax of LOS is the pathophysiologic mechanism of achalasia.\(^10\) The consequent result is the impaired flow of ingested food into the stomach and subsequent stasis of food and secretions in the oesophagus.

In children, specifically in infants, symptoms are unspecific. Dysphagia is the most common symptom in adults while chronic vomiting seems to be more common in children, as in this study and others previously reported.\(^{[1][13]}\) This could be explained by the intermittent nature of dysphagia at the beginning of the disease and the impossibility in young children to express it. An additional half of patients in this series had an associated disease including four cases of Alagrove’s syndrome or triple-A syndrome, two cases of achalasia and alacrima without ACTH insensitivity and one case of mental retardation. There were two siblings diagnosed with OA. The frequency of consanguineous unions, associated diseases and family-related forms of achalasia make this study interesting and support the hypothesis that OA is a hereditary disease transmitted by an autosomal recessive trait.\(^{[12]}\)

One of the limitations in our study could be the diagnosis process. The oesophageal manometry, which is the key test for the diagnosis has been performed in only two of our patients. In a systematic review of the literature, Pacilli et al.\(^{[13]}\) reported a rate of oesophageal manometry in 76% of patients, whereas barium swallow test was performed in 97% of patients. According to Morera and Nurko,\(^{[14]}\) the classic description of a non-relaxing high-pressure LOS in patients with achalasia is rarely found in children. They reported that the LOS function in paediatric patients with achalasia is heterogeneous (partial relaxations are common and normal relaxations may be present). Manometry is, therefore, necessary but not indispensable for the diagnosis of achalasia in children.

In this series, surgery was the first treatment of all patients. Hence, we cannot conclude about the superiority of LHM in comparison with other treatments of OA. However, the low rate of perioperative and postoperative complications

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**Table 1: The demographic, clinical features, surgical management and follow-up of the 14 patients**

| Case number | Age at surgery (year) | Sexe | Presenting symptoms | DOS (month) | Associated diseases | Diagnostic method | Surgical technique | Peroperative Complication | FUP (month) | Outcome |
|-------------|-----------------------|------|---------------------|-------------|---------------------|-------------------|-------------------|------------------------|------------|---------|
| 1           | 9                     | Female | V, D, P, W, C    | 72          | -                   | B, E, M          | LHM+Nissen        | Perforation (conversion) | 110        | Asymptomatic |
| 2           | 8                     | Male   | V, D, P, W        | 12          | -                   | B, E             | OHM+Nissen        | No                     | 110        | Asymptomatic |
| 3           | 1.6                   | Female | V, W, C           | 18          | -                   | B, E             | OHM+Nissen        | No                     | 86         | Asymptomatic |
| 4           | 4                     | Male   | V, D, C           | 8           | Mental retardation  | B, E             | LHM+Nissen        | Perforation (conversion) | 74         | Asymptomatic |
| 5           | 2.2                   | Male   | V, D              | 27          | 3A                  | B, E             | LHM+Dor           | No                     | 62         | Asymptomatic* |
| 6           | 7                     | Female | V, W              | 60          | -                   | B, E             | LHM               | No                     | 26         | Asymptomatic |
| 7           | 7                     | Male   | V, W, C           | 84          | 3A                  | B, E             | LHM+Dor           | No                     | 32         | Asymptomatic |
| 8           | 7                     | Female | V, D, P, W, C    | 36          | 3A                  | B, E             | LHM+Dor           | No                     | 25         | Asymptomatic |
| 9           | 1.3                   | Male   | V, W              | 14          | -                   | B, E             | LHM+Dor           | No                     | 22         | Asymptomatic* |
| 10          | 0.9                   | Female | V, W              | 9           | -                   | B, E             | LHM+Dor           | Hepatic bleeding (conversion) | 22         | Asymptomatic |
| 11          | 8                     | Male   | V, W              | 90          | 2A                  | B, E             | OHM+Nissen        | No                     | 12         | Dysphagia |
| 12          | 9                     | Female | V, W, C           | 48          | -                   | B, E             | LHM+Dor           | No                     | 12         | Asymptomatic |
| 13          | 2                     | Female | V                 | 12          | 2A                  | B, E             | LHM+Toupet        | No                     | 6          | Asymptomatic |
| 14          | 6                     | Female | V                 | 19          | 3A                  | B, E             | LHM+Dor           | No                     | 6          | Asymptomatic |

*Asymptomatic after balloon dilation. DOS: Duration of symptoms, FUP: Follow-up period, 3A: Achalasia, alacrima and adrenal insufficiency, 2A: Achalasia and alacrima, LHM: Laparoscopic Heller myotomy, OHM: Open Heller myotomy, V: Chronic vomiting, D: Dysphagia, W: Weight loss, P: Chest pain, C: Cough, B: Barium oesphagram, E: Endoscopy, M: Manometry
in the present study as in the literature\textsuperscript{[11,15-18]} shows that the HM should be regarded today as the most safe and effective treatment of OA in children. The surgical technique was described by Ernest Heller in 1913.\textsuperscript{[13]} For many years, this procedure has been performed by an open approach, either through a thoracotomy or a laparotomy. The first minimally invasive myotomy in the United States was performed with a left thoracoscopic approach in 1991 by Shimi et al.\textsuperscript{[19]} Thus, the last three decades have witnessed a progressive evolution in the surgical treatment of OA, with a shift from open to a minimally invasive HM.\textsuperscript{[20]}

There is a true debate about OA management in children not only through the low incidence of the disease but also more importantly, due to the absence of any significant randomised controlled trials in children. Despite the multiple treatment strategies, no modality has been found to be unequivocally better than the other.\textsuperscript{[17]} Medical treatment has been attempted in the form of calcium channel blockers and nitrates, with minimal effect, especially in children. This therapy is only recommended for short-term symptomatic relief pending definitive surgical treatment.\textsuperscript{[21]} The use of pneumatic dilation and botulinum toxin injection over surgery has been debated in the literature, especially in children. Both pneumatic dilation and botulinum toxin injection have demonstrated improvement in the symptoms related to achalasia, but neither treatment has long-term efficacy and requires repeated procedures.\textsuperscript{[21]}

Lee et al.\textsuperscript{[26]} reported that all oesophageal dilation patients developed recurrent symptoms; 93% required additional intervention in the form of repeated dilations or HM. Meyer et al.\textsuperscript{[10]} using botulinum toxin, as a primary therapy in half of their patients, had a temporary impact. Many patients (about one-third of this group) ultimately necessitated a second myotomy after the first. According to Zagory et al.,\textsuperscript{[22]} HM is superior to balloon dilation or botulinum injection in children with achalasia. Currently, the LHM is the most effective method for childhood achalasia in many centres.\textsuperscript{[5,16,21-23]} About an additional fundoplication to laparoscopic myotomy, there is no consensus to date. Nevertheless, it seems that there is no difference in the incidence of gastro-oesophageal reflux and dysphagia between patients with or without fundoplication.\textsuperscript{[8]}

For them, the routine use of an additional fundoplication might not be justified. As far as we are concerned, we systematically perform a Dor fundoplication, and our results are encouraging. Between 2007 and 2011, five patients were operated for OA in our institution. Three underwent laparoscopic HM including two mucosal perforations and necessitated conversion to OHM. Amongst the nine patients of the series operated by LHM between 2014 and 2016, there was no perforation. The only perioperative complication was hepatic bleeding which led to a conversion. The particularity of this patient is that he was the youngest of this series (11 months at surgery) and had an important weight loss (lower than 5 kg weight at the surgery).

In children, the risk factors for intraoperative mucosal perforation during LHM remain unclear. Tsuboi et al. found in adult, an increased risk of mucosal perforations where the surgeon’s operative experience was lower than five cases, prolonged operative time and increased intraoperative blood loss.\textsuperscript{[24]} They found an overall incidence of mucosal perforation of 15.4% comparable to our study (18.1%). However, this rate of mucosal perforation is much higher than many studies in children.\textsuperscript{[4,5,16,17,22,25,26]} This rate could be explained by the fact that, before 2010, laparoscopic surgery was less used in our institution. However, since 2011, there has been a generalisation of minimally invasive surgery in digestive, thoracic, and urologic pathologies. The learning curve of our surgeons has thereby strongly improved. That could also explain the absence of mucosal perforation in the nine patients operated after 2011. It is therefore evident that the main factor of mucosal perforation is surgeon expertise in coelioscopic surgery.

**Conclusion**

The diagnosis of achalasia is suspected in children with chronic vomiting, dysphagia, chronic cough and weight loss. It is confirmed by a combination of barium oesophagram, upper endoscopy with or without oesophageal manometry. Laparoscopic myotomy with or without fundoplication provides both short- and long-term symptomatic relief with decreased hospital stay and less complication rate. It is the most effective surgical approach in children.

**Acknowledgements**

The authors thank the patients for their consent and ethics committee for approval of the project of publication of this article.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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