Treatments for pediatric achalasia: Heller myotomy or pneumatic dilatation?

Summary

Aim. — The treatment of achalasia consists of reducing distal esophageal obstruction by either Heller myotomy surgery or endoscopic pneumatic dilatation. The aim of the present study was to evaluate the short- and middle-term results of these procedures in children.

Methodology. — For technical reasons, children under six years old (n = 8) were treated by surgery only, whereas patients over six years old (n = 14) were treated by either Heller myotomy or pneumatic dilatation.

Results. — Of the children aged under six years, 75% were symptom-free at six months and 83% at 24 months of follow-up. Of the patients aged over six years, complete remission was achieved by Heller myotomy in 44.5% vs. 55.5% by pneumatic dilatation after six months, and in 40% vs. 65%, respectively, after 24 months. Both pneumatic dilatation and Heller myotomy showed significant rates of failure.

Conclusion. — These results suggest that pneumatic dilatation may be considered a primary treatment in children over six years old. Also, where necessary, Heller myotomy and pneumatic dilatation may be used as complementary treatments.

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Treatments for pediatric achalasia

Introduction

Idiopathic esophageal achalasia is a primary motor disorder characterized by ineffective peristalsis in the body of the esophagus and inadequate relaxation of the LES. This pathology is relatively rare, with an estimated incidence of about 1 per 100,000 births [1,2]; the diagnosis is usually made during adulthood.

The clinical symptoms are dysphagia that is usually intermittent, anorexia, vomiting and regurgitation, all of which may be mistaken for gastroesophageal reflux and retrosternal pain [3]. It rapidly leads to failure of weight growth in children. The diagnosis is made according to the results of esophageal manometry, which allows esophageal motor disturbances to be recorded, including the absence of propagated esophageal contractions in response to drinking water and esophageal aperistalsis, and the lack of, or incomplete, LES relaxation (in other words, the LES is mostly hypertonic).

However, esophageal achalasia may be part of a more general pathology, such as the Allgrove (triple-A) syndrome, comprising alacrima, adrenal insufficiency and achalasia [4].

The treatment consists of lowering LES pressure by pharmacological, surgical or endoscopic means [5]. Pharmacological treatments such as nitro compounds and intrasphincteric injection of botulinum toxin result in transitory effects and, nowadays, are used only in very mild-to-moderate forms of the condition [5].

Surgical treatment by HM involves a 4- to 6-cm longitudinal incision in the distal esophageal smooth muscle and extending a few centimeters onto the gastric cardia. An antireflux procedure is often carried out to reduce postoperative reflux, considered the main side-effect of this surgical intervention. The other reference treatment in adults is PD, which involves rupturing the muscle fibers with an inflatable balloon. Several dilatations may be required, each time using a larger balloon diameter. The main complication of PD is esophageal perforation, with reported rates of 1–3% in adults [5].

However, these surgical and endoscopic treatments have primarily been evaluated only in adults. In pediatric patients, HM has long been considered the reference method, and was initially performed by thoracotomy, then laparotomy and, now, laparoscopy [6], whereas PD is a more recent treatment [7–9]. For this reason, its efficacy has not been thoroughly studied.

The present study involved 22 children treated with HM and/or PD at two tertiary pediatric gastroenterology centers. As PD is not possible in children under six years of age for technical reasons (the size of the balloon), we divided our patients into two categories: those under six years old were treated by HM only (n = 8); and those over six years old were treated by either HM and/or PD (n = 14). The short- and middle-term effectiveness of the treatments in these two groups were evaluated.

Patients and methods

Patients

Altogether, 22 children were treated for idiopathic esophageal achalasia by HM and/or PD between 1990 and 2007 at two French tertiary pediatric gastroenterology centers, and followed-up for at least six months after treatment. The most frequently observed initial clinical symptoms were: dysphagia; vomiting and regurgitation; cough or dyspnea after meals; retrosternal pain; ptyalism; and weight loss and/or growth retardation. The diagnosis was made on the basis of manometric criteria: the absence of propagation of contraction waves in the esophagus; and a failure of relaxation of the LES. Barium esophagograms showed abnormal enlargement of the esophagus, barium retention and sticture of the cardia.

After treatment, the follow-up study of the children was longitudinal and based on the presence of the following clinical criteria: dysphagia; regurgitation and/or vomiting; pain; and respiratory signs. The growth parameters taken into consideration were height and weight.

The responses to treatment were classified as: success, where patients were free of symptoms; partial improvement, where patients had only occasional dysphagia and/or vomiting; and failure, where patients had daily dysphagia.

Pneumatic dilatation

PD was carried out under general anesthesia from 2002 to 2007, and the patients’ stay in hospital lasted 24–48 h after treatment to confirm proper resumption of oral feeding and
the absence of complications. Prior to the PD procedure, all patients underwent upper digestive endoscopy. The Rigiflex® pneumatic dilatation balloon used was either 30 or 35 mm in diameter. The procedure was repeated 1–3 times per session at varying pressures—from 2 to 10 PSI, which were maintained for no more than 60 s at maximum pressure—until the X-ray control showed the disappearance of the cardia impression. In cases where two dilatation sessions were necessary, the air pressure for inflating the balloon and the dilatation period were both increased.

Heller myotomy

Recently, laparotomy and then laparoscopy have been used to perform a modified HM procedure [10]. The procedure has also included fundoplication, using Dor, hemi-Toupet or Nissen antireflux valves; the intragastric extension was 3–4 cm in length. No upper endoscopy was performed at the time of surgery.

Treatment choice

Of the 22 treated children with achalasia, 13 had surgery, five had PD, two had PD followed by HM and two had HM followed by PD. Each treatment case was discussed at an interdisciplinary staff meeting, including the surgeons and pediatric gastroenterologists at the two centers. Treatment was based on the following criteria: age; feasibility of carrying out PD; and effect of previous treatment. PD was not performed in children who were under six years old or who weighed under 20 kg, as the dilatation balloon was not adapted for pediatric use, and its introduction into the esophagus of a small child could lead to trauma. This meant that the children aged under six years (n = 8) underwent modified HM [5], while those aged over six years first underwent either HM (n = 7) or PD (n = 7), when the latter became available after 2002.

Statistical analysis

Yates’ chi-square test was used for comparisons of proportions. The Mann–Whitney test was used to compare the distributions of patients who were successfully treated and those who were not responsive to treatment in the two studied groups. P values < 0.05 were considered significant.

Results

Patients

A total of 22 children (12 boys and 10 girls) with esophageal achalasia were treated by either PD or HM surgery, or both (Tables 1 and 2). The average age at the time of diagnosis was 7.8 years (the youngest was one month old, and the oldest was 16 years of age). Eight children were aged under six years (patients 1–8; age range: 3 months to 4 years) and 14 were aged over six years (patients 9–22; age range: 6.5–16 years). One child (patient 19) had Allgrove syndrome. Five children under six years old had the following associated pathologies: Hirschsprung’s disease and Ondine’s curse (patient 1); moyamoya disease (patient 2); autoimmune thrombocytopenia (patient 5); and chronic bronchopneumopathy (patient 7). Patients 3 and 4 were siblings. All of the children except patient 11—who had cough and dyspnea after eating—suffered from dysphagia or vomiting episodes. Five children had ptyalism, and five complained of thoracic pain. The average follow-up was 52 months (range: 7 months to 17 years). The time from symptom onset to diagnosis was, on average, 9.5 months (range: < 1 month to 4 years). Growth retardation was similar in both the PD and HM groups (—1.2 SD and —1.5 SD in the weight curve, respectively; —0.5 SD and —0.6 SD in the height curve, respectively).

Of the 22 treated children, 13 had previously received calcium-channel blockers. In one case (patient 11), nifedipine treatment had to be stopped because of hypotension, whereas the drug either was or became ineffective in the others. Endoscopic treatment by injection of botulinum toxin had not been used.

Therapeutic results

Heller myotomy

Seventeen children were treated by HM. In two of these cases, HM was a second-line treatment after failure of PD (Table 1). The average age at the time of surgery was 21 months (range: 4 months to 4 years) in the children aged under six years, and 11.5 years (range: 6.5–17 years) in the PD group. Postoperatively, inhalation pneumopathy occurred in one case (patient 12). Dumping syndrome, a well-known complication after fundoplication in children [11], was observed in three cases (patients 1, 9 and 20); the diagnosis was made after symptomatic hypoglycemia. In cases where the Nissen fundoplication wraps were excessively tight (n = 4), hydrostatic balloon dilatation was carried out. The time-lag between fundoplication surgery and hydrostatic balloon dilatation was two months in three cases and eight months in the remaining case.

Six months after HM, 58% of the patients were symptom-free and 23% showed partial improvement (persistent intermittent dysphagia). At 24 months after treatment, 64% of the patients were still free of symptoms (Table 1). Three children were followed-up for eight years, and one up to now has still shown no symptoms. There was no statistically significant difference (P = 0.18) between the responses to HM in the children aged under six years (75% had no symptoms six months later) and those over six years of age (45% had no symptoms six months later).

Pneumatic dilatation

Nine children over six years of age underwent PD and, in seven cases, it was the primary treatment for achalasia and the only treatment in five children (Table 2). The average age of the patients at the time of treatment was 13 years (range: 8.5–18 years). One (n = 5) or two (n = 4) PD sessions were carried out; the second session was performed only when the first had failed.

At six months of follow-up, 55.5% of these patients had no symptoms and, at 24 months, two-thirds of the followed-up patients had still not relapsed. Furthermore,
Table 1  Results of the Heller myotomy (HM) surgical procedure.

<table>
<thead>
<tr>
<th>Patient number</th>
<th>Age at HM</th>
<th>HM as first intention</th>
<th>HM as second intention</th>
<th>Surgical procedure</th>
<th>Second surgical procedure</th>
<th>Postsurgical complications</th>
<th>Results at 6 months</th>
<th>Results at 12 months</th>
<th>Results at 18 months</th>
<th>Results at 24 months</th>
<th>Results at 4 years</th>
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<td>S</td>
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<td>S</td>
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<td>3</td>
<td>14 months</td>
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<td>S</td>
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<td>S</td>
<td>S</td>
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<tr>
<td>4</td>
<td>4 months</td>
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<td></td>
<td>Laparotomy</td>
<td></td>
<td>S</td>
<td>S</td>
<td>S</td>
<td>S</td>
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<tr>
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<td>2 years</td>
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<td>Laparotomy</td>
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<td>S</td>
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<tr>
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<td>2 years</td>
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<td></td>
<td>Laparotomy</td>
<td></td>
<td>S</td>
<td>S</td>
<td>S</td>
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<td>7</td>
<td>3.5 years</td>
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<td>S</td>
<td>S</td>
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<tr>
<td>8</td>
<td>4 years</td>
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<td>Dumping syndrome</td>
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<td>I</td>
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<td>8 years</td>
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<td></td>
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<td>Infection pneumopathy</td>
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<td></td>
<td>Laparotomy</td>
<td></td>
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<td>S</td>
<td>S</td>
<td>S</td>
<td>S</td>
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<tr>
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<td>11.5 years</td>
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<td></td>
<td>Laparotomy</td>
<td></td>
<td>S</td>
<td>S</td>
<td>F</td>
<td>F</td>
<td>F</td>
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<tr>
<td>19</td>
<td>15 years</td>
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<td></td>
<td>Laparotomy</td>
<td></td>
<td>F</td>
<td>F</td>
<td>F</td>
<td>F</td>
<td>F</td>
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<tr>
<td>20</td>
<td>15 years</td>
<td>Yes</td>
<td></td>
<td>Laparotomy</td>
<td></td>
<td>Dumping syndrome</td>
<td>F</td>
<td>F</td>
<td>F</td>
<td>F</td>
<td></td>
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<tr>
<td>21</td>
<td>17 years</td>
<td>No</td>
<td>Yes</td>
<td>Laparoscopy</td>
<td></td>
<td>F</td>
<td>10 S; 3 F; 4 PI</td>
<td>7 S; 4 F; 2 PI</td>
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<td></td>
<td>1 S; 2 F</td>
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</tbody>
</table>

Total Mean: 15/17  2/17  3 patients  4 patients
7 years

S: success; F: failure; PI: partial improvement.
no post-dilatation complications and, most notably, no esophageal perforations were reported.

Discussion

Twenty-two children with esophageal achalasia (eight aged under six years, and 14 aged over six years) were treated by either HM or PD, or both. Only those children who were symptom-free after treatment were considered to be in remission. After six months, 57% of the children had no symptoms and, after two years, 64% were symptom-free. These rates of success reflect the difficulties of treating achalasia not only in adults (80—90% of patients had either partial or complete remission at two years) [12—14], but in children as well (70—90% had either partial or complete remission at two years) [15—17].

In the present study, all of the HM procedures included fundoplication, and none of the children in this series experienced gastroesophageal reflux. However, the addition of an antireflux procedure after HM is controversial, as reported in other previous studies [18—22].

For technical reasons (balloon size), in children aged under six years old, surgery was the only possible treatment. Also, PD cannot be used in children weighing under 20 kg. However, achalasia in infants may be different from that in older children. In the present study, five infants began to show symptoms of achalasia at ages 3—7 months. However, achalasia can be either a disorder on its own or part of a more general pathology. Indeed, patients 3 and 4 (siblings) had a familial form; patient 1 had Hirschsprung’s disease and Ondine’s curse; patient 2 suffered from moyamoya disease and had experienced cerebrovascular accidents; and patient 5 had transient autoimmune thrombocytopenia. Some cases of transitory achalasia in infancy were reported, notably in infants with low birth weights [23,24] and in children with the Pierre Robin malformation sequence [23]. The results of their manometry tests were similar to those of older children with achalasia. However, some spontaneously regressing abnormalities highlighted a distinction between achalasia in very young children (< 6 years old) and adults [24]. In such infants, any associated co-morbidities should be investigated. Also, the use of medical treatments or intrasphincter injection of botulinum toxin [5] may allow time for the possible resolution of symptoms to occur, thereby avoiding HM surgery. Nevertheless, in cases of medication treatment failure, surgery is the only form of treatment available for children of this age.

In the present study, the children aged over six years were treated with either PD or HM, depending on the feasibility of PD and the effects of previous treatment. At six months, the results of HM (in children aged > 6 years) were similar to those of PD — 44.5% rate of success after HM vs. 55.5% after PD — and, at 24 months, these rates were 40% vs. 65%, respectively. No complications were observed after PD. However, given the absence of sufficient case evidence of the use of PD in children in the literature [8,10,25—27], HM has been the main treatment option of achalasia in children [15,16,20,28].

Nevertheless, in our experience, PD appears to be a feasible and effective treatment for achalasia in children aged over six years and/or weighing over 20 kg. Indeed, in such
cases, PD may be considered a first-line treatment in preference to HM. In the present study, two patients were dilated either once or twice, depending on the functional results after the first PD session. In adults, two or three PD sessions are usually attempted before considering the result of endoscopic treatment a failure [13,29,30].

In the present study, two children underwent HM followed by PD. In one case (patient 12), PD was ineffective, and the patient was treated again with a Heller–Dor procedure. Patient 20 (who had Allgrove syndrome) underwent HM at age 15 years, but it failed to improve the symptomatology, although PD after 12 months of follow-up was successful. Two children underwent HM after PD failure: patient 11 showed a reduction in symptoms; patient 21 showed no improvement with either treatment (PD followed by HM). These data suggest that HM and PD can be used as complementary treatments in children as well as in adults [31].

Prognostic factors related to treatment response were also assessed in the present study. However, no differences were found with either patient’s age at diagnosis or treatment, duration of symptoms, presence or absence of associated co-morbidities or severity of the initial clinical presentation. Initial and post-treatment LES measures were not predictive of a successful outcome in our small pediatric series, whereas such a correlation has been suggested in adults [13,14,29,30,32]. Increasing the number of pediatric patients and prospective studies (as carried out in adults) as well as the use of “high-resolution manometry” [33] are needed to confirm our present findings.

Conclusion

Idiopathic esophageal achalasia is a rare pathology, and the available treatments (PD and HM) can only deal with the symptoms. The present study included 22 children with achalasia treated with HM and/or PD. For those children aged under six years, HM remains the treatment of first choice, due to technical reasons, following medical treatment failure. In patients over six years of age, the results with HM and PD were similar (50% complete remission after 24 months of follow-up). This suggests that, in such patients, PD may be considered a first-line treatment before HM. Also, no factors predictive of the response to treatment could be associated co-morbidities or severity of the initial clinical presentation. Initial and post-treatment LES measures were not predictive of a successful outcome in our small pediatric series, whereas such a correlation has been suggested in adults [13,14,29,30,32]. Increasing the number of pediatric patients and prospective studies (as carried out in adults) as well as the use of “high-resolution manometry” [33] are needed to confirm our present findings.

Conflict of interest

None.

References


