Improving the surgery for sigmoid achalasia: long-term results of a technical detail

Enrico Faccani a, Sandro Mattioli a,*, Maria Luisa Lugaresi a, Massimo Pierluigi Di Simone a, Tommaso Bartalena b, Vladimiro Pilotti a

a Department of Surgery, Intensive Care, and Organ Transplantation, Division of Esophageal and Pulmonary Surgery Villa Maria Cecilia e San Pier Damiano Hospitals, Cotignola and Faenza (Ravenna), University of Bologna, Bologna, Italy

b Doctorate in Pneumo-Cardio-Thoracic Sciences of Medical and Surgical Interest, University of Bologna, Bologna, Italy

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Abstract

Objective: Heller myotomy results for the treatment of sigmoid achalasia are worse than those achieved for fusiform achalasia. We retrospectively examined two groups of sigmoid achalasia patients, in which we performed (1) the standard Heller–Dor procedure (no pull-down) and (2) the Heller–Dor plus a technique apt to obtain the verticality of the oesophageal axis (pull-down). We verified whether the latter technique improved long-term results. Materials and methods: We considered 33 patients affected by primitive oesophageal sigmoid achalasia operated upon consecutively (1979–2005). Diagnosis was based on symptoms, manometry, radiology and endoscopy. After 1987, we routinely isolated 360° of the gastro-oesophageal junction and the lower oesophagus and applied U stitches at the right side of the lower oesophagus to pull down and rotate the gastro-oesophageal junction toward the right. Fifteen patients underwent the no pull-down and 18 patients underwent the pull-down technique. Postoperative follow-up included objective clinical and instrumental evaluation (questionnaire filled by a surgeon including the assessment of symptoms and endoscopic reflux oesophagitis according to a semi-quantitative scale) and subjective evaluation (self-evaluation SF-36 questionnaire). Results: The mean follow-up period was 89 months (range 12–261 months). The postoperative dysphagia score was significantly improved in the entire group. Excellent results were present in 12 patients (36.4%), good in 11 (33.3%), fair in 3 (9.1%) and insufficient in 7 patients (21.2%). No statistically significant differences were observed between the two groups with regard to the postoperative symptoms and oesophagitis. Postoperative radiological measurements of oesophageal diameter and residual barium column were significantly improved in the whole group and within each group with respect to the radiological variables measured preoperatively (p = 0.000). In the comparison of the two groups, statistically significant differences were observed with regard to mean oesophageal diameter (p = 0.030) (pull-down, 4 ± 0.9 cm; no pull-down, 4.7 ± 0.6 cm) and residual barium column (p = 0.048) (pull-down, 6.2 ± 3.4 cm; no pull-down, 9.6 ± 5.8 cm). Conclusions: The Heller–Dor operation is effective in the presence of sigmoid achalasia. The clinical objective and subjective evaluations show a trend toward the improvement of results with the pull-down technique. Stronger statistical significance would probably be obtained from a larger case series.

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1. Introduction

Surgical therapy is the gold standard for the treatment of oesophageal achalasia. The Heller myotomy with anterior fundoplication is a good surgical procedure that has achieved satisfactory results in more than 80% of patients [1–5] and significant improvement in health-related quality of life [6–11].

Long-term results in the presence of a sigmoid achalasic oesophagus, which represents the most advanced stage of disease, are not as good as those achieved when dilation of the oesophagus is less pronounced and the organ preserves its axial shape [12]. Some surgeons recommend myotomy as the initial treatment and reserve oesophageal resection for patients with persistent symptoms [13]. Others believe that marked oesophageal dilation and redundancy predict the im possibility of improving emptying by means of simple myotomy and recommend oesophagectomy as the first-line treatment [12,14–16].

The aim of this study was to retrospectively analyse the long-term results of a patient cohort affected by sigmoid achalasia subjected to Heller myotomy associated with a Dor

* Corresponding author. Address: Università degli Studi di Bologna, Dipartimento di Discipline Chirurgiche, Rianimatorie e dei Trapianti, Via Massarenti 9, 40138 Bologna, Italy. Tel.: +39 051 6364870; fax: +39 051 347431.

E-mail address: sandro.mattioli@unibo.it (S. Mattioli).

the lower oesophageal sphincter (LOS) associated with abolishing the two components, oesophageal and gastric, of gastric myotomy according to Heller, and was aimed at pull-down group), which consists of an anterior oesophago-

range 35—76 years), a standard procedure was performed (no left, outside of the oesophageal axis. Prior to this date, the and the distal oesophagus was particularly kinked toward the 46>

vs. 33 patients affected with sigmoid achalasia, who were diagnosed when the diameter of the oesophagus was >6 cm and the distal oesophagus was kinked toward the left, outside of the oesophageal axis.

Since 1987, verticalisation of the oesophageal axis (pull-down technique) was routinely associated with the Heller—Dor procedure when the oesophageal diameter was >6 cm and the distal oesophagus was particularly kinked toward the left, outside of the oesophageal axis. Prior to this date, the pull-down technique was performed only in one case.

In 15 patients (7 men and 8 women, mean age 61 years, range 35—76 years), a standard procedure was performed (no pull-down group), which consists of an anterior oesophago-

gastric myotomy according to Heller, and was aimed at abolishing the two components, oesophageal and gastric, of the lower oesophageal sphincter (LOS) [18] associated with the anterior hemifundoplication according to Dor [19]. The phreno-oesophageal membrane was divided anteriorly and the anterior wall of the stomach was pulled down by the first assistant. The pull-down technique was conducted (pull-down group) in 18 patients (7 men and 11 women, mean age 61 years, range 23—91 years). The gastro-oesophageal junction was fully isolated, the oesophagus was circled by a string and the lower mediastinal oesophagus was isolated for at least 6 cm. Prior to performing the Heller—Dor procedure, two or more U intramuscular stitches were applied at the level of the oesophageal curling on the right side of the oesophagus. The anterior and posterior vagus nerves were isolated distally to completely mobilise the lower oesophagus and the gastro-oesophageal junction. (b) The sutures were tied in order to shorten the right profile of the oesophagus and to rotate toward the right side the gastro-oesophageal junction. (c) After the pull-down technique is completed, the anterior oesophago-gastric myotomy according to Heller is performed. (d) Anterior hemi-

dfundoplication according to Dor.

During the clinical examination performed 3 months after surgery, an objective evaluation and a subjective evaluation (SF-36 questionnaire) were conducted (including an assessment of reflux symptoms, dysphagia and the evidence of reflux oesophagitis).

Symptoms associated with dysphagia and gastro-oesophageal reflux were evaluated according to a semi-quantitative scale. For dysphagia (D), the classification criteria proposed by Van Trappen and subsequently modified were used (D0 = absence of symptoms; D1 = sticking of solid foods or liquids two to four times a month; D2 = sticking of solid foods or liquids two to four times a week; D3 = sticking of solid foods or liquids on a daily basis). To assess the gastro-oesophageal reflux symptoms (RS), the modified Visik criteria were adopted (RS0 = absence of symptoms; RS1 = spontaneous or postural retrosternal heartburn or pain and/or regurgitation occurring two to four times a month; RS2 = spontaneous or postural retrosternal heartburn or pain or regurgitation occurring two to four times a week associated or not with occasional aspiration; RS3 = spontaneous or postural retrosternal heartburn or pain and regurgitation occurring on a daily basis associated or not with frequent aspiration) [5,17,20].

Radiological examination was performed with the patient standing upright for the four orthogonal projections and lying down for the prone OPS projection with a constant focus-film distance of 105 cm. The maximal oesophageal diameter and the air fluid barium column height were measured in centimetres when the cardia closed after barium swallow [19]. Postoperative measurements were compared to the preoperative ones.

The oesophagogastroscopic examination (EGDS) was completed by taking biopsies aimed at determining the presence of reflux oesophagitis (OE), its complications (Barrett’s oesophagus, stenosis, ulcers, etc.) and areas suspected for dysplasia or tumour [17,20].

Oesophagitis was assessed by adopting the modified Savary—Miller endoscopic classification of reflux oesophagitis and the criteria proposed by Ismail—Beigi [17] (E0 = normal; E1 = hyperaemia, oedema and/or histology positive for reflux oesophagitis; E2 = single or multiple non-confluent erosions;
E3 = multiple confluent erosions; E4 = deep ulcers, stenosis, Barrett’s oesophagus).

The presence of Barrett’s oesophagus was diagnosed macroscopically and microscopically on the basis of the histological identification of the columnar-lined oesophagus beyond the Z line. Until 1995, it was classified according to the Bremner criteria: (stage 1) slight replacement, (stage 2) development of Barrett’s mucosa extending for less than 3 cm, (stage 3) development of Barrett’s mucosa extending for more than 3 cm and circumferentially [17,20]. The presence of Barrett’s oesophagus was subsequently classified by extension as short Barrett’s oesophagus (<3 cm) and long Barrett’s oesophagus (>3 cm) [21]. Dysplasia was defined as mild, moderate or severe [17,20].

The SF-36 questionnaire is comprised of eight domains with multiple-choice answers, each measuring specific health domains: physical functioning (PF), restrictions in activities due to physical or emotional health problems, role physical (RP) and role emotional (RE), bodily pain (BP), general health (GH), vitality (energy/tiredness) (VT), mental health (MH) and social functioning (SF).

The questionnaires are designed to be self-administered and were sent to the patients by post or completed independently when the patients attended the hospital for their clinical visit.

On the basis of the assessment of the symptoms and reflux oesophagitis, the overall results of the operation were classified as excellent to insufficient according to a semi-quantitative scale (D0, RS0, OE0 = excellent; D1, RS1, OE1 = good; D2, RS2, OE2-4 = fair; D3, RS3, OE2-4 = poor) [20].

2.1. Statistical analysis

The Mann–Whitney U-test was used to compare the ordinal qualitative variables between the two groups of patients. Wilcoxon signed ranks test was used to compare pre- and postoperative data. The χ²-test was used to evaluate nominal qualitative variables.

Data were expressed as mean values unless stated otherwise. Student’s t-test for unpaired and paired data was adopted for the analysis of radiological data as appropriate.

The relationship between the objective and subjective data was assessed by linear regression analysis.

A probability of <5% was considered statistically significant (p < 0.05).

Statistical analyses were performed using SPSS 12.00 software package (SPSS Inc., Chicago, IL).

3. Results

Preoperative severe dysphagia was present in all 33 patients, while reflux symptoms were present in 20 patients: slight in 14 and moderate in 5. Preoperative reflux oesophagitis was absent in all patients (Table 1).

Mean preoperative radiological oesophageal diameter was 6.4 ± 0.5 cm (range 5—9 cm) and mean preoperative residual barium column was 20 ± 4.4 cm (range 14—30 cm).

Preoperatively, no statistically significant differences were observed between the two groups of patients with regard to mean age, sex distribution, mean preoperative radiological diameter (pull-down, 6.3 ± 0.3 cm; no pull-down, 6.5 ± 0.7 cm) (p = 0.399) and residual barium column (pull-down, 19.2 ± 1.4 cm; no pull-down, 21 ± 6.3 cm) (p = 0.233) (Table 1).

The mean postoperative stay was 5.6 ± 1.6 days (range 4—10 days). In the no pull-down group, the mean stay was 5.1 ± 1.2 days (range 4—7 days), while in the pull-down group, it was 5.8 ± 1.8 days (range 4—10 days).

Perioperative mortality was 0 and morbidity was 6% (two pull-down patients) without the need for revision surgery. In one case, a condition of severe dysphagia associated with regurgitations was resolved through medical therapy with prokinetics, and in the other patient, radiological blinding leakage of contrast medium at the myotomy site without any associated symptom healed after a period of liquid diet.

The mean follow-up was 89 months in the entire group (range 12—261 months), while it was 101 months (range 12—261 months) in the no pull-down group and 78 months (range 12—234 months) in the pull-down group (not significant).

In Fig. 2, the box plot graphic of the score of pre- and postoperative dysphagia in the whole group is shown (see Fig. 2). Dysphagia was absent (D0) in 17 patients (51.5%) (8 no

| Table 1 | \hline
| Preoperative and postoperative objective and instrumental data (see text) |
| --- | --- | --- | --- |
| | Pull-down | No pull-down |
| Preoperative dysphagia | | |
| 0 | — | — |
| 1 | — | — |
| 2 | — | — |
| 3 | 18 (100%) | 15 (100%) |
| Preoperative reflux symptoms | | |
| 0 | — | — |
| 1 | 7 (38.9%) | 7 (46.2%) |
| 2 | 2 (11.1%) | 3 (20%) |
| 3 | — | — |
| Preoperative reflux oesophagitis | | |
| 0 | 18 (100%) | 15 (100%) |
| 1 | — | — |
| 2 | — | — |
| 3 | — | — |
| Preoperative oesophageal diameter | Mean ± SD | 6.3 ± 0.3 cm | 6.5 ± 0.7 cm |
| | Mean ± SD | 19.2 ± 1.4 cm | 21 ± 6.3 cm |
| Postoperative dysphagia | | |
| 0 | — | — |
| 1 | 4 (22.2%) | 6 (40%) |
| 2 | 2 (11.1%) | — |
| 3 | 3 (16.7%) | 1 (6.7%) |
| Postoperative reflux oesophagitis | | |
| 0 | 16 (88.9%) | 13 (86.7%) |
| 1 | 1 (5.6%) | 1 (6.7%) |
| 2 | — | — |
| 3 | 1 (5.6%) | 1 (6.7%) |
| Postoperative oesophageal diameter | Mean ± SD | 4 ± 0.9 cm | 4.7 ± 0.6 cm |
| | Mean ± SD | 6.2 ± 3.4 cm | 9.6 ± 5.8 cm |
pull-down, 9 pull-down), mild (D1) in 10 patients (30.3%) (6 no pull-down, 4 pull-down), moderate (D2) in 2 (6%) (2 pull-down) and severe (D3) in 4 (12.2%) (1 no pull-down, 3 pull-down) (Table 1). Statistically significant differences were detected in the comparison between the pre- and postoperative dysphagia score in the entire group \((p = 0.000)\) (Table 1).

Reflux symptoms were absent (RS0) in 18 (54.6%) patients, slight (RS1) in 12 (36.4%) (3 no pull-down, 9 pull-down), moderate (RS2) in 2 (6%) (1 no pull-down, 1 pull-down) and severe (RS3) in only 1 patient (3%) (no pull-down) (Table 1).

The appearance of reflux oesophagitis, which was absent before the operation, occurred in four patients: one (3%) mild (OE1) (pull-down), one (3%) moderate (OE2) (one no pull-down) and two (6%) severe (OE3) (one no pull-down and one pull-down) (Table 1).

The overall outcome of patients was excellent (OE0-D0-RS0) in 12 patients (36.3%), good (OE0-D1-RS1) in 11 (33.3%), fair (OE1-D2-RS2) in 3 (9.2%) and insufficient (OE2-4-D3-RS3) in 7 patients (21.2%).

In the group of pull-down patients (18 cases), the overall outcome was excellent in 8 (44.4%), good in 6 (33.3%), fair in 2 (11.1%) and insufficient in 2 patients (11.1%).

In the group of no pull-down patients (15 cases), the overall outcome was excellent in 4 (26.7%), good in 5 (33.3%), fair in 1 (6.7%) and insufficient in 5 patients (33.3%).

In comparison to the objective overall outcome, no statistically significant differences were observed between the two groups of patients.

In Fig. 3a the score of the two groups of patients on the self-evaluation performed using the SF-36 questionnaire is depicted. Statistically significant differences between the two groups of patients were detected with regard to the bodily pain domain (mean no pull-down, 96.2; mean pull-down, 71.8) \((p = 0.034)\) and the general health domain (mean no pull-down, 61.7; mean pull-down, 55.4) \((p = 0.044)\).

By applying linear regression analysis to the objective and subjective data, it was possible to show a statistically significant relationship between the postoperative objective assessment of dysphagia and the general health domain of the SF-36 questionnaire \((p = 0.0041)\) (Fig. 3b).

In Fig. 4, the box plot graphic of the pre- and postoperative mean oesophageal diameter (Fig. 4a) and mean residual barium column in the entire group is presented (Fig. 4b). Statistically significant differences were observed between pre- and postoperative measurements \((p = 0.000)\).

In Fig. 5(a, b) the pre- and postoperative diameter and barium column measurements are displayed in the no pull-down and pull-down groups. In both the groups, the pre- and postoperative measurements were statistically different \((p = 0.000)\) (see Fig. 5a, b).

The preoperative mean values of oesophageal diameter and air fluid barium column were similar in the no pull-down and pull-down groups. However, after surgery, the oesophageal diameter \((p = 0.030)\) and residual barium column \((p = 0.048)\) mean values were significantly smaller in the pull-down group (mean oesophageal diameter 4.7 ± 0.6 cm, range 3.5–6.5 cm; mean postoperative residual barium column 6.2 ± 3.4 cm, range 0–12 cm) with respect to the no pull-down group (mean oesophageal diameter 4.7 ± 0.6 cm, range 3.5–6.5 cm; mean residual barium column was 9.6 ± 5.8 cm, range 0–22 cm) (see Figs. 4a, b and 6).

A statistically significant relationship between the postoperative oesophageal diameter and the postoperative objective assessment of dysphagia \((p = 0.0330)\) was obtained by means of linear regression analysis in the whole group (Fig. 7). A similar significant relationship was observed in the pull-down group \((p = 0.0031)\) but not in the no pull-down group.

4. Discussion

Sigmoid oesophagus characterised by a widened (>6 cm diameter) and tortuous oesophageal body resulting in a
sigmoid-shaped appearance represents the most advanced stage of achalasia [22].

In the past, patients with sigmoid achalasia were thought best treated with oesophagectomy rather than myotomy [12,14,15] because it was felt that the dilated and often tortuous aperistaltic oesophagus does not empty sufficiently to improve dysphagia, even when the LOS is disrupted.

Recently, many surgeons have elected to perform the Heller myotomy with anterior fundoplication as a first-choice treatment option, especially after the advent of minimally invasive surgery. Few clinical studies have been published in the literature regarding the surgical treatment and long-term outcomes of sigmoid achalasia. Two recent studies have demonstrated good postoperative results in patients with sigmoid achalasia treated with Heller myotomy [23,24]. The obvious advantage of Heller myotomy is the avoidance of morbidity and mortality associated with oesophagectomy. Patti et al. [23] performed a laparoscopic Heller myotomy on patients with a dilated oesophagus (>6 cm) with straight axis and sigmoid-shaped configuration. They neither reported increased difficulty in performing the surgery nor increase in complications, with 92% of the patients reporting excellent or good relief of dysphagia [23].

Fig. 4. (a) Graphic box plot representation of the preoperative (black bar) and postoperative (white bar) oesophageal diameter scores in the whole group. (b) Graphic box plot representation of preoperative (black bar) and postoperative (white bar) oesophageal residual barium column scores in the entire group.

Fig. 5. (a) Graphic box plot representation of the preoperative (left side) and postoperative (right side) oesophageal diameter scores in the pull-down (grey bar) and no pull-down (white bar) groups. (b) Graphic box plot representation of preoperative (left side) and postoperative (right side) scores of the oesophageal residual barium column in the pull-down (grey bar) and no pull-down (white bar) groups.

Fig. 6. (a) Preoperative barium oesophagogram shows a dilated oesophagus with sigmoid appearance of its lower third. Oesophageal angle between proximal and distal oesophagus long axis is approximately 115°. (b) Postoperative oesophagogram after Heller—Dor myotomy plus pull-down technique shows reduced calibre of the oesophagus and straightening of the oesophageal angle to 150°.
Likewise, Mineo and Pompeo [24] studied 14 patients with sigmoid oesophagus treated with a Heller myotomy. With a median follow-up of 85 months, excellent or good results were reported by 72%. Postoperative dysphagia and regurgitation scores decreased significantly and were equivalent to postoperative scores from a non-dilated oesophagus group undergoing Heller myotomy. Additionally, oesophageal width was found to narrow with time, on an average 10 mm in 24 months. Health-related quality of life evaluated with the SF-36 questionnaire showed statistically improved general health, social function and mental health [24].

This study evaluates the long-term results of patients who underwent surgical therapy for oesophageal sigmoid achalasia with a carefully coded follow-up continued for more than 25 years by our group [5,17,20].

Dysphagia, gastro-oesophageal reflux symptoms and reflux oesophagitis that persist or appear after the operation are the parameters most commonly used to evaluate the results of oesophageal achalasia surgical treatment [17,23,25].

In the present study we considered only patients affected by sigmoid achalasia who underwent in the first instance the Heller—Dor operation. However, it may be interesting to report that 4 of the 33 patients had been offered oesophagectomy in consideration of the size of their oesophagus or their young age and in one case because he had recurrent infection secondary to oesophageal stasis, but they did prefer a more conservative surgery. In the same period 10 more patients underwent oesophagectomy, 9 for cancer implanted in their mega-oesophagus after one or more myotomies and 1 for postsurgical scar stenosis of the gastro-oesophageal junction.

The long-term overall outcomes of the 33 patients, obtained with a mean follow-up of 88.5 months of the objective evaluations of symptoms and oesophagitis, were excellent or good in 23 patients (69.7%), fair in 3 (9.1%) and insufficient in 7 patients (21.2%). Postoperative dysphagia score and radiological measurements of oesophageal diameter and residual barium column improved significantly.

The insufficient overall outcomes in seven patients (21.2%) were due to the persistence of severe dysphagia and the appearance of reflux oesophagitis, absent before the operation and always associated with reflux symptoms.

The overall outcome of the objective evaluation of symptoms and oesophagitis in the pull-down group showed a positive trend with respect to those obtained in the no pull-down group. Also, these data may not have been significant because of the small sample size.

In the analysis of the self-evaluation assessment using the SF-36 questionnaire, significantly better scores were observed in the bodily pain and general health domains in the no pull-down group with respect to pull-down patients.

The no pull-down patients appeared to perceive symptoms less severely than pull-down patients, as detected by evaluating variables associated with bodily pain and general health domains.

However, significantly worse objective parameters of disease were noted, as detected by the assessment of radiological variables.

A possible interpretation of the disagreement between the objective and subjective evaluation could be that no pull-down patients with a longer follow-up (mean 101 months) with respect to those of the pull-down group (mean 78 months) could have developed a trend to tolerate subjective sensations due to visceral pain (bodily pain domain) and the state of general health (general health domain) perceptions.

A strong inter-relationship between the objective and subjective evaluation criteria, highlighted by the statistically significant relationship detected by means of the linear regression analysis between the postoperative dysphagia score and general health domain of the SF-36 questionnaire, was observed. The postoperative score of dysphagia increased inversely relative to a decrease in the general health domain score.

The postoperative radiological parameters, oesophageal diameter and residual barium column significantly decreased after surgery with respect to those preoperatively measured in the entire group and inside each group. In the comparison between the two groups, the mean postoperative radiological values of the pull-down group were significantly better relative to those of the no pull-down group; preoperatively, no significant differences were detected between the two groups.

These radiological objective values prove that the pull-down technique associated with Heller—Dor procedure is more effective than the Heller—Dor procedure alone for the surgical treatment of oesophageal sigmoid achalasia.

Further evidence of the strong inter-relationship between the objective clinical and instrumental variables is presented by the significant relationship between postoperative oesophageal diameter and postoperative objective assessment of dysphagia. These data show that the oesophageal diameter increased progressively in direct relation to increases in the dysphagia score. A similar significant relationship was observed in the pull-down group (p = 0.0031) but not in the no pull-down group.

In conclusion, the Heller—Dor operation does represent a reasonable option for the first-instance surgical treatment of sigmoid achalasia; the pull-down technique associated with a Heller myotomy and anterior hemifundoplication according to Dor may be the primary treatment option in patients affected with sigmoid achalasia.
The objective overall clinical and instrumental results suggest that the pull-down technique is more effective than the Heller—Dor procedure alone.

We offer oesophagoscopy for cases of sigmoid achalasia (a) in patients younger than 55 years, (b) with severe mucosal inflammation or moderate-to-severe dysplasia [20]. The frequency of cancer arising in sigmoid achalasia is not definitively defined; however, the data collected to date (17) may not force surgeons to oesophagoscopy in the absence of dysplasia, mainly if the surgeon has inadequate specific experience. According to empirical criteria, we routinely perform oesophagoscopy associated with multiple target and random biopsies of oesophageal mucosa at least every 5 years in cases of persisting macro- and/or microscopic mucosal inflammation due to food stasis or postoperative gastro-oesophageal reflux [17]. After 5 years, the follow-up protocol should be primarily endoscopic and histologic for the early identification of reflux oesophagitis and dysplasia. In patients with sigmoid oesophagus, endoscopy should be performed after 3–5 days of a liquid diet and after lavage of the oesophagus [17].

A larger multicentric study would serve to confirm the data provided with this study and to elucidate more on this interesting topic.

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