Long-term Evaluation of Modified Lateral Anorectal Myomectomy for Low-Segment Hirschsprung Disease

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Objectives: To provide a simple myomectomy technique for low-segment Hirschsprung disease and evaluate the efficacy of the new modification.

Design: Case series of 19 patients followed up for 12 to 56 months (mean, 39.1 months).

Setting: Tanta University Hospital, Tanta, Egypt.

Participants: Nineteen patients aged 4 months to 10 years complaining of chronic constipation, with radiological and clinical data suggestive of low-segment Hirschsprung disease proven by histological examination.

Intervention: Modified lateral anorectal myomectomy.

Main Outcome Measures: Clinical and radiological improvement measured by postoperative barium enema, bowel habits, and patient’s relief of symptoms.

Results: Seventeen of 19 patients improved clinically and 13 showed radiological improvement 3 years postoperatively. There was poor response in 2 patients, who were subjected to further Soave procedures.

Conclusion: Modified lateral anorectal myomectomy is an effective and technically simple procedure in patients suspected of having low-segment Hirschsprung disease.

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Hirschsprung disease (HD) occurs in 1 in 5000 live births, as a result of arrest of the embryogenic invasion of the gut by ganglion cells cephalocaudally between the 6th and 12th weeks of gestation. About 30% of all HD patients have agangliosis restricted to the anal canal and lower rectum. Anorectal myomectomy is an effective and less extensive operation for this group. Moreover, the procedure provides good muscle biopsy specimens for diagnosis.

Various procedures have been described for the surgical treatment of this type of HD: posterior excisional anorectal myotomy, submucous sphincterectomy, transanal posterior myectomy, or the posterior sagittal approach. Although these techniques have nearly the same principle, they differ in the technical details.

Anorectal myomectomy could also be useful in the treatment of other causes of chronic constipation in children, as it is a safe procedure and leads to satisfactory results in approximately 90% of cases.

Our study was carried out to evaluate the new modification of lateral anorectal myomectomy for this group of patients, looking especially at clinical outcome and the postoperative radiological changes in barium enema examination.

RESULTS

The study group included 12 boys and 7 girls with a mean age of 28 months (range, 4 months to 10 years). In 3 cases other congenital anomalies were present (2 ventricular septal defects and 1 subcoronal hypospadias).

Preoperative barium enema examination showed rectal dilatation in all cases, while the narrow segment was detected in 11 cases with low cone position. The length of the muscle strip ranged from 3.5 to 7 cm. Histopathological examination of the muscle strips revealed aganglionosis in all cases; normal ganglia were seen at the proximal end of the strip only in 11 cases. Postoperative radiological results are presented in Table 1.

No immediate postoperative complications were reported. During follow-up,
PATIENTS AND METHODS

Nineteen children were selected among infants and children (younger than 12 years) who presented to the pediatric surgical clinic of Tanta University Hospital, Tanta, Egypt. They were screened on the basis of clinical history, examination results, and preoperative barium enema study. The group suspected of having low-segment HD by clinical and radiological data was included in the study. Each subject’s guardian gave informed consent preoperatively.

Each patient underwent preoperative barium enema without preparation, then underwent modified lateral anorectal myomectomy for diagnostic and therapeutic purposes as described by El-Banna et al:

- Short preoperative colonic preparation with enemas.
- Under general anesthesia, with the patient in lithotomy position, an anal speculum was applied and opened in the sagittal plane to stretch the left lateral wall of the anal canal. A small transverse incision was made at the mucocutaneous junction at the 3-o'clock position for 2 to 2.5 cm in length.
- Dissection of the internal sphincter from the mucosa medially and from the part of the external sphincter laterally was performed at the intersphincteric plane. The dissection was extended proximally to include the rectal muscle as much as possible.
- Excision of a 0.5- to 1-cm-wide strip from the dissected muscle, with its distal end including a part of the internal anal sphincter.
- Hemostasis and temporary anal packing with sutureless healing.
- Oral feeding with fluids was begun 6 hours postoperatively and the patient was discharged from the hospital 24 hours later.
- Histopathological examination of the muscle biopsy specimen.
- Follow-up protocol included a questionnaire regarding bowel habits, complications, and symptom relief, administered monthly in the first 6 months and every 3 months thereafter. A postoperative barium enema was performed 2 months after myomectomy, after 6 to 12 months, then once yearly for the next 3 years to evaluate the results of surgery.
- Follow-up ranged from 12 to 56 months. The results of the study were compared with literature reports of other techniques of anorectal myomectomy.

5 patients developed recurrence of symptoms within the first 3 months, which was treated with anal dilatation. Two of these patients had treatment failure (1 developed postmyomectomy enterocolitis) and underwent further Soave procedures within 1 year of the myomectomy, without any technical difficulties caused by the previous myomectomy. The other 3 patients responded well to dilatation, but 1 developed a perianal abscess at the myomectomy site. This patient underwent drainage but developed a low perianal fistula that was surgically excised 6 months later.

The 3 patients with preoperative encopresis completely recovered. No patient experienced incontinence or anal stricture following the procedure. Patient satisfaction regarding symptom relief and regular bowel habits is presented in Table 2.

COMMENT

Chronic constipation in infants and children is becoming more prevalent. It is time to take another look at the role of anorectal myomectomy in HD.

Constipation was the main symptom in our study, which is in agreement with previous studies. However, there is a group of HD patients with nonclassic forms of the disease regarding time of presentation (7 of our patients had constipation when solid food was introduced after a normal neonatal course) or regarding a good clinical response to minor procedure. Preoperative barium enemas revealed the cone only in 11 cases, while the cone was seen in 4 of 7 patients studied by Backwinkel et al and in 2 of 11 in Nissan et al. Six to 12 months postoperatively, 10 of 19 patients showed radiological improvement and 13 of 18 after 3 years in our study, while Nissan et al reported improvement in 7 of 11 cases 1 year after myomectomy.

Histopathological examination of the excised muscle strips demonstrated the transitional zone of nor-

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### Table 1. Radiological Data 6 to 12 Months and 3 Years Postoperatively

<table>
<thead>
<tr>
<th>Postoperative Barium Enema Results</th>
<th>No. of Cases at 6 to 12 mo</th>
<th>No. of Cases at 3 y</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>Decreased RCD*</td>
<td>6</td>
<td>5</td>
</tr>
<tr>
<td>Same as preoperatively</td>
<td>9</td>
<td>5</td>
</tr>
<tr>
<td>Not done</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>19</td>
<td>19</td>
</tr>
</tbody>
</table>

*RCD indicates rectocolonic dilatation.

### Table 2. Response of Patients to the Operative Procedure Regarding Symptom Relief and Bowel Habits

<table>
<thead>
<tr>
<th>Time of Postmyomectomy Evaluation</th>
<th>Response</th>
<th>1 mo</th>
<th>3 mo</th>
<th>1 y</th>
<th>3 y</th>
</tr>
</thead>
<tbody>
<tr>
<td>Excellent</td>
<td>15 (79)</td>
<td>12 (63)</td>
<td>14 (74)</td>
<td>15 (83)</td>
<td></td>
</tr>
<tr>
<td>Fair</td>
<td>1 (5)</td>
<td>3 (15)</td>
<td>3 (16)</td>
<td>1 (6)</td>
<td></td>
</tr>
<tr>
<td>Poor</td>
<td>3 (16)</td>
<td>4 (22)</td>
<td>2 (10)</td>
<td>2 (11)</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>19 (100)</td>
<td>19 (100)</td>
<td>19 (100)</td>
<td>19 (100)</td>
<td></td>
</tr>
</tbody>
</table>

*Measured by criteria for frequency and quality of defeation. Excellent indicates defeation more than once daily without symptoms; fair, defeation at least once every 2 days without symptoms; and poor, defeation once every 3 days or less with symptoms. All values are given as number (percentage) of patients.
nal ganglia in 11 of 19 cases, which is similar to the report of Udassin et al.\textsuperscript{14} Although the existence of low-segment HD is still controversial to some surgeons, our results confirmed its presence as the length of the myomectomy strips was over 3 cm, consistent with the results of Sawin et al.\textsuperscript{3} Some patients improved even though the muscle biopsy specimen showed no ganglia along its whole length. Possibly the remaining aganglionic segment is too short to cause further symptoms or this group represents a subgroup of sphincteric dysfunction group among HD patients. Seventeen patients had a good clinical response without laxative use and 2 patients showed poor improvement and underwent further Soave procedures; however, our failure rate is still less than that of Scobie and Mackinlay\textsuperscript{15} or Thomas et al.\textsuperscript{16}

Anal stricture\textsuperscript{13} and incontinence\textsuperscript{17} have both been reported as complications of myomectomy; however, no case of soiling, stricture, or incontinence was seen in our patients.

The 2 patients who underwent the Soave procedure were doing very well 3 and 3.5 years postoperatively because the aganglionic rectal cuff was already myomectomized. Some authors have suggested that myomectomy may be needed after definitive procedures, as recommended after endorectal pull-through,\textsuperscript{18,19} in the form of a secondary operation when symptoms recur,\textsuperscript{20} or as an additive step to the anterior resection for long-segment cases.\textsuperscript{21}

The usefulness of diagnostic myomectomy to provide the full-thickness muscle biopsy specimen is established. Further intervention with other procedures is not affected by the modified myomectomy and a considerable group of patients responded well to this procedure.

Although the number of our patients was small, this new modification of lateral anorectal myomectomy is an excellent diagnostic procedure. It is also useful as a primary therapeutic procedure for selected short- and ultrashort-segment cases of HD (low type), as a secondary technique following definitive techniques in long-segment cases, or even as a preliminary procedure with follow-up, as proved beneficial to this selected group of patients with HD.

**REFERENCES**