Adults Born with High Anorectal Atresia—How Do They Manage?

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PURPOSE: We are interested in the way patients, who underwent surgery for high anorectal atresia, control their defecation. Considering that some patients, despite newer operative techniques, always will suffer from minor or major soiling we attempted to find some guidelines for post-operative support for future patients. METHOD: Fifty-eight patients (median age, 26 (range, 18.1-56.9) years) were personally interviewed. RESULTS: Regulating defecation is done in five different modes: 16 patients have stools after urge, 15 control their stools mainly by going to the toilet at regular times, 18 perform bowel-irrigations or use enemas, 2 have loss of feces continuously, and 7 patients have an ileostomy or colostomy. More than one-half of patients influence their defecation by diet. Of the patients with anal defecation, 6 never soil, 39 sometimes soil small amounts, and 6 often soil seriously. Eighteen patients occasionally suffer from constipation. There is no mode of defecation regulation outstanding in preventing soiling or constipation. However, patients who do not regulate defecation somehow suffer from serious soiling. Most patients are content with their level of cleanliness.1 We are interested in how patients are able to control/regulate defecation to cope with their incontinence and we hope to learn from the older generation how they manage. Questions occur if and how these patients are able to achieve control over their defecation? How do they cope with the [possible] problem of soiling and how do they manage to evacuate feces sufficiently?

During the last ten years, operative treatment has changed drastically and is performed at the moment exclusively by well-trained pediatric surgeons. Still a part of this newer generation of anal atresia patients will be incontinent for feces. Despite newer operative techniques, we have to know how patients are able to regulate their defecation to cope with their incontinence, and we hope to learn from the older generation how they manage. Questions occur if and how these patients are able to achieve control over their defecation? How do they cope with the [possible] problem of soiling and how do they manage to evacuate feces sufficiently?

PATIENTS AND METHODS

From clinical records of pediatric surgical centers in the Netherlands and from summons by patient orga-
nizations, 80 patients who are 18 years of age and older and who were operatively corrected for high anorectal atresia are gathered. Patients were approached by phone or by mail and asked to join a study looking for the consequences on their lives of being born with a high anorectal atresia. Ten patients refused for reasons of not wanting to talk about their problems, too busy, having had bad experiences with hospitals, etc., eight refused without giving a reason, one was lost to follow-up after initially consenting to participate, one was hospitalized at time of investigation because of cardiac failure, and two mentally handicapped patients were excluded from the study. Finally, 58 patients entered the study, and their experiences are described here.

The study group consists of 39 males and 19 females who have a median age of 26 (range, 18.1–56.9) years. The first operation to correct the high anorectal atresia was done by perineal (n = 12), abdominoperineal (n = 38), or sacroabdominoperineal (n = 6) approach. In two patients the exact approach could not be traced. Twenty-five patients underwent surgery more than once to improve fecal continence (Table 1).

In a personal interview, a detailed questionnaire is completed. Questions pertained to the way defecation is regulated. Of the patients who have anal defecation (i.e., without an ileostomy or colostomy), soiling and constipation are evaluated from questions about frequency and amount of loss of feces, frequency of bowel movements, and consistency of feces. Also, the feeling of urge (always, sometimes, never), discrimination (complete, incomplete), and holding back (more than 5 minutes, between 1 and 5 minutes, impossible) are investigated in these patients. All patients are asked whether they are content with their current level of cleanliness (not, moderate, considerable, very).

Patients are classified according to their main mode of defecation regulation. For each group, soiling and constipation are described to evaluate the effectiveness of used mode and contentedness with their level of cleanliness.

RESULTS

Regulation of Defecation (All 58 Patients)

Defecation is regulated in five different modes: 16 patients have stools after urge, 15 patients control their stools by going to the toilet at regular times (regularity; some of whom in combination with going after urge), 18 patients use bowel-irrigations or enemas to control defecation, 2 patients have loss of feces continuously and wear inlays/diapers, and 7 patients have an ileostomy or colostomy. Besides, more than one-half of all patients reported avoiding or eating constipating or laxative foodstuffs, i.e., taking extra dietary measures as well. Five patients use laxative or constipating oral medication sometimes, whereas daily use of oral medication is reported by two patients (1 uses bisacodyl in addition to regulating defecation by regularity, and 1 uses loperamidehydrochloride in addition to the use of enemas). Nobody uses oral medication as the only manner of regulating defecation. Four patients reported the use of enemas sometimes in addition to their main mode of regulation. Frequency of using bowel irrigations or enemas as main mode of regulation varies from daily or every other day (n = 14) to once or twice per week (n = 4).

Soiling and Constipation (51 Patients with Anal Defecation)

Six patients never experience soiling. However, 39 patients sometimes experience small amounts of soiling and 6 patients often have serious soiling. Overall, 33 patients never experience any form of constipation, whereas 18 patients have occasionally difficulties with evacuation of feces. Three patients use bowel irrigations or enemas mainly to prevent constipation. Absolute numbers and percentages for soiling and constipation according to each mode of regulation are given in Table 2.
Table 2.
Controlled Defecation on Toilet vs. Uncontrolled Defecation in Underwear and Constipation (in 51 Patients with Anal Defecation)

<table>
<thead>
<tr>
<th></th>
<th>All in Toilet/Never Soils (%)</th>
<th>Mostly in Toilet/Sometimes Soils (%)</th>
<th>Less than One-Half in Toilet/Often Soils (%)</th>
<th>Constipation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Urge</td>
<td>1 (6)</td>
<td>13 (81)</td>
<td>2 (13)</td>
<td>10 (63)</td>
</tr>
<tr>
<td>Regularity</td>
<td>3 (20)</td>
<td>11 (73)</td>
<td>1 (7)</td>
<td>8 (53)</td>
</tr>
<tr>
<td>Bowel irrigation/enemas</td>
<td>2 (11)</td>
<td>15 (83)</td>
<td>1 (6)</td>
<td>14 (78)</td>
</tr>
<tr>
<td>No regulation</td>
<td>—</td>
<td>—</td>
<td>2 (100)</td>
<td>1 (50)</td>
</tr>
<tr>
<td>Total</td>
<td>6 (12)</td>
<td>39 (76)</td>
<td>6 (12)</td>
<td>33 (65)</td>
</tr>
</tbody>
</table>

Table 3.
Numbers of Patients Who Are Content with Their Current Level of Cleanliness According to Their Mode of Control

<table>
<thead>
<tr>
<th>Soiling</th>
<th>All in Toilet/ Never Soils</th>
<th>Mostly in Toilet/ Sometimes Soils</th>
<th>Less than One-Half in Toilet/ Often Soils</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Urge</td>
<td>1 (1)</td>
<td>10 (13)</td>
<td>2 (2)</td>
<td>13 (16)</td>
</tr>
<tr>
<td>Regularity</td>
<td>2 (3)</td>
<td>9 (11)</td>
<td>1 (1)</td>
<td>12 (15)</td>
</tr>
<tr>
<td>Bowel irrigation/enemas</td>
<td>2 (2)</td>
<td>14 (15)</td>
<td>— (1)</td>
<td>16 (18)</td>
</tr>
<tr>
<td>No regulation</td>
<td>—</td>
<td>—</td>
<td>1 (2)</td>
<td>1 (2)</td>
</tr>
<tr>
<td>Colostomy or ileostomy</td>
<td>x</td>
<td>x</td>
<td>x</td>
<td>6 (7)*</td>
</tr>
<tr>
<td>Total</td>
<td>5 (6)</td>
<td>33 (39)</td>
<td>4 (6)</td>
<td>48 (58)*</td>
</tr>
</tbody>
</table>

* Of one patient, it is unknown whether she is content with her level of cleanliness. Numbers between parentheses reflect total number of patients in the group.

Effectiveness of Regulation (51 Patients with Anal Defecation)

Effectiveness of regulation of defecation is evaluated with respect to soiling and constipation. No differences between mode of regulation and effectiveness were observed. With respect to soiling, all groups show a considerable number of patients who experience some soiling and one or two patients who have serious soiling. Both patients who do not regulate their defecation have serious soiling. Patients who perform bowel irrigations or use enemas suffer the least from constipation (Table 3).

Urge, Discrimination, and Holding Back (51 Patients with Anal Defecation)

Twenty-nine patients reported always feeling the urge to defecate, 16 patients experience it sometimes, and 6 patients never experience any feelings of urge. Complete discrimination of stools and flatus is experienced by 24 patients, although in 27 patients discrimination is impaired. Nineteen patients indicated they were able to hold back feces longer than five minutes, 16 were able to do so between one and five minutes, and 16 were not able to hold back feces at all. We could not find a correlation between the presence of urge, discrimination, and holding back of feces and chosen mode of regulation of defecation.

Contentedness with Current Level of Cleanliness (All 58 Patients)

Of the whole group, 48 patients were (reasonably or very) content with their level of cleanliness, although 10 patients indicated they were not satisfied or only moderately content (Table 3). Of the ten patients who were not content, four had the idea that they tried everything to obtain a better level of cleanliness but did not consider colostomy and four thought they did not try everything and even considered colostomy once. One patient admitted he did not try everything to achieve better cleanliness and did not consider colostomy. One patient who did not soil at all was not
content, because he has to prevent soiling by using the toilet once or twice after defecation.

**DISCUSSION**

In this series of 58 adult patients who had formerly undergone surgery for high anorectal atresia, we found five different main modes of regulating defecation. As we reported previously, these patients came to their final mode of defecation regulation at a median age of 15 (range, 5-31) years. Except for patients with an ileostomy or colostomy, no matter what mode of regulation is chosen, many patients sometimes soil small amounts, and a few often soil seriously. However, patients who do nothing to regulate defecation soil much more frequently. When patients sometimes suffer from soiling, this occurs, especially when the consistency of feces is (very) loose or under special circumstances like stress, sports-activities, or heavy lifting. In many cases, evacuation did not seem to be an important problem. In view of the anatomic defects, the impaired feeling of urge, discrimination, and ability of holding back feces, one can argue whether these results are optimal for these patients or that they could be better.

Most patients with anal defecation indicate that they are content with the current level of cleanliness. Apparently the patients themselves consider the way they regulate defecation at present as acceptable. Even four of the six patients who suffer greatly from soiling are content. The fact that patients are content with their current level of cleanliness is for some a reflection of acceptance of their handicap, because they suppose that there seems no better way. Others, however, have taken a defeatist attitude. They told us they think that new medical advice is of no use.

Of our population, only one patient is successful in remaining clean without any adjustment to prevent soiling. This indicates that the vast majority of patients with high anorectal atresia (almost) daily will be confronted with preventing the loss of feces and being aware that soiling might happen. Therefore, these patients should be considered as people who have to cope with a (major or minor) handicap. However, the adults in our study had to learn their defecation regulation, more than one-half of them without professional support, by trial and error. Also, Vandvik and Ødegård reported that toilet training in children with anal atresia often is less consistent than in children without anatomic defects, because it is unclear for the parents what can be expected or the parents hope the child will "outgrow" it. These facts indicate that, in the past, patients with high anal atresia did not receive adequate instructions and have been left to their own resources. Knowing this, we wonder whether it would be possible to improve defecation regulation and minimize soiling for more patients. Perhaps optimal results could also be achieved at younger ages than currently possible. We suggest that results will improve by (re)-teaching patients to empty their bowel appropriately, according to a defecation regulation program, optimally adjusted to each individual patient. For some patients, it might be helpful to learn optimum self-control by a behavior modification program in a multidisciplinary approach (medical and behavioral), for example, as described by Shepherd et al. This biofeedback could be a useful additional method to teach patients to apply proper defecation dynamics and holding-up techniques. Other patients might be helped by a structured bowel management program using bowel irrigation or enemas. Peña described his experience with patients who suffer from fecal incontinence after a previous operation for imperforate anus. He postulated that 95 percent of his patients improved dramatically their quality of life and most of the time remain clean by applying a well-instructed bowel management program. An easier and more effective way might be the newer form of antegrade colonic enema by a Malone stoma.

To achieve better fecal continence, one could also consider additional surgery. However, we learned from the literature and from our own experiences that results of additional surgery such as levatorplasty or graciloplasty are uncertain. Nevertheless, some patients may improve by additional posterior sagittal anorectoplasty operation. Recently, Baeten et al. reported a new technique using electrostimulated graciloplasty, claiming continence in five of nine patients with anorectal atresia. However, this method still must prove its value over a longer period of time.

When all conservative treatments have been tried and continence-improving operations have been considered or performed and still results are not acceptable to the patient, a final option for some patients could be a colostomy. However, this decision must be well-considered, because it is more or less a definitive solution. On one hand, one can imagine that patients who used to suffer greatly from soiling and tried everything to achieve better fecal continence, even by undergoing additional surgery, are content with their colostomy (as is the case with 2 patients in our study who decided to have a colostomy at age 19). On the
other hand, one must keep in mind that patients who receive a colostomies at younger ages do not have the opportunities to try to regulate defecation in a more natural way (as is case with 5 patients in our study who received a ileostomy or colostomy between their 3rd and 14th year of life). Moreover, they are not able to make this decision for themselves.

CONCLUSIONS

No matter what mode of regulation is chosen by patients with high anorectal atresia, many patients with anal defecation suffer from soiling more or less. Nevertheless, most patients are content with their level of cleanliness. In view of the trial and error method and the lack of professional support in learning defecation regulation in these patients, we assume that effectiveness can be improved for some of them. It seems important to us to offer patients a structured learning program with professional support, so patients may learn to defecate according to a stepwise protocol, starting with the most natural way of defecation. Each step should be carefully evaluated at regular intervals with respect to effectiveness on soiling and constipation. In our opinion, this seems the most adequate way to find the optimal mode (earlier and better) of defecation regulation for patients with (high) anorectal atresia.

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REFERENCES