Unraveling childhood constipation: Pathophysiology, diagnostics and treatment

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CHAPTER 4

The value of fluoroscopic defecography in the diagnostic and therapeutic management of defecation disorders in children

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Submitted
ABSTRACT

Background and Objective: Defecography is a study to assess anorectal function during evacuation. We aimed to investigate the value of fluoroscopic defecography in directing diagnostic and therapeutic management in children with defecation disorders.

Patients and Methods: All fluoroscopic defecography studies performed (2003-2009) in children with defecation problems and normal anorectal motility studies were reviewed. Results were classified in 3 groups; 1) Normal pelvic floor function; 2) Pelvic floor dyssynergia: incomplete relaxation of pelvic musculature, inconsistent change in anorectal angle and incomplete voluntary evacuation; 3) Structural abnormality: excessive pelvic floor descent, with an intra-rectal intussusception, rectocele, or rectal prolapse.

Results: 18 patients (13 boys, median age 9.1 years) were included. Indication for fluoroscopic defecography was chronic constipation in 56%, fecal incontinence in 22% and rectal prolapse in 22%. Defecography showed pelvic floor dyssynergia in 9 patients, a structural abnormality was found in 4 and defecography showed a normal pelvic floor function in 5 patients (28%). In 12 patients (67%) the outcome of fluoroscopic defecography directly influenced therapeutic management. After defecography 4 children (22%) were referred for anorectal biofeedback treatment, 4 children (22%) for surgery, 2 children (11%) for an additional MR defecography; 1 patient to psychology department and medication was changed in 1 patient. In 6 patients (33%) the result did not change the management. In 9 patients (75%) the change of management was successful.

Conclusions: Fluoroscopic defecography can be a useful tool in understanding the pathophysiology and it may provide information that impacts management of children with refractory defecation disorders.
INTRODUCTION

Defecation disorders represent a common problem in children, with one third of the children followed-up beyond puberty continuing to be symptomatic. The pathophysiology is multifactorial and remains incompletely understood. A commonly accepted hypothesis is that abnormalities in the function of pelvic floor muscle activity may lead to obstructed defecation. Children not responsive to medical treatment merit further investigation in order to better understand their underlying pathophysiology. Therefore, several imaging modalities have been proposed including abdominal x-rays to monitor the transit of radio-opaque markers, barium enema, and MRI of the spine. However, pelvic floor dysfunction is hard to diagnose with the above-mentioned studies.

Defecography is a dynamic radiologic test performed during voluntary evacuation of the rectum, to assess the anorectal function at rest and during defecation. Since the initial description by Wallden in 1952, the technique has been further improved and simplified, but a consensus is still lacking regarding the optimal examination technique. Although defecography may increase our understanding of the pelvic floor pathophysiology, literature in pediatric patients is lacking and fluoroscopic defecography has not yet proven its value in the management of children with defection disorders.

This study aims to investigate the role of fluoroscopic defecography in understanding the pathophysiology of defecation disorders in children and to describe its value in directing diagnostic and therapeutic management.

METHODS

Patients

Patients were retrospectively selected based upon the following inclusion criteria: 1) Constipation and/or fecal incontinence (FI) unresponsive to medical treatment; 2) Completion of clinically indicated anorectal manometry and defecography studies performed at Nationwide Children’s Hospital, Columbus, Ohio; 3) Age ≤ 18 year. In all patients, the manometry was performed without sedation, with a water perfused catheter. Patients with abnormal anorectal motility upon manometry, defined as the absence of an intact recto-anal inhibitory reflex, were excluded. The study protocol was approved by the Institutional Review Board of Nationwide Children’s Hospital.

Defecography

Bowel preparation was not carried out before defecography. With the patient in the left decubitus position, a thick, stool-like semisolid, barium paste (Anastrast, Lafayette Pharmaceuticals) was injected into the rectum, using a caulking gun, until the patient experienced an urge to defecate due to rectal distention. Once contrast was inserted, the patient was placed upright on a specialized radiolucent commode (Figure 1). In the lateral projection, spot radiographs of the anorectal region were obtained at rest, during pelvic floor contraction (squeeze), and at valsalva against contracted pelvic floor (straining). Cine images (3 frames per sec) were obtained during expulsion of contrast (defecation).
Measurements
Measurements obtained during defecography were acquired according to previously published standards\(^8,18,19\) and included: 1) Anorectal angle (ARA): the angle between the axis of the anal canal and the posterior rectal wall, measured during rest and straining; 2) Pelvic floor descent: the perpendicular distance from the anorectal junction to the pubococcygeal line (most inferior portion of the pubic symphysis to the last horizontal coccygeal joint); 3) Evacuation of the rectum; 4) Morphologic changes of the rectal wall, such as rectal rectocele, rectal prolapse and rectal intussusception.

Categories
The defecography results were, according to previously published adult literature,\(^2,15,20-23\) classified into 3 groups: 1) Normal study: ARA of 80-120° at rest and an increase of 20-45° during straining, relaxation of the pelvic floor with a descent of 1.5-3 cm, complete evacuation of the rectum, and no morphological abnormalities of the rectal wall; 2) Pelvic floor dyssynergia: incomplete relaxation of the pelvic musculature, inconsistent change in the ARA (absence of change or paradoxical decrease in ARA) and incomplete voluntary evacuation of the rectum; 3) Structural abnormality: excessive pelvic floor descent in combination with a rectocele, rectal prolapse, or intrarectal intussusception.

Figure 1: The specialized radiolucent commode
Data collection and analysis
Data regarding demographics, medical history and recommendations after the defecography study were collected from the medical charts. The defecography studies and the radiation dose during the study were retrospectively analyzed by one experienced pediatric radiologist. The estimated effective dose was calculated by multiplying the calculated dose area product (DAP-mGy*cm²) by the appropriate conversion coefficient. The value of the defecography in understanding the etiology of defecation disorders and the value in the therapeutic and diagnostic management of our patients are descriptive. Success of the change in management following the defecography study was defined as an improvement of symptoms without occurrence of complications in patients with a surgical intervention.

RESULTS
Demographics
A total of 18 patients were included, 13 boys and 5 girls, with a median age of 9.1 years (range 3-16 years). Prior to defecography, the median duration of symptoms was 6 years (range 1.5–10 years). The indication for defecography was long-lasting constipation in 10 children (one of these patients had a tethered cord and another patient had an imperforated anus). In 4 children the indication was intractable FI (one patient had a tethered cord and an imperforated anus). In the other 4 patients the indication for defecography was recurrent rectal prolapse. In all patients, maximal medical treatment, including high dose of different oral laxatives, retrograde enemas, cleanouts, and behavioral interventions had failed. Two patients had a previous surgical intervention (cecostomy, colostomy) for intractable constipation. All patients had normal anorectal motility, measured with anorectal manometry prior to defecography.

Defecography results
The entire study took a maximum of 15 minutes, with a median fluoroscopic time of 1.2 min (range 0.43-5.43 min). The cooperation of 15 patients was good. In 3 children (3-5 years of age), there were difficulties completing the study due to age-related lack of cooperation. Five patients had a normal defecography study: 3 with constipation and 2 with FI. The defecography showed pelvic floor dyssynergia in 9 patients: 6 with constipation, 2 with FI and 1 patient with rectal prolapse (Table 1). All patients with dyssynergia had incomplete (n=5) or no relaxation (n=4) of the pelvic floor musculature. Five patients had incomplete evacuation of the rectum, 4 patients no evacuation of the rectum at all. In 4 patients defecography revealed a structural abnormality: an intra-rectal intussusception was seen in 3 patients and the fourth patient had a combination of an intussusception, rectal prolapse and anterior rectocele. Figures 2-4 depict study examples of the three categories.
Table 1: Defecography results by initial indication for defecography study

<table>
<thead>
<tr>
<th></th>
<th>Constipation</th>
<th>FI</th>
<th>Rectal prolapse</th>
</tr>
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<tbody>
<tr>
<td>Normal defecography</td>
<td>3</td>
<td>2</td>
<td>-</td>
</tr>
<tr>
<td>Pelvic floor dyssynergia</td>
<td>6</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Structural abnormality</td>
<td>1</td>
<td>-</td>
<td>3</td>
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FI = fecal incontinence

Figure 2: Normal defecation.
10 year old boy with constipation and a history of tethered cord repair. (A) Lateral view of the rectum at rest demonstrates a normal anorectal angle (ARA) of 115 degrees. PCL = pubococcygeal line. (B) Lateral view during squeeze demonstrates normal elevation of the pelvic floor (AJR- anorectal junction) and decrease in the ARA to 92 degrees. (C) Lateral view during straining shows normal descent of the pelvic floor (AJR) and widening of the ARA. (D) Lateral view during evacuation of barium paste demonstrates normal funnel-shaped anorectum and widening of the anal canal (arrow).
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Figure 3: Pelvic floor dyssynergia.
16 year old girl with chronic constipation. (A) Lateral view of the rectum at rest shows a normal ARA of 82 degrees. (B) Lateral view during attempted defection demonstrates paradoxical decrease in anorectal angle (ARA) to 58 degrees. The patient is unable to evacuate barium paste.

Figure 4: Rectal prolapse.
9 year old boy with a history of rectal prolapse. (A) Normal lateral view of the rectum at rest. (B) Lateral view of the rectum during defection demonstrates full thickness anterior wall intussusception (arrow) and narrowing of the anal canal (double arrow). (C) Lateral view at termination of defection demonstrates rectal prolapse (arrows).

Management change
Figure 5 depicts the change in diagnostic or therapeutic management based on the defecography result. In 6 patients the result of the defecography did not change the management. In 12 patients the outcome of defecography directly influenced clinical management. Four patients were referred for anorectal biofeedback treatment, three with pelvic floor dyssynergia, and one with a normal study. Four patients underwent surgical treatment following the defecography. A cecostomy was placed for the treatment of constipation in 2 patients. One patient underwent a rectosigmoidectomy (Altemeier
procedure) for rectal prolapse, and one patient received peri-anal Thiersch sutures. Two patients were referred to a specialized center for additional MR defecography, due to inconclusive results on the fluoroscopic defecography. In the first patient the fluoroscopic defecography study showed an incomplete relaxation of the pelvic floor musculature and a narrow anal canal. The additional MR defecography revealed an enterocele, which was managed conservatively. In the second patient an anterior rectocele, large anterior intussusception, rectal prolapse, and a narrow anal canal were seen on the fluoroscopic defecography. The MR defecography showed overall weakness of the pelvic floor, with a significant rectal prolapse, sigmoidocele, and cystocele. This patient was treated surgically with the Frijkman Goldberg procedure (abdominal sutured rectopexy with sigmoid resection). One patient was referred to the psychology department, because of major depressive disorder. In one patient, the prior described medication to treat constipation was changed, following a normal defecography study. In 9 out of the 12 patients the change in management was successful, after a median follow-up time of 12.5 months (range 1-50 months). There was improvement of symptoms and no complications in the patients with a surgical intervention. In 3 patients the change of therapy was not successful, and symptoms did not improve. All three patients presented initially with constipation. Two were initially referred for biofeedback, and in one the management did not change after defecography. After a mean time of 9 months, all 3 patients received a cecostomy for delivering antegrade enemas.

Figure 5: The change in management based on the defecography result.
*cecostomy, **1 peri-anal Thiersch sutures, and 1 resection of prolapse
The mean radiation exposure during the defecography studies was 3.55 mSv per patient, with a median of 2.32 mSv and a range between 0.11 and 10.86 mSv.

**DISCUSSION**

Defecography can be performed to evaluate anorectal and pelvic floor function in children with defecation disorders not responding to medical treatment. In our study population of 18 children, fluoroscopic defecography demonstrated pelvic floor dyssynergia in 50%, a structural abnormality was found in 22%, while 28% of the subjects had a normal defecography study. The defecography findings led to a change in clinical management in 67% of the patients. In 75% of those symptoms improved as a consequence of the changes made after the defecography. These results suggest that defecography may be a useful diagnostic tool to understand the pathophysiology of defecation disorders in children and that defecography may direct medical or surgical treatment in a selected subgroup of children with refractory constipation.

Despite controversies regarding clinical relevance and the advice that results should be interpreted with caution,24 defecography remains the only dynamic method, that visually evaluates the process of defecation.13,25 The strength of fluoroscopic defecography is that it allows assessment of the rectal function with the patient in sitting position during the procedure, to achieve a more physiological defecation.26,27 Defecography has been considered a relatively simple and inexpensive procedure that can provide valuable information about dynamics of the anorectal system.28 A combination of defecography with anorectal manometry has been suggested for the diagnostic workup of children with severe, intractable constipation.29

We divided the study patients into three groups. The main indication for defecography was long-lasting severe constipation. The other indications were intractable FI and rectal prolapse. It should be noted that this study group represents a skewed population. The patients had symptoms for many years and had failed multiple medical treatments. Four patients from this study population were referred for biofeedback treatment after pelvic floor dyssynergia was identified on defecography. In 2 patients biofeedback was successful, whereas in 2 patients biofeedback had no benefit. Paradoxical contraction or failure to relax the pelvic floor during attempts to defecate is described as pelvic floor dyssynergia and commonly observed in children with constipation.30

Biofeedback training (BFT) can be used in these children to teach them how to tighten and relax their perianal muscles in order to pass bowel movements more efficiently. A recently published Cochrane review, however, concluded that there is no evidence that biofeedback training adds any benefit to conventional treatment in the management of childhood constipation.31 This is in contrast with data in adults with constipation and dyssynergic defecation, where biofeedback was effective and superior to laxatives.32 Because BFT has been considered a relatively well tolerated treatment and available in most hospitals, it can be worth trying BFT in children before considering more radical
treatment options. In this study group, patients in whom BFT was not successful had more severe evidence of dyssynergia compared to patients in whom BFT was successful. Thus, BFT might be considered when the patient has more subtle dyssynergia characteristics during defecography. Secondly, cooperation with biofeedback training is essential. So it is very important that the physician discusses the treatment with the patient and parents and estimates if the (especially younger) patient will be cooperative during BFT. It is certainly possible that only a small subgroup of pediatric patients benefits from biofeedback and that tests not commonly performed, such as defecography may aid the clinician to identify such patients.

We found defecography to be well tolerated in the majority of patients. In this series the cooperation of patients was good and patients only experienced some discomfort during the filling of the rectum with contrast paste, due to rectal distension. We had difficulties completing the study in three children. In order to perform a defecography in a child, it is essential that the patient is cooperative and relaxed\(^2^6\). The minimum age when a defecography study is feasible has been debated, but the current opinion is that defecography is feasible starting at the age of 4 years or when the child has had at least one to two years of toilet training.\(^1^3\) It has been stated that patients’ embarrassment at the setting of the study may inhibit normal defecation of the contrast material.\(^2^4\) Literature is lacking regarding the psychological burden associated with defecography in pediatric patients. However, Deutekom et al. report that the burden for defecography was low in adults and comparable with other endo-anal function tests.\(^3^3\)

One of the disadvantages of defecography is the radiation exposure. We found that a wide range of radiation dose was used in our case series, with a mean and median exposure of respectively 3.55 and 2.32 mSv during the defecography studies. In order to put these data in perspective, it should be noted that the dose due to background radiation is 3 mSv/year, the radiation dose during a chest X-ray is 0.2 mSv, and a barium enema is two to three times that of defecography.\(^1^3,3^4\) Radiation dosage can be kept as low as reasonably achievable by careful attention to measures of radiation protection, using pulsed fluorography as sparingly as possible and using a special radiolucent commode. Although the radiation exposure is relatively high, the risk may be considered acceptable in children with severe, intractable constipation, when the use of defecography may impact treatment and may avoid other equally invasive diagnostic tests or therapeutic procedures.\(^3^4\) In recent years, MR defecography has been proposed as an alternative technique to provide insight into pelvic floor and rectal function.\(^3^5-3^7\) Advantages of MR defecography are the absence of radiation exposure, the superior soft tissue visualization, and the global view of the other compartments. However, MR defecography, and especially open MR devices that allow sitting position, is limited in availability, and more expensive than fluoroscopic defecography.\(^1^9,3^5,3^6,3^8\) Wherever MR defecography is available such test should have the preference, due to his advantages compared to fluoroscopic defecography. Referring a patient to a center with the capability to perform an MR defecography may be appropriate in some patients with inconclusive fluoroscopic defecography results.
Prior studies in adult patients with defecation disorders are inconclusive regarding the value of fluoroscopic defecography in clinical decision-making. Some investigators reported that defecography had substantial diagnostic and therapeutic benefits and was useful to establish a definitive diagnosis in 43–53% of cases.\textsuperscript{39,40} In another study, defecography had an overall accuracy of 83.3%, and was stated to be a reliable tool in clinical decision making in constipated patients.\textsuperscript{41} In contrast, inconsistent methodology, wide interobserver variations in the measurements of defecography parameters and problems in interpretation have been reported as well.\textsuperscript{13,16,24,39,42} The variation between observers in the measurement of the ARA is due to the fact that some investigators favor the use of the central axis of the rectum, whereas others prefer referring to the posterior rectal wall to determine the ARA.\textsuperscript{15,26,42,43} In our study the ARA was defined as the angle between the axis of the anal canal and the posterior rectal wall. Regarding the inconsistent methodology, a variety of different contrast mediums are used for defecography. The texture of the barium paste aims to replicate the consistency of stool.\textsuperscript{13} Although there is no consensus regarding the type of the barium paste among different hospitals, altering the viscosity of the barium contrast medium used for defecography does not seem to substantially affect the subsequent radiographic findings.\textsuperscript{44} Evaluation of defecography requires experience in the method. It has been advised that studies should only be performed in centers where there are a sufficient number of patients.\textsuperscript{25} No specific recommendation regarding the minimum of studies that has to be performed has been described in the literature. In this study, defecography studies were all performed and analyzed by one pediatric radiologist.

This study had several limitations. First, this was a retrospective study. Second, patients were selected from a tertiary referral center, a factor that might have resulted in selection bias. A limitation of the technique used in this study, was the lack of pre-contrast filling of the small bowel, and the lack of vaginal marking for location. These limitations may limit diagnostic capabilities. To compare results and improve diagnostic reproducibility in future research, it would be important to develop normal pediatric values for defecography measurement and to standardize definitions and techniques.

In conclusion, this study suggests that fluoroscopic defecography can be a useful tool in understanding the pathophysiology of defecation disorders and it may provide information that impacts the management of children with refractory defecation disorders.
REFERENCES


