REVIEW

A systematic review on bowel management and the success rate of the various treatment modalities in *spina bifida* patients

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Study design: Systematic review.

Objectives: To determine the different treatment modalities aimed at achieving fecal continence in *spina bifida* (SB) patients and their effectiveness.

Setting: International literature.

Method: Electronic databases were searched ('Pubmed', 'Web of science', 'CINAHL' and 'Cochrane') identifying studies published since the mid-eighties and screened for relevance according to the Centre for Reviews and Dissemination procedure guidelines. A total of 37 studies were selected for inclusion.

Results: Studies on toilet sitting, biofeedback, anal plug, retrograde colon enemas (RCE) and antegrade colon enemas were found. Fecal continence was achieved in 67% of SB patients using conservative methods ($n = 509$). In patients using RCE ($n = 190$) an 80% continence rate was reached. Patients following surgical treatment ($n = 469$) reached an 81% continence rate, however, 23% needed redo surgery because of complications. Better fecal continence was associated with an improved quality of life, which was negatively influenced by the amount of time spent on bowel management.

Conclusion: Evidence favors an individually tailored stepwise approach with surgery as a final step in case of failure of conservative measures. Continued specialized support throughout life remains important to maintain continence. Cross-over and comparative trials are needed in order to optimize treatment.

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Keywords: *spina bifida*; fecal pseudo-continence; bowel management; constipation

INTRODUCTION

Spina bifida (SB) or meningocele is a complex neuroembryological disorder resulting from a variable degree of incomplete closure of the posterior neural tube. Clinical presentation is highly variable and depends on the localization of the defect along the spinal cord and the degree of incomplete closure. These patients present with a spectrum of impairments but the primary functional deficits are lower-limb paralysis and sensory loss, bladder and bowel dysfunction and cognitive dysfunction.¹

In the majority of patients the lower regions of the spine are affected, resulting in dysfunction of the distal gastrointestinal tract: rectum, anus and anal sphincter. Voluntary control of defecation requires normal rectal sensation, peristalsis and normal anal sphincter function. Two primary involuntary reflexes, the intrinsic and parasympathic reflex, located at sacral level 2–4, initiate defecation. The pudendal nerve, responsible for voluntary defecation, controls the opening and closing of the external anal sphincter. Nerve damage above the S2 level will impair both involuntary reflexes and pudendal nerve function. Loss of the involuntary reflexes perturbs rectal sensation and initiation of defecation. Damage of the pudendal nerve leads to partial or total loss of the voluntary sphincter control. Perturbation of these processes will result in bowel incontinence, which is often associated with constipation in SB patients.^{2,3} Fecal incontinence in SB patients is reported to be present in 28–53%, regardless of the therapy used.^{4–7}

Fecal and urinary incontinence importantly affect quality of life (QoL) in SB patients and form a major barrier to attending school, obtaining employment and sustaining relationships. Krogh *et al.*⁴ report that 66% of SB patients older than 6 years with fecal incontinence perceive incontinence as having a negative influence on their social activities. Lie *et al.*⁸ report that 75% of SB patients with urinary incontinence regard incontinence as a stress factor.

REVIEW QUESTION

A systematic literature review was performed regarding treatment modalities for constipation and/or fecal incontinence in SB patients and their success rate. Treatment success is defined as fecal continence that corresponds to stool losses less than once a month.

REVIEW METHOD

The review is performed following the guidelines of the Centre for Reviews and Dissemination procedures.⁹

Study selection

Inclusion criteria:

- Original papers with full paper available
- Written in English
- SB patient cohort larger than 20
- SB patients using any form of bowel management

Exclusion criteria:

- No data on outcome of treatment for SB patients
- Review, opinion or editorial pieces

Study identification

The following databases were used for study identification: 'Pubmed' and 'Web of science', 'CINAHL' and 'Cochrane' database. A search was performed in March 2013. The following Mesh terms were used: 'spina bifida and bowel management', 'spina bifida and fecal incontinence', 'spina bifida and enema', 'myelomeningocele and bowel management', 'myelomeningocele and fecal incontinence' and 'myelomeningocele and enema'. The search covered studies published since 1986 as this was the start of important bowel management evolutions, with the introduction of retrograde colon enemas (RCE) by Shandling¹⁰ and some years later the antegrade colon enema (ACE) by Malone.¹¹

This database search resulted in a selection of 37 papers (see flowchart).^{4–7,10,12–43}

A limitation of the review is the lack of search in gray literature as well as the absence of expert contact to attain more information on the subject.

Data extraction was performed independently by two authors to avoid selection bias in the process. From the full copies retrieved, two articles were not retained by both authors.

RESULTS

A total of 37 studies were included in the review.^{4–7,10,12–43}

Most studies were observational studies with retrospective or prospective data on SB patient cohorts in a single or multiple center setting. Only two were randomized clinical trials on the use of electrical stimulation, of which one was double-blind controlled²³ and one was patient-blinded.²⁶ All studies had a low grade of recommendation.⁴⁴

Clinical data were collected using questionnaires,^{6,7,12–15,17,19,23–25,29,30,34,35,37,43} neurogenic bowel dysfunction scales,^{20,26,45} QoL questionnaires^{4,16,21,22,27,28,31,32,38,41} or child behavior checklists.^{4,31} The collection methods were interviews during follow-up visits,^{6,15,20,25,43} or telephone contacts^{4,7,12,17,23,32,38} or both.^{16,31,37,41}

The follow-up period of SB patients after starting bowel management varied from 3,^{15,19,20,22,23,41,42} over 12,^{12,13,21,25,31} 30^{16–18,27–30,34,39} and 60 months^{24,32,33,35,36,40,43} to 120 months.^{38,43}

To explore different treatment modalities for bowel management in SB and their effectiveness the studies were grouped according to the treatment modality: conservative bowel management (16 studies), electrical stimulation (4 studies) and non-conservative (surgical) bowel management (17 studies).

Conservative bowel management

The studies on conservative bowel management are summarized in Table 1. From the 16 studies, 1 reported on toilet sitting, 2 on biofeedback, 1 on anal plug use, 4 on a stratified treatment strategy and 8 on RCE.

Most studies on bowel management used the above-mentioned strict continence definition (no stool loss or stool loss less than once a month).^{4,6,12,16–21} Some studies gave no definition^{7,13,15} or used a less-strict definition.^{5,10,14,22} The results for fecal pseudo-continence achievement using conservative bowel management varied from 36¹³ to 100%.¹⁰ In order to calculate the combined continence rate, only the studies using the strict continence definition of stool losses less than once a month were used. Fecal pseudo-continence was achieved in 67% (341/509).

Looking only at the studies on RCE ($n=8$), fecal pseudo-continence was achieved in 80% (144/190). Two studies reported on the effect of RCE on constipation. Mattsson *et al.*¹⁹ reported absence of constipation when using RCE, whereas Ausili *et al.*²⁰ reported 60% (36/60) constipation relief using RCE. In both studies, all SB patients suffered from constipation at the start. The irrigation fluid used was saline in 3,^{10,16,18} tap water in 4^{17,19,21,22} and one did not mention the fluid type used.²⁰ The irrigation volumes used were sometimes fixed body weight-related quantities (20 ml kg⁻¹)^{10,16,17} or varied between 300¹⁹ and 616 ml.²¹

The time spent on treatment was not often reported^{7,12–15,17,20} and if reported varied from 15^{5,10,16,21,22} over 30 min¹⁸ to 60 min.¹⁹ Krogh *et al.*⁴ described a threefold increase in treatment time when comparing RCE with digital evacuation.

Although achieving fecal pseudo-continence is the primary treatment goal, patient satisfaction is an important secondary goal. Results on satisfaction could not be compared as all reporting studies used different questionnaires and scales. Some studies reported an improvement of satisfaction or a high satisfaction rate associated with successful bowel management.^{16,17,19–21} The study of Shoshan *et al.*¹⁵ described a significant reduction of daily life impairment by fecal incontinence after bowel management intervention. Finally, implementing a stepwise fecal incontinence treatment protocol improved fecal pseudo-continence and in parallel resulted in better socialization, less need for support by the caregiver and less negative

Table 1 Summary of papers on conservative bowel management in SB patients

Author	Sample size, age, type	FU/TT	FI definition	Study design	Results—effectiveness	Results—satisfaction
King <i>et al.</i> ¹²	<i>n</i> = 40 SB Age: 18 months–29 years LL: 12 T, 25L, 2S FI: 35/40 Constipation: nm	FU: 15 m TT: not mentioned (nm)	Stool loss <once per month	Single center Retrospective chart review Prospective bowel program: daily, regular, consistently timed, reflex-triggered bowel evacuation Phone FU every 2 weeks and during FU visits	Continence: start 5/40 (12.5%), 15 m: 24/40 (60%) 24/40 compliant, 19/24 (79%) continent 11/40 non-compliant, none continent (<i>P</i> < 0.0001).	Not evaluated
Whitehead <i>et al.</i> ¹³	<i>n</i> = 33 SB Age: 5–16 years LL: T10–S2 no mental impairment FI: nm Constipation: nm	FU: 12 m TT: nm	Not defined	Single center Prospective-controlled trial. Daily symptom log, 1 m before and during 19 SB behavior modification: 10 times toilet sitting every evening 14 biofeedback every 2 weeks and behavior modification.	Biofeedback: (before) 5.38–1.93 (12 m) accidents per week 5/14 (36%) continent Behavior: (before) 5.77–2.3 (12 m) accidents per week 4/19 (21%) became continent. No significant difference between groups	Not evaluated
Ponticelli <i>et al.</i> ¹⁴	<i>n</i> = 73 SB (67 congenital) Age: 7–25 years LL: L5–S1 lesions FI: 52 Constipation 57	FU: nm TT: nm	Not defined	Single center Prospective-controlled trial Questionnaire evacuation habits 10 biofeedback sessions 12 conventional treatment (laxatives, stimulants and enema) 30 no treatment	Biofeedback: 2/10 improved, 4/10 full bowel control Conventional treatment: 7/12 improved. No statistical analysis done	Not evaluated
Shoshan <i>et al.</i> ¹⁵	20 SB Age: 4–29 years LL: T4–L5 FI: 17 diapers, 3 pads Constipation: nm	FU: 5w TT: nm	Not defined	Single center Self-controlled clinical trial. Daily record. FU visits at week 0, 1, 2 and 5. Anal plug use start at week 1. Effect on FI scale 0–4 (0 = not bothersome, 4 = very bothersome).	15/20 completed the study Accidents per week start: 4(0–28), 5w 0(0–8) (<i>P</i> = .002)	Baseline 50% FI severely impedes daily life. 5w 40% slight interference Significant reduction of FI scale (<i>P</i> = 0.001).
Malone <i>et al.</i> ⁷	<i>n</i> = 109 SB Age: 9–47.8 years 68 wheelchair LL: nm FI: 55 Constipation: nm	FU: nm TT: nm	Not defined	Multicenter Questionnaires Random cohort <2000 patient database 109/144 responded	94/104 regular toileting 26/104 manual evacuation 25/104 laxatives 13/103 suppositories 55/104 (53%) regular soiled	Not evaluated
Krogh <i>et al.</i> ⁴	<i>n</i> = 125 SB Age: 2–18 years LL: nm FI: 55 Constipation: nm	FU: nm TT: RCE: 133' Digital: 61 times No method 132'	Stool loss <once month	Multicenter 184 item questionnaire (tested for reproducibility and validity) and validated CBCL 125/208 (100 > 4 years old) responded	25/125 digital evacuation 13/125 suppositories 35/125 RCE 35/125 laxatives 55/100 (children >4 years old)(55%) FI	10/42 FI major influence on QoL in 2–5-year-olds 21/46 FI major influence on QoL in 6–10-year olds 17/37 FI major influence on QoL in 11–18-year-olds
Verhoef <i>et al.</i> ⁶	<i>n</i> = 350 SB Age: 16–25 years 70 wheelchair LL: L5–S1 lesions FI: nm Constipation: nm	FU: nm TT: 58/ 179 > 15 min a day	Stool loss <once per month	Multicenter Data collected from interviews and neurophysiological testing, retrospective medical history 179 responded from 350	31/179 laxatives 49/179 RCE 27/179 manual evacuation 61/179 (34%) FI	47/61 (77%) perceived FI as a problem
Vande Velde <i>et al.</i> ⁵	<i>n</i> = 80 SB Age: 5–18 years 33 wheelchair LL: 26 < S2, 22 L5–S1, 32 > L4 FI: nm Constipation: nm	FU: nm TT: RCE and ACE 21 min a day (8.5–35 min per day)	Stool loss <once per week	Single center Descriptive cohort study Stepwise therapeutic strategy	5/80 regular toileting (no FI) 13/80 manual evacuation (38% FI) 24/80 RCE (13% FI) 16/80 ACE (no FI) 22/80 (27%) FI	Not evaluated
Shandling and Gilmour, ¹⁰	<i>n</i> = 112 SB Age: 4–20 years LL: nm FI: 112 Constipation: nm	FU: nm TT: 15–20 min	Stool loss <4 times per month	Single center RCE with balloon catheter with saline water 20 ml kg ⁻¹ every 24–48 h	4 dropped out 5 returned to RCE after initially dropping out 100% continence rate	Not evaluated

Table 1 (Continued)

Author	Sample size, age, type	FU/TT	FI definition	Study design	Results—effectiveness	Results—satisfaction
Liptak and Revell ¹⁶	<i>n</i> = 31, 30 SB Age: 3–19 years LL: T-S4 FI: 18/25 Constipation: 14/25	FU: 30 months TT: 21 min	No stool loss	Single center Prospective clinical trial. After bowel cleaning, start RCE with balloon catheter, every 24–48 h with saline water 20 ml kg ⁻¹ . Standardized questionnaire over telephone or by visit. Satisfaction rate 1–4 (1 = extremely dissatisfied, 4 = extremely satisfied).	6 dropped out first 3 weeks 9 dropped out after 18 months FI dropped from 72 (18/25) to 29% (7/25) at 18 months and to 6% (1/16) after 30 months (<i>P</i> < 0.01)	Mean satisfaction score increased from 1.1–2.8 after 18 months and 3.3 after 30 months (<i>P</i> < 0.01)
Schöller-Gyüre <i>et al.</i> ¹⁷	<i>n</i> = 53 SB Age: 7 months–22 years LL: nm FI: 14 Constipation: 14	FU: 33 months TT: nm	No stool loss	Single center Case review and questionnaire. RCE with cone catheter with tap water 20 ml kg ⁻¹ every 24 h. Frequent telephone contact.	41 returned questionnaire 27/41 (66%) complete fecal continence. 6/41 RCE painful, 3/41 RCE unpleasant	Parental satisfaction high in 63%, good in 37%. Major disadvantage is time and energy to perform RCE (51%), daily burden on family (39%)
Eire <i>et al.</i> ¹⁸	<i>n</i> = 33 SB Age: 5–22 years LL: nm FI: nm Constipation: nm	FU: 30 months TT: Median 30 min (15–45 min)	No stool loss	Single center Selected and well-motivated patients Retrospective case review. After desimpaction, start RCE with balloon catheter saline water, median 500 ml every 24 h and at continence, every 48 h	32/33 became continent 2/33 were independent	Not evaluated
Mattsson and Gladh ¹⁹	<i>n</i> = 40 SB Age: 10 months–11 years LL: nm FI: 40 Constipation: 40	FU: 4 months, up to 8 years after RCE TT: 12–60 min	No stool loss	Single center Parental questionnaire (eight questions). RCE with cone catheter with tap water, median 300 ml every 24 h Plasma sodium levels in 28 SB before start and after 1 month or 1 year Manometry in 28 SB before and after 1–3 years	5 dropped out 35/40 were continent All free of constipation 1/40 was independent	35/40 found RCE satisfactory All found RCE time consuming 36/40 parents reported general improvement in well-being
Ausili <i>et al.</i> ²⁰	<i>n</i> = 60 SB Age: 8–17 years LL: nm FI: 16 Constipation: 60	FU: 3 months TT: nm	Stool loss < once per month	Single center Prospective clinical trial. Validated questionnaire (NBD score, range 0–47 (47 = severe bowel dysfunction) ⁴⁵) and QoL Visit at start and after 3 months RCE with balloon catheter	36/60 (60%) relief of constipation 12/16 (75%) relief of fecal incontinence NBD decreased from 17.5 to 8.5 after treatment (<i>P</i> < 0.001)	Parents reported an improvement on QoL and degree of satisfaction NBD score improved from 17.5 to 8.5 (<i>P</i> < 0.001)
Pereira <i>et al.</i> ²¹	<i>n</i> = 40, 28 SB Age: 6–25 years LL: nm FI: nm Constipation: nm	FU: 12 months TT: average 15–30 min	No stool loss Pseudo-continence: no stool loss with treatment	Single center Prospective clinical trial Standard questionnaire on bowel function and QoL (rate 0–10, 0 = great reduction, 10 = great improvement) RCE with balloon catheter with average 616 ml tap water, every 3 days	35 returned questionnaire Pseudo-continence rose from 10/35 to 28/35 (80%) 16/35 partially or totally independent Significantly less time spent than conventional bowel management	Mean grade of satisfaction was 7.3
Choi <i>et al.</i> ²²	<i>n</i> = 53 SB Age: 3–13.8 years LL: nm FI: nm Constipation: nm	FU: 4 months TT: 15.9 min	Stool loss < once per week	Single center Prospective clinical trial Survey questionnaire on bowel symptoms, QoL and general characteristics (40 items) Stepwise bowel program: first polyethylene glycol 3350 at 0.5 g kg ⁻¹ per day, if failure start RCE with cone or balloon catheter with tap water every 48–72 h	6/53 (11%) success 43/47 (81%) success Bowel care time decreased from 27 to 15.9 min (<i>P</i> = 0.003) FI per week decreased from 6.9 to 0.5 defecations per week (<i>P</i> = 0.004)	Significant reduction in impact on travel and socialization (<i>P</i> = 0.006) Significant reduction in caregiver support and emotional impact (<i>P</i> < 0.001)

Abbreviations: ACE, antegrade colon enema; CBCL, child behavior checklist; FI, fecal incontinence; FU, follow-up; NBD, neurogenic bowel dysfunction; LL, lesion level; QoL, quality of life; RCE, retrograde colon enema; SB, *spina bifida*; TT, therapy time.

emotional impact.²² However, two studies reported RCE to be 'a daily burden'. In both studies, the time spent and the energy needed to perform RCE were perceived as a major obstacle.^{17,19} Non-compliance and non-motivation could be a result of dissatisfaction with the treatment choice. King *et al.*¹² reported a non-compliance rate of 28%.

Electrical stimulation

The studies on electrical stimulation are summarized in Table 2.

Due to different stimulation techniques and result reports, the studies are not comparable. Most studies (3/4) used the above stated definition of pseudo-continence.^{23–25} Only the study of Kajbafzadeh *et al.*²⁶ used a less-strict pseudo-continence definition. The achieved pseudo-continence rate ranged from 50²⁴ to 70%.²⁵ An increased spontaneous stool frequency was reported by two studies.^{23,26} Only one study reported a 73% decrease of constipation. The study of Marshall and Boston²³ was the only double-blind randomized placebo-controlled trial reporting an important placebo effect (no actual data available).

Non-conservative, surgical bowel management

The studies are summarized in Table 3. During a surgical procedure, the appendix (or ileum if appendix is not available) is used to create a catheterizable stoma. Besides using the appendix or creating a stoma a cecostomy can be made laparoscopically or percutaneously. A stoma or cecostomy can be placed both right- or left-sided. In this review, mainly results on right-sided stoma were found.

Most studies (10/17) on surgical bowel management or ACE used the strict definition for fecal pseudo-continence (no stool loss or stool loss less than once a month).^{28,30–32,35–39,43} Some studies (5/17) gave no definition for fecal pseudo-continence^{27,29,33,40,41} or a less-strict definition.⁴² One study evaluated incontinence with a 5-point Likert scale.³⁴ The reports on achieving fecal pseudo-continence using non-conservative bowel management varied from 60 (only adults)⁴³ to 94%.^{35,36} Again only the studies using the above-mentioned strict definition of pseudo-continence were used to calculate the overall effectiveness of non-conservative treatment. Fecal pseudo-continence was achieved in 81% (378/469). No study reported on constipation. The washout fluid used was saline in 4,^{27,32,37,42} tap water in another 4^{31,35,36,39} and a combination of both in 2 studies.^{34,43} Five studies did

Table 2 Summary of papers on electrical stimulation in SB patients

Author	Sample	FU/defecation time	Definition FI	Study design	Findings—effectiveness	Findings—satisfaction
Marshall and Boston ²³	n = 50 SB Age: 4–18 years LL: nm FI: nm Constipation: nm	FU: 6 weeks TT: nm	Not defined	Single center Randomized double-blind placebo-controlled trial Daily 1 h at home ES stimulation on skin: 26 treatment, 24 sham Regular telephone contact to stimulate compliance 1 week diary	49% more spontaneous stools in active ES but not significant Important placebo effect No adverse effects	Not evaluated
Palmer <i>et al.</i> ²⁴	n = 55 SB Age: 2–14 years LL: nm FI: nm Constipation: nm	FU: 12–72 months TT: nm	Improvement: - less defecations - better rectal sensation - ability to hold consciously	Single center Prospective clinical trial Home transrectal ES 30 min per day, 5 days per week	20/55 (36%) complete success (improvement of all parameters) 30/55 (55%) moderate success (improvement of any parameter) 71% complete bowel continence No adverse effects	Inclusion of parental subjective opinion decreased the success rate of this therapy
Han <i>et al.</i> ²⁵	n = 24 SB Age: 4–13 years LL: nm FI: nm Constipation: nm	FU: 15.8 months TT: nm	No stool loss	Single center Retrospective case review Daily 1 h at home transurethral ES, 5 days per week for 4 weeks FU cycles: 2 weeks every 3–6 months One week diary before treatment and subsequent cycles	Mean episodes of fecal incontinence decreased significantly from 7.3–4.8 a week ($P < 0.05$) Complete fecal continence in 50% (12/24) Urinary tract infections in 10 patients during transurethral ES	Not evaluated
Kajbafzadeh <i>et al.</i> ²⁶	n = 30 SB Age: 3–12 years LL: nm FI: nm Constipation: nm	FU: nm TT: nm	Stool loss < once per week	Single center Randomized controlled trial Home cutaneous ES for 20 min, 3 times per week: 15 treatment, 15 sham Bowel habit diary, NBD: range 0–47 (47 = severe bowel dysfunction) ⁴⁵ Manometry before and 6 months after treatment	Constipation is decreased in 11/15 (73%) and remained in 8/15 (53%) after 6 months Significant increase of stool frequency from 2.5 to 4.7 stools per week after treatment Significant improvement manometry measures No adverse effects	Not evaluated

Abbreviations: ES, electrical stimulation; FU, follow-up; LL, lesion level; NBD, neurogenic bowel dysfunction; SB: *spina bifida*.

Table 3 Summary of papers on non-conservative bowel management in SB patients

Author	Sample	FU/ defecation time	Definition fecal continence	Study design	Findings—effectiveness Findings—complications	Findings—satisfaction
Hensle <i>et al.</i> ²⁷	<i>n</i> = 27 SB Age: 10–31 years 22 wheelchair LL: nm FI: nm Constipation: nm	FU: 9–30 months TT: 30 min	Not defined	Single center Retrospective case review Malone ACE, eight concomitant urologic surgery Saline water 300 ml Standard QoL questionnaire	Two dropped out 19/25 (76%): complete bowel control 10/27 complications 3/27 redo surgery	25/27 reported substantial improvement in their QoL
Shankar <i>et al.</i> ²⁸	<i>n</i> = 40, 27 SB Age: 6–21 years 14 wheelchair LL: nm FI: nm Constipation: nm	FU: 21 months TT: nm	Stool loss < once per month	Single center Retrospective case review Malone ACE every 48 h QOLI score: 0–5 (5: ideal) CTT and manometry in 34	4 dropped out 17/27 (63%) success 23/40 complications 11/40 redo surgery	Mean QOLI: 3.5, all report some improvement in QoL QOLI significantly lower in SB wheelchair group (<i>P</i> = 0.033)
Webb <i>et al.</i> ²⁹	<i>n</i> = 57, 43 SB Age: 5–30 years LL: nm FI: nm Constipation: nm	FU: 30 months	Not defined	Single center Retrospective case review Malone ACE every 72 h phosphoda or soapy water, 21 concomitant urologic surgery	2 dropped out 40/43: good to excellent results 8/57 complications 3/57 redo surgery	Overall patient satisfaction is good to excellent
Curry <i>et al.</i> ³⁰	<i>n</i> = 273, 108 SB Age: 7.5–29.9 years LL: nm FI: nm Constipation: nm	30 months	Full: totally clean or minor leakage on night of washout	Multicenter retrospective pro-forma Malone ACE or button (19/273)	68/108 (63%) full success 111/27 three complications	Not evaluated
Aksnes <i>et al.</i> ³¹	<i>n</i> = 20 SB Age: 6.3–17 years LL: nm FI: 16 Constipation: 4	FU: 16 months TT: 50 min (30–75)	No stool loss	Single center Prospective clinical trial Malone ACE 900 ml tap water Standardized questionnaire before and 6 months after ACE, CBCL, YSR, SPPA 1–4 score (4 = describes me very well) FU visit and telephone	16/20 gained independency 16/20 complete continence 6/20 redo surgery	SPPA: postoperative improved self-esteem (<i>P</i> = .04) and close friends (<i>P</i> = .006) CBCL and YSR no difference pre- and postoperative
Dey <i>et al.</i> ³²	<i>n</i> = 62, 31 SB Age: 3.8–21.4 years LL: nm FI: nm Constipation: nm	FU: 65 months TT: 53 min (15–180)	Stool loss < once per month	Single center Retrospective casenote review Appendix ACE 550 ml saline water Questionnaire: 32 responded from 51 still using ACE	11 dropped out 27/32 (84%) continent 35/62 complications 22/62 redo surgery	Median satisfaction score: 9 (10 = completely satisfied) Significant correlation between continence and satisfaction (<i>P</i> = 0.04)
Casale <i>et al.</i> ³³	<i>n</i> = 275 SB Age: mean 11 years LL: nm FI: nm Constipation: nm	FU: 45–120 months TT: nm	Not defined	Single center Retrospective chart review 215 Malone ACE Single surgery compared with combined urologic surgery (<i>n</i> = 158)	94% continence, no difference between groups 27% complications, no difference between groups	Not evaluated
Lemelle <i>et al.</i> ³⁴	<i>n</i> = 423 SB Age: 10–47 years 145 wheelchair LL: nm FI: nm Constipation: nm	FU: 36 months TT: 50 min (15–90)	5-point Likert scale (1 = permanent, 2 = frequent, 3 = occasional, 4 = rare, 5 = never)	Multicentre Clinical interview and medical chart review Right ACE: 40, left ACE: 7, 1.2L tap or saline water Conventional treated group (<i>n</i> = 382) compared to ACE group (<i>n</i> = 41)	6 dropped out ACE group is significant younger ACE group significant less FI Left ACE tends to shorter defecation time and worse FI	Not evaluated
Bani-Hani <i>et al.</i> ³⁵	<i>n</i> = 236, 199 SB Age: 2–36 years LL: nm FI: nm Constipation: nm	FU: 50 months TT: nm	No stool loss	Single center Retrospective chart review Malone ACE daily 642 ml tap water Evaluation of use of additives: - immediate FI: larger volume or more time on toilet - midday FI: bowel cleanout and add 17 mg MiraLAX	196/236 (83%) continence rose to 221/236 (94%) using additives	Not evaluated

Table 3 (Continued)

Author	Sample	FU/ defecation time	Definition fecal continence	Study design	Findings—effectiveness Findings—complications	Findings—satisfaction
Cain <i>et al.</i> ³⁶	<i>n</i> = 236, 199 SB Age: 2–36 years LL: nm FI: nm Constipation: nm	FU: 50 months TT: nm	No stool loss	Single center Retrospective chart review Malone ACE daily, 642 ml tap water	221/236 (94%) continent 98/236 complications 51/236 redo surgery	Not evaluated
Wong <i>et al.</i> ³⁷	<i>n</i> = 64, 41 SB Age: 6–15y LL: nm FI: nm Constipation: nm	FU: nm TT: 5–60 min	No stool loss	Single center Retrospective chart review Interviews and telephone, structured questionnaire score 0–24 (0 = full continence, 24 = complete incontinence) Chait cecostomy glycerin or saline water every 48–72 h	FI score in SB improved from 18 to 7 28/64 complications	Not evaluated
Yardley <i>et al.</i> ³⁸	<i>n</i> = 61, 27 SB Age: 15.5–35.1 years LL: nm FI: nm Constipation: nm	FU: 132 months TT: nm	Stool loss < once per month	Single center Postal/telephone questionnaire	25 dropped out 11/16 (69%) SB continent 5/36 complications 2/36 redo surgery	Satisfaction score of ACE users: 4 (1 = very unsatisfied, 5 = very satisfied) High dissatisfaction with transitional care
Matsuno <i>et al.</i> ³⁹	<i>n</i> = 25 SB Age: 4.1–23.1 years 5 wheelchair LL: nm FI: nm Constipation: nm	FU: 27.5 months TT: 37.7 min (20–60)	No stool loss	Single center Comparing RCE to ACE retrospectively - 13 patients RCE - 12 patients ACE tap water, daily to every 48 h age is significantly (<i>P</i> = 0.009) younger in RCE compared with ACE	10/13 (77%) RCE pseudo-continent 8/12 (75%) ACE pseudo-continent 3/13 RCE independent 8/12 ACE independent	Not evaluated
Bar-Yosef <i>et al.</i> ⁴⁰	<i>n</i> = 21 SB Age: 6–22 years LL: nm FI: nm Constipation: nm	FU: 56 months TT: nm	Not defined	Single center Retrospective chart review Artificial urinary sphincter and Malone ACE	19/21 (90%) full fecal continence 3/21 complications 2/21 redo surgery	Not evaluated
Ok and Kurzrock ⁴¹	<i>n</i> = 23 SB families Age: nm LL: nm FI: nm Constipation: nm	FU: 6 months TT: 45 min	Not defined	Single center FICQoL: 51 item questionnaire before and 6 months after ACE procedure - 23 families before surgery - 18 families after surgery	Diaper need decreased Accident number improved from 3.9 to 0.3 per week (<i>P</i> < 0.01)	Significant improvement in caretaker anxiety, depression and bother
Siddiqui <i>et al.</i> ⁴²	<i>n</i> = 105, 40 SB Age: median 11.1 years LL: nm FI: 21 (10/40 SB) Constipation: 38 Mixed: 46	FU: 6 months TT: 51.7 ± 3.5 min	Stool loss < once per week	Single center Retrospective chart review Percutaneous or surgical ACE 847 ml, GoLYTELY or saline water every 24–48 h 20 concomitant urologic surgery	30/40 (75%) SB continent 74/117 complications 39/117 redo surgery Significant increase in total complications with poor outcome in percutaneous ACE	Not evaluated
Vande Velde <i>et al.</i> ⁴³	<i>n</i> = 40 SB 25 children age: 5–17 years 15 adults age: 18–38 years 16 wheelchair LL: 8S, 30L, 2T FI: nm Constipation: nm	Children FU: 60 months TT: 35 min (15–60) Adults FU: 132 months TT: 60 min (30–120)	No stool loss	Single center Retrospective cohort description Questionnaire at clinic Comparing RCE and ACE use in children and adults - child 18 RCE, 7ACE - adult 4 RCE, 11 ACE	5 dropped out 19/25 (76%) children continent 9/15 (60%) adults continent 5/25 children independent 8/15 adults independent Significant relation between follow-up and continence	2/6 children perceived incontinence as social problem compared with 4/6 adults did

Abbreviations: ACE, antegrade continence enema; CBCL, child behavior checklist; CTT, colonic transit time; FICQoL, fecal incontinence and constipation QoL; LL, lesion level; QoL, quality of life; QOLI, QoL improvement; RCE, retrograde continence enema; SB, *spina bifida*; SPPA, self-perception profile for adolescents; TT, therapy time; YSR, youth self-report.

not mention the type of fluid used.^{28,30,33,40,41} Most authors do not mention the volumes used (9/17).^{28–30,33,37–41} When mentioned, volumes varied between 300²⁷ and 1500 ml (in adults).⁴³ The time spent for treatment was not reported in nine studies.^{28–30,33,35,36,38–40} If reported, it varied from 30^{27,37,43} to 50 min.^{31,32,34,41–43}

Complications are an important issue in case of surgical treatment. The complication rate varied widely. It was not clear whether studies reported all complications in a comparable way. The follow-up time of the different studies is different, also leading to a different complication rate. The most frequently reported complications were stomal stenosis, wound infections and perforations. Redo surgery for complications varied from 5^{29,38} over 10%^{27,36,40} to an even higher rate of 30%.^{28,31,32,42} Overall redo surgery was necessary in 23% (142/616) of patients.

Comparison of the satisfaction rate was not attempted due to the differences in questionnaires and scales used. An improved satisfaction or a high satisfaction rate when using ACE was reported in six studies.^{27–29,31,32,38} One study described a significant improvement in anxiety, depression and bother of both caretakers and patients.⁴¹ However, most studies report a drop-out rate of 5²⁹ to 30%.³⁸ This could be perceived as dissatisfaction of the used strategy. Lack of transition care is, as reported by one study, also an important reason for dissatisfaction. This issue was also reflected in a difference of pseudo-continence rate between children and adults as described in one study, with children achieving higher rates associated with stricter follow-up.⁴³

DISCUSSION

Most studies included in the review report on case series or patient cohorts without controls and therefore no conclusions are drawn regarding best mode of treatment. Further on, data were collected retrospectively in most studies and only a few used standardized questionnaires. Follow-up since starting treatment varied widely. Only two randomized trials both on electrical stimulation were identified. One was a double-blind placebo-controlled study, including, however, a very small patient group without clear end points. Therefore, stating recommendations or drawing conclusions on the different types of treatments used is tentative.

The results indicated that several treatment strategies can achieve pseudo-continence. Large colon washouts, retrograde or antegrade (surgical), however, seem to provide the best overall outcome. The RCE-treated patient group ($n=190$) is smaller and has a shorter follow-up period compared with the patients treated by surgery. Despite the good pseudo-continence results, most authors state that surgery remains the final step in bowel management because of the amount of complications and drop-outs associated with this treatment modality. Some also consider ACE procedures in case of urological interventions. RCE, should be considered as a first-line treatment when both patients and parents are willing to invest in fecal pseudo-continence. Rigorous follow-up and problem solving in case of treatment failure with extra attention for transition care from adolescence to adulthood is of major importance in becoming and staying fecal pseudo-continent.

For all the treatment strategies used, the balance between successful therapy and the daily time investment will influence the compliance. Several studies tried to measure the impact of both fecal incontinence and treatment issues on social life and daily burden. The wide variety in disease severity as well as the multitude of health issues involved make the QoL measurement especially difficult.

Most centers use a stepwise and individually tailored protocol in the treatment of fecal incontinence with ACE surgery as a last step.

These programs are largely experience based and hardly evidence based. A common relevant definition of fecal pseudo-continence is needed to compare study results. In order to ameliorate results and gain insight into which treatment suits which patient, multicenter comparative trials will be needed in patients with comparable impairments, using standardized QoL outcome measurements. Cross-over trials are needed to compare the effect of different irrigation modalities in both RCE and ACE.

CONCLUSION

Irrigations both retrograde and antegrade are valuable treatment options in becoming fecal continent for SB patients. Surgery is currently used as final step in achievement of pseudo-continence. The burden of treatment can be important and should be accounted for. More research is needed to evaluate different techniques, long-term results of treatment and prediction of success or failure using clearly defined fecal pseudo-continence as a primary goal and standardized QoL evaluation as a secondary goal. Motivational support and strict follow-up of SB patients regarding fecal incontinence have an important role in the outcome.

DATA ARCHIVING

There were no data to deposit.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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