

Guidelines for the Care of People with Spina Bifida

Bowel Function and Care

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Introduction

Managing bowel function can be one of the biggest and most important challenges accompanying the diagnosis of Spina Bifida. Spina Bifida results in the loss of normal motor and sensory control in the gastrointestinal tract and anorectal dysfunction. This is defined as a neurogenic bowel dysfunction (NBD). Nervous system lesions above the conus medullaris result in upper motor NBD leading to failure to evacuate the bowel, resulting in constipation or impaction. Lesions at or below the level of the conus medullaris result in lower motor NBD, resulting in failure to contain stool and thus fecal incontinence. Attention to these pathophysiologic causes of neurogenic bowel will lead to a better understanding, better program adherence, and increase the probability of attaining the goals of continence without constipation.^{3,10,24}

In the first 10 clinics in the National Spina Bifida Patient Registry (NSBPR), it was found that less than 30% were continent of stool. Further defining the problem, select NSBPR clinics identified males, non-Hispanic blacks, those with higher lesion levels, and those with public insurance were less likely to have bowel continence. The secondary complications from NBD in Spina Bifida extend beyond constipation and incontinence as these contribute to urinary incontinence, urinary tract infections, shunt malfunction, potential for skin breakdown, hemorrhoids, anal fissures, loss of social and work opportunities, and decreased quality of life.^{4-5,11,17-18,26}

Proactive, systematic, and rational approaches can lead to continence and a more functional lifestyle.^{7,12-24} The following step-by-step guidelines were developed to emphasize management leading to the specific goal of bowel continence without constipation. The guidelines should be considered from least invasive to most invasive.

Tailoring to the individual, considering upper or lower motor bowel dysfunction, is important in the success of the bowel program.^{7,24} In working with school-age children, consider use of school staff to aid in tracking. The school nurse plays a vital role in assisting the child to reach educational goals and manage health concerns.⁹ These guidelines should be followed with the guidance of a health care professional with expertise in bowel management in Spina Bifida.

Outcomes

Primary

1. Maintenance of social continence as appropriate for age level.

Secondary

1. Maximization of independence with managing bowel program.
2. Maximized knowledge and compliance with diet and bowel program.

Tertiary

1. Minimization of constipation.

0-11 months

Clinical Question

1. What evidence exists that prevention of constipation in the first year of life improves the outcome of bowel management in later childhood?

Guidelines

1. Monitor stool frequency, consistency, and amounts.^{7,17}
2. Use dietary management, in particular breastfeeding if possible, as it is easier to digest and offers better restoration of the microbiome after surgery.²⁵ (clinical consensus)
3. Consider dietary management (fiber and fluids) before pharmacologic adjuncts (sennoside), and/or rectal stimulants (glycerin suppositories) to manage constipation.^{7,24,26}
4. Use barrier creams to protect perineal area from breakdown as needed.² (Integument (Skin) Guidelines).

1-2 years 11 months

Clinical Questions

1. Is there evidence to support the benefit of toilet training at the same developmental stage as peers without dysfunction?

Guidelines

1. Discuss toilet training and habit training with parents.^{7,17,24,26}
2. Establish goal of working toward bowel continence.^{7,17,24,26}
3. Focus on fiber, fluids, exercise, and timed bowel movements after meals.^{6,13,24}
4. Consider two-pronged approach of oral and rectal interventions to meet the goal of bowel continence without constipation.^{3,6-7,14,24,26}
5. Use dietary management (fiber and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation and fecal incontinence.^{3,6-7,14,24,26}
6. Use barrier creams to protect perineal area from breakdown as needed.²
7. Refer to a Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)

3-5 years 11 months

Clinical Questions

1. Is there evidence that “habit training,” or forced evacuation with stimulants such as suppositories or enemas, increases social continence?

Guidelines

1. Discuss consequences of constipation and bowel incontinence (including shunt malfunction, urinary tract infections (UTIs), skin breakdown, social isolation.^{4,7,11,18-20,24,26})
2. Establish the goal of bowel continence and institute the bowel continence program using guidelines below.^{6-7,13,17,24,26}
3. Focus on fiber, fluids, exercise, and timed bowel movements after meals.^{6,13,24}
4. Consider two-pronged approach of oral and rectal interventions to meet the goal of bowel continence without constipation or fecal incontinence.^{3,6-7,14,24,26}
5. Use dietary management (fiber and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation and fecal incontinence.^{3,6-7,14,24,26}
6. Use barrier creams to protect perineal area from breakdown as needed.²

7. Refer to a Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)

6-12 years 11 months

Clinical Questions

1. What is the evidence that the Malone Antegrade Continence Enema (MACE) procedure or continent cecostomy is an effective form of bowel management in children with refractory incontinence?
2. What are the most effective protocols for MACE?
3. What is the evidence that electrical stimulation (trans-rectal or intravesicular) provides benefit for increased bowel continence?

Guidelines

1. Discuss consequences of constipation and bowel incontinence (including shunt malfunction, urinary incontinence, UTIs, skin breakdown, social isolation)^{4,7,11,18-20,24,26} and focus on developing independent management skills. (Self-Management and Independence Guidelines)
2. Establish the goal of bowel continence and institute the bowel continence program using the guidelines below.^{7,17,24,26}
3. Assist the child with learning how to minimize and manage bowel accidents.^{9,26}
4. Use barrier creams to protect perineal area from breakdown as needed.²
5. Keep a bowel habit diary to better understand triggers for incontinence and overall patterning to direct a choice of options for bowel management.^{7,9,17,24,26}
6. Focus on fiber, fluids, exercise, and timed bowel movements after meals.^{6,13,24}
7. Consider twofold attack of oral and rectal interventions to meet the goal of bowel continence without constipation or fecal incontinence.^{3,6-7,14,24,26}
8. Use dietary management (fiber, fiber supplements, and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation.^{3,6-7,14,24,26}
9. Discuss other options for treatment if the above have failed, including cone enema or other transanal irrigation, cecostomy, or MACE.^{1,3,5,14-15,21,24,26}
10. Refer to a Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)

13-17 years 11 months

Clinical Questions

1. What support is needed by teens with Spina Bifida to be successful in maintaining their bowel program?
2. Is there evidence that hormonal fluctuations impact continence?

Guidelines

1. Discuss consequences of constipation and bowel incontinence (including shunt malfunction, urinary incontinence, UTIs, skin breakdown, social isolation)^{4,7,11,18-19,2- ,24,26} and focus on developing independent management skills. (Self-Management and Independence Guidelines)
2. Establish or maintain the goal of bowel continence and institute or maintain the bowel continence program using the guidelines below.^{7,17,24,26}
3. Assist the child with learning how to minimize and manage bowel accidents.^{7,9- 10,17,24,26}
4. Use barrier creams to protect perineal area from breakdown as needed.²
5. Keep a bowel habit diary to better understand triggers for incontinence and overall patterning to direct a choice of options for bowel management.^{7,9,17,24,26}
6. Focus on fiber, fluids, exercise, and timed bowel movements after meals.^{6,13,24}

7. Consider a twofold attack of oral and rectal interventions to meet the goal of bowel continence without constipation.^{3,6-7,14,24,26}
8. Use dietary management (fiber, fiber supplements, and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation.^{3,6-7,14,24,26}
9. Discuss other options for treatment if the above have failed, including cone enema or other transanal irrigation, cecostomy, or antegrade continence enema (Malone).^{1,3,6,14-15,21,24,26}
10. Refer to Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)
11. Access support services for personal care, if needed.^{9,12,26}

18+ years

Clinical Questions

1. What impact does pregnancy have on bowel management or on use of a cecostomy or MACE? (Women's Health Guidelines)
2. Does early chronic constipation impact management of constipation in adult years?
3. Is there a change in bowel function later in life that should be addressed with a more aggressive bowel program? Does menopause result in changes?

Guidelines

1. Discuss consequences of constipation and bowel incontinence (including shunt malfunction, urinary incontinence, UTIs, skin breakdown, social isolation).^{4,7,11,18-19,20,24,26}
2. Establish the goal of bowel continence and institute the bowel continence program using timing, suppositories, pharmacologic agents or enemas as needed.^{7,17,24,26}
3. Assist the adult with learning how to minimize and manage bowel accidents.^{7,9-10,17,24,26}
4. Use barrier creams to protect perineal area from breakdown as needed.²
5. Keep a bowel habit diary to better understand triggers for incontinence and overall patterning to direct a choice of options for bowel management.^{7,9,17,24,26}
6. Discuss management of bowel program as it may impact sexual relations.^{4,11,26}
7. Focus on fiber, fluids, exercise, and timed bowel movements after meals.^{6,13,24}
8. Consider a twofold attack of oral and rectal interventions to meet goal of bowel continence without constipation or fecal incontinence.^{3,6-7,14,24,26}
9. Use dietary management (fiber, fiber supplements, and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol, lubiprostone, or other prescription), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation.^{3,6-7,14,24,26}
10. Discuss other options for treatment if the above have failed, including cone enema or other transanal irrigation, cecostomy, or MACE.^{1,3,6,14-15,21,24,26}
11. Refer to a Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)
12. Access support services for personal care if needed.^{9,12,26}

Research Gaps

1. Are there benefits to probiotic use in the population with Spina Bifida? Discuss consequences of constipation and bowel incontinence (including shunt malfunction, urinary incontinence, UTIs, skin breakdown, social isolation).^{4,7,11,18-19,20,24,26}
2. What is the evidence that electrical stimulation (trans-rectal or intravesicular) provides benefit for bowel continence in Spina Bifida?

3. Does an individualized, stepped approach to bowel management in patients with Spina Bifida lead to less constipation and incontinence?
4. What factors contribute to a successful bowel program in the population with Spina Bifida?
5. What challenges do pregnancy or menopause create for an established bowel program?
6. What are the relative efficacies of interventions for management of incontinence and constipation in the setting of neurogenic bowel in Spina Bifida by age group?
7. What evidence exists that prevention of constipation in the first year of life improves the outcome of bowel management in later childhood?
8. Is there evidence to support the benefit of toilet training a child with Spina Bifida at the same developmental stage as peers without dysfunction?
9. Is there evidence that "habit training," or forced evacuation with stimulants such as suppositories or enemas, increases social continence?
10. What is the evidence that the MACE procedure or continent cecostomy is an effective form of bowel management in children with refractory incontinence?
11. What are the most effective protocols for cecostomies?
12. What support is needed by teens with Spina Bifida to be successful in maintaining their bowel program?
13. Is there evidence that hormonal fluctuations impact continence?
14. Does early chronic constipation impact management of constipation in adult years?
15. Is there a change in bowel function later in life that should be addressed with a more aggressive bowel program?

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