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Constipation Management in Congenital Myopathy

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The information provided in this booklet is not intended to replace medical advice from a physician. Always consult your medical providers on decisions related to your health.



Photograph of a woman's torso, wearing a light blue dress, and holding a digitally drawn image of the large and small intestine.
Photography by Stacy Cossette, Digital illustration by Lucinda Lee

Understanding Constipation in Congenital Myopathy

Constipation in congenital myopathy (CMyo) is common, chronic, and often severe, yet misunderstood and sometimes not treated properly. Obtaining the appropriate care when it's needed can be difficult because many GI clinicians do not routinely manage the complex relationship between myopathic dysfunction and gastrointestinal motility, often missing how the neuromuscular disease impacts the GI system. In CMyo, constipation is often due to a set of systemic neuromuscular issues presenting as a GI symptom. When clinicians apply standard patient protocols that rely heavily on fiber and harsh stimulants, they fail to account for the unique neuromuscular limitations of the skeletal muscles, diaphragm, abdominal wall, and smooth muscle physiology.

For example, the diaphragm plays an important role in GI motility. When the diaphragm is weak, the intra-abdominal pressure gradient system required for bowel evacuation is inadequate. Plus, the resulting constipation can further restrict diaphragm movement, making it more difficult to obtain adequate breaths to ventilate the lungs. While the nervous system and the smooth muscles of the gut are usually intact, patients often lack sufficient intra-abdominal pressure because of muscle weakness. Effective constipation management in individuals with CMyo requires an approach that recognizes the underlying muscle disorder and should include coordinated, multidisciplinary care.

Patients with CMyo are at significantly increased risk of complications related to constipation, and

the commonly used treatments, compared with the general population. As a result, careful monitoring during episodes of constipation or diarrhea is recommended, including assessment of hydration status and blood tests for electrolytes, vitamins, and minerals.

Standard laboratory reference ranges are based on population averages and may not fully reflect an individual's physiology, particularly in the context of CMyo. Patients should discuss their personal baseline values and condition-specific factors with their clinician when interpreting results.

Because myopathy severity and individual physiology vary widely, these guidelines should be used as a flexible framework rather than a rigid prescription. Effective constipation management requires tailoring strategies to each patient's specific needs and clinical presentation, ideally in partnership with their medical team.

Our purpose is to provide guidance on proactive management strategies for chronic constipation and rescue options for acute impaction in individuals with CMyo, with consideration for intake restrictions, mobility differences, and ventilatory concerns.

Recognizing the Signs: What constipation could look like in a CMyo patient

- ◆ Delayed stomach emptying (gastroparesis)
- ◆ Reduced appetite
- ◆ Feeling full after eating very little
- ◆ Infrequent bowel evacuations (less than 3 per week)
- ◆ Hard, lumpy, or small pieces of stool coming out, possibly every day
- ◆ Liquid stool, which is seeping around impacted stool
- ◆ Softer stool that is still difficult to get out
- ◆ Straining on the toilet to get something out
- ◆ If something does come out, feeling like there's more that just won't come out
- ◆ Bloating, distention of the abdomen
- ◆ Abdominal cramping or discomfort
- ◆ Rectal discomfort
- ◆ Increased work to breathe
- ◆ Shallower breaths
- ◆ Reflux or vomiting
- ◆ Nausea
- ◆ Behavioral signs in nonverbal patients such as agitation



Young white woman in a power wheelchair clutching her stomach in pain. Image generated by Gemini AI, 2026.

MYTH: Having diarrhea means you are not constipated.

FACT: When hard stool is present, liquid stool can leak around the impaction, a condition commonly called overflow diarrhea or paradoxical diarrhea. The passage of liquid stool often leads to the mistaken assumption that the patient requires a reverse in treatment, although a significant hardened stool impaction still remains. It is important to recognize this misconception, since increasing bulking agents or taking anti-diarrheal medications will make symptoms worse.

What Causes Constipation in CMyo?

Constipation in CMyo usually results from a combination of some of the following:

- ◆ Impaired abdominal wall & diaphragm muscle function leading to poor defecatory force
- ◆ Poor coordination of pelvic floor muscles with abdominal muscles
- ◆ Reduced mobility that leads to decreased colonic motility
- ◆ Disuse atrophy worsening muscle weakness
- ◆ Scoliosis compressing the intestines
- ◆ Smooth muscle weakness of the GI tract may be a factor for some patients
- ◆ Energy deficits
- ◆ Hypoventilation (underbreathing)
- ◆ Positive airway pressure mechanical ventilation use that results in altered intra-abdominal pressure dynamics
- ◆ BPAP device pushing air into the stomach & intestines that alters the bowel's rhythm (peristalsis)
- ◆ Low volume and/or nutritionally inadequate feedings
- ◆ Dietary choices that create constipation
- ◆ Chronic dehydration
- ◆ Feeding constraints due to chewing and swallowing impairment
- ◆ Constipating medications and supplements

Medication Side Effects:

Anticholinergics, which are often used to manage secretions/drooling, significantly dry out the gut and slow motility.

Pain Medications: Opioid medications commonly slow the intestines and can cause severe constipation. Examples include oxycodone, hydrocodone, morphine, fentanyl, and tramadol.

Supplements: Iron or calcium supplements, often prescribed for bone density or anemia in non-ambulatory patients, are highly constipating.

KEY POINT

Constipation in CMyo is multifactorial, not simple fiber deficiency, which is a common clinical misconception that can lead to ineffective treatment for many patients.

Interventions: Prevention, Maintenance, and Rescue Therapies

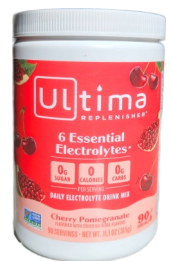
This section covers interventions, from maintenance therapy through rescue therapy. The goal in maintenance therapy for bowel health is to keep things moving, preventing stool backup and impaction. For many CMyo patients, bulking-up the stool with fiber is counterproductive. Stimulant laxatives can be too harsh for frequent use, although some patients may experience good results from occasional use, such as once per week. The best results are usually obtained with daily management that is proactive, gentle, consistent, and sustainable for the lifetime. Therefore, the foundation of a maintenance plan begins with thoughtful, daily dietary choices that can help keep stool moist and support more consistent movement through the digestive system.

Chronic Management Approaches

1. Dietary strategies that promote consistent bowel function:

✓ Hydration

- ◆ **Water:** When the body is even mildly dehydrated, the colon pulls extra water out of stool to compensate, so drink plenty throughout the day.
- ◆ **Electrolyte powder** added to water: Some CMyo patients find these drinks more effective for hydration and better tolerated than large volumes of plain water. Ultima brand is recognized for its completeness, taste, and quality.
- ◆ **Prune juice:** Contains sorbitol, a natural sugar alcohol that draws water back into the bowel, keeping stool moist and soft.
- ◆ **Hot tea or coffee** (decaf or caffeinated): Having tea or coffee first thing after waking each morning, before eating stimulates the bowels. Choose tea for a mild stimulating effect and coffee for a moderate stimulating effect.
- ◆ **Hydrating foods:** Soups, gelatin desserts, and yogurt are carriers for fiber and fluid.



Photograph by Lucinda Lee



Digital illustration of a bowl of soup. Illustration by Lucinda Lee

✓ Foods that encourage motility

- ◆ **Kiwi fruit:** Contains actinidin, an enzyme that improves motility.
- ◆ **Oatmeal and creamy rice cereals:** A soluble fiber that attracts moisture into the stool.
- ◆ **Peeled fruits and well-cooked vegetables:** Easier to digest with better motility.
- ◆ **Pureed foods & smoothie drinks:** Blending fruits and vegetables with plenty of fluid helps ensure good hydration and a balanced fiber intake. You can remove peels before pureeing or blending, when appropriate.
- ◆ **Magnesium-rich foods:** Creamy nut butter, ground & saturated chia, and pureed legumes.
- ◆ **Fermented foods:** Greek yogurt, kefir, refrigerated pickles, fermented soybeans (i.e. miso, tempeh), sour cream, cottage cheese, aged cheese, buttermilk, acidophilus milk, tofu, and soymilk.



A strawberry smoothie surrounded by various fruits. Image generated by Gemini AI 2026.

✗ What to avoid

- ◆ Raw vegetables, whole grains, and peelings are insoluble fibers and counterproductive, acting like an anchor to hold food in-place, causing and worsening constipation.

It's important to maintain the right balance of fat, protein, and carbohydrates daily. Five mini-meals per day is usually better tolerated than three big meals. Find the right balance for your body between volume of food consumed and triggers of colonic movement.

Quick Tips:

- Opt for warm or hot instead of cold liquids and foods.
- Inadequate fluid leads to worse constipation.

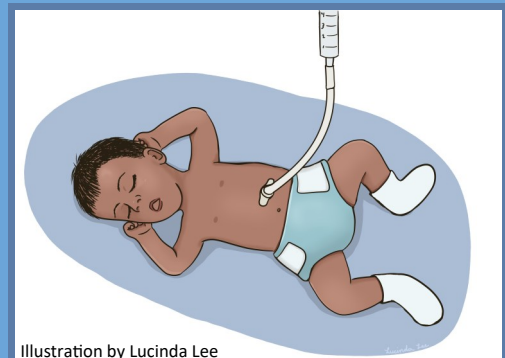


Illustration by Lucinda Lee

GASTROSTOMY TUBES

When oral intake is insufficient, a feeding tube may be needed. For G-tube users all intake must be finely pureed and well-hydrated.

Mix thoroughly with water or formula and flush the tube well after administration to prevent clogging of the tube.

2. Medications that promote consistent bowel function:

Adding a low maintenance dose of polyethylene glycol or partially hydrolyzed guar gum can also be useful for daily management.

- **Polyethylene glycol** (PEG / Miralax) works well in daily doses to keep stools hydrated and softer.
 - ◆ Small doses are adjusted based on the diet that day
 - ◆ Can be used daily for years to keep stools consistently soft
 - ◆ Well tolerated
- **Partially hydrolyzed guar gum** (PHGG) is best as tiny daily maintenance doses rather than “rescue” dosing.
 - ◆ Mix a small amount into water, smoothies, yogurt, cereal, coffee, etc.
 - ◆ Ferments slowly resulting in less gas
 - ◆ Improves stool frequency and consistency
 - ◆ Safe for daily, lifelong use, but start low and gradually increase the amount to your optimal level

MYTH: Probiotics can help constipation.

FACT: There is currently no clinical evidence to support a benefit of probiotics in maintaining bowel function or managing constipation in patients with CMyo.

3. Body Movement and Positioning

Your intestines also rely on your body’s motion to move stool along. Even for those with significant weakness, shifting positions, stretching, isometrics, or seated limb exercises can help trigger the bowel’s natural contractions. Movement encourages blood flow to the digestive tract, helping with its function. Body core engagement helps push stool toward the rectum. If a patient lacks anti-gravity movement due to severe muscle weakness, then a caregiver could help with range-of-motion and stretching maneuvers, known as passive movement. Standing frames are also helpful in preventing constipation in those who lack the strength to stand on their own. Physical therapy is recommended as a life-long strategy to optimize your health, including your bowel health. Pelvic floor therapy may be helpful as it can be difficult for patients to coordinate the anal sphincter relaxing along with the abdominal muscles contracting in a coordinated fashion. Biofeedback is suggested to train muscle coordination to help with better evacuation of stool.

Mechanical & Positional Tips

1. Scheduled toileting

- ◆ Same time daily
- ◆ After feeding to take advantage of the gastro-colic reflex

2. Positioning

- ◆ Foot support with squatting posture for emptying bowels
- ◆ Sit up while toileting; avoid reclined positions

- ◆ Abdominal support garments can help some patients

3. Abdominal massage

- ◆ Gentle, clockwise colon-tracking massage
- ◆ Evidence supports benefit in neuromuscular disease

Using a footstool (like an ergonomic Squatty Potty) while sitting on the toilet can make bowel movements easier and more effective by putting your body in a more natural position. Raising your knees above your hips and leaning slightly forward helps straighten the rectum, allowing stool to pass more easily. Imaging studies show that this position opens the rectum more than just sitting upright, and it also reduces the amount of pressure needed in your abdomen, meaning less straining. Many people who use a footstool often find they can go more quickly, with less effort, and empty more completely.



“Comparison of Squatty Potty and alternative market options” from PEGA Bidet, 2025

Advanced Management Approaches

Daily management strategies are essential for preventing constipation and promoting regular bowel function, but some people may continue to experience symptoms despite these efforts. When constipation persists or becomes more severe, additional treatment options may be needed. The following are some advanced management approaches that can help support bowel function when dietary measures, hydration, movement, and basic maintenance therapies are not enough.

◆ **Lactulose**

- A laxative made from lactose to manage chronic constipation
- A synthetic sugar in syrup-like form appropriate for oral feeders or diluted tube feedings.
- Caution: may cause gas and bloat

◆ **Polyethylene glycol** (PEG / Miralax) in a higher dose than usual

- Non-stimulant powder added to ample liquid before consuming to help soften stool
- Appropriate for oral and tube feeders

◆ **Prucalopride** (Motegrity) for adult patients who can safely swallow

- Taken daily as a maintenance drug
- Tablets must be swallowed whole, never crushed
- Total gut prokinetic, can enhance gut motility from esophagus to rectum to help propel stool along
- Caution: may cause depression or suicidal ideation in rare instances

◆ **Linzess** (Linaclotide) for adults and children over age 6

- Taken daily as a maintenance drug
- Capsules can be swallowed whole or opened and the contents put into a soft food or water, take before eating anything
- Acts locally within the GI tract
- Bowel movements often occur in 1 to 7 days

◆ **Magnesium hydroxide** (Milk of Magnesia)

- At low doses, softens stool and adds volume through osmosis
- Well tolerated
- Bowel movements often occur 2-6 hours after dosing

◆ **Magnesium citrate**

- Use low dose (15-30 mL) if combined with any of the above treatments
- Non-stimulant liquid helps soften stool
- Appropriate for oral and tube feeders
- Stronger/harsher than PEG / Miralax
- Bowel movements often occur in 4-8 hours after dosing

Repeated or large doses of magnesium-based products can lead to a build-up of the mineral in the body, which is why you should take the lowest effective dose. Medication risks are greater in patients with CMYo compared to the healthy population. Please consult your specialist for guidance.

If upper GI symptoms, such as severe reflux or slow stomach emptying, are chronic concerns, try a medication that accelerates gastric emptying.

When to Seek Medical Evaluation

If abdominal distension, bloating, or discomfort persist, a medical work up may be appropriate. This can include a review of symptoms, history of the problem, and a physical exam including palpation of the abdomen and rectal exam by a trained clinician. Imaging studies may also be useful, such as abdominal ultrasound or X-ray, which can verify retained or impacted stool. These findings provide objective evidence of ongoing constipation and guide appropriate treatment.



Black woman in a power wheelchair using non-invasive ventilation.
Image generated by Gemini AI 2026.

SPECIAL CONSIDERATIONS FOR MECHANICALLY VENTILATED PATIENTS

Proactive daily bowel management is medically necessary for patients with breathing impairment because constipation can 1) alter intra-abdominal pressure, 2) reduce diaphragmatic movement, 3) worsen ventilation efficiency, and 4) increase reflux and aspiration risk.

Rescue Treatment Approaches

Lower-GI Rescue Strategies

When patients are already experiencing stool impaction, maintenance therapy is ineffective. Sometimes the rectum is plugged with stool, so removing lower colon fecal matter would be the next step. The goal of rescue therapy is rapid evacuation of the stool, not more stool softening or bulking.

Preferred acute rescue options, sometimes called “Rectal Therapy”, for when stool is in the rectum (from least to most powerful effect):

1. **Rectal glycerin suppository** is a gentler stimulant laxative delivered directly into the rectum.

- ♦ Gentle stimulant effect, draws water into the rectum, triggers a local response, useful when hard stool is already near the rectum
- ♦ Slide the suppository between the stool ball and rectal lining
- ♦ Usually works in 5-30 minutes with minimal urgency and discomfort
- ♦ If this is ineffective, move-on to one of the next steps in this category

2. **Mini-enema** (2.25 fluid ounces) into the rectum.

- ♦ Works rapidly, 2-15 minutes
- ♦ Has a mechanical stretch effect that triggers motility

3. **Rectal bisacodyl suppository** is a stimulant laxative delivered directly into the rectum.

- ♦ Usually works in 10-30 minutes
- ♦ Works directly on stool that has plugged the rectum
- ♦ Cramping is normal; that means it’s working
- ♦ If no fecal matter is discharged after 60 minutes, another saline enema may be tried, or escalate to the next step

Because treatment doses are tailored to body weight and individuals with CMyo are often underweight, many favor smaller-volume options like mini-enemas, low-volume PEG, and diluted magnesium.

4. **Bowel irrigation** systems with manual disimpaction and removal as a last resort for stool trapped in the rectum.

- ♦ Both require training to perform correctly and safely
- ♦ Manual pump enema systems (e.g., Peristeen Anal Irrigation System; PAI), also known as rectal irrigation or transanal irrigation (TAI), are used in patients with a history of extensive bowel management programs, neuromuscular bowel dysfunction, and chronic constipation.
- ♦ Tap water or saline enema using a kit purchased online or from your clinician.

Quick Tip:

- Prioritize rectal rescue methods over oral stimulants when you are unsure , or if you know, that the fecal impaction is toward the rectum just to ensure that lower impaction is cleared before stimulating the upper colon.

Upper-GI Rescue Strategies

The following suggestions are ordered from mild to powerful stimulant action.

- Oral Senna is a mild stimulant that usually produces a bowel movement in 6-12 hours.
- Oral Bisacodyl is a stimulant that takes up to 6-12 hours to produce a bowel movement.
- Castor oil is a powerful stimulant that usually produces a bowel movement in 2-6 hours. Ask your medical specialist if castor oil is appropriate for you as it can cause intense cramping, nausea, and rapid fluid loss.

MYTH: Using stimulant laxatives repeatedly as rescue therapy in CMyo patients leads to increased fatigue or bowel dependence.

FACT: There is no convincing evidence to support this concern.

When Rescue Therapy Isn't Working

If rectal interventions fail to produce good results, it may indicate a more significant lower bowel obstruction or a buildup of stool higher in the digestive tract. Likewise, if oral rescue therapies do not result in meaningful stool evacuation, additional medical evaluation may be needed.

Persistent abdominal distension, worsening bloating, ongoing pain, vomiting, increased breathing difficulty, or continued inability to pass stool despite rescue therapy should not be ignored. Imaging studies such as abdominal X-rays or ultrasound may help identify retained stool, impaction, or other complications and guide further treatment.

Large amounts of retained stool can occasionally lead to serious complications, including bowel obstruction or volvulus (twisting of the intestine), which requires urgent medical attention. If symptoms are worsening or rescue measures are not working, contact your GI specialist, primary care provider, urgent care clinic, or emergency department, as appropriate.

Why Constipation Should Be Taken Seriously

Chronic constipation is sometimes not treated as the serious medical condition that it is. In people with CMyo, constipation can affect far more than bowel function. Stool retention can increase intra-abdominal pressure, restrict diaphragmatic movement, worsen ventilation efficiency, increase reflux and aspiration risk, and contribute to discomfort and fatigue.

When constipation is severe or prolonged, pressure within the colon can reduce blood flow to the intestinal wall, leading to inflammation and tissue damage. One particularly serious complication of this is stercoral colitis, a condition associated with a high mortality rate if left untreated. This complication is most common in patients with reduced mobility or immobility.

Patients with CMyo are also more vulnerable to complications related to constipation. Low muscle mass, limited nutritional reserves, reduced nutritional intake, high energy demands of respiratory work, and stimulant side-effects such as dehydration or electrolyte imbalances make careful monitoring especially important. Effective constipation management for CMyo patients requires an individualized approach tailored to the specific needs of this population that recognizes the unique challenges of living with this condition.



Four people are conversing outside. From left to right: a young Black boy who has a tracheostomy sits in a power wheelchair. His mother stands behind him. In the middle, a young Hispanic woman sits in a mobility scooter looking towards the mother and son. On the right, an older white woman is in a power wheelchair looking at the others and smiling. Image generated by Gemini AI 2026.

10 Things Every CMyo Patient Should Know About Constipation

1. **Constipation in CMyo is usually more than a fiber problem.** Muscle weakness, breathing mechanics, mobility limitations, and nutritional factors often play important roles.
2. **Liquid stool can still mean constipation.** Overflow or paradoxical diarrhea occurs when liquid stool leaks around an impaction.
3. **Careful health monitoring is necessary.** Often done through blood panels, hydration, electrolyte, vitamin, and mineral levels should be monitored, especially during significant constipation or diarrhea.
4. **Daily prevention is easier than rescue treatment.** Consistent hydration, nutrition, movement, and maintenance therapies are the foundation of bowel health.
5. **The best treatment plan is individualized.** Constipation treatment for CMyo patients must be tailored to the specific needs of this population and the individual person.
6. **Patients with weak abdominal muscles may respond better to rectal therapies.** People with CMyo often have weak abdominal muscles making oral treatments that stimulate intestinal muscle contraction less effective. Therefore, rectal agents may be more reliable.
7. **Swallowing difficulties should be considered when choosing medication formulations.** Liquid, powdered, crushable, compounded, and tube-compatible options may be necessary for patients with swallowing impairment.
8. **Many common medications can worsen constipation.** Review all medications and supplements with your healthcare team. There are also online resources that can help (see Appendix for examples).
9. **Start low and adjust gradually.** Escalating therapies too quickly can increase cramping. To avoid this, stagger osmotic and stimulant therapies and always start on a low-dose.
10. **Constipation should be taken seriously.** Severe constipation can affect breathing, nutrition, comfort, and overall health.

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Cover image generated by Gemini AI.

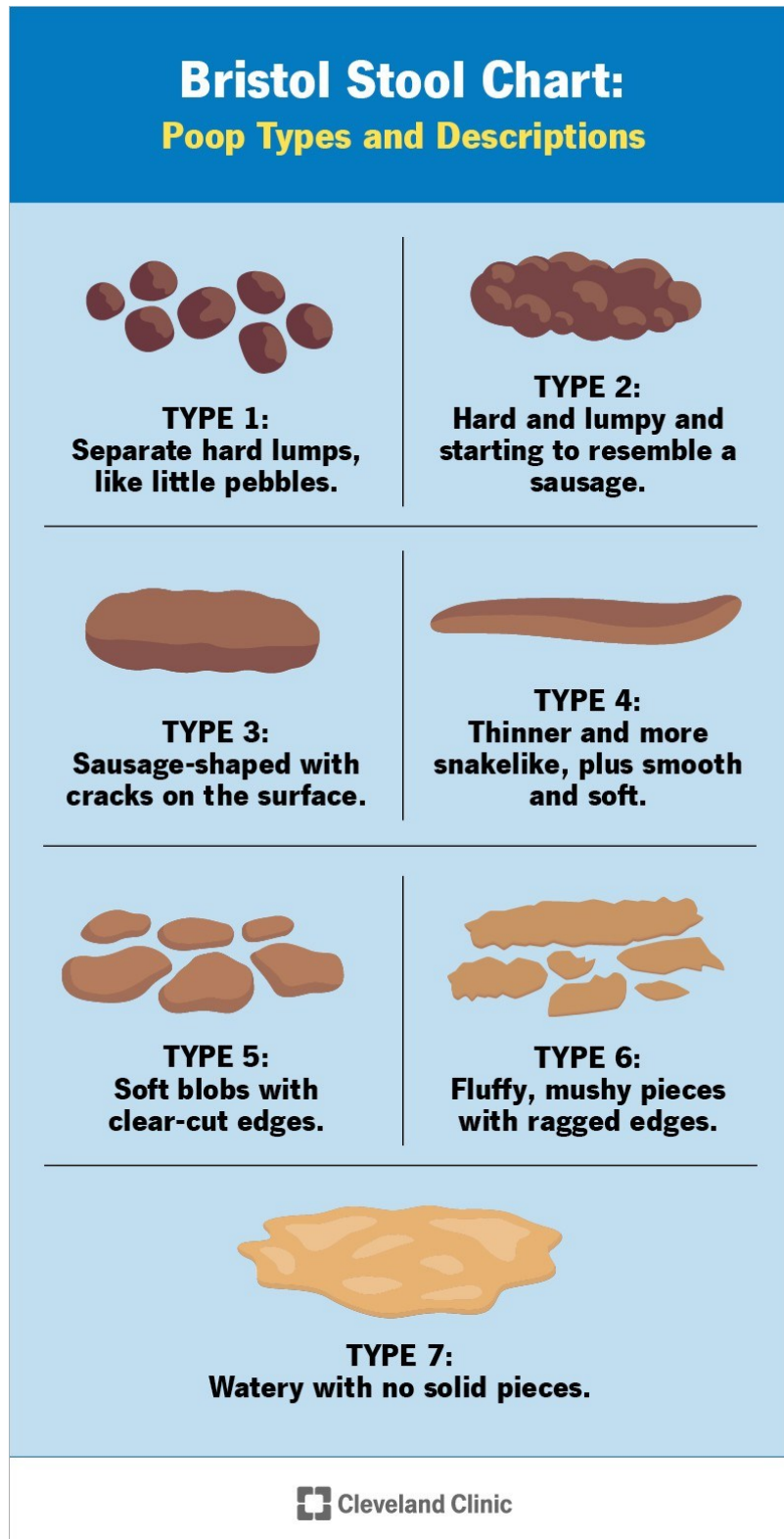
Conflict of Interest Statement

The authors and contributors declare no financial, commercial, or professional conflicts of interest related to the content of this guidebook. The information presented is intended solely for educational purposes and reflects the collective knowledge, experiences, and perspectives of the contributors.

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Appendix A: Bristol Stool Chart



The Bristol stool chart is designed to help patients track trends in their bowel movements by classifying the shape and consistency of feces into seven types. Types 1–2 indicate constipation, 3–4 are considered ideal, and 5–7 are trending to diarrhea.

Appendix B: Bowel Tracking Log

Printable chart for tracking bowel movements.

DATE	TIME	BRISTOL STOOL TYPE	AMOUNT	PAIN	OTHER SYMPTOMS
			☐ small ☐ medium ☐ large	☐ yes ☐ no	
			☐ small ☐ medium ☐ large	☐ yes ☐ no	
			☐ small ☐ medium ☐ large	☐ yes ☐ no	
			☐ small ☐ medium ☐ large	☐ yes ☐ no	
			☐ small ☐ medium ☐ large	☐ yes ☐ no	
			☐ small ☐ medium ☐ large	☐ yes ☐ no	
			☐ small ☐ medium ☐ large	☐ yes ☐ no	

Appendix C: Additional Resources

The Big 8 Constipation-Causing Medications

<https://www.goodrx.com/conditions/constipation/the-big-8-constipation-causing-medications>

The Care of Congenital Myopathy: A Guide for Families

Kindle and Paperback (English) — <https://www.amazon.com/Care-Congenital-Myopathy-Guide-Families/dp/B0GDSL1ZCK>

PDF (English) — <https://buildingstrength.org/wp-content/uploads/sites/542/2026/07/CMFG-English-Version.pdf>

PDF (Spanish) — <https://buildingstrength.org/wp-content/uploads/sites/542/2026/07/CMFG-Spanish-Version.pdf>

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