

spinal muscular atrophy

# A guide to nutrition for patients with spinal muscular atrophy

This guide provides practical advice and information to help you make informed choices about your diet and manage your symptoms effectively



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### Introduction

Living with Spinal Muscular Atrophy (SMA) presents unique challenges, and nutrition plays a vital role in maintaining your overall health and well-being. This guide, developed based on a comprehensive review of medical studies and expert recommendations, aims to empower you with the knowledge to make informed decisions about your diet and manage your SMA symptoms effectively. We'll explore the connection between SMA and nutrition, discuss general dietary guidelines, delve into specific nutritional needs, and provide practical advice, meal planning tips, and recipes.

# Understanding spinal muscular atrophy and its impact on nutrition

Spinal Muscular Atrophy (SMA) is a genetic condition that affects the motor neurons, the nerve cells responsible for transmitting signals from the brain to the muscles. This can lead to muscle weakness, atrophy (shrinkage), and difficulties with movement. SMA can affect individuals of all ages, and its severity varies. While the primary impact of SMA is on the muscles, it can also indirectly affect other bodily functions, including digestion, metabolism, and bone health [1].

SMA can affect the muscles involved in swallowing, leading to a condition called dysphagia. Dysphagia can make it difficult to consume enough calories and fluids, increasing the risk of malnutrition and dehydration [2]. In some cases, it can also lead to aspiration, where food or liquid enters the airway and lungs, potentially causing aspiration pneumonia [2].

Furthermore, SMA can affect the enteric nervous system (ENS), which is a network of nerves in the digestive tract. This can lead to various gastrointestinal symptoms, such as constipation, gastroesophageal reflux (GERD), and delayed gastric emptying [3]. These digestive issues can further complicate nutritional management.

It's important to remember that proper nutrition is not just about consuming enough calories. It's about providing your body with the essential nutrients it needs to function optimally. Good nutrition can support lung health, improve breathing, and enhance your overall quality of life [4].

# General dietary guidelines for people with spinal muscular atrophy

While individual needs vary, some general dietary guidelines can help you make healthier choices:

- **Prioritize nutrient-rich foods:** Focus on a balanced diet that includes fruits, vegetables, whole grains, lean protein, and healthy fats [6]. These foods provide essential vitamins, minerals, and antioxidants to support your body's functions.
- **Choose whole grains:** Opt for whole-grain bread, cereals, and pasta over refined grains. Whole grains are higher in fiber, which can help with digestion and prevent constipation, a common issue for people with SMA [7].
- **Include lean protein:** Protein is essential for maintaining muscle mass and strength. Good sources include poultry, fish, beans, lentils, and low-fat dairy products [8].
- **Incorporate healthy fats:** Include sources of healthy fats like olive oil, avocados, nuts, and seeds in your diet. These fats are important for overall health and help your body absorb essential vitamins [1].
- Embrace quercetin-rich foods: Quercetin is a natural compound found in various fruits and vegetables that has antioxidant and anti-inflammatory properties. Some studies suggest that quercetin may have potential benefits for people with SMA [6]. Include foods like apples, berries, onions, and leafy greens in your diet.
- **Limit added sugars:** Minimize your intake of sugary drinks, candies, and desserts. These foods provide empty calories and may contribute to weight gain [9].

- **Reduce sodium intake:** Limit processed foods, canned soups, and salty snacks. High sodium intake can contribute to fluid retention and high blood pressure [9].
- **Stay hydrated:** Drink plenty of fluids throughout the day. This is especially important if you have difficulty swallowing or experience constipation [10].
- Eat smaller, more frequent meals: This can help with digestion, prevent reflux, and maintain stable blood sugar levels [7].

# Specific nutritional needs for spinal muscular atrophy patients

SMA can affect your body's ability to metabolize nutrients, so it's essential to pay attention to specific needs:

#### Vitamin D and Calcium

These are crucial for bone health, as individuals with SMA are at an increased risk of osteoporosis and fractures [1]. Ensure you get enough vitamin D through sunlight exposure, fortified foods, or supplements, and consume calcium-rich foods like dairy products, leafy greens, and fortified plant milk.

#### **Essential Fatty Acids**

These fats are vital for brain development and overall health. Include sources like fatty fish (salmon, tuna), avocados, and plant-based oils (flaxseed, canola) in your diet [10].

#### Fiber

Adequate fiber intake is essential for digestive health and preventing constipation. Aim for your age plus 5 grams of fiber per day [11]. Good sources include fruits, vegetables, whole grains, and legumes.

#### Hydration

Staying hydrated is crucial for overall health, especially if you have difficulty swallowing or experience constipation [10]. Aim for 1 ½ ounce of fluid per pound of body weight for adults and ½ ounce per pound for children [12].

#### Protein

Protein is essential for maintaining muscle mass and overall health, but requirements can vary widely among individuals with SMA depending on disease progression, physical activity, and any concurrent health conditions.

- For individuals with higher muscle activity levels or those with milder forms of SMA, protein intake may be in the range of 1.5-2g per kg of body weight, as this can support muscle strength and growth.
- For individuals with lower activity levels or advanced SMA, the body's need for protein might differ. In some cases, excessive protein intake may not be beneficial and could even stress the kidneys or contribute to other metabolic concerns. Therefore, protein intake should be carefully monitored and adjusted, especially for those with kidney issues, metabolic disorders, or other complicating factors.

Consulting with a registered dietitian is crucial to determine the appropriate protein intake based on your specific needs, especially if you have underlying conditions like renal disease or metabolic concerns that require special dietary considerations.

### Practical advice and meal planning tips for spinal muscular atrophy patients

- Modify food textures: If you have difficulty chewing or swallowing (dysphagia), consider modifying food textures [1]. Choose softer foods, puree or mash fruits and vegetables, and thicken liquids as needed. Dysphagia can be caused by muscle weakness in the throat and mouth, as well as damage to the bulbar motor neurons that control chewing, speech, and swallowing [2].
- Adaptive utensils: If you have difficulty with fine motor skills, consider using adaptive utensils to make eating easier [12].
- **Meal timing:** Avoid lying down flat for at least 30 minutes after eating to reduce the risk of reflux [7].
- **Food safety:** Be mindful of food safety, especially if you have a weakened immune system [12]. Wash your hands thoroughly before meals, and ensure food is cooked properly.
- Avoid prolonged fasting: Do not go for extended periods without eating, as this can lead to low blood sugar, muscle breakdown, and other complications [9]. Aim for no more than 6 hours between meals for SMA Type I and 8-12 hours for SMA Types II/III [10].

#### Sample meal plans

Here are some sample meal ideas that incorporate the principles discussed above:

#### **Breakfast options**

#### • For normal chewing/swallowing:

- Scrambled eggs with avocado and soft whole-wheat toast
- Greek yogurt with soft berries and honey
- o Oatmeal with ground flaxseeds, banana, and almond butter
- Smoothie with Greek yogurt, banana, spinach, and protein powder

#### • For dysphagia or difficulty chewing:

- Pureed oatmeal with mashed banana and almond butter
- Greek yogurt blended with fruit puree and honey
- Smoothie with protein powder, nut butter, and soft fruits

#### **Mid-morning snack**

#### • For normal chewing/swallowing:

- Cottage cheese with soft peaches or mango
- Mashed avocado on soft whole-grain crackers
- Soft-boiled egg with mashed sweet potatoes

#### • For dysphagia:

- Pureed fruit smoothie with Greek yogurt
- Cottage cheese blended with soft fruit puree
- Soft mashed avocado mixed with mild yogurt

#### **Lunch options**

#### • For normal chewing/swallowing:

- Soft turkey and cheese sandwich on whole-grain bread
- Lentil soup with soft whole-wheat bread
- Quinoa salad with soft-cooked vegetables and shredded chicken

#### • For dysphagia:

- Pureed lentil soup with soft whole-grain bread (dipped for easier swallowing)
- Blended turkey and vegetable soup
- Mashed quinoa with soft vegetables and protein puree

#### Afternoon snack

#### • For normal chewing/swallowing:

- Hummus with soft pita bread
- Nut butter on soft whole-wheat crackers
- Mashed avocado with soft-cooked sweet potato

#### • For dysphagia:

- o Blended hummus with mashed soft pita
- Nut butter blended into a smoothie
- Soft avocado and yogurt blend

#### **Dinner options**

#### • For normal chewing/swallowing:

- Grilled salmon with mashed sweet potatoes and steamed broccoli
- Chicken stir-fry with soft-cooked vegetables and brown rice
- Soft turkey meatballs with whole-wheat pasta and pureed marinara sauce

#### • For dysphagia:

- Blended salmon with mashed potatoes and soft vegetables
- Pureed chicken and soft vegetable mix with rice porridge
- Blended turkey and marinara over mashed pasta

#### **Evening snack**

#### • For normal chewing/swallowing:

- Soft cheese with whole-wheat crackers
- A soft-boiled egg with smooth guacamole
- Chia pudding with mashed berries

#### • For dysphagia:

- Blended chia pudding with soft banana puree
- Mashed guacamole with mild soft cheese
- Smooth yogurt mixed with soft protein sources

#### **Recipes**

Here are a few simple and nutritious recipes suitable for individuals with SMA:

#### **Creamy Tomato Soup** [16]

- Mix 2 cans of cream of tomato soup, 1 can of water, 1 cup half and half, 1/2 cup milk, 1/4 cup sour cream, and 1/4 cup shredded cheese in a large saucepan.
- Stir over low heat until the cheese melts.
- Reduce heat to very low and heat for 20-30 minutes to blend flavors, stirring frequently. Do not boil.
- Puree if needed.

#### **Shepherd's Pie** [16]

- Heat oven to 375°F.
- Brown 1 pound ground meat in a large skillet and drain.
- Mix 2 cups mashed potatoes, 4 ounces cream cheese, ½ cup shredded cheddar cheese, and 1 clove garlic until well blended.
- Combine meat, 1 cup mixed vegetables, and 1 can of gravy; spoon into a 9-inch square baking dish.
- Cover with potato mixture and remaining cheddar cheese.
- Bake for 20 minutes or until heated through.

#### **Maple Oatmeal** [16]

- Pour 1 cup oatmeal into a microwaveable bowl.
- Stir in 1 cup milk. Mix well.
- Microwave uncovered for 1-1/2 to 2 minutes until thickened.
- Let stand until desired consistency.
- Add 1 tablespoon butter and 2 tablespoons maple syrup.

# Importance of working with a healthcare professional

It's crucial to work with a healthcare professional, such as a registered dietitian or nutritionist, to develop a personalized nutrition plan tailored to your specific needs [15]. They can assess your nutritional status, identify any deficiencies, and provide guidance on appropriate calorie intake, food choices, and meal planning [17]. Early assessment and intervention by a registered dietitian can help optimize your nutritional status and prevent complications associated with malnutrition or other dietary issues [5].

### Conclusion

Nutrition is fundamental to managing your health and well-being with SMA. This guide, compiled from various medical studies and reputable sources, provides a foundation for understanding your nutritional needs. By following the guidelines and practical advice presented here, you can make informed choices that support your overall health and help you live your best life. Remember that individual needs vary, and personalized guidance from a registered dietitian or nutritionist is essential to create a nutrition plan that addresses your specific requirements and helps you manage SMA symptoms effectively. With the right approach to nutrition, you can improve your overall health, enhance your quality of life, and navigate the challenges of SMA with greater confidence.

### References

- 1. Fatty Acids, Nutrition, and SMA: Can Changes in Diet Help? | mySMAteam <a href="https://www.mysmateam.com/resources/fatty-acids-nutrition-and-sma-can-changes-in-diet-help">https://www.mysmateam.com/resources/fatty-acids-nutrition-and-sma-can-changes-in-diet-help</a>
- 2. Difficulty Swallowing With Spinal Muscular Atrophy | mySMAteam, <a href="https://www.mysmateam.com/resources/difficulty-swallowing-with-spinal-muscular-atrophy">https://www.mysmateam.com/resources/difficulty-swallowing-with-spinal-muscular-atrophy</a>
- 3. Nutritional, Gastrointestinal and Endo-Metabolic Challenges in the Management of Children with Spinal Muscular Atrophy Type 1 PubMed Central https://pmc.ncbi.nlm.nih.gov/articles/PMC8308588/
- 4. Cure SMA Nutrition Basics <a href="https://www.curesma.org/wp-content/uploads/2020/02/02252020\_Nutrition">https://www.curesma.org/wp-content/uploads/2020/02/02252020\_Nutrition\_Basics\_vWeb.pdf</a>
- 5. NUTRITION BASICS Cure SMA Canada <a href="https://curesma.ca/wp-content/uploads/2017/04/fsmac\_nutrition2013-1.pdf">https://curesma.ca/wp-content/uploads/2017/04/fsmac\_nutrition2013-1.pdf</a>
- 6. Key to a Healthy SMA Diet is Variety <a href="https://hcp.smanewstoday.com/2019/02/19/key-to-a-healthy-sma-diet-is-variety/">https://hcp.smanewstoday.com/2019/02/19/key-to-a-healthy-sma-diet-is-variety/</a>
- 7. SMA Diet and Eating Tips | mySMAteam <a href="https://www.mysmateam.com/resources/sma-diet-and-eating-tips">https://www.mysmateam.com/resources/sma-diet-and-eating-tips</a>
- 8. MDA Virtual Learning: Nutrition with a Neuromuscular Disease YouTube, <a href="https://www.youtube.com/watch?v=Hqu0qFiP-oA">https://www.youtube.com/watch?v=Hqu0qFiP-oA</a>

9. NUTRITIONAL CARE GUIDELINES FOR CHILDREN WITH SMA OR NEUROMUSCULAR DISORDERS DURING ACUTE ILLNESS OR FASTING Proactive Nutrition - Columbia Neurology, https://www.neurology.columbia.edu/file/1458/download?token=fRjntiwb

10. Getting the Most Out of What You Eat! Nutrition for Oral Feeders - Cure SMA,

https://www.curesma.org/wp-content/uploads/2019/07/nutrition-2018-conference.pdf

- 11. Nutrition is critically important for maintaining muscle mass and strength and minimizing fatigue in SMA patients Columbia Neurology <a href="https://www.neurology.columbia.edu/file/1457/download?token=533WREX">https://www.neurology.columbia.edu/file/1457/download?token=533WREX</a> N
- 12. Common complications in spinal muscular atrophy (SMA) type 1 after nusinersen treatment The Turkish Journal of Pediatrics <a href="https://turkjpediatr.org/article/download/4527/3270/12688">https://turkjpediatr.org/article/download/4527/3270/12688</a>
- 13. Nutrition Cure SMA https://www.curesma.org/nutrition/
- 14. FYI: High Calorie And Easy to Chew Recipes | The ALS Association <a href="https://www.als.org/navigating-als/resources/fyi-high-calorie-and-easy-chew-recipes">https://www.als.org/navigating-als/resources/fyi-high-calorie-and-easy-chew-recipes</a>
- 15. Nutritional Practices at a Glance: Spinal Muscular Atrophy Type I Nutrition Survey Findings https://pmc.ncbi.nlm.nih.gov/articles/PMC4334580/