CURE SMA

CARE SERIES BOOKLET

A SOURCE OF INFORMATION AND SUPPORT FOR INDIVIDUALS LIVING WITH SPINAL MUSCULAR ATROPHY AND THEIR FAMILIES.

NUTRITION BASICS

cure SMA

Make today a breakthrough.

WHY NUTRITION IS IMPORTANT

Nutrition from food provides energy the body needs to breathe and move. It also provides fuel for life-promoting health and growth. As an individual with spinal muscular atrophy (SMA), or the caregiver of an individual with SMA, it's important to keep nutrition at the top of your mind.

Proper nutrition provides:

- Improved Growth: Gaining weight while growing in height is essential for good health. Having enough energy (calories) helps keep the body growing by supporting lung tissue and the heart muscle.
- Better Breathing: Growth in length helps with breathing, as it gives more room for the chest to expand. This is important because breathing problems are the leading cause of illness for individuals with SMA.
- Preventing Illness: Adequate nutrition helps to prevent/fight colds and viruses that could turn into a potentially life-threatening lower respiratory infection in the lungs (pneumonia).
- Improved Motor Function: Too much weight or too little weight can cause individuals with SMA to decline in both strength and ability to move.
- Enhanced Quality of Life: Poor nutrition can cause physical symptoms to become worse.
Good nutrition can translate into feeling your best.

BUILDING BLOCKS OF GOOD NUTRITION

Everything that happens in the body requires energy. Calories are the measure of energy in foods. The three basic sources of calories are carbohydrates, proteins, and fats. All play an important role in a balanced diet.

Carbohydrates are sugars, starches, and fibers. The digestive system breaks down carbohydrates to create glucose (blood sugar). Glucose is the most efficient source of energy for the body.

Proteins are made up of amino acids that help the body grow and repair damaged tissues.

Fats are essential for growth, healthy skin, and protecting the organs. They are especially important for brain and eye development in infants and small children. Fats also help the body absorb vitamins A, D, E, and K.

The Calorie Needs for Individuals with SMA

Every individual's calorie needs are different. Factors that affect calorie needs include ability to move arms and legs, ability to breathe and eat and the amount of muscle mass. For example, with more movement and activity, more energy will be used and more calories will be needed.

Infants and children with SMA need enough calories to gain weight and grow in length. Adults and older teenagers (who are no longer growing) need enough calories to maintain a healthy weight.



Vitamins, Minerals, and Fluid

Vitamins and minerals are essential for normal growth and development, even though they don't provide calories.

There are two basic types of vitamins:

Fat-soluble vitamins (A, D, E, and K) are stored in the body's fatty tissues.

Water-soluble vitamins (all the rest) are used right away and the excess is not stored in the body.

Some important minerals include:

Sodium, potassium, magnesium, and chloride are electrolytes that balance the amount of water and acid or pH level in the body. They help ensure that the nerves, muscles, heart, and brain work properly.

Calcium
Important for bone strength and health.

Iron
Important for oxygen transport throughout the body.

Zinc, selenium
Help the body with wound healing, skin health, and growth.

Water is the most important substance in the body. It is needed for every function. Getting enough fluids is important for respiratory function and preventing constipation. Many individuals with SMA avoid drinking enough fluids because of dependence on others for using the bathroom. Take note of how much is being consumed each day.

General fluid recommendations:

Adults – about ½ oz. per pound of body weight

Children – about 1 ½ oz. per pound of body weight

Signs of dehydration or that more fluids may be needed include:

- Urine is dark yellow or has a foul odor.
- Thirst.
- Dry lips.
- Thick respiratory secretions or mucous plugs.
- Stool is hard and dry.

During illness, fluid needs may increase. Consult your medical team.

NUTRITION FOR INDIVIDUALS WITH SMA

Who Specializes in SMA Nutrition?

It is important to work with a registered dietitian who is familiar with, or ready to learn about the nutritional needs of individuals with SMA.

Is There a Right Kind of Diet for Individuals with SMA?

There are many opinions about what makes a healthy diet for children and adults, and just as many opinions about the best diet for individuals with SMA.

There are essentially no scientific studies that specify which foods or nutritional supplements are best for optimal health in SMA. However, a registered dietitian with experience in SMA will help determine the proper balance of proteins, fats and carbohydrates, taking into account individual needs and preferences.

Specific foods that may increase reflux:

- High fat foods
- Spicy foods
- Acidic foods (citrus fruit, tomato, vinegar)
- Caffeinated drinks
- Chocolate
- Peppermint
- Alcoholic drinks

NUTRITION RELATED PROBLEMS ASSOCIATED WITH SMA

Type of Issue	Presentation	Challenges
Feeding Difficulty	Depending on severity of SMA, difficulty chewing and/or swallowing, choking.	Mouth opening/bite, tongue control, head control and positioning, muscle weakness, chewing muscle fatigue.
Aspiration	Small amounts of food or liquid enter the windpipe instead of the esophagus (tube to the stomach). Can also occur due to reflux of stomach contents back into the esophagus and mouth and then into the lungs.	Can cause difficulty breathing and pneumonia (a sudden event that may be life-threatening).
Gastrointestinal (GI) Problems/Discomfort	Diarrhea, bloating, spitting up, vomiting after meals. Bad breath, regurgitation of foods, abdominal distention.	May contribute to undernutrition.
Under Nutrition	Difficulty growing and gaining weight.	Increased risk of infection. Difficulty with wound healing. Tendency for pressure sores. Associated with decline in motor function. Decrease the effectiveness of new treatments.
Obesity/Over Nutrition	Excess weight gain and lower calorie needs.	Increased burden of care and decreased quality of life. Increased pain and associated complications with hips and back. Increased risk of diabetes and hypertension. Can be associated with loss of motor function (e.g., loss of walking).
GERD (Gastroesophageal Reflux Disease)	Reflux causes stomach contents to move back up the esophagus. More common with more severe SMA symptoms.	Increased risk of aspiration (saliva, drink, or food going into the lungs). Harm to the lining of the esophagus. Increased risk of reflux with overweight/obesity and specific foods.
Constipation	Infrequent and/or hard-to-pass bowel movements. Aggravated by inadequate intake of whole grains, fruits, and vegetables and/or inadequate fluid intake.	Abnormal gastrointestinal motility (ability to move bowels). Low abdominal wall muscle tone. Increased respiratory efforts and distress.
Low or High Blood Sugar	Prolonged fasting in some individuals who have severe SMA symptoms, especially those who are undernourished or ill.	Blood sugar is an immediate source of energy. If low, body turns to alternate sources of energy from muscle and fat breakdown.

METHODS AND MEASUREMENTS ASSESSING SMA NUTRITION

What Do I Need to Know About Nutritional Assessment?

Nutritional assessment of individuals with SMA is built on the same basic principles as the assessment of individuals without SMA.

Infants and children with SMA are expected to grow over time. Infants will have their weight, length and head circumference measured at every clinic visit. Children will have their weight and length measured at every clinic visit. Adults may have only their weight measured after achieving full adult height. Infants and children will have their growth compared to growth charts from the Center for Disease Control and Prevention or the World Health Organization (up to 2 years of age). Individuals who are not able to stand will have their height estimated with a length board, by arm span, or segmentally with a tape measure or other tool.

It is important to compare weight to height when assessing nutritional status. This is called weight-for-length in infants and body mass index (BMI) in older children and adults. Since individuals with SMA tend to have smaller muscles, it can be difficult to compare their growth to standard weight-for-length and BMI charts.

For example, an individual with SMA who cannot walk and is very weak may actually be overweight or obese at the 50th percentile for BMI, while a BMI at the 3rd percentile may be appropriate.

The healthcare team will track growth trends over time using the standard charts, keeping in mind that it may be best for individuals with SMA to grow with their weight-for-length or BMI at the lower end of the normal range.

Remember, every family's growth history and every individual with SMA is unique and should not be expected to gain weight or grow taller at the exact same rate as any other individual. Change to nutritional plans may be needed if there is sudden weight gain, unintentional weight loss, or poor growth.

Be sure to bring it to your physician's or dietitian's attention if you notice:

- Rapid changes in weight. For example, clothing is suddenly too tight or too loose.
- Changes in appearance that concern you.
- Height or weight that falls off a growth curve.
- Sudden weight changes.

Your physician or dietitian will complete a physical exam, and review your weight history and length/height history, to determine if you're at a healthy weight.

What Other Tests Might Be Recommended?

Your physician may suggest alternate methods to measure body composition due to the differences in fat and muscle mass with SMA. Bloodwork may be recommended to assess nutritional status. Swallow tests and tests for GI motility may be recommended.

OTHER MEASURES TO ASSESS BODY COMPOSITION

Test	Measures	Important Considerations
Skinfold Thickness	Estimates fat stored at the triceps (upper arm) or other areas. These specified areas may be sensitive to changes in nutritional status.	Should be measured along with Mid Upper Arm Circumference and tracked over time. The dietitian should be a trained skinfold examiner.
Mid Upper Arm Circumference	Measures around the arm at the mid-point of the upper arm (between shoulder and elbow).	Should be done along with triceps skinfold and/or tracked over time.
DEXA Scan (Dual Energy X-ray Absorptiometry)	Assess body composition. Uses low-level radiation in order to distinguish between fat, fat-free mass and bone. Benefits include, increased precision of body composition measurements.	Can scan the whole body or just one particular region.

OTHER TESTS

Bloodwork	Assess nutritional status.	Tests: 25-OH vitamin D, iron status; other nutrition labs as needed.
Swallow Study	Uses an X-ray video to watch swallowing mechanisms and diagnose any difficulties.	Can assess ability to swallow different consistencies and textures.
Upper Gastrointestinal (UGI) Xray Series	Assess reflux. An X-ray of the mouth, throat, esophagus, stomach and the small intestine.	A negative result does not rule out reflux.

MANAGING NUTRITION IN SMA

KNOW THE SIGNS OF ASPIRATION

Aspiration occurs when small amounts of food or liquids enter the trachea (windpipe) instead of the esophagus (tube to the stomach). Some individuals with SMA only aspirate when they are ill.

A few things to watch for include:

- **Coughing/choking:** This could signal reflux of stomach contents back up into the esophagus and then into the lungs.
- **Vomiting:** A gastrointestinal illness can make an individual more likely to aspirate.
- **Cold/respiratory sickness:** Chewing and swallowing can be especially difficult when ill.

When is a Feeding Tube Necessary?

Your physician or dietitian may recommend tube feeding if you are:

- Unable to swallow safely.
- Unable to meet all nutrient needs by mouth.
- Not growing well (child) or cannot maintain weight (adult).
- Unable to drink enough fluid.
- Ill and not able to eat or drink temporarily.
- Before or after surgery.

How Does a Feeding Tube Work?

Tube feeding (also called enteral feeding) provides nutrition directly through a tube leading into the stomach. The kind of tube feeding depends on individual needs. Your doctor will help to decide whether the need is temporary or longer-term, and whether to insert a tube through the nose or through an incision in the abdomen.

Your physician will decide how to insert the tube by considering:

- The length of time the tube feeding might be needed.
- How well your gastrointestinal tract is functioning.
- What would be most comfortable.
- Whether a risk of aspiration from feedings or secretions exists.

Feeding tubes can be used for:

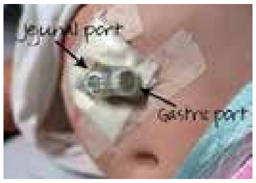
- Nighttime tube feeds with oral feeds during the day.
- Bolus feeds after meals to top off calories.
- Bolus for snacks between meals.

For individuals with swallowing/aspiration risks, feeding tubes can:

- Provide all nutrition and fluid needs.
- Run continuously during the day.
- Be pumped over shorter periods, depending on risk.
- Provide bolus feeds, depending on risk, for hydration/fluids only, medications and fluids, as well as during illness/fatigue or on days when not eating well.

How Do You Know What Feeding Tube Option to Use?

*Please note: *Both NG and NJ tubes can be placed easily and replaced as necessary. They can be a little uncomfortable, so they are not good solutions for long-term use.*

	Type of Tube	How It's Used	Considerations
	**Nasogastric (NG), Nasoduodenal (ND) or Nasojejunal (NJ) Tube**	A small flexible tube is inserted through the nose down to the stomach or the small intestine. These tubes can be easily placed and removed, if necessary.	• Temporary solution for swallowing and feeding problems. • Can be placed by a physician or RN without surgery. • Correct placement is verified by X-ray. • In some cases, families can be taught how to place these tubes at home. • Complications include sinusitis, sore throat, tube blockage, or dislodgement.
	**Percutaneous Gastrostomy (PEG) or Gastrostomy (G) Tube**	A small tube is placed surgically, through the skin directly into the stomach from the outside. A PEG tube has long tubing on the outside and a G-tube has a low profile "button."	• Long term solutions for swallowing and feeding problems. • Larger tube diameter can tolerate a variety of formulas and feeding plans. • PEG tubes are more commonly placed in older teenagers and adults. • After being placed, G-tubes and PEG's are essentially the same. • Complications include: infection around the site, excess drainage, and the tube becoming dislodged.
	**Gastrostomy-Jejunostomy (G-J) Tube**	A small tube is placed surgically, directly into the stomach and has two ports – one that goes into the stomach and one that goes into the small intestine (jejunum). J-tube feedings may be recommended to help prevent aspiration from gastroesophageal reflux.	• Long term solution for swallowing and feeding problems. • Formula must be given at a slower rate through the J-port and requires a longer time on the feeding pump. • Sometimes requires a more specialized formula. • Requires extra appointments because the J-tube needs to be replaced every 3 months in intervention radiology (IR). • In some cases, you may still be able to give medications, vitamins/minerals, and small amounts of water through the G-port. • Complications include: infection around the site, excess drainage and the tube becoming dislodged.

How Are Tube Feedings Given?

After an NG, NJ or G-tube (see chart for details) is placed, a registered dietitian will help make a plan for the feedings and a physician will write a prescription for formula and tube feeding supplies if needed. The first question that needs to be considered is how the feeding will be given.

The most common tube feeding methods are bolus feeding or continuous feeding.

Bolus feedings are administered through a syringe, gravity bag, or a pump into the feeding tube over a short time, typically 5 to 20 minutes, and can be given over a longer time. Bolus feedings are only given to individuals who can move food successfully through the digestive system.

Typically, bolus feedings are given through a G-tube but not a J-tube. The larger diameter of the G-tube allows larger volumes of formula to pass into the stomach at a faster rate. The stomach then releases small amounts to the small intestine over time. Your physician or dietitian may recommend several bolus feedings per day to meet nutritional needs. Complications of bolus feeding may include nausea, vomiting, diarrhea, abdominal distention, or aspiration.

Continuous feedings are provided through a feeding pump over longer periods of time. The amount of time spent feeding may range from 12 to 24 hours per day. Feedings to the small intestine or jejunum are given continuously because the small intestine tolerates small amounts of formula at a time.

Although continuous feedings may make an individual less mobile, they may be appropriate if they:

- Have a gastrointestinal problem that keeps food from passing through the system easily.
- Need a slower feeding because he/she cannot tolerate larger volumes of formula at once.
- Are at high risk for aspiration.
- Are fed by tube in the jejunum.

Complications of continuous feeding may include nausea, vomiting, diarrhea, or abdominal distention.

Some nutrition plans may include both bolus and continuous feedings.

What Type of Tube Feeding Formula is Best?

There are three types of tube feeding formulas that vary based on the type of protein used. They are called intact or whole protein, semi-elemental (peptides), or elemental formulas (amino acids). You'll be able to recognize them by their brand names:

	Infant	Pediatric	Adult
Whole Protein/ Real Food	• Breast Milk	• Compleat Pediatric Organic Blends • Pediasure Harvest • Nourish	• Compleat Organic Blends • Liquid Hope • Real Food Blends (not complete)
Whole Protein/ Standard	• Enfamil NeuroPro • Similac ProAdvance • Nestle Gerber Good Start	• Compleat Pediatric • Pediasure • Boost Kids Essentials • Kate Farms Pediatric	• Ensure • Boost • Kate Farms
Peptides	• Nutramigen • Alimentum	• Pediasure Peptide • Peptamen Junior • Kate Farms Pediatric Peptide	• Peptamen • Kate Farms Peptide
Amino Acids	• Elecare Infant • Neocate Infant • PurAmino	• Elecare Junior • Neocate Splash • Vivonex Pediatric • Tolerex	• Vivonex • Tolerex

You will want to consult with a trained dietitian to determine what kind of formula is best for the individual. Some factors to consider include:

- Food allergies.
- Bowel function.
- Reflux.
- Type and size of feeding tube.
- Overall balance of nutrients.

There are many ways to use a feeding tube, and eating may still be possible. Placing a G-tube (or PEG) doesn't commit you to using it all the time. Placement of a G-tube allows you to have it for times of need, or to supplement feedings. It does not mean eating is not possible (if it is safe to do so). It is important to continuously consult your healthcare team to ensure adequate nutrition and safety.

COMMON NUTRITION QUESTIONS

How Can I Help with Acid Reflux?

Acid reflux, caused by GERD (Gastroesophageal Reflux Disease), is a common problem in individuals with SMA. The more severe the SMA, the more common GERD is. Acid reflux can cause uncomfortable heartburn and lead to more serious problems such as, aspiration of the stomach contents into the windpipe (where the food travels all the way back up to the throat).

To help with reflux:

- **Eat small meals –** six or more small meals throughout the day.
- **Limit greasy, high fat foods –** Foods high in fat leave the stomach more slowly.
- **Avoid irritating foods –** Foods that are more likely to cause reflux include chocolate, peppermint, spicy foods, and acidic foods (soft drinks, citrus, tomatoes, etc.).
- **Incline after meals –** For infants or individuals unable to sit up, try propping up his or her head/shoulders during/after feeding.
- **Consider medication –** If the above measures aren't working, talk to your physician or GI specialist. They may recommend:
    - Medication that increases the speed at which food moves through the digestive system.
    - Acid blocking medications to prevent damage to the esophagus and aversion to food.

What is a Nissen Fundoplication?

Sometimes doctors recommend a special operation for serious acid reflux. It is more likely to be needed by individuals with more severe SMA.

A Nissen fundoplication makes it more difficult for stomach acid to travel backwards from the stomach to the esophagus.

This procedure can be performed at the same time as a G-tube procedure. Newer laparoscopic techniques (often requiring only a small incision in the stomach) allow the two procedures to be done at once, reducing recovery time. Since a Nissen fundoplication has a higher risk of complications, it is important to make the decision carefully.

A pH probe study can show how often an individual has acid reflux. The procedure involves placing a small tube (similar to an NG tube, but smaller) down into the esophagus. The tube is used to monitor the degree to which acid flows back into the esophagus for up to a 24-hour period. The study involves drinking apple juice every four hours. At the end, your physician will have a better idea of the degree of reflux when the individual is in different positions and circumstances.

How Does SMA Make an Individual Prone to Constipation?

Individuals with SMA have weaker muscles in the abdomen, and therefore constipation is more likely. This can cause the body to struggle to move food through the digestive system. Individuals with SMA are more likely to have less fiber and fluids in their diet. To prevent constipation, individuals with SMA should consume foods naturally high in fiber and drink plenty of fluids.

Constipation can lead to abdominal discomfort. It can also cause an individual to feel bloated, spit up, have bad breath, or vomit after meals. Having to work harder to push bowel movements out can cause breathing problems in some individuals with severe SMA. For some, constipation gets better when fiber and/or fluid is increased. Some types of constipation may require medications, please consult your medical team.

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How Much Fiber is Enough?

1. For children: Add 5 grams to child's age.

   Example: A 3-year-old needs 8 grams of fiber a day.
2. For adults: 20-30 grams per day depending on overall calorie needs.
3. Choose the right foods - High fiber foods include whole grains (such as oats, corn, whole wheat, quinoa) and fruits and vegetables (carrots, apples, celery, oranges, dried fruits, etc.). Try soft cooked or canned versions of these foods or mix them into smoothies. Raw fruits and vegetables can be hard to chew or swallow.

Should I Be Concerned About Weight?

Obesity (being very overweight) is a serious health concern. It can make physical activity more challenging, breathing or being lifted more difficult, and take away from self-image.

These alone are good reasons for staying in a normal weight range. However, when an individual has SMA, being obese can increase the risk of some very serious health conditions:

- Diabetes and hypertension.
- Greater pain.
- Loss of mobility.

NUTRITION DURING ILLNESS OR SURGERY

Because of limited muscle mass (total weight of muscle in their body), many individuals with SMA find it difficult to get better after an illness. Since many illnesses bring fever and a lack of appetite, the body may not get the nutrients it needs to function properly. When this happens, the body begins to break down muscle to perform normal body functions.

This may be one of the reasons why individuals with SMA lose so much strength during an illness and why they take longer to recover after illness or a surgery. Their low muscle mass is being called on to do even more for the body. If the body is unable to get enough energy from muscle, the individual may become seriously ill with hypoglycemia (low blood sugar). If the individual is hospitalized and unable to be fed by mouth or feeding tube, your doctor may recommend use of IV or "parenteral nutrition," also known as PPN or TPN. This IV solution includes sugars, amino acids, vitamins and minerals. Consult your healthcare team.

Conclusion and Resources

Nutrition is a complex topic for anyone, but especially for individuals with SMA. Due to the unique challenge SMA presents, it is important to assess growth and nutritional status, and then choose interventions that best meet the individual's nutritional challenges.

CURE SMA

Cure SMA is a non-profit organization and the largest worldwide network of families, clinicians, and research scientists working together to advance SMA research, support affected individuals/caregivers, and educate the public and professional communities about SMA.

Cure SMA is a resource for unbiased support. We are here to help all individuals living with SMA and their loved ones, and do not advocate any specific choices or decisions. Individuals and caregivers make different choices regarding what is best for their situation, consistent with their personal beliefs. Parents and other important family members should be able to discuss their feelings about these topics, and to ask questions of their SMA care team. Such decisions should not be made lightly, and all options should be considered and weighed carefully. All choices related to SMA are highly personal and should reflect personal values, as well as what is best for each individual and their caregivers.

Remember that your healthcare team and Cure SMA are here to support you. To continue learning, please see other available Care Series booklets:

- Breathing Basics
- Caring Choices
- Genetics of SMA
- Musculoskeletal System
- Understanding SMA

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