



# Nutrition Basics

Fostering Health and Growth for Spinal Muscular Atrophy

SMA CARE SERIES



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breakthrough.

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# Introduction

We need energy to survive and to thrive. Food provides the energy our body needs to breathe in life-giving oxygen, to pump blood throughout our body, and to carry out vital processes in the body. Nutrition provides fuel for life—promoting health and growth!

As the parent of a child with SMA, it's important to keep this in mind. In whatever form you feed your child, whether by mouth, by feeding tube, or through continuous feed, you are nurturing and caring for them...providing them the very best chance at the best health they can attain.

But food also provides one of the most important connections that a family makes. We call it mealtime!

## THE FAMILY THAT EATS TOGETHER...

Eating together as a family is a symbol of love, connection and communication. Mealtimes provide quality time for the family. Family mealtimes provide time for parents to listen and to talk with their children, while children gain insight into their parents' lives. Together, parents and children build trust and enjoy discussions that reflect family values and build a sense of heritage.

For families with a child with SMA, this is just as important. Whether eating by mouth or being tube fed, that child can be part of the family mealtime. With thoughtful preparation, family time can be special for all.

## LEARNING TOGETHER...

As you go through this booklet, you'll see that providing for your child's nutrition isn't always simple. SMA may present many nutritional challenges. But by educating yourself, talking to other parents, and getting help from a registered dietitian you can stay ahead of the learning curve.



**Having a family mealtime can help your child develop intellectually, physically and emotionally!**

- **Intellectually—**  
Watching adults interact with each other.
- **Physically—**  
Learning about healthy foods.
- **Emotionally—**  
Building confidence and self-esteem as they share in family time.

Source: West Virginia University Extension Service

# Nutrition 101 – Mastering the Basics

Nourishing your child begins with an understanding of how feeding provides what the body needs...to grow and thrive.

## WHAT ARE THE BASICS OF GOOD NUTRITION?

Everything that happens in the body requires energy from calories—whether pumping blood, breathing or moving. Calories are the measure of potential energy in a food. The three basic sources the body uses to provide calories are carbohydrates, proteins and fats.

**Carbohydrates** are sugars, starches and fiber. The simplest carbohydrate is glucose, also known as blood sugar. The digestive system breaks down complex carbohydrates (starches) to create blood sugar. Glucose then flows through the blood to every cell in the body.

**Proteins** are made up of amino acids and other compounds. They help the body grow and repair damaged tissues. If they are not used immediately, proteins can be stored as fats in the body. These fats help provide backup energy if someone is sick or unable to eat.

**Fats** are made up of fatty acids and serve as back-up energy storage for the body. Fats help the body absorb vitamins A, D, E and K.

## WHAT ELSE DOES MY CHILD NEED?

By choosing foods high in vitamins and minerals, you provide other building blocks for your child's body. If your child does not get enough of a particular vitamin or mineral, health problems can result.

**Vitamins** are essential for normal growth and development. These include Vitamin A, C, D, E and K as well as all the B vitamins.<sup>1</sup> There are two basic types of vitamins:

- **Fat-soluble vitamins** are stored in the body's fatty tissues.
- **Water-soluble vitamins** are used right away. Any extra moves straight through the body. B12 is the only water-soluble vitamin the body is able to store.

**Minerals** are also found in foods and are essential for health. Some of these include water, sodium, potassium, chloride, calcium, magnesium, iron, copper, and zinc. Health problems can result if the body does not receive enough minerals.

### Why does sugar make my child cranky?

Simple carbohydrates (like candy and cola) enter the blood quickly. The body reacts by releasing insulin to make sure the blood sugar doesn't go too high. Not too long afterwards, the blood sugar may drop too low. This rollercoaster ride of blood sugar levels may cause your child to feel nervous and cranky.

You can prevent this through a balanced diet full of complex carbohydrates (like oats, corn, wheat and rice). Complex carbohydrates don't have this effect since they take longer to digest. They also provide more energy and more nutritional value!<sup>2</sup>

<sup>1</sup> The B vitamins include (thiamine, riboflavin, niacin, pantothenic acid, biotin, B6, B12, and folic acid.)

<sup>2</sup> Adapted from *The Yale Guide to Children's Nutrition*, quoted on [www.howthingswork.com/innovation/edible-innovations/food2.htm](http://www.howthingswork.com/innovation/edible-innovations/food2.htm).

# Building Blocks – Understanding Nutrition for SMA Kids

## WHY IS NUTRITION SO IMPORTANT?

As we've already seen, good nutrition is essential to health and growth! But proper nutrition is complex, especially for children with SMA. Since we are still waiting for clinical studies, there is much we still need to learn. Parents and dietitians have found that children with SMA receive many benefits from good nutrition including:

- **Improved Growth** — Gaining weight while growing in height is essential to good health. Having enough energy (calories) and protein helps keep the body growing by supporting lung tissue and the heart muscle.
- **Better Breathing** — Growth in length helps with breathing, providing more room for the chest to expand. This is important because breathing problems are the leading cause of illness for children with SMA.
- **Preventing Illness** — Adequate nutrition helps to prevent/fight colds and viruses that could turn into a potentially threatening lower respiratory infection in the lungs (pneumonia).<sup>3</sup>
- **Improved Motor Function** — Too much weight or too little weight can cause children with SMA to decline in both strength and ability to move.
- **Better Quality of Life** — Poor nutrition can cause physical symptoms to become worse. Good nutrition can translate into better living!



## WHO SPECIALIZES IN SMA NUTRITION?

With nutrition such an important part of care for your child, it's important to work with a registered dietitian who has experience working with patients with SMA.

### Getting help.

It is important to work with a registered dietitian who is familiar with the nutritional needs of children with SMA. The ultimate goal is to help children grow and thrive who have SMA type I or II, and to prevent overnutrition and/or obesity for children and adults with SMA type II who are stronger and SMA type III.

<sup>3</sup> For more information, see "Breathing Basics: Respiratory Care for Children with Spinal Muscular Atrophy", part of the SMA Care Series.

## IS THERE A *RIGHT* KIND OF DIET FOR CHILDREN 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 children with SMA. The body has many ways of converting foods of all kinds into energy to supply our needs. The body then stores excess food energy in the liver, muscle and body fat for future use.

We can live healthy lives on all kinds of diets—from a high fat, low carbohydrate diet<sup>4</sup>, to a very low fat, high protein diet, to a vegetarian or vegan diet. For scientists to prove the “best” foods, they would need volunteers willing to eat foods they are not accustomed to for an extended period of time. When you add in the individual nature of health and nutrition, this proves very difficult.

This is even more challenging when it comes to children with SMA. There are essentially no good scientific studies that specify which foods or nutritional supplements might be better. That being said, a nutritionist can help you determine the proper balance of proteins, fats and carbohydrates, recommend foods that your child might better tolerate, and help to make sure your child is taking in enough vitamins and minerals.

## WHAT NUTRITION RELATED PROBLEMS ARE ASSOCIATED WITH SMA?

It is important to take a close look at your child’s nutrition, because there are many nutrition-related challenges facing children and adults with SMA. The chart, *Nutrition Related Problems Associated with SMA*, explains these issues in more detail, as well as related challenges.



<sup>4</sup> Or “ketogenic” diet.

## Nutrition Related Problems Associated with SMA

Type of issue	Presentation	Challenges
<b>Feeding issues</b>	Depending on the type and severity of SMA, feeding challenges include difficulty chewing and/or swallowing and choking issues.	Mouth opening/bite, tongue control, head control and positioning, facial muscle weakness, range/fatigue in chewing muscles.
<b>Aspiration</b>	Small amounts of food or liquid enter the trachea (windpipe) instead of the esophagus (tube to the stomach).	Can also occur due to stomach contents flowing back into the esophagus and then into the lungs. Can cause pneumonia and difficulty breathing.
<b>Abdominal problems/discomfort</b>	Diarrhea, bloating, spitting up, vomiting after meals, bad breath, regurgitation of feeds, and/or abdominal distention.	May contribute to undernutrition.
<b>Undernutrition</b>	Difficulty growing and gaining weight is common in children with SMA type I or possibly in those with type II.	Increased risk of infection. Difficulty with wound healing. Tendency for pressure sores.
<b>Obesity/overnutrition</b>	Commonly seen in those with milder SMA type II and type III. Could increase the burden of care and/or decrease the quality of life.	Increased pain & associated complications with hips and back. Increased risk of diabetes and hypertension.
<b>GERD</b> Gastroesophageal Reflux Disease	Most common in individuals with type I, followed by type II and less common with type III.	Causes increased danger of aspiration and pain.
<b>Constipation</b>	Infrequent bowel movements. Tied to reduced intake of fiber and/or inadequate fluid intake.	Abnormal gastrointestinal motility (ability to move bowels).
<b>Yeast overgrowth</b>	Results when the body's bacterial flora is out of balance. Presentations include: thrush; orange tinge in eyebrows, hair and around g-tube; cheesy smell on hands and feet.	Can contribute to sweating, bloating and constipation.
<b>Low or high blood sugar</b>	Present in some children who have SMA type I, with prolonged fasting.	Blood sugar is an immediate source of energy. If low, the body breaks down muscles to get energy.

*For suggestions on how to manage these health problems, see pages 19-24 in this booklet, **Obstacles to Nutrition—Facing Feeding Challenges.***

# Methods and Measurements – Assessing SMA Nutrition

## WHAT DO I NEED TO KNOW ABOUT NUTRITIONAL ASSESSMENT?

On every trip to the doctor, your child is measured and weighed. This enables your physician to compare your child's progress to growth charts from the Center for Disease Control and Prevention (CDC). These charts include weight for age, length for age, weight for length, body mass index (BMI) for age, and head circumference for age.

The nutritional assessment of children with SMA is built on the same basic principles as the assessment of children without SMA. So, let's start by discussing normal growth.

### Growth in infancy.

A normal infant experiences rapid growth from birth to six months. Although it is normal for an infant to lose six percent of his/her body weight during the first seven days of life, a full term, normal infant will usually:

- Double their body weight by 4 to 5 months; triple by 12 months of age.
- Increase their body length by nearly 50% by 12 months of age.
- Increase head circumference by 35% by 12 months of age.
- Measure between the 5th and 95th percentiles for weight, length and circumference.



### Anthropometric:

The study of human body measurements especially on a comparative basis.





<b>Measurements for Assessing Infant Nutrition (up to 3 years of age)</b> <i>For differences in SMA children see pages 10-12</i>	
<b>Weight for age</b>	Weighed without diaper or clothing. Measured via a calibrated digital or beam balance to the nearest 0.1 kilogram of body weight. Weight is tracked over time to ensure proper growth. Weight less than the 5th percentile may indicate undernutrition. Weight greater than the 95th percentile may indicate obesity.
<b>Length for age (linear growth)</b>	Measured using an inflexible length board with fixed headboard and moveable footboard. The head is held still and the knees are flattened to fully extend the legs. Length less than the 5th percentile may mean undernutrition or greater than the 95th percentile may indicate obesity.
<b>Weight for length</b>	Charted on growth charts by using both weight and length. Ideal weight for length for a healthy child is in the 50th percentile. Less may indicate undernutrition.
<b>Head circumference (brain growth)</b>	Measured with a flexible, plastic coated or disposable tape measure. The tape measure is placed above the eye sockets and around the back of the head in a full circle. Less than the 5th percentile may point to undernutrition or greater than the 95th percentile may indicate catch-up growth or medical problems.

**Growth Charts**

From birth to 36 months, growth charts are helpful to assess:

- Weight for age and rate of growth
- Length for age and rate of growth
- Weight for length
- Head circumference for age





## FROM TODDLER TO YOUNG ADULT

As children age, the measures and ways physicians take these measures changes slightly. To determine whether a child is growing properly, their physician will commonly look at their weight for age, height for age, and body mass index.

Measurements for Assessing Older Children (age 2 to 20)	
<b>Weight for age</b>	<p>Weighed in light clothing or stocking feet.</p> <p>Track progress over time to monitor speed of growth.</p> <p>Weight less than the 5th percentile may indicate undernutrition or greater than the 95th percentile may indicate obesity.</p>
<b>Height for age</b>	<p>Measured by standing as soon as the child can stand unassisted.</p> <p>Height less than the 5th percentile may indicate undernutrition or greater than the 95th percentile may indicate obesity.</p>
<b>BMI for age (Body mass index)</b>	<p>Determines if weight is appropriate for height.</p> <p>Calculated by <math>(\text{weight in kg})/(\text{height in meters squared})</math>.</p> <p>Being in the 85-95th percent may indicate risk of overweight; greater than the 95th percentile may indicate risk of obesity.</p>

### How is nutritional assessment different for a child with SMA?

Now that we've looked at how physicians measure normal children, let's take a look at the differences for a child with SMA.



## MEASURING WEIGHT

Since children with SMA tend to have less muscle, it can be difficult to judge the "right" amount of weight. Although nutritionists commonly use muscle weight (BMI) to identify undernutrition in children without SMA, they need different measures for the child with SMA.

Due to having less muscle, children with SMA have a different proportion of weight to height. Although these will vary from child to child, here are some general guidelines:

- **Average:** Somewhere around the 5th to 10th percentile of weight for height (for a child with SMA type II).
- **Overweight:** Children who are above the 50th to 75th percentile of weight for height.
- **Underweight:** More difficult to measure. The physician can best determine this by examining the child.

Infants with SMA will usually fall fairly quickly below the normal growth curve for weight, within the first few weeks or months. Some children with SMA type III may follow the normal curve into early childhood.

## USING WEIGHT FOR LENGTH

Weight can also be tracked on a **weight for length growth chart**. This chart determines the growth percentile by considering both your infant's weight and length. Ideal weight for length for a healthy child is the 50th percentile. In children without SMA, weight for length of less than the 50th percentile may indicate that they are not getting enough nourishment.

**In children with SMA**, a weight for length between the 3rd and 25th percentile is considered normal as long as the length is between the 3rd and the 97th percentile.

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**In children with SMA**, a weight for length between the 3rd and 25th percentile is considered normal as long as the length is between the 3rd and the 97th percentile.

## MEASURING HEIGHT

Since many children with SMA cannot stand to have their height measured, and since they may have scoliosis and contractures, caregivers may use other methods. These include:

- **Stadiometer** — Children who cannot stand on their own can be measured with this moveable headboard attached to a measuring board. The child lies down to have this measurement.
- **Arm spans** — Using a flexible tape measure with a child's arms extended may be a good estimation of height. A child with SMA will need someone to help extend his or her arms for this measure. BMI is not accurate (and not recommended) for children with SMA because of less muscle and more fat mass.

### Scoliosis:

A side-to-side curving of the spine.

### Contracture:

Distorted muscle or connective tissues due to a spasm, scar or paralysis.

### Beyond the charts

Although growth charts are helpful, each child is different.

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

- Notice big jumps. For instance, your child is growing out of his/her clothes really fast or he/she seems to be getting thinner.
- Think your child appears "different" in a way that concerns you.
- See a big deviation from the growth charts.

Your physician will use a physical exam along with growth charts to see whether your child is growing properly for them.

## WHAT OTHER TESTS MIGHT MY CHILD’S CAREGIVER REQUEST?

Since BMI isn’t a good measure for your child, your physician may suggest some other tests. These include skinfold tests, DEXA scans, and bloodwork. Your healthcare provider may even recommend a swallow study, a moving x-ray of a child’s swallow, and/or an UGI (upper gastrointestinal series), an x-ray used to measure reflux.

Tests Used to Assess SMA Nutrition		
Test	Measures	Important considerations
<b>Skinfold thickness</b>	Estimates fat stores at the triceps or other areas. These specified areas may be sensitive to changes in nutritional status.	Pediatric equations should be used for accuracy and better interpretation of results.  The dietitian should be a trained skinfold examiner.
<b>DEXA Scan</b> (Dual energy x-ray absorptiometry)	Measures the density or thickness of your child’s bones. Uses low-level radiation in order to distinguish between fat, fat free mass and bone.	Can scan the whole body or just one particular region.  Better determinant of body fat for children with SMA than BMI. Gives a more precise measurement of body composition.
<b>Bloodwork</b>	Used to assess nutritional status.	
<b>Swallow Study</b>	Uses a “moving x-ray” to watch your child’s swallowing mechanism and diagnose any difficulties.	Different consistencies of liquids and solids can be tested.
<b>UGI Series</b> (Upper Gastrointestinal)	Used to assess reflux.  An x-ray of the pharynx, esophagus, stomach and the duodenum (upper-most part of the small intestine).	Note: A negative result does not rule out reflux.

### What might a baseline evaluation of my child’s nutritional status include?

1. **Skinfold tests**
2. **DEXA scan**
3. **Bloodwork including:**
  - **CBC (complete blood count: checks for iron deficiency)**
  - **CMP (complete metabolic panel, includes electrolytes, calcium, magnesium, phosphorus, blood sugar)**
  - **Quantitative amino acids**
  - **Carnitine profile**
  - **Prealbumin**
  - **Vitamin D 25, OH**
  - **Blood glucose**
  - **Essential fatty acids**
4. **Bone mineral density**
5. **Swallow study**
6. **UGI**

# What, When and How Much? – Managing Nutrition in SMA

## WHAT KIND OF CALORIE NEEDS DO CHILDREN WITH SMA HAVE?

Infants and children with SMA are less active, have less muscle mass, and use less energy. *Therefore, they need fewer calories.* Fewer calories are needed for them to achieve an acceptable rate of weight gain or growth in length (using a National Center for Health Statistics Growth Chart).

Each child is different, but you can anticipate that your child will most likely need **20 to 50% less calories than a child** without SMA.<sup>5</sup> Aim for 10 to 20% of their total calorie intake to come from protein. The goal is to help your child maintain somewhere between the 3rd and the 25th percentile of length/weight on your physician's growth charts for healthy children.

## WHAT FOODS CAN BE ESPECIALLY TROUBLESOME FOR CHILDREN WITH SMA?

Certain types of foods are more difficult to chew and swallow than others, especially when there is muscle weakness in the jaw, tongue and throat muscles. Avoid foods that prove difficult for your children to chew and swallow. For example, avoid giving your child large or tough pieces of meat or sticky foods like thick cheese spread or peanut butter. These may be difficult to chew and/or swallow. Instead offer pureed or finely diced meat and soft cubes of cheese. If certain fruits and/or vegetables are too difficult for your child to chew, offer soft cooked or canned fruits or vegetables.

Some children have difficulty swallowing. Thin, clear liquids like water or juice could easily be inhaled into the airway. Try offering liquids in the form of a thin milkshake. Milkshakes can be fun and help prevent aspiration. (See Obstacles to Nutrition, on pages 19-24, for more information on aspiration.)



**Don't forget about the fluids!**

**Even if your child has reduced body mass, they still need fluids! They sweat and drool, and need a replacement of liquids.**

<sup>5</sup> This may not hold true for some younger children with type III, as they use more energy to get around.

## WHEN MIGHT MY CHILD NEED SUPPLEMENTAL FEEDING?

Although little is known about the best kinds of food in SMA, more is known about the timing of feeding, particularly when it comes to periods of fasting (extended times without food). The goal of supplemental feeding is to help your child feel better during the day.

Why is this needed? During a time when a child naturally goes without food – say from the evening meal to the next morning – the body still has work to do. So the body gets some of the energy it needs by burning muscle protein. This is a natural process, where energy flows into and back out of the muscles, like the tides of the ocean, after and between meals.

In children with SMA, particularly those with less muscle, this flow of energy in and out of muscle may be more difficult for the body to do. Long periods of going without food, like when your child is ill, can stress their body.

You might find that your child feels better and has more energy if you limit fasting. This can be done by adding a late night supplemental feed, or by adding a continuous drip of feedings via a G-tube, through the night. The goal isn't just to increase the number of calories, but to eliminate the long nighttime fast that makes it more difficult for them to function during the daytime.

## WHEN IS A FEEDING TUBE NECESSARY?

Your physician or dietitian may recommend tube feeding if your child is:

- Taking in less nutrients than he/she needs.
- Unable to eat by mouth.
- Struggling with digestion.
- Falling far behind on growth.
- Going through an illness or surgery.



## HOW DOES A FEEDING TUBE WORK?

Tube feeding (also called enteral feeding) allows a solution full of nutrients to be fed to your child through a tube leading into his/her stomach. The kind of tube feeding depends on your child's needs. Your doctor will help 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 (laparoscopy).

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

- The length of time the tube feeding might be needed.
- How well your child's GI tract is functioning.
- What would be most comfortable for your child.
- Whether your child has a risk of aspiration from feedings or secretions from his/her mouth or throat.

See the chart, *Understanding Feeding Tubes*, for information on types of tubes and considerations.





### Understanding Feeding Tubes

Type of tube	How it's used	Considerations
<b>Nasogastric Tube</b> (NG tube)	Passes through the nose to the stomach.  Used for short-term nutrition.	Can be completed at bedside by a physician, RN or registered dietitian. Correct placement is verified by x-ray.
<b>Nasojejunal Tube</b> (NJ tube)	Similar to an NG tube, but extends all the way into the small intestine.  Used for short-term nutrition.	Preferred if there is risk for aspiration, or if reflux, persistent nausea or vomiting are present.
Both NG and NJ tubes can be placed easily and replaced as necessary. A little uncomfortable, not good solutions for long-term use. Complications include: sinusitis, sore throat, tube blockage or dislodgement.		
<b>Gastrostomy Tube</b> (G-tubes)	Guided into the stomach through the abdominal wall with local anesthesia in a hospital or outpatient setting.  For long-term (more than 3 to 4 weeks).	Large tube diameter means less clogging. Can receive nutrient solutions and medications faster.  Skin level or button G-tube is less noticeable and causes less disruption to a child's activities.
<b>Percutaneous Gastrostomy</b> (PEG)	A tube that is placed from the inside out, using a special scope that is inserted into the stomach through the mouth.	Still placed under anesthesia. Preferred by some physicians and/or institutions.
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.		





## HOW ARE TUBE FEEDINGS GIVEN?

Just as you would take time to prepare food for a healthy child, and give care to the way you feed him or her, tube feeding can become a normal and loving routine. It's just a different approach that allows you to meet your child's need for nutrition and nurturing. Mouth feeding and tube feeding have the same goals: giving your child energy, ensuring good nutrition, and promoting wound healing and illness recovery.

After your child receives a NG, NJ or G-tube (see chart for details), a registered dietitian or physician will write a prescription for the feedings. The first question that needs to be considered is how the feeding will be done. The most common tube feeding methods are bolus feeding, intermittent drip feeding or continuous drip feeding.

**Bolus feedings** are administered through a syringe (similar to the syringes that come with children's medicine) into the feeding tube over a short time, typically 5 to 20 minutes. This type of feeding does not require a feeding pump and can be more convenient and less expensive. Bolus feedings are only given to children who can move food successfully through the digestive system.

Typically, bolus feedings are given with a G-tube. The larger diameter allows larger volumes of formula to pass into your child's stomach at a faster rate. Your physician or dietitian may recommend several bolus feedings per day to meet your child's nutritional needs. Complications of bolus feeding include nausea, vomiting, diarrhea, abdominal distention or aspiration.

**Intermittent drip feedings** can be administered through gravity drip or through a feeding pump. Several feedings are typically provided each day, each taking approximately 20-60 minutes. Like bolus feedings, drip feedings allow your child to be more mobile and have freedom outside of feeding time. Complications of intermittent drip may include nausea, vomiting, diarrhea, bloating or aspiration.

**Continuous feedings** are provided through a feeding pump over long periods of time. The amount of time spent feeding may range from 60 minutes to 24 hours per day. Although continuous feedings may make your child less mobile, they may be appropriate if he/she:

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

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



**Mealtime is an important social occasion...and tasty food is fun to eat**

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, and does not mean that eating is not possible. For instance, placing a small dollop of frosting from a birthday cake, on your child's pacifier can help her or him take part in an important family event.

## WHAT TYPE OF TUBE FEEDING FORMULA IS BEST FOR MY TYPE I CHILD?

There are three types of tube feeding formulas that each rely on a different protein source. They are called polymeric (whole protein), semi-elemental (peptides) or elemental formulas (amino acids). You'll be able to recognize them by their brand names:

- **Whole protein formulas** — Pediasure and Boost Kids Essentials.
- **Semi-elemental formulas** — Peptamen and Pediasure Peptide (formerly Vital Junior).
- **Elemental formulas** — Tolerex, Vivonex, Elecare and Neocate products.

You'll want to consult with a trained pediatric dietitian to determine what kind of formula is best for your child. Some questions they might consider include:

- **Does this formula contain too much fat** — Some of these formulas provide more than 30% of calories from fat, which may be too high after the first year of life.
- **Does this formula contain little fat?** — Tolerex contains very little fat. Additional fat sources may need to be added like flax or safflower oil. Vivonex also contains less fat, although more than Tolerex. Your dietitian may also suggest mixing a lower fat formula with a higher fat product such as Neocate Junior.
- **Does the formula contain adequate protein?** — Some children with SMA have low amino acid levels. They may require additional amino acid supplementation.
- **Does the formula contain adequate vitamins and minerals?** — Because calorie needs are lower, it can be hard to meet vitamin and mineral needs with formula alone. A separate vitamin and minerals supplement is often needed.

# Obstacles to Nutrition – Facing Feeding Challenges

Making sure your child is well nourished doesn't just mean considering what they eat, it also means considering how he or she eats. The very nature of SMA raises some feeding challenges. Understanding these can help you plan ahead and be aware of warning signs.

## WHAT EATING/DRINKING PROBLEMS CAN SMA CAUSE/CONTRIBUTE TO?

In children who are very weak, swallowing may be difficult either because of trouble moving food around the mouth with the tongue, tightness or weakness of jaw muscles, or difficulty protecting the airway. These eating and drinking difficulties could cause your child to accidentally “spill” food or drink into their airway.

Children with SMA also tend to struggle moving food through the GI system. Weak abdominal muscles can contribute to conditions such as acid reflux (food going backwards from the stomach to the windpipe) or constipation. Because nutrition is so important to your child with SMA, you'll also want to be aware of signs of conditions such as low blood sugar and yeast overgrowth.

## WHAT CAUSES ASPIRATION?

Aspiration occurs when small amounts of food or liquids enter the trachea (windpipe) instead of the esophagus (tube to the stomach). Aspiration can also occur:

- When stomach contents back up into the esophagus and then into the lungs.
- With vomiting during a stomach illness. Some children only aspirate when they are ill.
- During times of respiratory illness, when chewing and swallowing become especially difficult.

Not all children who aspirate during eating or drinking show obvious signs such as coughing or choking. This is known as **silent aspiration**. Silent aspiration may be suspected in individuals with a history of frequent respiratory problems, even without other signs of a cold (like nasal congestion or runny nose).

### Signs of possible aspiration

- Coughing or choking during eating and/or drinking.
- Acid reflux.
- Frequent respiratory problems without other signs of a cold (silent aspiration).

### To help prevent aspiration:

- Avoid foods difficult to chew or swallow.
- Offer liquids with the consistency of a thin milkshake.
- Consult with your dietitian or physician.



## HOW CAN I HELP PREVENT ASPIRATION?

Knowing the signs of aspiration and ways to prevent aspiration is the best place to start. Your child may be at more risk if he/she is very weak, even if it is due to a short-term illness. By knowing the signs ahead of time, you can get the child the help he or she needs more quickly.

Be especially alert if your child has trouble moving food around the mouth with their tongue, or if he or she has tight or weak jaw muscles. Any of these could make it difficult for your child to protect his or her airway and put him or her at risk for aspiration.

### Know the signs.

Again, 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 your child more likely to aspirate.
- **Cold/respiratory sickness:** Chewing and swallowing can be especially difficult when ill.

### Avoid hard-to-swallow foods.

Certain consistencies of foods are more difficult to chew and swallow than others when there is muscle weakness in the jaw, tongue and throat muscles. Use common sense and avoid foods that are difficult for your child to chew and swallow. These might include large or tough pieces of meat or sticky foods like thick cheese spread or peanut butter.

You may find that your child is more likely to aspirate thin, clear, liquids like water or juice. If so, try offering your child liquids that are slightly thicker, such as the consistency of a thin milkshake.

### Watch for signs of silent aspiration.

Not all children who aspirate show obvious signs such as coughing or choking. If your child has a history of frequent respiratory problems, when they do not have other signs of a cold such as nasal congestion or runny nose, you might ask your physician about the possibility of silent aspiration.

## HOW CAN I HELP MY CHILD WITH ACID REFLUX?

Acid reflux, caused by GERD (Gastroesophageal Reflux Disease) is a common problem in individuals with SMA. It is most common in children with type I, followed by type II, and less common in individuals with type III. To help your child with reflux:

- **Think small meals** — Try preparing your child six or more small meals throughout the day.
- **Follow a low-fat diet** — Fat causes food to leave the stomach more slowly. Children with SMA older than 1-2 years should not eat a diet with more than 30% of their total calories from fat. During illness, the amount of fat should be even less, no more than 15% to 20% of total calories.
- **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** — If your child is an infant or unable to sit up, you might try propping up his or her head/shoulders at a 20 to 30 degree angle 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 this special operation for serious acid reflux. Acid reflux can not only cause uncomfortable heartburn, but can 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.

A Nissen fundoplication makes it more difficult for stomach acid to travel backwards from the stomach to the esophagus. It is more likely to be needed by children with SMA type I.

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.

### How much fat content?

For your child with SMA older than 1-2 years, aim for 25% to 30% of calories coming from fat.

Let's say your child is on a 1,200-calorie diet. Use this formula:

$1,200 \text{ (calories)} \times .30 \text{ (30\% from fat)} = 360 \text{ (total calories that can come from fat)}$

$360 \text{ (fat calories)} \div 9 \text{ (calories per gram of fat)} = 40 \text{ fat grams.}$

So you should aim for 33 to 40 grams of fat a day. Be sure to look for the fat gram content on food labels.

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

## HOW DOES SMA MAKE MY CHILD PRONE TO CONSTIPATION? HOW CAN I HELP?

Children with SMA benefit from foods with high fiber (see sidebar) to help prevent constipation. They are more likely to suffer from constipation because they have weak muscles in their abdomen. This can cause the body to struggle to move food through the digestive system. In addition, children with SMA are more likely to have less fiber and fluids in the diet.

Constipation can lead to abdominal discomfort. It can also cause your child 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 children with SMA type I. For some children, constipation gets better when they get more fiber and/or fluid. You might try offering prune juice.

**Note:** If prune juice is too harsh for your child, try pear, apple or white grape juice. If the problem continues, your doctor may recommend Miralax or other laxatives, depending on your child's muscle strength.

## SHOULD I BE CONCERNED IF MY CHILD IS OVERWEIGHT?

Obesity (being very overweight) is a serious health concern that increases the amount of care you'll need to give your child. It can also take away from self-image. Being overweight also makes physical activity more challenging, and can make it more difficult to breathe or be lifted.

### Make sure your child gets enough fiber!

#### 1. Determine the amount needed — Add 5 to your child's age.

Example: A 3 year old needs 8 grams of fiber a day.

#### 2. Choose the right foods — High fiber foods include whole grain cereals/ breads and fruit and vegetables (carrots, apples, celery, oranges, dried fruits, juices, etc.) Try soft cooked or canned versions of these foods or mix them into smoothies and batters. Raw fruits and vegetables can be hard to chew or swallow.

These alone are good reasons for parents to help their child stay in a normal weight range. However, when a child has SMA, being obese can increase the risk of some very serious health conditions:

- Increased risk of diabetes and hypertension.
- Greater pain.
- Risk of losing the ability to walk, for patients with SMA type III.

Children with SMA who are very weak are more likely to have trouble processing fat. Aim to keep your child's total fat intake between 25 to 30% of his/her calories after 1-2 years of age. (Some children who are very weak may need less than this amount.) *Children with SMA may also require a supplement called Carnitine to help the body process fat. Talk to your dietitian or physician if you have questions about these issues.*

## **HOW CAN I RECOGNIZE A YEAST INFECTION?**

Some parents report issues with yeast in their children with SMA. The sign/symptoms include:

- Orange tint in eyebrows, hair and around G-tube.
- Cheesy smell on hands and feet.
- Increased sweating, bloating or constipation.

Please seek medical attention if you suspect your child has a yeast infection.

## **WHAT IF I SUSPECT BLOOD SUGAR ISSUES?**

Some children with SMA can experience blood sugar issues. If you suspect this, you may want to ask your physician to check your child's blood glucose level. A dietitian can help analyze your child's diet. Your dietitian will want to check to see if your child is receiving:

- Less than 30% fat;
- Less than 10-20% protein, and;
- Greater than 50-60% carbohydrate.

If these are out of balance, your dietitian may suggest changing your child's diet to decrease the carbohydrate and increase the fat and protein. The dietitian may also want to find out the amount of time your child is waiting between feedings. Going more than 6 to 8 hours between a feeding may cause low blood sugar. If these changes do not help, your physician may want to do further testing/evaluation.



## WHAT DO I NEED TO KNOW ABOUT NUTRITION DURING MY CHILD'S ILLNESS OR SURGERY?

Because of limited muscle mass (total weight of muscle in their body), many children with SMA find it difficult to get better after an illness. Since many childhood 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 children 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, robbing their overall strength. And, if the body is unable to get enough energy from muscle, your child may become seriously ill with hypoglycemia (low blood sugar).

If your child is hospitalized and only receiving IV sugar (glucose), they can develop low blood sugar. Your doctor may recommend use of "parenteral nutrition," also known as PPN or TPN. This IV solution includes sugars, amino acids and vitamins.

Talk to your physician or dietitian if your child cannot tolerate any form of feeding for a significant amount of time due to illness or surgery. They may recommend supplemental feeding to support your child.



### After illness or surgery

**When your child has had a long illness or is recovering from surgery, supplemental feeding might help them regain function more quickly.**

**Check with your dietitian or physician for advice.**



# Finding the Balance — Preventing Undernutrition or Overnutrition

## HOW DOES TYPE OF SMA AFFECT MY CHILD'S NUTRITIONAL CHALLENGES?

Depending on the type of SMA your child has, they may be at risk of either undernutrition or overnutrition. For instance...

- Children with severe SMA type I or type II are more prone undernutrition, which causes the failure to thrive.
- Children with strong SMA type II or type III are more likely to suffer from overnutrition, which leads to being overweight or even obesity.

## SEVERE SMA TYPE I AND WEAKER TYPE II — PREVENTING UNDERNUTRITION

If you have a child with severe SMA, it is critical to make sure they get enough nourishment and gain enough weight. Children who are significantly underweight or undernourished are at risk for additional problems due to their SMA. For instance, they could be more likely to get infections, have trouble healing a wound, or develop a tendency to get pressure sores.

Children with SMA type I eventually require tube feedings because of muscle weakness and the inability to consume food by mouth. Some children with SMA type II require nightly tube feedings in addition to their regular diet. By making sure your child gets proper nutrition, you give your child the best chance of establishing or regaining functioning and strength.

Even though there is concern of undernutrition in children with severe SMA type I and type II, increased fat may cause digestion and other problems. Because of this, high fat diets are not recommended. It is important to keep total fat intake within 25 to 30% of his/her calories in fat. Sometimes, an even lower fat diet is needed.

**Note:** This fat intake recommendation (25 to 30%) only applies to children older than 1-2 years. The AAP recommends 30-50% fat for infants.

### Undernutrition:

Too little food or from the inability of the body to convert or absorb it.

### Overnutrition:

Taking in too much food, especially in unbalanced amounts.

### Failure to thrive:

A child that is too low in height and weight because he or she failed to grow or because he or she has dropped several percentiles (for instance from the 75th to the 25th percentile).

## **STRONG SMA TYPE II AND III — PREVENTING OVERNUTRITION**

For stronger children with SMA type II or type III, preventing obesity, and all the accompanying health risks, is a major concern. Strong children with SMA type II can usually eat enough to maintain a healthy weight/length, but also need to be watched to make sure they don't become overweight.



Children with SMA type III are most at risk for gaining too much weight. The calorie requirements for a SMA type III child could be as low as half of what a normal child may need.

It is best for children who have strong SMA type II and III to get an evening snack, in addition to eating three meals a day. This prevents them from going too long without food.



### **How can I help my child maintain a good nutritional status?**

A skilled pediatric dietitian will help you monitor your child's growth and eating. They can help you assess your child's nutritional status, develop a plan, and help you respond to challenges that come along the journey.



# Conclusion and Resources

As you've learned in these pages, nutrition is a complex issue for anyone, but especially for infants and children with SMA. While we wait for more studies to be completed that give us more conclusive evidence about the best way to meet the nutritional needs of children with SMA, there is much we have learned. Due to the unique challenge SMA presents, it is important to assess your child's growth and nutritional status, and then choose interventions that best meet whatever nutritional challenges he or she may face.

Although nutrition is a moving target for the families with children who have SMA, there is much to be encouraged about. With the help of a registered dietitian, your physician, and other support professionals, you can make intelligent decisions that help improve your child's quality of life and resilience to infection and sickness.

Here are a few resources that can help you on this journey.

**Cure SMA website:**  
**[www.cureSMA.org](http://www.cureSMA.org)**

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Mahan, K and Escott-Stump, S. *Krause's Food and Nutrition Therapy*, 12th edition. Saunders, Elsevier: St. Louis, MO, 2008.

Shils, M, Olson, J, Shike, M, and Ross, A, eds. *Modern Nutrition in Health and Disease*, 9th edition. Lippincott Williams & Wilkins: Philadelphia, PA, 1999.

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# About Cure SMA

Cure SMA is dedicated to the treatment and cure of spinal muscular atrophy (SMA)—a disease that takes away a person’s ability to walk, eat, or breathe. It is the number one genetic cause of death for infants.

Since 1984, we’ve directed and invested in comprehensive research that has shaped the scientific community’s understanding of SMA. We are currently on the verge of breakthroughs in treatment that will strengthen our children’s bodies, extend life, and lead to a cure.

We have deep expertise in every aspect of SMA—from the day-to-day realities to the nuances of care options—and until we have a cure, we’ll do everything we can to support children and families affected by the disease.

Learn more about how you can help us reach a treatment and cure at [www.cureSMA.org](http://www.cureSMA.org).

# Contacting Cure SMA



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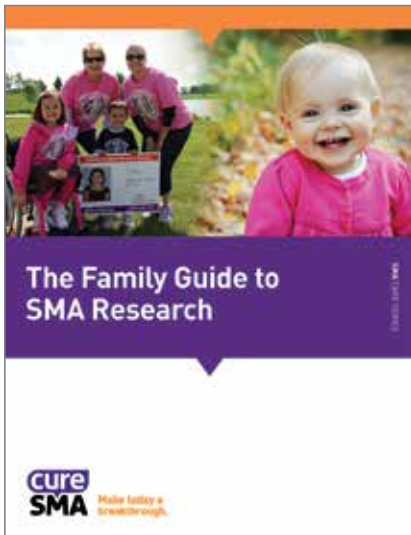


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We're on the Web!  
[www.cureSMA.org](http://www.cureSMA.org)

# Other Publications From Cure SMA



## Family Guide to SMA Research

This guide helps to answer questions such as what the key areas of SMA research are and what the costs for conducting SMA drug development and clinical trials are. Clear definitions and graphics are included to help explain and illustrate how SMA drugs begin and the key steps involved in developing new therapies for Spinal Muscular Atrophy.

The topics in this new booklet cover the following important areas of SMA research:

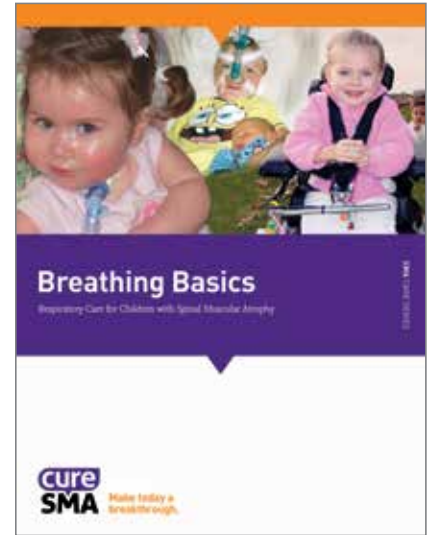
- SMA Researchers.
- Drug Research.
- Gene Therapy.
- Stem Cells.
- Clinical Trials.
- Government Research and the FDA.



## Cure SMA Family Support and Patient Services

This booklet has details on the following Cure SMA programs:

- 1) Programs For Newly Diagnosed Families:
  - Including our special type I programs such as: Care Packages; Sheep Skin Blankets; wagons.
- 2) Cure SMA Equipment Pool.
- 3) Medical Care.
- 4) Daily Living.
- 5) Local Support.
- 6) How to Keep up to Date.
- 7) The Annual SMA Conference:
  - Including the Cure SMA Newly Diagnosed Conference Program.

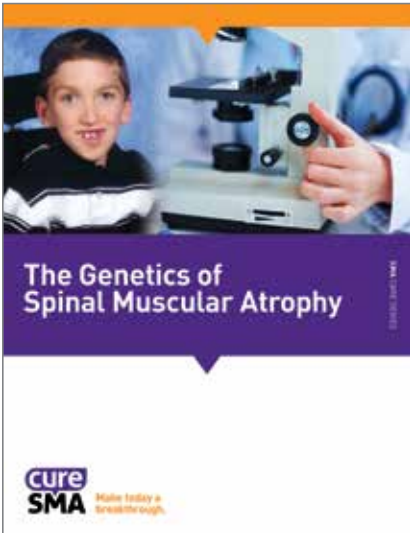


## Breathing Basics

This new booklet is focused on the critical aspects of respiratory care for children with Spinal Muscular Atrophy. The booklet was authored by Mary Schroth, M.D., a member of the Cure SMA Medical Advisory Council, and a leading expert on respiratory care for SMA patients.

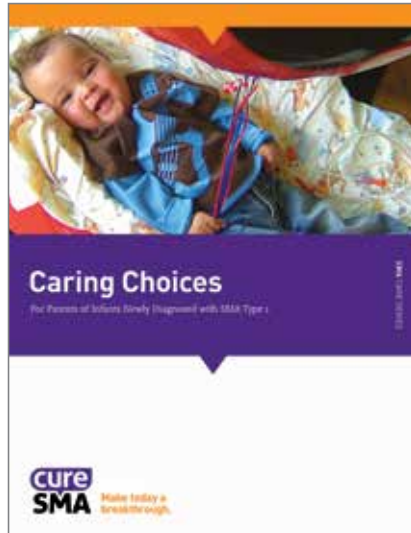
This booklet reviews the following important topics:

- Why is respiratory care so important in SMA?
- What are common respiratory problems in children with SMA?
- Elements of respiratory care management in SMA
- What are special needs of children with SMA type I, type II and type III?
- What respiratory equipment will you need at home?



### The Genetics of Spinal Muscular Atrophy

Confused about genes, proteins, DNA and how SMA is diagnosed? Read this helpful pamphlet. It includes definitions, explanations and diagrams from genetics expert Louise Simard, Ph.D. and the Cure SMA Medical Advisory Council.



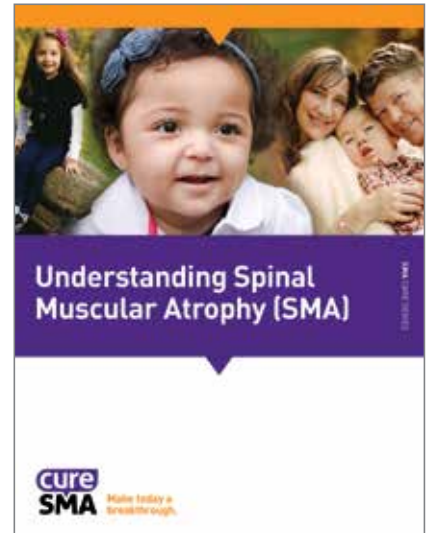
### Caring Choices

This booklet is focused on caring choices for parents of infants newly diagnosed with Spinal Muscular Atrophy type I.

Topics review the basics of the main care options for newly diagnosed SMA type I:

- What is Non-Invasive
- What is Non-Invasive Respiratory Care?
- What is Invasive Respiratory Care?
- What is Palliative Care?

And where you can go for support and guidance.



### Understanding Spinal Muscular Atrophy (SMA)

**For electronic copies:**  
Download this booklet from the Cure SMA web site at [www.cureSMA.org](http://www.cureSMA.org). Go to the support & care publications section on our website.

**For print copies:**  
Please contact the Cure SMA national office at [info@curesma.org](mailto:info@curesma.org).

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Cure SMA does not, as an organization, support or endorse any particular treatment or therapy. Information contained in this booklet is for informational and educational purposes only. All medical information presented should be discussed with a qualified physician.

**If you would like a hard copy mailed to you please email us at [info@curesma.org](mailto:info@curesma.org) or call 800.886.1762**



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