

# **Nutrition Support in Spinal Muscular Atrophy**

2016 Spinal Muscular Atrophy (SMA)  
Symposium on Optimizing Care

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September 30, 2016

# Goals

1. Briefly summarize the Nutrition Section listed in the 2007 Standard of Care (SOC)
2. Share a brief summary on what parents in the SMA Community are doing to fill in for the missing gaps in the SOC
3. Summarize further research opportunities with outcome measures for furthering the quality of nutrition care for SMA patients

# Consensus Statement for Standard of Care in Spinal Muscular Atrophy

Ching H. Wang, MD, PhD, Richard S. Finkel, MD, Enrico S. Bertini, MD, Mary Schroth, MD, Anita Simonds, MD, Brenda Wong, MD, Annie Aloysius, MRCSLT, HPC, Leslie Morrison, MD, Marion Main, MCSP, MA, Thomas O. Crawford, MD, Anthony Trela, BS, and Participants of the International Conference on SMA Standard of Care

Journal of Child Neurology  
Volume 22 Number 8  
August 2007 1027-1049  
© 2007 Sage Publications  
10.1177/0883073807305788  
<http://jcn.sagepub.com>  
hosted at  
<http://online.sagepub.com>

## Consensus on Gastrointestinal and Nutritional Care

### Overview of Gastrointestinal and Nutritional Complications in Spinal Muscular Atrophy

# 2007 SMA Standard of Care

# Nutrition

## Growth

- Calorie goals for each individual child to follow his or her growth curves for length/height, weight, weight/height
- Percutaneous gastrostomy tube placement with Nissen fundoplication when infant/child at least risk from complications when inadequate oral feeding or safety is of concern

# 2007 SMA Standard of Care

# Nutrition

## Nutrition

- No special diet or formulas
- No protein restriction or excess
- No fat restriction or excess
- Meet fluid needs
- Meet DRI for vitamins and minerals

## Bulbar dysfunction

- Feeding or swallowing dysfunction symptoms in infants/children include weak suck/bite, limited mouth opening, fatigue, prolonged feeding
- Video fluoroscopic swallow study if concerns about swallow safety or aspiration
- Risk of oropharyngeal aspiration of liquids, solid food, and saliva. Concern of silent aspiration, which may lead to pulmonary infections

## Gastroesophageal dysmotility

- **Constipation** caused by poor tone in abdominal muscles and immobility
  - Bowel regimen important in type 2 to aid chronic constipation or impaction
- **Delayed gastric emptying**
  - Evaluation by gastroenterologist
- **Gastroesophageal reflux** can exacerbate feeding problems and increase risk of aspiration
  - Prokinetic agent for delayed emptying
  - Presurgical evaluation for gastrostomy placement/Nissen
- Meet fiber and fluid needs. Caution with high fiber diet as it may contribute to abdominal bloating. Preventing abdominal distension from bloating will aid in sustaining respiration

## Acute illness

Avoid prolonged fasting. Meet full caloric needs in 4-6 hours of admission either enterally or parenterally

- Metabolic decompensation (especially type 1 and 2)
- Prevent hypoglycemia

Review

## Describing nutrition in spinal muscular atrophy: A systematic review

Georgia E. Moore <sup>a</sup>, Amara W. Lindenmayer <sup>a</sup>, Grace A. McConchie <sup>a</sup>, Monique M. Ryan <sup>b,c,d</sup>,  
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Received 6 November 2015; received in revised form 6 May 2016; accepted 6 May 2016

Reviewing the evidence base for nutrition in SMA reveals consistent limitations across all areas, with little high-quality evidence to guide practice. The heterogeneity of studies with regard to outcomes measured makes it very difficult to draw definitive conclusions. Interventional studies are scanty, so extrapolation from observational studies is necessary. However, these observation studies are not without merit. Given the obstacles that SMA researchers face, observational studies may comprise the strongest and most accurate evidence available to assist this fragile group. Future research efforts should be directed towards longitudinal studies documenting the long term nutritional consequences of SMA as well as high-quality clinical trials investigating targeted nutrition therapies. In order to obtain adequate sample sizes, collaboration across multiple sites will be critical.

This systematic review presents a comprehensive synthesis of the current nutrition literature in SMA. Nutritional management of patients with SMA is complex but there is little high-quality evidence to guide practice. Growth, body composition and energy requirements are likely different in persons with SMA, but further research is needed before nutritional guidelines can be developed. Findings from this review may assist clinicians in making informed clinical decisions about the nutritional care of children with SMA.





ScienceDirect

Neuromuscular Disorders 26 (2016) 395–404



www.elsevier.com/locate/nmd

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### Summary

- Growth and body composition
- Nutritional issues
- Nutrition management strategies

### Discussion

- Of 39 studies retrieved, focusing predominantly on types I and II, overall quality of evidence was low
- Very little known about energy requirements
- No well designed studies prospective studies on use of elemental diet

# Parent Community



The image shows a Facebook login banner. At the top, the Facebook logo is on the left, and the login fields for "Email or Phone" and "Password" are on the right, with a "Log In" button and a "Forgot account?" link. Below the header, on the left, is a grid of 15 Facebook 'f' icons in various colors and styles. In the center is a yellow awareness ribbon with a red rose in the center, containing the text "FIGHT SMA CURE". To the right of the ribbon, the text "SMA AWARENESS RIBBON" is written in a bold, red, serif font. Below this, two paragraphs of text describe the ribbon's symbolism: "The Ivory Ribbon represents the innocence and purity of all those affected by SMA." and "The Rose stands for love, inner - strength and courage."

facebook

Email or Phone

Password

Log In

Forgot account?

**SMA AWARENESS RIBBON**

The Ivory Ribbon represents the innocence and purity of all those affected by SMA.

The Rose stands for love, inner - strength and courage.

# Parent Community Concerns

## SMA Type 1

- Usually dependent on tube feedings as sole source of nutrition
  - Frequently say their child is not tolerating standard formula
  - Advocate for a low fat diet
    - Elemental (amino acid) formula and/or breastmilk past 1yr of age

## SMA Type II or III

- Usually modified consistency oral diet and/or supplemented with tube feedings.
  - Frequently say, “Will the amino acid diet benefit my child with type 2 or 3 ?”
  - Some parents advocate for elemental formula while providing normal table food

# SMA Support Inc.

Supporting Families With SMA

To Have A Diet "Expert" contact you with regards to the Amino Acid Diet for your child, Please Send Us an Email with the following

information: 1). Your Name, 2). Contact Information, 3) Age of Child, 4) Type of SMA, 5) Current Weight and Height of child, 6) Whether they eat orally or not, 7) Do they have a g-tube or not, and 8) What does their current diet consists of in general. The appropriate helper for your specific child's needs will contact you and provide more information on how the Amino Acid Diet can be applied in your specific situation.

## WHAT IS THE AMINO ACID DIET?

*This information is based on opinions and experiences compiled from parents of children with SMA and should not necessarily be relied upon as an alternative to medical advice from professional health care providers.*

### ABOUT THE DIET

The Amino Acid Diet is a nutritional approach that many SMA families believe has had a positive impact on their child's quality of life.

Using this dietary approach, milk and soy based formulas are eliminated and substituted with a low-fat, elemental free form amino acid formula. Elemental

Many children with SMA seem very reactive to the proteins and fat in dairy and soy formulas. Their reactions are often attributed to their SMA progression. However, when this protein and fat are replaced with free form amino acids and appropriately reduced amounts of fat, children with SMA experience improved respiratory health and fewer metabolic complications during illness. Most children with Type 1 SMA have an immediate reduction in airway secretions. Constipation, which can be a major complication, is more manageable or even eliminated.

Some children have improvement in strength and regained function.

Parents using these formulas vary their child's particular diet based on tolerance, age, weight and length, and severity of SMA, but there are some basic guidelines that most children respond to favorably.

Children with SMA do not tolerate large amounts of amino acids or fat due to their reduced muscle mass. This reduction of muscle mass prohibits the proper absorption, storage and utilization of amino acids and fat, which could result in toxic accumulation in the bloodstream.

# The “Amino Acid” diet

Recommended for children one year and older

Elemental formula with water and electrolytes

- No intact protein or semi-elemental cow milk/soy milk based formulas

Protein limited to 7-10% total calories

- No animal proteins such as dairy, meat, egg

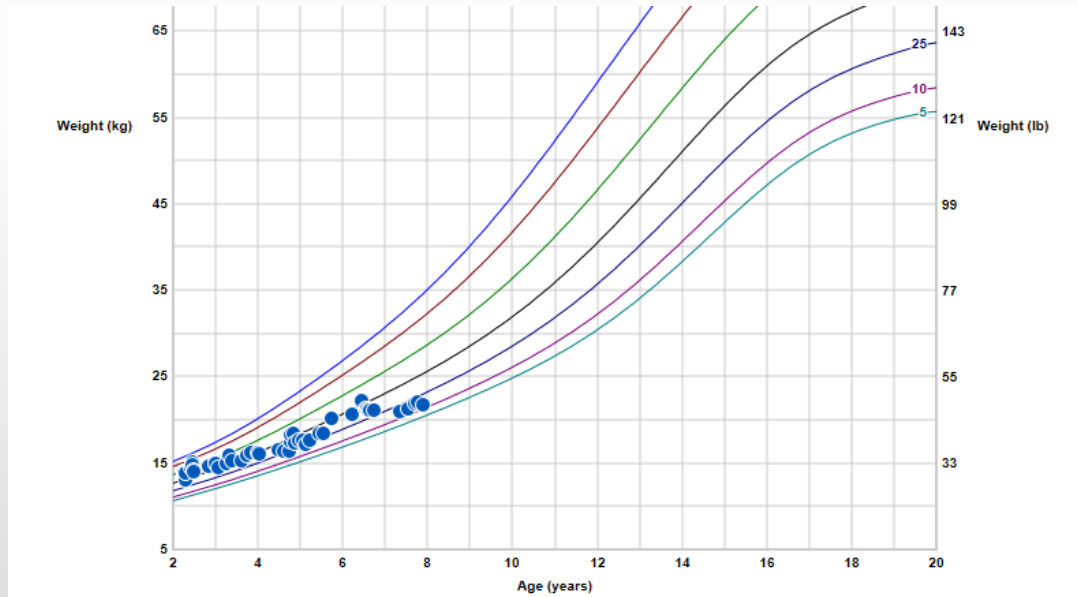
Fat limited to <30% total calories

Juices, puree fruit and vegetables, grains

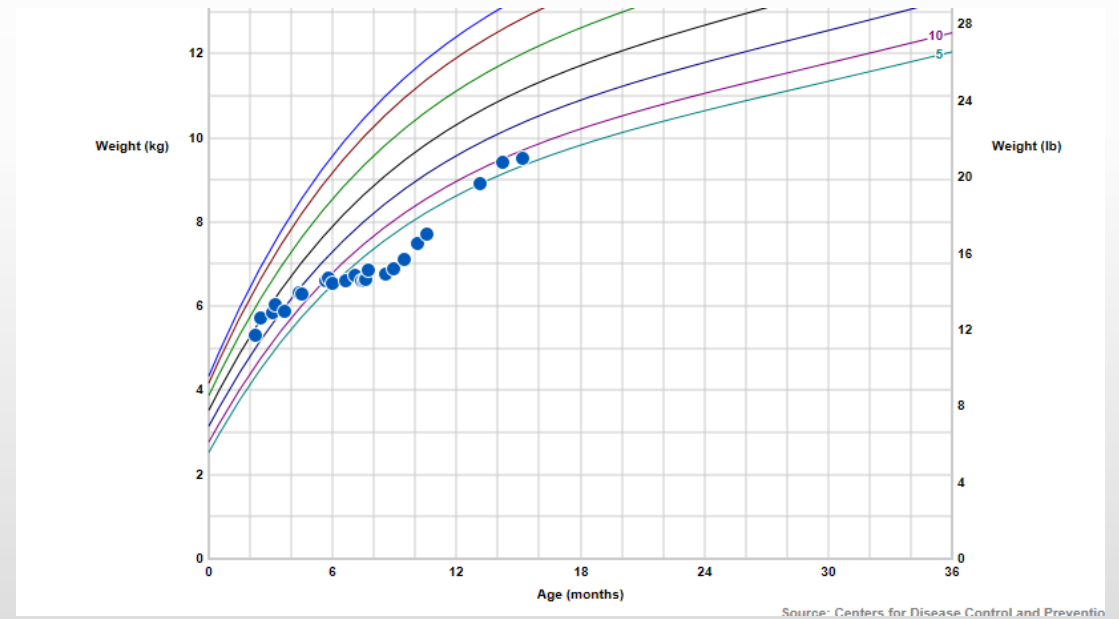
Human milk

# Growth Chart comparison

## Growth of Type I patient tolerating standard pediatric formula



## Growth chart of Type I patient on continuous modified Amino acid diet advocated by parent



# Future Considerations

The image shows a Facebook page for 'Spinal Muscular Atrophy News'. The page header includes the Facebook logo and login fields for 'Email or Phone' and 'Password', with a 'Log In' button and a 'Forgot account?' link. The profile picture is a DNA double helix. The page name is 'Spinal Muscular Atrophy News' with the handle '@SpinalMuscularAtrophyNews'. A navigation menu on the left includes 'Home', 'About', 'Photos', and 'Likes'. The main content area features a large word cloud with the word 'Hope' as the central focus. Other words in the cloud include 'Research News', 'Spinal Muscular Atrophy', 'SMN1 Gene', 'Cure', 'SMN2 Activation', 'SMA Type 1', 'Drugs in development', and 'Gene Therapy'. Below the word cloud are interaction buttons for 'Like', 'Message', 'Share', and 'More'. At the bottom, there is a post header for 'Spinal Muscular Atrophy News' dated 'May 22' and a category label 'Health/Wellness Website'.

# Future considerations

Gastrointestinal/Nutrition group strong recommendations  
for multidisciplinary approach

## Future areas of study:

1. Use of **elemental formulas**-support/refute perceived benefits of optimal growth and decreased oral secretions
2. Need for reduced fat intake, in consideration of concern for **mitochondrial fatty acid oxidation abnormalities**
3. Need for **protein supplementation** beyond dietary recommended intake- related to muscle wasting/atrophy
4. Need for checking **biochemical tests** to screen metabolic/mitochondrial fatty acid abnormalities
5. Use of **body composition** in the assessment of nutritional status and creation of a population-specific growth chart



# Future considerations

- Total calorie needs
- Glucose, protein and fat metabolism
- **Acute illness guidelines** based on glucose control, protein and fat metabolism rate research
- Supplements- creatine
- Bone health -affected by ambulatory status
- Lab monitoring and frequency- Comprehensive metabolic panel, Vitamin D, fasting and non-fasting glucose tolerance profiles

# Conclusion

1. Summarized the Nutrition Section listed in the 2007 Standard of Care (SOC)
2. Shared a brief summary on what parents in the SMA Community are doing to fill in for the missing gaps in the SOC
3. Summarized further research opportunities with outcome measures for furthering the quality of nutrition care for SMA patients