Nutrition Support in Spinal Muscular Atrophy

2016 Spinal Muscular Atrophy (SMA) Symposium on Optimizing Care

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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, MRCST, 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

Consensus on Gastrointestinal and Nutritional Care

Overview of Gastrointestinal and Nutritional Complications in Spinal Muscular Atrophy
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

- 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

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2007 SMA Standard of Care

Nutrition

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

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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
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.
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 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
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

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Future Considerations
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

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