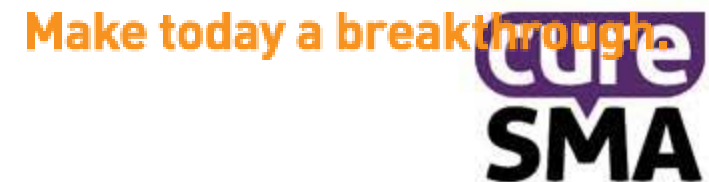


Tube Feeding and SMA: Recommendations and Practices

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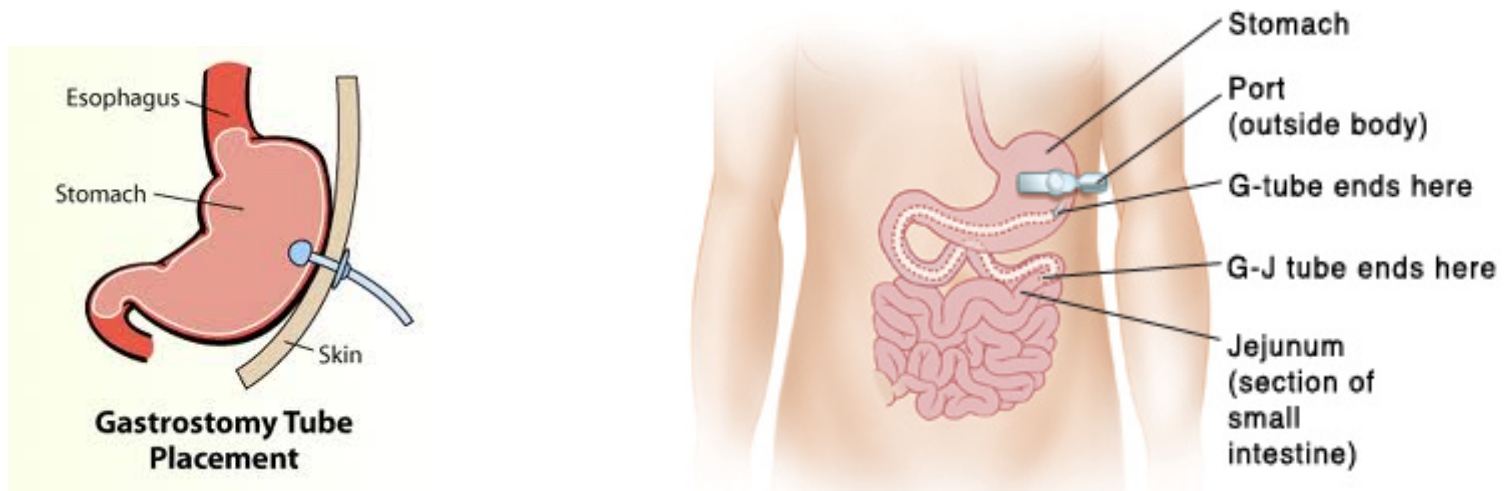
Outline

- Common nutrition issues
- Understanding growth
- Understanding nutrient intake
- Special diets & supplements
- Feeding issues and intolerance
- Possible nutrition implications of new therapies
- How a dietitian helps

Feeding and Swallowing Problems

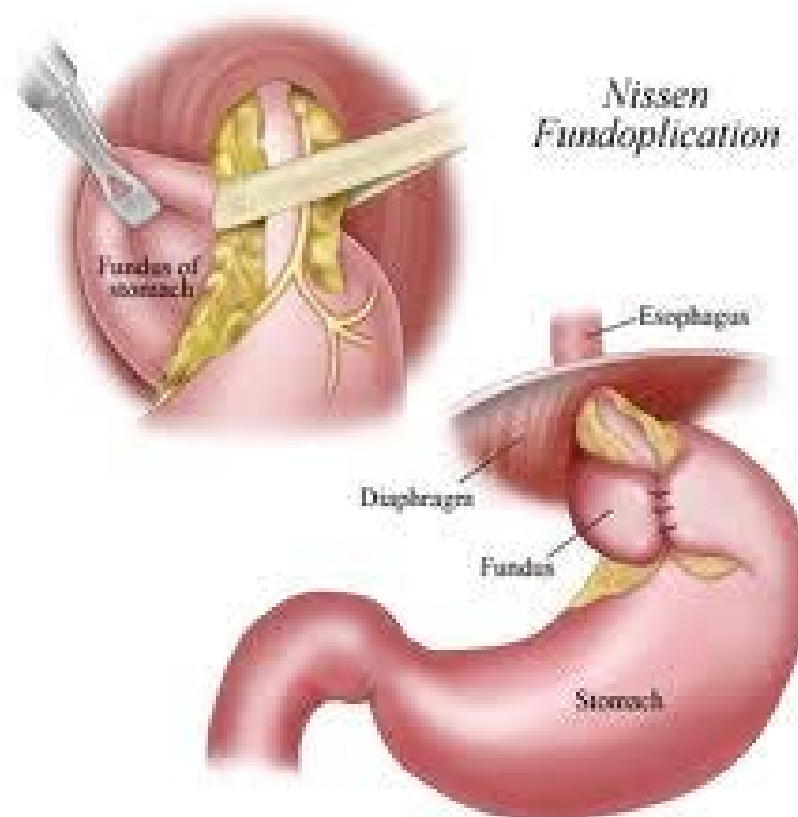
- Weakened swallowing muscles, poor head control
→concern for safe swallowing
- High risk for aspiration => food/formula goes to lungs rather than stomach
- Recommend swallow study :
 - Presentation with SMA symptoms in infancy
 - Symptoms (coughing with feeds, fatigue, pneumonia, long feeding times-things change fast)
- Modifications, Feeding tube for nutrition

Feeding tubes



Nissen Fundoplication

- For those with aspiration/reflux concerns
- Speak with GI



Different Ways Feeding Tubes are Used

- Night time tube feeds with oral feeds during the day.
- Bolus feeds after meals to top off calories.
- Bolus for snacks between meals
- To provide majority of nutrition for those with swallowing/aspiration risks
 - can run continuously over 24 hours or during the day or
 - pumped over shorter periods, depending on risk,
 - bolus feeds, depending on risk.
- For hydration/fluids only
- For medications and fluids
- During illness/fatigue or on days not eating well.

Timing of Feeds

- Individualized based on strength and tolerance
- May need to move to continuous or slower feeds for:
 - GI dysmotility
 - Volume tolerance
 - Illness

Gastroesophageal Reflux

- Symptoms
 - Spitting up or vomiting after feeds
 - Chest or abdominal discomfort
 - Arching back
 - Bad Breath
 - Refusal of feeds
- Solutions
 - Nissen fundoplication
 - Jejunal tube placement
 - Positioning
 - Smaller feed volume, more often
 - Slow feeding rate
 - Dietary modifications
 - Medicine

Constipation

- Symptoms
 - Abdominal distention and bloating
 - Irritability
 - Sweating, red face
 - Respiratory distress
- Causes
 - Abnormal gastrointestinal motility
 - Reduced intake of dietary fiber
 - Inadequate fluid intake
 - Low muscle tone of abdominal wall

Constipation

- Solutions
 - Increase water!
 - Pureed fruits/vegetables
 - Juice- apple, pear, white grape, prune
 - Dose based on age/weight
 - Probiotics (all strains are not the same)
 - Medication for constipation, GI dysmotility

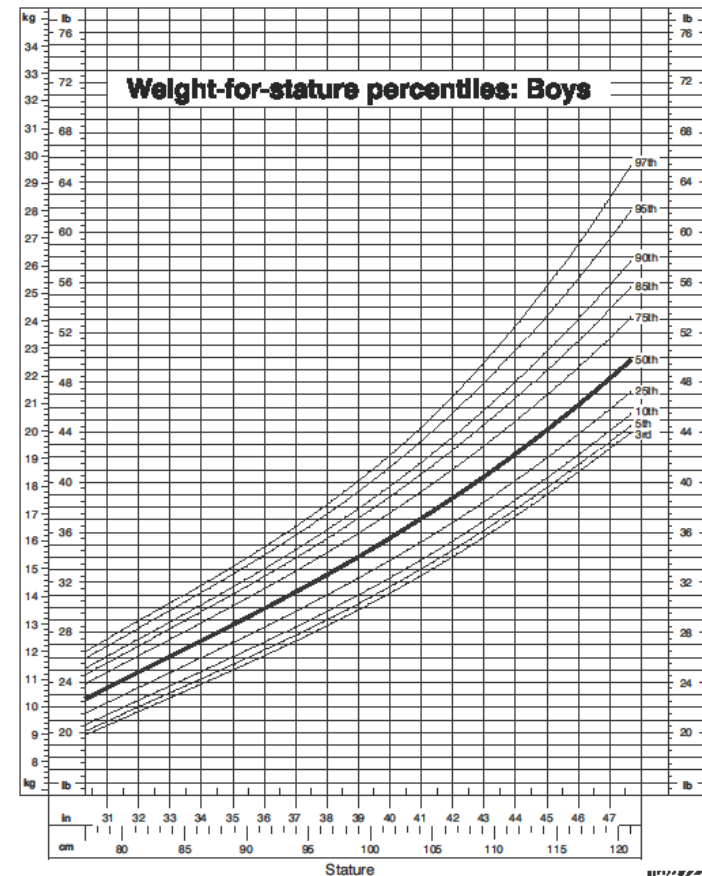
FOR ACUTE CONSTIPATION MEDICATION IS
NECESSARY

Growth

Weight/Length

- Growth chart
 - Follow trends
- Charts are not good tools by themselves.
 - Increased fat mass
 - less muscle mass
 - $>50^{\text{th}}$ %ile wt for length may be overfat
 - $<5^{\text{th}}$ %ile wt may be appropriate

CDC Growth Charts: United States



Published May 30, 2000 (modified 11/21/00).
SOURCE: Developed by the National Center for Health Statistics in collaboration with
the National Center for Chronic Disease Prevention and Health Promotion (2000).

SAFER • HEALTHIER • PEOPLE

Growth

- Excess weight gain can further reduce mobility and strength.
- Poor weight gain can reduce strength and immunity.
- Diet changes or non-ideal growth may require more frequent assessment.
- Important to monitor over time.

DIET

- Nutrients to grow:
 - Calories
 - Protein
 - Carbohydrates
 - Fat
 - Vitamins, and minerals

Calories

- Sources of calories
 - Protein
 - Carbohydrates
 - Fat
- People with SMA need less
- Lower lean muscle mass
- Depends on activity, work of breathing, etc.
- Clinician adjusts calories higher or lower based on growth

Protein

- Children require protein for adequate growth. Adults need less.
- In general: 0.45-0.9 g protein per pound weight (1-2 g protein/kg).
 - Infants require more protein.
 - Others may need more in certain cases.
 - Those on amino acid formulas need at least 20% more.
- Protein sources
 - Commercial formula
 - Blenderized foods-peas, chicken, fish
 - Supplemental protein mixes

Fat

- Recommended fat intakes
 - Infants-35-50%
 - Children older than 2 yrs- 20-30%
- Fat sources-
 - Commercial formula
 - Healthy fats- avocado, fish, flaxseed, plant oils
- Essential Fats-
 - Not made in body
 - Must come from food
- Essential fat sources-
 - Omega 3: walnut, fish, flaxseed, canola oil
 - Omega 6: safflower oil, egg, poultry

Fat

- Too much fat can:
 - slow gastric emptying
 - increase reflux
- Too little fat stunts:
 - brain development
 - eye development
 - growth
- Signs of essential fatty acid deficiency:
 - scaly skin
 - hair loss
 - poor wound healing
 - increased risk of infection
 - poor growth

Vitamins and Minerals

- No research specific to SMA
- Follow Dietary Reference Intakes (DRI) for healthy children/adults
- Age and gender specific
- http://www.nationalacademies.org/hmd/~media/Files/Activity%20Files/Nutrition/DRI-Tables/2_%20RDA%20and%20AI%20Values_Vitamin%20and%20Elements.pdf?la=en

Fluids

- For hydration
 - Often sweat a lot
 - Increased respiratory losses
- Prevent constipation
- Usually recommend 100-135 ml fluid per kg body weight for infants and younger children.
 - Adjusted as tolerated
- Adequate?
 - Check frequency
 - Urine color
 - Labs

Nutrients at Risk for Deficiency

- Omega 3 & 6 Essential fatty acid (EFA)
- Iron (iron deficiency anemia is common)
- Calcium
- Fat Soluble Vitamins- (ADEK)
- Magnesium
- Folate

Supplements

- May require a multivitamin or individual supplement to meet intake needs.
- Avoid Mega-doses or high intakes of supplements
 - Higher intakes can have side effects
 - Niacin-flushing, itching
 - Calcium-kidney stones
 - B12-covers up folate deficiency
- Carnitine Supplementation?
 - Fat transporter concentrated in muscle
 - May recommend if low carnitine levels
- Work with a dietitian to determine supplement doses.

Sodium Intake

- Only add if recommend by doctor/dietitian and use under medical supervision.
 - Pedialyte
 - Iodized table salt - contains iodine which can help increase iodine intake.
 - Morton Lite salt – if potassium needed.
 - Himalayan Salt-several minerals, less iodine

Calcium

- Take calcium separately from iron and fiber
- Break up dose to 200-300 mg at a time
- Calcium carbonate-with food
- Calcium citrate-food not needed
- Ask dietitian if a calcium + is needed
 - Cal-Mag- Zinc, Cal Plus, Cal +vit D solutions

Vitamin D

- Many children with SMA have low vitamin D levels
- Low vitamin D-increases fracture risk
- Supplement or sunlight (Watch sunscreen)
- Amount depends on age and vitamin D level
- Infants exclusively receiving breast milk
- Check 25, hydroxy Vitamin D levels
 - Every year, more often if low

Calcium and Vitamin D Recommendations

Age Group	Calcium (mg)	Vitamin D* (IU)
Infants	200-260	400
Toddlers	700	600
Children/Teens	1000-1300	600
Adults	1000-1200	600-800

* Recommended vitamin D intakes may be higher based on lab results.

Other asked about supplements

- Omega 3
- Probiotics
- Vitamin C
- Elderberry Extract
- Quercetin
- Curcumin



Nutrition Tests

- Labs as indicated depending on diet
- Blood tests:
 - Complete Blood Count (CBC) and iron
 - Protein status lab- such as albumin, prealbumin, PQAA
 - Comprehensive Metabolic Panel (CMP)
 - 25 hydroxyvitamin D (annually)
 - Free and total carnitine
 - Essential Fatty Acid Profile (esp if very low fat diet)
 - Other nutrition labs as needed(i.e. zinc, phosphorus, iron panel, ferritin)
- DXA scan (bone health, body composition)

Dietitian's Role

- Evaluate physical nutritional status
- Help with:
 - Formula and feeding schedule to:
 - optimize nutrition
 - minimize adverse GI symptoms
 - Fluid
 - Vitamin/mineral needs
- Every 3-6 months, when younger
- Older-at least annually

Diet: Formulas/Foods

- Very individualized based on individual's tolerance and individual/parental choice.
- Many formula types/diets:
 - Breast milk
 - Formulas
 - Homemade or commercially blenderized foods
 - Amino Acid diet

Special Diets-Human Breast Milk

- **PROS**
 - Very well tolerated
 - Immune factors, enzymes
 - Can be pumped and frozen 1 yr
 - May be benefits in SMA, no research
- **CONS**
 - Added stressor to mother
 - Pumping is a time commitment
 - Reflects mother's health and intake
 - Where are you getting milk from?
 - May need to concentrate



Formula Types

- Regular (intact protein)
 - often see sensitive or versions used for spit up
 - Whey protein-easier to digest
 - Can be milk or soy-based
- Hydrolyzed
 - more hypoallergenic, protein is broken down
- Elemental (Amino Acid)
 - synthetic amino acids, some more hypoallergenic than others

Special Diets-Amino Acid Diet

- Free amino acid (elemental) formula
- Pureed baby or blenderized food
- Dairy free milk, juice, and/or breast milk
- Water
- Often need multivitamin/mineral
- Supplements
 - Specific oils to provide essential fatty acids
- Many versions-one size DOES NOT fit all!

Amino Acid Diet

– Pros:

- Lower fat diet minimizes reflux.
- Variety of foods blenderized.
- Many patients and caregivers feel it improves health and strength.

– Cons:

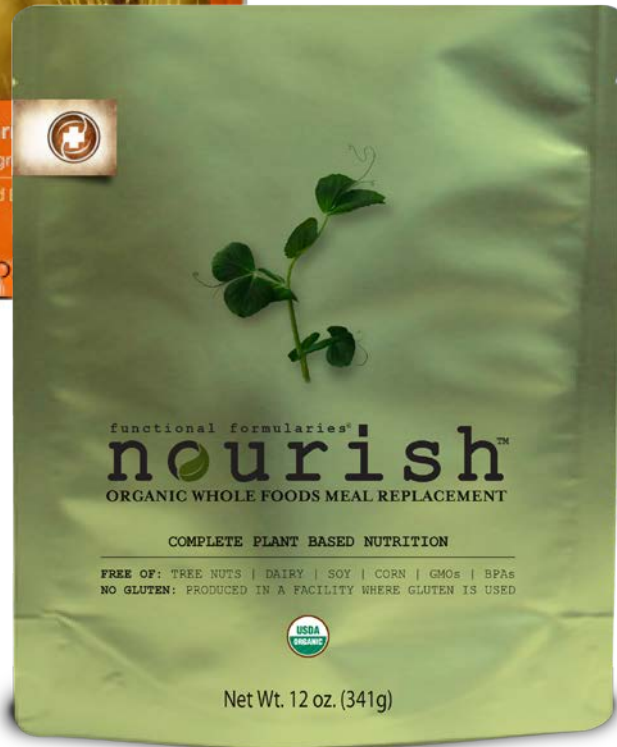
- More time/dedication to prepare.
- Limited protein/fat intake = ↑ sugar/carb intake
 - May affect blood glucose and triglyceride levels
- Amino acids rapidly digested → more frequent feeds
- May result in other nutrient deficiencies/imbbalances if not monitored closely.
- Expensive if not covered by insurance.

Special Diets- Homemade Blenderized Foods

- Can puree non-formula foods (fruit, vegetables, baby foods)
 - Blend with formula and/or water or other fluid to thin consistency. Water flushes necessary!
 - May require high powered blender
 - Food safety is a top concern-contamination
 - Day feeds only
 - Feed within short period of time
 - EXPERIMENT!
 - Resources:
 - Complete Tube Feeding- EA O’Gorman
 - <http://www.foodfortubies.org/>
 - Homemade Blended Formula Handbook-Klein & Morris



Special Diets-Commercially Available Blenderized Formulas



New Formulas-2018



Food Intolerance Signs/Symptoms

- Gagging, vomiting
- Eczema, rash
- Diarrhea
- Swollen belly, tummy ache
- Increased heart rate
- Delayed stomach emptying
- Increased secretions
- Progression of disease can increase intolerance
- Illness can increase intolerance

No Prolonged Fasting During Illness

- While well, fasting times individual
 - health and nutrition status
- Non sitters typically can fast 6-8 hours
- Sitters typically can fast 10-16 hours.

- SICK DAY PLAN
 - Decreased fasting times
 - May need to dilute formula with rehydration solution for short time.
 - May need more frequent feeds, formula change short term
 - **Seek medical attention for prolonged fasting, vomiting, diarrhea**

Fasting for Surgery

- Appropriate pre-surgical fasting time depends on:
 - Individual nutrition status
 - Strength
 - Situation/Type of surgery
 - Institutional protocol
 - Institution protocols
 - Some make first surgery of the day
 - Some give IV dextrose, monitor, and start enteral feeds ASAP
 - Some give parenteral nutrition
- Prior to surgery, discuss with care team.

Nutrition Implications with New Therapies

Observations so far:

- Spinraza may not improve lost swallow function
- BUT better digestion:
 - Less need for continuous feeding > less time on pump
 - Easier to use blended foods – able to bolus by syringe
- Overall increase in nutrient needs (calories, protein, fat) with increased strength/activity

- There is no one diet for SMA.
- Find what individual diet works best for you/your child/your family.
- Diets change over time.

Thank you!

- Break into groups for discussion/questions.