

Tube Feeding and SMA: Recommendations and Practices

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Make today a breakthrough.

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Outline

- Common nutrition issues
- Understanding growth
- Understanding nutrient intake
- Special diets & supplements
- Feeding issues and intolerance
- How a dietitian helps

Feeding and Swallowing Problems

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

Feeding tube options

- Nasogastric (NG tube)
- Nasojejunal (NJ tube)



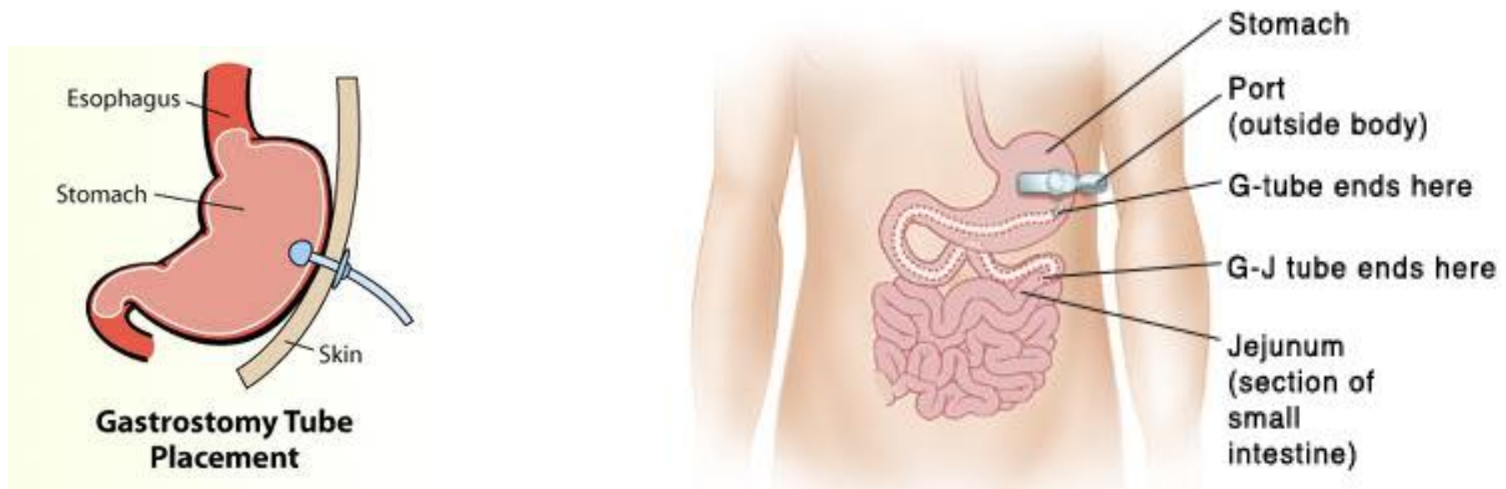
Short term

- Gastrostomy (G tube)
- Gastrojejunal (GJ tube)



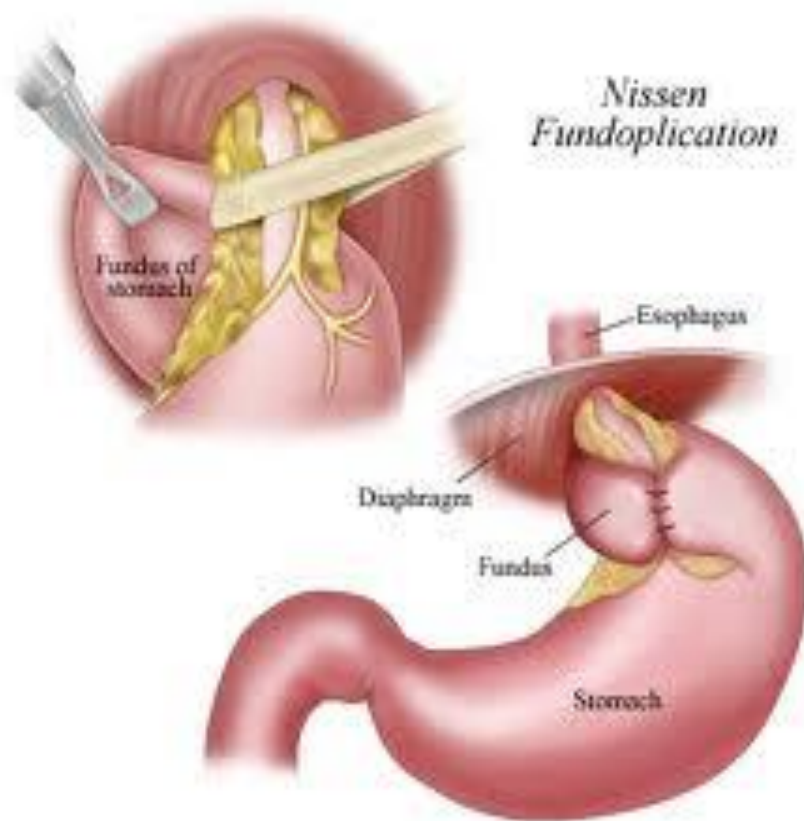
Longer term

Feeding tubes



Nissen Fundoplication

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



Feeding Terms

- Bolus-given at once, typically several feeds divided throughout day.
 - Gravity
 - Syringe
 - Pump
- Continuous- feeds given using a pump over a longer time period, i.e.
 - 20-24 hrs
 - Nocturnal feeds-overnight (8-12 hrs)
- Can be a combination of bolus/ continuous

Timing of Feeds

- Stronger SMA able to tolerate bolus feeds
 - Every 3-4 hours
- Progress to continuous, depending on symptoms
 - 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
 - Positioning
 - Smaller feed volume, more often
 - Slow feeding rate
 - Dietary modifications
 - Medicine

Constipation

- Causes
 - Abnormal gastrointestinal motility
 - Reduced intake of dietary fiber
 - Inadequate fluid intake
 - Low muscle tone of abdominal wall

Constipation

- Symptoms
 - Abdominal distention and bloating
 - Irritability
 - Sweating, red face
 - Respiratory distress

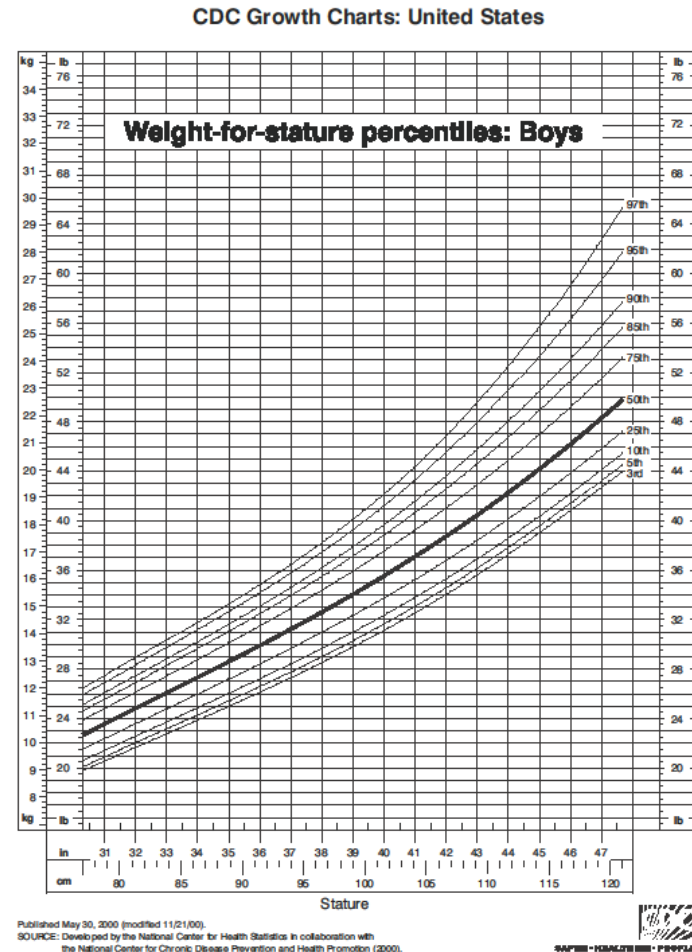
Constipation

- Solutions
 - Increase water!
 - Pureed fruits/vegetables
 - Juice- apple, pear, white grape, prune
 - Probiotics
 - Medication for constipation, GI dysmotility

FOR ACUTE CONSTIPATION MEDICATION IS
NECESSARY

Growth

- Weight
 - A growth chart is helpful to follow trends.
 - Charts are not good tools by themselves.
 - Increased fat mass
 - less muscle mass
 - >50th %ile wt for length may be overfat
 - <5th %ile wt may be appropriate



Growth

- Length-follow trend over time
- Alternate measurements if length hard to get:
 - Arm Span
 - Segmental Arm Span
 - Segmental Length
 - Ulnar length
 - Knee height

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
- Children 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 elemental formulas need at least 20% more.
- Protein sources
 - Formula “milk”
 - Blenderized foods-peas, chicken, fish
 - Supplemental protein mixes

Fat

- Too much fat can slow gastric emptying/increase reflux
- Too little fat stunts brain/eye development
- Recommended fat intakes
 - Infants-35-50%
 - Children older than 2 yrs- 20-30%
- Fat sources
 - Formula “milk”
 - Healthy fats- avocado, fish, flaxseed, plant oils
 - Carnitine Supplementation?
 - Fat transporter concentrated in muscle
 - May recommend if low carnitine levels
 - Carnitine use is not allowed for some drug trials

Vitamins and Minerals

- No research specific to SMA
- Follow Dietary Reference Intakes (DRI) for healthy children/adults
- Age and gender specific
- <http://www.iom.edu/Activities/Nutrition/SummaryDRIs/DRI-Tables.aspx>

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-very low fat diet (ADEK)
- Vitamin D, Vitamin K
- 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
- Work with a dietitian to determine supplement doses.

Sodium Intake

- Low calorie needs may mean lower sodium intake from formula
- 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
- Helps build strong bones
- Supplement or sunlight (Watch sunscreen)
- Amount depends on age and vitamin D level
- Especially need if exclusively receiving breast milk
- Check 25, hydroxy Vitamin D levels
 - Every 1-2 years, 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.

Most asked about supplements

- Omega 3's
- Probiotics
- Vitamin C
- Vitamin D
- Elderberry Extract
- Multivitamins
- CoQ10



Nutrition Tests

- Labs at least annually (more often as indicated)
- Blood tests:
 - Complete Blood Count (CBC) and iron
 - Protein status lab- such as albumin, prealbumin, PQAA
 - Comprehensive Metabolic Panel (CMP)
 - 25 hydroxyvitamin D (1-2 years)
 - Free and total carnitine
 - Essential Fatty Acid Profile (esp if very low fat diet)
 - Other nutrition labs as needed(i.e. zinc, phosphorus)
- DXA scan (bone health, body composition)

Dietitian's Role

- Evaluate physical nutritional status
- Help with:
 - Formula and feeding schedule to optimize nutrition
 - Fluid
 - Vitamin/mineral needs
 - Diet tolerance

Diet: Formulas/Foods

- Very individualized based on patient's tolerance and patient/parental choice.
- Many formula types/diets:
 - Breast milk
 - Regular>soy>hydrolyzed>elemental formulas
 - Amino Acid diet
 - Homemade blenderized diet

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 worsen tolerance

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
- Increasingly used by older children with SMA



- **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
- Soy
 - often used for lactose intolerance. If milk allergy, hydrolyzed is often best
- Hydrolyzed
 - more hypoallergenic, protein is broken down
- Elemental
 - synthetic amino acids, some more hypoallergenic than others

Special Diets-Amino Acid Diet

- Elemental formula
- Pureed baby or blenderized food
- Dairy free milk, juice, and/or breast milk
- Water
- Often need multivitamin/mineral
- Supplements
 - May include healthy oils
- 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.
 - May be inadequate in protein/fat intake.
 - May result in other nutrient deficiencies/imbbalances if not monitored closely.
 - Expensive if not covered by insurance.
 - If too high in carbohydrates, may result in blood sugar/triglyceride issues

Special Diets – 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



Common Blenderized Foods

- Stage 2 baby food fruits, vegetables
 - Wide selection, easy to use
- Spinach or kale
 - Many vitamins/minerals and low calorie
- Avocado
 - Source of vitamin K, fat
 - Higher calories



Common Blenderized Foods

- Sweet potato
 - Complex carb; source of vit A, B, C, minerals
- Legumes
 - Complex carb, fiber, minerals
 - Can clog tube
- Other fruits
 - High in antioxidants, vit C, fiber
 - Fresh/frozen-watch for added sugar



No Prolonged Fasting

- Non sitters should not fast longer than 6-8 hours
- Sitters should have feeding schedule to avoid fasting much longer than 10-14 hours.
- Fasting times may depend on health and nutrition status
- **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**