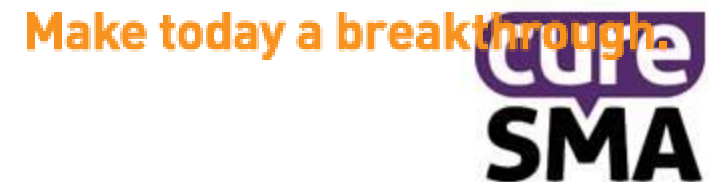


# Tube Feeding and SMA: Recommendations and Practices

2017 Annual SMA Conference  
Disney World, FL  
July 1, 2017



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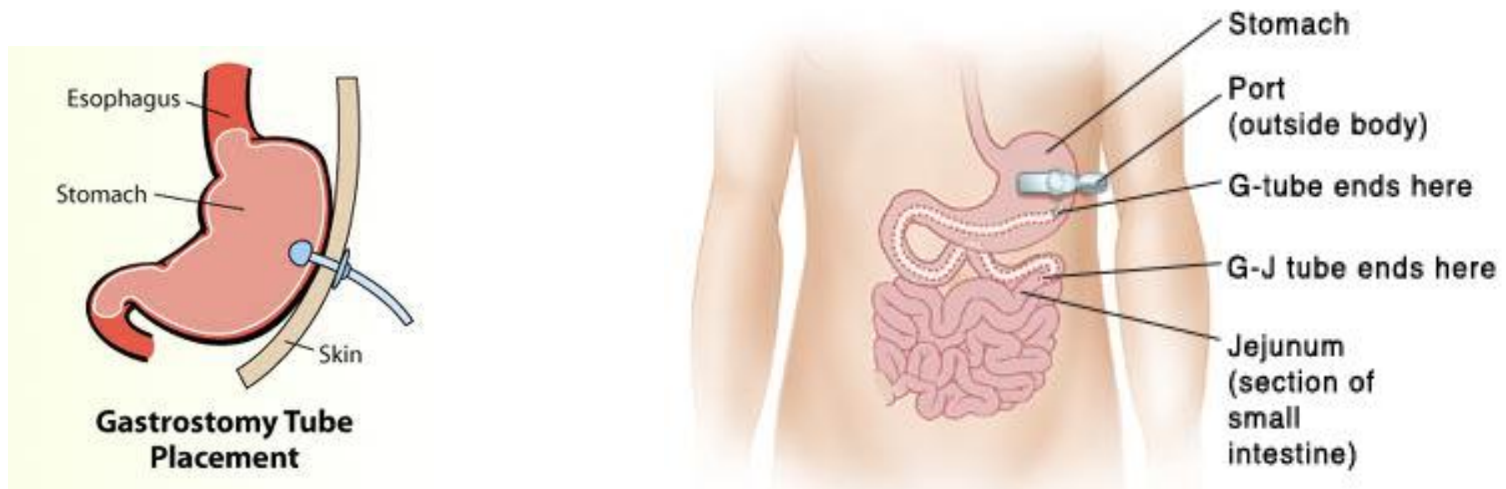
# Outline

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

# Feeding and Swallowing Problems

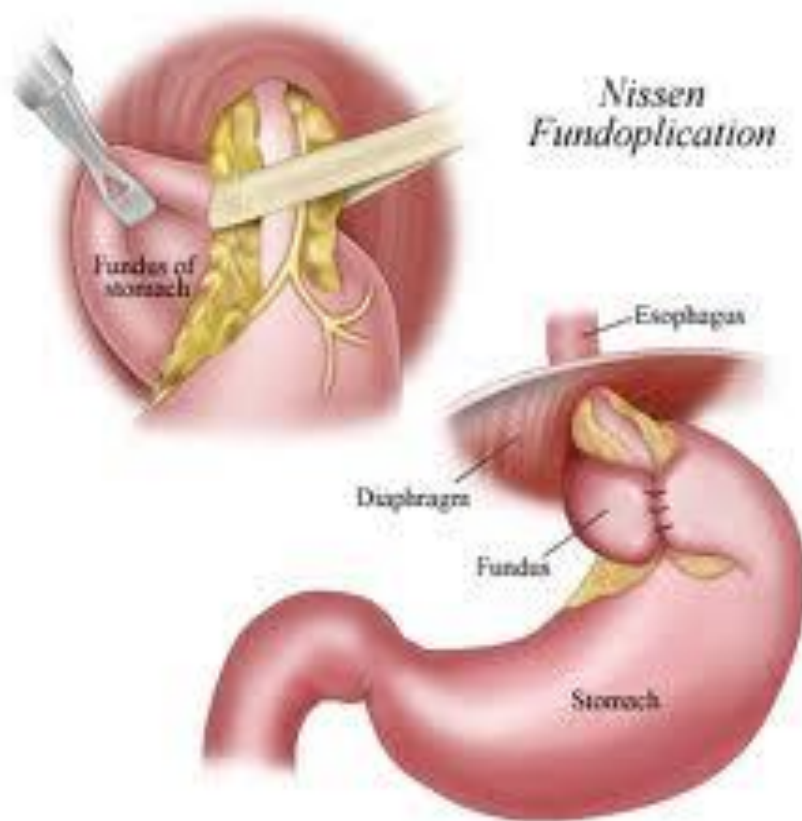
- Weak swallowing muscles
- Poor head control preventing safe swallowing
- High risk for aspiration => food/formula goes to lungs rather than stomach
- Recommend swallow study :
  - Presentation with SMA in infancy
  - Symptoms (coughing with feeds, fatigue, pneumonia, long feeding times-things change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

# Feeding 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

- 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

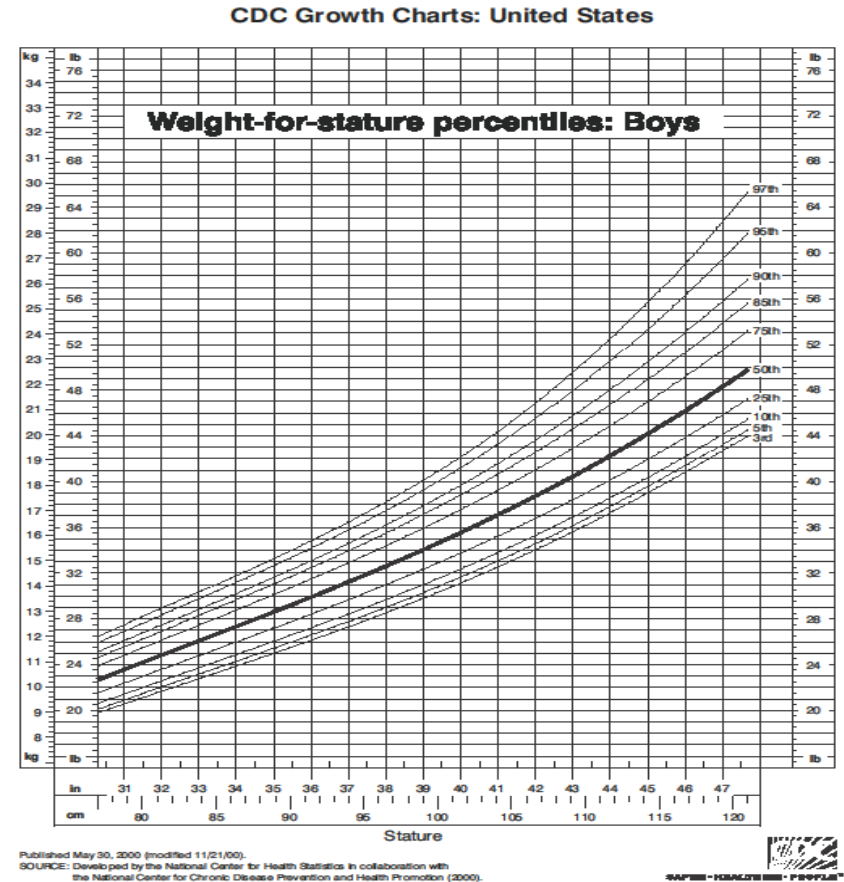
- Solutions
  - Increase water!
  - Pureed fruits/vegetables
  - Juice- apple, pear, white grape, prune
  - Probiotics
  - Medication for constipation, GI dysmotility

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



# 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 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

# 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](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
- 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



# 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 individual's tolerance and individual/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**

- 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.

# SMA Conference Survey

**Please complete your conference survey at this link:**

<https://www.surveymonkey.com/r/2017SMAAnnualConference>

**Or fill out the paper survey in your conference folder.**

- **All participants who complete a survey by 10:30 am on Sunday July 2nd, will have their name entered into a raffle for a brand new iPad!**
- **The winners will be drawn and announced on Sunday, July 2nd at the Closing General Session/It's a Wonderful Life.**