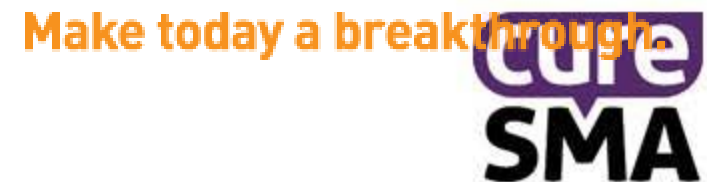


# Tube Feeding and SMA: Recommendations and Practices

2015 Annual SMA Conference  
Kansas City, Missouri  
June 20, 2015



- Erin Seffrood MS, RD, CSP, CD

American Family Children's Hospital

[eseffrood@uwhealth.org](mailto:eseffrood@uwhealth.org)

- Stacey Tarrant RD, LD

Boston Children's Hospital

[Stacey.Tarrant@childrens.harvard.edu](mailto:Stacey.Tarrant@childrens.harvard.edu)

- Rebecca Hurst Davis MS, RD, CSP, CD

University of Utah [becky.hurst@utah.edu](mailto:becky.hurst@utah.edu)

# Disclaimer

- The purpose of this presentation is not to give you a specific diet.
- Our goals are to outline the nutrition and growth information to enable you to work with your physician and dietitian to find the diet that works for your child/you.

# Outline

- Common nutrition issues
- Understanding growth
- Understanding nutrient intake
- Special diets & supplements
- Feeding issues and intolerance
- How a dietitian helps
- Different approaches to tube feeding



# 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
- Type I
  - Swallow study at dx

# Feeding tube options

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



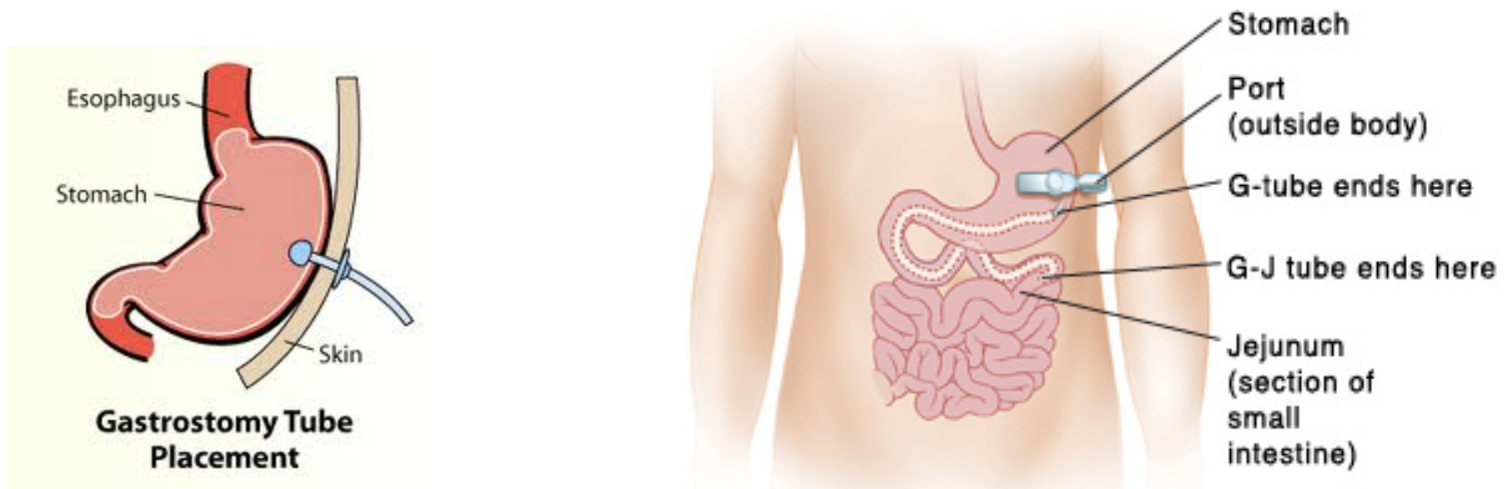
Short term

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



Longer term

# Feeding tubes



# Feeding Terms

- Bolus-given at once, typically several feeds divided throughout day
- Continuous- feeds given using a pump throughout the day. Usually 20-23 hours daily.
- Intermittent Continuous-Feeds given at a steady pump rate over a shorter time.
  - ie night time or 2 hour daytime feeds 3xday



# Gastroesophageal Reflux - Symptoms

- Spitting up or vomiting after feeds
- Chest or abdominal discomfort
- Arching back
- Bad Breath
- Refusal of feeds

# Gastroesophageal Reflux - Solutions

- Nissen fundoplication
- Positioning
- Decreasing volume of feeds
- Decreasing fat content
- 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 fluids
  - Fiber\* – but start slowly and give plenty of water!
    - Pureed fruits,veggies
  - Juice- apple, pear, white grape, prune
  - Medication for constipation, GI dysmotility

\*sometimes not well-tolerated in weaker patients

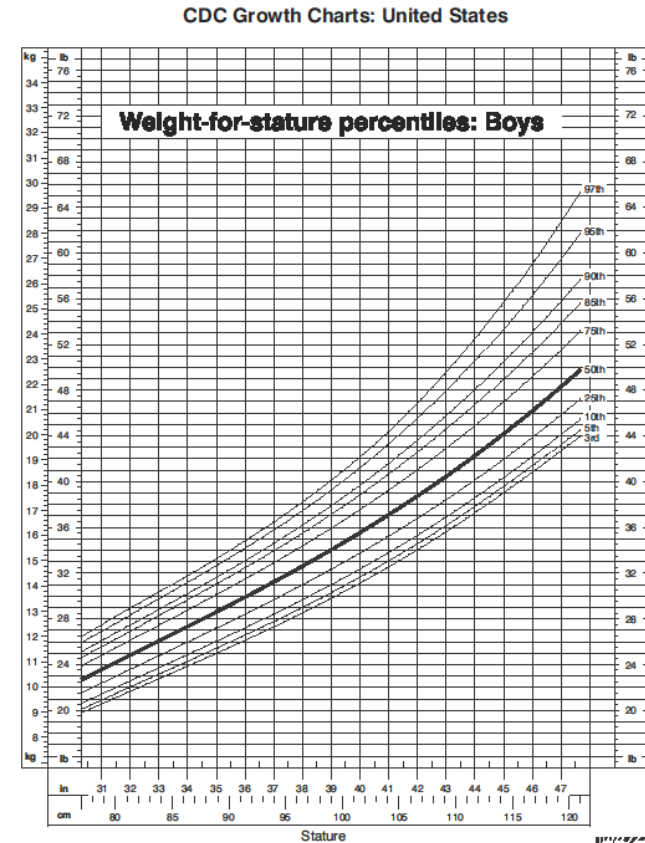
FOR ACUTE CONSTIPATION MEDICATION IS  
NECESSARY

# Growth

SMA Type I and II

- **Weight**

- A growth chart is helpful to follow trends.
- Charts are not good tools to predict over- or under-nutrition by themselves.
  - Increased fat mass
  - less muscle mass



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).



# Growth

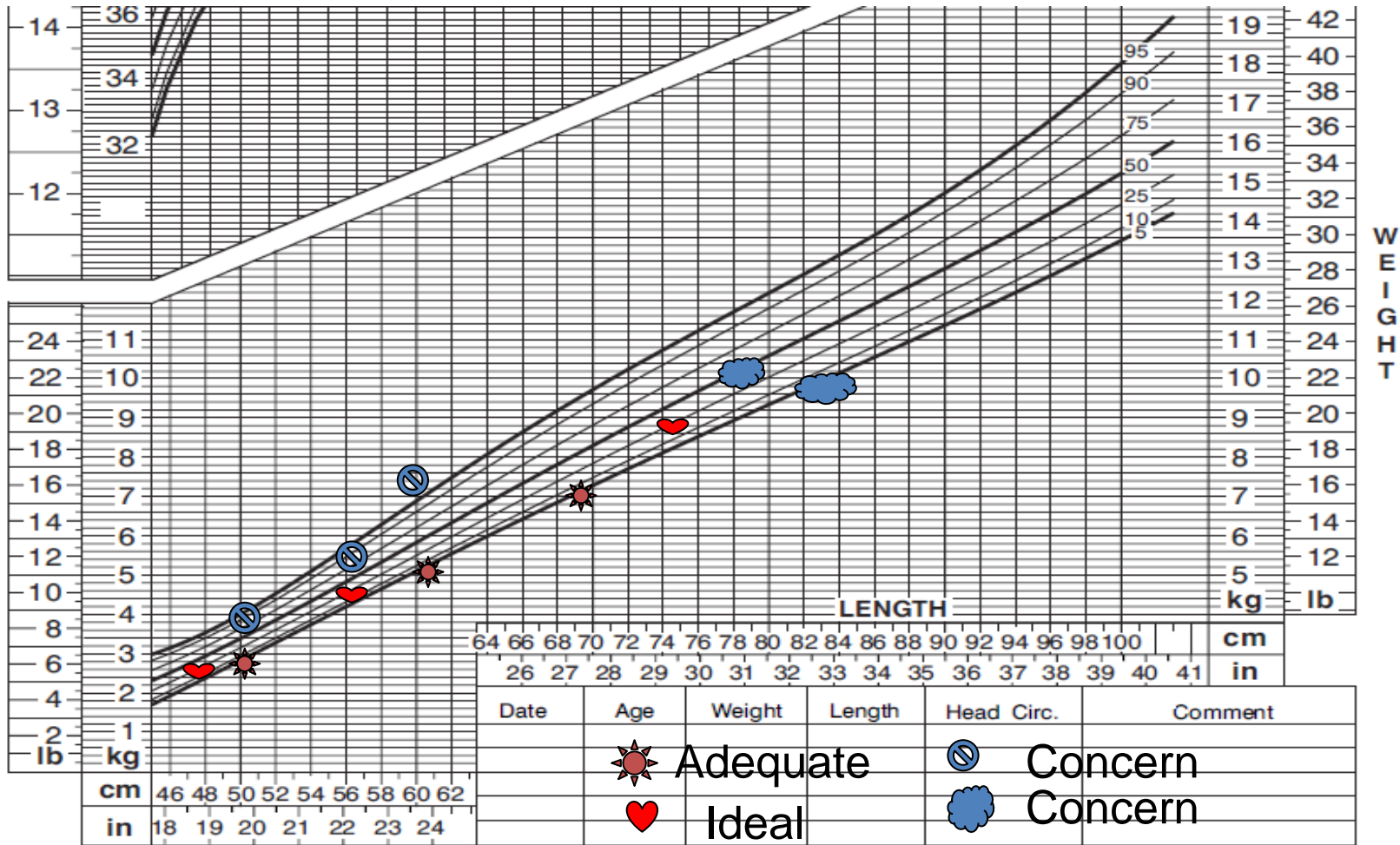
- Length
- Alternate measurements if length hard to get:
  - Arm Span
  - Segmental Arm Span
  - Segmental Length
  - Ulnar length
  - Knee height

# 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 weight checks.



# Weight for Length Examples



# Other growth measurements

- Head circumference –up to 2 years
- Body composition measurements
  - Help determine muscle/fat reserves
    - triceps or other skinfold measurements
    - arm circumference/ abdominal circumference
    - BIA
    - DXA scans

# DIET

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

# Calories

- Children with SMA need less
- Lower lean muscle mass
- Depends on activity, work of breathing, etc.
- Regular follow-up with nutritionist to adjust as needed
- Adjust 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 may require up to 20% more
- Protein sources
  - Formula, “milk”
  - Blenderized foods-peas, chicken
  - Supplemental protein mixes

# Fat

- High fat intake can slow gastric emptying/increase reflux
- Recommend that children older than 1-2 years limit fat to 20-30% of energy intake
- Infants require more fat
  - Brain/eye development 35-50% AAP
- Carnitine is given to help the body use the fat in the diet.
  - Regularly check carnitine levels in the blood.
  - Carnitine use is not allowed for some drug trials
- Fat sources-
  - formula, other “milk”
  - Healthy oils- flaxseed, canola, safflower, walnut oil

# Vitamins and Minerals

- No research in SMA
- Follow Dietary Reference Intakes (DRI)  
<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.
  - Adjusted as tolerated
- Adequate?
  - Check frequency of diapers
  - Urine color
  - Labs



# Nutrients at Risk for Deficiency

- Most common inadequate intakes:
  - Omega 3 & 6 Essential fatty acid (EFA)
  - Iron (iron deficiency anemia is common)
  - Calcium, 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.

# Salt-Sodium Chloride

- Since sodium is an electrolyte, only add if recommend by doctor/dietitian and use under medical supervision.
- Iodized table salt also contains iodine which can help increase iodine intake.
- 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
- Can buy over the counter/
- Can get from sunlight (Watch sunscreen)
- Amount depends on age and vitamin D level
- Infants-liquid vitamin D3 drops, especially need if exclusively receiving breast milk
- Check 25, hydroxy Vitamin D levels
  - Every 1-2 years, more often if low

# Most asked about supplements

- Omega 3's
- Probiotics
- Vitamin C
- Elderberry Extract
- Multivitamins
- Curcumin
- Quercetin



# 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 growth: length, weight, head growth, other body measures over time
- Help with:
  - Formula and feeding schedule to optimize nutrition
  - Fluid
  - Vitamin/mineral needs
  - Diet tolerance



# Diet: Formulas/Foods

- No studies on optimal diet for SMA
- 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,
- increased secretions, residuals
  - If bolus, check residuals before each feed
  - If continuous, check residuals as needed.
  - With SMA progression or illness, previous formulas → intolerable
  - Residuals can increase with illness.



# 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
- Supplements
  - May include healthy oils



# 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 work involved to balance diet and “feed” your child.
- May result in nutrient deficiencies if not monitored closely.
- Expensive if not covered by insurance.
- There is no research for this diet in SMA.

# 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 yet
- Increasingly, older children with SMA using



- 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
- If exclusive: Need to add Vit D; add iron 4-6 months

# Special Diets- Blenderized Foods

- Can puree non-formula foods and put through tube
  - Such as fruit, vegetables, baby foods
  - Wash fruits, vegetables well. Consider organic if not peeled (such as strawberries).
  - Blend with formula and/or water or other fluid to thin consistency. Water flushes necessary!
  - Caution: Increased food safety risk.
  - May require high powered blender
  - EXPERIMENT!



# 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

- Children with Type I should not fast longer than 6-8 hours
- Children with Type II should have feeding schedule to avoid fasting longer than 12 hours.
- Night-time feeds-good way to get supplemental calories, but not necessary!
- Reduced muscle mass-less nutrition reserves

# 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

# Sick Days

- When ill, our bodies break down substances at a faster rate.
- Especially limit fasting when ill
  - Weaker SMA- fats may break down differently in certain situations
- During illness:
  - Residuals can increase
  - Secretions can thicken and increase
  - May need short term changes
    - a more broken down formula
    - more frequent, continuous feeds
  - May need to dilute formula with rehydration solution for short time.
  - If prolonged fasting, diarrhea, vomiting, and/or fevers, seek medical attention!

# University of Utah

- After g-tube placement
  - Consider tolerance of previous formula
  - If formula intolerant, move to hydrolyzed formula.
  - Recommend formula based on tolerance, function, and type of feeding.
  - Do not recommend a no-fat formula unless also getting breastmilk or an additional formula to provide fat
  - Often dilute formula for fluid needs and tolerance

# University of Utah

- Eventually, type I needs to be on a more broken down formula
  - Symptoms
  - Parental choice
  - No benefit to early introduction
  - No harm either
  - May recommend changing feeding type, concentration, timing, moving to continuous feeds, before changing formula
  - Prefer elemental formulas for longer term feeds, not bolus.

# University of Utah

- Type I- hydrolyzed formulas, blenderized formulas with intact protein, elemental formulas.
- Type II- often hydrolyzed or blenderized formulas with intact protein.
  - Occasionally, with malabsorption issues will recommend an elemental formula for longer time feeds.

# Boston Children's

- Multidisciplinary SMA clinic day monthly:
  - Neuromuscular MD
  - Genetic Counselor
  - Pulmonary
  - PT
  - Orthopedics
  - Dietitian
  - GI



# Boston Children's

- Home Ventilation Program
  - MD
  - Nurse Practitioner
  - Registered Respiratory Therapist

# Boston Children's

- Type I: G-tube/Nissen recommended ASAP after diagnosis if consistent with goals of care.
- Formula based on tolerance – many type 1 children on intact protein formulas.
- Support families' nutrition goals .

# American Family Children's

- Type I: usually recommend feeding tubes at diagnosis (proactive approach)
  - May start with a nasojejunal feed if the feeding tube placement is delayed
  - Choose to feed into the intestine to prevent aspiration
  - When a permanent feeding tube is placed, we also perform a nissen fundoplication at the same time.
- Choose a formula based on tolerance
  - For infants this may be typically breast milk, standard infant formula
  - We aim for 45-50% calories from fat during infancy

# American Family Children's Hospital

- Type I
  - Prior to the first birthday, we would talk about different options for formulas
  - Most often go with a combination of amino acid based formulas mixed with water and possibly an electrolyte solution:
    - Tolerex \*
    - Pediatric Vivonex
  - Based on nutrient needs, we may also add pureed fruits/vegetables, juice, non-dairy milks, and oils

\*We never give Tolerex alone as it is too low in fat; often combined with Pediatric Vivonex, oil, and/or human milk; aim for 20-25% fat

# American Family Children's Hospital

- Type I
  - If you are being fed into the stomach and are strong enough, we would start with bolus feeds (given over 1-2 hours) during the day along with a continuous feed overnight
  - If you are being fed into the jejunum, you would give continuous feeds (given over 18-22 hours)

# American Family Children's Hospital

- Type II
  - If you are still eating orally during the day, we would often start with an intact, standard formula like Pediasure® or Nutren Junior® given overnight
  - If this is not well tolerated, we would then try a partially hydrolyzed formula (Pediasure Peptide® or Peptamen Junior®)
  - If you do not pass the swallow study and have trouble with reflux/volume tolerance, we would likely start with a lower fat amino acid based formula

# SMA Conference Survey

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

<https://www.surveymonkey.com/s/2015AnnualSMAConference>

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

**All participants who complete the survey will receive a raffle ticket to win an iPad! Winner will be announced on Sunday, June 21 at the Closing General Session. All completed surveys will also be entered into a drawing for a chance to win a trip, including airfare generously donated by ISIS Pharmaceuticals, to either The 2016 or The 2017 Annual SMA Conference**

