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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)
  \[\text{Short term}\]
- Gastrostomy (G tube)
- Gastrojejunal (GJ tube)
  \[\text{Longer term}\]
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
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

![Graph showing weight for length examples with categories Adequate, Ideal, and Concern marked on the chart.]

<table>
<thead>
<tr>
<th>Date</th>
<th>Age</th>
<th>Weight</th>
<th>Length</th>
<th>Head Circ.</th>
<th>Comment</th>
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- ★ ★ ★ Adequate
- ❤ ❤ ❤ Ideal
- ✈ ✈ ✈ Concern
- ✈ ✈ ✈ Concern
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!
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
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 is typically breast milk, standard infant formula
  - We aim for 45-50% calories from fat during infancy
• 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)
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.
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.