Nutricia Metabolics Webinar Series

Part II: Feeding Issues and Formula Transition in Older Children with Inborn Errors of Metabolism

May 22, 2017

Ligia Brochi, RD, CSP, LDN
Dena M. Dunn, PsyD

Children's Hospital of Philadelphia

About Us

Ligia Brochi
Registered Dietitian

Dena M. Dunn
Clinical Psychologist

Children's Hospital of Philadelphia
Pediatric Feeding and Swallowing Center
Multidisciplinary Feeding Team

The opinions reflected in this Webinar are those of the speaker and independent of Nutricia North America.

NUTRICIA Metabolics Inspiring Futures

Objectives

- Identify possible feeding issue in patients with Inborn Errors of Metabolism (IEM)
- Outline psychosocial feeding challenges for patients with IEM
- Provide behavioral strategies for dietitians and families to increase children's acceptance of formula or food by mouth
- Identify when to refer patients with feeding disorders to a multidisciplinary team
- Discuss a case report and relate learning to one's own practice

Metabolism & Genetic Disorders
Medical Nutrition Therapy

- Disorders of Protein Metabolism:

  PKU, HCU, Tyrosinemia, GA, PA, MMA, Cobalamin defects, IVA, MSUD, Urea Cycle Disorders

- Fatty Acid Oxidation Disorders

  LCHAD, VLCAD, CPT, TFP, CACT

- Carbohydrate Metabolism

  Galactosemia, HFI

Diagnosis and Life Course

- Newborn
  Newborn Screening
- Infancy
- Childhood
- Later in life

Early diagnosis and intervention of most of these disorders leads to a better outcome

Physiologic Feeding Challenges of Metabolic Patients

- Poor sucking reflex
- Vomiting / intolerance
- Breathing / Cardiac insufficiency
  (i.e. need for supplemental oxygen)
- Lethargy
- Oral motor delays
- Anorexia
- Poor growth
- Muscle hypotonia
- Cognitive Impairment
- Vision / hearing loss

Nutritional Challenges: Formula Intake

- Multiple formulas
- Mixing instructions
- Large volumes
- Taste fatigue
- Infant bottle vs. cup
- Emotional attachment

Nutritional Challenges: Strict Diet

WHAT

- Formula requirements
- Log food intake
- Increase calories of foods (growth)

WHEN

- Avoid long periods of fasting (catabolism)

HOW MUCH

- Weighing / measuring food

Psychosocial Feeding Challenges of Metabolic Patients

1. Meal planning and preparation
   - Specific nutrition regimen (weighing food, reading labels, food diary, medications)
2. Patient compliance
   - School-aged children often eat meals outside of the home without strict adult supervision
3. Parent child relationship stressors
   - Rebellion and testing limits is developmentally typical at this age; children may choose to disobey in the form of regimen refusal

Feeding challenges for typically developing school-aged children

- Food preferences
- Peer comparison and social referencing
- Social norms about eating
- Body image
- Context for eating: lunch room, time limited, etc.
- Stigma

Risk factors for developing feeding difficulties

Complex nutritional needs for metabolism patients

+

Developmentally typical feeding and eating behavior for school-aged children

+

Biopsychosocial variables =

Increased risk for feeding difficulties and disordered eating

Formula Transition in Older Children

Age 1-3 years, or later if calorie intake from foods is insufficient

High potential for development of feeding disorders at this phase

- "Cold turkey"
- Offer different cup
- Add flavoring
- New formula mixed into water, sport drinks, or current formula (i.e. Phenylade GMP Mix-In)

Periflex Early years (20 g PE)	=	Periflex Early Years (10 g PE) 1 pack of Phenylade GMP Mix-in (10 g PE)

- Titrate slowly (mix old formula with new formula)

Formula Transition in Older Children: Slow Titration

Titrating current formula (F1) to new formula (F2):

- Maintain same total grams of protein

Step1: 75% F1 : 25% F2

Step 2: 50% F1 : 50% F2

Step 3: 25% F1 : 75% F2

Step 4: (goal) 100% F2

- Watch decrease in total calories
  - Risk for Metabolic Decompensation
  - Patients with Feeding difficulties may need modular added
  - Keep longer periods on each step

Optimize Formula acceptance: Nutrition Strategies

- Add flavoring (i.e. tang, syrups, extracts, fresh fruit)
- Smaller frequent servings
- Mix formula with an acidic agent to counteract the bitter amino acid taste (i.e. lemon pudding, lemon sorbet)

Optimize Formula acceptance: Nutrition Strategies (cont'd)

- Add a splash of seltzer for carbonation, bubbles may help (e.g. sparkling citrus flavored beverages)

- Immediately after formula, offer sweet or tangy fruit of preference to neutralize the after taste (i.e. pineapple; maraschino cherries, starburst, lollipops, fruit leathers ups, frozen grapes/mangoes, sour candy)

Optimize Acceptance of New Foods: Nutrition Strategies

- Roasting vegetables accentuates their natural sweet flavor
- Sour counteracts bitter flavor: add lemon juice and zest to vegetables
- Vary texture or temperature (e.g. some kids who don't eat cooked peas will eat them frozen out of the bag)

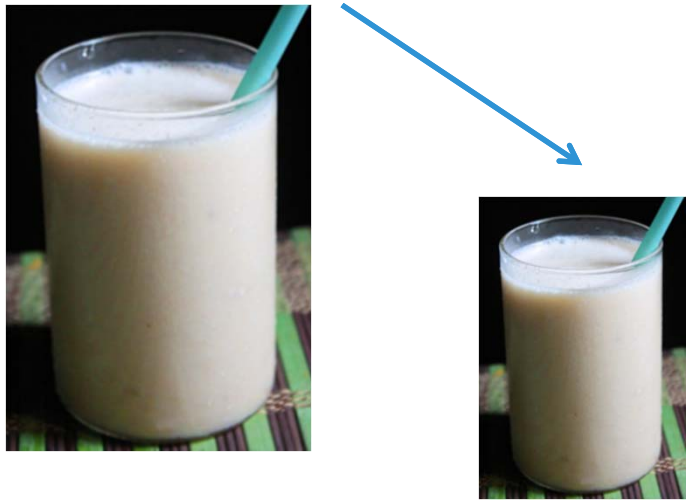
Optimize Acceptance of New Foods: Nutrition Strategies (cont'd)

- Be creative! Make food visually appealing, and vary visual presentations
- Involve children in menu planning and meal preparation
- Use "appetite" to your advantage when introducing new foods: start dinner with 2 bites of non-preferred foods, and save favorite foods for last, set a meal structure

Structured meal time

- Decrease volume of formula to allow time for meals

- Adjust schedule:



Meal #1 + Formula

break (2-3 hours)

Meal #2 + Formula

break (2-3 hours)

Meal #3 + Formula

break (2-3 hours)

Formula – boost calorie as needed

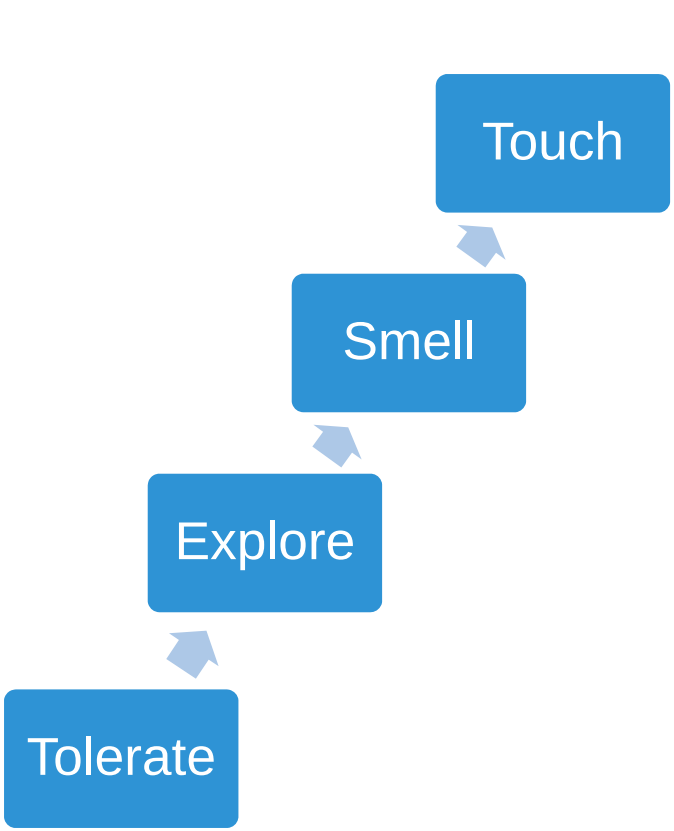
Behavioral Strategies to Minimize Oral Aversion

- Patient education and honesty (Try not to trick young children into eating or drinking)

- Family “buy in” and positive attitude
- Modeling positive feeding behavior
- Developing reward systems or sticker charts
- Providing praise and attention for positive feeding behavior and ignore negative mealtime behaviors (i.e. spitting, refusing, making excuses)

Behavioral Strategies to Minimize Oral Aversion

Sensory Feeding Progression



Taste

- **Kisses food**
- **Licks lips, licks food**
- **Bites pieces, spits out immediately**
- **Bites pieces, holds in mouth for a short time (a few seconds) and spits out**
- **Bites, chews a few times, and spits out**
- **Chews and swallows with drink**
- **Chews and partially swallows**
- **Chews and swallows independently!**

Strategies to Optimize Food Acceptance

- Have at least one preferred food on the plate
- Limit the desperate action of offering too many options
- “You don’t have to like it. You just have to taste it”
- Don’t give up, continue exposing to a wide variety of flavors
- Keep presenting, NOT pushing
- Consider giving some control (i.e. taco night)

When feeding becomes disordered: When do I refer out?

- Is feeding or eating significantly affecting the patient's ability to comply with nutrition recommendations?
- Are you as a solo practitioner (RD) struggling to manage a case because of behavioral or social issues or challenges with compliance?

When feeding becomes disordered: When do I refer out? (cont'd)

- Does the patient or family seem stressed or unable to cope beyond what is "normal" or expected?
- Has the child's feeding behavior significantly changed recently (i.e. Increased refusal, decreased acceptance)?

Multidisciplinary treatment for feeding disorders

- Feeding team evaluation and consultation
- Oral motor feeding therapy (speech language pathologist)
- Outpatient behavioral feeding therapy (psychologist)
- Partial hospitalization/Day hospital program
- Inpatient treatment

Case Study

“Katie”

2 year old female with a Disorder of Amino Acid Metabolism

Recurrent Metabolic decompensation, Developmental Delay, Recurrent otitis media, and NG-tube supplementation

Anthropometrics:

Weight: 15.1 kg >95th% Height: 89.1 cm 75-90th% BMI: 19.0 Kg/M2 90-95th %

Feeding concerns:

- Limited and Inconsistent oral acceptance
- Has selected more foods out of her diet post hospitalization

Katie was referred for a comprehensive feeding assessment by a multidisciplinary feeding team consisting of medical, nutrition, occupational therapist, speech pathologist and psychologist

Case Study

Current Diet:

10 grams of protein from foods + nurses twice a day ~ 6-7 oz (2 grams protein)

Formula: F1 + water = 17 ounces

SUGGESTED SCHEDULE

Meal#1 (8 am): Offer foods (3 g protein) in high chair for 10 min + 3 oz Formula (sippy cup)

Meal#2 (12 pm): Offer foods (3 g protein) in high chair for 10 min + 3 oz Formula

Snack (3 pm): Offer foods (1 g protein)

Meal#3 (6 pm): Offer foods (3 g protein) in high chair for 10 min + 3 oz Formula

Bedtime: 8 oz Formula

Case Study

Current Diet:

10 grams of protein from foods + nurses twice a day ~ 6-7 oz (2 grams protein)

Formula: F1 + water = 17 ounces (25 cal/oz) (430 kcal, 15 g PE)

Slowly transition from F1 to F2:

Step 1: F1 (12 g PE) + F2 (3 g PE) + water = 17 oz
Step 2: F1 (10 g PE) + F2 (5 g PE) + water = 16 oz
Step 3: F1 (9 g PE) + F2 (6 g PE) + water = 15 oz
Step 4: F1 (7.5 g PE) + F2 (7.5 g PE) + water = 14 oz
Step 5: F1 (5 g PE) + F2 (10 g PE) + water = 12 oz

New plan: F2 + water = 8 oz (25 cal/oz) (205 kcal, 15 g PE)

- When Katie stops nursing, increase protein from food goal to 12 grams per day.
- Continue to provide whole cow's milk or Pediasure, 1 ounce = 1 gram protein, to ensure Katie meets her daily protein allowance.
- If Katie is consistently unable to meet her daily protein goal, will consider add nonfat dry milk powder or cow's milk to the daily formula batch

Case Study

Oral Feeding

- Continue with her current diet of soft solids, and chewable textures.
- Begin to slowly transition Katie from the bottle, and suggest giving her morning formula via the sippy cup

Self Feeding

- Continue to allow Katie the opportunity to progress her self feeding skills by allowing her to use utensils during low stress times such as snack time.
- Continue to feed Katie during meal times and provide her with her own spoon to practice self feeding skills.

Behavioral Feeding Recommendations

- Establish a meal time routine for Katie of 3 meals of day
- Limit mealtimes to 10 Minutes. The time expectation for Katie can start at a 10 minutes and increase as Katie's tolerance for meal time increases
- Eliminate feeding during play times when Katie is not sitting in a chair
- “Catch Katie being good at mealtimes:” Provide praise for Katie when demonstrates positive feeding behavior and ignore any negative behaviors

THANK YOU!

Questions?

NUTRICIA Metabolics Inspiring Futures

Feedback, Please!
Certificate of Attendance

For those interested in obtaining a Certificate of Attendance:

1. For viewers of the live Webinar: a link to a survey will pop-up as you exit.
2. If not, please go to: https://www.surveymonkey.com/r/FeedingIEM
3. Complete the survey and an event code will be available at the end of the survey.
4. Go to www.NutriciaLearningCenter.com and enter the event code.
   Your certificate will be automatically emailed to you.

For question on this Webinar or Nutricia's products, please email:

NutritionServices@nutricia.com

or call:

1-800-365-7354 (option 2)