

Nutricia Metabolics Webinar Series



3 PART WEBINAR SERIES:

**FEEDING &
DIET PRACTICES**

Part One:
***Infant and Toddler
Feeding in IEM:
A Time of Transition***

Infant and Toddler Feeding in Inborn Errors of Metabolism: A Time of Transition

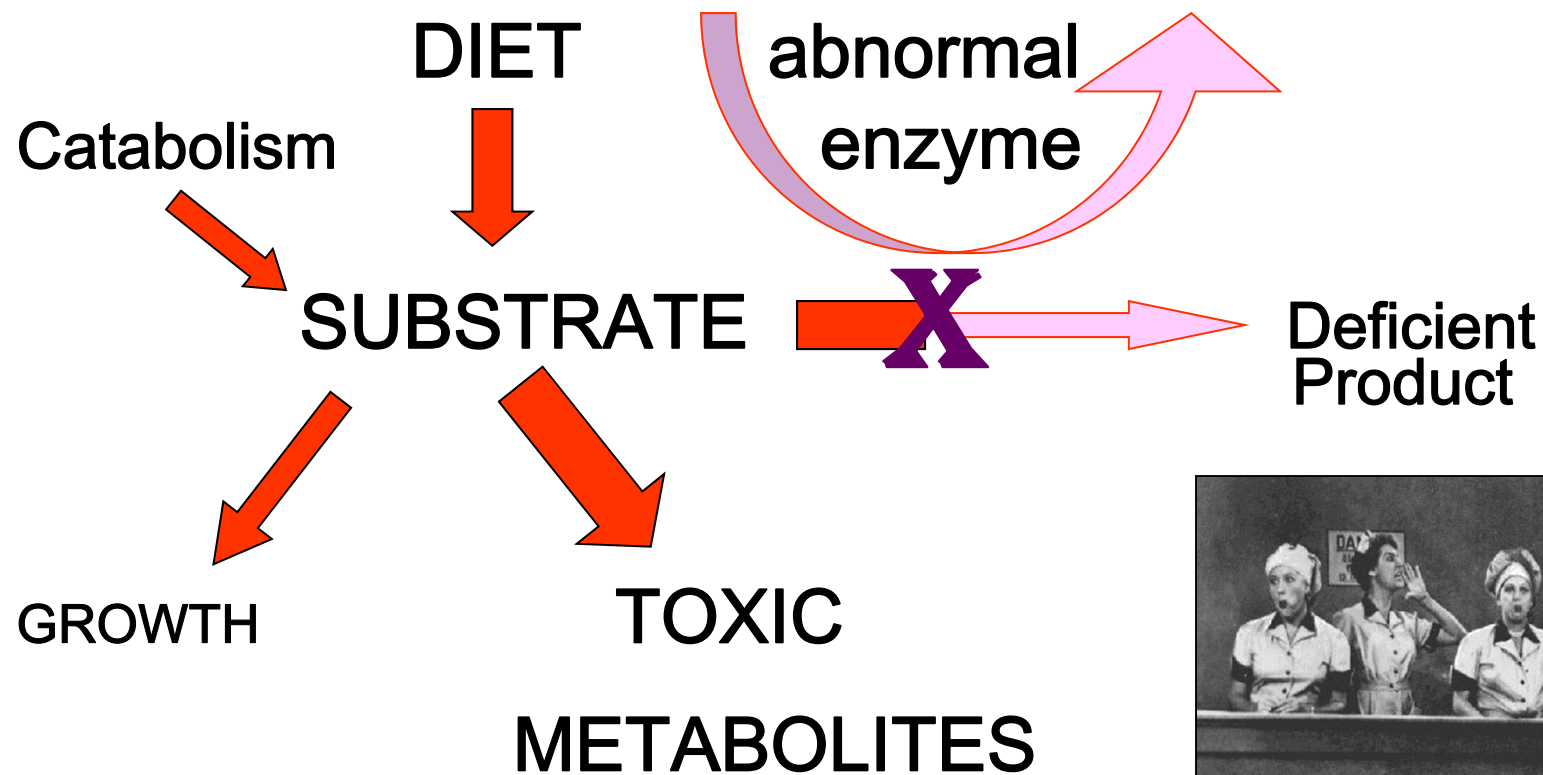
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The opinions reflected in this Webinar are those of the speaker and independent of Nutricia North America.

Abnormal Metabolic Pathway





Counseling the Family



New Diagnosis

- First contacts → lasting impressions
- *Time and repetition* → acceptance and competence
- Good communication
- Mentor families – support groups – resources



Counseling the Family



Ongoing

- More listening than talking – non-directive questions
- Family circumstances matter
- Uncomplicated explanations and instructions
 - “Avoid unnecessary exactitude”
 - Clear – straightforward – realistic

Keep “Mom” Calm



Instill confidence – form a partnership



The First Prescription



Recommended Intake of Protein and Energy for an Infant

AGE	Protein (g/kg/day)	Energy <i>DRI</i> (Kcal/kg/day)
0 to <3 mo	2.5-3.0	95-145
3 to <6 mo	2.0-3.0	95-145
6 to <9 mo	2.0-2.5	80-135
9 to <12 mo	2.0-2.5	80-135
1 to <4 years	1.5-2.1	900-1800

Adapted from Acosta and DRI

- Protein requirements higher when medical food is primary source
- Refer to GMDI Guidelines and Toolkits

Translating the Diet Prescription



For the infant

- Intact protein – provides affected substrate for anabolism without exceeding enzyme capacity
 - May be <25% of normal intake
 - Not too much and not too little
- Medical food – provides remaining protein and calories free of substrate & supplements enzyme product
 - Amino acid based – free of whole protein
 - Protein hydrolysates or GMP based for some disorders
 - Protein free – CHO and fat for Kcalories
- Mixed to provide adequate fluid
- Space feedings throughout day



Hints from Experience: Feeding Infants



Bottle feeding

- Round all final numbers
- Weighing powder – most accurate
- **Or *carefully*** measure scoops/Tbsp/tsp
 - Understanding of “household measurements”
- Make formula volume to even number mL or fl oz
- Early rapid growth requires frequent formula adjustments
 - Educate mom about what to expect
 - Assure mom you will be guiding formula RX



Know Formula Choices: Infant and Medical Food



- Nutrient profiles*
 - Amino acids, protein, calories, vit/min, DHA/ARA
 - MCT content for FAODs
 - Higher for VLCADD, LCHAD – lower for MCADD
 - Powder or ready-to-feed
- Begin with current infant formula when possible
 - WIC formulary where appropriate
- Infant formula changes – partner with pediatrician
 - Soy or more elemental formula
 - Resist adding cereal if restricting protein – HBV formula protein replaced with lower BV cereal protein

* See GMDI Guideline tables

Breastfeeding and Metabolic Conditions



1. Calculate breast milk amount for grams intact protein or mg individual amino acid allowed (ex. mg Phe)
2. Calculate calories in amount of breast milk from #1
3. Calculate medical food for remaining calories needed
4. Be sure protein needs are also met
5. Calculate water to dilute medical food to 20 cal/oz
5. Number medical food bottles/day = $\frac{\text{daily volume medical food (from \#4)}}{\text{usual ounces/feeding}}$

Breast Milk Estimates



Volume Breast Milk for Age

Age (mo)	Breast milk (mL) (average)	Breast milk (mL) (range)
0.2	640	510–740
0.5	660	530–780
1.0	700	560–840
2.0	750	610–920
3.0	830	670–1,000
4.0	900	720–1,060
5.0	940	760–1,160
6.0	1000	790–1,200

Nutrients Per 100 ml Breast Milk

	Colostrum (1–5 d)	Transitional (6–10 d)	Mature (>10 d)
Energy(cal)	67	74	75
Protein (g)	2.3	1.6	1.1
Phe (mg)	104	63	41

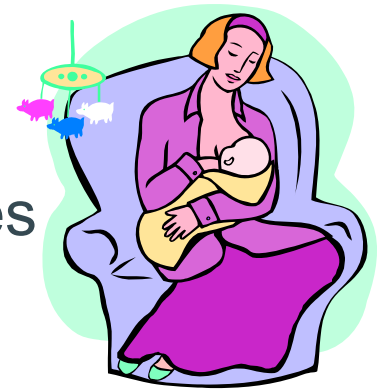
Tables from Greve,
JADA 1994

Breastfeeding



Two ways to do it:

1. Pump breast milk & add to medical food
or alternate medical food and breast milk bottles
 - + nutritional benefits
 - lose ease of nursing from breast
2. Feed from breast – alternate breast and medical food bottle feedings **or** start each feeding with medical food bottle, finish with ad lib from breast
 - + nutritional benefits, ease of nursing
 - less precise, frequent monitoring



Hints from Experience: Feeding Infants



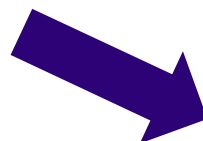
Breast feeding

- Establishing breast feeding is hard – support mom
- Milk production responds to feeding frequency and length
- Breast milk ↑ with ↑ ad-lib feedings – accommodates intact protein needed for growth
- Success depends on:
 - Infant's tolerance – acuity of disease
 - Compliant mom
 - Intake imprecise – so need frequent labs & weights
 - Reliable interaction with RD

Transitioning



How to get from here



To here?



Transitioning the Diet Prescription



1. Gradually add intact protein from foods: cereals, fruit, vegetables
2. **AND** reduce intact protein from infant formula by equal amounts
3. Medical food remains major source of protein & calories – nutritional adequacy

BABIES AGE:	WHEN BABIES CAN:	SERVE:
Birth through 3 Months 	• Only suck and swallow	LIQUIDS ONLY • Breastmilk • Infant formula with iron
4 months through 7 months 	• Draw in upper or lower lip as spoon is removed from mouth • Move tongue up and down • Sit up with support • Swallow semisolid foods without choking • Open the mouth when they see food • Drink from a cup with help, with spilling	ADD SEMISOLID FOODS • Infant cereal with iron • Strained vegetables* • Strained fruit* <i>*may be started later in the age range</i>
8 months through 11 months 	• Move tongue from side to side • Begin spoon feeding themselves with help • Begin to chew and have some teeth • Begin to hold food and use their fingers to feed themselves • Drink from a cup with help, with less spilling	ADD MODIFIED TABLE FOODS • Mashed or diced soft fruit • Mashed or soft cooked vegetables • Mashed cooked beans or peas • Pieces of soft bread • Crackers • Breastmilk, iron-fortified formula, or fruit juice in a cup

➔ **Formula – metabolic Rx**
8-12 feedings, 2-6 oz/feeding

➔ **Formula – metabolic Rx**
4-6 feedings, 4-8 oz/feeding
Begin Solids
Cereals: baby food to soft cooked
2-3 servings, 1-2 Tbsp/serving
Fruit/vegetables: plain strained baby food
to mashed/very soft table food
2-3 servings, 1-2 Tbsp/serving

➔ **Formula – metabolic Rx**
3-4 feedings: 7-8 oz/feeding
Increase Solids
Starches: baby food or soft cooked
cereals, soft breads and starches
4 servings, 1-2 Tbsp/serving
Fruit/vegetables: mashed or soft fruit and
vegetables, soft finger foods
4 servings, 2-3 Tbsp/serving

Introducing Solids – When?



Readiness

- 4-6 months
- Holds head steady – sits upright with support
- Absent tongue thrust when spoon placed on tongue
- Watches others eat
- Reaches for food
- Opens mouth for food



Introducing Solids: How to Start



Baby food

- Test readiness with very low protein fruit or veggies (ex. applesauce, pears)
- Dry baby food cereal – rice less allergenic
 - 1 Tbsp $\approx$ 0.3 g protein
 - Mix with formula, water
 - Satiety
- Strained fruit/vegetables
- Spread intact protein sources through day



Introducing Solids: How to Continue



Progress to allowances

- Transition responsibility
- More complicated for parent – individualize
 - Count mg AA or g protein
 - Label reading
 - Food records
- Provide resources



**Goal of feeding in 1st year of life is to teach eating skills
– formula still provides nutritional needs**

Introducing Solids: How to Continue



Baby food

- Advance variety as tolerated
- Introduce one new food *or* texture at a time
- Combine “new” food with well liked foods
- Discourage “mixed meal” jarred foods



Introducing Table Food



Table food

- 9 – 12 mo
- Increase food textures as tolerated
 - Cooked cereals – mashed fruit, vegetables
 - Very soft “chunky” foods
- Encourage self feeding
- Introduce finger foods
 - Interesting, develops motor skills, allows choices
- Avoid choking hazards – can’t be “gummed”
- Low protein specialty foods – for acceptance



Transitioning Medical Foods



- 1st level appropriate for infant's rapid growth
- 2nd level appropriate for child's slower growth
 - Fewer calories and higher protein/g powder

When to change – *suggestions*:

- Avg. 1-3 years of age
- Infant formula has been discontinued
- Continue 1st level if growth compromised
- Change to 2nd level if need less volume
- Consider GMP after 1 year – better nutrient profile?
- Individualize!

Transitioning Medical Foods



How to change – *options:*

- Titrate – keep total grams protein or calories same
 - Incremental exchange of 1st level for 2nd level
 - “Cold turkey”
- Mom is partner – infant acceptance decides
- Individualize

Weaning From the Bottle to a Cup



Hints from experience:

- Formula may taste different in cup
- Closed cup to minimize smell
- Colder temperature may be more acceptable
- Use cup for formula only – not water
- Encourage – but child and parent are partners



Feeding Solid Foods: Hints from Experience



- It will be messy!!
- “One bite rule” – don’t have to count
- Portioning – weighing most precise but accurate measuring usually enough
- Low appetite – offer food before formula
- Learn satiety cues
- Don’t force feed
 - Recommend temporary “higher” foods
- Reassess and review at 6 month visit



A Simplified PKU Diet for Very Young Children



- Can start when introducing solids
- Goal – decrease Phe from intact protein by $\approx 30\%$
- Don't count “free foods”
 - Most fruit & vegetables, low protein foods
 - Phe <75 mg/100 g

Example:

1. Incrementally add ≈ 15 mg Phe from food
(ex. 1 Tbsp rice cereal)
2. *AND* subtract 20 mg Phe from formula
3. Until formula only contains medical food
4. Monitor blood Phe frequently



Communication Bernstein and Rohr, and GMDI Guidelines



Case Study: GW starting solids



6 month old male with PKU

- Early and well treated
 - Current blood Phe 250 $\mu\text{mol/L}$
 - Historical blood Phe range 200-640 $\mu\text{mol/L}$
 - Normal growth – 75th %
 - Current wgt: 8 kg
-
- Formula meets recommended nutrients for age
 - Phe 300 mg, protein 20 g, 800 kcal, 20 kcal/fl oz
 - Frequent illness from day care exposure + vomiting
 - GI eval: hyperactive gag and esophageal irritation
 - Rx for meds, change to gentle ease formula



Case Study: GW starting solids



Readiness: sits upright with support, automatic tongue thrust absent, interest in other's food

- Transition of Phe from formula to food:
- 1 Tbsp dry rice cereal morning and night (34 mg Phe)
- Infant formula reduced by 34 mg Phe
- Medical food increased for total protein and calories
- Started strained applesauce or pears
- Gag reflex and vomiting better, but remain



Case Study: GW starting solids



10-12 months of age

- Titrated Phe from baby foods – 150 mg/d (50% allowance)
- Refused further *reliable* increase
- Only baby food cereal, limited strained fruits/vegetables
- ST for delayed speech and OT for feeding aversion
- Refusal of “chunky” or finger food
- Some hyper-active gag reflex and reflux remain
- Good growth and large motor skills



Case Study: GW starting solids



18 months of age

- OT – chewing and swallowing techniques, self feeding skills (finger food, spoon) – progress
- *Continue to:*
 - Increase Phe from food (by 5-10% increments)
 - Advance food texture and increase variety
 - Low protein pasta for taste acceptance
 - Increase Phe density
 - (ex. combine low protein pasta with “regular” pasta)
 - Nutritional needs from infant formula + medical food

Case Study: GW starting solids



Evaluation

- Good growth and large motor skills
- Delayed speech and feeding skills
 - Poor acceptance of new food tastes and textures

Plan

- Postpone transition to toddler formula
 - Nutrition support despite inconsistent food intake
- Postpone weaning to cup
 - First achieve current feeding goals
- Evaluation for autistic spectrum



Special Considerations



- Chronic reflux risks esophageal inflammation/erosion
 - Causes formula or food refusal
 - May need GI evaluation, medications
- Taste, texture, fluid aversions common in some disorders
 - Ex. MMA, PA
- Anxiety in parents increases with severity of disorder and risk of metabolic consequences

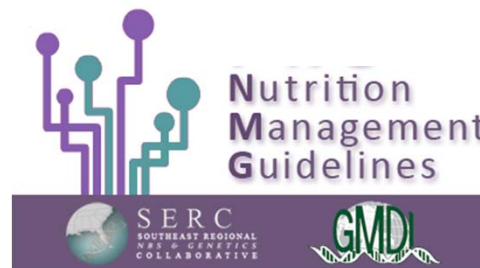


Resources



**See GMDI Guidelines and Toolkits for
References and Resources**

www.GMDI.org or www.sercc.org



THANK YOU!



Questions?

Feedback, Please!

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