

NUTRITION MANAGEMENT OF CHILDREN AND ADULTS WITH EPIDERMOLYSIS BULLOSA

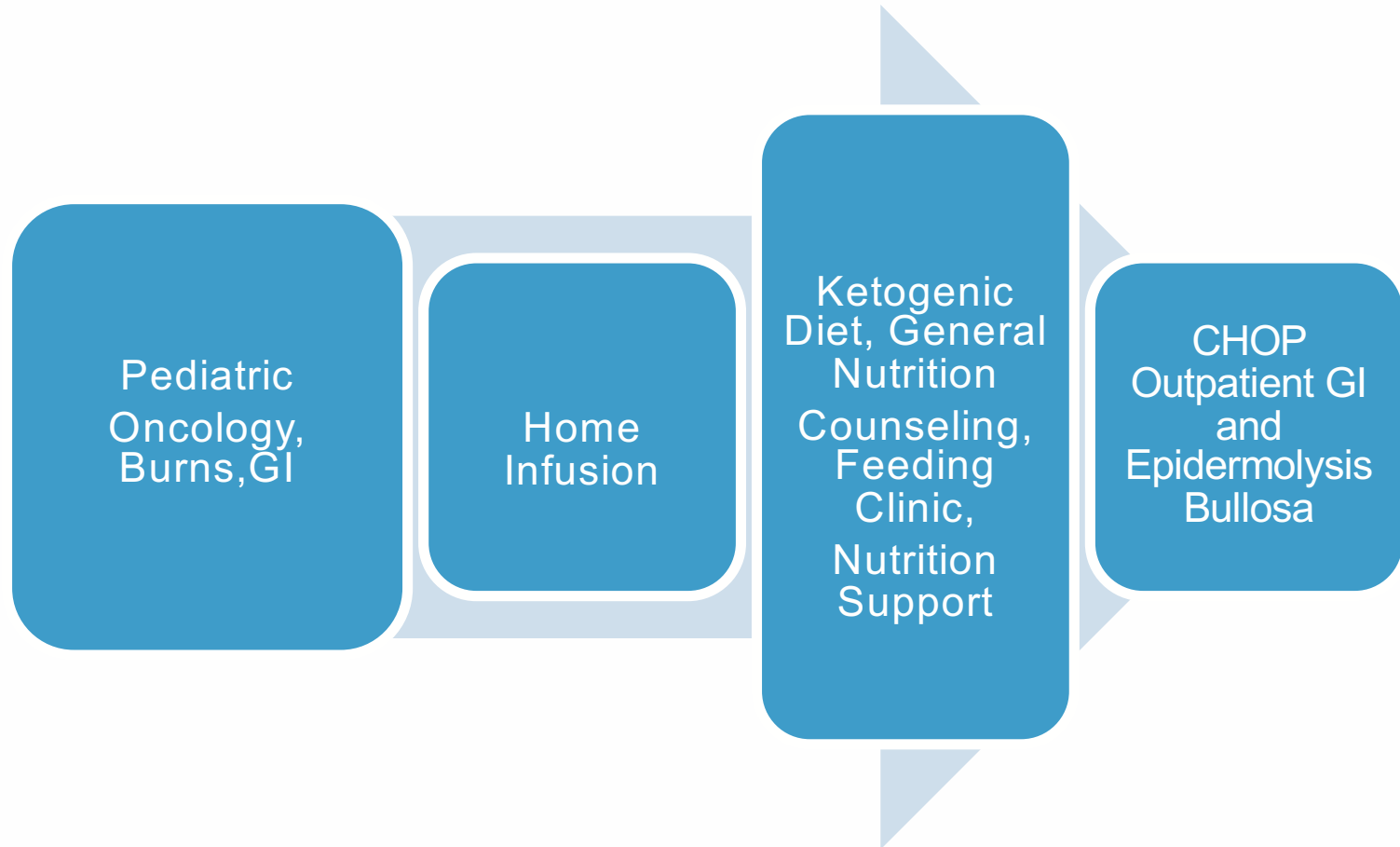


Colleen Vicente, RD, LDN, CNSC

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MY BACKGROUND



TOPICS

- Calorie and Protein Requirements
- Oral Nutrition Supplements
- Tube Feedings
 - Blenderized Tube Feedings
- Vitamins and Minerals
- Anti-Inflammatory Diet



WHY IS NUTRITION SO IMPORTANT FOR EB PATIENTS?



Promote adequate growth and development

Provide nutrients for proper healing of skin lesions

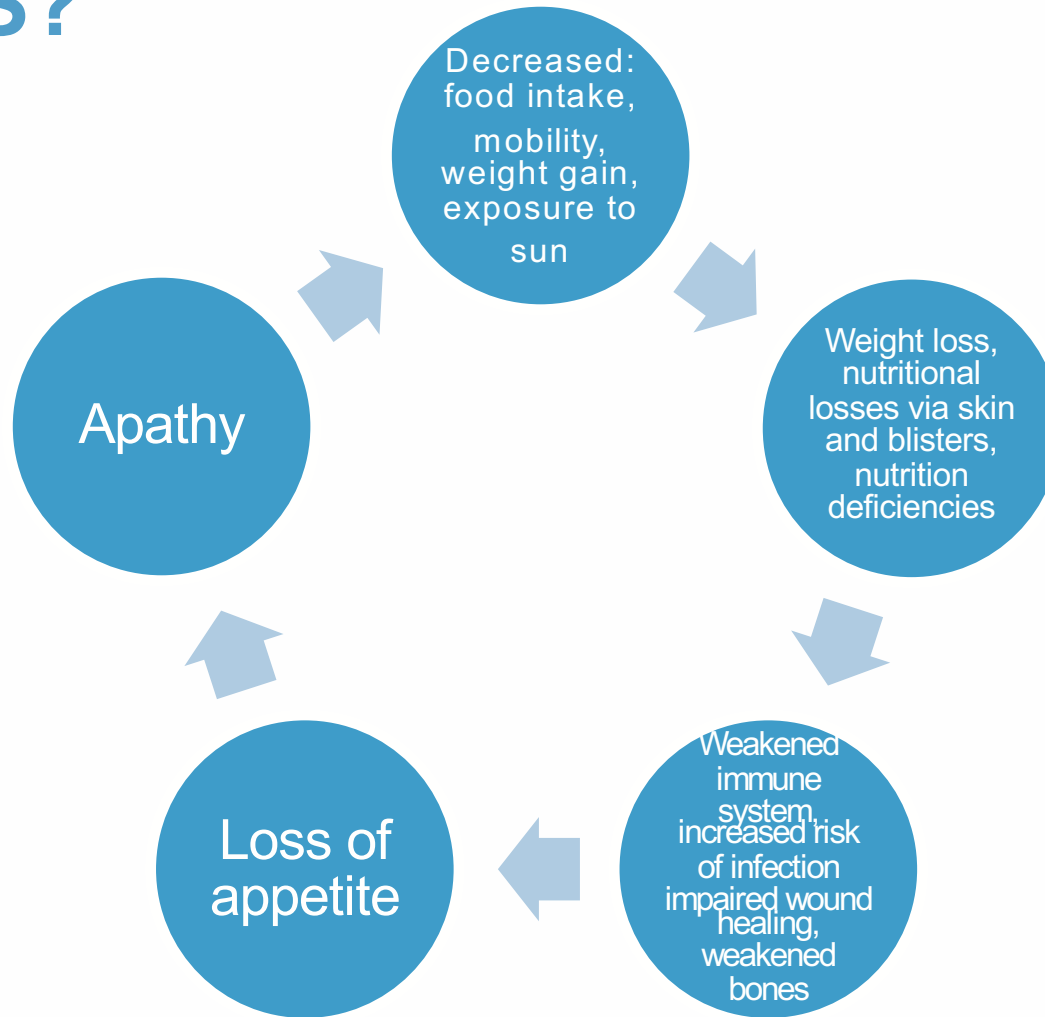
Strengthen the immune system to fight infection

Provide energy

Aid with certain side effects of EB such as constipation

Improve overall quality of life

HOW DOES EB IMPACT NUTRITION STATUS?



COMMON GI SIDE EFFECTS

Constipation

- Increase intake of fiber
- Ensure adequate water intake
- Laxatives may need to be started under medical supervision

Chewing and Swallowing Difficulties

- High calorie pureed meals
- High calorie/protein nutritional supplements/ nutritional shakes
- Tube feedings

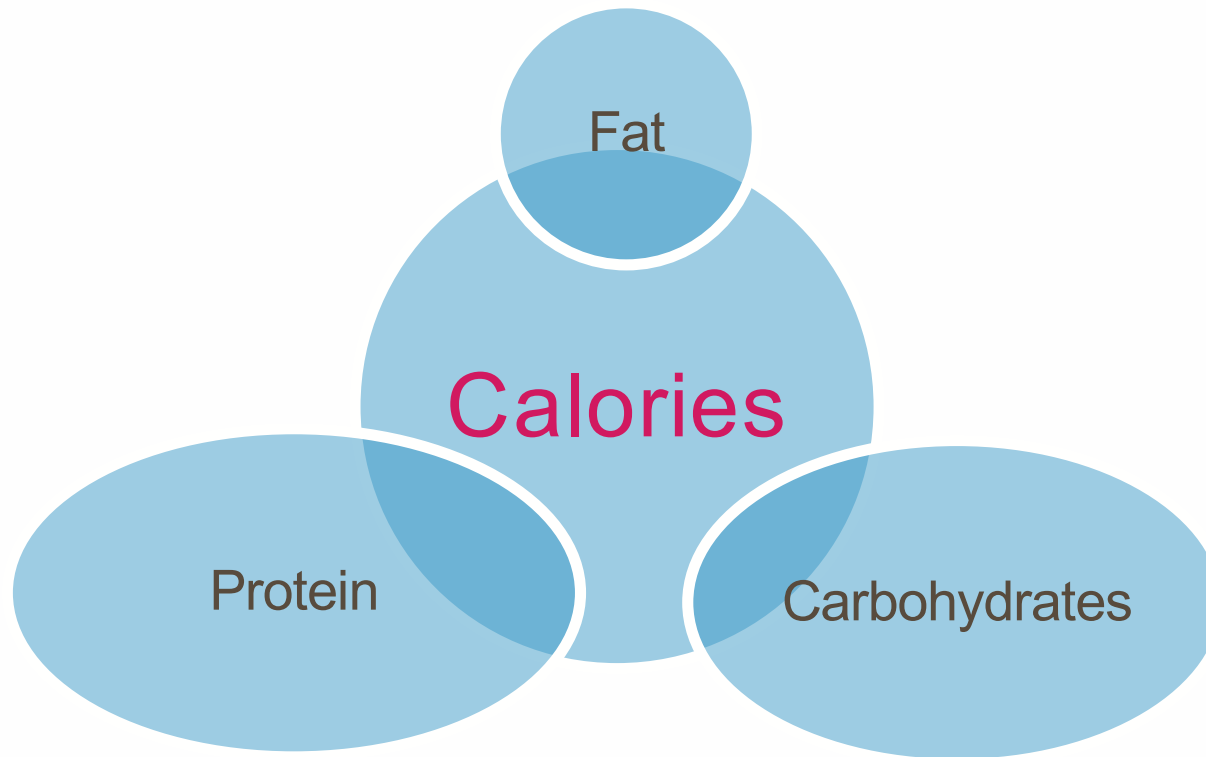
Malabsorption related to internal wounds

- Regular monitoring of labs followed by vitamin supplementation per doctor or RD recommendation

CALORIE AND PROTEIN REQUIREMENTS

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- Basal Metabolic Rate – baseline calorie needs at rest (breathing, digestion etc)



CALORIE AND PROTEIN REQUIREMENTS

- Infants can require 10% or more calories than their typical needs (recommend breastmilk fortified using a soothe or sensitive formula)
- Calorie needs for children and adults can be 1.5 – 2 times greater than baseline requirements
- Protein needs may be 2 or 3 times the recommended dietary allowance for infants, children and adults
- Small frequent meals are often better tolerated



STRATEGIES FOR INCREASING CALORIES

- *Calorie boosting- adding small amounts of a food that contain significant calories per serving to a meal or snack.*



CALORIE BOOSTING EXAMPLES

- Add **avocado** to toast
- Add **mayo** to tuna or salmon salad
- Add **butter** to oatmeal, rice or pasta
- Add **dry pudding mix** to batter
- Add **greek yogurt** to smoothie
- Add **quinoa** to oatmeal or smoothie
- Add **granola** to greek yogurt



ORAL NUTRITION SUPPLEMENTS

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- Beneficial in providing extra calories, protein and micronutrients
- Helpful addition to regular diet if intake is decreased
- Variety of options for all ages
- Things to consider:
 - Calorie /protein content
 - With or without fiber
- May also use modulers



ORAL NUTRITION SUPPLEMENTS

Pediatrics	Adults	Modulars
Pediasure or Boost Kids Essential	Ensure or Boost (Ensure Compact)	Duocal
Boost Breeze or Ensure Clear	Boost Breeze or Ensure Clear	Beneprotein
Orgain for Kids	Orgain	Nutrisource fiber
Carnation Instant Breakfast	Carnation Instant Breakfast	Benecalorie



ORAL NUTRITION SUPPLEMENTS

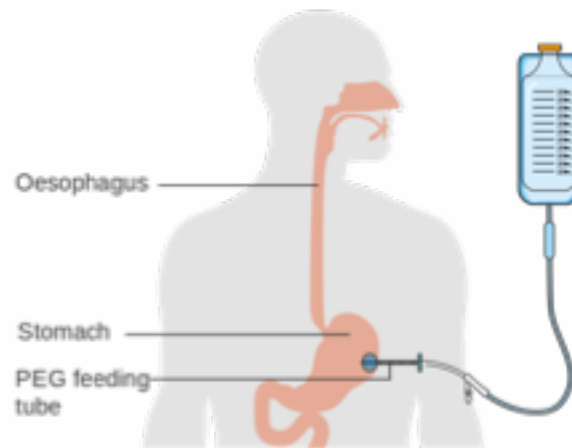
- If you or your child is “tired” of drinking the supplement...
 - Freeze into popsicles
 - Add dry pudding mix and make pudding
 - Add into a smoothie or milk shake
 - Use as the base for oatmeal or cereal



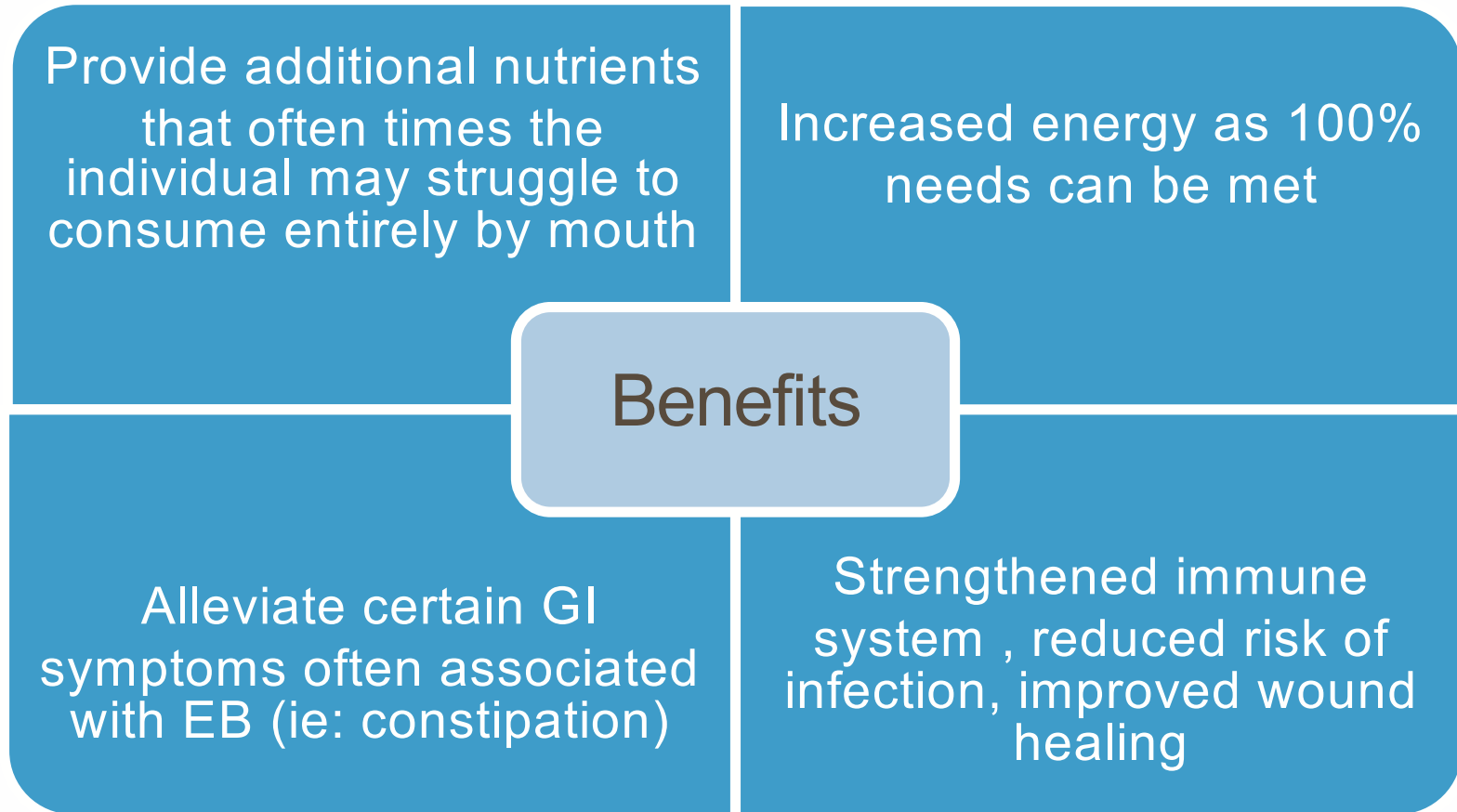
TUBE FEEDINGS

TUBE FEEDINGS

- **G Tube:** most common, typically better tolerated
- **NG Tube:** less common, temporary and often times irritates skin on face
- Can be given overnight or during the day



TUBE FEEDINGS



ENTERAL NUTRITION



Methods of
Delivery



Bolus

Gravity

Syringe

Pump

Continuous
overnight

COMMON TUBE FEEDING FORMULAS

Standard

- Pediasure
- Boost Kids Essential
- Ensure
- Boost
- Promote
- Nutren Jr./ Nutren

Concentrated

- Pediasure 1.5
- Boost Kids Essential 1.5
- Ensure Plus
- Boost Plus
- Nutren Jr. 1.5/Nutren 1.5

Semi Hydrolyzed

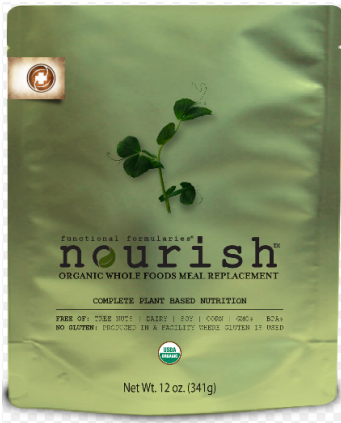
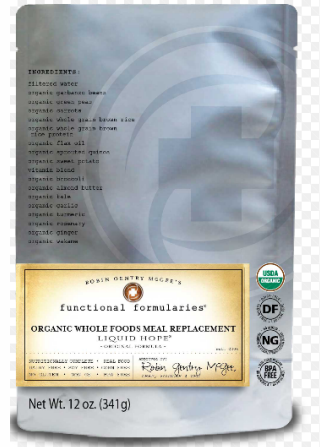
- Pediasure Peptide (1.0 and 1.5)
- Peptamen Jr (1.0 and 1.5)
- Peptamen HP
- Vital (1.0 and 1.5)

BLENDERIZED TUBE FEEDINGS

- *Defined as whole foods that are liquefied in a blender with water, juice, broth, or various types of milk, and administered by syringe bolus in feeding tubes.*



BLENDERIZED TUBE FEEDINGS



HOME BLENDED TUBE FEEDS

- Made with natural ingredients within the home setting
- Since many EB individuals have elevated needs (ie: protein, iron, zinc, fiber), RD can provide specific recipe for that patient's needs
- Home blended recipes typically given via bolus method to prevent clogging
- Ingredients may include: fruits, vegetables, chicken, potato, quinoa, oils, seeds, water, milk or milk alternative



SAMPLE HOME BLEND RECIPE

2 C organic chicken broth

1.5 C filtered water

1 C cooked quinoa

4T almond butter

1 hard boiled egg

1T goat butter

1 C goat kefir

1/2 C goat milk powder

1T olive oil

1T Udo-3-6-9 oil

1/2 C applesauce

1 banana

1 pitted prune

Fresh grated ginger and cinnamon

BLENDERIZED TUBE FEEDINGS

- Ensure you work closely with a dietitian when using these like you would with any standard tube feeding formula
- If this is an individual's sole source of nutrition, additional vitamin supplementation may be needed

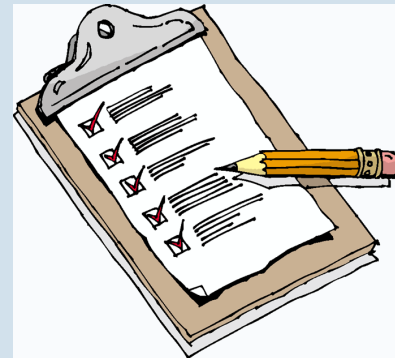


VITAMINS AND MINERALS

NUTRITION-RELATED LABS

Check every 6-12 months

- Comprehensive Metabolic Profile with Magnesium and Phosphorus
- Vitamin D
- Complete Blood Count
- Iron Panel with Ferritin
- Zinc, Selenium
- C- Reactive Protein
- Vitamin B1
- Carnitine
- Vitamin B12 with Methylmalonic acid
- Folate



VITAMINS AND MINERALS



Multivitamin

Iron – anemia secondary to skin wounds

Vitamin D/ Calcium – bone health

Vitamin C – strengthen immune system

Zinc – improved wound healing



ANTI INFLAMMATORY DIET

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- Rich in fresh foods and less processed foods
- Includes high fiber foods, healthy fats and lean protein
- Examples: Nuts, seeds, avocado, fatty fish (salmon), eggs, yogurt, cheese, milk, legumes, lean white meats (chicken), fruits, vegetables, tofu



ANTI-INFLAMMATORY DIET: TYPICAL DAY



Breakfast	Lunch	Snack (2-3 snacks per day)	Dinner
Whole grain cereal or oatmeal with berries and 8 oz milk	Grilled chicken on whole grain wrap with lettuce, tomato and avocado with calcium fortified orange juice	Guacamole and cheese with whole grain pita chips or banana with almond butter	Salmon burger with baked sweet potatoes and sautéed vegetables

TAKE AWAY!



- Nutrition is a priority given elevated nutrient needs coupled with inadequate intake
- Aim on calorie boosting and small frequent meals
- Consider an oral supplement in-between meals
- Tube feedings are often implemented and beneficial
 - Bolus vs. Continuous
 - Variety of formulas/blenderized products
- Additional vitamins may be required
 - Avoid starting until labs are checked and reviewed by MD and RD
- Focus on fresher and less processed foods

THANK YOU FOR LISTENING!

QUESTIONS?