NUTRITION MANAGEMENT OF CHILDREN AND ADULTS WITH EPIDERMOLYSIS BULLOSA

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MY BACKGROUND

- Pediatric Oncology, Burns, GI
- Home Infusion
- Ketogenic Diet, General Nutrition Counseling, Feeding Clinic, Nutrition Support
- CHOP Outpatient GI and Epidermolysis Bullosa
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?

Apathy

Decreased: food intake, mobility, weight gain, exposure to sun

Weight loss, nutritional losses via skin and blisters, nutrition deficiencies

Weakened immune system, increased risk of infection, impaired wound healing, weakened bones

Loss of appetite
## COMMON GI SIDE EFFECTS

<table>
<thead>
<tr>
<th>Condition</th>
<th>Recommendations</th>
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</thead>
</table>
| 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.

- Avocado
- Butter/Oil
- Cheese
- Mayo
- Heavy cream
- Greek Yogurt

- Promotes weight gain
- Adds additional 100-150 calories
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
ORAL NUTRITION SUPPLEMENTS

• 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

<table>
<thead>
<tr>
<th>Pediatrics</th>
<th>Adults</th>
<th>Modulars</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pediasure or Boost Kids Essential</td>
<td>Ensure or Boost (Ensure Compact)</td>
<td>Duocal</td>
</tr>
<tr>
<td>Boost Breeze or Ensure Clear</td>
<td>Boost Breeze or Ensure Clear</td>
<td>Beneprotein</td>
</tr>
<tr>
<td>Orgain for Kids</td>
<td>Orgain</td>
<td>Nutrisource fiber</td>
</tr>
<tr>
<td>Carnation Instant Breakfast</td>
<td>Carnation Instant Breakfast</td>
<td>Benecalorie</td>
</tr>
</tbody>
</table>
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

Provide additional nutrients that often times the individual may struggle to consume entirely by mouth.

Increased energy as 100% needs can be met.

Alleviate certain GI symptoms often associated with EB (ie: constipation).

Strengthened immune system, reduced risk of infection, improved wound healing.
ENTERAL NUTRITION

Methods of Delivery

- Bolus
- Pump

Delivery Methods:
- Gravity
- Syringe
- Continuous overnight
# Common Tube Feeding Formulas

<table>
<thead>
<tr>
<th>Standard</th>
<th>Concentrated</th>
<th>Semi Hydrolyzed</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Pediasure</td>
<td>• Pediasure 1.5</td>
<td>• Pediasure Peptide (1.0 and 1.5)</td>
</tr>
<tr>
<td>• Boost Kids Essential</td>
<td>• Boost Kids Essential 1.5</td>
<td>• Peptamen Jr (1.0 and 1.5)</td>
</tr>
<tr>
<td>• Ensure</td>
<td>• Ensure Plus</td>
<td>• Peptamen HP</td>
</tr>
<tr>
<td>• Boost</td>
<td>• Boost Plus</td>
<td>• Vital (1.0 and 1.5)</td>
</tr>
<tr>
<td>• Promote</td>
<td>• Nutren Jr. 1.5/Nutren 1.5</td>
<td></td>
</tr>
<tr>
<td>• Nutren Jr./Nutren</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
**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
<table>
<thead>
<tr>
<th>Ingredient</th>
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</thead>
<tbody>
<tr>
<td>2 C organic chicken broth</td>
</tr>
<tr>
<td>1.5 C filtered water</td>
</tr>
<tr>
<td>1 C cooked quinoa</td>
</tr>
<tr>
<td>4T almond butter</td>
</tr>
<tr>
<td>1 hard boiled egg</td>
</tr>
<tr>
<td>1T goat butter</td>
</tr>
<tr>
<td>1 C goat kefir</td>
</tr>
<tr>
<td>1/2 C goat milk powder</td>
</tr>
<tr>
<td>1T olive oil</td>
</tr>
<tr>
<td>1T Udo-3-6-9 oil</td>
</tr>
<tr>
<td>1/2 C applesauce</td>
</tr>
<tr>
<td>1 banana</td>
</tr>
<tr>
<td>1 pitted prune</td>
</tr>
<tr>
<td>Fresh grated ginger and cinnamon</td>
</tr>
</tbody>
</table>
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
ANTI INFLAMMATORY DIET

• 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

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