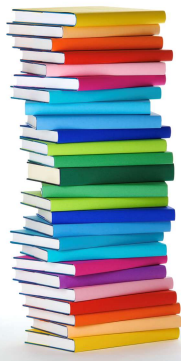



Success Stories:
Ready-to-feed AAF for the Dietary Management of SBS, EoE, and FPIES




1



Learning Objectives

- Understand the role of dietary management in short bowel syndrome (SBS), eosinophilic esophagitis (EoE), and food protein-induced enterocolitis syndrome (FPIES)
- Recognize clinical scenarios in which a ready-to-feed amino acid-based formula (AAF) may be indicated


2




Short Bowel Syndrome (SBS)

Ruba Abdelhadi, MD, CNSC, NASPGHAN-F

Professor of Pediatrics, University of Missouri-Kansas City School of Medicine
 Director, Nutrition Support and Nutrition Services Programs at Children's Mercy Hospitals and Clinics



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Disclosures

- Honorarium provided by Nutricia
- I serve on the Clinical Advisory Board of the Global Enteral Device Supplier Association (GEDSA) – non-profit
- I serve as Director on the National Board of Physician Nutrition Specialists (NBPNS) – non-profit
- **None pose any conflict of interest for this presentation**

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What is Short Bowel Syndrome?

- **Malabsorption** resulting from anatomical or functional loss of a significant length of the small intestine
 - bowel length, mucosal integrity, motility, perfusion
- Possible consequences:
 - Fluid electrolyte disturbances, diarrhea, dehydration
 - Malnutrition, micronutrient deficiencies
 - Poor growth, development
 - Parenteral nutrition-related complications and nutrition-related organ disease
 - Poor quality of life
 - High morbidity
 - Poor survival probability
- **Incidence** = approximately 24.5 per 100,000 live births per year
- **Prevalence** has increased over the past several decades

Walls, et al 2004 and Merrill et al 2017

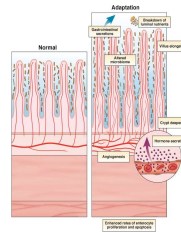
5



Nutrition Management

Goals

- Facilitate intestinal autonomy
- Promote intestinal adaptation
- Enhance enterohepatic circulation
- Promote an adapted absorptive surface




Enteral Nutrition

- **Early Stage:** Human milk is best!
- Best formula choice for infants
 - Extensively hydrolyzed formula (eHF)
 - AAF
 - ↑ Tolerance to feeds
 - ↑ Advancement of feeds
 - ↑ Ability to wean down parenteral nutrition


Cellular and Molecular Gastroenterology and Hepatology 2018; 24(2):380-391
 https://www.mdpi.com/1422-0067/19/2/380

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Case Study: SBS & AAF Supplementation 

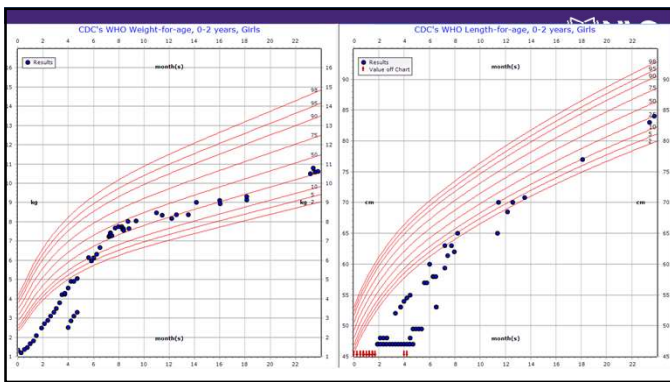
- Now 13 yr old female history of complex gastroschisis and jejunal atresia leading to SBS
- She has had multiple abdominal surgeries including 3 Serial Transverse Enteroplasty Procedure (STEP) procedures
- Her last intestinal measurement was approximately 150 cm of small bowel
 - Ileum connected to descending colon and rectum

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Case Study: SBS & AAF Supplementation 

- Born at 34 weeks 6 days gestation
- BW 2.91 kg, BL 47.5 cm
- Received parenteral nutrition (PN) since birth
- Received enteral nutrition (EN) pumped human milk until 2 months of age
- Developed bloody stools with concern for cow milk protein intolerance (CMPI)
 - Switched to pumped human milk (dairy-free, soy-free, egg-free, nut-free) – unsuccessful
- Switched to powdered infant AAF until 1 year of age then powdered junior AAF after 1 year of age

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Case Study: SBS & AAF Supplementation 

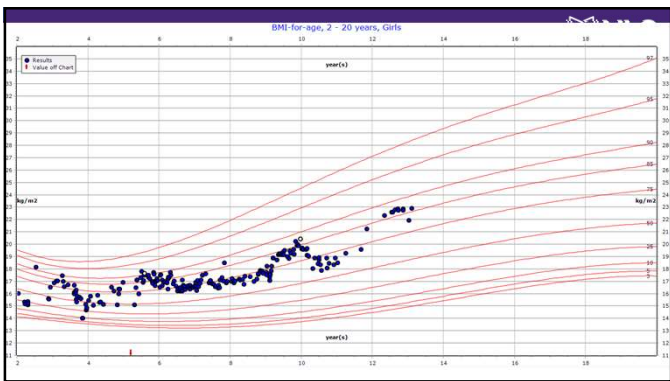
- From age 1 year to 9 years of age:
 - Continued junior powdered AAF via G Tube after 1 year of age
 - Ongoing Challenges in PN support
 - Line infections
 - Hyperactive child, line breaking
 - History of left subclavian vein thrombosis on prophylactic Lovenox
 - Challenges in weaning down PN
- Continued PN support
 - 50% PN, 50% EN

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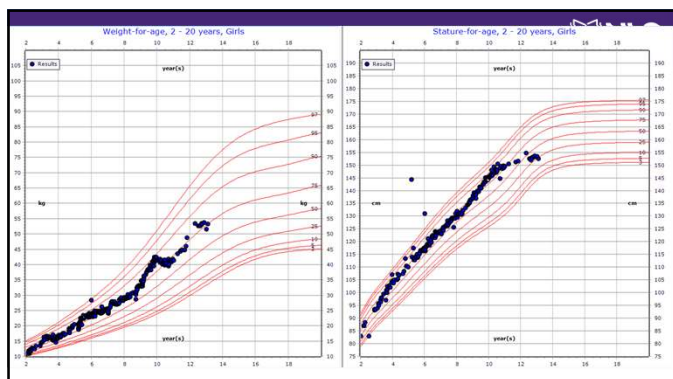
Case Study: SBS & AAF Supplementation 

- Developed line infection and sepsis at age 10 years
 - Infectious disease consult - central line was removed
- Ongoing challenges in advancement of EN feeds
 - G Tube dislodgement & site problems
 - Challenges with administering feeds in a hyperactive child
- Ongoing challenges in advancement of oral diet
 - Oral aversion
 - Feeding difficulties
 - Limited food variety, poor palatability
- Trial of ready-to-feed (RTF) AAF, orally
 - Eventually consuming 2-3 cartons per day PO

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NLC
NUTRICIA LEARNING CENTER

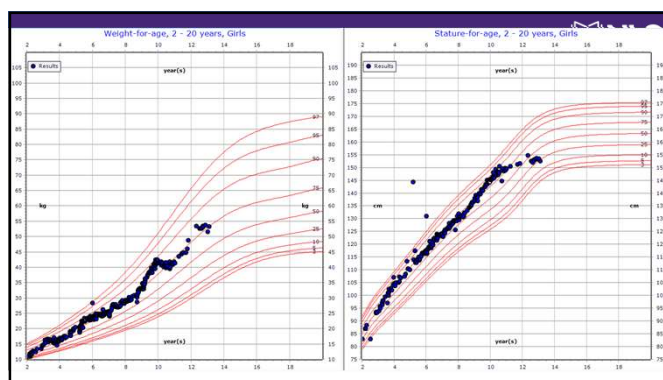
Nutrition and Medical Management

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Case Study: SBS & AAF Supplementation **NLC**
NUTRICIA LEARNING CENTER

- Discontinued PN support
 - Discharged home with close follow up
- Outpatient follow up
 - Monitored stool output
 - Prepared family to anticipated weight loss
- Offered family and child psychology support
 - Medically fragile child
- Encouraged offering RTF AAF to siblings
- Encouraged mealtime with family and school lunch

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Medical Management **NLC**
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- Medications
 - Improve gastric accommodation
 - Improve appetite
 - Slow down transit time

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Nutrition Management **NLC**
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- Lack of interest in foods with appropriate nutritional value
- RTF AAF:
 - Preferred Orange-Pineapple flavor
 - Tasted like real juice
 - Gave the child and family a sense of normalcy
 - Convenient on the go
 - Our hospital Home Care were able to provide it
 - Gave her the power to be in charge

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Eosinophilic Esophagitis (EoE)

Alison Cassin, MS, RD, CSP, LD
 Cincinnati Children's Hospital



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EoE is clearly defined

- Definition from 2011 consensus recommendations:
 - "Eosinophilic Esophagitis (EoE) represents a chronic, immune/antigen-mediated esophageal disease characterized clinically by symptoms related to esophageal dysfunction and histologically by eosinophil-predominant inflammation."

EoE if >15 eos / hpf (400x microscope field)



Magnification: 400x = "HPF"



Estimated prevalence of 56.7/100,000

♂♂♀
 2/3 of EoE patients are male

1. Noel RJ, et al. NEJM 2018; 2004; 2. Spergel et al. Allergy 2003; 3. Funda, et al. Gastroenterology 2007; 133:3342-51; 4. Dellon, et al. Ann Gastroenterol 2013; 108:979-95; 5. Durkin, et al. Pract Gastroenterol 2018;42:40-51. Illustration courtesy of Nutricia Medical and Scientific Affairs, North America.

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Signs & symptoms of EoE vary by age

INFANTS	TODDLERS	CHILDREN	TEENS	ADULTS
Poor growth	Feeding difficulties	Vomiting/retching	Gastroesophageal reflux symptoms (heartburn, reflux)	Abdominal pain
			Chest pain	Dysphagia
				Food impaction
Nonesophageal symptoms				Esophageal symptoms

Possible signs and symptoms of EoE^{1,2}
 Predominant symptoms by age, not comprehensive^{4,5}

- **Infants and Toddlers:** FTT, feeding difficulties
- **School age:** vomiting, abdominal pain, reflux
- **Adolescents/Adults:** Dysphagia & food impaction

1. Noel RJ, et al. NEJM 2018; 2004; 2. Spergel et al. Allergy 2003; 3. Funda, et al. Gastroenterology 2007; 133:3342-51; 4. Dellon, et al. Ann Gastroenterol 2013; 108:979-95; 5. Durkin, et al. Pract Gastroenterol 2018;42:40-51. Illustration courtesy of Nutricia Medical and Scientific Affairs, North America.

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Esophageal inflammation can impact feeding

- Not sufficient to ask, "do you have trouble swallowing?"

UNUSUAL EATING BEHAVIORS:
 Slow eating, lots of fluid with meals,¹ and preference for pureed textures are seen in children¹

1. Licozras, et al. J Allergy Clin Immunol. 2011; 2. Dellon E, et al. Gastroenterology. 2018.

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Management of the EoE Patient

```

    graph TD
      A[EoE Management Options] --> B[Dietary management]
      A --> C[Esophageal dilation]
      A --> D[Pharmacologic management]
      B --> B1[Elimination diet  
Empiric or Test Directed]
      B --> B2[Elemental diet (AAF)]
      D --> D1[Steroids]
      D --> D2[PPIs]
    
```


1. Licozras, et al. J Allergy Clin Immunol. 2011; 2. Dellon E, et al. Gastroenterology. 2018.

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Case Study: EoE & AAF Supplementation 


- 4 yo male
 - Feeding difficulties as a toddler
 - Feeding therapist suspected medical cause of food aversion
 - Limited food variety
 - Preferred foods = chocolate milk and oyster crackers

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Case Study: EoE & AAF Supplementation 

- Atopic: IgE-mediated egg and tree nut allergies
- Adequate linear growth, but poor weight gain
 - BMI z-score = -1.43
- Underwent EGD w/ biopsies confirming EoE diagnosis

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EoE: Initial management options 

□ Pharmacologic? *or...* □ Empiric diet elimination?


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EoE: Initial management options 

- Pharmacologic
 - Fluticasone?
 - Too young
 - Budesonide?
 - Refused to take




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Empiric six food elimination diet (SFED) 




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SFED: nutritional challenges 


1. Needed to support growth & development
2. Limited repertoire of accepted foods:
 - crackers, cereal, chips, gluten-free pretzels, banana chips
3. Early satiety, grazing pattern

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Practical challenges with diet elimination 


- Meal planning and preparation efforts
- **Shopping inconvenience:** specialty grocery stores & online
Wolf AW, Huang KZ, Durban R, et al. The six-food elimination diet for eosinophilic esophagitis increases grocery shopping cost and complexity. Dysphagia. 2016;31:765-70.
- High cost of allergen-free convenience foods
- Allergy-friendly foods aren't always
 - Portable & convenient
 - Nutrient dense


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Problem / Solution 

- **Poor diet quality**
 - ▣ Deficient in calories, protein, micronutrients
- **Ready-to-feed amino acid-based formula**
 - ▣ Two 8 fl oz servings daily provided:
 - 27% of calorie needs
 - 75% of protein needs


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SFED success 




- Outcome: “passed” SFED Trial
 - ▣ EGD/biopsy results - normal
 - ▣ Symptom improvement

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With AAF, the SFED did NOT impair growth 

Age (years)	Milestone	Weight kg (%ile)	Weight z-score
4	EoE diagnosis	13.8 (7)	-1.51
4.5	After successful 3-month initiation of the SFED supplemented with AAF	15.0 (10)	-1.27
7	Feeding therapy graduate	20.2 (16)	-1.01

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Food Protein-Induced Enterocolitis Syndrome (FPIES)



Raquel Durban, MS, RD, LD/N
 The Carolina Asthma & Allergy Center

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Disclosures 

- ▣ Honorarium provided by Nutricia
- ▣ Consultant positions: Mead Johnson Nutrition, Abbott Nutrition, AstraZeneca
- ▣ Speakers Bureau: Mead Johnson Nutrition, Abbott Nutrition, Danone
- ▣ Advisory board affiliations: I-FPIES, FAACT
- **None pose any conflict of interest for this presentation**

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FPIES Definitions




- FPIES is a non-IgE, cell mediated food allergic disorder that manifests with predominately gastrointestinal symptoms:
 - Delayed emesis 1-4 hours after food ingestion
 - May include:
 - Lethargy
 - Pallor
 - Diarrhea
- Absence of cutaneous and respiratory allergic symptoms
- Prevalence
 - 0.51% in children (17 years and under)
 - 0.22% of US Adult population


1. Ann Allergy Asthma Immunol 2020;125:14-16. 2. J Allergy Clin Immunol 2017;139:1111-26. 3. Nisalak-Wegzyn et al. JACI OCT 2019

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Management of the FPIES Patient



- Avoidance of the trigger food for ~18 months, reintroduction
 - Most common triggers:
 - Milk, soy, rice, oat
 - Other unusual triggers, ex. avocado, sweet potato, beef, banana, pea
- Children with FPIES can continue to breast feed
 - Summary Statement 21: Do not recommend routine maternal dietary elimination of offending triggers while breast-feeding if the infant is thriving and remains asymptomatic
- Extensively hydrolyzed cow's milk-based formula
 - 10-20% require AAF



1. J Allergy Clin Immunol 2017;139:1111-26

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Key considerations



- Diagnosis of infancy - typically
- Building the diet, rather than eliminating
- Feeding skills/milestones
- Time of rapid growth and development
*Taste, Temperature, Texture
 Color, Consistency, Creativity
 Resistance, Resilience, Repeat*


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Patient History



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History of Present Illness



<ul style="list-style-type: none"> □ Vaginal birth, 38.5 weeks gestation □ Female <ul style="list-style-type: none"> ■ 10th percentile weight for age ■ 5th percentile length for age □ Exclusively breast fed (EBF) until 2 months of age <ul style="list-style-type: none"> ■ Of note, no maternal dietary restrictions ■ 3-4 ounces cow milk (CM) based formula introduced <ul style="list-style-type: none"> ■ 45-60 minutes post ingestion, 1 episode of vomiting 	<ul style="list-style-type: none"> □ Returned to EBF until 4 months of age <ul style="list-style-type: none"> ■ 10th percentile weight for age ■ 25th percentile ■ 4-5 ounces of CM based formula was fed <ul style="list-style-type: none"> ■ 1-2 hours post ingestions, at least 10 episodes of vomiting <ul style="list-style-type: none"> ■ Lethargy ■ Rehydrated at Ped office, NO ER
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Nutrition and Medical Management



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Nutrition Management



- Allergist recommends milk and soy avoidance
 - Skin prick test negative to milk
 - Prescribed eHF formula to supplement BF
 - Vomiting resolved on eHF
 - No maternal restrictions
- RD assessed feeding readiness
 - Selection of foods to be introduced over 5-7 days based on cultural preference and risk assessment
 - Vitamin D3 400 IU per day

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Nutrition Management



- Continued to introduce foods with appropriate growth and nutritional adequacy at 1 yo
 - ~ 10th percentile weight for age
 - ~ 50th percentile length for age
- At 1 y/o, transitioned to ready-to-feed amino acid-based formula r/t persistent FPIES to milk pending OFC.

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Formula Selection




- RTF AAF was selected due to:
 - Nutrient profile
 - Convenience
 - Taste preference over plant-based milk
 - Peers and sibling drink juice boxes
- 8 to 16 ounces per day
 - 240-480 calories, preferred tropical and orange-pineapple flavors
 - Met vitamin D and calcium recommendations
 - Variability in amount to provide flexibility with caregivers

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Conclusion




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- Growth continued to trend well and diet expanded with only avoidance of milk and soy
 - Versatile in types, texture, color, temperature of food
- With increased options, self weaned to at least 8 ounces of RTF AAF per day (did not exceed 16 ounces) only preferred one flavor in a plain cereal



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- At 21 months of age patient did not pass OFC to milk
 - 4 episodes of vomiting
- She began to avoid safe foods but increased acceptance of RTF AAF as a "safe" option.
 - Ideas presented for alternative preparations such as smoothies, popsicles, stove top grains, meatballs
- She continues of RTF AAF at a rate of 16-24 ounces per day r/t decreased solid food intake post OFC.
Weight trends maintained
Additional allied health: feeding therapist and play therapist

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Thank You!



Q&A

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