Dietetic Reference Guide for Cystic Fibrosis

Pediatric Pulmonary Center

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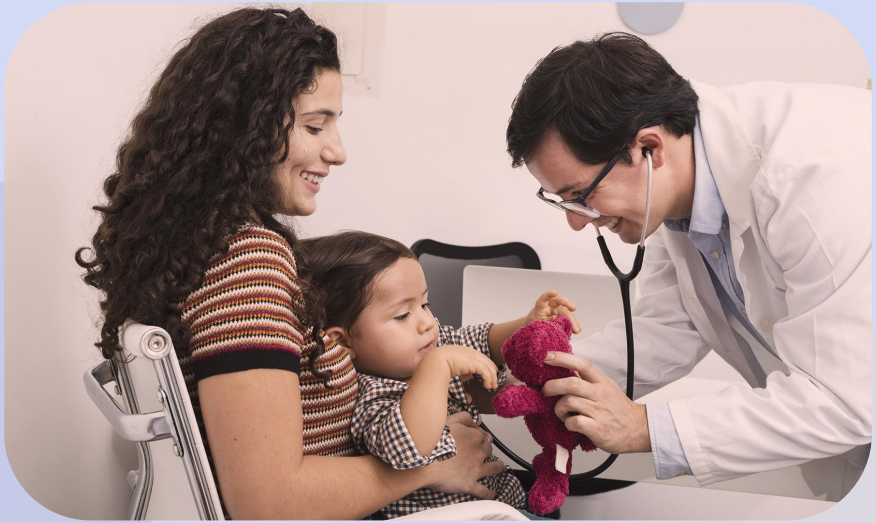
Nutrition Trainee



About Me/Challenges

1. Big ideas/Little time
2. Did not have a dietitian in CF Clinic
3. Schedule Changes

Failed Capstone Attempts

1. Feeding protocol for infants (DONE)
2. Solve Food Insecurity- (Outside my scope)
3. Provide access to dietitians with a resource list (Easily Outdated)

Needs Assessment

Access to a dietitian with experience with CF is difficult to find.
Every CF-related source recommends consulting a dietitian
Lack of dietitian in Clinic

Methods

- Literature Search
- Google Search
  - Inpatient/outpatient/private
- Facebook CF Chat Group
- Job Postings
- Ask Dr. Daines
- Interview w/TMC Dietitians

FINDINGS FROM NEEDS ASSESSMENT

LACK of Pediatric Dietitians in Tucson

Google provided very limited options for dietitians for the pediatric population

Open Positions for Ped Dietitians

CF Clinic, Meal Time Connection, TMC one all have been mentioned as needing a dietitian

Lack of Updated CF knowledge

CF is a complicated diagnosis and has had many advancements in treatment of care

Dr. Daines

Dietitians with CF speciality may not exist in Tucson

Lack of Peds Training in School

There were very few opportunities to focus on pediatrics throughout my education

Specific Accessibility

Researching CF is overwhelming as it affects so many systems and has multiple components

CAPSTONE MISSION

To create a reference guide made specifically for dietitians with limited training or experience with Cystic Fibrosis. This guide will provide a quick reference with the most relevant information, up-to-date recommendations, and important calculations needed in the treatment of Cystic Fibrosis in pediatrics.

Dietetic Reference Guide Overview

Cystic Fibrosis (CF)

A genetic, autosomal recessive condition that is caused by a mutation in the CFTR gene which is responsible for the permeability of the chloride channel. There are around 2,000 known mutations with varying degrees of severity classified in classes 1 (most severe)- 5 (least severe).

High Nutritional Risk:

- Hypermetabolism
- Malabsorption due to exocrine pancreatic insufficiency (EPI)
- Liposoluble vitamin deficiency
- CF-Related diabetes
- Gastroesophageal reflux
- Decreased bone density

Cystic Fibrosis Nutritional Guide's Main Points

01 Increased Nutrient Needs

02 Pancreatic Enzymes (PERT)

03 Malabsorption Supplements Labs

04 Cystic Fibrosis Related Diabetes

Energy Needs

Needs are individualized and determined by other CF-related factors that can increase nutrient needs.

Recommendations include:

Energy needs: 110–200% of RDI
Protein: 150-200% of RDI or higher

*Advancements in modulators like Trikafta have increased pancreatic absorption ability. More research is needed to determine energy needs with medical advancements.

Exocrine Pancreatic Insufficiency (EPI)

- Starts in utero due to thickened secretions obstructing intrapancreatic ducts.
- Infants with a new CF diagnosis
  - 75% have pancreatic insufficiency
  - 85% of infants will develop it within 3-4 months of age.

Pancreatic Enzyme Replacement Therapy (PERT):

- Pancreatic Enzyme Calculations
- PERT Oral Intake dosing table
- PERT Tube Feed dosing table
- Information about Administration

Vitamin and Mineral Deficiencies

- Fat Soluble Vitamins ADEK Deficiencies
- Additional Vit D supplementation Chart
- Additional Vit K supplementation
- Iron, Zinc, Calcium Deficiencies
- Sodium deficiencies
- RDA/ UL Charts provided
- Recommended to assess Labs annually

Cystic-Fibrosis-Related Diabetes (CFRD)

Management of CFRD shares features with DM 1 and 2 but differs as CFRD has insulin resistance and insulin deficiency.

Annual screening by age 10 or earlier

- **Fasting glucose >/=126mg/dL**
- **2-hour glucose >/=200mg/dL**
- **HbA1C >/= 6.5%**

*HbA1C could be falsely low in patients with CF, while CF patients w/o CFRD diabetes could have high glucose levels.

*CFRD should be screened for if there is significant weight loss or poor weight gain, elevated serum glucose, and unexplained decline in lung function

Modulators

Enhance or restore functional protein expression of certain CF-causing mutations, significantly expanding life expectancy.

- **Trikafta®** (elexacaftor/tezacaftor/ivacaftor)- approved for people with CF ages 2 and older who have at least one copy of the F508del mutation or at least one copy of 177 specified mutations.
- **Symdeko®** (tezacaftor/ivacaftor)
- **Orkambi®** (lumacaftor/ivacaftor
- **Kalydeco®** (ivacaftor)

Newborn and Infant

CF Foundation Guidelines for Growth:

Use WHO for 0-24 months
Use CDC Growth Charts for >2 yrs old

CF Patients Recommended

- 0-12mo: Maintain weight-for-length 50th percentile
- 12-24mo: Maintain weight-for-length 75th percentile
- >2 yrs: Maintain a BMI 50th percentile

EXPECTED GROWTH VELOCITY:

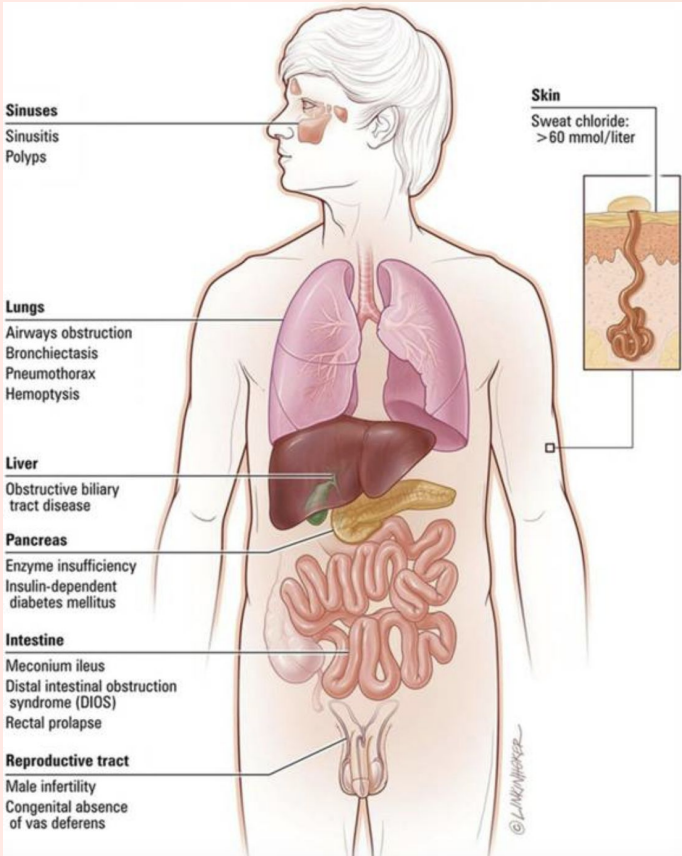
AGE	WEIGHT (g/day)	HEIGHT (cm/week)	Head Circumference (cm/wk)
0-4 months	23-34	0.8-0.93	0.38-0.48
4-8 months	10-16	0.37-0.47	0.16-0.2
8-12 months	6-11	0.28-0.37	0.08-0.11
12-16 months	5-9	0.24-0.33	0.04-0.08
16-20 months	4-9	0.21-0.29	0.03-0.06
20-24 months	4-9	0.19-0.26	0.02-0.04
2-4 yrs	2-3 kg/yr	5.5-9 cm/yr	
4-6yrs	2-3kg/yr	5-8.5cm/yr	
6-10 yrs Boys Girls	2-3 kg/yr	 4-6 cm/yr 4.5-6.5cm/yr	
>10 yrs	2-3kg/yr	8-14cm/yr	

Diagnosis of Pediatric Malnutrition

Indicators	Mild	Moderate	Severe
Weight gain velocity (<2 yr)	<75% of expected wt gain for age	<50% of expected wt gain for age	<25% of expected wt gain for age
Weight Loss (>2 yr)	5% below UBW	7.5% below UBW	10% below UBW
Deceleration in Wt/Length or BMI/age	Decline of 1 Z score	Decline of 2 Z scores	Decline of 3+ Z scores
Inadequate nutrient intake	51-75% of est. energy/pro needs	26-50% of est. energy/pro needs	</= 25% of est. energy/pro needs

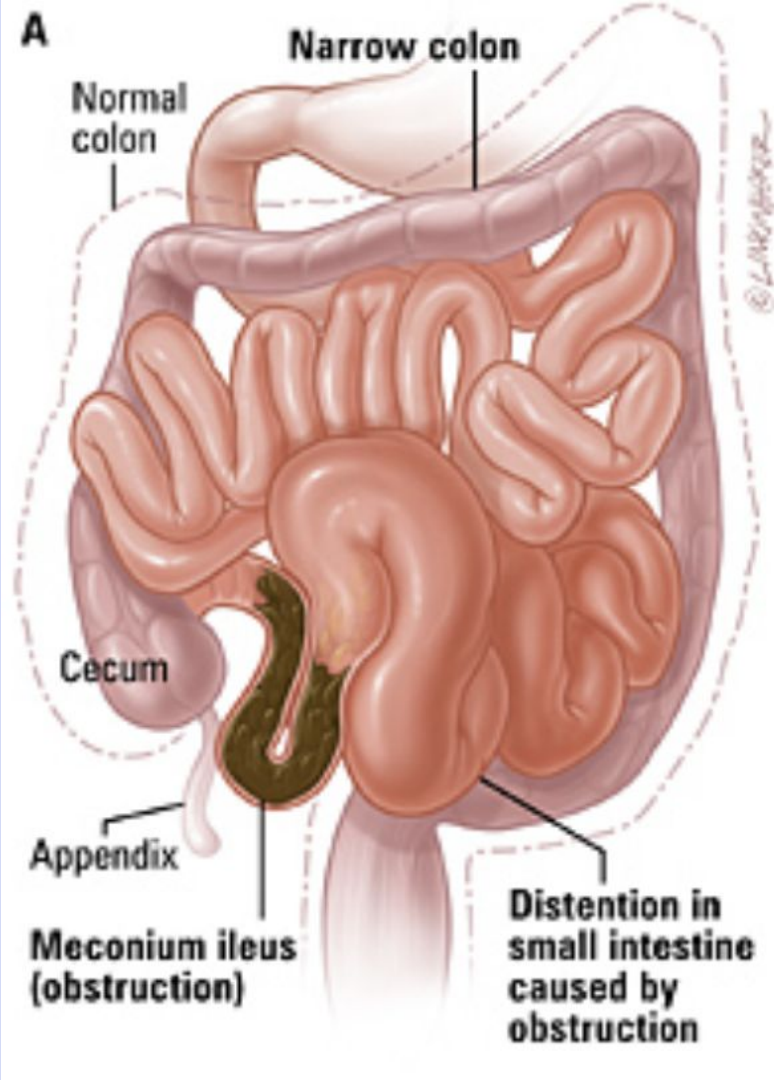
Common complications with CF newborn:

- Meconium ileus
- Distal Intestinal Obstruction Syndrome (DIOS)
- Intestinal obstruction with or without intussusception or volvulus
- Obstructive jaundice
- Edema with hypoproteinemia, anemia and hypoprothrombinemia
- Hypoprothrombinemia
- Failure to thrive
- Salty taste or salt loss syndromes
- Rectal prolapse



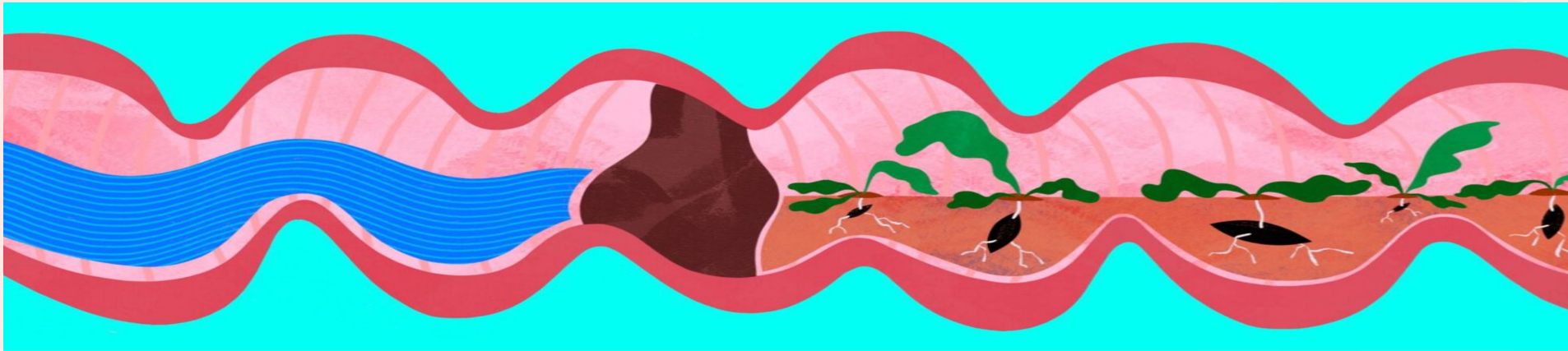
Meconium Ileus

- A bowel obstruction that occurs when the meconium in your child's intestine is even thicker and stickier than normal meconium.
- Treatment- Nonsurgical treatment includes enema but may need surgery which entails a bowel resection and ileostomy placement.



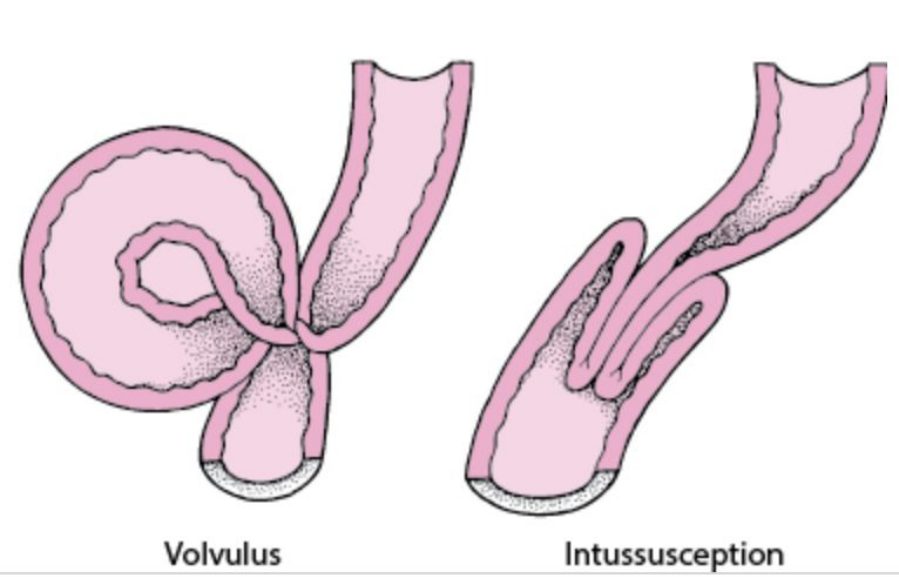
Distal Intestinal Obstruction Syndrome (DIOS)

Occurs when the bowel becomes partially or completely blocked. It usually occurs where the small intestine joins the large intestine.



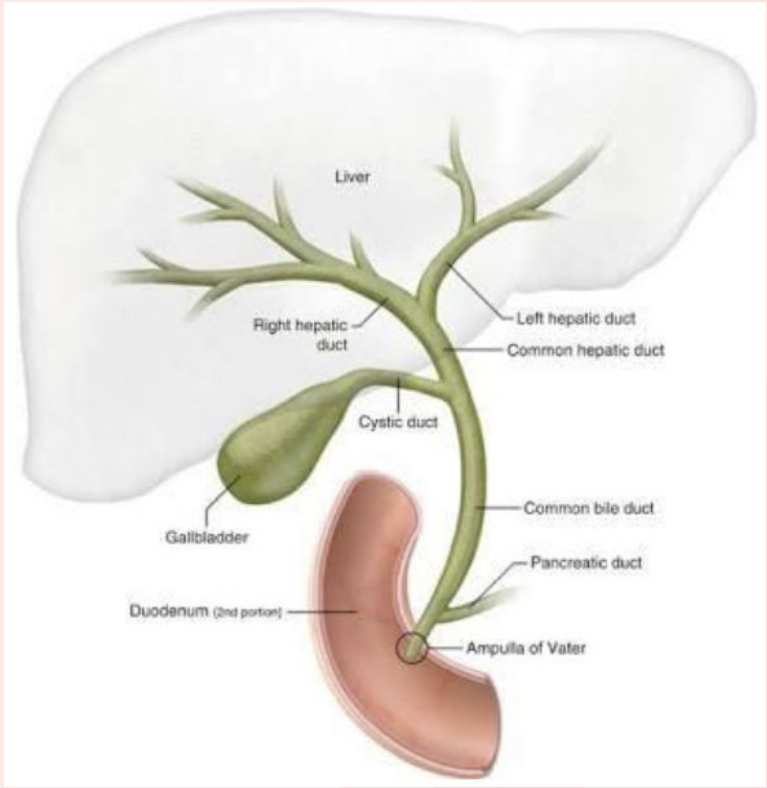
Intestinal Obstruction with or without Intussusception or Volvulus

Recurrent, crampy abdominal pain associated with a right lower quadrant fecal mass is often a feature of CF. Mal-digested food, combined with dehydrated, viscid intestinal mucus, may lead to the formation of a fecal mass at the ileocecal junction.



Obstructive Jaundice

A specific type of jaundice caused by a narrowed or blocked bile duct or pancreatic duct, preventing the normal drainage of bile from the bloodstream into the intestines



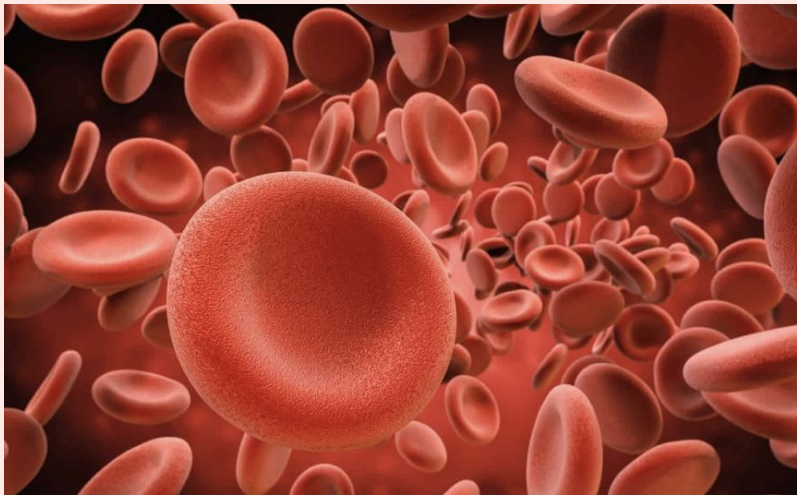
Edema with Hypoproteinemia, Anemia and Hypoprothrombinemia

Constellation of these three may be the earliest presentation of malabsorption syndrome secondary to pancreatic insufficiency in CF

Hypoprothrombinemia

Characterized by a deficiency of the blood-clotting substance prothrombin, resulting in a tendency to prolonged bleeding.

PERT and Vitamin K supplementation is required for correction.



Failure to Thrive

CF should always be considered in the differential diagnosis of failure to thrive in childhood, especially in children who have a history of frequent, loose, bulky stools and whose velocity of weight gain is decreasing.

Salty Taste or Salt-Loss Syndromes

In the absence of salt supplementation, the child may present with heat prostration (Depletion of electrolytes). CF can also lead to chronic hyperchloremic metabolic alkalosis.

- **Hyperchloremic Metabolic Alkalosis -** Results from either low chloride intake or excessive chloride wasting.

Sodium Supplementation

Sodium (Na) needs are increased in CF due to higher concentrations of Na in sweat and increase in warmer climates or with physical activity.

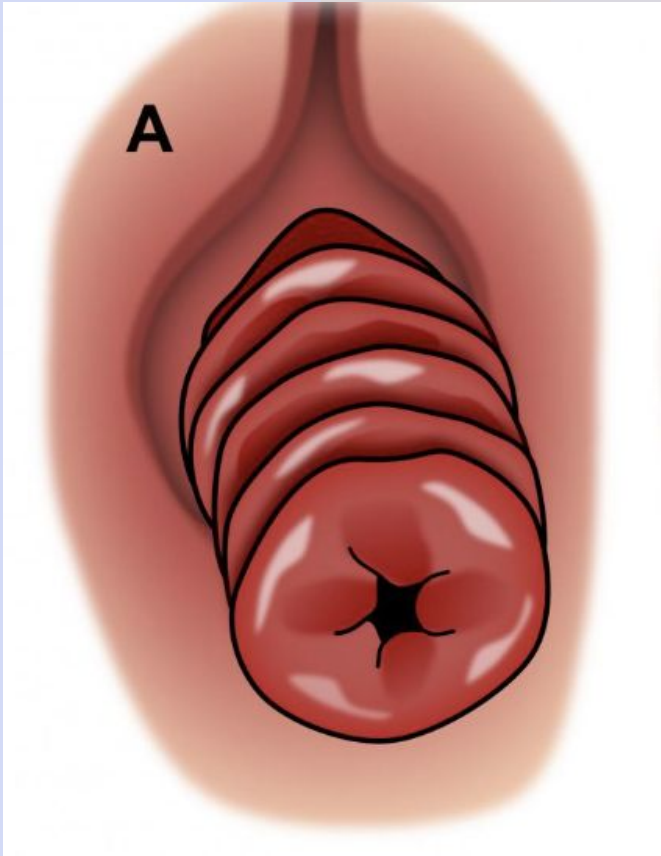
Recommendations: 2-4mEq Na/Kg/day

(Na) supplementation by age:

- Infant 0-6mo: 1/8th tsp. per day
- Infants 6-12mo: 1/4th tsp. per day
- Children >1yr: Liberal salt diet

Rectal Prolapse

Secondary to steatorrhea, bulky stools, and poor muscle tone may occur in up to 20% of patients with CF in the first few years of life.



Additional Signs and Symptoms

- Recurrent pneumonia or bronchiolitis
- Pulmonary infections with Staphylococcus and Pseudomonas

Sustainability

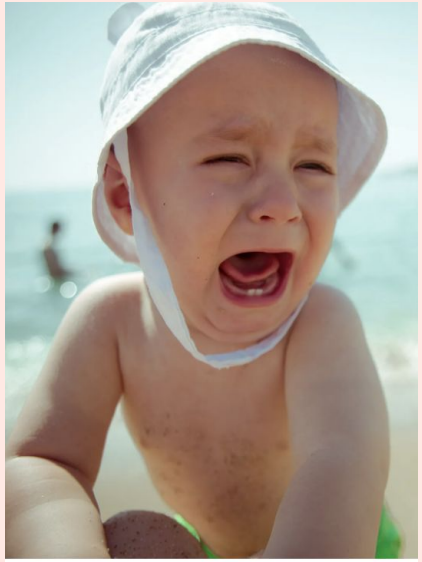
- Used by future nutrition trainees
- Updated by future nutrition trainees
- Will continue to evolve

Strengths of Guide

- Can be personalized- Made to be updated and adapted
- Contains updated info since May of 2023
- Will help avoid the oversight of high risk factors
- Provides a starting point for anyone who may have a CF patient with limited knowledge

Limitations of Guide

- Limited review and feedback from other dietitians
- Mainly based on two different hospital protocols
  - Texas Children's Hospital 13th Edition
  - Banner Clinical Nutrition Guide



Questions Or Suggestions ????

THANK YOU!!!

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