Early Nutrition in Cystic Fibrosis:
Challenges and Strategies from Birth to 18 Months

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DISCLOSURES

- Consultant for Nutricia
- Honorarium provided by Nutricia

None pose any conflict of interest for this presentation

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OBJECTIVES

1. Describe the basic genetics of cystic fibrosis, the newborn screening process, and early diagnostic indicators, with a focus on malnutrition risk.
2. Explain how altered gastrointestinal physiology in CF impacts nutritional status and the gut microbiome.
3. Identify key micro- and macronutrient requirements in infants with CF.
4. Apply appropriate tools for nutritional physical exam (NPFE) and laboratory monitoring in infants with CF.
5. Review a case study illustrating the nutritional management journey from diagnosis through 18 months of age.

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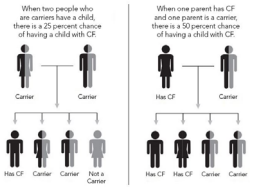
CYSTIC FIBROSIS (CF) INHERITANCE

AN OVERVIEW

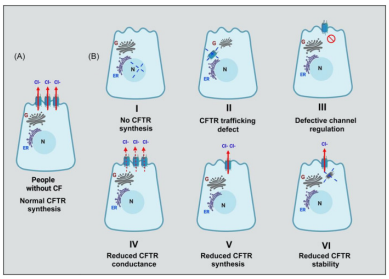
- Autosomal recessive
- >2,500 mutations of cystic fibrosis transmembrane receptor (CFTR) have been identified
- Mutations are categorized into 6 different classes
  - Class I and II – no CFTR at cell surface
  - Classes III, IV, V, VI – some CFTR at cell surface with decreased function



1. Cystic Fibrosis Foundation. Carrier Testing for Cystic Fibrosis. Accessed August 6, 2025. https://www.cff.org/intro-cf/carrier-testing-cystic-fibrosis

4

CFTR MUTATION CLASSES



2. Vavrina K, Griffin TB, Jones AM, Schindler T, Bui TN, Sankararamen S. Evolving nutrition therapy in cystic fibrosis: adapting to the CFTR modulator era. *Nutr Clin Pract*. 2025;1-13. Doi:10.1002/ncp.11332

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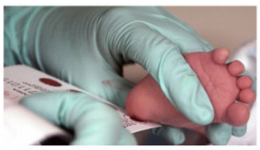
NEWBORN SCREENING

Step 1:

- Blood spot collection typically completed within the first 24-48 hours of life
- Blood is tested for levels of immunoreactive trypsinogen (IRT) a chemical made by the pancreas
  - Typically elevated in newborns with CF
  - Can be elevated for other reasons
  - *IRT may be repeated for confirmation*



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NEWBORN SCREENING

- Step 2:
  - Genetic testing completed for infants with an IRT greater than predetermined cutoff
  - Specific variant analysis (not whole gene sequencing)
    - Which variants and how many analyzed varies by state
    - State determines which variants to look for based on prevalence in that region
      - Example: Florida has large Hispanic population; NBS includes variants more commonly seen in that population
- Step 3:
  - Sweat testing
    - Completed at CF Foundation-accredited care center
    - Measures the amount of salt in the infants sweat
    - >60 mmol/L is indicative of CF

https://www.cff.org/intro-cf/sweat-test

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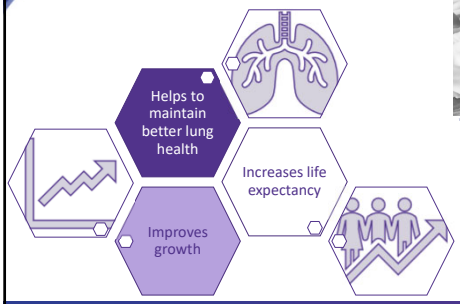
IMPACT OF EARLY DIAGNOSIS



3. Sathe, Meghana et al. Meconium Ileus in Cystic Fibrosis. *Journal of Cystic Fibrosis.* 2017; 16: S32 – S39.

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IMPACT OF EARLY DIAGNOSIS

Clinical Signs and Symptoms

- Meconium ileus - ~20% of infants with CF
- Frequent, foul-smelling stools with or without the presence of oil and/or mucous
- Insatiable hunger
- Gastrointestinal distress – gas, bloating, acid reflux
- Gastrointestinal complications – rectal prolapse, intussusception, volvulus, etc.
- Protein-calorie malnutrition and micronutrient malnutrition resulting in Failure to Thrive

3. Sathe, Meghana et al. Meconium ileus in Cystic Fibrosis. *Journal of Cystic Fibrosis.* 2017; 16: S32 – S39.

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NUTRITION IMPLICATIONS OF CYSTIC FIBROSIS

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ALTERED PHYSIOLOGY

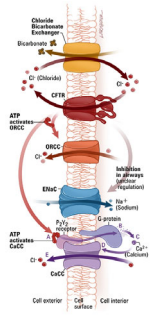
Dysfunction of CFTR (and that of other channels effected by CFTR) results in:

Electrolyte losses

- Chloride
- Sodium
- Bicarbonate
- Potassium

Systemic impact

- Lungs
- Pancreas
- Sweat glands
- Gastrointestinal tract
- Reproductive organs
- Salivary glands
- Kidneys
- Hepatobiliary system



6. Thomas L. Ratchford, Jeffrey H. Tackman and Dhiren R. Patel. Gastrointestinal pathophysiology and nutrition in cystic fibrosis. *Expert Rev Gastroenterol Hepatol.* 2018 Sep;12(9):853-862. doi: 10.1080/17474124.2018.1502463.
3. Deschamp AR, Chen Y, Wang WF, Raisi M, Hatch J, Sanders DB, Ranganathan SC, Ferkol T, Perkins D, Finn P, Davis SD. The association between gut microbiome and growth in infants with cystic fibrosis. *J Cyst Fibros.* 2023 Nov;22(6):1050-1056. doi: 10.1016/j.jcf.2023.08.003.
9. Johns Hopkins Cystic Fibrosis Center. CFTR Effects on Other Channels. Updated 2021. Accessed August 6, 2025. https://hopkinscf.org/knowledge-cftr/

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Poll #1

How often do you manage patients with CF

- A Daily
- B 1-2x weekly
- C Monthly
- D Rarely or not at all

ENDF = energy- and nutrient-dense formula

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IMPACT ON GASTROINTESTINAL (GI) TRACT

Thick sticky mucus in the GI tract, pancreas, gallbladder, and liver among other organ systems leads to:

- Impaired transit of digestive enzymes and bicarbonate from pancreas to GI tract
- Impaired bile production and transit to GI tract
- Exocrine pancreatic insufficiency
- Poor absorption of fat-soluble vitamins
- Increased acidity of GI tract

Slowed intestinal transit time

The overall environment contributes to poor nutritional status and gut dysbiosis

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MICRONUTRIENT NEEDS

- **All vitamins and minerals should be given with food and pancreatic enzyme replacement therapy (PERT)**
- Increased importance of replacing fat-soluble **vitamins A, D, E, and K**

Micronutrient:	Considerations:
Zinc	• increased losses in stool that correlate with fecal fat losses[1] • Serum levels are not reliable[1] • 6-month trial in infants who are not meeting growth standards despite optimized nutrition and PERT • 1 mg/kg/day
Magnesium	• Low magnesium levels are increasingly recognized • Increased renal loss due to aminoglycoside effect on proximal convoluted tubules • Decreased absorption • Malnutrition • No routine recommendations for monitoring
Fluoride	• Consider in infants 6 months to 2 years if community water supply contains less than 0.3 parts per million • Supplement with 0.25 mg/day

1. Ramsey B, Robinson KA, Rosenfeld M, Sawicki GS, Sabadosa KA, Spira TJ, et al. Cystic fibrosis foundation evidence-based guidelines for management of infants with cystic fibrosis. J Pediatr. 2009;155:S73-S93.
2. Borowitz D, Baker RD, Stallings V. Consensus report on nutrition for pediatric patients with cystic fibrosis. J Pediatr Gastroenterol Nutr. 2002;35:246-59.
3. Adair SM, Rowan MH, Burt BA, Kumar JV, Levy SM, Pendrys DG, et al. Recommendations for Using Fluoride to Prevent and Control Dental Caries in the United States. 2001 Centers for Disease Control and Prevention Website: https://www.cdc.gov/mmwr/preview/mmwrhtml/rr5014a1.htm. Accessed August 6, 2021.
11. Sankararaman, S., Hendrix, S. J., & Schindler, T. (2022). Update on the management of vitamins and minerals in cystic fibrosis. *Nutrition in clinical practice : official publication of the American Society for Parenteral and Enteral Nutrition*, *37*(5), 1078–1087.

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MICRONUTRIENT NEEDS – (CONT.)

Micronutrient:	Considerations:
Iron	• CF specific vitamins do not contain iron • Monitor hemoglobin and hematocrit for anemia • Ferritin is an acute phase reactant and may be falsely elevated • Low serum ferritin is consistent with deficiency
Calcium	• CF specific vitamins do not contain calcium • Assess dietary intake yearly • Calcium homeostasis often leads to normal serum levels • Hypoalbuminemia can result in falsely low levels • Hypercalcemia possibly related to hypervitaminosis D • Consider medication interactions and type of calcium supplement (carbonate, citrate, gluconate, etc.)

7. Leonard A, Schindler T, Padula L, Altman K, Vavrina K, Chin M. Cystic Fibrosis Nutrition 101: Getting Started. 9th Edition. Cystic Fibrosis Foundation; 2022.

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MICRONUTRIENT NEEDS – (CONT.)

Micronutrient:	Considerations:
Sodium	• Large amounts lost in sweat • Amount in human milk and formula may not meet the needs of an infant with CF • There is no added salt in baby foods • 0-6 months – 1/8 teaspoon daily • 6-12 months – ¼ teaspoon daily • 1+ years – liberal addition

8.Leonard LD, Robinson KA, Rosenfeld M, Orenn KD, Sabadosa KA, Spear SL et al. Cystic fibrosis foundation evidence-based guidelines for management of infants with cystic fibrosis. J Pediatr 2009;155:S73-S93.
9. Borowitz D, Baker RD, Stallings V. Consensus report on nutrition for pediatric patients with cystic fibrosis. J Pediatr Gastroenterol Nutr 2002;35:246–59.
10. Adair SM, Rowan MH, Burt BA, Kumar V, Levy SM, Pendry DG, et al. Recommendations for Using Fluoride to Prevent and Control Dental Caries in the United States 2001 Centers for Disease Control and Prevention Website: https://www.cdc.gov/mmwr/preview/mmwrhtml/rr5014a1.htm Accessed August 6, 2025

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CFTR MODULATOR THERAPY

Therapy is specific to genotype and age

Two therapies available for 0-18 months

May alter nutrition recommendations based on individual response

Lumacaftor/Ivacaftor:
- Patients with 2 copies of delF508
- ~50% of pwCF
- ≥ 1 year of age

Ivacaftor:
- Patients with Class III and/or R117H (Class IV)
- ~6% of pwCF
- ≥ 1 month of age

11. Kerem E, Corey M, Kerem BS, Rommens J, Markiewicz D, Levison H, Tsui LC, Durie P. The relation between genotype and phenotype in cystic fibrosis--analysis of the most common mutation (delta F508). N Engl J Med. 1990 Nov 29;323(22):1517-22. doi: 10.1056/NEJM199011293232203. PMID: 2233922.

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NUTRITION FOCUSED PHYSICAL EXAM (NFPE)
ANTHROPOMETRICS

Weight
- Naked or in a clean, dry diaper
- Use infant scale
- Measure to the nearest gram

Length
- Use length board with fixed headboard and adjustable footboard
- Need two people to complete measurement
- Measure to the nearest 0.1 cm

Infant Length Measurement Video - YouTube

12. DeTalia C, ed. *The Practitioner's Guide to Nutrition-Focused Physical Exam of Infants, Children, and Adolescents: An Illustrated Handbook*. Silver Spring, MD: American Society for Parenteral and Enteral Nutrition; 2019.

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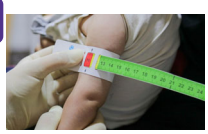
NUTRITION FOCUSED PHYSICAL EXAM (NFPE)
ANTHROPOMETRICS CONTINUED

Head Circumference
- Non-flexible tape
- Measure to the nearest 0.1 cm

Mid-Upper Arm Circumference (MUAC)
- ≥6 months of age
- Non-flexible tape
- Measure to the nearest 0.1 cm



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MUAC IN CYSTIC FIBROSIS

The authors found that MUAC detected a higher percentage of malnutrition than did BAZ
(47% vs 10%)

The study showed improvement in MUAC to be more sensitive compared to BAZ or weight changes with a nutrition intervention

There is a need for evidence-based recommendations on frequency of MUAC measurements as well as expected improvement in body composition over time

Currently there are no standardized practices

Phone RY. et al. *Nutr Clin Pract*. 2020;35(6):1094-1100.

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Poll #2

Do you use MUAC as part of your CF nutrition assessment?

- A Yes, I use MUAC at least annually
- B I use MUAC only when clinically indicated
- C I am aware of MUAC, but do not use clinically
- D This is the first time I've learned of using MUAC in CF

ENDF = energy- and nutrient-dense formula

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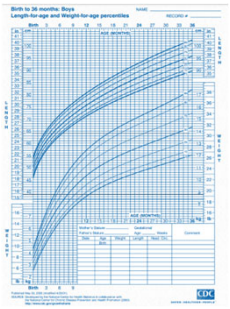
NUTRITION FOCUSED PHYSICAL EXAM (NFPE)
MEDICAL HISTORY

Chart review

- Growth chart percentiles and trajectory
- Calculate rate of weight gain and compare to age and sex matched counterparts
- Look for any recent illnesses, antibiotic use, trips to emergency department, concerns discussed at pediatrician visits, etc.
- Assess laboratory data for indication of inflammation, micronutrient deficiencies or signs of excessive intake, and fluid status



12. DeTallo C, ed. *The Practitioner's Guide to Nutrition-Focused Physical Exam of Infants, Children, and Adolescents: An Illustrated Handbook.* Silver Spring, MD: American Society for Parenteral and Enteral Nutrition; 2019

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NUTRITION FOCUSED PHYSICAL EXAM (NFPE)
INTERVIEW

Have you noticed any changes in how much or how often they are eating? How long has change been present?	Are clothes that previously fit well starting to appear loose?	Have you noticed any changes in the appearance of their skin, hair, or nails?
Are they meeting age-appropriate milestones?	Have you noticed a change in the frequency, appearance, or smell of stools?	Have there been any changes in sleep patterns?

12. DeTallo C, ed. *The Practitioner's Guide to Nutrition-Focused Physical Exam of Infants, Children, and Adolescents: An Illustrated Handbook.* Silver Spring, MD: American Society for Parenteral and Enteral Nutrition; 2019.

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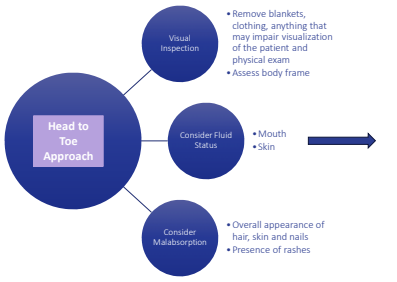
NUTRITION FOCUSED PHYSICAL EXAM (NFPE)



Fluid Status		
	Normal	**Dehydrated**
Eyes	Moist, able to produce tears	Membranes appear dry, eyes appear sunken
Mouth	Tongue moist, saliva production normal	Tongue is dry and cracked
Skin	Turgor test- Returns to original position quickly	Skin returns to original position slowly

12. DeTallo C, ed. *The Practitioner's Guide to Nutrition-Focused Physical Exam of Infants, Children, and Adolescents: An Illustrated Handbook*. Silver Spring, MD: American Society for Parenteral and Enteral Nutrition; 2019.

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NUTRITION FOCUSED PHYSICAL EXAM (NFPE)
HEAD-TO-TOE APPROACH CONTINUED

Head and Face – Muscle and Fat			
	Orbital (fat pads)	**Buccal (fat pads)**	**Temple (muscle)**
Normal	Slightly bulging fat pads	Full, round cheeks	Well-defined, flat
Mild-Moderate Malnutrition	Slightly dark circles, somewhat hollow	Flat cheeks	Slightly depressed
Severe Malnutrition	Dark circles, hollow appearance, saggy skin	Hollow, sunken cheeks	Hollow, sunken

12. DeTallo C, ed. *The Practitioner's Guide to Nutrition-Focused Physical Exam of Infants, Children, and Adolescents: An Illustrated Handbook*. Silver Spring, MD: American Society for Parenteral and Enteral Nutrition; 2019.

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NUTRITION FOCUSED PHYSICAL EXAM (NFPE)
HEAD-TO-TOE APPROACH CONTINUED

Head and Face – Nutrients and Fluid					
	Hair	**Eyes**	**Oral Cavity**	**Neck**	**Face**
Physical Sign	Loss, thinning, or lightening of color	Pale conjunctiva, burning, itching, photophobia, dull, dry membranes with white patches	Dry, cracked, red lips; bleeding gums; dry mouth, inflamed mucosa, glossitis, beefy red tongue or poor dentition	Acanthosis nigricans	Hirsutism
Possible Nutrient Findings	Protein, zinc, biotin, essential fatty acids, copper, or selenium deficiency	Iron, folate, Vitamin B12, copper, riboflavin, or Vitamin A deficiency	Riboflavin, niacin, Vitamin B complex, Vitamin C, zinc, iron, folate, or fluoride deficiency. Dehydration, high intake of simple carbohydrates, or bulimia	Insulin resistance	Obesity, polycystic ovary syndrome

12. DeTallo C, ed. *The Practitioner's Guide to Nutrition-Focused Physical Exam of Infants, Children, and Adolescents: An Illustrated Handbook*. Silver Spring, MD: American Society for Parenteral and Enteral Nutrition; 2019.

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NUTRITION FOCUSED PHYSICAL EXAM (NFPE)
HEAD-TO-TOE APPROACH CONTINUED

Upper Body- Muscle and Fat				
	Upper Arm (fat pad)	Clavicle Bone (muscle)	Acromion Bone (muscle)	Scapular Bone and Spine (muscle)
Normal	Ample fat tissue	May be visible but not prominent	Rounded curves at arms, shoulders, and neck	Bones not prominent, no depressions
Mild-Moderate Malnutrition	Some fat tissue to pinch, not ample	Some protrusion	Acromion process slightly protruding, not square	Bones may show slightly, mild depression around scapula
Severe Malnutrition	Very little space, fingers touch	Protrudes and is prominent	Squared, bones prominent	Scapula is prominent, spine depression is significant

12. DeTalla C, ed. *The Practitioner's Guide to Nutrition-Focused Physical Exam of Infants, Children, and Adolescents: An Illustrated Handbook*. Silver Spring, MD: American Society for Parenteral and Enteral Nutrition; 2019.

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NUTRITION FOCUSED PHYSICAL EXAM (NFPE)
HEAD-TO-TOE APPROACH CONTINUED

Ribs and Midaxillary Line - Fat	
	Thoracic and Lumbar (fat)
Normal	Chest is full and round, ribs not prominent, minimal visualization of iliac crest
Mild-Moderate Malnutrition	Ribs visualized with slight depressions between them, iliac crest slightly visible
Severe Malnutrition	Ribs prominent with loss of intercostal tissue, iliac crest prominent

12. DeTalla C, ed. *The Practitioner's Guide to Nutrition-Focused Physical Exam of Infants, Children, and Adolescents: An Illustrated Handbook*. Silver Spring, MD: American Society for Parenteral and Enteral Nutrition; 2019.

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NUTRITION FOCUSED PHYSICAL EXAM (NFPE)
HEAD-TO-TOE APPROACH CONTINUED

Lower Body - Muscle			
	Anterior Thigh (muscle)	Patellar (muscle)	Posterior Calf (muscle)
Normal	Well rounded, no depressions	Kneecap not prominent, muscle protrudes	Well-developed, rounded
Mild-Moderate Malnutrition	Slight depressions	Kneecap slightly more prominent	Smaller muscle
Severe Malnutrition	Considerable depressions	Kneecap is prominent; little muscle around knee	Thin overall, very little muscle definition

12. DeTalla C, ed. *The Practitioner's Guide to Nutrition-Focused Physical Exam of Infants, Children, and Adolescents: An Illustrated Handbook*. Silver Spring, MD: American Society for Parenteral and Enteral Nutrition; 2019.

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NUTRITION FOCUSED PHYSICAL EXAM (NFPE)
HEAD-TO-TOE APPROACH CONTINUED

Full Body- Nutrients and Fluid				
	Arms/Legs	**Back**	**Nails**	**Skin**
Physical Sign	Corkscrew hair	Lanugo	Spoon-shaped, dull, mottled, poor blanching	Pallor, dry/scaly, dermatitis, nonhealing wounds
Possible Nutrient Findings	Vitamin C deficiency	Energy deficiency	Iron, protein, Vitamin A, or Vitamin C deficiency	Iron, folate, Vitamin B12, Vitamin A, essential fatty acid, zinc, Vitamin C, or protein deficiency

12. DeTalia C, ed. *The Practitioner's Guide to Nutrition-Focused Physical Exam of Infants, Children, and Adolescents: An Illustrated Handbook*. Silver Spring, MD: American Society for Parenteral and Enteral Nutrition; 2019

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NFPE RESOURCE
INCORPORATING NUTRITION FOCUSED PHYSICAL EXAM INTO THE PHYSICAL ASSESSMENT OF INFANTS

Scan here to download

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DETERMINING ENERGY & PROTEIN REQUIREMENTS

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EQUATIONS FOR ENERGY AND PROTEIN

Age	Estimated energy requirements (kcals/day)	Protein (g/kg/day)
0-3 months	(89 x weight (kg) - 100) + 175 kcal	1.5*
4-6 months	(89 x weight (kg) - 100) + 56 kcal	1.5*
7-12 months	(89 x weight (kg) - 100) + 22 kcal	1.5*
13-36 months	(89 x weight (kg) - 100) + 20 kcal	1.1**

*Adequate intake; **Recommended daily allowance
Energy needs may be 1.2-2x greater than baseline calculation
Protein needs may be 1.5-2 x greater than baseline calculation

Weight (kg)	Fluid Needs
1-10	100 mL/kg
11-20	50 mL/kg * (weight – 10 kg) + 1000 mL
>20	20 mL/kg * (weight – 20 kg) + 1500 mL

Holliday-Segar Method
Fluid needs may be greater than baseline calculation

13. Beer S, Bunting K, Canada N, Rich S, Spoede E, Turybury K, ed. Texas Children's Hospital Pediatric Nutrition Reference Guide. 11th edition. 2016; 19-24.

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CONSIDERATIONS FOR MACRONUTRIENT NEEDS

Potentially increased energy and protein needs secondary to

- malabsorption
- work of breathing
- higher metabolic demand
- and/or infections

Is the patient getting enough?

- Routine monitoring with close clinic follow ups
- Accurate anthropometric measurements

7. Leonard A, Schindler T, Padula L, Altman K, Vavrina K, Chin M. *Cystic Fibrosis Nutrition 101: Getting Started*. 9th Edition. Cystic Fibrosis Foundation; 2022.

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WHEN IS ENTERAL NUTRITION INDICATED

Consider if other interventions have failed and patient exhibits one or more of the following:

- Consistent suboptimal weight gain despite apparently adequate oral intake
- Oral intake is not safe for patient
- Oral intake is maximized to patient's ability
- Disease state/illness limiting oral intake for prolonged period of time
- Unable to maintain established length-for-age percentile
- Weight-for-length below 10%ile
- Decline in major growth chart percentiles

Consideration to start enteral nutrition should be a multidisciplinary approach including the patient and family

7. Leonard A, Schindler T, Padula L, Altman K, Vavrina K, Chin M. *Cystic Fibrosis Nutrition 101: Getting Started*. 9th Edition. Cystic Fibrosis Foundation; 2022.

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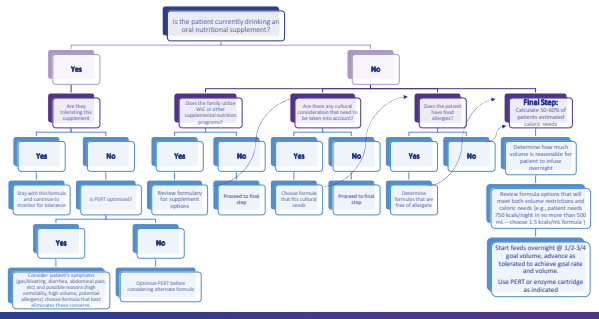
EXAMPLE DECISION TREE FOR DETERMINING ENTERAL FORMULA



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Poll #3

The most common obstacle I am faced with in managing infants with cystic fibrosis:

- A. Require concentrated formula or fortified breastmilk to achieve growth and weight gain standards.
- B. Have some form of EBM/formula intolerance necessitating a more specialized formula such as hydrolyzed infant formula
- C. Require acid suppression to improve oral intake
- D. Grow well without any intervention
- E. Other

1. ENDF = energy- and nutrient-dense formula

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MNT GOALS: CFF GUIDELINES

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CYSTIC FIBROSIS FOUNDATION (CFF) GUIDELINES

GROWTH AND FOLLOW UP

- Data from the CF Foundation Patient Registry indicate that higher body mass index (BMI) percentiles at 2 years of age are strongly associated with better pulmonary function later in childhood
- Recommendation that children reach weight-for-length of 50th percentile by 2 years of age using CDC growth charts

Age at Visit	24-48 hrs of Dx	1 week – 1 mo	2 Mo	3 mo	4 mo	5 mo	6 mo	8 mo	10mo	Q 2-3 mo in 2nd yr of life
Assess weight gain, calorie intake, PERT dosing, CF specific vitamin	Start PERT and CF specific vitamins	●	●	●	●	●	●	●	●	●
Encourage Human Milk Feeding		●	●	●	●	●				
Salt Supplementation	1/8 tsp salt						↑ ¼ tsp salt			Continue salt supplementation
Hx and PE with weight, length, OFC		●	●	●	●	●	●	●	●	●

14. Infant Care Guidelines: Cystic Fibrosis F, Borowitz D, Robinson KA, et al. Cystic Fibrosis Foundation evidence-based guidelines for management of infants with cystic fibrosis. The Journal of pediatrics. 2009;155(6 Suppl):S73-93.
15. Cystic Fibrosis Foundation. *Clinical Care Schedule for Newborns to 5 year-olds with CF*. Accessed August 13, 2025. https://www.cff.org/medical-professionals/infant-care-clinical-care-guidelines

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UNIQUE EXAMPLES & NUTRITION CHALLENGES

Challenges

- Family resistance to nutritional supplements
  - Mom strongly desires to exclusively breastfeed
  - Want to avoid extensively processed foods
  - Do not understand clinical concerns for weight gain/growth

Overcoming barriers

- Meet family where they are
- Show compassion and patience when responding to all questions and concerns
- Discuss options
  - Supplemental nursing system
  - Milk skimming
  - Starting with only one feeding of nutrition supplement daily
  - Provide a variety of supplement options and rationale for each
  - Optimize PERT
  - Add acid suppresion
  - PCP weight checks and close follow up

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WHAT IS AN ENERGY- AND NUTRIENT-DENSE FORMULA?

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ENDF: CLINICAL FEATURES

ENDFS ARE USED GLOBALLY FOR INFANTS WITH UNIQUE MEDICAL OR DIETARY CONDITIONS

- 30 kcal/fluid oz to support high energy needs and fluid restriction
- 2.6 g of protein per 100 kcals
- Lower osmolality (AAP suggests <450 mOsm/kg)
- Ready to feed/sterile
- Nutritionally complete
- Can be used to supplement infants consuming breastmilk
- Well-tolerated and supports growth

American Society for Parenteral and Enteral Nutrition. Nutrition Management of Term Infants with Growth Failure. www.nutritioncare.org. Published 2022. Accessed June 11, 2024.
Goday PS, Lewis JD, Sang Cr Jr, et al. Energy- and protein-enriched formula improves weight gain in infants with malnutrition due to cardiac and noncardiac etiologies. *JPEN J Parenter Enteral Nutr.* 2022;46(6):1270-1282. doi:10.1002/jpen.2308

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ENERGY- AND NUTRIENT-DENSE FORMULA INDICATIONS

For the dietary management of		
term infants and young children		0-18 months Up to 19lbs 13oz (9kg)
with or at risk of		
growth failure	increased energy requirements	fluid restrictions
due to conditions such as:		
Congenital Heart Disease	Chronic Lung Disease	Cystic Fibrosis
Neurological Syndromes	Respiratory Syncytial Virus	Non-Disease Related Failure-to-Thrive

ENDF: energy- and nutrient-dense formula

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POLL # 4

Have you utilized an ENDF? If so, in which populations?

A. I have utilized an ENDF for patients with respiratory conditions
B. I have utilized ENDF in infants with cystic fibrosis
C. I have heard of ENDF but have not utilized it
D. This is my first time learning about ENDF

ENDF = energy- and nutrient-dense formula

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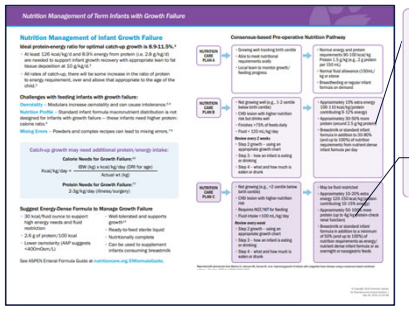
ENDF is Becoming the Standard of Care in the US

ASPEN RECOGNIZES ENDF FOR TERM INFANTS WITH GROWTH FAILURE



- Approximately 10% extra energy 100-110 kcal/kg (protein contributing 9-12% energy)
- Approximately 30-50% more protein (around 2.5 g/kg protein)
- Breastmilk or standard infant formula in addition to 30-80% (and up to 100%) of nutrition requirements from nutrient dense infant formula per day

- May be fluid restricted
- Approximately 10-20% extra energy 120-150 kcal/kg (protein contributing 10-15% energy)
- Approximately 50-100% more protein (up to 4g/kg protein-check renal function)
- Breastmilk or standard infant formula in addition to a minimum of 50% (and up to 100%) of nutrition requirements as energy/ nutrient dense infant formula or as overnight or nasogastric feeds

American Society for Parenteral and Enteral Nutrition. (May 30, 2024.) ASPEN Pathway for Term Infants With Growth Failure. https://nutritioncare.org/wp-content/uploads/2024/12/Infant-Growth-Failure-Factsheet.pdf

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IMPROVED NUTRITIONAL STATUS IN NEONATE WITH CYSTIC FIBROSIS AND NON-COMPACTION CARDIOMYOPATHY AFTER INITIATION OF ENERGY- AND NUTRIENT-DENSE FORMULA

Ryan Juel, MS, RD, LD, CACFD
Clinical Dietitian, Morgantown, WV

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PATIENT HISTORY

Baby Boy M

Patient Hx:

- 39 weeks 0 day
- Meconium ileus
- Feeding intolerance
- Positive Newborn screen for CF – homozygous delF508
- Pancreatic insufficiency requiring PERT
- Non-compaction cardiomyopathy

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MEDICAL COURSE & NUTRITION MANAGEMENT

Hospital Course Day 1

Presented to NICU on DOL 2

NPO 2/2 nonbilious emesis, feeding intolerance, failure to pass meconium

Birthweight 3.43 kg (weight-for-age 43rd%ile, z-score -0.18)

Weight at admission 3.26 kg (weight-for-age 28th%ile, z-score -0.5)

Weight loss is within expected range for age

PLAN:

- Consult Pediatric Surgery
- Fluoro gastrografin enema

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MEDICAL COURSE & NUTRITION MANAGEMENT (CONT)

Hospital Course Day 2

Enema concerning for meconium plugs

Taken for Ex-lap – meconium ileus confirmed, meconium partially evacuated from small and large bowel, and diverting double barrel ileostomy completed

No bowel resection

PLAN:

- TPN
- NPO
- Monitor ostomy output to determine when to restart feeds
- Follow CF testing results

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MEDICAL COURSE & NUTRITION MANAGEMENT (CONT)

Hospital Course Days 3-5

Newborn screen returned confirming CF diagnosis (homozygous delF508)

Mom and nurses reporting increased cough

Positive ostomy output on day 4 s/p ileostomy

PO trophic feeds of expressed breastmilk (EBM) 5mL q3h started (Mom pumping)

PLAN:

- Continue TPN
- Cautiously increase PO feeds as tolerated
- CF team to follow

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MEDICAL COURSE & NUTRITION MANAGEMENT (CONT)

Hospital Course Day 6-58

Slow increase of PO feeds, tolerated well

PERT initiated at 50 mL/feeding

TPN decreased with progression of PO feeds to meet goal of 120-130 kcal/kg/day

EBM fortified up to 22 kcal/oz with standard infant formula

Feeding intolerance (emesis) and cough persistent

Developed oral thrush and enzymes held until resolved due to acidity from applesauce increasing oral irritation

- During this time feeds oral feeds were limited and did not exceed 50 mL/feeding

Average weight gain ~25 grams/day (83% of goal)

PLAN:

- Reanastomosis
- Continue to transition to full PO feeding
- Start vitamin and salt supplementation when TPN discontinued

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MEDICAL COURSE & NUTRITION MANAGEMENT (CONT)

Hospital Course Day 59-Transfer

Successful ostomy take down

Variable post feeding emesis

Cough still present

PERT brand changed 2/2 concern for intolerance

Mom continuing to pump – most feeds are 100% EBM

Supplemental formula changed to a partially hydrolyzed formula one bottle a day fortified to 22 kcal/oz (concern for intolerance of initial standard infant formula)

Failed discharge oximetry screening -> echocardiogram showed dilated cardiomyopathy and heart failure -> transferred to Peds CICU

Diagnosed with left ventricular non-compaction cardiomyopathy

Average weight gain ~9 grams/day (30% of goal)

PLAN:

- Transfer to Pennsylvania for cardiac management and transplant workup

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MEDICAL COURSE & NUTRITION MANAGEMENT (CONT)

Outpatient Clinic Visit #1

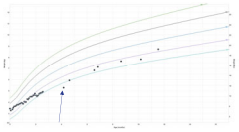
Baby Boy M is now 4 months old

Responded well to medical management of LVNC and discharged from Pittsburgh Children's Hospital

Nutrition recommendations at discharge were to continue with partially hydrolyzed formula 5-6 oz/feeding fortified to 26 kcal/oz

At time of visit, Baby Boy M was taking 4oz/feeding without fortification 2/2 poor tolerance (gas, acid reflux, and excessive spit up and stooling)

Average weight gain 8 grams/day
(27% of goal; weight-for-age 2nd%ile, z-score= -2.01)



PLAN:

Optimize PERT

Trial Energy and Nutrient Dense Formula (ENDF)

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MEDICAL COURSE & NUTRITION MANAGEMENT (CONT)

Outpatient Clinic Visit #2

Tolerating ENDF well
Taking 6 oz/feeding
Stool frequency decreased from 8-10/day to 2-3/day
Spit up and gas decreased – continued to have s/s acid reflux
Average weight gain 15 grams/day
(50% of goal; weight-for-age 6th%ile, z-score= -1.58)

Baby Boy M is now 6 months old

PLAN:

Optimize PERT
Weight adjust PPI (started in between visits)
Continue ENDF
Introduce solid foods with calorie boosters (olive, avocado, MCT oil recommended)

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MEDICAL COURSE & NUTRITION MANAGEMENT (CONT)

Outpatient Clinic Visit #3

Continues to tolerate ENDF well
Taking 6-8 oz/feeding
Enjoying/tolerating baby foods and yogurt
No acid reflux or other GI concerns
Average weight gain 13 grams/day
(65% of goal (adjusted goal for age); weight-for-age 6th%ile, z-score= -1.59)

Baby Boy M is now 8 months old

PLAN:

PERT regimen adjusted to reflect meals/snacks
Continue ENDF
Family transferring outpatient care to transplant facility due to heart condition

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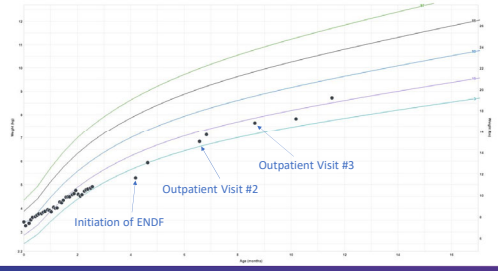
Baby Boy M Weight-for-age Growth Chart

WHO 0-2 years, boys



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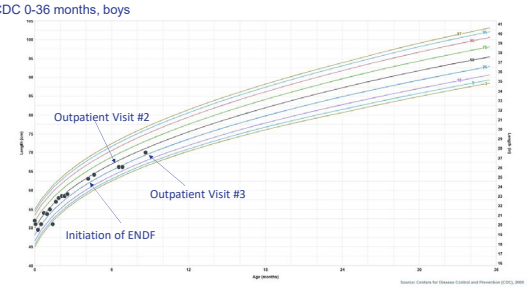
BABY BOY LENGTH-FOR-AGE GROWTH CHART

CDC 0-36 months, boys



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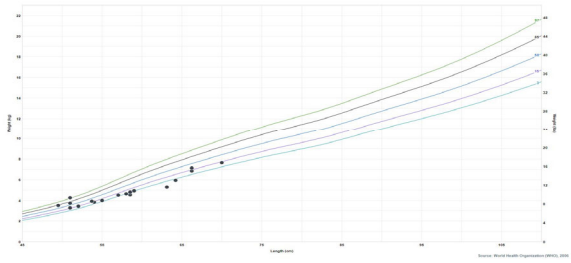
Baby Boy M weight-for-length Growth Chart

WHO 0-2 years, boys



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MEDICAL COURSE & NUTRITION MANAGEMENT (CONT)

8 month follow up

Baby Boy M was transitioned to pediatric standard supplement at 1 year of age

Mom reports he is meeting/exceeding milestones and growth expectations

ENDF was successfully used age 4 month-16 months?

Baby Boy M is now 16 months old

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DISCUSSION QUESTIONS: FOOD FOR THOUGHT

MANAGEMENT OF INFANTS WITH CF

- How do your current practices align with or differ from the recommendations presented?
- What are your most common barriers to achieving weight gain and liner growth in this population?
- How are new medications affecting your nutritional management?
- ENDF …

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QUESTIONS & DISCUSSION

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Thank you for your attendance!

Please fill out the survey to obtain your CE Certificate:

https://www.surveymonkey.com/r/HM5MBXY

Or Scan the QR Code

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