

PPC Trainee Pediatric Nutritional Reference Guide

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THE UNIVERSITY OF ARIZONA
COLLEGE OF MEDICINE TUCSON

**Pediatric Pulmonary
Center**

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Monitoring Patient Growth

Growth Expectations

Age	Males		Females	
	Grams/Day	cm/month	Grams/day	cm/month
0-1 month	36	4.5	30	4.5
1-2 months	35	4	30	3.5
2-3 months	27	3	27	3
3-4 months	23	2.5	20	2
4-5 months	17	2	17	2
5-6 months	13	1.5	13	2
6-9 months	11	1.5	10	1.3
9-12 months	8	1.3	8	1.3
12-18 months	7	1.1	7	1.1
18-24 months	7	1	7	0.8
2-6 years	6	0.6	5.5	0.4
6-7 years	5.5	0.5	5.5	0.5
7-8 years	7	0.5	8	0.5
8-9 years	8	0.5	9.5	0.5
9-10 years	9.5	0.5	11	0.4
10-11 years	11	0.5	11	0.5
11-12 years	12	0.5	12	0.6
12-13 years	14	0.6	12	0.5
13-14 years	15	0.7	9.5	0.3
14-15 years	15	0.5	7	0.2
15-16 years	12	0.3	5.5	0.1
16-17 years	9.5	0.2	3	0-0.1
17-18 years	7	0.1	4	0
18-19 years	3	0-0.1	3	0
19-20 years	7	0-0.1	3	0

References:

- (1) Adapted age 0-24 months from WHO Growth Velocity Standards 2013 (2) Adapted from age 2-20 years from CDC Growth Charts

Last Reviewed: 04/19/16 by Lindsey Vaughn, RD

Cystic Fibrosis

Expected Rate of Weight Gain (g/d) for Birth – 24 months

Age Range	Males	Females
Birth - 1 month	30	26
1-2 months	35	29
2-3 months	26	23
3-4 months	20	19
4-5 months	17	16
5-6 months	15	14
6-9 months	10-13	10
9-24 months	7-10	7-10

Adapted from *Nutrition in Cystic Fibrosis*, Yen and Radmer Leonard, Figure 6.1

Expected Rate of Weight Gain (g/d) for Ages 2-12

Age Range (years)	Males	Females
2-3	4.7-6.7	5.2-8.2
3-4	5.2-8.1	5.3-9.0
4-5	5.9-9.5	5.9-10.0
5-6	6.3-10.4	6.3-10.9
6-7	6.5-11.7	6.9-12.2
7-8	7.1-13.4	7.9-14.2
8-9	8.0-15.5	9.4-16.6
9-10	9.3-17.4	10.7-18.8
10-11	10.9-18.8	11.9-20.0
11-12	12.6-19.3	12.2-19.7

Adapted from *Nutrition in Cystic Fibrosis*, Yen and Radmer Leonard

Determining height gained cm/month:

Change from current visit – previous visit / # of months since last visit

EX: 141 cm – 133.5 cm / 10 months = **gain of .75 cm/month**

Determining weight gained gm/day

Change from current visit – previous visit x 1000 / # days between visits

EX: 34.9 kg – 32.1 kg = 2.8 x 1000 = 2,800/288 days = **gain of 9.7gm/day**

Estimated Energy Requirements (EER) Equations

Infants and Toddlers:

0-3 months	$(89 * \text{weight}[\text{kg}] - 100) + 175$
4-6 months	$(89 * \text{weight} [\text{kg}] - 100) + 56$
7-12 months	$(89 * \text{weight} [\text{kg}] - 100) + 22$
13-36 months	$(89 * \text{weight} [\text{kg}] - 100) + 20$

Boys:

3-8 Years	$EER = 88.5 - (61.9 * \text{age} [\text{yr}]) + [\text{PA} * ((26.7 * \text{wt}[\text{kg}]) + (903 * \text{ht} [\text{m}]))] + 20$
9-18 Years	$EER = 88.5 - (61.9 * \text{age} [\text{yr}]) + [\text{PA} * ((26.7 * \text{wt} [\text{kg}]) + (903 * \text{ht} [\text{m}]))] + 25$

Girls:

3-8 Years	$EER = 135.3 - (30.8 * \text{age} [\text{yr}]) + [\text{PA} * ((10 * \text{wt} [\text{kg}]) + (934 * \text{ht} [\text{m}]))] + 20$
9-18 Years	$EER = 135.3 - (30.8 * \text{age} [\text{yr}]) + [\text{PA} * ((10 * \text{wt} [\text{kg}]) + (934 * \text{ht} [\text{m}]))] + 25$

Physical Activity (PA) Coefficient

Boys	Girls
Sedentary = 1	Sedentary = 1
Low Activity = 1.13	Low Activity = 1.16
Active = 1.26	Active = 1.31
Very Active = 1.42	Very Active = 1.56

Reference: The Institute of Medicine Equation was published in September 2002

Last Revised: 07/07/15, 08/18/16 by Lindsey Vaughn, RD

Examples

Infants/toddlers	4 months old male	6.6 kg	63 cm	$((89 * 6.6) - 100) + 56$ = 545 kcal /day (round to the nearest 5)
School Age w/ low activity physical coefficient (1.16)	10-year-old girl	32 kg	138 cm	$135.3 - (30.8 * 10) + ((1.16 * ((10 * 32) + (934 * 1.38))) + 25 =$ 1720 kcal/day (round to nearest 5)

Protein and Fluid Needs

Guidelines for Protein

Age	Grams of protein/kg/day
0-12 months	1.5
1-3 years	1.1
4-13 years	0.95
14-18 years	0.85
Adults	0.8
Pregnancy/Lactation	1.1

Guidelines for Fluid: Holliday-Segar Method

Weight	Fluid Calculation
<10kg	Weight (kg) * 100 ml
10-20kg	[Weight (kg) -10] *50 + 1000 ml
>20kg	[Weight (kg) – 20] * 20 + 1500 ml

EXAMPLES

8 kg	8kg x 100 ml = 800 ml
15kg	(15kg – 10) *50 + 1000 ml = 1250 ml
30kg	(30kg – 20) * 20 + 1500 ml = 1700 ml

References:

- (1) Dietary Reference Intakes: The Essential Guide to Nutrient Requirements, <http://www.nap.edu/catalog/11537.html> (page 144)
- (2) Journal of Pediatric Pharmacology and Therapeutics > v.14(4): Oct-Dec 2009 > PMC3460795, <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3460795/>

Last Reviewed: 04/19/16 by Lindsey Vaughn, RD

Calculating Estimated Nutrition Needs for Cystic Fibrosis:

Note: Patients on modulators may require different nutritional recommendations; evaluate current BMI%tile to assess the need for additional nutrition

- To determine caloric needs, calculate the EER of the patient based on their age category. Multiply EER by 1.2 and 1.5 to determine caloric range for CF patient
- To determine protein needs, calculate RDA for protein based on client age category. Multiply calculated RDA by 1.2 and 1.5 to determine protein range for CF patient
- Fluid needs can be calculated from above fluid recommendations

Pediatric Malnutrition Classifications

Note: These indicators are a “work in progress” and should be used along with other clinical assessment tools and clinical judgment

Z-score: refers to the number of standard deviations above or below the mean value of what is being observed or studied. Z-scores that are positive are above the mean, while those that are negative are below

z-score of 0: 50thtile on a growth chart

z-score \pm 1.0: 15thtile or 85thtile on growth chart

z-score \pm 2.0: 3rdtile or 97thtile on growth chart

Choose the Highest Acuity Indicator

Single Data Point available	<i>Mild Malnutrition</i>	<i>Moderate Malnutrition</i>	<i>Severe Malnutrition</i>
Weight for height z-score (<2 years):	-1 to 1.9 z score	-2 to 2.9 z score	\leq -3 z score
BMI for age z-score: (>2 years):	-1 to 1.9 z score	-2 to 2.9 z score	\leq -3 z score
Length or height z-score:			\leq -3 z score*
Mid-upper arm circumference	-1 to 1.9 z score	-2 to 2.9 z score	\leq -3 z score

Two or More data points available	<i>Mild Malnutrition</i>	<i>Moderate Malnutrition</i>	<i>Severe Malnutrition</i>
Weight gain velocity (<2 years)	<75 th tile of norm for expected weight gain	<50% of norm for expected weight gain	<25% of norm for expected weight gain
Weight loss (2-20 years)	5% usual body weight	7.5% of usual body weight	10% of usual body weight
Deceleration in weight-for-length/height z score	Decline of one z score	Decline of 2 z score	Decline of 3 z score
<i>Inadequate nutrient intake**</i>	<i>Use this indicator cautiously without other diagnostic criteria</i>		

Chronicity: **<3 months = acute** **>3 months = chronic**

Percent weight change: weight change (in grams) divided by starting weight (in grams) x 100. For example: Patient lost 480 grams from starting weight of 12.2kg. 480 divided by 12200 = 3.9% of UBW

Growth Velocity: Divide total grams of weight gain from previous data divided by number of days

*Severe malnutrition based on length: do not use this criterion in patients with chronic disease such as chronic kidney disease, CP, CF, prematurity. Increased calories may increase fat deposition and not length increase.

****Nutrient Intake:** Be cautious in classifying malnutrition based on this indicator, especially when a patient is NPO < 5-7 days. It may need to be charted in a PES statement such as “Inadequate intake related to acute injury and inability to eat as evidenced by no oral intake for 36 hours.” Use clinical judgement with use of this indicator

References: Nutrition Services Dept., ks, kt, mk 2017, From Consensus Statement of the Academy of Nutrition and Dietetics/American Society for Parenteral and Enteral Nutrition, 2014

PES Statement Examples:

Problem (P)	Etiology (E): related to	Signs/Symptoms (S): as evidenced by
Malnutrition (mild, moderate, severe)	Illness/medical condition, inflammation, psychological factors, dietary condition	z-scores or percentages
Increased (nutrient needs, calorie needs, protein needs)	Increased metabolic demand, increased nutrient demand	Cystic fibrosis
Overweight	Increased caloric intake vs expenditure	BMI of (<i>percentile</i>)

Reading Growth Charts

Interpreting Plotted Measurements

Note: WHO growth charts should be used for all children from birth to 2 years of age to monitor growth in the United States. Use CDC growth charts for children 2-19 years of age to monitor growth in the US

- Curved lines on the growth charts show the percentiles that indicate the child's growth rank
- **EXAMPLE:** If dotted line is plotted on the 95th percentile on the CDC BMI-for-age growth chart, this indicates that 5 out of 100 children of the same age and gender in the reference population have a higher BMI-for-age.

WHO growth chart standards: 2nd and 98th percentiles are used as the outer most percentile cut offs for abnormal growth

CDC growth chart standards: 5th and 95th percentiles are used as the outer most percentile cut offs for abnormal growth

Plotting Measurements:

1. Find child's age on horizontal axis, when plotting weight-for-length find the child's length on the horizontal axis. Use a straight edge to draw a vertical line up from that point
2. Find the appropriate measurement (weight, length, stature, BMI) on the vertical axis. Use a straight edge to draw a horizontal line from that point until it intersects with the vertical line of the first plot line
3. Make a small dot where the two lines intersect

Reference: Center for Disease Control and Prevention. *Use and Interpretation of the WHO and CDC Growth Charts for Children from Birth to 20 Years in the United States*. CDC. Last updated May 2013.

<https://www.cdc.gov/nccdphp/dnpao/growthcharts/resources/growthchart.pdf>

BMI %tile

Underweight	<5 th %tile
Normal/healthy (CF recommended)	5-85 th %tile
Overweight	85-95 th %tile
Obese	>95 th %tile

CFF Recommendations for Weight-for-stature Assessment

Age Group	Recommendation weight-for-stature goal
<2 years	≥50 th percentile wt-for-length
2-20 years	≥50 th percentile BMI
≥20 years (female)	BMI ≥22
≥20 years (male)	BMI ≥23

Reference Growth Charts

Male

Boys Birth to 36 months: <https://www.cdc.gov/growthcharts/data/set1clinical/cj41l017.pdf>

Boys 2-20 years: <https://www.cdc.gov/growthcharts/data/set1clinical/cj41l021.pdf>

Boys BMI for age: <https://www.cdc.gov/growthcharts/data/set1clinical/cj41l023.pdf>

Female

Girls Birth to 36 months: <https://www.cdc.gov/growthcharts/data/set1clinical/cj41l018.pdf>

Girls 2-20 years: <https://www.cdc.gov/growthcharts/data/set1clinical/cj41l022.pdf>

Girls BMI for age: <https://www.cdc.gov/growthcharts/data/set1clinical/cj41l024.pdf>

Cystic Fibrosis Nutrition

Energy Needs: 120-150% of RDA for energy to allow for normal growth for age (multiply calculated EER by 1.2-1.5 to obtain caloric intake range)

Protein: multiply calculated RDA by 1.2-1.5 to obtain protein intake range (see pg 6 for protein requirements)

Fluids: based on age patient age range, see table 2 on page 6

Fat: 35-45% of total caloric intake

Routine Labs

Note: Results vary based on lab standards

Lab	Normal Range
Vitamin A	20-60mcg/dL
Vitamin D	≥30ng/mL
Vitamin E	3-18.4ug/mL
Vitamin K	0.2-3.2ng/mL
HgA1c	<5.7%
ALT	
AST	
Prothrombin time (Pt/INR)	9.4-12.5 H
Complete Metabolic Panel (CMP)	
Complete Blood Count	
Oral Glucose Tolerance Test (OGTT)	

Patient Goals Examples:

- Maintain BMI >50th%tile
- Increase/decrease gm/day weight goals
- Increase/decrease cm/month height goals
- Physical activity goals

Monitoring/Evaluation Examples:

- Height/weight/BMI trends
- Stooling trends
- Vitamin (A, D, E, K) lab trends

Pediatric Vitamin/Mineral Requirements

Vitamin

Infants*

Age	Vitamin A (ug/d)	Vitamin C (mg/d)	Vitamin D (ug/d)	Vitamin E (mg/d)	Vitamin K (ug/d)	Vitamin B6 (mg/d)	Vitamin B12 (ug/d)	Choline (mg/d)
0-6 months	400	40	10	4	2.0	0.1	0.4	125
6-12 months	500	50	10	5	2.5	0.3	0.5	150

*Based on Adequate Intake (AI)

Children 1-8 years old

Age	Vitamin A (ug/d)	Vitamin C (mg/d)	Vitamin D (ug/d)	Vitamin E (mg/d)	Vitamin K (ug/d)	Vitamin B6 (mg/d)	Vitamin B12 (ug/d)	Choline (mg/d)
1-3 years	300	15	15	6	30*	0.5	0.9	200*
4-8 years	400	25	15	7	55*	0.6	1.2	250*

Bold = Recommended Daily Allowance (RDA)

*Adequate Intake (AI)

Males 9-18 years old

Age	Vitamin A (ug/d)	Vitamin C (mg/d)	Vitamin D (ug/d)	Vitamin E (mg/d)	Vitamin K (ug/d)	Vitamin B6 (mg/d)	Vitamin B12 (ug/d)	Choline (mg/d)
9-13 years old	600	45	15	11	60*	1.0	1.8	375*
14-18 years old	900	75	15	15	75*	1.3	2.4	550*

Bold = Recommended Daily Allowance (RDA)

*Adequate Intake (AI)

Females 9-18 years old

Age	Vitamin A (ug/d)	Vitamin C (mg.d)	Vitamin D (ug/d)	Vitamin E (mg/d)	Vitamin K (ug/d)	Vitamin B6 (mg/d)	Vitamin B12 (ug/d)	Choline (mg/d)
9-13 years old	600	45	15	11	60*	1.0	1.8	375*
14-18 years old	700	65	15	15	75*	1.2	2.4	400*

Bold = Recommended Daily Allowance (RDA)

*Adequate Intake (AI)

Minerals

Infants*

Age	Calcium (mg/d)	Magnesium (mg/d)	Sodium (mg/d)	Iron (mg/d)	Zinc (mg/d)	Selenium (ug/d)	Copper (ug/d)	Fluoride (mg/d)
0-6 months	200	30	110	0.27	2	15	200	0.01
7-12 months	260	75	370	11	3	20	220	0.5

*Based on Adequate Intake (AI)

Bold = Recommended Dietary Allowance (RDA)

Children 1-8 years old

Age	Calcium (mg/d)	Magnesium (mg/d)	Sodium (mg/d)	Iron (mg/d)	Zinc (mg/d)	Selenium (ug/d)	Copper (ug/d)	Fluoride (mg/d)
1-3 years	700	80	800*	7	3	20	340	0.7*
4-8 years	1,000	130	1,000*	10	5	30	440	1*

*Adequate Intake (AI)

Males 9-18 years old

Age	Calcium (mg/d)	Magnesium (mg/d)	Sodium (mg/d)	Iron (mg/d)	Zinc (mg/d)	Selenium (ug/d)	Copper (ug/d)	Fluoride (mg/d)
9-13 years	1,300	240	1,200*	8	8	40	700	2*
14-18 years	1,300	410	1,500*	11	11	55	890	3*

*Adequate Intake (AI)

Bold = Recommended Dietary Allowance (RDA)

Females 9-18 years old

Age	Calcium (mg/d)	Magnesium (mg/d)	Sodium (mg/d)	Iron (mg/d)	Zinc (mg/d)	Selenium (ug/d)	Copper (ug/d)	Fluoride (mg/d)
9-13 years	1,300	240	1,200*	8	8	40	700	2*
14-18 years	1,300	360	1,500*	15	9	55	890	3*

*Adequate Intake (AI)

Bold = Recommended Daily Allowance (RDA)

Vitamin D Recommendations in Cystic Fibrosis

Vitamin D Status in CF

	US Reference Range	SI Unit
Deficiency	<20 ng/mL	<50 nmol/L
Insufficient	21-29 ng/mL	50-75 nmol/L
Sufficient	>30 ng/mL	>75 nmol/L

Reference: (1) Daley T, Hughan K, Rayas M, Kelly A, Tangpricha V. Vitamin D deficiency and its treatment in cystic fibrosis. *J of Cystic Fibrosis*. 2019;18(2):S66-S77

Recommendations

Assessment of Vitamin D status and Overall Treatment Goals

- Annually assess Vitamin D status
- Treat with cholecalciferol (D₃) to maintain serum 25(OH)D levels at least 30 ng/mL
- Check serum 25(OH)D levels at least 3 months after a dosage change
- Take a once daily Vitamin D₃ supplement or its weekly equivalent to maintain serum levels >30 ng/mL

Treating Infants from Birth to 12 Months

- Initial dose of 400-500 IU vitamin D₃ per day
- 25(OH)D levels less than 10 ng/mL, assess for rickets and manage urgently
- 25(OH)D levels that are at least 20 ng/mL but less than 30 ng/mL (and adherence to current supplemental prescription is confirmed): increase dosage to 800-1,000 IU per day
- 25(OH)D levels less than 20 ng/mL or persistent serum 25(OH)D levels of at least 20 ng/mL (with confirmed adherence to current vitamin regimen): increase to a maximum of 2,000 IU per day
- If unable to achieve a serum 25(OH)D level of at least 30 ng/mL after treatment with 2,000 IU per day vitamin D₃, consult a specialist on Vitamin D therapy

Treatment of Children Older than 12 Months to 10 Years

- Initial dose of 800-1,000 IU vitamin D₃ per day
- Serum 25(OH)D levels that are at least 20 ng/mL but less than 30 ng/mL w/ confirmed adherence to current vitamin regimen: increase dose to 1,600-3,000 IU per day
- 25(OH)D levels less than 20 ng/mL or patient has persistent levels of at least 20 ng/mL w/ confirmed adherence to current vitamin regimen: increase dose to a maximum of 4,000 IU per day

- Consult vitamin D specialist if unable to achieve serum 25(OH)D levels at 30 ng/mL or above with dosage of 4,000 IU per day

Treatment of Children above 10 Years to Adulthood

- Initial dose of 800-2,000 IU per day
- Serum 25(OH)D levels that are at least 20 ng/mL but less than 30 ng/mL w/ adherence to current vitamin regimen: increase dose to 1,600-6,000 IU per day
- 25(OH)D levels less than 20 ng/mL or those with a persistent serum level of 20 ng/mL w/ adherence to current vitamin regimen: increase to a maximum of 10,000 IU per day
- Consult vitamin D specialist if unable to achieve serum 25(OH)D levels at 30 ng/mL or above with dosage of 10,000 IU per day

Reference: (1) Daley T, Hughan K, Rayas M, Kelly A, Tangpricha V. Vitamin D deficiency and its treatment in cystic fibrosis. *J of Cystic Fibrosis*. 2019;18(2):S66-S77

Infant Sodium Requirements

Age of Infant	Sodium Requirements (per day)
Birth – 6 months	1/8 teaspoon
6 -8 months	1/4 teaspoon
8 – 10 months	1/4 teaspoon

Reference: (1) Yen EH, Leonard AR. *Nutrition in cystic fibrosis: a guide for clinicians*. Humana Press; 2015.

How is sodium supplementation provided?

- Small amounts in each feeding for the day so as not to create adversity to foods due to salty taste
- **Bottle-fed infants:** add a small amount of salt requirement to 3-4 bottles until total daily amount is distributed
- **Breast fed infants:**
 - Add to fruit puree for pancreatic enzyme dosing
 - Add to expressed breast milk in a bottle
- After 6 months salt can start being mixed in with solid foods
- Additional salt supplementation may be necessary during times of illness, extreme heat, or exercise

Medications

Cystic Fibrosis Mutations

Mutation (Class)	Description	~ percentage of patient cases
Normal	CFTR protein is created and moves to the cell surface, allowing for exchange between chloride and water	No mutation
Class I	No functional CFTR protein created	~22% of mutations
Class II	CFTR protein is created, but misfolds so is unable to make its way to the cell surface	~88% of mutations
Class III	CFTR protein is created and moves to the cell surface, however the channel gate does not open properly	~6% of mutations
Class IV	CFTR protein is created and moves to the cell surface, however channel function is faulty	~6% of mutations
Class V	Normal CFTR protein is created and moves to the cell surface but in insufficient quantities	~5% of mutations

Adapted from: (1) Cystic Fibrosis Foundation. *Know your CF mutations infographic*. CFF. Last updated September 2017. <https://www.cff.org/What-is-CF/Genetics/Know-Your-CFTR-Mutations-Infographic.pdf>

Modulator Therapy

Modulator	Description	Mutation	Age
Kalydeco (ivacaftor)	Binds to defective protein at cell surface and opens chloride channel, regulating the amount of fluids at the cell surface	Class III G551D gating mutation	4 months and older with at least one copy of 97 specified mutations (see reference)
Trikafta (elexacaftor/ivacaftor /tezacaftor)	Aids the F508del-CFTR protein form the correct shape so that it can make its way to the cell surface	Class II mutation Helps the majority of CF patients	12 years and older with at least one copy of F508del mutation or one copy of 177 specified mutations (see reference)

Orkambi (lumacaftor/ivacaftor)	Aids the F508del-CFTR protein to form into the right shape, as well as aids in the opening of the chloride channel to allow enough chloride to flow through	Class II mutation At least 2 copies of F508del mutation	2 years and older with at least 2 copies of the F508del mutation
Symdeko (tezacaftor/ivacaftor)	Helps CFTR protein form the right shape, traffics it to the cell surface, and aids in it staying there longer. Less side effects and drug interactions compared to Orkambi	Class II mutation At least 2 copies of F508del mutation	6 years and older with 2 copies of F508del mutation OR 6 years and older with at least one copy of 154 specified mutations (see reference)

Adapted from: (1) Cystic Fibrosis Foundation. *CFTR modulator therapies*. CFF. <https://www.cff.org/Life-With-CF/Treatments-and-Therapies/Medications/CFTR-Modulator-Therapies/>

In clinic notes for patients on modulator therapy:

- Recommend the lower end of caloric and protein requirements (use the EER, do not multiply by factor of 1.2-1.5) as these patients have an easier time putting on weight
- If patient is overweight/obese (or trending that way), do NOT increase EER
- Always check activity level of patient to determine energy needs
- Best clinical judgment depending on specific patient is needed to determine adequate energy requirements

Supplements

Supplement	Vitamin A	Vitamin D	Vitamin E	Vitamin K
MVW Complete Formulation[®], drops, chewable, soft gels, D3000 soft gels	9254 IU/1 mL 16,000 IU/ 1 chewable 32,000 IU/ 2 soft gels 32,000 IU/2 soft gels (D3000)	1500 IU/ 1 mL 1500 IU/ 1 chewable 3000 IU/ 2 soft gels 6000 IU/ 2 soft gels (D3000)	100 IU/ 1 mL 200 IU/ 1 chewable 400 IU/ 2 soft gels 400 IU/ 2 soft gels (D3000)	1000 mcg/1 mL 1000 mcg/1 chewable 1600 mcg/2 soft gels 1600 mcg/2 soft gels (D3000)
AQUADEKS[®], drops, chewable, soft gels	5751 IU/1 mL 18,167 IU/ 2 chewable 36,334 IU/ 2 soft gels	600 IU/1 mL 1200 IU/ 2 chewable 2400 IU/ 2 soft gels	50 IU/1 mL 100 IU/ 2 chewable 300 IU/ 2 soft gel	400 mcg/1 mL 700 mcg/2 chewable 1400 mcg/2 soft gels
Vitamax[®], drops, chewable	3170 IU/1mL 5,000 IU/1 chewable	400 IU/ 1 mL 400 IU/ 1 chewable	50 IU/ 1 mL 200 IU/ 1 chewable	300 mcg/1 mL 200 mcg/1 chewable
Poly-Vi-Sol[®], drops, Centrum[®], chewable, tablet	750 IU/ mL 3500 IU/ 1 chewable 7,000 IU/ 2 tablets	400 IU/1 mL 400 IU/ 1 chewable 800 IU/2 tablets	5 IU/ 1 mL 30 IU/ 1 chewable 60 IU/ 2 tablets	10 mcg/1 chewable 50 mcg/2 tablets

Reference: (1) Yen EH, Leonard AR. *Nutrition in cystic fibrosis: a guide for clinicians*. Humana Press; 2015

Adapted from Table 5.1 “Nutrient content of CF-specific multivitamins with zinc and select over-the-counter products”

Pancreatic Enzymes

What are Enzymes?

- Comes in capsule form, containing tiny “beads” of digestive enzyme. Enteric coating on the outside of beads allows for them to dissolve in the small intestine
 - Functions:
 - Absorption of fat, carbohydrates, and protein
 - Promote weight gain
 - Promote nutrient absorption
- Given before meals and snacks, dosage dependent on age, pancreatic enzyme used, and particular pancreatic function of client
 - INFANTS & SMALL CHILDREN: capsules can open, and beads mixed in with a food such as applesauce; can offer breastmilk or formula to infant after beads have been given
- Adherence to enzyme regimen is very important in ensuring the best nutritional outcomes for the patient, and reduce the risks associated with malabsorption

FDA Approved Pancreatic Enzymes

Brand Name	Amounts in Units per Dosage		
	Lipase	Protease	Amylase
Creon	<u>3,000</u>	9,500	15,000
	<u>6,000</u>	19,000	30,000
	<u>12,000</u>	38,000	60,000
	<u>24,000</u>	76,000	120,000
	<u>36,000</u>	114,000	180,000
Pancreaze	<u>4,200</u>	10,000	17,500
	<u>10,500</u>	25,000	43,750
	<u>16,800</u>	40,000	70,000
	<u>21,000</u>	37,000	61,000
Pertzye (bicarbonate buffered)	<u>8,000</u>	28,750	30,250
	<u>16,000</u>	57,500	60,500
Ultresa	<u>13,800</u>	27,600	27,600
	<u>20,700</u>	41,400	41,400
Zenpep	<u>3,000</u>	10,000	16,000
	<u>5,000</u>	17,000	27,000
	<u>10,000</u>	34,000	55,000
	<u>15,000</u>	51,000	82,000
	<u>20,000</u>	68,000	109,000
	<u>25,000</u>	85,000	136,000
	<u>40,000</u>	136,000	218,000

Bowel Movements

Indicator	Normal	Malabsorption
Frequency	1 or 2 times daily	3 or more daily, or none if obstructed
Color	Brown	Very light brown
Shape	Solid	Loose, fills toilet bowl
Amount	1 flush	Many flushes
Smell	Mild	Strong
Sink or Float	Sink	Float
Oil or Mucus	No signs of oil or mucus	Oils slicks (red or orange in color) or mucus

Other signs of Malabsorption w/ Pancreatic Insufficiency

- Slow/no weight gain and/or growth but continuing to eat a lot of food
- Hungry all the time
- Frequent gas
- Belly pain/cramps
- Feeling bloated or full

Reference:

- (1) Cystic Fibrosis Foundation. *Enzyme card*. MVW Nutritionals. Last updated 2016. <https://www.cff.org/Life-With-CF/Daily-Life/Fitness-and-Nutrition/Nutrition/Taking-Care-of-Your-Digestive-System/Enzyme-Card.pdf>
- (2) Cystic Fibrosis Foundation. *Nutrition: pancreatic enzyme replacement in people with cystic fibrosis*. CFF; 2006. <https://www.cff.org/PDF-Archive/Pancreatic-Enzyme-Replacement.pdf>

Further Reference Articles & Websites

Website:

1. Used to calculate # of days between patient visits (used in calculating grams gained per day in clinic):
<https://www.timeanddate.com/date/durationresult.html?m1=7&d1=13&y1=2020&m2=11&d2=23&y2=2020&ti=on>
2. Full List of Vitamin/Mineral Recommendations:
 - a. Vitamins:
<https://www.ncbi.nlm.nih.gov/books/NBK56068/table/summarytables.t2/?report=objectonly>
 - b. Minerals:
https://www.ncbi.nlm.nih.gov/books/NBK545442/table/appJ_tab3/?report=objectonly

Additional Reference Articles:

1. ***A good article on general information related to Cystic Fibrosis:***
 - a. Sullivan JS and Mascarenhas MR. Nutrition: prevention and management of nutritional failure in cystic fibrosis. J of Cystic Fibrosis. 2017;16:S87-S93.
2. ***Vitamin D Article:***
 - a. Daley T, Hughan K, Rayas M, Kelly A, Tangpricha V. Vitamin D deficiency and its treatment in cystic fibrosis. J of Cystic Fibrosis. 2019;18(2):S66-S73
3. ***CF Modulators and their effects on gut microbiome and inflammation:***
 - a. Ooi CY, Syed SA, Rossi L, et al. Impact of CFTR modulation with Ivacaftor on gut microbiota and intestinal inflammation. Sci Rep. 2018;8(1):17834.
4. ***CF-related Diabetes and Nutritional Considerations:***
 - a. Kaminski BA, Goldsweig BK, Sidhaye A, et al. Cystic fibrosis related diabetes: nutrition and growth considerations. J of Cystic Fibrosis. 2019;18(2):S32-S37
5. ***Further information on Pancreatic Enzymes:***
 - a. Singh VK and Schwarzenberg SJ. Pancreatic insufficiency in cystic fibrosis. J of Cystic Fibrosis. 2017;16:S70-S78.