Cystic Fibrosis Nutrition (Peds)

Kimberly Stephenson, RD

CHEAT SHEET

NC Children’s Hospital, February 2011

a. CLASSIFICATION

<table>
<thead>
<tr>
<th>Nutritional Status</th>
<th>BMI %ile or WT/HT %ile</th>
<th>Height %ile</th>
</tr>
</thead>
<tbody>
<tr>
<td>OUTSTANDING</td>
<td>≥ 50th</td>
<td>&gt;50th%ile and &gt;genetic potential</td>
</tr>
<tr>
<td>ACCEPTABLE</td>
<td>25th to 49th</td>
<td>&gt;50th%ile and &gt;genetic potential</td>
</tr>
<tr>
<td>AT RISK</td>
<td>10th to 24th</td>
<td>&lt; genetic potential</td>
</tr>
<tr>
<td>URGENT NEED</td>
<td>&lt; 10th</td>
<td>&lt; 5th%ile</td>
</tr>
</tbody>
</table>

b. DIET

1) High calorie, high protein, high fat, high salt diet
2) Calories:
   1. DRI x (1.2 if Outstanding) or (1.3 if Acceptable) or (1.4 if At Risk) or (1.5 if Urgent Need)
   2. Infants 130-150 cals/kg
3) Protein: DRI x 1.5 to 2.0
4) Oral Supplements: add for extra calories (Pediasure 250cals, Boost Kid Essentials 1.5 375cals, EnsurePlus 365cals, Scandishake 600cals, VHC 560cals, Carnation Instant Breakfast Plus 360cals, Resource Breeze 250cals)
5) Restrict tyramine if on LINEZOLID (Zyvox) until 2 weeks after d/c med. Tyramine handout @ http://intranet.unchealthcare.org (Select Patient Ed→Food Drug→MAO Inhib. Mark out “An MAO Inhibitor is an antidepressant… less sad & depressed.” CF pts use for lung therapy).
6) Restrict oxalate if have calcium-oxalate kidney stone. Need adequate fluid, RDI Ca+, no Na+ restriction. Oxalate handout @ NCM online (Select Client Educ→Renal→Urin Stone, pg3).
7) Appetite stimulants
   a) Periactin (generic cyproheptadine)
      i) Children >5years: 4mg BID-QID, comes as a 4mg tablet
      ii) Children <5years, 0.25-0.5 mg/kg/day given as a divided dose (BID), comes as 2mg/5ml suspension
      iii) Side effects: sleepiness, moodiness. To avoid sleepiness, give evening dose only for 1 week or until wears off, then increase as tol to BID or TID.
   b) Megace
      i) Rarely used in pediatrics
      ii) Suspension 200mg/5ml or tablet 10mg
      iii) Begin 200mg/day, max 800mg/day. Usually get good effect at 400mg/day.
      iv) Side effects: elevated glucose, adrenal suppression, amenorrhea for women, decreased sex drive, breast enlargement in males, decreased bone density
   c) Marinol (generic dronabinol)
      i) Rarely used in pediatrics
      ii) Capsule comes as 2.5mg, 5mg, 10mg
      iii) Begin 2.5mg QD-BID, max dose 20mg/day

3. ENZYMES

a. Avoid fibrosing colonopathy by keeping enzyme dose in safe range

<table>
<thead>
<tr>
<th>Max Dosing Ranges</th>
<th>2500-3000 units of lipase/kg/MEAL and 10,000-15,000 units of lipase/kg/DAY</th>
</tr>
</thead>
<tbody>
<tr>
<td>The Calculation</td>
<td>(Enzyme # after name: 5,10, etc) x (#caps at meal OR daily) x (if non-Creon multiply by 1000) x (Weight in kg)</td>
</tr>
</tbody>
</table>

b. Take enzymes immediately prior to meals and snacks

c. Use brand name products only (Pancreaze®, Creon®, Zenpep®);
   1. “Generics” are not AB rated equivalents; illegal to substitute for brand names
   2. Write on prescriptions: “Brand name medically necessary; Do not substitute”
**PANCREATIC ENZYMES**

<table>
<thead>
<tr>
<th>Product</th>
<th>Manufact.</th>
<th>Lipase</th>
<th>Protease</th>
<th>Amylase</th>
<th>Special Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Creon 6000</td>
<td>Abbott</td>
<td>6,000</td>
<td>19,000</td>
<td>30,000</td>
<td></td>
</tr>
<tr>
<td>Creon 12000</td>
<td>Abbott</td>
<td>12,000</td>
<td>30,000</td>
<td>60,000</td>
<td></td>
</tr>
<tr>
<td>Creon 24000</td>
<td>Abbott</td>
<td>24,000</td>
<td>76,000</td>
<td>120,000</td>
<td></td>
</tr>
<tr>
<td>Pancreaze 4200</td>
<td>J&amp;J (McNeil)</td>
<td>4,200</td>
<td>10,000</td>
<td>17,500</td>
<td>Capsules, enteric-coated microtablets, porcine origin</td>
</tr>
<tr>
<td>Pancreaze 10500</td>
<td>J&amp;J (McNeil)</td>
<td>10,500</td>
<td>25,000</td>
<td>43,750</td>
<td></td>
</tr>
<tr>
<td>Pancreaze 16800</td>
<td>J&amp;J (McNeil)</td>
<td>16,800</td>
<td>40,000</td>
<td>70,000</td>
<td></td>
</tr>
<tr>
<td>Pancreaze 21000</td>
<td>J&amp;J (McNeil)</td>
<td>21,000</td>
<td>37,000</td>
<td>61,000</td>
<td></td>
</tr>
<tr>
<td>Zenpep 5000</td>
<td>Eurand</td>
<td>5,000</td>
<td>17,000</td>
<td>27,000</td>
<td>Delayed-release capsules, porcine origin,</td>
</tr>
<tr>
<td>Zenpep 10000</td>
<td>Eurand</td>
<td>10,000</td>
<td>34,000</td>
<td>55,000</td>
<td></td>
</tr>
<tr>
<td>Zenpep 16800</td>
<td>Eurand</td>
<td>15,000</td>
<td>51,000</td>
<td>82,000</td>
<td></td>
</tr>
<tr>
<td>Zenpep 20000</td>
<td>Eurand</td>
<td>20,000</td>
<td>68,000</td>
<td>109,000</td>
<td></td>
</tr>
</tbody>
</table>

**d. Adolescents will swallow capsule whole**

**e. Infants/children will open capsule and swallow enteric coated beads within; can mix with any baby fruit puree, applesauce, jelly, ketchup. Do not chew beads. Infants – can use pacifier.**

**f. Antacid/PPI will help enzymes activate (Zantac, Prilosec, Nexium)**

**4. VITAMINS**

a. **Recommendations**

<table>
<thead>
<tr>
<th>Needs for CF:</th>
<th>Vitamin A (IU)</th>
<th>Vitamin D (IU)</th>
<th>Vitamin E (IU)</th>
<th>Vitamin K (mg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-1 yr</td>
<td>1500</td>
<td>400</td>
<td>40-50</td>
<td>0.3 – 0.5mg qd (300-500mcg)</td>
</tr>
<tr>
<td>1-3 yrs</td>
<td>5000</td>
<td>400</td>
<td>80-150</td>
<td></td>
</tr>
<tr>
<td>4-8 yr</td>
<td>5000-10,000</td>
<td>400-800</td>
<td>100-200</td>
<td></td>
</tr>
<tr>
<td>&gt;8 yr</td>
<td>10,000</td>
<td>800</td>
<td>200-400</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Lab to Check:</th>
<th>Vitamin A Goal &gt;25 mcg/dL</th>
<th>(25OH) Vit D Goal Total &gt;30</th>
<th>Vitamin E Per lab range</th>
<th>PT (indirect measure) or PIVKA-II (direct msr)</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>ADD for Deficiency</th>
<th>Vitamin A Cholecalciferol 400units daily x 3months.</th>
<th>Give 50,000 ergocalciferol (Drisdol) twice weekly x 3months. If normal then give maintenance dose of 50,000units once monthly forever. *If still low, can increase to 3times weekly, or QD. **If still low, change to cholecalciferol 1000-3000 units QD.</th>
<th>200 units daily x 1-3mo</th>
<th>a. If PT high, give 5mg/d (2.5mg if under 1 year) until d/c antibiotics</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;5yrs</td>
<td>10,000 units daily x1-3mo</td>
<td></td>
<td></td>
<td>b. If PT normal and on antibiotics, give 5mg (2.5mg if under 1 year) twice weekly as prophylaxis. D/C when d/c antibiotics.</td>
</tr>
<tr>
<td>&gt;5yrs</td>
<td>20,000 units daily x 1-3mo</td>
<td></td>
<td></td>
<td>c. May need more with CF liver disease: 10-20mg/day</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Deficiency Symptoms</th>
<th>Night Blindness</th>
<th>Bone Disease</th>
<th>Hemolytic Anemia</th>
<th>↑ Bleeding/Brusing**</th>
</tr>
</thead>
</table>

*Also consider sun exposure 15min twice weekly, no sunscreen.

**Deficiency ↑es with antibiotic use that kills vitamin K-producing gut flora

b. Preparations available - ADEK (or equivalent) is an oral multi-nutrient preparation containing high levels of the fat-soluble vitamins A, D, E, and K. These vitamins are present in their water miscible forms, which are thought to improve their absorption. They also contain vitamins C, B complex vitamins, folic acid, and zinc to supplement dietary intake.
### Vitamin Preparations

<table>
<thead>
<tr>
<th>Manufacturer</th>
<th>ADEK/AquADEK</th>
<th>VITAMAX</th>
<th>SourceCF ABDEK (formulary)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Scandipharm</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>VITAMIN PREPARATIONS</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>MVI + Zn and CoQ10 in Aquadek</td>
<td>Multivitamin + zinc</td>
<td>Multivitamin + zinc</td>
<td></td>
</tr>
<tr>
<td>Products</td>
<td>ADEK: chewable tablet</td>
<td>Liquid or chewable tablet</td>
<td>Liquid, chew tab or gelcap</td>
</tr>
<tr>
<td>Flavor</td>
<td>ADEK chew: unflavored</td>
<td>Liquid: cherry, grape, orange</td>
<td>Liquid: citrus</td>
</tr>
<tr>
<td>Dosing</td>
<td>Liquid: 0-1 year = 0.5ml BID</td>
<td>Tablets: 3-9 years = 1 tab qd</td>
<td>10+ years = 1 tab BID</td>
</tr>
<tr>
<td>Availability</td>
<td>Any pharmacy</td>
<td>Only at CF mail order pharm.</td>
<td>CF Pharm, manufacturer, and local pharmacy</td>
</tr>
<tr>
<td>Programs</td>
<td>None</td>
<td>Free w/ Creon’s CareForward Program (non-Medicaid, non-Tricare):</td>
<td>Free w/ Creon CareForward Program or Zenpep ZPoints (non-Medicaid, non-Tricare)</td>
</tr>
<tr>
<td></td>
<td></td>
<td><a href="http://www.cfcareforward.com">www.cfcareforward.com</a></td>
<td><a href="http://www.cfcareforward.com">www.cfcareforward.com</a></td>
</tr>
</tbody>
</table>

Note: CF pts may also be able to receive free vitamins with other inhaled mail-order meds (ask RN).

### Vitamin Contents

<table>
<thead>
<tr>
<th>Vitamin</th>
<th>ADEK/AquADEK</th>
<th>VITAMAX</th>
<th>SourceCF ABDEK (formulary)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vitamin A (palmitate + beta-carotene)</td>
<td>5751 IU (87% beta-carotene)</td>
<td>3170 IU (all retinol palmitate)</td>
<td>4627 IU (75% beta-carotene)</td>
</tr>
<tr>
<td>Vitamin D</td>
<td>400 IU (cholecalciferol)</td>
<td>400 IU (cholecalciferol)</td>
<td>500 IU</td>
</tr>
<tr>
<td>Vitamin E</td>
<td>50 IU (d-alpha-tocopherol)</td>
<td>50 IU (dl- or tocopheryl acetate)</td>
<td>50 IU (dl- or tocopheryl acetate)</td>
</tr>
<tr>
<td>Vitamin K</td>
<td>400 mcg</td>
<td>300 mcg</td>
<td>100 mcg</td>
</tr>
<tr>
<td>Vitamin C</td>
<td>150 mcg</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Vitamin B1</td>
<td>150 mcg</td>
<td>15 mcg</td>
<td>15 mcg</td>
</tr>
<tr>
<td>Vitamin B2</td>
<td>0</td>
<td>3 mcg</td>
<td>3 mcg</td>
</tr>
<tr>
<td>Niacin</td>
<td>6 mg</td>
<td>0.6 mg</td>
<td>0.6 mg</td>
</tr>
<tr>
<td>Folic Acid</td>
<td>10 mcg</td>
<td>10 mcg</td>
<td>0.6 mg</td>
</tr>
<tr>
<td>Pantothenic Acid</td>
<td>10 mcg</td>
<td>10 mcg</td>
<td>0.6 mg</td>
</tr>
<tr>
<td>Zinc</td>
<td>3 mg</td>
<td>3 mg</td>
<td>0</td>
</tr>
</tbody>
</table>

**VITAMIN PREPARATIONS**

<table>
<thead>
<tr>
<th>Vitamin</th>
<th>ADEK/AquADEK</th>
<th>VITAMAX</th>
<th>SourceCF ABDEK (formulary)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vitamin A (palmitate + beta-carotene)</td>
<td>9000 IU (60% beta-carotene)</td>
<td>10000 IU (88% beta-carotene)</td>
<td>18167 IU (92% beta carotene)</td>
</tr>
<tr>
<td>Vitamin D</td>
<td>400 IU (cholecalciferol)</td>
<td>400 IU (cholecalciferol)</td>
<td>800 IU</td>
</tr>
<tr>
<td>Vitamin E</td>
<td>150 IU (succinate)</td>
<td>200 IU (dl- or tocopherol)</td>
<td>150 IU</td>
</tr>
<tr>
<td>Vitamin K</td>
<td>150 mcg</td>
<td>200 mcg</td>
<td>100 mcg</td>
</tr>
<tr>
<td>Vitamin C</td>
<td>60 mg</td>
<td>60 mg</td>
<td>100 mcg</td>
</tr>
<tr>
<td>Vitamin B1</td>
<td>60 mg</td>
<td>60 mg</td>
<td>100 mcg</td>
</tr>
<tr>
<td>Vitamin B2</td>
<td>60 mg</td>
<td>60 mg</td>
<td>100 mcg</td>
</tr>
<tr>
<td>Niacin</td>
<td>60 mg</td>
<td>60 mg</td>
<td>100 mcg</td>
</tr>
<tr>
<td>Folic Acid</td>
<td>300 mcg</td>
<td>300 mcg</td>
<td>100 mcg</td>
</tr>
<tr>
<td>Pantothenic Acid</td>
<td>100 mcg</td>
<td>100 mcg</td>
<td>100 mcg</td>
</tr>
<tr>
<td>Zinc</td>
<td>7.5 mg (oxide)</td>
<td>7.5 mg (oxide)</td>
<td>7.5 mg (oxide)</td>
</tr>
</tbody>
</table>

**Flavor**

- ADEK chew: unflavored
- ADEK chew: citrus
- ADEK chew: black cherry
- Liquid: cherry, grape, orange
- Liquid: bubble gum

**Dosing**

- Liquid: 0-1 year = 0.5ml BID
- 1-3 years = 1ml BID
- Tablets: 3-9 years = 1tab qd
- 10+ years = 1 tab BID

**Availability**

- Any pharmacy
- Only at CF mail order pharm.
- CF Pharm, manufacturer, and local pharmacy

**Programs**

- None
- Free w/ Creon’s CareForward Program (non-Medicaid, non-Tricare):
  - www.cfcareforward.com
  - 1-855-care4wd
- Free w/ Creon CareForward Program or Zenpep ZPoints (non-Medicaid, non-Tricare):
  - www.cfcareforward.com
  - www.zenpep.com

**Note:** CF pts may also be able to receive free vitamins with other inhaled mail-order meds (ask RN).
5. MINERALS

<table>
<thead>
<tr>
<th>Usual Needs</th>
<th>Sodium</th>
<th>Calcium</th>
<th>Zinc</th>
</tr>
</thead>
<tbody>
<tr>
<td>For 0-6 months: 1/8 tsp QD table salt (1 salt pack). For 6-12 months: ¼ tsp QD. After 1yr, salt foods.</td>
<td>At least DRI for age (800-1300mg/d), &gt;4 servings per day of hi Ca+ foods</td>
<td>At least RDA for age</td>
<td></td>
</tr>
</tbody>
</table>

| Supplements | | |
|-------------|--------|---------|------|
| With exercise, hot weather. Table salt | 500-1500mg/d if diet low | Add 25-50mg/d x6mos if FTT. Infant or toddler, dose 1-2mg/kg/d. A 220mg ZnSO4 cap = 50mg elemental (OTC). Dissolve 1cap in 5ml water. Then 1ml = 10mg elemental zn. |

| Deficiencies | | |
|-------------|--------|---------|------|
| Hyponatremic dehydration | Osteopenia/osis | Poor growth, short stature |

6. TUBE FEEDING

a. Calories: Provide 1/3 to 1/2 of total calorie needs for supplemental feeds
b. Formula
   1. Older children and adults use 2cal/ml formula for max calories/volume (Nutren2.0, TwoCalHN, VHC 2.25)
   2. 1-5 years old use 1.0 to 1.5 cal/ml pediatric formula
   3. Infants use infant formula up to max concentration of 30cal/oz
   4. If severe steatorrhea, liver disease, or short bowel may need product with hydrolyzed protein and high MCT oil (Pregestimil for infants, Peptamen Junior for 1-5 years, or Peptamen1.5 for older children)
c. Schedule
   1. Overnight continuous – tailor hours to sleep schedule
   2. Daytime bolus – morning and evening and/or afternoon. Works well for toddlers that get tangled in nighttime gtube tubing and for adults who must wake in the middle of the night to care for their own children.
d. Enzyme dose calculation
   1. For ≤12hour infusion, give usual mealtime dose PO of enzymes at both the beginning and end of infusion
   2. For >12hour continuous infusion
      a. Using enteric-coated enzymes, give usual mealtime dose of enzyme every 4-6hours
      b. Give 1800 units of lipase per gram of LCT in formula, in divided doses every 4 hours.
      Calculation = (Total grams of LCT in 24hrs of formula x 1800) / (6 if q4h) -or- 4 (if q6h)

   NOTE: Enzymes must be held when tube feeds are held!
e. Enzyme administration method
   1. Preferred method is PO
   2. If unable to give PO – see protocols on N: drive (summaries are listed below)
      a. If Mic-key GT button is ≥16F, can give intact Creon® brand enzyme beads via GT
         i. Pour into tubing, follow with water flush
      b. Crush beads and mix with formula
         i. Use with any enteral feeds (including NGT, OGT, JT) ≥ 14 Fr
         ii. Open capsule and crush beans with a mortar and pestal.
         iii. Start with 1 enzyme per can of formula and titrate as needed
         iv. Must use semi-elemental formula Peptamen 1.5 (others congeal)
         v. Swirl crushed beads into formula can, wait 30 minutes before adding to bag to prevent clogging.
   c. UNC official policy for inpatients is to use bicarbonate solution:
      i. Flush enteral access device with water, includes gastric (NGT, OGT, PEG, mic-key button), duodenal, or jejunal
      ii. Obtain an empty 60 mL oral syringe. Remove plunger and add 10 mL of water. For small pediatric patients, a smaller size syringe with less volume should be considered.
iii. Open 1 Creon capsule (24,000 unit size, but others can be used) and add capsule contents to the syringe.

iv. Add 1 sodium bicarbonate 648 mg tablet to the syringe (don’t need to crush). [Note: Can also mix 2 capsules of Creon w/ 2 tabs bicarb in same amount of water. We haven’t tested other combinations.]

v. Let mixture sit for about 15 minutes, shaking once or twice. Do not let sit for longer than 30 minutes.

vi. Administer via enteral access device.

vii. Flush with an additional 10 mL of water following administration.

7. INFANT FORMULA
   a. Generally, thrive with MBM or standard formula once enzymes initiated (PI)
   b. May need increased calories per ounce (max 30cal/oz)
   c. Use protein hydrolysate/high MCT oil formulas in severe FTT, SBS, liver dz (Pregestimil®)

8. BONE DISEASE
   a. Osteopenia/porosis common in children and adults
   b. Risks: malabsorption, FTT, hepatobiliary dz, ↓ wt-bearing activity, steroid use
   c. Check Total (25OH) Vitamin D levels and supplement for levels <30.
      1. 0-5yrs: 400units cholecalciferol daily – use OTC liquid (Enfamil’s D-vi-sol 400/ml), tab (crush) or soft gel (pierce).
      2. >5yrs: 50,000 IU ergocalciferol twice weekly for 3 months and recheck.
      3. Those who replete after twice/weekly therapy should then continue maintenance dose.
      4. If still low, ↑ frequency of dosing to 3times weekly or daily if very low (6-10range)
      5. If still low, change to cholecalciferol (this is in all OTC products) 1000-3000 units/day and/or 50,000units (Replesta brand) weekly.
   d. Consider DEXA scan for >8yrs with: pre/post lung transplant, ESLD, bone fracture, chronic steroids, delayed puberty, FTT
   e. Therapy: supplement Ca+, vitamin D, vitamin K as needed, wt-bearing activity, consider anti-resorptive drug therapy (bisphosphonates, IV pamidronate)

9. CF RELATED DIABETES
   a. Screen yearly and at admit via random glucose (goal <126)
   b. Screen >10yo with 2hour OGTT (lab draw only)
   c. Therapy: Insulin + Carbohydrate Counting. Do not restrict diet except no regular sodas.
   d. Diagnosis:
      i. 2-h OGTT plasma glucose >200 mg/dl (11.1 mmol/l)
      ii. FPG ≥126 mg/dl (7.0 mmol/l)
      iii. A1C ≥ 6.5% (A1C<6.5% does not rule out CFRD because this value is often spuriously low in CF.)
      iv. Classical symptoms of diabetes (polyuria and polydipsia) in the presence of a casual glucose level >200 mg/dl (11.1 mmol/l)

10. FECAL FAT STUDIES
    a. Fecal Elastase: (PE-1) determines pancreatic insufficiency, requires random stool sample
       1. Child must be >2weeks old. Can be on enzymes. First choice for initial dx of PI.
       2. Results: <100=Severe PI, 100-200=Mild/Moderate PI, 200-480 Low PS, >480 = PS
    b. Qualitative fecal fat: "spot check" of one stool sent to lab
       1. May need series to get accurate measure
       2. Receive qualitative result: normal, slight, moderate increase
    c. Quantitative fecal fat: 72hr collection of stool with (detailed) calorie count
       1. Requires accurate collection of stool (>200g); difficult w/ diapers (absorbed)
       2. Requires accurate food record - total grams of fat eaten per day
       3. Hang “ATTENTION–Fecal Fat Study” sheet (N: drive) on outer bathroom door to alert staff
       4. Ignore lab range; calculate "coefficient of fat absorption" (CFA). < 93% = malabsorption.
          Calculation of CFA% = (Average grams of fat eaten/24hr) - (grams of stool fat/24hr) x 100
11. EDUCATION RESOURCES
   a. www.cff.org  Search on “nutrition” or “enzymes” or “bone”
   b. www.portcfc.org Password/login granted by CF Center Director

12. OTHERS
   a. Essential Fatty Acid Deficiency (EFAD) - ↓ tetraene:triene ratio, assoc. w/ FTT. More common with short bowel syndrome, liver disease. We have treated with fish oil, IV lipids.
   b. Pubertal assessment - treat delayed puberty with nutrition and/or hormones

13. New Diagnoses
   a. All
      i. Fecal elastase after child is greater than 2 weeks old to determine PS/PI
      ii. If pancreatic insufficient or 2 CFTR mutations associated with PI
         1. Begin enzymes at 500-1000 units lipase/kg/feeding
         2. Begin CF specific vitamin at age-appropriate dose.
      iii. Table salt
         1. Infants up to 6 months: 1/8th teaspoon once daily; titrate as needed. (Or 1 salt packet).
         2. Infants 6 months up to 1 year: ¼ teaspoon once daily or divided.
         3. Children over 1 year: salt all table foods; Gatorade as needed.
         4. Goal is 2-4 mEq/kg/day of supplemental sodium
      iv. Enroll in enzyme-specific programs for non-medicaid/care, non-TriCare)
         1. Creon
            a. Solvay Cares program
               i. www.cfcareforward.com Enrollment forms in clinic.
               ii. For non-medicaid, non-tricare patients only
               iii. Provides free vitamins (vitamax or SourceCF brand) with creon purchase
               iv. Provides free supplement (Pediasure, EnsurePlus, Enlive)
               v. $50/month copay assistance
      2. Pancreaze
         a. Instant Savings Card used at pharmacy for up to $50 off their copay at the point of sale. We have been unable to obtain these.
         b. http://www.pancreaze.net/savingscardhcp
      3. Zenpep
         a. Zpoints Program
            i. www.zenpep.com; info in clinic
            ii. Earn points with filled prescription. Redeem for supplements, vitamins, amazon gift card, Hypersal
            iii. Free Source CF liquid, chewable, of softgel
            iv. Nutrabalance shakes or cookies
            v. $50/month copay assistance
   a. Newborns
      i. MBM or formula of choice. Increase caloric density as needed to meet calorie needs.
      ii. Teaching on nutrition goals
         1. Use age-specific handouts from CFF (on N: drive)
         2. Use pancreatic enzyme handout from CFF (on N: drive)
         3. Show enzyme beads and how to administer, store
         4. Review growth goal of wt/ht >50%ile; 20-30g/day weight gain in newborns

REFERENCES:
3. Clinical Practice Guidelines for Cystic Fibrosis, 1997; Cystic Fibrosis Foundation, Bethesda, MD.