Nutrition in Cystic Fibrosis

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Objectives

• Cystic fibrosis (CF) - genetics, disease background
• Reasons for malnutrition unique to CF
  - pancreatic insufficiency
  - recurrent infections
  - CF related diabetes
• Nutritional status and its relation with clinical outcomes
• Evidence for nutritional strategies in CF
Inheritance of Cystic Fibrosis

- Genetic disorder - inherited in autosomal recessive manner

Both parents are carriers for CF

Child must inherit both genes to have CF

1 in 4 children will have CF
2 will be carriers for CF
1 will be normal
Genetics of CF

- Caused by mutation in CFTR gene, chromosome 7
- CFTR = cystic fibrosis transmembrane regulator
- Encodes chloride channel and ion conductance regulator → defect in epithelial ion transport in various organs
Hallmarks of CF

Chronic respiratory disease

- Pancreatic insufficiency
- Fat malabsorption
- Failure to thrive

Elevation of sweat chloride

Health Problems with Cystic Fibrosis

- Sinus Problems
- Nose Polyps (growths)
- Frequent lung infections
- Enlarged heart
- Trouble breathing
- Gallstones
- Abnormal pancreas function
- Trouble digesting food
- Salty sweat
- Fatty BM's
Cystic Fibrosis today

- 30,000 affected individuals in US
  - 1 in 2500 Caucasians
  - 1 in 15,000 African Americans
- 1 in 25 unaffected Caucasians are carriers

- Median age: 15.9 yrs
- Median predicted survival: 36.5 years

- Respiratory disease is the major cause of morbidity and mortality in CF
<table>
<thead>
<tr>
<th>Authors</th>
<th>No of Pat</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bhakoo et al. 1968</td>
<td>01</td>
<td>Histopathology</td>
</tr>
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<td>Goodchild 1974</td>
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<td>Maya PP et al 1980</td>
<td>03</td>
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<td>Jagdish JS. 1989</td>
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<td>Prasad ML et al 1990</td>
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<td>Bowlers 1993</td>
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<td>Spencer et al 1994</td>
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<td>Kabra et al 2000</td>
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<td>Singh et al 2002</td>
<td>18</td>
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<td>Ashavaid TF et al 2003</td>
<td>5</td>
<td>Sweat test and mutations</td>
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<td>Kabra et al 2003</td>
<td>120</td>
<td>Sweat &amp; mutations</td>
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<tr>
<td>Ashavaid et al 2005</td>
<td>39</td>
<td>Sweat &amp; mutations</td>
</tr>
</tbody>
</table>

Indian Pediatrics 2002; 39: 813-818.
CF at AIIMS: 1995-2010

- Total diagnosed till 2010: 355
- Died: 45
- Lost to follow up: 95
- Irregular follow up: 110
- Regular follow up: 100
- Oldest patient: 34 years
- Median age 2010: 10 years
- Age > 15 years: 20%

Slide and data from Dr Sushil K. Kabra, AIIMS, Delhi
CF at AIIMS: place of origin

CF at AIIMS: place of residence

Slide and data from Dr Sushil K. Kabra
AIIMS, Delhi

Indian Pediatr 2003; 40: 612-619
Magnitude of the problem in Indian population

- Estimated frequency of disease in immigrant Indians
  - in UK 1: 10000 – 12000
  - in USA 1: 40750 (compared with 1:3300 Caucasian)

- Carrier rates in India
  - 950 cord blood samples were tested for carrier status of D F 508 mutations.
  - 4 samples were positive for D F 508
  - Delta F 508 vary from 19-34% in CF patients

- The frequency of CF in India is about 1:40000

J Cyst Fibrosis 2006; 5: 43-46,
Estimates of CF in India

- Population of India: 100,000,000 (one billion)
- Birth rates: 20 per thousand per year
- Total births per annum: 20,000,000
- Number of children with CF per annum

<table>
<thead>
<tr>
<th>Frequency</th>
<th>Estimated CF cases per year</th>
</tr>
</thead>
<tbody>
<tr>
<td>1:2500</td>
<td>8000</td>
</tr>
<tr>
<td>1:10000</td>
<td>2000</td>
</tr>
<tr>
<td>1:40000</td>
<td>500</td>
</tr>
</tbody>
</table>

- Majority are likely not diagnosed, and do not receive proper treatment

Slide and data from Dr Sushil K. Kabra AIIMS, Delhi
CF patients followed at Apollo Hospital, Hyderabad

- Total: 8 children; 6 boys, 2 girls
- Ages: 6 mths, 3 yrs, 7 yrs, 8 yr, 10 yr, 10 yr, 11 yr, 16 yr

- Except the infant, all have bronchiectasis of the lungs due to repeated lung infections
- One has advanced lung disease on home oxygen
- All are pancreatic insufficient on enzymes
- 2 have normal height and weight
- Rest are underweight and have reduced height for age
CF: clinical course and complications

- Chronic cough, pulmonary exacerbations of CF
- Recurrent respiratory infections, pneumonia

- **Pancreatic insufficiency**
  - exocrine- (α cells) fat malabsorption
  - endocrine- (β cells) CF related diabetes (CFRD)

- **Liver disease**- biliary cirrhosis, hypoalbuminemia
CF: Pancreatic insufficiency

Malabsorption, steatorrhea

Malnutrition – edema

Enlarged liver

Rectal prolapse
Relationship between Nutrition and CF

FEV1% at age 6

Lung function

Konstan et. al. Pediatr Pulmonol 2000
Patients who have increase in WFA from Age 3 to 6 years have higher FEV1 at age 6 year

Konstan et al, Pediatr Pulmonol 2000
Nutritional Status and Survival

Dietary advice, enzyme supplementation and treatment of LRTI may improve the outcome of CF.

*Pediatric Pulmonology 1999; supplement 19: 337*
Basically …..

“The individual with CF must work diligently to consume adequate energy in order to meet increased needs caused by the increased work of breathing and altered digestive absorption”
CF nutrition: issues

- Chronic sinusitis - altered smell and taste
- Gastroesophageal reflux, heartburn, reduced appetite
- Chronic cough, phlegm
- Increased caloric requirement due to recurrent illnesses
- Higher REE, related to pulmonary disease

- At risk for CF related diabetes
- Fat malabsorption, need pancreatic enzyme replacement therapy, fat soluble vitamin supplementation
- CF related liver disease, low albumin

(Need to consider other routes of feeding? Nasogastric feeds, Gtube feeds)
So what does this mean....for the nutritionist

- Need to provide more than estimated caloric requirement (120-150% RDA)
- Need high fat diet (40% fat), fat soluble vitamins ADEK
- Need sufficient protein for growth

- Need to overcome energy deficit, promote growth and development
  
  - **Supplement salt, excessive losses in sweat.**
  - Provide iron, micronutrients, fatty acids
  - (Pancreatic enzymes with each meal and snack)
  - (Monitor sugars, screen for CF related diabetes)
Energy expenditure in CF

- Increased REE (resting energy expenditure) in CF
- Related to genotype *
- Related to decline in lung function/severity of lung disease**
- Related to gender


CF: Infants

- Highest growth rate and needs
- May be malnourished at diagnosis, catch up growth needs

Breastfeeding or formula?

- Breast milk lower in protein, but many benefits.
- Breastfed for > 6 months - beneficial *
  (anthropometric parameters, infections, hospitalizations: may protect decline of pulmonary function.
  BF should be promoted

CF infants: Elemental formula?

- May use elemental formulas for infants with gut resection, short gut syndrome
- Results from a randomized study failed to support the use of a hydrolyzed formula for the routine care of infants newly diagnosed with CF.

CF: Infants

- Human milk fortifiers for preterms
- Medium chain triglyceride oil added to feeds

- Enzymes given with soft solid even in neonatal period !!

- Will need salt supplement !!
- Wean at appropriate age
- Control gastroesophageal reflux (worse with chronic cough)
Nutrition considerations with age

**Toddlers**
- variable intake
  - “picky eaters”

**School age**
- school, activities interfere with meals
- need to assume more responsibility
- peer pressure

**Adolescents**
- More independent
- peer pressure
- body image
- pubertal requirements

**Adults**
- increased requirement due to declining lung function
  - CF related diabetes
- comorbid conditions
Recommendation for caloric intake in CF patients is 110 and 200% of the estimated average requirement (EAR) for age groups and gender, of which 35-40% energy should be from fat. It is questionable whether the advice is met.

1726 Completed 3-day dietary food records of 234 CF patients (111 girls) and 2860 completed two non-consecutive 24-hour dietary assessments of healthy controls (1411 girls) were studied.

The dietary intake in CF patients was compared with that of healthy controls by using independent sample t tests.
Caloric intake in CF children varied highly with age (88-127% EAR), which is in the lower range of the recommended 110-200% EAR.

CF patients had a significantly higher caloric intake than controls.

Most CF children had fat intake of 35 energy% or more - which was significantly higher than in controls.

Consumption of saturated fat, was well above 10% of the total energy intake.

Fat intake does generally meet recommendations, but this resulted in a considerable consumption of saturated fat; a reduction of the latter seems appropriate.
Oral calorie supplements for cystic fibrosis *

- There were no significant differences between persons receiving supplements or dietary advice alone for change in weight, height, body mass index, z score or other indices of nutrition or growth

Oral calorie supplements for cystic fibrosis *

• Oral calorie supplements do not confer any additional benefit in the nutritional management of moderately malnourished children with CF, over and above the use of dietary advice and monitoring alone.

• While nutritional supplements may be used, they should not be regarded as essential.

• Further randomized controlled trials are needed to establish the role of short-term oral protein energy supplements in people with CF and acute weight loss and also for the long-term nutritional management of adults with CF or advanced lung disease, or both.

Exocrine function of the pancreas

Exocrine
The pancreas produces enzymes that help digest our food

- Amylase
- Protease
- Lipase

ENZYMES

- Starch
- Protein
- Fat

NUTRIENTS
Exocrine Pancreatic Insufficiency (EPI)

**Signs and symptoms of EPI**

- Malabsorption leading to malnutrition
- Vitamin A, D, E, K and zinc deficiency

**Food intake**
PERT

- Oral pancreatic enzyme replacement therapy (porcine derived lipase, amylase, protease)

- Dosage based on fat content of food, and body weight
- **500-2500 units of lipase /kg body weight per meal**
  (1/2 this dose with snacks)

- Not to exceed 10,000 units/kg body weight lipase per day
- Ideally taken with meals
PERT (pancreatic enzyme replacement therapy)

-Produced from porcine pancreatic tissue
-Lipase, amylase, protease
-Enzymes- microspheres, enteric coated, coat dissolves at pH of 6
-Non-enteric coated powder (Viokase)
-CF patients have deficient bicarbonate secretion, increased acid production- therefore **antacids can** increase efficacy of enzymes
-CFF discourages generic enzymes because although same dose of lipase, different coating may not be biologically equivalent
Fat malabsorption in CF

- Despite advances in CF care intestinal fat malabsorption remains a persistent feature,
- Difficulty of achieving complete correction of fat malabsorption in clinical practice despite remarkable benefits resulting from exogenous pancreatic enzyme replacement therapy (PERT)

- Various defective mechanisms in CF, abnormal, intraluminal and intracellular factors.
- Relationship between essential fatty acid deficiency (EFAD) and intestinal fat transport

  *Mechanisms of lipid malabsorption in Cystic Fibrosis: the impact of essential fatty acids deficiency*

Fat malabsorption in CF patients on pancreatic enzymes

- Study investigating whether fat malabsorption in CF patients treated with PERT is caused by
  - insufficient lipolysis of triacylglycerololks or
  - by defective intestinal uptake of long-chain fatty acids.

- Results suggest that continuing fat malabsorption in CF patients receiving PERT is not likely due to insufficient lipolytic enzyme activity, but rather to
  - incomplete intraluminal solubilization of long-chain fatty acids
  - reduced mucosal uptake of long-chain fatty acids, or both.

Role of medium chain triglycerides in CF


- 15 CF children (aged 4 to 17 years) and persistent failure to thrive supplemented on an out-patient basis by a daily oral intake of 35 g of medium chain triglycerides (MCT) fat.

- Followup 6 months after initiation of the MCT diet - frequency of the bowel movements was reduced and abdominal discomfort disappeared but no weight gain was observed.

- No significant changes in either serum cholesterol or serum triglycerides were detected over the 6-month period.
- No clinical signs of an essential fatty acid deficiency
MCT

- MCT digestion starts in the stomach (by gastric lipase) in addition to pancreatic and intestinal lipases unlike LCT digested only by pancreatic lipase.

- Rapidly absorbed by enterocytes
  Directly reaches the liver via hepatic portal vein unlike LCT which undergo lymphatic circulation.

- (Additional benefit in lung disease: MCT – being fat source has low respiratory quotient compared to carbohydrates, poses least load on the respiratory system)
Effect of supplementing medium chain triglycerides with linoleic acid-rich monoglycerides on severely disturbed serum lipid fatty acid patterns in patients with cystic fibrosis.


7 CF patients with fatty acid composition indicating essential fatty acid deficiency, were given a mixture of medium chain triglycerides (MCTs) with linoleic acid-rich monoglycerides (LAMs) as food fat (about 1-1.25 g/kg body weight/day).

Treatment resulted in a significant increase of the previously reduced fraction of linoleic acid in all lipid classes. Thus, supplementing a MCT-containing diet with LAMs in cystic fibrosis patients results in a considerable amelioration of the previously disturbed fatty acid composition (FAC) of the major serum lipid classes.

Steinkamp G et al

**The effects of an oral energy supplement rich in linoleic acid in CF**

- In contrast to the control group (dietary counselling alone), the patients with supplemented diets achieved significant increases of
  - energy
  - weight for height
  - body fat

- as well as the initially low values of plasma phospholipid linoleic acid and its main metabolite arachidonic acid

- CF Patients with cystic fibrosis with low body weight and poor EFA status benefit from EFA-rich energy supplements and can synthesize arachidonic acid from the precursor linoleic acid.
Omega 3 and Omega 6 fatty acids

- Diet rich in fat (including animal fat) has omega-6 fatty acids which may adversely affect CF patients by encouraging inflammation in the lungs.

- In contrast, omega-3 fats appear to be of clinical benefit in CF. Reduction of sputum, improved lung function, a decrease in inflammation, and a decreased need for antibiotics have been observed in patients who have taken omega-3 fatty acid supplements.

Increasing intake of plant sources of omega-3 fats (e.g., flax seed and flax oil) and monounsaturated fats (e.g., olive oil) has been suggested as an approach to improving fatty acid nutrition in CF patients.
Fat soluble vitamins (ADEK) in CF

- Fat soluble vitamin deficiency recognized in CF
- PERT does not always correct this, hence oral supplementation needed

**Cochrane reviews on CF and supplementation of:**

- Vit A: no randomized trials (2012)
- Vit D: no evidence of benefit or harm in the limited number of small-sized published trials (2012)
- Vit K: Evidence from RCTs on benefits of routine vitamin K supplementation for people with CF is currently weak (2012)
- Vit E: (protocol alone, no review, 2012)

- Recommend – follow current guidelines / expert consensus
- Annual testing: serum vitamin levels, A, D, E, and Prothrombin time (PT) for K
PERT- update

- In 2010, the FDA required manufacturers of PERT to have approval for marketing, rescinding the distribution of PERT that had been available for decades without definitive studies of efficacy and safety.
- All preparations tests demonstrated superiority over placebo-controlled portions of the clinical trials.
- Side-effects no different compared with placebo.
- Additional PERTs are being evaluated including a nonporcine preparation which may be available in the future.
- Some patient variability to response continues, so clinicians need to continue to titrate dose and preparations based on weight gain and patient response.

New PERT-Lipoprotamase – nonporcine PERT

Liprotamase Enzymes

- **Lipase (32,500 U USP)**
  - Crystallized and crosslinked to increase low pH stability (Lipase-CLEC [crossed-linked enzyme crystal])

- **Protease (25,000 U USP)**
  - Crystallized to prevent proteolysis in the capsule over product shelf life

- **Amylase (3,750 U USP)**
  - Amorphous
New PERT- Lipoprotamase


Zinc deficiency in CF

- Poor absorption, and increased endogenous fecal zinc losses

- Zinc deficiency can affect Vitamin A absorption, transport, utilization

- Normal children - supplement 1mg/kg/day upto 10-15 mg/day

- *No specific dosing recommendations for CF*
### CF in Indian Children Micronutrient Deficiency

<table>
<thead>
<tr>
<th>Lab parameter</th>
<th>Total</th>
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</thead>
<tbody>
<tr>
<td>Serum zinc &lt;70 ppm</td>
<td>17(31.48%)</td>
</tr>
<tr>
<td>Serum calcium &lt;9 mg/dl</td>
<td>5 (9%)</td>
</tr>
</tbody>
</table>

Many Indian CF Children are deficient in Zinc

Need to study effect of zinc supplement on morbidity

Slide and data from Dr Sushil K. Kabra AIIMS, Delhi
Iron Profile of Indian CF Patients

<table>
<thead>
<tr>
<th>Lab parameter</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>MCV &lt;85</td>
<td>32 (59%)</td>
</tr>
<tr>
<td>Hb&lt;10</td>
<td>6 (11.11%)</td>
</tr>
<tr>
<td>Serum iron &lt;75</td>
<td>45 (83.33%)</td>
</tr>
<tr>
<td>% saturation &lt;20</td>
<td>42 (77.78%)</td>
</tr>
<tr>
<td>Serum iron mean median</td>
<td>47.65±15.34</td>
</tr>
<tr>
<td>Median</td>
<td>42</td>
</tr>
</tbody>
</table>

Majority of children with CF are deficient in iron and may need iron supplementation

Slide and data from Dr Sushil K. Kabra AIIMS, Delhi
CF related diabetes

- Increasing incidence with increasing age
- Usually associated with a decline in lung function
- Insulin deficiency - pancreatic endocrine insufficiency
  Different entity than Type I and Type II DM

- Ketoacidosis is rare
- Not associated with macrovascular disease as Type I DM
- Therefore fat content of diet should not be restricted, and may need to be as high as 40%
- Avoid excessive carbohydrates
Impaired glucose Tolerance Test in Children with CF

<table>
<thead>
<tr>
<th>Age group (Number of patients) (N 28)</th>
<th>Abnormal GT number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;10 yrs (9)</td>
<td>1 (11)</td>
</tr>
<tr>
<td>10-12 yrs (9)</td>
<td>2 (22)</td>
</tr>
<tr>
<td>&gt;12 yrs (10)</td>
<td>3 (30)</td>
</tr>
</tbody>
</table>

Slide and data from Dr Sushil K. Kabra AIIMS, Delhi
Complementary therapies in CF

- Vitamin A, vitamin C, vitamin E
- Zinc
- Omega 3 fatty acids, docosahexaenoic acid (DHA)
- Garlic
- Ginseng, and
- Curcumin.


*Complementary therapies in cystic fibrosis: nutritional supplements and herbal products.*

*Braga SF, Almgren MM.*
Cucurmin

- Turmeric is a spice derived from the rhizomes of *Curcuma longa*, a member of the ginger family.
- Curcuminoids are *polyphenolic compounds* that give turmeric its yellow color.
- Curcumin is the principal curcuminoid in turmeric.
Cucurmin

- Mechanisms implicated in inhibition of tumorigenesis: diverse and appear to involve a combination of antiinflammatory, antioxidant, immunomodulatory, proapoptotic, and antiangiogenic properties via pleiotropic effects on genes and cell-signaling pathways at multiple levels

- (The potentially adverse sequelae of curcumin's effects on proapoptotic genes, particularly p53, represent a cause for current debate)

Curcumin: preventive and therapeutic properties in laboratory studies and clinical trials. Strimpakos AS, Sharma RA.
Curcurmin and CF

- A study done jointly at Yale University and the Hospital for Sick Children in Toronto, showed that curcumin corrects the DeltaF508 CFTR physiological defect in mice.

- Clinical benefits also were demonstrated, with CF mice treated with curcumin having a better survival rate than those not treated.

- Until the safety and efficacy of curcumin in individuals with cystic fibrosis has been evaluated in clinical trials, the Cystic Fibrosis Foundation does not recommend the use of curcumin as a therapy for cystic fibrosis.
Nutrition therapy in CF

Overcome the energy deficit and promote normal growth and development for CF patients

- **Unrestricted diet**
  - High fat, high protein diet
  - Enteral feeds for moderate-severe malnutrition

- **Pancreatic enzyme supplements**
- **Fat soluble vitamins** - ADEK
- **Water soluble vitamins**, zinc, iron, essential fatty acids
- **Complementary therapies**
Nutrition in CF

• Medical management of CF is multifaceted
• **Role of nutrition in improving mortality has been established.**
• **Early identification of nutrition issues with appropriate intervention** plays a vital role in pulmonary health, and is key to longevity.
65 Roses® is a registered trademark of the Cystic Fibrosis Foundation.

“65 roses”

Did You Know?

Cystic fibrosis is sometimes called “65 roses.” The nickname came from a little boy who overheard his mom talking about the condition on the phone. He thought that each time his mom said “cystic fibrosis,” she was talking about 65 roses.
<table>
<thead>
<tr>
<th>Age Group</th>
<th>At Risk</th>
<th>Nutritional Failure</th>
</tr>
</thead>
<tbody>
<tr>
<td>0–2 years old</td>
<td>Weight for length 10%–25%</td>
<td>Height Percentile &lt;5%, or weight for length &lt;10% or IBW &lt;90%</td>
</tr>
<tr>
<td>2–20 years</td>
<td>BMI percentile 10th–25th</td>
<td>BMI percentile &lt;10% or IBW &lt;90%</td>
</tr>
<tr>
<td>Adults</td>
<td>BMI 19–20</td>
<td>BMI &lt;19 or percent IBW &lt;90%</td>
</tr>
</tbody>
</table>
### Table 4
**Dosing Pancreatic Enzymes (24)**

<table>
<thead>
<tr>
<th>Age Group</th>
<th>Dose</th>
<th>Adjusting Dose</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infants</td>
<td>2000–4000 Units lipase/120 ml formula or with each nursing OR 450–900 units lipase/gram of fat</td>
<td>Increase by 2000–2500 Units lipase per feed as volume increases or if symptoms of malabsorption return</td>
</tr>
<tr>
<td>Children &lt;4 years</td>
<td>1,000–2,000 Units lipase/kg/meal OR 500–4,000 Units lipase/gm fat</td>
<td>Snacks: 1/2 meal dose Compare U lipase per fat gram when weight dose appears above range.</td>
</tr>
<tr>
<td>Adults and children &gt;4</td>
<td>500–2000 Units lipase/kg/meal OR 500–4,000 Units lipase/gm fat</td>
<td>Snacks: 1/2 meal dose Compare U lipase per fat gram when weight dose appears above range.</td>
</tr>
</tbody>
</table>

**Note:**
- Doses of lipase greater than 2500 units/kg/meal (10,000 units/kg/d) are not recommended (23).
- >6,000 units lipase/kg/meal has been associated with colonic strictures in children <12 years (23, 24).
### Table 8
Vitamin Recommendations for Cystic Fibrosis (4,15)

<table>
<thead>
<tr>
<th>Age*</th>
<th>Vit A (IU)</th>
<th>Vit E (IU)</th>
<th>Vit D (IU)</th>
<th>Vit K (mcg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;8 years</td>
<td>5,000–10,000</td>
<td>200–400</td>
<td>400–8002</td>
<td>300–500</td>
</tr>
<tr>
<td>&gt;18 years</td>
<td>10,000</td>
<td>400–800</td>
<td>400–8002</td>
<td>300–500</td>
</tr>
<tr>
<td>To correct deficiencies</td>
<td>10,000–20,000</td>
<td>400–12,000</td>
<td>800–16002</td>
<td>5–10 mg/week or daily4</td>
</tr>
</tbody>
</table>

1. d-α-tocopherol
2. D2 or D3
3. Calcifediol (25-OHD)
4. Frequency of supplementation is dependent on response to therapy.

**Biochemical Testing:**
- Vitamin E—Serum α tocopherol
- Vitamin D—Serum 25-OHD
- Vitamin A—Serum retinol, retinol binding protein
  - Monitor serum retinyl esters – elevation indicates toxicity
- Vitamin K - PIVKA-II level or prothrombin time

*Refer to reference 15 for vitamin recommendations for children <8 years.
<table>
<thead>
<tr>
<th>Product</th>
<th>Vitamin A IU</th>
<th>Vitamin D IU</th>
<th>Vitamin E IU</th>
<th>Vitamin K (mcg)</th>
<th>Availability</th>
<th>Cost (CF Services Pharmacy)</th>
</tr>
</thead>
</table>
| CF Foundation Recommendations | 5,000–10,000 | 400–800      | 200–800      | 300–500         | Rx CF Services Pharmacy  
Local pharmacies               | $21.95/60     |
| ADEK Chewables              | 9,000        | 400          | 150          | 150             | Rx CF Services Pharmacy  
Local pharmacies               | $12.95/90     |
| Vitamax Chewables           | 5,000        | 400          | 200          | 150             | Rx CF Services Pharmacy  
Local pharmacies               | $18.95/90 gelcaps  
$29.95/90 chewables          |
| ABDEK Soft Gel Capsules     | 9,000        | 400          | 200          | 500             | Rx CF Services Pharmacy  
Local pharmacies               | $12.95/90     |
Table 14

Nutrition Management of Cystic Fibrosis Related Diabetes

1. Maintain high calorie intake with both simple and complex carbohydrates
2. Cover all simple carbohydrates with insulin
3. Maintain high fat diet (40%) for weight maintenance
4. Restrict sugary beverages unless they are counted as part of the carbohydrates and covered appropriately with insulin
Special situations

- Puberty
- Pregnancy
- Post surgical
- Post-transplant
Supplemental enteral tube feeding is widely used throughout the world to improve nutritional status in people with cystic fibrosis. The methods mostly used, nasogastric or gastrostomy feeding, are expensive and may have a negative effect on self-esteem and body image. Reported use of enteral tube feeding suggests that it results in nutritional and respiratory improvement; but, efficacy has not been fully assessed by randomised controlled trials. It is acknowledged, however, that performing a randomised controlled trial would be difficult due to the ethics of withholding an intervention in a group of patients whose nutritional status necessitates it.