**GETTING MORE CALORIES AND PROTEIN**

- Add grated cheese to soups, sauces, casseroles, vegetables, mashed potatoes, rice, noodles or meat loaf.
- Spread peanut butter on bread products or use it as a dip for raw vegetables and fruit. Add peanut butter to sauces or use on waffles.
- Serve cottage cheese with canned or fresh fruit.
- Use whole milk, half and half, cream, or enriched milk in cooking or beverages.
- Choose breaded meat, chicken, and fish instead of broiled or plain roasted.

**FOR MORE INFORMATION...**

[Image of the Cystic Fibrosis Foundation logo]

Provides current information, research, and treatment practices for cystic fibrosis.

Call: (800) FIGHT CF (344-4823)

www.cff.org

**NUTRITION FOR THE CYSTIC FIBROSIS PATIENT**

**Increase Calories**

**Increase Protein**

Tips to manage your diet and optimize your health...
**Cystic fibrosis** is a potentially life-threatening disease that causes thick, sticky mucus to build up in the lungs and digestive tract. A major organ affected is the pancreas.

Your pancreas is an organ that sits in your stomach behind your belly. The pancreas helps digest your food by releasing enzymes that break down the major nutrients in food including proteins, fats, and carbohydrates.

Thick mucus build-up in the pancreas from cystic fibrosis can lead to serious problems and pancreatic insufficiency. You may have the following problems:

- Stools that contain mucus, are foul smelling, or float
- Gas, bloating, or distended belly
- Problems getting enough protein, fat, and calories, in your diet

Because of these problems people with cystic fibrosis need to eat **high calorie and high protein foods** throughout the day. People with cystic fibrosis may have a **hard time maintaining weight**. Even if your weight is normal, you may not be getting enough nutrition. Children with cystic fibrosis may have difficulty growing and developing properly.

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**ENZYMES, VITAMINS, AND SALT**

- Most people with cystic fibrosis take pancreatic enzymes. These enzymes help your body digest nutrients.
- Taking your enzymes with all meals and snacks will help reduce bloating, gas, and foul smelling stools.
- Work with your doctor to increase or decrease enzymes based on your symptoms.
- Make sure you are taking a multivitamin that contains Vitamin A, D, E, & K.

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**EATING PATTERNS**

- Eat whenever you are hungry. This may mean eating several small meals throughout the day.
- Keep a variety of nutritious snacks around. Try to snack on something every hour.
- Make an effort to eat regularly, even if you only eat a few bites. Or try a nutritional supplement or milkshake.
- Be flexible. If you aren’t hungry at dinner time, make breakfast, mid-morning snacks, and lunch your main meals.
Cystic Fibrosis

- Cystic fibrosis is the most common inherited disease
- CF is an autosomal recessive gene
  - Defective gene must be inherited from both parents
- CF results from a gene mutation of the CFTR protein
- This mutation causes the exocrine glands of the digestive and respiratory tracts produce thick mucus
- This leads to lung infections and digestion problems
Unaffected "Carrier" Father

Unaffected "Carrier" Mother

Unaffected 1 in 4 chance

Unaffected 2 in 4 chance

Affected 1 in 4 chance
Lifespan

- The median age for a person with CF is 37 years
- CF patients most commonly die of respiratory failure
Prevalence

- 30,000 people have CF in the United States
- Incidence in ethnic groups:
  - Caucasians 1 in 3,000
  - African Americans 1 in 17,000
  - Asian Americans 1 in 31,000
- Around 1,000 cases of new CF are diagnosed each year in the US
- 1 in 31 Caucasian have the defective CF gene
Healthy CFTR Function

- A thin layer of secretions from exocrine glands helps keep the glands lubricated
  - Examples: sweat, tears, saliva, digestive juices, and mucus
- The CFTR protein is a chloride channel in epithelial membranes that produces these secretions
- CFTR regulates electrolyte and water movement across membranes
- In the lungs, the salty thin mucus kills pathogens we inhale
Etiology

- CF results from a mutation of the CFTR protein which is located on the long arm of chromosome 7.
- The CFTR mutation causes a defect in the chloride channel, which also disrupts sodium and water transport.
- This leads to not enough water outside of the cells.
- The result is dehydrated, thick, viscous secretions.
CFTR Mutations

- More than 1600 mutations of this gene have been found
- Most of these mutations are caused by a change in a single amino acid
- The mutations can cause changes in the production, structure, or stability of the chloride channel
Affected Organs

- Respiratory tract
- Sweat, salivary glands
- Intestine
- Pancreas
- Liver
- Reproductive tract
- Almost all exocrine glands
Pulmonary Complications

- Acute and chronic bronchitis
- Brochiectasis
- Pneumonia
- Atelectasis
- Peribrochial and parenchymal scarring
**Digestive Complications**

- Meconium ileus
- Pancreatic insufficiency
- Steatorrhea
- Hemolytic anemia
- Acute and chronic pancreatitis
- CF-related diabetes mellitus
- Hepatobiliary disease
- Biliary cirrhosis
- Liver disease
Co morbidities

- Diabetes mellitus/glucose intolerance
- Pulmonary infections
- Airway hyperreactivity
- Hypoxemia
- Pneumothorax
- Pulmonary hypertension
- Cor pulmonale
- Malnutrition, growth retardation, low BMI
- Intestinal obstruction

- Gastroesophageal reflux
- Hepatobiliary disease
- Coagulopathy
- Acute and chronic sinus infections
- Nasal polyps
- Urinary stress incontinence
- Delayed onset of puberty, azoospermia, and infertility
- Osteopenia and osteoarthritis
Prevention

- There is no way to prevent CF
- Genetic screening may detect 60-90% of carriers
Risk Factors

- Having Caucasian parents
- Both parents being carriers of the defective cystic fibrosis gene
Severity: Environmental

- Smoking
- Infectious exposures
- Poor nutrition
Severity: Medical Management

- Decreased self-management of disease
- Care at a Cystic Fibrosis Foundation approved center
- Timing of diagnosis

- Genetic modifiers also determine the severity of CF
Environmental Exposures
- Microorganisms
- Tobacco smoke
- Nutrition

Genetics
- CF mutations
- Modifier genes
- Ethnicity

Medical Treatment
- Adherence
- Access to care
- Clinical variations
Pathophysiology
Respiratory System

Function:
1. Gas exchange (major function)
2. Protective barrier against inhaled particles and microorganisms
3. Synthesize surfactant
4. Regulate body acid-base balance
5. Synthesize arachidonic acid
6. Convert angiotensin I to angiotensin II
Exocrine Pancreas

Function:
1. Secrete digestive enzymes into duodenum
2. Secrete bicarbonate to neutralize acidic chyme from stomach
Pathophysiology

- Obstruction of Exocrine Glands and Ducts:
  - secretion of abnormally thick, tenacious mucus by exocrine glands

- Infection and Inflammation

- Affected Organs:
  - lungs, pancreas, intestine, reproductive organs, sweat glands, salivary glands, liver
Cystic fibrotic lungs become permanently colonized with bacteria, leading to a vicious cycle of chronic infection.
Mucus Function

- Healthy epithelial cells:
  - Watery mucus that acts as a lubricant and protects the body’s tissues
  - Normal airway mucus consists of water, glycoproteins, and electrolytes

- CF affected epithelial cells:
  - Insufficient salt and water on outside of cells
  - Thick and sticky mucus that clogs passages and ducts
  - Mucus is thick and difficult to move

- Lungs:
  - CF lungs become chronically colonized with pathogens
  - Develop increased inflammatory responses:
    1. Reduced concentrations of the anti-inflammatory factor
    2. Increased breakdown of anti-inflammatory protein in the bronchoalveolar fluid

- Pancreas:
  - Abnormal pancreas secretions leading to maldigestion and malabsorption
  - Autodigestion of pancreas
The severity of cystic fibrosis symptoms are different from person to person. The most common symptoms are:

- very salty-tasting skin
- persistent coughing at times producing phlegm
- frequent lung infections; pneumonia or bronchitis
- wheezing or shortness of breath
- poor growth/weight gain in spite of a good appetite
- frequent greasy, bulky stools or difficulty in bowel movements
- small, fleshy growths in the nose called nasal polyps
Signs and Symptoms: Lungs

- **Pulmonary complications:**
  - Problems breathing
  - Acute and chronic bronchitis, bronchiectasis, atelectasis, hemoptysis, and peribronchial and parenchymal scarring
  - Pneumonia and infection by Staph aureus and Pseudomonas aeruginosa (80% ages 25-34)

- **Advanced Stages:**
  - Cor pulmonale (right ventricular enlargement)
Signs and Symptoms: Pancreas

- **Exocrine Pancreas:**
  - Plugs of thick mucus reduce the quantity of digestive enzymes released from the pancreas into the small intestine
  - Enzyme insufficiency causes maldigestion of food and malabsorption of nutrients
  - Decreased bicarbonate secretion can further reduce digestive enzyme activity
  - Bulky, foul smelling stools, cramping and intestinal obstruction, and rectal prolapse

- **Endocrine Pancreas:**
  - As the disease progresses damage to the endocrine portion causes impaired glucose tolerance leading to CF-related diabetes mellitus
Signs and Symptoms

- **GI Tract:**
  - Excess mucus in SI causes nutrient malabsorption

- **Liver:**
  - 1-2% of CF patients develop liver cirrhosis

- **Reproductive Organs:**
  - Female CF-patients show slightly reduced fertility
  - 97% of male CF-patients are infertile
Nutritional Implication

- Obstruction of glands and ducts cause complications in the affected organs which leads to a suppression of physical growth
Severity and Genotype

- **CF gene mutations:**
  - More than 1,600 different mutations of the CF gene
  - The severity of CF symptoms is partly based on the types of CF gene mutations a person with the disease has

- **Other gene mutations:**
  - A defect in the CFTR protein is only one contributing factor of the disease
  - Not all CF-patients have identical clinical symptoms, even when they carry the same CF gene mutations
  - Other genetic and/or environmental factors influence the effect of the CFTR defect on the disease phenotype
  - Predominantly located in genes affecting the host defense and inflammation systems
Lung Disease Severity

- **More severe lung disease:**
  - High levels of TNF
  - Low levels of glutathione S-transferase
  - Auto-antibodies to the bactericidal permeability increasing protein which increases the risk of infection

- **Earlier diagnosis/ severe signs and symptoms:**
  - Homozygous for CF related mutations

- **Later diagnosis/ mild signs and symptoms:**
  - Heterozygous for CF related mutations
Severity of Pancreas Malfunction

- **Severe Pancreas Malfunction:**
  - Pancreatic insufficient patients
  - CFTR channels with severe mutations do not support bicarbonate (HCO$_3^-$) transport
  - Some of these cases exhibit normal or even increased chloride conductance.

- **Mild Pancreas Malfunction:**
  - Pancreatic sufficient patients
  - CFTR channels are associated with reduced but still measurable bicarbonate (HCO$_3^-$) transport
Medical Diagnosis

- Three Main Considerations:
  1. Family history of CF
  2. Physical examination
  3. CF specific testing
     - Sweat test
     - Secretin stimulating test
     - Neo-natal CF screening

- Criteria for Diagnosis:
  - Presence of chronic lung disease, failure to thrive, and malabsorption or family history of CF
  - Positive sweat test
Medical Diagnosis

- **Sweat Test:**
  - Most reliable diagnostic tool of CF; gold standard
  - Measures the level of sodium and chloride in sweat

- **Genetic Test:**
  - Blood sample of cheek cells are sent to a lab
  - Test for most common CF mutations
  - Used if results from a sweat test are unclear
Sweat Electrolytes Test

- Why the test is performed:
  - Principal diagnostic test for CF that measures the level of chloride in sweat
  - Elevated levels are indicative of CF
  - Performed when CF symptoms are present, such as poor nutrition, repeated sinus or respiratory infections, foul-smelling stools, or infertility (in men)

- How the test is performed:
  - Colorless, odorless chemical that causes sweating is applied to a small area on an arm or leg
  - Electrode sends an electrical current to the area to stimulate sweating
  - Clean the stimulated area and collect the sweat
Sweat Test Results

- **For children who are less than six months old:**
  - Chloride levels at or above 60 mmol/L means the child has CF
  - Chloride levels between 30 and 59 mmol/L are considered borderline and need to be examined on a case-by-case basis
  - Chloride levels below 30 mmol/L are considered negative for CF

- **For people over the age of six months:**
  - Chloride levels at or above 60 mmol/L means the person has CF
  - Chloride levels between 40 and 59 mmol/L are borderline
  - Chloride levels below 40 mmol/L are considered negative for CF

- **Factors affecting lab values:**
  - Normal value ranges may vary slightly among different laboratories
  - Some conditions, such as dehydration or edema can affect the test results
Some CF-patients, however, clearly show a CF phenotype but exhibit normal or borderline sweat chloride concentrations. This results often in a delayed or missed diagnosis of cystic fibrosis. Alternative diagnostic tests needed therefore to be developed.
Secretin Stimulating Test

- Why the test is performed:
  - Measures the ability of the pancreas to release digestive enzymes in response to secretin
  - Secretin is produced by the small intestine when partially digested food has moved into the SI from the stomach

- How the test is performed:
  - Tube is inserted through the nose and into the stomach, then into the first part of the SI
  - Secretin is given intravenously
  - Over the next 1 - 2 hours, the contents of the material released from the pancreas into the duodenum are removed through the tube.
Neo-natal CF Screening

- Why the test is performed:
  - Most states conduct newborn screening for CF
  - Treatment can be initiated to prevent malnutrition and growth delay

- How the test is performed:
  - Sample of baby’s blood is taken and then examined for increased levels of immunoreactive trypsinogen (IRT), a protein produced by the pancreas that is linked to CF

- CF Negative:
  - If the test results are negative then the child does not have CF (unless they have symptoms of CF- further testing should be conducted)

- CF Positive:
  - Positive test results mean the child may have CF but further testing is needed for clear diagnosis
  - Not all children who test positive have CF
  - Test results do not indicate severity of CF
Treatment and Medications
Airway Clearance Techniques

• loosen thick, sticky lung mucus so it can be cleared
• Coughing & huffing
• Chest physical therapy (CPT) or Postural Drainage & Percussion (PD&P)
  • PD: sits or lies in various positions that drain mucus from lung parts
  • P: Chest is clapped/vibrated to dislodge mucus. Gravity will move mucus from small to large airways where it can be coughed up
  • Electrical palm precursors ad vibrators also used
• PD&P performed before meals or 1.5-2 hours after eating
ACT continued...

- Oscillator or Vest
  - Inflatable vest attached to machine that vibrates @ very high frequency
  - Loosens and thins mucus
  - Stop every 5 minutes to cough & huff

- Oscillating Positive Expiratory Pressure
  - Breath into a device that vibrates the large/small airways
  - Mucus thins and gets dislodged
  - Cough & huff
Inhaled Medication

- Antibiotics
- Mucolytics
- Bronchodilators

Why inhaled medication?
- Systemic toxicity can occur over long periods of time
- Reaches airway quicker and with more ease

CF patients use a nebulizer
Antibiotics

- Oral, aerosol, or IV
- Aerosol or inhaled preferred
- **TOBI- tobramycin**
  - Effective against Pseudomonas aeruginosa
  - Inhibits protein synthesis
  - 300 mg, 2X a day, intermittent cycle (28 days on 28 days off)
  - Dosage same for all patients regardless of age or weight
  - Taken before or after eating
  - 15-20 minutes per treatment
  - Side effects: bronchoconstriction, tinnitus
- Why TOBI?
  - Improves lung function
  - Decreases the # of days in a hospital
  - Reduces the need for IV antibiotics
Other antibiotics

- Colistimethate sodium
  - Damages cell membranes
  - Effective against gram-negative bacteria
  - Has been prescribed to CF patients for 20 years
  - Side effects: bronchoconstriction, inflammation
  - However, studies show that tobramycin is more effective
- Aztreonam lysine (Cayston)
  - Recently reformulated for inhalation delivery (had been prescribed parenterally for 25 years)
  - Improves lung function
  - Reduces bacteria in sputum
- Liposomal ciprofloxacin
- Levofloxacin
- Polymyxins
- Aminoglycosides
- Azithromycin (oral antibiotic)
Mucolytics

- **Pulmozyme (dornase alfa)**
  - Recombinant human DNA (rhDNase)
  - Thins mucus
  - Dosage differs for patient, 2.5 mg daily improved lung function in study
  - Side effects: voice alteration and rash

- **Hypertonic Saline**
  - Hydrates viscous mucus
  - Improve clearance and lung function

- **Studies have showed that dornase alfa is more effective, however large individual differences in response were found**

- **Denufosol tetrasodium**

- **Lancovutide**
Anti-inflammatory drugs

- Inhaled corticosteroids are used in CF to reduce endobronchial inflammation and reduce systemic effects
- Shown to impair childhood growth
- Multiple studies have shown no benefit or harm from corticosteroid use in CF patients
Oral corticosteroids can be very useful in acute exacerbations of CF lung disease b/c of powerful anti-inflammatory effects.

Macrolides have shown significant improvement in FEV1 and FVC.

Suppress pro-inflammatory cytokines and alter function of neutrophils.

Azithromycin.
Bronchodilators

- Used for inflamed, narrow, or hyperactive airways
- 80% of CF patients prescribed with inhaled bronchodilators
- Relaxes the muscles and dilates the airways of the lung
- Help with mucus clearance
- Given through a nebulizer
- Beta-agonists
- Albuterol (Ventolin HFA)
- Salbutamol
- Salmeterol
How does this fit together?
Order for treatments

1. Bronchodilator
2. Mucolytics (dornase alfa or hypertonic saline)
3. Airway clearance techniques- Chest therapy
4. Antibiotics (TOBI)
Pancreatic Enzyme Replacement Therapy (PERT)

- 90% CF patients experience pancreatic insufficiency
- Can lead to maldigestion and malabsorption
- Enteric-coated enzyme microspheres
- Typically a combination of lipase, amylase and protease
- Pancrelipase
  - 500 lipase units/kg of body weight/meal to start
  - Dosage increased depending on response and amount of fat in diet
- Side effects: diarrhea, constipation, upset stomach, vomiting
- Brand names: Creon, Lipram, Pancreaze, Ultrase, Zenpep
PERT cont’d

- Not every CF patient needs PERT
- Depends on amount of fat malabsorption
- Fecal Fat test to diagnose
- Normal value <7 grams in 24 hours
Lung Transplants

- Recommended when CF patients develop severe lung disease
- 900 lung transplants performed each year in the US for CF
- More people apply than can receive them
- 50% of CF patients who receive lung transplants are alive after 5 years
- Immunosuppressive drugs need to be taken daily for life
Is exercise good?

- Aids in mucus lung clearance
- Slows lung function decline
- Enhances feeling of well-being
- Improves respiratory muscle strength
- Higher exercise capacity is associated with improved survival
- However, need individualized exercise programs
- Take CF-specific risks into consideration:
  - Dehydration
  - Excessive salt losses
  - Bronchoconstriction
  - Hypoxemia
Alternative Therapies

- Some research shows health benefits by increasing intake of antioxidants
- Increase fruit and vegetable intake or supplements
- Non-cystic fibrosis lungs appear to have more natural antioxidants
- Inflammation and infection cycle in CF could be related to lack of natural antioxidant
Gene Therapy

- CFTR gene cloned in 1989
- Most trials so far have not achieved therapeutic correction of CF
- Vector molecules delivered to the airway are rapidly lost by mucociliary clearance
- Thickened mucus also enhances the barrier
- Few cells take up the vector and express the gene

However…
On January 31, 2012

- FDA approved Kalydeco for CF patients with the G551D mutation
- 4% of CF patient population have this mutation
- In this mutation, the defected protein moves to its proper place, but does not function properly
- Acts like a locked gate, preventing the flow of Na and fluid
- Kalydeco unlocks the gate and restores function
- Improves lung function, lowers sweat chloride levels, helps patients gain weight
- Kalydeco is also being studied in trial in combination with another oral medication, VX-809, in people with the Delta F508 mutation of CF
Postural drainage is a technique for loosening mucus in the airway so that it may be coughed out.

Tapping is performed in certain areas with the patient in different positions.
Role of CF on Nutrition

- Thick mucus is secreted, affecting many systems involved in digestion or breakdown of food. This includes:
  - Salivary glands
  - Intestine
  - Pancreas: 85-90% of people with CF have pancreatic insufficiency
  - Liver
Role of CF on Nutrition

- The amount of digestive enzymes released from the pancreas is dramatically reduced due to thick mucus plugs.
- Digestive enzyme activity is further reduced by the decreased bicarbonate secretion.
- Enzyme insufficiency causes maldigestion and malabsorption.
- Decreased bile resorption due to damage to the ducts of the liver leads to fat malabsorption.
Role of CF on Nutrition

- Excessive mucus lining the intestinal tract may cause problems with nutrient absorption. Associated complications include:
  - Bulky, foul-smelling stools
    - Irregular stools discourage people from eating a regular, healthy diet
  - Cramping and intestinal obstruction
    - This can lead to rectal prolapse
  - Liver involvement
    - Blockage of ducts, and therefore resorption
Role of CF on Nutrition

- Damage of the endocrine portion of the pancreas can cause impaired glucose tolerance and development of CF related diabetes mellitus
- Diabetes is considered one of the most common complications associated with CF
- Prevalence can be as high as 50% in adult patients
Effect on Appetite/ Eating Habits

- An individual with Cystic Fibrosis may experience a decreased desire to eat or an inability to keep food down due to:
  - Gastrointestinal discomfort
  - Impaired sense of smell and taste
  - Coughing and cough induced vomiting
  - Onset of Distal Intestinal Obstruction Syndrome (DIOS)
    - Also known as recurrent intestinal impaction
    - Abdominal pain and distension, bloating, nausea and anorexia, bowel obstruction
MNT

- High risk for malnutrition due to maldigestion, malabsorption, and disease complications
- Dyspnea, coughing, vomiting, GI discomfort, anorexia, impaired sense of smell and taste, and glucosuria interfere with nutrient retention
- Growth failure is often present before diagnosis in infants
- Growth is nearly normal for individuals with adequate energy and nutrient intakes
- Inadequate sodium levels can lead to lethargy, vomiting, and dehydration
MNT

- Goals:
  - Control maldigestion and malabsorption (enzyme therapy)
  - Provide adequate nutrients to promote optimal growth. Increase calories, increase protein
  - Provide adequate nutrients to maintain weight for height and optimal pulmonary function
  - Prevent nutritional deficiencies
  - Protein requirements are increased, but if energy needs are met the protein requirements will usually be met as well
  - Protein should make up 15%-20% total calories consumed, or at levels consistent with DRIs
  - Fat intake should provide 35%-40% of total calories
MNT

- Diet must be determined individually per patient
- Diet must be coordinated with any medications or other treatments
- Supplementation by feeding tube can be administered when necessary
- Decreasing activity levels is not recommended. Energy intake should be increased instead
- Due to increased work load of breathing, more energy is required.
- Individuals at high risk include: infants, children, adolescents, pregnant/ lactating women
Cystic Fibrosis

Etiology

Autosomal recessive inheritance

Cystic Fibrosis Gene
Cystic fibrosis transmembrane receptor (CFTR)

Pathophysiology

Obstruction of Glands and Ducts
Secretion of abnormally thick, tenacious mucus by exocrine glands

Affected Organs
- Respiratory organs
- Pancreas
- Reproductive organs
- Sweat glands
- Salivary glands
- Intestine
- Liver

Pathophysiology

Suppression of Physical Growth
Short stature and low body weight

Medical Management
- Genotyping
- Oral or IV antibiotics
- Aerosol antibiotics
- Inhaled medications
- Chest and physical therapy

Nutrition Management
- Nutrition status assessment
- Control malabsorption with pancreatic enzymes
- Meet increased energy requirements
- Vitamin and mineral supplementation

Algorithm content developed by John J.B. Anderson, PhD, and Sanford C. Garner, PhD. Updated by Donna H. Mueller, PhD, RD, FADA, LDN, 2006.
Multivitamin Use

- Water-soluble vitamins are usually adequately absorbed as long as enzyme supplements are taken.
- Fat-soluble vitamins are usually inadequately absorbed, even with enzyme supplementation.
Multivitamin Use

- Low vitamin E levels are associated with hemolytic anemia and abnormal neurologic findings.
- Decreased vitamin D levels may be related to decreased bone mineral content.
Multivitamin Use

Supplements of fat-soluble vitamins (A, D, E, and K), are recommended
Prognosis

- Survival rates have been improved due to scientific advancements and improvements in diagnostic and treatment procedures.
- Median age of CF patients is now 37 years.
- The number of people surviving to, or being diagnosed at 18 years of age or older is 42%.
- Women with CF have been able to deliver and breastfeed healthy infants.
- There is currently no cure, but with treatments, therapies, and good nutrition, quality and length of life is increased.
Prognosis

- For an individual on enzyme replacement medications, and correctly ensuring airway clearance one to four times a day, the quality of life can be rather good.
- Proper nutrition and strict adherence to medical council can increase the level of comfort in an individual.
Case Study
Case Study

- Lily Johnson
- Age: 14
- Sex: Female
- Ethnicity: Caucasian
- Chief complaint: Caught a cold that has gotten worse, treatments not working
- Patient hx: Diagnosed with CF at 6 months, has been hospitalized several times for respiratory infections
- Tx: High-frequency chest compression vest for 1 hour 2x per day
- Meds:
  - Pancrease: 1-3 caps after meals
  - Prevacid: 20 mg daily
  - Humabid: ½ tablet every 12 hours
  - Multivitamin
  - Proventil PRN
Nutritional Assessment

- Diagnosed with acute pneumonia
- Treatment plan:
  - Bed rest
  - Regular diet as tolerated
  - IV: D5 at 50 mL/hr
  - Vancomycin 10 mg/kg IV every 6 hours
Nutritional Assessment

- **Anthropometric**
  - Ht: 5’5”
  - Wt: 102 lbs
  - UBW: 110-115 lbs
  - BMI: 17
  - IBW: 82%
  - % Weight Change: 7.3%-11.3% (severe)
Nutritional Assessment

- Biochemical
  - Low: Transferrin, HCT
  - High: HbA1C, WBC
- Clinical
  - Thin, flushed
  - Pharynx reddened with postnasal drainage
  - Severe weight loss, underweight
  - Decreased breath sounds, percussion hyperresonant, rhonchi and rales present
Nutritional Assessment

- Dietary Assessment
  - Appetite has not been very good the last few days
  - Never drinks milk
  - Tries to avoid fried foods because they give her diarrhea
  - Inadequate intake due to decreased appetite
  - 24 hour recall
Nutritional Assessment

- Genetic Assessment:
  - Grandmother with type 2 diabetes
  - Great aunt with CF
- History:
  - Uneventful disease course
  - Hospitalized several times for respiratory infections
  - Receives routine medical care from local physician
Nutritional Assessment

- Subjective Global Assessment:
  - Weight loss: severe in the last 3 months
  - Physical findings: Thin, flushed, decreased breath sounds and rales
  - GI symptoms: bowel sounds present, nontender
  - Oral intake: inadequate
Nutritional Assessment

- PES statement
  - Inadequate oral intake related to under consumption of calories as evidenced by severe weight loss
- Intervention:
  - Increase caloric consumption
  - Education of healthy diet
  - Education on fat content in foods
- Monitoring and evaluation:
  - Assess weight gain
  - Assess understanding of education on fat content
  - Watch for diabetes
Nutritional Assessment

- **Estimated Calorie Needs While On Bed Rest:**
  - REE: $12.2 \times 46 \text{ kgs} + 746 = 1312 \text{ kcal}$
  - TEE: $1312 \times (1.3 + 0.3) = 2099 \text{ kcal}$
  - DER: $2099 \times (0.93/0.85) = 2288 \text{ kcal}$

- **Estimated Calorie Needs While Active:**
  - REE: $1312 \text{ kcal}$
  - TEE: $1312 \times (1.7 + 0.3) = 2624 \text{ kcal}$
  - DER: $2624 \times (0.98/0.85) = 2860 \text{ kcal}$
Nutritional Assessment

- Protein Requirements
  - (1g/kg) 46kgs = 46 grams of protein
- Actual Caloric Intake:
  - 1585 kcal/day
- IV D5
  - 1200 ml/day
Nutritional Assessment

- Bed rest:
  - 2288 kcal
  - D5@ 50ml/hr = 204 kcal
  - 2084 kcal from food
Sample Diet

- **Breakfast**
  - 2 pancakes
  - 1 Tbsp. butter
  - 2 Tbsp. syrup
  - ½ c. strawberries
  - 1 c. soy milk
- **Morning snack**
  - 1 c. yogurt
  - ½ c. granola
- **Lunch**
  - 3 ½ oz. turkey
  - 1 oz. cheese
  - 1 Tbsp. mayo
  - 3 tsp. mustard
- **Afternoon snack**
  - 1 oz. pretzels
  - 1 oz. cheese stick
- **Dinner**
  - 1 c. spaghetti
  - ½ c. spaghetti sauce with meat
  - 1 c. broccoli
  - 1 c. grape juice
Diet Rationale

- Provides the recommended amount of calories
- Soy milk can be tried to increase dairy consumption
- Extra snacks are given to increase calorie intake
References

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