Cystic Fibrosis Assessment

Cystic fibrosis (CF) is an autosomal recessive genetic disorder that affects the lungs, pancreas, gastrointestinal, and reproductive tracts and is considered a lethal genetic illness. In patients with CF, there is a mutation in the protein that regulates sodium and chloride channels in the body. As a result, secretions found in the lungs, pancreas, intestines, and other organs become abnormally thick and sticky. Buildup of mucus in the affected organs can eventually lead to organ failure and even death. Patients with CF experience recurrent pulmonary infections, pulmonary congestion, pancreatic insufficiency, delayed growth, and poor weight gain. Early diagnosis and advances in therapy have greatly improved patient outcomes.



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Thickened Mucus
Thick Mucus
Cystic fibrosis transmembrane regulator (CFTR), the protein responsible for regulating sodium and chloride channels in the body, is altered causing these channels to become blocked in patients with CF. Organs throughout the body continue to produce secretions; however, because the channels are blocked, the secretions are low in sodium chloride and water. This causes the patient’s secretions to be thicker and stickier than usual.

Recurrent Pulmonary Infections
Recurrent-clock at Lungs with Bacteria
Thickened mucus and decreased functioning of cilia allows mucus to stay in the lungs, creating an environment for infection. Infections are typically caused by organisms, such as Staphylococcus aureus and Pseudomonas, which are chronic in nature. Antibiotic resistance may develop in these patients.

Pulmonary Congestion
Lungs with Congested-traffic
Inflammation and mucus buildup in the bronchioles (bronchiolitis) initially occurs in patients with CF. Bronchiolitis, if chronic, will eventually lead to permanent widening of the airway, known as bronchiectasis.

Pancreatic Insufficiency
Pancreas Damaged
Mucus secreted in the pancreas is also abnormally thick, causing the exocrine ducts to become plugged. Insufficient production of pancreatic enzymes prevents adequate absorption of nutrients such as fat, protein, and fat-soluble vitamins. Patients with CF may also develop CF-related diabetes, due to destruction of insulin-producing islet cells in the pancreas. For this reason, it is important to closely monitor blood glucose levels to prevent dangerous complications related to hyperglycemia.

Steatorrhea
Steak-diarrhea
Malabsorption of fat will cause a patient’s stool to become bulky and foul-smelling. The stool will also float due to its increased lipid content.

Fat-Soluble Vitamin Deficiencies
Bacon Viking-ship Tattered
Inability to absorb fat-soluble vitamins, such as vitamin D, can lead to osteoporosis and osteopenia if left untreated.

Salty Taste to Skin
Salty Taste to Skin-suit
Patients with CF cannot absorb sodium chloride from their sweat as it passes through the sweat glands. As a result, these patients excrete four times the normal amount of sodium chloride in their sweat.

Meconium Ileus
Meconium-macaroni Eels
One of the first signs of CF in infants is meconium ileus. Meconium, an infant’s first stool, becomes thickened in the intestines and can become stuck. When this happens, the bowel becomes obstructed, and the meconium is unable to pass.

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Delayed Growth and Poor Weight Gain

Delayed-sign Growth-chart and Poor Weight-scale

Inadequate absorption of nutrients due to insufficient production of pancreatic enzymes can lead to delayed growth and poor weight gain in children.

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