CFTR is involved in the regulation of transepithelial ion transport and water–electrolyte homeostasis in many organ systems.Current guidelines recommend annual screening for cystic fibrosis–related diabetes with oral glucose–tolerance testing.24 Cystic fibrosis–related liver disease ranges from steatosis associated with increased alanine aminotransferase (ALT), aspartate aminotransferase (AST), and bilirubin levels to focal biliary fibrosis or to severe cholestasis and advanced liver cirrhosis with portal hypertension warranting liver transplantation.Noncirrhotic portal hypertension without synthetic liver dysfunction, which is common in persons with cystic fibrosis, may not warrant liver transplantation.In the sweat glands, normal CFTR activity results in chloride ion absorption from primarily isotonic perspiration; CFTR dysfunction in persons with cystic fibrosis causes impaired chloride absorption in the sweat–gland ducts and consequently elevated sweat chloride concentrations.1,2 The absence or dysfunction of CFTR in airway epithelium leads to decreased chloride and bicarbonate secretion at the apical membrane, the inability of alternative chloride channels such as TMEM16A (also called anoctamin 1 and ANO1) to compensate, and persistent sodium absorption through loss of CFTR–mediated inhibition of the epithelial sodium channel, which causes absorption of airway–surface fluid.9 The consequences of this fluid imbalance in the lungs are thickened secretions and reduced mucociliary transport, resulting in mucus retention and plugging of airways.Chronic lung disease with progressive decline in lung function and ultimately respiratory failure continues to be the major cause of death, but cystic fibrosis is a multiorgan disease In classic cystic fibrosis, thickened secretions cause pancreatic autodigestion and fatty replacement of the organ.

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