

Table 1		
Systems affected in cystic fibrosis		
System	Problem	Clinical consequence
Respiratory	Defective mucociliary clearance due to depleted airway surface liquid and thick sticky mucus	Recurrent/chronic infections Wet productive cough Bronchiectasis Persistent chest radiograph abnormalities Reduced lung function
Gut	Reduced luminal hydration with viscous mucus resulting in thickened stool and bowel blockage	Meconium ileus Distal intestinal obstruction syndrome Rectal prolapse
Pancreas	Pancreatic duct blockage from thick mucus prevents enzymes from entering the small intestine	Malabsorption Undernutrition Faltering growth Fat soluble vitamin deficiencies
	Pancreatic autodigestion	Acute and chronic pancreatitis
	Islet cell and beta cell dysfunction from inflammation and fibrosis	CF related diabetes, usually in teenagers and young adults
Liver	Excessive mucus build-up in bile ducts resulting in inflammation and fibrosis	CF related liver disease, may require liver transplantation
Sinus	Thick mucus causing persistent sinus congestion, infection and inflammation	Recurrent sinusitis Nasal polyps
Skin	Excessive salt loss in sweat, volume depletion and secondary hyperaldosteronism	Pseudo-Bartter syndrome (hyponatraemic, hypokalaemic, hypochloraemic metabolic alkalosis)
Reproductive	Congenital bilateral absence of vas deferens causing obstructive azoospermia	Male infertility
Other	Chronic illness	Muscle pains and arthralgia Osteoporosis
	Chronic cough causing pelvic floor weakness	Urinary incontinence