movement; the CFTR appears to function as a chloride channel. Children with CF demonstrate an increase in sodium and chloride in both saliva and sweat. This characteristic is the basis for the sweat chloride diagnostic test. The sweat electrolyte abnormality is present from birth, continues throughout life, and may be unrelated to the severity of the disease or the extent to which other organs are involved.

The primary factor, and the one that is responsible for many of the clinical manifestations of the disease, is mechanical obstruction caused by the increased viscosity of mucous gland secretions (Fig. 21-10). Instead of forming a thin, freely flowing secretion, the mucous glands produce a thick mucoprotein that accumulates and dilates them. Small passages in organs (such as the pancreas and bronchioles) become obstructed as secretions precipitate or coagulate to form concretions in glands and ducts. The earliest postnatal manifestation of CF is often **meconium ileus** in the newborn, in which the small intestine is blocked with thick, puttylike, tenacious, mucilaginous meconium.