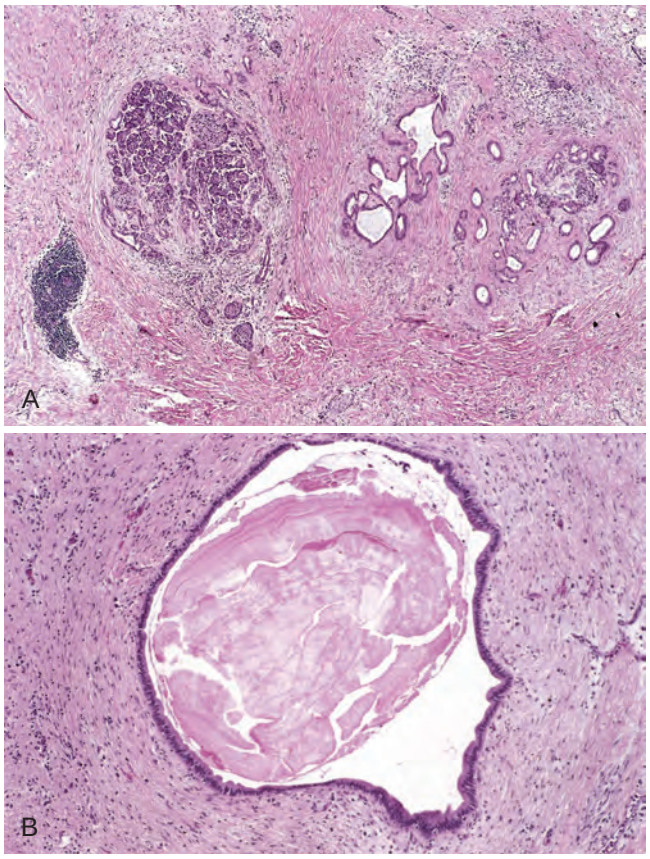

Figure 19-6 Chronic pancreatitis. **A,** Extensive fibrosis and atrophy has left only residual islets *(left)* and ducts *(right)*, with a sprinkling of chronic inflammatory cells and a few islands of acinar tissue. **B,** A higher power view demonstrating dilated ducts with inspissated eosinophilic ductal concretions in a person with alcoholic chronic pancreatitis.

Clinical Features. Chronic pancreatitis may present in many different ways. It may follow repeated bouts of acute pancreatitis. There may be repeated attacks of mild to moderately severe abdominal pain, or persistent abdominal and back pain. Attacks may be precipitated by alcohol abuse, overeating (which increases demand on the pancreas), or the use of opiates and other drugs that increase the tone of the sphincter of Oddi. In other patients the disease may be entirely silent until pancreatic insufficiency and diabetes mellitus develop due to destruction of the exocrine and endocrine pancreas.

The diagnosis of chronic pancreatitis requires a high degree of suspicion. During an attack of abdominal pain there may be mild fever and mild-to-moderate elevations of serum amylase. When the disease has been present for a long time, however, the dropout of acinar cells may be so great as to eliminate these diagnostic clues. Gallstone-induced obstruction may be evident as jaundice or elevations in serum levels of alkaline phosphatase. A very helpful finding is visualization of calcifications within the pancreas by computed tomography and ultrasonography. Weight loss and edema due to low albumin from malabsorption caused by pancreatic exocrine insufficiency also support the diagnosis.

Although chronic pancreatitis is usually not an immediately life-threatening condition, the long-term outlook for individuals with chronic pancreatitis is poor, with a 20- to 25-year mortality rate of 50%. Pancreatic exocrine insufficiency, chronic malabsorption, and diabetes mellitus can all lead to significant morbidity and contribute to mortality. In other patients **severe chronic pain** is a dominant problem. **Pancreatic pseudocysts** (described later) develop in about 10% of patients. As already mentioned, patients with hereditary pancreatitis have a 40% lifetime risk of developing pancreatic cancer; whether this increased cancer risk extends to other forms of chronic pancreatitis is unclear.

KEY CONCEPTS

- Chronic pancreatitis is characterized by *irreversible injury* of the pancreas leading to fibrosis, loss of pancreatic parenchyma, loss of exocrine and endocrine function, and high risk of developing pseudocysts
- Chronic pancreatitis is most often caused by
  - Repeated bouts of acute pancreatitis
  - Chronic alcohol abuse
  - Germline mutations in genes such as *CFTR* (the gene encoding the transporter that is defective in cystic fibrosis), particularly when combined with environmental stressors
  - Clinical features include intermittent or persistent abdominal pain, intestinal malabsorption, and diabetes

Nonneoplastic Cysts

A variety of cysts can arise in the pancreas. Most are nonneoplastic pseudocysts (discussed later), but congenital cysts and neoplastic cysts also occur.

Congenital Cysts

Congenital cysts are unilocular, thin-walled cysts that are believed to result from anomalous development of the pancreatic ducts. They range in size from microscopic lesions to 5 cm in diameter, and are lined by a glistening, uniform cuboidal epithelium or, if the intracystic pressure is high, by a flattened and attenuated cell layer. Congenital cysts are enclosed in a thin, fibrous capsule and are filled with a clear serous fluid. Congenital cysts may be sporadic, or part of inherited conditions such as *autosomal-dominant polycystic kidney disease* (Chapter 20) and *von Hippel-Lindau disease* (Chapter 28). Cysts in the kidney, liver, and pancreas frequently coexist in polycystic kidney disease. In von Hippel-Lindau disease vascular neoplasms are found in the retina and cerebellum or brain stem in association with congenital cysts (and also neoplasms) in the pancreas, liver, and kidney.

Pseudocysts

Pseudocysts are localized collections of necrotic and hemorrhagic material that are rich in pancreatic enzymes and lack an epithelial lining (hence the prefix "pseudo"). Pseudocysts account for approximately 75% of cysts in the pancreas. They usually arise following a bout of acute pancreatitis, particularly one superimposed on chronic alcoholic pancreatitis. Traumatic injury to the pancreas can also give rise to pseudocysts.