



The Pancreas

Ralph H. Hruban • Christine A. Iacobuzio-Donahue

CHAPTER CONTENTS

Congenital Anomalies 883
 Pancreatitis 884
 Acute Pancreatitis 884
 Chronic Pancreatitis 888
 Nonneoplastic Cysts 889
 Congenital Cysts 889
 Pseudocysts 889

Neoplasms 890
 Cystic Neoplasms 890
 Pancreatic Carcinoma 892
 Precursors to Pancreatic Cancer 892
 Pathogenesis 892

Acinar Cell Carcinoma 895
 Pancreatoblastoma 895

The adult pancreas is a transversely oriented retroperitoneal organ extending from the C-loop of the duodenum to the hilum of the spleen (Fig. 19-1A). Although the organ gets its name from the Greek *pankreas* (“all flesh”), it is in fact a complex lobulated organ with distinct exocrine and endocrine components.

The *exocrine pancreas* constitutes 80% to 85% of the organ and is composed of acinar cells that secrete enzymes needed for digestion. Acinar cells are pyramidally shaped epithelial cells containing membrane-bound granules rich in proenzymes (zymogens), including trypsinogen, chymotrypsinogen, procarboxypeptidase, proelastase, kallikreinogen, and prophospholipase A and B. Upon secretion, these proenzymes and enzymes are carried by a series of ductules and ducts to the duodenum, where they are activated by proteolytic cleavage in the gastrointestinal tract (described later).

The *endocrine pancreas* is composed of about 1 million clusters of cells, the islets of Langerhans, scattered throughout the gland. The islet cells secrete insulin, glucagon, and somatostatin and constitute only 1% to 2% of the organ. Diseases of the endocrine pancreas are described in detail in Chapter 24.

Congenital Anomalies

The complex process by which the dorsal and ventral pancreatic primordia fuse during pancreatic development is prone to “imperfections” that frequently gives rise to congenital variations in pancreatic anatomy. The pancreas normally arises from the fusion of dorsal and ventral outpouchings of the foregut. The body, the tail, and the superior/anterior aspect of the head of the pancreas, as well as the accessory duct of Santorini, are derived from

the dorsal primordium. Normally, the ventral primordium gives rise to the posterior/inferior part of the head of the pancreas and drains through the main pancreatic duct into the papilla of Vater.

Pancreas Divisum. *Pancreas divisum* is the most common congenital anomaly of the pancreas, with an incidence of 3% to 10%. In most individuals, the main pancreatic duct (the duct of Wirsung) joins the common bile duct just proximal to the papilla of Vater, and the accessory pancreatic duct (the duct of Santorini) drains into the duodenum through a separate minor papilla (Fig. 19-1A). *Pancreas divisum* is caused by a failure of fusion of the fetal duct systems of the dorsal and ventral pancreatic primordia. As a result, the bulk of the pancreas (formed by the dorsal pancreatic primordium) drains into the duodenum through the small-caliber minor papilla (Fig. 19-1B). The duct of Wirsung in persons with *divisum* drains only a small portion of the head of the gland through the papilla of Vater. Although controversial, it has been suggested that inadequate drainage of the pancreatic secretions through the minor papilla, especially when combined with genetic defects that also increases susceptibility to pancreatitis (described later), predisposes individuals with *pancreas divisum* to chronic pancreatitis.

Annular Pancreas. Annular pancreas is a band-like ring of normal pancreatic tissue that completely encircles the second portion of the duodenum. Annular pancreas can produce duodenal obstruction.

Ectopic Pancreas. Pancreatic tissue that is aberrantly situated, or ectopic, is found in about 2% of careful routine postmortem examinations. The favored sites for ectopia are the stomach and duodenum, followed by the jejunum,