present in the pylorus, inflammation and scarring may lead to obstruction. Because the rests may be present within any layer of the gastric wall, they can mimic invasive cancer. *Gastric heterotopia,* small patches of ectopic gastric mucosa in the small bowel or colon, may present with occult blood loss due to peptic ulceration of adjacent mucosa.

Meckel Diverticulum

A true diverticulum is a blind outpouching of the alimentary tract that communicates with the lumen and includes all three layers of the bowel wall. *The most common true diverticulum is the Meckel diverticulum, which occurs in the ileum.*

Meckel diverticulum occurs as a result of failed involution of the vitelline duct, which connects the lumen of the developing gut to the yolk sac. This solitary diverticulum extends from the antimesenteric side of the bowel (Fig. 17-2). The "rule of 2s" is often used to help remember characteristics of Meckel diverticula, which

- Occur in approximately 2% of the population
- Are generally present within 2 feet (60 cm) of the ileocecal valve
- Are approximately 2 inches (5 cm) long
- Are twice as common in males
- Are most often symptomatic by age 2 (only approximately 4% are ever symptomatic).

The mucosal lining of Meckel diverticula may resemble that of normal small intestine, but ectopic pancreatic or gastric tissue may also be present. The latter may secrete acid, cause peptic ulceration of adjacent small intestinal mucosa, and present with occult bleeding or abdominal pain resembling acute appendicitis or obstruction.

Less commonly, congenital diverticula occur in other parts of the small intestine and ascending colon. Virtually all other diverticula are acquired and either lack muscularis entirely or have an attenuated muscularis propria. The most common site of acquired diverticula is the sigmoid colon (discussed later).

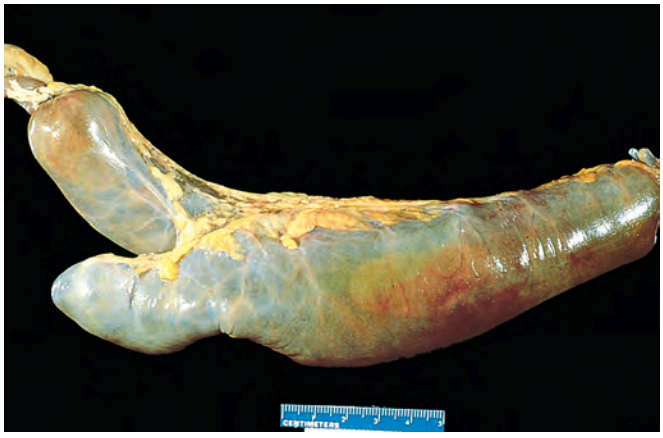

Figure 17-2 Meckel diverticulum. The blind pouch is located on the antimesenteric side of the small bowel.

Pyloric Stenosis

Congenital hypertrophic pyloric stenosis is three to five times more common in males and occurs once in 300 to 900 live births. Monozygotic twins have a high rate of concordance, with a 200-fold increased risk if one twin is affected. Incidence of congenital hypertrophic pyloric stenosis is also increased in dizygotic twins and siblings of affected individuals, although here the risk is only increased by 20-fold, reflecting a complex multifactorial pattern of inheritance. Consistent with a genetic basis, Turner syndrome and trisomy 18 also confer an increased risk of congenital hypertrophic pyloric stenosis. While the underlying mechanisms are not understood, erythromycin or azithromycin exposure, either orally or via mother's milk, in the first 2 weeks of life has been linked to increased disease incidence.

Congenital hypertrophic pyloric stenosis generally presents between the third and sixth weeks of life as new-onset regurgitation, projectile, nonbilious vomiting after feeding, and frequent demands for re-feeding. Physical examination reveals a firm, ovoid, 1 to 2 cm abdominal mass. In some cases abnormal left to right hyperperistalsis is evident during feeding and immediately before vomiting. This constellation of findings stems from hyperplasia of the pyloric muscularis propria, which obstructs the gastric outflow tract. Edema and inflammatory changes in the mucosa and submucosa may aggravate the narrowing. Surgical splitting of the muscularis (myotomy) is curative. Acquired pyloric stenosis occurs in adults as a consequence of antral gastritis or peptic ulcers close to the pylorus. Carcinomas of the distal stomach and pancreas may also narrow the pyloric channel due to fibrosis or malignant infiltration.

Hirschsprung Disease

Hirschsprung disease occurs in approximately 1 of 5000 live births. It may be isolated or occur in combination with other developmental abnormalities; 10% of all cases occur in children with Down syndrome and serious neurologic abnormalities are present in another 5%.

Pathogenesis. The enteric neuronal plexus develops from neural crest cells that migrate into the bowel wall during embryogenesis. Hirschsprung disease, also known as congenital aganglionic megacolon, results when the normal migration of neural crest cells from cecum to rectum is arrested prematurely or when the ganglion cells undergo premature death. This produces a distal intestinal segment that lacks both the Meissner submucosal and the Auerbach myenteric plexus ("aganglionosis"). Coordinated peristaltic contractions are absent and functional obstruction occurs, resulting in dilation proximal to the affected segment.

The mechanisms underlying defective neural crest cell migration in Hirschsprung disease are unknown, but a genetic component is present in nearly all cases, and 4% of patients' siblings are affected. Heterozygous loss-of-function mutations in the receptor tyrosine kinase *RET* account for the majority of familial cases and