

Figure 17-43 Juvenile polyposis. **A**, Juvenile polyp. Note the surface erosion and cystically dilated crypts. **B**, Inspissated mucous, neutrophils, and inflammatory debris can accumulate within dilated crypts.

MORPHOLOGY

The polyps of Peutz-Jeghers syndrome are most common in the small intestine, although they may occur in the stomach and colon, and, with much lower frequency, in the bladder and lungs. Grossly, the polyps are large and pedunculated with a lobulated contour. Histologic examination demonstrates a characteristic arborizing network of connective tissue, smooth muscle, lamina propria, and glands lined by normal-appearing intestinal epithelium (Fig. 17-44). The arborization and presence of smooth muscle intermixed with lamina propria are helpful in distinguishing polyps of Peutz-Jeghers syndrome from juvenile polyps.

Clinical Features. Because the morphology of Peutz-Jeghers polyps can overlap with that of sporadic hamartomatous polyps, the presence of multiple polyps in the small intestine, mucocutaneous hyperpigmentation, and a positive family history are critical to the diagnosis. Detection of *STK11* mutations can be helpful diagnostically in patients with polyps who lack mucocutaneous hyperpigmentation. However, the absence of *STK11* mutations does not exclude the diagnosis, since mutations in other presently unknown genes can also cause the syndrome.

Neoplastic Polyps

Any neoplastic mass lesion in the GI tract may produce a mucosal protrusion, or polyp. This includes adenocarcinomas, neuroendocrine (carcinoid) tumors, stromal tumors, lymphomas, and even metastatic cancers from distant sites. *The most common neoplastic polyps are colonic adenomas, which are precursors to the majority of colorectal adenocarcinomas.*

Adenomas are intraepithelial neoplasms that range from small, often pedunculated, polyps to large sessile lesions. There is a small male predominance, and they are present in approximately 30% of adults living in the Western world by age 60. Because these polyps are precursors to colorectal adenocarcinoma, it is recommended that all adults in the United States undergo surveillance by age 50. Patients at increased risk, including those with a family history of colorectal adenocarcinoma, are typically screened colonoscopically at least 10 years before the youngest age at which a relative was diagnosed. The preferred approach to surveillance varies, but colonoscopy is most common.

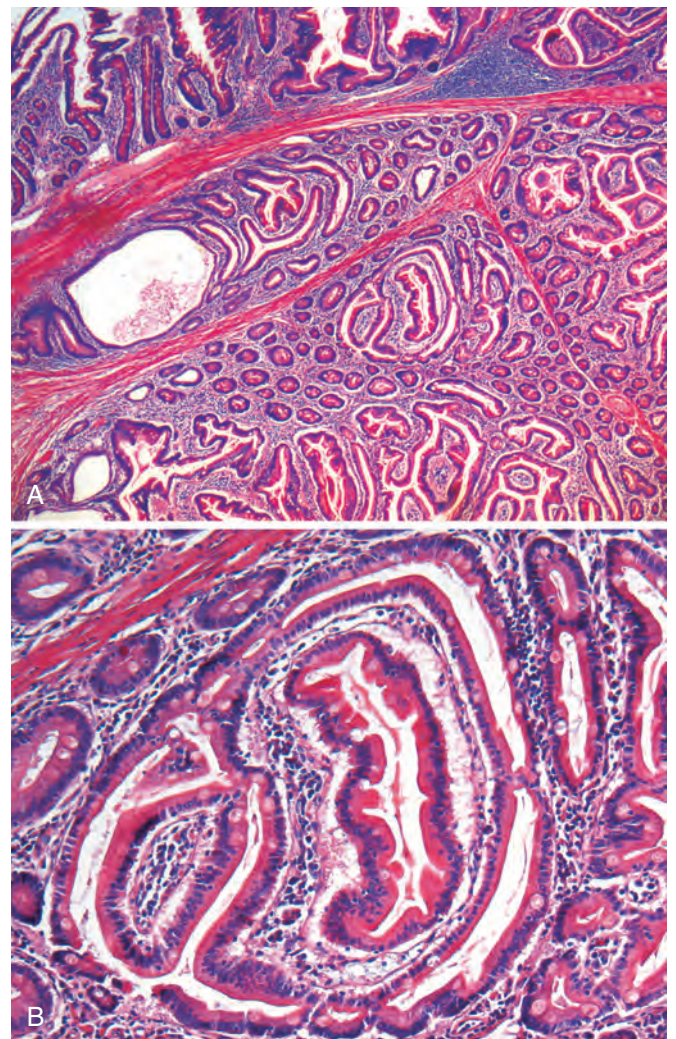


Figure 17-44 Peutz-Jeghers polyp. **A**, Polyp surface (top) overlies stroma composed of smooth muscle bundles cutting through the lamina propria. **B**, Complex glandular architecture and the presence of smooth muscle are features that distinguish Peutz-Jeghers polyps from juvenile polyps. Compare to Figure 17-42.