Appendiceal cancer is very rare with approximately 1-2 cases per 1 million individuals. Appendiceal cancers can occur at any age, with the peak occurrence in the 6th decade (average age of 50 at diagnosis). Most studies report that men and women are affected in equal numbers, while a few have suggested a slightly increased frequency in women. Peritoneal carcinomatosis is the spread and growth of cancer cells in the abdominal cavity. Cancers that are most frequently associated with peritoneal carcinomatosis include the gastrointestinal (colon, rectal, appendiceal, gastric, pancreas, small bowel and gallbladder) and the gynecologic (ovarian, primary peritoneal, and uterine) cancers. Other cancers that can spread to the abdominal cavity include breast, esophagus, and melanoma. Gastrointestinal stromal tumors (GISTs) are a subtype of sarcoma of the intestine that can also present with multiple cancer tumors in the abdomen. Peritoneal mesothelioma is a cancer that originates in the lining of the abdominal cavity (peritoneum) and presents with signs and symptoms of peritoneal carcinomatosis. Because there are no unique features of appendiceal cancer on imaging studies such as ultrasound, CT scan, PET scan or MRI, the actual diagnosis of appendiceal cancer cannot be made until a tumor specimen is examined by a pathologist. This is frequently accomplished at the time of appendectomy for appendicitis, surgery for an intestinal blockage or presumed ovarian cancer, or through a diagnostic tumor biopsy performed for an abnormal clinical or radiographic finding such as a palpable tumor or tumors seen on an imaging study. The finding of a dilated, mucin filled appendix on CT scan or MRI should prompt concern for an appendiceal tumor and an appendectomy should be considered. The different types of appendiceal tumors and cancers can be distinguished by the appearance of the cells under the microscope and by staining them for specific markers. Goblet cell carcinoid tumors tend to be easier to identify because of the unique combination of neuroendocrine and epithelial cells.