

APPENDIX CANCER: Little organ. Big problems.

➤ What is appendix cancer?

Appendix cancer originates from the cells lining the lumen of the appendix. It is often not discovered until it has spread outside of the appendix, often throughout the pelvis and abdomen. Although it is a rare cancer, its incidence has significantly increased in recent years, though specific causes have not been identified.

The Appendix Cancer PMP Research Foundation (ACPMP) is focused on epithelial appendiceal cancers, which include low-grade appendiceal mucinous neoplasm (LAMN), high-grade appendiceal mucinous neoplasm (HAMN), adenocarcinoma, goblet cell adenocarcinoma, and signet ring cell. The different types of appendix cancer exhibit different behaviors; some are slowly progressive while others can be highly aggressive. All are potentially life-threatening and require the attention of a physician with significant expertise in treating appendix cancer.

When appendix cancer spreads to the peritoneum, it often produces a jelly-like substance called mucin. Pseudomyxoma peritonei (PMP) is the progressive accumulation of mucin and mucus-secreting tumor cells within the abdomen and pelvis. Most cases of PMP arise from an appendiceal tumor that has ruptured the appendix and spread to surrounding peritoneal surfaces, although rare cases arise from tumors located in other organs. Rupture of the appendix can produce symptoms of acute appendicitis, but sometimes, appendiceal mucinous tumors leak mucin and tumor cells for weeks or months before being discovered. As mucin and mucinous tumor cells accumulate, the abdomen swells, and the patient can experience an array of generalized symptoms.

➤ What to do if you find it?

As this is a rare cancer that requires specific treatment, referral to a surgical oncologist with expertise in treating appendix cancer and peritoneal malignancy is critical, as delayed or wrong treatment can be detrimental to patient outcomes.

A surgical oncologist with expertise in treating appendix cancer will do additional imaging and testing to determine whether the patient is a good candidate for a procedure called CRS/HIPEC. This is a complex surgical procedure that consists of cytoreductive surgery with or without heated intraperitoneal chemotherapy, depending on the completeness of the cytoreductive surgery. Many patients who undergo CRS/HIPEC with a specialist have very good long-term outcomes.

**From Laura Lambert, MD, FACS,
Surgical Oncologist at Huntsman
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Chair of the ACPMP Medical Advisory
Board**

- Do least amount of surgery necessary to:**
1. Get a diagnosis
 2. Fix the problem
 3. Assess extent of disease – particularly on small bowel and mesentery
 4. Describe detailed findings in op note
 5. Take pictures
 6. Refer to peritoneal malignancy program
 7. Offer hope



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