

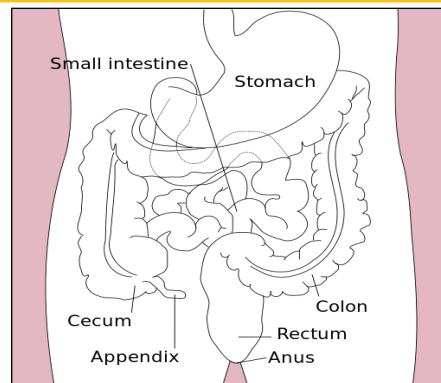


Appendiceal Mucinous Neoplasms



Appendiceal mucinous neoplasms have been classified as either low-grade or high-grade based on their cytologic features. Low-grade and high-grade neoplasms are a histologic subtype of appendiceal mucinous neoplasms. The behavior codes for these mucinous neoplasms are determined by the microscopic analysis of cytologic features conducted by a pathologist. Appendiceal mucinous neoplasms with only pushing borders are LAMN or HAMN. Low-grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy accounting for 1% of gastrointestinal neoplasms and is found in less than 0.3% of appendectomy specimens.

https://commons.wikimedia.org/wiki/File:Stomach_colon_rectum_diagram-en.svg



Gross examination of LAMN may be unremarkable or may appear as a mucin-filled, cystically dilated tissue. The appendix wall may appear thin, fibrotic, hyalinized, or calcified with a smooth or granular appearance. The diagnosis can be quite challenging, particularly in female patients where primary appendiceal and ovarian mucinous neoplasms share common atypical clinical and imaging findings.

[https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6440557/#:~:text=Low%2Dgrade%20appendiceal%20mucinous%20neoplasm%20\(LAMN\)%20is%20a%20rare,to%20pseudomyxoma%20peritonei%20\(PMP\)](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6440557/#:~:text=Low%2Dgrade%20appendiceal%20mucinous%20neoplasm%20(LAMN)%20is%20a%20rare,to%20pseudomyxoma%20peritonei%20(PMP))

https://documents.cap.org/protocols/Appendix_5.0.0.0.REL_CAPCP.pdf

Solid Tumor Rules - Colon

Rule H5 Code **low grade appendiceal mucinous neoplasm (LAMN) and high grade appendiceal mucinous neoplasm (HAMN) 8480** when:

- Diagnosis date is 1/1/2022 forward **AND**
- Behavior is stated to be in situ/non-invasive **OR**
- Behavior is not indicated

Note 1: ICD-O-3.2 lists LAMN as behavior of /1. The standard setters have agreed LAMN should be collected and assigned a behavior code of /2 beginning with cases diagnosed 1/1/2022 forward.

Note 2: A diagnosis of LAMN or HAMN does not require the tumor to be greater than 50% mucinous in order to be coded 8480.

Note 3: If the pathologist indicates LAMN or HAMN is invasive or has a malignant behavior, continue through the rules

ICD-O-3.2 LAMN/HAMN Coding Updates - Effective 1/1/2022

8480/2	Low grade appendiceal mucinous neoplasm (LAMN) (C181)	Beginning with cases diagnosed 1/1/2022 forward, LAMN should be assigned code of /2. LAMN diagnosed prior to 1/1/2022 is not reportable.
8480/3	High grade appendiceal mucinous neoplasm (HAMN) (C181)	New behavior/term

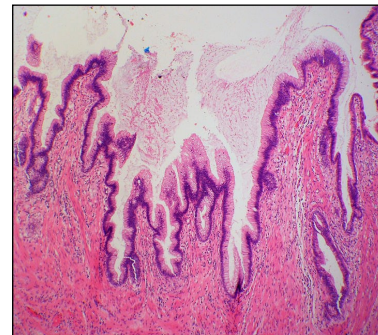
https://seer.cancer.gov/tools/solidtumor/2023/Colon_STM.pdf



Low Grade Appendiceal Mucinous Neoplasm (LAMN)

Low grade appendiceal mucinous neoplasms (LAMN) have a unique characteristic. The LAMN lacks an infiltrative or destructive invasion feature. Instead the LAMN can extend into and sometimes through the appendiceal wall by a pushing front. Spread in this neoplasm does not indicate malignancy. The histology code and behavior code of a LAMN is 8480/2 unless the pathologist indicates a presence of invasion.

The most worrisome complication of this particular neoplasm is seeding of mucin into the adjacent peritoneum leading to pseudomyxoma peritonei (PMP). Seeding into the peritoneum occurs in the late stages of the disease.



<https://www.sciencedirect.com/science/article/pii/S0046817722001770#bib6>

[https://commons.wikimedia.org/wiki/File:Appendix_LowGradeMucinousNeoplasm_MucinousNeoplasm_LowGrade_HP_\(2\)_PA.JPG](https://commons.wikimedia.org/wiki/File:Appendix_LowGradeMucinousNeoplasm_MucinousNeoplasm_LowGrade_HP_(2)_PA.JPG)

CAP Protocol (June 2022)

pTis LAMN is applicable only to LAMN. Low-grade appendiceal mucinous neoplasm confined to the muscularis propria; Acellular mucin or mucinous epithelium may invade into the muscularis propria. (T1 and T2 are not applicable to LAMN; Acellular mucin or mucinous epithelium that extends into the subserosa or serosa should be classified as T3 or T4a, respectively.)

If the histologic features of the appendiceal primary qualify for LAMN, the histologic type in the tumor synoptic should be selected as LAMN even if there is peritoneal involvement

High Grade Appendiceal Mucinous Neoplasm (HAMN)

The high grade appendiceal mucinous neoplasm (HAMN) or mucinous adenocarcinoma without invasion also contain the same unique pushing characteristic. On rare occasion the HAMN can resemble an LAMN that show areas of high-grade cytologic features. This would support an HAMN with the behavior code of 8480/2 unless the pathologist indicates the presence of invasion. Therefore a HAMN can be coded with a behavior of /2 or /3 depending on the evaluation by the pathologist.

https://documents.cap.org/protocols/Appendix_5.0.0.0.REL_CAPCP.pdf

CAP Protocol (June 2022)

High-grade appendiceal mucinous neoplasms (HAMN) are staged similar to mucinous adenocarcinoma.

Pseudomyxoma Peritonei (PMP)

Pseudomyxoma peritonei (PMP) is a clinical condition caused by cancerous cells that produce abundant mucin or gelatinous ascites. This disease is most commonly caused by an appendiceal primary cancer.

The term PMP is not a histologically based term, PMP symbolizes a clinical syndrome and is not factored in the staging of a LAMN.

Pseudomyxoma peritonei now has a **two-tiered system** that classifies pseudomyxoma peritonei as either **high-grade** or **low-grade**. Pseudomyxoma peritonei is usually associated with **mucinous** tumors of the appendix and is rarely associated with ovarian mucinous tumors.

- High-grade pseudomyxoma peritonei is malignant /3
- Low-grade pseudomyxoma peritonei is not malignant /1

https://seer.cancer.gov/tools/solidtumor/2023/Colon_STM.pdf

https://en.wikipedia.org/wiki/Pseudomyxoma_peritonei