

## Mucinous tumours of the appendix and pseudomyxoma peritonei (PMP)

Pseudomyxoma peritonei (PMP) is clinically defined as the slow and relentless accumulation of intraperitoneal mucin from peritoneal implants of a mucinous tumour. This is considered a malignant condition. Tumours of the appendix are different from tumours in the rest of the large bowel. They more frequently demonstrate mucinous differentiation with local spread in the peritoneal cavity. The vast majority of PMP originate from a perforated mucinous tumour of the appendix.

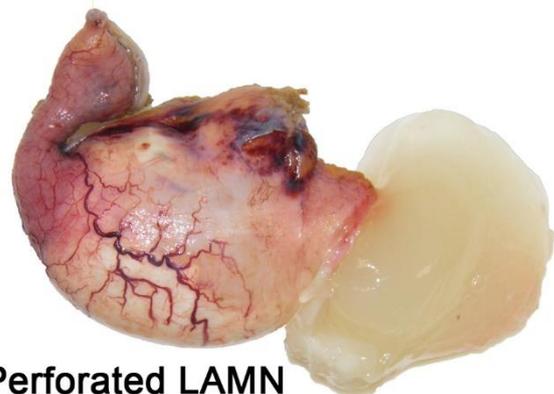
### Classification and nomenclature of mucinous tumours of the appendix

Current terminology	Histological features	Risk of PMP
Low-grade appendiceal mucinous neoplasm (LAMN)	Flat or undulating mucinous epithelium with low grade cytological atypia; loss of muscularis mucosae and submucosal fibrosis; pushing invasion with no desmoplasia	<ul style="list-style-type: none"> <li>• No perforation – very low</li> <li>• Perforation with acellular mucin deposits – low (5%)</li> <li>• Perforation with cellular mucin deposits – high (33%)</li> </ul>
High-grade appendiceal mucinous neoplasm	Same architecture as LAMN with high grade cytological atypia	Unknown, probably at least similar to LAMN
Mucinous adenocarcinoma	Infiltrative invasion with desmoplasia in the wall and high grade cytological atypia	Very high
Poorly differentiated adenocarcinoma with signet ring cells	Mucinous carcinoma with <50% of signet ring cells	Very high
Signet ring cell carcinoma	> 50% signet ring cells	Very high

Note: Some mucinous and signet ring cell carcinomas may arise from a goblet cell carcinoid tumour.



**LAMN - No perforation**

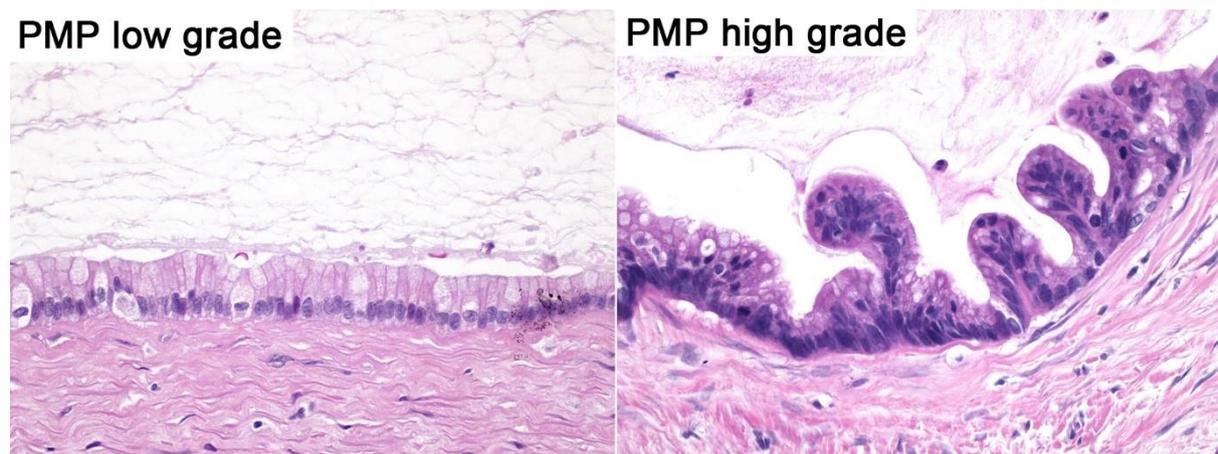


**Perforated LAMN**

## Classification and nomenclature (grading) of PMP

Current terminology	Distinctive features	Other nomenclature	5-year survival
Acellular mucin	No epithelial cells identified	<ul style="list-style-type: none"> <li>• Low-grade mucinous neoplasm/adenocarcinoma</li> <li>• AJCC grade G1</li> <li>• Disseminated peritoneal adenomucinosis</li> </ul>	63-91%
Low-grade mucinous carcinoma peritonei	Mucinous epithelium with low-grade cytological atypia		
High-grade mucinous carcinoma peritonei	Mucinous epithelium with high-grade cytological atypia	<ul style="list-style-type: none"> <li>• High-grade mucinous adenocarcinoma</li> <li>• AJCC grade G2</li> <li>• Peritoneal mucinous carcinomatosis with intermediate features</li> </ul>	23-61%
High-grade mucinous carcinoma peritonei with signet ring cells	PMP with signet ring cells	<ul style="list-style-type: none"> <li>• High-grade mucinous adenocarcinoma with signet ring cells</li> <li>• AJCC grade G3</li> <li>• Peritoneal mucinous carcinomatosis with intermediate features</li> </ul>	23%

Note: The grade of the primary mucinous appendiceal tumour and of the PMP may differ.



According to the 7<sup>th</sup> edition of the TNM classification, a mucinous tumour of the appendix with peritoneal spread beyond the right lower quadrant (PMP) is classified M1a = Stage IV disease, further stratified into IVA for low-grade peritoneal disease and IVB for high-grade peritoneal disease (in the absence of other metastasis).

### References

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