Sponsored by ACPMP APPENDIX CANCER PSEUDOMYXOMA PERITONEI RESEARCH FOUNDATION

Tumours of the Appendix Histopathology Reporting Guide

ICCR

Family/Last name

Given name(s)

Date of birth DD – MM – YYYY

Patient identifiers

Date of request DD – MM – YYYY

Accession/Laboratory number

Elements in **black text** are CORE. Elements in **grey text** are NON-CORE.

☐ indicates multi-select values ◯ indicates single select values

SCOPE OF THIS DATASET

CLINICAL INFORMATION (Note 1)

- ◯ Information not provided
- ◯ Information provided (select all that apply)
  - ☐ Appendicitis
  - ☐ Appendiceal mass/tumour
  - ☐ Preoperative therapy, *specify*
  - ☐ Other clinical information, *specify*

OPERATIVE PROCEDURE (Note 2)

- ◯ Not specified
- ◯ Appendicectomy
- ◯ Appendicectomy with partial caecectomy
- ◯ Ileocaecectomy
- ◯ Right colectomy
- ◯ Other, *specify*

TUMOUR SITE (select all that apply) (Note 3)

- ☐ Proximal appendix
- ☐ Distal appendix
- ☐ Entire length of the appendix
- ☐ Other, *specify*

MACROSCOPIC APPEARANCE (select all that apply) (Note 4)

- ☐ Perforation of the appendix
- ☐ Presence of mucin on the surface
- ☐ Other, *specify*

TUMOUR DIMENSIONS (Note 5)

Greatest dimension mm

Additional dimensions mm x mm

- ◯ Cannot be assessed, *specify*

BLOCK IDENTIFICATION KEY (Note 6)

(List overleaf or separately with an indication of the nature and origin of all tissue blocks)

HISTOLOGICAL TUMOUR TYPE (Note 7)

(Value list based on the World Health Organization Classification of Digestive System Tumours, 6th Edition (2026))

- ◯ Mucinous adenoma
- ◯ Adenoma, intestinal-type
  - ◯ Low grade
  - ◯ High grade
- ◯ Appendiceal mucinous neoplasm
  - ◯ Low grade
  - ◯ High grade
- ◯ Appendiceal mucinous neoplasm with extra-appendiceal spread
- ◯ Adenocarcinoma, not otherwise specified (NOS)
  - ◯ Signet ring cell adenocarcinoma
  - ◯ Mucinous adenocarcinoma
  - ◯ Non-mucinous adenocarcinoma
  - ◯ Undifferentiated carcinoma
- ◯ Goblet cell adenocarcinoma
- ◯ Neuroendocrine neoplasms[a]
  - ◯ Neuroendocrine carcinoma, NOS
    - ◯ Large cell
    - ◯ Small cell
  - ◯ Mixed neuroendocrine-non-neuroendocrine neoplasm (MiNEN)
- ◯ Other, *specify*

[a] *Neuroendocrine tumours are not covered in this dataset.*

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HISTOLOGICAL TUMOUR GRADE (Note 8)

Appendiceal mucinous neoplasms

- ◯ Low grade
- ◯ High grade
- ◯ Cannot be assessed, *specify*

Other

- ◯ G1: Well differentiated
- ◯ G2: Moderately differentiated
- ◯ G3: Poorly differentiated
- ◯ Cannot be assessed, *specify*

APPENDIX PERFORATION AT OR AWAY FROM TUMOUR (Note 9)

- ◯ Not identified
- ◯ Perforation at tumour
- ◯ Perforation away from tumour

EXTENT OF INVASION (Note 10)

- ◯ Cannot be assessed
- ◯ Tumour confined to mucosa
- ◯ Tumour or mucin extends into submucosa
- ◯ Tumour or mucin extends into muscularis propria
- ◯ Tumour or mucin extends to subserosa and/or mesoappendix
- ◯ Tumour or mucin invades visceral peritoneum
- ◯ Tumour directly invades other adjacent organ(s)/ structure(s), *specify*
- ◯ Other, *specify*

LYMPHOVASCULAR INVASION[b] (Note 11)

- ◯ Not identified
- ◯ Indeterminate
- ◯ Present

[b] *Core for all invasive tumours; non-core for low grade mucinous neoplasm (LAMN) and high grade appendiceal mucinous neoplasm (HAMN).*

PERINEURAL INVASION[b] (Note 12)

- ◯ Not identified
- ◯ Indeterminate
- ◯ Present

MARGIN STATUS (Note 13)

- ◯ Cannot be assessed
- ◯ Not involved
  - Distance of tumour from closest margin ____ mm
  - Specify closest margin
- ◯ Involved
  - Specify margin

LYMPH NODE STATUS (Note 14)

- ◯ No nodes submitted or found

Number of lymph nodes examined

- ◯ Not involved
- ◯ Involved
  - Number of involved lymph nodes

TUMOUR DEPOSITS[b] (Note 15)

- ◯ Not applicable
- ◯ Cannot be assessed
- ◯ Not identified
- ◯ Present
  - Number of tumour deposits

ADDITIONAL FINDINGS (select all that apply) (Note 16)

- ◯ None identified
- ☐ Appendicitis
- ☐ Ulcerative colitis
- ☐ Crohn disease
- ☐ Diverticulosis
- ☐ Neuroendocrine tumour[a]
- ☐ Other, *specify*

[a] *Neuroendocrine tumours are not covered in this dataset.*

ANCILLARY STUDIES (Note 17)

Mismatch repair (MMR) immunohistochemistry[c]

- ◯ Not performed
- ◯ Performed
  - ◯ Not interpretable
  - ◯ Proficient
  - ◯ Deficient, *specify*

Microsatellite instability (MSI) testing[c]

- ◯ Not performed
- ◯ Performed
  - ◯ MS-stable
  - ◯ MSI-high

Other, *record test(s), methodology and result(s)*

Representative blocks for ancillary studies, *specify those blocks best representing tumour and/or normal tissue for further study*

[c] *Core for invasive carcinoma only.*

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ANCILLARY STUDIES continued (Note 17)

HIGH GRADE NEUROENDOCRINE NEOPLASMS ONLY[a]

Neuroendocrine markers

- ◯ Not performed
- ◯ Performed (select at least two of the following core options)
  - ☐ Synaptophysin
  - ☐ Chromogranin-A
  - ☐ INSM1
  - ☐ Other, *specify*

Specify test(s) performed and result(s) if available

Ki-67 proliferation index

- ◯ Not performed
- ◯ Performed
  - ____ %

p53

- ◯ Not performed
- ◯ Performed
  - ◯ Abnormal, *specify test(s) performed and result(s) if available*
  - ◯ Wildtype pattern

RB1

- ◯ Retained
- ◯ Deficient

Low molecular weight cytokeratin(s)

- ◯ Positive
- ◯ Negative

[a] *Neuroendocrine tumours are not covered in this dataset.*

PERITONEAL METASTASES (Note 18)

- ◯ Not identified
- ◯ Present
  - ◯ Acellular mucin only
  - ◯ Mucin with mucinous neoplastic cells, *specify grade*
  - ◯ Metastatic adenocarcinoma, non-mucinous
  - ◯ Goblet cell adenocarcinoma

NON-PERITONEAL METASTASES (Note 19)

- ◯ Not known
- ◯ Present, *specify site(s)*

PATHOLOGICAL STAGING (UICC TNM 9th edition)[d] (Note 20)

TNM Descriptors (only if applicable) (select all that apply)

- ☐ m - multiple primary tumours
- ☐ y - post-therapy
- ☐ r - recurrent

Primary tumour (pT)

- ◯ TX[e] Primary tumour cannot be assessed
- ◯ T0 No evidence of primary tumour
- ◯ Tis Carcinoma in situ: intraepithelial or invasion of lamina propria
- ◯ Tis (LAMN)[f] Low grade appendiceal mucinous neoplasm confined to the muscularis propria; acellular mucin or mucinous epithelium may invade into the muscularis propria
- ◯ T1 Tumour invades submucosa[g]
- ◯ T2 Tumour invades muscularis propria[g]
- ◯ T3 Tumour invades subserosa or mesoappendix
- ◯ T4 Tumour perforates visceral peritoneum, including mucinous peritoneal tumour or acellular mucin on the serosa of the appendix or mesoappendix and/or directly invades other organs or structures[h]
  - ◯ T4a Tumour perforates visceral peritoneum, including mucinous peritoneal tumour or acellular mucin on the serosa of the appendix or mesoappendix
  - ◯ T4b Tumour directly invades other organs or structures

Regional lymph nodes (pN)

- ◯ NX[e] Regional lymph nodes cannot be assessed
- ◯ N0 No regional lymph node metastasis
- ◯ N1 Metastasis in 1-3 regional lymph nodes
  - ◯ N1a Metastasis in 1 regional lymph node
  - ◯ N1b Metastases in 2-3 regional lymph nodes
  - ◯ N1c Tumour deposit(s), i.e., satellites,[i] in the subserosa, or in non-peritonealised pericolic or peri-rectal soft tissue *without* regional lymph node metastasis
- ◯ N2 Metastasis in 4 or more regional lymph nodes

Distant metastasis (pM)

- ◯ M1 Distant metastasis microscopically confirmed
  - ◯ M1a Intraperitoneal acellular mucin only
  - ◯ M1b Intraperitoneal metastasis only, including mucinous epithelium
  - ◯ M1c Non-peritoneal metastasis

[d] *Reproduced with permission. Source: UICC TNM Classification of Malignant Tumours, 9th Edition, eds by James Brierley, Meredith Giuliani, Brian O'Sullivan, Brian Rous, Elizabeth Van Eycken. 2025, Publisher Wiley (incorporating any errata published up until 18 July 2026).*

[e] *TX and NX should be used only if absolutely necessary.*

[f] *The Tis(LAMN) category does not apply to HAMNs; HAMN are staged using the appendiceal adenocarcinoma T categories.*

[g] *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.*

[h] *Direct invasion in T4 includes invasion of other intestinal segments by way of the serosa, e.g., invasion of ileum. Tumour that is adherent to other organs or structures, macroscopically, is classified cT4b. However, if no tumour is present in the adhesion, microscopically, the classification should be pT1, 2 or 3.*

[i] *Tumour deposits (TDs) represent discrete tumour nodules of any shape, contour or size in peri-rectal and peri-colonic fat, away from the leading edge of the tumour, within the lymph drainage area of the primary carcinoma. TDs can originate from different histological structures, including lymph nodes, vessels and nerves. Therefore, TDs may contain foci of extramural vascular invasion (EMVI) and perineural invasion (PNI). The feature distinguishing a TD from EMVI and PNI is the presence of unequivocal tumour extension from the vessel or nerve into the surrounding fat or fibroconnective tissue. When tumour outgrowth from EMVI and/or PNI is present, the diagnosis of TDs and EMVI/PNI should be denoted separately in the report. If the tumour involves an identifiable lymph node, it is considered as lymph node metastasis and not as TDs even if the tumour extends into the perinodal fat.*

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Definitions

CORE elements

CORE elements are those which are essential for the clinical management, staging or prognosis of the cancer. These elements will either have evidentiary support at Level III-2 or above (based on prognostic factors in the National Health and Medical Research Council (NHMRC) levels of evidence[1]). In rare circumstances, where level III-2 evidence is not available an element may be made a CORE element where there is unanimous agreement by the Dataset Authoring Committee (DAC). An appropriate staging system, e.g., Pathological TNM staging, would normally be included as a CORE element.

Molecular and immunohistochemical testing is a growing feature of cancer reporting. However, in many parts of the world this type of testing is limited by the available resources. In order to encourage the global adoption of ancillary tests for patient benefit, International Collaboration on Cancer Reporting (ICCR) includes the most relevant ancillary testing in ICCR Datasets as CORE elements, especially when they are necessary for the diagnosis. Where the technical capability does not yet exist, laboratories may consider temporarily using these data elements as NON-CORE items.

The summation of all CORE elements is considered to be the minimum reporting standard for a specific cancer.

NON-CORE elements

NON-CORE elements are those which are unanimously agreed should be included in the dataset but are not supported by level III-2 evidence. These elements may be clinically important and recommended as good practice but are not yet validated or regularly used in patient management.

Key information other than that which is essential for clinical management, staging or prognosis of the cancer such as macroscopic observations and interpretation, which are fundamental to the histological diagnosis and conclusion e.g., macroscopic tumour details, may be included as either CORE or NON-CORE elements by consensus of DAC.

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Scope

The dataset has been developed for the pathological reporting of carcinomas and mucinous neoplasms of the appendix. It includes specimens designated appendectomy with or without segmental resection. Biopsy specimens are excluded. Neuroendocrine carcinomas (NECs), mixed neuroendocrine-non-neuroendocrine neoplasms (MiNENs) and amphicrine-like carcinoma are included. Neuroendocrine tumours (NETs) and non-epithelial malignancies are excluded.

In development of this dataset, the DAC considered evidence up until July 2026.

The authors of this dataset can be accessed here.

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Note 1 – Clinical information (Core and Non-core)

Clinical information can be provided by the clinician on the pathology request form. Pathologists can search for additional information from previous pathology reports, if any, or accessible medical records. Whether the patient presented with abdominal pain/appendicitis or with features clinically concerning for an underlying mass should be recorded. It is worth noting that many appendiceal tumours can present with symptoms of appendicitis, particularly appendiceal mucinous neoplasms and goblet cell adenocarcinoma, although a significant number present with features concerning for neoplasm.[2-6] In fact, presentation as appendicitis correlates with better prognosis in patients with goblet cell adenocarcinoma.[7,8] The mimics of mucinous neoplasm such as interval appendectomy specimens, post-inflammatory mucosal hyperplasia and appendiceal diverticular disease most often present with features of appendicitis or treated appendicitis, and only rarely does the surgeon raise concern for neoplasia, whereas appendiceal mucinous neoplasms often present with a clinical concern for underlying tumour.[9-12]

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Note 2 – Operative procedure (Core)

The operative procedure pertains to the appendix primary tumour; additional specimens that are submitted, such as spleen or peritoneal sampling/debulking in patients with disseminated disease, are separately recorded and described according to their nature. The type of specimen in appendiceal neoplasia affects the likelihood of an involved surgical resection margin.[13,14] Furthermore, the nature of the operative procedure has been shown in several studies to have prognostic importance in mucinous and non-mucinous adenocarcinomas of the appendix, with right hemicolectomy being superior to appendectomy,[14-20] although some studies showed no significant difference in survival in patients with high stage tumours or well differentiated tumours.[20,21] Right hemicolectomy has also shown to improve survival compared to appendectomy in Stage II goblet cell adenocarcinoma.[13,14] However, one study showed no survival advantage to right hemicolectomy in goblet cell adenocarcinoma.[22]

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Note 3 – Tumour site (Non-core)

Location of the tumour along the appendix may affect the clinical presentation; for example, an early paper on appendiceal adenocarcinoma described tumours at the base of the appendix presenting with symptoms of appendicitis.[23] However, in a review of published cases before 1956, another group of authors found no correlation between tumour location and the co-existence of acute appendicitis.[24] Tumours that involve the base may be more likely to involve the margin in appendectomy specimens than tumours that involve the distal half of the appendix. However, location of the tumour along the appendix has not been shown to affect prognosis, independent of the stage of the tumour.[25,26]

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Note 4 – Macroscopic appearance (Non-core)

The macroscopic appearance of the tumour should be recorded, particularly gross perforation and the presence or absence of mucinous tumour on the appendiceal serosal surface. In patients with appendiceal mucinous neoplasms, rupture of the appendix confers a risk of developing pseudomyxoma peritonei.[27] Other macroscopic features such as cystic change (e.g., mucocele), solid areas, polypoid lesions, infiltrative type lesions, or thickening of the appendix may be noted. In some cases, the tumour may not be grossly evident.

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Note 5 – Tumour dimensions (Core and Non-core)

Recording tumour size, based on a combination of macroscopic and microscopic assessment, allows correlation with pre-operative imaging and surgical assessments. Assessment of tumour dimensions should, if possible, exclude any inflammatory component. No prognostic significance has been attached to tumour size for appendiceal cancer and size does not directly influence tumour staging.[4] In one study of non-mucinous adenocarcinomas, tumour size less than 20 millimetres (mm) had a statistically significantly lower risk of having lymph node metastases on univariate analysis but not on multivariate analysis.[15] Goblet cell adenocarcinoma is notorious for being grossly inapparent and difficult to measure. Not surprisingly, studies have shown no correlation between size and prognosis in patients with goblet cell adenocarcinoma.[2,3]

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Note 6 – Block identification key (Non-core)

The origin/designation of all tissue blocks should be recorded. This information should ideally be documented in the final pathology report and is particularly important when further internal or external review arises. The reviewer needs to have unequivocal description of the origin of each block in order to provide an informed specialist opinion. If this information is not included in the final pathology report, it should be available on the laboratory computer system and relayed to the reviewing pathologist. It is highly encouraged to have a digital image (photograph) of the specimen and record of the key of the tumour blocks.

Recording the origin/designation of tissue blocks also facilitates retrieval of blocks for further immunohistochemical or molecular analysis, research studies, or clinical trials.

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Note 7 – Histological tumour type (Core)

Histologic type is a major determinant of tumour biology, prognosis, and patient management.[28] All carcinomas and mucinous neoplasms of the appendix should be classified based on the 6th edition World Health Organization (WHO) Classification of Digestive System Tumours, 2026 (Table 1).[28] Appendiceal mucinous neoplasms generally do not metastasise to lymph nodes, and therefore right hemicolectomy is generally not required. However, they frequently metastasise to the peritoneal cavity where peritoneum-directed therapy becomes relevant. Among invasive adenocarcinomas, mucinous adenocarcinoma generally has a better prognosis than non-mucinous adenocarcinoma.[6,29] In some studies, this difference is only

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among Stage IV tumours.[30,31] A study of patients with appendiceal tumours and peritoneal metastases found that histologic tumour type was associated with prognosis on multivariate analysis, with non-mucinous adenocarcinoma having a worse prognosis than appendiceal mucinous neoplasms.[32] Among non-mucinous adenocarcinomas, signet ring cell carcinomas often present at higher tumour stage and are associated with a worse outcome than non-signet ring cell carcinomas.[30,33] Conversely, non-signet ring cell morphology is a lower risk for lymph node metastases than signet ring cell morphology.[15]

Table 1: 6th edition of the World Health Organization Classification of tumours of the appendix.[28]

Descriptor	ICD-O codes[a]
Benign epithelial tumours and precursors	
Mucinous adenoma	84800/0
Adenoma, intestinal-type, low-grade	81441/0
Adenoma, intestinal-type, high-grade	81441/2
Malignant epithelial tumours	
Low-grade appendiceal mucinous neoplasm	84800/2
High-grade appendiceal mucinous neoplasm	84801/2
Appendiceal mucinous neoplasm with extra-appendiceal spread	84803/3
Adenocarcinoma, not otherwise specified (NOS)	81400/3
Signet ring cell adenocarcinoma	84900/3
Mucinous adenocarcinoma	84800/3
Non-mucinous adenocarcinoma	82520/3
Undifferentiated carcinoma	80200/3
Goblet cell adenocarcinoma	82430/3
Neuroendocrine carcinoma, NOS	82460/3
Large cell neuroendocrine carcinoma	80130/3
Small cell neuroendocrine carcinoma	80410/3
Mixed neuroendocrine-non-neuroendocrine neoplasm (MiNEN)	81540/3

[a] These morphology codes are from the International Classification of Diseases for Oncology, fourth edition, (ICD-O-4.0).[34] Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour: /2 for carcinoma in situ and grade lll intraepithelial neoplasia; /3 for malignant tumours, primary site; and /6 for malignant tumours, metastatic site. Behaviour code /6 is not generally used by cancer registries.

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Note 8 – Histological tumour grade (Core)

Various tumour types have their own grading systems, including low grade mucinous neoplasm (LAMN)/high grade appendiceal mucinous neoplasm (HAMN), mucinous adenocarcinoma and goblet cell adenocarcinoma.[28] It is also worth noting that pseudomyxoma peritonei has its own grading system, separate from the primary tumour.[28]

Tumour grade has been shown across many studies to affect the likelihood of lymph node involvement as well as prognosis. In one study, univariate analysis showed that tumour grade is significantly associated with

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overall and disease specific survival in primary appendiceal adenocarcinoma and goblet cell adenocarcinoma.[6] Among appendiceal non-mucinous adenocarcinomas, poor and undifferentiated carcinomas conferred high risk for lymph node metastases compared to well or moderately differentiated tumours and showed worse disease specific survival.[15,35] In other studies of mucinous and non-mucinous adenocarcinoma, tumour grade correlates with overall survival and disease specific survival,[16] including among Stage IV tumours.[30] In goblet cell adenocarcinoma, tumour grade correlates with prognosis, regardless of the classification system.[8,36-39] In patients with goblet cell adenocarcinoma, grade may be an important prognostic factor in terms of survival, but the data are limited.[40]

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Note 9 – Appendix perforation at or away from tumour (Non-core)

Perforation of the appendix at, or away from, tumour incorporates both macroscopic and microscopic examination. Perforation of an appendiceal adenocarcinoma at the site of the tumour has been shown to be a poor prognostic finding, compared to perforation distal to the tumour.[25] In patients with appendiceal cancer and appendicitis, rupture of the appendix (presumably due to appendicitis rather than tumour) did not affect survival.[18] In appendiceal mucinous neoplasms, perforation away from the tumour has not been specifically studied; perforation of the appendix confers risk of pseudomyxoma peritonei by allowing mucin and neoplastic cells to escape,[27] and that risk can be further estimated by the identification of mucin and/or neoplastic mucinous epithelial cells on the appendiceal serosa or beyond.[41-43] In goblet cell adenocarcinoma, one study found perforation of the appendix to be prognostically significant on univariate but not multivariate analysis.[37]

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Note 10 – Extent of invasion (Core)

Appendiceal mucinous neoplasms that are confined to the appendix (LAMN/HAMN), with an intact appendiceal serosa, have been shown across several studies to be cured by appendectomy in the vast majority of cases.[43-45] However, exceptional cases of peritoneal recurrence have been reported in patients with tumours apparently confined to the appendix (i.e., neither mucin nor neoplastic cells outside the appendix).[46] In cases with perforation of the appendiceal serosa, the presence of acellular mucin on the serosa is associated with a low risk of recurrence, whereas neoplastic cells in mucin on the serosa confers high risk of recurrence as pseudomyxoma peritonei.[4,41,43] Therefore, it is imperative that pathologists report the presence of mucin on the appendiceal serosa, and distinguish between acellular mucin and cellular mucin. Both scenarios are designated as pT4a, and therefore pathologic T stage is insufficient to convey this important information. When an appendix contains a non-mucinous tumour and shows distal perforation due to appendicitis or obstruction, it may be appropriate to stage the tumour based on its extent of invasion separately from the serosal mucin, if that serosal mucin can be attributed to obstruction. In appendiceal mucinous tumours (LAMN/HAMN), it may be impossible to determine if neoplastic cells escaped into the peritoneum whenever there is serosal mucin, and it is appropriate to stage the tumour based on the spread of mucin as pT4a.

Among adenocarcinomas, one study of non-mucinous adenocarcinoma showed that T stage (T1 versus >T1) was independently associated with the risk of lymph node metastasis.[15] T stage also correlates with overall survival, progression free survival, and disease specific survival in appendiceal adenocarcinoma.[6,16,35] Similarly, T stage has been shown to correlate with the likelihood of lymph node metastasis in goblet cell adenocarcinoma[13] and to correlate with progression free survival and overall survival.[6,22] In other studies, T

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stage did not correlate with prognosis; patients with Stage IV disease had a worse prognosis than patients with Stage I to III.[7,8]

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Note 11 – Lymphovascular invasion (Core)

Lymphovascular invasion is not an expected finding in appendiceal mucinous neoplasms, but its presence has been shown to be an adverse prognostic finding in appendiceal adenocarcinoma.[20,29] Furthermore, the presence of lymphovascular invasion predicts lymph node involvement in appendiceal mucinous and non-mucinous adenocarcinoma, including goblet cell adenocarcinoma.[2,15,47,48] Some studies of patients with goblet cell adenocarcinoma showed lymphovascular invasion was significant on univariate analysis, although it was no longer significant on multivariate analysis.[8,37,49] In one study of patients with Stage I to III goblet cell adenocarcinoma, lymphovascular invasion did not predict recurrence.[7]

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Note 12 – Perineural invasion (Core)

Perineural invasion is highly unlikely in appendiceal mucinous neoplasms. In contrast, it is commonly present in goblet cell adenocarcinomas. In goblet cell adenocarcinoma, perineural invasion has been shown to be a negative prognostic factor, at least on univariate analysis.[2,8,49] However, in a study on goblet cell adenocarcinomas, perineural invasion did not predict recurrence in patients with Stage I-III disease.[7] Another large series of goblet cell adenocarcinomas found no prognostic difference in patients with and without perineural invasion.[37]

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Note 13 – Margin status (Core)

The margins of an appendectomy specimen include the proximal transection margin and, when it can be identified, the mesenteric margin. However, the status of the proximal transection margin is usually the margin of interest. Acellular mucin at the proximal margin should be reported but not classified as a positive margin. Most studies that have examined the outcome of appendiceal mucinous tumours with proximal margin involvement have concluded that tumours that involve the appendiceal margin will not recur and additional resection specimens do not show residual tumour.[4,42,45,50-52] One study found that a positive margin was associated with increased frequency of disease recurrence or death, although confounding factors such as perforation were not considered.[53] In the setting of either invasive adenocarcinoma or goblet cell adenocarcinoma, involvement of the proximal appendiceal margin warrants consideration of additional resection.

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Note 14 – Lymph node status (Core)

The likelihood of lymph node involvement differs based on tumour type, stage, and tumour grade. Appendiceal mucinous neoplasms are unlikely to show lymph node involvement.[32,54] Well-differentiated mucinous adenocarcinoma has a low likelihood of lymph node involvement, whereas high grade mucinous adenocarcinoma and signet ring cell carcinomas have higher likelihood of lymph node involvement.[54-56] Tumour T stage also correlates with the likelihood of nodal metastases.[54,56] This understanding may inform the extent of surgical resection as well as the number of lymph nodes considered adequate for appropriate pathologist stage. Finding fewer than 10 lymph nodes in a right colectomy specimen has been reported to correlate with reduced overall survival for patients with appendiceal adenocarcinoma, presumably due to missed occult metastases.[55] Whereas, finding fewer than 12 lymph nodes was found to be associated with worse prognosis in a study by Xie et al (2016).[57] In appendiceal adenocarcinoma, including mucinous adenocarcinoma, signet ring cell carcinoma, and goblet cell adenocarcinoma, lymph node involvement has been shown to be an adverse prognostic finding.[13,20,29,35,54,56]

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Note 15 – Tumour deposits (Core)

Tumour deposits have not been studied in appendiceal adenocarcinoma, but they should be reported given their established significance in colorectal adenocarcinoma. Tumour deposits are less relevant in appendiceal mucinous neoplasms or mucinous adenocarcinomas, since the distinction between a tumour deposit and mucinous carcinoma peritonei is ambiguous. In those settings, discontinuous mucinous tumour deposits should be interpreted as peritoneal dissemination of a mucinous tumour and infiltration of the underlying connective tissue. This element is non-core (optional) for LAMN and HAMN.

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Note 16 – Additional findings (Non-core)

The presence of other pathologic findings in the appendix may be recorded. Additional mucosal lesions such as sessile serrated lesion, conventional colorectal type adenoma, or mucinous adenoma may be included under ‘other’. Since it is difficult in the appendix to distinguish sessile serrated lesion from mucosal hyperplasia secondary to obstructing tumours, and given the small size of the appendix, it is unusual to have a benign polyp separate from the invasive tumour and its precursor. In cases of goblet cell adenocarcinoma, in which a conventional precursor is generally not present, overlying mucosal benign polyps may be noted, although it is challenging to discriminate mucosal obstructive changes from sessile serrated lesions or even low grade appendiceal mucinous neoplasms. NETs should only be included as an additional finding if they are incidental and pose no significant risk to the patient.

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Note 17 – Ancillary studies (Core and Non-core)

Mismatch repair (MMR) immunohistochemistry and/or microsatellite instability (MSI) testing should be performed on all invasive carcinomas, although MMR deficiency in appendiceal adenocarcinoma is rare.[58-60] RAS, GNAS, and TP53 are frequently mutated in appendiceal adenocarcinoma and have shown prognostic significance in some studies.[61] In patients with peritoneal metastases, molecular studies for RAS, GNAS, and TP53 may be useful for patient prognostication and management, and should be considered.[62,63]

If a pure or mixed neuroendocrine neoplasm is suspected on morphology, positivity for at least two markers of neuroendocrine differentiation by immunohistochemistry is required (core) to confirm neuroendocrine differentiation, usually synaptophysin, chromogranin A and/or INSM1. As with other gastrointestinal tract and pancreatic neuroendocrine neoplasms, assessment of proliferation index by Ki-67 immunohistochemistry is a core component as it is fundamental to grading of the neuroendocrine component.[28] For the differential diagnosis with grade 3 NET, p53 (core) and Rb1 (non-core) can be applied. Low molecular weight cytokeratins can be used (non-core). MMR stains may also be considered for NECs (non-core). A Ki-67 proliferation index <55% is associated with better overall survival in NECs.[64] TP53 mutations are found in over 69% of NEC compared to 10-15% of NET grade 3 (G3),[65,66] while RB1 loss is found in 40-60% of NEC and only very rarely in NET G3.[67]

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Note 18 – Peritoneal metastases (Core)

Peritoneal metastasis is a common mode of spread of appendiceal malignancies. The presence or absence of mucin with or without tumour cells is essential to appropriately manage patients with these malignancies. The presence of acellular mucin in the peritoneal cavity has been shown to carry a lower risk for progressive disease compared to cellular mucinous tumour deposits, and to have a better prognosis than cellular mucin deposits.[32,68-70] In contrast, the presence of mucinous epithelial cells within peritoneal mucin is a progressive disease, and the peritoneal tumour grade has prognostic value in that setting.[68,71-75] In cases in which the peritoneal tumour grade and the primary tumour grade are discordant, both tumour grades should be recorded. In this setting, the grade of the peritoneal tumour correlates with prognosis.[76,77] It should be noted that peritoneal metastases of mucinous neoplasms ('pseudomyxoma peritonei') have a different grading system than primary tumours.[28]

Patients with non-mucinous adenocarcinoma or goblet cell adenocarcinoma who develop peritoneal metastases have a relatively poor prognosis.[40] One study showed no difference in overall survival in patients with peritoneal metastases of non-mucinous adenocarcinoma and goblet cell adenocarcinoma.[32]

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Note 19 – Non-peritoneal metastases (Core and Non-core)

Data regarding the significance of non-peritoneal metastases is limited, but the consensus of the DAC was that reporting the presence of non-peritoneal metastases should be a core element. The reporting of the sites where present is non-core.

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Note 20 – Pathological staging (Core)

The pathologic stage should be assessed according to the agreed criteria of the Union for International Cancer Control (UICC)/American Joint Committee on Cancer (AJCC) 9th edition Cancer Staging Manuals as it provides critical information for patient management and prognosis.[78,79] In SEER data, Stage IV appendiceal adenocarcinoma is associated with worse prognosis.[80] In goblet cell adenocarcinoma, distant spread is associated with a worse prognosis compared to local or regional disease.[22]

Reporting of pathological staging categories (pT,pN,pM) is based on the evidence available to the pathologist at the time of reporting. As indicated in UICC/AJCC TNM9,[78,79] the final stage grouping of a patient's tumour is based on a combination of pathological staging and other clinical and imaging information.

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References

1 Merlin T, Weston A and Tooher R (2009). Extending an evidence hierarchy to include topics other than treatment: revising the Australian 'levels of evidence'. *BMC Med Res Methodol* 9:34.

2 Nonaka D, Papaxoinis G, Lamarca A, Fulford P, Valle J and Chakrabarty B (2018). A study of appendiceal crypt cell adenocarcinoma (so-called goblet cell carcinoid and its related adenocarcinoma). *Hum Pathol* 72:18–27.

3 Pham TH, Wolff B, Abraham SC and Drelichman E (2006). Surgical and chemotherapy treatment outcomes of goblet cell carcinoid: a tertiary cancer center experience. *Ann Surg Oncol* 13(3):370–376.

4 Aleter A, El Ansari W, Toffaha A, Ammar A, Shahid F and Abdelaal A (2021). Epidemiology, histopathology, clinical outcomes and survival of 50 cases of appendiceal mucinous neoplasms: Retrospective cross-sectional single academic tertiary care hospital experience. *Ann Med Surg (Lond)* 64:102199.

5 Palmer K, Weerasuriya S, Chandrakumaran K, Rous B, White BE, Paisey S, Srirajaskanthan R and Ramage JK (2022). Goblet Cell Adenocarcinoma of the Appendix: A Systematic Review and Incidence and Survival of 1,225 Cases From an English Cancer Registry. *Front Oncol* 12:915028.

6 Chen I, Liu X, Kovar-Peltz S, Conrad SJ, Chen HH and Liao X (2023). Clinicopathological spectrums and prognosis of primary appendiceal adenocarcinoma, goblet cell adenocarcinoma, and low-grade appendiceal mucinous neoplasms. *Pathology* 55(3):375–382.

7 Tsang ES, McConnell YJ, Schaeffer DF, Lee L, Yin Y, Zerhouni S, Schaff K, Speers C and Kennecke HF (2018). Outcomes of Surgical and Chemotherapeutic Treatments of Goblet Cell Carcinoid Tumors of the Appendix. *Ann Surg Oncol* 25(8):2391–2399.

8 Lee LH, McConnell YJ, Tsang E, Zerhouni S, Speers C, Kennecke H and Schaeffer DF (2015). Simplified 2-tier histologic grading system accurately predicts outcomes in goblet cell carcinoid of the appendix. *Hum Pathol* 46(12):1881–1889.

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© 2026 International Collaboration on Cancer Reporting Limited (ICCR).

9 Lowes H, Rowaiye B, Carr NJ and Shepherd NA (2019). Complicated appendiceal diverticulosis versus low-grade appendiceal mucinous neoplasms: a major diagnostic dilemma. *Histopathology* 75(4):478–485.

10 Hissong E, Goncharuk T, Song W and Yantiss RK (2020). Post-inflammatory mucosal hyperplasia and appendiceal diverticula simulate features of low-grade appendiceal mucinous neoplasms. *Mod Pathol* 33(5):953–961.

11 Malvar G, Peric M and Gonzalez RS (2022). Interval appendicitis shows histological differences from acute appendicitis and may mimic Crohn disease and other forms of granulomatous appendicitis. *Histopathology* 80(6):965–973.

12 Mostyka M, Yantiss RK, Chen Z and Tseng-Chen Y (2022). Interval Appendectomy Specimens. *Arch Pathol Lab Med* 147(5):546–551.

13 Kowalsky SJ, Nassour I, AlMasri S, Paniccia A, Zureikat AH, Choudry HA and Pingpank JF (2021). Omission of Right Hemicolectomy May be Safe for Some Appendiceal Goblet Cell Adenocarcinomas: A Survival Analysis of the National Cancer Database. *Ann Surg Oncol* 28(13):8916–8925.

14 Marks VA, Kerekes D, Butensky S, Ahuja N, Johnson C, Turaga K and Khan SA (2023). Role of colectomy in the management of appendiceal tumors: a retrospective cohort study. *BMC Gastroenterol* 23(1):398.

15 Straker RJ, 3rd, Grinberg SZ, Sharon CE, Shannon AB, Fraker DL, Shanmugan S, Miura JT and Karakousis GC (2022). Pathologic Factors Associated with Low Risk of Lymph Node Metastasis in Nonmucinous Adenocarcinoma of the Appendix. *Ann Surg Oncol* 29(4):2334–2343.

16 Asare EA, Compton CC, Hanna NN, Kosinski LA, Washington MK, Kakar S, Weiser MR and Overman MJ (2016). The impact of stage, grade, and mucinous histology on the efficacy of systemic chemotherapy in adenocarcinomas of the appendix: Analysis of the National Cancer Data Base. *Cancer* 122(2):213–221.

17 Nitecki SS, Wolff BG, Schlinkert R and Sarr MG (1994). The natural history of surgically treated primary adenocarcinoma of the appendix. *Ann Surg* 219(1):51–57.

18 Lenriot JP and Huguier M (1988). Adenocarcinoma of the appendix. *Am J Surg* 155(3):470–475.

19 Hesketh KT (1963). The management of primary adenocarcinoma of the vermiform appendix. *Gut* 4(2):158–168.

20 AlMasri SS, Hammad AY, Singhi AD, Paniccia A, Zureikat AH, Celebrezze JP, Jr., Choudry HA and Nassour I (2023). Appendectomy Is Oncologically Equivalent to Right Hemicolectomy for Well-Differentiated T1 Appendiceal Adenocarcinoma. *Dis Colon Rectum* 66(1):67–74.

21 Turaga KK, Pappas S and Gamblin TC (2013). Right hemicolectomy for mucinous adenocarcinoma of the appendix: just right or too much? *Ann Surg Oncol* 20(4):1063–1067.

22 El Asmar ML, Mortagy M, Chandrakumaran K, Cecil T and Ramage J (2024). Right Hemicolectomy and Appendicectomy as Treatments for Goblet Cell Adenocarcinoma: A Comparative Analysis of Two Large National Databases. *Curr Oncol* 31(7):3855–3869.

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© 2026 International Collaboration on Cancer Reporting Limited (ICCR).

23 Uihlein A and McDonald JR (1943). Primary carcinoma of the appendix resembling carcinoma of the colon. *Surg Gynec & Obst* 76:711–714.

24 Sieracki JC and Tesluk H (1956). Primary adenocarcinoma of the vermiform appendix. *Cancer* 9(5):997–1011.

25 Didolkar MS and Fanous N (1977). Adenocarcinoma of the appendix: a clinicopathologic study. *Dis Colon Rectum* 20(2):130–134.

26 Gilhome RW, Johnston DH, Clark J and Kyle J (1984). Primary adenocarcinoma of the vermiform a ppendix: report of a series of ten cases, and review of the literature. *Br J Surg* 71(7):553–555.

27 Sueda S, Young S, Sung M, Hotta M, O'Connor V and Leung AM (2020). Predictors of Progression of Appendiceal Mucinous Neoplasm to Pseudomyxoma Peritonei. *Am Surg* 86(10):1379–1384.

28 WHO Classification of Tumours Editorial Board (ed) (2026). *WHO Classification of Tumours, Digestive System Tumours, 6th Edition*, IARC, Lyon.

29 Acs M, Gerken M, Zustin J, Blaj S, Isgandarova S and Piso P (2023). Prolonged Survival in Peritoneal Metastatic Appendiceal Carcinoma Patients Treated With Combined Cytoreductive Surgery and Hyperthermic Intraperitoneal Chemotherapy. *J Surg Res* 283:839–852.

30 Overman MJ, Fournier K, Hu CY, Eng C, Taggart M, Royal R, Mansfield P and Chang GJ (2013). Improving the AJCC/TNM staging for adenocarcinomas of the appendix: the prognostic impact of histological grade. *Ann Surg* 257(6):1072–1078.

31 Legué LM, van Erning FN, Creemers GJ, de Hingh I, Lemmens V and Huysentruyt CJ (2020). The prognostic relevance of histologic subtype in appendiceal adenocarcinoma. *Eur J Surg Oncol* 46(3):433–438.

32 Garach NR, Kusamura S, Guaglio M, Bartolini V, Deraco M and Baratti D (2021). Comparative study of mucinous and non-mucinous appendiceal neoplasms with peritoneal dissemination treated by cyoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC). *Eur J Surg Oncol* 47(5):1132–1139.

33 Enblad M, Egerszegi PP, Birgisson H, Sjöblom T, Glimelius B and Folkesson J (2023). Signet Ring Cell Colorectal and Appendiceal Cancer: A Small Signet Ring Cell Component Is Also Associated with Poor Outcome. *Cancers (Basel)* 5(9):2497.

34 International Agency for Research on Cancer (IARC) (2026). *International Classification of Diseases for Oncology, Fourth edition ICD-O-4*. Available from: tumourclassification.iarc.who.int/icd-o-4/ (Accessed 20 August 2026).

35 McClelland PH, Gregory SN, Nah SK, Hernandez JM, Davis JL and Blakely AM (2024). Predicting Survival in Mucinous Adenocarcinoma of the Appendix: Demographics, Disease Presentation, and Treatment Methodology. *Ann Surg Oncol* 31(9):6237–6251.

36 Burke AP, Sobin LH, Federspiel BH, Shekitka KM and Helwig EB (1990). Goblet cell carcinoids and related tumors of the vermiform appendix. *Am J Clin Pathol* 94(1):27–35.

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37 Yozu M, Johncilla ME, Srivastava A, Ryan DP, Cusack JC, Doyle L, Setia N, Yang M, Lauwers GY, Odze RD and Misdraji J (2018). Histologic and Outcome Study Supports Reclassifying Appendiceal Goblet Cell Carcinoids as Goblet Cell Adenocarcinomas, and Grading and Staging Similarly to Colonic Adenocarcinomas. *Am J Surg Pathol* 42(7):898–910.

38 Tang LH, Shia J, Soslow RA, Dhall D, Wong WD, O'Reilly E, Qin J, Paty P, Weiser MR, Guillem J, Temple L, Sobin LH and Klimstra DS (2008). Pathologic classification and clinical behavior of the spectrum of goblet cell carcinoid tumors of the appendix. *Am J Surg Pathol* 32(10):1429–1443.

39 Taggart MW, Abraham SC, Overman MJ, Mansfield PF and Rashid A (2015). Goblet cell carcinoid tumor, mixed goblet cell carcinoid-adenocarcinoma, and adenocarcinoma of the appendix: comparison of clinicopathologic features and prognosis. *Arch Pathol Lab Med* 139(6):782–790.

40 Shyu S, Choudry H, Hall L, Pingpank J, Holtzman M, Bartlett D and Pai RK (2020). Clinicopathological analysis of appendiceal goblet cell adenocarcinoma with peritoneal metastasis: World Health Organization grade predicts survival following cytoreductive surgery with intraperitoneal chemotherapy. *Histopathology* 77(5):798–809.

41 Yantiss RK, Shia J, Klimstra DS, Hahn HP, Odze RD and Misdraji J (2009). Prognostic significance of localized extra-appendiceal mucin deposition in appendiceal mucinous neoplasms. *Am J Surg Pathol* 33(2):248–255.

42 Baumgartner JM, Srivastava A, Melnitchouk N, Drage MG, Huber AR, Gonzalez RS, Bell P, Wu E, Resnick M, Turaga K, Poli E, Esquivel J, Deneve J, Kelly KJ, Veerapong J and Lowy AM (2021). A Multi-institutional Study of Peritoneal Recurrence Following Resection of Low-grade Appendiceal Mucinous Neoplasms. *Ann Surg Oncol* 28(8):4685–4694.

43 Pai RK, Beck AH, Norton JA and Longacre TA (2009). Appendiceal mucinous neoplasms: clinicopathologic study of 116 cases with analysis of factors predicting recurrence. *Am J Surg Pathol* 33(10):1425–1439.

44 Polydorides AD and Wen X (2022). Clinicopathologic parameters and outcomes of mucinous neoplasms confined to the appendix: a benign entity with excellent prognosis. *Mod Pathol* 35(11):1732–1739.

45 Lakmal C, Chakrabarty B, Tan C, Larby L, Kochhar R, Fish R, Sutton P, Wild J, Clouston H, Selvasekar C, Renehan AG, Wilson M, O'Dwyer ST and Aziz O (2024). The risk of developing pseudomyxoma peritonei from a non-perforated low grade appendiceal mucinous neoplasm found at appendicectomy. *Eur J Surg Oncol* 50(10):108600.

46 Sipok A, Dort JM, Visioni A and Bijelic L (2022). Retrospective Review of Outcomes in Non-Invasive Mucinous Appendiceal Neoplasms with and without Peritoneal Spread: A Cohort Study. *Curr Oncol* 29(12):9125–9134.

47 Shannon AB, Goldberg D, Song Y, Paulson EC, Roses RE, Fraker DL, Kelz RR, Miura J and Karakousis GC (2020). Predictors of lymph node metastases in patients with mucinous appendiceal adenocarcinoma. *J Surg Oncol* 122(3):399–406.

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48 Grotz TE, Royal RE, Mansfield PF, Overman MJ, Mann GN, Robinson KA, Beaty KA, Rafeeq S, Matamoros A, Taggart MW and Fournier KF (2017). Stratification of outcomes for mucinous appendiceal adenocarcinoma with peritoneal metastasis by histological grade. *World J Gastrointest Oncol* 9(9):354–362.

49 Reid MD, Basturk O, Shaib WL, Xue Y, Balci S, Choi HJ, Akkas G, Memis B, Robinson BS, El-Rayes BF, Staley CA, Staley CA, Winer JH, Russell MC, Knight JH, Goodman M, Krasinskas AM and Adsay V (2016). Adenocarcinoma ex-goblet cell carcinoid (appendiceal-type crypt cell adenocarcinoma) is a morphologically distinct entity with highly aggressive behavior and frequent association with peritoneal/intra-abdominal dissemination: an analysis of 77 cases. *Mod Pathol* 29(10):1243–1253.

50 Arnason T, Kamionek M, Yang M, Yantiss RK and Misdraji J (2015). Significance of proximal margin involvement in low-grade appendiceal mucinous neoplasms. *Arch Pathol Lab Med* 139(4):518–521.

51 Wong M, Barrows B, Gangi A, Kim S, Mertens RB and Dhall D (2020). Low-Grade Appendiceal Mucinous Neoplasms: A Single Institution Experience of 64 Cases With Clinical Follow-up and Correlation With the Current (Eighth Edition) AJCC Staging. *Int J Surg Pathol* 28(3):252–258.

52 Ibrahim E, Akrmah M and Ligato S (2023). Does a Positive Appendiceal Resection Margin in Low-Grade Appendiceal Mucinous Neoplasms, Warrant Additional Surgery? Our Institution Experience and Literature Review. *Ann Surg Oncol* 30(12):7189–7195.

53 Fournier K, Rafeeq S, Taggart M, Kanaby P, Ning J, Chen HC, Overman M, Raghav K, Eng C, Mansfield P and Royal R (2017). Low-grade Appendiceal Mucinous Neoplasm of Uncertain Malignant Potential (LAMN-UMP): Prognostic Factors and Implications for Treatment and Follow-up. *Ann Surg Oncol* 24(1):187–193.

54 Takeyama H, Murata K, Takeda T, Fujii M, Kagawa Y, Kawachi H, Yamaguchi T, Noura S, Masuishi T, Inoue A, Takii Y, Suto T, Sakamoto K, Tei M, Kishimoto M, Yao T and Sugihara K (2022). Clinical Significance of Lymph Node Dissection and Lymph Node Metastasis in Primary Appendiceal Tumor Patients After Curative Resection: a Retrospective Multicenter Cohort Study. *J Gastrointest Surg* 26(1):128–140.

55 Lopez-Ramirez F, Sardi A, King MC, Nikiforchin A, Falla-Zuniga LF, Barakat P, Nieroda C and Gushchin V (2024). Sufficient Regional Lymph Node Examination for Staging Adenocarcinoma of the Appendix. *Ann Surg Oncol* 31(3):1773–1782.

56 Webb C, Chang YH, Pockaj BA, Gray RJ, Stucky CC and Wasif N (2021). Lymph node positivity and association with long-term survival for different histologies of appendiceal cancer. *J Surg Oncol* 124(1):88–96.

57 Xie X, Zhou Z, Song Y, Li W, Diao D, Dang C and Zhang H (2016). The Management and Prognostic Prediction of Adenocarcinoma of Appendix. *Sci Rep* 6:39027.

58 Taggart MW, Galbincea J, Mansfield PF, Fournier KF, Royal RE, Overman MJ, Rashid A and Abraham SC (2013). High-level microsatellite instability in appendiceal carcinomas. *Am J Surg Pathol* 37(8):1192–1200.

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59 Tokunaga R, Xiu J, Johnston C, Goldberg RM, Philip PA, Seeber A, Naseem M, Lo JH, Arai H, Battaglin F, Puccini A, Berger MD, Soni S, Zhang W, Hwang JJ, Shields AF, Marshall JL, Baba H, Korn WM and Lenz HJ (2019). Molecular Profiling of Appendiceal Adenocarcinoma and Comparison with Right-sided and Left-sided Colorectal Cancer. *Clin Cancer Res* 25(10):3096–3103.

60 Misdraji J, Burgart LJ and Lauwers GY (2004). Defective mismatch repair in the pathogenesis of low-grade appendiceal mucinous neoplasms and adenocarcinomas. *Mod Pathol* 17(12):1447–1454.

61 Zhu X, Salhab M, Tomaszewicz K, Meng X, Mathew C, Bathini V, Switzer B, Walter O, Cosar EF, Wang X, Lambert LA and Hutchinson LM (2019). Heterogeneous mutational profile and prognosis conferred by TP53 mutations in appendiceal mucinous neoplasms. *Hum Pathol* 85:260–269.

62 Wald AI, Pingpank JF, Ongchin M, Hall LB, Jones H, Altpeter S, Liebdzinski M, Hamed AB, Derby J, Nikiforova MN, Bell PD, Paniccia A, Zureikat AH, Gorantla VC, Rhee JC, Thomas R, Bartlett DL, Smith K, Henn P, Theisen BK, Shyu S, Shalaby A, Choudry MHA and Singhi AD (2023). Targeted Next-Generation Sequencing Improves the Prognostication of Patients with Disseminated Appendiceal Mucinous Neoplasms (Pseudomyxoma Peritonei). *Ann Surg Oncol* 30(12):7517–7526.

63 Gibson J, Pengelly RJ, Mirandari A, Boukas K, Stanford S, Cecil TD, Mohamed F, Dayal SP, Tzivanakis A, Moran BJ, Mirnezami A, Carr NJ and Ennis S (2024). Targeted Genetic Sequencing Analysis of 223 Cases of Pseudomyxoma Peritonei Treated by Cytoreductive Surgery and Hyperthermic Intraperitoneal Chemotherapy Shows Survival Related to GNAS and KRAS Status. *Cancer Med* 13(20):e70340.

64 Milione M, Maisonneuve P, Spada F, Pellegrinelli A, Spaggiari P, Albarello L, Pisa E, Barberis M, Vanoli A, Buzzoni R, Pusceddu S, Concas L, Sessa F, Solcia E, Capella C, Fazio N and La Rosa S (2017). The Clinicopathologic Heterogeneity of Grade 3 Gastroenteropancreatic Neuroendocrine Neoplasms: Morphological Differentiation and Proliferation Identify Different Prognostic Categories. *Neuroendocrinology* 104(1):85–93.

65 Venizelos A, Elvebakken H, Perren A, Nikolaienko O, Deng W, Lothe IMB, Couvelard A, Hjortland GO, Sundlöv A, Svensson J, Garresori H, Kersten C, Hofsli E, Detlefsen S, Krogh M, Sorbye H and Knappskog S (2021). The molecular characteristics of high-grade gastroenteropancreatic neuroendocrine neoplasms. *Endocr Relat Cancer* 29(1):1–14.

66 Sciarra A, Libera L, Destro A, Maragliano R, Furlan D, Guastafierro V, Marchiori D, Vanoli A, Gonzalez RS, Letovanec I, Lazzi S, La Rosa S and Uccella S (2026). Molecular Profiling of Digestive Grade 3 Neuroendocrine Tumors Reveals a Shared Molecular Framework with Lower-Grade Tumors, Marked Heterogeneity, and Therapeutic Opportunities. *Endocr Pathol* 37(1):23.

67 Chen R, Tihy M, Lacroix L, de Rycke O, Lacombe C, Just PA, Cazes A, Svrcek M, de Montpreville VT, Antoine M, Scoazec JY, de Mestier L, Couvelard A and Cros J (2025). Performance of Immunohistochemistry to Detect Rb/p53 Alterations in High-Grade Neuroendocrine Neoplasms: Staining Patterns and Comparison with Genetic Testing. *Endocr Pathol* 36(1):40.

68 Baratti D, Kusamura S, Milione M, Bruno F, Guaglio M and Deraco M (2018). Validation of the Recent PSOGI Pathological Classification of Pseudomyxoma Peritonei in a Single-Center Series of 265 Patients Treated by Cytoreductive Surgery and Hyperthermic Intraperitoneal Chemotherapy. *Ann Surg Oncol* 25(2):404–413.

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69 Sugarbaker PH, Liang JJ, Deng T and Chang D (2024). Cellularity of mucus is a prognostic variable in low-grade appendiceal mucinous neoplasms. *Eur J Surg Oncol* 50(7):108373.

70 Roxburgh CS, Fenig YM, Cercek A, Shia J, Rassam RM, Paty PB and Nash GM (2019). Outcomes of Low-Grade Appendiceal Mucinous Neoplasms with Remote Acellular Mucinous Peritoneal Deposits. *Ann Surg Oncol* 26(1):118–124.

71 Shetty S, Natarajan B, Thomas P, Govindarajan V, Sharma P and Loggie B (2013). Proposed classification of pseudomyxoma peritonei: influence of signet ring cells on survival. *Am Surg* 79(11):1171–1176.

72 Bradley RF, Stewart JHt, Russell GB, Levine EA and Geisinger KR (2006). Pseudomyxoma peritonei of appendiceal origin: a clinicopathologic analysis of 101 patients uniformly treated at a single institution, with literature review. *Am J Surg Pathol* 30(5):551–559.

73 Davison JM, Choudry HA, Pingpank JF, Ahrendt SA, Holtzman MP, Zureikat AH, Zeh HJ, Ramalingam L, Zhu B, Nikiforova M, Bartlett DL and Pai RK (2014). Clinicopathologic and molecular analysis of disseminated appendiceal mucinous neoplasms: identification of factors predicting survival and proposed criteria for a three-tiered assessment of tumor grade. *Mod Pathol* 27(11):1521–1539.

74 Ronnett BM, Yan H, Kurman RJ, Shmookler BM, Wu L and Sugarbaker PH (2001). Patients with pseudomyxoma peritonei associated with disseminated peritoneal adenomucinosis have a significantly more favorable prognosis than patients with peritoneal mucinous carcinomatosis. *Cancer* 92(1):85–91.

75 Ronnett BM, Zahn CM, Kurman RJ, Kass ME, Sugarbaker PH and Shmookler BM (1995). Disseminated peritoneal adenomucinosis and peritoneal mucinous carcinomatosis. A clinicopathologic analysis of 109 cases with emphasis on distinguishing pathologic features, site of origin, prognosis, and relationship to "pseudomyxoma peritonei". *Am J Surg Pathol* 19(12):1390–1408.

76 Memon AA, Godbole C, Cecil T, Dayal S, Moran B, Tzivanakis A, Mohamed F and Carr NJ (2022). Overall Survival is More Closely Associated with Peritoneal than Primary Appendiceal Pathological Grade in Pseudomyxoma Peritonei with Discordant Pathology. *Ann Surg Oncol* 29(4):2607–2613.

77 Rauwerdink P, Al-Toma D, Wassenaar ECE, Raicu MG, Laclé MM, Milne AN, Kuijpers KC, Huysentruyt CJR, Poelmann FB, van Ramshorst B, Elias SG, Kranenburg O, Borel Rinkes IHM, Witkamp AJ, Wiezer MJ, van Grevenstein HMU and Boerma D (2024). Reclassification of Appendiceal Mucinous Neoplasms and Associated Pseudomyxoma Peritonei According to the Peritoneal Surface Oncology Group International Consensus: Clinicopathological Reflections of a Two-Center Cohort Study. *Ann Surg Oncol* 31(13):8572–8584.

78 Brierley JD, Giuliani M, O'Sullivan B, Rous B and Van Eycken E (eds) (2025). *Union for International Cancer Control. TNM Classification of Malignant Tumours, 9th Edition*, Wiley, USA.

79 Overman MJ, Kakar S, Carr NJ, Hanna NN, Holowatyj AN, Gress D, Williams A, Goldberg RM (2022). *AJCC Cancer Staging System: Appendix: Version 9 of the AJCC Cancer Staging System*. Washington MK (ed). American College of Surgeons.

80 Onyemkpa C, Davis A, McLeod M and Oyasiji T (2019). Typical carcinoids, goblet cell carcinoids, mixed adenoneuroendocrine carcinomas, neuroendocrine carcinomas and adenocarcinomas of the appendix: a comparative analysis of survival profile and predictors. *J Gastrointest Oncol* 10(2):300–306.

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