



Guidelines

German S2k-guideline on diagnostics, treatment and surveillance of low-grade appendiceal mucinous neoplasms (LAMN)

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ABSTRACT

The German guideline for low-grade appendiceal mucinous neoplasms (LAMN) and pseudomyxoma peritonei (PMP) offers comprehensive recommendations for diagnosis, treatment, and surveillance of these rare tumours. Developed by the German Society of General and Visceral Surgery (DGAV) alongside 14 other medical societies or task groups, this S2k-guideline addresses the need for standardised care in the absence of high-quality randomized controlled trials due to the rarity of LAMN and PMP. The guideline covers classification and staging of LAMN according to WHO and TNM systems, emphasising histological analysis and surgical protocols aimed at preventing intra-abdominal perforation. Diagnostic recommendations include imaging (MRI or CT) and preoperative tumour marker assessment, along with screening colonoscopies to rule out synchronous colorectal malignancies in specific age groups. Therapeutic guidelines focus on the importance of treatment in specialised centres with expertise in cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC). CRS combined with HIPEC is recommended for patients with PMP, with an emphasis on multidisciplinary team involvement and psycho-oncological support. The guideline outlines post-treatment surveillance, recommending six-monthly imaging and tumour marker evaluations for five years. It highlights the importance of considering fertility preservation in patients undergoing cytoreductive surgery and HIPEC. This consensus-based guideline

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aims to enhance the quality and consistency of care for patients with LAMN and PMP, offering a structured approach despite limited clinical trial data.

1. Introduction

Low-grade appendiceal mucinous neoplasms (LAMN) are rare tumours arising in the appendix, characterised by a unique spread pattern that leads to mucinous deposits in the peritoneal cavity, known as pseudomyxoma peritonei (PMP). The risk factors for LAMN remain unidentified, but advanced-stage LAMN is known to cause PMP more frequently. Treatment is challenging due to the rarity of these tumours and the lack of high-quality studies. Over an eight-year period, only 612 patients were included in the German Cancer Registry (ZfKD) [1], highlighting the rarity of LAMN. However, most general surgeons will likely encounter LAMN and/or PMP at some point in their careers, often incidentally.

Guidelines provide up-to-date and easily accessible information for surgeons, oncologists, general practitioners, and other healthcare professionals as well as patients. In Germany, the "Association of the Scientific Medical Societies" (AWMF) has created a platform for medical societies to publish guidelines according to specific regulations and standards. Depending on the level of evidence and systematic methodology used, guidelines are classified into four categories:

S1: Recommendations made by an expert group. Most S1-guidelines are developed by a single medical society, with a low level of evidence and no structured development process.

S2k: Consensus-based guidelines. A guideline committee, including representatives from all relevant medical societies and patient representatives, reaches a formal consensus on all recommendations, which are graded according to specific standards.

S2e: Evidence-based guidelines. Patients are involved in the guideline development process, and recommendations are based on systematic literature reviews and standardised evaluation methods.

S3: Evidence- and consensus-based guidelines. These involve patient participation, systematic literature reviews, and the use of structured consensus-building methods such as the Delphi process.

To develop evidence-based guidelines, the majority of supporting literature should be of high quality, such as randomized controlled trials (RCTs) [2]. However, due to the rarity of LAMN and PMP, very few RCTs have been published. As a result, the decision was made to develop an S2k-guideline.

2. Development of the guideline

The need for a guideline on LAMN and PMP was proposed by members of a peritoneal malignancy center to the *German Society of General and Visceral Surgery* (DGAV). The DGAV acknowledged the necessity of developing a guideline on LAMN and subsequent PMP and agreed to serve as the leading medical society for its development. In addition to the two initiators of the guideline, four further representatives were designated.

Guideline coordinators approached medical societies and invited them to participate in the guideline's development. During the first meeting, all representatives had the opportunity to propose additional medical societies or task groups that could provide valuable contributions. As a result, 14 medical societies or task groups, represented by 18 designated members, participated in the guideline's development. A list of participating medical societies and their representatives is provided in [Table 1](#).

The timeline, voting rights, and other organisational matters were addressed in the kick-off meeting.

Although systematic literature research is not mandatory for an S2k-guideline, the authors aimed to incorporate as much evidence-based information as possible. Therefore, literature searches were conducted

using the PubMed, Embase, and Cochrane databases, and the findings were shared among all representatives. The result of the literature search is provided in the appendix end-note file. Based on their areas of expertise, representatives worked on different sections of the guideline. After reviewing the literature, recommendations were formulated.

Each recommendation was then subjected to an anonymous vote, where representatives could either approve it or propose amendments. Recommendations that received over 95 % approval did not require further discussion in the final meeting. Those with less than 95 % approval were discussed under neutral moderation by an AWMF representative.

Given the number of experts involved, a single vote against a recommendation resulted in its rejection and the need for revision. Through this process, amendments were made, and all recommendations ultimately achieved more than 95 % approval. The acceptance rates were established during the initial kick-off meeting.

As a result:

- Six recommendations were accepted in the first vote.
- Eighteen recommendations were accepted after modifications and a second vote in the final meeting.
- Three new recommendations were introduced, discussed, and voted on following additional comments and deliberation.

A detailed table outlining the recommendations and the primary and secondary voting results can be found in the [supplements \(Tables 3 and 4\)](#).

In total, 28 recommendations were developed, covering pathological classification, diagnostics, treatment of LAMN and PMP, and post-

Table 1

Representatives of the medical societies or task groups of same.

Representative	Medical Society / Task Group
Prof. Dr. Dirk Arnold	Association Medical Oncology (AIO) – Task group of the German Cancer Society (DKG)
Prof. Dr. Daniela E. Aust	Oncological Pathology – Task group of the German Cancer Society (DKG)
PD Dr. Johannes Betge	German Society for Gastroenterology, Digestive and Metabolic Diseases
PD Dr. Ines Brecht	Society for Paediatric Oncology / Haematology
Prof. Dr. Christoph-Thomas Germer	German Society for General and Visceral surgery (DGAV)
Herr Bernd Grouven	Patients Representative Support Group Peritoneal Cancer
Prof. Dr. Philipp Harter	German Society of Gynaecology and Obstetrics (DGOG)
Prof. Dr. Stefan Kasper-Virchow	Association Medical Oncology (AIO) – Task group of the German Cancer Society (DKG)
Herr Kai Koslowski	Patients Representative
Dr. Franziska Köhler	German Society for General and Visceral surgery (DGAV)
Prof. Dr. Hannes Philipp Neeff	German Society for General and Visceral surgery (DGAV)
Prof. Dr. Jens Neumann	German Society for Pathology Association of German Pathologists
Prof. Dr. Dr. Pompiliu Piso	German Society for General and Visceral surgery (DGAV)
Prof. Dr. Beate Rau	Oncological Rehabilitation and Social Medicine – Task group of the German Cancer Society (DKG)
Prof. Dr. Dominik Schneider	Society for Paediatric Oncology / Haematology
Prof. Dr. Andreas G. Schreyer	German Radiological Society (DRG)
Dr. Maria Kröplin	German Society for General and Visceral surgery (DGAV)
Prof. Dr. Armin Wiegering	Association Surgical Oncology (ACO) – Task group of the German Cancer Society (DKG)

treatment surveillance. The guideline was then presented to the steering committees of all participating medical societies and received their approval. Subsequently, it was published on the AWMF website in February 2024, along with a concise version in both German and English. As per policy, the guideline remains valid for five years.

3. Pathological classification

Recommendation 1. : Mucinous neoplasms of the appendix should be classified according to the current WHO classification.

The World Health Organization (WHO) provides specific criteria for the safe diagnosis of LAMN and its differentiation from high-grade appendiceal mucinous neoplasms (HAMN) and mucinous adenocarcinoma. These criteria include filiform or villous mucinous epithelium with tall cytoplasmic mucin vacuoles, varying degrees of extracellular mucin, fibrosis, hyalinization, and calcification of the appendiceal wall, as well as broad pushing margins. Furthermore, the primary tumour and peritoneal metastases should be classified individually according to the three-tier classification [3].

Recommendation 2. *Low-grade appendiceal mucinous neoplasms should be classified according to the current TNM-classification.*

TNM classification for LAMN differs significantly from that of other tumours, as shown in Table 2. While pTis refers to tumours confined to the appendix, there are no pT1 or pT2 classifications for LAMN. pT3 describes infiltration of the subserosal tissue, while pT4 includes tumour perforation and mucinous deposits within the right lower quadrant. Metastatic disease is defined by tumour deposits beyond the right lower quadrant.

Recommendation 3. : *Histological specimens of low-grade appendiceal mucinous neoplasms should be representative and the appendiceal wall should be captured orthograde.*

Recommendation 4. : *If pseudomyxoma peritonei is present, representative embedding should be performed. The following can be used as a guide: 1 block/cm of altered tissue.*

These recommendations were included in the guideline to ensure adequate diagnosis of both primary and metastatic disease.

Recommendation 5. : *Currently, mutation analysis has no role in prognostic assessment and prediction of low-grade appendiceal mucinous neoplasms and pseudomyxoma peritonei. Mutation analysis can be used for the differential diagnosis of reactive to low-grade findings and in selected patients with unresectable pseudomyxoma peritonei, for the evaluation of individual tumour therapy in accordance with the recommendations of a molecular multidisciplinary meeting.*

While molecular analysis plays a crucial role in other tumours, where treatment options are guided by specific mutations (e.g., microsatellite

stability (MSS) in colorectal cancer [4]), no targeted therapy has been established for LAMN and PMP to date. However, frequent KRAS and GNAS mutations observed in LAMN and PMP [5] may offer therapeutic potential in the future.

4. Diagnostics

Recommendation 6. *If a low-grade appendiceal mucinous neoplasms/pseudomyxoma peritonei is suspected preoperatively, e.g. by ultrasound findings, contrast-enhanced MRI abdomen-pelvis with diffusion-weighted imaging (especially in children and adolescents) or alternatively contrast-enhanced CT abdomen-pelvis should be performed.*

While LAMN and subsequent PMP are pathological diagnoses, lesions of the appendix are often detected incidentally during ultrasound. To further investigate and rule out PMP, additional radiological imaging is frequently performed. Both CT and MRI are suitable for detecting PMP, but MRI appears to be more sensitive. Moreover, MRI does not involve radiation, making it the preferred imaging modality, particularly for children and adolescents [6].

Recommendation 7. : *If a low-grade appendiceal mucinous neoplasm without evidence of pseudomyxoma peritonei is diagnosed postoperatively and adequate treatment has already been provided by surgery (see chapter on Therapy), no immediate staging including radiological imaging is indicated; this is performed later as part of surveillance (see chapter on Surveillance). To provide better comparability during further surveillance, immediate radiological imaging can be performed.*

Small tumours can easily be missed during an appendicectomy. In these cases, if PMP was not present intraoperatively, no further treatment is required, and surveillance should be initiated. When preoperative focused ultrasound imaging has been performed, additional postoperative imaging (MRI/CT) may be beneficial for improved comparability in future assessments.

Recommendation 8. *If a low-grade appendiceal mucinous neoplasm / pseudomyxoma peritonei is suspected preoperatively, the tumour markers CEA, CA19-9 and CA 125 ought to be determined in the preoperative work-up to differentiate from other neoplasms.*

While tumour markers cannot confirm the diagnosis of LAMN and subsequent PMP, they serve as a useful tool for surveillance. Therefore, preoperative tumour marker assessment can be beneficial. Additionally, tumour markers can aid in differentiating LAMN from other malignancies, such as ovarian tumours [7].

Recommendation 9. *Screening colonoscopy to rule out synchronous colorectal neoplasms should be performed according to the recommendations of the S3 guideline "colorectal carcinoma" in patients with low-grade appendiceal mucinous neoplasms and pseudomyxoma peritonei.*

Bowel screening programs play a crucial role in preventing and

Table 2
TNM-classification of LAMN and consecutive PMP.

	Definition
Tis	Tumour confined to the appendix with acellular mucin or mucinous epithelium confined to the muscularis propria
T1	Not applicable in LAMN
T2	Not applicable in LAMN
T3	Tumour or acellular mucin extends onto the subserosal soft tissue
T4	Tumour extends to the serosa of the appendix or mesoappendix including perforation of cellular or acellular mucin
T4a:	Tumour perforation including cellular or acellular mucin within the right lower quadrant
T4b:	Tumour invades other organs or structures
N-Status	n/a
M0	No metastatic disease
M1	Metastatic disease
M1a:	Intraperitoneal acellular mucin
M1b:	Intraperitoneal cellular mucin
M1c:	Metastases beyond the abdominal cavity

detecting colorectal cancer at an early stage [4]. While appendiceal tumours may occasionally be visualised during colonoscopy, they are often not detected. Before performing surgery in LAMN/PMP patients, synchronous colorectal malignancies should be ruled out through colonoscopy. The current German guideline recommends initiating screening at age 50 for asymptomatic patients [4]. There is no evidence supporting more frequent colonoscopies or screening at a younger age.

Recommendation 10. *Instead of a surgical biopsy for histological confirmation of low-grade appendiceal mucinous neoplasms / pseudomyxoma peritonei, treatment **should** be performed in the first instance (see chapter on Therapy).*

If LAMN is suspected, surgery (e.g., appendectomy) should be performed rather than a biopsy, which carries the risk of tumour perforation. In cases of LAMN with PMP, treatment (e.g., CRS and HIPEC) should be initiated directly instead of performing a biopsy first.

5. Therapy

Recommendation 11. *All patients diagnosed with low-grade appendiceal mucinous neoplasms / pseudomyxoma peritonei **should** be discussed in a multidisciplinary team meeting, who are experienced in the management of these conditions. When imaging suggests low-grade appendiceal mucinous neoplasms / pseudomyxoma, multidisciplinary discussion **should** be done prior to any treatment.*

Discussing cases in multidisciplinary team meetings and making treatment plans within these meetings has been shown to improve the quality of care and overall survival across various tumours, including colorectal cancer [8]. A similar benefit can be expected for rare tumours like LAMN and PMP.

Recommendation 12. *Patients diagnosed with low-grade appendiceal mucinous neoplasms / pseudomyxoma peritonei **ought** to be referred to / treated in hospitals with expertise in the treatment of same. The following criteria appear reasonable to prove expertise: multidisciplinary discussion of at least 50 patients with malignant diseases of the peritoneum per year, and frequent cytoreductive surgeries and HIPEC procedures (25/year). Expertise **can** be demonstrated through publicly accredited certification.*

Retrospective studies have shown that highly specialised procedures like CRS and HIPEC have lower mortality and morbidity rates when performed by experienced surgeons. Additionally, higher rates of complete cytoreduction, measured by the completeness of cytoreduction score (CC-Score), can be achieved [9–13]. In Germany, accreditation by the German Society of General and Visceral Surgery serves as proof of expertise [14].

Recommendation 13. *Patients with low-grade appendiceal mucinous neoplasms / pseudomyxoma peritonei **should** be offered psycho-oncological care.*

A diagnosis of a malignant disease presents challenges in various aspects of a patient's life, and psycho-oncological care can provide valuable support in managing these difficulties. The German S3-guideline further elaborates on this topic [15].

Recommendation 14. *If low-grade appendiceal mucinous neoplasm/pseudomyxoma peritonei is suspected on imaging, surgical exploration and resection **should** be performed at a hospital with sufficient expertise (see Recommendation 12) after preoperative diagnostics. In individual cases, postponing the surgery **can** be considered for specific circumstances.*

LAMNs are known to be non-invasive and slow-growing tumours, but CRS and HIPEC remain the only curative treatment, significantly prolonging overall survival. Therefore, CRS and HIPEC should be performed in a timely manner. However, in some cases, immediate surgery is not feasible (e.g., during pregnancy or in the presence of a synchronous malignancy with higher mortality risk). In these patients, surgery can be

postponed.

Recommendation 15. *Patients with low-grade appendiceal mucinous neoplasms with pTis or pT3 tumours, without evidence of extra-appendicular mucin (M0), corresponding to a maximum UICC stage IIa and complete resection with an appendectomy (R0 status), are fully treated with an appendectomy.*

Localised LAMNs without evidence of PMP have a very low risk of recurrence. Most studies report a 0 % recurrence risk for pTis or pT3 tumours. Therefore, more extensive surgery beyond appendectomy is not justified. [16–18].

Recommendation 16. *Patients with low-grade appendiceal mucinous neoplasms pT4a und pT4b without evidence of extra-appendicular mucin beyond the right lower quadrant, and complete resection (R0 status) do not require a right hemicolectomy, omentectomy, parietal peritonectomy, or systemic treatment. These patients **should** undergo surveillance. If mucin is present on the appendiceal surface or in the right lower quadrant (cellular mucin or cellularity not determined), HIPEC and local cytoreductive surgery **can** be considered. The treatment decision is individual and needs to be discussed with the patient.*

Overall, the risk of synchronous PMP is higher in patients with advanced LAMN (pT4) compared to those with locally confined tumours [19]. If PMP is not present at diagnosis, the risk of recurrence appears to be low. LAMN does not metastasise via the lymphatic system or bloodstream; therefore, oncological resections such as right hemicolectomy and lymphadenectomy are not justified. HIPEC and local CRS may be considered as individualised treatments for patients with mucin on the appendiceal surface but should not be performed routinely."

Recommendation 17. *If a low-grade appendiceal mucinous neoplasm is suspected intraoperatively while performed an exploratory laparoscopy (i.e. for suspected appendicitis or ovarian pathology), the main goal **should** be to prevent intra-abdominal perforation of the neoplasm. If appendectomy cannot be performed laparoscopically in a safe manner, conversion to open surgery **should** be performed. Furthermore, the whole abdominal cavity **should** be inspected.*

Preventing intra-abdominal perforation of LAMN is crucial to avoid mucin contamination in the abdominal cavity. In patients with small tumours, laparoscopic appendectomy can be performed safely without perforation. However, in cases with dilated or enlarged appendices, this poses a greater challenge. Thorough exploration of the abdominal cavity should always be conducted to rule out PMP.

Recommendation 18. *In patients with pseudomyxoma peritonei due to low-grade appendiceal mucinous neoplasms, complete cytoreductive surgery and HIPEC **should** be performed in hospitals with expertise in the treatment of these patients (see Recommendation 12). If cytoreduction without HIPEC was performed without prior knowledge of the diagnosis of pseudomyxoma peritonei secondary to low-grade appendiceal mucinous neoplasm, patients **should** be referred to a hospital with expertise (see Recommendation 12) for individual evaluation of HIPEC.*

Morbidity and mortality are significantly lower when CRS or oncological surgery is performed by experienced surgeons [20]. Additionally, higher rates of complete cytoreduction can be achieved, leading to improved overall survival [9–13].

Recommendation 19. *In the case of M1a-situation and completely resected mucin, as well as R0-resection at the appendix base, cytoreductive surgery and HIPEC **can** be renounced.*

When LAMN and PMP are incidentally discovered during surgery for acute appendicitis, CRS and HIPEC are often not readily available. In such cases, the primary focus should be on removing all visible mucin along with the primary tumour. If no residual mucin remains in the abdominal cavity, further CRS and HIPEC may not be necessary. However, ensuring that all visible tumour deposits (mucin) have been

completely removed is crucial in making this decision.

Recommendation 20. *If mucin is present intraoperatively, a representative sample for histopathological evaluation **should** be taken.*

The risk of recurrence largely depends on the cellularity of intra-abdominal mucin. To accurately assess this risk, mucin samples must be examined.

Recommendation 21. *In symptomatic patients with irresectable pseudomyxoma peritonei (obstructive symptoms), tumour debulking **can** be considered. In these cases of irresectable disease, HIPEC has not been proven to prolong overall survival or symptom-free survival in the literature.*

The primary goal of curative treatment for PMP patients is to achieve complete cytoreduction. In cases of unresectable disease, surgery is generally not recommended. However, an exception exists for symptomatic patients who frequently experience obstructive symptoms, anorexia, respiratory difficulties, or abdominal pain. In these cases,

surgical options, including tumour debulking, may be considered. The use of HIPEC in cases where only incomplete cytoreduction is possible has not been proven to improve overall or disease-free survival.

Recommendation 22. *In patients with pseudomyxoma peritonei, the Sugarbaker peritoneal carcinomatosis index (PCI) **should** be calculated as well as the completeness of cytoreduction score (CC-score) after resection.*

To standardise and quantify the extent of peritoneal deposits, Paul Sugarbaker, a pioneer in peritoneal malignancy surgery, developed an index to accurately describe peritoneal disease. The abdomen is divided into 13 regions, and the extent of peritoneal involvement is then assessed [21] (see Figure 1).

Recommendation 23. *Patients with pseudomyxoma peritonei with involvement of the uterus and/or adnexal regions should be treated in cooperation with gynaecology. Besides oncological goals, endocrine function including preservation of fertility **should** be considered.*

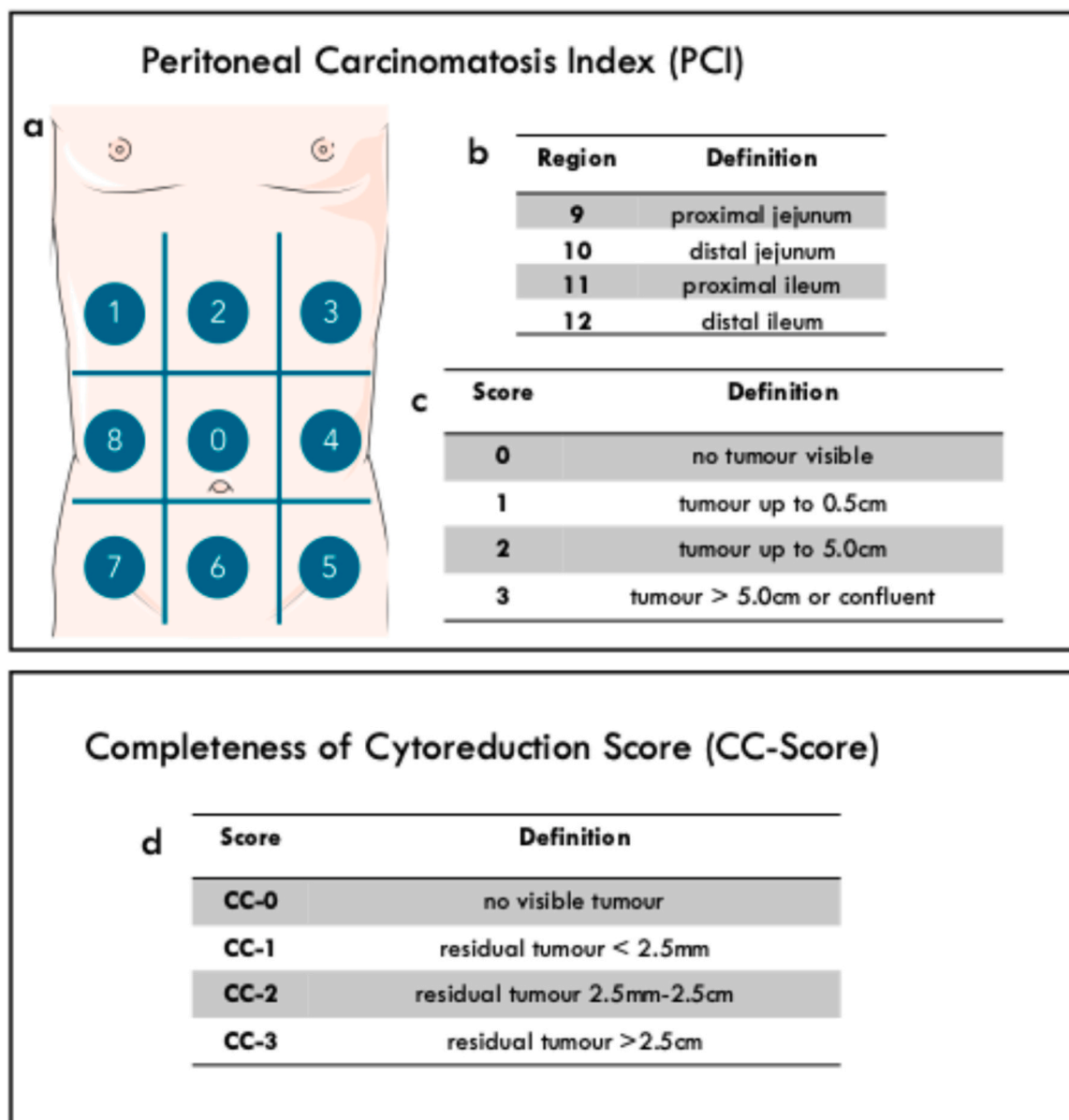


Fig. 1. Peritoneal Carcinomatosis Index (PCI) (a-c) and Completeness of Cytoreduction Score (CC-Score) (d) first introduced by Sugarbaker and Jacquet [21]: the abdominal cavity is divided into 12 regions: a - region 0–8 and b - region 9–12. Tumour burden is measured for each region individually (c) resulting in a maximum PCI of 39 which would describe confluent tumour on all peritoneal surfaces. Post CRS residual tumour burden is measured to determine the completeness of cytoreduction (d). CC-0 and CC-1 are frequently described as complete cytoreduction, whereas CC-2 and CC-3 are rather tumour debulking surgeries.

Recommendation 24. *If HIPEC is planned in patients without disease of the reproductive organs (regardless of gender), they **should** be offered fertility counselling prior to performing this procedure.*

Women are slightly more frequently affected by LAMN and PMP compared to men. While LAMN is typically treated with an appendectomy, PMP management often requires bowel resections and (bilateral) salpingo-oophorectomy and/or hysterectomy [22], which can lead to impaired fertility or infertility. Additionally, adhesions following CRS and the potential gonadotoxic effects of HIPEC are further reasons why patients should seek counselling from a gynaecologic oncology specialist prior to treatment.

The S2k-guideline on *Fertility Counselling in Oncological Patients* does not specifically address HIPEC but provides general recommendations [23]. Given that LAMN and PMP are slow-growing tumours, fertility preservation measures - such as egg or embryo freezing - should be considered.

Finally, treatment priorities should be discussed before CRS and HIPEC. In some patients, the desire to have children may take precedence over complete tumour removal. These personal preferences should be acknowledged and respected.

6. Surveillance

Recommendation 25. *Surveillance of patients with low-grade appendiceal mucinous neoplasms or pseudomyxoma peritonei **ought** to include tumour markers CEA, CA 19-9 and CA 125.*

Recommendation 26. *Surveillance of patients with low-grade appendiceal mucinous neoplasms **ought** to include radiological imaging. If there is no contraindication, a contrast enhanced MRI of the abdomen and pelvis including diffusion-weighted imaging **ought** to be performed. Alternatively, a contrast-enhanced CT of the abdomen and pelvis **can** be performed.*

Recommendation 27. *In the follow-up of patients with low-grade appendiceal mucinous neoplasms/ pseudomyxoma peritonei, the importance of screening colonoscopies for early detection of colorectal carcinoma **should** be highlighted. In asymptomatic patients who are followed up post low-grade appendiceal mucinous neoplasm/pseudomyxoma peritonei, there is no evidence to support screening colonoscopies before the age of 50.*

There is no validated surveillance regimen following appendectomy for localised LAMN or CRS and HIPEC for PMP. Most studies recommend monitoring tumour markers (CEA, CA19-9, and CA 125) alongside cross-sectional imaging. As previously mentioned, MRI should be the preferred imaging modality, as it offers similar sensitivity to CT without radiation exposure.

Tumour markers are valuable indicators of recurrence and play a crucial role in surveillance. Since tumour recurrence is always extraluminal, endoscopic surveillance is not considered feasible. Given that most patients fall within the same age group as those at risk for colorectal cancer, screening colonoscopies should be recommended according to the S3-guideline for colorectal cancer [4].

Recommendation 28. *Surveillance for low-grade appendiceal mucinous neoplasms or pseudomyxoma peritonei **ought** to be performed for 5 years in six-monthly intervals. In individual cases, surveillance **can** be prolonged.*

In oncological surveillance protocols, a five-year duration is most commonly recommended. The experts on this guideline committee also agreed on a five-year surveillance regimen. However, given that LAMN and PMP are slow-growing conditions, and cases of PMP developing long after the primary diagnosis have been reported, extended surveillance may be warranted.

The addendum to recommendation 28 states that surveillance can be prolonged in select patients. To determine which individuals may benefit from extended monitoring, risk factors such as preoperatively

elevated tumour markers and the completeness of cytoreduction should be carefully evaluated.

7. Conclusion

Guidelines serve as valuable tool for providing information to a broad audience. The S2k-guideline “Diagnostics, Treatment and Surveillance of low-grade appendiceal mucinous neoplasms (LAMN)” represents the first German guideline on this topic and marks a significant advancement in improving the management of these rare conditions.

Managing rare tumours like LAMN and PMP presents challenges in treatment, accurate pathological evaluation, and appropriate follow-up. A lack of knowledge in any of these areas can lead to either overtreatment - such as performing an unnecessary oncological right hemicolectomy for a patient with non-perforated LAMN - or undertreatment, where inadequate surveillance results in PMP being diagnosed at an advanced, potentially irresectable stage.

Internationally, the Peritoneal Surface Oncology Group International (PSOGI) published a guideline in 2020 on the treatment of appendiceal tumours and PMP, including high-grade tumours and adenocarcinomas. Unlike the S2k-guideline, which aims for broad consensus, the PSOGI guideline presents a range of expert opinions. However, despite their differing approaches, both guidelines align in their core recommendations, highlighting a strong consensus among leading experts in the field [24].

Some recommendations in this guideline - such as providing psycho-oncological support for LAMN and PMP patients or offering fertility counselling before CRS and HIPEC - may seem intuitive. However, they are often overlooked in treatment planning, despite their significant impact on quality of life. Specialised centers that regularly manage these conditions typically adopt a holistic approach, ensuring that patients receive not only evidence-based treatment but also comprehensive supportive care.

The German S2k-guideline for LAMN and PMP addresses a critical gap in the management of these complex conditions. Developed through a collaborative, expert-driven process, it provides practical, consensus-based recommendations in the absence of extensive randomized controlled trials. The emphasis on specialised care, individualised treatment strategies, and structured surveillance ensures a comprehensive approach to improving patient outcomes.

Looking ahead, advancements in molecular profiling and targeted therapies may further refine these guidelines. For now, however, this framework serves as an essential resource for healthcare providers in Germany and beyond, standardising care and improving the management of LAMN and PMP.

CRedit authorship contribution statement

Kröplin Maria A.: Writing – review & editing, Validation, Investigation, Formal analysis, Data curation. **Wiegner Armin:** Writing – review & editing, Writing – original draft, Validation, Supervision, Software, Project administration, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Aust Daniela:** Validation, Investigation, Formal analysis, Data curation. **Betge Johannes:** Writing – review & editing, Validation, Investigation, Formal analysis, Data curation. **Arnold Dirk:** Validation, Investigation, Formal analysis, Data curation. **Grouven Bernd:** Writing – review & editing, Validation, Investigation, Formal analysis, Data curation. **Harter Philipp:** Writing – review & editing, Validation, Investigation, Formal analysis, Data curation. **Brecht Ines B.:** Writing – review & editing, Validation, Investigation, Formal analysis, Data curation. **Germer Christoph-Thomas:** Writing – review & editing, Validation, Investigation, Formal analysis, Data curation. **Neeff Hannes Philipp:** Writing – review & editing, Validation, Investigation, Formal analysis, Data curation. **Köhler Franziska:** Writing – review & editing, Writing – original draft, Visualization, Validation, Project administration, Methodology,

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Declaration of Competing Interest

All Authors declare no conflict of interest.

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Appendix A. Supporting information

Supplementary data associated with this article can be found in the online version at [doi:10.1016/j.ejca.2025.115430](https://doi.org/10.1016/j.ejca.2025.115430).

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