

1



Find a Specialist

It is highly recommended that appendix cancer and PMP patients seek out a specialist who deals with these conditions on a regular basis before commencing treatment.

Check out acpmp.org/find-a-specialist to find one near you.

2



Educate Yourself

Learn about appendix cancer and PMP, particularly about diagnosis, treatment and terminology. Check out videos from our educational events to learn more about these diseases and optimal treatment.

Go to acpmp.org/helpful-links to learn more.

3



Participate in the ACPMP Patient Registry

ACPMP's Patient Registry serves as a platform for patients around the world to share information about appendix cancer and PMP in order to build an international resource to be used by scientists in future research. Visit acpmpregistry.iamrare.org

4



Explore the Clinical Trial Database

Review ACPMP's list of active clinical trials and discuss with your physician. Ask your physician about genetic & molecular profiling of your tumor to guide treatment : acpmp.org/clinical-trials.

5



Receive Updates & Connect with Others

Follow ACPMP on social media and receive regular updates from ACPMP by joining our mailing list at acpmp.org/email-signup. Scan the QR code for all resources and ways to connect with other patients and caregivers.

The ACPMP Research Foundation is an IRS-designated 501(c)3 charitable organization (EIN # 26-2890160)

UNDERSTANDING APPENDIX CANCER & PSEUDOMYXOMA PERITONEI

Overview, Symptoms, Diagnosis, and Treatment

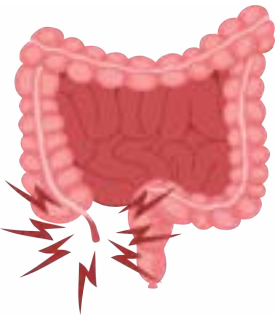
Overview

Appendix cancer starts with a tumor in the cells of the appendix.

There are many different types of tumors:

1. Low-grade mucinous neoplasm of the appendix (LAMN),
2. High-grade mucinous neoplasm of the appendix (HAMN),
3. Goblet cell carcinoid (defined by a unique combination of two types of cancer cells – neuroendocrine [carcinoid] and epithelial [adenocarcinoma]), adenocarcinoma, further classified as:
 - a. well-differentiated,
 - b. moderately-differentiated,
 - c. poorly-differentiated and
 - d. signet ring cell (SRC).

Appendiceal tumors frequently spread inside the abdominal cavity. Depending on the type of tumor, this can lead to a condition called peritoneal carcinomatosis or peritoneal surface malignancy.



Pseudomyxoma peritonei (PMP) is the progressive accumulation of mucus-secreting or mucinous tumor cells within the abdomen and pelvis after an appendiceal tumor bursts through the wall and spreads mucinous cells throughout the surrounding surfaces. As mucinous tumor cells accumulate, the abdominal area becomes swollen and digestive function becomes impaired.

In very rare cases, pseudomyxoma peritonei can arise from tumors located in organs other than the appendix, but the vast majority arise from appendiceal tumors.

Incidence, Symptoms & Diagnosis

Incidence

Cancers and tumors of the appendix are extremely rare with an estimated incidence of 2-9 per 1,000,000 people. The average age of onset is between 50 and 55 years, and they affect men and women equally.

Symptoms

Appendix cancer and PMP are often misdiagnosed because in many cases, relatively common symptoms such as abdominal pain, increased abdominal girth, bloating, hernia, ascites, and ovarian cysts or tumors or infertility in women are misunderstood for years. Many women are initially misdiagnosed with ovarian cancer.

Diagnosis

For many patients, appendix cancer and/or PMP are discovered during an unrelated surgical procedure. The following diagnostic tests are also commonly used in diagnosing appendix cancer and PMP:

- CT scan of the chest, abdomen and pelvis.
- MRI of the abdomen and pelvis (special protocols and expertise at reading MRIs are required to get the maximal benefit of this technology).
- Diagnostic laparoscopy.
- Blood tests (tumor markers). Some tumor markers in the blood have been shown to be indicative of appendix cancer and/or PMP activity in some (but not all) patients, particularly CEA, CA-125, & CA 19-9.

Treatment

Treatment varies depending on the stage of disease and the subtype, and whether the cancer is localized to the appendix or has spread. For LAMN that have not spread in the abdomen, an appendectomy is all that is required. For moderately- differentiated, poorly-differentiated and signet ring cell appendiceal cancers, surgical removal of the right side of the colon (right hemicolectomy) is recommended to ensure that all the disease has been removed and to test the regional lymph nodes for any cancer cells.



For tumors and cancers that have spread into the abdominal cavity or into the local lymph nodes, intravenous chemotherapy using existing colon cancer regimens and/or additional surgery to remove the cancer is recommended, in particular cytoreductive surgery (CRS) accompanied by heated intraperitoneal chemotherapy (HIPEC), delivered directly into the abdomen.

Cytoreductive Surgery (CRS) & Heated Intraperitoneal Chemotherapy (HIPEC)

CRS/HIPEC is the standard of care for peritoneal surface malignancies of appendiceal origin. This procedure should be performed at an experienced HIPEC center. CRS/HIPEC has been shown to provide long-term survival in up to 90% of patients with low-grade pathology who are properly diagnosed and undergo this treatment early enough in the disease process to offer maximum benefit.

When patients are misdiagnosed, they are less likely to benefit from CRS/HIPEC, and even with proper diagnosis many patients are never even offered it. This treatment, unavailable to our patient population a generation ago, has turned what used to be an almost certain death sentence into hope for thousands of patients around the world.

Meet The Appendix Cancer PMP Research Foundation

MISSION & WHO WE ARE.

- The Appendix Cancer PMP Research Foundation is a 501(c)3 charitable organization whose core mission is to support and fund promising research and education that moves us toward a cure for appendix cancer and PMP.
- We also support the appendix cancer & PMP community by providing the latest, most accurate information.

Medical Advisory Board Members:

- Dr. Laura Lambert (Chair), Huntsman Cancer Institute
- Dr. Andrew Lowy, San Diego Moores Cancer Center
- Dr. Reetesh Pai, Pittsburgh School of Medicine/UPMC
- Dr. Armando Sardi, Mercy Medical Center
- Dr. John Paul Shen, Anderson Cancer Center
- Dr. Konstantinos Votanopoulos, Levine Cancer

Scientific Advisory Board Members:

- Dr. Michael Overman, Anderson Cancer Center
- Dr. Konstantinos Votanopoulos, Levine Cancer
- Dr. Patrick Wagner, Allegheny Health Network

WHAT WE DO.

- ACPMP has awarded **over 2 million dollars** in **research grants** and provides funding for physicians to present their research at relevant medical conferences.
- We provide, contingent upon qualifying submissions, at least **two \$50,000, 2-year Catalyst Grants each year**. In addition, in 2025 we launched a **2-year \$150,000 Changemaker Grant**. ACPMP looks forward to offering at least one Changemaker Grant in 2026.
- ACPMP partners with institutions treating appendix cancer and PMP to **host patient/caregivers & families and healthcare professionals focused educational webinars & symposiums each year**. These events give patients and physicians the opportunity to hear about the latest developments and research in the treatment of these diseases.
- **ACPMP plays a leading role in the dissemination of accurate information regarding appendix cancer & PMP** both to the patient and medical communities.
- Create initiatives and resources for patients, caregivers and healthcare professionals with information in managing and treating this disease.
- Created a **"Find a Specialist" Tool** which includes physicians in the US with experience in treating appendix cancer and PMP.
- Created a **Clinical Trial Repository** to help patients identify clinical trials of potential interest.

WHY WE DO IT.

- Prior to the formation of the ACPMP Research Foundation, there was no centralized organization dedicated to raising funds for appendix cancer and PMP research in an objective way.
- Private donations for appendix cancer research were primarily made to individual doctors rather than towards competitive, RFP-driven funding based on the quality of the science.
- From the patient/caregiver perspective, research often appeared to be redundant or theoretical with little coordination between researchers.
- Movement appeared slow to change clinical practice or improve curability.
- The rarity of appendix cancer and PMP translates into minimal government funded research.
- Appendix cancer and PMP patients continue to be misdiagnosed and/or not offered the optimum treatment for these diseases.

GET INVOLVED.

For more information, please visit our website and join our contact list using the QR codes below:

**Visit
our website
acpmp.org**



**Join the
ACPMP
mailing list**



(833) 227-6773 | info@acpmp.org

ACPMP RESEARCH FOUNDATION

APPENDIX CANCER/PMP PATIENT REGISTRY

The Appendix Cancer/PMP Patient Registry collects critical information to help build a better understanding of this rare cancer and drive new therapies forward.

“The ACPMP Patient Registry will provide a complete picture of each patient’s experience with appendix cancer,” said Deborah Shelton, Executive Director of the Appendix Cancer PMP (ACPMP) Research Foundation. “We are launching this initiative to help collect as much patient data as possible to for use by researchers and medical experts to advance research and one day, find a cure for all.”

The Appendix Cancer PMP (ACPMP) Research Foundation in partnership with the National Organization for Rare Disorders (NORD) has launched the ACPMP Research Foundation Appendix Cancer/Pseudomyxoma Peritonei Patient Registry (ACPMP Patient Registry) to provide a secure platform for patients and caregivers worldwide to share information about Appendix Cancer and Pseudomyxoma Peritonei (PMP).

By participating in this registry, you can contribute valuable data that will help scientists and researchers better understand these rare diseases. The registry collects information through electronic surveys about the patient experience and disease progression. All data submitted is anonymized and securely stored to protect your privacy.

The ACPMP Patient Registry is a natural history study designed to support future research, accelerate the development of new diagnostics and treatments, and empower the Appendix Cancer and PMP community through shared knowledge.

Participation is free, voluntary, and open to individuals with Appendix Cancer and/or PMP, as well as their caregivers or guardians. You may withdraw from the registry at any time.

We’re pleased to share that Dr. Erin Ward of the University of Utah Huntsman Cancer Institute has agreed to serve as Chief Medical Advisor for the ACPMP Patient Registry, continuing her valued collaboration on this critical initiative.



Scan the QR code or click the link to access the patient registry.

[ACPMP.ORG/PATIENTREGISTRY](https://www.acpmp.org/patientregistry)

