

Radiation Therapy Center

Of Morris Hospital



MORRIS
HOSPITAL
& HEALTHCARE CENTERS

Patient Services Newsletter

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Appendix cancer is cancer that starts in the cells lining the inside of the appendix. There are a number of different types of cancers of the appendix. The different types are determined by which type of cells in the appendix become cancerous and what the cells look like under the microscope. The different types are associated with different behaviors (i.e. likelihood of spreading to other organs or other parts of the body, rate of growth, ability to be completely removed with surgery etc.) and therefore the types of treatment that are offered. Appendix cancers are the most common cause of pseudomyxoma peritonei (or “PMP”)

How are Appendix Cancer and PMP diagnosed?

For many patients, appendix cancer and/or PMP are discovered during an unrelated surgical procedure at which time the surgeon observes the mucinous tumors or “studding” on the peritoneum or abdominal organs. The following diagnostic tests are also commonly used in diagnosing appendix cancer and PMP:

- *CT scan of the chest, abdomen and pelvis.* Note that CT scans are not perfect pictures, and for some patients visible signs of tumor are not present on a CT scan. CT scans also require the judgment of a radiologist who can interpret the scan to identify the signs of appendix cancer or PMP such as fluid buildup or tumor “studding”.
- *MRI of the abdomen and pelvis.* There is a growing interest in the use of MRI of the abdomen and pelvis to diagnose and follow appendix cancers and PMP. Special protocols and expertise at reading MRIs are required to get the maximal benefit of this technology. Like CT scans, MRIs are not perfect pictures and they both require tumors to be big enough to be seen on the images.
- *Diagnostic laparoscopy.* A thin tube with a camera at its tip is inserted through “keyholes” in the abdomen to observe the abdominal cavity. Again, proper diagnosis depends upon the surgeon performing the procedure. Laparoscopic diagnosis of appendix cancer and PMP presents additional challenges in that failure to follow appropriate protocols may result in the spreading or seeding of tumors. If appendix cancer and/or PMP are suspected, a specialist in the disease should be brought in to take the proper precautions.
- *Blood tests (tumor markers).* Some tumor markers in the blood have been shown to be indicative of appendix cancer and/or PMP activity, particularly CEA, CA-125, and CA 19-9. Unfortunately, tumor markers are not accurate or indicators for everyone. Some patients will have no elevation in their tumor markers despite having extensive, aggressive tumor.

APPENDIX CANCER AND PSEUDOMYXOMA PERITONEI

FACT SHEET



OVERVIEW OF APPENDIX CANCER & PSEUDOMYXOMA PERITONEI (PMP)

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]),
- (4) 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 0.15-0.9 per 100,000 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, and 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 SRC 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.



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Company Fruit Salad



Ingredients

- 4 medium Golden Delicious apples, diced
 - 4 medium Red Delicious apples, diced
 - 2 cups seedless green grapes, halved
 - 2 cups seedless red grapes, halved
 - 1 can (20 ounces) pineapple chunks, drained
 - 1 can (11 ounces) mandarin oranges, drained
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- **dressing:**

- 3 ounces cream cheese, softened
 - 1/2 cup sour cream
 - 1/2 cup mayonnaise
 - 1/2 cup sugar
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Directions

1. In a large bowl, combine the first 6 ingredients. In a small bowl, beat dressing ingredients until smooth. Pour over fruit; toss gently to coat.

Test Kitchen Tips

- Experiment with different fruit-and-nut flavor combinations in your fruit salads, such as peaches and pecans; apples and walnuts; cherries and almonds; or pineapple and macadamia nuts.
- When making a fruit salad, toss the juice from a small can of pineapple into the bowl. The juice keeps the other fruit from browning, and bits of pineapple in the salad give it a tropical zing.

Nutrition Facts

3/4 cup: 161 calories, 7g fat (2g saturated fat), 11mg cholesterol, 48mg sodium, 25g carbohydrate (22g sugars, 2g fiber), 1g protein.