

## Appendix Cancer

### What is Appendix Cancer?

Cancer of the appendix, also called appendiceal carcinoma, is a relatively rare condition in the gastrointestinal (GI) system. According to the American Cancer Society, it affects less than 1000 people each year and accounts for about five percent of all GI cancers in the United States.

The appendix is a tube-like structure, three to six inches long, that is attached to the colon. It is located in the lower right part of the abdomen and serves no known purpose.

Appendix cancers are usually found after patients have surgery for acute appendicitis or when an abdominal mass is seen on a computerized tomography (CT) scan for an unrelated condition.

There are two main types of appendix cancer: carcinoid and non-carcinoid. Carcinoid tumors are the most common type accounting for approximately 66 percent of all cases. It typically develops at the tip of the appendix. Most patients have no symptoms until the cancer spreads to other organs. If diagnosed early, when the cancer is contained only in the area where it started, this type of appendix cancer has a good prognosis for successful treatment. The average age of being diagnosed with carcinoid appendix cancer is approximately 40.

Mucinous cystadenocarcinoma is the most common form of non-carcinoid tumors representing about 20 percent of all cases. These tumors develop from the epithelial cells lining the inside of the appendix and are slow-growing. Most of the cells creating these tumors produce a jelly-like material called mucin. Over time, the tumor cells and mucin may spread within the abdominal cavity, which contains the stomach, spleen, liver, gallbladder, pancreas, small intestine, kidneys, ureters and most of the colon. If left untreated, the tumor cells and mucin can build up and lead to bowel obstruction, weight and muscle loss and loss of appetite. This type of cancer rarely spreads beyond the abdominal cavity.

Other rare types of appendix cancer include colonic-type adenocarcinoma and signet ring adenocarcinoma, also commonly called signet ring cell cancer. Colonic-type adenocarcinoma accounts for about 10 percent of appendix tumors. This type of cancer can look and behave like the more common form of colorectal cancer. Signet ring cell cancer is very rare and considered to be one of the more aggressive types.

### What are the Risk Factors for Appendix Cancer?

A risk factor is anything that increases the chance of developing a specific cancer. There are no known risk factors for appendix cancer, nor is there a significant risk of inheriting it from a blood relative.

## **How is Appendix Cancer Diagnosed?**

Common symptoms of appendix cancer include fatigue, loss of appetite and weight loss. Other symptoms may include feeling bloated, quickly feeling full after eating, difficult bowel movements, frequent urination, shortness of breath sometimes accompanied by an increased heart rate, abdominal and low back pain, indigestion, reflux, gas, nausea and/or vomiting. Due to pressure on the groin area from the cancer, a hernia may also develop.

Cancer of the appendix is commonly found after the patient is treated for suspected appendicitis or bloating around the abdomen or during surgery to repair a hernia or another issue.

## **What is Staging?**

After your diagnosis, other tests and exams are done to determine the size of the cancerous tumor and whether the disease has spread to other parts of the body. This process is called staging. Your doctor needs to know the stage of the disease in order to plan the best treatment for you.

A CT scan along with previous pathology and surgical reports are most commonly used to determine the stage of this cancer. Depending on your case, your doctor may order additional tests. Because these tumors are often slow-growing, positron emission tomography (PET) scans are not always helpful to determine the stage of this cancer.

If the cancer is in an early stage, it is contained in the area where it started or only involves the regional lymph nodes.

Late stage or metastases means the cancer has spread to distant organs or lymph nodes. In cancers of the appendix, this is often associated with signs of ascites (the buildup of fluid in the abdomen).

## **How is Appendix Cancer Treated?**

Treatment depends on the cancer's type and stage as well as the patient's overall health.

### **Surgery**

If caught at an early stage, appendix cancer can be treated with surgery alone. Common types of surgery for this type of cancer are appendectomy, hemicolectomy and cytoreductive.

An appendectomy is the removal of the appendix and is usually the only treatment needed if the tumor is smaller than one and a half centimeters.

A hemicolectomy is the removal of a portion of the colon next to the appendix, nearby blood vessels and lymph nodes following an appendectomy. This is usually used for a tumor larger than two centimeters.

Cytoreductive surgery, also known as tumor debulking, may be performed to treat later stages of cancer after it has spread in the abdominal cavity. The surgeon will attempt to remove as much of the bulk as possible. Although the surgery will not remove every tumor cell, it can provide symptom relief for patients. Sometimes this surgery will be combined with heated intraperitoneal chemotherapy (HIPEC). HIPEC is a highly concentrated, heated chemotherapy treatment delivered into the open abdomen to help kill any remaining tumor cells by bathing all areas of the abdominal cavity. This procedure may also involve removing other involved organs such as part of the intestines, gallbladder, spleen, ovaries and uterus.

## **Systemic Chemotherapy**

Systemic chemotherapy delivers drugs through the bloodstream in order to reach cancer cells throughout the body. This is usually done intravenously using a needle placed into a vein. The drugs used kill cancer cells by stopping their ability to grow and divide. There is no standard of care for use of chemotherapy in treatment of appendix cancer. However, drugs used to treat colorectal cancer have shown to be beneficial. Chemotherapy can be given after surgery to decrease the chance of cancer returning, before surgery to reduce the size of the tumor or when surgery is not an option.

## **Managing Side Effects**

Ascites or the buildup of fluid in the abdominal cavity may cause bloating and pressure-like pain. If appropriate, the fluid can be removed by inserting a drainage tube.

Pain can be controlled with appropriate medicines. Tell your doctor if you feel pain.

Nausea/vomiting and loss of appetite are common side effects from the cancer itself and/or chemotherapy treatments. Small frequent meals are a good way to maintain calorie intake. If you are having difficulty maintaining your weight, a feeding tube may be temporarily or permanently placed in the abdomen for nutritional support.

Constipation can be caused by pain medicine or by the cancer. Maintaining normal bowel movements and good hydration is important. Your dietitian can help you with this.

## **Potential Complications**

Some of the common complications from appendix cancer are bowel obstruction (blockage of the bowel), abdominal pain and blood clots.

## **Follow-Up Care**

Routine blood work is needed to make sure your electrolytes and blood counts are normal, and your liver and kidneys are functioning properly. Stay well hydrated by drinking plenty of water. You will have a CT scan periodically to assess if the tumor is responding to treatment.

## **Resources**

The American Cancer Society offers free booklets, support groups and programs. You can call 800.227.2345 or visit [www.cancer.org](http://www.cancer.org).

The Cancer Information Service (CIS), supported by the National Cancer Institute, is a free telephone service providing current information on cancer prevention, detection, diagnosis, treatment and rehabilitation. CIS also makes referrals for counseling, home care services, hospice and screening programs. Bilingual counselors are available. You can call 800.422.6237 or visit [cis.nci.nih.gov](http://cis.nci.nih.gov).

Cancer.Net is sponsored by more than 30,000 oncology doctors. This doctor-approved resource helps patients and families make informed health decisions. You can call 888.651.3038 or visit [www.cancer.net/cancer-types/appendix-cancer](http://www.cancer.net/cancer-types/appendix-cancer).

The "Shooting for a Cure" sporting clay tournament was established in 2009 to raise funds for appendix cancer research at MD Anderson Cancer Center. For more information, visit [www.shootoutcancer.org](http://www.shootoutcancer.org).