

Appendiceal Cancer

What is appendiceal cancer?

Cancer of the appendix, also called appendiceal cancer, is a relatively rare cancer in the gastrointestinal (GI) system that affects about 1500 people each year.

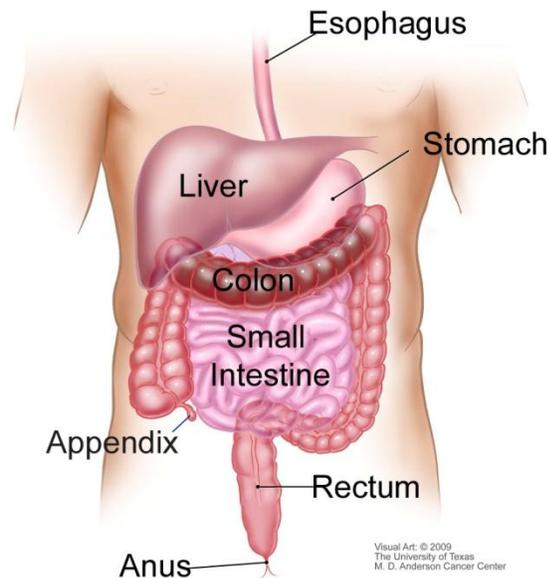
The appendix is a tube-like structure three to six inches long that is attached to the colon. It is located in the lower right part of the abdomen and serves no known purpose.

Appendiceal 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 appendiceal cancer: carcinoid and non-carcinoid epithelial tumors.

Carcinoid tumors are the most common form of appendiceal cancers accounting for approximately 66 percent of all appendix tumors. An appendix carcinoid tumor 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 good prognosis for successful surgery.

Mucinous cystadenocarcinoma is the most common non-carcinoid appendiceal carcinoma representing about 20 percent of all appendix cancer cases. These tumors develop from the epithelial cells lining the inside of the appendix and are slow-growing. They rarely spread beyond the abdominal cavity (area of the body containing the stomach, spleen, liver, gallbladder, pancreas, small intestine, kidneys, ureters and most of the colon). Most of the cells creating these tumors produce a jelly-like material known as mucin. Over time, the tumor cells and mucin may spread within the abdominal cavity. If left untreated, the tumor cells and mucin build up. This build up can cause a bowel obstruction, weight loss, muscle loss and loss of appetite.



Appendix and nearby organs

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Other rare types of appendix cancer include:

- Colonic-type adenocarcinoma – This cancer accounts for about 10 percent of appendiceal tumors. This type of cancer can cause similar symptoms and acts like common colorectal cancer.
- Signet-ring cell adenocarcinoma – This type is very rare and is considered one of the more aggressive types of appendiceal cancer.

What are the risk factors for appendiceal cancer?

A risk factor is anything that increases the chance of developing a specific cancer. There are no known risk factors for appendiceal cancer, nor is there a significant risk of inheriting this type of cancer from a blood relative. The average age of being diagnosed with carcinoid appendix cancer is approximately 40.

How is appendiceal cancer diagnosed?

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

Cancer of the appendix is commonly found:

- after the patient is treated for symptoms of suspected appendicitis,
- during surgery to repair a hernia,
- during surgery for another reason, or
- when the patient is treated for unexplained bloating around the abdomen.

What is staging?

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

A CT scan and previous pathology and surgical reports are most commonly used to determine the stage of the 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.

Early stage – The cancer is contained in the area where it started or only involving regional lymph nodes.

Late stage/metastases – The cancer has spread to distant organ or lymph nodes. In appendiceal cancer cases, this is often associated with signs of ascites (fluid build up in the abdomen).

How is appendiceal cancer treated?

Treatment options depend on the cancer type, staging and to the patient's overall health.

Surgery

- If caught at an early stage appendiceal cancer can be treated with surgery alone. Types of surgeries include Appendectomy - The removal of the appendix and is usually the only treatment needed if the tumor is smaller than 1.5 centimeters.
- Hemicolectomy - The removal of a portion of the colon next to the appendix, nearby blood vessels and lymph nodes. This is usually used for a tumor larger than 2 centimeters.
- Cytoreductive surgery – This is also known as tumor debulking surgery and may be preformed to treat later stages of cancer after it has spread to 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). During surgery, chemotherapy is delivered into the open abdomen to help kill any remaining tumor cells by bathing all areas of the abdominal cavity. The surgery can involve removing involved organs such as part of the intestines, gallbladder, spleen, ovaries, uterus or others. If you have this surgery, your health care team will explain your treatment plan to you in detail.

Chemotherapy

Systemic chemotherapy is cancer fighting drug(s) delivered through a vein to reach cancer cells throughout the body. The drugs are used to kill cancer cells by stopping their ability to grow and divide. There is no standard of care for use of chemotherapy in treatment of appendiceal 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,
- for late stage cancer to reduce the size of the tumor before surgery, or
- when surgery is not an option.

Other treatment options are being studied.

Managing Side Effects

Ascities (a type of fluid) may build up in the abdominal cavity causing bloating and pressure-like pain. If appropriate, the fluid can be removed. If palliative (comfort) care is required, a G-tube and/or J-tube may be inserted. A G-tube is used to deflate the stomach and a J-tube provides nutrition.

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.

Constipation can be caused by pain medicine or the tumor(s). 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 clot.

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 and other issues.

Resources

American Cancer Society

800-227-2345

<http://www.cancer.org>

Across the United States, this voluntary organization offers free booklets, support groups and programs.

Cancer Information Service

800-422-6237

<http://cis.nci.nih.gov/>

Supported by the National Cancer Institute, this free telephone service provides current information on cancer prevention, detection, diagnosis, treatment and rehabilitation. CIS also makes referrals for counseling, home care services, and hospice and screening programs. Bilingual counselors are on duty to help.

Cancer.Net

888-651-3038

<http://www.cancer.net/cancer-types/appendix-cancer>

Cancer.Net is sponsored by more than 30,000 oncology doctors. This doctor-approved resource helps patients and families make informed health decisions.

Shooting For a Cure

<http://www.shootoutcancer.org>

Established in 2009, the “Shooting for a Cure” sporting clay tournament was created to raise funds for appendiceal cancer research at MD Anderson Cancer Center.