

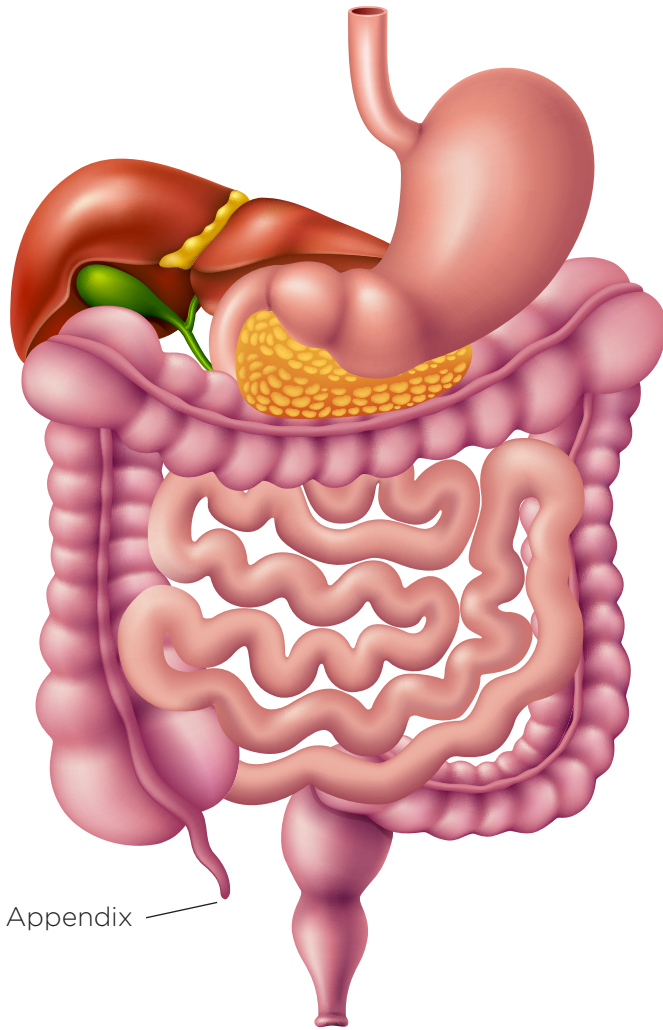


Appendiceal (Appendix) Cancer

Appendiceal Cancer

Is a disease in which malignant (cancer) cells form in the appendix. The appendix is a small pouch connected to your colon. Appendix cancer is rare and treated in different ways depending on the tissue type. Appendix tumors may be found when not expected during removal of the appendix or another surgery.

- **Neuroendocrine** appendix tumors (Carcinoids) are reviewed in a separate Neuroendocrine Tumors guide.
- **Gastrointestinal Stromal Tumors** are reviewed in a separate guide.
- **Adenocarcinoma** is a tissue type of appendix cancer that behaves like others such as colon cancer.
 - **Goblet cell adenocarcinoma** is a specific type of adenocarcinoma that can be fast-growing.
 - **Signet-ring adenocarcinoma** is a rare and fast-growing type of appendix cancer.
- **Low-grade appendiceal mucinous neoplasm (LAMN)** is a type of rare appendix cancer. Depending on the type of cells, they may be considered low grade or high grade. High grade cell-types are more likely to spread to other parts of the body.
 - A condition called **pseudomyxoma peritonei** occurs if the tumor breaks out of the appendix and covers the space inside the abdomen.



Risk Factors for Appendix Cancer

Anything that increases your chance of getting a disease is called a risk factor. Having a risk factor does not mean that you will get cancer; not having risk factors does not mean that you will not get cancer. Risk factors for appendix cancers are not known. Older age is the main risk factor for most cancers. The chance of getting cancer increases as you get older.

Signs and Symptoms

If there are signs and symptoms for appendiceal cancer they are often overlooked. The symptoms below can also be caused by other conditions:

- Pain or discomfort in the lower right section of your abdomen
- Bloating
- Fluid build-up in the abdomen, called ascites
- Acid back-up called reflux
- Poor appetite
- Shortness of breath
- Problems digesting food
- Constipation or diarrhea

Tests Used to Detect (Find) and Monitor Appendix Cancer

There is no screening test for people who do not have symptoms for appendix cancer. The following tests and procedures may be used to find and monitor cancer.

Carcinoembryonic Antigen (CEA) Assay

A test that measures the level of CEA in the blood. CEA is released into the bloodstream from both cancer cells and normal cells. A higher than normal amount may be a sign of cancer or other health problems. This test is considered non-specific. Some people who have appendix cancer never have a high CEA. It can help monitor treatment or signal a return of cancer.

CT Scan (Computed Tomography)

A series of detailed pictures of areas inside the body are taken from different angles. The pictures are made by a computer linked to an x-ray machine. A dye may be given into a vein or swallowed to help the organs or tissues show up more clearly.

MRI (Magnetic Resonance Imaging)

A magnet, radio waves, and a computer are used to make a series of detailed pictures. A substance called gadolinium is injected through a vein. The gadolinium collects around the cancer cells so they show up brighter in the picture.

PET Scan (Positron Emission Tomography Scan)

A small amount of radioactive glucose (sugar) is injected into a vein. The PET scanner rotates around the body and makes a picture of where glucose is being used in the body. Cancer cells show up brighter in the picture because they are more active and take up more glucose than normal cells do.

Certain Factors Affect Prognosis (Chance of Recovery) and Treatment Options

The prognosis (chance of recovery) and treatment options depend on the type of tissue and stage (extent) of the cancer. Appendix cancer can be cured if it is found and diagnosed early.

The process used to find out whether the cancer has spread to other parts of the body is called **staging**. It is important to know the stage in order to plan treatment.

Cancer can spread through tissue, the lymph (filter and transport) system, and the blood. When cancer spreads to another part of the body, it is called **metastasis**. Cancer cells break away from where they began (the primary tumor) and travel through the lymph system or blood. A metastatic tumor is the same type of cancer as the primary tumor. For example, if appendix cancer is found in the liver, it is appendix cancer, not liver cancer.

A condition commonly caused by some cancers is known as peritoneal carcinomatosis. This is when the appendix cancer has started to spread within the abdomen, causing small tumors to grow in your peritoneum (the tissue that lines your abdominal wall and inner organs).

Treatment Option Overview

Surgery

Surgery (removing the cancer in an operation) is the most common treatment for appendix cancer. Further treatment may be needed during or after surgery, which can include radiation or drug therapy. For some types and in an early stage, surgery may be all that is needed.

Hyperthermic Intraperitoneal Chemotherapy (HIPEC) may be used to treat certain cancers in the abdomen and pelvis. Following surgical removal of all cancer that can be seen (this is called cytoreduction), HIPEC is given while you are still asleep and under general anesthesia. With HIPEC therapy, chemotherapy (chemo) is put directly into the belly (abdominal cavity) and circulated using a pump.

Hyperthermic – refers to chemotherapy heated to 42°C (107°F), above normal body temperature

Intraperitoneal – refers to chemotherapy delivered to the lining of the abdominal cavity.

Radiation Therapy

Radiation therapy is a cancer treatment that uses high-energy x-rays or other types of radiation to kill cancer cells or keep them from growing.

Chemotherapy

Chemotherapy is a treatment that uses drugs to stop the growth of cancer cells, either by killing the cells or by stopping the cells from dividing.

Chemotherapy can be given in different ways:

- Systemic chemotherapy can be given by mouth or in an IV. The treatment can affect all cells.
- Local (intraperitoneal) chemotherapy is focused on one part of the body. For appendix cancer, it can be given during a surgery or later through a tube in the abdomen. When the chemotherapy is warmed higher than the body temperature, it is called HIPEC (heated intraperitoneal chemotherapy).

Clinical Trials

Clinical trials are done to find out if new cancer treatments are safe and effective or better than the standard treatment.

People who take part in a clinical trial may receive:

- The standard drugs alone or
- The standard drugs plus the new treatment being studied

Taking part in a clinical trial helps improve the way cancer will be treated in the future. Even when clinical trials do not lead to effective new treatments, they often answer important questions and help move research forward.

Some clinical trials only include people who have not yet received treatment. Other trials test treatments for those whose cancer has not gotten better. There are also clinical trials that test new ways to stop cancer from coming back or reduce the side effects of cancer treatment.

Many of today's standard treatments for cancer are based on earlier clinical trials.

Ask if there is a clinical trial right for you.

Follow-Up Tests May Be Needed

Some of the tests that were done to diagnose the cancer or to find out the stage of the cancer may be repeated. It is extremely important to keep your follow-up appointments with your cancer provider. This will help show any early sign of cancer recurrence, and more testing can be ordered if needed.

Support is available for coping with changes that may have happened as a result of cancer treatment. Your healthcare team can offer ideas as well as a plan of care for long-term follow-up.

Common Questions

What does the pathology report say?

What is the stage of my cancer?

What are my goals for treatment?

What are my treatment choices?

What kind of support services are available for me about finances, emotions, spiritual questions, etc.?

My Health Care Team	Contact Information
Surgeon:	
Medical Oncologist:	
Radiation Oncologist:	
Primary Care Doctor:	
Navigator	
Nurse:	
Registered Dietitian Nutritionist:	
Other:	
Other:	
Other:	
Other:	

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Adapted from Appendiceal Cancer originally published by the National Cancer Institute
This information is not intended as a substitute for professional medical care.
Always follow your health care provider's instructions.

