Appendix Cancer

What is the Appendix?
The **appendix** is a pouch-like tube that is attached to the cecum (first section of the large intestine or colon). The appendix averages 4 inches in length and is part of the gastrointestinal (GI) tract. It is generally thought to have no special function in the body. Surgical removal of the appendix causes no health problems.

![Diagram of the appendix and its relation to the large intestine and small intestine]

What is Appendix Cancer?
Appendix cancer happens when cells in the appendix become abnormal and multiply without control. These cells form a growth of tissue, called a tumor. Another name for this type of cancer is appendiceal cancer.

Types of Appendix Cancers:
- **Malignant Carcinoid**: A carcinoid tumor starts in the hormone-producing cells that are normally present in small amounts in almost every organ of the body. An appendix carcinoid tumor most often occurs at the tip of the appendix. Over one-half of appendix tumors are carcinoids. Most carcinoids are small and can often be treated successfully with surgery.
- **Mucinous Cystadenocarcinoma and Pseudomyxoma Peritonei (PMP)** - These are 20% of appendix cancers. This type of tumor produces a jelly-like substance called mucin that can fill the abdominal cavity.

- **Colonic-Type Adenocarcinoma** - These are 10% of appendix cancers and occur at the base of the appendix. This type looks and acts like colorectal cancer.

- **Signet Ring Cell Adenocarcinoma** - This type is very rare and is more aggressive and difficult to treat.

**Is this a common cancer?**
Cancers of the appendix are rare. There are less than 1,000 cases per year.

**What are the risk factors?**
- Smoking tobacco
- Gender: Women are more likely to develop carcinoids
- Family history of appendix cancers
- Age: Average age at diagnosis is age 40
- Certain health conditions

**What are the symptoms?**
People with appendix cancer may experience some or none of the following symptoms:
- Appendicitis
- Fluid in the abdomen (ascites)
- Bloating/Increase in the size of the abdomen
- Pain in the abdomen or pelvis
- Changes in bowel function

**What is the prognosis?**
The prognosis (chance of recovery) depends on:
- Type of tumor
- If it has spread to other parts of the body
- Whether the cancer can be completely taken out by surgery
- Whether the cancer is a new diagnosis or has come back
- Your overall health

**How is it diagnosed?**
- **History and Physical Exam** - review of your symptoms, health habits, past illnesses and previous treatments.
- **Biopsy** - The removal of a small amount of tissue that is checked under a microscope to learn if it is cancer and where it came from. Often it is diagnosed unexpectedly by a surgery for appendicitis.
- **CT Scan** - A series of computerized pictures of the inside of the body taken after drinking or injecting into a vein a special dye.
- **MRI** - Magnetic Resonance Imaging uses a magnet, radioactive waves and a computer to take pictures of the inside of the body.
How is appendix cancer staged?
The stage describes a cancer’s growth or spread. This helps to determine what kind of treatment is best and can help predict prognosis.

One tool doctors use to describe the stage is the **TNM** system.
- **Tumor (T)** - How large is the tumor and where is it located?
- **Node (N)** - Has the tumor spread to the lymph nodes?
- **Metastasis (M)** - Has the cancer spread to other organs in the body?

Appendix cancer can be described by stage:
- **Localized** - Cancer is found in the appendix, colon, rectum small intestine and/or stomach only.
- **Regional** - Cancer has spread from the appendix, colon, rectum, stomach and/or small intestine to nearby tissues or lymph nodes.
- **Metastasis** - Cancer has spread to other parts of the body.

How is it treated?
- **Surgery** - The removal of the tumor and surrounding tissue during an operation. It is the most common treatment for appendix cancer.

- **Types of surgery include:**
  - **Appendectomy** - Removal of the appendix
  - **Hemicolectomy** - Removal of a portion of the colon next to the appendix.
  - **Debulking surgery** - For later stages of appendix cancer this may be done. The surgeon removes as much of the tumor “bulk” as possible. Sometimes this is followed by chemotherapy. If the cancer produces mucous, much of this is removed and often relieves bloating.

- **Chemotherapy** - The use of drugs to kill cancer cells, usually by stopping the cancer cells’ ability to grow and divide.
  - **Local/Intraperitoneal Chemotherapy** - This is done by a surgeon after a debulking surgery. Chemotherapy is heated and given directly into the abdominal cavity. This is called **HIPEC**. The chemotherapy is given through a tube. Once the chemotherapy is completed, the tube is taken out.
  - **Systemic chemotherapy** - Chemotherapy medicine is injected intravenously (IV) and enters the bloodstream to reach cancer cells throughout the body. This is called **systemic** chemotherapy.

- **Radiation Therapy** - Rarely used in the treatment of appendix cancer. This is a treatment that uses high energy x-rays or other types of radiation to kill cancer cells, or slow their growth.
Clinical Trials
You may have the chance to take part in a clinical trial. Clinical trials are controlled research studies done to find out if new cancer treatments are safe and effective, or better than the standard treatments. Clinical trials are voluntary and help find better treatments for cancer.

Follow-up Tests
During your treatments you will need blood tests and scans to see how well the treatment is working. These tests help guide decisions to keep going, stop, or change treatments. This is called restaging.

Blood tests and scans will be done from time to time after you have finished your treatments. They can show if your condition has changed or the cancer has recurred, or come back.