All About Small Bowel Cancer

What is the small bowel?

The small bowel, also known as the small intestine, is the portion of the digestive tract that connects the stomach and the large bowel (colon). There are three distinct parts of the small bowel: 1) the duodenum, 2) the jejunum and 3) the ileum. (Note: Although these three parts can all be grouped under the term "small bowel", there are several notable differences between them, such as blood supply, degree of attachment, surrounding structures, etc.)

In order to fit inside a person's body, the small and large intestines are folded up into a very compact form. However, if you were to unfold the small bowel and measure its full length, it would be about 15 to 20 feet long! In total, the small bowel makes up about 75% of the entire digestive system.

The small bowel plays a critical role in the break down and absorption of food, so that important vitamins, minerals and nutrients can be absorbed into the body.

What is small bowel cancer?

Small bowel cancer occurs when cell in the small bowel begin to grow out of control. These cells can then invade nearby tissues and spread throughout the body. Large collections of cancer cell are called tumors. There are four main types of small bowel cancer, depending on the appearance of the cells under a microscope and the "cell of origin" (cell type in which the cancer starts):

- **Adenocarcinoma**: Represents 30-40% of all small intestine cancers. It typically starts in the lining or inside layer of the bowel, and usually occurs in the duodenum. Like adenocarcinomas of the colon or rectum, these tumors are thought to arise from a benign growth, known as an adenoma, in the small bowel. Adenocarcinoma of the small bowel is more common in males and occurs around 50-70 years of age. This type of small bowel cancer is more likely to be diagnosed at a late stage. People with Crohn's disease may present at an earlier age.
- **Sarcoma**: Constitutes 10-15% of small bowel cancers. The typical subtype is leiomyosarcoma, which starts in the muscle wall of the small bowel and usually occurs in the ileum. One uncommon subtype is gastrointestinal stromal tumor (GIST), which can occur in any of the three parts of the small bowel. GIST tumors make up about 7-15% of cancers of the small bowel.
- **Carcinoid**: Accounts for 35-42% of small bowel cancers. Carcinoid cancers of the small bowel tend to occur in the ileum. Carcinoid tumors are a type of neuroendocrine tumor.
- **Lymphoma**: Responsible for 15-20% of small intestine cancers. Small intestine lymphoma starts in the lymph tissue of the small bowel and usually occurs in the jejunum. The typical subtype is non-Hodgkin's lymphoma; more specifically MALT lymphomas, large B cell lymphoma, immunoproliferative small intestinal disease, and Burkitt's lymphoma.

Occasionally, small bowel cancer may actually be metastatic cancer, meaning that it has spread to the small bowel from a primary cancer located elsewhere in the body. In that case, it is treated as the primary cancer would be – if it has spread from the lung, it is treated as a lung cancer.

There are also several benign tumors that can arise from the small bowel including: adenomas, leiomyomas, fibromas, and lipomas.

What causes small bowel cancer and am I at risk?

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Despite the amazingly long length of the small bowel compared to the rest of the digestive tract, cancer of the small bowel is very rare. This includes either cancers starting in the bowel or cancers spreading there from another body site. Despite comprising 75% of the digestive tract and 90% of the surface of the digestive tract, small bowel cancers account for only 1-2% of all gastrointestinal cancers. It is estimated each year there were 10,190 new cases of small bowel cancer. This represents 0.6% of all new cancer cases diagnosed in the year. It is more prevalent in African American men and women than in other races.

Unfortunately, the cause of most small bowel cancers is unknown. However, there are some risk factors that might increase a person's chance of developing small bowel cancer. Older age may also be a risk factor. Small bowel cancers also occur more commonly in those over the age of 55. Some studies have found an increased risk associated with smoking and alcohol consumption. Those with Celiac disease may be at a higher risk for a small bowel lymphoma, called enteropathy-associated T-cell lymphoma, as well as have a higher risk of small bowel adenocarcinoma. Survivors of colon cancer also have a higher risk of developing small bowel cancer, as do those with Crohn's disease.

There are also several genetic syndromes associated with a higher risk of developing small intestine adenocarcinoma. These include familial adenomatous polyposis (FAP), hereditary nonpolyposis colorectal cancer (HNPCC), Peutz-Jeghers syndrome (PJS), MYTYH-associated polyposis and Cystic Fibrosis (CF). Those with these genetic syndromes should consult with their healthcare team and a geneticist to more accurately gauge their risk for developing small bowel cancer and any screening needs.

Finally, immunosuppression associated with HIV/AIDS or organ transplantation can result in a higher risk of small bowel lymphoma.

**How can I prevent small bowel cancer?**

There are no known ways to prevent small bowel cancer. Since tobacco use is a potential risk factor for developing small bowel cancer, it is important to quit smoking or never start smoking. It is also important to eat a healthy diet and exercise regularly to reduce your cancer risk.

Those with FAP who have many polyps and are considered at especially high risk for developing small bowel cancer should consult with their healthcare team about the benefits of surgical intervention (for example: a Whipple procedure) to remove parts of the small bowel/gastrointestinal system before cancer can develop.

**What screening tests are available?**

There are no routine screening tests available for small bowel cancer. Be sure to tell your healthcare provider if you have a history of FAP so you can be closely monitored.

**What are the signs of small bowel cancer?**

Given how rare small bowel cancer is compared to many other cancerous and noncancerous diseases, it is already a difficult diagnosis. On top of that, the symptoms of small bowel cancer are usually pretty nonspecific, adding to the difficulty of diagnosis.

Common symptoms include:

- crampy abdominal (belly) pain
- blood mixed in the stools
- dark/tarry/black stools (from "digested" blood)
- weight loss
- diarrhea
- jaundice
- weakness
- fatigue
- anemia (decreased red blood cell counts)
Again, these symptoms could be caused by a number of medical conditions. If symptoms such as the ones listed above are severe, worsening, or persistent, medical attention should be sought so that a timely and accurate diagnosis can be made. Anytime blood is in the stools or the stools are black, medical attention should be sought.

A far less common presenting symptom is bowel obstruction, in which the tumor blocks the passage of food products through the bowel. The blockage could be complete or partial. Bowel obstruction can cause sharp belly pain, sensation of abdominal bloating, vomiting, and, of course, constipation. In the worst case scenario, the bowel blockage can actually cause the bowel to suddenly rupture, leading to severe pain and shock (dangerous drop in blood pressure). This is a medical emergency requiring surgical intervention.

With carcinoid tumors, people can experience watery diarrhea, flushing, wheezing and decreased blood pressure. Specific details on carcinoid tumors are provided in a separate overview.

How is small bowel cancer diagnosed?

Your healthcare provider will take a complete medical history and perform a physical examination. They will likely order some basic blood tests, especially if there is a history of blood loss in the stool or diarrhea.

Additional tests to diagnose small bowel cancer can include:

- Stool sample to test for blood
- Endoscopy or colonoscopy: looks inside the duodenum and the upper part of the jejunum (from above, through the throat and past the stomach) or the lower part of the ileum (from below). A biopsy can also be taken through the endoscope to confirm diagnosis.
- Enteroscopy can investigate more of the small bowel but is more invasive and requires special equipment and training. There is also a pill with a small camera, which can be used to assess the bowel (capsule endoscopy). The pill is swallowed and then takes pictures periodically, which are sent wirelessly to a computer where they can be viewed.
- Barium x-rays: also known as small bowel follow-through. During this procedure, the patient first drinks barium liquid (which looks white on x-ray) and then gets abdomen x-rays to follow the passage of the barium through the entire small bowel. Enteroclysis, a special kind of barium study, which uses double contrast (both barium and methylcellulose) to get a better picture of the intestine can also be used. This technique can also be used with CT (CAT) scans.
- Radiology tests: CT scans, ultrasound scans and other kinds of x-rays, which sometimes pick up a small bowel tumor, but more importantly look at other parts of the body to see if the cancer may have spread.
- MRI may be ordered if liver involvement is suspected, which can occur with carcinoid tumors.

These methods are methods of guiding the clinical diagnosis, based on clinical studies (physical exam, radiology studies, etc.). They can identify a small bowel cancer about 50% of the time, the remainder are found through surgery. An actual pathologic diagnosis requires biopsy and microscopic evaluation of tissue from the suspected mass by a pathologist. However, it can be quite difficult to clearly visualize - and then access - the tightly folded-up small bowel, and biopsies cannot always be done. In these cases, the pathologic diagnosis may have to be made as part of a surgical operation.

How is small bowel cancer staged?

In order to guide treatment and offer some insight into prognosis, small bowel cancer is staged. Healthcare providers use the TNM system (also called tumor - node - metastasis system). This system describes the size and local invasiveness of the tumor (T), which, if any, lymph nodes are involved (N), and if it has spread to other more distant areas of the body (M). This is then interpreted as a stage somewhere from I (one) denoting more limited disease to IV (four) denoting more advanced disease. Generally, the higher the stage, the more serious the cancer. The TNM breakdown is quite technical, but is provided at the end of this article for your reference. Your healthcare provider will use the results of the diagnostic work up to assign the TNM result.

How is small bowel cancer treated?

This section focuses on the treatment of adenocarcinomas of the small bowel, as it is most common. Treatment varies somewhat based on the tumor type, size and location of the tumor, involvement of lymph nodes and metastasis.
**Surgery**

The main treatment option for small bowel cancer is surgery to remove the tumor (excision) and to reconnect the remaining bowel (anastomosis). Surgery may also be needed for relief of bowel obstruction, (i.e., intestinal bypass when the obstructing tumor itself cannot be removed). The success of the surgery as a cure depends on the extent to which the surgeon can remove the entire mass during the operation. This depends on the location of the mass within the bowel as well as the amount of bowel that is involved.

In some cases, it could be necessary to remove part of the stomach, colon, the gall bladder or surrounding lymph nodes in order to do a more complete and effective surgery. The type of surgery used is largely dependent on the size of the tumor and its location.

Sometimes when a significant part of the bowel has to be removed in order to get the tumor out, there is not always enough bowel left over to reconnect. In these cases, the top end (proximal part) of the bowel is brought up to the wall of the abdomen and connected to a hole in the skin called a stoma. Attached to the stoma is a bag that is worn under one’s clothes and collects the stool. This procedure is called an ileostomy, and may be temporary or permanent.

Depending upon the extent of the surgery, special diets, vitamins, supplements, etc. may be needed to help with food digestion and absorption. Any such dietary changes post-surgery would likely be permanent. The surgical team and a dietician will provide information about these changes if they are relevant to you.

**Radiation**

*Radiation treatment* plays a smaller role than surgery in the management of small bowel cancers. In some cases, it can be used post-operatively if there is tumor left behind or if there are close surgical margins (the cancer cells were close to the edge of the tissue removed), to "clean up" any microscopic tumor cells. It may also be used to help palliate (relieve) symptoms from advanced disease, such as blood loss or pain from the tumor. Radiation therapy for bowel cancer can be delivered either by external beam or, less commonly, brachytherapy (implants, usually at time of surgery).

**Chemotherapy**

*Chemotherapy* may be given either in combination with radiation therapy/surgery or by itself as a single treatment in select cases. Several chemotherapy medications may have some efficacy in the treatment of small bowel cancer. However, given how rare small bowel cancer is, very few clinical trials have been completed showing the benefits of chemotherapy in the treatment of small bowel cancer. Some of the chemotherapy medications used in the treatment of small bowel cancer include: capecitabine, oxaliplatin, fluorouracil, irinotecan, and leucovorin. These medications are typically used in combination.

Chemotherapy may take a primary role in the treatment for small bowel cancers classified as lymphomas.

**Targeted Therapy**

One use of non-standard oncologic therapy for this tumor is interferon. *Interferon* is a medication used for some types of small bowel cancers, typically carcinoids. This medication work by activating the body’s immune system to fight the cancer.

For gastrointestinal stromal tumors (GIST) of the small bowel, there is a large role for the targeted therapy medications including imatinib, sunitinib, and regorafenib.

**Clinical Trials**

There are clinical research trials for most types of cancer, and every stage of the disease. Clinical trials are designed to determine the value of specific treatments. Trials are often designed to treat a certain stage of cancer, either as the first form of treatment offered, or as an option for treatment after other treatments have failed to work. They can be used to evaluate medications or treatments to prevent cancer, detect it earlier, or help manage side effects. Clinical trials are extremely important in furthering our knowledge of disease. It is through clinical trials that we know what we do today, and many exciting new therapies are currently being tested. Talk to your provider about participating in clinical trials in your area. You can also explore currently open clinical trials using the OncoLink Clinical Trials Matching Service.

**Follow-Up Care and Survivorship**
Once a patient has been treated for small bowel cancer, you will need to be closely followed for a recurrence. At first, you will have follow-up visits fairly often. The longer you are free of disease, the less often you will have to go for checkups. Your healthcare provider will tell you when he or she wants follow-up visits, and/or CT scans, depending on your case. It is very important that you let your healthcare provider know about any symptoms you are experiencing and that you keep all of your follow-up appointments.

Fear of recurrence, relationship issues, financial impact of cancer treatment, employment issues, and coping strategies are common emotional and practical issues experienced by small bowel cancer survivors. Your healthcare team can identify resources for support and management of these challenges faced during and after cancer.

Cancer survivorship is a relatively new focus of oncology care. With some 15 million cancer survivors in the US alone, there is a need to help patients transition from active treatment to survivorship. What happens next, how do you get back to normal, what should you know and do to live healthy going forward? A survivorship care plan can be a first step in educating yourself about navigating life after cancer and helping you communicate knowledgeably with your healthcare providers. Create a survivorship care plan today on OncoLink.

**Resources for More Information**

**CancerCare**

Cancer Care provides counseling, support groups, education and financial assistance.

https://www.cancercare.org/diagnosis

**Imerman Angels**

Imerman Angels provides you with one-on-one support through their cancer support community.

https://imermanangels.org

**Appendix: Complete Small Bowel Cancer Staging**

AJCC, Cancer Staging Manual, 8th Edition

<table>
<thead>
<tr>
<th>Primary Tumor (T)</th>
<th>Description</th>
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<tr>
<td>Tx</td>
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<tr>
<td>T0</td>
<td>There is no evidence of a primary tumor</td>
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<tr>
<td>Tis</td>
<td>Carcinoma in situ / High grade dysplasia</td>
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<tr>
<td>T1a</td>
<td>Tumor limited to the lamina propria (innermost layer of the small bowel)</td>
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<tr>
<td>T1b</td>
<td>Tumor invades the submucosa</td>
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<tr>
<td>T2</td>
<td>Tumor involves the muscularis propria</td>
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<tr>
<td>T3</td>
<td>Tumor has grown through the muscularis propria and into the subserosa or into the tissues surrounding the small bowel</td>
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<td>T4</td>
<td>Tumor has invaded other organs or has grown through the lining of the visceral peritoneum</td>
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<td>N0</td>
<td>No regional lymph node metastasis</td>
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<tr>
<td>N1</td>
<td>Involvement of 1-2 regional lymph nodes</td>
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<tr>
<td>N2</td>
<td>Involvement of 3 or more lymph nodes</td>
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<td>Distant metastasis cannot be evaluated</td>
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<tr>
<td>M0</td>
<td>No distant metastasis</td>
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<td>M1</td>
<td>Distant metastasis present</td>
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<table>
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