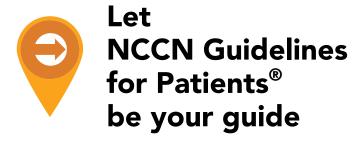




It's easy to get lost in the cancer world



- ✓ Step-by-step guides to the cancer care options likely to have the best results
 - ✓ Based on treatment guidelines used by health care providers worldwide
 - ✓ Designed to help you discuss cancer treatment with your doctors



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- Explain the cancer care options likely to have the best results

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These NCCN Guidelines for Patients are based on the NCCN Guidelines® for Esophageal and Esophagogastric Junction Cancers, Version 1.2022 – December 21, 2021.

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Sponsored by Esophageal Cancer Awareness Association (ECAA)

ECAA are comprised of all volunteers who support those involved in the esophageal cancer journey as well as raise awareness to learn the signs and symptoms of esophageal cancer. ecaware.org

Endorsed by Esophageal Cancer Education Foundation (ECEF)

ECEF is honored to join the chorus of those endorsing the good work by the NCCN and their Guidelines for Patients. ECEF takes its responsibility serious in providing information and support to not only patients, but also their families and care givers. Adding these Guidelines to the arsenal of information already on our website, and to our support group calls, is a welcome and much needed resource. The more information patients, their families and caregivers have, the more informed their decisions will be. That is the ultimate goal of ECEF, to walk the journey with patients who have this disease and support of those most in need. fightec.org



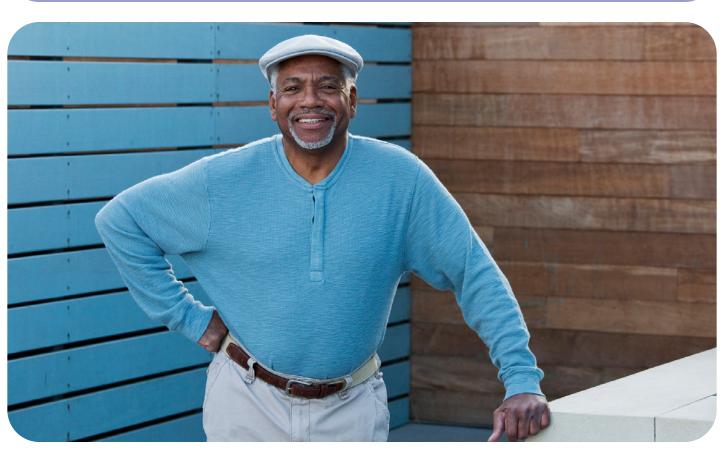
NCCN Guidelines for Patients® Esophageal Cancer, 2022

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1 Esophageal cancer basics

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1

The esophagus is a long, muscular tube through which food passes from the throat to the stomach. Esophageal cancer starts when abnormal cells grow out of control in the esophagus wall.

The esophagus

The esophagus is a long, muscular tube through which food passes from the throat to the stomach. It is located behind the trachea (windpipe) and in front of the spine.

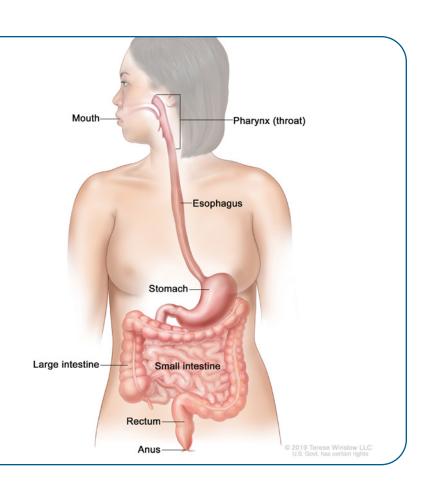
The esophagus is part of the digestive system. The digestive system takes in and breaks down food, absorbs nutrients, and removes waste from the body. It includes the esophagus, stomach, small intestine, colon, and rectum.

Food and drink enter the mouth and move through the esophagus into the stomach. When you swallow, small contractions, called peristalsis, along with gravity and pressure, move food down the esophagus and into the stomach. Except during the act of swallowing, the esophagus is normally empty.

The esophagus is about 10 inches long and is about 1 inch wide. It has two sphincters, circular muscles that normally remain closed except during the act of swallowing. One sphincter is located at the top of the esophagus and the other at the bottom, between the esophagus and the stomach.

The digestive tract

The digestive or gastrointestinal (GI) tract is part of the digestive system. Food enters the mouth and passes through the esophagus into the stomach. After being broken down into a liquid, food enters the small intestine. The large intestine prepares unused food to be moved out of the body.



Parts of the esophagus

Esophagus wall

The esophagus wall contains 4 layers:

- 1. Mucosa inner membrane that is in contact with food. It has 3 parts:
 - Epithelium innermost lining and is normally made up of flat, thin cells called squamous cells
 - Lamina propria thin layer of connective tissue under the epithelium
 - Muscularis mucosa very thin layer of muscle under the lamina propria
- Submucosa layer of connective tissue just below the mucosa that contains blood vessels and nerves. In some parts of the esophagus, this layer also includes glands that secrete mucus.
- 3. Muscularis propria thick layer of muscle under the submucosa. It contracts in a coordinated way to push food down the esophagus from the throat to the stomach.
- 4. Tunica adventitia outermost layer, made of connective tissue.

Upper esophagus

The upper part of the esophagus is made of striated muscle. Striated muscle generates force and contracts. It is under your control (voluntary).

Middle esophagus

The middle part of the esophagus is a mixture of striated and smooth muscle.

Lower esophagus

The lower part of the esophagus consists only of smooth muscle. Smooth muscle is not under your control. It is involuntary.

Esophagogastric junction

The esophagogastric junction (EGJ) is the place where the stomach and the esophagus meet, found just beneath the diaphragm.

Esophageal cancer

Cancer of the esophagus (or esophageal cancer) starts when cells in the esophagus begin to grow out of control.

There are 2 types of esophageal cancer:

- Esophageal adenocarcinoma (EAC) starts in the mucus-making cells of the esophagus. Adenocarcinomas are often found in the lower esophagus, but can occur in the mid-esophagus as well.
- Esophageal squamous cell carcinoma (ESCC) starts in the thin, flat cells found in the inner lining of the esophagus. Squamous cell carcinoma (SCC) is often found in the upper and middle esophagus, but can occur in the lower esophagus as well.

Today, most esophageal cancers in North America and Western Europe are adenocarcinomas. Squamous cell carcinoma is more common in Eastern Europe and Asia. Treatment is based on the type and location of esophageal cancer, in addition to staging information such as the tumor size and lymph node involvement.

1

Some tumors that start in the stomach and cross over into the area between the esophagus and stomach (esophagogastric junction) are treated as esophageal cancers and not as stomach cancers. However, this depends on the exact location of the tumor.

Barrett esophagus

In Barrett esophagus (BE), the squamous cells that line the lower part of the esophagus have changed or been replaced with abnormal cells. Those with Barrett esophagus are at risk of developing adenocarcinoma of the esophagus.

How cancer spreads

Esophageal cancer usually starts in the innermost layer and grows outward through and along the layers of the esophagus wall. Cancer can spread to nearby lymph nodes, veins, arteries, and organs such as the liver, pancreas, lung, and spleen. It might also grow into nearby lymphatic or blood vessels, and from there spread to nearby lymph nodes or to other parts of the body.

Esophageal cancers tend to develop slowly over many years. Before cancer develops, precancerous changes often occur in the inner lining (mucosa) of the esophagus. Since these early changes rarely cause symptoms, they often go undetected.

Those with esophageal cancer can be grouped into 3 main categories, depending on how far the cancer has spread.

- Early-stage cancer has not grown beyond the first layer (mucosa) of the esophagus wall. The tumor is often very small (2 centimeters or less) and is not in any lymph nodes.
- Locoregional or locally advanced cancer has invaded other layers of the esophagus wall and/or spread to the lymph nodes or organs near or in direct contact with the esophagus.
- Metastatic cancer has spread to other parts of the body. The most common metastatic sites are the liver, distant lymph nodes, lung, bone, and brain.

1

Key points

- The esophagus is part of the digestive system. The digestive system takes in and breaks down food, absorbs nutrients, and removes waste from the body.
- Most esophageal cancers start in cells that line the inside of the esophagus and secrete mucus. These esophageal cancers are called adenocarcinomas.
- Esophageal squamous cell carcinoma starts in the thin, flat cells found in the inner lining of the esophagus.
- The esophagus wall is made up of 4 main layers: mucosa, submucosa, muscularis propria, and tunica adventitia.
- Esophageal cancers tend to develop slowly over many years. Before cancer develops, pre-cancerous changes often occur in the inner lining (mucosa) of the esophagus.
- Early-stage esophageal cancer has not grown beyond the first layer (mucosa) of the esophagus wall. The tumor is often small and is not in any lymph nodes.
- Locoregional or locally advanced esophageal cancer has invaded other layers of the esophagus wall and/or spread to the lymph nodes or organs near or in direct contact with the esophagus.
- Cancer can spread to distant parts of the body through the blood or lymphatic system. This is called metastatic esophageal cancer. Distant metastases could be in the liver, distant lymph nodes, lung, bone, and brain.

4 layers of the esophagus wall

1. Mucosa – Inner membrane that is in contact with food.

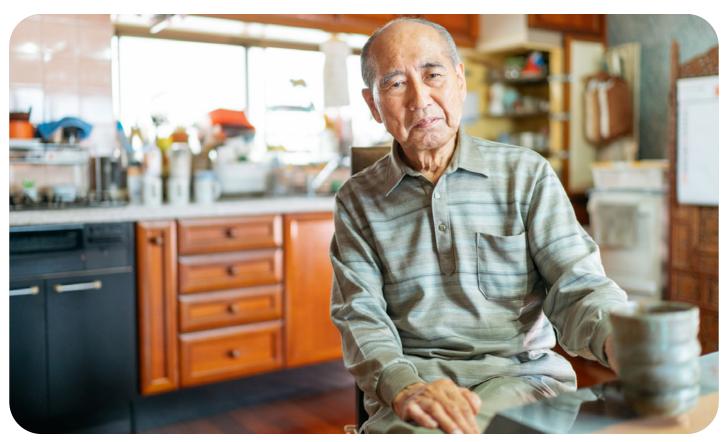
Mucosa consists of 3 layers:

- Epithelium A thin layer of squamous cells that forms the interior lining.
- Lamina propria A type of connective tissue found under the epithelium.
- Muscularis mucosa A thin strip of muscle that separates the mucosa from the submucosa.
- **2. Submucosa** A layer of connective tissue, blood vessels, and nerve cells.
- **3. Muscularis propria** A thick layer of muscle that helps move food through the esophagus.
- **4. Tunica adventitia** The outermost layer, consisting of connective tissue.

2 Diagnosing esophageal cancer

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Treatment planning starts with testing. This chapter presents an overview of the tests you might receive and what to expect.

Test results

Results from blood tests, imaging studies, and biopsy will be used to determine your treatment plan. It is important you understand what these tests mean. Ask questions and keep copies of your test results. Online patient portals are a great way to access your test results.

Keep these things in mind:

- Bring someone with you to doctor visits, if possible.
- Write down questions and take notes during appointments. Don't be afraid to ask your care team questions. Get to know your care team and help them get to know you.
- Get copies of blood tests, imaging results, and reports about the specific type of cancer you have.
- Organize your papers. Create files for insurance forms, medical records, and test results. You can do the same on your computer.
- Keep a list of contact information for everyone on your care team. Add it to your phone. Hang the list on your refrigerator or keep it in a place where someone can access it in an emergency. Keep your primary care physician informed of changes to this list.



Create a medical binder

A medical binder or notebook is a great way to organize all of your records in one place.

- Make copies of blood tests, imaging results, and reports about your specific type of cancer. It will be helpful when getting a second opinion.
- Choose a binder that meets your needs.
 Consider a zipper pocket to include a pen, small calendar, and insurance cards.
- Create folders for insurance forms, medical records, and tests results. You can do the same on your computer.
- Use online patient portals to view your test results and other records. Download or print the records to add to your binder.
- Organize your binder in a way that works for you. Add a section for questions and to take notes.
- Bring your medical binder to appointments. You never know when you might need it!

General health tests

Medical history

A medical history is a record of all health issues and treatments you have had in your life. Be prepared to list any illness or injury and when it happened. Bring a list of old and new medicines and any over-the-counter medicines, herbals, or supplements you take. Tell your doctor about any symptoms you have. A medical history will help determine which treatment is best for you. It is sometimes called a health history.

Family history

Some cancers and other diseases can run in families. Your doctor will ask about the health history of family members who are blood relatives. This information is called a family history. You can ask family members about their health issues like heart disease, cancer, and diabetes, and at what age they were diagnosed.

Physical exam

During a physical exam, your health care provider may:

- Check your temperature, blood pressure, pulse, and breathing rate
- Check your weight
- Listen to your lungs and heart
- Look in your eyes, ears, nose, and throat
- Feel and apply pressure to parts of your body to see if organs are of normal size, are soft or hard, or cause pain when touched. Tell your doctor if you feel pain.

Feel for enlarged lymph nodes in your neck, underarm, and groin. Tell your doctor if you have felt any lumps or have any pain.

Nutritional assessment

You should meet with a nutrition expert before starting treatment. The nutritionist or dietician can assess the impact of the cancer on your health. Esophageal cancer can make you lose your appetite. You may also feel full after eating very little or have difficulty swallowing certain foods or liquids. These changes may have caused you to lose too much weight or make you feel weak and tired.

It is important that you receive adequate and sustained nutrition before you start treatment. You might receive food through a plastic tube that is placed through the skin of the abdomen into stomach or small intestine.

During and after treatment, your treatment team will monitor for weight loss and other signs you aren't getting enough nutrition.

For possible tests, see Guide 1.

Guide 1

Possible tests: Esophageal cancer

Medical history and physical exam. Screen for family history.

Upper GI endoscopy and biopsy

CT of chest and abdomen with oral and IV contrast. CT of pelvis with contrast as needed.

FDG-PET/CT evaluation (skull base to mid-thigh) if no evidence of metastatic disease

Complete blood count (CBC) and comprehensive chemistry profile

Endoscopic ultrasound (EUS) if no evidence of metastatic unresectable disease

Endoscopic resection (ER) is essential for the accurate staging of early-stage cancers (T1a or T1b). Early-stage cancers can best be diagnosed by ER.

Biopsy of metastatic disease as needed

Testing for MSI, MMR, and PD-L1 for metastatic disease or if suspected

HER2 for metastatic adenocarcinoma or if suspected

Bronchoscopy, if tumor is at or above the carina with no evidence of metastatic disease

Assign Siewert category

Nutritional assessment and counseling

Smoking cessation advice, counseling, and medicine as needed

Blood tests

Blood tests check for signs of disease and how well organs are working. They require a sample of your blood, which is removed through a needle placed into your vein.

Complete blood count

A complete blood count (CBC) measures the levels of red blood cells, white blood cells, and platelets in your blood. Your doctor will want to know if you have enough red blood cells to carry oxygen throughout your body, white blood cells to fight infection, and platelets to control bleeding.

Comprehensive chemistry profile

A comprehensive chemistry profile provides important information about how well your kidneys and liver are working, among other things. It is usually part of a comprehensive metabolic panel (CMP). A CMP measures 14 different substances in your blood.

CA 19-9 and CEA

Cancer antigen 19-9 (CA 19-9) and carcinoembryonic antigen (CEA) are occasionally made by tumors and can be detected in the blood. These tumor markers may be followed if elevated at the time of diagnosis.

Imaging tests

Imaging tests take pictures of the inside of your body. Images can be made with scanning machines or scoping tools. Imaging tests may show if the tumor involves any veins, arteries, and other organs. A radiologist, an expert in interpreting imaging tests, will write a report and send this report to your doctor. Your doctor will discuss the results with you.

CT scan

A computed tomography (CT or CAT) scan uses x-rays and computer technology to take pictures of the inside of the body. It takes many x-rays of the same body part from different angles. All the images are combined to make one detailed three-dimensional (3D) picture.

A CT scan of your chest, abdomen, and/ or pelvis may be one of the tests to look for cancer. In most cases, contrast will be used. Contrast material is used to improve the pictures of the inside of the body. Contrast materials are not dyes, but substances that help enhance and improve the images of several organs and structures in the body. It is used to make the pictures clearer. The contrast is not permanent and will leave the body in your urine immediately after the test.

Tell your doctors if you have had allergic reactions to contrast in the past. This is important. You might be given medicines, such as Benadryl® and prednisone, to avoid the effects of those allergies. Contrast might not be used if you have a serious allergy or if your kidneys aren't working well.

PET scan

A positron emission tomography (PET) scan uses a radioactive drug called a tracer. A tracer is a substance injected into a vein to see where cancer cells are in the body and if they are using sugar to grow. Cancer cells show up as bright spots on PET scans. However, not all tumors will appear on a PET scan. Also, not all bright spots are cancer. It is normal for the brain, heart, kidneys, and bladder to be bright on PET. When a PET scan is combined with CT, it is called a PET/CT scan. It may be done with one or two machines depending on the cancer center. An FDG-PET scan uses 18-fluorodeoxyglucose as its tracer.

Ultrasound

An ultrasound (US) uses high-energy sound waves to form pictures of the inside of the body. A probe will be pressed onto your abdomen. This is similar to the sonogram used for pregnancy. Ultrasound is painless and does not use x-rays, so it can be repeated as needed. It can show small areas of cancer that are near the surface of the body. Sometimes, an ultrasound or CT is used to guide a biopsy.

Endoscopy procedures

Some imaging tests use a thin, tube-shaped tool called a scope that is inserted into the body to take pictures. One end of the scope has a small light and camera lens to see inside your body. The image is sent to a television monitor. This will help guide your doctor in a biopsy, stent placement, or other tasks. The scope is guided into the body through a natural opening, such as the mouth, nose, or anus. It may also be inserted through a small surgical cut.

More than one type of scope may be used for imaging tests. The type of scope often used for esophageal cancer is called an endoscope. An endoscope is often guided into the body through the mouth. Endoscopy is an important tool in the diagnosis, staging, treatment, and care of those with esophageal cancer. Before an endoscopy, you will be given medicine to help you relax or sleep during the procedure.

Tests with scopes might include:

- Endoscopic ultrasound (EUS)
- Esophagogastroduodenoscopy (EGD)
- Bronchoscopy
- Laparoscopy

Endoscopic ultrasound

An ultrasound (US) uses high-energy sound waves to form pictures of the inside of the body. Endoscopic ultrasound (EUS) uses both imaging and an endoscope to see how deep the tumor has grown into the esophagus wall. Signs of cancer within lymph nodes and other nearby organs can also be detected. An EUS in an important part of cancer diagnosis and staging.

Upper endoscopy or EGD

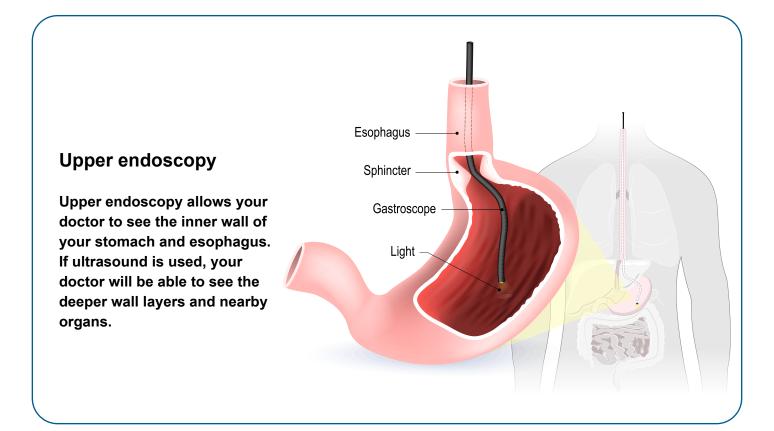
In an upper gastrointestinal (GI) endoscopy or esophagogastroduodenoscopy (EGD), a device is guided down the throat into the esophagus, stomach, and upper parts of the small intestine (duodenum). An EGD is used to inspect the lining of these organs and to look for any signs of cancer or other abnormalities such as dilated blood vessels or ulcers. Biopsies are usually performed during an EGD. A dilating (stretching) procedure can be also be done if a tumor is obstructing the esophagus. After the endoscopy, your throat may feel sore and you may feel some swelling.

Bronchoscopy

During a bronchoscopy, a device is inserted through the nose or mouth to examine the inside of your airway, including the trachea and bronchi. Bronchoscopy may be used to detect cancer from the esophagus or to perform some treatment procedures.

Laparoscopy

Laparoscopy (key-hole surgery) is a type of surgical procedure that inserts a camera (laparoscope) through a small cut in the abdomen. A tool can take tissue samples or remove tumors. Laparoscopy is done under general anesthesia. This is a controlled loss of wakefulness from drugs.



Biopsy

A biopsy removes a sample of tissue or fluid during an upper endoscopy. Several samples may be taken from the tumor, wall of your esophagus or stomach, and lymph nodes. The samples will be sent to a pathologist, an expert in examining cells under a microscope to find disease.

Other types of biopsies may include:

- Fine-needle aspiration (FNA) or core biopsy (CB) uses needles of different sizes to remove a sample of tissue or fluid. An ultrasound (US) may guide the FNA for diagnosis.
- Brushings or washings involve removing tumor or cell samples with a small brush at the end of an endoscope.
- Liquid biopsy uses a sample of blood for testing.

Biopsy of metastases

A metastasis is the spread of cancer to an area of the body such as the lung, liver, kidney, bone, or distant lymph nodes. A biopsy of the metastasis may be needed to confirm the presence of cancer. If there is more than one metastasis, each site may be biopsied. The type of biopsy used depends on the location of the suspected metastases and other factors.

Biomarker testing

A sample from a biopsy of your tumor may be tested to look for specific DNA (deoxyribonucleic acid) mutations/alterations, protein levels, or other molecular features. This information is used to choose the best treatment for you. It is sometimes called molecular testing or tumor profiling.

Biomarker testing includes tests of genes or their products (proteins). It identifies the presence or absence of mutations and certain proteins that might suggest treatment. Proteins are written like this: NTRK. Genes are written like this: *NTRK*.

Immunohistochemistry (IHC), fluorescence in situ hybridization (FISH), polymerase chain reaction (PCR), or next-generation sequencing (NGS) are types of tests used to look for biomarkers.

Testing for HER2 status, microsatellite (MS) status, programmed death ligand 1 (PD-L1) expression, and neurotrophic tropomyosin-related kinase (*NTRK*) gene fusions are important for the treatment and management of metastatic esophageal cancer.

Liquid biopsy

Some mutations can be found by testing circulating tumor DNA (ctDNA) in the blood. In a liquid biopsy, a sample of blood is taken to look for cancer cells or for pieces of DNA from tumor cells.

Those who have metastatic or advanced esophageal cancer and are unable to undergo a traditional biopsy might have a liquid biopsy. Sometimes, testing can quickly use up a tumor sample and a liquid biopsy might be an option in this case.

Tumor mutation burden

When there are 10 or more mutations per million base pairs of tumor DNA, it is called tumor mutational burden-high (TMB-H). TMB-H can be used to help predict response to cancer treatment using immune checkpoint inhibitors that target the proteins PD-1 or PD-L1.

Tumor mutation testing

A sample of your tumor or blood may be used to see if the cancer cells have any specific DNA mutations. This is a different type of DNA testing than the genetic testing for mutations you may have inherited from your parents. In tumor mutation testing, only the tumor is tested and not the rest of your body. Some mutations such as *NTRK* gene fusions can be targeted with specific therapies.

HER2

Human epidermal growth factor receptor 2 (HER2) is a protein involved in normal cell growth. It is found on the surface of all cells. When amounts are high, it causes cells to grow and divide. This is called HER2 positive, overexpression, or amplification.

There might be higher amounts of HER2 in your esophageal cancer. A sample of your tumor might be tested for HER2. If your tumor makes too much HER2, you might receive a targeted therapy called trastuzumab (Herceptin®) or a biosimilar, or trastuzumab deruxtecan (Enhertu®). A biosimilar is a drug that is very much like one that has been approved by the U.S. Food and Drug Administration (FDA). It must be used in the exact same way and at the same dose as the other drug.

MSI-H/dMMR

Microsatellites are short, repeated strings of DNA. When errors or defects occur, they are fixed by mismatch repair (MMR) proteins. Some cancers prevent these errors from being fixed. This is called microsatellite instability (MSI) or deficient mismatch repair (dMMR). When cancer cells have more than a normal number of microsatellites, it is called microsatellite instability-high (MSI-H). This is often due to dMMR genes.

NTRK gene fusions

In a tumor with an *NTRK* gene fusion, a piece of the *NTRK* gene and a piece of another gene fuse, or join. This activates the *NTRK* gene in a way that causes uncontrolled cell growth. Larotrectinib (Vitrakvi®) and entrectinib (RozlytrekTM) might be used to target advanced or metastatic cancer that is *NTRK* gene fusion-positive.

PD-L1

Programmed death-ligand 1 (PD-L1) is an immune protein. If this protein is expressed on the surface of cancer cells, it can cause your immune cells to ignore the cancer and suppress the anti-tumor immune response. If your cancer expresses the PD-L1 protein, you might have treatment that combines chemotherapy and a so-called checkpoint inhibitor therapy. This is designed to activate your immune system to better fight off the cancer cells.

FISH

Fluorescence in situ hybridization (FISH) is a method that involves special dyes called probes that attach to pieces of DNA, the genetic material in a person's cells.

Immunohistochemistry

Immunohistochemistry (IHC) is a special staining process that involves adding a chemical marker to cancer or immune cells. The cells are then studied using a microscope.

Next-generation sequencing

Next-generation sequencing (NGS) is a highthroughput method used to determine a portion of a person's DNA sequence. This method would only be used if enough tumor tissue remains after other biomarker testing has been completed.

PCR

A polymerase chain reaction (PCR) is a lab process that can make millions or billions of copies of your DNA (genetic information). PCR is very sensitive. It can find 1 abnormal cell among more than 100,000 normal cells. These copies called PCR product might be used for NGS.

Genetic risk testing

Genetic testing is done using blood or saliva (spitting into a cup). The goal is to look for gene mutations inherited from your genetic parents called germline mutations. Some mutations can put you at risk for more than one type of cancer. You can pass these genes on to your children. Also, family members might carry these mutations. Tell your doctor if there is a family history of cancer. Depending on your family history or other features of your cancer, your health care provider might refer you for hereditary genetic testing to learn more about your cancer. A genetic counselor will speak to you about the results.

Hereditary syndromes

Certain genetic (inherited) syndromes may put someone at risk for developing esophageal cancer. A syndrome is a group of signs or symptoms that occur together and suggest the presence of or risk for a disease. A genetic risk assessment will identify if you carry a cancer risk and if you may benefit from genetic testing, additional screening, or preventive interventions. Depending on the genetic risk assessment, you might undergo genetic testing and genetic counseling.

Hereditary syndromes most closely related to esophageal and EGJ cancers include:

- Esophageal cancer, tylosis with nonepidermolytic palmoplantar keratoderma (PPK), and Howel-Evans syndrome
- Familial Barrett esophagus (FBE)
- Bloom syndrome (BS)
- Fanconi anemia (FA)

Performance status

Performance status (PS) is a person's general level of fitness and ability to perform daily tasks. Your state of general health will be rated using a PS scale called ECOG (Eastern Cooperative Oncology Group) or the Karnofsky Performance Status (KPS).

ECOG PS

The ECOG PS scores range from 0 to 4.

- > PS 0 means you are fully active.
- PS 1 means you are still able to perform light to moderate activity.
- PS 2 means you can still care for yourself but are not active.
- PS 3 means you are limited to the chair or bed more than half of the time.

PS 4 means you need someone to care for you and are limited to a chair or bed.

In esophageal cancer, PS might be referred to as good or poor. Good PS is usually PS 0 or PS 1.

Karnofsky PS

The KPS score ranges from 0 to 100.

- 10 to 40 means you cannot care for yourself.
- > 50 to 70 means you cannot work and need some help to take care of yourself.
- 80 to 100 means you can carry out daily tasks.

Performance status

Performance status (PS) is a person's ability to perform daily tasks.



Key points

- Tests are used to find cancer, plan treatment, and check how well treatment is working.
- A medical history and physical exam inform your doctor about your overall health.
- Getting good nutrition is important before starting treatment. You should meet with a nutritionist before starting treatment.
- Blood tests check for signs of disease and how well organs are working.
- Imaging tests take pictures of the inside of your body. Images can be made with scanning machines or scoping tools.
- A biopsy removes a sample of tissue or fluid during an endoscopy.
- A sample from a biopsy of your tumor may be tested to look for specific DNA (deoxyribonucleic acid) mutations, protein expression levels, or other molecular features. Some mutations and proteins can be targeted with specific therapies.
- Genetic testing might be done to look for gene mutations inherited from your birth parents called germline mutations.
- Performance status (PS) is a person's general level of fitness and ability to perform daily tasks.
- Online portals are a great way to access your test results.

Imaging and other
tests are not
always accurate. A
multidisciplinary
team should review
the results.

3 Staging

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Cancer staging is used to make treatment decisions. It describes the size and location of the tumor, if the tumor has grown through the layers of the esophagus wall, and if cancer has spread to lymph nodes, organs, or other parts of the body. This chapter explains esophageal cancer stages.

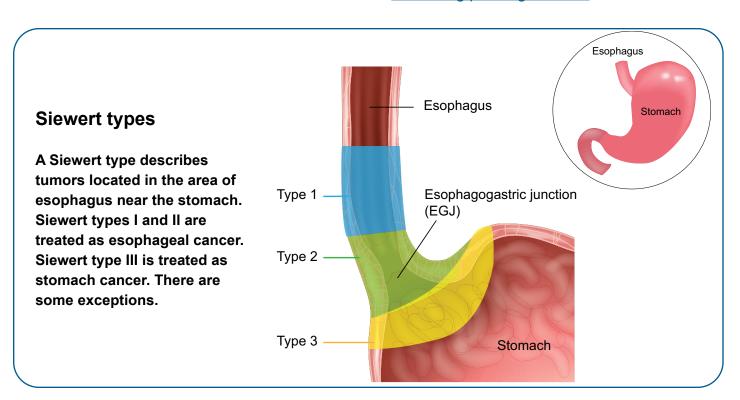
Siewert types

Siewert types are helpful in telling the difference between a stomach and esophageal cancer. A Siewert type describes tumors located in the area of esophagus near the stomach. This area is called the esophagogastric junction (EGJ) and the cardia of the stomach. The EGJ is found just beneath the diaphragm.

EGJ tumors located within 2 cm of the stomach (Siewert Types I and II) are staged as esophageal adenocarcinoma. Cancers whose center is more than 2 cm below the EGJ, even if the EGJ is involved, will be staged using the stomach cancer TNM and stage groups.

- Siewert Type I The tumor center located within 1 to 5 cm above the EGJ and stomach cardia
- Siewert Type II The tumor center is located within 1 cm above and 2 cm below the EGJ.
- Siewert Type III The tumor center is located between 2 to 5 cm below the EGJ, which infiltrates the EGJ and the lower esophagus from below.

Siewert Type III are considered and treated as stomach (gastric) cancers. For more information, see NCCN Guidelines for Patients®: Stomach Cancer, available at NCCN.org/patientquidelines.



Staging

Esophageal cancer staging is often done twice.

- Clinical stage (c) is the rating given before any treatment. It is based on a physical exam, biopsy, and imaging tests. An example might look like cN2 or cM1.
- Pathologic stage (p) or surgical stage is determined by examining tissue removed during surgery. An example might be pN2. If you are given drug therapy before surgery, then the stage might look like ypT3.

A cancer stage is a way to describe the extent of the cancer at the time you are first diagnosed. The American Joint Committee on Cancer (AJCC) created a staging system to determine how much cancer is in your body, where it is located, and what subtype you have. AJCC is just one type of staging system.

Staging is based on a combination of information to reach a final numbered stage. Often, not all information is available at the initial evaluation. More information can be gathered as treatment begins. Doctors may explain your cancer stage in different ways than described next.

TNM scores

The tumor, node, metastasis (TNM) system is used to stage esophageal cancer. In this system, the letters T, N, and M describe different areas of cancer growth. Based on imaging and pathology results, your doctor will assign a score or number to each letter. The higher the number, the larger the tumor or the more the cancer has spread to lymph nodes or

other organs. These scores will be combined to assign the cancer a stage. A TNM example might look like this: T1N0M0 or T1, N0, M0.

The TNM letters represent the following:

- T (tumor) Depth and spread of the main (primary) tumor in the wall of esophagus
- N (node) If cancer has spread to nearby (regional) lymph nodes
- M (metastasis) If cancer has spread to distant parts of the body or metastasized

A specific diagnosis of esophageal squamous cell carcinoma (SCC) or adenocarcinoma is needed for staging and treatment purposes. Mixed adenosquamous carcinomas and carcinomas are staged using the TNM system for SCC.

Grade

Grade describes how abnormal the tumor cells look under a microscope (called histology). Higher-grade cancers tend to grow and spread faster than lower-grade cancers. GX means the grade can't be determined, followed by G1, G2, and G3. Well differentiated (G1) means the cancer cells look similar to normal cells. Poorly differentiated (G3) means the cancer cells look very different compared to normal cells. G3 is the highest grade for esophageal cancers.

- GX Grade cannot be determined
- > **G1** Well differentiated
- > **G2** Moderately differentiated
- G3 Poorly differentiated or undifferentiated

Numbered stages

Numbered stages are based on TNM scores. Stages range from stage 1 to stage 4, with 4 being the most advanced. Doctors write these stages as stage I, stage II, stage III, and stage IV. For example, stage 1 might be T1, N0, M0.

Other terms might be used instead of numbered cancer stages. This book will use the following terms to describe esophageal cancer:

- Resectable Tumor can be removed completely with surgery.
- Unresectable Tumor cannot be removed with surgery. It might involve nearby veins and arteries making it unsafe to remove.
- Locoregional or locally advanced Tumor might be any size and could be in any layer of the stomach. Cancer might be in nearby lymph nodes, organs, and tissues.
- Metastatic Cancer that has spread to other parts of the body, including distant lymph nodes. The most common metastatic sites are the liver, distant lymph nodes, lung, bone, and brain.

T = Tumor

A tumor can grow through the layers of the esophagus wall and into nearby structures.

- Tis High-grade dysplasia (HGD) is cancer that has not grown beyond the epithelium
- T1 Tumor invades the lamina propria, muscularis mucosa, or submucosa
 - T1a Tumor invades the lamina propria or muscularis mucosa
 - T1b Tumor invades the submucosa.
- > **T2** Tumor invades the muscle layer called muscularis propria.
- > **T3** Tumor invades the adventitia.
- T4 Tumor has grown all the way through the wall of the esophagus into nearby structures.
 - T4a Tumor has grown into nearby structures, such as the pleura, pericardium, azygos vein, diaphragm, or peritoneum. The pleura covers the lungs and lines the interior wall of the chest cavity. The pericardium is the sac that surrounds the heart. The azygos vein is a blood vessel that carries blood from the back walls of the chest and abdomen to the heart. The peritoneum lines the abdominal wall and covers most of the organs in the abdomen.
 - T4b Tumor has grown into nearby structures, such as the aorta, spinal column, heart, or airway (trachea).

Tis and T1 tumors are often considered earlystage cancer. Esophageal cancers are rarely found this early.

N = Regional lymph node

There are hundreds of lymph nodes throughout your body. They work as filters to help fight infection and remove harmful things from your body. Regional lymph nodes are found near the esophagus. Cancer found in a regional lymph node is called a lymph node metastasis. This is different than a distant metastasis, which is found farther from the main tumor in the esophagus.

Lymph, a clear fluid containing cells that help fight infections and other diseases, drains through channels from the wall of the esophagus into lymphatic vessels in the mucosa and submucosa. From here, lymph drains into lymph nodes outside and along the esophagus. These lymph nodes drain into the thoracic duct. It is possible for cancerous cells to travel through lymph to other parts of the body.

Since the lymphatic network is concentrated in the submucosa, metastases can be found in early-stage cancers.

Lymph nodes must be removed to confirm cancer. The removal of lymph nodes is called lymph node dissection (lymphadenectomy). It might be referred to as a nodal dissection. At least 15 lymph nodes should be removed.

- N0 No cancer found in nearby lymph nodes.
- ▶ N1 1 or 2 nearby nodes have cancer.
- ▶ N2 3 to 6 nearby nodes have cancer.
- N3 7 or more nearby lymph nodes have cancer.

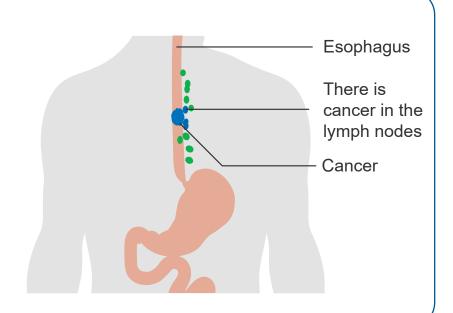
M = Metastasis

Cancer that has spread to distant parts of the body is shown as M1. The most common metastatic sites are the liver, distant lymph nodes, lung, bone, and brain.

Regional lymph nodes

A tumor can be found anywhere in the esophagus. Cancer may be found in lymph nodes along the esophagus.

https://commons.wikimedia.org/wiki/File:Diagram_showing_oesophageal_cancer_in_the_lymph_nodes_(N_staging)_CRUK_174.svg



Clinical stage

The clinical (before surgery) stage is based on the endoscopic ultrasound (EUS) and other imaging or biopsy results. These tests are done before any treatment as part of an initial diagnosis. Surgery is needed to know exactly how much cancer is in the body. Clinical stages for esophageal squamous cell carcinomas can be found in Guide 2.

Clinical stages for esophageal adenocarcinomas can be found in Guide 3.

Guide 2 Squamous cell carcinoma stages: Clinical (c)		
Stage 0	• Tis, N0, M0	
Stage 1	• T1, N0 or N1, M0	
Stage 2A	T2, N0 or N1, M0T3, N0, M0	
Stage 3	• T3, N1, M0 • T1 to T3, N2, M0	
Stage 4A	• T4, N0 to N2, M0 • Any T, N3, M0	
Stage 4B	• Any T, Any N, M1	

Guide 3 Adenocarcinoma stages: Clinical (c)		
Stage 0	• Tis, N0, M0	
Stage 1	• T1, N0, M0	
Stage 2A	• T1, N1, M0	
Stage 2B	• T2, N0, M0	
Stage 3	T2, N1, M0T3, N0 or N1, M0T4a, N0 or N1, M0	
Stage 4A	T1 to T4a, N2, M0T4b, Any N, M0Any T, N3, M0	
Stage 4B	• Any T, Any N, M1	

Pathologic stage

The pathologic (after surgery) stage is based on information gained after surgery to remove part or all of the esophagus and nearby lymph nodes. This gives a more accurate picture of how far the cancer has spread and is used to determine your treatment options after surgery. The removal of tumor tissue and nearby lymph nodes is an important part of pathologic staging.

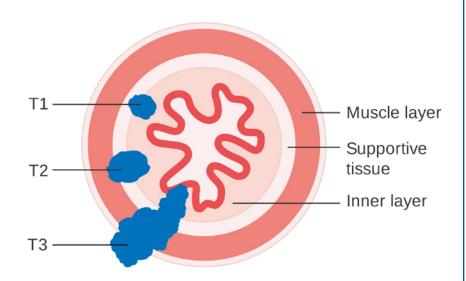
Pathologic stages for esophageal squamous cell carcinomas can be found in Guide 4.

Pathologic stages for esophageal adenocarcinomas can be found in Guide 5.

Esophageal cancer

Esophageal cancer starts in the cells that line the esophagus. Cancer grows through and along the layers of the esophagus wall.

https://commons.wikimedia.org/wiki/File:Diagram_showing_T1,T2_and_T3_stages_of_oesophageal_cancer_CRUK_277.svg



Guide 4 Pathologic (pTNM) stage with grade (G) and location: Squamous cell carcinoma		
Stage 0	• Tis, N0, M0, any location	
Stage 1A	• T1a, N0, M0, GX or G1, any location	
Stage1B	 T1a, N0, M0, G2 or G3, any location T1b, N0, M0, GX or G1 or G2, any location T2, N0, M0, G1, any location 	
Stage 2A	 T2, N0, M0, GX or G2 or G3, any location T3, N0, M0, G1 or G2 or G3, lower esophagus T3, N0, M0, G1, upper or middle esophagus 	
Stage 2B	 T3, N0, M0, G2 or G3, upper or middle esophagus T3, N0, M0, GX, lower or upper or middle esophagus T3, N0, M0, any grade, location unknown T1, N1, M0, any grade, any location 	
Stage 3A	 T1, N2, M0, any grade, any location T2, N1, M0, any grade, any location 	
Stage 3B	 T2, N2, M0, any grade, any location T3, N1 or N2, M0, any grade, any location T4b, N0 or N1, M0, any grade, any location 	
Stage 4A	 T4a, N2, M0, any grade, any location T4b, N0 or N1 or N2, M0, any grade, any location Any T, N3, M0, any grade, any location 	
Stage 4B	Any T, Any N, M1, any grade, any location	

Guide 5 Pathologic (pTNM) stage and grade (G): Adenocarcinoma		
Stage 0	• Tis, N0, M0	
Stage 1A	• T1a, N0, M0, GX or G1	
Stage1B	• T1a, N0, M0, G2 • T1b, N0, M0, GX or G1 or G2	
Stage1C	• T1, N0, M0, G3 • T2, N0, M0, G1 or G2	
Stage 2A	• T2, N0, M0, GX or G3	
Stage 2B	• T1, N1, M0, any grade • T3, N0, M0, any grade	
Stage 3A	• T1, N2, M0, any grade • T2, N1, M0, any grade	
Stage 3B	 T2, N2, M0, any grade T3, N1 or N2, M0, any grade T4b, N0 or N1, M0, any grade 	
Stage 4A	 T4a, N2, M0, any grade T4b, N0 or N1 or N2, M0, any grade Any T, N3, M0, any grade 	
Stage 4B	Any T, Any N, M1, any grade	

Key points

- Staging is used to make treatment decisions. Staging describes how much cancer is in your body, where it is located, and what subtype you have.
- The tumor, node, metastasis (TNM) system is used to stage esophageal cancer.
- Esophageal cancer staging is often done twice, before and after surgery.
- The clinical stage (c) of esophageal cancer is based on the results of testing before endoscopic resection or surgery. It is written as cTNM.
- The pathologic stage (p) of esophageal cancer is based on the results of endoscopic resection or surgery. It is written as pTNM.
- Regional lymph nodes are found near the esophagus.
- Cancer that has spread to distant parts of the body is metastatic cancer.
- The most common metastatic sites are the liver, distant lymph nodes, lung, bone, and brain.
- Doctors may explain your cancer stage in different ways to make it less confusing.



Let us know what you think!

Please take a moment to complete an online survey about the NCCN Guidelines for Patients.

NCCN.org/patients/response

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There is more than one treatment for esophageal cancer. This chapter describes treatment options and what to expect. Together, you and your doctor will choose a treatment plan that is best for you.

Treatment team

Those with esophageal cancer should seek treatment at experienced cancer centers.

Treating esophageal cancer takes a team approach. Treatment decisions should involve a multidisciplinary team (MDT). An MDT is a team of doctors, health care workers, and social care professionals from different professional backgrounds who have knowledge (expertise) and experience with your type of cancer. This team is united in the planning and implementing of your treatment. Ask who will coordinate your care.

Some members of your care team will be with you throughout cancer treatment, while others will only be there for parts of it. Get to know your care team and help them get to know you.

Depending on your diagnosis, your team might include the following specialists:

- A gastroenterologist is an expert in diseases of the digestive tract.
- A pathologist analyzes the cells, tissues, and organs removed during a biopsy or surgery and provides cancer diagnosis, staging, and information about biomarker testing.

Get to know your care team and help them get to know you.

- A diagnostic radiologist interprets the results of x-rays and other imaging tests.
- An interventional radiologist performs needle biopsies, endoscopies, and ablation procedures, and places ports for treatment. An endoscopist is trained in the use of an endoscope.
- A surgical oncologist performs operations to remove cancer, including the esophagus.
- A thoracic surgical oncologist performs operations to remove organs that involve the chest, including the esophagus.
- A medical oncologist treats cancer in adults using systemic therapy.
- A radiation oncologist prescribes and plans radiation therapy to treat cancer.
- An anesthesiologist gives anesthesia, a medicine so you do not feel pain during surgery or procedures.
- Palliative care nurses and advanced practice providers help provide an extra layer of support with your cancer-related symptoms.

- 4
- Residents and fellows are doctors who are continuing their training, some to become specialists in a certain field of medicine.
- Oncology nurses provide your handson care, like giving systemic therapy, managing your care, answering questions, and helping you cope with side effects. Sometimes, these experts are called nurse navigators.
- Nutritionists and dietitians can provide guidance on what foods are most suitable for your condition.
- Psychologists and psychiatrists are mental health experts who can help manage issues such as depression, anxiety, or other mental health conditions that can affect how you feel.
- Social workers help people solve and cope with problems in their everyday lives.
- A Research team helps to collect research data if you are in a clinical trial

Your physical, mental, and emotional wellbeing are important. You know yourself better than anyone. Help other team members understand:

- How you feel
- What you need
- What is working and what is not

Keep a list of names and contact information for each member of your team. This will make it easier for you and anyone involved in your care to know whom to contact with questions or concerns.



If you smoke or vape

If you smoke tobacco or use e-cigarettes, it is very important to quit. Smoking can limit how well cancer treatment works. Smoking greatly increases your chances of having side effects during and after surgery. It also increases your chances of developing other cancers.

Nicotine is the chemical in tobacco that makes you want to keep smoking. Nicotine withdrawal is challenging for most smokers. The stress of having cancer may make it even harder to quit. If you smoke, ask your doctor about counseling and medicines to help you quit.

For online support, try these websites:

- SmokeFree.gov
- <u>BeTobaccoFree.gov</u>
- CDC.gov/tobacco

Endoscopic therapy

Some treatments use a thin, tube-shaped tool called a scope that is inserted into the body to take pictures. One end of the scope has a small light and camera lens to see inside your body. The image is sent to a television monitor. This will help guide your doctor in a biopsy, treatment, or other tasks. An endoscope is often guided into the body through the mouth.

The goal of endoscopic therapy is the remove or destroy early-stage disease and Barrett esophagus (BE). Endoscopic resection and/or ablation is preferred for early-stage cancer.

Endoscopic resection

For those with early-stage (T1a or T1b) esophageal cancer or Barrett esophagus, endoscopic mucosal resection (EMR) or endoscopic submucosal dissection (ESD) is essential for accurate staging and diagnosis. Early-stage esophageal cancer has not grown beyond the first layer (mucosa) of the esophageal wall. The tumor is often very small (2 centimeters or less) and is not in any lymph nodes.

Endoscopic mucosal resection

Endoscopic mucosal resection (EMR) is a procedure to remove pre-cancerous, early-stage cancer or other abnormal tissues (lesions) from the esophageal wall. During EMR, the endoscope is passed down your throat to reach the lesion or tumor in your esophagus. The lesion can be removed through suction or cutting it away. Talk to your doctor to learn more.

Endoscopic submucosal dissection

Endoscopic submucosal dissection (ESD) uses an endoscope to locate the tumor in the esophageal wall. A tool is inserted through the endoscope that injects fluid between the tumor and layer of the esophageal wall. Then a tool lifts and cuts away the tumor from the esophageal wall.

Ablation

Ablation uses extreme cold or extreme heat, radio waves, microwaves, or chemicals to destroy cancer cells. It can destroy small tumors and metastases with little harm to nearby tissue. You might have multiple treatments to destroy the tumor or metastasis. Ablation might be used if you are not healthy enough for surgery.

There are many types of ablation used to destroy cancer cells. Those most commonly used to treat an esophageal tumor include:

- Cryotherapy or cryosurgery kills cancer cells by freezing them with a very cold substance.
- Radiofrequency ablation (RFA) kills cancer cells by heating them with highenergy radio waves.
- Photodynamic therapy (PTD) uses drugs that become active when exposed to light. These activated drugs may kill cancer cells.

Surgery

Surgery is an operation or procedure to remove cancer from the body. Often, surgery is the main or primary treatment to remove the cancer from the esophagus. This is only one part of a treatment plan. Surgery can also provide supportive care by easing pain or discomfort. This is called palliative surgery.

When preparing for surgery, seek the opinion of an experienced surgeon. The surgeon should be an expert in performing your type of surgery. Surgery for esophageal cancer should be done at a high-volume center that does at least 15 to 20 esophageal surgeries each year. Hospitals that perform many surgeries often have better results. You can ask for a referral to a hospital or cancer center that has experience in treating your type of cancer.

The removal of the cancer through surgery can be accomplished in different ways depending on the specific circumstances, such as the size and location of the tumor, and if there is cancer in any surrounding organs and tissues. Surgery is based on the safest and best way to remove the cancer.

Open surgery

Open surgery, including thoracotomy and laparotomy, removes tissue through surgical incisions. These larger incisions let your doctor directly view and access the tumor in your esophagus to remove it. Open surgery may take several hours or longer. After the surgery, you will need to stay in the hospital for several days or longer to recover.

Surgery and
endoscopic procedures
are best performed
in high-volume
esophageal centers by
experienced surgeons
and endoscopists.

Minimally invasive surgery

Minimally invasive surgery (key-hole surgery) uses several small incisions. Small tools are inserted through each incision to perform the surgery. One of the tools, called a videooscope, is a long tube with a video camera at the end. The camera lets your doctor see your esophagus and other tissues inside your chest and abdomen. Other tools are used to remove the tumor. Minimally invasive surgery, also called thoracoscopic and laparoscopic surgery, can also be done using robotic arms to control the surgical tools. This is called robot-assisted laparoscopic surgery.

Tumor resection

Imaging tests will be ordered to see if your cancer is resectable (can be removed completely by surgery) or unresectable (cannot be removed completely by surgery). Sometimes, imaging and scoping tests cannot clearly show one way or the other.

Goal of surgery

The goal of surgery or tumor resection is to remove all of the cancer. To do so, the tumor is removed along with some normal-looking tissue around its edge called the surgical margin. The surgical margin may look normal, but cancerous cells may be found when viewed under a microscope by a pathologist. A clear or negative margin (R0) is when no cancer cells are found in the tissue around the edge of the tumor. In a positive margin, cancer cells are found in normal-looking tissue around the tumor.

You may receive treatment before surgery called neoadjuvant or preoperative therapy. Neoadjuvant therapy will help reduce the size of the tumor and the amount of cancer in the body.

Surgical margins

The goal of surgery is a cancer-free surgical margin with reasonable functional outcome. This is your ability to perform daily tasks of living. After surgery, you may receive treatment such as radiation, chemoradiation, or systemic therapy to kill any remaining cancer cells.

- In a clear or negative margin (R0), no cancerous cells are found in the tissue around the edge of the tumor.
- In an R1 positive margin, the surgeon removes all the visible tumor, but the microscopic margins are still positive for tumor cells. Despite best efforts this can happen.
- In an R2 positive margin, the surgeon is unable to remove all the visible tumor or there is metastatic disease (M1).

A negative margin (R0) is the best result. Your surgeon will look carefully for cancer not only along the surgical margin, but in other nearby areas. An intraoperative pathology consultation is often used by surgeons. This includes inspecting the resected esophageal tissue for cancer location and distance to margins, examining by microscope frozen sections of proximal and distal margins, and examining by microscope for possible intra-abdominal metastasis such as liver or peritoneal metastasis. Intraoperative pathology consultation serves an important role in guiding the surgery.

Despite best efforts, it is not always possible to find all of the cancer. Sometimes, surgeons can't safely remove the tumor with a cancerfree margin.

You might have more than one surgery. You might also have a wound drain to prevent fluid from collecting in the body after surgery. These drains are usually removed a few days after surgery.

Esophagectomy

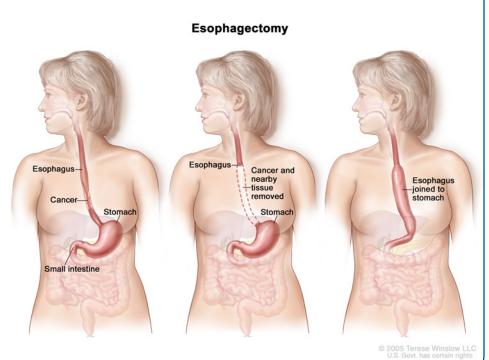
An esophagectomy removes part, most, or all of the esophagus. An esophagectomy is a big operation, even when it uses a minimally invasive approach. The surgery crosses two or three body cavities — abdomen, chest, and neck — and usually takes 4 to 6 hours. Therefore, eating a healthy diet and exercising before surgery is important. When you are in your best possible physical condition before surgery, it is more likely that you will experience a quicker and easier recovery after surgery.

Types of esophagectomy include:

- A transhiatal esophagectomy (THE) is performed through creating an opening on the neck and abdomen.
- A transthoracic esophagectomy (TTE) is performed through creating an opening on the abdomen and the chest, and occasionally in the neck as well.
- Minimally invasive laparoscopic and thoracoscopic or robotic surgical approaches

Before and after an esophagectomy

These images show before and after an esophagectomy. In this case, a large portion of the esophagus has been removed. The stomach has been pulled up and joined with the esophagus. Sometimes, instead of the stomach, a piece of the small intestine or colon is used to replace the missing part of the esophagus.



Reconstruction conduits

A conduit is a tube-shaped tissue, such as the stomach, the colon, or small intestine, used to replace or reconstruct the part of the esophagus removed during an esophagectomy. A stomach (gastric) conduit is preferred and is the most commonly used conduit for esophageal reconstruction. The stomach is a large, durable organ with abundant blood supply.

The type of esophageal resection is dictated by the location of the tumor, the available choices for conduit, and the surgeon's experience and preference, as well as your preference.

Other procedures

Esophagogastrectomy

In an esophagogastrectomy, the top part of the stomach and distal part of the esophagus are removed. A nutritionist or dietician provides guidance on what foods are most suitable for you before and after this surgery.

Gastrectomy

A gastrectomy removes all or part of the stomach. Surgery that removes part of the stomach is called gastric resection. Gastric resection should include the removal of regional lymph nodes (lymphadenectomy). For more information on a gastrectomy, see NCCN Guidelines for Patients: Stomach Cancer, available at NCCN.org/patientguidelines.

Lymph node dissection

The removal of lymph nodes or groups of lymph nodes is called lymph node or nodal dissection. It might be referred to as a lymphadenectomy. At least 15 regional nodes should be removed and tested for cancer.

G-tube

A gastrostomy tube (G-tube) is a soft, plastic tube placed through the skin of the abdomen directly into the stomach. It allows air and fluid to leave the stomach and can be used to give medicines and fluids, including liquid food. Giving food through a gastrostomy tube is a type of enteral nutrition. It is also called a percutaneous endoscopic gastrostomy (PEG) tube.

J-tube

A jejunostomy tube (J-tube) is a soft, plastic tube placed through the skin of the abdomen into the midsection of the small intestine. The tube delivers food and medicine until you are healthy enough to eat by mouth.

Systemic therapy

Systemic therapy works throughout the body. Types include chemotherapy, targeted therapy, and immunotherapy. Systemic therapy might be used alone or with other therapies. Goals of systemic therapy should be discussed before starting treatment. Your wishes about treatment are important.

- When systemic therapy or chemoradiation is given <u>before surgery</u>, it is called neoadjuvant or **preoperative therapy**.
- When systemic therapy is given before and after surgery, it is called perioperative therapy.
- When systemic therapy or radiation therapy is given <u>after surgery</u>, it is called adjuvant or **postoperative therapy**.
- When systemic therapy is given for advanced disease, it may be called palliative therapy.

Warnings!

You might be asked to stop taking or avoid certain herbal supplements when on a systemic therapy. Some supplements can affect the ability of a drug to do its job. This is called a drug interaction. It is critical to speak with your care team about any supplements you may be taking.

Some examples include:

- Turmeric
- Gingko biloba
- Green tea extract
- > St. John's Wort

Certain medicines can also affect the ability of a drug to do its job. Antacids, heart medicine, and antidepressants are just some of the medicines that might interact with a systemic therapy. Therefore, it is important to tell your doctor about any medications, vitamins, over-the-counter (OTC) drugs, herbals, or supplements you are taking. Bring a list with you to every visit.

Chemotherapy

Chemotherapy kills fast-growing cells throughout the body, including cancer cells and some normal cells. When chemotherapies are combined, it is called multi-agent or combination chemotherapy. Chemotherapy might be used before and after surgery. It might be used with radiation (called chemoradiation).

Some examples of chemotherapies include:

- Capecitabine (Xeloda®)
- Carboplatin
- Cisplatin
- Docetaxel (Taxotere®)
- > Fluorouracil
- Irinotecan (Camptosar[®])
- Oxaliplatin (Eloxatin®)
- Paclitaxel (Taxol[®])

Chemoradiation

Treatment that combines chemotherapy with radiation therapy is called chemoradiation. Chemotherapy may improve how well radiation works, and that is why they are sometimes used together. Chemoradiation may be used to control symptoms caused by a tumor, to shrink the tumor before surgery, to prevent the return of cancer after surgery, or as the primary (main) treatment for the tumor.

Targeted therapy

Targeted therapy focuses on specific or unique features of cancer cells. Targeted therapies seek out how cancer cells grow, divide, and move in the body. These drugs stop or inhibit the action of molecules that help cancer cells grow and/or survive.

Some examples of targeted therapies include:

- Ramucirumab (Cyramza®) targets VEGF receptors.
- Entrectinib (Rozlytrek®) and larotrectinib (Vitrakvi®) are examples of TRK inhibitors used for NTRK gene fusion-positive tumors.
- Trastuzumab (Herceptin®), trastuzumab deruxtecan (Enhertu®), or a biosimilar target HER2 overexpression.

It is important to tell your care team about all side effects so they can be managed.

Immunotherapy

Immunotherapy increases the activity of your immune system. By doing so, it improves your body's ability to find and destroy cancer cells. Immunotherapy can be given alone or with other types of treatment.

Examples of immunotherapies:

- Nivolumab (Opdivo®) targets PD-L1.
- Pembrolizumab (Keytruda®) targets
 MSI-H or dMMR tumors, or high tumor
 mutation burden (TMB-H).
- Dostarlimab-gxly (Jemplerli) targets MSI-H or dMMR tumors.

For more information on checkpoint inhibitors and immunotherapy side effects, see *NCCN Guidelines for Patients: Immunotherapy Side Effects*, available at NCCN.org/ patientquidelines.

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Radiation therapy

Radiation therapy (RT) uses high-energy radiation from x-rays, photons, protons, electrons, and other sources to kill cancer cells and shrink tumors. RT can be given alone or with other treatments. Treatment may focus on individual tumors, a small area/region of the body, or specific lymph nodes. RT can be used as the primary treatment for esophageal cancer. RT can also be given before or after surgery to treat or slow the growth of cancer, especially if the surgical margins have cancer cells. RT may also be used as supportive care or palliative care to help ease pain or discomfort caused by cancer, or to control bleeding caused by a tumor.

There are 2 main types of radiation treatment:

- External beam radiation therapy (EBRT) uses a machine outside of the body to aim radiation at the tumor(s) or areas of the body.
- Internal radiation is placed inside the body as a solid like seeds. This is called brachytherapy.

A four-dimensional (4D) CT scan might be used to plan RT. A 4D-CT records multiple images over time. It allows playback of the scan as a video, so that internal movement can be tracked and observed.



Finding a clinical trial

In the United States

NCCN Cancer Centers
NCCN.org/cancercenters

The National Cancer Institute (NCI)

<u>cancer.gov/about-cancer/treatment/clini-</u> <u>cal-trials/search</u>

Worldwide

The U.S. National Library of Medicine (NLM)

clinicaltrials.gov/

Need help finding a clinical trial?

NCI's Cancer Information Service (CIS)

1.800.4.CANCER (1.800.422.6237) <u>cancer.gov/contact</u>

Clinical trials

A clinical trial is a type of medical research study. After being developed and tested in a laboratory, potential new ways of fighting cancer need to be studied in people. If found to be safe and effective in a clinical trial, a drug, device, or treatment approach may be approved by the U.S. Food and Drug Administration (FDA).

Everyone with cancer should carefully consider all of the treatment options available for their cancer type, including standard treatments and clinical trials. Talk to your doctor about whether a clinical trial may make sense for you.

Phases

Most cancer clinical trials focus on treatment. Treatment trials are done in phases.

- Phase I trials study the dose, safety, and side effects of an investigational drug or treatment approach. They also look for early signs that the drug or approach is helpful.
- Phase II trials study how well the drug or approach works against a specific type of cancer.
- Phase III trials test the drug or approach against a standard treatment. If the results are good, it may be approved by the FDA.
- Phase IV trials study the long-term safety and benefit of an FDA-approved treatment.

Who can enroll?

Every clinical trial has rules for joining, called eligibility criteria. The rules may be about age, cancer type and stage, treatment history, or general health. These requirements ensure that participants are alike in specific ways and that the trial is as safe as possible for the participants.

Informed consent

Clinical trials are managed by a group of experts called a research team. The research team will review the study with you in detail, including its purpose and the risks and benefits of joining. All of this information is also provided in an informed consent form. Read the form carefully and ask questions before signing it. Take time to discuss with family, friends, or others whom you trust. Keep in mind that you can leave and seek treatment outside of the clinical trial at any time.

Start the conversation

Don't wait for your doctor to bring up clinical trials. Start the conversation and learn about all of your treatment options. If you find a study that you may be eligible for, ask your treatment team if you meet the requirements. If you have already started standard treatment you may not be eligible for certain clinical trials. Try not to be discouraged if you cannot join. New clinical trials are always becoming available.

Frequently asked questions

There are many myths and misconceptions surrounding clinical trials. The possible benefits and risks are not well understood by many with cancer.

Will I get a placebo?

Placebos (inactive versions of real medicines) are almost never used alone in cancer clinical trials. It is common to receive either a placebo with a standard treatment, or a new drug with a standard treatment. You will be informed, verbally and in writing, if a placebo is part of a clinical trial before you enroll.

Are clinical trials free?

There is no fee to enroll in a clinical trial. The study sponsor pays for research-related costs, including the study drug. You may, however, have costs indirectly related to the trial, such as the cost of transportation or child care due to extra appointments. During the trial, you will continue to receive standard cancer care. This care is billed to—and often covered by—insurance. You are responsible for copays and any costs for this care that are not covered by your insurance.

Supportive care

Supportive care is health care given during all cancer stages. It aims to prevent, reduce, and relieve suffering, and to improve quality of life. Supportive care might include pain relief (palliative care), emotional or spiritual support, financial aid, or family counseling. Tell your care team how you are feeling and about any side effects so they can be managed. Best supportive care, supportive care, and palliative care are often used interchangeably.

It is very important to take care of yourself by eating well, drinking plenty of fluids, exercising, and doing things that make you feel energized. Strength is needed to sustain you during treatment.

After surgery, you may have problems such as getting full too fast, heartburn and reflux, stomach discomfort, bloating, nausea, diarrhea, or dumping syndrome. Some people also experience discomfort or "sticking" when swallowing foods. Nutrition plays an important role in managing these symptoms.

Advance care planning

Advance care planning is making decisions now about the care you would want to receive if you become unable to speak for yourself. Advance care planning is for everyone, not just for those who are very sick. It is a way to ensure your wishes are understood and respected.

Advance care planning starts with an honest discussion with your doctor. Ask your doctor about the course your cancer will take called a prognosis. Find out what you might expect if your cancer spreads or worsens. Discuss the medicines or therapies that will give you the

best quality of life. Include family and friends in your advance care planning. Make your wishes clear. It is important that everyone understands what you want.

You don't have to know the exact details of your prognosis. Just having a general idea will help with planning. With this information, you can decide if there is a point where you might want to stop cancer treatment. You can also decide what treatments you would want for symptom relief, such as radiation therapy, surgery, or medicine. You can change your advance care plan at any time. It might be helpful to have this talk with your doctor and/

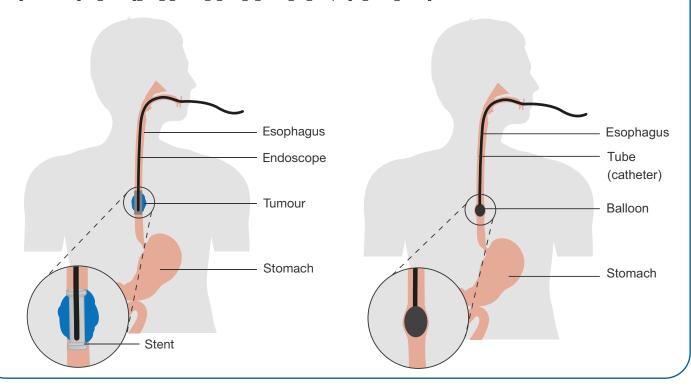
or friends and family more than once during your treatment. What you want today might change tomorrow. Making your wishes clear will ensure everyone knows what you want.

Bleeding

Bleeding is common in those with esophageal cancer. It may be caused by the tumor or a result of treatment. An endoscopic treatment, radiation therapy (if not done before), and an angiography with embolization might be used to treat (stop) bleeding.

Blocked esophagus An esophageal stent is a tube that widens the esophagus so food can pass into the stomach (image on left). Special expanding (dilating) balloons or bougies can relieve a blockage or a narrowing of the esophagus (image on right).

 $Left: https://commons.wikimedia.org/wiki/File: Diagram_showing_an_oesophageal_stent_being_put_in_CRUK_495.svg. \ Right: https://commons.wikimedia.org/wiki/File: Diagram_showing_how_a_balloon_is_used_to_stretch_the_oesophagus_CRUK_492.svg$



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An angiography might be done when a blood vessel has narrowed or suddenly becomes blocked and does not allow blood to flow. In an angiography, a catheter (thin plastic tube) is inserted into an artery through a small incision in the skin and guided to the area with the use of x-rays. A contrast material is injected through the tube and x-ray images produce a picture of the blood vessel called an angiogram.

Embolization is the process of blocking blood flow through a blood vessel. This is performed by placing various materials through the angiography catheter while it is inside the blood vessel. The material can be a coil, small beads, or liquid medicine that causes the blood to clot and block the flow of blood.

Blocked esophagus

A tumor may block the esophagus, the esophagogastric junction (EGJ), or the stomach. An esophageal stent is a metal or plastic tube that widens the esophagus so food can pass into the stomach. Special expanding (dilating) balloons or bougies can relieve a blockage caused by a tumor or treatment-related narrowing (stricture) of the esophagus.

Distress

Distress is an unpleasant experience of a mental, physical, social, or spiritual nature. It can affect how you feel, think, and act. Distress might include feelings of sadness, fear, helplessness, worry, anger, and guilt.

Depression, anxiety, and sleeping problems are common in cancer. Talk to your doctor and with those whom you feel most comfortable about how you are feeling. There are services and people who can help you. Support and counseling services are available.

For more information, see NCCN Guidelines for Patients: Distress During Cancer Care, available at NCCN.org/patientguidelines.

Fatigue

Fatigue is extreme tiredness and inability to function due to lack of energy. Fatigue may be caused by cancer or it may be a side effect of treatment. There are treatments for fatigue. Let your care team know how you are feeling and if fatigue is getting in the way of doing the things you enjoy. Eating a balanced diet, exercise, yoga, and massage therapy can help. You might be referred to a nutritionist or dietitian to help with fatigue.

Nausea and vomiting

Nausea and vomiting are a common side effect of treatment. You will be given medicine to prevent and treat nausea and vomiting.

For more information, see *NCCN Guidelines* for *Patients: Nausea and Vomiting*, available at NCCN.org/patientquidelines.

Pain

Tell your care team about any pain or discomfort. You might meet with a palliative care specialist or with a pain specialist to manage pain.

Pain is common in those with a tumor causing a blockage or for metastatic esophageal cancer. Severe abdominal pain can occur when the tumor grows into nearby nerves or presses against other organs. This pain is treated with around-the-clock medicine such as morphine or other opioids (narcotics). Sometimes, non-narcotic medicines are used to treat pain.

Some people may benefit from palliative radiation therapy, with or without systemic therapy, to help relieve the pain. During this treatment, a radiation beam is focused on the tumor.

Treatment side effects

All cancer treatments can cause unwanted health issues. Such health issues are called side effects. Side effects depend on many factors. These factors include the drug type and dose, length of treatment, and the person. Some side effects may be harmful to your health. Others may just be unpleasant.

Ask for a complete list of side effects of your treatments. Also, tell your treatment team about any new or worsening symptoms. There may be ways to help you feel better. There are also ways to prevent some side effects.

Keep a pain diary

A pain diary is a written record that helps you keep track of when you have pain, how bad it is, what causes it, and what makes it better or worse. Use a pain diary to discuss your pain with your care team. You might be referred to a specialist for pain management.

Include in your pain diary:

- The time and dose of all medicines
- · When pain starts and ends or lessens
- Where you feel pain
- Describe your pain. Is it throbbing, sharp, tingling, shooting, or burning? Is it constant, or does it come and go?
- Does the pain change at different times of day? When?
- Does the pain get worse before or after meals? Does certain food or drink make it better?
- Does the pain get better or worse with activity? What kind of activity?
- Does the pain keep you from falling asleep at night? Does pain wake you up in the night?
- Rate your pain from 0 (no pain) to 10 (worst pain you have ever felt)
- Does pain get in the way of you doing the things you enjoy?

Trouble swallowing

Difficult or painful swallowing is called dysphagia. Treatment such as ablation, photodynamic therapy, or placement of a stent may help relieve pain and discomfort caused by dysphagia.

Trouble eating

Sometimes side effects from surgery, cancer, or other treatments might cause you to feel not hungry or sick to your stomach (nauseated). You might have a sore mouth. Healthy eating is important during treatment. It includes eating a balanced diet, eating the right amount of food, and drinking enough fluids. A registered dietitian who is an expert in nutrition and food can help. Speak to your care team if you have trouble eating or maintaining your weight.

For long-term relief of anorexia, dysphagia, or malnutrition, placement of a feeding tube might help.

Your wishes about treatment are always important. Talk to your care team and make your wishes known.

Tube feeding

Tube feeding is a special liquid food mixture containing protein, carbohydrates (sugar), fats, vitamins, and minerals, given through a tube. You will likely have tube feeding after surgery to allow your esophagus to heal. The tube is temporary and will be removed. However, some may need tube feeding for longer periods to maintain weight or for pain or difficulty swallowing.

Liquid nutrition might be given though:

- A nasogastric tube (NG tube) inserted through the nose.
- A gastrostomy tube (G-tube) placed through the skin of the abdomen directly into the stomach. It is also called a percutaneous endoscopic gastrostomy (PEG) tube.
- A jejunostomy tube (J-tube) placed through the skin of the abdomen into the small intestine.

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Key points

- Surgery is a main or primary treatment for esophageal cancer.
- A resectable tumor can be removed with surgery. An unresectable tumor cannot be removed with surgery.
- Systemic therapy works throughout the body. It includes chemotherapy, targeted therapy, and immunotherapy.
- Targeted therapies can block the ways cancer cells grow, divide, and move in the body.
- Immunotherapy uses your body's natural defenses to find and destroy cancer cells.
- Radiation therapy (RT) uses high-energy radiation from x-rays, gamma rays, protons, photons, and other sources to kill cancer cells and shrink tumors.
- A clinical trial is a type of research that studies a treatment to see how safe it is and how well it works.
- Supportive care is health care that relieves symptoms caused by cancer or its treatment and improves quality of life. Supportive care is always given.
- All cancer treatments can cause unwanted health issues called side effects. It is important for you to tell your care team about all your side effects so they can be managed.
- Eating a balanced diet, drinking enough fluids, exercise, yoga, and massage therapy can help manage side effects.

- Tube feeding is a special liquid food mixture given through a tube. You will likely have tube feeding after surgery to allow your esophagus to heal.
- Pain may be treated with medication, or radiation with or without systemic therapy.
 A pain diary might help you manage pain.
- A registered dietitian who is an expert in nutrition and food can help if it is hard for you to eat food or maintain weight.
- If you smoke or vape, it is important to quit for the best treatment results.

5 Squamous cell carcinoma

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Esophageal squamous cell carcinoma starts in the squamous cells of the epithelium, the innermost lining of the esophagus and part of the mucosa. Treatment is based on the size and location of the tumor and your overall health. Together, you and your doctor will choose a treatment plan that is best for you.

Esophageal squamous cell carcinoma (ESCC) starts in the thin, flat cells found in the inner lining of the esophagus. Squamous cell carcinoma (SCC) is often found in the upper and middle esophagus. Location is defined by the position of the center of the tumor in the esophagus.

Squamous cell carcinoma location might be described as the cervical (neck region) or noncervical area of the esophagus. This chapter is for those with early-stage or locoregional (locally advanced) ESCC.

Early stage

Very small early-stage tumors are staged using a biopsy sample from an endoscopic resection. This is called the pathologic stage (pTNM).

Early-stage tumors include:

- pTis abnormal cells in the epithelium layer of the mucosa
- pT1a tumor invades lamina propria or muscularis mucosa layer of the mucosa

 pT1b - tumor invades the second layer of the esophagus wall (the submucosa)

Before starting treatment, you might be given liquid nutrition through a tube. Treatment is based on the size, grade, and location of the tumor and your overall health.

Surgery is an option

If you are healthy enough for surgery and do not have any other serious health issues that prevent surgery, then endoscopic therapies (preferred) or an esophagectomy might be an option.

- For pTis tumor, endoscopic therapies are preferred. These include endoscopic resection (ER), ER followed by ablation, or ablation. Esophagectomy is also an option.
- For pT1a tumor, ER or ER followed by ablation is preferred. Esophagectomy is also an option.
- For pT1b tumor that is node-negative, esophagectomy is the option.

Treatment after a successful (tumor completely removed with negative margins) endoscopic therapy is surveillance. Surveillance consists of testing on a regular basis to watch for signs that cancer has returned. It includes upper GI endoscopy (or EGD) testing at regular intervals. Other tests might be done.

Treatment after an esophagectomy is based on surgical margin results.

Surgery may not be an option

If you are not healthy enough for surgery or do not want surgery, then treatment will be endoscopic resection and/or ablation.

- For pTis tumor, endoscopic resection (ER), ER followed by ablation, or ablation are the options.
- For pT1a tumor, ER or ER followed by ablation is the option.
- For pT1b tumor that is node-negative, ER or ER followed by ablation is the option. Treatment might be followed by definitive chemoradiation. Definitive treatment is defined as the best treatment after all choices have been considered.

Treatment after a successful (tumor completely removed with negative margins) endoscopic therapy is surveillance. Surveillance includes upper GI endoscopy (or EGD) testing at regular intervals. Other tests might be done.

T = Tumor

A tumor can grow through the layers of the esophagus wall and into nearby structures.

- Tis tumor is found in the epithelium (or innermost) layer of the mucosa (first layer).
- T1a tumor is found in the lamina propria or muscularis mucosa layers of the mucosa.
- **T1b tumor** is found in the submucosa (second layer).
- **T2 tumor** is found in the muscularis propria (third layer).
- **T3 tumor** is found in the adventitia (outer layer). It has grown through the other layers of the esophagus wall.
- T4 tumor has grown all the way through the wall of the esophagus into nearby structures.

Tis and T1 tumors are often considered earlystage cancer. Esophageal cancers are rarely found this early.

T2, T3, and T4 tumors are often referred to as locoregional disease. Cancer may be in lymph nodes or structures near or in direct contact with the esophagus.

In metastatic disease, the tumor can be any size, cancer may be in lymph nodes, and cancer has spread to other parts of the body.

Locoregional

In locoregional disease, the tumor has grown into the second layer (submucosa) or other layers of the esophagus. Cancer may be in the lymph nodes. Cancer found in lymph nodes is called node-positive or nodal disease. Lymph nodes must be removed to confirm cancer.

Locoregional disease is staged before surgery. This is called clinical stage (cTNM). It is based on the endoscopic ultrasound (EUS) and other imaging or biopsy results. Before starting treatment, you might be given liquid nutrition through a tube. Treatment is based on the size, grade, and location of the tumor.

cT1b to **T2**, **N0**

Surgery might be an option if the tumor is not in the cervical esophagus, the tumor is less than 3 centimeters (cm), it has not grown beyond the second layer (submucosa) of the esophagus wall, and there is no cancer in the lymph nodes. Treatment after an esophagectomy will be based on the surgical margin results.

cT2 or node-positive tumors

For tumors 3 cm or more, node-positive tumors, or tumors that have grown into or beyond the third layer (muscularis propria) of the esophagus wall, options include:

- For non-cervical esophagus, preoperative chemoradiation followed by an esophagectomy, or definitive chemoradiation
- For cervical esophagus, definitive chemoradiation

After chemoradiation, you will have tests such as an FDG-PET/CT, CT, and upper GI endoscopy with biopsy to see if any cancer remains. If no disease remains, then you will enter surveillance or have an esophagectomy. An esophagectomy is preferred if disease remains.

cT4b

For tumor that has grown through the layers of the esophagus wall into nearby structures (cT4b), treatment is definitive chemoradiation. Definitive treatment is defined as the best treatment after all choices have been considered. Chemotherapy alone might be possible if cancer is found in the trachea (windpipe), large vessels, spine, or heart. After chemoradiation, you will have tests such as an FDG-PET/CT, CT, and upper GI endoscopy with biopsy to see if any cancer remains. An esophagectomy is possible if disease remains. If no disease remains, then you will enter surveillance.

Surgery is not an option

Not everyone is healthy enough for or wants surgery. For those who do not want surgery, chemoradiation is an option. If you are not able to tolerate chemoradiation, then palliative radiation therapy or palliative care with best supportive care are options. Palliative and best supportive care aim to manage symptoms, improve quality of life, and extend life.

After an esophagectomy

A sample of your tumor and lymph nodes removed during surgery will be tested and staged before starting the next treatment.

No chemoradiation before surgery

The following lists the treatment options after an esophagectomy for those who did not have chemoradiation before surgery.

R₀

In a clear or negative surgical margin (R0), no cancerous cells are found in the tissue around the edge of the tumor (resection margins). Cancer may be in the regional lymph nodes. If you did not receive chemoradiation before surgery, then you will enter surveillance, where you will be monitored for the return of cancer.

R1

In an R1 positive margin, the surgeon removes all the visible tumor, but the microscopic margins are still positive for tumor cells. Despite best efforts this can happen. Treatment is fluoropyrimidine-based chemoradiation.

R2

In an R2 positive margin, the surgeon is unable to remove all the visible tumor or there is metastatic disease (M1). Treatment is fluoropyrimidine-based chemoradiation or palliative care. Palliative and best supportive care aim to manage symptoms, improve quality of life, and extend life.

Chemoradiation before surgery

The following lists the treatment options after an esophagectomy for those who had preoperative (before surgery) chemoradiation.

R₀

In a clear or negative surgical margin (R0), no cancerous cells are found in the tissue around the edge of the tumor (resection margins). Cancer may be in the regional lymph nodes. If no cancer is found, then you will enter surveillance, where you will be monitored for the return of cancer. If cancer remains or is found in lymph nodes, then you will be offered nivolumab (Opdivo®).

R1

In an R1 positive margin, the surgeon removes all the visible tumor, but the microscopic margins are still positive for tumor cells. Despite best efforts this can happen. Treatment is observation until progression or palliative care. Observation is sometimes referred to as watch-and-wait. Palliative and best supportive care aim to manage symptoms, improve quality of life, and extend life.

R2

In an R2 positive margin, the surgeon is unable to remove all the visible tumor or there is metastatic disease (M1). Treatment is observation until progression or palliative care. Palliative care and best supportive care aim to manage symptoms, improve quality of life, and extend life.

Follow-up care

After treatment, you will receive follow-up care to watch for signs that cancer has returned called recurrence. It is important to keep any follow-up doctor visits and imaging test appointments.

If your cancer was successfully treated, then follow-up care might include:

- Medical history and physical exam every 3 to 6 months for 1 to 2 years, every 6 to 12 months for 3 to 5 years, and annually thereafter
- Chemistry profile and complete blood count (CBC), as needed
- Imaging scans as needed
- Upper GI endoscopy (EGD) and biopsy, as needed
- Dilation of esophagus for anastomotic stenosis
- Nutritional assessment and counseling

Contact your doctor if you have any new or worsening symptoms.

Need help paying for medicine or treatment?

Ask your care team what options are available.

Key points

- Esophageal squamous cell carcinoma (ESCC) starts in the squamous cells of the epithelium, the innermost lining of the esophagus and part of the mucosa.
- Treatment is based on the size and location of the tumor, your overall health, and goals for treatment.
- Very small early-stage tumors have not grown beyond the second layer (submucosa) of the esophagus wall. These are staged using a biopsy sample from an endoscopic resection. This is called the pathologic stage (pTNM).
- In locoregional disease, the tumor has grown into the submucosa or other layers of the esophagus. Cancer may be in the lymph nodes.
- Cancer found in lymph nodes is called node-positive or nodal disease.
- Locoregional disease is staged before surgery. This is called clinical stage (cTNM). It is based on the endoscopic ultrasound (EUS) and other imaging or biopsy results.
- Not everyone is healthy enough for or wants surgery. Not everyone can tolerate chemoradiation. There are other treatment options.
- A sample of your tumor and lymph nodes removed during an esophagectomy will be tested and staged before starting the next treatment.
- Palliative and best supportive care aim to manage symptoms, improve quality of life, and extend life.

- Chemoradiation might be given before (preoperative) or after (postoperative) surgery, or as definitive treatment.
- Definitive treatment is defined as the best treatment after all choices have been considered.
- After treatment, you will receive followup care. It is important to keep any follow-up doctor visits and imaging test appointments. Contact your doctor if you have any new or worsening symptoms.

6 Adenocarcinoma

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Esophageal adenocarcinoma (EAC) starts in the mucus-making cells of the esophagus. Treatment is based on the size and location of the tumor and your overall health. This chapter is for those with early-stage or locoregional (locally advanced) EAC. Together, you and your doctor will choose a treatment plan that is best for you.

Early stage

Very small early-stage tumors are staged using a biopsy sample from an endoscopic resection. This is called the pathologic stage (pTNM).

Early-stage tumors include:

- pTis abnormal cells
- pT1a tumor invades lamina propria or muscularis mucosa layer of the mucosa
- > pT1b tumor invades submucosa

Before starting treatment, you might be given liquid nutrition through a tube. Treatment is based on the size, grade, and location of the tumor and your overall health.

If you are healthy enough for surgery and do not have any other serious health issues that prevent surgery, then an esophagectomy might be an option. For those who do not want surgery, chemoradiation is an option.

Surgery is an option

If you are healthy enough for surgery and do not have any other serious health issues that prevent surgery, then endoscopic therapies (preferred) or an esophagectomy might be an option.

- For pTis tumor, endoscopic therapies are preferred. These include endoscopic resection (ER), ER followed by ablation, or ablation. Esophagectomy is also an option.
- For pT1a tumor, ER or ER followed by ablation is preferred. Esophagectomy is also an option.
- For a superficial pT1b, ER followed by ablation or esophagectomy.
- For pT1b tumor that is node-negative, then an esophagectomy.

Treatment after an endoscopic therapy is surveillance. Surveillance includes upper GI endoscopy (EGD) testing at regular intervals. Other tests might be done.

Surgery may not be an option

A tumor that cannot be completely removed with surgery is called unresectable. For those who have an unresectable tumor, who are medically unable to tolerate major surgery, or who do not want surgery, then treatment will be endoscopic resection and/or ablation.

- For pTis tumor, endoscopic resection (ER), ER followed by ablation, or ablation are the options.
- For pT1a tumor, ER or ER followed by ablation is the option.

For pT1b tumor that is node-negative, ER or ER followed by ablation is the option. Treatment might be followed by definitive chemoradiation. Definitive treatment is defined as the best treatment after all choices have been considered.

Treatment after an endoscopic therapy is surveillance. Surveillance includes upper GI endoscopy (EGD) testing at regular intervals. Other tests might be done.

Surveillance consists
of testing on a
regular basis to
watch for signs that
cancer has returned.

Locoregional

In locoregional disease, the tumor has grown into the second layer (submucosa) or other layers of the esophagus. Cancer may be in the lymph nodes. Cancer found in lymph nodes is called node-positive or nodal disease. Lymph nodes must be removed to confirm cancer.

Locoregional disease is staged before surgery. This is called clinical stage (cTNM). It is based on the endoscopic ultrasound (EUS) and other imaging or biopsy results. Before starting treatment, you might be given liquid nutrition through a tube. Treatment is based on the size, grade, and location of the tumor.

cT1b to **T2**, **N0**

An esophagectomy might be an option if the tumor is less than 3 centimeters (cm), it has not grown beyond the third layer (muscularis propria) of the esophagus wall, and there is no cancer in the lymph nodes. Treatment after an esophagectomy will be based on the surgical margin results.

cT2 or node-positive tumors

For tumors 3 cm or more, node-positive tumors, or tumors that have grown into or beyond the third layer (muscularis propria) of the esophagus wall, options include:

- Preoperative chemoradiation (preferred). Depending on the results, it might be followed by an esophagectomy.
- Definitive chemoradiation for those who do not want an esophagectomy or who are not medically able to have surgery
- Perioperative chemotherapy (which is chemotherapy then esophagectomy, followed by chemotherapy)

After chemoradiation, you will have tests such as an FDG-PET/CT, CT, and upper GI endoscopy with biopsy to see if any cancer remains.

cT4b

For tumor that has grown through the layers of the esophagus wall into nearby structures (cT4b), treatment is definitive chemoradiation. Definitive treatment is defined as the best treatment after all choices have been considered. Chemotherapy alone might be possible if cancer is found in the trachea (windpipe), large vessels, spine, or heart. After chemoradiation, you will have tests such as an FDG-PET/CT, CT, and upper GI endoscopy with biopsy to see if any cancer remains. An esophagectomy is possible if disease remains. If no disease remains, then you will enter surveillance.

Surgery is not an option

Not everyone is healthy enough for or wants surgery. For those who do not want surgery or are not healthy enough for surgery, chemoradiation is an option. If you are not able to tolerate chemoradiation, then palliative radiation therapy or palliative with best supportive care are options. Palliative and best supportive care aim to manage symptoms, improve quality of life, and extend life.

Before an esophagectomy

Drug choices for preoperative chemoradiation or chemotherapy are based on the type of esophageal cancer, the location of the tumor, and if you might have chemoradiation or systemic therapy after surgery. Ask your doctor about the preferred drugs and why one might be chosen over another.

After an esophagectomy

A sample of your tumor and lymph nodes removed during surgery will be tested and staged before starting the next treatment.

No chemoradiation before surgery

The following lists the treatment options after an esophagectomy for those who did not have chemoradiation or chemotherapy before surgery.

R0

In a clear or negative surgical margin (R0), no cancerous cells are found in the tissue around the edge of the tumor (resection margins). Cancer may be in the regional lymph nodes. If you did not receive chemoradiation before surgery, then you will enter surveillance, where you will be monitored for the return of cancer.

- If no cancer is found in the lymph nodes, then you will enter surveillance. Fluoropyrimidine-based chemoradiation or chemotherapy are possible options depending on the size of the removed tumor.
- If cancer is found in the lymph nodes, then surveillance, fluoropyrimidine-based chemoradiation, or chemotherapy are possible options.

R1

In an R1 positive margin, the surgeon removes all the visible tumor, but the microscopic margins are still positive for tumor cells. Despite best efforts this can happen. Treatment is fluoropyrimidine-based chemoradiation.

R2

In an R2 positive margin, the surgeon is unable to remove all the visible tumor or there is metastatic disease (M1). Treatment is fluoropyrimidine-based chemoradiation or palliative care.

Chemoradiation before surgery

The following lists the treatment options after an esophagectomy for those who had chemoradiation or chemotherapy before surgery.

R₀

In a clear or negative surgical margin (R0), no cancerous cells are found in the tissue around the edge of the tumor (resection margins). Cancer may be in the regional lymph nodes.

- If no cancer is found in the lymph nodes, then you will enter observation. Observation is sometimes referred to as watch-and-wait. Chemotherapy might be given if you had it before surgery.
- If cancer is found in the lymph nodes, then you will be offered nivolumab if you had chemoradiation before surgery. Observation is also possible. Chemotherapy might be given if you had it before surgery.

R1

In an R1 positive margin, the surgeon removes all the visible tumor, but the microscopic margins are still positive for tumor cells. Despite best efforts this can happen. Chemoradiation is possible only if you did not have it before surgery. Other options include observation until progression or another surgery.

R2

In an R2 positive margin, the surgeon is unable to remove all the visible tumor or there is metastatic disease (M1). Treatment is chemoradiation if you did have it before surgery or palliative care. Palliative care and best supportive care aim to manage symptoms, improve quality of life, and extend life.

Follow-up care

After treatment, you will receive follow-up care to watch for signs that cancer has returned. It is important to keep any follow-up doctor visits and imaging test appointments.

If your cancer was successfully treated, then follow-up care might include:

- Medical history and physical exam every 3 to 6 months for 1 to 2 years, every 6 to 12 months for 3 to 5 years, and annually thereafter
- Chemistry profile and complete blood count (CBC), as needed
- Imaging scans as needed
- Upper GI endoscopy (or EGD) and biopsy, as needed
- Dilation of esophagus for anastomotic stenosis
- Nutritional assessment and counseling

Contact your doctor if you have any new or worsening symptoms.

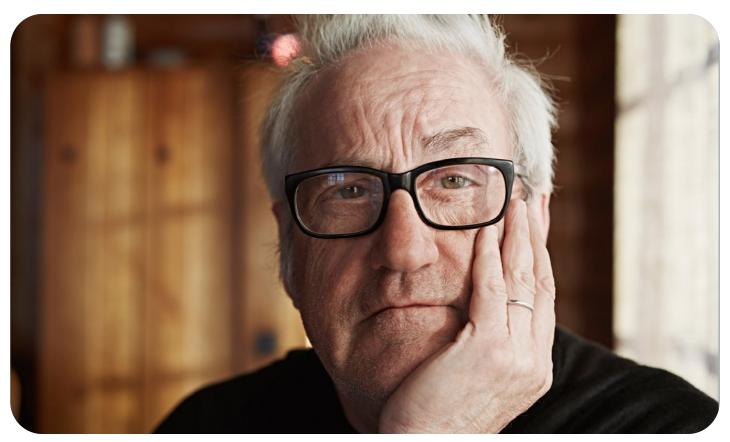
Key points

- Esophageal adenocarcinoma (EAC) starts in the mucus-making cells of the esophagus.
- Treatment is based on the size and location of the tumor, your overall health, and goals for treatment.
- Very small early-stage tumors are staged using a biopsy sample from an endoscopic resection. This is called the pathologic stage (pTNM).
- In locoregional disease, the tumor has grown into the submucosa or other layers of the esophagus. Cancer may be in the lymph nodes.
- Cancer found in lymph nodes is called node-positive or nodal disease. Lymph nodes must be tested to confirm disease.
- Locoregional disease is staged before surgery. This is called clinical stage (cTNM). It is based on the endoscopic ultrasound (EUS) and other imaging or biopsy results.
- Not everyone is healthy enough for or wants surgery. Not everyone can tolerate chemoradiation. There are other treatment options.
- A sample of your tumor and lymph nodes removed during an esophagectomy will be tested and staged before starting the next treatment.
- Chemoradiation might be given before (preoperative) or after (postoperative) surgery, or as definitive treatment.

- Palliative and best supportive care aim to manage symptoms, improve quality of life, and extend life.
- After treatment, you will receive followup care. It is important to keep any follow-up doctor visits and imaging test appointments. Contact your doctor if you have any new or worsening symptoms.

Recurrent or metastatic disease

65	Overview
66	Squamous cell carcinoma
69	Adenocarcinoma
70	Key points



This chapter discusses treatment options for recurrent or metastatic disease. Recurrence is the return of cancer. Cancer that has spread to distant sites in the body is called metastatic disease. Together, you and your doctor will choose a treatment plan that is best for you.

Overview

When cancer returns near the esophagus, it is called locoregional recurrence. Surgery might be an option for those with a locoregional resectable tumor and those who are healthy enough for surgery. Palliative care is also an option.

Esophageal cancer that has spread to distant sites in the body is called metastatic disease. It might be referred to as stage 4 cancer. Not all stage 4 cancer is metastatic. The most common metastatic sites are the liver, distant lymph nodes, lung, bone, and brain. The goal of treatment is to reduce the amount of cancer, called cancer burden, and to prevent the further spread of cancer.

For unresectable locoregional recurrence, metastatic disease, or those who aren't healthy enough for surgery, treatment will focus on palliative and best supportive care. Systemic therapy might be given. Options are based on your performance status (PS). PS is a person's general level of fitness and ability to perform daily tasks.

Order of treatments

Most people with esophageal cancer will receive more than one type of treatment. Below is an overview of the order of treatments.

- Preoperative or neoadjuvant (before)
 therapy is given to shrink the tumor before
 primary treatment (surgery).
- Perioperative therapy is systemic therapy, such as chemotherapy, given before and after surgery.
- Primary treatment is the main treatment given to rid the body of cancer. Surgery is often the main treatment for resectable esophageal cancer. You must be healthy enough for surgery.
- Postoperative or adjuvant (after) therapy is given after primary treatment to rid the body of any cancer cells left behind from surgery. It is also used when the risk of cancer returning (recurrence) is felt to be high.
- Definitive treatment is defined as the best treatment after all choices have been considered
- **First-line therapy** is the first set of systemic (drug) treatment given.
- Second-line therapy is the next set of treatment given if cancer progresses during or after systemic therapy.

Talk to your doctor about your treatment plan and what it means for your stage and type of esophageal cancer.

Squamous cell carcinoma

Locoregional recurrence

It is possible to have cancer return in the remaining esophagus or to have new cancer such as squamous cell carcinoma in another organ. Locoregional recurrence is cancer that has returned to or near the esophagus. Cancer might be in regional lymph nodes.

Had surgery before, but not chemoradiation

Treatment options include chemoradiation (preferred), surgery, chemotherapy, or palliative and best supportive care.

Had chemoradiation before, but not surgery

- If the tumor is resectable and you are healthy enough for surgery, then an esophagectomy is an option.
- If the tumor is unresectable, you are not healthy enough for surgery, or do not want surgery, then treatment will focus on palliative and best supportive care.

Metastatic disease

For metastatic disease, treatment is based on your performance status (PS). If you are able to take care of yourself (PS 0, 1, or 2) and don't have any other serious health issues, then systemic therapy might be an option with palliative and best supportive care. For those with a PS of 3 or 4, treatment focuses on palliative and best supportive care.

A biosimilar is a drug that is very much like one that has been approved by the FDA. It must be used in the exact same way and at the same dose as the other drug.

Systemic therapy

Systemic therapy might be an option depending on your performance status. Before starting systemic therapy, you might have microsatellite and PD-L1 testing if not done before and squamous cell carcinoma is suspected.

For systemic therapy options, see Guide 6 and Guide 7.

Guide 6

First-line therapy options: Unresectable, recurrent, or metastatic disease

For HER2 overexpression positive adenocarcinomas:

- Fluoropyrimidine (fluorouracil or capecitabine) and oxaliplatin and trastuzumab
- Fluoropyrimidine (fluorouracil or capecitabine) and cisplatin and trastuzumab

Preferred options

For HER2 overexpression negative tumors:

- For adenocarcinoma only: Fluoropyrimidine (fluorouracil or capecitabine), oxaliplatin, and nivolumab
- Fluoropyrimidine (fluorouracil or capecitabine), oxaliplatin, and pembrolizumab
- Fluoropyrimidine (fluorouracil or capecitabine), cisplatin, and pembrolizumab
- Fluoropyrimidine (fluorouracil or capecitabine) and oxaliplatin
- Fluoropyrimidine (fluorouracil or capecitabine) and cisplatin

For HER2 overexpression positive adenocarcinomas:

- Fluoropyrimidine (fluorouracil or capecitabine), cisplatin, trastuzumab, and pembrolizumab
- Fluoropyrimidine (fluorouracil or capecitabine), oxaliplatin, trastuzumab, and pembrolizumab

Other recommended

- Fluorouracil and irinotecan
- Paclitaxel with or without cisplatin or carboplatin
- · Docetaxel with or without cisplatin
- Fluoropyrimidine (fluorouracil or capecitabine)
- · Docetaxel, cisplatin or oxaliplatin, and fluorouracil
- Docetaxel, carboplatin, and fluorouracil

Notes:

Oxaliplatin is generally preferred over cisplatin due to lower toxicity.

An FDA-approved biosimilar might be used for trastuzumab.

Leucovorin might be added to fluorouracil-based regimens.

Guide 7

Next-line therapy options: Unresectable, recurrent, or metastatic disease

For esophageal squamous cell carcinoma:

- Nivolumab
- Pembrolizumab for second-line therapy for cancer with certain PD-L1 expression levels

Preferred options

For adenocarcinoma:

- · Ramucirumab and paclitaxel
- Fam-trastuzumab deruxtecan-nxki for HER2 overexpression positive adenocarcinoma
- Trifluridine and tipiracil for third-line or subsequent therapy for esophagogastric junction (EGJ) adenocarcinoma

Other:

- Docetaxel
- Paclitaxel
- Irinotecan
- Fluorouracil and irinotecan (leucovorin might be added)

Other recommended

For adenocarcinoma:

- Ramucirumab
- Fluorouracil and irinotecan with ramucirumab (leucovorin might be added)
- · Irinotecan and ramucirumab
- · Irinotecan and cisplatin
- · Docetaxel and irinotecan

Used in some cases

- Entrectinib or larotrectinib for *NTRK* gene fusion-positive tumors
- Pembrolizumab for MSI-H or dMMR tumors
- Pembrolizumab for TMB high (10 or more mutations per megabase) tumors
- Dostarlimab-gxly for MSI-H or dMMR tumors

Notes:

Systemic therapy options are based on prior therapy and performance status (PS).

Adenocarcinoma

Locoregional recurrence

It is possible to have cancer return in the remaining esophagus or to have new cancer in another organ. Locoregional recurrence is cancer that has returned to or near the esophagus. Cancer might be in regional lymph nodes.

Had surgery before, but not chemoradiation

Treatment options include chemoradiation (preferred), surgery, chemotherapy, or palliative and best supportive care.

Had chemoradiation before, but not surgery

- If the tumor is resectable and you are healthy enough for surgery, then an esophagectomy is an option.
- If the tumor is unresectable, you are not healthy enough for surgery, or do not want surgery, then treatment will focus on palliative and best supportive care.

Metastatic disease

For metastatic disease, treatment is based on your performance status (PS). If you are able to take care of yourself (PS 0,1, or 2) and don't have any other serious health issues, then systemic therapy might be an option alone with palliative and best supportive care. For those with a PS of 3 or 4, treatment focuses on palliative and best supportive care.

Did you know?

The terms "chemotherapy" and "systemic therapy" are often used interchangeably, but they are not the same.
Chemotherapy, targeted therapy, and immunotherapy are all types of systemic therapy.

Systemic therapy

Systemic therapy might be an option depending on your performance status. Before starting systemic therapy, you might have microsatellite, PD-L1, and HER2 testing if not done before and squamous cell carcinoma is suspected.

For systemic therapy options, see Guide 6 and Guide 7.

7

Key points

- It is possible to have cancer return in the remaining esophagus or to have new cancer in another organ.
- Locoregional recurrence is cancer that has returned to or near the esophagus.
 Cancer might be in regional lymph nodes.
- Esophageal cancer that has spread to distant sites in the body is called metastatic disease. It might be referred to as stage 4 cancer. Not all stage 4 cancer is metastatic.
- The most common metastatic sites are the liver, distant lymph nodes, lung, bone, and brain.
- The goal of treatment is to reduce the amount of cancer, called cancer burden, and to prevent the further spread of cancer.
- An unresectable tumor cannot be completely removed with surgery.
- Treatment for an unresectable and/or metastatic disease is palliative and best supportive care with or without systemic therapy, depending on your performance status (PS).



We want your feedback!

Our goal is to provide helpful and easy-to-understand information on cancer.

Take our survey to let us know what we got right and what we could do better:

NCCN.org/patients/feedback

8Survivorship

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After treatment, you will be monitored for any new or ongoing health issues. It is important to keep any follow-up doctor visits and imaging test appointments. Maintain your weight, eat a healthy diet, exercise, limit alcohol, and if you smoke or vape, seek help to quit.

Monitoring

In addition to monitoring for the possible return of cancer called recurrence, you should seek good routine medical care, including regular doctor visits for preventive care and cancer screening. Routine esophageal cancer-specific tests such as imaging, endoscopy, or tumor tests are not recommended after 5 years. It is important to keep any follow-up doctor visits and imaging test appointments.

General health

Esophageal cancer survivors are monitored for long-term side effects. Side effects can be managed. Talk to your doctor about how you are feeling.

In general:

- Maintain a healthy body weight.
- Adopt a physically active lifestyle. The goal is at least 30 minutes of moderateintensity activity most days of the week.
- > Eat a mostly plant-based diet.
- Limit alcohol.
- If you smoke or vape, seek help to quit.

For more information on survivorship, see NCCN.org/patientquidelines.

Diarrhea

Diarrhea is frequent and watery bowel movements. Your care team will tell you how to manage diarrhea and may recommend medicines to stop the diarrhea. It is important to drink lots of fluids. Changes to your diet might help.

Difficulty swallowing

Dysphagia or difficulty swallowing may be a result of an esophagectomy. When part of the esophagus is surgically removed, the two remaining ends are sewn or stapled together (anastomosed). An anastomotic stricture (or stenosis) is a narrowing of the esophagus after an esophagectomy.

Dumping syndrome

Dumping syndrome occurs when food empties into the small intestine too quickly. This may happen within 30 minutes after eating a meal (early dumping syndrome) or within 2 to 3 hours of eating (late dumping syndrome).

Symptoms of early dumping syndrome include palpitations, diarrhea, nausea, and cramps. Late dumping syndrome tends to cause dizziness, hunger, cold sweats, and faintness.

To help manage the symptoms of dumping syndrome:

- Eat often throughout the day
- Avoid drinking liquids with meals
- Eat a diet high in protein and fiber and low in simple carbohydrates and sugars

Fatigue

Fatigue is extreme tiredness and inability to function due to lack of energy. There are treatments for fatigue. Let your care team know how you are feeling and if fatigue is getting in the way of doing the things you enjoy. A balanced diet, exercise, yoga, and massage therapy can help. You might be referred to a nutritionist or dietitian to help with fatigue.

Fullness after meals and eating issues

Eat small portions and eat more often to cope with feeling full after meals. Minimize high fat and high fiber content in food. Also, avoid drinking liquids with meals. Continue to drink fluids in between meals. You might be referred to a gastroenterology for ongoing symptoms.

Heart issues

Because of the location of the esophagus, radiation therapy for esophageal cancer may cause heart issues. You should work with your primary care doctor to monitor your blood pressure, cholesterol, blood sugar, and weight.

Indigestion

Indigestion is a general term that describes discomfort in your upper abdomen. Indigestion is also called dyspepsia or an upset stomach.

To prevent indigestion:

- Avoid foods that increase acid production such as citrus juices, tomato sauces, and spicy foods.
- Avoid foods that lower gastroesophageal sphincter tone such as caffeine, peppermint, and chocolate.

Neuropathy

Neuropathy is a nerve problem that causes pain, numbness, tingling, swelling, or muscle weakness in different parts of the body. It usually begins in the hands or feet and gets worse over time. Neuropathy caused by chemotherapy is called chemotherapy-induced neuropathy.

Reflux

Reflux is the backward flow of liquid from the stomach into the esophagus. Avoid lying flat after eating. Use a foam wedge (triangular) pillow in bed and sleep in a slightly upright position at night. Talk to your doctor before taking any over-the-counter (OTC) medicine.

Weight loss

Your weight will be monitored for changes. Weight loss is expected in the first 6 months after an esophagectomy. In addition, you will be monitored to make sure you are getting enough nutrition and your body is absorbing the nutrition from the food you eat.

Healthy eating is very important. It includes eating a balanced diet, eating the right amount of food, and drinking enough fluids. Eat often and avoid fluids with meals.

A registered dietitian who is an expert in nutrition and food can help if you have trouble eating, absorbing nutrition, or maintaining weight.

Cancer screening

Schedule cancer screenings and vaccinations as recommended by your doctor based on your age, risk, and other factors.

Screenings for cancer include:

- Breast
- Colorectal
- Lung
- Prostate

Key points

- Surgery and other treatments for esophageal cancer can cause health problems. Your health will be monitored.
- A nutritionist or dietician provides guidance on what foods are most suitable for your condition.
- Continue to see your primary health care provider on a regular basis and have preventive cancer screenings as recommended by your doctor.
- Maintain a healthy weight and lifestyle.
- Eat often throughout the day.
- Avoid drinking liquids with meals.
- Eat a mostly plant-based diet that is high in protein and fiber and low in simple carbohydrates and sugars.
- > Limit alcohol.
- If you smoke or vape, seek help to quit.

9 Making treatment decisions

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It's important to be comfortable with the cancer treatment you choose. This choice starts with having an open and honest conversation with your doctor.

It's your choice

In shared decision-making, you and your doctors share information, discuss the options, and agree on a treatment plan. It starts with an open and honest conversation between you and your doctor.

Treatment decisions are very personal. What is important to you may not be important to someone else.

Some things that may play a role in your decision-making:

- What you want and how that might differ from what others want
- > Your religious and spiritual beliefs
- Your feelings about certain treatments like surgery or chemotherapy
- Your feelings about pain or side effects such as nausea and vomiting
- Cost of treatment, travel to treatment centers, and time away from school or work
- Quality of life and length of life
- How active you are and the activities that are important to you

Think about what you want from treatment. Discuss openly the risks and benefits of specific treatments and procedures. Weigh options and share concerns with your doctor.

If you take the time to build a relationship with your doctor, it will help you feel supported when considering options and making treatment decisions.

Second opinion

It is normal to want to start treatment as soon as possible. While cancer can't be ignored, there is time to have another doctor review your test results and suggest a treatment plan. This is called getting a second opinion, and it's a normal part of cancer care. Even doctors get second opinions!

Things you can do to prepare:

- Check with your insurance company about its rules on second opinions. There may be out-of-pocket costs to see doctors who are not part of your insurance plan.
- Make plans to have copies of all your records sent to the doctor you will see for your second opinion.

Support groups

Many people diagnosed with cancer find support groups to be helpful. Support groups often include people at different stages of treatment. Some people may be newly diagnosed, while others may be finished with treatment. If your hospital or community doesn't have support groups for people with cancer, check out the websites listed in this book.

Questions to ask your doctors

Possible questions to ask your doctors are listed on the following pages. Feel free to use these questions or come up with your own. Be clear about your goals for treatment and find out what to expect from treatment.

Questions to ask about testing and staging

1.	What tests will I have? How often will they be repeated? Will my insurance pay for these tests?
2.	When will I have a biopsy? Will I have more than one? What are the risks?
3.	How will my biopsy be performed? What else might be done at this time?
4.	How soon will I know the results and who will explain them to me?
5.	Who will talk with me about the next steps? When?
6.	What biomarker tests will I have? When? Will I have genetic testing?
7.	What will you do to make me comfortable during testing?
8.	Is my cancer resectable or unresectable? What does this mean?
9.	Is my cancer early stage, locally advanced, or metastatic?
10.	Is the cancer in any other areas like my liver, lungs, brain, or bone?
11.	What does my cancer stage mean in terms of length of survival and quality of life?

Questions to ask your doctors about their experience

1.	. What is your experience treating esophageal cancer?				
2.	What is the experience of those on your team?				
3.	Do you only treat esophageal cancer? What else do you treat?				
4.	How many patients like me (of the same age, gender, race) have you treated?				
5.	Will you be consulting with experts to discuss my care? Whom will you consult?				
6.	How many procedures like the one you're suggesting have you done?				
7.	Is this treatment a major part of your practice?				
8.	How many of your patients have had complications? What were the complications?				
9.	How many esophageal cancer surgeries have you done? How many per year?				
10	10. Who will manage my day-to-day care?				

Questions to ask about options

- 1. What will happen if I do nothing?
- 2. How do my age, overall health, and other factors affect the options?
- 3. Am I a candidate for a clinical trial? Can I join a clinical trial at any time?
- 4. Which option is proven to work best for my cancer, age, and other risk factors?
- 5. Does any option offer a long-term cancer control? Are the chances any better for one option than another? Less time-consuming? Less expensive?
- 6. Which treatment will give me the best quality of life? Which treatment will extend life? By how long?
- 7. What are our options if the treatment stops working?
- 8. Can I stop treatment at any time? What will happen if I stop treatment?
- 9. Is there a social worker or someone who can help me decide?
- 10. Is there a hospital or treatment center you can recommend for esophageal cancer treatment? Can I go to one hospital for surgery and a different center for systemic or radiation therapy?

Questions to ask about treatment

- 1. What are my treatment choices? What are the benefits and risks? Which treatment do you recommend and why?
- 2. How will my age, performance status, cancer stage, and other health conditions limit my treatment choices?
- 3. Does the order of treatment matter?
- 4. How long do I have to decide about treatment?
- 5. Does this hospital or center offer the best treatment for me?
- 6. When will I start treatment? How long will treatment take?
- 7. How much will the treatment cost? How much will my insurance pay for this treatment?
- 8. What are the chances my cancer will return? How will it be treated if it returns?
- 9. I would like a second opinion. Is there someone you can recommend?
- 10. How will treatment affect my ability to eat and digest food? Will I be able to do things I enjoy?

Questions to ask about surgery

- 1. If my cancer is resectable, how much of my tumor will be removed? How much of my esophagus will be removed?
- 2. What other organs or tissues might be removed during surgery? What will this mean in terms of my survival and recovery?
- 3. What kind of surgery will I have? Will I have more than one surgery?
- 4. Does my cancer involve any veins or arteries? How might this affect surgery?
- 5. What are the chances you can remove the whole tumor and I will have a negative margin?
- 6. What happens if during surgery you find you can't remove the tumor?
- 7. How long will it take me to recover from surgery? When will I be able to return to work?
- 8. How much pain will I be in? What will be done to manage my pain?
- 9. What is the chance that this surgery will shorten my life?
- 10. What other side effects can I expect from surgery? What complications can occur from this surgery? How will surgery affect my ability to eat and digest food?
- 11. What treatment will I have before, during, or after surgery? What does this treatment do?
- 12. Will I need tube feeding? How long will I need tube feeding?

Questions to ask about food and nutrition

1. What changes will I need to make to my diet after surgery? How can I prepare?
2. What changes should I make to my diet now?
3. Who can help me with meal planning?
4. Should I keep a food diary?
5. I often do not feel well enough to cook or prepare meals. What do you recommend?
6. What can I do if other members of my household do not want to prepare my meals? Or eat the same foods?
7. How can you help if I have trouble paying for food? Or don't have access to the food you are suggesting I eat?

Questions to ask about radiation therapy

1.	What type of radiation therapy (RT) will I have?
2.	What will you target?
3.	What is the goal of this RT?
4.	How many treatment sessions will I require? Can you do a shorter course of RT?
5.	Do you offer this type of RT here? If not, can you refer me to someone who does?
6.	What side effects can I expect from RT?
7.	Should I eat or drink before RT?
8.	Will I be given medicine to help me relax during RT?
9.	What should I wear?

Questions to ask about side effects

- 1. What are the side effects of systemic therapy? Surgery? A stent? Radiation therapy?
- 2. What are the side effects of esophageal cancer?
- 3. How long will these side effects last? Do any side effects lessen or worsen in severity over time?
- 4. What side effects should I watch for? What side effects are expected and which are life threatening?
- 5. When should I call the doctor? Can I text? What should I do on weekends and other non-office hours?
- 6. What emergency department or ER should I go to? Will my treatment team be able to communicate with the ER team?
- 7. What medicines can I take to prevent or relieve side effects?
- 8. What can I do to help with pain and other side effects?
- 9. Will you stop treatment or change treatment if there are side effects? What do you look for?
- 10. What can I do to lessen or prevent side effects? What will you do?
- 11. What medicines may worsen side effects of treatment?

Questions to ask about clinical trials

1.	What clinical trials are available for my type and stage of esophageal cancer?	
2.	What are the treatments used in the clinical trial?	
3.	What does the treatment do?	
4.	Has the treatment been used before? Has it been used for other types of cancer?	
5.	What are the risks and benefits of this treatment?	
6.	What side effects should I expect? How will the side effects be controlled?	
7.	How long will I be in the clinical trial?	
8.	Will I be able to get other treatment if this doesn't work?	
9.	How will I know if the treatment is working?	
10. Will the clinical trial cost me anything? If so, how much?		
_		

Resources

American Cancer Society (ACS)

cancer.org/cancer/esophagus-cancer.html

<u>cancer.org/content/dam/cancer-org/cancer-ontrol/en/worksheets/pain-diary.pdf</u>

Cancer Care

cancercare.org

Cancer Support Community

cancersupportcommunity.org/living-cancer

Chemocare

chemocare.com

Esophageal Cancer Education Foundation

fightec.org

Esophageal Cancer Awareness Association

ecaware.org

MedlinePlus

medlineplus.gov/esophagealcancer.html

My Survival Story

mysurvivalstory.org

National Cancer Institute

cancer.gov/types/esophageal

National Coalition for Cancer Survivorship

canceradvocacy.org/toolbox

National Financial Resource Directory

- Patient Advocate Foundation

<u>patientadvocate.org/explore-our-resources/</u> <u>national-financial-resource-directory</u>

National Hospice and Palliative Care Organization

CaringInfo.org

NCCN Virtual Reimbursement Resource Room and App

NCCN.org/reimbursement

OncoLink

oncolink.org

Patient Access Network Foundation

panfoundation.org

Radiological Society of North America

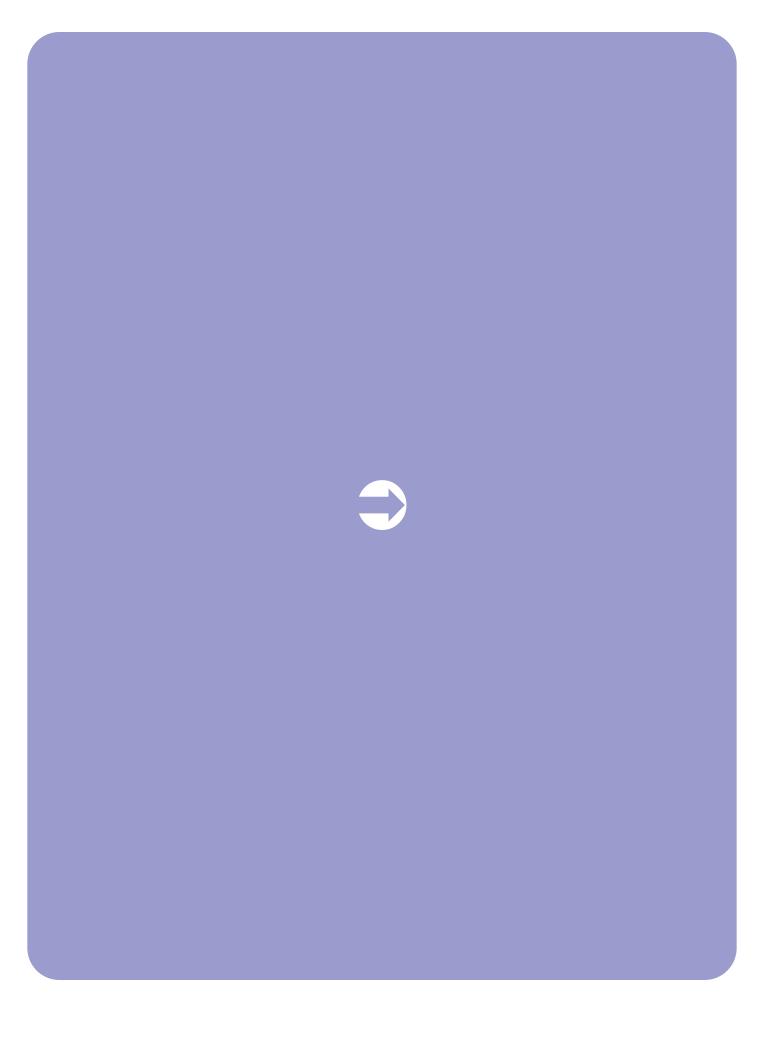
radiologyinfo.org

Smart Patients

smartpatients.com/communities/esophagealcancer

Testing.com

testing.com



Words to know

abdomen

The belly area between the chest and pelvis.

adenocarcinoma

Cancer of cells that form glands and may produce mucus.

anastomotic stricture (or stenosis)

Narrowing of the esophagus after an esophagectomy.

Barrett esophagus (BE)

A condition in which the cells lining the lower part of the esophagus have changed or been replaced with abnormal cells that could lead to cancer of the esophagus.

best supportive care

Treatment that improves quality of life.

biopsy

A procedure that removes fluid or tissue samples to be tested for a disease.

biosimilar

A drug that is very much like one that has been approved by the U.S. Food and Drug Administration (FDA). It must be used in the exact same way and at the same dose as the other drug.

brachytherapy

A treatment with radiation from an object placed near or in the tumor. Also called internal radiation.

bronchi

The two airways extending from the windpipe into the lungs.

bronchoscope

A device that is guided through the nose or mouth to examine the inside of the trachea, bronchi, and lungs

cancer grade

A rating of how much cancer cells look like normal cells.

cancer stage

A rating of the growth and spread of cancer.

carina

A ridge at the base of the trachea (windpipe) that separates the openings of the right and left main bronchi (the large air passages that lead from the trachea to the lungs).

chemoradiation

Treatment that combines chemotherapy with radiation therapy.

chemotherapy

Drugs that kill cancer cells by damaging or disrupting the making of the genetic code.

clinical stage (c)

Rating the extent of a tumor based on tests before treatment.

clinical trial

Research on a test or treatment to assess its safety or how well it works.

colon

The longest part of the large intestine.

complete blood count (CBC)

A test of the number of blood cells.

comprehensive chemistry profile

A panel of tests that gives information about the health and functions of the kidneys and the liver. Usually ordered as part of a comprehensive metabolic panel (CMP).

computed tomography (CT)

A test that uses x-rays to view body parts.

conduit

Tube-shaped tissue, such as the stomach or part of the colon or small intestine, that

Words to know

replaces the part of the esophagus removed during an esophagectomy.

contrast

A substance put into your body to make clearer pictures during imaging tests.

definitive treatment

The best treatment after all choices have been considered.

diaphragm

The thin muscle below the lungs and heart that separates the chest from the abdomen and helps a person to breathe.

digestive system

A set of organs that breaks down food for the body to use.

digestive tract

A set of tube-shaped organs that breaks down food for the body to use. Part of the digestive system.

duodenum

First part of the small intestine.

dysphagia

Difficult or painful swallowing.

Eastern Cooperative Oncology Group (ECOG) Performance Scale

A rating scale of one's ability to do daily activities.

endoscope

A thin, long tube fitted with tools that is guided down the mouth.

endoscopic mucosal resection (EMR)

Removal of early tumors with a snare that has been guided down the throat.

endoscopic resection (ER)

Treatment that removes early tumors with a tool guided down the throat.

endoscopic submucosal dissection (ESD)

Removal of early tumors with a special knife that has been guided down the throat.

endoscopic ultrasound (EUS)

A device guided down your throat to make pictures using sound waves.

epithelium

Cells that line the esophagus wall.

esophagastroduodenoscopy (EGD)

Use of a thin tool guided down the throat into the esophagus and stomach. Also called an upper endoscopy or upper gastrointestinal (GI) endoscopy.

esophagectomy

A surgery that removes all or part of the esophagus.

esophagogastrectomy

A surgery that removes the esophagus and some of the stomach.

esophagogastric junction (EGJ)

The area where the esophagus and stomach join.

esophagus

The tube-shaped organ between the throat and stomach.

external beam radiation therapy (EBRT)

Radiation therapy received from a machine outside the body.

fine-needle aspiration (FNA)

Removal of a tissue sample with a thin needle.

fluoroscopy

An x-ray procedure that makes it possible to see internal organs in motion.

gastrectomy

A surgery that removes some or all of the stomach.

gastroenterologist

A doctor who's an expert in digestive diseases. This system contains organs that break down food for the body to use.

gastrointestinal (GI) tract

The group of organs through which food passes after being eaten. Also called digestive tract.

gastrostomy tube (G-tube)

A feeding tube that is inserted through a cut in the skin of the abdomen directly into the stomach. Also called a percutaneous endoscopic gastrostomy (PEG) tube.

hereditary

Passed down from birth parent to child through coded information in cells.

high-grade dysplasia (HGD)

Abnormal cells that are likely to become cancer cells.

human epidermal growth factor receptor 2 (HER2)

A protein on the surface of a cell that sends signals for the cell to grow.

jejunostomy tube (J-tube)

A feeding tube that is inserted through a cut into the middle intestine (jejunum).

jejunum

The middle part of the small intestine.

imaging

A test that makes pictures (images) of the insides of the body.

immune system

The body's natural defense against infection and disease.

immunotherapy

A treatment with drugs that help the body find and destroy cancer cells.

infection

An illness caused by germs.

interventional radiologist

A doctor who is an expert in imaging tests and using image-guided tools to perform minimally invasive techniques to diagnose or treat disease.

intestine

The organ that food passes through after leaving the stomach.

intravenous (IV)

A method of giving drugs by a needle or tube inserted into a vein.

Karnofsky Performance Status (KPS)

A rating scale of one's ability to do daily activities.

lamina propria

Connective tissue within the mucosa of the esophagus wall.

laparoscopy

Use of a thin tool inserted through a cut made into the belly area.

lymph

A clear fluid containing white blood cells.

lymph node

A small group of special disease-fighting cells located throughout the body.

lymph node dissection

A type of surgery that removes some diseasefighting structures called lymph nodes.

medical oncologist

A doctor who's an expert in cancer drugs.

metastasis

The spread of cancer cells from the first (primary) tumor to a new site.

microsatellite instability (MSI)

Errors made in small, repeated DNA parts during the copy process because of an abnormal repair system.

microsatellite instability-high (MSI-H)

Mutations in 30% or more microsatellites.

minimally invasive procedure

A procedure that uses small incisions or a tool placed into the opening of the body to reduce damage to body tissue.

mucosa

The first, inner layer of the esophagus wall. It is made up of 3 layers: epithelium, lamina propria, and muscularis mucosa.

muscularis mucosa

A thin layer of muscle separating the mucosa from the submucosa of the esophagus wall.

muscularis propria

The third layer of the esophagus wall made mostly of muscle.

mutation

An abnormal change.

nasogastric tube (NG tube)

Inserted through the nose and into the stomach to provide liquid nutrition.

observation

A period of testing for changes in cancer status while not receiving treatment.

pathologic stage (p)

A rating of the extent of cancer based on microscopic review after treatment.

pathologist

A doctor who's an expert in examining tissue and cells to find disease.

pelvis

The area of the body between the hip bones.

percutaneous endoscopic gastrostomy (PEG)

A procedure that inserts a feeding tube into the stomach through a small cut in the skin.

positron emission tomography-computed tomography (PET/CT)

A test that uses radioactive material and x-rays to see the shape and function of body parts.

primary treatment

The main treatment used to rid the body of cancer.

primary tumor

The first mass of cancer cells.

radiation oncologist

A doctor who's an expert in radiation treatment.

radiation therapy (RT)

A treatment that uses high-energy rays.

radiologist

A doctor who is an expert in imaging tests.

recurrence

The return of cancer after a cancer-free period.

resectable

Cancer that can be removed with surgery.

side effect

An unhealthy or unpleasant physical or emotional response to treatment.

simulation

The steps needed to prepare for treatment with radiation therapy (RT).

small intestine

The digestive organ that absorbs nutrients from eaten food.

squamous cell carcinoma (SCC)

A type of cancer that starts in thin and flat cells that line the surface of organs like the esophagus.

stricture

Narrowing of the esophagus. Might be caused by a tumor or treatment.

submucosa

The second layer of the esophagus wall made mostly of connective tissue, blood vessels, and nerve cells.

subtype

A smaller group within a type of cancer that is based on certain cell features.

supportive care

Health care that includes symptom relief but not cancer treatment. Also called palliative care.

surgical margin

The normal-looking tissue around the edge of a tumor that is removed during surgery.

surgical oncologist

A surgeon who's an expert in performing surgical procedures in cancer patients.

surveillance

Testing after treatment ends to check for the return of cancer.

targeted therapy

Drugs that stop the growth process specific to cancer cells.

trachea

The airway between the throat and airway into the lungs. Also called the windpipe.

tumor marker

A substance found in body tissue or fluid that may be a sign of cancer.

tunica adventitia

The outermost layer of the esophagus wall.

ultrasound (US)

A test that uses sound waves to take pictures of the insides of the body.

unresectable

Cancer that can't be removed by surgery.

upper endoscopy

Use of a thin tool guided down the throat into the esophagus and stomach. Also called an upper gastrointestinal (GI) endoscopy or esophagogastroduodenoscopy (EGD).

widespread metastatic disease

The spread of cancer from the first tumor to many new sites in the body.



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NCCN Contributors

This patient guide is based on the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Esophageal and Esophagogastric Junction Cancers, Version 1.2022. It was adapted, reviewed, and published with help from the following people:

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Esophageal Cancer

2022

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