



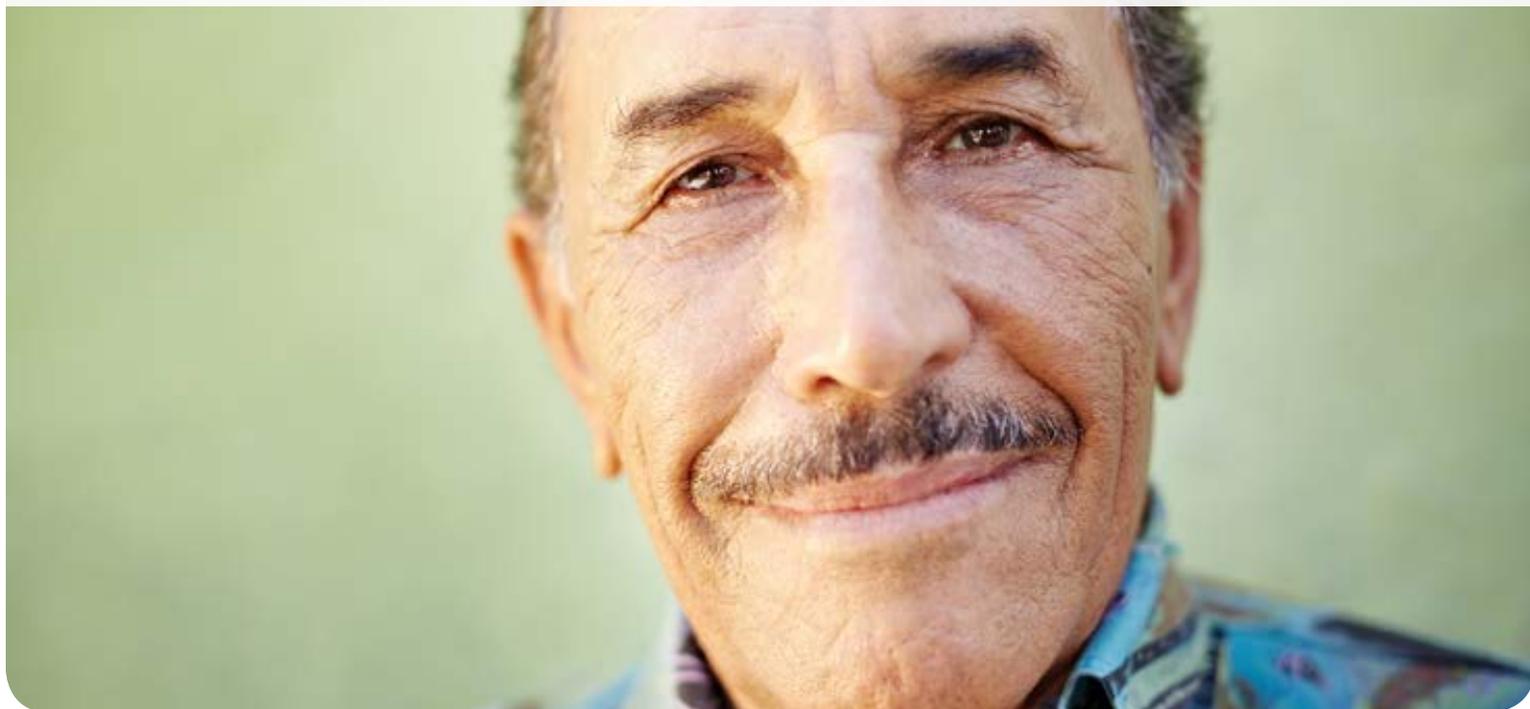
NCCN  
GUIDELINES  
FOR PATIENTS®

2018

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# Esophageal Cancer

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## LEARNING that you have cancer can be overwhelming.

The goal of this book is to help you get the best care. It explains which cancer tests and treatments are recommended by experts in esophageal cancer.

The National Comprehensive Cancer Network® (NCCN®) is a not-for-profit alliance of 27 leading cancer centers. Experts from NCCN have written treatment guidelines for doctors who treat esophageal cancer. These treatment guidelines suggest what the best practice is for cancer care. The information in this patient book is based on the guidelines written for doctors.

This book focuses on the treatment of esophageal cancer in adults. Key points of the book are summarized in the related [NCCN Quick Guide™](#). NCCN also offers patient books on lung cancer, melanoma, and many other cancer types. Visit [NCCN.org/patients](https://www.nccn.org/patients) for the full library of patient books, summaries, and other resources.

# About



These patient guidelines for cancer care are produced by the National Comprehensive Cancer Network® (NCCN®).

The mission of NCCN is to improve cancer care so people can live better lives. At the core of NCCN are the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®). NCCN Guidelines® contain information to help health care workers plan the best cancer care. They list options for cancer care that are most likely to have the best results. The NCCN Guidelines for Patients® present the information from the NCCN Guidelines in an easy-to-learn format.

Panels of experts create the NCCN Guidelines. Most of the experts are from NCCN Member Institutions. Their areas of expertise are diverse. Many panels also include a patient advocate. Recommendations in the NCCN Guidelines are based on clinical trials and the experience of the panelists. The NCCN Guidelines are updated at least once a year. When funded, the patient books are updated to reflect the most recent version of the NCCN Guidelines for doctors.

For more information about the NCCN Guidelines, visit [NCCN.org/clinical.asp](http://NCCN.org/clinical.asp).

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NCCN Foundation was founded by NCCN to raise funds for patient education based on the NCCN Guidelines. NCCN Foundation offers guidance to people with cancer and their caregivers at every step of their cancer journey. This is done by sharing key information from leading cancer experts. This information can be found in a library of NCCN Guidelines for Patients® and other patient education resources. NCCN Foundation is also committed to advancing cancer treatment by funding the nation's promising doctors at the center of cancer research, education, and progress of cancer therapies.

For more information about NCCN Foundation, visit [NCCNFoundation.org](http://NCCNFoundation.org).

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### **Esophageal Cancer Awareness Association (ECAA)**

The ECAA strongly supports NCCN's efforts to provide accurate and easy to read information to anyone affected by esophageal cancer. Patients, family and friends can better understand this disease thanks to these comprehensive guidelines and resources to help move forward with treatment. [www.ecaware.org](http://www.ecaware.org)

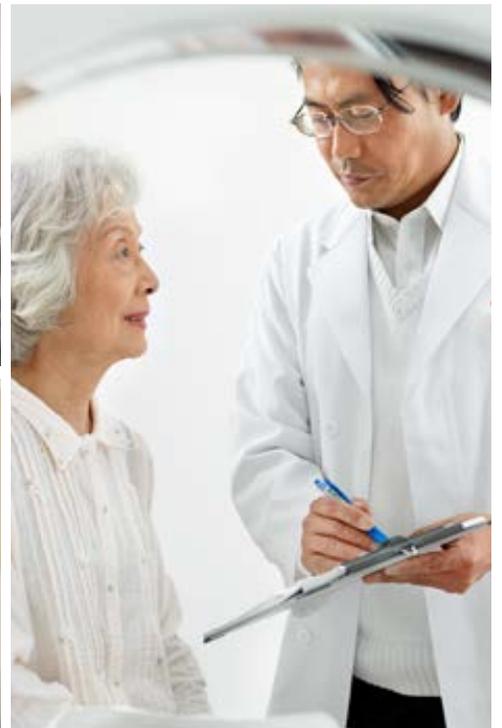
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### **Endorsed by Esophageal Cancer Action Network, Inc. (ECAN)**

ECAN is proud to endorse the NCCN Guidelines for Patients. ECAN understands how critically important it is for patients and families to understand all of their options when facing the life and death decisions so often confronted in Esophageal Cancer. These Guidelines illuminate complex issues as they help answer the important questions that can direct patients on a path to appropriate, life-saving care. [www.ecan.org](http://www.ecan.org)

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Jan & Paula Buckner  
The Honorable Ellen O. Tauscher  
Marcie Reeder in honor of Dr. Lawrence A. Rodgers



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### Who should read this book?

This book is about treatment for esophageal cancer. People with esophageal cancer and those who support them—caregivers, family, and friends—may find this book helpful. It may help you discuss and decide with doctors what care is best.

### Are the book chapters in a certain order?

Starting with **Part 1** may be helpful. It explains what esophageal cancer is. Read **Part 2** to learn about how esophageal cancer is staged. **Part 3** talks about the health tests you may need to take in order to plan the best treatment for you. The main ways to treat esophageal cancer are explained in **Part 4**. The two main kinds of esophageal cancer are squamous cell carcinoma and adenocarcinoma. Recommendations for treating these cancer types are presented in **Parts 5 and 6**. The last chapter is **Part 7**. This chapter offers help for making treatment decisions.

### Does this book include all options?

This book includes treatment options for most people. Your treatment team can point out what applies to you. They can also give you more information. While reading, make a list of questions to ask your doctors.

The treatment options are based on science and the experience of NCCN experts. However, their recommendations may not be right for you. Your doctors may suggest other options based on your health and other factors. If other options are given, ask your treatment team about anything you don't understand.

### Help! What do the words mean?

In this book, many medical words are included. These are words you will likely hear from your treatment team. Most of these words may be new to you, and it may be a lot to learn.

Don't be discouraged as you read. Keep reading and review the information. Feel free to ask your treatment team to explain a word or phrase that you don't understand.

Words that you may not know are defined in the text or in the *Dictionary*. *Acronyms* are also defined when first used and in the Glossary. Acronyms are short words formed from the first letters of several words. One example is DNA for **d**eoxyribonucleic acid.

# 1

## Esophageal cancer basics

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You've learned that you have esophageal cancer. It's normal to feel shocked and confused. This chapter goes over the basics of cancer and how it affects the esophagus. This first look at your cancer will help prepare you for next steps.

## The esophagus

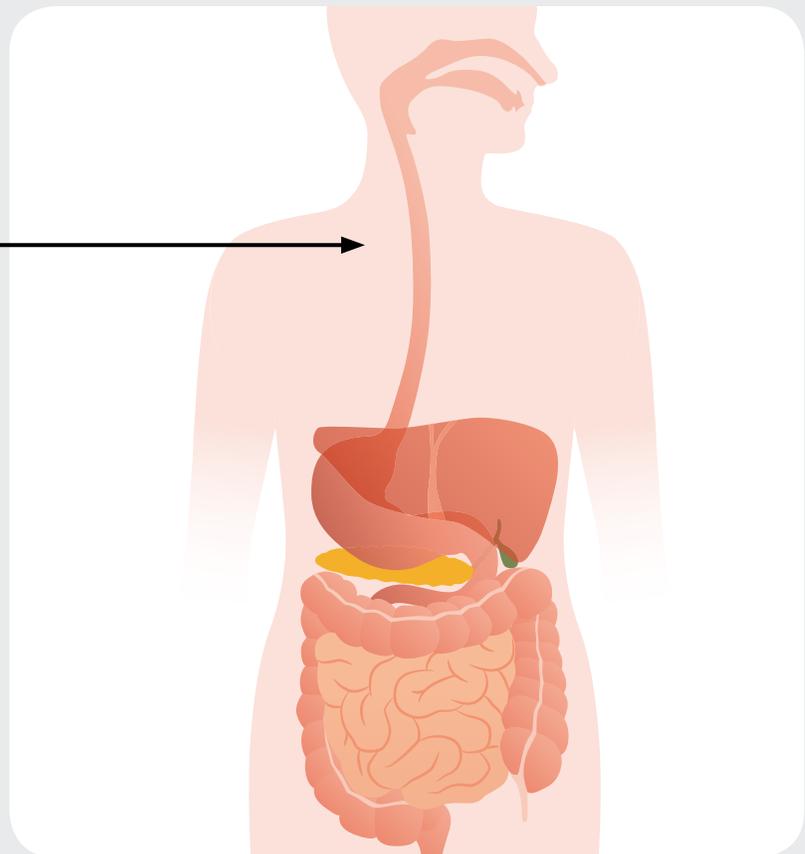
The esophagus is part of your digestive system. It is the hollow, muscular tube that moves food and liquids from your mouth to your stomach. **See Figure 1** for a picture of the esophagus in the body.

The most important facts for you to know about the esophagus are:

- The wall of the esophagus has four layers.
- Cancer starts on the inside (layer 1) and grows outward through layers 2, 3, and 4.
- There are 2 main kinds of esophageal cancer—squamous cell carcinoma and adenocarcinoma.
- Squamous cell carcinoma is usually found in the middle or top part of the esophagus.
- Adenocarcinoma is usually found at the bottom of the esophagus, near the stomach.

**Figure 1. The esophagus**

The esophagus moves food from your mouth to your stomach. Cancer starts on the inside and grows through the four layers of the esophagus wall.



## How cancer works

Your body is made of over 30 trillion cells. All cells have built-in rules that tell them how to act. These rules, or instructions, are called genes. Genes are a part of your DNA (**d**eoxyribonucleic **a**cid). Changes (called mutations) in genes cause normal cells to become cancer cells.

Cancer cells don't act like normal cells. **See Figure 2.** The three most important differences between cancer cells and normal cells are:

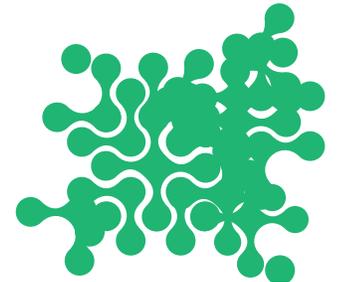
- **Normal cells** grow and then divide to make new cells when needed. They also die when old or damaged. **Cancer cells** make new cells that aren't needed and don't die quickly when old or damaged. Over time, cancer cells form a lump called a tumor.
- **Normal cells** listen to signals from nearby cells telling them to “stop” when they get too close. **Cancer cells** ignore the “stop” signals from nearby cells and invade nearby tissues.
- **Normal cells** stay in the area of the body where they belong. For example, stomach cells stay in the stomach. **Cancer cells** can travel to other parts of your body (metastasize). They can then grow and make more tumors in the new area of your body.

**Figure 2. Key differences between normal cells and cancer cells**



**Normal cells**

- ✓ **Make new cells as needed; die if old/damaged**
- ✓ **Stop when they get too close to other cells**
- ✓ **Stay where they belong in the body**



**Cancer cells**

- ➔ **Grow out of control, forming a tumor over time**
- ➔ **Ignore other cells and invade nearby tissues**
- ➔ **Can spread and make new tumors**

## Review

- The esophagus moves food from the throat to the stomach.
- The wall of the esophagus has four layers. Cancer starts on the inside (the first layer) and grows towards the outside (the fourth layer).
- Squamous cell carcinoma usually starts in the middle or top part of the esophagus.
- Adenocarcinoma usually starts at the bottom of the esophagus, near the stomach.
- Cancer cells form a tumor over time because they don't die like normal cells.
- Cancer cells can spread to other parts of your body and make new tumors.



When you or a loved one is diagnosed with cancer, it affects everyone. Try and find as much information and a support group/network if possible.

– Rhonda

Wife of cancer survivor

# 2

## Cancer staging

- 12 The TNM system
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You have likely heard the word “stage” when talking about cancer. The stage of a cancer is a snapshot of how far it has grown. Your doctors use the stage to decide which tests and treatments will help you most. This chapter explains the system used to stage esophageal cancer.

## The TNM system

The AJCC (**A**merican **J**oint **C**ommittee on **C**ancer) staging system is used to stage esophageal cancer. This system uses three key pieces of information about your cancer in order to give it a stage:

- **T:** The size of your tumor
- **N:** The number of lymph nodes that have cancer (if any)
- **M:** Whether the cancer has spread to other parts of your body (metastasized)

### T = Tumor

Esophageal tumors first grow through the four layers of the esophagus wall, and then into the rest of the body. To describe how far through the esophagus wall the tumor has grown, a number from 1 to 4 (and sometimes a letter) is used. The higher the number, the bigger the tumor. Here are the tumor stages for esophageal cancer:

- **Tis** means that there are abnormal cells on the inside of the esophagus. They could turn into cancer.
- **T1a** means that a lump of cancer cells (a tumor) has started growing on the inside of your esophagus. The tumor hasn't grown very far through the esophagus wall.
- **T1b** means that the tumor has grown through the first layer of the esophagus wall and has entered the second layer.
- **T2** means that the tumor has entered the third layer of the esophagus wall.
- **T3** means that the tumor has reached the last and outer layer of the esophagus wall.
- **T4a** means that the tumor has grown all the way through the esophagus and into the lining of nearby body parts.
- **T4b** means that the tumor has grown all the way through the esophagus and into important nearby body parts.

**See Figure 3** for a picture of what the tumor stages mean.

**N = Node**

There are hundreds of lymph nodes in your body. They work as filters to help fight infection and to remove harmful things from the body. It is important to know if cancer has spread to any lymph nodes. Doctors use a number from 0 to 3 to describe whether the cancer has reached any lymph nodes. The higher the number, the more lymph nodes have cancer. If your doctors don't know if any lymph nodes have cancer, an X is used instead of a number. See below for the N numbers and what they mean.

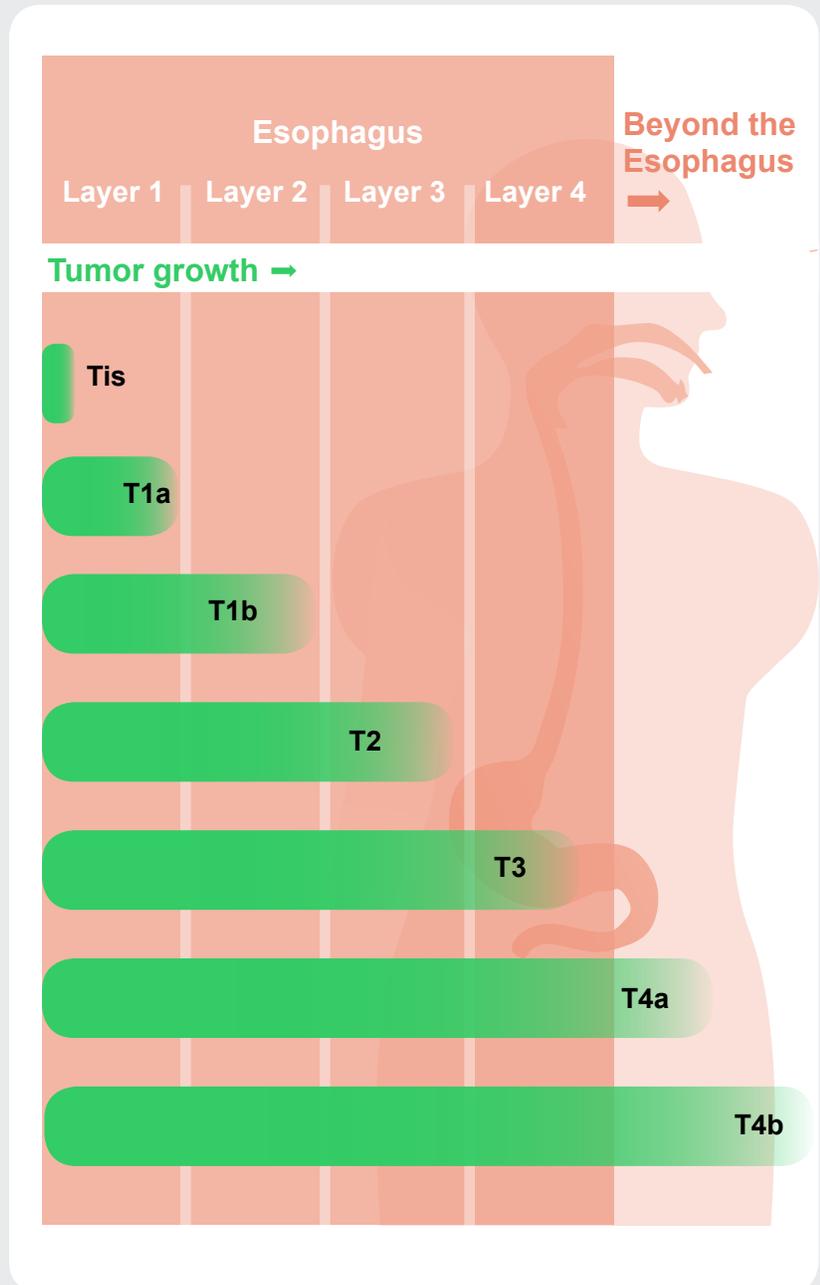
- **NX** means that it is unknown if you have any lymph nodes with cancer.
- **N0** means that you don't have any nearby lymph nodes with cancer.
- **N1** means that you have 1 or 2 lymph nodes with cancer.
- **N2** means that you have 3 to 6 lymph nodes with cancer.
- **N3** means that you have 7 or more lymph nodes with cancer.

**M = Metastasis**

Cancer can spread to areas of the body far from where it started. This process is called "metastasis." Knowing whether the cancer has spread far from the esophagus is an important part of choosing which treatments can be used. To describe whether the cancer has spread far (metastasized), either a 0 or a 1 is used. If your doctors don't know if the cancer has spread, an X is used instead of 0 or 1. The M values are described next.

**Figure 3. Esophageal tumor growth chart**

Esophageal tumors first grow through the 4 layers of the esophagus wall, and then into the rest of the body. To describe how far through the esophagus wall the tumor has grown, a number from 1 to 4 (and sometimes a letter) is used.



- **MX** means that it is unknown if your cancer has spread far from your esophagus.
- **M0** means that the cancer hasn't spread from your esophagus.
- **M1** means that the cancer has spread in the bloodstream away from your esophagus into other areas of your body.

### G = Grade

The next piece of information used to stage cancer is called its “grade.” The grade is a rating of how fast your doctors think the cancer will grow and spread. To figure out the grade, a sample of your tumor will be studied in a laboratory by a pathologist. The pathologist will compare the cancer cells to normal cells. The more different they look, the faster the cancer is expected to spread.

- **GX** means that the grade can't be determined. This may happen if the sample being tested isn't big enough.
- **G1** means that the cancer cells look similar to healthy cells.
- **G2** means that the cancer cells are somewhat different than healthy cells.
- **G3** means that the cancer cells barely look like healthy cells.

### Putting it all together

We just learned about these four key pieces of information your doctors need to know about your cancer:

- Tumor size
- Whether any lymph nodes have cancer
- Whether the cancer has spread to other parts of your body
- How fast the cancer is expected to grow

These four things are then combined to give the cancer a stage. **See Figure 4.** When staging squamous cell carcinoma, the location of the tumor in the esophagus (top, middle, or bottom) is also factored in. Stages are numbered using roman numerals. There are five main stages (0, I, II, III, IV). Stages I through IV are also broken down into two or more sub-stages. Letters are used to describe the sub-stages. For example, there is a stage IIB squamous cell carcinoma and a stage IC adenocarcinoma.

It is important to know that two people with esophageal cancer may be the same stage, even though their cancers are not exactly the same. In other words, there is more than one definition of a stage. For example, if you have squamous cell carcinoma and the primary tumor has entered the third layer of the esophagus wall and there is cancer in one nearby lymph node, your cancer is stage IIIA. If someone else's primary tumor has only entered the second layer of the esophagus wall (which means it's smaller), but there is cancer in two nearby lymph nodes, that person's cancer is also stage IIIA. So, different combinations of physical characteristics can be the same stage of cancer.

In general, people with earlier cancer stages have better outcomes, but not always. It is important to

keep in mind that some people will do better than expected for their stage, and some people will do worse. Cancer is often staged twice. The first rating is done before treatment and is called the clinical (or baseline) stage. If you have surgery, the second rating is done after surgery, and is called the pathologic stage.



#### Figure 4. Esophageal cancer staging

Your doctors combine important information about your cancer to give it a rating, called a stage. The stage tells them how much the cancer has grown so far.

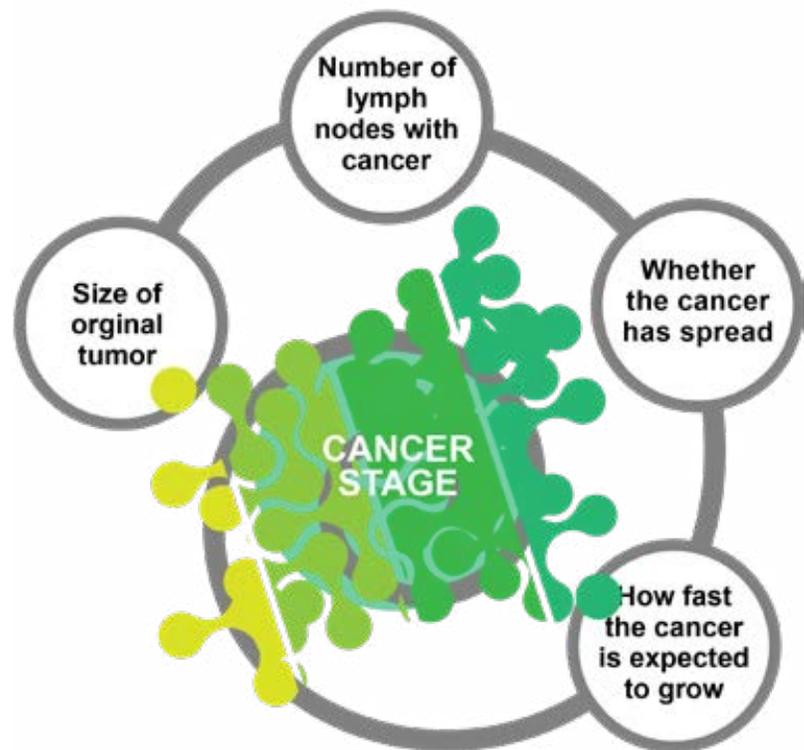


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## Squamous cell carcinoma stages

Stages I, II, III, and IV squamous cell carcinoma are described in [Guides 1 through 4](#).

The next pages describe the five stages of esophageal squamous cell carcinoma. The earliest stage is stage 0. Stage 0 means that there are abnormal cells on the lining of the esophagus. These cells could become cancer, and so they need to be treated. You may hear this stage called “high-grade dysplasia” (HGD).

### Guide 1. Stage I squamous cell carcinoma

#### Stage IA

##### Description

- The tumor is in the first layer of the esophagus wall, and
- There is no cancer in nearby lymph nodes or in distant sites, and
- The cancer cells are grade I, or the grade can't be determined, and
- The tumor is at the top, middle, or bottom of the esophagus (any location).

#### Stage IB

Description 1	Description 2	Description 3
<ul style="list-style-type: none"> <li>• The tumor is in the <u>first</u> layer of the esophagus wall, and</li> <li>• The cancer cells are grade 2 or grade 3, and</li> </ul>	<ul style="list-style-type: none"> <li>• The tumor has entered the <u>second</u> layer of the esophagus wall, and</li> <li>• The cancer cells are grade I, grade 2, grade 3, or the grade can't be determined, and</li> </ul>	<ul style="list-style-type: none"> <li>• The tumor has entered the <u>third</u> layer of the esophagus wall, and</li> <li>• The cancer cells are grade I, and</li> </ul>
<ul style="list-style-type: none"> <li>• There is no cancer in nearby lymph nodes or in distant sites, and</li> <li>• The tumor is at the top, middle, or bottom of the esophagus (any location).</li> </ul>		

## Guide 2. Stage II squamous cell carcinoma

## Stage IIA

Description 1	Description 2	Description 3
<ul style="list-style-type: none"> <li>The tumor has entered the <u>third</u> layer of the esophagus wall, and</li> <li>The cancer cells are grade 2, grade 3, or the grade can't be determined, and</li> <li>The tumor is at the top, middle, or bottom of the esophagus (any location), and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor has entered the <u>fourth</u> and outer layer of the esophagus wall, and</li> <li>The cancer cells are any grade, and</li> <li>The tumor is at the <u>bottom</u> of the esophagus, and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor has entered the <u>fourth</u> and outer layer of the esophagus wall, and</li> <li>The cancer cells are grade I, and</li> <li>The tumor is at the <u>top or middle</u> of the esophagus, and</li> </ul>
<ul style="list-style-type: none"> <li>There is no cancer in nearby lymph nodes or in distant sites.</li> </ul>		

## Stage IIB

Description 1	Description 2	Description 3	Description 4
<ul style="list-style-type: none"> <li>The tumor has entered the <u>first or second</u> layer of the esophagus wall, and</li> <li>There is cancer in 1 nearby lymph node, but not in distant sites, and</li> <li>The cancer cells are any grade, and</li> <li>The tumor may be at the top, middle, or bottom of the esophagus (any location).</li> </ul>	<ul style="list-style-type: none"> <li>The tumor has entered the <u>fourth</u> and outer layer of the esophagus wall, and</li> <li>There is no cancer in nearby lymph nodes or in distant sites, and:</li> </ul>		
	<ul style="list-style-type: none"> <li>The cancer cells are grade 2 or grade 3, and</li> <li>The tumor is at the <u>top or middle</u> of the esophagus.</li> </ul>	<ul style="list-style-type: none"> <li>The grade of the cancer cells can't be determined, and</li> <li>The tumor may be at the top, middle, or bottom of the esophagus (any location).</li> </ul>	<ul style="list-style-type: none"> <li>The cancer cells are any grade, and</li> <li>The location of the tumor can't be determined.</li> </ul>

## Guide 3. Stage III squamous cell carcinoma

### Stage IIIA

Description 1	Description 2
<ul style="list-style-type: none"> <li>The tumor has entered the <u>second</u> layer of the esophagus wall, and</li> <li>There is cancer in 2 nearby lymph nodes, but not in distant sites, and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor has entered the <u>third</u> layer of the esophagus wall, and</li> <li>There is cancer in 1 nearby lymph node, but not in distant sites, and</li> </ul>
<ul style="list-style-type: none"> <li>The cancer cells are any grade, and</li> <li>The tumor may be at the top, middle, or bottom of the esophagus (any location).</li> </ul>	

### Stage IIIB

Description 1	Description 2	Description 3
<ul style="list-style-type: none"> <li>The tumor has entered the <u>third</u> layer of the esophagus wall, and</li> <li>There is cancer in 2 nearby lymph nodes, but not in distant sites, and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor has entered the <u>fourth</u> and outer layer of the esophagus wall, and</li> <li>There is cancer in 1 or 2 nearby lymph nodes, but not in distant sites, and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor has grown through the esophagus wall and into the <u>lining</u> of nearby body parts, and</li> <li>There is cancer in 0 or 1 nearby lymph nodes, but not in distant sites, and</li> </ul>
<ul style="list-style-type: none"> <li>The cancer cells are any grade, and</li> <li>The tumor may be at the top, middle, or bottom of the esophagus (any location).</li> </ul>		

## Guide 4. Stage IV squamous cell carcinoma

### Stage IVA

Description 1	Description 2	Description 3
<ul style="list-style-type: none"> <li>The tumor has grown through the esophagus wall and into <u>the lining</u> of nearby body parts, and</li> <li>There is cancer in 2 nearby lymph nodes, but not in distant sites, and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor has grown through the esophagus wall and <u>into important nearby body parts</u>, and</li> <li>There is cancer in 0, 1, or 2 lymph nodes, but not in distant sites, and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor is any size, and</li> <li>There is cancer in 3 nearby lymph nodes, but not in distant sites, and</li> </ul>
<ul style="list-style-type: none"> <li>The cancer cells are any grade, and</li> <li>The tumor may be at the top, middle, or bottom of the esophagus (any location).</li> </ul>		

### Stage IVB

Description
<ul style="list-style-type: none"> <li>The tumor is any size and in any location (top, middle, or bottom), and</li> <li>The cancer is in any number of lymph nodes, and</li> <li><b>The cancer has spread to areas of your body far from the esophagus (<i>metastasized</i>).</b></li> </ul>

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## Adenocarcinoma stages

The next pages describe the five stages of esophageal adenocarcinoma. The earliest stage is stage 0. Stage 0 means that there are abnormal cells on the lining of the esophagus. These cells could become cancer, and so they need to be treated. You may hear this stage called “high-grade dysplasia”

(HGD). Stages I, II, III, and IV are described in [Guides 5 through 8](#).

### Guide 5. Stage I adenocarcinoma

#### Stage IA

##### Description

- The tumor is in the first layer of the esophagus wall, and
- There is no cancer in nearby lymph nodes or in distant sites, and
- The cancer cells are grade I, or the grade can't be determined.

#### Stage IB

##### Description 1

- The tumor is in the first layer of the esophagus wall, and
- The cancer cells are grade 2, and

##### Description 2

- The tumor has entered the second layer of the esophagus wall, and
- The cancer cells are grade I, grade 2, or the grade can't be determined, and

- There is no cancer in nearby lymph nodes or in distant sites.

#### Stage IC

##### Scenario 1

- The tumor has entered the first or second layer of the esophagus wall, and
- The cancer cells are grade 3, and

##### Scenario 2

- The tumor has entered the third layer of the esophagus wall, and
- The cancer cells are grade I or grade 2, and

- There is no cancer in nearby lymph nodes or in distant sites.



## Guide 7. Stage III adenocarcinoma

### Stage IIIA

Description 1	Description 2
<ul style="list-style-type: none"> <li>The tumor has entered the <u>first or second</u> layer of the esophagus wall, and</li> <li>There is cancer in 2 nearby lymph nodes, but not in distant sites, and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor has entered the <u>third</u> layer of the esophagus wall, and</li> <li>There is cancer in 1 nearby lymph node, but not in distant sites, and</li> </ul>
<ul style="list-style-type: none"> <li>The cancer cells are any grade.</li> </ul>	

### Stage IIIB

Description 1	Description 2	Description 3
<ul style="list-style-type: none"> <li>The tumor has entered the <u>third</u> layer of the esophagus wall, and</li> <li>There is cancer in 2 nearby lymph nodes, but not in distant sites, and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor has entered the <u>fourth</u> and outer layer of the esophagus wall, and</li> <li>There is cancer in 1 or 2 nearby lymph nodes, but not in distant sites, and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor has grown through the esophagus wall and into <u>the lining</u> of nearby body parts, and</li> <li>There is cancer in 0 or 1 nearby lymph nodes, but not in distant sites, and</li> </ul>
<ul style="list-style-type: none"> <li>The cancer cells are any grade.</li> </ul>		

## Guide 8. Stage IV adenocarcinoma

### Stage IVA

Description 1	Description 2	Description 3
<ul style="list-style-type: none"> <li>The tumor has grown through the esophagus wall and into <u>the lining</u> of nearby body parts, and</li> <li>There is cancer in 2 nearby lymph nodes, but not in distant sites, and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor has grown through the esophagus wall and <u>into important nearby body parts</u>, and</li> <li>There is cancer in 0, 1, or 2 lymph nodes, but not in distant sites, and</li> </ul>	<ul style="list-style-type: none"> <li>The tumor is any size, and</li> <li>There is cancer in 3 nearby lymph nodes, but not in distant sites, and</li> </ul>
<ul style="list-style-type: none"> <li>The cancer cells are any grade.</li> </ul>		

### Stage IVB

Description
<ul style="list-style-type: none"> <li>The tumor is any size and in any location (top, middle, or bottom), and</li> <li>The cancer is in any number of lymph nodes, and</li> <li><b>The cancer has spread to areas of your body far from the esophagus (<i>metastasized</i>).</b></li> </ul>

## Review

- The TNM (tumor, node, metastasis) system and the cancer grade are used to figure out what stage of cancer you have. For squamous cell carcinoma, the location of the tumor in the esophagus is also used.
- The cancer grade is a rating of how fast your doctors think the cancer will grow and spread. It is based on how much the cancer cells look like healthy cells.
- There are 5 stages of esophageal cancer: 0, I, II, III, and IV.
- Cancer is often staged twice. The clinical stage is based on tests given before treatment. The pathologic stage is based on the results of surgery.
- Some people will do better than expected for their stage, and some people will do worse.

# 3

## First steps

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25 Testing before treatment

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30 Your treatment team

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31 Good nutrition

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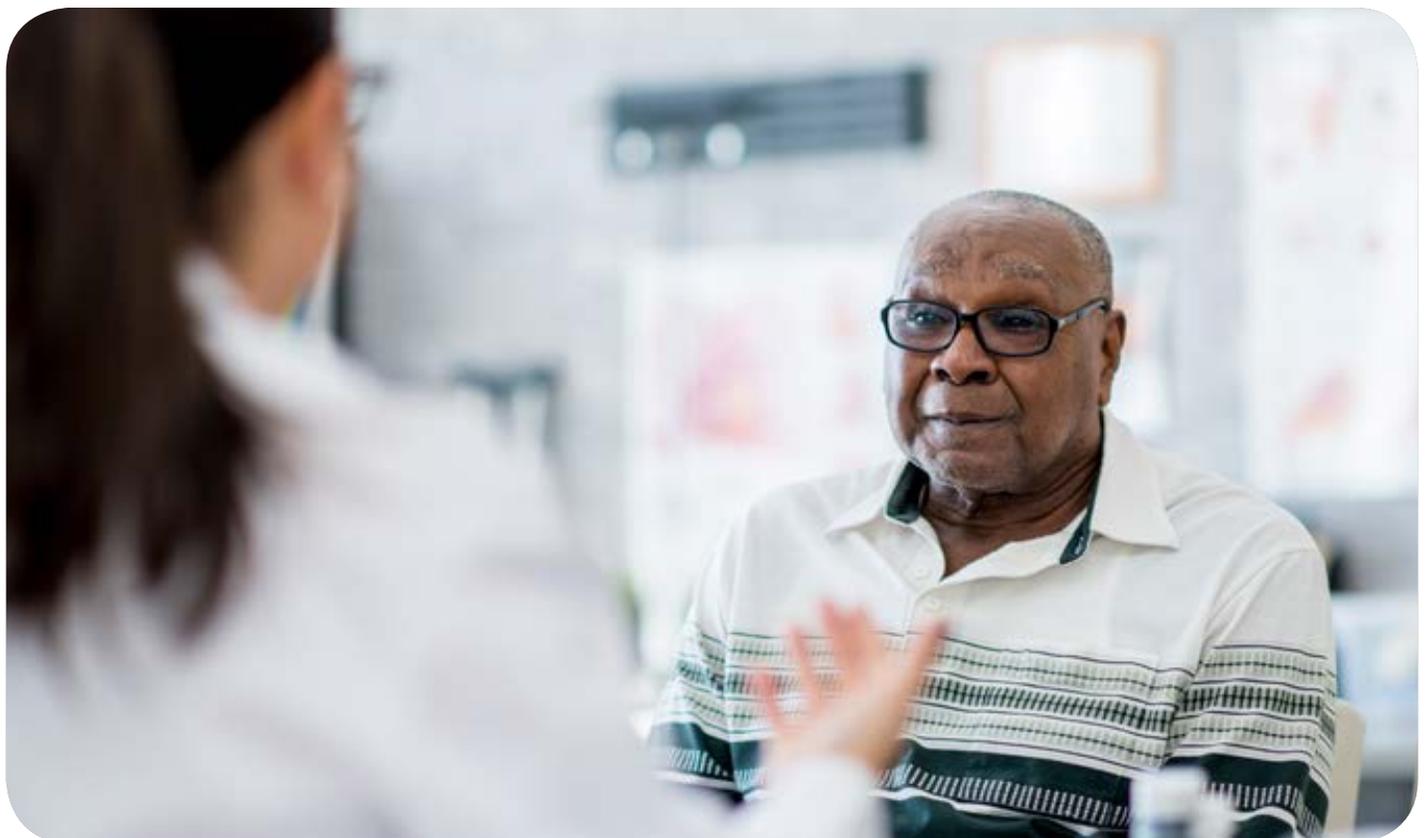
32 Quit smoking!

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32 Distress screening

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32 Review



Your doctors will make a treatment plan just for you. First, they need to gather information about your unique cancer and your general health. This chapter goes over the tests you may need to have done and other steps needed to create your treatment plan.

## Testing before treatment

### Health history and physical exam

Your doctor will need to know a lot about your past and current health. He or she will ask you about:

- Illnesses, diseases, and surgeries you've had
- Medicines that you take (prescription or over-the-counter)
- Your lifestyle (your diet, how much exercise you get, and whether you smoke or drink alcohol)
- Symptoms that could be related to esophageal cancer, such as chest pain, heartburn, trouble swallowing, and unplanned weight loss

Your doctor will also do a physical exam of your body to look for general signs of disease. He or she will likely do the following things during the exam:

- Check your vital signs (blood pressure, heart rate, breathing rate, and body temperature)
- Assess your overall appearance
- Listen to you breathe to check your lungs
- Examine your head and neck
- Feel and/or listen to the organs in your abdomen, including your liver and stomach



## SNAPSHOT

### Barrett's esophagus

- ✓ A condition where the cells that line the lower part of the esophagus become damaged
- ✓ Most likely caused by long-term acid reflux
- ✓ Can lead to esophageal adenocarcinoma over time
- ✓ People with Barrett's esophagus should have regular exams to look for signs of cancer

Some health problems run in families. Your doctor will want to know if you have a family history of cancer, or of other diseases that can raise your risk of getting cancer. Barrett's esophagus, Bloom syndrome, tylosis, and Fanconi anemia are health conditions that are strongly linked to esophageal cancer. If you have any of these disorders, your doctor will likely refer you to a genetic counselor. A genetic counselor is an expert in changes within genes that are related to disease.

### Blood tests

A CBC (**complete blood count**) is a very common blood test. It gives important information about the numbers and kinds of cells in the blood, especially red blood cells, white blood cells, and platelets.

Another blood test is called a chemistry profile. It measures the amount of certain substances in the blood, such as metabolites, electrolytes, fats, and proteins. This test gives important information about how well your kidneys, liver, and other organs are working.

### CT scan

Your doctor will likely want to do a CT (**computed tomography**) scan of your chest, stomach, and maybe your pelvis. A CT scan is a more detailed kind of x-ray. It takes a lot of pictures, or images, from different angles. A computer then combines the images to make 3-D (**three-dimensional**) pictures.

### PET/CT scan

Sometimes CT is combined with another imaging test called PET (**positron emission tomography**). When used together, it is called a PET/CT scan. PET uses small amounts of radioactive materials called radiotracers. About an hour before the scan, you will be injected with a sugar radiotracer. The radiotracer gives off a small amount of energy that can be seen by the imaging machine. Cancer appears brighter in the pictures because cancer cells use sugar more quickly than normal cells.

### Upper endoscopy and biopsy

An upper GI (**gastrointestinal**) endoscopy lets your doctor see inside your esophagus and stomach without making any cuts into your body. An endoscope is a long, skinny tube with a light and a video camera at one end. The endoscope is put in your mouth and guided down your throat into the esophagus. The video camera is connected to a monitor that allows your doctor to see what the camera sees. **See Figure 6.**



## What to expect: CT scan

- ✓ You will lie face-up on a table that moves through a tunnel-like machine. **See Figure 5.**
- ✓ Contrast dye ("contrast" for short) will be used to see everything better.
- ✓ The dye will be injected into your vein and mixed with a liquid you drink.
- ✓ The contrast may cause you to feel flushed or get hives.
- ✓ You will be alone during the scan, but a technician will be nearby. You will be able to hear and talk to the technician.
- ✓ You may hear buzzing or clicking during the scan.
- ✓ Tell your doctor if you get nervous in tight spaces.

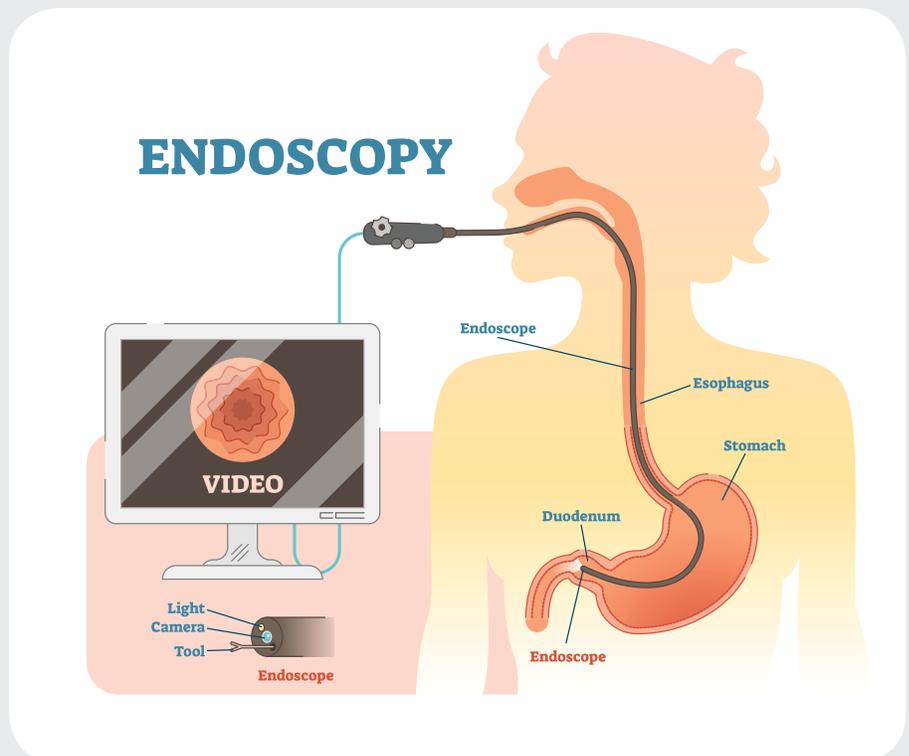
**Figure 5.**  
**CT machine**

A CT scan is a more detailed kind of x-ray. It takes a lot of pictures, or images, from different angles. A computer then combines the images to make 3-D pictures.



**Figure 6.**  
**Upper GI endoscopy**

An endoscope is a long, skinny tube with a light and a video camera at one end. The video camera is connected to a monitor that allows your doctor to see your esophagus.



Your doctor will make note of where the tumor is, how big it is, and how much it is blocking your esophagus. He or she will also look for Barrett's esophagus and areas that look suspicious for cancer. Tiny pieces (samples) of the tumor and the suspicious areas will be taken out to be tested. This is called a biopsy. To remove the samples, the doctor will use a tool that looks like tiny tweezers or salad tongs.

### Endoscopic ultrasound

EUS (endoscopic ultrasound) uses both imaging and an endoscope to see how deep the tumor has grown into the esophageal wall. The ultrasound device bounces sound waves off organs to make pictures. Signs of cancer within lymph nodes and other nearby organs can also be seen.

Like for an upper endoscopy, you will likely be sedated for EUS. The EUS endoscope will be guided down your esophagus. If it looks like the cancer has spread, the endoscope can be used to do an FNA (fine-needle aspiration). An FNA is a type of biopsy. A needle will be inserted through the wall of your esophagus and into nearby tissue to get samples. Your doctor may take samples from your lymph nodes or organs next to your esophagus. An FNA may be done as long as the needle doesn't go through the primary tumor or major blood vessels.

After the biopsy, the endoscopist will provide clinical staging information. He or she will report the depth of tumor growth (**T** value) and whether cancer is present in your lymph nodes (**N** value). This information will help your doctor plan the best treatment for you.

### Bronchoscopy

The carina is supportive tissue at the base of your windpipe (trachea). If the tumor is at or above the carina, a bronchoscopy can be used to see if the tumor has grown into your trachea or airways (bronchi). This test is much like an endoscopy except that the scope is guided down your trachea.



## What to expect: Upper endoscopy

- ✓ Most people are sedated for this kind of endoscopy. This means you will be awake, but will be given medicine to make you sleepy and relaxed.
- ✓ The medicine takes time to wear off, so you won't be able to drive right away. Plan to have someone drive you home.
- ✓ You may have some swelling and sound hoarse afterwards.

Like endoscopes, bronchoscopes are open in the middle and have a light and camera at one end. The light and camera allow your doctor to guide the tube down your nose or mouth and see inside your body. A small brush, needle, or tongs can be put down the open part of the bronchoscope to collect samples. Or, liquid might be sprayed into the airway and suctioned back up to get the samples. After the biopsy, you may feel some swelling and sound hoarse.

### Laparoscopy

If your tumor is an adenocarcinoma and is in the area where the esophagus and stomach meet, your doctor may want to do a laparoscopy. This is a type of surgery that allows your doctor to look for diseases inside your abdomen. Your abdomen is the area between your chest and your pelvis. Laparoscopy is able to find signs that the cancer has spread to the lining of your abdomen or liver.

Laparoscopy is done under general anesthesia. It involves the use of a tool called a laparoscope, which is much like an endoscope. The tube-like part of the laparoscope will be inserted through a tiny cut in your abdomen. Your doctor will be able to look for signs of cancer and obtain fluid for cancer testing (peritoneal washings).

Laparoscopy is done in an operating room and takes about 30 minutes. You will be able to go home about one hour after the procedure is completed and can go back to work the next day. After the surgery, you may feel tired and may have some pain. You may also have a small scar after the cut has healed.

### Tumor biomarker testing

Does your doctor think—or know—that the cancer has spread to other parts of your body (metastasized)? If so, you will be offered some tests to find out if certain newer treatments might help you.

Just like each person's DNA is unique, each person's cancer is unique. This means that a treatment that helps one person might not help you. To find out if certain treatments might help you, your doctor may offer you tumor marker testing. This is also called biomarker (short for biological marker) testing.

Tumor markers can be substances, like molecules or proteins, that are made by your body because you have cancer. Tumor markers can also be processes, such as the way your DNA “acts” that makes it unique. To find out if your cancer has any markers,

the tumor removed by biopsy or during surgery is tested in a laboratory. The three main tumor markers for adenocarcinoma that has spread (metastasized) are described below.

### HER2 testing

HER2 (**h**uman **e**pidermal growth factor **r**eceptor **2**) is a protein found on the surface of cells. It is made by the *HER2* gene. Some esophageal adenocarcinomas have too much of the HER2 protein, which causes the cancer to grow and spread more quickly than it normally would. If testing shows that your adenocarcinoma has too much HER2, your doctor may want to add a drug to your chemotherapy treatment. The drug is called trastuzumab.

### PD-L1 testing

Your immune system has important white blood cells called T cells. T cells' main job is to attack harmful things in your body, like bacteria, viruses, and cancer. They do this with the help of a protein on their surface called PD-1 (**p**rogrammed **c**ell **d**eath-**1**). Cancer cells have a different protein on their surface called PD-L1 (**p**rogrammed **d**eath-**l**igand **1**). When PD-1 and PD-L1 meet, it is called an immune checkpoint. The T cell is “told” to leave the cancer cell alone instead of attacking it.

A PD-L1 test measures how much PD-L1 a tumor makes. Knowing this helps your doctor decide if treatment with drugs called immune checkpoint inhibitors may help you. Immune checkpoint inhibitors stop these two proteins from meeting. This means that the T cells will do their job and attack the cancer cells. If your tumor is an adenocarcinoma and is positive for PD-L1, it doesn't mean that treatment with an immune checkpoint inhibitor is definitely right for you. There are other factors that your doctors will weigh to find out if this treatment is best for you.

**Microsatellite instability or Mismatch repair testing**

Some people have a problem with their genes that makes them unable to fix damaged DNA. In normal cells, a process called MMR (**mismatch repair**) fixes errors that happen when the DNA divides and makes a copy of itself. If a cell's mismatch repair system isn't working right, errors build up and cause the DNA to become unstable. This is called MSI (**microsatellite instability**).

There are two kinds of laboratory tests for this genetic defect. Depending on which method is used, the result will either be MSI-H (**microsatellite instability high**) or dMMR (**mismatch repair deficient**) if you have this genetic defect. Both results mean the same thing.

Like PD-L1 testing, the goal of testing for this tumor marker is to find out if treatment with an immune checkpoint inhibitor may help you. If your cancer is MSI-H or dMMR, it doesn't mean that treatment with an immune checkpoint inhibitor is definitely right for you. There are other factors that your doctors will weigh to find out if this treatment is best for you.

## Your treatment team

Treating esophageal cancer takes a team of doctors and other experts. It is important that everyone involved in your care meets often to make decisions about your care. NCCN experts recommend that meetings take place every week or every other week.

Your treatment team will probably include a:

- Pathologist – an expert in testing tissue to find disease,
- Radiologist – an expert in reading imaging tests,
- Oncology surgeon – an expert in cancer surgery,
- Medical oncologist – an expert in cancer drugs,
- Gastroenterologist – an expert in digestive diseases,
- Radiation oncologist – an expert in using radiation to treat cancer, and an
- Interventional radiologist – an expert in using minimally invasive procedures to diagnose and treat cancer.

The following professionals may also be a part of your treatment team:

- Integrative medicine doctor – an expert in mind-body treatments,
- Nutritionist – an expert in healthy foods and drinks,
- Nurse – an expert trained to care for the sick,
- Social worker – an expert in meeting social and emotional needs, and
- Supportive/palliative care specialists – experts in improving quality of life.

## Good nutrition

Esophageal cancer can make swallowing food difficult or painful. This is called dysphagia. Surgery and other cancer treatments may be too dangerous if you are weak from not eating enough. To help you get as healthy as possible before treatment, you will likely meet with a nutritionist. The nutritionist can help you find ways to eat and drink better.

If you are unable to get enough nutrients by eating and drinking on your own, you may need to have a feeding tube put in. There are different kinds of feeding tubes.

- ▶ A jejunostomy tube (“J-tube” for short) is a soft, plastic tube placed through the skin into the small intestine. NCCN experts prefer J-tubes for patients with esophageal cancer who need tube feeding. **See Figure 7.**
- ▶ Another type called a nasogastric tube is placed through the nose, down the esophagus, and into the stomach.
- ▶ A third type called a PEG (percutaneous endoscopic gastrostomy) tube is placed through the skin directly into the stomach.

### Figure 7. Jejunostomy tube (J-tube)

A J-tube is a soft, plastic tube placed through the skin of the stomach into the small intestine. It is used to deliver nutrients to patients who can't eat well enough on their own.

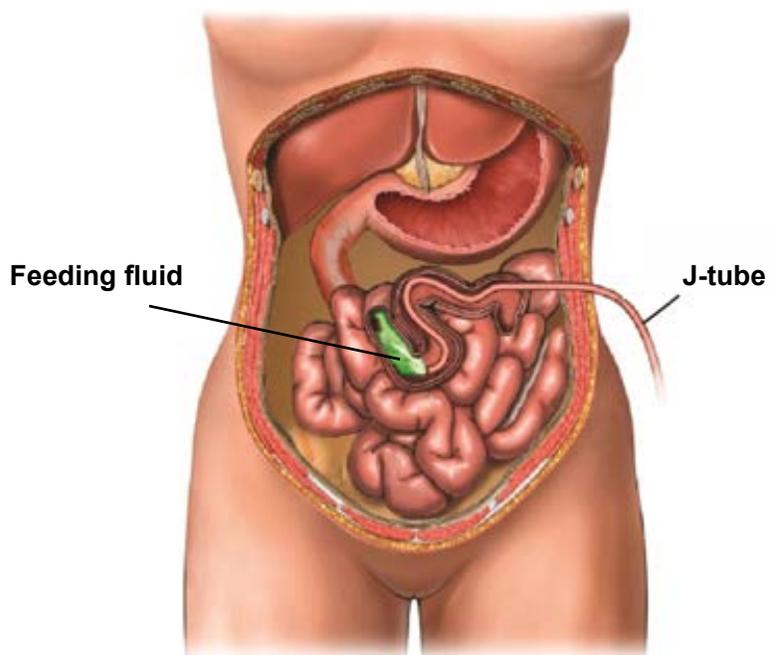


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## Quit smoking!

If you are a smoker, it is very important to quit. Quitting is important because smoking can limit how well cancer treatment works. Smoking also raises the chances of having side effects after surgery.

Your doctor will have ways to help you quit. So, if you smoke, ask your doctor about counseling and drugs to help you stop. There are more options now than ever. The stress of esophageal cancer could make it even harder to quit, so ask your doctor for help.

## Distress screening

Distress is an unpleasant experience of a mental, physical, social, or spiritual nature. It can affect how you feel, think, and act. It can include feelings of sadness, fear, helplessness, worry, anger, guilt, and so forth. Everyone with cancer has some distress at some point in time. It is to be expected.

Feeling distressed may be a minor problem or it may be more serious. You may be so distressed that you can't do the things you used to do. Serious or not, it is important that your treatment team knows how you feel. They may ask you to complete a list of screening questions to assess how distressed you are. Read the [NCCN Guidelines for Patients®:Distress](#) to learn more.



If needed, your treatment team can get you help. Help can include support groups, talk therapy, or medication. Some people also feel better by exercising, talking with loved ones, or relaxing. There may also be helpful community resources, such as support groups and wellness centers.

## Review

- ▶ You will need to do some tests to help your doctors plan the best treatment for you.
- ▶ If your cancer has spread (metastasized), your doctors may offer you tumor biomarker testing to find out if newer treatments might help you.
- ▶ Treating esophageal cancer takes a team of experts.
- ▶ Getting good nutrition is important before starting treatment.
- ▶ To get the best treatment, it is very important to quit smoking.

# 4

## Overview of cancer treatments

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34 Endoscopic therapies

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34 Surgery

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37 Radiation therapy

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38 Chemotherapy

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40 Targeted therapy

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41 Immunotherapy

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42 Clinical trials

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43 Review



In this chapter, the main ways to treat esophageal cancer are described. Knowing what a treatment is will help you understand your options. Not every person will need every treatment described in this chapter. The treatments you get depend a lot on the stage of your cancer.

## Endoscopic therapies

Endoscopic therapies are used to remove very small esophageal tumors and areas of abnormal tissue that could become cancer. The two main endoscopic therapies used to treat early esophageal cancer are called resection and ablation. Both types use an endoscope and one or more tools to remove or kill the cancer cells.

### Endoscopic resection

There are two ways to resect, or remove, small esophageal tumors. In both methods, liquid is first injected under the tumor. The liquid acts as a cushion and “lifts” the tumor up. Then, the tumor is removed in one of two ways.

- **EMR (endoscopic mucosal resection):** The cancer cells are removed with a lasso-shaped tool called a snare. This method is best for removing tumors that haven’t reached deeper layers of the esophageal wall. **See Figure 8.**
- **ESD (endoscopic submucosal dissection):** The doctor uses a knife to break up the healthy tissue under the tumor. This allows the whole tumor to be removed in one piece, which helps stop it from coming back. This method is best for removing tumors that have grown deeper into the esophageal wall.

### Endoscopic ablation

Another way to treat very small tumors using an endoscope is called ablation. Ablation destroys cancer cells using heat, cold, or light-activated drugs. The three main types of ablation are:

- **RFA (radiofrequency ablation)** kills cancer cells using heat from electrodes that are passed through an endoscope. This is the most common type of ablation.
- **Cryoablation** kills cancer cells by freezing them with liquid nitrogen that is sprayed through an endoscope.
- **PDT (photodynamic therapy)** kills cancer cells using drugs that become active when exposed to light.

## Surgery

Surgery is one of the major treatments used for esophageal cancer. The main type of surgery used is called an esophagectomy. The goal of surgery is to remove the entire tumor and some normal-looking tissue around it. The normal-looking tissue is called the surgical margin.

### Esophagectomy

An esophagectomy is a surgery to remove some (or all) of your esophagus and the nearby lymph nodes. **See Figure 9.** How much of the esophagus is removed depends on:

- How big the tumor is
- Where it is in your esophagus
- If the cancer has entered your stomach

### Figure 8. Endoscopic resection

Esophageal tumors that have not grown into deeper layers of the esophageal wall may be removed with endoscopic resection. This treatment removes tumors with tools inserted through an endoscope.



Fluid is injected under the tumor



A cutting tool removes the tumor

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### Figure 9. Before and after esophagectomy

Surgery removes any of your esophagus that has cancer and then attaches your stomach to your remaining esophagus.

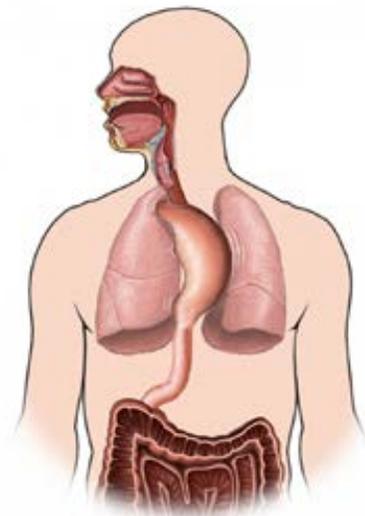
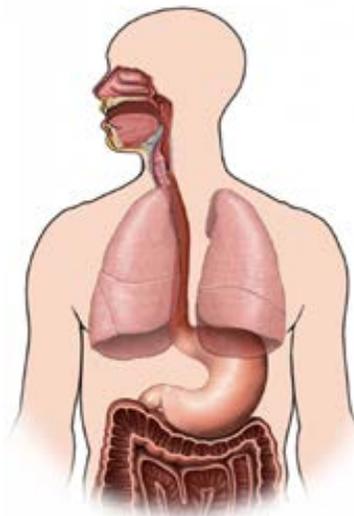


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If the cancer is only in your esophagus and not in your stomach, the part of your esophagus with the cancer will be removed. Then, your stomach will be pulled up into your chest and attached to the rest of your esophagus.

If cancer has grown into your stomach, then the surgeon will take out part of your esophagus and the top part of your stomach. The rest of your stomach will then be attached to the rest of your esophagus. This is called an esophagogastrectomy.

Standard open esophagectomy removes tissue through large surgical cuts. There are two common types of open surgery. An Ivor Lewis transthoracic esophagectomy removes tissue through cuts in your chest and stomach. A McKeown esophagectomy involves cuts in the chest, stomach, and neck.

Minimally invasive esophagectomy removes tissue through a few small cuts. It is done with either the Ivor Lewis or McKeown approach. A laparoscope will be inserted through a small cut into your belly. Through this cut, work on your stomach can be done. A thoracoscope, which is much like a laparoscope, will also be inserted into a small cut made between your ribs. This cut allows the surgeon to work in the chest.

After the cancer is removed, your stomach will need to be attached to your remaining esophagus. It may be directly attached or a piece of your intestine may be used to connect the two organs. As you heal from surgery, you will receive food from a J-tube that is inserted through your belly wall and into your intestine.



## What to expect: Radiation therapy

- ✓ You will first have a planning session called a simulation. You will be placed in the treatment position, and a CT scan will be done. You will need to lie on your back and stay very still. You may get fitted for a prop to help you stay still during the radiation sessions. The CT scan images are used to make your radiation plan. The plan describes the best radiation dose for you, as well as the number of sessions you will need.
- ✓ You may feel very tired after radiation therapy. It may also be uncomfortable or difficult for you to swallow.

## Radiation therapy

Radiation therapy uses high-energy waves similar to x-rays to kill cancer cells. Radiation therapy and chemotherapy are usually used together to treat esophageal cancer. This combined treatment is called chemoradiation.

The type of radiation used most often is called EBRT (**external beam radiation therapy**). In EBRT, a large machine aims radiation at the part of your body where the tumor is. The radiation passes through your skin and other tissue to reach the tumor. **See Figure 10.**

There is more than one kind of EBRT. For esophageal cancer, 3D-CRT (**three-dimensional conformal radiation therapy**) is often recommended. In 3D-CRT, the radiation beams match the shape of your tumor to help prevent damage to the healthy tissue around the tumor.

Sometimes, important organs are too close to the tumor to use 3D-CRT. These include the heart, lungs, liver, and kidneys. In this case, IMRT (**intensity-modulated radiation therapy**) may be used. IMRT is more precise and is better at avoiding vital organs. IMRT works by aiming multiple beams of radiation at the tumor from many angles. The beams match the shape of your tumor and the intensity of each beam is varied to help the radiation to match the shape of the tumor.

**Figure 10. A patient having radiation therapy**

A large machine aims radiation at the part of your body where the tumor is. The radiation passes through your skin and other tissue to reach the tumor.



## Chemotherapy

Chemotherapy (“chemo” for short) is treatment with drugs to kill cancer cells. You might have it as part of your treatment for early or advanced esophageal cancer. Chemotherapy and radiation therapy are usually used together to treat esophageal cancer. This combined treatment is called chemoradiation.

Most chemotherapy drugs for esophageal cancer are liquids that are slowly injected into a vein. The drugs travel in your bloodstream to treat cancer throughout your body. Treatments that affect the whole body are called “systemic.” You are most likely to have a combination of two or three chemotherapy drugs. Chemotherapy can be given in different settings. Many people get chemotherapy at cancer centers. **See Figure 11.**

Chemotherapy is given in cycles of treatment days followed by days of rest. This allows your body to recover before the next cycle. For example, you might receive chemotherapy every day for 1 week, followed by 3 weeks with no chemotherapy. These 4 weeks make up one cycle. Cycles vary in length depending on which drugs are used. Often, a cycle is 14, 21, or 28 days long.

Chemotherapy and other drugs used to treat esophageal cancer are listed in [Guide 9](#).

**Figure 11. Chemotherapy treatment room**

**Chemotherapy is often given in treatment rooms, which allow several people to receive treatment at the same time.**



## Guide 9. Drugs for esophageal cancer

### Chemotherapy drugs

Generic name	Brand name (sold as)
capecitabine	Xeloda®
carboplatin	--
cisplatin	--
docetaxel	Taxotere®
epirubicin	Ellence®
etoposide	Etopophos®
fluorouracil (5-FU)	--
irinotecan	Camptosar®
oxaliplatin	Eloxatin®
paclitaxel	Taxol®

### Targeted therapy drugs

Generic name	Brand name (sold as)
ramucirumab	Cyramza®
trastuzumab	Herceptin®, Ogivri®

### Immunotherapy drugs

Generic name	Brand name (sold as)
pembrolizumab	Keytruda®



## What to expect: Chemotherapy

- ✓ Side effects of chemotherapy depend on many things and are different for everyone.
- ✓ Common side effects include nausea, not feeling hungry, diarrhea, hair loss, and mouth sores.
- ✓ Some chemotherapy drugs can damage your sensory nerves. Symptoms of this include numbness, tingling, and pain in fingers and toes. You may also be sensitive to cold and feel pain to light touch. This side effect can last a long time for some people.

## Targeted therapy

There are very small particles in your body called molecules. Some of the molecules help cancer to grow and spread. Targeted therapies are drugs that target—and stop—these molecules from helping the cancer grow.

Not everyone with esophageal cancer will be treated with a targeted therapy. There are two targeted therapies available for treating some advanced cancers found in the area where the stomach and esophagus meet (the EGJ [esophagogastric junction]). Treatment with a targeted therapy is only given if other treatments have failed.

### Ramucirumab

Tumors need new blood vessels to grow. VEGF (vascular endothelial growth factor) is a protein that causes new blood vessels to form. Some cancer cells have receptors for VEGF. This means that some cancer cells have a “landing pad” where VEGF can “land” (attach) and create new blood vessels for the tumor to get bigger.

Ramucirumab shuts down the landing pad so that VEGF can't attach and no new blood vessels can be made. It is used alone or with a chemotherapy drug called paclitaxel.

### Trastuzumab

In Part 2 we learned about HER2 (human epidermal growth factor receptor 2). To recap, HER2 is a protein found on the surface of cells. Some esophageal adenocarcinomas have too much HER2 protein, which causes the cancer to grow and spread more quickly than it normally would.

Trastuzumab helps kill the cancer cells that have too much HER2. If testing shows that your adenocarcinoma has too much HER2, your doctor may want to add trastuzumab to your chemotherapy.



## SNAPSHOT

### Targeted therapy

- ✓ This type of therapy is used to treat some advanced cancers in the area where the esophagus and stomach meet .
- ✓ Like chemotherapy, targeted therapies are given by infusion. This means they are injected into a vein and then enter your bloodstream.
- ✓ Ramucirumab is given alone or with chemotherapy; trastuzumab is given with chemotherapy.
- ✓ Some common side effects of ramucirumab are high blood pressure, diarrhea, nosebleeds, and fatigue.
- ✓ The first dose of trastuzumab may make you feel like you have the flu. This is less common after the second and third doses.

## Immunotherapy

The immune system is your body's natural defense against infection and disease. A newer type of cancer treatment called immunotherapy increases the activity of your immune system. By doing so, it improves your body's ability to find and destroy cancer cells.

### **Pembrolizumab**

Your immune system has important white blood cells called T cells. T cells' main job is to attack harmful things in your body, like bacteria, viruses, and cancer. They do this with the help of a protein on their surface called PD-1. Cancer cells have a different protein on their surface called PD-L1. When PD-1 and PD-L1 meet, it is called an immune checkpoint. The T cell is "told" to leave the cancer cell alone instead of attacking it.

A type of drug called an immune checkpoint inhibitor stops these two proteins from meeting. This means that the T cells will do their job and attack the cancer cells. Pembrolizumab is an example of an immune checkpoint inhibitor. If your tumor tested positive for certain tumor markers, you may be offered treatment with pembrolizumab. There are other factors that your doctors will weigh to find out if treatment with pembrolizumab is right for you.



When I was diagnosed it totally took me my surprise, leaving me very confused about next steps. The first step for me was to compile an excellent medical team who I felt comfortable with and trusted with my life. The support of family and friends was what got me through a very rigorous treatment plan and surgery. It is so very important to 'never lose hope'! There is life after cancer.

– Karen

Esophageal cancer survivor

## Clinical trials

New tests and treatments aren't offered to the public as soon as they're made. They first need to be studied. A clinical trial is a type of research study that involves people.

Clinical trials study how safe and helpful tests and treatments are. When found to be safe and helpful, they may become tomorrow's standard of care. Because of clinical trials, the tests and treatments in this book are now widely used to help people with esophageal cancer.

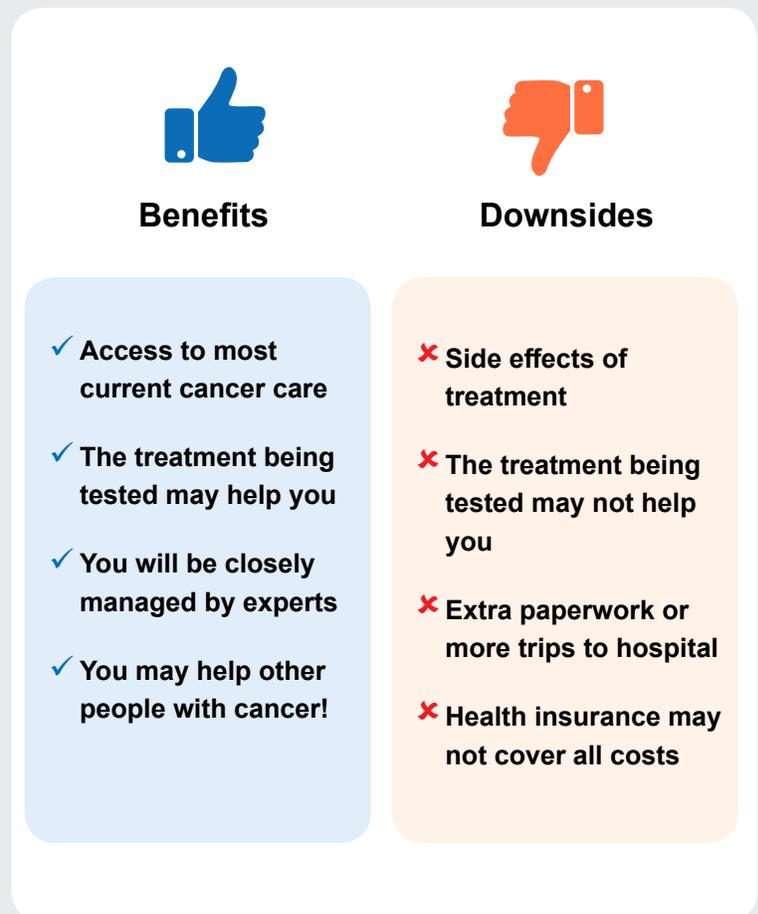
Joining a clinical trial can have both upsides and downsides. **See Figure 12** for some of the things you may want to think about when deciding to join a clinical trial. You will need to weigh the pros and cons and decide what is right for you.

To join a clinical trial, you must meet the conditions of the study. Patients in a clinical trial are often alike in terms of their cancer and general health. This is to know that any progress is because of the treatment and not because of differences between patients.

To join, you'll need to review and sign a paper called an informed consent form. This form describes the study in detail. The study's risks and benefits should be described and may include others than those described above.

Ask your treatment team if there is an open clinical trial that you can join. There may be clinical trials where you're getting treatment or at other treatment centers nearby. You can also find clinical trials through the websites listed in Part 7.

**Figure 12. Possible benefits and downsides of joining a clinical trial**



## Review

- ▶ Endoscopic therapies are used for treating very small esophageal tumors.
- ▶ An esophagectomy is major surgery to remove some (or all) of the esophagus, along with nearby lymph nodes.
- ▶ Radiation therapy uses high-energy rays to kill cancer cells or stop new cancer cells from being made.
- ▶ Chemotherapy is treatment with drugs to kill cancer cells. Most chemotherapy drugs for esophageal cancer are liquids that are slowly injected into your vein.
- ▶ Treatment with targeted therapy or immunotherapy may be an option for some patients with metastatic adenocarcinoma in the area where the esophagus and stomach meet.
- ▶ Clinical trials give people access to new tests and treatments that they wouldn't normally get. These new tests and treatments may, in time, be approved by the FDA.

# 5

## Treatment guide: Squamous cell carcinoma

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This chapter describes the treatment options for squamous cell carcinoma of the esophagus. Treatment options are grouped by the extent of the cancer. Your doctors may suggest other treatments than those listed here based on your health and personal wishes.

## Early cancer

Before beginning treatment, your doctor will assess if you are able to have surgery by testing your lungs, heart, and nutritional intake. Before beginning any treatment, your entire treatment team should give input on your cancer and treatment options. If they think you might be too weak for some treatments, you may need to get nutrients through a feeding tube. This will help you get as strong as possible for treatment.

### Patients able to have surgery

This section presents options for treating early squamous cell carcinomas in people able to have surgery. The treatment options in this section are for tumors that haven't grown beyond the second layer of the esophagus wall, and for when there is no cancer in lymph nodes. This includes the following tumor sizes:

- Tis
- T1a tumors
- Some T1b tumors

There are two main ways to treat Tis and T1a tumors—endoscopic therapies and surgery. Endoscopic therapies are preferred by NCCN experts for treating most Tis and T1a tumors. Endoscopic therapies include resection and ablation.



### SNAPSHOT

## Esophageal squamous cell carcinoma

- ✓ Starts in flat, thin cells that line the surface of the esophagus
- ✓ Usually found in the middle or top part of the esophagus
- ✓ Risk factors include tobacco use, alcohol use, and regularly drinking very hot drinks

Endoscopic treatment of these very early cancers is shown in [Guide 10](#).

Surgery (esophagectomy) is another option for some patients with Tis or a T1a tumor. Surgery may be the best treatment if the tumor has grown over a large area, or if your doctor doesn't think endoscopic therapies will work well. Surgery for Tis and T1a tumors is described in [Guide 11](#). If you do have surgery as your primary treatment, you may receive chemotherapy and radiation (chemoradiation) to kill any remaining cancer cells.

If your tumor is in the T1b category, it means that it has grown through the first layer and into the second layer of the esophagus wall. Treatment of T1b squamous cell carcinomas depends on whether

Guide 10. Endoscopic treatment of early squamous cell carcinomas

Tumor	Endoscopic treatment options	What's next?
Tis	 <p>Endoscopic resection alone <b>OR</b> Ablation <b>OR</b> Endoscopic resection followed by ablation</p>	<p>Monitoring for return of cancer:</p> <ul style="list-style-type: none"> <li>• History and physical exam every 3–6 months for 1–2 years, every 6–12 months for 3–5 years, then once a year after that.</li> <li>• Upper GI endoscopy every 3 months for the first year, then every 6 months for the second year, and then once a year after that.</li> <li>• Blood tests as needed.</li> </ul>
T1a	 <p>Endoscopic resection alone <b>OR</b> Endoscopic resection followed by ablation</p>	

Guide 11. Surgical treatment of early squamous cell carcinomas

Tis, T1a, and T1b tumors with no cancer in lymph nodes

Results of surgery	Next treatment	What's next?
All cancer was removed	→	Start follow-up care
Some cancer remains	→ Chemotherapy and radiation	→ Start follow-up care
A lot of cancer remains or cancer has spread far	 <p>Chemotherapy and radiation <b>OR</b> Supportive care</p>	→ Start follow-up care
		→ See <i>Advanced Cancer</i> section

cancer was found in nearby lymph nodes during an endoscopic ultrasound. If the endoscopic ultrasound found no cancer in nearby lymph nodes, surgery (esophagectomy) is recommended. Treatment of T1b tumors with no cancer in lymph nodes is shown in Guide 11. Treatment of all other T1b squamous cell carcinomas is covered in the next section.

**Patients unable to have surgery**

Patients with early cancer who are unable to have surgery are treated with endoscopic therapies. If you have Tis or a T1a tumor, NCCN experts prefer endoscopic treatment over surgery for most patients, anyway. See Guide 12 for your treatment options if you can't have surgery.

**Guide 12. Treatment of early squamous cell carcinomas in patients unable to have surgery**

Tumor	Endoscopic treatment options	What's next?
Tis	 <p>Endoscopic resection alone  <b>or</b>                      Ablation  <b>or</b>                      Endoscopic resection followed by ablation</p>	 <p>Start follow-up care</p>
T1a	 <p>Endoscopic resection alone  <b>or</b>                      Endoscopic resection followed by ablation</p>	 <p>Start follow-up care</p>
T1b, no cancer in lymph nodes	 <p>Endoscopic resection alone  <b>or</b>                      Endoscopic resection followed by ablation</p>	 <p>Start follow-up care  <b>or</b>                      Chemotherapy and radiation (for some patients)</p>

## Invasive cancer

### Patients able to have surgery

This section presents options for treating T1b, T2, T3, and T4a tumors when there is (or may be) cancer in nearby lymph nodes. There is more than one option for treating these tumors in patients healthy enough for surgery. The options are listed below.

- **Chemoradiation, then surgery (if possible).** In this option, chemotherapy and radiation are given first to try to shrink the tumor before surgery. This is called preoperative chemoradiation. Then, if possible, surgery is used to remove the area of the esophagus where the tumor is. This treatment option is for tumors that aren't near your neck. [See Guide 13](#) for this treatment option.
- **Surgery (esophagectomy).** Surgery is a primary treatment option for some T1b and T2 tumors. To have surgery as the primary treatment, there are a few rules (see below). Surgical treatment of T1b and T2 tumors is presented in [Guide 14](#).
  - The T1b or T2 tumor should be very small and not near your neck.
  - The cancer cells should look a lot like normal cells (this means they are likely to grow and spread slowly).
  - There should be no cancer in the lymph nodes.
- **Chemoradiation alone.** Chemoradiation given to try to cure cancer without using other treatments is called “definitive chemoradiation.”

This is the best treatment for you if:

- the tumor is in your neck area, or
- you are healthy enough to have surgery, but don't want to have surgery

T4b is the biggest esophageal tumor category. T4b tumors have grown through all 4 layers of the esophagus wall and into important nearby body parts, like the windpipe or the aorta. Most T4b tumors are treated with definitive chemoradiation. Depending on how well the chemoradiation worked, surgery may be an option.

If the cancer has entered your windpipe, heart, or the large vessels that bring blood to and from the heart, treatment with chemotherapy alone may be better for you. Your doctor will consider whether treatment with definitive chemoradiation or chemotherapy alone is more appropriate for your specific situation. [See Guide 15](#) for treatment of T4b squamous cell carcinomas.

### Patients unable to have surgery

If your treatment team feels that you aren't strong enough for surgery, other treatment options are available. If they think you are strong enough to have chemotherapy and radiation (chemoradiation), then that is the best option. After that, you would start follow-up care.

If you are not able to have chemoradiation, you will likely begin supportive care. Another option is to have radiation therapy alone before beginning supportive care. Radiation therapy at this stage is called “palliative” by doctors. The goal of this type of radiation therapy isn't to cure the cancer. The goal is to control any symptoms of the cancer or to prevent symptoms from occurring in the first place. It may help you have a better quality of life.

Guide 13. Preoperative chemoradiation for invasive squamous cell carcinomas T1b, T2, T3, and T4a tumors

Result of chemoradiation	Next treatment	Result of surgery	What's next?
No signs of cancer	Surgery	All cancer was removed	Start follow-up care
		Some cancer remains	Follow-up care OR Supportive care
		A lot of cancer remains, or the cancer has spread far	Follow-up care OR Supportive care
	Start follow-up care		Start follow-up care
Cancer still in same area	Surgery (NCCN preferred treatment)	All cancer was removed	Start follow-up care
		Some cancer remains	Follow-up care OR Supportive care
		A lot of cancer remains OR cancer has spread far	Follow-up care OR Supportive care
	Supportive care		see <i>Advanced Cancer</i> section
Cancer can't be removed with surgery OR has spread far	Supportive care		see <i>Advanced Cancer</i> section

Guide 14. Surgical treatment of selected T1b/T2 squamous cell carcinomas

Treatment	Result of surgery	Next treatment	What's next?
Surgery (esophagectomy)	All cancer was removed	→	Start follow-up care
	Some cancer remains	→ Chemotherapy and radiation	→ Start follow-up care
	A lot of cancer remains or cancer has spread	→ Chemotherapy and radiation or Supportive care	→
→			See <i>Advanced Cancer</i> section

Guide 15. Treatment of T4b squamous cell carcinomas

Treatment	Result of chemoradiation	Next treatment	What's next?
Definitive chemoradiation	No signs of cancer	→	Start follow-up care
	Cancer still in same area	→ Surgery	→ Start follow-up care
		→ Supportive care	→ See <i>Advanced Cancer</i> section
Cancer has spread far	→ Supportive care	→ See <i>Advanced Cancer</i> section	
Chemotherapy (for some patients)	→	Supportive care	→ See <i>Advanced Cancer</i> section

**Follow-up care**

Follow-up care is started when there are no signs of cancer after treatment. [Guide 16](#) lists follow-up care for invasive squamous cell carcinomas. Imaging tests are based on the stage of your cancer and which treatments you’ve had. For all invasive cancers, updates of your medical history and physical exams should be done regularly.

Blood tests are only done when needed. Surgery and radiation can narrow your esophagus. So, you may have trouble swallowing afterward. If this happens, your esophagus can be stretched using a small balloon or tube guided down your throat to the right spot. After treatment, it may also be helpful to meet with a nutritionist to make sure you are eating enough, especially enough healthy foods.

**Long-term follow-up care**

Esophageal cancer and its treatment can have long-term side effects. These long-term side effects range from being bothersome (acid reflux) to very serious (damage to your heart from chemotherapy). It is important to be aware of the side effects you might have so that you can talk to your doctor about them. [Guide 17](#) is a list of possible long-term side effects and ways that you (or your doctor) can help manage them.

An important part of life after cancer is trying to stay as healthy as possible. Your doctor will talk to you about things you can do to get (and stay) healthy.

**Guide 16. Follow-up care for invasive squamous cell carcinomas**

Tumor size	Treatment you’ve had	Follow-up care
<ul style="list-style-type: none"> <li>• T2</li> <li>• T3</li> <li>• T4a</li> <li>• T4b</li> </ul>	Chemotherapy AND radiation BUT NOT surgery	<ul style="list-style-type: none"> <li>• History and physical exam every 3–6 months for 1–2 years, every 6–12 months for 3–5 years, then once a year after that</li> <li>• Upper GI endoscopy every 3–6 months for the first 2 years, then every 6 months for the third year, then as needed</li> <li>• You may also have a CT scan every 6 months for up to 2 years</li> <li>• Widening of esophagus if needed</li> <li>• Blood tests as needed</li> <li>• Checks to make sure you’re getting enough nutrients</li> </ul>
<ul style="list-style-type: none"> <li>• T2</li> <li>• T3</li> <li>• T4a</li> <li>• T4b</li> </ul>	Chemotherapy AND radiation AND surgery	<ul style="list-style-type: none"> <li>• History and physical exam every 3–6 months for 1–2 years, every 6–12 months for 3–5 years, then once a year after that</li> <li>• You may have a CT scan every 6 months for up to 2 years</li> <li>• Widening of esophagus if needed</li> <li>• Blood tests as needed</li> <li>• Checks to make sure you’re getting enough nutrients</li> </ul>

## Guide 17. Long-term follow-up care for invasive squamous cell carcinomas

Issues or side effects you may have	Things that you or your doctor can do to help
Losing too much weight or not getting enough nutrients	<p>In the first 6 months after surgery, you are likely to lose weight because of eating less. This means that you may not be getting enough nutrients. Your doctor:</p> <ul style="list-style-type: none"> <li>• will monitor your weight and your nutrition;</li> <li>• may measure your vitamin B, folic acid, vitamin D, and calcium levels;</li> <li>• may refer you to a nutrition expert; and</li> <li>• will ask you about other aspects of your physical and emotional health that may be affecting your diet.</li> </ul>
Food doesn't move fast enough to the intestines ("delayed gastric emptying")	<ul style="list-style-type: none"> <li>• Try to eat 5 <u>small</u> meals throughout the day.</li> <li>• Try not to eat a lot of high-fiber or high-fat foods.</li> <li>• If your symptoms keep coming back, your doctor may refer you to an expert in digestive issues.</li> </ul>
Food moves too fast to the intestines ("dumping syndrome")	<ul style="list-style-type: none"> <li>• Eat 5 <u>small</u> meals throughout the day.</li> <li>• Try to eat a lot of protein and fiber. Try not to eat many sweets, like baked goods, cookies, or sugary cereals.</li> <li>• Try not to drink anything with your meals.</li> </ul>
Acid reflux symptoms	<ul style="list-style-type: none"> <li>• Try not to lie down flat after eating.</li> <li>• Use a foam wedge (triangle-shaped) pillow in bed, and try not to sleep totally flat.</li> <li>• Drugs called proton pump inhibitors may help some people with acid reflux.</li> </ul>
Trouble swallowing after surgery	<p>Tell your doctor if you are having trouble swallowing. He or she will look for possible causes. There may be ways to help.</p>
If you had diabetes or high blood pressure	<ul style="list-style-type: none"> <li>• Blood pressure and blood sugar levels may go down in the months after surgery because you're losing weight. So, your doctor will monitor you to see if your medication(s) needs to be changed or stopped.</li> </ul>
Damage to your heart from radiation	<ul style="list-style-type: none"> <li>• Your cancer doctor and your primary care doctor should work together to monitor and manage your risk for heart issues. Hypertension, diabetes, high cholesterol, and obesity can raise your risk of getting heart disease.</li> <li>• You may be referred to a cardiologist.</li> </ul>
Nerve problems from chemotherapy	<p>Chemotherapy can cause pain, numbness, tingling, or sensitivity in your hands and feet. If you have bad nerve pain, there is a drug called duloxetine that might help.</p>
Fatigue	<ul style="list-style-type: none"> <li>• Try to exercise when you have the energy, and rest when you need to.</li> <li>• Your doctor will ask you about other aspects of your physical and mental health that may be making you tired.</li> </ul>

The most important steps you can take towards living a healthy life are:

- Be active! Try to exercise most days of the week.
- Eat a healthy diet with lots of fruits and veggies.
- The less alcohol you drink, the better.
- If you are a smoker, quit!

Eating healthy and exercising will help you achieve (and keep) a healthy body weight. Being at a healthy weight is very important to your overall health. Some of the issues in Guide 17 may make it harder for you to be as active or as healthy as you'd like. Your doctor will have ideas on small changes you can make to work around any side effects you may have.

After cancer, your primary care doctor will play an important part in your lifelong health. Your cancer treatment team and your primary care doctor should work together to make sure that all aspects of your health are managed. This includes cancer-related care and non-cancer-related care. All of your doctors will agree that you should continue to be screened for other kinds of cancers that affect adults, like lung, prostate, colon, and breast cancers.

## If cancer comes back

The return of cancer after a cancer-free period is called a recurrence. [Guide 18](#) lists the treatment options for cancer that returns during follow-up care. Options for treating recurrent cancer are based on two things:

- The area where the cancer returned
- Which treatment(s) you've already had

Cancer that returns to the esophagus area is called a locoregional recurrence. The steps of treating this kind of recurrence are described in Guide 18.

Cancer that returns to other parts of your body far from the esophagus is called a metastatic recurrence. This kind of recurrence is addressed in the next section, *Advanced cancer*.

## Advanced cancer

Cancer that that can't be cured with treatment, or that has spread to other areas of your body, is called advanced cancer. Options to manage advanced cancer are based on your performance status, which is your ability to do activities. Your doctor will rate your performance status using one of two scales:

### ECOG (Eastern Cooperative Oncology Group) Performance Scale

- A score of 0 means you are fully active.
- A score of 1 means you are able to do all self-care activities but are unable to do hard physical work.
- A score of 2 means you are able to do all self-care activities and spend most of waking time out of bed but you are unable to do any work.
- A score of 3 means you are unable to do all self-care activities and any work and spend most of waking time in bed.
- A score of 4 means you are fully disabled.

### KPS (Karnofsky Performance Status)

- A score of 0 to 49 means you are unable to care for yourself.

- A score of 50 to 79 means you are unable to work and some assistance is needed.
- A score of 80 to 100 means you are able to do your normal work and activities.

The main options for managing advanced cancer are supportive care (also called palliative care) and

chemotherapy. The options are based on your ability to do activities (performance status). Supportive care is an option for all advanced cancers. If your health hasn't seriously limited your activities, chemotherapy is also an option to slow down cancer growth. [See Guide 19](#). Chemotherapy and supportive care are addressed next.

### Guide 18. Squamous cell carcinoma that returns to esophagus area

Treatment you've had		Next treatment options			What's next?	
Surgery but not chemotherapy and radiation			Chemotherapy and radiation OR		CT scan	Start follow-up care. If cancer returns, begin supportive care.
			Surgery OR			
			Chemotherapy alone OR			
			Supportive care			
Chemotherapy and radiation but not surgery	Can have surgery		Surgery		CT scan	Start follow-up care. If cancer returns, begin supportive care.
	Can't have surgery		Supportive care			See <i>Advanced Cancer</i> section

## Guide 19. Managing advanced squamous cell carcinoma

Performance status	Management options
Your KPS score is 60% or higher <b>OR</b> Your ECOG performance score is 0, 1, or 2	<ul style="list-style-type: none"> <li>• Chemotherapy to slow cancer growth</li> <li>• Supportive care</li> <li>• Clinical trials</li> </ul>
Your KPS score is 0% to 59% <b>OR</b> Your ECOG performance score is 3 or 4	<ul style="list-style-type: none"> <li>• Supportive care</li> <li>• Clinical trials</li> </ul>

### Chemotherapy for advanced cancer

If your health hasn't seriously limited your activities, chemotherapy is an option to slow down cancer growth. Your doctor will choose a chemotherapy regimen based on your health and the side effects of treatment. Regimens consisting of two drugs have less severe side effects than three-drug regimens. If you are given 5-FU, leucovorin may be added to limit side effects of the chemotherapy. No matter what regimen you receive, you should be assessed for side effects on a regular basis.

If your cancer doesn't respond to first-line regimens, your doctor will give you a different regimen. The regimen that is best for you depends on your prior treatment and performance status. Most preferred regimens have been shown in clinical trials to control cancer growth better than other treatments.

### Supportive care

Because the cancer can't be cured, the goal of supportive care is to make you more comfortable and to help keep the cancer under control. Supportive care may also help you live longer, improve your eating, and help you feel better overall. When used

for advanced cancers, supportive care is often called palliative care.

People without advanced cancer also receive supportive care. Some aspects of supportive care, such as symptom control, are useful for many people with any stage of esophageal cancer. Symptom control is addressed next.

Supportive care alone (without chemotherapy) is an option if you have an ECOG score of 3 or more or a KPS score of less than 60%. You may receive other types of care to improve your quality of life. Keep reading to learn about symptom control.

## Help with symptoms

Cancer or its treatment can cause unpleasant and sometimes harmful symptoms. One of the most common symptoms of esophageal cancer is trouble with food passing through the esophagus. This is called dysphagia. Your doctor will assess what you can and can't swallow and what is causing the dysphagia.

Dysphagia is often caused by the tumor blocking the passage. However, sometimes it is caused by the tumor impairing the muscles of the esophagus or by scarring from radiation. Treating dysphagia depends on what's causing it. Treatment options for a blocked esophagus are described in the next section.

Bleeding is another common symptom, but it's not as common as dysphagia. Bleeding may be caused by the cancer or the cancer treatment. Endoscopic treatment that uses heat, cold, lasers, or injections may stop bleeding from the tumor surface. External radiation therapy may stop ongoing blood loss.

Other symptoms related to esophageal cancer include pain and nausea, with or without vomiting. These symptoms may be caused by the tumor blocking the passage of the esophagus. Treatment options for a blocked esophagus are described in the next section. Pain may be controlled with radiation therapy, chemotherapy, and pain medication. There are medicines and other methods that may help stop nausea and vomiting.

You may have other symptoms that aren't listed here. Talk to your treatment team about your symptoms. There may be ways to help you feel better.

## Blocked esophagus

The cancer may block food and liquids from passing through your esophagus. Treatment is based on how much of the esophagus is blocked.

A **complete blockage** doesn't allow any food or liquids to pass through your esophagus. Treatment options include endoscopic methods, radiation therapy, chemotherapy, and sometimes surgery. Dilation and stents are endoscopic methods described below. External radiation is the common radiation method, but internal radiation (brachytherapy) may also be an option. You may also get a feeding tube to make sure you are getting enough nutrients.

A **severe blockage** allows only liquids to pass through your esophagus. One treatment option is to stretch open (dilate) your esophagus. Your esophagus can be stretched using a small balloon or tube guided down your throat to the right spot.

Another option for a severe blockage is a stent. A stent is a thin metal or plastic tube. It can be placed in your esophagus with endoscopic tools while you are sedated. The stent will expand in the passage and remain in your body to allow food to pass through. Stents are not used for patients who may undergo surgery to treat the cancer.

Other options for a severe blockage are the same as for a complete blockage. External and internal radiation therapy work well to unblock the esophagus. Symptom relief from radiation is slower but more long-lasting compared to endoscopic methods.

A **moderate blockage** allows liquids and semisolid foods to pass through your esophagus. Applesauce and cottage cheese are examples of semisolid foods. Treatment options are the same as for complete and severe blockage.

### Clinical trials

A clinical trial is a type of research study that involves people. NCCN believes that the best management for any cancer patient is in a clinical trial. Ask your treatment team if there is an open clinical trial that you can join. Clinical trials are discussed in more detail at the end of Part 4. You can use the websites in Part 7 to find clinical trials near you.

## Review

- Endoscopic therapies are preferred over surgery for Tis and T1a tumors. Surgery may be an option, though, if the Tis or T1a tumor has grown over a large area, or if your doctor doesn't think endoscopic therapies will work.
- If there is no cancer in nearby lymph nodes, surgery is recommended for treating T1b tumors. After surgery you may receive chemotherapy and radiation (chemoradiation) to kill any remaining cancer cells.
- The main treatments for invasive squamous cell carcinoma not in the neck area are chemotherapy and radiation, followed by surgery. Treatment with only chemotherapy and radiation (no surgery) is an option for tumors in the neck area, and for people who can't have (or don't want to have) surgery.
- Most T4b tumors are treated with chemotherapy and radiation to try to cure the cancer. Chemotherapy alone is an option for some patients whose cancer has entered very important organs.
- Tumors in the neck area cannot be treated first with surgery. These tumor types are treated with definitive chemotherapy and radiation to try to cure the cancer.
- When there are no signs of cancer after treatment, follow-up care begins. This involves testing to check for new cancer growth.
- Cancer that returns near to where the esophagus is (or was) may be curable with surgery or chemoradiation.
- Cancer that can't be cured can be managed with supportive care. Treatment with chemotherapy to slow down the cancer growth is also an option for some people with advanced squamous cell carcinoma. Participation in clinical trials is another option and is especially encouraged by NCCN.

# 6

## Treatment guide: Adenocarcinoma

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This chapter walks you through the options for treating an esophageal adenocarcinoma. The treatments are grouped by the extent of the cancer. Your doctors may suggest treatments not listed here based on your health and personal wishes.

## Early cancer

Before beginning treatment, your doctor will assess if you are able to have surgery by testing your lungs, heart, and nutritional intake. Before beginning any treatment, your entire treatment team should give input on your cancer and treatment options. If they think you might be too weak for some treatments, you may need to get nutrients through a feeding tube. This will help you get as strong as possible for treatment.

### Patients able to have surgery

This section presents options for treating early adenocarcinomas in people healthy enough to have surgery. The treatment options in this section are for tumors that haven't grown beyond the second layer of the esophagus wall, and for when there is no cancer in lymph nodes. This includes the following tumor sizes:

- Tis (abnormal cells that could become cancer)
- T1a tumors
- Superficial (not deep) T1b tumors
- T1b tumors with no cancer in lymph nodes



### SNAPSHOT

## Esophageal adenocarcinoma

- ✓ Most common type of esophagus cancer in the U.S.
- ✓ Usually found at the bottom of the esophagus, near the stomach
- ✓ More common in men than women
- ✓ Risk factors include obesity, GERD, and Barrett's esophagus

### Tis and T1a tumors

Endoscopic treatment is preferred by NCCN experts for treating most Tis and T1a tumors. Endoscopic treatment includes endoscopic resection (ESD and EMR) and ablation. Endoscopic treatment of Tis and T1a tumors is presented in [Guide 22](#).

Surgery (esophagectomy) is another option for some patients. Surgery may be the best treatment if the Tis or T1a tumor has grown over a large area, or if the tumor isn't expected to respond well to endoscopic therapies. Surgical treatment of early adenocarcinomas is shown in [Guide 23](#).

## Guide 20. Endoscopic treatment of early adenocarcinomas

Tumor	Endoscopic treatment options	What's next?
Tis	 <ul style="list-style-type: none"> <li>Endoscopic resection alone</li> <li>OR</li> <li>Ablation</li> <li>OR</li> <li>Endoscopic resection followed by ablation</li> </ul>	 <p>Monitoring for return of cancer:</p> <ul style="list-style-type: none"> <li>• History and physical exam every 3–6 months for 1–2 years, every 6–12 months for 3–5 years, then once a year after that.</li> <li>• Upper GI endoscopy every 3 months for the first year, then every 6 months for the second year, and then once a year after that.</li> <li>• Blood tests as needed.</li> </ul>
T1a	 <ul style="list-style-type: none"> <li>Endoscopic resection alone</li> <li>OR</li> <li>Endoscopic resection followed by ablation</li> </ul>	
Superficial (not deep) T1b	 <ul style="list-style-type: none"> <li>Endoscopic resection followed by ablation</li> </ul>	 <p>Monitoring for return of cancer:</p> <ul style="list-style-type: none"> <li>• Upper GI endoscopy every 3 months for the first year, then every 4–6 months for the second year, and then once a year after that.</li> <li>• Endoscopic ultrasound may be done at same time as upper GI endoscopy</li> <li>• You may also have a CT scan once a year for up to 3 years, and then as needed after that.</li> <li>• Blood tests as needed.</li> </ul>

**T1b tumors**

Treatment of a T1b tumor depends on a few things:

- how deep into the second layer of the esophagus wall the tumor has grown, and
- if an endoscopic ultrasound found cancer in nearby lymph nodes.

If the tumor has just grown a tiny bit into the second layer, this is called a “superficial” T1b tumor. The treatment options for superficial T1b tumors are similar to those for Tis and T1a tumors. Endoscopic treatment ([Guide 20](#)) and surgery ([Guide 21](#)) are options.

T1b tumors that are not superficial are treated differently. The main difference is that endoscopic treatment is not an option. If an endoscopic ultrasound didn't find any cancer in your lymph nodes, surgery is the recommended treatment. Treatment of T1b tumors with no cancer in lymph nodes is shown in [Guide 22](#).

**Patients unable to have surgery**

Patients with early cancer who are unable to have surgery are treated with endoscopic treatments. If you have Tis or a T1a tumor, NCCN experts prefer endoscopic treatment over surgery for most patients, anyway. See [Guide 20](#) for your treatment options if you can't have surgery.

## Guide 21. Surgical treatment of early adenocarcinomas

Tis, T1a, and superficial (not deep) T1b tumors

Results of surgery		Next treatment		What's next?
All cancer was removed and cancer <b>wasn't</b> found in lymph nodes			→	Start follow-up care
All cancer was removed and cancer <b>was</b> found in lymph nodes	↳	Chemotherapy and radiation OR Chemotherapy alone	→	Start follow-up care
Some cancer remains	→	Chemotherapy and radiation	→	Start follow-up care
A lot of cancer remains or cancer has spread far	↳	Chemotherapy and radiation	→	Start follow-up care
		OR Supportive care	→	See <i>Advanced Cancer</i> section

## Guide 22. Treatment of T1b adenocarcinomas with no cancer in lymph nodes

Treatment	Results of surgery	Next treatment	What's next?
Surgery	All cancer was removed	No cancer in lymph nodes	→ Start follow-up care
		Cancer in lymph nodes	↳ Chemotherapy and radiation → Start follow-up care
	↳ Chemotherapy → Start follow-up care		
	Some cancer remains	→ Chemotherapy and radiation → Start follow-up care	
	A lot of cancer remains or cancer has spread far	↳	Chemotherapy and radiation → Start follow-up care
			Supportive care → see <i>Advanced Cancer</i> section

## Invasive cancer

### Patients able to have surgery

#### T1b, T2, T3, and T4a tumors

This section presents options for treating T1b, T2, T3, and T4a tumors when there is (or may be) cancer in nearby lymph nodes. There are several primary treatment options for invasive adenocarcinomas in patients healthy enough for surgery:

- ▶ **Chemotherapy and radiation, then surgery (if possible).** This is the treatment preferred by NCCN experts. This treatment option is shown in [Guide 23](#).
- ▶ **Chemotherapy, then surgery, then chemotherapy again.** Another option is to receive chemotherapy before and after surgery. This is called “perioperative chemotherapy.” See [Guide 24](#) for the steps in this treatment option.
- ▶ **Chemotherapy, then surgery.** Instead of having chemotherapy before and after surgery, you may just have it before. This is called “preoperative chemotherapy.” This option is also described in [Guide 24](#).
- ▶ **Surgery (esophagectomy).** Surgery is a primary treatment option for some T1b and T2 tumors. To have surgery as the primary treatment, there are a few rules (see below). The steps of this treatment path are explained in [Guide 25](#).
  - The T1b or T2 tumor should be very small.
  - The cancer cells should look a lot like normal cells (this means they are likely to grow and spread slowly).

- There should be no cancer in the lymph nodes.

- ▶ **Only chemotherapy and radiation (no surgery).** Chemoradiation given to try to cure cancer without using other treatments is called “definitive chemoradiation.” This option is for patients who are healthy enough, but who don’t want to have surgery.

#### T4b tumors

T4b is the biggest esophageal tumor category. T4b tumors have grown through all 4 layers of the esophagus wall and into important nearby body parts, like the windpipe or the aorta. Most T4b tumors are treated with definitive chemoradiation (chemotherapy and radiation) to try to cure the cancer. Depending on how well the chemoradiation worked, surgery may be an option.

If the cancer has entered your windpipe, heart, or the large vessels that bring blood to and from the heart, treatment with chemotherapy alone may be a better option for you. Your doctor will consider whether treatment with definitive chemoradiation or chemotherapy alone is more appropriate for you.

Treatment of T4b tumors in patients healthy enough for surgery is covered in [Guide 26](#).

Guide 23. Preoperative chemoradiation for invasive adenocarcinomas  
T1b, T2, T3, and T4a tumors

Result of chemoradiation	Next treatment	Result of surgery	What's next?
No signs of cancer	Surgery (NCCN-preferred treatment)	All cancer was removed	Start follow-up care
		Some cancer remains	Start follow-up care OR You may have another surgery
		A lot of cancer remains OR cancer has spread far	Supportive care (see <i>Advanced Cancer</i> section)
	Start follow-up care		Start follow-up care
Cancer still in same area	Surgery (NCCN-preferred treatment)	All cancer was removed	Start follow-up care
		Some cancer remains	Start follow-up care OR You may have another surgery
		A lot of cancer remains OR cancer has spread far	Supportive care (see <i>Advanced Cancer</i> section)
	Supportive care		See <i>Advanced Cancer</i> section
Cancer can't be removed with surgery OR has spread far	Supportive care		See <i>Advanced Cancer</i> section

Guide 24. Treatment of invasive cancer with chemotherapy and surgery

Primary treatment	Results of surgery	Next treatment	What's next?
Chemotherapy, then surgery	All cancer was removed and cancer <b>wasn't</b> found in lymph nodes	Observation	Start follow-up care
		Chemotherapy (if received peri-operatively)	
	All cancer was removed and cancer <b>was</b> found in lymph nodes	Observation	Start follow-up care
		Chemotherapy and radiation	
		Chemotherapy (if received perioperatively)	
	Some cancer remains	Chemotherapy and radiation	Start follow-up care
	A lot of cancer remains or cancer has spread far	Chemotherapy and radiation OR Supportive care	Start follow-up care
			See <i>Advanced Cancer</i> section

Guide 25. Surgical treatment of selected T1b/T2 adenocarcinomas

Treatment	Result of surgery	Next treatment	What's next?
Surgery (esophagectomy)	All cancer was removed and cancer <b>wasn't</b> found in lymph nodes	→ T1b tumors: observation	Start follow-up care
		→ T2 tumors: observation, OR chemotherapy and radiation in some cases	
	All cancer was removed and cancer <b>was</b> found in lymph nodes	→ Chemotherapy and radiation	
		→ Chemotherapy	
	Some cancer remains	→ Chemotherapy and radiation	
A lot of cancer remains or cancer has spread	→ Chemotherapy and radiation	Start follow-up care	
	→ Supportive care	See <i>Advanced Cancer</i> section	

Guide 26. Treatment of T4b adenocarcinomas

Treatment	Result of chemoradiation	Next treatment	What's next?
Definitive chemoradiation	No signs of cancer	→	Start follow-up care
	Cancer still in same area	→ Surgery	Start follow-up care
		→ Supportive care	see <i>Advanced Cancer</i> section
Cancer has spread far	→ Supportive care	See <i>Advanced Cancer</i> section	
Chemotherapy (for some patients)	→	Supportive care	See <i>Advanced Cancer</i> section

## Patients unable to have surgery

If your treatment team feels that you aren't strong enough for surgery, other treatment options are available. If they think you are strong enough to have chemotherapy and radiation (chemoradiation), then that is the best option. After that, you would start follow-up care.

If you are not able to have chemoradiation, you will likely begin supportive care. Another option is to have radiation therapy alone before beginning supportive care. Radiation therapy at this stage is called "palliative" by doctors. The goal of this type of radiation therapy isn't to cure the cancer. The goal is to control any symptoms of the cancer or to prevent symptoms from occurring in the first place. It may help you have a better quality of life.

## Follow-up care

Follow-up care is started when there are no signs of cancer after treatment. [Guide 27](#) lists follow-up care for invasive adenocarcinomas. Imaging tests are based on cancer staging and which treatments you've had. For all invasive cancers, updates of your medical history and physical exams should be done regularly.

Blood tests are only done when needed. Surgery and radiation can narrow your esophagus. So, you may have trouble swallowing afterward. If this happens, your esophagus can be stretched using a small balloon or tube guided down your throat to the right spot. After treatment, it may also be helpful to meet with a nutritionist to make sure you are eating enough, especially enough healthy foods.

## Long-term follow-up care

Esophageal cancer and its treatment can have long-term side effects. These long-term side effects range from being bothersome (acid reflux) to very serious (damage to your heart from chemotherapy). It is important to be aware of the side effects you may

have so that you can talk to your doctor about them. [Guide 28](#) is a list of the issues you might have and ways that you (or your doctor) can help manage them.

An important part of life after cancer is trying to stay as healthy as possible. Your doctor will talk to you about things you can do to get (and stay) healthy. The most important steps you can take towards living a healthy life are:

- Be active! Try to exercise most days of the week.
- Eat a healthy diet with lots of fruits and veggies.
- The less alcohol you drink, the better.
- If you are a smoker, quit!

Eating healthy and exercising will help you achieve (and keep) a healthy body weight. Being at a healthy weight is very important to your overall health. Some of the issues in [Guide 28](#) may make it harder for you to be as active or as healthy as you'd like. Your doctor will have ideas on small changes you can make to work around any side effects that you may have.

After cancer, your primary care doctor will play an important part in keeping you healthy for the rest of your life. Your cancer treatment team and your primary care doctor should work together to make sure that all aspects of your health are managed. This includes cancer-related care and non-cancer related care. All of your doctors will agree that you should continue to be screened for other kinds of cancer that you are at average risk of getting.

## Guide 27. Follow-up care for invasive adenocarcinomas

Tumor size	Treatment you've had	Follow-up care
<ul style="list-style-type: none"> <li>• T2</li> <li>• T3</li> <li>• T4a</li> <li>• T4b</li> </ul>	Chemotherapy and radiation BUT NOT surgery	<ul style="list-style-type: none"> <li>• History and physical exam every 3–6 months for 1–2 years, every 6–12 months for 3–5 years, then once a year after that</li> <li>• Upper GI endoscopy every 3–6 months for the first 2 years, then every 6 months for the third year, then as needed</li> <li>• You may also have a CT scan every 6 months for up to 2 years</li> <li>• Widening of esophagus if needed</li> <li>• Blood tests as needed</li> <li>• Checks to make sure you're getting enough nutrients</li> </ul>
<ul style="list-style-type: none"> <li>• T2</li> <li>• T3</li> <li>• T4a</li> <li>• T4b</li> </ul>	Chemotherapy and radiation AND surgery	<ul style="list-style-type: none"> <li>• History and physical exam every 3–6 months for 1–2 years, every 6–12 months for 3–5 years, then once a year after that</li> <li>• You may have a CT scan every 6 months for up to 2 years</li> <li>• Widening of esophagus if needed</li> <li>• Blood tests as needed</li> <li>• Checks to make sure you're getting enough nutrients</li> </ul>

## Guide 28. Long-term follow-up care for invasive adenocarcinomas

Issues or side effects you may have	Things that you or your doctor can do to help
Losing too much weight or not getting enough nutrients	<p>In the first 6 months after surgery, you are likely to lose weight because of eating less. This means that you may not be getting enough nutrients. Your doctor:</p> <ul style="list-style-type: none"> <li>• will monitor your weight and your nutrition;</li> <li>• may measure your vitamin B, folic acid, vitamin D, and calcium levels;</li> <li>• may refer you to a nutrition expert; and</li> <li>• will ask you about other aspects of your physical and emotional health that may be affecting your diet.</li> </ul>
Food doesn't move fast enough to the intestines ("delayed gastric emptying")	<ul style="list-style-type: none"> <li>• Try to eat 5 <u>small</u> meals throughout the day.</li> <li>• Try not to eat a lot of high-fiber or high-fat foods.</li> <li>• If your symptoms keep coming back, your doctor may refer you to an expert in digestive issues.</li> </ul>
Food moves too fast to the intestines ("dumping syndrome")	<ul style="list-style-type: none"> <li>• Eat 5 <u>small</u> meals throughout the day.</li> <li>• Try to eat a lot of protein and fiber. Try not to eat many sweets, like baked goods, cookies, or sugary cereals.</li> <li>• Try not to drink anything with your meals.</li> </ul>
Acid reflux symptoms	<ul style="list-style-type: none"> <li>• Try not to lie down flat after eating.</li> <li>• Use a foam wedge (triangle-shaped) pillow in bed, and try not to sleep totally flat.</li> <li>• Drugs called proton pump inhibitors may help some people with acid reflux.</li> </ul>
Trouble swallowing after surgery	<p>Tell your doctor if you are having trouble swallowing. He or she will look for possible causes. There may be ways to help.</p>
If you had diabetes or high blood pressure	<ul style="list-style-type: none"> <li>• Blood pressure and blood sugar levels may go down in the months after surgery because you're losing weight. So, your doctor will monitor you to see if your medication(s) needs to be changed or stopped.</li> </ul>
Damage to your heart from radiation	<ul style="list-style-type: none"> <li>• Your cancer doctor and your primary care doctor should work together to monitor and manage your risk for heart issues. Hypertension, diabetes, high cholesterol, and obesity can raise your risk of getting heart disease.</li> <li>• You may be referred to a cardiologist.</li> </ul>
Nerve problems from chemotherapy	<p>Chemotherapy can cause pain, numbness, tingling, or sensitivity in your hands and feet. If you have bad nerve pain, there is a drug called duloxetine that might help.</p>
Fatigue	<ul style="list-style-type: none"> <li>• Try to exercise when you have the energy, and rest when you need to.</li> <li>• Your doctor will ask you about other aspects of your physical and mental health that may be making you tired.</li> </ul>

## If cancer comes back

The return of cancer after being cancer-free is called a recurrence. Options for treating recurrent cancer are based on two things:

- The area where the cancer returned
- Which treatment(s) you've already had

Cancer that returns to the esophagus area is called a locoregional recurrence. Treatment of this kind of recurrence is explained in [Guide 29](#). Cancer that returns to other parts of your body far from the esophagus is called a metastatic recurrence. Cancer that returns to other parts of your body far from the esophagus is called a metastatic recurrence. Metastatic cancer is addressed in the next section, *Advanced cancer*.

## Advanced cancer

Cancer that can't be cured with treatment, or that has spread to other areas of your body, is called advanced cancer. Options to manage advanced cancer are based on your performance status, which is your ability to do activities. Your doctor will rate your performance status using one of two scales:

ECOG (Eastern Cooperative Oncology Group) Performance Scale

- A score of 0 means you are fully active.
- A score of 1 means you are able to do all self-care activities but are unable to do hard physical work.
- A score of 2 means you are able to do all self-care activities and spend most of waking time out of bed but you are unable to do any work.

- A score of 3 means you are unable to do all self-care activities and any work and spend most of waking time in bed.
- A score of 4 means you are fully disabled.

KPS (Karnofsky Performance Status)

- A score of 0 to 49 means you are unable to care for yourself.
- A score of 50 to 79 means you are unable to work and some assistance is needed.
- A score of 80 to 100 means you are able to do your normal work and activities.

The main options for managing advanced cancer are supportive care (also called palliative care) and chemotherapy. The options are based on your ability to do activities (performance status). Supportive care is an option for all advanced cancers. If your health hasn't seriously limited your activities, systemic therapy is an option to slow down cancer growth. [See Guide 30](#). Systemic therapy and supportive care are addressed next.

### Systemic therapy

Doctors use the term "systemic" when talking about a cancer treatment for the whole body. Chemotherapy is the type of systemic treatment most often used for advanced esophageal cancer. If your health hasn't seriously limited your activities, chemotherapy is an option to slow down cancer growth.

Your doctor will choose a chemotherapy regimen based on your health and the side effects of treatment. Regimens that have two drugs have less severe side effects than three-drug regimens. If you are given 5-FU, leucovorin may be added to limit side effects of the chemotherapy. No matter what regimen you receive, you should be assessed for side effects on a regular basis. If your cancer doesn't respond

Guide 29. Adenocarcinoma that returns to esophagus area

Treatment you've had		Next treatment options			What's next?	
Surgery but not chemotherapy and radiation			Chemotherapy and radiation OR		CT scan	Start follow-up care. If cancer returns, begin supportive care.
			Surgery OR			
			Chemotherapy alone OR			
			Supportive care			
Chemotherapy and radiation but not surgery	Can have surgery		Surgery		CT scan	Start follow-up care. If cancer returns, begin supportive care.
	Can't have surgery		Supportive care			See <i>Advanced Cancer</i> section

Guide 30. Managing advanced adenocarcinoma

Performance status	Management options
Your KPS score is 60% or higher <b>OR</b> Your ECOG performance score is 0, 1, or 2	<ul style="list-style-type: none"> <li>• Chemotherapy to slow cancer growth</li> <li>• Supportive care</li> <li>• Clinical trials</li> </ul>
Your KPS score is 0% to 59% <b>OR</b> Your ECOG performance score is 3 or 4	<ul style="list-style-type: none"> <li>• Supportive care</li> <li>• Clinical trials</li> </ul>

to first-line regimens, your doctor will give you a different regimen. The regimen that is best for you depends on your prior treatment and performance status.

You may recall from earlier chapters that some esophageal adenocarcinomas have too much HER2 protein, which causes the cancer to grow and spread more quickly than it normally would. If HER2 testing wasn't already done, and if your doctor thinks the cancer has metastasized, HER2 testing may be done at this point. The goal is to see if targeted therapy might help you. Targeted therapy is a drug treatment for advanced esophageal adenocarcinomas. Trastuzumab is a targeted therapy. It should be given with first-line chemotherapy if the cancer cells have too much HER2, but not if you're also taking an anthracycline. Epirubicin is an anthracycline.

### Supportive care

Because the cancer can't be cured, the goal of supportive care is to make you more comfortable and to keep the cancer under control. Supportive care may also help you live longer, improve your eating, and help you feel better overall. When used for advanced cancers, supportive care is often called palliative care.

People without advanced cancer also receive supportive care. Some aspects of supportive care, such as symptom control, are useful for many people with any stage of esophageal cancer. Symptom control is addressed next.

Supportive care alone (without systemic therapy) is an option if you have an ECOG score of 3 or more or a KPS score of less than 60%. You may receive other types of care to improve your quality of life. Keep reading to learn about symptom control.

### Help with symptoms

Cancer or its treatment can cause unpleasant and sometimes harmful symptoms. One of the most

common symptoms of esophageal cancer is trouble with food passing through the esophagus. This is called dysphagia. Your doctor will assess what you can and can't swallow and what is causing the dysphagia.

Dysphagia is often caused by the tumor blocking the passage. However, sometimes it is caused by the tumor impairing the muscles of the esophagus or by scarring from radiation. Treating dysphagia depends on what's causing it. Treatment options for a blocked esophagus are described in the next section.

Bleeding is another common symptom, but not as common as dysphagia. Bleeding may be caused by the cancer or the cancer treatment. Endoscopic treatment that uses heat, cold, lasers, or injections may stop bleeding from the tumor surface. External radiation therapy may stop ongoing blood loss.

Other symptoms related to esophageal cancer include pain and nausea, with or without vomiting. These symptoms may be caused by the tumor blocking the passage of the esophagus. Treatment options for a blocked esophagus are described in the next section. Pain may be controlled with radiation therapy, chemotherapy, and pain medication. There are medicines and other methods that may help stop nausea and vomiting.

You may have other symptoms that aren't listed here. Talk to your treatment team about your symptoms. There may be ways to help you feel better.

### Blocked esophagus

The cancer may block food and liquids from passing through your esophagus. Treatment is based on how much of the esophagus is blocked.

A **complete blockage** doesn't allow any food or liquids to pass through your esophagus. Treatment options include endoscopic methods, radiation therapy, chemotherapy, and sometimes

surgery. Dilation and stents are endoscopic methods described below. External radiation is the common radiation method, but internal radiation (brachytherapy) may also be an option. You may also get a feeding tube to make sure you are getting enough nutrients.

A **severe blockage** allows only liquids to pass through your esophagus. One treatment option is to stretch open (dilate) your esophagus using a small balloon or tube guided down your throat to the right spot.

Another option for a severe blockage is a stent. A stent is a thin metal or plastic tube. It can be placed in your esophagus with endoscopic tools while you are sedated. The stent will expand in the passage and remain in your body to allow food to pass through. Stents are not used for patients who may undergo surgery to treat the cancer.

Other options for a severe blockage are the same as for a complete block. External and internal radiation therapy work well to unblock the esophagus. Symptom relief from radiation is slower but more long-lasting compared to endoscopic methods.

A **moderate blockage** allows liquids and semisolid foods to pass through your esophagus. Applesauce and cottage cheese are examples of semisolid foods. Treatment options are the same as for complete and severe blockage.

### Clinical trials

A clinical trial is a type of research study that involves people. NCCN believes that the best management for any cancer patient is in a clinical trial. Ask your treatment team if there is an open clinical trial that you can join. Clinical trials are discussed in more detail at the end of Part 4. You can use the websites in Part 7 to find clinical trials near you.

## Review

- Endoscopic therapies are preferred for most Tis and T1a adenocarcinomas. Surgery is a better option, though, if the Tis or T1a tumor has grown over a large area, or if your doctor doesn't think endoscopic treatment will work well.
- T1b tumors that haven't grown very far into the second layer of the esophagus wall are treated with endoscopic treatment or with surgery. Treatment of all other T1b tumors depends on whether there is cancer in nearby lymph nodes. If there isn't, surgery is recommended.
- T1b, T2, T3, and T4a adenocarcinomas that may have spread to nearby lymph nodes are often treated with chemotherapy and radiation (chemoradiation), followed by surgery to remove any remaining cancer. Other treatment options are also available.
- When there are no signs of cancer after treatment, testing to check for new cancer growth should be done. Medical history and physical exams are needed. You may also receive blood, imaging, and tests using an endoscope.
- Cancer that returns after local treatment near to where the esophagus is (or was) may be curable with surgery or chemoradiation.
- Cancer that is unable to be cured can be managed with supportive care. Treatment with chemotherapy and/or targeted therapy are also options for some people with advanced adenocarcinomas. Participation in clinical trials is another option and is especially encouraged by NCCN.

# 7

## Making treatment decisions

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Having cancer is very stressful. There is a lot to learn in what feels like a short time. This chapter can help you make decisions that are in line with your beliefs, wishes, and values.

## It's your choice

The role patients want in choosing their treatment differs. You may feel uneasy about making treatment decisions because you don't know much about cancer. Stress, pain, and medications can also limit your ability to make good decisions. Or, you may simply think that your judgment isn't any better than your doctors'.

Letting others decide which option is best may make you feel more at ease. But, whom do you want to make the decisions? You may rely on your doctors alone to make the right decisions. However, your doctors may not tell you which to choose if you have more than one good option. You can also have loved ones help. They can gather information, speak on your behalf, and share in decision-making with your doctors. Even if others decide which treatment you will receive, you still have to agree by signing a consent form.

On the other hand, you may want to take the lead or share in decision-making. Most patients do. In shared decision-making, you and your doctors share information, weigh the options, and agree on a treatment plan. Your doctors know the science behind your plan, but you know your concerns and goals. By working together, you are likely to get a higher quality of care and be more satisfied. You'll likely get the treatment you want, at the place you want, and by the doctors you want.

## Questions to ask your doctors

You may meet with experts from different fields of medicine. Try to talk with each expert. Prepare questions before your visit and ask questions if the person isn't clear. You can also record your talks and get copies of your medical records. It may be helpful to have your spouse, partner, or a friend with you at these visits. They can help to ask questions and remember what was said. Below are some suggested questions to ask.

## What's my diagnosis and prognosis?

It's important to know that there are different types of cancer. Cancer can greatly differ even when people have a tumor in the same organ. Based on your test results, your doctors can tell you which type of cancer you have. He or she can also give a prognosis. A prognosis is a prediction of the pattern and outcome of a disease. Knowing the prognosis may affect what you decide about treatment.

1. Is this cancer common?
2. What is the cancer stage? Does this stage mean the cancer has spread far?
3. Is this a fast- or slow-growing esophageal cancer?
4. What other tests results are important to know?
5. How often are these tests wrong?
6. Would you give me a copy of the pathology report and other test results?
7. How likely is it that I'll be cancer-free after treatment?

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## What are these other options based on?

1. Do your suggested options include clinical trials? If yes, please explain why.
2. How do my age, health, and other factors affect my options?
3. Which option is proven to work best?
4. Which options lack scientific proof?
5. What are the benefits of each option? Does any option offer a cure? Are my chances any better for one option than another? Less time-consuming? Less expensive?
6. What are the risks of each option? What are possible complications? What are the rare and common side effects? Short-lived and long-lasting side effects? Serious or mild side effects? Other risks?
7. What can be done to prevent or relieve the side effects of treatment?
8. What are my chances that the cancer will return?

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## Weighing your options

Deciding which option is best can be hard. Doctors from different fields of medicine may have different opinions on which option is best for you. This can be very confusing. Your spouse or partner may disagree with which option you want. This can be stressful. In some cases, one option hasn't been shown to work better than another, so science isn't helpful. Some ways to decide on treatment are discussed next.

### 2<sup>nd</sup> opinion

After finding out you have cancer, 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 2<sup>nd</sup> opinion, and it's a normal part of cancer care.

Getting a 2<sup>nd</sup> opinion doesn't mean you don't trust the first doctor. In fact, most doctors that are diagnosed with cancer will see more than one doctor before beginning treatment. What's more, some health plans require a second opinion. If your health plan doesn't cover the cost of a second opinion, you have the choice of paying for it yourself.

If the two opinions are the same, you may feel better about the treatment you accept to have. If the two opinions differ, think about getting a third opinion. Choosing your cancer treatment is a very important decision. It can affect your length and quality of life.

### Support groups

Besides talking to health experts, it may help to talk to patients who have walked in your shoes. Support groups often consist of people at different stages of treatment. Some may be in the process of deciding while others may be finished with treatment. At support groups, you can ask questions and hear about the experiences of other people with esophageal cancer.

### Compare benefits and downsides

Every option has benefits and downsides. Consider these when deciding which option is best for you. Talking to others can help identify benefits and downsides you haven't thought of. Scoring each factor from 0 to 10 can also help since some factors may be more important to you than others.

## Websites

### American Cancer Society

[www.cancer.org/cancer/esophaguscancer/index](http://www.cancer.org/cancer/esophaguscancer/index)

### Esophageal Cancer Awareness Association

[www.ecaware.org](http://www.ecaware.org)

### Esophageal Cancer Action Network, Inc.

[www.ecan.org](http://www.ecan.org)

### National Coalition for Cancer Survivorship

[www.canceradvocacy.org/toolbox](http://www.canceradvocacy.org/toolbox)

### National Cancer Institute

[www.cancer.gov/types/esophageal](http://www.cancer.gov/types/esophageal)

### NCCN

[www.nccn.org/patients](http://www.nccn.org/patients)

### U.S. National Library of Medicine Clinical Trials Database

[www.clinicaltrials.gov](http://www.clinicaltrials.gov)

## Review

- Shared decision-making is a process in which you and your doctors plan treatment together.
- Asking your doctors questions is vital to getting the information you need to make informed decisions.
- Getting a second opinion, attending support groups, and comparing benefits and downsides may help you decide which treatment is best for you.



I am a 3-year esophageal cancer stage 3 survivor and was so fortunate to have found ECAA during my journey. I am giving back now by volunteering in the organization and I am also on the Board of Directors. No one has to fight EC alone - ECAA is here for you too!

– Mary Jo

Esophageal cancer survivor

# Glossary

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80 Dictionary

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85 Acronyms

# Dictionary

## **abdomen**

The belly area between the chest and pelvis.

## **ablation**

A treatment that destroys very small tumors with heat, cold, lasers, or chemicals. Also called ablative therapy.

## **adenocarcinoma**

Cancer of cells that line organs and make fluids.

## **Barrett's esophagus**

The presence of stomach cells within the inner wall of the esophagus.

## **biopsy**

Removal of small amounts of tissue or fluid to be tested for disease.

## **bronchi**

The two airways extending from the windpipe into the lungs.

## **bronchoscope**

A device that is guided down the throat to work inside the airways.

## **bronchoscopy**

A procedure to work inside the airways with a device that is guided down the throat.

## **cancer grade**

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

## **cancer stage**

A rating of the outlook of a cancer based on its growth and spread.

## **carina**

The supportive tissue at the base of the windpipe.

## **chemotherapy**

Cancer drugs that stop the cell life cycle so cells don't increase in number.

## **clinical stage**

The rating of the extent of cancer before treatment is started.

## **clinical trial**

A type of research involving people that assesses health tests or treatments.

## **complete blood count (CBC)**

A lab test that includes the number of blood cells.

## **computed tomography (CT)**

A test that uses x-rays from many angles to make a picture of the insides of the body.

## **contrast**

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

## **cryoablation**

Treatment that kills cancer cells by freezing them. Also called cryosurgery.

## **deoxyribonucleic acid (DNA)**

A chain of chemicals inside cells that contains coded instructions for making and controlling cells.

## **diaphragm**

A sheet of muscles below the ribs that helps a person to breathe.

## **digestive system**

A set of organs that changes food into small parts for the body to use as energy.

## **dysphagia**

Difficult or painful swallowing.

## **Eastern Cooperative Oncology Group (ECOG) Performance Scale**

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

## **electrode**

A small device that transmits electricity.

## **endoscope**

A device that is passed through a natural opening to do work inside the body.

## **endoscopic mucosal resection (EMR)**

A procedure to remove an early tumor with a cap or snare on a device that is passed through a natural opening.

## **endoscopic resection**

A procedure that removes an early tumor with a device passed through a natural opening.

**endoscopic submucosal dissection (ESD)**

A procedure that removes an early tumor with a special knife on a device that is passed through a natural opening.

**endoscopic ultrasound (EUS)**

A procedure that takes detailed pictures of the digestive tract and nearby tissue with a device passed through a natural opening.

**epithelium**

Tissue that lines the inner wall of the digestive tract.

**esophagectomy**

An operation that removes all or part of the esophagus.

**esophagogastrectomy**

An operation that removes the esophagus and some of the stomach.

**esophagus**

The tube-shaped organ between the throat and stomach.

**external beam radiation therapy (EBRT)**

A cancer treatment with radiation received from a machine outside the body.

**fine-needle aspiration (FNA)**

A procedure that removes tissue samples with a very thin needle.

**follow-up care**

Health care that starts once treatment has ended and there are no signs of cancer.

**gastroenterologist**

A doctor who's an expert in digestive diseases.

**gastroesophageal reflux disease (GERD)**

A health condition in which stomach contents often flow back into the esophagus.

**esophagogastric junction (EGJ)**

The area where the esophagus and stomach join.

**general anesthesia**

A drug-induced, sleep-like state for pain relief.

**gene**

Coded instructions in cells for making new cells and controlling how cells behave.

**genetic counselor**

An expert in diseases that are passed down in families.

**high-grade dysplasia (HGD)**

Abnormal cells that are likely to become cancer cells.

**hives**

A skin rash caused by your body attacking a foreign substance that is not harmful.

**human epidermal growth factor receptor 2 (HER2)**

A protein on the edge of a cell that send signals to the cell to grow.

**inferior pulmonary vein**

A vessel that returns blood to the heart from the lungs.

**intensity-modulated radiation therapy (IMRT)**

Treatment with radiation that uses small beams of different strengths.

**integrative medicine doctor**

An expert in mind-body treatments.

**intestine**

The organ that food passes through after leaving the stomach.

**jejunostomy tube (J-tube)**

A feeding tube that is inserted through a cut into the gut.

**Karnofsky Performance Status (KPS)**

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

**liquid nitrogen**

The chemical, nitrogen, that has been cooled to a non-solid state.

**local anesthesia**

A drug-induced loss of feeling in a small area of the body.

**lymph**

A clear fluid containing white blood cells.

**lymph node**

A small, bean-shaped disease-fighting structure.

**medical history**

A report of all your health events and medications.

**medical oncologist**

A doctor who's an expert in cancer drugs.

**metastasis**

The spread of cancer from the first tumor to a new site.

**minimally invasive esophagectomy**

An operation that removes the esophagus with tools inserted through small cuts into the body.

**nasogastric tube**

A feeding tube that is inserted down the nose and into the stomach.

**nodule**

A small mass of tissue.

**nutritionist**

A health care worker who completed education in food and diet.

**observation**

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

**oncology surgeon**

A doctor who's an expert in operations that remove cancer.

**pathologic stage**

A rating of the extent of cancer based on tests given after treatment.

**pathologist**

A doctor who's an expert in testing cells and tissue 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.

**photodynamic ablation**

A treatment with a laser that turns on a drug inside the tumor.

**physical exam**

A study of the body by a health expert for signs of disease.

**positron emission tomography (PET)**

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

**positron emission tomography/computed tomography (PET/CT)**

A test that uses two picture-making methods to show the shape and function of tissue.

**primary tumor**

The first mass of cancer cells.

**radiation oncologist**

A doctor who's an expert in treating cancer with radiation.

**radiation therapy**

A treatment that uses high-energy rays.

**radiofrequency ablation**

Treatment that destroys very small tumors with heat.

**radiologist**

A doctor who's an expert in reading imaging tests.

**radiotracer**

A substance that releases energy, which is seen in tissue with a special camera.

**recurrence**

The return of cancer after a cancer-free period.

**sedative**

A drug that helps a person to relax or go to sleep.

**side effect**

An unhealthy or unpleasant physical or emotional response to treatment.

**simulation**

The steps needed to prepare for treatment with radiation.

**small intestine**

A digestive organ that absorbs nutrients from eaten food.

**squamous cell carcinoma**

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

**standard open esophagectomy**

An operation to remove the esophagus through large cuts made into the body.

**supportive care**

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

**surface receptor**

A protein within the cell membrane to which substances can attach.

**surgical margin**

The normal-looking tissue around a tumor that was removed during an operation.

**targeted therapy**

A drug treatment that impedes the growth process specific to cancer cells.

**thoracoscope**

A device that is passed through a small cut in the skin to do work inside the chest.

**three-dimensional conformal radiation therapy (3D-CRT)**

A treatment with radiation that uses beams matched to the shape of the tumor.

**trachea**

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

**upper gastrointestinal (GI) endoscopy**

A procedure to do work in the first parts of the digestive tract with a device guided down the throat. Also called an esophagastroduodenoscopy (EGD).

**vascular endothelial growth factor (VEGF)**

A molecule that triggers the growth of blood vessels.

# Acronyms

**3D-CRT**

three-dimensional conformal radiation therapy

**AJCC**

American Joint Committee on Cancer

**CBC**

complete blood count

**CT**

computed tomography

**DNA**

deoxyribonucleic acid

**EBRT**

external beam radiation therapy

**ECOG**

Eastern Cooperative Oncology Group

**EGJ**

esophagogastric junction

**EMR**

endoscopic mucosal resection

**ESD**

endoscopic submucosal dissection

**EUS**

endoscopic ultrasound

**FDA**

Food and Drug Administration

**FNA**

fine-needle aspiration

**GERD**

gastroesophageal reflux disease

**GI**

gastrointestinal

**HER2**

human epidermal growth factor receptor 2

**HGD**

high-grade dysplasia

**IMRT**

intensity-modulated radiation therapy

**J-tube**

jejunostomy tube

**KPS**

Karnofsky Performance Status

**MMR**

mismatch repair

**MSI**

microsatellite instability

**PD-1**

programmed cell death-1

**PD-L1**

programmed death-ligand 1

**PDT**

photodynamic therapy

**PEG**

percutaneous endoscopic gastrostomy

**PET**

positron emission tomography

**PET/CT**

positron emission tomography/computed tomography

**RFA**

radiofrequency ablation

**TNM**

tumor, node, metastasis

**VEGF**

vascular endothelial growth factor definition

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For disclosures, visit [www.nccn.org/about/disclosure.aspx](http://www.nccn.org/about/disclosure.aspx).

# NCCN Member Institutions

Fred & Pamela Buffett Cancer Center  
Omaha, Nebraska  
800.999.5465  
[nebraskamed.com/cancer](http://nebraskamed.com/cancer)

Case Comprehensive Cancer Center/  
University Hospitals Seidman Cancer  
Center and Cleveland Clinic Taussig  
Cancer Institute  
Cleveland, Ohio  
800.641.2422 • UH Seidman Cancer Center  
[uhhospitals.org/seidman](http://uhhospitals.org/seidman)  
866.223.8100 • CC Taussig Cancer Institute  
[my.clevelandclinic.org/services/cancer](http://my.clevelandclinic.org/services/cancer)  
216.844.8797 • Case CCC  
[case.edu/cancer](http://case.edu/cancer)

City of Hope Comprehensive  
Cancer Center  
Los Angeles, California  
800.826.4673  
[cityofhope.org](http://cityofhope.org)

Dana-Farber/Brigham and  
Women's Cancer Center  
Massachusetts General Hospital  
Cancer Center  
Boston, Massachusetts  
877.332.4294  
[dfbwcc.org](http://dfbwcc.org)  
[massgeneral.org/cancer](http://massgeneral.org/cancer)

Duke Cancer Institute  
Durham, North Carolina  
888.275.3853  
[dukecancerinstitute.org](http://dukecancerinstitute.org)

Fox Chase Cancer Center  
Philadelphia, Pennsylvania  
888.369.2427  
[foxchase.org](http://foxchase.org)

Huntsman Cancer Institute  
at the University of Utah  
Salt Lake City, Utah  
877.585.0303  
[huntsmancancer.org](http://huntsmancancer.org)

Fred Hutchinson Cancer  
Research Center/Seattle  
Cancer Care Alliance  
Seattle, Washington  
206.288.7222 • [seattlecca.org](http://seattlecca.org)  
206.667.5000 • [fredhutch.org](http://fredhutch.org)

The Sidney Kimmel Comprehensive  
Cancer Center at Johns Hopkins  
Baltimore, Maryland  
410.955.8964  
[hopkinskimmelcancercenter.org](http://hopkinskimmelcancercenter.org)

Robert H. Lurie Comprehensive Cancer  
Center of Northwestern University  
Chicago, Illinois  
866.587.4322  
[cancer.northwestern.edu](http://cancer.northwestern.edu)

Mayo Clinic Cancer Center  
Phoenix/Scottsdale, Arizona  
Jacksonville, Florida  
Rochester, Minnesota  
800.446.2279 • Arizona  
904.953.0853 • Florida  
507.538.3270 • Minnesota  
[www.mayoclinic.org/cancercenter](http://www.mayoclinic.org/cancercenter)

Memorial Sloan Kettering  
Cancer Center  
New York, New York  
800.525.2225  
[mskcc.org](http://mskcc.org)

Moffitt Cancer Center  
Tampa, Florida  
800.456.3434  
[moffitt.org](http://moffitt.org)

The Ohio State University  
Comprehensive Cancer Center -  
James Cancer Hospital and  
Solove Research Institute  
Columbus, Ohio  
800.293.5066  
[cancer.osu.edu](http://cancer.osu.edu)

Roswell Park Comprehensive  
Cancer Center  
Buffalo, New York  
877.275.7724  
[roswellpark.org](http://roswellpark.org)

Siteman Cancer Center at Barnes-  
Jewish Hospital and Washington  
University School of Medicine  
St. Louis, Missouri  
800.600.3606  
[siteman.wustl.edu](http://siteman.wustl.edu)

St. Jude Children's Research Hospital  
The University of Tennessee  
Health Science Center  
Memphis, Tennessee  
888.226.4343 • [stjude.org](http://stjude.org)  
901.683.0055 • [westclinic.com](http://westclinic.com)

Stanford Cancer Institute  
Stanford, California  
877.668.7535  
[cancer.stanford.edu](http://cancer.stanford.edu)

University of Alabama at Birmingham  
Comprehensive Cancer Center  
Birmingham, Alabama  
800.822.0933  
[www3.ccc.uab.edu](http://www3.ccc.uab.edu)

UC San Diego Moores Cancer Center  
La Jolla, California  
858.657.7000  
[cancer.ucsd.edu](http://cancer.ucsd.edu)

UCSF Helen Diller Family  
Comprehensive Cancer Center  
San Francisco, California  
800.689.8273  
[cancer.ucsf.edu](http://cancer.ucsf.edu)

University of Colorado Cancer Center  
Aurora, Colorado  
720.848.0300  
[coloradocancercenter.org](http://coloradocancercenter.org)

University of Michigan  
Rogel Cancer Center  
Ann Arbor, Michigan  
800.865.1125  
[mcancer.org](http://mcancer.org)

The University of Texas  
MD Anderson Cancer Center  
Houston, Texas  
800.392.1611  
[mdanderson.org](http://mdanderson.org)

University of Wisconsin  
Carbone Cancer Center  
Madison, Wisconsin  
608.265.1700  
[uwhealth.org/cancer](http://uwhealth.org/cancer)

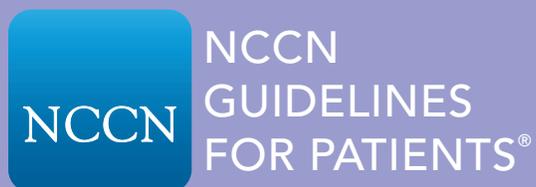
Vanderbilt-Ingram Cancer Center  
Nashville, Tennessee  
800.811.8480  
[vicc.org](http://vicc.org)

Yale Cancer Center/  
Smilow Cancer Hospital  
New Haven, Connecticut  
855.4.SMILOW  
[yalecancercenter.org](http://yalecancercenter.org)

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# Esophageal Cancer

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