National Comprehensive Cancer Network®

2026

NCCN Guidelines for Patients®

Cancer care recommendations from leading experts at the National Comprehensive Cancer Network® (NCCN®)

Adrenal Tumors



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NCCN Guidelines for Patients®

The essential guide for people facing cancer.

Based on care recommendations from leading cancer experts.

Explains high-quality cancer care provided at state-of-the-art cancer centers.

Reviewed and revised every year.

Did you know that top cancer centers across the United States work together to improve cancer care? This alliance of leading cancer centers is called the National Comprehensive Cancer Network® (NCCN®).

Because cancer care is always evolving, NCCN develops and frequently updates evidence-based cancer care recommendations used by health care providers worldwide. These recommendations are known as the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®).

The NCCN Guidelines for Patients plainly explain these expert recommendations, so you can talk with your care team about the best care for you.

These NCCN Guidelines for Patients are based on the NCCN Guidelines® for Neuroendocrine and Adrenal Tumors, Version 1.2026 — April 21, 2026.

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1
About adrenal tumors

5 What is an adrenal tumor?

6 What are the types of adrenal tumors?

6 How are adrenal tumors diagnosed and treated?

7 How can I get the best care?

The adrenal glands sit on top of the kidneys and produce hormones used throughout the body. Adrenal tumors can form in one or both adrenal glands.

Most adrenal tumors are benign (not cancer) and don't cause symptoms. This chapter gives an overview of adrenal tumors and some of the conditions related to them.

What is an adrenal tumor?

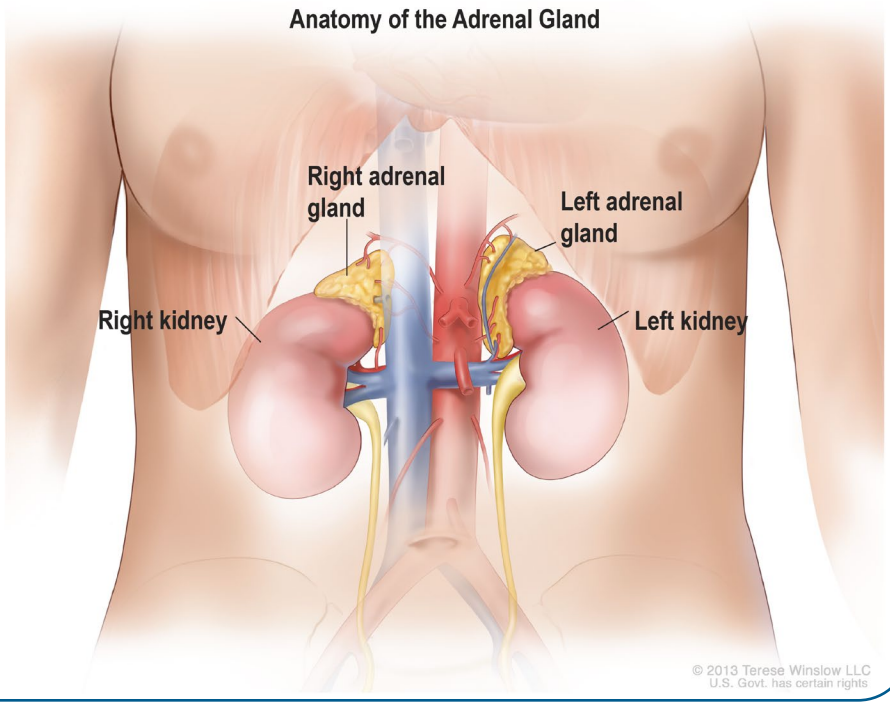
A tumor is a group of abnormal cells. Adrenal tumors start in the adrenal glands.

Most adrenal tumors are benign. That means they are not cancer. Malignant tumors are cancerous.

The adrenal glands are important to the body's endocrine system. The endocrine system is made up of tissues and organs that produce hormones. Hormones are substances that help control how the body works.

Some hormones control how your body turns what you eat and drink into energy. Others manage blood pressure. Others control body features, such as hair growth and body shape. Others control the body's response to stress.

There are 2 adrenal glands, one on top of each of our kidneys. The kidneys are in the lower back, under the rib cage.



Hormones and adrenal tumors

Some, but not all, adrenal tumors produce hormones. This can cause symptoms. Symptoms depend on the type of hormone released by the tumor. These types of tumors are usually benign, but some are cancerous, or malignant, and can spread to other parts of the body.

Conditions caused by hormones made by adrenal tumors

In these guidelines, you'll read about 2 of the conditions related to adrenal tumors:

- **Primary aldosteronism** is when 1 or both adrenal glands make too much of the hormone aldosterone. Read Chapter 4 to learn about this condition.
- **Hypercortisolism** is when 1 or both adrenal glands make too much of the hormone cortisol. Read Chapter 5 to learn about this condition.

What are the types of adrenal tumors?

Adrenal gland-related tumors discussed in these guidelines are described here.

Adrenocortical carcinoma

Adrenocortical carcinoma (ACC) is cancer that forms in the outer layer of the adrenal gland. Hypercortisolism may be a sign of ACC.

Paraganglioma

Paraganglioma (PGL) is a rare type of tumor that forms near blood vessels and nerves outside of the adrenal glands.

Adrenal conditions and tumors discussed in these guidelines:

- ✓ Primary aldosteronism
- ✓ Hypercortisolism
- ✓ Adrenocortical carcinoma
- ✓ Paraganglioma
- ✓ Pheochromocytoma

Pheochromocytoma

Pheochromocytoma (PCC) is a rare tumor that develops in the center of the adrenal gland. The tumor grows from cells that produce hormones in the inner layer of the gland.

PGL and PCC are treated in the same way.

How are adrenal tumors diagnosed and treated?

Most adrenal tumors are found through imaging tests done for an unrelated issue.

Many of these tumors can just be observed, but some require surgery to remove the tumor along with the whole adrenal gland.

This guide will discuss in greater detail how adrenal tumors are diagnosed and treated. It will provide an overview of testing and treatment and what to expect during testing and treatment.

How can I get the best care?

Advocate for yourself. You have an important role to play in your care. Many people feel

more satisfied when they actively take part in planning their cancer care.

The NCCN Guidelines for Patients will help you play a larger role in your care. Discuss the recommendations in this guide with your care team. Ask questions about your options and share your goals and concerns.

Don’t know what to ask? You’re not alone. That’s why we include suggested questions to ask at the end of chapter.

Keep reading to find the best care for you.

How this guide can help you

Making decisions about cancer care is stressful. There’s a lot to learn, and you don’t know what the future holds.

Use this guide to get the information and support you need.

Patients, doctors, and other health care professionals trust the NCCN Guidelines for Patients. This guide uses clear, everyday language to explain current cancer care recommendations made by respected experts in the field. Their recommendations are based on the latest research and practices at leading cancer centers.

Your health is unique to you, so your cancer care should be, too. As you read this guide, you’ll learn which treatments are likely to provide the best results for you. And you’ll be better prepared to talk with your care team.

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Testing for adrenal tumors

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10 Blood and urine tests

11 Imaging tests

12 Biopsy

12 Genetic testing

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13 Questions to ask

If an adrenal tumor is found on an imaging scan, testing will be based on if you have a history of cancer or if there is a risk that you have cancer that has spread. Either way, your health care team will want to gather as much information as they can.

Tests are used to diagnose conditions and find out more about adrenal tumors. They're also used to plan symptom management and treatment and to see how your tumor is responding to treatment. You can have tests at different stages—before, during, or after treatment.

Tests you may have are highlighted in **Guide 1**. You'll learn more about these tests in this chapter.

Guide 1 **Tests you may have if an adrenal tumor is found**
Medical and family history, physical exam
Blood and urine tests
Imaging tests
Biopsy
Genetic testing

General health tests

Some general health tests are described next.

Medical history

A medical or health history is a record of all health issues and treatments you've had in your life. It includes past surgeries and current health conditions.

Family history

Some tumors, cancers, and other diseases can run in families. Your doctor will ask about the health history of family members who are blood relatives.

Physical exam

Your health care team may:

- Check your temperature, blood pressure, pulse, and breathing rate
- Check your height and weight
- Listen to your lungs and heart
- Look in your eyes, ears, nose, and throat
- Feel and apply pressure to parts of your body to see if organs are of normal size, are soft or hard, or cause pain when touched
- Feel for enlarged lymph nodes in your neck, underarm, and groin

Blood and urine tests

Blood and urine tests check for signs of disease and how well organs are working.

They're also used to look for hormones and other chemicals produced by your tumor.

- Blood tests require a sample of blood, which is removed through a needle placed into a vein in your arm.
- For a urine test, you'll be asked to fill a small cup with urine (pee).

Blood and urine samples are sent to a lab for testing. Some blood and urine tests you may have are described next.

Biochemical tests

Biochemical tests measure hormones, proteins, and sugar (glucose) in blood and urine.

Complete blood count

A complete blood count measures the levels of red blood cells, white blood cells, and platelets in your blood. It's a key test that gives a picture of your overall health.

Comprehensive metabolic panel

A comprehensive metabolic panel measures substances in your blood. It provides important information about how well your kidneys and liver are working, among other things.

Adrenal vein sampling

Adrenal vein sampling measures how much hormone is produced by each adrenal gland. A needle and catheter (plastic tube) are inserted into the adrenal veins, and blood is taken directly from each adrenal gland.

This test is used to diagnose primary aldosteronism (Conn syndrome), a condition that causes the adrenal glands to produce too much aldosterone.

It's important to tell your health care team if there is a history of cancer in your family.



Imaging tests

You'll likely have imaging tests. Imaging tests take pictures of the inside of your body. They show the primary tumor, or where the cancer started, and look for cancer or tumors in other parts of the body.

In this section, you'll learn about imaging tests you may have.

CT scan

A computed tomography (CT or CAT) scan uses x-rays and computer technology to take pictures of the inside of the body. It takes many x-rays of the same body part from different angles.

Other less common types of CT scans may be used in very specific cases.

MRI scan

A magnetic resonance imaging (MRI) scan uses radio waves and powerful magnets to take pictures of the inside of the body.

Some things to know about CT and MRI

Contrast material

Contrast is a substance that's sometimes used to improve the quality of imaging tests, such as CT and MRI scans. Contrast might be taken by mouth (oral) or given through a vein (intravenously, or IV). The types of contrast vary and are different for CT and MRI. After the tests:

- Oral contrast is passed with your next bowel movement (poop).
- IV contrast leaves the body in the urine (pee) immediately after the test.

CT machine

A CT machine is large and has a tunnel in the middle. During the test, you will lie on the table, which moves slowly through the tunnel.



CT or MRI in phases

When CT or MRI is done in phases, there are 2 scans—1 with contrast and 1 without contrast. The scan with contrast is done first. IV contrast is given, and multiple sets of pictures are taken as the contrast moves through the area of the body near the tumor. This allows your care team to see whether the tumor involves any veins, arteries, or organs.

PET scan

PET (positron emission tomography) scans use radioactive tracers (or radiotracers) injected into a vein before imaging. This helps to see where cancer cells are in the body.

A radiotracer called fluorodeoxyglucose (FDG) is often used with PET scans. FDG-PET scans can help determine if tumors are becoming more aggressive or when there is cancer in the bones. An FDG-PET scan might be combined with certain PET scans that use other kinds of radiotracers to provide more details about the tumor.

Biopsy

When a sample of tissue or group of cells is removed for testing, it's called a biopsy.

Biopsies are rarely done to diagnose adrenal tumors. Instead, they may be done to learn more about cancer in the body. You may have a:

- Needle biopsy, guided by imaging (such as ultrasound, CT, or MRI)
- ctDNA biopsy, which tests small pieces of DNA from tumor cells that are released into the blood

Immunohistochemistry

Immunohistochemistry is a special staining process used to study cells from a biopsy or tumor sample.

Genetic testing

In some people, depending on your family history or tumor features, your health care team might refer you for genetic testing.

Genetic testing is done using blood or saliva (spitting into a cup or taking a cheek swab).

The goal is to look for gene mutations inherited from your biological (birth) parents. Some genetic mutations can put you at risk of more than one type of tumor or cancer. These mutations can be passed on to your children. Other family members might also carry these mutations.

Hereditary cancer risk

Some genetic syndromes put people at risk for developing certain tumors and cancers. A syndrome is a group of signs or symptoms that occur together and suggest the presence or risk of a disease. A hereditary syndrome may also be found in blood (biological) relatives.

A genetic cancer risk assessment will detect if you have a genetic risk factor for tumors or cancer and if you may benefit from genetic testing, additional screening, or preventive interventions.

Depending on the genetic risk assessment, you might have genetic testing and genetic counseling to see if you have a hereditary syndrome that puts you at risk for developing adrenal tumors.

Key points

- Tests are used to find tumors or conditions, plan treatment, and check how your tumor or cancer is responding to treatment.
- Blood and urine tests in addition to imaging studies are used to determine what treatments are best for your type of tumor or condition.
- Blood and urine tests look for signs of disease and how well your organs are working. These tests also measure hormone, blood sugar (glucose), and protein levels.
- Imaging tests take pictures of the inside of your body and are used to learn more about the type, size, and location of tumors.
- A biopsy is the removal of a sample of tissue or group of cells for testing. Biopsies are rarely done to diagnose adrenal tumors.
- Genetic testing looks for gene mutations inherited from your biological (birth) parents.

Questions to ask

- What tests will I have?
- What tests will be done on the adrenal condition or tumor?
- Who will explain the test results to me?
- What other information do you need before I start treatment?
- Do you recommend I have genetic testing and counseling for inherited cancer risk? Why or why not?



Let us know what you think!

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

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Types of treatment

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There's more than one treatment for adrenal tumors and conditions. This chapter describes treatment options and what to expect. Together, you and your care team will choose a treatment plan that's best for you.

Results from blood, urine, tumor, and imaging tests will guide your treatment plan. Many factors play a role in how a tumor or cancer responds to treatment.

Most adrenal tumors are benign (not cancer) and don't need treatment. They are observed with imaging studies and blood and urine tests. However, some adrenal tumors and conditions do require treatment.

Treatment for adrenal tumors is usually a combination of therapies starting with surgery and might include systemic therapy or radiation therapy.

Your care team

Treating cancer takes a team approach. Treatment decisions should involve a multidisciplinary team of health care and psychosocial care professionals from different professional backgrounds who have knowledge and experience in your type of cancer. This team is united in planning and implementing your treatment. Ask who will coordinate your care.



Your care team

Depending on the type and location of your adrenal tumor, your care team might include the following specialists.

- An **oncologist** – A doctor who diagnoses and treats cancer. Types of oncologists include medical, radiation, and surgical oncologists.
- An **endocrinologist** – A doctor who diagnoses and treats disorders of the endocrine glands.
- A **pathologist** – A doctor who analyzes the cells, tissues, and organs removed during a biopsy or surgery.
- A **radiologist** – A doctor who interprets the results of CT scans, MRIs, and other imaging tests and performs image-guided procedures as needed.
- A **surgeon** – A doctor who performs surgical procedures.

Adrenalectomy

An adrenalectomy is surgery to remove the adrenal gland. It's also called a resection. The tumor and any areas where the cancer has spread, including lymph nodes or nearby structures, may be removed. There are 2 types of adrenalectomy:

- **Minimally invasive surgery** uses several small incisions. Small tools are inserted through each incision to perform the surgery.
- **Open surgery** removes tissue through one large surgical cut. The cut lets your surgeon directly view and access the tumor in your adrenal gland. Open surgery is often done when the tumor might be in lymph nodes or other nearby organs, or if cancer is suspected.

Systemic therapy

Systemic therapy is therapy that works throughout the body. Some types of systemic therapy include chemotherapy, targeted therapy, and immunotherapy.

All systemic treatments listed in this guide are recommended and appropriate. When helpful, NCCN experts also assign a level of preference to their recommendations for systemic therapies:

- **Preferred therapies** have the most evidence they may work better and be safer than other therapies.
- **Other recommended therapies** can provide effective results but may have less evidence, more side effects, or may not work quite as well as preferred therapies.
- **Therapies used in certain cases** work best for individuals with specific cancer features or health circumstances.

The goal of surgery is to remove all of the cancer

After surgery, you may receive treatment, such as systemic or radiation therapy, to kill any remaining cancer cells.



Systemic therapy recommendations depend on the tumor type.

Chemotherapy

Chemotherapy kills fast-dividing cells throughout the body, including cancer cells and some normal cells. Some things to know about chemotherapy include:

- A single agent regimen is when only 1 chemotherapy medicine is used.
- A combination or multi-agent regimen is when 2 or more chemotherapy medicines is used.
- Some chemotherapy medicines are given through a vein (intravenously, or IV) or injected under the skin with a needle. Others may be given as a pill that is swallowed.
- Most chemotherapy is given in cycles of treatment days followed by rest days. This allows the body to recover before the next cycle.

Targeted therapy

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

Therapy with radiopharmaceuticals

A radiopharmaceutical contains a radioactive substance that gives off radiation. These may be used for certain types of rare tumors. (See Chapter 7.)

Immunotherapy

Immunotherapy helps your immune system fight cancer cells. It can be given alone or with other types of treatment. Immunotherapy that might be used to treat adrenal tumors includes pembrolizumab, nivolumab, and ipilimumab.

Immunotherapy can cause side effects. You can learn about these side effects and how they're managed in *NCCN Guidelines for Patients: Immunotherapy side effects: Immune checkpoint inhibitors*.

Radiation therapy

Radiation therapy uses high-energy radiation from x-rays (photons), protons, and other sources to kill cancer cells, shrink tumors, or treat liver metastases. Radiation is typically delivered from outside of the body by a computerized device, which can shape the treatment to closely fit the location and size of the tumor.

Other therapies

Tumors found in the liver or lungs may be treated with surgery or one or both of the following therapies:

- **Ablation** uses extreme cold, extreme heat, radio waves, microwaves, or chemicals to destroy cancer cells.
- **Arterially directed therapy** or embolization treats tumors by injecting particles, chemotherapy, or radioactive beads directly into the blood vessels that supply the tumor(s).

Clinical trials

You may also be able to obtain treatment through a clinical trial. A clinical trial is a type of medical research study. After being developed and tested in a lab, potential new ways of treating cancer need to be studied in people.

If found to be safe and effective in a clinical trial, a drug, device, or treatment approach may be approved by the U.S. FDA.

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

Phases

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

- **Phase 1** trials study the safety and side effects of an investigational drug or treatment approach.
- **Phase 2** trials study how well the drug or approach works against a specific type of cancer.
- **Phase 3** trials test the drug or approach against a standard treatment. If the results are good, it may be approved by the FDA.
- **Phase 4** trials study the safety and benefit of an FDA-approved treatment.

Who can enroll?

It depends on the clinical trial's rules, called eligibility criteria. The rules may be about age, cancer type and stage, treatment history, or general health. They ensure that participants are alike in specific ways and that the trial is as safe as possible for the participants.

Informed consent

Clinical trials are managed by a research team. This group of experts will review the study with you in detail, including its purpose and the risks and benefits of joining. All of this information is also provided in an informed consent form. Read the form carefully and ask questions before signing it. Take time to



Finding a clinical trial

In the United States

NCCN Cancer Centers
NCCN.org/cancercenters

The National Cancer Institute (NCI)
cancer.gov/about-cancer/treatment/clinical-trials/search

Worldwide

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

Need help finding a clinical trial?

NCI's Cancer Information Service (CIS)
1.800.4.CANCER (1.800.422.6237)
cancer.gov/contact

discuss it with people you trust. Keep in mind that you can leave and seek treatment outside of the clinical trial at any time.

Will I get a placebo?

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

Are clinical trials free?

There's no fee to enroll in a clinical trial. The study sponsor pays for research-related costs, including the study drug. But you may need to pay for other services, like transportation or childcare, due to extra appointments. During the trial, you will continue to receive standard cancer care. This care is often covered by insurance.

Supportive care

Supportive care is an important part of cancer care. The goal is to improve your quality of life during and after cancer treatment. Supportive care is for everyone with cancer and their families, not just for those at the end of life.

Supportive care includes a wide range of services. Supportive care prevents or manages the symptoms of cancer and cancer-related fatigue. It also addresses the mental, social, emotional, and spiritual concerns faced by people with cancer.

Supportive care provides help with additional needs, such as:

- Making treatment decisions
- Coordinating your care
- Paying for care
- Planning for advanced care and end of life

"Connecting with other people with adrenal tumors is vital to my well-being. This journey can feel lonely at times, and it helps so much to share thoughts and experiences with people who also struggle sometimes and understand."



Read more about the types of support you may receive in *NCCN Guidelines for Patients: Palliative Care*, available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.

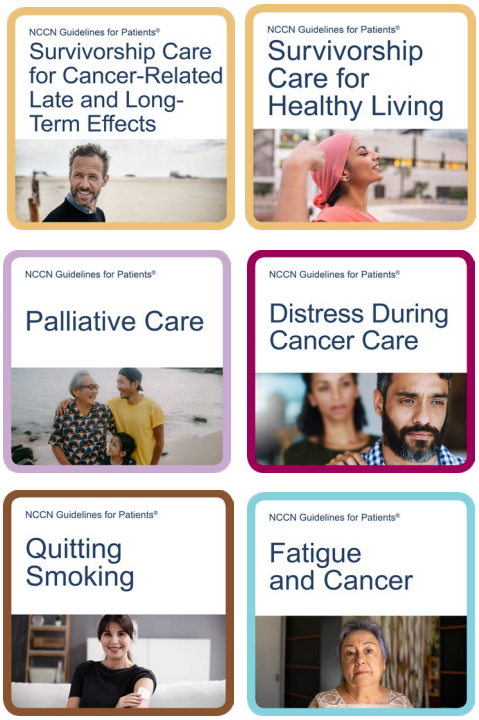
Survivorship

A person is a cancer survivor from the time of diagnosis until the end of life. After treatment, your health will be monitored for side effects and the return of cancer. This is part of your survivorship care plan. It's important to keep any follow-up care and imaging test appointments. Seek routine medical care, including regular doctor visits for preventive care and cancer screening.

A personalized survivorship care plan will contain a summary of possible long-term effects of treatment, called late effects, and a list of follow-up tests. Find out how your primary care provider will coordinate with specialists for your follow-up care.

Supportive care resources

More information on supportive care is available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



Key points

- Surgery is the main or primary treatment for adrenal tumors with limited or no spread.
- Systemic therapy is drug treatment that works throughout the body.
- Chemotherapy, targeted therapy, and immunotherapy are kinds of systemic therapy.
- Radiation therapy uses high-energy radiation from x-rays (photons), protons, and other sources to kill cancer cells.
- A clinical trial is a type of research that studies a treatment to see how safe it is and how well it works.
- Supportive care is health care that relieves symptoms and improves quality of life.
- Survivorship begins at diagnosis and continues through a person's life.

Questions to ask

- What are my treatment options?
- What treatment(s) do you recommend and why?
- What are the treatment side effects?
- Does the order of treatments matter?
- How long will treatment take?



Take our survey and help make the NCCN Guidelines for Patients better for everyone!

NCCN.org/patients/comments

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Primary aldosteronism

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24 Questions to ask

Primary aldosteronism occurs when the adrenal glands produce too much of the hormone aldosterone. Without treatment, this condition increases your risk of cardiovascular diseases, including heart attack and stroke.

This chapter provides information on the testing and treatment of primary aldosteronism, also known as hyperaldosteronism or Conn syndrome.

Adrenal glands and aldosterone

The adrenal glands make the hormone aldosterone. This hormone helps balance potassium and sodium in the body. Primary aldosteronism is a non-cancerous condition that occurs when one or both adrenal glands make too much aldosterone.

Too much aldosterone causes the body to hold sodium and lose potassium (hypokalemia), leading to high blood pressure (hypertension).

Without treatment, this condition increases your risk of cardiovascular diseases, including heart attack and stroke. Also, low potassium levels can cause heart rhythm changes.

Treatment for primary aldosteronism

Treatment depends on if you can and want to have surgery to remove the adrenal gland. This is called an adrenalectomy. You and your health care team will decide if surgery is an option for you. Next steps are described in **Guide 2**.

Guide 2
Treatment for primary aldosteronism

If you want and can have surgery	If 1 gland is producing too much aldosterone	→	You'll have an adrenalectomy
	If both glands are producing too much aldosterone	→	Your symptoms will be managed with medicine
If you do not want surgery or can't have surgery	Your symptoms will be managed with medicine		

If you do not have surgery

You'll likely be treated with medicine, such as spironolactone (Aldactone) or eplerenone (Inspra), to lower your blood pressure and increase potassium levels.

If you can and want to have surgery

You'll likely have adrenal vein sampling to measure hormone levels of both adrenal glands. To learn more about adrenal vein sampling, revisit Chapter 2.

If the results of adrenal vein sampling show too much aldosterone is being produced in:

- One gland, you'll have an adrenalectomy. Minimally invasive surgery is preferred.
- Both glands, you'll likely be treated with spironolactone or eplerenone to lower your blood pressure and increase potassium levels.

Key points

- Primary aldosteronism occurs when one or both adrenal glands make too much aldosterone, the hormone that helps to balance potassium and sodium in the body.
- The main symptom of primary aldosteronism is high blood pressure (hypertension) and low potassium levels (hypokalemia).
- Without treatment, primary aldosteronism increases your risk of cardiovascular diseases, including heart attack and stroke.
- Adrenal vein sampling is a test used to determine if too much aldosterone is being produced by one or both adrenal glands.
- Surgery to remove an adrenal gland, called an adrenalectomy, may be part of your treatment plan.
- Symptoms may be managed with spironolactone or eplerenone.

Questions to ask

- What tests will I have and what can I expect?
- Do you recommend treatment with medicine or surgery? Why?
- If my condition is being treated with medicine, what kind of specialist will monitor my care?
- Will I take this medicine for the rest of my life?
- What are the options if the medicine doesn't lower my risk of cardiovascular issues?

5
Hypercortisolism

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In this chapter, you'll learn about hypercortisolism, a condition that can happen when an adrenal tumor releases too much cortisol in the body. Cortisol is a hormone made and released by the adrenal glands.

If a tumor is causing hypercortisolism, decisions about treatment will be made based on the size of the tumor and possibly information learned from imaging studies. Without treatment, this condition can increase the risk of blood clots and problems with electrolytes, minerals the body needs to function.

Adrenal glands and cortisol

The adrenal glands make the hormone cortisol. Cortisol helps the body use glucose (a sugar), protein, and fats. A tumor of the adrenal gland can cause hypercortisolism.

Hypercortisolism occurs when there is too much cortisol in your body. Treatments for this condition are listed in **Guide 3**.

Hypercortisolism can happen with or without Cushing syndrome, which is explained next.

Cushing syndrome

When the body is exposed to hypercortisolism over a long period of time, Cushing syndrome can develop.

Cushing syndrome can cause facial roundness, weight gain around the middle body and upper back, thinning of the arms and legs, easy bruising, and stretch marks.

Guide 3
Treatment for adrenal tumors producing too much cortisol

If the tumor is smaller than 4 cm* and appears benign	You'll have an adrenalectomy
If the tumor is 4 cm* or larger and appears malignant	If the tumor hasn't spread (is not metastatic), you'll have an adrenalectomy
	If the tumor has spread (metastatic), you'll be treated for adrenocortical carcinoma

* Some cancer centers use 6 cm.

Tumor testing

Imaging and other tests will be done to learn more about the tumor. Treatment is based on if the tumor:

- Is smaller than 4 cm, or 4 cm or larger
- Has spread to any nearby areas
- Looks benign or malignant on imaging scans

Tumors are usually measured in centimeters (cm).

A **benign** tumor is a tumor that is **not cancer**.

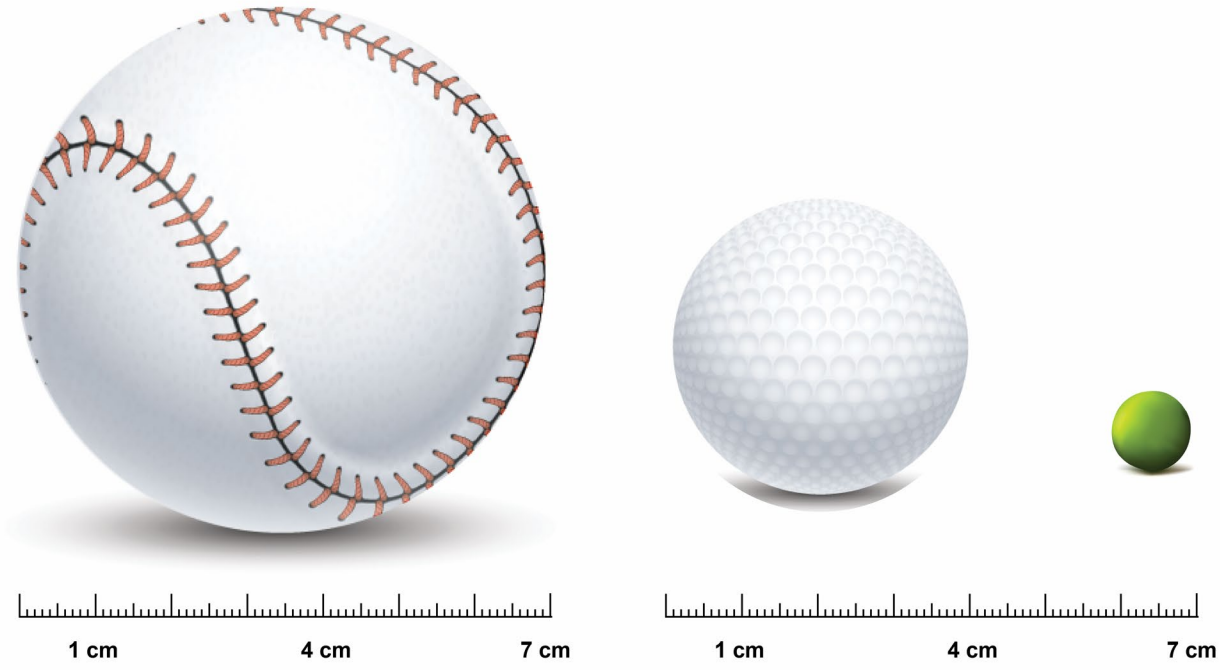
A **malignant** tumor **is cancer**.

Treatment for tumors causing hypercortisolism

If the tumor is smaller than 4 cm

If the tumor is small and doesn't look like cancer, the adrenal gland will be removed. This is called an adrenalectomy. Minimally invasive surgery is preferred.

Adrenal tumors are measured in centimeters: A baseball is 7 cm, a golf ball is 4 cm, and a pea is 1 cm.



If the tumor is 4 cm or larger

If the tumor is large and your team suspects it might be cancer, you'll have additional imaging tests, including:

- FDG-PET/CT
- Chest CT, possibly with contrast

You'll have one of the following imaging studies:

- CT without contrast, and possibly another with contrast. Additional imaging may be done.
- MRI, with and without contrast

After imaging

If the tumor hasn't grown into nearby areas

You'll have an adrenalectomy. Minimally invasive surgery is preferred. Your surgeon will remove the adrenal gland that has the tumor and possibly other areas where the cancer has spread.

If there is metastatic disease

If the tumor has spread, or metastasized, you'll be treated for adrenocortical carcinoma, which will be discussed in the next chapter.

Key points

- Cortisol is a hormone made and released by the adrenal glands. It helps the body use glucose (a sugar), protein, and fats.
- Adrenal gland tumors are usually not cancer.
- A tumor in the adrenal gland can cause hypercortisolism. This is when your body is exposed to too much cortisol.
- When the body is exposed to hypercortisolism over a long period of time, a condition called Cushing syndrome can develop.
- Treatment for hypercortisolism is based on the size of the adrenal tumor, if the tumor has spread to any nearby areas, and if the tumor looks benign or malignant on imaging.
- Treatment is usually surgery to remove the tumor and adrenal gland. Nearby lymph nodes or other structures might be removed.
- If adrenal tumors have spread, they're treated as adrenocortical carcinoma.

Questions to ask

- What treatment(s) do you recommend and why?
- What can I expect from treatment?
- Will I have surgery and, if so, what type of surgery?
- What will be removed during surgery?
- Will I have any treatment for follow-up?

6
Adrenocortical carcinoma

30 About adrenal tumors of the gland’s outer layer

31 Testing and treatment

32 After treatment

33 Key points

33 Questions to ask

Adrenocortical carcinoma (ACC) is cancer that forms in the outer layer of the adrenal gland, called the adrenal cortex. These tumors often make and release excess hormones. The usual treatment for ACC is surgery to remove the adrenal gland, tumor, or metastases. Treatment aims to prevent the spread of cancer.

About adrenal tumors of the gland's outer layer

Adrenocortical carcinoma (ACC) is cancer that forms in the cortex (outer layer) of the adrenal glands. Not all ACCs cause symptoms.

Most ACCs cause the adrenal gland to make extra hormones, usually cortisol and androgen. Tumors can secrete more than a single hormone.

Some ACCs don't affect hormone production, but they can grow large and press on nearby organs and tissues.

Inherited conditions

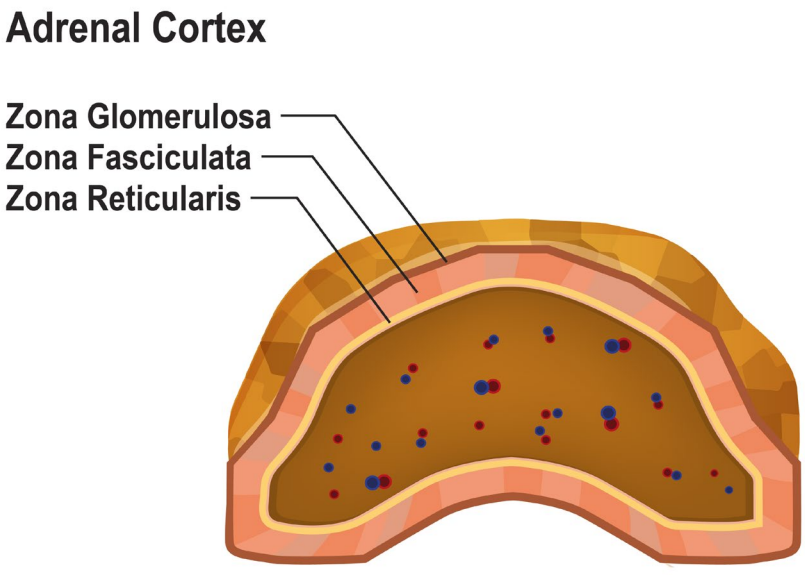
ACC has been linked to some cancers, as well as genetic conditions, such as:

- Li-Fraumeni syndrome
- Lynch syndrome
- Multiple endocrine neoplasia type 1
- Familial adenomatous polyposis

Genetic counseling and testing for inherited genetic syndromes might be done if genetics is suspected to be the cause of ACC.

Adrenal gland

Adrenocortical carcinoma forms in the cortex, the outer layer of the adrenal gland.



Testing and treatment

If ACC is suspected

If your care team thinks you have ACC, you'll have imaging scans and other tests to learn more about the tumor.

Tests will be done to see if the tumor is producing hormones, such as cortisol, aldosterone, and androgen.

If ACC is confirmed and your care team determines the tumor:

- Can be removed surgically, you'll have an adrenalectomy (removal of adrenal gland)
- Can't be removed or they think the cancer may have metastasized, you'll have additional testing.
  - If additional testing determines that cancer has not spread from where it started, you'll have an adrenalectomy. Open surgery is preferred. The tumor and lymph nodes may be removed.

For local cancer

After surgery, if there's a high risk that cancer might return:

- A clinical trial is recommended if available
- You may have radiation therapy to the area where the tumor was removed
- Mitotane, a medicine that kills cancer cells, may be given after other treatments

Making treatment decisions

Treatment decisions are made, in part, based on where cancer is found. So,

Local means cancer is limited to the site where it started.

Regional means cancer has spread to nearby lymph nodes, tissues, or organs.

Locoregional means cancer is both in the site where it started and has spread nearby.

Metastatic means cancer has spread to distant parts of the body.

For locoregional cancer that can't be removed or is metastatic

You may be treated with one or more of the following:

- Observation, with imaging
- Surgery to remove the main tumor and any cancer that has spread
- Radiation, ablation, or other therapy
- Systemic therapy in a clinical trial, if possible

Systemic therapy recommendations are listed in **Guide 4**.

After treatment

You'll be monitored for signs that cancer has returned, grown, or spread. You may have imaging tests, such as CT or MRI scans, with intervals ranging from every 3 months to once per year for up to 10 years. You may also have other kinds of tests. Tests may be done after 10 years if your care team thinks they're needed.

"When first diagnosed, trust and let your doctors do the research and testing—and report them to you."

Guide 4
Adrenocortical carcinoma (ACC): Recommended systemic therapies

Preferred therapies	• Carboplatin with Etoposide (Etopophos) • Cisplatin with Etoposide • (Doxorubicin, Mitotane, or both might be added to either regimen)
Other recommended therapies	• Cabozantinib (Cabometyx) • Mitotane (Lysodren) • Pembrolizumab (Mitotane might be added)
Therapies used in certain cases	• Nivolumab with Ipilimumab • Osilodrostat (Isturisa) (to manage symptoms)

Key points

- Adrenocortical carcinoma (ACC) is a cancer that forms in the cortex (outer layer) of the adrenal glands.
- ACC can cause the adrenal glands to make extra hormones.
- ACC has been linked to some genetic conditions.
- Primary treatment for ACC depends on whether the tumor can or cannot be removed with surgery.
- Additional treatment decisions are made based on whether the cancer has spread or not.
- If cancer has spread to distant parts of the body, you may have surgery, radiation, or systemic therapy.
- After treatment, you'll have follow-up tests every 3 months to once per year for up to 10 years.

Questions to ask

- Can the tumor be removed with surgery? Why or why not?
- What might be removed during surgery?
- If surgery isn't an option, what treatment(s) do you recommend and why?
- What are the treatment side effects?
- Is there a clinical trial available?

7 Paraganglioma and pheochromocytoma

35 About these rare tumors

36 Testing and treatment

38 Multiple endocrine neoplasia type 2

39 Key points

39 Questions to ask

Paraganglioma (PGL) is a neuroendocrine tumor that forms near the adrenal glands. The adrenal glands are part of the endocrine system.

Pheochromocytoma (PCC) is a type of paraganglioma that forms in the center of the adrenal gland, causing the gland to make too much adrenaline.

In this chapter, we'll discuss testing and treating these tumors. We'll also discuss multiple endocrine neoplasia type 2 (MEN2), an inherited condition that increases the risk of developing PGL and PCC.

About these rare tumors

This section provides an overview of paragangliomas, pheochromocytomas, and inherited conditions that might put you at increased risk of these rare tumors.

These tumors can grow slowly. But they can grow fast. Even when they are considered benign, they may be treated like cancer.

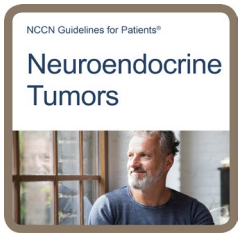
Neuroendocrine means related to the **nervous system** (brain, spinal cord, nerves) and the **endocrine system**.

Paraganglioma

Paraganglioma (PGL) is a rare type of tumor that forms near blood vessels and nerves outside of the adrenal glands. The nerve cells affected are part of the nervous system.

Paragangliomas can be found anywhere along the spine. However, this chapter will only discuss paragangliomas found near or outside of the adrenal gland. Paragangliomas can spread to other areas of the body, such as the lymph nodes, bones, lungs, or liver.

Learn about other neuroendocrine tumors at *NCCN Guidelines for Patients: Neuroendocrine Tumors*, available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



Pheochromocytoma

Pheochromocytoma (PCC) is a rare tumor that develops in the center of the adrenal gland.

Adrenaline and noradrenaline

The tumor grows from cells that produce hormones, including adrenaline and noradrenaline.

These hormones trigger your body's fight-or-flight response to act quickly to a threat. Pheochromocytoma causes these hormones to be released, even when you are not in danger. That's why it can cause:

- High blood pressure
- Pounding headaches
- Irregular heartbeat
- Sweating

Inherited conditions

Some rare hereditary conditions can increase the risk of PGL and PCC, including:

- Hereditary PCC/PGL syndrome
- Multiple endocrine neoplasia type 2 (MEN2)
- Neurofibromatosis type 1
- von Hippel-Lindau syndrome

If you are diagnosed with MEN2, you'll have genetic counseling and testing. You'll be evaluated for cancer and other issues that MEN2 can be related to (including PCC). You can read more about MEN2 later in this chapter.

Testing and treatment

Tests, such as imaging scans and blood, urine, and tumor mutation tests, are used to diagnose a PGL or PCC and plan treatment. Genetic counseling and testing for inherited genetic syndromes might be done if a genetic cause is suspected. Most people with PGL or PCC are offered genetic testing.

Additional testing will be done if the tumor has spread or more tumors have been found near the original site.

Before treatment

Before starting treatment:

- You'll likely be given alpha blockers to lower your blood pressure.
- You'll be asked to eat a high-salt diet and drink a lot of fluids for a few days before procedures.
- One or more medicines might be added to the alpha blockers:
  - Dihydropyridine calcium channel blocker
  - Beta blocker
  - Metyrosine

Treatments you may have

Treatment is based on whether the tumor can or cannot be removed with surgery, and if tumors have spread to other areas of the body, such as the lymph nodes, bones, lungs, or liver.

For a PGL or PCC that can be removed

Minimally invasive adrenalectomy, to remove the adrenal gland, is recommended whenever possible.

For a PGL or PCC that can't be removed

If the tumor has spread locally

Your condition will be monitored for any changes or symptoms. This is called observation. Or, if the tumor is producing hormones, you'll continue taking the alpha blocker, and you may:

- Be treated in a clinical trial
- Have radiation and possibly surgery to reduce the tumor size
- Have systemic therapy, or radiation therapy to treat metastases or metastases causing symptoms.

If the tumor has spread to distant parts of the body

Your condition will be observed for any changes or symptoms. Or, if the tumor is giving off hormones, you'll continue taking the alpha blocker, and you may:

- Be treated in a clinical trial
- Have surgery to reduce the tumor size
- Have systemic therapy or radiation therapy to treat metastases or metastases causing symptoms.

Good to know:
Pheochromocytomas rarely spread.

Systemic therapy may be used when a tumor can't be removed. Recommended therapies are listed in **Guide 5**.

About systemic therapy recommendations for PGL and PCC

Here are a few things to know about the recommendations in **Guide 5**.

Preferred therapy

Enrolling in a clinical trial is the preferred therapy for PGL and PCC.

Other recommended therapies

CVD stands for cyclophosphamide, vincristine, and dacarbazine.

About tumors with somatostatin receptors

Some tumors have receptors on their surface for the hormone somatostatin. They're called somatostatin receptor (SSTR)-positive tumors.

Some therapies are recommended only for tumors that have these somatostatin receptors. Here's how these therapies work:

- **PRRT** is peptide receptor radionuclide therapy. Peptides are short proteins. With PRRT, lab-made somatostatin peptides work with a radionuclide (a radioactive element) to target the cancer cells.
- **SSAs** are somatostatin analogues. An SSA is similar to the somatostatin the body makes and is used to help manage tumor side effects, like diarrhea and flushing (redness of the face, neck, and chest).

Therapies used in certain cases

Selpercatinib can be used for tumors that have a certain biomarker and haven't responded to systemic treatment.

After treatment

Tests are done at specific times after treatment to check on any changes in your condition. This is called surveillance. You can expect to have blood and urine tests and to have your blood pressure monitored every 3 months to a year. You may also have imaging tests. CT or MRI is most common. But you may have imaging tests in addition to or instead of these, especially if further therapies are being considered.

Multiple endocrine neoplasia type 2

If you are diagnosed with multiple endocrine neoplasia type 2 (MEN2), you'll be evaluated for PCC. You will have blood and urine tests. To learn more about next steps, see the "Testing and treatment" section earlier in this chapter.

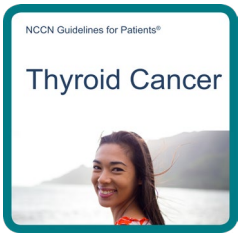
You'll also be evaluated for:

- **Medullary thyroid carcinoma**. This is a rare type of thyroid cancer. Testing will include blood tests and imaging of the thyroid and neck lymph nodes.
- **Parathyroid gland problems**. Parathyroid glands are small glands in the back of the thyroid that make a hormone that controls blood calcium. Problems with these glands can cause levels of calcium to get too high.
  - If calcium levels are too high, testing will include blood and urine tests and imaging scans. Treatment is surgical removal of the parathyroid.

Guide 5
Pheochromocytoma and paraganglioma: Recommended systemic therapies

Preferred therapy	Clinical trial
Other recommended therapies	• Belzutifan (Welireg) • Cabozantinib (Cabometyx) • Sunitinib (Sutent) • CVD or Temozolomide (Temodar)
	If the tumor is SSTR-positive: • PRRT with Lutetium Lu 177 dotatate (Lutathera) • SSAs (Octreotide LAR or Lanreotide)
Therapies used in certain cases	Selpercatinib (Retevmo)

More information on treatments for medullary thyroid carcinoma and parathyroid gland problems is available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



Key points

- Paraganglioma (PGL) is a rare type of tumor that forms near blood vessels and nerves outside of the adrenal gland.
- Pheochromocytoma (PCC) is a rare tumor that develops in the center of the adrenal gland causing the gland to overproduce hormones.
- Multiple endocrine neoplasia type 2 (MEN2) and some other inherited conditions can be related to PGL and PCC.
- You'll have blood and urine tests and imaging scans to diagnose a PGL or PCC.
- PCC can cause symptoms, including high blood pressure, headaches, irregular heartbeat, and sweating.
- Surgery and systemic therapy may be part of treatment.
- Before treatment for PCC, you may be prescribed medicine (alpha blockers) to lower your blood pressure.

Questions to ask

- What type of tumor do I have?
- Is the tumor producing any hormones?
- What are my treatment options?
- What treatment(s) do you recommend and why?
- What are the treatment side effects?

8
Other resources

41 What else to know

41 What else to do

41 Where to get help

42 Questions to ask

Want to learn more? Here's how you can get additional help.

What else to know

This guide helps you know your options so you can make informed decisions and improve your cancer care. But it's not the only resource that you have.

Ask for as much information and help as you need. Many people are interested in learning more about:

- Finding an oncologist or doctor who's an expert in adrenal tumors
- Making treatment decisions
- The details of treatment and its side effects
- Getting financial help
- Coping with other health challenges

What else to do

Your health care center can help you with next steps. It often has on-site resources to help meet your needs and find answers to your questions. Health care centers can also inform you of resources in your community.

In addition to help from your providers, the resources listed in the next section provide support for many people like yourself.

Where to get help

Look through the list below and visit the provided websites to learn more about these organizations.

Bone Marrow & Cancer Foundation
bonemarrow.org

Cancer*Care*
cancercare.org

Imerman Angels
Imermanangels.org

Neuroendocrine Cancer Awareness Network
netcancerawareness.org

Neuroendocrine Tumor Research Foundation
netrf.org

The Healing NET Foundation
thehealingnet.org

Triage Cancer
triagecancer.org

Questions to ask

- Who can I talk to about help with housing, food, and other basic needs?
- What help is available for transportation, childcare, and home care?
- What other services are available to me and my caregivers?
- How can I connect with others and build a support system?
- Who can I talk to if I don't feel safe at home, at work, or in my neighborhood?



We want your feedback!

Our goal is to provide helpful and easy-to-understand information on cancer. Take our survey to let us know what we got right and what we could do better.
NCCN.org/patients/feedback



Words to know

adrenalectomy
Surgery to remove one or both adrenal glands.

adrenal gland
A small organ on top of each kidney that makes hormones.

adrenocortical carcinoma (ACC)
A rare type of adrenal cancer.

aldosterone
A hormone that helps balance potassium and sodium in the body.

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

carcinoma
A cancer of cells that line the inner or outer surfaces of the body.

chemotherapy
Medicine that kills cancer cells by damaging or disrupting the making of the genetic code.

clinical trial
A type of research that assesses health tests or treatments.

computed tomography (CT)
A test that uses x-rays from many angles to make pictures of the inside of the body.

Conn syndrome
A condition that occurs due to too much aldosterone being produced by the adrenal glands. Also called primary aldosteronism.

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

cortisol
A hormone that controls blood sugar, metabolism, and other functions in the body.

Cushing syndrome
A condition caused by the release of excess cortisol in the body over a long period of time.

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

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

hormone
A chemical in the body that triggers a response from cells or organs.

hypercortisolism
A condition caused by the release of excess cortisol in the body.

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

immunotherapy
Treatment with medicine that help the body's own immune system find and destroy cancer cells.

lymph node
A small, bean-shaped disease-fighting structure.

magnetic resonance imaging (MRI)
A test that uses radio waves and powerful magnets to make pictures of the insides of the body.

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

multiple endocrine neoplasia syndrome type 2 (MEN2)
An inherited condition that increases the risk of developing paragangliomas and pheochromocytomas.

mutation
An abnormal change in cells.

paraganglioma (PGL)
A tumor that arises from neuroendocrine tissues found along the spine.

peptide receptor radionuclide therapy (PRRT)
A type of systemic therapy that is used to treat tumors with somatostain receptors.

pheochromocytoma (PCC)
A tumor that forms in the center of the adrenal gland that causes it to make too much adrenaline.

positron emission tomography (PET)
A test that uses radioactive material to see the shape and function of body parts.

primary aldosteronism
A condition that occurs due to too much aldosterone being produced by the adrenal glands. Also called Conn syndrome or hyperaldosteronism.

radiation therapy (RT)
Treatment that uses high-energy rays or related approaches to kill cancer cells.

somatostatin
A hormone that attaches to receptors and controls the endocrine system and nervous system.

somatostatin analogue (SSA) therapy
Treatment to slow down or stop tumor hormone production.

somatostatin receptor (SSTR)
Receptors found on some types of tumors that cause the tumor to release extra hormones.

supportive care
Health care that includes symptom relief but not cancer treatment.

surgery
An operation to remove or repair a part of the body.

targeted therapy
Drug treatment that impedes the growth process specific to cancer cells.

NCCN Contributors

This patient guide is based on the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Neuroendocrine and Adrenal Tumors, Version 1.2026. It was adapted, reviewed, and published with help from the following people:

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Cancer care recommendations from leading experts at the
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