NCCN GUIDELINES FOR PATIENTS®

2022

Adrenal Tumors

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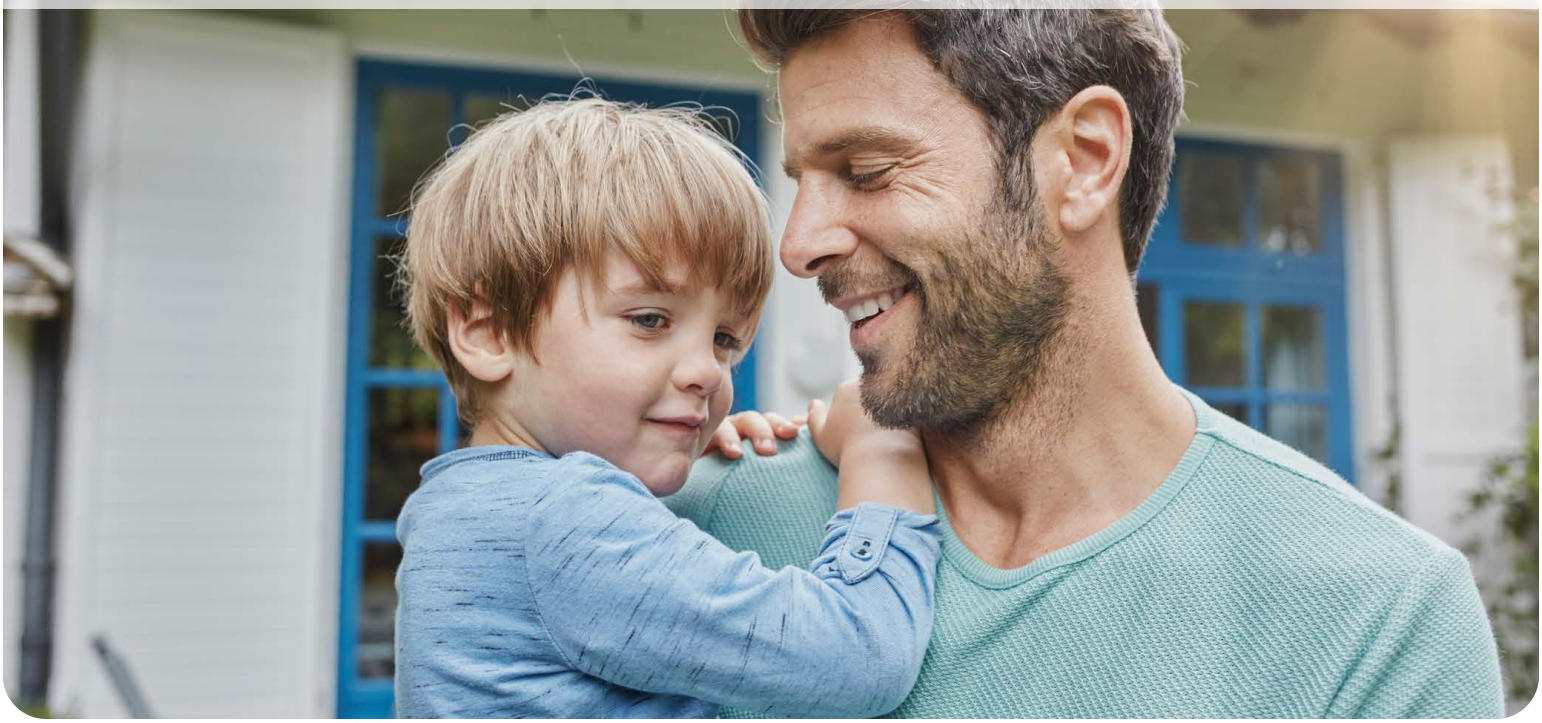

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These NCCN Guidelines for Patients are based on the NCCN Guidelines® for Neuroendocrine and Adrenal Tumors, Version 1.2022 — May 23, 2022.

NCCN Foundation seeks to support the millions of patients and their families affected by a cancer diagnosis by funding and distributing NCCN Guidelines for Patients. NCCN Foundation is also committed to advancing cancer treatment by funding the nation's promising doctors at the center of innovation in cancer research. For more details and the full library of patient and caregiver resources, visit NCCN.org/patients.

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Adrenal basics

Adrenal tumors form in one or both adrenal glands. Adrenal glands produce hormones that affect your body in different ways. This chapter reviews the basics of adrenal tumors.

Adrenal glands

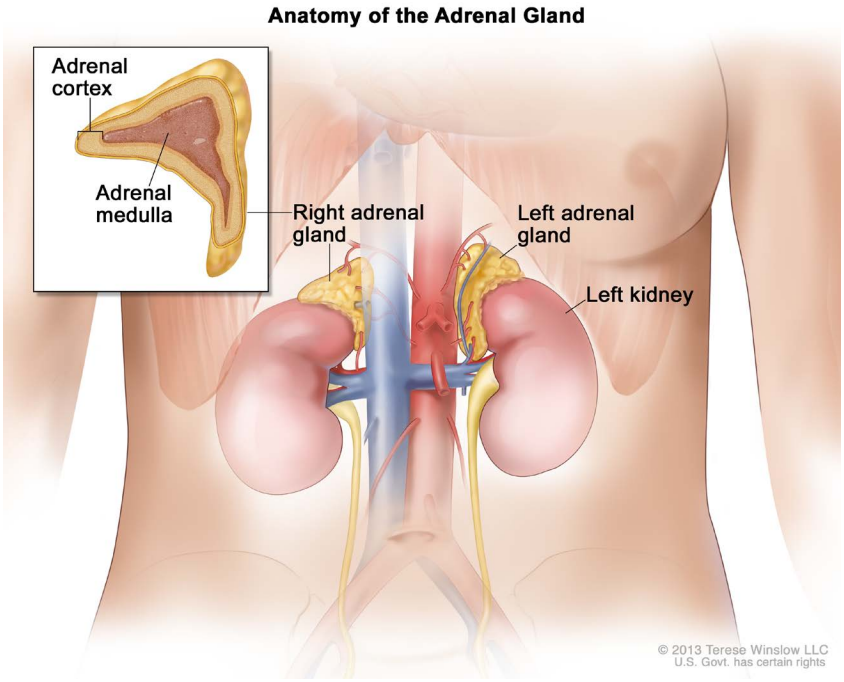
Your body has 2 adrenal glands. They are located on top of each kidney. Adrenal glands are small, yellow glands that are important to the body's endocrine system. The endocrine system is made up of tissues and organs that produce hormones. Hormones are chemical substances that are made in one part of the body and carried through the bloodstream to other parts of the body where they help control how organs or cells work.

Each adrenal gland has 2 main parts:

- **Adrenal cortex** – This is the outer part of the adrenal gland. The adrenal cortex makes 3 main hormones: cortisol, aldosterone, and dehydroepiandrosterone (DHEA). These hormones control metabolism, blood pressure, and body features (such as hair growth and body shape).
- **Adrenal medulla** – This is the inner part of the adrenal gland. The medulla makes 3 hormones: epinephrine, norepinephrine, and dopamine. These hormones control the body's response to stress. This includes the "fight or flight" surge of adrenaline.

Adrenal glands

Adrenal glands are small, triangle-shaped glands found on the top of the kidneys. These glands make hormones that help to regulate your immune system, response to stress, metabolism, and other functions.



Adrenal tumors

Adrenal tumors can occur in one or both adrenal glands. Most adrenal gland tumors are benign (not cancer). Adrenal tumors often do not cause symptoms and are found through an imaging test such as a CT or MRI scan performed for another reason.

Adrenal tumors are categorized in the following ways:

- **Benign (not cancer) adrenal tumors** do not metastasize (spread) to other parts of the body. This type of tumor is often found by chance through imaging tests done for an unrelated issue.
- **Functional adrenal tumors** are usually benign, although some are capable of becoming cancerous and spreading. Benign functional tumors can still produce hormones and may be found during tests for hormone-related symptoms.
- **Malignant adrenal tumors** are cancerous tumors. They are rare, with about 300 to 500 people being diagnosed each year. Certain genetic conditions may increase the risk for these tumors.

Types

Adrenal tumor categories are further broken down into types. They include the following:

- **Adenoma** – This is the most common type of adrenal gland tumor, also referred to as adrenocortical adenoma. It is a noncancerous tumor that can be functioning or nonfunctioning. If an adenoma is small and/or does not produce excessive hormones, and is not causing symptoms, it may not need treatment. An adenoma that produces too much of the hormone aldosterone is called an aldosteronoma. Whereas, an adenoma that produces too much of the hormone cortisol (stress hormone) causes a condition called Cushing syndrome.
- **Adrenocortical carcinoma (ACC)** – Although it is the most common type of adrenal gland cancer, it is very rare and affects approximately 1 to 2 people out of every 1 million. Adrenocortical carcinoma begins in the adrenal cortex. It can be a functioning or nonfunctioning tumor. If the tumor is functioning, it may produce more than 1 hormone.
- **Neuroblastoma** – This rare type of adrenal tumor is cancerous. It affects children ages 5 and under, though it is most often found in infants. Neuroblastoma can spread to other parts of the body, including the bones.
- **Pheochromocytoma** – This type of tumor begins in the adrenal medulla and is characterized by symptoms of high blood pressure, headache, rapid heartbeat, and sweating. Most pheochromocytomas are benign, but can be cancerous.

Adrenocortical carcinoma

Adrenocortical carcinoma (ACC) is an adrenal cancer that starts in one or both of the adrenal glands. It can occur at any age. But it's most likely to affect children ages 5 and under, and adults in their 40s and 50s. If caught early, adrenal cancer can be cured. However, if the cancer has spread beyond the adrenal glands, cure becomes less likely. Treatment can be used to delay progression or recurrence.

Signs and symptoms

Half of those with adrenal cancer will have symptoms caused by hormones produced by the tumor. Others may experience symptoms if the tumor has grown so large that it is pressing on nearby organs. If you have any of the signs or symptoms listed below, make sure to discuss them with your health care provider.

Symptoms of adrenal cancer may include:

- Weight gain
- Muscle weakness
- Pink or purple stretch marks on the skin
- Hormonal changes such as excess facial hair, hair loss on the head, irregular periods, enlarged breast tissue, and shrinking testicles
- Nausea
- Vomiting
- Abdominal bloating
- Back pain
- Fever
- Loss of appetite
- Loss of weight without trying

Risk factors

There are no known causes for most adrenal tumors. However, some genetic (hereditary) conditions can put you at risk for developing an adrenal tumor. Hereditary conditions are mutations passed down from biological parent to child through genes. Those with one of the conditions listed below might have a higher risk for developing an adrenal tumor. This accounts for only a small portion of cases.

- Beckwith-Wiedemann syndrome
- Carney complex
- Familial adenomatous polyposis (FAP)
- Hereditary paraganglioma-pheochromocytoma (PGL/PCC) syndrome
- Li-Fraumeni syndrome
- Lynch syndrome
- Multiple endocrine neoplasia type 1 (MEN 1)
- Multiple endocrine neoplasia type 2 (MEN 2)
- Neurofibromatosis type 1 (NF1)
- Von Hippel-Lindau syndrome (VHL)

Some of these conditions can also put one at risk for developing a kidney tumor. For more information, read the *NCCN Guidelines for Patients: Kidney Cancer,* available at NCCN.org/patientguidelines.

Key points

- Adrenal glands are small, yellow glands that are important to the body's endocrine system.
- The endocrine system is made up of tissues and organs that produce hormones.
- Hormones are chemical substances that are made in one part of the body and are carried through the bloodstream to other parts of the body where they help control how organs or cells work.
- Adrenal tumors are tumors that can be found in the adrenal glands in one or both adrenal glands. Most tumors found on the adrenal glands are benign (not cancer).
- Adrenal cancer is a rare cancer that starts in one or both of the adrenal glands.
- Adrenal cancer, also called adrenocortical cancer (ACC), can occur at any age. But it's most likely to affect children ages 5 and under, and adults in their 40s and 50s.
- Half of those with adrenal cancer will have symptoms caused by hormones from the tumor. Others may experience symptoms if the tumor has grown so large that it is pressing on nearby organs.
- There are no known causes for most adrenal tumors. However, hereditary conditions have been linked to an increased risk of developing an adrenal tumor.

2
Testing

Treatment planning starts with testing. Testing is used to diagnose and plan treatment for adrenal tumors. This chapter presents an overview of the tests you might receive and what to expect.

Test results

Adrenal tumors will be diagnosed based on a series of test results. Your diagnosis will determine your treatment plan. It is important you understand what these tests mean.

Keep these things in mind:

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

Create a medical binder

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

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

General health tests

Medical history

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

Family history

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

Physical exam

During a physical exam, a health care provider may:

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

What is your family health history?

Some cancers and other diseases run in families—those who are related to you through genes passed down from parent to child. This information is called a family health history. You can ask family members about their health issues like heart disease, cancer, and diabetes, and at what age they were diagnosed. For relatives who have died, ask about the cause and age of death.

Start by asking your parents, siblings, and children. Next, talk to half-siblings, aunts and uncles, niece and nephews, grandparents, and grandchildren.

Write down what you learn about your family history and share this information with your health care provider.

Some of the questions to ask include:

- Do you have any chronic diseases, such as heart disease or diabetes, or health conditions such as high blood pressure or high cholesterol?
- Have you had any other diseases, such as cancer or stroke?
- How old were you when each of these diseases and health conditions was diagnosed?
- What is our family's ancestry—from what countries did our ancestors originate?

Imaging tests

Imaging tests take pictures of the inside of your body. A radiologist, an expert in interpreting test images, will write a report and send this report to your doctor. Your test results will be discussed with you.

CT scan

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

Contrast

Contrast materials are not dyes, but substances that help enhance and improve the images of several organs and structures in the body. Contrast is used to make the pictures clearer. It might be taken by mouth (oral) or given through a vein (IV). The contrast is not permanent and will leave the body in your urine after the test. The types of contrast vary but are different for CT than for MRI.

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

MRI scan

A magnetic resonance imaging (MRI) scan uses radio waves and powerful magnets to take pictures of the inside of the body. It does not use x-rays. Contrast might be used. An MRI may be used as an initial test, to check treatment results, and to see if the cancer has spread to other parts of the body.

PET scan

A positron emission tomography (PET) scan uses a radioactive drug called a tracer. A tracer is a substance injected into a vein to help see cancer cells in the body. PET scans are performed about an hour after the tracer is injected. PET scans are combined with CT or MRI scans (PET/CT or PET/MRI) to determine where the cancer cells are located. This combined test is considered to be more accurate than either scan alone. Types of PET scans that can be helpful for those with adrenal tumors include FDG-PET and SSTR-PET.

FDG-PET/CT scan

An FDG-PET scan uses a radioactive type of sugar (18-fluorodeoxyglucose) as its tracer. Active cancer cells use sugar faster than normal cells. This will make the cells look brighter in pictures. Many adrenal tumors are not fast-growing and not very bright on FDG-PET scans. FDG-PET scans can help determine if neuroendocrine tumors are becoming more aggressive.

SSTR-PET/CT scan

A somatostatin receptor (SSTR) antagonist is a protein found on the surface of cells that bind to a hormone called somatostatin. Somatostatin helps to control other hormones in the body. SSTRs are found on many different types of cells. A SSTR-PET uses a tracer that binds to these SSTRs.

Blood tests

Blood tests are used to look for signs of hormone secretion or to see how well you are responding to certain treatments.

Biochemical tests

Biochemical tests measure substances like hormones in blood, urine, and/or saliva that may be made by the adrenal tumor. Symptoms may be seen when your body makes too much of a hormone. Your doctor may consider these tests if you may have an adrenal tumor or syndromes caused by a tumor. More information on biochemical tests can be found in Guide 1.

Guide 1
Biochemical tests

Tumor or syndrome	Symptoms	Possible tests
Pheochromocytoma or paraganglioma	• Hypertension • Tachycardia • Sweating • Fainting	• Plasma free or 24-hour urine fractionated metanephrines and normetanephrines with/without serum or 24-hour urine catecholamines • Serum and urine catecholamines or methoxytyramine
Hypercortisolemia	• Weight gain • Stretch marks • Hypertension • Excessive hair growth • Hyperglycemia • Depression	• Overnight dexamethasone suppression • Midnight salivary cortisol • 24-hr urinary free cortisol • If positive, retest • Plasma ACTH
Primary aldosteronism	• Hypertension • Low potassium	• Suppressed renin/renin activity with an elevated plasma aldosterone level • If positive, retest
Suspected or confirmed adrenocortical carcinoma	• Weight gain • Stretch marks • Hypertension • Excessive hair growth • Hyperglycemia • Depression	• Overnight dexamethasone suppression • Midnight salivary cortisol • 24-hour urinary free cortisol • If positive, retest • Plasma ACTH • Testosterone • DHEA-S

Adrenal vein sampling

Adrenal vein sampling (AVS) is a procedure where blood is taken from both adrenal glands to measure how much hormone is being produced by each gland. A needle and catheter (plastic tube) is inserted into the vein of your groin. Contrast is used to map your veins before the doctor places the tube.

Genetic tests

Genetic testing is done using blood or saliva (spitting into a cup). The goal is to look for gene mutations. Not all mutations are inherited. Mutations inherited from your biological parents are called germline mutations. Some mutations can put you at risk for more than one type of cancer. You can pass these germline mutations on to your children. Also, other family members may have these mutations.

Tell your doctor if there is a family history of cancer or tumors. Depending on your family history or other features of your cancer, your health care provider might refer you for genetic testing to learn more about inherited risks for cancer. You may meet with a genetic counselor to discuss genetic testing options and/or to explain your test results.

Hereditary conditions

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

Biopsy

Biopsies are rarely done for adrenal tumors. It might be done to rule out pheochromocytoma. A biopsy refers to obtaining a sample of tissue or group of cells for testing. Your sample should be reviewed by a pathologist who is an expert in the diagnosis of adrenal tumors. This review is often referred to as histology, histopathology, or hematopathology review. The pathologist will note the overall appearance and the size, shape, and type of your cells. Tests will be done on the biopsied cells.

Image-guided needle biopsy

An image-guided needle biopsy refers to a biopsy that uses an imaging tool (such as ultrasound or CT) to guide the needle during the procedure. This type of biopsy allows your doctor to biopsy suspicious areas that are not easily seen or felt through your skin.

Key points

- Adrenal tumors can be diagnosed based on a series of test results. Your diagnosis will determine your treatment plan.
- Your doctor will ask about the health history of family members who are blood relatives. This information is called a family history.
- A computed tomography (CT or CAT) scan uses x-rays and computer technology to take pictures of the inside of the body.
- A magnetic resonance imaging (MRI) scan uses radio waves and powerful magnets to take pictures of the inside of the body. It does not use x-rays.
- Contrast materials are not dyes, but substances used to make imaging pictures clearer.
- Biochemical tests measure substances like hormones in blood, urine, and/or saliva that may be made by the adrenal tumor.
- Some genetic (hereditary) conditions can put you at risk for developing an adrenal tumor. Genetic testing if often offered to those with adrenal cancer or tumors.
- A biopsy obtains a sample of tissue or group of cells for testing. Biopsies are rarely done for adrenal tumors.

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

3
Treatment overview

This chapter presents an overview of the treatment types for adrenal tumors. Not every person will receive every treatment listed in this chapter. Together, you and your doctor will choose a treatment plan that is best for you.

Treatment team

Treatment decisions should involve a multidisciplinary team (MDT). An MDT is a team of doctors, health care workers, and social care professionals from different professional backgrounds who have knowledge (expertise) and experience with adrenal tumors. This team is united in the planning and implementing of your treatment.

The MDT may include pathologists, endocrinologists, radiologists (including nuclear medicine specialists), nutritionists, and medical, radiation, and surgical oncologists.

Ask who will coordinate your care.

Your physical, mental, and emotional well-being are important. You know your body better than anyone. Help other team members understand:

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

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

Your treatment team

Treating adrenal tumors takes a team of experts. Your treatment team may include:

Diagnostic radiologist: interprets the results of CTs, MRIs, and other imaging tests.

Endocrinologist: specializes in the diagnosis and treatment of disorders of the endocrine system.

Genetic counselor: helps people understand their risk or chance of developing cancer, tumors, or other diseases.

Oncologist: specializes in treating cancer.

Pathologist: analyzes the cells, tissues, and organs removed during a biopsy or surgery and provides cancer diagnosis and staging information.

Surgeon: performs procedures to remove tumors and cancer.

Surgery

Surgery is the primary treatment for most adrenal tumors. Primary treatment refers to the main treatment used to get rid of a tumor in the body. The method and extent of surgery depends on where the tumor is located and how far the disease has spread. Treatment will be based on whether the tumor can be removed by surgery (resectable) or cannot be removed by surgery (unresectable).

Adrenalectomy

An adrenalectomy refers to surgery where one (unilateral) or both (bilateral) adrenal glands are removed. During the procedure, your surgeon will remove the tumor and any areas where the cancer has spread. This can include lymph nodes or nearby structures.

There are 2 types of adrenalectomy:

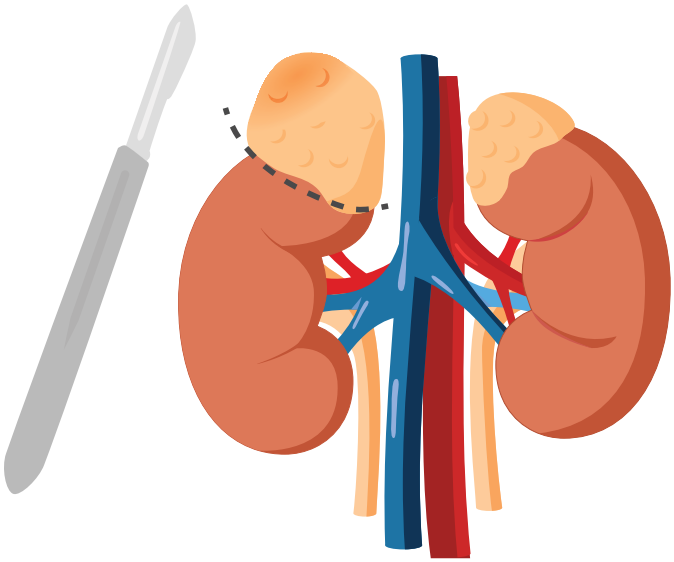
- **Open adrenalectomy** – This is used if the tumor is large, might be cancerous, or is likely to rupture. Your surgeon will make and open incision on your abdomen to remove the gland(s).
- **Laparoscopic adrenalectomy** – This is more commonly used. Your surgeon will make small punctures (either in your abdomen or back), then will use a laparoscope to remove the gland(s). A laparoscope is a thin tube with a camera and a light that can help your surgeon see inside your body.

Resection

A resection is a surgical procedure where a tumor is removed. During the procedure, the tumor along with a small amount of normal, healthy tissue around the tumor will be removed. This rim of normal, healthy tissue is referred to as a surgical margin. If the surgical

Adrenalectomy

During this procedure, one or both adrenal glands are removed.



margin has healthy tissue, then usually the tumor has been removed in its entirety.

A positive margin refers to cancer cells found at the edge of the tissue. This indicates that not all of the cancer has been removed. Despite best efforts this can happen.

Systemic therapy

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

- Systemic therapy given before surgery is referred to as **neoadjuvant therapy.**
- Systemic therapy given after surgery is referred to as **adjuvant therapy**.

Chemotherapy

Chemotherapy kills fast-growing cells throughout the body, including cancer cells and some normal cells. Chemotherapy drugs work in different ways to kill abnormal cells or to stop new ones from being made. When more than one chemotherapy drug is used, it is referred to as a combination regimen.

Chemotherapy is given in cycles of treatment days followed by days of rest. This allows the body to recover before the next cycle of chemotherapy. The length of treatment and rest days vary based on the chemotherapy drugs used.

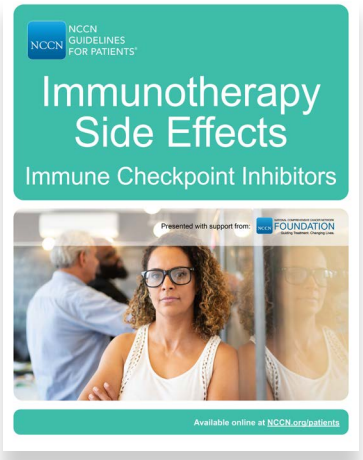
Targeted therapy

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

Immunotherapy

Immunotherapy increases the activity of your own immune system to fight cancer. The immune system is the body's natural defense against infection and disease. By changing how the immune system works, immunotherapy improves your body's ability to find and destroy cancer cells. Immunotherapy can be given alone or with other types of treatment.

For more information, read the *NCCN Guidelines for Patients: Immunotherapy Side Effects,* available at NCCN.org/patientguidelines.



Radiation therapy

Radiation therapy (RT) uses high-energy radiation from x-rays, photons, electrons, and other sources to kill cancer cells and shrink tumors. RT can be given alone or with other treatments. Treatment may focus on individual tumors, a small area/region of the body, or specific lymph nodes. RT may be used as supportive care or palliative care to help ease pain or discomfort caused by cancer.

Radiation therapy can also be given before, during, or after surgery to treat or slow the growth of cancer, especially if the surgical margins have cancer cells.

EBRT

External beam radiation therapy (EBRT) uses a machine outside of the body to aim radiation at the tumor(s) or areas of the body.

Common types of EBRT that may be used include:

- **Three-dimensional conformal radiation therapy (3D-CRT)** uses computer software and CT images to aim beams that match the shape of the tumor.
- **Intensity-modulated radiation therapy (IMRT)** uses small beams of different strengths to match the shape of the tumor.
- **Stereotactic body radiation therapy (SBRT)** uses high-energy radiation beams to treat cancers in five or fewer treatments.
- **Stereotactic radiosurgery (SRS)** uses special equipment to position the body and give one precise, large dose of radiation.

Stereotactic body radiation therapy

Stereotactic body radiation therapy (SBRT) is a type of external beam radiation therapy (EBRT).

SBRT may be used to:

- Relieve symptoms such as pain caused by cancer metastases
- Treat those who cannot have surgery as a primary treatment due to other health conditions
- Shrink tumors
- Prevent recurrence

With SBRT, you will receive high-dose radiation for 1 to 5 treatments. SBRT is very precise, and thereby reduces the chance of damage to nearby tissues. SBRT should be delivered at an experienced, high-volume center with technology that allows for image-guided radiation therapy or in a clinical trial.

Somatostatin analogs

Somatostatin analogs are drugs used to slow down or stop hormone production. Somatostatin analogs are made to have long-lasting activity in the body.

Somatostatin analogs that might be used to treat adrenal tumors include:

- Lanreotide (Somatuline Depot)
- Octreotide acetate (Sandostatin, Bynfezia Pen)
- Octreotide acetate (LAR) (Sandostatin LAR Depot)

Radioactive drugs

Nuclear medicine therapy uses radioactive molecules to target specific tumors. One example is peptide receptor radionuclide therapy (PRRT) with lutetium lu 177-dotatate (Lutathera). Lutathera is a combination of a somatostatin analog and a radioactive drug used to treat certain tumors that are somatostatin receptor-positive. It binds to a protein called somatostatin receptor, which is found on some adrenal tumor cells. Lutetium Lu 177-dotatate builds up in these cells and gives off radiation that may kill them.

Observation

Some people do not need immediate treatment. Observation is a plan that closely watches your condition. Observation may also be referred to as surveillance or watch-and-wait. During this time, you will have tests on a regular basis to look for any new or changing symptoms. You will not have any treatment during observation.

Observation

During a period of observation, you will receive tests to identify any changes, but you will not receive treatment.

Clinical trials

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

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

Phases

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

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

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

Who can enroll?

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

Informed consent

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

Start the conversation

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

Frequently asked questions

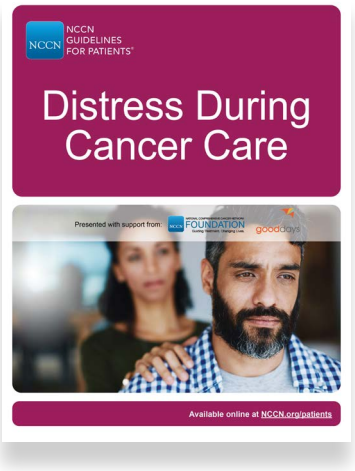
There are many myths and misconceptions surrounding clinical trials. The possible benefits and risks are not well understood by many with cancer.

Will I get a placebo?

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

Are clinical trials free?

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



General supportive care

Supportive care is health care that relieves symptoms and improves quality of life. It might include pain relief (palliative care), emotional or spiritual support, financial aid, or family counseling. Tell your care team how you are feeling and about any side effects. Best supportive care, supportive care, and palliative care are often used interchangeably.

Distress

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

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

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

Fatigue

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

Key points

- Treatment decisions should involve a multidisciplinary team (MDT) that has knowledge (expertise) and experience with adrenal tumors.
- Surgery is the primary treatment for adrenal tumors. The method and extent of surgery depends on where the tumor is located and how far the disease has spread.
- Systemic therapy is used in all stages of cancer treatment. Systemic therapy works throughout the body. Types include chemotherapy, targeted therapy, and immunotherapy.
- Radiation therapy (RT) uses high-energy radiation from x-rays, photons, electrons, and other sources to kill cancer cells and shrink tumors. RT can be given alone or with other treatments.
- Somatostatin analogs are drugs used to slow down or stop hormone production.
- Observation is a plan that closely watches your condition. Observation may also be referred to as surveillance or watch-and-wait.
- Everyone with cancer should carefully consider all of the treatment options available for their cancer type, including standard treatments and clinical trials.
- Supportive care is health care that relieves symptoms and improves quality of life. It might include pain relief (palliative care), emotional or spiritual support, financial aid, or family counseling.

4

Primary aldosteronism (Conn syndrome)

Primary aldosteronism is a condition caused by the adrenal glands producing 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 treatment of primary aldosteronism, also known as hyperaldosteronism or Conn syndrome.

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

Overview

Primary aldosteronism refers to a condition that occurs due to too much aldosterone being produced by the adrenal glands. Aldosterone is the hormone that helps to balance potassium and sodium in the body. Too much aldosterone will cause the body to hold sodium and lose potassium, leading to elevated blood pressure. Without treatment, this condition increases your risk of cardiovascular diseases, including heart attack and stroke. Also, low potassium levels can cause heart rhythm irregularities.

Primary aldosteronism is also referred to as hyperaldosteronism or Conn syndrome.

Symptoms

The main symptom of primary aldosteronism is high blood pressure and low potassium levels.

Other symptoms include:

- Fatigue
- Headaches
- Muscle weakness
- Numbness
- Excessive urination
- Sporadic temporary paralysis

Treatment

Treatment for hyperaldosteronism depends on if excess aldosterone is being produced by one or both adrenal glands. This is determined by a test called adrenal vein sampling (AVS), which can measure hormone levels from each adrenal gland.

- If both adrenal glands are affected, high blood pressure (hypertension) and low potassium levels (hypokalemia) will be treated with medicine, either spironolactone or eplerenone.
- If only one adrenal gland is affected, an adrenalectomy—surgery where one or both adrenal glands are removed—is recommended.

Almost all aldosterone-producing adrenal tumors are benign and will be removed by a laparoscopic surgery. If there is concern that the tumor is cancerous, an open adrenalectomy will likely be recommended.

Key points

- Primary aldosteronism is a condition caused by too much aldosterone being produced by the adrenal glands.
- Aldosterone is 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, this condition increases your risk of cardiovascular diseases, including heart attack and stroke.
- Hyperaldosteronism may result from too much aldosterone being produced by one or both adrenal glands. Adrenal vein sampling (AVS) is used to determine this.
- If both adrenal glands are producing too much aldosterone, treatment will be with medications to lower your blood pressure and raise potassium levels. If too much aldosterone is being produced by just one adrenal gland, removal of that adrenal gland is recommended.

5
Hypercortisolism (Cushing syndrome)

Hypercortisolism occurs when your body is exposed to too much cortisol. Cortisol is a hormone produced during times when you are stressed. A tumor of the adrenal gland can cause too much cortisol. Hypercortisolism, also called Cushing syndrome, is a rare syndrome that can be fatal if left untreated.

Overview

Hypercortisolism occurs where there is too much cortisol in your body. Cortisol is a hormone that helps the body use glucose (a sugar), protein, and fats. A tumor of the adrenal gland can cause hypercortisolism. These tumors are usually benign (not cancer).

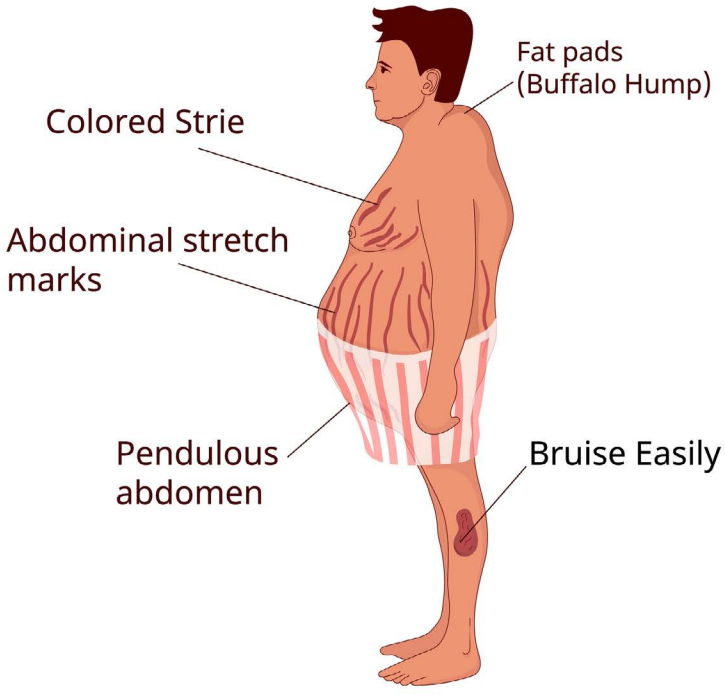
Symptoms

Hypercortisolism symptoms caused by an adrenal tumor include:

- Obesity and the development of stretch marks
- Unexplained weight gain often with a rounding of the face (“moon face”)
- Thinning of the arms and legs
- Increased fat found behind the neck
- Easy bruising
- Fatigue
- Muscle weakness
- High blood pressure
- Low blood potassium level
- High blood sugar

Cushing syndrome

Symptoms of Cushing syndrome (hypercortisolism) may cause more than minor discomfort. Speak with your care team about your symptoms. There are treatments available.



- Depression
- Irregular menstrual cycles
- Sexual dysfunction
- Osteoporosis

Testing

If your tumor is bigger than 4 centimeters (cm), has irregular features on CT and/or MRI, or cancer is suspected, you might have the following tests for clarification:

- FDG-PET/CT
- Chest CT with or without contrast
- Abdominal/pelvic CT or MRI with contrast
- Additional blood work

Treatment

Treatment for hypercortisolemia caused by an adrenal tumor is based on tumor size. Tumors are usually measured in cm. A golf ball is about 4 cm and a large pea is about 1 cm.

If your tumor is less than 4 cm and is suspected to be benign, you will receive an adrenalectomy (minimally invasive is preferred).

An adrenalectomy refers to surgery where one or both adrenal glands are removed. During the procedure, your surgeon will remove the tumor and any areas where the cancer has spread, such as lymph nodes or nearby structures.

If your tumor is larger than 4 cm, has irregular features on CT and/or MRI, or cancer is suspected, your treatment will depend on how far the tumor has spread.

If the tumor is localized (found in one part of your body), an open adrenalectomy is preferred. However, if the tumor has spread to other sites of the body (liver, lungs, and bones), chemotherapy may be recommended instead of surgery. For more information on non-surgical options, *see Chapter 6: Adrenocortical carcinoma* for treatment options.

Outlook

Symptoms of hypercortisolism can be difficult to live with and may worsen without treatment. Some symptoms such as hair loss, excessive weight gain and abdominal stretch marks, and physical changes such as “moon face” or “buffalo hump” may make you feel embarrassed or stop you from participating in social events. Please note, symptoms can be treated with medication. Work with your health care team by reporting any new or worsening symptoms or any concerns.

Key points

- Hypercortisolism occurs when your body is exposed to too much cortisol. Cortisol is a hormone produced when you are stressed.
- Treatment is based on tumor size and additional testing.
- If your tumor is suspected to be benign (not cancer), adrenalectomy is recommended. If the tumor is small, a minimally invasive adrenalectomy is preferred.
- If your tumor is suspected or known to be cancerous (based on its size, or characteristics on CT or MRI), open adrenalectomy is preferred. However, if the tumor has spread to other sites of the body (liver, lungs, and bones), chemotherapy may be recommended instead of surgery.
- Symptoms of hypercortisolism can be difficult to live with and may worsen without treatment. Some symptoms such as hair loss, excessive weight gain and abdominal stretch marks, and physical changes such as “moon face” or “buffalo hump” may make you feel embarrassed or stop you from participating in social events.
- Symptoms can be cured. Work with your health care team by reporting any new or worsening symptoms or any concerns.

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

6
Adrenocortical carcinoma

Adrenocortical carcinoma (ACC) is a rare type of cancer that forms in the adrenal cortex (outer layer) of the adrenal glands.

Overview

Adrenocortical carcinoma (ACC) is a rare type of adrenal gland cancer that forms in the cortex (outer layer) of the adrenal glands.

There are 2 types of ACC:

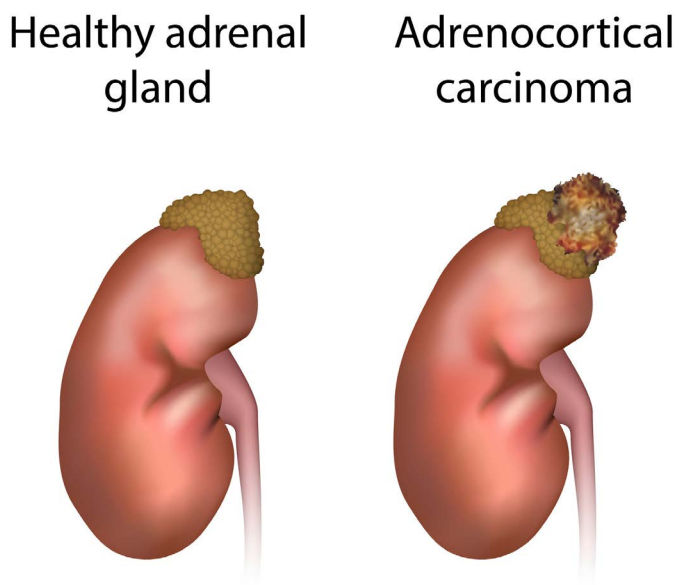
- **Functioning:** A functioning ACC tumor causes your adrenal gland to make more hormones than it normally should. Overproduction of cortisol and androgens are most common.
- **Nonfunctioning:** This type of tumor doesn't affect hormone production.

Symptoms of adrenal cancer can be related to hormone production and may include:

- Weight gain and increased abdominal fat
- Muscle weakness
- "Moon face" and "buffalo hump"
- Abdominal stretch marks (called striae)
- High blood pressure
- High blood sugar
- Trouble sleeping
- Deepening voice
- Increased hair growth (on the face)
- Change in pubic hair pattern
- Clitoral enlargement
- Pain in the abdomen or lower back
- Weight loss or loss of appetite

Adrenocortical carcinoma (ACC)

Adrenocortical carcinoma develops on the cortex (outer layer) of the adrenal glands.



Most ACC occurs randomly, not due to a genetic risk factor. In some cases, however, ACC has been linked to genetic conditions such as:

- Beckwith-Wiedemann syndrome
- Familial adenomatous polyposis (FAP)
- Li-Fraumeni syndrome
- Lynch syndrome
- Multiple endocrine neoplasia type 1 (MEN1)

Testing

You may receive the following tests to determine if you have ACC:

- FDG-PET/CT
- Chest CT with or without contrast
- Abdominal/pelvic CT or MRI with contrast
- Biochemical tests

Additional testing may include:

- Genetic counseling and testing for inherited genetic syndromes
- Microsatellite instability-high (MSI-H), mismatch repair deficient (dMMR), or tumor mutational burden-high (TMB-H) tumor testing.

Treatment

Primary treatment for ACC depends on whether the tumor can be removed with surgery (resectable) or cannot be removed with surgery (unresectable). If the tumor is localized, resection is usually possible. If the tumor has spread to other organs, chemotherapy is generally recommended instead of surgery.

If you want and are able to have surgery, an open adrenalectomy is recommended. After surgery, you might have external beam radiation therapy (EBRT). After all treatment is completed, you will be monitored for any

Guide 2
Adrenocortical carcinoma: Surveillance tests as needed

Tumor type	Timeframe and tests
Localized disease	Every 12 weeks to 1 year (up to 5 years): • Chest CT with or without contrast and abdominal CT or MRI with contrast • Biochemical tests
Unresectable or metastatic disease	Every 12 weeks to 1 year (up to 5 years): • Chest CT with or without contrast • Abdominal/pelvic CT or MRI with contrast or FDG-PET/CT

changes. For a list of follow-up tests, see Guide 2.

If you have unresectable or metastatic disease, you may be treated with one or more of the following:

- Observation, in some cases
- Resection of primary tumor and metastases, in some cases
- Local therapy (SBRT, thermal ablative therapies, liver-directed therapy)
- Systemic therapy (chemotherapy with or without mitotane)
- Enrollment in a clinical trial

Surveillance

Surveillance tests are done at specific times after treatment to check if the cancer has returned. These tests can help find cancer early so you can start treatment again, if needed. Surveillance tests may include general health tests such as a medical history and a physical exam, and blood, urine, and imaging tests. For a list of recommended testing, see Guide 2.

Key points

- Adrenocortical carcinoma (ACC) is a rare type of cancer that forms in the cortex (outer layer) of the adrenal glands.
- Most ACC occurs randomly (not due to a genetic risk factor), but genetic testing is often offered to those with ACC.
- Testing will help plan treatment.
- Primary treatment for ACC depends on whether the tumor can be removed with surgery (resectable) or cannot be removed with surgery (unresectable).
- Surveillance tests are done at specific times after treatment to check if the cancer has returned. These tests can help find cancer early so you can start treatment again, if needed.

7
Pheochromocytoma and paraganglioma

Paraganglioma is a tumor that arises from neuroendocrine tissues found along the spine. Pheochromocytoma is a type of paraganglioma that forms in the center of the adrenal gland causing the adrenal gland to make too much adrenaline. Paragangliomas and pheochromocytomas can occur in individuals with other inherited disorders, such as von Hippel-Lindau syndrome.

Overview

Paraganglioma

Paraganglioma is a rare type of tumor that forms near blood vessels and nerves outside of the adrenal glands that can cause symptoms similar to a pheochromocytoma. The nerve cells affected are part of the peripheral nervous system (outside of the brain and spinal cord). These tumors are also referred to as extra-adrenal pheochromocytomas.

Paragangliomas can be found anywhere along the spine from the base of the skull to the pelvis. However, this chapter will only discuss paragangliomas found near/outside the adrenal gland.

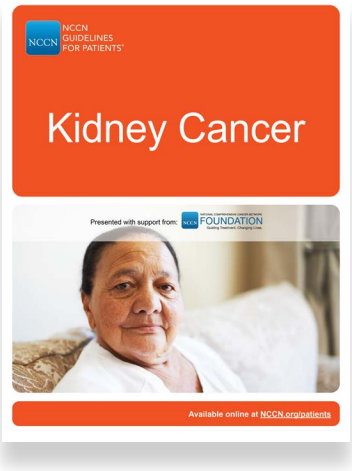
Pheochromocytoma

Pheochromocytoma is a rare, usually benign (not cancer) tumor that develops in the center of the adrenal gland. In pheochromocytoma, the tumor grows from chromaffin cells. Chromaffin cells produce hormones including adrenaline (epinephrine) and noradrenaline (norepinephrine).

Adrenaline and noradrenaline hormones trigger your body's fight-or-flight response to threat. Your body's response includes an increase in blood pressure and a rapid heartbeat. The hormones prepare your body to react quickly to a threat. Pheochromocytoma causes more of these hormones to be released, even when you are not in a threatening situation.

A pheochromocytoma (or "pheo" for short) can cause high blood pressure, pounding headaches, irregular heartbeat, flushing of the face, sweating, nausea, and vomiting.

For information on kidney-related paragangliomas and pheochromocytomas, see *NCCN Guidelines for Patients: Kidney Cancer,* available at NCCN.org/patientguidelines.



Symptoms

Some people will develop symptoms, while others will not. The most common symptom is high blood pressure (hypertension). A common feature with these types of adrenal tumors is a frequent but sporadic (irregular) attack of symptoms. These intense symptoms are referred to as “paroxysmal attacks.”

These attacks may worsen as the tumor grows. Some people have attacks because of physical or emotional stress. Others are woken up from sleep by symptoms.

Symptoms may include:

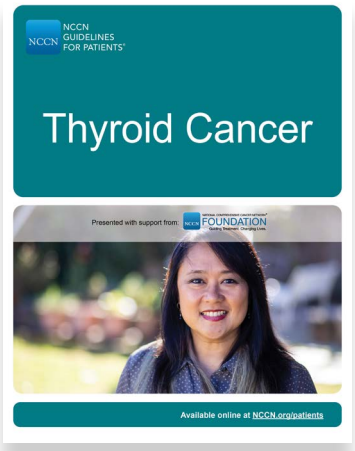
- High blood pressure
- Headaches
- Rapid heartbeat
- Sweating

Risk factors

There are some rare hereditary conditions that increase the risk for pheochromocytoma or paraganglioma.

Some of these inherited conditions include:

- **Multiple endocrine neoplasia type 2 (MEN2)** – refers to a genetic condition that increases your risk of developing tumors in the endocrine system (particularly in the thyroid, parathyroid, and adrenal glands). For treatment of medullary thyroid cancer and parathyroid tumors, see the *NCCN Guidelines for Patients: Thyroid Cancer*, available at NCCN.org/patientguidelines.
- **Von Hippel-Lindau disease (VHL)** – this condition produces tumors at multiple sites, including the central nervous system, endocrine system, pancreas, and kidneys.
- **Neurofibromatosis 1 (NF1)** – refers to multiple tumors in the skin (neurofibromas), pigmented skin spots, and tumors of the optic nerve.
- **Hereditary paraganglioma-pheochromocytoma (PGL/PCC) syndrome** – refers to inherited disorders that cause pheochromocytomas or paragangliomas.
- **Carney-Stratakis dyad** – refers to a rare, inherited condition that causes tumors in the GI tract or nervous tissue in the head, neck, and torso.
- **Carney triad** – refers to a rare condition that causes 3 kinds of tumors: paraganglioma, gastrointestinal stromal tumors (GIST), and pulmonary chondroma.



Testing

The following blood, urine, and imaging tests are recommended to determine if you have pheochromocytoma or paraganglioma:

- Plasma free or 24-hour urine fractionated metanephrines and normetanephrines with or without serum or 24-hour urine catecholamines
- Adrenal protocol CT (abdomen/pelvis)
- Genetic counseling and testing for inherited genetic conditions

Additional testing may include:

- Abdominal/pelvic multiphasic CT or MRI
- SSTR-PET/CT or SSTR-PET/MRI
- FDG-PET/CT (skull base to mid-thigh)
- Chest CT with or without contrast
- MIBG (iodine-123 meta-iodobenzylguanidine) scan

Treatment

Before treatment, you will likely be prescribed specific types of pills to lower your blood pressure and heart rate. These typically include alpha- and beta-blockers. You will also be asked to take a high salt diet and drink a lot of fluids for a few days before procedures such as surgery.

One or more of the following might be added:

- Dihydropyridine calcium channel blockade
- Metyrosine

If your tumor is resectable, it means it can be removed with surgery. Minimally invasive surgery is recommended whenever possible, and it is safe and feasible.

If your tumor is unresectable, asymptomatic, or slow-growing**,** your treatment will need to be individualized to help control tumor growth and manage any symptoms you are experiencing.

- If your tumor is secreting, a clinical trial is preferred. Other treatments might include systemic therapy, radiation therapy, or radioactive drug therapy.

If you have distant metastases, your treatment will need to be individualized to help slow tumor growth and manage any symptoms you are experiencing.

- If your tumor is secreting, a clinical trial is preferred. Other treatments might include systemic therapy, surgery to remove metastases, radiation therapy, or radioactive drug therapy

Surveillance

Surveillance tests are done at specific times after treatment to check if the cancer has returned. These tests can help find cancer early so you can start treatment again, if needed. Surveillance tests may include general health tests such as a medical history and a physical exam, and imaging tests. For a list of recommended testing, see Guide 3.

Guide 3
Pheochromocytoma and paraganglioma: Surveillance tests

Resectable disease (after surgery)	**Every 12 weeks to 12 months** • Physical exam, blood pressure, and biochemical tests • Possible chest CT with or without contrast and abdominal/pelvic CT or MRI with contrast
	Over 1 year up to 10 years • Physical exam, blood pressure, and biomarkers • Possible chest CT with or without contrast and abdominal/pelvic CT or MRI with contrast
	Over 10 years • Surveillance as needed
Unresectable disease or distant metatases	**Every 12 weeks to 12 months** • Physical exam, blood pressure, and biochemical tests • Chest/abdominal/pelvic CT with contrast • Chest CT with or without contrast and abdominal/pelvic MRI without contrast • MIBG (iodine-123 meta-iodobenzylguanidine) scan with SPECT/CT prior to considering radionuclide therapy • FDG-PET/CT for bone-dominant disease • SSTR-PET/CT or SSTR-PET/MRI prior to considering radionuclide therapy

Key points

- Pheochromocytoma is a rare, usually benign tumor that develops in the center of the adrenal gland causing the adrenal gland to make too much adrenaline.
- Paraganglioma is a rare type of tumor that forms near blood vessels and nerves outside of the adrenal gland.
- Some people will develop symptoms, while others will not. The most common symptom is hypertension (high blood pressure).
- You may be at risk for pheochromocytoma or paraganglioma if you have a rare inherited condition. Tumors associated with these disorders are more likely to be malignant (cancerous).
- You will have blood, urine, and imaging tests to diagnose a pheochromocytoma or paraganglioma.
- Before treatment, you will likely be prescribed specific types of pills to lower your blood pressure and heart rate. These typically include alpha- and beta-blockers. You will also be asked to take a high salt diet and drink a lot of fluids for a few days before procedures such as surgery.
- Surveillance tests are done at specific times after treatment to check if the cancer has returned. These tests can help find cancer early so you can start treatment again, if needed.

8
Making treatment decisions

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

It's your choice

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

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

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

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

Think about what you want from treatment. Discuss openly the risks and benefits of specific treatments and procedures. Weigh options and share concerns with your doctor. If you take the time to build a relationship with your doctor, it will help you feel supported when considering options and making treatment decisions.

Second opinion

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

Things you can do to prepare:

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

Support groups

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

Questions to ask your doctors

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

Questions to ask about diagnosis and testing

1. What type of adrenal gland tumor do I have?
2. Is the tumor cancerous?
3. Is the tumor functional or nonfunctional? What does this mean?
4. Where is the tumor located? Is the tumor located inside and/or outside of the adrenal gland?
5. Is there a cancer center or hospital nearby that specializes in adrenal tumors?
6. What tests are needed? What other tests do you recommend?
7. How do I prepare for testing?
8. How soon will I know the results and who will explain them to me?
9. Would you give me a copy of the pathology report and other test results?
10. Will you explain my pathology report (laboratory test results) to me?
11. Who will talk with me about the next steps? When?

Questions to ask about treatment options

1. Will my age, health, and other factors affect my treatment options?
2. Is there a better treatment option based on my age and other risk factors?
3. What is the goal of treatment?
4. What are the possible side effects of each treatment?
5. Is there an option that is less expensive?
6. Who can help me if I am concerned about managing the costs of care?
7. Can I stop treatment at any time?
8. What will happen if I stop treatment?
9. What support services are available to me?
10. Could the treatment affect my fertility? If so, should I speak to a fertility specialist before treatment?
11. How often will I need follow-up visits after I finish treatment?
12. Whom should I call with questions or concerns?

Questions to ask about surgery

1. Do I need to have surgery? If yes, what type do you recommend?
2. Will the entire adrenal gland be removed?
3. How long will the procedure take?
4. How long will I be in the hospital?
5. How long will it take me to recover?
6. How much pain will I be in? What will be done to manage my pain?
7. What other side effects can I expect?
8. What are the possible long-term effects of having this surgery?

Questions to ask about clinical trials

1. What clinical trials are available for me?
2. Has the treatment been used before?
3. What are the risks and benefits of this treatment?
4. What side effects should I expect? How will the side effects be controlled?
5. How long will I be in the clinical trial?
6. Will I be able to get other treatment if this doesn't work?
7. How will I know if the treatment is working?
8. Will the clinical trial cost me anything? If so, how much?
9. What type of long-term follow-up care will I have?

Questions to ask about follow-up care

1. What is the chance that the tumor will come back? What are potential signs and symptoms?
2. What long-term side effects or late effects are possible based on the treatment I received?
3. What follow-up tests will I need, and how often will I need them?
4. How do I get a treatment summary and follow-up care plan to keep in my personal records?
5. Who will be leading my follow-up care?
6. What support services are available to me after treatment? To my family?

Questions to ask your doctors about their experience

1. Who will be part of my health care team, and what does each member do?
2. Who will be leading my overall treatment?
3. What is your experience in treating people with adrenal tumors?
4. Who else will be on my treatment team?
5. What other diagnostic tests or procedures will I need?
6. I would like to get a second opinion. Is there someone you recommend?
7. How many patients like me (of the same age, gender, race) have you treated?
8. Will you be consulting with experts to discuss my care? Whom will you consult?
9. How many procedures like the one you're suggesting have you done?
10. Is this treatment a major part of your practice?
11. How many of your patients have had complications? What were the complications?
12. Who will manage my day-to-day care?

Resources

American Cancer Society
cancer.org/cancer/adrenal-cancer.html

American Society of Clinical Oncology (ASCO)
cancer.net

Carcinoid Cancer Foundation
carcinoid.org

Chemocare
chemocare.com

Endocrine Society
endocrine.org/patient-engagement/endocrine-library

MedlinePlus
medlineplus.gov

National Cancer Institute
cancer.gov/types/adrenocortical/hp/adrenocortical-treatment-pdq

National Coalition for Cancer Survivorship
canceradvocacy.org

National Financial Resource Directory
- Patient Advocate Foundation
patientadvocate.org/explore-our-resources/national-financial-resource-directory

National Hospice and Palliative Care Organization
nhpco.org

OncoLink
oncolink.org

Patient Access Network Foundation
panfoundation.org

Radiological Society of North America
radiologyinfo.org

Testing.com
testing.com

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

NCCN.org/patients/comments

Words to know

adrenal cortex
The outer layer of the adrenal gland that makes androgen and corticosteroid hormones.

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 cancer that forms in the adrenal cortex (outer layer) of the adrenal glands.

biochemical test
A test to measure the level of chemicals in the body.

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

bronchopulmonary
A term used for the airways that lead to the lungs (bronchi) and the lungs (pulmonary).

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

bronchus
One of the airways that extends into the lungs. The plural form is bronchi.

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

cancer stage
A rating of the outlook for people with cancer based on the cancer's growth and spread.

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

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

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

colon
The hollow organ in which eaten food turns from a liquid into a solid form.

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

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

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, a hormone, in the body. Also called hypercortisolism.

deoxyribonucleic acid (DNA)
A chain of chemicals in cells that contains coded instructions for making and controlling cells. Also called the "blueprint of life."

duodenum
A part of the digestive tract that receives food from the stomach and mixes it with digestive juices.

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

external beam radiation therapy (EBRT)
A cancer treatment with radiation delivered from a machine outside the body.

first-line therapy
The first drug or set of drugs given to treat cancer.

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

genetic assessment
A lab test of abnormal coded instructions in cells that are passed down within a family.

hereditary
Passed down from 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, a hormone, in the body. Also called Cushing syndrome.

hypertension
A condition in which the force of the blood against the artery walls is too high. Also referred to as high blood pressure.

hypothalamus
A part of the brain that works with the nervous system and glands that make hormones in the body.

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

immune system
The body's natural defense against infection and disease.

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

liver
The largest organ and gland in the body with many vital functions.

lung
One of a pair of organs that consists of airways and air sacs.

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.

medical history
A report of all your health events and medications.

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

multiple endocrine neoplasia (MEN)
An inherited syndrome that causes tumors to grow in the glands of the endocrine system.

mutation
An abnormal change in cells.

neoadjuvant treatment
A treatment given before the main treatment to reduce the cancer. Also called preoperative treatment if given before an operation.

neuroendocrine cells
A cell that receives signals from the nervous system to make and release hormones into the blood.

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

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

pancreas
An organ that makes fluids that help digest food and hormones that control blood sugar.

paraganglioma
Tumor that arises from neuroendocrine tissues found along the spine.

pelvis
The body area between the hipbones.

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

pheochromocytoma
Tumor that forms in the center of the adrenal gland that causes it to make too much adrenaline.

pituitary gland
An organ in the brain that controls certain body functions and other hormone glands. Also called the “master gland.”

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.

primary treatment
The main treatment used to rid the body of cancer.

primary tumor
The first mass of cancer cells.

progression
The growth or spread of cancer after being tested or treated.

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

radiopharmaceutical
A drug that carries a certain amount of radioactive material.

serotonin
A hormone that sends signals between nerve cells and controls things like mood, sleep, and memory.

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

somatostatin receptor scintigraphy
A type of imaging scan used to assess carcinoid tumors that have somatostatin receptors.

sporadic
Occurring at irregular intervals or only in a few places.

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

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

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

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

thymus
A gland that is behind the breastbone.

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

ulcer
A sore on the skin or mucous membrane in the body.

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

x-ray
A test that uses small amounts of radiation to make pictures of the insides of the body. Also called a plain radiograph.

NCCN Contributors

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800.789.7366 • pennmedicine.org/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/services/cancer-services
866.223.8100 • CC Taussig Cancer Institute
my.clevelandclinic.org/departments/cancer
216.844.8797 • Case CCC
case.edu/cancer

City of Hope National Medical Center
Los Angeles, California
800.826.4673 • cityofhope.org

Dana-Farber/Brigham and Women's
Cancer Center | Massachusetts General
Hospital Cancer Center
Boston, Massachusetts
617.732.5500 • youhaveus.org
617.726.5130
massgeneral.org/cancer-center

Duke Cancer Institute
Durham, North Carolina
888.275.3853 • dukecancerinstitute.org

Fox Chase Cancer Center
Philadelphia, Pennsylvania
888.369.2427 • foxchase.org

Fred & Pamela Buffett Cancer Center
Omaha, Nebraska
402.559.5600 • unmc.edu/cancercenter

Fred Hutchinson Cancer
Research Center/Seattle
Cancer Care Alliance
Seattle, Washington
206.606.7222 • seattlecca.org
206.667.5000 • fredhutch.org

Huntsman Cancer Institute
at the University of Utah
Salt Lake City, Utah
800.824.2073 • huntsmancancer.org

Indiana University
Melvin and Bren Simon
Comprehensive Cancer Center
Indianapolis, Indiana
888.600.4822 • www.cancer.iu.edu

Mayo Clinic Cancer Center
Phoenix/Scottsdale, Arizona
Jacksonville, Florida
Rochester, Minnesota
480.301.8000 • Arizona
904.953.0853 • Florida
507.538.3270 • Minnesota
mayoclinic.org/cancercenter

Memorial Sloan Kettering
Cancer Center
New York, New York
800.525.2225 • mskcc.org

Moffitt Cancer Center
Tampa, Florida
888.663.3488 • moffitt.org

O'Neal Comprehensive
Cancer Center at UAB
Birmingham, Alabama
800.822.0933 • uab.edu/onealcancercenter

Robert H. Lurie Comprehensive Cancer
Center of Northwestern University
Chicago, Illinois
866.587.4322 • cancer.northwestern.edu

Roswell Park Comprehensive
Cancer Center
Buffalo, New York
877.275.7724 • roswellpark.org

Siteman Cancer Center at Barnes-
Jewish Hospital and Washington
University School of Medicine
St. Louis, Missouri
800.600.3606 • siteman.wustl.edu

St. Jude Children's
Research Hospital/
The University of Tennessee
Health Science Center
Memphis, Tennessee
866.278.5833 • stjude.org
901.448.5500 • uthsc.edu

Stanford Cancer Institute
Stanford, California
877.668.7535 • cancer.stanford.edu

The Ohio State University
Comprehensive Cancer Center -
James Cancer Hospital and
Solove Research Institute
Columbus, Ohio
800.293.5066 • cancer.osu.edu

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410.955.8964
www.hopkinskimmelcancercenter.org

The University of Texas
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Ann Arbor, Michigan
800.865.1125 • rogelcancercenter.org

University of Wisconsin
Carbone Cancer Center
Madison, Wisconsin
608.265.1700 • uwhealth.org/cancer

UT Southwestern Simmons
Comprehensive Cancer Center
Dallas, Texas
214.648.3111 • utsouthwestern.edu/simmons

Vanderbilt-Ingram Cancer Center
Nashville, Tennessee
877.936.8422 • vicc.org

Yale Cancer Center/
Smilow Cancer Hospital
New Haven, Connecticut
855.4.SMILOW • yalecancercenter.org

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