CONDITION

Adrenocortical carcinoma

Learn about adrenocortical carcinoma, including hormone-related symptoms, diagnosis, surgery, medicines, possible complications, and follow-up care.

WHAT IT IS
Cancer of the adrenal cortex

BODY SYSTEM
Hormone system

MAIN SYMPTOMS
Weight gain, high blood pressure, hormone-related changes, abdominal or back pain

WHAT CAUSES IT
Usually unknown, sometimes inherited

HOW IT IS DIAGNOSED
Examination of surgical or biopsy tissue

MAIN TREATMENT
Surgery for removable cancer

CAN IT BE CURED?
Sometimes - recurrence possible after complete surgery

IS IT AN EMERGENCY?
Sometimes - repeated vomiting, confusion or collapse

LONG-TERM OUTLOOK
Recurrence or spread possible after complete surgery

Overview

Adrenocortical carcinoma, or ACC, is a cancer that begins in the adrenal cortex, the outer layer of an adrenal gland. The two adrenal glands sit above the kidneys and make hormones that help regulate blood pressure, metabolism, stress responses, and sexual development.

ACC is different from an adrenal adenoma, which is noncancerous. Most adrenal growths found on imaging are not cancer. Doctors assess a growth's size, appearance, hormone activity, and changes over time. A definite diagnosis often depends on examining tissue removed during surgery.

ACC may produce excessive adrenal hormones, invade nearby structures, or spread to areas such as the lungs, liver, lymph nodes, or bones. It may be found after hormone-related symptoms, abdominal discomfort, or an unrelated scan.

Care is often coordinated by specialists experienced in adrenal tumors, including endocrinologists, surgeons, oncologists, radiologists, pathologists, and genetic counselors.

Symptoms

Symptoms depend on whether the tumor produces hormones, presses on nearby organs, or has spread. Some people have few symptoms initially.

Excess cortisol can cause Cushing syndrome. Possible effects include:

Tumors that produce androgens may cause increased facial or body hair, acne, scalp hair loss, a deeper voice, or irregular periods in women. Children may develop pubic hair or other signs of early sexual development.

Estrogen-producing tumors can cause breast enlargement or tenderness in men and abnormal uterine bleeding in women. Excess aldosterone may cause high blood pressure, weakness, or muscle cramps due to low potassium.

A growing mass may cause persistent pain or fullness in the abdomen, side, or back. Other possible symptoms include reduced appetite, unintended weight loss, nausea, or an abdominal lump.

These symptoms can have many causes and do not confirm ACC. Discuss new or persistent symptoms with a healthcare professional.

Seek emergency care for severe chest pain, sudden breathing difficulty, fainting, confusion, or uncontrolled bleeding. People with adrenal insufficiency also need emergency help for severe vomiting, profound weakness, severe abdominal pain, confusion, or collapse, particularly during illness or after injury. These may indicate an adrenal crisis caused by dangerously low cortisol.

- Weight gain, especially around the abdomen or face
- Weakness in the hips or shoulders
- Easy bruising, wide purple stretch marks, or slow wound healing
- High blood pressure or blood sugar
- Bone loss, mood changes, menstrual changes, infections, or blood clots

Causes and risk factors

The exact cause of most ACC is unknown. The cancer develops when cells in the adrenal

cortex acquire changes that disrupt normal control of cell growth.

A small proportion of cases are associated with inherited cancer-predisposition syndromes, including Li-Fraumeni syndrome, Lynch syndrome, multiple endocrine neoplasia type 1, familial adenomatous polyposis, and Beckwith-Wiedemann syndrome. Having one of these syndromes does not mean someone will develop ACC, and most people with ACC have no known inherited syndrome.

Genetic counseling may be appropriate when ACC occurs at a young age or personal and family histories suggest an inherited condition. A genetic counselor can explain how testing may affect care and whether relatives should consider evaluation.

No lifestyle change is known to prevent most cases. People should not assume the cancer resulted from something they did or did not do.

Diagnosis

Evaluation usually includes a medical history and physical examination. The clinician may ask about weight, blood pressure, muscle strength, menstrual periods, hair growth, sexual development, medicines, and family history.

Blood and urine tests can measure cortisol, adrenal androgens, and other steroid hormones. Aldosterone and renin may be checked in people with high blood pressure or low potassium.

Doctors may also test for excess catecholamines to rule out pheochromocytoma, a different adrenal tumor that requires special precautions before surgery or biopsy.

CT scans provide detailed images of the adrenal gland and nearby organs. MRI may further assess blood vessels, the liver, or surrounding tissues. Imaging of the chest, abdomen, and pelvis helps determine whether cancer has spread. PET scanning may be useful in selected cases.

Needle biopsy is not routine for every suspicious adrenal mass. It may not reliably distinguish ACC from a noncancerous adrenal cortical tumor and can complicate later surgery. If a tumor appears removable, surgery may be recommended without biopsy. Biopsy may be considered when cancer has spread and the result would change treatment.

Pheochromocytoma must be excluded before adrenal biopsy because disturbing one can trigger a dangerous hormone surge.

A pathologist examines tissue obtained through surgery or biopsy. The report may describe invasion, cell division, dead tumor tissue, and whether cancer reaches the surgical edges. Expert review can be helpful because adrenal tumors are difficult to classify.

Staging considers whether cancer is confined to the adrenal gland, has invaded nearby structures or lymph nodes, or has spread to distant organs. Stage, hormone production, completeness of surgery, and microscopic features guide treatment and follow-up.

Treatment and management

Treatment should be individualized by a team experienced in ACC. Decisions depend on stage, whether the cancer can be removed completely, hormone production, previous treatment, overall health, and personal goals.

Surgery is the main treatment for removable ACC. The operation generally removes the affected adrenal gland and surrounding fatty tissue. Nearby organs or lymph nodes may also be removed if involved. Surgeons aim to remove the tumor in one piece without breaking its capsule because spillage may increase recurrence risk.

An open operation is often used when ACC is strongly suspected, especially for large tumors or those involving nearby structures. Minimally invasive surgery may be considered in carefully selected cases by an experienced surgeon.

Mitotane may be offered after surgery when clinical or pathology findings indicate a meaningful recurrence risk. It is not appropriate for everyone. Mitotane damages adrenal cortical cells and changes how the body processes hormones and other medicines. Monitoring includes drug levels, hormone function, liver function, cholesterol, and side effects. Possible effects include nausea, diarrhea, poor appetite, tiredness, dizziness, confusion, and coordination problems.

Mitotane often causes adrenal insufficiency. People taking it commonly need prescribed steroid like prednisone or hydrocortisone replacement and clear instructions for illness, injury, or surgery. It also has important drug interactions, so the care team should review all prescription medicines, nonprescription products, and supplements.

For cancer that cannot be removed or has spread, treatment may include mitotane alone or chemotherapy. A commonly used combination is etoposide, doxorubicin, and cisplatin with mitotane. These medicines may slow or shrink cancer but can cause substantial side effects.

Radiation therapy may control cancer in a specific area, reduce pain, or address a high risk of local recurrence. Surgery, ablation, or other focused procedures may sometimes treat a limited number of metastases. Immunotherapy, targeted treatments, or newer combinations may be available in selected cases or clinical trials.

Hormone excess often requires separate treatment. Medicines may reduce cortisol production, block hormone effects, or manage blood pressure and potassium. Close

monitoring is necessary because hormone levels can change with cancer treatment.

Supportive care can address pain, nausea, nutrition, sleep, emotional distress, fertility concerns, and treatment's practical effects at any stage.

Complications

ACC may return near the original site or spread to other organs even after apparently complete surgery. Recurrence risk varies according to stage and pathology findings.

Hormone excess can cause complications independently of tumor growth. High cortisol may contribute to diabetes, high blood pressure, bone loss, infection, muscle weakness, mood changes, and blood clots. Excess aldosterone can cause low potassium and abnormal heart rhythms. Androgen or estrogen excess can affect fertility, sexual function, menstrual periods, and physical characteristics.

Treatment complications may include bleeding, infection, injury to nearby organs, or adrenal insufficiency after surgery. Depending on the medicines used, chemotherapy can affect blood counts, kidneys, nerves, hearing, or heart function. Radiation effects depend on the treated area.

Adrenal insufficiency can become life-threatening during severe illness, injury, or surgery. People at risk should understand their steroid replacement and emergency-dosing plans and carry medical identification if advised.

Follow up

Follow-up usually includes scheduled visits, imaging, and hormone tests to detect recurrence and manage treatment effects. Timing depends on the cancer's stage, treatment, hormone activity, and time since diagnosis.

If the original tumor produced hormones, the same markers may help monitor for recurrence. People taking mitotane need ongoing blood tests and medicine reviews. Monitoring may continue after treatment stops because mitotane can remain in the body for an extended period.

Contact the cancer team promptly about persistent abdominal or back pain, a growing lump, unexplained weight loss, new hormone-related changes, or worsening side effects. These require timely assessment but are not always emergencies.

Seek emergency care for severe breathing difficulty, chest pain, fainting, confusion, or possible

adrenal crisis symptoms such as repeated vomiting, severe weakness, very low blood pressure, or collapse.

Long-term care may address bone health, cardiovascular risk, fertility or sexual health, emotional well-being, and genetic counseling. Keep copies of pathology, operation, imaging, and treatment reports, especially when care involves several centers.

Sources

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