Adrenocortical carcinoma is a rare disease in which malignant (cancer) cells form in the outer layer of the adrenal gland.

There are two adrenal glands. The adrenal glands are small and shaped like a triangle. One adrenal gland sits on top of each kidney. Each adrenal gland has two parts. The outer layer of the adrenal gland is the adrenal cortex. The center of the adrenal gland is the adrenal medulla. The adrenal cortex makes important hormones that. :Balance the water and salt in the body. Help keep blood pressure normal. Help manage the body's use of protein, fat, and carbohydrates.

Cause the body to have masculine or feminine characteristics.

The adrenal medulla makes hormones that help the body react to stress.

Adrenocortical carcinoma is also called cancer of the adrenal cortex. A tumor of the adrenal cortex may be functioning (makes more hormones than normal) or nonfunctioning (does not make hormones). The hormones made by functioning tumors may cause certain signs or symptoms of disease.

Cancer that forms in the adrenal medulla is called pheochromocytoma. See the PDQ summary on Pheochromocytoma Treatment for more information.

Having certain genetic conditions increases the risk of developing adrenocortical carcinoma. Anything that increases your risk of getting a disease is called a risk factor. Having a risk factor does not mean that you will get cancer; not having risk factors doesn't mean that you will not get cancer. People who think they may be at risk should discuss this with their doctor.

Risk factors for adrenocortical carcinoma include having the following hereditary diseases:

Li-Fraumeni syndrome.
Beckwith-Wiedemann syndrome.
Carney complex.
Possible signs of adrenocortical carcinoma include pain in the abdomen and certain physical changes.

These and other symptoms may be caused by adrenocortical carcinoma:

A lump in the abdomen.
Pain the abdomen or back.
A nonfunctioning adrenocortical tumor may not cause symptoms in the early stages. A functioning adrenocortical tumor makes too much of a certain hormone (cortisol, aldosterone, testosterone, or estrogen). Having too much of a certain hormone may cause the following symptoms:

Too much cortisol

Weight gain in the face, neck, and trunk of the body and thin arms and legs.
Growth of fine hair on the face, upper back, or arms.
A round, red, full face.
A lump of fat on the back of the neck.
A deepening of the voice and swelling of the sex organs or breasts in both males and females.
Muscle weakness.
High blood sugar.
High blood pressure.

Too much aldosterone

High blood pressure.
Muscle weakness or cramps.
Frequent urination.
Feeling thirsty.

Too much testosterone (in women)

Growth of fine hair on the face, upper back, or arms.
Acne.
Balding.
A deepening of the voice.
No menstrual periods.
Men who make too much testosterone do not usually have symptoms.

Too much estrogen (in women)

Irregular menstrual periods in women who have not gone through menopause.
Menstrual bleeding in women who have gone through menopause.

Too much estrogen (in men)

Growth of breast tissue.
Lower sex drive.
Impotence.

These and other symptoms may be caused by adrenocortical carcinoma. Other conditions may cause the same symptoms. A doctor should be consulted if any of these problems occur.

Imaging studies and tests that examine the blood and urine are used to detect (find) and diagnose adrenocortical carcinoma.

The tests and procedures used to diagnose adrenocortical carcinoma depend on the patient's symptoms. The following tests and procedures may be used:

Physical exam and history: An exam of the body to check general signs of health, including checking for signs of disease, such as lumps or anything else that seems unusual. A history of the patient's health habits and past illnesses and treatments will also be taken.

Twenty-four-hour urine test: A test in which urine is collected for 24 hours to measure the amounts of cortisol or 17-ketosteroids. A higher than normal amount of these in the urine may be a sign of disease in the adrenal cortex.

Low- dose dexamethasone suppression test: A test in which one or more small doses of dexamethasone is given. The level of cortisol is checked from a sample of blood or from urine that is collected for three days.

High-dose dexamethasone suppression test: A test in which one or more high doses of dexamethasone is given. The level of cortisol is checked from a sample of blood or from urine that is collected for three days.

Blood chemistry study: A procedure in which a blood sample is checked to measure the amounts of certain substances, such as potassium or sodium, released into the blood by organs and tissues in the body. An unusual (higher or lower than normal) amount of a substance can be a sign of disease.

Blood tests: Tests to measure the levels of testosterone or estrogen in the blood. **Higher than normal amounts of these hormones may be a sign of adrenocortical carcinoma.**

CT scan (CAT scan): A procedure that makes a series of detailed pictures of areas inside the body, taken from different angles. The pictures are made by a computer linked to an x-ray machine. A dye may be injected into a vein or swallowed to help the organs or tissues show up more clearly. This procedure is also called computed tomography, computerized tomography, or computerized axial tomography.

MRI (magnetic resonance imaging): A procedure that uses a magnet, radio waves, and a computer to make a series of detailed pictures of areas inside the body. This procedure is also called nuclear magnetic resonance imaging (NMRI). An MRI of the abdomen is done to diagnose adrenocortical carcinoma.

Adrenal angiography: A procedure to look at the arteries and the flow of blood near the adrenal gland. A contrast dye is injected into the adrenal arteries. As the dye moves through the blood vessel, a series of x-rays are taken to see if any arteries are blocked.

Adrenal venography: A procedure to look at the adrenal veins and the flow of blood near the adrenal gland. A contrast dye is injected into an adrenal vein. As the contrast dye moves through the vein, a series of x-rays are taken to see if any veins are blocked. A catheter (very thin tube) may be inserted into the vein to take a blood sample, which is checked for abnormal hormone levels.

PET scan (positron emission tomography scan): A procedure to find malignant tumor cells in the body. A small amount of radioactive glucose (sugar) is injected into a vein. The PET scanner rotates around the body and makes a picture of where glucose is being used in the body. Malignant tumor cells show up brighter in the picture because they are more active and take up more glucose than normal cells do.

Certain factors affect the prognosis (chance of recovery) and treatment options.

The prognosis (chance of recovery) and treatment options depend on the following:

The stage of the cancer; the size of the tumor and whether it is in the adrenal gland only or has spread to other places in the body- Whether the tumor can be completely removed in surgery; whether the cancer has been treated in the past and the patient's health.

Adrenocortical carcinoma may be cured if treated at an early stage.

After adrenocortical carcinoma has been diagnosed, tests are done to find out if cancer cells have spread within the adrenal gland or to other parts of the body.

The process used to find out if cancer has spread within the adrenal gland or to other parts of the body is called staging. The information gathered from the staging process determines the stage of the disease. It is important to know the stage in order to plan treatment. The following tests and procedures may be used in the staging process:

CT scan (CAT scan): A procedure that makes a series of detailed pictures of areas inside the body, such as the abdomen or chest, taken from different angles. The pictures are made by a computer linked to an x-ray machine. A dye may be injected into a vein or swallowed to help the organs or tissues show up more clearly. This procedure is also called computed tomography, computerized tomography, or computerized axial tomography.

MRI (magnetic resonance imaging) with gadolinium: A procedure that uses a magnet, radio waves, and a computer to make a series of detailed pictures of areas inside the body. A substance called gadolinium may be injected into a vein. The gadolinium collects around the cancer cells so they show up brighter in the picture. This procedure is also called nuclear magnetic resonance imaging (NMRI).

Adrenal angiography: A procedure to look at the arteries and the flow of blood near the adrenal gland. A contrast dye is injected into the adrenal arteries. As the dye moves through the blood vessel, a series of x-rays are taken to see if any arteries are blocked.

Adrenal venography: A procedure to look at the adrenal veins and the flow of blood near the adrenal gland. A contrast dye is injected into an adrenal vein. As the contrast dye moves through the vein, x-rays are taken to see if any veins are blocked. A catheter (very thin tube) may be inserted into the vein to take a blood sample, which is checked for abnormal hormone levels.

Cavagram: A procedure to look at the inferior vena cava and the flow of blood through the inferior vena cava. A contrast dye is injected into a blood vessel. As the contrast dye moves through the blood vessel to the inferior vena cava, a series of x-rays are taken to see if there are any changes to the inferior vena cava and the flow of blood through the inferior vena cava.

Ultrasound exam: A procedure in which high-energy sound waves (ultrasound) are bounced off internal tissues or organs, such as the vena cava, and make echoes. The echoes form a picture of body tissues called a sonogram.

Adrenalectomy: A procedure to remove the entire adrenal gland. A tissue sample is viewed under a microscope by a pathologist to check for signs of cancer.

The following stages are used for adrenocortical carcinoma:

Stage I

In stage I, the tumor is 5 centimeters or smaller and is found only in the adrenal gland.

Stage II

In stage II, the tumor is larger than 5 centimeters and is found only in the adrenal gland.

Stage III

In stage III, the tumor can be any size and may have spread to fat or lymph nodes near the adrenal gland.

Stage IV

In stage IV, the tumor can be any size and has spread to fat or organs and to lymph nodes near the adrenal gland; or to other parts of the body. Adrenocortical carcinoma commonly spreads to the lung, liver, bones, and peritoneum (the tissue that lines the abdominal wall and covers most of the organs in the abdomen).

Recurrent Adrenocortical Carcinoma

Recurrent adrenocortical carcinoma is cancer that has recurred (come back) after it has been treated. The cancer may come back in the adrenal cortex or in other parts of the body.

There are different types of treatment for patients with adrenocortical carcinoma.

Different types of treatments are available for patients with adrenocortical carcinoma. Some treatments are standard (the currently used treatment), and some are being tested in clinical trials. Before starting treatment, patients may want to think about taking part in a clinical trial. A treatment clinical trial is a research study meant to help improve current treatments or obtain information on new treatments for patients with cancer. When clinical trials show that a new treatment is better than the standard treatment, the new treatment may become the standard treatment.

Clinical trials are taking place in many parts of the country. Information about ongoing clinical trials is available from the NCI Web site. Choosing the most appropriate cancer treatment is a decision that ideally involves the patient, family, and health care team.

Three types of standard treatment are used:

Surgery

Surgery to remove the adrenal gland (adrenalectomy) is often used to treat adrenocortical carcinoma. Sometimes the nearby lymph nodes are also removed.

Radiation therapy

Radiation therapy is a cancer treatment that uses high-energy x-rays or other types of radiation to kill cancer cells or keep them from growing. There are two types of radiation therapy. External radiation therapy uses a machine outside the body to send radiation toward the cancer. Internal radiation therapy uses a radioactive substance sealed in needles, seeds, wires, or catheters that are placed directly into or near the cancer. The way the radiation therapy is given depends on the type and stage of the cancer being treated.

Chemotherapy

Chemotherapy is a cancer treatment that uses drugs to stop the growth of cancer cells, either by killing the cells or by stopping them from dividing. When chemotherapy is taken by mouth or

injected into a vein or muscle, the drugs enter the bloodstream and can reach cancer cells throughout the body (systemic chemotherapy). When chemotherapy is placed directly into the spinal column, an organ, or a body cavity such as the abdomen, the drugs mainly affect cancer cells in those areas (regional chemotherapy). The way the chemotherapy is given depends on the type and stage of the cancer being treated.

Mitotane may be used to treat adrenocortical carcinoma. Mitotane stops the adrenal cortex from making hormones and relieves symptoms caused by the hormones.

Treatment Options for Recurrent Adrenocortical Carcinoma

Treatment of recurrent adrenocortical carcinoma may include the following as palliative therapy to relieve symptoms and improve the quality of life:

Surgery to remove the tumor, surgery to remove the cancer from places where it has spread, a clinical trial of chemotherapy or biologic therapy.