



ADRENOCORTICAL CANCER
SUPPORT AND INFORMATION



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ADRENOCORTICAL CARCINOMA – ACC

PATIENT INFORMATION
FOR ADULT PATIENTS

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INTRODUCTION

“It is never easy to accept that you have cancer. A rare one, I think, makes it even more difficult”

ACC patient

Being diagnosed with any type of cancer can be scary, but when you have a rare cancer like ACC you can also feel alone and frustrated. It might be hard to find a medical specialist who can help or anyone else who has been through the same as you. You might not be able to find information to help inform you of what to do or where to go. This booklet has been written to help answer some of the questions that you may have about your cancer.

If you need further information or support please do not hesitate to call or email ACC Support UK (www.accsupport.org.uk). Further details of this and other useful organisations can be found at the end of this booklet.

WHAT IS CANCER?

The body is made up of hundreds of millions of living cells. Normal body cells grow, divide, and die in an orderly way. During the early years of a person's life, normal cells divide faster to allow the person to grow. After the person becomes an adult, most cells divide only to replace worn out, damaged, or dying cells.

Cancer begins when cells in a part of the body start to grow out of control. There are many kinds of cancer, but they all start because of this out-of-control growth of abnormal cells. Cancer cell growth is different from normal cell growth. Instead of dying, cancer cells keep on growing and form new cancer cells. These cancer cells can grow into (invade) other tissues, something that normal cells cannot do. Being able to grow out of control and invade other tissues is what makes a cell a cancer cell.

Carcinoma is the medical term used to describe tumours that are cancerous and malignant (having the ability to spread and invade other tissue).

In most cases the cancer cells form a tumour. Tumours can be benign (not cancer) or malignant (cancer).

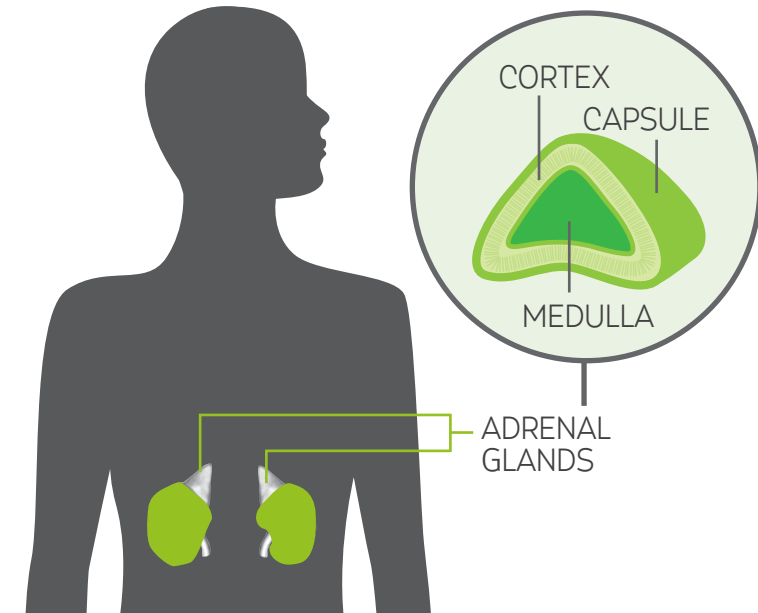
When cancer cells from solid tumours get into the bloodstream or lymph vessels, they can travel to other parts of the body. There they begin to grow and form new tumours that replace normal tissue. This process is called *metastasis*.

No matter where a cancer may spread, it is always named for the place where it started. For instance, breast cancer that has spread to the liver is still called breast cancer, not liver cancer. Likewise, prostate cancer that has spread to the bone is called metastatic prostate cancer, not bone cancer.

Different types of cancer can behave very differently. For example, lung cancer and breast cancer are very different diseases. They grow at different rates and respond to different treatments. That is why people with cancer need treatment that is aimed at their own kind of cancer.

Within this booklet, where we use the term 'tumour', we are talking about a malignant growth within the adrenal gland, known as Adrenocortical Carcinoma or ACC for short. We discuss what it is, how it is diagnosed and treated and how you can get the best information, support and self-care advice you may need.

WHAT ARE THE ADRENAL GLANDS?



The body has two walnut-sized adrenal glands, one above each of the kidneys ('ad-renal' means 'next to the kidney'). Even though the glands are small they are important as they produce several *hormones* (the body's chemical messengers) that are important for life.

The *adrenal medulla* (the inner area of the adrenal gland) makes a number of hormones called *catecholamines*, mainly *adrenaline* and *noradrenaline*. These hormones help the body to maintain blood pressure and deal with sudden stress or threats. The *adrenal cortex* (outer area of the gland) makes hormones called *steroids*, mainly *cortisol* (also known as *glucocorticoid*) and *aldosterone* (also known as *mineralocorticoid*). These steroids help the body to maintain blood pressure as well as salt and sugar levels. Cortisol is also an important messenger in our bodies' response to stress and illness.

WHAT IS ACC?

There are several different types of tumours that can occur in the adrenal gland. They can develop in either the outer part of the gland (the cortex) or the inner part of the gland (the medulla). Tumours can be benign (not cancer) or malignant (cancer).

Benign tumours of the cortex are called adrenocortical adenomas, and malignant tumours are called adrenocortical carcinomas (ACC).

Adrenocortical carcinoma (ACC) is often known simply as adrenal cancer and affects 1-2 people per million per year, making it a rare form of cancer.

ACC in adults tends to occur in people in their 50s and 60s and is more common in women than in men. Most ACCs are sporadic (meaning that they do not run in families), but they may sometimes be part of a condition that is congenital (present at birth) and/or familial (passed down in families).

WHAT ARE THE SYMPTOMS OF ACC?

Adrenocortical carcinomas (ACCs) are usually found because of the symptoms they cause but sometimes they are also found by chance if a patient has a scan for other reasons. ACCs may make higher than normal amounts of cortisol and aldosterone and may also secrete hormones that a healthy adrenal would not produce. These may include the male hormone *testosterone* and the female hormone *oestradiol* that cause symptoms and even body changes. Some ACCs get very large and although these don't usually make active hormones (they are non-functioning), they may cause symptoms by pressing on other organs in the body.

"I started feeling poorly [with] intermittent bloating and bathroom changes so wasn't concerned until symptoms became constant and I felt a lump in my stomach area"

There are 2 types of ACC:

HORMONE SECRETING

This is when the tumour makes more than normal amounts of steroid hormones in addition to those already made by the normal adrenal gland cells.

High levels of these hormones cause symptoms that may include:

- diabetes
- high blood pressure
- sexual dysfunction,
- muscle weakness and wasting,
- weight gain,
- excess facial or body hair in women
- baldness in women

- deepening of the voice in women
- soreness and increase in the size of the breasts in men
- easy bruising,
- acne,
- early puberty in children,
- osteoporosis,
- reduced immunity (impaired response to infections),
- change in body shape
- mood changes.

"I started getting hair on my chest, on my back, the hair on my head started getting very thin. It got so bad that I started wearing wigs. I started gaining weight, plus I would have these episodes where I would just feel like passing out."

NON-FUNCTIONING

These tumours are likely to be found when symptoms occur due to the size of the tumour. Symptoms might include pain or swelling, weight loss or signs that the disease has spread outside of the adrenal gland. They do not cause the symptoms of high hormone levels, although sometimes these hormones are found in the blood or urine. Sometimes tumours are found by chance, such as during a scan while looking at something unrelated.

These tumours are described as *'incidental'*.

Most adrenal *nodules* (small tumours) discovered by chance are harmless but larger tumours (usually more than 4cm) need careful checking for cancer.

WHAT CAUSES ACC?

Scientists do not know exactly what causes most ACCs. However, over the past few years, they have made great progress in learning how certain changes in a person's *DNA* can cause cells in the adrenal gland to become cancer. *DNA* carries the instructions for nearly all the things our cells do, and *DNA* changes can become risk factors for getting certain diseases including some types of cancer.

Most *DNA* changes that are seen in cancers happen during life rather than having been passed down in families. These changes may be caused by exposure to radiation, lifestyle or cancer-causing chemicals. But many of these changes happen for no known reason.

Most ACCs do not run in families; however, a number of rare conditions can increase the risk of developing ACC. Therefore, if a person with ACC has other members of the family who have been diagnosed with cancer aged 50 years or less, they should ask for a referral to their Regional Genetics Service.

HOW IS ACC FOUND?

Very often, ACC tumours are not found until they have grown quite large. This is different for children since they react more to the hormones these tumours make. Sometimes, ACC may be found early by coincidence when having tests or scans for other unrelated health problems. Usually, ACC is found due to the symptoms caused by either the hormones they make or because the tumour has grown large and is pressing on other organs in the body.

WHAT TESTS ARE NEEDED?

These days, the first *imaging* test (scan) to be used is likely to be a *CT scan*. Other tests may be used to find out more about the size and spread of the tumour to nearby structures such as *lymph nodes* or to distant areas such as the liver and lungs. These tests help doctors to 'stage' the disease, i.e. how advanced it is, and to plan the right treatment or combination of treatments. Urine and blood tests are also essential to look for hormones that the tumour may be making.

One or more of the following tests may be done if the doctor thinks you may have an ACC:

CT scans (computed tomography): A *CT scan* uses x-rays to make detailed pictures of the inside of your body. Instead of taking just one x-ray, the doughnut-shaped CT scanner takes many pictures as it moves around you. A computer then combines these into a picture of a slice of your body. CT scans can show the adrenal glands and can often confirm whether a tumour is present, how large it is, and whether it has spread to nearby sites. CT scans also show the organs near the adrenal glands, as well as lymph nodes and distant organs. This test can help show if the cancer has spread to the liver or other organs. Before any pictures are taken, or after the first scan, you may also have an IV (intravenous) line put into your arm through which a *contrast dye* can be given. This helps the outline structures in your body look clearer. The contrast dye can cause some redness and a warm feeling that

may last from hours to days. A few people are allergic to the dye. Rarely, more serious reactions like breathing problems and low blood pressure can happen. Medicine can be given to prevent and treat these problems. Be sure to tell the doctor if you have ever had a reaction to any contrast substance used for CT scans before. CT scans take longer than regular x-rays and you need to lie still on a table while they are being done.

PET scan (positron emission tomography): For a *PET scan*, a type of radioactive sugar is injected into your vein. The sugar collects in cancer cells and shows up in the pictures. This test is useful for finding ACC that has spread outside of the adrenal glands.

MRI (magnetic resonance imaging): *MRI scans* use radio waves and strong magnets to take pictures. The MRI sometimes gives similar information to a CT scan but may be more helpful because it can show views from different angles. In long term follow-up, MRIs may be used to avoid the repeated exposure to radiation from a CT scan. An MRI scan can take up to an hour to perform. Due to the tube-like shape of the MRI scanner, you might feel quite confined during your scan. If you have problems with tight spaces, tell the doctor before your MRI is set up; you may be able to have the test done using a less confined open MRI scanner. The machine makes loud thumping and clicking noises during the scan as the magnet switches on and off. You will be given earplugs or headphones, sometimes with music, to help block this out.

Chest x-ray: This can show if the cancer has spread to the lungs. This is also often done to check on your general health status before an operation

Ultrasound: This test uses sound waves to make pictures of organs inside your body. A computer shows the picture on a screen. This test can show if there is a tumour in the adrenal gland or liver. This would not be the first choice of test as a CT scan would be preferred.

Blood and urine tests: If there is a suspicion that the tumour is making increased amounts of hormones, then this can be measured in the bloodstream and/or in a urine collection as part of an initial hormone screening. Some hormone tests are done after patients have taken a synthetic steroid tablet called *dexamethasone*. It may also be possible to measure hormones that do not cause any effect on the body. All of these

tests are equally useful after surgery to check that all of the tumour has been removed and, later on, to see if there is any evidence of the tumour returning (follow-up screening). In addition, it may be necessary to do more common blood tests to assess your general level of health.

- Full blood count
- Liver and kidney function
- Hormonal work-up
 - Glucocorticoid levels
 - Dexamethasone suppression test
 - 24 hour urine collection test for free urinary cortisol
 - Basal cortisol (serum)
 - Basal ACTH (plasma)
- Sexual steroids and steroid precursors
 - Serum DHEA-S, 17=OH-progesterone, Androstenedione and Testosterone
 - 17-beta-oestradiol (serum, men and postmenopausal women only),
 - 24 hour urine collection to measure steroid metabolites
- Mineralocorticoid levels
 - Potassium (serum), aldosterone/renin ratio (only in those with arterial hypertension with or without low potassium levels)
 - Catecholamine levels
 - Normetanephrines, metanephrines and methoxytyramines (plasma)
 - Alternatively; fractionated metanephrine production (24 hour urine collection)

These tests or some of these tests may not be needed if it is already certain that you have ACC.

Biopsy: A *biopsy* is when a sample of tissue is removed with a fine needle to see whether cancer cells are in it (Fine Needle Aspiration or FNA). FNA should NOT be done in suspected cases of ACC since it usually does not give enough tissue to tell the difference between cancerous and non-cancerous growths. In addition, it is important that the tumour remains intact and piercing it by a biopsy may lead to tumour spread.

STAGING OF ACC

Doctors use 'staging' to describe the extent of disease. The 'stage' of disease guides the doctors when planning treatment and also gives an idea of likely long term survival. Staging is done using the results of imaging tests together with further information obtained after surgery. A *pathologist* will test the tissue after surgery, to help stage the disease using a *Weiss* score and a *Ki67* level. It is vital that the staging tests are carried out so that the right treatment option can be offered to you. The stage of ACC depends upon the size of the tumour and how far it may have spread. If you want to know more about how your cancer is staged, please talk to your nurse or doctor.

TREATING ACC

For ACC, the treatment that offers the best chance of a cure is timely surgery by a specialist surgeon experienced with ACC surgery. This can be achieved if the tumour is localised (just in one place). Follow-up is essential however even if 'curative surgery' is thought to have been achieved. Even after complete removal the tumour can recur, and your doctor would want to ensure that he/she caught any recurrence in good time. Unfortunately, ACCs can be quite fast growing. They are often diagnosed in the later stages (when they are quite advanced) and this means it can be difficult to cure them. Nevertheless, even if surgery cannot remove all of the tumour, it may sometimes be helpful to reduce the volume of the tumour(s) to allow other treatments to be used. Surgery can also be a useful option after other drug treatment has helped to shrink the tumour.

ACCs are rare and **treatment should be carried out by specialist teams**. Specialist centres that offer surgery and other therapies for adrenal cancer will have a 'multidisciplinary team' that meet regularly to discuss ACCs. These days, all decisions about surgery and other treatments should be taken by a team including a number of doctors of different specialities rather than one individual doctor. You should make sure that you have been referred to a multidisciplinary team experienced in caring for patients with ACC and other adrenal tumours. .

As part of your treatment, your doctor may suggest:

- Surgery
- Radiotherapy
- Chemotherapy
- Mitotane
- Combination treatments
- Clinical Trials

MULTI-DISCIPLINARY TEAMS (MDTs)

The care for an ACC patient can be complex. However, for the patient the journey can include not only a wide range of tests, treatments, but also a whole host of emotions and contact with many different healthcare professionals.

The very fact that there are often several treatment options from diagnosis and throughout the patient journey, means that collaboration between many key healthcare professional groups is essential in order to make the best clinical decisions for individual patients.

This type of collaboration is usually called an MDT (multidisciplinary team). MDTs are now used across the world in the care of cancer and rare disease patients.

An MDT will usually be led by a particular specialist with expertise and interest in ACC and other adrenal tumours. This specialist is commonly an endocrinologist or oncologist and is often the person that patients will see at their hospital appointments.

An ACC patient may see some or all of the following people:

- Oncologist (Cancer Specialist)
- Surgeon
- Endocrinologist (Hormone Specialist)
- Radiology staff (scans)
- Dietitian
- Nurse Specialist (including Macmillan Nurses)
- Pain Team
- General Practitioner/Practice Nurse
- Counselling Staff
- Various Technicians

- Clinic Staff
- Hospital Staff
- Palliative/Supportive Care Team (in hospital, at home or in a hospice)

Patients can feel more confident in the knowledge that all aspects of their care have been discussed and that the best possible treatment plan will be made. **An effective and experienced MDT is a very important aspect for care** when striving to achieve the best quality of life and the best outcome for ACC patients.

SUPPORTIVE / PALLIATIVE CARE

If a cure is unlikely or not possible, you will be entitled to specialist end-of-life care. This is also known as palliative or supportive care. A variety of services are available to help you to live as well as possible until you die and to die with dignity. You can receive end of life care at home, or in care homes, hospices or hospitals, depending on your needs and preference. For more information on palliative care, visit the following NHS website: <https://www.nhs.uk/conditions/end-of-life-care/>.

SURGERY

If your cancer is diagnosed early enough, surgery to remove the cancer and the adrenal gland is the first choice of treatment and can cure the cancer. This operation is called an *adrenalectomy*. If your surgeon thinks the cancer may have spread locally, they will remove the tissues immediately surrounding the adrenal gland together with nearby lymph nodes. When tumours are large they may have grown into surrounding structures such as the liver or kidney. In these cases surgery may still be possible but the operation will be larger. Even when the tumour has spread (metastasised) to other organs, it may still be appropriate to remove the initial tumour first and then remove or treat other metastases later.

TYPES OF SURGERY

The type of surgery that your surgeon will use will depend on the type and size of the tumour and also your individual needs. There are different surgical approaches that can be performed to remove malignant ACCs.

Open adrenalectomy - Most frequently, surgeons will make either a horizontal, diagonal or vertical cut to open the tummy, to remove the tumour. Since the tumours are often large, the incisions need to be large too. Sometimes surgeons use an incision in the side just below the ribs.

Laparoscopic Adrenalectomy - in this 'key-hole surgery' operation, a number of small cuts are made in the tummy through which operating instruments and a camera are inserted. This is the preferred approach for small benign (non-cancerous) tumours since it is a smaller operation and generally causes less pain and a shorter hospital stay than other approaches. However, for ACC this is not usually either possible or appropriate and ACCs are usually better treated

by a conventional open operation. Laparoscopic adrenalectomy may sometimes be used in cases where the tumour is fairly small and it is not clear if it is a cancer or not.

COMPLICATIONS AND CONSEQUENCES OF SURGERY

Hormone production - if one adrenal gland is removed, the other gland is usually able to take over making all the hormones you need. Some patients can experience a delay in this happening, and may need replacement steroid hormone tablets for a period of time, until the remaining adrenal gland is working fully. These tablets will also be needed if you are given mitotane after surgery, since this drug stops the remaining gland from working.

Infection – as with any surgery, there is always a risk of infection in the wound or of the chest.

Bleeding – this can happen during or after surgery. Sometimes you may wake from the surgery with a drain from the wound to help with this.

MITOTANE (LYSODREN)

Your doctor may suggest a drug called *mitotane* (Lysodren). It works by killing both normal adrenal cells and cancer cells. It can also stop the remaining adrenal gland from working, meaning that replacement steroid hormone medicines are needed while mitotane is being used. Mitotane can be given in four different settings:

Treatment of persistent or recurrent disease

If the initial surgery is unable to remove all of the tumour or the tumour returns later, Mitotane has been proven to reverse the disease and can sometimes lead to complete *remission* for years. Most doctors would be cautious about claiming that it can ‘cure’ the condition in this situation. Mostly, Mitotane will be continued for life.

Adjuvant therapy

There is evidence that mitotane may be useful in preventing cancer returning after ‘curative surgery’, i.e. if all tumour tissue appears to have been successfully removed. This applies in particular to ACCs considered to have a relatively high risk of returning as judged by examining the tumour tissue obtained at surgery under the microscope. In this situation, it is usually recommended to start a course of two to three years of mitotane treatment, which should start within 12 weeks of the surgery. If the tumour is judged to have a relatively lower risk of returning, then usually the patient is given the choice between close follow-up (observation only) and treatment with mitotane as for this situation no clear evidence of what to do currently exists. In all instances, patients after apparently complete removal of the ACC will be followed by regular blood and urine tests and usually CT scans.

Primary therapy

In some patients the disease is advanced and no surgical treatment can be performed. In this situation, chemotherapy may be used to shrink the tumour(s) and this can be monitored by a scan. In such a situation your doctors may not only recommend chemotherapy but also mitotane; however, this depends on individual circumstances.

Controlling hormone secretion

Sometimes steroid hormones that cause problems, such as too much cortisol or too much testosterone, remain high after surgery or when surgery cannot be done. Mitotane may be helpful to control the hormone levels and prevent distressing symptoms.

Mitotane may be used alongside chemotherapy. Mitotane can also treat the symptoms of advanced adrenocortical carcinoma. You take mitotane each day as a number of tablets (often 6-12).

Side effects of Mitotane

Side effects are common and include nausea, tiredness and feeling dizzy. Nausea can often be controlled with anti-sickness pills and other side effects can usually be controlled by using a lower dose of mitotane.

"I've been tolerating mitotane fairly well so far, mostly nausea and fatigue that waxes and wanes... good days and bad."

Mitotane kills normal adrenal cells and the normal hormones that these would make need to be replaced with tablets. All patients on mitotane require replacement of the normal cortisol levels using hydrocortisone tablets. Hydrocortisone needs to be taken at all times even if mitotane is temporarily stopped or paused. When mitotane is being used in the adjuvant setting (as a temporary therapy to prevent tumour recurrence) and it is stopped after two to three years, the function of the adrenal gland may sometimes recover, but

hydrocortisone needs to be continued until blood tests prove that the patient does not need it anymore. Patients on replacement hydrocortisone must be aware of the 'sick day rules' of this medication (see box).

ADRENAL CRISIS AND SICK DAY RULES

The following signs may indicate an adrenal crisis which requires immediate treatment:

- Extreme weakness, feeling terrible, vomiting, headache
- Light-headedness or dizziness on sitting up or standing up
- Feeling very cold, uncontrollable shaking;
- back, limb or abdominal pain
- Confusion, drowsiness

Treat by:

- injecting yourself (or the person you are caring for) with your hydrocortisone ampoule in your emergency injection kit (100mg)
- seeking immediate medical attention - call 999, stating "adrenal crisis" and "adrenal insufficiency"

Sometimes patients on mitotane also need replacement of the blood pressure hormone aldosterone, and for this reason will also have to take fludrocortisone tablets. Your doctor will check whether you need fludrocortisone by measuring your blood pressure whilst sitting and standing and by carrying out blood tests.

Mitotane is effective if the level in the body is controlled to a particular level (14-20mg/L). This needs blood samples to be taken every 1-2 months to make sure that the level is high enough to kill the cancer cells, but not to cause side effects. Your doctor will advise you on side effects to be expected from mitotane treatment. If mitotane blood levels are too high the function of the brain can be affected causing problems often described as "trouble talking and trouble walking". This side effect is fully reversible when mitotane treatment is temporarily stopped or the dose is reduced.

There is a separate and more detailed information leaflet about mitotane treatment which your doctor will give to you if you are considered for mitotane treatment.

RADIO THERAPY

Radiotherapy uses radioactive beams to kill cancer cells and prevent or delay disease recurrence. It is usually only used when a large ACC has spread beyond the adrenal glands, in particular to the bones. In this instance, Radiotherapy often helps to control the growth of the bone lesions and to ease pain. An MRI scan is used to plan the radiation field, and then the treatment is given by pointing the radiation beam via 3 targets to focus on the area of disease. This is quite painless, and is usually given for 5 days a week over 5 weeks, giving 25 treatments altogether. Each treatment usually lasts half an hour, and most patients can carry on their normal life throughout although they may tire more easily than usual.

Side Effects of Radiotherapy

- Sore skin (like sunburn)
- Feeling tired
- Feeling sick
- Diarrhoea

If you feel very suddenly tired and sick, you should contact your medical team straight away as this can be a sign of anaemia (low levels of red blood cells).

CHEMOTHERAPY

Chemotherapy uses anti-cancer (cytotoxic) drugs to destroy cancer cells. It is given by slow injection through a vein. You may have just one drug or a combination of more than one drug. Chemotherapy is usually only used to treat ACC that has spread to other parts of the body. It is given in a number of cycles of treatment, with time for recovery from side effects between each one. Your doctor will discuss how many cycles of chemotherapy you may need. Chemotherapy can be used in several different ways:

Primary chemotherapy (treatment with chemotherapy only)

Chemotherapy is given on its own when it is expected to control or cure the cancer; it can be given as a short-term treatment or for long-term disease control.

Adjuvant Chemotherapy (chemotherapy given after surgery)

This is used in this manner to decrease the risk of the cancer coming back. This is done even when no clear evidence of cancer can be found, but certain factors (e.g. metastasis to the lymph nodes, large tumour size) predict an increased risk of cancer recurrence.

Neoadjuvant Chemotherapy (chemotherapy given before surgery)

This is used to shrink a tumour before surgery, which may allow the surgeon to perform a smaller surgery and/or remove all visible tumour.

Combined Chemotherapy

This is the practice of using chemotherapy together with other treatments, such as radiation or surgery or mitotane. Therapies are combined to obtain a greater response rate than could be achieved with a single type of treatment. Today,

using combinations of treatments is common for most cancers. Your doctor will discuss which options are suitable for you.

Some of the chemotherapy drugs used to treat ACC are:

- Cisplatin
- Doxorubicin (Adriamycin)
- Paclitaxel (Taxol)
- Fluorouracil (5FU)
- Vincristine
- Etoposide (VP16)
- Gemcitabine
- Capecitabine
- Streptozotocin

Combination Therapy

In some cases the use of one single chemotherapy treatment is not as effective as combining two different treatments together. Examples of combination treatments are:

- Etoposide + Doxorubicin + Cisplatin + Mitotane (Lysodren)
- Streptozotocin + Mitotane

- Etoposide + Cisplatin + Mitotane
- Gemcitabine + Capecitabine

Side Effects of Chemotherapy

Short Term - Chemotherapy is very toxic and because it affects both cancer cells and healthy cells in the body, it can cause a number of side-effects, including:

- Feeling tired
- Feeling sick (and being sick)
- Hair loss
- Pain (head and muscle aches)
- Diarrhoea or constipation

Most side effects get better within a few days after each cycle of treatment. In addition, other medicines may be given to help lessen them.

Long Term - Most side effects go away after treatment. However, some may continue, come back, or develop later. For example, some types of chemotherapy may cause permanent damage to the heart, lung, liver, kidneys, or reproductive system. Some people have concentration and memory problems for months or years after treatment.

FOLLOW-UP TESTS AFTER TREATMENT

For patients who have had the whole tumour removed, the following tests may be done afterwards on a regular basis:

- CT/MRI scan, 3 monthly for 2 years, then 3-6 monthly for further 3 years
- Ongoing testing beyond 5 years is suggested but can be adapted according to each individual case
- Regular hormone screening using blood tests

For patients who have not had the whole tumour removed, or who have disease that has spread and cannot be removed by surgery, scans and blood tests may be done on a regular basis according to the extent of the disease in each individual case.

CLINICAL RESEARCH

Research is a step-by-step process that involves collecting and examining information. Research into adrenocortical carcinomas is vital to improve our understanding of the disease and how it can be treated.

Research goals include:

- Understanding what causes ACC
- Understanding how ACC grows
- Finding more effective scans and tests for diagnosis and management of ACC
- Finding new treatments, and ensuring that current treatments are being used so as to provide the best outcome for the patient

ACCs are a rare form of cancer, and there are only a small number of dedicated medical teams around the world who treat patients every day. It is important that these teams are given the resources to carry out research within their centres, so that our understanding of this disease and how to treat it continues to grow.

In clinical trials, patients agree to try new therapies (under careful supervision) in order to help doctors find the best treatments with the fewest side effects. Clinical trials may also test the value of new tests to diagnose disease and to help the doctors to detect earlier when the tumour is coming back.

If patients want to take part in a clinical trial, they should discuss this with their specialist, who will know what trials are recruiting patients and whether they are eligible.

To decide who is eligible, all studies are run on strict inclusion and exclusion criteria in order to ensure the safety of patients.

Criteria may include things like age and site or stage of disease. **It can be frustrating for patients to discover that they are not eligible**, but no medical professional is able to influence any decisions based on these criteria.

No one should ever include a patient in a clinical trial without his or her knowledge. A doctor, nurse or other researcher will ask for permission (consent), and they cannot enter a patient into the trial unless that patient has given this.

To help patients decide whether they want to take part, the researchers should tell them all about the study:

- what it is trying to find out
- how they will be treated
- what they will have to do
- what possible side effects may be if testing a new drug.

Even after consent has been given, a patient may leave the trial without giving a reason at any time. If a patient is having a new treatment as part of a trial and then leaves the trial, he or she may not be able to continue having the new treatment. In this situation, patients would be given the appropriate standard treatment for their type of cancer.

You can find out more about current trials at:

Current Controlled Trials

This website allows users to search, register and share information about randomised controlled trials. Covers England, Scotland and the US but not easy to search. <http://www.controlled-trials.com/>

ClinicalTrials.gov

This is a register of federally and privately supported clinical trials conducted in the United States and around the world.

<http://www.clinicaltrials.gov>

You can also search for research trials available to patients in the UK and Europe on these websites:

www.macmillan.org.uk

www.cancerhelp.org.uk

www.ctu.mrc.ac.uk (The Medical Research Council Clinical Trials Unit)

www.ncrn.org.uk (National Cancer Research Network)

www.eortc.be/ (European Organisation for Research and Treatment of Cancer)

www.ensat.org (European Network for the Study of Adrenal Tumours)

EMOTIONAL WELLBEING

Living with a rare cancer is challenging. Some people cope better than others, but most people will have periods of low mood at some point along the way. It is now better recognised that overall health depends upon both physical and emotional health. For this reason, both AMEND and the Neuroendocrine Cancer UK offer a free telephone counselling service. In addition, our Counsellors are sometimes available for face-to-face sessions at our free events. See our websites for more details.

It should also be remembered that hormones secreted in large quantities by ACCs can also cause mood changes. Partners and family members of patients should bear this in mind as much as the pressure of having cancer. Patients should discuss any such concerns with their doctor.

AMEND has produced some resources that patients with any rare disease or cancer should find useful. 'Dealing with Diagnosis', 'Living with Uncertainty' and 'Looking after Yourself' are available to download for free from the Resources section of AMEND's website or in hard copy on request. A series of podcasts and an introductory video on the relaxation method, Mindfulness, are also free to access via our website and YouTube Channel (AMEND3).

"It's nice just to have a cuppa and a chat with other people in a similar position"

USEFUL INFORMATION

FREE PRESCRIPTIONS: In the UK, you are entitled to free prescriptions for all your medicines if you need to take lifelong hydrocortisone.

You should obtain a FP92A leaflet from your doctor and fill in parts 1 and 2. Your doctor will then sign it and send it on. You will then receive a Medical Exemption Certificate, which you must show to your pharmacist when collecting medicines. You can find more information on Medical Exemption Certificates on the following website: www.nhsbsa.nhs.uk/exemption-certificates/medical-exemption-certificates

MEDICALERT Emblem®: We recommend that anyone taking lifelong medicines obtain and wear a MedicAlert® identification emblem. The emblem contains summary information of your medical condition and a 24-hour Helpline number for emergency medical staff to call in order to obtain detailed information on your medical condition from the MedicAlert database. This helps emergency medical staff to give treatments in full knowledge of your underlying condition and current medicines. Emblems come in a range of styles so that there is something for everyone, even children. Join online at www.medicalert.org.uk. Other medical identification products are available.



Neuroendocrine Cancer UK

Patient information, support and specialised nurse advice line

0800 434 6476

<http://neuroendocrinecancer.org.uk/>



AMEND (Association for Multiple Endocrine Neoplasia Disorders)

Patient information, support and counselling service

01892 516076

www.amend.org.uk



Macmillan Cancer Support

National cancer support charity

www.macmillan.org.uk



Maggie's Centres

Drop-in centres offering free practical, emotional and social support to people with cancer and their families and friends.

www.maggiescentres.org/

ACTH	Adrenocorticotrophic Hormone made by the pituitary gland to tell the adrenal glands how much stress hormone to make
Adjuvant therapy	A treatment given after an initial treatment
Adrenal gland	A pair of walnut-sized organs found above the kidneys that make stress hormones
Adrenal cortex	The outer layer of the adrenal gland
Adrenal medulla	The inner layer of the adrenal gland
Adrenalectomy	Surgery to remove the adrenal gland (and any tumour within the gland)
Adrenaline / Epinephrine	'Fight or flight' hormone made by the inner part of the adrenal gland (medulla)
Aldosterone	Adrenal gland hormone that helps maintain fluid and salt levels in the body
Benign	A condition, tumour, or growth that is not cancer.
Biopsy	Using a fine needle (often guided by an ultrasound probe) to remove tissue from inside the body for tests
Bone density	The amount of calcium and other minerals in a bone. Assessing bone density is used to detect and/or monitor osteoporosis and its treatment.
Cancer	The abnormal and uncontrolled growth of cells, which can lead to loss of cell structure and function. Cells that become cancer can invade nearby cells and travel to other parts of the body.
Carcinoma	The medical word for cancer
Catecholamines	Hormones made by your adrenal glands that are released when you are physically or emotionally stressed – these include dopamine, norepinephrine (noradrenaline) and epinephrine (adrenaline)

Chemotherapy	Cancer treatment using chemicals	Incidental	Found by chance while looking for something else
Contrast dye	A substance radiologists use that acts like a dye (but without permanent stain) to allow certain parts of the body to show up more clearly on scans. It can help tell one structure from another.	Laparoscopic	Surgery through several small cuts in the skin. Also known as 'key-hole' surgery
Corticosteroids	Steroid hormones that are either made by the body (in the outer part of the adrenal gland – the cortex) or are man-made. There are 2 key types: glucocorticoids and mineralocorticoids. Manmade corticosteroids mimic the actions of naturally occurring corticosteroids and may be used to replace corticosteroids in people with adrenal glands that are unable to make enough.	Lymph nodes	Oval or bean shaped organs (1 - 30 mm in diameter) that help form part of the lymphatic system
Cortisol	The main hormone made by the outer part of the adrenal gland (cortex)	Lymphatic system	A network of tissues and organs that help rid the body of toxins, waste and other unwanted materials. Its main function is to transport lymph, a fluid containing infection-fighting white blood cells, throughout the body.
CT scan	Combines a series of X-ray images taken from different angles around your body and uses computer processing to create cross-sectional images (slices) of the bones, blood vessels and soft tissues inside your body. CT scan images provide more details than plain X-rays do.	Malignant	A condition, tumour or growth that is cancerous.
Cytotoxic	Substances that are toxic to living cells	Metastasis (pl. metastases)	Cancer that has spread to other sites in the body
Dexamethasone	This medication is a corticosteroid hormone (glucocorticoid) that is used for a number of health conditions including cancer. It may also be used as a test for adrenal gland disorders	Mineralocorticoid	A steroid hormone that helps to regulate the balance of salt and water in the body
DNA	Short for deoxyribonucleic acid; the carrier of genetic information, stored in every cell in the body	Mitotane	An oral chemotherapy drug used to treat cancer of the adrenal glands called adrenal cortical carcinoma
Glucocorticoid	A steroid hormone that can suppress inflammation and immunity and assist in the breakdown of fats, carbohydrates, and proteins	MRI scan	A type of scan that uses strong magnetic fields and radio waves to produce detailed images of the inside of the body.
Hormones	The body's chemical messengers made by glands	Neoadjuvant therapy	A treatment given before another treatment to improve or allow that second treatment to work more effectively – for example chemotherapy as a first step to try to shrink a tumour so that surgery can take place.
Imaging	Scans	Nodules	Small swelling, growth or tumour; that can be benign or malignant
		Noradrenaline / Norepinephrine	'Fight or flight' hormone made in the inner part of the adrenal gland (medulla). It is vital in controlling blood pressure, and also plays a role in affecting mood, sleep patterns and concentration
		Oestrodial	Female sex hormone – it is the main oestrogen made by

the body and is involved in the development and function of the female reproductive system, breast development and bone density.

Pathologist A doctor who interprets and diagnoses the changes caused by disease in the body's cells and tissues.

PET Scan A scan that produces detailed 3-dimensional images of the inside of the body by detecting the radiation given off by a substance (radiotracer) injected into your arm. The tracer collects in specific parts of the body and by analysing these areas it is possible to work out how well certain body areas it is possible to work out how well certain body functions are working and identify any abnormalities. Different tracers are used for different conditions and cancers.

Radiotherapy A form of cancer treatment that uses X-ray radiation to destroy cancer cells

Recurrence A cancer that has come back, usually after a period of time during which the cancer could not be detected. The cancer may come back to the same place as the original (primary) tumour or to another place in the body.

Remission A decrease in or disappearance of signs and symptoms of cancer. In partial remission, some, but not all, signs and symptoms of cancer have disappeared. In complete remission, all signs and symptoms of cancer have disappeared, although cancer may still be in the body.

Risk factor Something that increases the chance of developing a disease. Some examples of risk factors for cancer are age, a family history of certain cancers, use of tobacco products, being exposed to radiation or certain chemicals, infection with certain viruses or bacteria, and certain genetic changes.

Stage / Staging The extent of a cancer in the body. Staging is usually based on the size of the tumour, whether lymph nodes contain cancer, and whether the cancer has spread from the original site to other parts of the body.

Steroids One of a large group of chemical substances classified by its structure. Steroids include drugs used to relieve swelling and inflammation, such as prednisone and cortisone, and certain hormones made by the body, such as testosterone and oestradiol.

Testosterone A (steroid) hormone that stimulates development of male secondary sexual characteristics, made mainly in the testes, but also in the ovaries and adrenal cortex.

Ultrasound scan (US) A scan that uses high-frequency sound waves to create an image of part of the inside of the body – such as that used in pregnancy to see the developing baby.

Weiss Score The Weiss score is a scoring system used in histology to assess ACC. It has a set of points (9 in total) that is used to look at the cell's appearance, activity and behaviour in order to confirm the diagnosis. A score of 3 or more may confirm the cell as ACC.

X-Ray Scan An X-ray is usually a quick and painless scan commonly used to produce images of the inside of the body. They're mainly used to look at the bones and joints, although they're sometimes used to detect problems affecting soft tissue, such as internal organs. For example a chest x-ray may be done to check for signs of infection and / or other abnormalities.

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- Professor Wiebke Arlt, CEDAM, University of Birmingham
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This book has been written for ACC patients with the help of a medical advisory team. The aim of this book is to answer those questions, sometimes in great detail, that one may come across after receiving a diagnosis of ACC. It is not for use in self-diagnosis. It contains detailed information on tests, surgery and potential symptoms associated with ACC. However, it is possible that not all of this information will be relevant to you. This book is not intended to replace clinical care decisions and you should always discuss any concerns you have with your specialist. Every care has been taken to ensure that the information contained in this book is accurate. Nevertheless, ACC Support UK cannot accept responsibility for any clinical decisions.

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ADRENOCORTICAL CANCER
SUPPORT AND INFORMATION

ACC Support UK is a joint venture between two well-respected patient organisations. Through working together, AMEND and Neuroendocrine Cancer UK provide information resources and support services to ACC patients and their families and carers in the UK. Both organisations provide free psychological Counselling services and moderate a private Facebook Group for those affected by ACC which is helpful for peer support. Neuroendocrine Cancer UK provides a specialist nurse helpline for patients and AMEND provides opportunities for ACC patients to meet others at a variety of free events. Both organisations work together to provide up-to-date information resources to help patients to navigate through their ACC journey.

Please visit the ACC Support UK website for more information and to join for free: www.ACCsupport.org.uk

Find us on Facebook <https://www.facebook.com/ACCSupportUK/>

Follow us on Twitter (@ACCSupportUK)

Scan to go to our website:

