Understanding Adrenocortical Carcinoma

A Guide for Patients

A diagnosis of adrenocortical carcinoma can bring many questions and concerns. This guide is to help you better understand this rare disease, the treatment options, and the different steps of care.

Inside, you will find clear, reliable information to help you move forward with confidence and know that you are not alone.

This educational brochure was created by ESTEVE
and Hannah Franjou (illustrator) in collaboration with ENS@T,
Let's Cure ACC, and patients and their families.
We would like to extend sincere thanks to the patients
and families for their valuable insights and time.

The content of this educational brochure is based on major open access publications and currently available data on adrenocortical carcinoma.

1. European Society of Endocrinology Clinical Practice Guidelines on the management of adrenocortical carcinoma in adults, in collaboration with the European Network for the Study of Adrenal Tumors. European Journal of Endocrinology 2018. doi: 10.1530/EJE-18-0608

2. Adrenocortical carcinomas and malignant phaeochromocytomas: ESMO-EURACAN Clinical Practice Guidelines for diagnosis, treatment and follow-up. Annals of Oncology 2020. doi: 10.1016/j.annonc.2020.08.2099

3. Adrenocortical carcinoma: a practical guide for clinicians. Lancet Diabetes and Endocrinology 2025. doi: 10.1016/S2213-8587(24)00378-4

4. Let's Cure ACC website https://letscureacc.com/

ENS@T is the European Network for the Study of Adrenal Tumours. https://ensat.org/

ESTEVE is a pharmaceutical company that provides treatments in highly specialised areas. https://www.esteve.com/

Let's Cure ACC is the international patient advocacy group dedicated solely to adrenal carcinoma. https://letscureacc.com/

This brochure is designed for you and your loved ones.
It aims to support you and provide the clearest information possible on adrenocortical carcinoma.
This brochure also aims to help you to prepare any questions you may wish to ask your physician at your next medical visit.

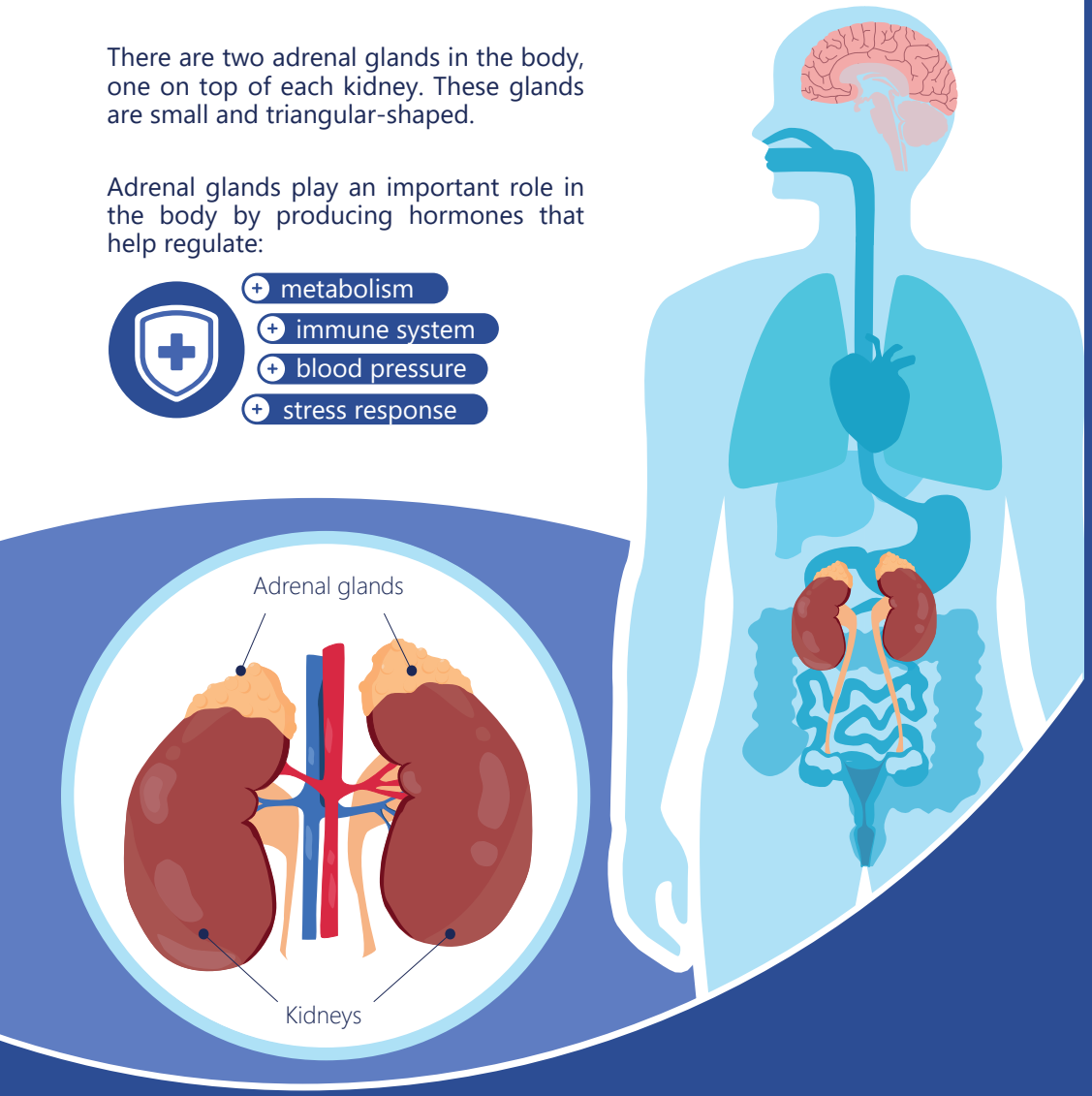
At the end of this brochure, you will find an extensive list of terms to help you get a better understanding of adrenocortical carcinoma.

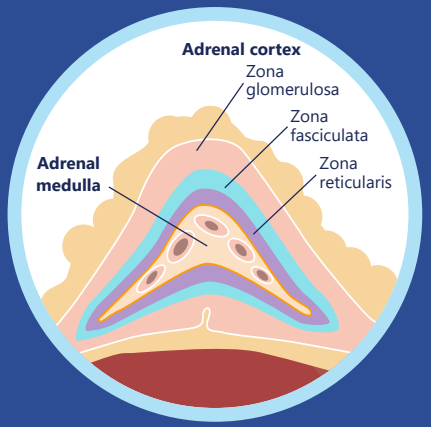
1 ADRENAL GLANDS: WHAT THEY ARE AND WHAT THEY DO

There are two adrenal glands in the body, one on top of each kidney. These glands are small and triangular-shaped.

Adrenal glands play an important role in the body by producing hormones that help regulate:

- metabolism
- immune system
- blood pressure
- stress response





Each adrenal gland consists of 2 parts:

The adrenal medulla *(inner part)*

The medulla produces catecholamines or stress hormones:

- adrenaline (epinephrine), and
- noradrenaline (norepinephrine).

These trigger the "fight-or-flight" response.

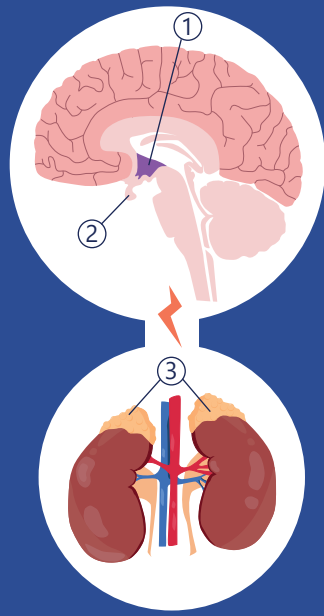
The adrenal cortex *(outer part)*

The cortex produces three groups of steroid hormones:

- aldosterone, a mineralocorticoid secreted by the zona glomerulosa, which regulates the salt and water balance in the body.
- androgens and oestrogens, sex hormones secreted by the zona reticularis, which affect reproductive development and secondary sex characteristics, and
- cortisol, a glucocorticoid secreted by the zona fasciculata, which manages metabolism and the body's response to stress.

The secretion of glucocorticoids is regulated by a hormone called ACTH (adrenocorticotropic hormone), which is produced by a small gland, the pituitary gland, situated at the base of the brain.

The interaction between these different parts of the body is called the hypothalamic-pituitary-adrenal (HPA) axis, which is our body's main way of responding to stress. It consists of three parts (the hypothalamus, the pituitary gland, and the adrenal cortex) that each release hormones to eventually increase the level of cortisol in the body.

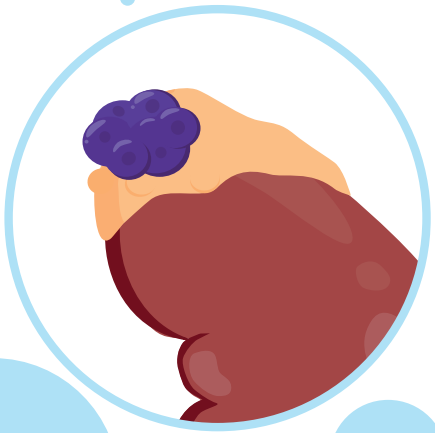


① Hypothalamus

② Pituitary gland

③ Adrenal glands

2 WHAT IS ADRENOCORTICAL CARCINOMA?



Adrenocortical carcinoma (ACC) is a rare endocrine tumour in which malignant (cancer) cells form in the adrenal cortex. ACC has an estimated annual incidence (i.e., rate of new cases) of 0.5-2 cases per million individuals.

Adrenocortical carcinoma can occur in both adults and children.

ACC is typically discovered in one of three ways:

1. If there are no symptoms, ACC may be discovered incidentally (by chance) during an imaging procedure (e.g., MRI) performed for other reasons.

2. It can be discovered because of non-specific symptoms like back-pain or side-pain, bloating or nausea, or vomiting. These symptoms are related to the size of the tumour and the resulting pressure on other organs. Cases without hormone over-production are called "non-secreting" or "non-functional" ACC.

3. It may be discovered when lab tests show high levels of certain hormones produced by the adrenal glands, e.g., cortisol, which causes Cushing's syndrome. When there is an excess production of hormones, it is called "secreting" or "functional" ACC.

ACC WITH HORMONAL OVERPRODUCTION

An overproduction of hormones occurs in 60-70% of ACC patients. This excess production of hormones has a negative impact on the HPA axis, which normally regulates steroid hormone production in the body.
In 50-70% of cases, cortisol is the hormone that is overproduced; this results in Cushing's syndrome, which is often the first sign that something is wrong with your adrenal glands (once tests have been done to ensure your pituitary gland is working properly).

CUSHING'S SYNDROME

Signs and symptoms of Cushing's syndrome may include, but are not limited to:

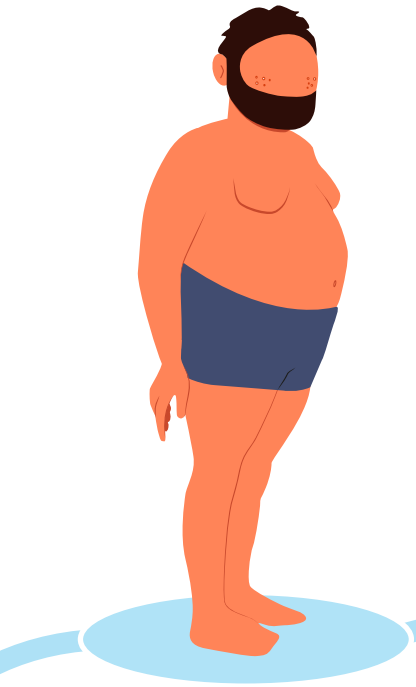
- Obesity
- Flushed, rounded face with pudgy cheeks *(moon face)*
- Fatty, rounded hump high on the back just below the neck *(buffalo hump)*
- Purple stretch marks *(especially on the abdomen, thighs, breasts, and arms)*
- Thinning, fragile skin that bruises easily
- Irregular or absent menstrual periods for those who have a menstrual cycle
- Delayed growth *(in children and teens)*
- Virilization: development of male traits *(body and pubic hair, acne, deeper voice, enlarged clitoris in females).*
- Mood swings
- High blood pressure
- Osteoporosis
- Diabetes

OTHER HORMONE-RELATED SYMPTOMS

Although excess cortisol is the most common hormone abnormality in ACC, these tumours can also produce excessive amounts of other hormones normally released by the adrenal cortex.

Aldosterone is one of these. This hormone helps regulate blood pressure and electrolyte balance. When overproduced, it can lead to such symptoms as:

- High blood pressure
- Muscle problems
- Weakness



If the ACC tumour produces excess androgens, it can cause virilization in women, with the appearance of male characteristics such as increased facial and body hair growth, acne or deepening of the voice.
Similarly, an excess of oestrogens in men can cause gynecomastia (abnormal but non-cancerous enlargement of breast tissue).

Comprehensive blood tests will be performed to determine hormone levels.
Patients with ACC-induced Cushing's syndrome often show abnormal levels of multiple adrenal hormones, not just cortisol.

The exact cause of adrenocortical carcinoma is still unknown. However, in rare cases, it may develop from inherited (genetic) disorders, such as Li-Fraumeni syndrome. These conditions are caused by specific gene mutations and can be passed down through families.

3 HOW IS THE DIAGNOSIS OF ADRENOCORTICAL CARCINOMA CONFIRMED?

Depending on your symptoms, your doctor will do a physical exam, imaging (scans), biological tests (blood, urine), and maybe a biopsy (tissue sample) to confirm the diagnosis of ACC and to assess whether the tumour has spread to other parts of your body (metastases).

IMAGING

Different imaging (scan) techniques can be used. They show the size of the tumour, where it is located, and whether it has spread to other parts of your body. The most commonly used techniques are a CT (computed tomography) scan, MRI (magnetic resonance imaging), or a special form of PET (positron emission tomography) scan called FDG-PET.

LABORATORY TESTS

Blood and 24-hour urine tests are done to check hormone levels.

HISTOPATHOLOGY

The diagnosis of ACC is confirmed by analysing a small sample of the tumour that is removed during surgery. A pathologist then studies the cells under a microscope. A scoring system called the Weiss score determines how likely the tumour is to be malignant (cancerous). This helps distinguish ACC from other types of adrenal tumours and is a key tool in confirming the diagnosis.

4 YOUR MEDICAL TEAM

A MULTIDISCIPLINARY TEAM

ACC is a rare, endocrine tumour, so you need to be taken care of in an expert center with a team of specialists from different disciplines, including an endocrinologist, an oncologist, a radiologist, a surgeon, a nurse, a pathologist, a nutritionist, a radiotherapist, and a nuclear medicine physician. This team will work together, sharing their expertise and providing a holistic and coordinated approach to your treatment.

THE IMPORTANCE OF EXPERT CENTERS

Specialist cancer centers provide everything the patient needs. They ensure that each diagnosis is confirmed by an expert multidisciplinary team and that every patient benefits from an individualised treatment plan. These centres also participate in clinical trials, meaning that patients have access to the latest innovations in treatment options. Your role as a patient is crucial to the success of clinical research. You may be asked to provide consent for your medical data or any surplus tissue removed during surgery to be used for research. This will not require you to undergo any additional exams or procedures. By participating in registries and clinical studies, you will make a valuable contribution to research on future treatment options and to improving our understanding of this rare disease.

EXISTING EXPERT NETWORKS

ENS@T (European Network for the Study of Adrenal Tumors) is a collaborative European initiative focused on improving the understanding, diagnosis, and treatment of adrenal tumour, including rare types like adrenocortical carcinoma. A list of these centers is available on the ENS@T website.

EURACAN (European Reference Network for Rare Adult Solid Cancers) is a specialized network established by the European Commission to improve care for patients with rare adult solid tumours, like adrenocortical carcinoma.

https://ensat.org/page-1317257
https://www.euracan.eu/patients

5 TREATMENT OPTIONS

There are different treatment options which depend on the medical care available in your country and the clinical characteristics of your ACC. Here is a brief overview.

1. SURGERY

Wherever it is possible, surgery is the most effective and most widely recommended treatment, as it can greatly affect your overall outcome. The procedure includes removing the primary tumour, metastases (when accessible), and any tumours that may have come back (recurred). Surgery must be performed by a specialist with experience in this type of cancer. Once your tumour has been removed, a pathologist will study it under a microscope. Based on their observations, your expert team will determine the most appropriate treatment for you.

2. MITOTANE

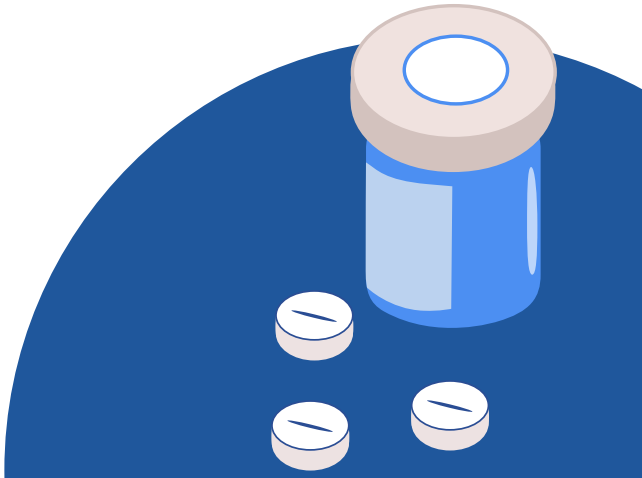
Mitotane is the only drug specifically approved for the treatment of advanced ACC by both the European Medicines Agency (EMA) and the U.S. Food and Drug Administration (FDA). It is also recommended by the main international clinical practice guidelines (ESE-ENS@T and ESMO-EURACAN).

Mitotane works in two main ways:
-It destroys adrenal cancer cells (this is called the adrenolytic effect).

-It reduces the excess production of hormones such as cortisol, which occurs frequently with ACC tumours.

This dual action can help control the tumour and its symptoms but may also affect the healthy adrenal gland, meaning your body may produce inadequate quantities of certain hormones (a condition called adrenal insufficiency). You may therefore be required to take glucocorticoid replacement therapy (hydrocortisone) and maybe mineralocorticoid replacement therapy (fludrocortisone).

Treatment with mitotane must be overseen by an experienced endocrinology and/or oncology specialist. Your prescribing physician will give you precise instructions on how and when to take mitotane tablets.



3. CHEMOTHERAPY

Chemotherapy is used to destroy cancer cells even though it may also damage healthy cells. There are different ways of taking chemotherapy but in the case of ACC, the drugs are injected directly into the body (intravenously).
Chemotherapy is the recommended treatment for advanced or recurrent ACC. Chemotherapy combined with mitotane gives the best response rates.
The most frequently used chemotherapy for ACC is a combination of three drugs—etoposide, doxorubicin, and cisplatin—and is known as EDP.

4. RADIOTHERAPY

Radiotherapy, also known as radiation therapy, is a localised treatment that targets a specific part of your body. It uses radiation to destroy cancer cells. However, like chemotherapy, it may also affect healthy cells. If surgery was unable to eradicate all the cancer, radiotherapy in combination with mitotane may be recommended.

5. INTERVENTIONAL RADIOLOGY

In certain cases of advanced ACC, local radiological therapies such as thermo- or thermal ablation can be used to treat metastatic lesions (e.g., lung, liver, bone), to reduce the risk of onset of symptoms and block disease progression.

6. OTHER THERAPIES

Other oral drugs, called steroidogenesis inhibitors (e.g., ketoconazole, metyrapone, and osilodrostat), are used to reduce the overproduction of cortisol.

Lastly, there are certain new drugs or different drug combinations that are not approved for use but only studied in clinical trials (e.g. immunotherapy and targeted therapies). You may be invited to take part in the ongoing clinical research. Should you accept, you will be given all the information you need and your consent will be requested.

A PERSONALISED TREATMENT APPROACH

To ensure you receive the most appropriate treatment for your ACC, the multidisciplinary team of specialists will work together to create a personal treatment plan for you; the exact sequence and type of treatment will depend on your individual situation.

The treatment plan may also include medical treatment after surgery; this is known as adjuvant therapy.

FERTILITY PRESERVATION

Although you will understandably be initially focussing on the diagnosis and treatment of your cancer, your healthcare providers will also tell you about the potential threats to your fertility. There is a wide array of options available and the topic should be addressed as early as possible (before treatment begins) by your healthcare providers.

6 FOLLOW-UP CARE VISITS

During the course of your treatment, your healthcare team will ask you to attend regular follow-up visits. The main objective will be to check your health status and talk about the different treatments. Your medical team might ask for additional tests, such as imaging, and blood and urine tests to be done.

MAKING THE MOST OF YOUR FOLLOW-UP APPOINTMENTS

The follow-up visits are a very good opportunity for you to provide feedback on your treatment, ask any questions you may have, and express your concerns. This discussion will enable the medical team to reassess your medical treatment, and adjust or change it according to your health status and latest test results. At the end of each visit, you and your doctor will discuss what the next steps in your care may involve and you will be given all the necessary advice and prescriptions.

Do not hesitate to reach out to your medical team in between follow-up visits

YOUR NOTES

7 WHAT YOU NEED TO KNOW ABOUT THE PROGNOSIS

Each ACC is a unique case, and your doctor is the best source of information on your particular case.

ACC can behave aggressively and often tends to come back after treatment. However, outcomes vary widely, and the increasing availability of expert centers, international collaborations, and ongoing advances in therapy and research are offering new hope for patients.

Outcomes vary according to multiple individual factors. Be sure to ask your physician about the 4 key factors listed below to get a better understanding of your specific situation and treatment options available to you.

1. EXCESS PRODUCTION OF THE HORMONE, CORTISOL *(assessed before surgery and during your follow-up)*

In 50-70% of hormone-secreting adrenocortical carcinoma, there is an overproduction of cortisol, which leads to Cushing's syndrome.
Cushing's syndrome is a life-threatening condition and results in increased health complications, such as cardiometabolic disorders, bone fractures, or psychological disorders.

Excess cortisol levels are mainly detected on blood and urine tests.

2. STAGE OF YOUR CANCER

- Stage I: Tumour ≤5 cm, confined within the adrenal gland
- Stage II: Tumour >5 cm, confined within the adrenal gland
- Stage III: Tumour has spread to areas near the adrenal gland (vena cava, renal vein)
- Stage IV: Metastatic disease (the tumour has spread to other parts of the body)

3. RESECTION STATUS AFTER SURGERY

Resection, also called "R", can be defined in 4 stages (R0, R1, R2, Rx). "R" status refers to the degree of resection (how much cancerous tissue has been removed).

- R0: resection is complete (whole tumour has been successfully removed) with clear margins
- R1: resection is incomplete, as some microscopic tumour tissue could not be removed
- R2: resection is incomplete, with macroscopic tumour tissue left (meaning the tissue can be seen by the naked eye)
- Rx: it is impossible to determine the resection status or it is unknown.

4. KI-67 PROLIFERATION INDEX OF TUMOUR

Ki-67, expressed as a percentage (%), shows how fast cancer cells in a tumour are growing and dividing. It is one of the most powerful tools for predicting the risk of recurrence. A high Ki-67 score means the cells are dividing rapidly and that the cancer is likely to grow and spread. This number also defines the grade of the tumour:

- low-grade tumours with a Ki-67 <10%
- intermediate grade tumours with a Ki-67 of 10–19%
- high-grade tumours with a Ki-67 ≥20%

Never hesitate to contact your doctor if you have any questions regarding your health, your treatment, or your prognosis. Knowledge is power - but only when you feel prepared.

8 ADRENOCORTICAL CARCINOMA IN KIDS AND TEENS

The special needs of this group of patients are addressed by paediatric physicians. There are fundamental differences in terms of development and genetic characteristics between tumours in children and in adults. Therefore, the findings regarding adrenocortical carcinoma cannot be uncritically transferred from adults to children.

Especially in childhood, ACC is often associated with so-called cancer predisposition syndromes, meaning a genetically inherited, familial occurrence of these tumours (e.g., Li-Fraumeni or Beckwith-Wiedemann syndromes). Genetic testing is generally recommended for all children with ACC. If a genetic predisposition is identified, it is advisable for family members to undergo genetic testing. Subsequent close monitoring may help reduce the risk of cancer.

In contrast to ACC tumours in adults, almost all tumours in children produce hormones (80% of cases are hormonally active). Primarily, the cortisol and male hormone precursors are secreted. The main conditions include Cushing's syndrome with symptoms such as reduced growth, weight gain, high blood pressure, skin changes, early puberty development, or abdominal pain.

Since paediatric ACC is very rare, experts worldwide collaborate closely, both clinically and scientifically. For paediatric tumours, the international working group "KIDS" of ENS@T-PACT has been set up. These experts work closely together with the Brazilian/American working group IC-PACT. Only in this way can children worldwide benefit from medical advances and expertise.
Please feel free to contact them to ask for advice and for a specific referral.

https://icpact-gm.com/
https://ensat.wildapricot.org/page-1317315

9 LIVING WITH ADRENOCORTICAL CARCINOMA

Both the initial diagnosis and subsequently, coming to terms with having ACC, can have a notable impact on your everyday life. It can affect your general well-being (making you tired) and your emotions. It will also require you to attend frequent hospital visits and undergo urine and blood tests, and imaging tests.

ACC is an extremely rare disease and you may feel like you are the only one dealing with this form of cancer. Although that feeling of isolation can be overwhelming, you are not alone. Start by identifying a circle of friends and/or family in whom you can trust. And never hesitate to contact a patient advocacy group if you feel you may benefit from talking to other patients in the same situation.

Let's Cure ACC is the only international patient advocacy group dedicated solely to adrenocortical carcinoma. Let's Cure ACC is a network of patients worldwide in contact with over 150 ACC specialists in more than 50 countries. It provides a unique opportunity for patients to connect with other individuals who understand exactly what they are going through. You will find people across the globe struggling with the same questions, challenges and need for support.

Contact Let's Cure ACC if you ever need to talk to patients on any topic. You are not alone any more!

https://letscureacc.com/contact-us/

SOME USEFUL TIPS

Being diagnosed with ACC or any other rare cancer or illness can trigger a multitude of emotions, such as anger, sadness, or fear. These feelings are all perfectly reasonable and understandable but we would like to offer a few useful tips that we hope will provide some comfort:

1. Do not be afraid to ask for help. Giving friends/family specific tasks is a great way to get help and to allow people who care about you to feel like they are supporting you.

2. Seek out appropriate online resources for the diagnosis. Always consult a qualified medical professional for advice about your condition.

3. Give yourself grace. Your house is a mess because you are so distracted with the news and/or the treatments. That is OK!

4. Prioritize rest if you need it. Pushing yourself to the point of exhaustion will only worsen your mental health.

5. Do not be afraid to seek psychological support.

6. Explore meditation/mindfulness.

7. If you are working during your treatment, find out what workplace adjustments can be made.

8. Engage in activities that you enjoy and create moments of happiness with your loved ones.

9. If you are able to exercise, even a short walk outdoors can be beneficial to your mental health.

10. If you have concerns about your emotions or mental health, share them with your loved ones and your medical team.

ACTH: Adrenocorticotropic hormone, or ACTH, is produced by the pituitary gland. Its main function is to trigger production of cortisol by the adrenal glands.

ADRENAL GLANDS: These are two small glands located on top of the kidneys. They produce hormones that help regulate heart rate, blood pressure, the immune system, and how the body responds to stress, but also other functions.

ADRENALINE: Also known as epinephrine, it is a hormone secreted by the medulla, i.e., the inner part of the adrenal glands. Adrenaline plays a role in the body's fight-or-flight response (the body's response to stress).

ADRENOCORTICAL CARCINOMA: Adrenocortical carcinoma is a rare cancer that originates in the adrenal cortex, the outer layer of the adrenal glands.

ALDOSTERONE: Aldosterone is a mineralocorticoid hormone secreted by the adrenal cortex. It regulates salt and potassium levels in the kidneys. If a patient's adrenal glands are not working properly, they will be given fludrocortisone.

ADJUVANT: Additional systemic treatment given after the initial surgery to reduce the risk of disease recurrence.

ADRENAL INSUFFICIENCY: Adrenal insufficiency occurs when the adrenal glands do not produce enough of the hormone cortisol.

ANDROGENS: Androgens are one of the 4 main classes of steroid hormones that stimulate the development and maintenance of male genitalia, facial and body hair, deepening voice and muscle mass. They are the masculinising hormones.

BECKWITH-WIEDEMANN SYNDROME: This is a growth disorder syndrome. It can cause physical differences and increase a child's likelihood of developing certain childhood cancers, including adrenocortical carcinoma.

CHEMOTHERAPY: Chemotherapy is a drug treatment that uses powerful chemicals to kill fast-growing cells in your body. Chemotherapy is most often used to treat cancer, since cancer cells grow and multiply much more quickly than most cells in the body.

CORTISOL: A glucocorticoid hormone secreted by the adrenal cortex. It can be replaced by hydrocortisone during treatment with mitotane (which reduces cortisol levels) or after surgical removal of both adrenal glands.

CT SCAN: A CT (computed tomography) scan is an imaging test that helps healthcare providers detect diseases and injuries within your body. It uses a series of X-rays and computer processing to create detailed images of your bones and soft tissue.

CUSHING'S SYNDROME: Cushing's syndrome is an accumulation of signs and symptoms caused by excess levels of cortisol in the body, typically due to an overproduction of cortisol by the body.

DOXORUBICIN: Doxorubicin is a chemotherapy drug used to treat various forms of cancer. It is usually used in combination with other chemotherapies.

ENDOCRINE SYSTEM: The endocrine system is in charge of creating and releasing hormones to maintain countless bodily functions. Endocrine tissue include the adrenal glands, the pituitary gland, the thyroid, the pancreas, and others. There are several conditions related to endocrine system issues, usually due to a hormone imbalance or problems directly affecting the tissue.

ENS@T: The European Network for the Study of Adrenal Tumours is a network of experts that work in the field of adrenocortical carcinoma.

ENS@T-PACT: The European Network for the Study of Adrenal Tumours for Paediatric Adrenocortical Carcinoma.

ESE: The European Society of Endocrinology (ESE) represents over 5,000 members and 50 national societies, supporting research and providing education to raise awareness and improve hormone health across Europe and the globe.

ESMO: ESMO is the European Society for Medical Oncology. This medical society represents more than 40,000 oncology professionals in 179 countries and territories. ESMO is a reference for oncology education and information.

ETOPOSIDE: Etoposide is another drug used to treat various types of cancer. It works by damaging the genetic material of cells and is given by mouth or by injection into a vein.

EURACAN: EURACAN is one of the 24 European Reference Networks (ERNs) funded by the European Commission and dedicated to rare diseases. EURACAN is the ERN for rare adult solid cancers.

FERTILITY PRESERVATION: Fertility preservation is the process of saving or protecting eggs, sperm, or reproductive tissue to enable someone who may have a medical condition or be undergoing certain medical treatments to use them to have biological children in the future.

FDG-PET scan: A positron emission tomography (PET) scan is an imaging procedure that uses an injectable drug (fluorodeoxyglucose), called a tracer, that highlights atypical activity such as cancer within the body. It is a complementary imaging test that is typically combined with CT or MRI scans.

FLUDROCORTISONE: This is a corticosteroid. It is a synthetic mineralocorticoid that is used when the body does not produce sufficient amounts. It controls salt and fluid levels and thus helps to stabilise blood pressure.

GLUCOCORTICOIDS: Steroid hormones secreted by the adrenal cortex. The main glucocorticoid hormone is cortisol.

HISTOPATHOLOGY: The study of tissue samples under a microscope.

HORMONES: Hormones are the body's messengers. They are produced by an endocrine gland and travel in the bloodstream to tissue or organs where they affect many different processes.

HYDROCORTISONE: Hydrocortisone is the name for the hormone cortisol when supplied as a medication.

HYPOTHALAMUS: A small structure deep within the brain that acts as your body's coordination center. Its primary function is to maintain the body in a stable state, known as homeostasis. It does so by directly influencing the autonomic nervous system (involuntary processes, e.g., heart rate, breathing, etc) or by managing hormones.

HYPOKALAEMIA: Low levels of potassium in the blood.

IC-PACT: International Collaboration for Paediatric Adrenocortical Carcinoma.

IMMUNOTHERAPY: Unlike chemotherapy, which specifically targets and destroys cancer cells, immunotherapy is a form of cancer treatment that activates the body's own immune system, thus improving its ability to recognize and eliminate cancer cells.

INCIDENTALOMA: Tumour discovered incidentally or by coincidence, usually as a result of tests carried out for another symptom or suspicion.

INTERVENTIONAL RADIOLOGY: Interventional radiology (IR) is a way to treat cancer and other conditions without major surgery.

Ki-67: This is a protein that is only found in cells that are actively dividing. It serves as a marker for how quickly the cancer cells are growing and dividing.

Let's Cure ACC: Is the international patient advocacy group dedicated solely to adrenal carcinoma.

LI-FRAUMENI SYNDROME: This is a rare hereditary condition that increases the risk of developing cancer.

METABOLISM: The physical, chemical or biological processes that keep the body alive.

METASTASIS: A collection of cancer cells that have migrated via the blood or lymph from the original site of disease.

MINERALOCORTICOIDS: A group of steroid hormones secreted by the adrenal cortex, which act on the kidneys to regulate the metabolism of sodium, potassium and water. The main mineralocorticoid is aldosterone, which can be replaced by fludrocortisone treatment.

NORADRENALINE: Also known as norepinephrine, noradrenaline plays an key role in the body's "fight-or-flight" response. Norepinephrine can be given as a drug to increase and maintain blood pressure levels in limited, short-term serious health situations.

OESTROGEN: This feminising hormone triggers the development and maintenance of female sexual characteristics.

PITUITARY GLAND: This is a small endocrine gland at the base of the brain. It produces many hormones and regulates the production of hormones in other endocrine glands.

RENIN: Renin is an enzyme. It helps control blood pressure, and ensures that sodium and potassium levels in your body remain healthy. Renin is secreted by special cells in the kidney; it is released into the bloodstream if blood pressure levels fall too low.

RADIOTHERAPY: You may be given radiotherapy, or radiation therapy, as part of your cancer treatment. This technique uses radiation, usually x-rays, to destroy cancer cells.

REPLACEMENT THERAPY: When the body's natural production of hormones is inadequate, a replacement therapy may be initiated. For example, patients on mitotane are also given hydrocortisone, and sometimes fludrocortisone, to compensate for the cortisol and aldosterone deficiency that results from this treatment.

STEROID HORMONES: Steroid hormones are secreted by the endocrine glands: the adrenal cortex secretes cortisol, the testes secrete testosterone, and the ovaries secrete oestradiol.

TARGETED THERAPY: Targeted therapy is a personalised type of cancer treatment. Oncologists identify then target the specific genetic changes that cause an individual's normal cells to become cancer cells. There are many kinds of targeted therapies. They are designed to stop cancer cells from growing and spreading but also to limit damage to normal, healthy cells.

TUMOUR PATHOLOGY: Tumour pathology is the study of tumours. Pathology reports of tumour cells removed during surgery provide crucial information on their characteristics and therefore on the diagnosis and staging of the cancer.

TUMOUR/CANCER STAGING: Staging is the process of classifying the disease in an individual, i.e., how much cancer is in a person's body, where it was first detected and if it has spread.

WEISS SCORE: The Weiss score is the gold standard method for distinguishing between a benign and a malignant adrenocortical tumour. It is based on nine criteria. A Weiss score ≥3 means the mass is cancerous and the diagnosis of ACC is confirmed. Scores of ≤2 define benign adrenal adenoma (a benign mass).

ZONA FASCICULATA: The middle layer of the adrenal cortex. It is primarily responsible for the production of glucocorticoids, such as cortisol.

ZONA GLOMERULOSA: The outermost layer of the adrenal cortex, primarily responsible for the production of aldosterone, a mineralocorticoid hormone.

ZONA RETICULARIS: The innermost layer of the adrenal cortex, primarily responsible for producing adrenal androgens and, to a lesser extent, estrogens.

WANT TO LEARN MORE ABOUT ADRENOCORTICAL CARCINOMA

at your own pace?

DOWNLOAD THIS BOOKLET

and find helpful information and support

ens@t
european network for the study of adrenal tumors

LET'S CURE ACC

ESTEVE

MAT-IG-NPR-0001