



Information for patients and their families and friends about

Adrenal insufficiency

Appendix to the 'Quality of Care Standard for Adrenal disorders in the Netherlands'

Introduction

This brochure has been written for adults with an adrenal insufficiency and also for those closest to them such as partners, parents and older children. Adrenal insufficiency is a disorder that affects the adrenal glands. This brochure explains the disorder and describes the treatment regime.

Although we use the pronoun 'you' throughout this brochure, it is also intended for people in the immediate circle of a person with adrenal insufficiency. Healthcare practitioners can also use this brochure as a source of information when they are treating you.

This information is not intended to replace the advice of a competent physician. If you have any queries or need advice, get in touch with your GP or your specialist.

This brochure describes the Dutch situation. Please check with your local healthcare professional if and how this can be applicable in your situation outside the Netherlands



The medical information in this brochure is based on the Dutch Quality of Care Standard for Adrenal Disorders (Dutch only) and the 'Diagnosis and Treatment of Primary Adrenal Insufficiency: An Endocrine Society Clinical Practice Guideline'.

Where this brochure speaks of 'hydrocortisone', this should be taken to mean whichever glucocorticosteroid medication the physician treating you has prescribed. Depending on your situation, this might be cortisone acetate, prednisone or dexamethasone for example.



Adrenal disorders are complex illnesses and explanations can be difficult to understand. If you come across unfamiliar words when reading this brochure, the website BijnierNET.nl includes a Thesaurus, an explanatory list of definitions (in Dutch only). For healthcare practitioners, the Thesaurus on www.adrenals.eu provides a comparative overview of applicable codes.

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Say hello to...
Lizzie
aged 40

Lizzie is 40 years of age and has adrenal insufficiency. She is a single mum with a daughter aged 20, and works as a cashier at a local supermarket. Lizzie has difficulty reading and writing. She finds it difficult to remember the name of the disorder she suffers from, and even more difficult to understand the consequences. And it took such a long time to get the correct diagnosis!

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The HPA axis

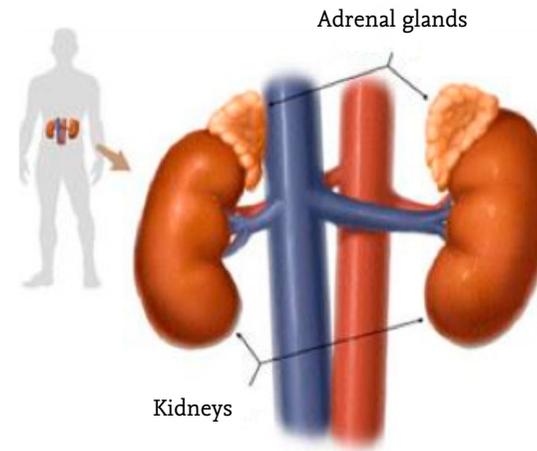
Adrenal insufficiency is a complex disorder. To properly understand what actually goes wrong, it is important that you understand the basics of how hormones work. That is what this paragraph is about: to explain what the HPA axis does.

HPA is the English abbreviation of 'hypothalamic-pituitary-adrenal axis'.

Everyone has one. It's a system that connects the hypothalamus, the pituitary gland and the adrenal glands. Doctors in many parts of the world use the English abbreviation: the HPA axis. The system consists of three components:

- the hypothalamus
- the pituitary gland
- the adrenal glands.

The hypothalamus and the pituitary gland are located in your head; the adrenal glands lie at the back of the abdominal cavity (your tummy).



The adrenal glands sit on top of the kidneys, just like hats.

Hypothalamus

The hypothalamus lies just above the pituitary gland. It plays an important part in the day-to-day functioning of your body (emotions, motivation and pleasure) and it sends instructions to the pituitary. The hypothalamus continually measures hormone levels in the blood. Is there enough cortisol, for example? If not, it will send an instruction to the pituitary gland using the hormone CRH. Hormones can therefore act as messengers and relay signals.

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Pituitary gland

The pituitary is a small organ, the size of a pea, that lies at the base of the brain. It plays a central role in regulating and adjusting the levels of various hormones in the body. The pituitary gland itself contains hormone-producing cells. When they receive instructions from the hypothalamus, these cells produce hormones such as ACTH and TSH. ACTH travels to the adrenal glands via the blood, and once there it stimulates the adrenal glands to produce cortisol.

Adrenal glands

The adrenal glands sit on top of the kidneys in the abdominal cavity. The kidneys are part of the renal system, the adrenal glands are adjuncts to the kidneys - hence their name. Everyone has two adrenal glands, one on each kidney, right and left. Healthy adrenal glands are about the size of your thumb.

Each adrenal gland consists of two components:

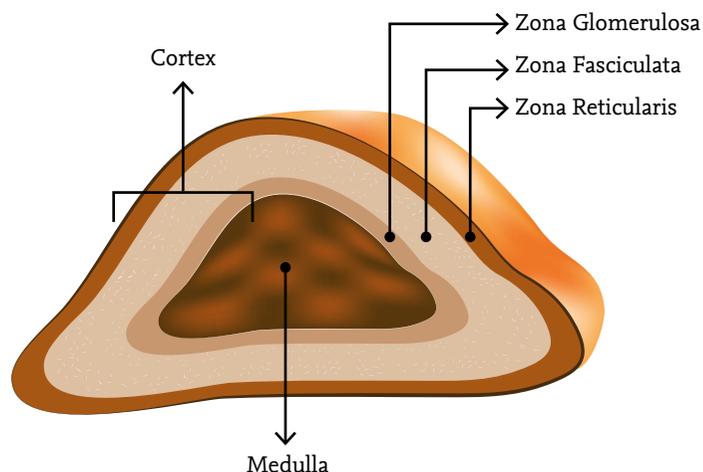
- The outside, called the cortex
- The inside, called the medulla.



The function of the adrenal glands is explained in more detail on the [BijnierNET/AdrenalNET](#) website.

The cortex is in turn made up of three layers. These layers are called:

- zona glomerulosa
- zona fasciculata and
- zona reticularis.



Cross-section of an adrenal gland

The various hormones are all produced within these three layers. Hormones are essential to our lives and they regulate everything our body does in different circumstances. The following hormones are produced in the **adrenal cortex**:

- cortisol,
- aldosterone,
- androgens (sex hormones).

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- There is a certain rhythm to the hypothalamic-pituitary-adrenal system. The hormones are released in pulses. The strength of the pulses decreases as the day progresses. That means that our adrenal glands produce a lot of cortisol in the morning, less towards the evening and none (or hardly any) at night. This system is complicated; it varies from person to person and it needs to be very precise. This makes it very difficult to replicate the system using medication.



The animated clip ‘The HPA axis’ on the Adrenals.eu website explains how this complex hormonal system works.

A well-functioning HPA axis is necessary for a normal everyday life. So what goes wrong in the case of people with adrenal insufficiency?

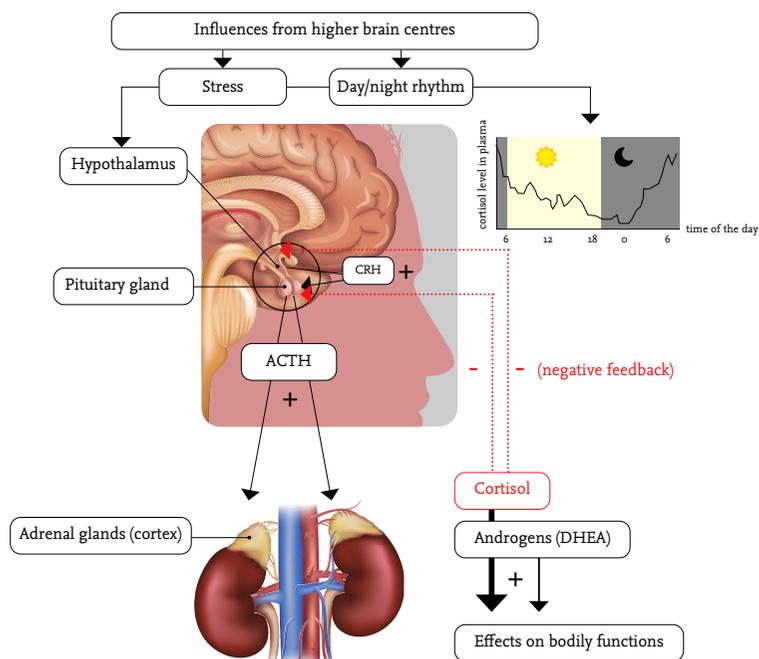


Diagram of the HPA axis

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What is adrenal insufficiency?

Adrenal insufficiency is a disorder which reduces the production of adrenal hormones. Adrenal insufficiency is a rare disorder that affects approximately 6000 to 7000 people in the Netherlands.

There is no real cure for adrenal insufficiency, but it can be treated effectively. The medicines hydrocortisone, fludrocortisone and sometimes DHEA are used for that purpose.

Three forms

There are three different forms of adrenal insufficiency.

1. Primary adrenal insufficiency -> a problem with the adrenal glands, usually caused by an autoimmune inflammation of the adrenal glands. It is estimated that 2000 to 2500 people in the Netherlands suffer from primary adrenal insufficiency.
2. Central adrenal insufficiency, of which there are two forms:
 - a. secondary adrenal insufficiency -> a problem with the pituitary gland. It is estimated (statistically) that between 2100 and 4700 people

in the Netherlands suffer from secondary adrenal insufficiency.

- b. tertiary adrenal insufficiency -> a problem with the hypothalamus.
3. Steroid-induced adrenal insufficiency -> a problem caused by the use of medication. A problem arises with the hypothalamus, the pituitary gland and the adrenal glands because of medication that contains cortisol or synthetic forms of cortisol. This happens mainly to people who have had to take prednisone for a long time, e.g. use eczema creams or inhalers (COPD). This is probably the most common form of adrenal insufficiency, but we don't know how many people are affected by it.



A simple explanation of adrenal insufficiency can be found in the infographic entitled 'What is adrenal insufficiency?' and the animated clip with the same name.

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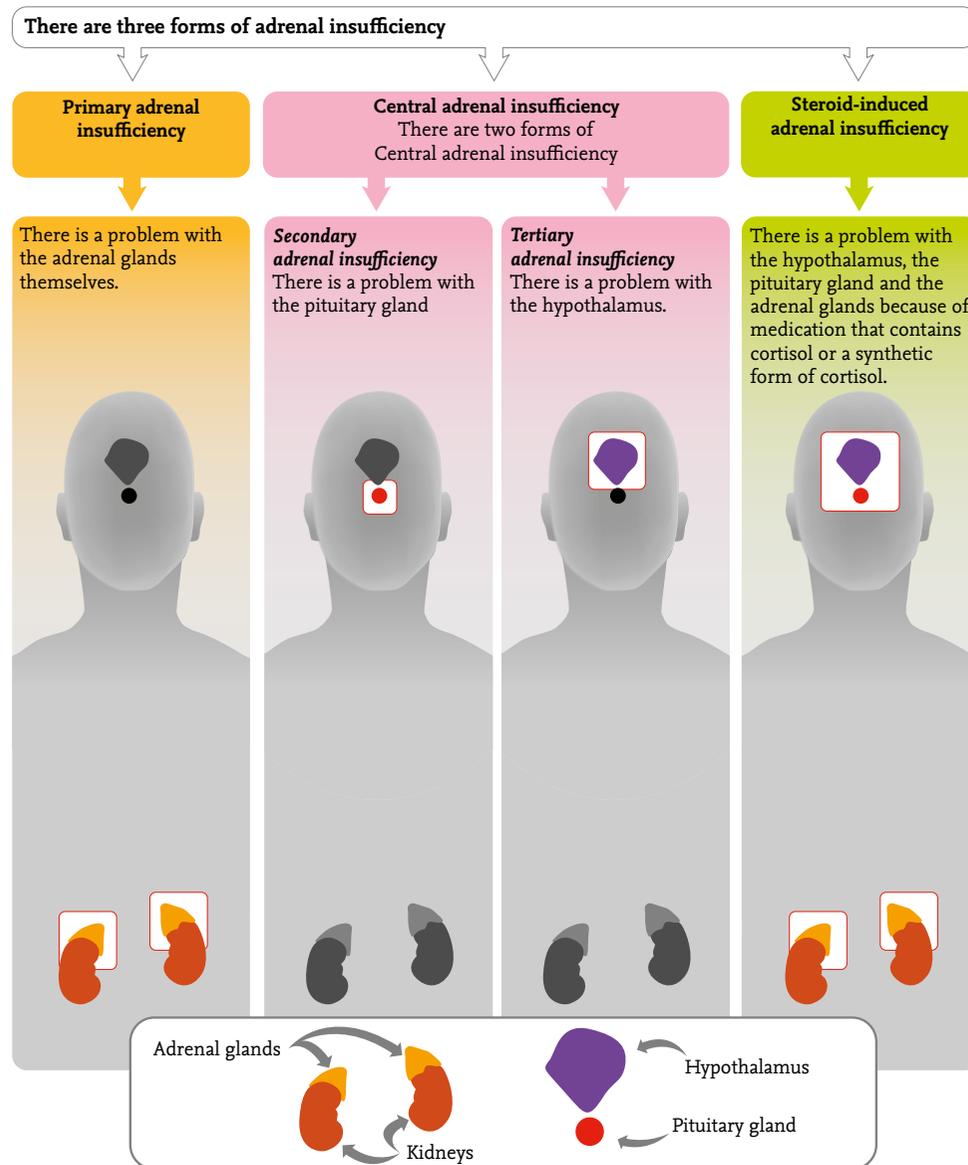
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What causes adrenal insufficiency?

Primary adrenal insufficiency is caused by damage occurring to the adrenal glands. Addison's disease is the most common cause of primary adrenal insufficiency. In the case of Addison's disease, the damage to the adrenal glands is caused by an inflammation of the adrenal glands as the result of an autoimmune disease. An autoimmune disease causes the body to make antibodies against itself. Those antibodies can lead to inflammation of a bodily organ; as a result that organ no longer functions properly. Other causes of primary adrenal insufficiency are infections, malignant tumours, severe bleeding or an operation to remove both adrenal glands.

Central adrenal insufficiency can be caused by any disorder, injury or operation that damages the hypothalamus or the pituitary gland (see Chapter 1 for further information about these organs). In some rare cases, primary and central adrenal insufficiency can also be caused by a predisposition or it can be part of a genetic syndrome.

Adrenal insufficiency caused by taking medication (steroid-induced adrenal insufficiency) occurs as a result of taking medication that contains glucocorticoids. These substances are used in many different sorts of medication, including creams, ointments or shampoos, injections, inhalers for lung diseases and tablets. These types of medication suppress the normal activity of the hypothalamus, the pituitary gland and the adrenal glands, and lead to adrenal insufficiency.

There are also other, less common causes of adrenal insufficiency. Examples of this are:

- infectious diseases such as HIV/AIDS
- metastases of cancer
- medicines such as mitotane
- genetic or familial (=inherited) disorders
- autoimmune reaction by the adrenal glands caused by immunotherapy
- inflammation of the pituitary gland caused by immunotherapy.

Those causes are not explained in this information brochure. If you need further explanation, please ask your GP or specialist.

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What are the symptoms of adrenal insufficiency?

A shortage of the hormone cortisol in the blood causes a wide range of symptoms. Symptoms can include:



1 Brown discoloration of the skin, mucous membranes in the mouth, and gums (only in the case of primary adrenal insufficiency).



2 Tiredness, lack of energy.



3 Weakness.



4 Nausea, vomiting, abdominal pain.



5 Weight loss, reduced appetite.



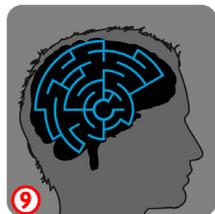
6 Pain in muscles and joints.



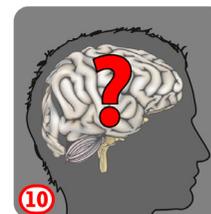
7 Salt craving (only in the case of primary adrenal insufficiency).



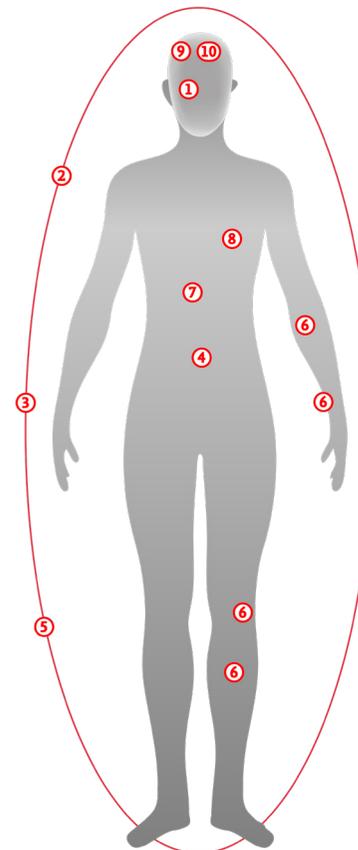
8 Low blood pressure.



9 Feeling gloomy.



10 Problems with memory, concentration, attention span.



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How is adrenal insufficiency diagnosed?

The diagnosis will be given on the basis of your symptoms and additional tests. Sometimes, the diagnosis becomes evident during an adrenal crisis (for an explanation, see page 17).

In other cases it can sometimes take quite a long time before the diagnosis is known. Let's focus on that for a moment first.

Delayed diagnosis

It is possible that you have felt unwell, or possibly even quite ill, for some time and that various doctors have been unable to pinpoint the precise diagnosis. This is because the symptoms are not exclusively linked to this particular adrenal disorder. This makes the diagnosis a treasure hunt, one that often begins with the family doctor. He will refer you to the hospital.

Unfortunately, adrenal insufficiency is often only diagnosed after a very long search. It is, after all, a rare disorder. When the diagnosis can only be given after what

- from your perspective - is a very long wait, we speak of a delayed diagnosis.

The intervening period is characterised by a great deal of uncertainty and doubt. Studies have shown that the average time that elapses between the beginning of symptoms and the diagnosis is around three years. Ultimately, the diagnosis is usually given by an endocrinologist or other internal specialist. In the case of autoimmune Addison, the functioning of the adrenal glands will slowly reduce even more and the symptoms will increase. The rate at which this happens varies from patient to patient.



In our mini documentary, (Dutch only) a patient explains how this long waiting time for the correct diagnosis has affected her personality and her life.

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Referral

In most cases, you will be referred to one or other medical specialist, depending on the symptoms you describe. Sometimes, the referral is on the advice of a physiotherapist, psychologist or dietician.

Working it out

Intake interview

The endocrinologist (or other specialist) will arrange an intake interview and carry out a physical examination.

First of all he will want to know whether you take any medication that contains glucocorticoids, such as creams or inhalers. This is, in fact, the most common cause of adrenal insufficiency, and doctors call this 'steroid-induced adrenal insufficiency'. If the adrenal insufficiency is caused by medication, an attempt will be made to reduce the medication. The adrenal glands will usually return to normal functioning after a short while.

You may be suffering from hypocortisolism: that means that the cortisol level in your blood is too low. Blood tests will be carried out to make sure.

For women

Oral contraceptives contain oestrogen. Oestrogen increases the binding protein for cortisol, cortisol-binding globulin (CBG). As a result, the concentration

of bound cortisol increases - and, with it, the overall cortisol - while the concentration of free cortisol - i.e. active, not bound to protein - remains unchanged. The total amount of cortisol is measured in the blood, and this will rise as a result of the pill. You may need to stop taking the pill for .XX. a number of month. Please consult your medical specialist.

How to find out if your cortisol level is too low

The hormone cortisol is important in any tests for adrenal insufficiency. The following four tests can be carried out to find out whether you have too little cortisol in your blood:

- early-morning cortisol level test
- ACTH test
- insulin tolerance test (ITT)
- metyrapone test

Early-morning cortisol level test

A lower than normal cortisol level (hypocortisolism) can be discovered if the blood is tested early in the morning; if the cortisol level is very low, this indicates an adrenal insufficiency. A further test is quite often necessary, because the level of cortisol in the blood may not provide a clear enough indication. That follow-up test is usually an ACTH test.

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The website of the Dutch Association for Clinical Chemistry & Laboratory Testing (NVKC) provides an [overview of the tests](#) (Dutch only) that are carried out in the laboratory. Unfortunately, the website is only in Dutch. If you type in the substance that is being tested for in your blood or urine, it provides you with a summary of what the test entails, when it is carried out and what the results mean.

If you have any further questions you can contact a clinical chemist via the [same website](#).

ACTH test

For this test, ACTH hormone is administered either by infusion (IV) or via an injection into muscle tissue (IM). After that, the cortisol level in the blood will be tested at set times. If your blood cortisol level does not rise sufficiently after the ACTH is administered, it is a sure sign that you have adrenal insufficiency.

Insulin tolerance test (ITT)

An Insulin Tolerance Test (ITT) checks whether you have enough of the hormone cortisol from the adrenal glands or the growth hormone from the pituitary gland. When an ITT is carried out, we can check whether your body is able to reduce the sugar level in your blood temporarily. This is done by administering

insulin. Insulin is a hormone that reduces the blood sugar level. Under normal circumstances, the body will respond to the insulin by increasing the production of cortisol and the growth hormone.

Metyrapone test

The metyrapone test is used as a test for central adrenal insufficiency. For this test, you will be admitted to the hospital on the evening before. The test starts between 11.30 p.m. and midnight, and the aim is to find out whether the pituitary gland can be stimulated to produce ACTH. You will be given some tablets to take. After that, you may not eat anything and you will be told to rest (or sleep) in bed because of possible side-effects.

Next morning, between 8 and 9 a.m., the lab will take blood samples so that they can test whether the levels of ACTH and other hormones have increased. After the blood test you can eat breakfast. Medical staff will decide whether you are then allowed to go home.

Additional tests in the case of hypocortisolism

If hypocortisolism is diagnosed, the next step is of particular importance. The aim of these additional tests is to pinpoint the cause of the hypocortisolism.

The following tests might be carried out:

- blood tests (electrolytes, glucose, cortisol, ACTH, aldosterone and renin levels, and general blood composition)

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- scan of the pituitary gland and/or the adrenal glands
- genetic testing, if indicated.

Blood tests (electrolyte, glucose, cortisol, ACTH, aldosterone and renin levels and general blood composition)

In the case of primary adrenal insufficiency, there is a very elevated level of the hormone ACTH in the body. Besides this, other deviations are often found in the blood such as reduced aldosterone, elevated renin and reduced DHEA-S. A reduced level of sodium and an elevated level of potassium is often found in the blood. That will prompt tests to check for the presence of antibodies working against the adrenal glands. If such antibodies are present, i.e. the antibody test is positive, this indicates Addison's disease.

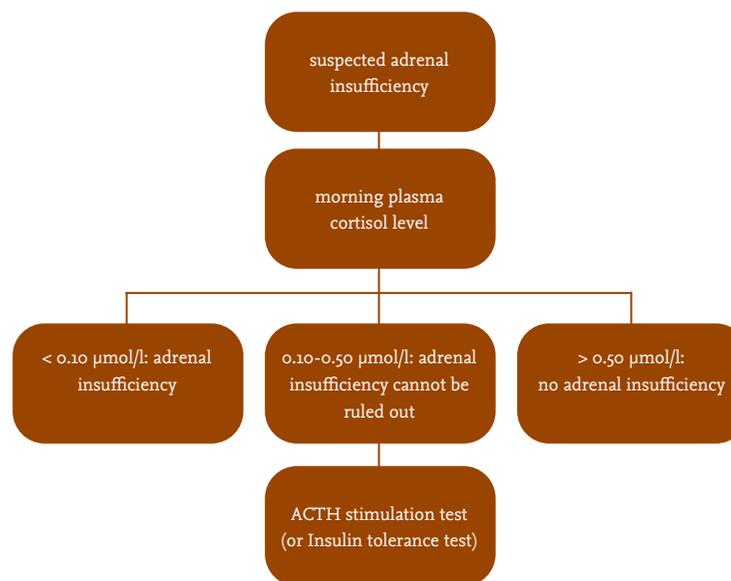
Scan of the pituitary gland and/or the adrenal glands

If no antibodies are discovered, a CT scan of the adrenal glands will be carried out. This will make it possible to check for the presence of calcification (often seen in patients with TB), bleeding, tumours or other abnormalities of the adrenal glands. If central adrenal insufficiency is suspected, an MRI scan of the pituitary gland will be carried out to check whether there is a tumour present, any inflammation or bleeding.

Genetic testing, if indicated

Genetic testing can be carried out when multiple cases of endocrine autoimmune diseases occur within a family group. You should discuss this with the physician treating you.

On the basis of the tests described above, an endocrinologist or other specialist can arrive at a precise diagnosis: adrenal insufficiency. This is an incurable disease but the symptoms can be treated successfully. That is what the next paragraph is about.



Schematic representation of the tests. Source for cut-off cortisol values: Smans LC, Zelissen PM. Front Horm Res. 2016;46: 146-58

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What treatment is available in the Netherlands?

Although the treatment for the three forms of adrenal insufficiency is largely identical, there are a number of differences.

The focus of the treatment lies on supplementing the hormones that your body is no longer producing for itself.

For this reason, you will need to take medicine: hydrocortisone (in one form or another); in the case of primary adrenal insufficiency, you will take fludrocortisone as well. DHEA is also sometimes prescribed. Doctors will help you find a suitable balance. In stressful situations, however, you will need to take a temporary higher dose. There will be quite a bit of trial and error before a good balance is achieved.

The medicinal treatment of the three forms is described in more detail below.

Primary adrenal insufficiency

(problem with the adrenal gland(s))

- Supplement the hormone cortisol (hydrocortisone)
- Supplement the hormone aldosterone (fludrocortisone)
- Sometimes: supplement with androgens (Prasterone/DHEA)

Central adrenal insufficiency

(problem with the hypothalamus or with the pituitary gland)

- Supplement the hormone cortisol with medicine containing hydrocortisone
- Sometimes: also supplement other hormones such as:
 - thyroid hormone (levothyroxine)
 - sex hormones oestrogen/testosterone
 - growth hormone (somatotropin)
 - antidiuretic hormone (minirin or desmopressin), a medicine that imitates the effect of vasopressin.

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Steroid-induced adrenal insufficiency

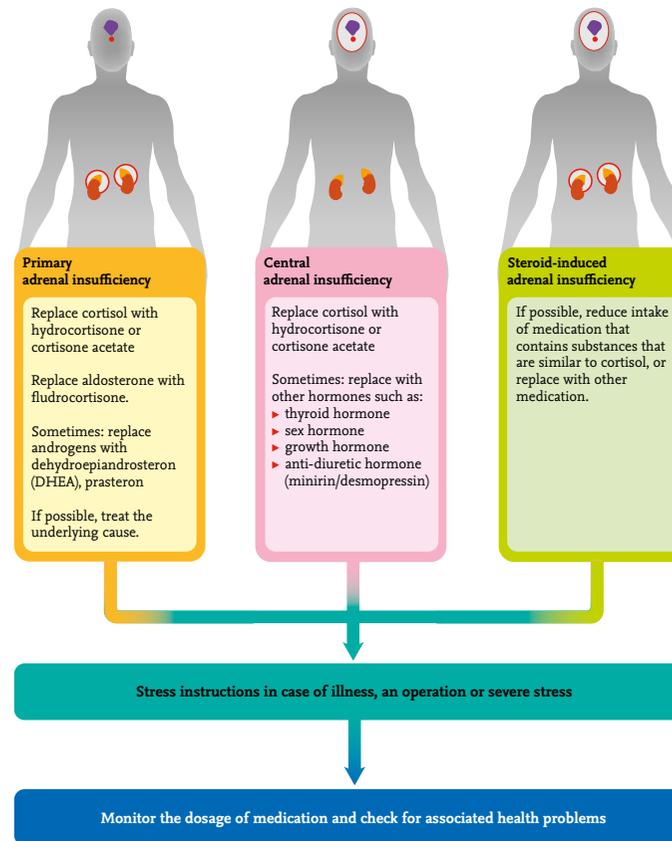
(problem caused by taking medication that contains substances that resemble cortisol)

- If possible, reduce intake of medication that contains substances that are similar to cortisol, or replace it with other medication.
- During the period that you are reducing the medication, you may be given hydrocortisone medicine.
- Even after you have reduced that medication, you may need to take extra hydrocortisone in times of stress.

In all the treatment regimes for adrenal insufficiency, taking the prescribed medicine(s) is of crucial importance. But there are other factors in play as well, because the hormones affect your entire well-being. You will start to feel much better immediately after the first dose of medicine.

After a while you will get to know your body better and together with your endocrinologist you will be able to decide on a personal hydrocortisone medication plan.

Another important aspect of the treatment is that you fully understand what the disease means and how it affects you, so that you can take action yourself in the event of illness, an operation or severe stress. In that way you can prevent an adrenal crisis. That will be the focus of the next paragraph.



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Adrenal crisis and stress instructions

An adrenal crisis is a potentially life-threatening situation for people with adrenal insufficiency.

An adrenal crisis occurs if people with an adrenal insufficiency have to cope with severe stress. Severe stress can occur when you are ill: when you have a fever, are vomiting and/or have diarrhoea for example. Attending a funeral or taking an important exam can also cause stress. During times of stress, your adrenal glands will not be able to produce enough **cortisol**, while your body needs more of it. It is then important to take extra medicine (hydrocortisone) as quickly as possible. If you are unable to do so, an adrenal crisis could occur.

If an adrenal crisis threatens, it is important that you do three things:

- The daily quantity of hydrocortisone must be doubled or trebled as set out in the stress instructions.
- When it is not possible to swallow extra tablets, or when they don't have the desired effect, a higher dose of hydrocortisone must be

administered by way of an injection into muscle tissue (IM) or an IV drip.

- You will also need to receive extra fluids via an IV drip.
- Once the crisis is dealt with, the underlying cause needs to be investigated and treatment started.

When your cortisol levels are too low, this can reduce your ability to make decisions such as when to call for help. If they are present, family or friends must step in and call a doctor and/or administer an emergency injection. Talk about this with close family members, or a good friend, and make sure they know what to do.

If the necessary measures are not taken promptly, a threatening adrenal crisis can lead to serious symptoms and problems such as reduced consciousness, low blood pressure, disruption to water and salt levels in the body, gastro-intestinal symptoms or even to death. Going through an adrenal crisis, or even the threat of one, can have a lasting impact on your life and increase the anxiety felt by yourself and your family and friends.

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The terms Addison crisis and adrenal crisis have the same meaning and are used interchangeably.



The [animated clip Addison crisis](#) provides a simplified explanation of the risk of an adrenal crisis and how to deal with it.

In order to be able to take action to prevent an adrenal crisis, your medical specialist or a specialist nurse will give you instructions. They will explain how to prepare an emergency injection (using a Solu-Cortef act-O-vial), how an adrenal crisis might occur and how you can recognize the danger. This is what is meant by the 'stress instructions'.

Stress instructions

The stress instructions help you take the necessary measures. They describe various situations and explain the changes you need to make (or don't need to make) to the amount of medication you are taking. In addition, the stress instructions include additional SOS-type measures so that it is more obvious to outsiders that you are dependent on hydrocortisone. Uniform stress instructions have been drawn up and these have been approved by the Dutch Association for Internal Medicine (NIV) and the Dutch Association of Endocrinologists. You can download the [stress instructions](#) here.

Example situations

Examples of stress situations are:

- illness with fever (high temperature)
- dental treatment
- taking an exam
- attending a funeral

The measures you need to take vary according to the patient and the circumstances, so you always need to consult the instructions you have been given. It is also advisable to make sure that at least one person from your immediate circle knows how to prevent an adrenal crisis. If you are no longer able to prepare and administer the injection yourself, that person will be able to help you and so avoid the long-term problems caused by delay. Actually experiencing an adrenal crisis can therefore have a significant impact on both you and any family and friends who are with you.

Reducing medication

You will also be given instructions about reducing the medication back to normal dosage levels after a crisis. You will do this over the course of 2 to 4 days, depending on the dosage and the symptoms. While you are reducing the dose, take your medication at the usual times.

If the total stress dose was less than 60 mg a day, you should reduce it by 10 mg a day until you are back on your usual maintenance dose.

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If the total stress dose was 60 mg a day or more, then you should reduce it by 20 mg a day until you bring the daily dose down to 40 mg, then by 10 mg a day until you are back on your usual maintenance dose.

For some patients, the rate of reduction needs to be slower: one step every two days. Consult your endocrinologist (or other medical specialist) or a nurse-specialist about this. If you have experienced a crisis in the past, it is possible that your doctor will recommend a slightly different reduction schedule.

Emergency injection

There are situations in which it is essential to immediately administer an emergency injection of 100 mg hydrocortisone. This would be the case, for example, if you are vomiting or you have watery diarrhoea (your medication will not be absorbed), or if you are only barely conscious.

It is then of vital importance that the injection is administered either into a muscle or under the skin. You will receive instructions for this as well, and if you have not already received them it would be advisable to ask your doctor or the nurse-specialist about it.



The animated clip about the Emergency Injection provides a clear explanation of how an emergency injection should be prepared. In the Netherlands, the emergency injection is available under the name Solu-Cortef act-O-vial.

If you have had an emergency injection you must always contact your specialist or endocrinologist to talk about the situation and decide what follow-up steps are necessary. Even if your family doctor has administered the emergency injection, it is always advisable to get in touch with your specialist or endocrinologist. The specialist will decide whether you need to visit the emergency room (A&E) at the hospital.

Visibility and recognition

You are dependent on hydrocortisone, and for that reason you need to take tablets. It is possible that you might find yourself in an unexpected situation in which you are unable to communicate your needs adequately. It is therefore important that it should be immediately evident that you are dependent on hydrocortisone.

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The following options exist:

- SOS emergency cards
- card holder to attach to car seat belt or shoulder bag
- In Case of Emergency (ICE) contact details on your mobile phone
- Adrenal APP on your mobile phone
- SOS medallion / bracelet
- letter from specialist/endocrinologist (in Dutch and/or English) to carry when you travel
- emergency kit containing emergency medication and written information (multilingual)
- Your family doctor can also play a role in the visibility of your condition. Together with the professional association of Dutch GPs and other primary healthcare providers, the Dutch Adrenal Society NVACP has developed a 'disruption flag'. Family doctors are being advised to include this as a 'keep on top' flag in your electronic patient record. This means that when you visit any after-hours GP post in the Netherlands, the duty doctor will immediately see in your record that you are dependent on hydrocortisone.

The specialist or endocrinologist will notify your family doctor about your disorder and explain the stress instructions.



Medical specialists are advised to add a standard sentence at the bottom of each letter to the patient's family doctor, stating that the patient is dependent on corticosteroids and that - in the event of an adrenal crisis or the threat of one - the measures set out in the stress instructions must be taken.

Arrangements have also been made with the Dutch ambulance service so that they know how to act if you need emergency treatment. The National Ambulance Protocol includes measures for acute adrenal insufficiency. It is important that ambulance crews can easily see or recognize that you have adrenal insufficiency.



There are a number of preparations you need to make if you travel abroad. A checklist has been published on the BijnierNET website (currently only in Dutch). The folder on Adrenal insufficiency and travel (Dutch only) also provides some useful information (Dutch, some English).

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APS-1 and APS-2

Two forms of autoimmune diseases can also lead to adrenal insufficiency. All the advice given in this brochure is also intended for people who are diagnosed with either APS-1 and/or APS-2.

In the USA, the disorder is known as Autoimmune Polyglandular Syndrome, and the abbreviation APS is used. In Britain, the disorder is known as Polyglandular Autoimmune Syndrome and the abbreviation PAS is used. Other abbreviations may be used in other countries

The cause of the problem lies in the fact that the body's autoimmune system reacts against its own organs, especially those organs that are involved in the production and distribution of hormones. This causes all sorts of abnormal hormonal reactions. These lead to symptoms of illness, which in turn lead you to visit your doctor. After various tests you might be told that you have APS-1 or APS-2.

Both diseases are explained briefly below.

APS-1 and the AIRE-1 gene

Autoimmune disease explained

Your immune system does not normally react against the body's own organs. The immune system is 'trained' to recognize 'alien' intruders, such as viruses and bacteria. The immune system is also responsible for the rejection symptoms following organ transplants: a donor kidney, for example, is recognized as being an alien organ (i.e. it was not produced by the body itself) and is then rejected. If the immune system falsely recognizes and attacks the body's own organs, we call that autoimmunity. Diseases such as rheumatism, lupus and multiple sclerosis are examples of autoimmune diseases.

Scientists suspect that most autoimmune diseases have a genetic basis. In the case of APS-1, it has been clearly demonstrated that the disease has a genetic basis. It has been found that when a person has two defective copies of a particular gene known as AIRE-1 (an abbreviation for autoimmune regulator), this disease will manifest itself. Anyone with one defective gene and one perfect copy will be a carrier of the disease, but will not become ill because of it. If both parents have one defective gene, their children can inherit

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the disease. The defective gene is found primarily (but certainly not exclusively) in Finland and within the population of Iranian Jews.

An in-depth study of the AIRE-1 gene is currently being carried out to help doctors better understand the cause of APS-1. A gene is the smallest unit of genetic code.

The AIRE-1 gene

From research into the AIRE-1 gene, scientists have learned that there is an abnormality if the person has APS-1. The sequence of information in the gene changes. As a result, the gene cannot carry out certain functions. The defective AIRE-1 protein allows cells to do things that would normally be 'forbidden', and this is what causes the autoimmune reactions.

APS-1 is not very common in the Netherlands, but that does not apply to APS-2.

APS-2

Doctors speak of APS-2 when endocrine autoimmune hormonal disorders occur in certain combinations. One such frequent combination is Addison's disease combined with a thyroid gland that works too slowly or too quickly and a vitamin B12 deficiency. There is also a known combination of diabetes mellitus and gluten intolerance, and combinations of other disorders are possible as well. You will be screened for such combinations, and if necessary you will be given additional treatment. Being aware of the possibility is also important.

It is important that you realize that you could also become affected by other disorders. The screening procedure helps to find effective treatment, but be aware of the possibility yourself. Other members of your family might also have a hormone disorder, an over- or under-active thyroid for example.

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Medication

Patients with both primary and central adrenal insufficiency are treated with hydrocortisone, because their own adrenal glands are no longer able to produce cortisol. Cortisol is of vital importance in our lives.

Given the importance of this medication, it is essential that you have access to an uninterrupted supply of hydrocortisone. You must always carry an extra supply of hydrocortisone for use in times of illness or severe stress. If you receive the medication in a Baxter roll, you should make sure that your pharmacist also supplies you with a separate quantity in a pill bottle.

Tablets

Hydrocortisone is now only supplied in tablet form in the Netherlands. In the past, doctors, patients, pharmacists and pharmaceutical companies had agreed that the capsules for each of the various doses would have a different colour. Instead of capsules, coloured tablets are now produced and supplied. These tablets are coated to mask the slightly bitter after-taste. Patients had complained that the original white tablets left a nasty taste which lasted for quite some time. Coated tablets reduce this problem.

We recommend that you discuss your medication requirements with your doctor, especially if you experience side-effects.

The emergency injection (Solu-Cortef act-O-vial) has a limited shelf life; it normally has to be used within 5 years. People who often take holidays in countries with a warm climate are advised to replace the vial each time they travel. You should also consider replacing the vial if you keep one in the glove compartment of your car. Ask your endocrinologist for a new prescription for an emergency injection. Solu-Cortef act-O-vial should be stored at room temperature, NOT in the refrigerator. The prescription will be marked 'manner of use known' or 'use as directed'. You can use an outdated emergency vial to practice preparing the injection, but do not inject it. It is important that you remain familiar with this skill.

The emergency injection is supplied complete with syringes and needles; your endocrinologist will write this on the prescription.

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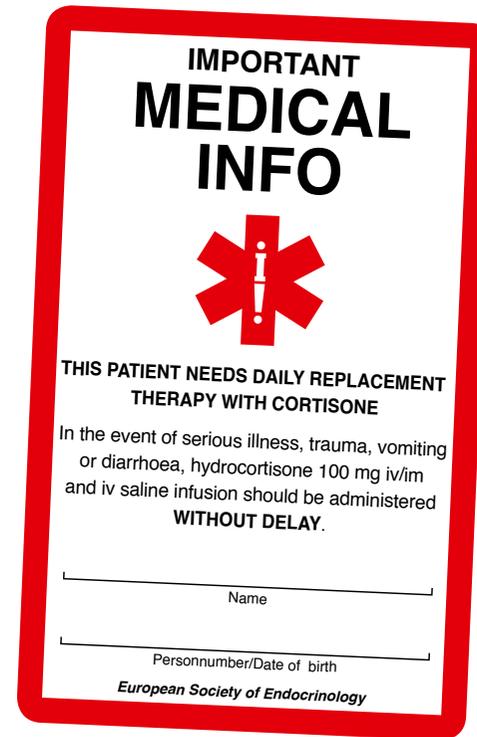
Your endocrinologist will also give you an extra prescription for the additional hydrocortisone pills to be taken in stressful situations. This prescription will be marked 'Take extra medication as necessary'.

Dutch citizens who take DHEA need to provide a letter of authorization once a year for a refund of the cost of the medication. The endocrinologist will fill in the application.

You should send this application to your health insurer. It is advisable to contact your health insurer about refunds before you actually start taking the medication.



The BijnierNET website includes a standard template that can be used for this purpose. If the insurance company fails to honour your authorization, and the costs are not refunded, you can lodge an appeal. Further information can be found under the theme [Medication](#) on the website (only in Dutch).



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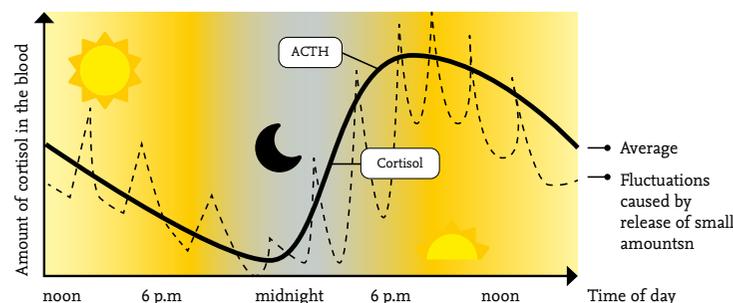
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Residual symptoms

The essence of the treatment of adrenal insufficiency is to counteract the shortage of cortisol. For this purpose you will take medicine in tablet form. Unfortunately, the treatment is far from perfect. Under normal circumstances, hormone production in the adrenal glands fluctuates at different times of the day and there are many factors that have an impact on the effect the adrenal hormones have. It is not possible to imitate this natural fluctuation with the medication we currently have available. You will notice this yourself, of course. Your experience and the symptoms are known as residual symptoms. Residual symptoms sometimes disappear, but that is not always the case. Then - unfortunately - they just become part of your illness.

Residual symptoms can partly be explained by the difference between the effects of the medication and your own personal and individual need for the hormone cortisol. The diagram illustrates that it is hardly possible to imitate the natural day-and-night rhythm.



Day-and-night rhythm of cortisol

The most common symptoms for people with adrenal insufficiency are tiredness, lack of energy and reduced vitality. Symptoms such as painful muscles and joints are often reported as well. Some symptoms have no connection with the adrenal insufficiency or the treatment. Sometimes it is not possible to find the cause of the symptoms. If you do experience residual symptoms, it is important that you discuss them with your doctor during your appointment.

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Symptoms can be caused by taking too much or too little hydrocortisone. This means that there will be **too much, or too little, cortisol** in your body. The problem is that having slightly too much or too little cortisol can lead to exactly the same symptoms (see page 28) A number of possible explanations are given below.

1. Hydrocortisone has a relative short half-life (about 90 minutes). This means that half of the active ingredient will already have disappeared from your blood 90 minutes after you take the medicine. Sometimes this means that you are undercompensating (undercompensating = insufficient dosage as a result of which a shortfall can occur) shortly before you take the next dose. The symptoms can be reduced by adjusting the timing and/or frequency of doses.
2. Other medication might be causing the hydrocortisone or cortisone acetate to break down more quickly, leading to under-compensation. Examples are: bosentan, carbamazepine, phenobarbital, phenytoin, primidone, rifampicin, rifabutin.
3. Not taking medication as instructed.
4. Modified working hours: the treatment needs be adjusted if you work evening and/or night shifts.
5. Sleep problems:
 - a. A high level of cortisol in the evening can give rise to sleep problems. This, in turn, can lead to tiredness. This symptom can sometimes be

relieved by reducing the amount of the evening dose, or by taking the last dose earlier before going to bed. There are, of course, other reasons why you might be having sleep problems. Some people sleep badly if the cortisol level is too low in the early hours of the morning.

- b. there can also be other reasons for the sleep problems.

Symptoms could be caused by a shortage of hormones other than cortisol, for example:

1. Secondary adrenal insufficiency: the pituitary gland might not be working properly
2. Primary adrenal insufficiency: associated autoimmune diseases, such as an under- or over-active thyroid, premature menopause, vitamin B12 deficiency, diabetes 1, etc.
3. Reduced androgens, especially in women: they feel less fit and have less energy. This occurs in combination with a reduced need for sex and feeling gloomy.

Some people who have residual symptoms and a reduced quality of life feel that adrenal insufficiency has a profound effect on their daily lives.



The series of mini documentaries (Dutch only) illustrates how different people cope with their illness. Having a look at them can help you get more insight into your own thoughts.

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Comorbidity

Once you are given the diagnosis 'adrenal insufficiency', the doctor, nurse-specialist or nurse will also give you information about the possibility of comorbidity. This subject will come up at each check-up: once a year for adults or every 3 months for children. If at any time between check-ups you experience symptoms that suggest comorbidity, it is recommended that you contact your doctor. He will then carry out additional tests.

People with adrenal insufficiency should be aware that they might be affected by the following forms of comorbidity.

- Possible shortages of (other) hormones
 - patients with central adrenal insufficiency can notice symptoms caused by a lack of other pituitary hormones
 - patients with primary adrenal insufficiency can also suffer from other autoimmune diseases.
- Possible residual symptoms and problems
 - tiredness
 - lack of energy
 - pain in muscles and joints
 - sleep problems
 - psychological problems
 - cardiovascular problems
 - greater chance of infections
 - osteoporosis (loss of bone density)
 - cognitive problems (e.g. memory, attention or concentration problems).

The following paragraph will focus on the psychological effects of the disorder and the treatment.

Comorbidity means that someone has more than one illness or disease at the same time, giving rise to additional symptoms and problems.

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Corticosteroids and the psyche

One of the most under-estimated aspects of having a shortage or a surplus of corticosteroids are the possible psychological consequences. Further information about this subject can be found on the [BijnierNET website](#).

Examples of the consequences of a lack, or particularly of a surplus of corticosteroids are:

- cognitive symptoms, such as finding it difficult to concentrate and memory problems
- mood problems, such as feeling gloomy or depressed, or being more emotional
- thought disorders, such as having phobias and/or unrealistic convictions
- personality changes.

You can recognize these disorders because people sometimes appear agitated and restless or indifferent, angry or sad, without any obvious reason, and they find it impossible to calm down and relax. This can make it seem as if that person is living in his or her own world.

There are similarities between the effects that both a shortage and a surplus in the hormone spectrum can have, and this makes it very difficult to gauge whether the hormones are being supplemented correctly, in quantities that suit the patient in question. And it's not only the patient himself who can experience a strangeness; their family and friends - such as partner, children or colleagues - can also notice changes. Support in the psychological and/or social domain is needed and could be part of the treatment offered, perhaps in the form of sessions with a psychologist or help from a social worker. As said before, the consequences associated with the disorder can also affect those dearest to you. It is therefore that any psychosocial support gives particular attention to those around the patient, so as to assure their well-being and keep a balance in their various relationships.



Watch the animated clip entitled '[the unknown effects of an adrenal disorder](#)' (Dutch only) or the mini documentary about '[relationships](#)'.

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Patient and Partner Education Programme for pituitary and/or adrenal disorders.

As a result of the disorder, patients can be confronted with changes in their daily lives. The aim of the Patient and Partner Education Programme (PPEP) for people with a pituitary and/or adrenal disorder is to provide patients and their partners with support in the psychological and social domain; this programme is currently offered by a number of Dutch hospitals.



PPEP Pituitary/Adrenal is a self-management programme that consists of eight weekly group meetings. Further information can be found here: [PPEP Pituitary/Adrenal in the PhD thesis written by Cornelia Andela.](#)

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On-going treatment (follow-up)

As there is no real cure for adrenal insufficiency, you will continue to be treated by an endocrinologist or other internal specialist. Once your medication has successfully been regulated, you will visit this doctor once a year. During your visit he will consider the following three questions with you:

- Is the treatment with medication working well, with as few problems as possible from under- or over-supplementation?
- Have you experienced any additional health problems?
- Do you need any non-medical or psychosocial assistance or support?

Patients who are being treated by a family doctor or another medical specialist should ask for an appointment with an endocrinologist or specialist in internal medicine at the hospital once a year.

A healthcare checklist has been developed especially for people with adrenal insufficiency. This checklist can help you prepare for your next appointment with the endocrinologist. Think of aspects such as:

- discussing the results of tests
- checking your current medication intake
- discussing any suspected side-effects or complications arising from the treatment
- have you experienced an adrenal crisis, and how did that work out?
- are you experiencing symptoms, limitations or health problems (either familiar or new)?
- discussing any issues that you are having to contend with and which reduce your quality of life. Consider things like memory problems, emotional outbursts, osteoporosis, pain in the joints and infections.
- information about possible additional tests or new treatments
- putting the stress instructions into practice

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- discussing any problems you experience at work, in other social contexts or within the family
- ask for extra support and assistance, if you wish to become pregnant for instance
- ask the specialist to provide your family doctor, and possibly other healthcare practitioners, with information about your situation.

AdrenalApp - diary

A diary option has been added to the AdrenalApp. By completing a checklist of 15 items on a regular basis, you can quickly see whether any changes have occurred. The items focus mainly on how you are feeling and on your quality of life. Examples of items are:

- I have experienced new symptoms
- I am feeling tired
- I am having trouble sleeping
- I am having mood swings

Unfortunately, the diary option only works for the Apple i-Phone at present.

If you were only diagnosed recently, it helps if you fill in this checklist every day.

If you have been receiving treatment for some time, and you have very few symptoms, you can just make a note of times when you have been under stress and describe your symptoms at those particular times.

You can print out the results and take them to your next appointment with the endocrinologist or nurse-specialist.

Experience has taught us that when you only have a check-up once a year, you are unlikely to be able to remember if, when or why you needed to increase your dose of hydrocortisone. The app's diary option can be a useful memory aid.

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How is patient care organised?

In the Netherlands, the various associations of healthcare practitioners, such as endocrinologists, and the patient associations have got together and drawn up **agreements about what constitutes 'good' healthcare** for people with adrenal insufficiency. These agreements have been combined in the Quality of Care Standard for Adrenal Disorders, which sets out quality criteria for good care.



Quality of Care Standard for Adrenal disorders (Dutch only)

They have agreed that it must be clear to you and your family (and perhaps close friends as well) who should be contacted during office hours - and how - if you encounter any problem or have any questions. Your contact person might be the endocrinologist himself, or a trainee doctor, or a nurse or nurse-specialist. Always ask for these details!

In the event of an acute problem, even if this occurs outside normal office hours, it must also be possible for you to consult a duty endocrinologist or similar specialist, trainee doctor or nurse-specialist. You will also be given the appropriate information for this purpose.

It has also been agreed that patients will be treated, by preference, in hospitals where endocrinologists work who have experience with adrenal disorders. Specifically: doctors who have experience in the diagnosis, treatment and care of patients with adrenal disorders and who provide care on the basis of the most up-to-date medical professional guidelines. Don't be afraid to ask the doctor treating you how much experience is represented in your hospital and your treatment team.

The hospital will carry out a periodic evaluation of the quality of care given to patients.

When they collect, record and transfer patient data, medical practitioners must respect the law, the rules governing their profession and national privacy regulations. Having said that, good care is dependent on an adequate exchange of data to and from primary healthcare practitioners, so a standardized template for medical records is essential. The family doctor of a patient with an adrenal disorder must be sufficiently informed about the patient's situation, because if the patient experiences any problems the family doctor is often the first person he turns to. Ask your family doctor to add a **'keep on top' flag to your electronic patient record.**

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Daily life

The consequences of having adrenal insufficiency differ from patient to patient and it is possible that you will experience little or no discomfort or hindrance from the disorder. But if you do experience negative effects that affect your daily life, they might also have a major impact on your partner, your family, colleagues and others close to you.

Issues that might play a role in this respect include:

- finding a job and/or keeping your job
- consequences for your social life
- the effect on your relationships, both intimate relationships and friendships
- your own personal development.

Stand up for yourself

Within the patient association, we like to encourage members to become articulate and conscious of their condition. You are welcome to join their ranks. It means that you will be an active participant in your health and healthcare alongside your endocrinologist. After a while you will be the person who knows your body the best; your healthcare professional is the one whose combined knowledge and experience give him insight into what usually works well in the treatment

of adrenal insufficiency. But you will find yourself confronted with unexpected incidents as time goes by and, as a result of having this disorder, they may affect you in a different way than someone who doesn't. So, remain alert and ensure that you can take the necessary measures or get someone to do that for you.



[Blog about the articulate and body-conscious patient](#) (only in Dutch)

Patient associations

Your healthcare professional can put you in touch with patient associations that you could join, so that you can have access to and benefit from the experience and information of other patients.

In the Netherlands, there are two patient associations:

- [Dutch Adrenal Society NVACP](#)
- [Dutch Pituitary Society](#)

Membership of a patient association brings a lot of benefits, so membership is a serious option for you to consider. In addition, the patient associations also act as lobbyists. They keep in touch with policy makers about adequate care, the supply of medicines and the

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provision of information about the various diseases. They also organise meetings so that patients can meet each other and/or learn about the latest developments. A number of Dutch health insurers will reimburse part if not all of the cost of membership of a patient association.

Work

A recent study brought to light that many people experience problems with work and relationships. People who have primary adrenal insufficiency can often continue working or become only partially incapacitated (NIVEL study, 2006). The problems mentioned above are more likely to arise in the case of Cushing's syndrome, particularly when the patient suffers from secondary adrenal insufficiency. On top of that, you might be facing an uphill struggle during the time when you are reducing your hydrocortisone intake until the adrenal gland(s) work normally once more. That also applies for people whose adrenal insufficiency is caused by taking medication that contains corticosteroids.

For many people, full participation in society is an important life goal. Work - a job - means:

- generating an income and, with it, economic independence
- personal growth and development
- building up and maintaining social contacts

- putting some structure in your life
- feeling that you are doing something meaningful or useful
- maintaining or acquiring social status
- improving your quality of life.

For the vast majority of people, having a job promotes their physical and mental well-being and improves the quality of their life. Research has shown that work has a positive impact on both the well-being and the recovery of people with a chronic disorder. It has to be assumed that this also applies to people with an adrenal disorder. One condition for work of any kind, however, is that the balance between the capabilities of the person and the demands of the job has to be carefully managed.

The endocrinologist or other medical specialist can play an important role in this respect by encouraging patients to discuss work-related problems. The endocrinologist helps the patient:

- to recover and maintain his health
- to become alert to signs of possible work-related health problems.

The issue of 'work' is an important aspect of healthcare. Discuss any questions you may have with your endocrinologist.

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If you are confronted with incapacity for work, whether complete or partial, there are various organisations that can help you.

This brochure is not the place for a detailed discussion of that issue, but any periods in which you have regular contact with an ergonomist (expert on labour matters), the doctors from the benefits agency, your own company doctor or the municipal welfare services are often accompanied by a great deal of stress, and that stress is not good for you. Be sure to make such people aware of that.

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Want to know more about adrenal insufficiency?

Further information about adrenal insufficiency can be found on the following websites (unfortunately, except for the first, they are mainly in Dutch):

- adrenals.eu
- www.bijniernet.nl
- [Dutch Adrenal Society NVACP](#)
- [Dutch Pituitary Society](#)

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The text of this brochure (Dutch and English versions alike) has been compiled with the greatest possible care. Despite this, they might contain errors. We request that you submit any additions or corrections to info@bijniernet.nl

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