



Great Ormond Street Hospital for Children NHS Foundation Trust: Information for Families

Cortisol deficiency and steroid replacement therapy

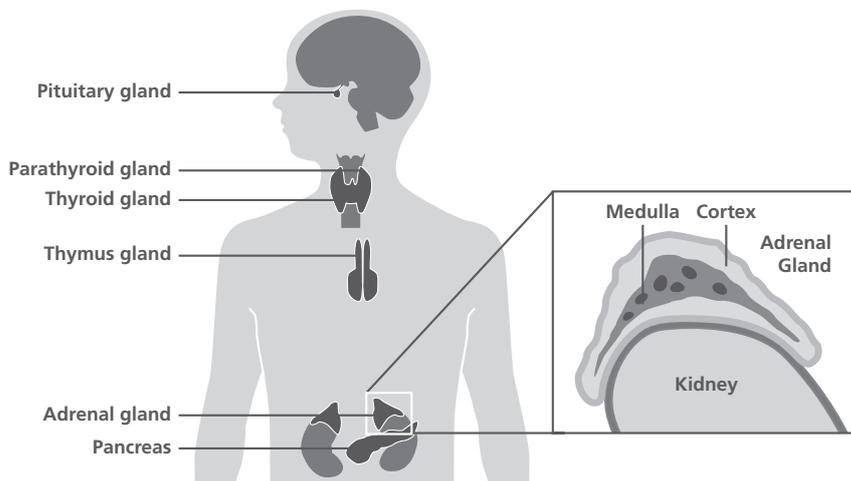
This leaflet explains about cortisol deficiency and how it is treated. It also contains information about how to deal with illnesses, accidents and other stressful events in children on cortisol replacement.

Where are the adrenal glands and what do they do?

The adrenal glands rest on the tops of the kidneys. They are part of the endocrine system, which organises the release of hormones within the body. Hormones are chemical messengers that switch on and off processes within the body.

The adrenal glands consist of two parts:

- the medulla (inner section) which makes the hormone 'adrenaline' which is part of the 'fight or flight' response a person has when stressed.
- the cortex (outer section) which releases several hormones.



The two most important ones are:

- **Aldosterone** – this helps regulate the blood pressure by controlling how much salt is retained in the body. If a person is unable to make aldosterone themselves, they will need to take a tablet called 'fludrocortisone'.
- **Cortisol** – this is the body's natural steroid and has three main functions:
 - helping to control the blood sugar level
 - helping the body deal with stress
 - helping to control blood pressure and blood circulation.

If a person is unable to make cortisol themselves, they will need to take a tablet to replace it. The most common form used is hydrocortisone, but other forms may be prescribed.



What is cortisol deficiency?

Cortisol deficiency occurs when the adrenal glands do not produce enough cortisol. This can happen for four main reasons:

- When the pituitary gland is unable to produce the chemicals needed to tell the adrenal glands to 'switch on' their cortisol production. The pituitary gland is the 'master gland' which controls other glands in the body.
- In a condition called congenital adrenal hyperplasia (CAH). CAH causes a blockage within the adrenal glands so they do not allow cortisol to be produced.
- If the adrenal gland itself fails (for example, in Addison's Disease) or is removed.
- The adrenal glands can stop producing cortisol because there are enough steroids in the body. The steroids should be withdrawn slowly to give the adrenal glands a chance to 'wake up' and start producing cortisol again.

For more information about the child's condition, please ask your doctor.

Cortisol deficiency is easily controlled with replacement therapy as hydrocortisone tablets given several times a day. However, if a person with cortisol deficiency becomes very stressed or unwell, either emotionally or physically, they are unable to increase the production of cortisol in their system to help the body cope and this could be life threatening.

In these circumstances, the amount of hydrocortisone given needs to be increased quickly. This is done by, either:

- increasing the dose of oral hydrocortisone taken as tablets

or by

- giving an injection into the patient's thigh (intramuscular hydrocortisone)

Increasing the dose of oral hydrocortisone is usually done by giving a double or treble dose, depending on how the child responds. If you have any concerns call your specialist treatment centre.

For instructions on giving intramuscular injections of hydrocortisone, please see our leaflet *How to give an emergency injection of Efcortisol®: Information for families*. You must always call an ambulance to take the child to hospital if they have had a hydrocortisone injection.

Does the medication have any side effects?

The doses of hydrocortisone and fludrocortisone are calculated to mimic normal production levels and so are unlikely to cause side effects. Side effects only tend to occur if someone takes steroids above the level usually produced by the body for a long period of time.



When to increase the child's hydrocortisone dose

It can be difficult to know when a child needs to increase their dose of hydrocortisone, but it may be necessary if they are ill, have an accident or injury or are going to have some medical treatments. The notes at the back of this booklet should help you decide. If you have any questions at any stage, contact your specialist treatment centre at Great Ormond Street Hospital or University College Hospital (contact numbers on the inside back cover) which provide a 24 hours/7 days a week service.

An extra dose will not do any harm, so if in doubt it is better to give it than not, and then contact the specialist treatment centre for advice about what to do next. If the child is given a hydrocortisone injection they must be admitted to hospital afterwards to be monitored. You should insist that this is for a minimum of 12 hours. This is because the injection lasts about 8 hours, and it is important to check that the child is stable on their normal oral dose before going home.

If your child is unwell, a further dose of hydrocortisone should be given at 4am – set your alarm clock! This should be double the usual morning dose.

It is important not to ignore diarrhoea and vomiting, especially if the child is also taking tablets such as fludrocortisone or DDAVP. These medicines can cause them to become dehydrated with imbalanced salt levels.

Vomiting

- It takes about an hour for oral hydrocortisone to be absorbed
- If the child vomits within an hour of taking the oral hydrocortisone, you should repeat the dose
- If the child is on milk feeds, it is important to try to give the medication one hour before the feed
- If the child is only sick once, an hour or longer after taking the oral hydrocortisone, you will not need to repeat it. However, you should think about why the vomiting has happened. It may be a one-off, or a warning of underlying illness
- Observe the child, and if they are sick again, give them double or treble the dose of oral hydrocortisone. Seek medical advice from your GP, local paediatric department, or your specialist treatment centre.
- If the child is unable to keep down the increased dose and/or is getting more unwell, you will need to give an intramuscular injection of hydrocortisone and call an ambulance to take the child to hospital immediately

Diarrhoea

- If the child develops diarrhoea, there is a risk that the oral hydrocortisone will pass through the gut too quickly and not be absorbed. You therefore need to double or treble the usual dose of oral hydrocortisone until the diarrhoea stops.



- You also need to seek medical advice to see if the child has picked up a stomach bug, which may need antibiotic treatment
- If the child has severe diarrhoea and is also vomiting, they will need hydrocortisone without delay. You should give the intramuscular injection and call an ambulance to take the child to hospital immediately

Coughs and colds

- If the child has a minor head cold with snuffles, runny nose and a cough but is otherwise well, there is usually no need to increase the oral hydrocortisone

Temperatures

- If the child has a temperature, this shows that they have some sort of infection and will need double or treble the dose of oral hydrocortisone. This will usually be for two to three days until the temperature is back to normal.
- If your child is unwell, a further dose of hydrocortisone should be given at 4am – set your alarm clock! This should be double the usual morning dose.

Antibiotics

- If the child has been prescribed antibiotics, you will need to give them double or treble the dose of oral hydrocortisone until the course of antibiotics is completed

Dentists

- If the child has a dental appointment for a check up or cleaning, they should not need any extra hydrocortisone
- If the child has an appointment for fillings or other treatments which require an injection, you should give them double or treble the dose of oral hydrocortisone for the 24 hour period around the appointment
- If the child unexpectedly needs an injection for fillings or other treatment, give them double or treble the dose of oral hydrocortisone as soon as possible, and continue for the next 24 hours
- Major dental work, like having teeth removed, should only be carried out in hospital. Please call your specialist treatment centre for advice before the appointment.

General anaesthetics

- If the child needs a general anaesthetic for any reason, they will need extra hydrocortisone given intravenously (IV) as they go to sleep. The endocrine registrars are available to advise your local team on the dose needed and can be contacted at your specialist treatment centre. Twenty-four hour advice is available for emergency admission.



Immunisations for childhood illnesses

- It is important that children who are on hydrocortisone replacement therapy have all their immunisations. If the child is taking medications other than hydrocortisone, you should ask your specialist treatment centre for advice
- You should give double or treble the dose of oral hydrocortisone for the 24 hour period around the injection, if you have any worries, please ring your specialist treatment centre for advice before the child is due to have the immunisation.
- Children may develop a high temperature and/or a rash several days after some immunisations, particularly the MMR vaccine. You should keep a close eye on the child after the immunisation and follow the advice under 'Temperatures' if they become unwell

Accidents and injuries

- If the child has a fall, bump or bruise, but immediately recovers and carries on what they were doing before, they may not need any extra hydrocortisone, but do contact your specialist treatment centre if you have any worries

- However, if they have a serious injury, for example, bump their head and become unconscious, break a limb or get burnt, you will need to give them an intramuscular injection of hydrocortisone and call an ambulance to take them to hospital immediately. The child may not necessarily need the extra dose of hydrocortisone, but it will do no harm. It is always better that they have the injection as more serious problems may occur if they do not get it when needed

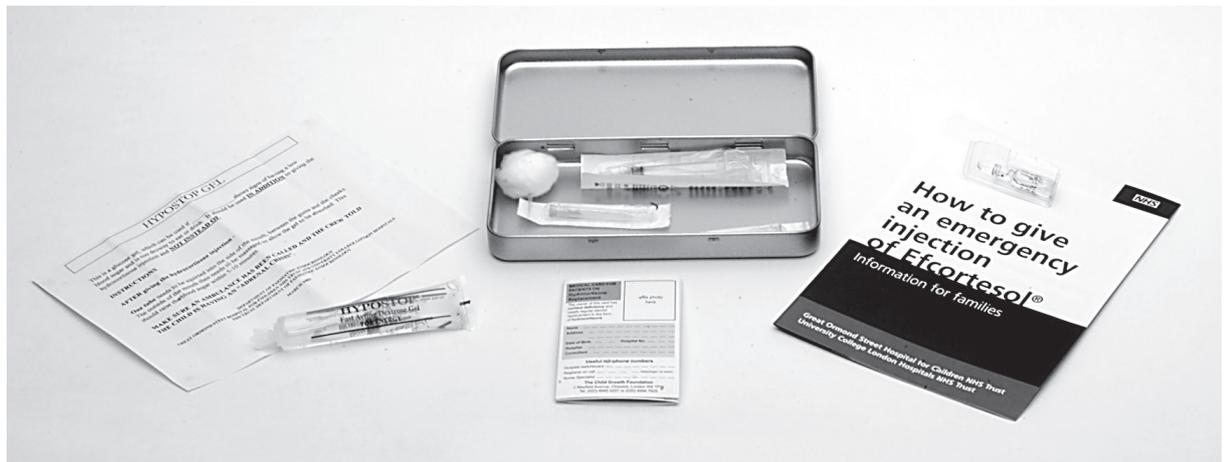
Unresponsive child/hypoglycaemia

- If for any reason you find the child with symptoms of hypoglycaemia (low blood sugar) for instance, they are:
 - pale
 - clammy
 - drowsy
 - confused
 - glazed
 - not responding as they would normallyyou should give them the intramuscular injection of hydrocortisone and call an ambulance to take the child to hospital immediately
- While you are waiting for the ambulance, you should give the child a sugary gel called Glucogel®. You give this by squirting the gel in the child's mouth between the gums and the inside of the cheek and then rub the cheek gently to help the gel become absorbed. You should never give an unresponsive child anything to eat or drink



Emergency kits

Your specialist treatment centre will issue you with emergency medication for your kit, one of which the child should carry at all times. Another should be kept at the child's nursery, school or college.



Each emergency kit should contain:

- 1x vial of hydrocortisone as Efcortisol®
- 1x 2ml syringe
- 2x blue needles
- 1x tube of Glucogel® gel with instructions for use
- 1x leaflet 'How to give an emergency injection of Efcortisol®'
- 1x steroid card filled in with child's details which you should keep in a specially labelled container.

You should check the expiry dates of Efcortisol® and Glucogel® and order replacements from your GP before they pass their expiry date.

Great Ormond Street Hospital for Children
NHS Foundation Trust

My Cortisol

Does your child carry an efcortisol emergency pack?

Download our free "My Cortisol" app

Available on the Apple App Store and Google Play Store



If you have any questions or queries, please call the following numbers:

Office hours (Monday to Friday 9am to 5pm)	Outside these hours
Clinical Nurse Specialist (GOSH) 020 7813 8214 (answerphone) or email endocrine.cns@gosh.nhs.uk	(GOSH) Call 020 7405 9200 and ask to speak to the 'On call Endocrinology Registrar'
Clinical Nurse Specialist (University College Hospital London) 0845 155 5000 ext 79204	(UCLH) Call 0845 155 5000 and ask to speak to the 'On-call Registrar for Paediatric Endocrinology'

Further information

We recommend that all children on steroid replacement therapy wear a medical identity bracelet or necklace at all times. Further information about these can be obtained from the clinical nurse specialists at either site, or the Health Information Centre at GOSH.

It is also important that they carry a 'Medical card for patients on hydrocortisone replacement' or a 'CAH steroid replacement' card at all times. You can get these cards from the clinical nurse specialists at either site or from the support organisations.

Special note for children with diabetes insipidus who are taking DDAVP

If the child needs:

Extra oral hydrocortisone for vomiting and/or diarrhoea, you should:

- Give them double or treble the dose of oral hydrocortisone
- Do not give them any more DDAVP
- Allow them to drink if thirsty
- Take them to hospital for a blood test to check their plasma electrolytes

Intramuscular hydrocortisone for any reason, you should:

- Give them an intramuscular injection of hydrocortisone
- Do not give them any more DDAVP
- Call an ambulance to take them to hospital immediately

Compiled by the Pharmacy & Endocrinology Departments
in collaboration with the Child and Family Information Group

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