

# Cortisol deficiency and steroid replacement therapy

Information for families

Great Ormond Street Hospital  
for Children NHS Foundation Trust

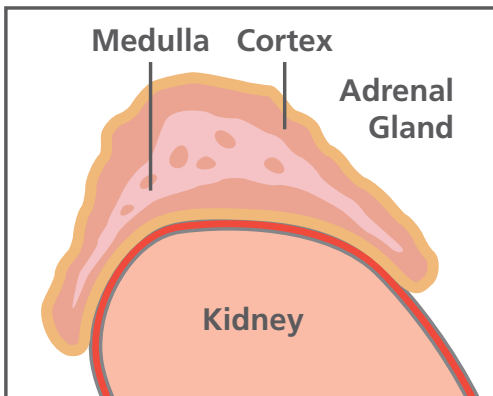
**This leaflet from Great Ormond Street Hospital (GOSH) and University College London Hospital (UCLH) explains about cortisol deficiency and about how to deal with illnesses, accidents and other stressful events for children on cortisol replacement.**

## **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 (diagram opposite page). Hormones are chemical messengers that switch on and off processes in 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 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.

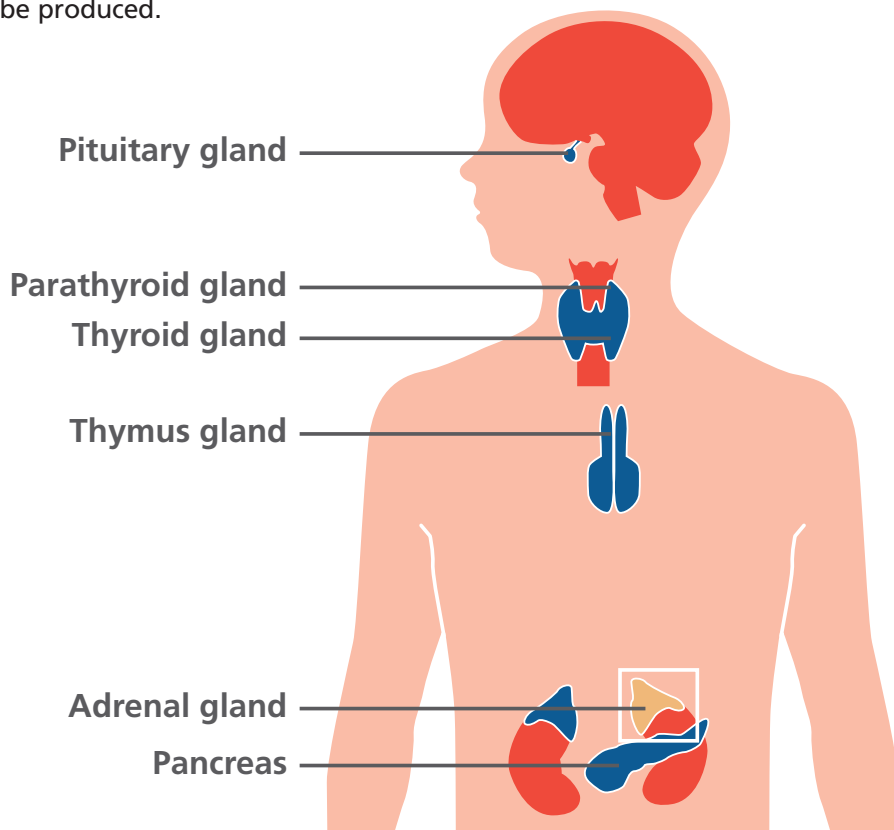
If the body does not make enough cortisol, a tablet version is given. This tablet is called 'hydrocortisone' and is given at regular intervals throughout the day. Without treatment your child will become very ill.

## 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.
- When there is a defect in the adrenal glands so they do not allow cortisol to be produced.
- If the adrenal gland itself fails or is removed.
- If adrenal glands stop producing cortisol because there are additional steroids in the body. The replacement steroid medicines should be withdrawn slowly to give the adrenal glands a chance to 'wake up' and start producing cortisol again.

For more information about your child's condition, please ask your doctor or nurse specialist.



## When to increase your child's hydrocortisone dose

If a person with cortisol deficiency becomes very unwell, they are unable to increase the production of cortisol in their system, which is needed to help the body cope and which 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
- giving an injection into the child's thigh (intramuscular hydrocortisone)

It can be difficult to know when your 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. Follow the flow chart you have been provided to help you decide if this is required

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 your child is unwell enough to require double dosing, an additional double normal morning dose of hydrocortisone should be given at 4am – set an alarm to remind you.

If your 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 effects of injection only lasts about six to eight hours and it is important to check that the child is stable on their normal oral dose before going home.



**Hydrocortisone Flow charts** available on GOSH website; search for 'cortisol deficiency'; scroll to useful documents; can print off flow charts required.

## Diarrhoea and/or vomiting

- It is important not to ignore diarrhoea and/or vomiting, especially if your child is also taking tablets such as fludrocortisone or DDAVP (desmopressin). Diarrhoea and/or vomiting can cause your child to become dehydrated with imbalanced salt levels. Please follow your flow chart for treating diarrhoea and/or vomiting.

## Coughs and colds

- If your 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 your child has a raised temperature, this shows that they have some sort of infection and will need double dose oral hydrocortisone and the additional 4am dose. This will usually be for two to three days until the temperature is back to normal.

## Antibiotics

- If your child has been prescribed antibiotics, you will need to give them double doses of oral hydrocortisone and the 4 am dose until they are better. This can then be reduced to normal maintenance dosing. However, your child must still finish the course of antibiotics as prescribed even if they are feeling better.



## Dentists

- If your child has a dental appointment for a check-up or cleaning, they should not need any extra hydrocortisone.
- If your child has an appointment for fillings or other treatments which require a pain killing injection, you should give them double the dose of oral hydrocortisone for the 24 hour period around the appointment.
- If your child unexpectedly needs an injection for fillings or other treatment for broken teeth for

example, give them double the dose of oral hydrocortisone as soon as possible, and continue for the next 24 hours.

- Major dental work, such as having teeth removed, should only be carried out in hospital. Please call your specialist treatment centre for advice before the appointment.

## General anaesthetics

- If your child needs a general anaesthetic for any reason, they will need extra hydrocortisone given intravenously (IV) as they go to sleep. The endocrine registrars at GOSH and UCLH are available to advise your local team on the dose needed and can be contacted by your treatment centre. Advice is available 24 hours a day for emergency admissions.
- At GOSH, your child will have an alert attached to their file to remind staff they are on hydrocortisone. It is still important to remind staff in all centres that the child needs their medication given regularly, on time and that it should not be omitted unless adequate IV cover is given.



## Immunisations for childhood illnesses

Immunisations are not contraindicated while on replacement hydrocortisone therapy and we would encourage families to complete all routine immunisations.

- If your child is taking medications other than hydrocortisone, you should ask your specialist treatment centre for advice.
- You should give double 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 your 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 your child after the immunisation and follow the advice under 'temperatures' if they become unwell.

## Accidents and injuries

- If your 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, or show signs of shock/hypoglycaemia for any reason, you will need to give them an intramuscular injection of hydrocortisone and call an ambulance to take them to hospital immediately.
- Your 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.

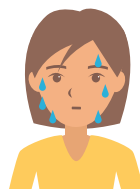


## Unresponsive child/ hypoglycaemia

If for any reason you find your child with symptoms of hypoglycaemia (low blood sugar) for instance, they are:

- pale
- clammy
- drowsy
- confused
- glazed
- not responding as they would normally

You should give them the intramuscular injection of hydrocortisone and call an ambulance to take them to hospital immediately.



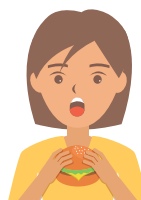
**SWEATING**



**PALLOR**



**IRRITABILITY**



**HUNGER**



**LACK OF  
COORDINATION**



**SLEEPINESS**

- While you are waiting for the ambulance, if your child is conscious you should give them a glucose gel. You give this by squirting the gel in their mouth between the gums and the inside of the cheek and then rub the cheek gently to help the gel become absorbed.
- If your child is unresponsive, never give them anything to eat or drink, including glucose gel.

## School packs

- We provide information packs for schools and are happy for schools to contact us for information. Please let the CNS team know about all changes of school.



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 doses of hydrocortisone and add in the 4am dose
- 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



## Emergency kits

Your specialist treatment centre will issue you with three emergency medication kits, one of which your child should carry at all times. Others should be kept at home and at your child's nursery, school or college.

You should make these up as emergency kits in a box or a tin that closes securely.

Each kit should contain:

- 1x vial of 100mg/1ml hydrocortisone sodium phosphate
- 1x 2ml syringe
- 2x blue needles
- 1x tube of glucose gel with instructions for use
- 1x leaflet 'How to give an emergency injection of hydrocortisone®'
- 1x steroid card filled in with child's details
- A copy of your ambulance protocol
- Some spare oral tablets of hydrocortisone

You should check the expiry dates of hydrocortisone tablets, injection and glucose gel and order replacements from your family doctor (GP) before they pass their expiry date.

## Further information

### Medical jewellery



- 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.
- If your child is just on hydrocortisone then the alert should say 'cortisol deficient; at risk of adrenal crisis'. If they are on other medications, please contact the Clinical Nurse Specialist (CNS) team for advice.

### Ambulance protocols

- With your permission your child will have an ambulance alert put in place for your home and their school address. It is essential you contact the CNS team to update any change to these addresses so the protocols can be resubmitted.



## App for your mobile phone

- **My Cortisol** – There is a free app available for Android and Apple devices to help with emergency care of children with cortisol deficiency.



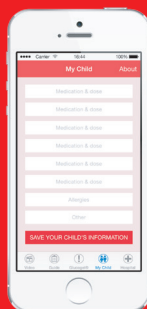
Great Ormond Street   
Hospital for Children  
NHS Foundation Trust

# 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**



## Travel

When travelling there are a few things to consider:

- Ensure you book in your child's travel vaccines with your GP and follow the immunisations advice earlier in the leaflet.
- You will need to request a customs letter. If you email [endocrine.cns@gosh.nhs.uk](mailto:endocrine.cns@gosh.nhs.uk) a minimum of two weeks before your holiday with all flight information including transfers, a customs letters and often an emergency protocol in the language of your destination can be sent to you.
- Ensure you have enough medication that if you needed to double dose the whole holiday you would not run short and add a few days extra in case of delays. This must be kept in hand luggage with an emergency hydrocortisone kit.
- When changing time zones, it is safest to give the normal morning dose then repeat eight-hourly (if on three times a day) or six-hourly (if on four times a day) until the morning of your destination's time zone and then commence at normal times and doses from that point.
- If your child is also on DDAVP please call the endocrine CNS team for advice
- Ensure your travel insurance company is aware of the child's cortisol deficiency as this will need to be registered as a pre-existing condition.
- Find out how to contact an ambulance and where the local hospital is located near your destination before you travel. If your child is unwell, you do not want to be finding out this information at that point.





## Results from admission

- Your child's results take time to collate and be reviewed. You will be contacted if changes to medications are needed. If no changes are needed, results will be discussed with you in your next clinic appointment.

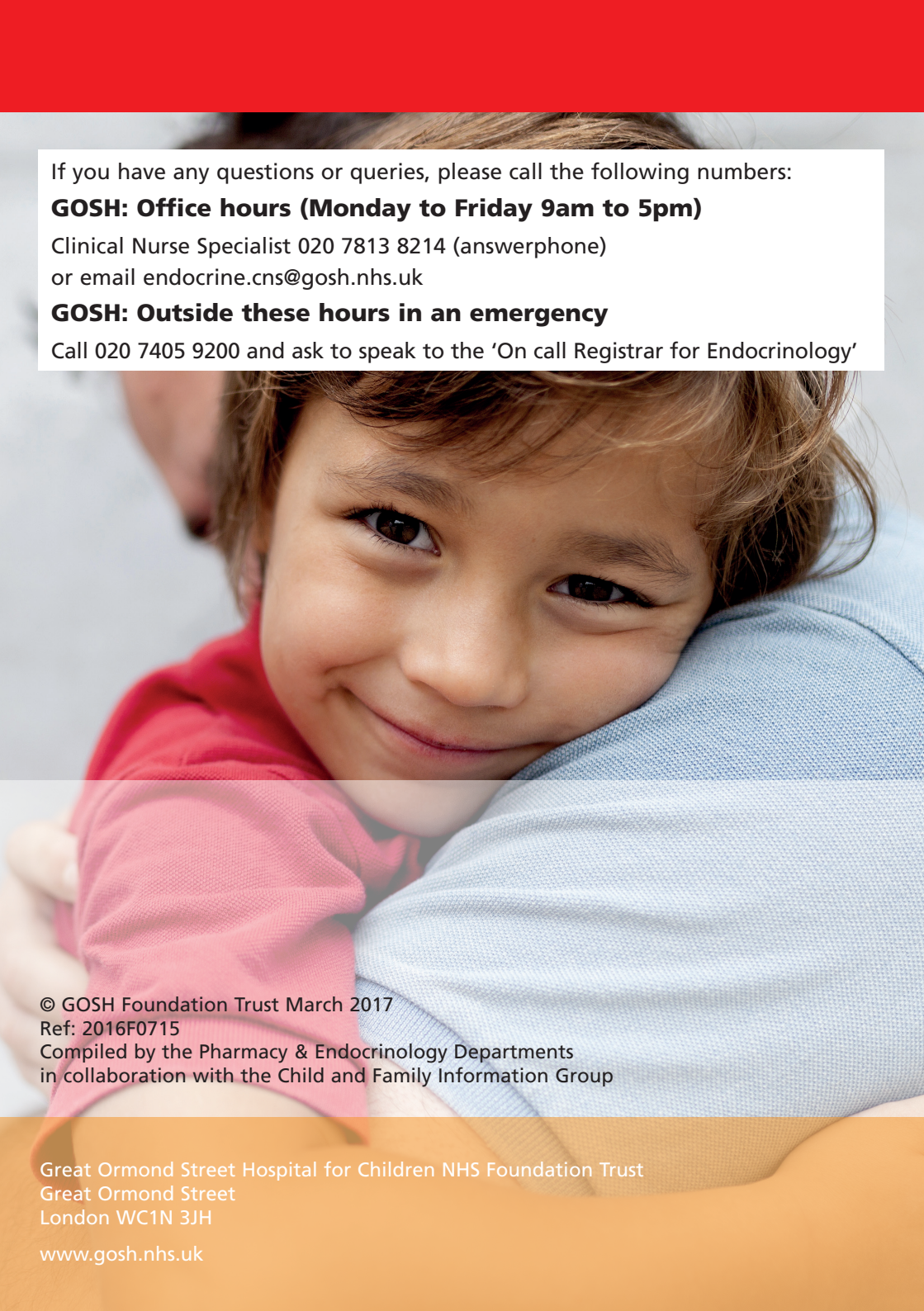
## Transition

- Many of our patients move to UCLH for their care once they reach adolescence. We work closely with the Endocrine team at UCLH to ensure transition is completed when it is appropriate for your child and happens as smoothly as possible. You will be given a leaflet introducing the service with contact information when the time comes.



## Notes

## Notes



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

**GOSH: Office hours (Monday to Friday 9am to 5pm)**

Clinical Nurse Specialist 020 7813 8214 (answerphone)  
or email [endocrine.cns@gosh.nhs.uk](mailto:endocrine.cns@gosh.nhs.uk)

**GOSH: Outside these hours in an emergency**

Call 020 7405 9200 and ask to speak to the 'On call Registrar for Endocrinology'

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Compiled by the Pharmacy & Endocrinology Departments  
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