

# **A Guide to Managing Adrenal Insufficiency and Steroid Replacement in Children and Young People**

## **Information for families**



## **A Guide to Managing Adrenal Insufficiency and Steroid Replacement in Children and Young People**

This leaflet explains about adrenal insufficiency (cortisol deficiency) and how it is treated. It also contains information about how to manage steroid replacement with illnesses, accidents and other stressful events in children.

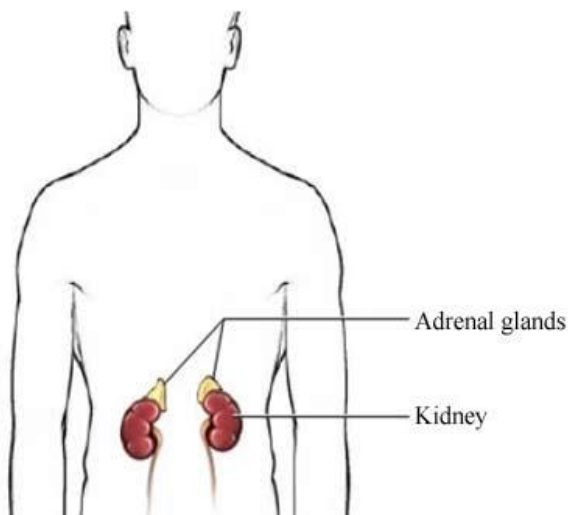
Steroid replacement is vital for your child to grow and develop normally. If the correct dose is used, there are no 'side-effects', as the treatment is merely replacing what the healthy adrenal cortex should normally produce. Individuals usually produce more cortisol in response to illness, injury and surgery. It is therefore very important that children and young people receiving steroid replacement therapy increase their normal dose of steroids to mimic this response.



## What are the adrenal glands and what do they do?

The adrenal glands rest on top of the kidneys. They produce important hormones (chemical messages) to control how the body works. One of these hormones is called cortisol. Cortisol is the body's natural steroid and has three main functions:

- helping to control the blood sugar level
- helping the body deal with physical stress
- helping to control blood pressure and blood circulation



If a person is unable to make cortisol (cortisol deficiency), they will need to take a tablet to replace it. The most common medication used is

hydrocortisone, but other forms may be prescribed.

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

If this happens, the amount of hydrocortisone given needs to be increased. This is done by, either:

- increasing the dose of hydrocortisone taken as tablets
- giving an injection into your child's thigh

**The information provided in this leaflet will guide you as to how to manage steroid replacement. However, if you are uncertain about your child's treatment, speak to your doctor or specialist nurse**

## General Advice

Your child has started on replacement steroids (hydrocortisone) as their adrenal glands are unable to produce enough cortisol. This medication is vital and should not be missed. Please ensure the following:

- Make sure that your child wears a steroid MedicAlert bracelet or equivalent (see picture). This lets others know in an emergency that your child is on steroids. It can be life saving in some cases
- That you and your child know the dose of hydrocortisone and that it is given three times per day (refer to your treatment plan – last page of this leaflet)
- Always have a spare supply of hydrocortisone available. Do not run out!
- As your child gets older, they will need a bigger dose of hydrocortisone. Your doctor will advise you about this.
- If you are uncertain about any aspect of your child's care and treatment, please ask your doctor or specialist nurse. It is important that you understand your child's condition and why they are taking steroids.



**MedicAlert bracelet**

## Knowing when to increase your child's hydrocortisone

When your child is unwell, has an accident or injury, or requires a medical investigation, they are likely to need an increase in their dose of hydrocortisone. An extra dose will not do any harm, so if in doubt it is better to give extra hydrocortisone and then contact Riverbank Ward for advice about what to do next.

If your child needs a hydrocortisone injection, then they should be taken to the nearest emergency department (A&E) immediately by paramedic ambulance for further assessment. If your child is less alert than usual, you should call a paramedic ambulance. The paramedics do not carry hydrocortisone for injection so if you haven't given the injection, please ensure you give the emergency kit to the

paramedic. Your child will be admitted to hospital for 12 hours. This is because the hydrocortisone injection takes about 8 hours to wear off and we need to ensure that it is safe for your child to go home.

## Vomiting

- If your child vomits within one hour of taking their hydrocortisone tablet, then give a second dose of hydrocortisone (repeat the last dose)
- If your child is sick again then give them their sick day dose or oral hydrocortisone. Seek medical advice early to ensure your child does not become more unwell
- With repeated/profuse vomiting, give the hydrocortisone injection and bring your child to hospital in a paramedic ambulance. Your child may need to stay in hospital to receive intravenous hydrocortisone until the vomiting has stopped.
- If your 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

## Diarrhoea

If your child develops diarrhoea, there is a risk that the oral hydrocortisone will pass through the gut too quickly and will not be absorbed. You therefore need to give them their sick day doses of oral hydrocortisone until the diarrhea stops.

If your child has severe diarrhoea and is also vomiting, you should give the intramuscular hydrocortisone injection and call an ambulance to go to hospital immediately.

## Infection and illness

**Coughs and colds:** If your child has a cough and cold without a temperature then there is no need to increase their dose of steroids. However, if you are uncertain about what to do, ask your doctor.

**High temperature and antibiotics:** Normal body temperature is 36.0 – 37.5°C. If your child has a high temperature due to an infection or is on antibiotics to treat infection, then give their sick day doses of hydrocortisone until 24 hours after the infection has cleared. If they are moderately or very unwell then increase their dose of steroids as instructed by the consultant. If your child is on antibiotics for longer than 10 days, ask your

doctor about how long to increase your child's dose of steroids. Also give your child an additional dose of hydrocortisone at 4 am at their sick day dose if they are very unwell. Always ask your doctor or specialist nurse if you are not sure.

**Chicken Pox:** Children on hydrocortisone replacement should take their sick day doses and also receive oral acyclovir (anti-viral medication). Acyclovir can be obtained from your GP.

How unwell	Hydrocortisone
<b><u>Mildly unwell</u></b>  Cold/cough without temp, can go to school	No change in dose
<b><u>Moderately unwell</u></b>  May keep off school, temp <38.5°C, may need an antibiotic	30mg/kg/m <sup>2</sup> in 4 divided doses
<b><u>Very unwell</u></b>  Off school, temp >38.5°C and needs an antibiotic, chicken pox	30mg/kg/m <sup>2</sup> in 4 divided doses

## Procedures

**Dental work:** If your child is having a dental check up or teeth cleaning they do not need to increase their dose of hydrocortisone. If your child needs a dental procedure that does not require a general anaesthetic, then give the sickness dose of hydrocortisone for 24 hours after the procedure. For dental work requiring a general anaesthetic, please see the information on general anaesthetics.

**Immunisations:** Children taking hydrocortisone should have all recommended childhood immunisations. If they are unwell or develop a temperature following their immunisation(s), then give the sickness dose of hydrocortisone until they improve.

**General anaesthetics:** If your child requires a general anaesthetic then they will need intravenous hydrocortisone which will be given before the operation or procedure (this includes MRI scans). Please ensure that the doctors and nursing staff know that your child is on steroid replacement. If their operation is in the morning your child should have their normal dose of hydrocortisone the night before their operation. If their operation is in the afternoon, your child should have their

normal morning dose of hydrocortisone. The specialist team will let you know what to do with your child's dose of steroids following their operation.

## Accidents and injuries

**Minor injuries:** If your child has a minor injury such as a bump, cut or graze from which they immediately recover and resume activities, then you do not need to increase their dose of hydrocortisone.

**Major injuries:** If your child has a significant injury e.g. fracture, head injury with altered level of consciousness or vomiting, or a serious burn, give the hydrocortisone injection and call a paramedic ambulance to bring your child to hospital.

**Unconscious:** If your child becomes unconscious, it is vital that you give the intramuscular injection of hydrocortisone immediately and then call a paramedic ambulance to bring them to hospital.

## Special considerations

**DDAVP (Desmopressin):** If your child is taking desmopressin (nasal spray, desmomelt, tablet) and needs to increase their dose of hydrocortisone because they are unwell, do not give further desmopressin until you have spoken to one of the

specialist team. Allow your child to drink freely. When your child is unwell, giving these medications together may result in a drop in their sodium level. They will therefore need to have their sodium level checked in hospital.

## Giving the hydrocortisone injection

The correct dose of hydrocortisone should be given by injection in emergencies. Remember that no harm can come from giving an unnecessary hydrocortisone injection. The dose that should be given is as follows:

- 25 mg for babies
- 50 mg for children aged 1—5 years
- 100mg for older children

The hydrocortisone solution should be drawn up into the syringe. The injection is given in the middle third of the outer side of the thigh where there are no important structures that can be damaged. The injection is given into the muscle (intramuscular). Your child's condition should improve quickly, but you should still take them to hospital if you have given the hydrocortisone injection.

Please use the pictures below as a guide to giving the hydrocortisone injection.

This photo shows what the ampoule and needle look like. If at all possible, wash your hands before starting.



### **Step 1: Breaking open the ampoule**

Locate the coloured dot on the side of the glass ampoule. Apply pressure to the dot and bend to break the ampoule. Use a tissue when breaking the ampoule to prevent cuts to your fingers.

### **Step 2: Drawing up the hydrocortisone**



- Open the syringe package

- Ensure that the needle is firmly attached to the syringe
- If the needle touches any surface including your hand it is no longer sterile & should be changed

### **Step 3: Flicking air out of the syringe**



It is very important to remove bubbles in the syringe. To do this hold the syringe at 90 degrees with the needle pointing upwards and tap the side of the syringe until the air bubbles have moved to the top. The air can then be removed by pushing up the syringe plunger until the air has passed through the neck of the syringe

#### Step 4: Giving the injection



- The injection should be given in the middle third of the outer side of the thigh
- Stretch the skin between your thumb and forefinger
- Hold the syringe at a 90 degree angle (like a pencil)
- It is important to hold the syringe straight to ensure that you inject into the muscle



- Push the needle into the skin with a firm quick action and inject the hydrocortisone
- Make sure to praise and reassure your child throughout the whole process. Apart from helping them to not feel scared, it

will also help to prevent tensing of the muscle and make the injection easier

#### Step 5: Call for an ambulance

Make sure that you ring for an ambulance or go straight to the nearest Accident & Emergency Department

#### My Cortisol app



Great Ormond Street Hospital have developed a free training app for giving an emergency injection of hydrocortisone. The app is a real time video showing the technique. The video is silent which solves concerns about non-English speakers using it and also if it's being used in a noisy environment. There is a tab with the still version of the procedure on it, a tab for parents to input doses, contact numbers and finally a tab with care instructions for a receiving A&E doctor. The My Cortisol app is available for both Apple and Android devices.

## Travelling and Hydrocortisone

Many of our patients now go abroad by airplane and so may encounter time differences between countries.

**European travel:** A time difference of 1 to 3 hours should not cause any problems. Do not change the time that you take your hydrocortisone

### To and from the USA

To:

- Morning dose as usual
- Usual second dose on arrival
- Evening dose before going to bed
- Normal treatment timings the next day

From:

- Evening dose as usual
- Normal treatment timings the next day

### To and from the Middle East/India

To:

- Evening dose as usual
- Half morning dose on arrival
- Normal treatment timings from the morning of arrival day

From:

- Evening dose as usual

- Repeat evening dose on boarding aircraft
- Usual morning dose on arrival in UK
- Second dose late afternoon
- Normal evening dose
- Normal treatment timings the next day

### To and from the Far East/Australia/New Zealand

For these journeys it is better to switch to an 8-hourly treatment programme.

To:

- Normal daily schedule up to departure
- Switch to 8 hourly regimen for flight duration

From:

- Continue 8 hourly until the morning after arrival then back to usual regimen

### Other things to remember before you go away

- Always carry extra tablets and do not put them all in the same place in case your luggage is lost/delayed
- Take and wear your Medic Alert Bracelet or equivalent
- Take your steroid/treatment card - either the standard

Department of Health Treatment Card, or the Hydrocortisone Replacement Therapy Card (Child Growth Foundation) or the Congenital Adrenal Hyperplasia Therapy Card (CAH Support Group). Make sure it is filled in and up to date.

- Your emergency supply of hydrocortisone for emergency use - make sure that it is still "in date." If not, request a new supply.
- Check that you have a sufficient supply of medication for the holiday duration. Getting medications abroad can be difficult sometimes and the formulations are not always the same.
- Don't forget your "Letter for Customs" as you will need this for traveling with needles and syringes. We can provide you with one of these.

## Dealing with emergencies abroad

The same sick-day rules apply with hydrocortisone:

- If your child is generally unwell then use your emergency sick day doses of hydrocortisone.
- If you need to give the intramuscular injection of hydrocortisone your child should be taken to the nearest hospital as soon as it is possible; let the doctor know that emergency hydrocortisone has already been administered and that your child is on steroid replacement therapy.
- Your child should not be discharged until the staff are satisfied that blood glucose and salt levels are normal.
- Remember to take your steroid card and medical details with you as this will help the local Doctors.



## My Treatment Plan

**Name:**

**DOB:**

**My Oral Hydrocortisone Replacement Dose is:**

Date	AM	MIDDAY	PM	MIDNIGHT
	mg	mg	mg	mg
	mg	mg	mg	mg
	mg	mg	mg	mg
	mg	mg	mg	mg

**Sickness Dose**

Dose	6AM	Midday	6PM	Midnight
mg	✓	✓	✓	✓

**My hydrocortisone injection mg expires:**

	Date	Date	Date
Home			
School			

## Useful contact numbers

Dr J West via Sarah Scotford (secretary)	+44 (0)1905 760734
Medical Doctor on-call (out-of-hours)	+44 (0)1905 763333
Riverbank Ward	+44 (0)1905 760588

**IN EMERGENCIES, PHONE 999 AND ASK FOR A PARAMEDIC  
AMBULANCE. DO NOT FORGET TO TELL THE PARAMEDICS THAT  
YOUR CHILD IS ON STEROID REPLACEMENT**

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### Acknowledgements

Worcestershire Acute Hospitals NHS Trust would like to thank the Endocrinology Team at Birmingham Children's Hospital for granting permission to use and adapt the adrenal insufficiency and steroid replacement patient information leaflet according to local service needs.