

A GUIDE FOR PARENTS/ GUARDIANS TO SUPPORT THE CARE OF THE CHILD WITH ADRENAL INSUFFICIENCY





Remember if you are unsure about anything about your child's care, please call a member of the endocrine team using the numbers below.

Contact details

Hospital: _____

Phone: _____

Doctor: _____

Phone: _____

Other useful numbers

Endocrine nurse specialist: _____

Endocrine appointments: _____

Out of hours contact number: _____

Family doctor: _____

Other: _____

Department of Endocrinology

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Introduction

Children who are adrenal insufficient usually require lifelong steroid replacement. During times of stress such as illness, trauma or surgery; they will require immediate treatment with extra hydrocortisone.

An adrenal crisis is a life-threatening emergency that requires immediate diagnosis and treatment.

This booklet aims to guide parents/guardians in managing their child with adrenal insufficiency during times of illness or stress, to promote optimal care and prevent adrenal crisis.

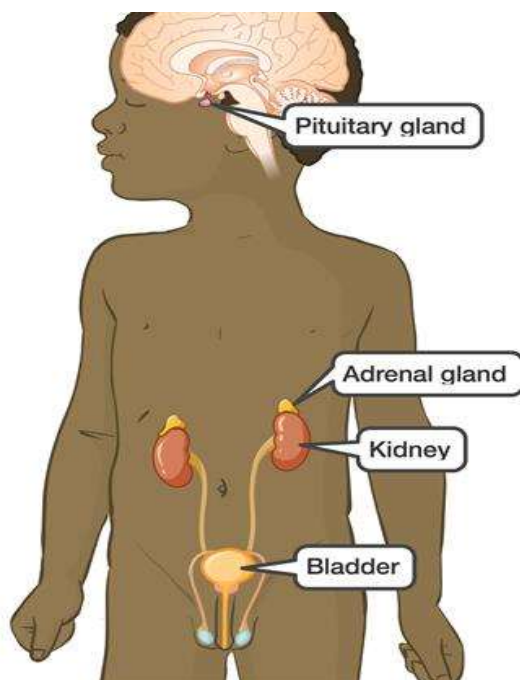
Where the adrenal glands are located and what they do?

The adrenal glands rest on top 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 certain processes in the body. The adrenal gland consist of two parts:

- The medulla (inner sections) 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 of which the two most important hormones are:
 - **Aldosterone** – This helps regulate 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:
 1. Helps to control the blood sugar level
 2. Helps the body deal with stress
 3. Helps to control blood pressure and blood circulation

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



Adrenal Insufficiency

Adrenal insufficiency is when the adrenal glands do not produce enough cortisol. This can happen for four main reasons:

1. When the pituitary gland is unable to produce the chemical 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
2. 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
3. If the adrenal gland itself fails or is removed
4. If the adrenal gland stops producing cortisol because there are additional cortisol-like steroids in the body, usually this is when a child is prescribed steroids for different medical reasons by a GP or paediatric doctor. The replacement steroid medication should be weaned slowly to give the adrenal glands a chance to 'wake up' and start producing cortisol again

When to increase your child's hydrocortisone dose / when to apply 'sick day rules'

The natural response of the body when unwell or faced with 'stress' is to increase the amount of cortisol that is produced. In a child with adrenal insufficiency this is not possible so extra hydrocortisone is required to mimic the body's natural response to 'stress'.

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

1. If your child can tolerate oral hydrocortisone, triple the daily oral dose
2. If your child is unable to tolerate oral Hydrocortisone, **hydrocortisone suppositories** can be given. They take effect after 10mins and the effect usually lasts up to 8hours. The endocrine team will inform you of the correct dose for your child
3. **Hydrocortisone intramuscular injections** can also be given if your child is unable to tolerate oral hydrocortisone. The endocrine team will inform you of the correct dose for your child

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.

An extra dose will not do them any harm, so if in doubt it is better to give a 'stress dose' of hydrocortisone. The following is a guide to increasing your child's oral hydrocortisone during illness or stress also known as **'SICK DAY RULES'**:

An Example of How to triple the oral dose of Hydrocortisone:

Add your child's total daily dose of hydrocortisone and multiply it by 3.

Divide this number by 4, this is the dose of oral hydrocortisone that needs to be given every 6 hours while your child is unwell.

For example: the child's daily dose is 20mgs in 24 hours.

AM		Midday		Night		Total Daily dose
7.5mg	+	7.5mg	+	5mg		= 20MG

$20\text{mg} \times 3 = 60\text{mg}$

$60\text{mg} \div 4 = 15\text{mg}$

15mg is the dose of oral hydrocortisone to be given every **6 hours** while your child is unwell

If required round each dose up to the nearest quarter tablet.

We recommend giving dose at 4am, 10am, 4pm and 10pm (or as near to these times as possible).

This mimics the body's natural rhythm, where cortisol concentrations begin to rise in the early hours of the morning. Continue to give this dose 4 times a day until the child is well.

If your child is unable to tolerate oral hydrocortisone then a hydrocortisone suppository or intramuscular injection of hydrocortisone should be given.

Recommendation for changes in dose / and when to apply 'sick day rules'

1. Vomiting and Diarrhoea

It is important not to ignore vomiting and/or diarrhoea. This may cause your child to become dehydrated and may cause low blood sugars. It is important to give regular sugary drinks if tolerated.

Vomiting

- If your child vomits within an hour of taking their hydrocortisone repeat the dose.
- If vomiting persists **triple the daily dose** of oral hydrocortisone as per 'sick day rules' if tolerated.
- If child is unable to keep down the increased dose of hydrocortisone tablets give a hydrocortisone suppository or intra muscular injection of hydrocortisone and seek immediate medical attention.

Diarrhoea

- If tolerating an oral dose of hydrocortisone **triple the daily dose** as per 'sick day rules'.
- If unable to tolerate oral dose give intramuscular injection of hydrocortisone immediately and seek medical attention.
It is not recommended to give hydrocortisone suppositories if your child has diarrhoea.

2. Coughs and colds

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

3. Temperatures

- If your child has a raised temperature (37.5°C and above), this shows that they have an infection and will need to **triple the daily dose** of oral hydrocortisone. If their condition is not improving, seek medical advice.

4. Antibiotics

- If your child is prescribed antibiotics, you will need to **triple the dose** of oral hydrocortisone until they are better and then reduce back to maintenance dosing. However your child must still finish the course of antibiotics as prescribed even if they feel better.

5. Dentists

- Please inform your dentist that your child has adrenal insufficiency prior to any treatment.
- For routine appointments, check-ups or cleaning, they should not require any extra hydrocortisone.
- If your child needs dental treatments such as fillings, dental extractions or major dental work, you will need to **triple the daily dose** of oral hydrocortisone for a 24 hour period around the appointment.

6. General anaesthetic

- It is important to inform medical team that your child is adrenal insufficient prior to undergoing a general anaesthetic.
- If your child needs a general anaesthetic for any reason, they will require extra hydrocortisone given intravenously by the medical team as they go to sleep.
- The endocrine team at CHI Crumlin are available to give your local team advice on the dose needed and can be contacted by your local treatment centre.
- Your child will have to remain on **triple the daily dose** postoperatively for approx. 24hours. You will be advised by the medical team on dose and duration required.

7. Immunisations for childhood illnesses

- Immunisations are not contraindicated while on replacement hydrocortisone therapy and we would encourage families to complete all routine immunisations.
- Children may develop a high temperature and/or rash several days after some immunisations particularly the MEN B and MMR vaccine. You should keep a close eye on your child after immunisation and follow advice under 'temperatures' if they become unwell.

8. Accidents and injury

- If your child has a fall, any bumps or bruises, but immediately recovers and carries on with what they were doing before, they may not need any extra hydrocortisone but you are advised to seek medical advice if concerned.

- However, if they have a serious injury, e.g. bump their head and become unconscious, broken bones, fractures or burns, they will need an **intramuscular injection of hydrocortisone**. You are advised to call an ambulance to take them to hospital immediately.
- Your child may not necessarily need an extra dose of hydrocortisone but it will do them 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

9. **Unresponsive child/hypoglycaemia**

If your child has symptoms of low blood sugars for instance, if they are:

- Pale
- Clammy
- Drowsy
- Confused
- Glazed
- Not responding as they normally would

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

- Also if your child is awake you should try to give them a sugary drink if they can tolerate it.

HYPOGLYCAEMIA



How to give hydrocortisone suppository

Suppositories are a form of medication that are inserted into the rectum (back passage) and they are used when it may be unsuitable to give medication to a child by mouth (vomiting, nausea).

This medication is designed specially to be given rectally and **SHOULD NOT BE GIVEN BY MOUTH.**

Do not give your child a suppository if they have diarrhoea

Administering a suppository:

- Suppositories can be administered in several positions.

Infant: lying on their back with the legs held upwards to ensure you can see the area clearly



Toddler / older child: The infant position can be used or lying on their side with the knees bent up towards the chest. Older children may be more comfortable to remain standing with one leg raised



- Remove the plastic wrapping from the suppository and lubricate the tip of the suppository with KY jelly/Vaseline



- Wash hands prior to giving suppository
- Gently insert the suppository into your child's **back passage** (the rectum) until the end of the suppository is no longer visible. Insert the pointed end first. Don't use any force when inserting suppository
- If possible, hold your child's buttocks together for a few minutes or ask them to lie still. This will encourage your child to hold onto the medication until it takes effect
- Medicines are absorbed via the rectum within minutes
- If you have inserted the suppository far enough into the back passage, it should not be passed. However, if the suppository is passed immediately after you insert it and it comes out intact, reinsert it

How to prepare and give intramuscular hydrocortisone injection



- Wash hands and check expiry date of medication.
- Hydrocortisone, Solu-Cortef® Powder for Solution for Injection, 100mg ampoule.
- Sterile water for injection 10ml ampoule.
- Syringe and needles.



1. Attach needle to syringe. Always keep guard on needle unless in use. If needle touches any surface such as your hands or table, it will no longer be sterile and should be changed for a new one.



2. Firmly break the top of the ampoule of water.



3. Remove guard



4. Insert the needle into water ampoule. Draw back plunger to pull water into syringe, 1mls of water is required to dissolve the powder. Discard ampoule



5. Push needle through rubber stopper of the Hydrocortisone vial



6. Gently inject water into the vial



7. Gently mix solution, then tip vial upside down with syringe still in place. Ensure that needle end is below fluid level to avoid getting air in syringe. Pull back plunger and draw up solution. Remove syringe from the vial.



8. Tip the syringe to point needle towards ceiling and gently tap side to dislodge any air bubbles to top of syringe, and then remove them by pushing plunger until the air has passed through the neck of the syringe



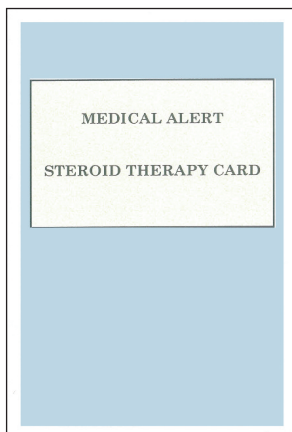
9. Divide the thigh in 3 parts and inject the medication in the middle third, within the upper outer area (as per picture). Stretch skin tight using thumb and forefinger. Hold syringe like a pencil and keep straight, push the needle into the skin with a quick firm action and push down on the plunger of the syringe to give the medication. Inject medication slowly: 1-2 seconds. If possible, leave needle in place for 5-10seconds after injecting the medication.

10. Discard used vial, needles and syringe into a sharps container



Essential requirements for children on steroid replacement therapy prior to discharge home

- Prescriptions to cover oral hydrocortisone tablets & supply of extra hydrocortisone tablets to cover 'sick days'.
- Prescription for Intramuscular Hydrocortisone ampoule (Solu-Cortef® Powder for Solution for Injection 100mg), 10ml ampoule for water for injection, syringes and needles with size and gauge indicated.
- Prescription for Hydrocortisone Suppositories and Guideline for administration.
- Reimbursement form signed and completed for hydrocortisone suppositories.
- Education competency checklist completed.
- **Medical Alert Steroid Therapy** – Your child needs to carry steroid therapy card at all times. You can get these cards from your Clinical Nurse Specialist (CNS).



- **Medi-alert bracelet-** we recommend that all children on steroid replacement therapy wear a medical alert bracelet or chain at all times.



- **My Cortisol** – there is a free app available for android and Apple devices to help with emergency care of children with adrenal insufficiency.



- **Emergency kit-** we recommend preparing a kit that contains equipment you will need in an event of an 'adrenal crisis' in a box or tin that closes securely.

TRAVEL

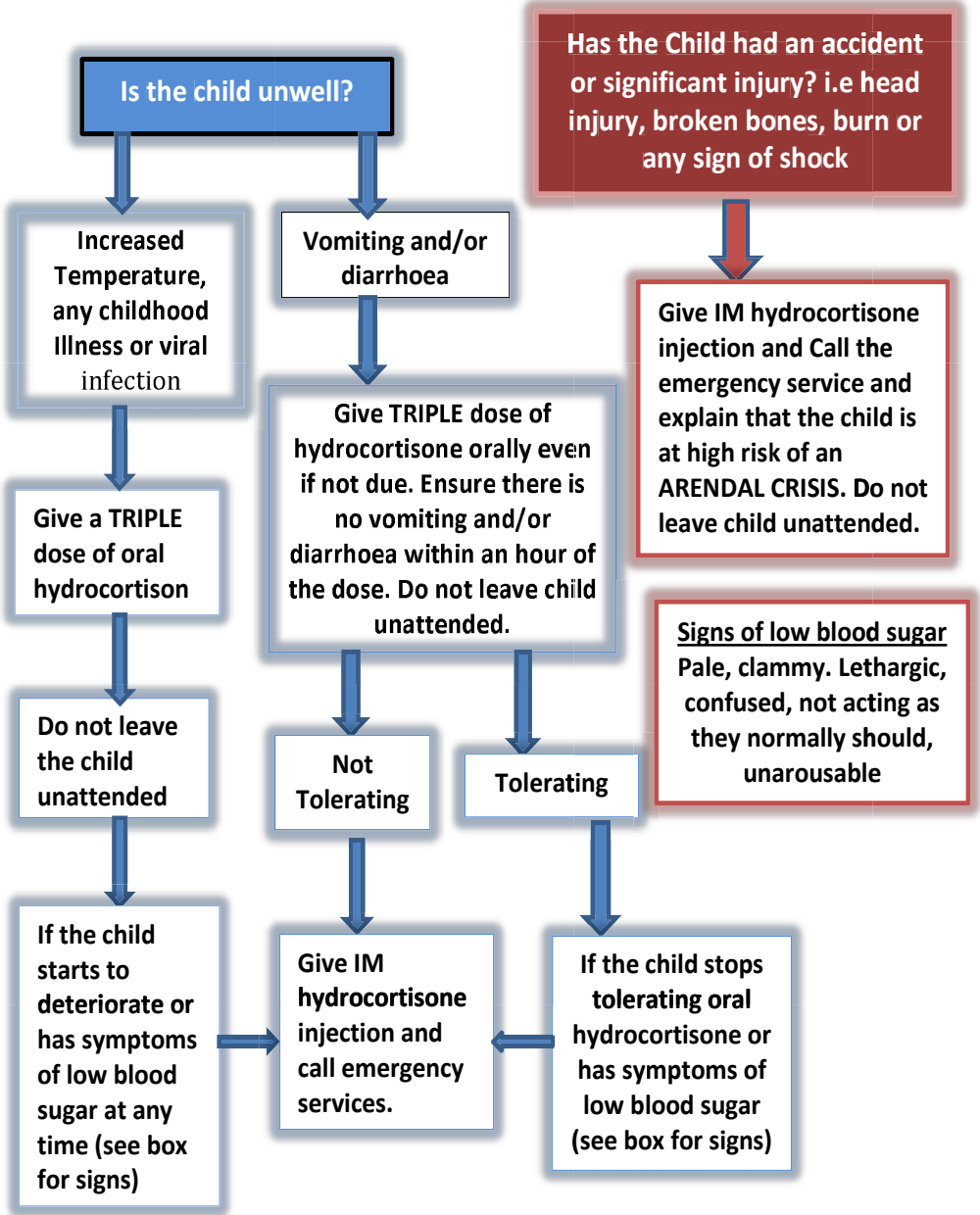
When travelling abroad there are a few things to consider:

- Ensure you book in your child's travel vaccines with your GP and follow the immunizations advice earlier in the leaflet
- You will need to request a letter for Security from the endocrine team well in advance of your holiday
- Ensure you have enough oral hydrocortisone for the whole holiday
- Bring an extra supply of oral hydrocortisone to cover triple dosing during the holiday in the event of any illness occurring
- Ensure to pack intramuscular injection and suppositories, and check all expiry dates
- Find out how to contact an ambulance and where the local hospital is located near your destination before you travel.





EMERGENCY PLAN FLOW CHART FOR CHILDREN WHO ARE ADRENAL INSUFFICIENT



Information for Teachers for Children with Adrenal Insufficiency
Student and Next of Kin contact

Name:			
Contact 1:		Phone no.	
Contact 2:		Phone no.	
Diagnosis:		Date of Birth:	
Medication:			

IMPORTANT MEDICAL INFORMATION

- Children who are adrenal insufficient usually require lifelong steroid replacement.
- During times of stress such as illness, trauma or falls with an injury or broken bones; they will require immediate treatment with extra hydrocortisone.
- Without treatment the child may become very ill suddenly. An adrenal crisis is a life threatening emergency that requires immediate treatment.
- If the child becomes unwell during school than the parents should be called to take the child home.
- The child should never be left unattended when unwell.
- If the child begins to deteriorate before parents arrive than an ambulance should be called and informed that the child is Adrenal insufficient and is at risk of having an adrenal crisis.
- When the child participates in any school outing, please ensure that all relevant medication is taken along and that at least two accompanying adults are fully informed of his/her condition.
- Adrenal insufficiency should not affect a child's day to day attendance and participation in school activities or outings.

