

## Signs of an adrenal crisis

Symptoms can vary in individuals but the following can be signs of an impending crisis:-

- \* Severe weakness.
- \* Dizziness.
- \* Floppiness.
- \* Failure to respond.
- \* Nausea and vomiting.
- \* Pallor and clammy sweating.
- \* Severe sweating with weakness.
- \* Cold hands and feet.
- \* Pins and needles or numbness in hands or feet.
- \* Tachycardia (fast heart beat).
- \* Hypotension (low blood pressure).
- \* Hypoglycaemia (low blood sugar).

## Glucogel for low blood sugar

- \* Glucogel® (used to be known as HypoStop) should be only used when you/your child is showing signs of low blood sugar levels.
- \* Glucogel® should be used **AFTER** you/your child has had the hydrocortisone injection.

## How to administer Glucogel

Gradually squirt the Glucogel® into the side of the mouth, between the gums and the cheek. Alternatively, squirt the Glucogel® onto your fingertip and apply it between the gums and cheek. Up to one-third of a 25g tube may be needed. Massage the cheek to allow the gel to be absorbed, this should raise the blood sugar level within 10 minutes.

## Important points

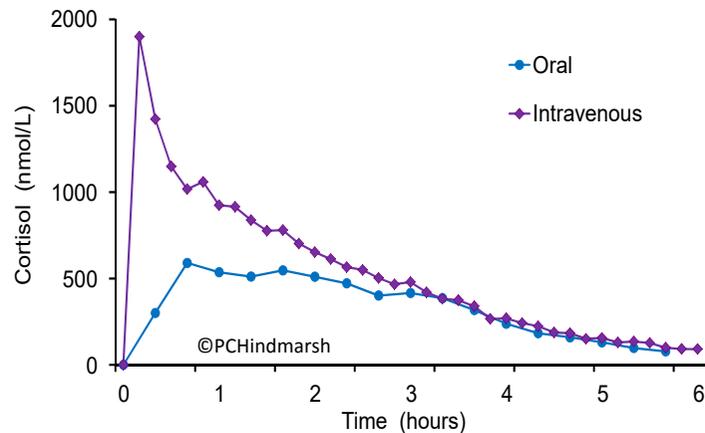
- \* The hydrocortisone injection will only last in the body, for 4-6 hours, depending on how long the body takes to use it up.
- \* It works quicker (peaks within approx. 20 minutes as opposed to approx. 2 hours for tablets) as it is administered directly into the muscle so it does not need to go through the stomach.

Note that a higher dose of hydrocortisone does not last longer in the blood stream than a lower dose, so it is not appropriate to give more than the recommended dose.

The reason it does not last longer is a bit complicated but it has to do with how the body removes medicines from the body and in fact adding more in, will cause the body to get rid of it quicker.

If too high a dose is given the body's natural response will be to excrete the excess as soon as possible.

The graph below shows the differences in the cortisol when measured in the blood after the same amount of hydrocortisone is given orally and IV.



**If you/your child should require emergency surgery a bolus of hydrocortisone must be given before surgery and protocol followed regarding hydrocortisone dosing during or after surgery. Please contact your endocrine team for advice on the protocol**

## Treating Diarrhoea

Diarrhoea is a particular problem due to the fluid losses. Again double/triple the dose of hydrocortisone is needed. Oral rehydration solutions such as Dioralyte should be used. It is important to make sure that you/your child is passing urine regularly.

Fludrocortisone dose must not be increased.

Seek medical advice early especially if there is:-

- \* A fever.
- \* Blood in the diarrhoea.
- \* You/your child becomes confused.
- \* The diarrhoea does not stop after 24 hours.

Do not use anti diarrhoeal drugs.

**ALWAYS REMEMBER: If in doubt it is safer to give the injection than not to give it, as it will do no real harm if given as a one off emergency dose, even if it is not really needed.**

Professor Peter Hindmarsh  
Professor of Paediatric Endocrinology  
Divisional Clinical Director  
for Paediatrics and Adolescents at UCLH  
<http://www.cahisus.co.uk>

*"The ideas expressed are independent of the editors' affiliations. Data provided is from current literature and should always be discussed with your endocrinologist"*



## Adrenal Hypoplasia Congenita

### A GUIDE FOR ILLNESS AND EMERGENCIES

**Professor Peter Hindmarsh**

*A handy reference guide in what to do in illness and emergencies. It is a good idea to keep your /your child's hospital emergency contact details in your phone.*

## WHAT TO DO IN ILLNESS AND INJURIES

We realise that when first faced with the diagnosis of you/your child having adrenal hypoplasia congenita (AHC) one of the most daunting and worrying issues is dealing with illness and emergencies, especially in babies and young toddlers who are not able to communicate on how they feel. We hope the following guidelines will help.

### So how do we deal with Emergencies and illness?

Remember that you know yourself/your child best but here are some steps, which we have put into Levels of illness that you can follow.

#### Level 1

- \* You/your child is unwell and has a temperature of 38°C or above.
- \* You/your child is taking fluids.

In this situation then the hydrocortisone dose should be doubled or trebled. If you are using twice a day hydrocortisone you may well need to increase the frequency of dosing to three times per day. This would mean adding in an extra dose at lunchtime and the amount would be double the dose that you use in the morning. To cover the early hours of the morning when the night time dose has worn off, we now advise an extra dose to be given around 4 am, this is to ensure there is cortisol around and will also go some way to preventing low blood glucose.

#### Summary

- \* Morning double/triple dose.
- \* Lunchtime double/triple morning dose.
- \* Bedtime double/triple dose.
- \* We recommend an **additional** dose equivalent to double the morning dose to be given at 4am.

Fluids that contain some sugar should be encouraged.

#### Level 2

- \* This would be same situation as in Level 1 but there is also associated vomiting.
- \* If you/your child has vomited 2-4 hours after the last dose of hydrocortisone then the last dose has probably been absorbed. But if you/your child vomits within 2 hours of the last dose:-
  - Give repeat double or triple dose orally. If this stays down, continue with double or triple dose orally.
  - If not and you/your child vomits it back within 2 hours then intramuscular hydrocortisone should be administered.

#### Intramuscular Hydrocortisone Doses For Emergency Use

Dose (mg)	Age range (years)
25	0 - 1
50	1 - 5
100	over 5

Following the administration of the intramuscular injection of hydrocortisone go to the nearest Accident and Emergency department as soon as it is possible and give a detailed explanation to the nurses and doctors that emergency hydrocortisone has already been administered. Show them your/your child's emergency letter and Adrenal Insufficiency Card.

You need to make sure that the medical team measure the following and you should not leave until you have the results:

- \* Urea and electrolyte concentrations to make sure salt levels are not low.
- \* Blood glucose concentration as this can go low.
- \* Blood Pressure which if low tells us if adrenal crisis is pending.

Should any of these levels be abnormal or in any situation of doubt then you/your child should be admitted for glucose, electrolyte and blood pressure monitoring along with possible need for intravenous fluids and hydrocortisone.

We recommend that after a hydrocortisone injection is given, a child should be admitted for observation for at least 12 hours.

The hydrocortisone injection should give cover for around 4 - 6 hours, so double or triple dose needs to be resumed after this time which will depend on how well you/your child has recovered, i.e. if you/ your child start to show signs of getting worse again after 4 hours, triple dose should be given.

#### Injuries and Accidents

- \* For mild bumps, scratches and bruises, there is no need to increase the dose.
- \* For sprains and other painful injuries, it will do no harm to give a double dose.
- \* For burns and serious injuries such as broken bones, head injury, severe fall with the possibility of a broken bone - give the intramuscular hydrocortisone injection.

If the patient is unconscious, give the hydrocortisone injection first, then dial 999 and state that patient/your child has 'Adrenal Insufficiency' See the leaflet on calling an ambulance for important information.

#### Why do we use the Intramuscular Injection?

It does sound drastic but like any person once they start vomiting it is hard for them to keep down fluids and in your/your child's case hydrocortisone. Also whatever makes them vomit may also interfere with the absorption of hydrocortisone from the stomach so the intramuscular injection bypasses this.

#### When to Call Ambulance:

You should call an ambulance when:

1. **AFTER** an emergency injection has been given and there has been no response.
2. If you/your child is involved in an accident.
3. There is loss of consciousness.
4. If you/your child is vomiting and cannot keep medicines down.
5. If you/your child is very unwell and you do not have an injection kit or unable to give the injection.

#### Calling an Ambulance:-

Many ambulance services in the UK are now following a 'pathways system' and each 999 call is triaged.

The 'pathway' followed for hypopituitarism and every other cortisol deficient disorder is:-

#### 'ADRENAL INSUFFICIENCY.'

The call handler will then ask for various details such as name, date of birth and what is wrong - it is most important that you state that the patient has **ADRENAL INSUFFICIENCY and NEEDS AN EMERGENCY HYDROCORTISONE INJECTION.**

**ADRENAL INSUFFICIENCY** is the key statement for the Ambulance Crew who will recognise it, respond accordingly and will act by giving emergency hydrocortisone.

At present if you have a protocol logged with emergency services this is flagged by the patient's address (not by their name) so it is important to make the emergency call handler aware that there is a protocol in place and to give the patients address if you have it.

When the ambulance arrives give a copy of our emergency letter or Adrenal Insufficiency Cards to the team.

Note that if an extra dose of hydrocortisone is given, this will not be harmful in any way.

#### On the Way to Hospital:

Give your endocrine team a call and advise them of the situation.

#### Medic Alert

Please note this reinforces the importance of wearing a medic alert with the wording **ADRENAL INSUFFICIENCY** and also carrying one of our Adrenal Insufficiency Cards.