



Emergency Treatment for Children with Cortisol and GH Deficiencies and Those Experiencing Recurrent Hypoglycaemia

PARENT INFORMATION PACK
(Including Emergency and School Information)



SERIES 1



SERIES 2



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SERIES 10



SERIES 11



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This booklet is intended to provide help when dealing with problems or difficulties associated with your child's condition and to provide information which will enable you to understand your child's treatment better and give you a basis for discussions with your child's specialist when necessary.

If you require further general information about *Emergency Treatment for Children with Cortisol and GH Deficiencies and Those Experiencing Recurrent Hypoglycaemia* you can contact the Child Growth Foundation.



THE CHILD GROWTH FOUNDATION

2 Mayfield Avenue
Chiswick
London W4 1PW

Telephones: 020 8995 0257 / 020 8994 7625
Fax: 020 8995 9075

EMERGENCY TREATMENT FOR CHILDREN WITH CORTISOL AND GH DEFICIENCIES AND THOSE EXPERIENCING RECURRENT HYPOGLYCAEMIA – Series 5 (February 1994) – Revised April 1996

Prepared by Rosemary Cordell (Child Growth Foundation). Edited by Mrs Vreli Fry (Child Growth Foundation) and Dr Richard Stanhope (Great Ormond Street Hospital for Sick Children and The Middlesex Hospital, London).

The British Society for Paediatric Endocrinology and Diabetes (BSPED) is an association of specialists who deal with hormone disorders in children.

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EMERGENCY TREATMENT FOR CHILDREN WITH CORTISOL AND GH DEFICIENCIES AND THOSE EXPERIENCING RECURRENT HYPOGLYCAEMIA

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INTRODUCTION

This information pack has been produced for parents with children who, for whatever reason, may require hydrocortisone or glucagon treatment during stress conditions such as hypoglycaemia, accidents or severe illness. This may apply to children with growth hormone deficiency (GHD), multiple pituitary hormone deficiency (MPHD), intrauterine growth retardation (IUGR) and congenital adrenal hyperplasia (CAH). For children with GHD or IUGR, the problem is usually low blood sugar levels (hypoglycaemia). For children with MPHD or CAH, the problem usually relates to cortisol deficiency, often in combination with hypoglycaemia.

It is hoped that this pack contains sufficient information to enable all those involved with the child's welfare to be confident that the condition is understood and that any problems which might arise can be handled effectively.

The pack aims to cater for all conceivable **possibilities**, NOT probabilities. It is important to emphasise that these children can and should be viewed as healthy and resilient in all normal situations but under stress conditions (accidents and severe illness) special precautions will probably be necessary.

We have designed this 'information pack' to be made available to both parents and school. The latter should be available in the pink section of the pamphlet.

For cross-referring to cortisol and hypoglycaemia in:

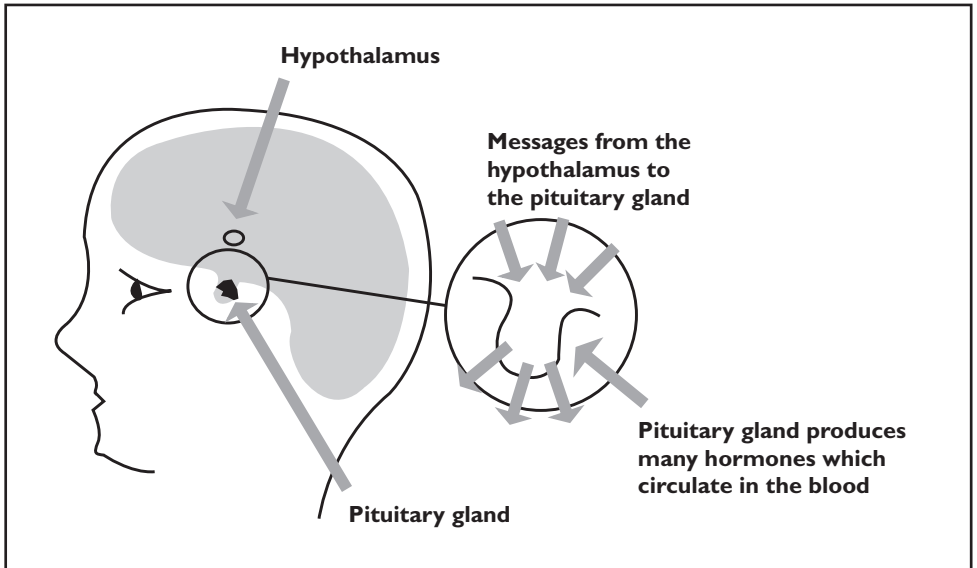
CAH	see Series No. 6	Congenital Adrenal Hyperplasia
IUGR	see Series No. 14	Intrauterine Growth Retardation and Other Low Birthweight Syndromes

A BRIEF EXPLANATION OF HORMONES

The **pituitary gland** is a small pea-sized gland at the base of the brain, which produces a number of hormones.

Hormones are the chemicals that carry messages from one cell to another via the bloodstream. The hormones produced by the front part of the pituitary gland include those responsible for growth (growth hormone), sexual development (gonadotrophins) and special hormones which stimulate the thyroid gland (thyroid stimulating hormone or TSH) and the adrenal glands (adrenocorticotrophic hormone or ACTH). The back part of the pituitary gland also produces a hormone (antidiuretic hormone – ADH) which regulates the amount of water passing through the kidneys.

There are many reasons why the pituitary gland might fail to produce hormones. The pituitary gland itself, or the brain connections to it, may have been damaged at birth or



by a severe head injury. Sometimes there are problems in the development of the foetus resulting in absence or underdevelopment of the gland. Sometimes brain or pituitary tumours or their treatment with surgery or irradiation cause failure of pituitary function.

When a child is lacking a particular hormone produced by the pituitary gland, he/she may be described as deficient according to that hormone, e.g. growth hormone deficiency. A deficiency in all the hormones means the child is Multiple Pituitary Hormone Deficient and the condition may be described by an abbreviation of this – MPHD (also called panhypopituitary).

All the hormones lacking in the hormone deficient child can be replaced by daily medication in the form of tablets and/or injections. Nowadays, careful monitoring of daily replacement doses effectively restores the normal growth and health of the child, who can lead an active and full life.

Most hormone deficient children are otherwise **normal** children – able to participate in all school activities. They should **not** be overprotected or treated differently from other children. Unfortunately, a few do have other disabilities affecting their schooling.

Hormone replacement therapy cannot be as efficient as natural hormone production where the body increases supplies of specific hormones when they are needed. Occasions arise, particularly when the body is under stress, when the child requires extra hormone medication **and this is perhaps the most important message in this information pack.**

There are two areas of concern for children with hormone deficiencies which may require emergency treatment with hydrocortisone or with glucagon during periods of stress or illness. These are:

- Hypoglycaemia associated with cortisol deficiency:
 - Congenital adrenal hyperplasia (CAH)
 - Multiple pituitary hormone deficiencies (MPHD)
 - Growth hormone deficiency (GHD)
- Other causes of hypoglycaemia
 - Growth hormone deficiency (GHD)
 - Intrauterine growth retardation (IUGR)

Different children will have different needs and these will have been explained to you for your child by your specialist. This pack aims to give practical information and advice on when and how to administer emergency treatments such as hydrocortisone and glucagon.

Hydrocortisone – normally given as tablets. However, if the child is unwell, vomiting or unconscious, treatment is given by intramuscular injection. The instructions for injecting are given on pages 11–13.

Glucagon – this acts to mobilise sugar stores from the liver into the blood stream, so helping to raise the blood sugar level. It is given by subcutaneous injection and in combination with the hydrocortisone to treat severe hypoglycaemia (low blood sugar). The instructions for injecting are given on page 14.

Note: In children with IUGR/Russell Silver syndrome, glucagon should **not** be given as they may not have sufficient store of sugar in the liver. In these children hydrocortisone only should be administered.

HYPOGLYCAEMIA (LOW BLOOD SUGAR)

Two hormones are important in the maintenance of normal blood-sugar levels: these are growth hormone and cortisol. When sudden illness or severe stress occurs, the body requires extra supplies of these hormones to keep up the level of blood sugar. If growth hormone or cortisol levels are not high enough, blood sugar levels may become low (**hypoglycaemia**) and the child feels very unwell and may lose consciousness.

Most children with only growth hormone deficiency do not have major problems with low blood sugar attacks but many children with deficiencies of both growth hormone and cortisol do have serious problems with blood glucose control.

Blood sugar (glucose) is essential for the brain to work properly. If sugar levels are too low the brain reacts by producing symptoms which may be *mild, moderate* or *severe*. Your growth specialist should instruct you how to recognise symptoms and, in some cases, to measure blood glucose levels at home.

Some of the stress factors which can cause hypoglycaemia are:

- Accident resulting in physical injury
- Infective illness, especially with a high temperature
- Vomiting (especially if hydrocortisone tablets are vomited)
- Missed meals
- Unusually prolonged extremely energetic activity
- Severe emotional stress

All children, of an age when they are separated from their parents, should carry a bracelet/necklace carrying details of their medical condition. If an emergency admission to hospital in an unconscious state is required, this information can be life-saving.

These can be obtained from:

MEDIC ALERT: 020 7833 3034

or contact the Child Growth Foundation

SYMPTOMS AND TREATMENT OF HYPOGLYCAEMIA

Mild hypoglycaemia

Children with slightly low blood sugar may feel dizzy, faint or hot but usually look pale. They may tingle, tremble, sweat, have a headache or palpitations.

The typical symptom of mild hypoglycaemia for

..... (print your child's name)

is

.....
.....
.....

(enter your child's symptoms or state 'no past history of hypoglycaemia')

At this stage hypoglycaemia should be treated with three glucose tablets (which should be available at school or carried by the child) or with Lucozade. Sugar, sweets or sweet drinks are also suitable alternatives and a glucose-gel called **Hypostop** may be given during such crises. Keep the child under observation until normal behaviour returns, then give two biscuits or a sandwich to maintain sugar levels until the next meal. If symptoms deteriorate, treat as **moderate hypoglycaemia**.

Moderate hypoglycaemia

Children with more serious hypoglycaemia also look pale and sometimes appear drunk, 'glassy-eyed', confused, unusually sleepy or very aggressive. These and any symptom which indicates a drift towards unconsciousness must be assumed to be due to moderate hypoglycaemia and if the two emergency injections are available, they should **BOTH** be administered without delay. (See instructions on hydrocortisone and glucagon administration.) If injections are not available, a strong sugar drink should be given (e.g. half a glass of Lucozade or Hypostop gel). If parents are confident that the child is recovering within 10–20 minutes, food can be given and the crisis may be over. However, if recovery is not complete or if the child vomits back the glucose, **a doctor or ambulance must be called**. Try to arrange admission directly to your nearest Accident and Emergency department as any delay may be serious. Once the acute problem has been sorted out, and if it is thought appropriate, transfer to your specialist unit could be arranged later.

Severe hypoglycaemia

In the unlikely event that the child is found in a coma and/or convulsing (fits), severe hypoglycaemia has developed. If available, appropriate staff or the parents must be summoned **immediately** to administer the two emergency injections – **before** transport to hospital. (See instructions on hydrocortisone and glucagon administration – pages 12–14.) Alternatively, glucose solution may be given rectally (i.e. via the bottom) and your specialist will advise you of the dose to be given as well as how to give it. **An ambulance must be called as hospitalisation is a matter of EXTREME URGENCY.**

Please note: For the MPHD or CAH child, severe hypoglycaemia is a very serious situation. Unlike diabetic children (who also suffer from hypoglycaemia), the hormone which is necessary to cope with the stress of hypoglycaemia (hydrocortisone) is not produced. Emergency admission to hospital will nearly always be necessary. Injection of glucagon and hydrocortisone will not necessarily remove the need for hospitalisation unless the carers are absolutely confident that recovery is rapid and complete.

Although more serious when it does occur, the MPHD or CAH child is not usually affected as frequently by hypoglycaemia as the diabetic child.

TO PREVENT HYPOGLYCAEMIA

When the child is under stress from illness

1. If the child is not feeling sick or vomiting, make sure food intake, particularly sweet foods or fluids, are taken every 2 hours during the day and perhaps once or twice at night.
2. If your child is receiving *growth hormone* treatment, make sure they continue their injections on a daily basis. An extra dose will do no harm and may successfully increase blood sugar.
3. Double or treble the usual morning dose of *hydrocortisone* and give this bigger dose **three times a day** until the child is well.
4. If your child is vomiting, give the emergency injection of *hydrocortisone* (25 mg, 50 mg or 100 mg intramuscularly) or ask a doctor to give it. The child should then be taken to the **nearest** Accident and Emergency Department.

Please Note:

5. Continued vomiting and hypoglycaemia **always** requires attention from a doctor. If you have been instructed in how to perform the injections, give the hydrocortisone and glucagon immediately, but also call a doctor. If in doubt, call the ambulance service to take you directly to the nearest Accident and Emergency department.
6. Once the situation is judged serious enough to have given the emergency injection of hydrocortisone, then the child should be transported to hospital.
7. If in any doubt, the hydrocortisone injection should be administered. In retrospect, if it is considered that such an injection was unnecessary, no harm would have been done by administering it.

ACTION IN EMERGENCY STRESS SITUATIONS

ACCIDENT resulting in unconsciousness

If the child has been involved in an accident and is unconscious, *glucagon* and *hydrocortisone* should be injected as quickly as possible and an ambulance should be called for emergency admittance to hospital.

ACCIDENT not involving unconsciousness

If the child is involved in a serious accident where severe injury results, *glucagon* and *hydrocortisone* should be injected to enable the child to cope with the possible results of stress, before he/she is sent to hospital.

If the child is involved in an accident which is not serious enough to need hospital treatment, please treat as for illness.

Please Note: In case of unforeseen accident, children with MPHD or CAH should wear an IDENTITY NECKLET/BRACELET.

ILLNESS without vomiting

If the child becomes ill at school, the safest treatment whilst waiting for the parents is to give three *dextrose* tablets or a *sweet drink*. If definite symptoms of hypoglycaemia develop, follow the guidelines under 'Symptoms and treatment of hypoglycaemia' (see page 5).

ILLNESS with repeated vomiting

If the child experiences repeated vomiting, the swallowed hormones may be lost. If *dextrose/sweet drink* and *hydrocortisone* treatment can be kept down, give this treatment, but if vomiting continues, the doctor or the hospital should be contacted for advice and *glucagon* and *hydrocortisone* should be injected (see pages 11–14). Glucose may also be given rectally which may be the preferred route if your child is fitting; your specialist will advise you on the dose and how to do this.

REMEMBER: It is not harmful to inject *glucagon* and *hydrocortisone* in a doubtful situation, whereas failure to inject when necessary could be very serious or even fatal.

SCHOOL

If your child has a history of frequent severe hypoglycaemia, it may be advisable to arrange for the administration of *glucagon* (not IUGR/RSS) and *hydrocortisone* in an emergency situation at school as well as at home. If the child's consultant considers school medical back-up advisable, the Local Education Authority should be contacted and the appropriate level of medical support in the school should be requested. It is not anticipated that this would be easily obtained but persistence can produce results. The Child Growth Foundation can offer advice on this procedure.

HOLIDAYS

Do not forget to take extra supplies of all the tablets and medicines. Do not forget the emergency injections. The stress and excitement of the holiday may result in your child needing more *hydrocortisone* than usual. Take a medical card (or letter from your specialist) to show doctors on holiday to explain about your child's treatment. For travelling through customs it is important to carry the medical card or a letter from your specialist to justify any drugs and, more importantly, the needles and syringes you may be carrying.

POINTS TO REMEMBER

For parents and teachers involved in the care of the child who has experienced hypoglycaemic attacks

- Glucose tablets or sugar should be available at all times. They must be easily accessible in the classroom and must be taken to swimming, sports sessions and outings.
- Regular eating is important – no meal should be missed. If a meal is rejected, some alternative, e.g. sugary drink, sweets, glucose tablets, must be taken.
- Do not keep the child late at school without informing the parents and ensuring that a snack is available.
- If you are the form teacher, please ensure that other teachers, helpers and dinner attendants know and understand the important points relating to the child's condition.
- When an MPHD or CAH child is unwell, he/she should never be sent to the secretary or medical room unaccompanied. If the child is sent home ill, it is essential that he/she is accompanied home and that a responsible adult is there to receive the child.
- Do not exclude the child from school outings but do ensure that at least two accompanying adults are fully informed of the child's condition.
- When the child participates in any school trip, do ensure that all relevant medication is taken along.
- MPHD and CAH children should always wear an identity necklet or bracelet or carry a medical card. The wearing or carrying of some medical identification is especially important when the child is on a school outing.
- If the child behaves abnormally, is seriously unwell or if frequent vomiting occurs, additional hormone injections or tablets may be required and he/she should be taken to the nearest hospital Accident and Emergency department.
- If in any doubt as to the condition or medical needs of the child and medical help is not 'at hand', **do not hesitate to call on the emergency services**. They would rather be called unnecessarily than not be called when treatment is essential.
- If emergency admission is required, it should be to the nearest hospital Accident and Emergency department/paediatric unit. Your specialist centre, in some cases, may be many miles away. To risk a long ambulance journey with a very sick child is a potential recipe for disaster. Telephone advice can always be obtained from the specialist centre and, if considered necessary, a transfer to the specialist centre can be arranged when the child's medical condition is more stable.

- Adolescents with MPHD or IUGR/RSS may develop hypoglycaemia with alcohol consumption, especially if vomiting occurs. Special counselling of such children on the dangers of excessive alcohol consumption should be given.

ADVICE TO PARENTS

It is important that you talk to the school when leaving the school information sheet so that the head teacher and staff clearly understand the needs of your child. In the case of large secondary schools you should consider re-visiting the school annually. If your child has experienced hypoglycaemia, the '**Points to Remember**' sheet should be included with the school sheet. (A second copy is included for this purpose.)

Where appropriate, please fill in your child's name and personal medical history including any symptoms of hypoglycaemia that have been experienced previously.

Please discuss this document with your own specialist, particularly any areas which are not clear to you. Remember that the Child Growth Foundation is always available to help with any problems or queries which may be raised by this document.

The medical back-up considered necessary by your child's specialist may not include all medications/injections mentioned in this pack. However, it is important to recognise that the possible problems described may become applicable to all MPHD and CAH children.

EMERGENCY DRUG ADMINISTRATION

Hydrocortisone

This hormone is vital in helping the body to overcome stress. It helps to keep the blood sugar and blood pressure at a safe level.

- *Hydrocortisone tablets*

Use these if the child is under stress, unwell or beginning to feel weak and sick. Double or treble the usual morning dose and continue to give this dose three times a day until the child is well and the stress situation has ceased.

- *Hydrocortisone injection*

Give this injection if the child is under stress and is beginning to vomit. If you cannot inject it yourself, ask a doctor to do it **urgently**.

Dose: Hydrocortisone 25 mg intramuscularly for babies/infants

Hydrocortisone 50 mg intramuscularly for children aged 1-5 years

Hydrocortisone 100 mg intramuscularly for children aged more than 5 years

Instructions: See pages 12–13.

Glucagon (NOT for IUGR/RSS children)

This hormone rapidly makes the body produce more sugar in the blood by releasing sugar stores in the liver.

Inject glucagon **before** hydrocortisone if the child is hypoglycaemic, then give hydrocortisone.

Dose: Glucagon 0.5 mg subcutaneously for children aged less than 10 years
 Glucagon 1.0 mg subcutaneously for children aged more than 10 years

Instructions: See page 14.

INSTRUCTIONS FOR INJECTIONS

Hydrocortisone

THIS SUBSTANCE SHOULD BE INJECTED INTRAMUSCULARLY.

Instructions for intramuscular injection:

Injection either 25 mg (for babies), 50 mg (for a child less than 15 years) or 100 mg (for older children) into the thigh as shown:

Injection

Intramuscular Injection

Stretch the skin tight using your thumb and forefinger. Hold the syringe like a pencil (**you must hold the syringe straight to make sure you inject into the muscle**) and push the needle into the skin with a quick firm action.

NOTE: Hydrocortisone for injection is also available as a ready-prepared solution. The dosage and method of injection will be the same as described above but the method of preparation will be different. Your doctor will advise you as to which is most appropriate for your child.



Divide the front part of the thigh into three parts between the hip bone and knee. Use the middle third on the side or front when the child is lying on its back

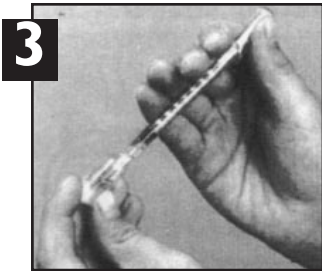
Hydrocortisone: preparation of substance



1 FIRMLY break the top of the ampoule of water.



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



3 Remove the guard and insert the needle into the water ampoule. Draw back the plunger to pull the water into the syringe. Discard the ampoule.



4 Push the needle through the rubber stopper of the hydrocortisone vial. Gently inject the water down the side of the bottle.

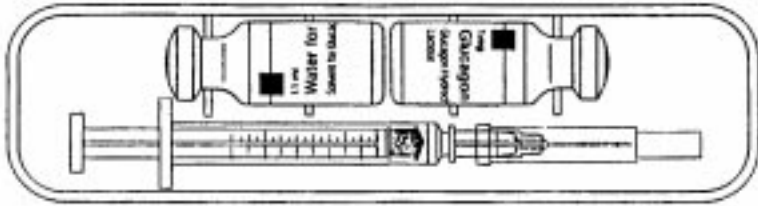


5 Make sure the solution has mixed then tip the vial upside down with the syringe still in place. Ensure that the needle end is below fluid level to avoid getting air in the syringe. Pull back the plunger and draw up the solution. Remove the syringe from the vial.



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

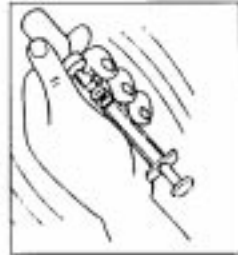
GLUCAGON (Hypoglycaemia Kit)



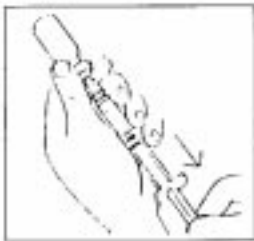
Use the disposable syringe provided to draw up the Water for Injections.



Inject the Water for Injections into the vial containing freeze-dried glucagon.



Without withdrawing the syringe, shake the vial until the contents are completely dissolved.



Draw up the solution into the syringe, check any air bubbles are removed, and inject it under the outer thigh or upper arm (subcutaneously). When the patient responds, administer oral carbohydrate (e.g. Dextrol, sweet biscuits or a sweet drink) to prevent a further 'hypo'.



Subcutaneous injection

Pinch the skin firmly between your thumb and forefinger. Hold the syringe like a pencil (either straight up or at an angle, whichever you prefer) and push the needle into the skin with a firm quick action.

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