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ACUTE ILLNESS

Dealing with acute illness in CAH exemplifies the fact that the parent is the expert. Parents are used to coping with CAH on a day to day basis and it is therefore very important that they take the initiative when their child is unwell.

Everyone suffers from colds, sore throats, coughs, vomiting etc. at some time. Children, especially babies, are more susceptible to these illnesses because their immune systems are less developed. One of nature's paradoxes is the response of cortisol to stress. On the one hand very large doses of steroids are needed to *dampen down* immunity and inflammation (for example in transplant recipients, sufferers from arthritis etc.), but on the other hand the body needs to make increased quantities of steroids to *combat* infection.

The problem in CAH is that the body cannot produce the extra cortisol required in response to stress, and this can result in the child becoming much more ill with an ordinary infection than would otherwise be the case. In young children especially this can cause blood sugar to fall (hypoglycaemia). In severe cases this can lead to unconsciousness and fitting. Another problem which can occur in children with CAH who do not have their steroid dose increased to cover stress is a worsening of the salt losing tendency, leading to vomiting and dehydration.

It is very rare for a child with CAH to die during an episode of acute illness. Far more commonly children are managed inadequately so that they are ill for longer than necessary, and require admission to hospital when with correct treatment this could have been avoided. Also, when hospital admission is necessary, this is often delayed.

The cornerstone of effective management of acute illness in CAH is for the parent **to increase the dose of oral hydrocortisone promptly** and to ensure that the child **receives an injection of hydrocortisone** if he/she is too ill to take the medicine properly, or keep it down because of vomiting.

WHEN DO YOU INCREASE HYDROCORTISONE?

Every centre advises slightly differently, and the exact "recipe" for hydrocortisone management isn't important. This is a rough account of the approach we advise in Glasgow:-

If the child has a mild illness (e.g. a cold) but is still well and able to play, go to school etc., then there is no need to increase the hydrocortisone dose.

For a mild to moderate illness, for example when the child has a cough and cold and is below par but still able to play, go to school etc., we advise doubling the daily dose.

For moderate to severe illness, in which the child is unwell and unable to play normally or go to school, then the dose should be tripled.

The increased dose is given for 1 to 2 days after the child has recovered. It should be noted that the fludrocotisone dose remains the same, it is only the hydrocortisone which is increased.

If the child is particularly unwell, and especially if there is drowsiness, vomiting or diarrhoea (which can interfere with the absorption of hydrocortisone) then an emergency injection is required. Unless there is a prompt response to this, the child should be taken to hospital.

N.B. When we say "double" the dose, what we mean is that twice the daily dose should be given. For example, if a child is on 5mg of hydrocortisone in the morning and 10mg in the evening equalling 15mg, then double the daily dose would be 30mg, perhaps given as 10mg, 3 times a day. This would be suitable for mild to moderate illness. A triple dose would be $15 \times 3 = 45\text{mg}$ daily, which could be given as 15mg, 3 times a day. In practice, parents learn to establish their own regime and become expert at judging by how much the hydrocortisone should be increased.

PARENTS MUST BE IN CHARGE

Time and time again, sensible parents who know perfectly well that their child needs a hydrocortisone injection or admission to hospital, face an inexperienced doctor who doesn't realise what CAH is all about, and somehow cannot manage the psychological hurdle of overriding the doctor and insisting on injection/admission. In Britain we have this extraordinary idea that the doctor knows best even when we know that, in certain instances, this is not so. This does not mean that family practitioners and junior medical staff aren't competent or caring. Rather, it is a reflection on the rarity of CAH and the unlikelihood of many doctors having treated a case, coupled with medical and parental difficulty coming round to the idea that in rare conditions especially, the parent knows best. Therefore, parents of children with CAH need to be informed, very polite and very firm!

The approach to acute illness should be backed up by written material from the hospital specialist and in Glasgow we have recently developed a CAH card for patients. The card gives details of current medication, what to do in acute illness, useful telephone and fax numbers (GP and hospital). It also has a section on what to do when the child is admitted to hospital, and this is most helpful for the junior staff and nurses.

HOLIDAYS

This is a notorious time for taking ill! Families must make sure that they take extra medication, a kit for injection and also a supply of paracetamol (avoiding over the counter medicines abroad which may contain aspirin, no longer recommended for children). Also, a letter from the hospital explaining the child's condition should be taken, with instructions as to what to do in emergency situations.

HYDROCORTISONE INJECTION

In Glasgow, the parents of newly diagnosed children are taught to give intramuscular hydrocortisone as standard treatment for 2 or 3 months, so as to completely familiarise them with the technique of injecting, which soon becomes second nature. If a parent does not feel confident about giving the child an injection, they must ensure that the injection is given promptly by the doctor or nurse.

TYPE OF MEDICATION

Hydrocortisone suspension

This causes quite a lot of confusion (between mg and ml) and there is uncertainty as to how consistent the dose of hydrocortisone is, especially if the bottle is not shaken properly. Also, the suspension has a short life. For these reasons we no longer recommend hydrocortisone suspension. 10mg hydrocortisone tablets can be divided and crushed up quite easily for younger children.

Hydrocortisone or other steroids (e.g. Prednisolone)?

Hydrocortisone is probably the best steroid for growing children and most adult males do fine on hydrocortisone. We change girls across to Prednisolone in the late stages of puberty and find that this gives them regular periods.

Whatever type of steroid is used, a minority of patients seem to get excessively hungry even on moderate doses, causing rapid weight gain. Sometimes the weight gain causes an increase in growth rate which can tempt the doctor to increase the steroid dose further, only making the problem worse. The specialist needs to draw a line between under-treatment and over-treatment and this can be very difficult.