

10 CRISIS MANAGEMENT

a) WHAT IS AN ADDISONIAN CRISIS?

“With Addison’s your body doesn’t react normally to trauma. So if you take risks and are severely injured, your chances of survival are more questionable than a person who has functioning adrenal glands. The potential of death is always there with Addison’s. I know this sounds strong but it is reality.” DIANA

An Addisonian crisis is what happens when an Addisonian faces extreme physical stress and does not get the extra steroid their body needs to meet that stress. It is a potentially life-threatening situation, which requires immediate emergency treatment.

The symptoms of extreme weakness, a serious drop in blood pressure and mental confusion will be familiar to many people from their experiences in the course of getting a diagnosis. A crisis is usually preceded by the symptoms of steroid insufficiency: headache, dizziness, nausea and vomiting.

Some people sail through life without ever experiencing a crisis. Others find they encounter one within days of being diagnosed. You need to be aware of the early symptoms of a crisis so that you can take action to prevent it developing into a full-scale emergency, *just in case* you find yourself in one of the situations described below.

The amount of extra steroid cover you require to avoid a crisis depends on the severity of the physical stress you are experiencing. The most common reason for Addisonians to require hospitalisation is because they do not take sufficient extra hydrocortisone early enough when they become seriously ill. The second most common reason is reducing their dose again too soon, while they are still in the middle of a serious infectious illness, such as influenza. This section explains some general rules of thumb for managing your medication in these situations. However, if you are in any doubt as to how to manage your dosage during

an illness, please ring your endocrinologist and ask them to advise you by phone.

As a general rule taking more steroid than your body strictly needs for a day or two is not harmful. Taking too much steroid for longer periods of time is harmful.

So you can prudently increase your dose for a short time if you think you are developing a serious illness.

Some serious illnesses require an emergency injection to prevent an Addisonian crisis.

All Addisonians are recommended to have an emergency injection kit at home, just in case you cannot get to a hospital or arrange to be seen by a medical practitioner quickly.

It is also prudent to have some anti-nausea tablets (or suppositories) at home, in case you experience vomiting.

Wearing a MedicAlert tag makes it easier for medical personnel to respond to your condition in an emergency. A letter from your doctor explaining the type of treatment you will need in an emergency is also helpful if you have to go to hospital suddenly.

“I had a crisis a year ago, the first in 25 years. I could not keep anything down and I felt worse and worse. Finally we went to the hospital and I told them, either I die in a bed or I die on the floor. Then they called an endocrinologist. He saved me. I was on a cortisone drip for four days. The main thing is, if you can get medication down you can pull yourself out of almost anything.” SUE

“I went into adrenal crisis because I got flu and my doctor did not advise me to increase medication under stress. Mine felt like a total body shut-down. Terrible nausea, dizziness, feeling like you can’t even move or speak from weakness. My heart went to 35 beats per minute and my breathing became extremely belaboured. I don’t want to worry you. A crisis like mine is completely preventable. At the first sign of infection increase your dose to double or triple and keep at that dose until you feel better. Then taper down to your regular dose.” DEDE

b) COLDS AND OTHER MINOR ILLNESSES

Coughs, colds and other minor illnesses, which do not involve a fever, do not usually need extra steroid cover. If you are in any doubt, check your temperature with a thermometer.

“I teach 140 children each day in high school. I have fewer colds and viruses than my husband who works in an office. I haven’t missed a day because of Addison’s.” SUE

c) SERIOUS ILLNESSES REQUIRING EXTRA STEROID COVER

Flu and other fevers

Any illness that involves a fever needs extra steroid cover. This means most flu bugs and some other infections. If you are prescribed antibiotics by your doctor, ask them if you need to increase your steroid dose at the same time, to combat the infection.

You are strongly advised to have a flu vaccination in the autumn each year because it can take Addisonians longer to recover from a serious illness than the general population.

Doctors’ recommendations vary slightly as to exactly what temperature constitutes a fever and by how much you should increase your steroid dose. As a general guide:

Double your normal daily dose when you have a temperature of more than 37.5° Celsius (99.5° Fahrenheit) or more than +1° Celsius above normal.

Treble your normal daily dose if your temperature rises to more than 39° Celsius (102° Fahrenheit).

Always seek medical help if your temperature reaches 40° Celsius (104° Fahrenheit).

As soon as your temperature returns to normal start to taper your steroid dose back to your normal daily dose.

You do not need to increase your fludrocortisone while you are ill because the higher dose of hydrocortisone will provide enough extra mineralcorticoid.

Inform your doctor if you are ill for more than 3 days.

“I pride myself on being wonderwoman and supermum all rolled into one. And that is okay until I get sick. Then it is such a let-down when I can’t do everything.” CAROLYN

Diarrhoea lasting for more than a day

Diarrhoea on its own can be considered an infection requiring you to double your dose for the duration and then taper back. Hydrocortisone is absorbed rapidly by the stomach so you should be able to continue taking oral medication unless your symptoms lead you to believe that food is passing

through your stomach in less than 30 minutes. Make sure you increase your fluid intake as you will be losing more fluids than usual. Remember that dehydration can be very destabilising for an Addisonian.

d) TAPERING YOUR DOSE BACK DOWN AFTER A SERIOUS ILLNESS

Doctors' recommendations vary as to how quickly you can taper your steroid dose back to normal after a serious illness, but all are agreed that it is desirable to get back to your normal dose as quickly as you can. As a general rule, the longer you have been taking a higher dose than normal, the more slowly you will find you need to taper back. After a short illness (say just a few days on double dose), it is usually safe to go straight back to your normal dose. After a longer period on a high dose (say several weeks), you may need to taper back over a number of days.

If you taper too slowly you can experience uncomfortable side-effects such as joint aches while you reduce your dose, as your body will have become habituated to the higher dose. If you taper too quickly your electrolyte balance (sodium/

potassium) can become destabilised. Experience will teach you how quickly you can taper your own dose after an illness. As a general guide to tapering down after a longer illness:

- *Start to reduce your dose as soon as possible, ie. as soon as your fever has gone.*
- *Start to reduce your dose by one-third to one-half of your normal daily dose.*
- *As you get closer to your normal daily dose, reduce by smaller amounts: eg. 5mg and then 2.5mg hydrocortisone per day.*
- *Remember that you do not need to adjust your fludrocortisone while you are ill, because the higher dose of hydrocortisone will provide enough extra mineralcorticoid.*
- *Inform your doctor that you have had a serious illness requiring you to increase your medication.*

“Mostly I am able to come off a high dose pretty quickly, for example when I have had to double my dose for illness I will cut back by 50% and feel no problem. But a few times I have had serious problems tapering. After an infection crisis where I needed two 100mg injections, I had to take 300% of my normal dose to get through the day and it seemed I could only decrease it by 5mg every two-three days or I started to go back into crisis.”

MARJORIE

e) SERIOUS ILLNESSES WHICH MAY REQUIRE AN EMERGENCY INJECTION

Stomach flu (gastric flu) and any illness involving vomiting

Vomiting needs careful management. Any illness involving vomiting can precipitate an Addisonian crisis as the body becomes dehydrated and medication is not easily absorbed. Individual tolerances for vomiting vary quite widely and some people need emergency treatment much more quickly than others. If you are experiencing continuous vomiting over several hours and you are not able to keep your medication down, you may need an emergency injection of hydrocortisone. You may then need intravenous fluids to repair the effects of dehydration.

As soon as vomiting begins:

- *Take an anti-nausea tablet (or suppository) if you have any to hand.*
- *Take extra hydrocortisone and see how long you can keep it down.*
- *Arrange to have an adult companion in the house in case your condition deteriorates rapidly.*
- *Arrange a house call from a doctor if necessary.*
- *Make preparations to give yourself an emergency injection should it become necessary, and to then go to the nearest hospital for intravenous fluids.*
- *Remember that if your illness progresses rapidly you may experience some mental confusion, so it is essential that a responsible adult is aware of your condition.*
- *If you need to go to hospital, explain that you are experiencing an adrenal crisis and require intravenous steroids and intravenous fluids to stabilise your condition.*

Shock and major accidents

Prolonged vomiting is the main illness requiring an emergency injection to prevent a crisis. Accidents, which trigger a state of shock, are the other main event requiring an emergency injection.

If you are involved in a serious road accident, or are injured in some other type of accident, you may start to go into shock. When this happens you may experience severe dizziness, a sudden drop in blood pressure and a state of mental confusion without a preceding period of nausea and vomiting. You will need an emergency injection to stabilise your condition as soon as possible.

Anyone who spends a lot of time travelling by car should arrange with their doctor to have an injection kit in the glove box, (preferably in an insulated container). If you go horse riding you should also make sure you have an injection kit with you.

“Even if you are not seriously injured you can go into shock at the drop of a hat. I was sitting at a red light when a mini-van rear-ended me and my car ended up half-way underneath the car in front. I had a fair amount of pain where my seatbelt had been so decided to get checked out at the hospital. About ten minutes before the ambulance got to the hospital I started feeling really bad. I realised my blood pressure was probably taking a dive so I told the paramedic I needed my emergency injection. He said he would have to call ahead and okay it with the hospital. In about five minutes my blood pressure had gone from 130/80 to 80/40. I told him not to worry about it and proceeded to whip down my pants and inject myself in a certain large muscle mass. By the time we got to the hospital my blood pressure was so low they couldn't find my pulse. Then the hydrocortisone kicked in and my blood pressure returned to normal. I've had Addison's for about six years and this was the first time I've had to use my emergency kit.” CHRIS

“I have had a 24 hour tummy bug on several occasions and have always dealt with it successfully at home. I take a small dose of hydrocortisone every two hours until I stop being sick and sip water continuously. The last time it happened I did call the doctor as I was getting a little confused, but so far I've not needed an emergency injection.” MARGO

f) GIVING YOURSELF AN EMERGENCY INJECTION

In the UK, an injection kit usually contains liquid hydrocortisone, which you draw up into the syringe to inject yourself. Always check the expiry date on your injection kit and keep it up to date.

When you first receive your injection kit, arrange to have a nurse or doctor show you how to give yourself an injection. Make sure you are able to practice this a few times. If it is some years since you were taught how to do an emergency injection and you have not had to use your kit in the meantime, you might like to ask for a refresher session.

Ideally, your partner or a regular companion should also know how to give you an injection. Good instructions on how to do it are available on a number of websites, including the Canadian website listed in section 11.

Some very rough emergency guidelines are:

■ *Hydrocortisone should be injected straight into muscle. The thigh or buttock is the best large muscle for this.*

- *Draw the solution up into the syringe.*
- *Make sure there is no air in the top of the syringe.*
- *Use one hand to hold the skin taut around your target injection site.*
- *Hold the needle like a dart and aim it straight down.*
- *Plunge the needle in as quickly as possible. It hurts less that way. (Speed is more important than skill).*
- *Squeeze the plunger down in a smooth movement so all the solution is eased into the muscle.*
- *Don't worry too much if you can't find a sterile swab for the skin beforehand.*

Some doctors are reluctant to issue Addisonians with their own injection kit, believing that they should simply seek emergency help from a hospital or the nearest medical practitioner in the event of serious injury or illness. If this is your doctor's view, try to identify the full range of situations in which you might need to use an injection kit and discuss these with them. For example, do you do a lot of driving? How often do you travel away from home? Do your holidays involve camping or hiking in remote areas, or overseas travel? In rush hour traffic, how long would it take you to get to your nearest hospital from work or home?

"I carry my syringe in a toothbrush holder. Film canisters are good for keeping the vial in. It's best to have a few syringes on hand, just in case. It's also good to do a couple of practice shots for yourself. I was terrified of giving myself injections until I was prescribed vitamin B12. Now I'm sticking needles in myself twice a week." JULIA

g) SURGERY REQUIRING EXTRA STEROID COVER

All surgery involves additional physical stress as your body repairs itself. How extensive the surgery is determines how much extra steroid cover you need, and for how long. Make sure you discuss your adrenal condition and the need for extra steroid cover beforehand with the surgeon or anaesthetist.

Try to schedule this discussion at least a day in advance so that your medical practitioners have time to check their manuals as to current best practice for your type of surgery and ensure they have all the supplies they need. In smaller hospitals, it may be prudent to take your own emergency injection kit with you, as not all hospitals or surgeries have large supplies of intravenous hydrocortisone on hand.

Dental surgery

Some endocrinologists do not believe that any extra steroid is needed for simple dental work involving a local anaesthetic. In practice, some Addisonians take a small top-up (say 5mg hydrocortisone) just before the procedure if they find dental work stressful.

Minor surgery with local anaesthetic

Again, some endocrinologists do not believe that any extra steroid is needed for minor surgery with

a local anaesthetic. Examples here would be the removal of a mole or small skin cancer. Again, in practice some Addisonians take a small top-up (say 5mg hydrocortisone) before a procedure involving anaesthetic.

Moderately stressful surgery

Here the experts agree on the need for extra steroid cover. Examples would be invasive procedures such as a barium enema, endoscopy, cataract surgery or major oral surgery. The textbook approach is a single 100mg intravenous dose of hydrocortisone just before the procedure. Then return to your normal daily medication.

Major surgery

This means any invasive surgery involving the abdomen or chest. Examples would be a caesarean section or heart operation. The textbook approach is 100mg intravenous hydrocortisone given just before anaesthesia, continued every 8 hours for 48 - 72 hours. Then taper rapidly to your normal medication. Where your anaesthesia means you cannot have anything to eat or drink for 8 hours before the surgery, see if you can have a saline drip for that period to prevent any risk of dehydration.

h) MEDICALERT AND OTHER USEFUL INFORMATION

Wearing a MedicAlert tag makes it easier for medical personnel to respond to your condition in an emergency. It should be easy to locate and say something along the lines of:

■ **Adrenal insufficiency. Steroid dependent.**

Informally, medical personnel will tell you that a bracelet is usually easier to locate than a necklace. Remember that paramedics such as ambulance staff are often not aware of what Addison's disease entails, because it is a rare condition.

A letter from your doctor stating what medical treatment you will require in the event of a crisis can be helpful, allowing busy staff at the accident and emergency desk to prioritise your case appropriately.

Because it is a rare condition, you cannot assume that all medical personnel will be familiar with Addison's disease or know how to treat an adrenal crisis. Any documentation you can offer to guide them in delivering the best emergency treatment will be useful. If you arrive at the accident and emergency department of a hospital without a MedicAlert, steroid card or doctor's letter, you may not receive treatment as promptly.

CRISIS MANAGEMENT: A SUMMARY

Have your own injection kit, anti-nausea tablets (or suppositories) and sufficient extra medication to cover a period of illness with you at home.

Make sure you and your partner or a regular companion know how to give an emergency injection of hydrocortisone.

If you have any doubts as to the severity of your illness, ring your doctor and ask for their advice.

The following conditions generally require you to double your normal daily dose:

- Fever of more than 37.5° Celsius (99.5° Fahrenheit)
- Diarrhoea lasting more than 24 hours

Gradually taper your steroid dose back down to your normal daily dose when you no longer need to double it.

The following conditions may require an emergency injection and intravenous fluids. Seek medical advice promptly in these cases:

- Severe vomiting
- Shock

Surgery generally requires extra steroid cover. Make sure you discuss your adrenal condition and the amount of extra steroid cover you will need beforehand with your surgeon or anaesthetist.

Wear a MedicAlert tag.

Crisis management means crisis avoidance whenever possible. Be prepared and be quick to seek help.

Remember to have a flu vaccination every autumn.

If you are unlucky enough to go into crisis and there is any uncertainty as to the treatment you need, insist on 100mg hydrocortisone by vein every 6 - 8 hours.