

LIVING WITH ADDISON'S DISEASE



An owner's manual for individuals with the disease

INTRODUCTION

Addison's disease is a rare chronic condition brought about by failure of the adrenal glands. Life-long, continuous treatment with steroid replacement therapy is required. This aims to replicate the necessary amounts of the missing hormones - cortisol and aldosterone - that the individual can no longer produce from the adrenal glands. With the right balance of daily medication, most people with Addison's disease are able to continue life much as it was before their illness.

This guide has been co-written by Sarah Baker and Katherine White, members of the UK Addison's Disease Self-Help Group, in collaboration with endocrinologist, Professor J A H Wass. It aims to provide you with sufficient information to allow you to work well with your doctors in fine-tuning steroid replacement therapy, and in making informed choices about some of the practicalities of managing the condition. A glossary of the terms and references used in the manual starts on page 21.

In common with fellow Addisonians, you will learn to manage your own health day-to-day. At some stage, you may experience illnesses or other situations requiring significant, short-term adjustments in medication. You will need to take extra medication if you get a serious infection or injury. Without this extra medication, you could experience potentially life-threatening symptoms known as an Addisonian crisis. This guide aims to help with both the everyday and the more unusual challenges that Addisonians can experience.

This guide is addressed principally to UK residents and reflects UK medical practices. Please remember that the information it provides is intended as a supplement to, not a substitute for, the advice of your doctor who knows the details of your medical history. Individual responses to the condition can vary widely and, in some instances, individuals may have experienced symptoms needing differing treatment to that described here.

Many of the specific issues addressed in this guide were raised in the most recent UK survey of the Addison's Disease Self-Help Group (ADSHG), conducted in 1996/7. The full results of that survey can be found on the ADSHG website listed in section 11.

Sarah Baker & Katherine White

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a) MANAGING YOUR LIFESTYLE

"I have had Addison's for twelve years and have not had a crisis needing medical assistance since my diagnosis. I have raised seven children, run a motor vehicle repair business and drive extensively. I am 55 years old, very active. Like many others I have few side effects and am happy with my quality of life."

JAMES

"My body cannot be normal, but I do pretty well. Our limitations really are few. I guess we just need to learn at what point we should go on or stop to give our bodies a break!"

SANDRA

"I have had Addison's for 15 years and have never had a crisis. I have had a full-time job, raised three children alone when my husband died 16 years ago, still play volleyball and golf, and just feel like I keep a fairly normal schedule for a 50 year old."

ROSEANNA

Individuals with Addison's disease can expect to have a normal life span, provided they manage their daily medication sensibly. Addisonians can bring up a family on their own, hold down a demanding job, run marathons or even become the President of the United States.

That is not to say that life as an Addisonian will always feel 'normal'. Most individuals find they have to become skilled at managing the fatigue that accompanies the condition. In the last UK survey of Addisonians (1996/7), around 40% of respondents said they had ongoing difficulties with fatigue and a lack of energy. There is more information on managing fatigue in section 3: *Medication* and in section 4: *Common concerns*.

A number of people find they develop related auto-immune conditions such as diabetes or hypothyroidism, requiring extra healthcare management. There is more information on related autoimmune conditions in section 2: *The Causes of Addison's*.

Addisonians must always be aware of their own health and ready to increase their dosage if they get sick or are seriously injured. A North American survey (1997) found that three-quarters of respondents had received emergency treatment at a hospital for their Addison's disease at some point, while around 15% had had to use their emergency injection kit at home. There is more information on what to do if you are seriously ill in section 10: *Crisis management*.

A few individuals are less lucky and find that their health never recovers sufficiently to allow them to return to full-time work after diagnosis. Sometimes this is because they experienced serious damage to their health (such as a stroke) brought

1 HOW 'NORMAL' CAN LIFE BE WITH ADDISON'S DISEASE?

about by a delayed diagnosis. Sometimes it is because they have multiple health conditions, which include adrenal failure. So not everyone can run marathons, just as not all of us could become President.

Most of us, however, continue to lead busy and rewarding lives, with the support of our family, friends and medical professionals. And if you do want to run a marathon, there is more information in section 6: *Exercise*.

"I have had Addison's for 20 years, and also hypothyroidism. I have also had diabetes for 40 years. I do work full time, and raised two girls, now 19 and 21. I was so glad to read about the experiences of others and discover that I really have an excuse for fatigue - not just lazy. My doctor never mentioned the exhaustion. I also get joint and back pain. Never incapacitating - just frustrating. But I do take good care of myself and have a wonderful doctor."

TRISH

"I work 9 to 5 and I have two boys. When I feel bad I feel exhausted and aching and can't get on with my life. People don't always feel great despite the medication."

GAIL

b) YOU AND YOUR DOCTORS

As an Addisonian, you are responsible for day-to-day management of your own health. You must take your daily medication at the right times and monitor your general health for any signs of serious illness where your medication might need increasing. Serious illnesses requiring extra medication are discussed in section 10 of this manual: *Crisis management*.

Most Addisonians are supported in their health management by two medical professionals: their General Practitioner and their endocrinologist.

In the UK, your General Practitioner (GP) is usually responsible for prescribing your current medication and for checking your general health status with you - including any problems unrelated to your Addison's.

In the UK, your GP will be able to authorise a Medical Exemption Certificate for you, allowing you to receive all prescription medication free of NHS charges, so please remember to ask them for this.

Your endocrinologist is generally responsible for reviewing your medication requirements and checking for any changes in your endocrine health. They will usually check for signs that your steroid medication may need adjusting up or down. Some Addisonians do also develop related autoimmune conditions, such as hypothyroidism or diabetes. Your endocrinologist will usually check for any signs of these. In the UK, most endocrinologists prefer to see their Addisonian patients every six to twelve months.

So, being a successful Addisonian means being a good communicator, as there are two medical professionals to keep informed about any important changes in your health.

Some Addisonians find it helps to keep a written record of their health status when there are important health issues they want to review with their GP or endocrinologist. At such times, a medical 'diary' can be a useful guide for your doctor, providing more specific information than you can recall in a general conversation. You will probably want to record the timing and amount of medication you take, your level of energy/fatigue and any other important health factors or changes.

"My GP and I work well as a team and he also works well with my endocrinologist. My GP has admitted honestly that he does not know enough about the disease to care for me on his own."

SANDRA

"I see my endocrinologist every six months and it is amazing the changes that can happen even in that short time."

JOAN

"My endocrinologist is great. I have kept a journal in the past with symptoms matched to activities including blood pressure. The doctor used this to help match trends in relation to medication. A journal makes your information more accurate and credible. And you must educate yourself as much as possible."

ALAN

2 THE CAUSES OF ADDISON'S AND RELATED AUTOIMMUNE DISEASES

a) PRIMARY AND SECONDARY ADRENAL DISEASE

Addison's disease is a rare condition where the adrenal glands gradually cease to function. The disease is not usually apparent until over 90% of the adrenal cortex has been destroyed, so that very little adrenal capacity is left. Symptoms of the disease, once advanced, can include severe fatigue and weakness, loss of weight, increased pigmentation of the skin, faintness and low blood pressure, nausea, vomiting, salt cravings, and painful muscles and joints.

The adrenal glands sit at the top of the kidneys, one on each side of the body, and have an inner core (known as the *medulla*) surrounded by the outer shell (known as the *cortex*). The inner medulla produces adrenaline, the 'fight or flight' stress hormone. The outer cortex produces the steroid hormones that are essential for life: cortisol and aldosterone. It also produces sex hormones known as adrenal androgens; the most important of these is DHEA.

Addison's disease is not an 'all or nothing' condition. In the early stages of the disease many individuals are still able to produce some cortisol and enough aldosterone. This is partly why individuals with the disease take varying amounts of medication and why the amount of medication you need may alter over the years.

Dr Thomas Addison first identified the disease in the mid 1800s while working at an inner-city London hospital. Then, the main cause of the disease was as a complication of tuberculosis. In third world countries today, tuberculosis remains an important cause of Addison's. HIV (AIDS) is now becoming another significant infectious disease causing adrenal failure among third world populations.

Among more affluent countries, the main cause of the disease today is *autoimmune adrenalitis*, where an over-active

immune system starts attacking the body's own organs. Autoimmune adrenalitis now accounts for around 70% of all cases, and affects more women than men. The cause of autoimmune adrenalitis is not known, in common with most other autoimmune diseases.

Other, much rarer causes of Addison's include certain fungal infections, adrenal cancer and adrenal haemorrhage (for example, following a car accident). In some cases it can result from the treatment needed for *Cushing's disease* (overproduction of adrenal hormones).

Certain rare hereditary diseases also cause adrenal insufficiency (such as adrenoleukodystrophy and congenital adrenal hyperplasia).

All of the conditions mentioned above are described as 'primary adrenal insufficiency', because they result from a disease process that has directly affected the adrenal glands.

Secondary adrenal insufficiency is sometimes informally described as 'Addison's', although it has a very different cause. Secondary adrenal insufficiency mostly occurs when a pituitary tumour (such as an *adenoma*) forms, although autoimmune destruction of the pituitary gland is also known. Secondary adrenal insufficiency is even rarer than primary Addison's disease.

Secondary loss of adrenal function occurs when the messenger hormone, which stimulates the adrenal glands into action, is no longer produced by the pituitary gland. The pituitary gland is located inside the skull, just behind the eyes and tucked below the grey matter of the brain. This messenger hormone is called *ACTH* and is responsible for the extra pigmentation found in primary Addison's. People with secondary adrenal failure do not experience the increased pigmentation found in primary Addison's, because their ACTH levels are declining.

Long-term use of high doses of steroid drugs to treat other illnesses (such as the high doses of prednisone which may be used to treat bowel disease or asthma) can also cause temporary or permanent loss of adrenal function. This is often referred to as *secondary adrenal suppression*.

b) RELATED AUTOIMMUNE CONDITIONS

Where Addison's disease is autoimmune, around half of those with the condition will develop another autoimmune disorder, usually another endocrine condition.

The most common endocrine disorder associated with Addison's is a thyroid problem, either *hyperthyroidism* or *hypothyroidism*. Most medical studies have found that just over one-fifth of those with autoimmune Addison's are likely to develop a thyroid problem of some kind.

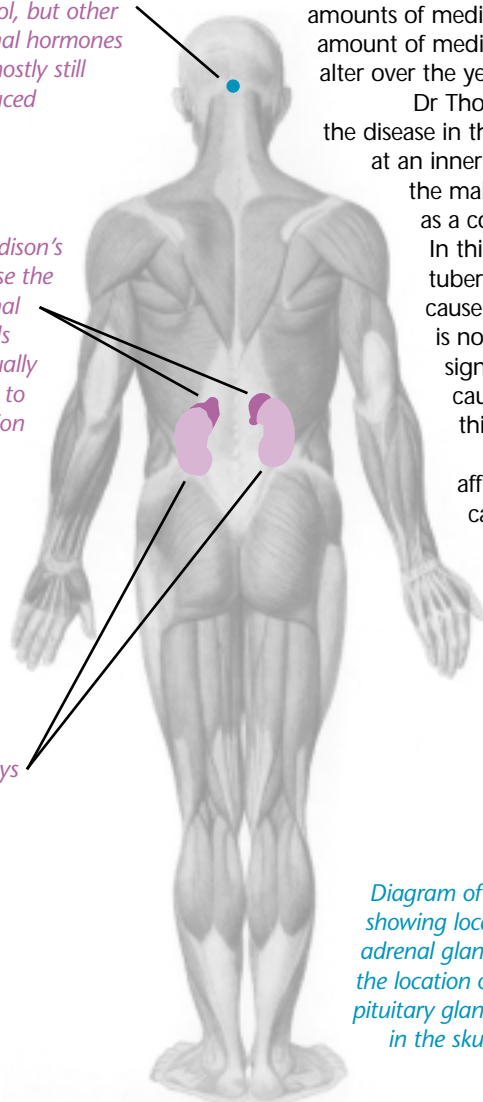
Other recognised associations include premature failure of the ovaries, insulin-dependent diabetes (*type 1 diabetes*) and parathyroid deficiency. These all occur less frequently than thyroid disorders.

In secondary adrenal insufficiency the pituitary gland no longer triggers the adrenals to produce cortisol, but other adrenal hormones are mostly still produced

In Addison's disease the adrenal glands gradually cease to function

Kidneys

Diagram of body showing location of adrenal glands and the location of the pituitary gland in the skull



A few people with autoimmune Addison's develop a combination of related autoimmune conditions, which are known as a *polyglandular autoimmune syndrome*.

Non-endocrine autoimmune diseases sometimes also occur in combination with autoimmune Addison's. Although these are seen less frequently than the endocrine conditions mentioned above, a very small number of people find they develop both endocrine and non-endocrine autoimmune conditions related to their Addison's.

Medical studies estimate that around 5% of individuals with autoimmune Addison's develop *pernicious anaemia* (vitamin B12 deficiency). Much smaller proportions are estimated to develop conditions such as *vitiligo* (loss of pigmentation from parts of the skin), *coeliac disease* (gluten allergy), *alopecia* (hair loss), *myasthenia gravis* (muscle wasting), *thrombocytopenia purpura* (loss of blood

platelets), *Sjogren's syndrome* (dry eyes and mouth) or rheumatoid arthritis.

Autoimmune Addison's is not usually a directly inherited condition. But a tendency to autoimmune diseases does seem to run in some families. Where autoimmune Addison's occurs on its own, some kind of family association with the condition can be traced in about one-third of cases. Where it occurs as part of a polyglandular syndrome, some kind of family history of related autoimmune diseases can usually be found in about half the cases.

Where a tendency to autoimmune endocrine disorders is inherited, it is often not the same condition as the parent/grandparent but some other related autoimmune condition, which appears in the next generation. For example, a grandmother with Addison's disease may see one of her grandchildren develop vitiligo or a thyroid condition.

a) WHAT IS AVAILABLE AND WHAT IT DOES¹

¹ In the UK, people with Addison's disease are entitled to free NHS prescription medication, by asking their GP to authorise a Medical Exemption Certificate for them.

The adrenal hormones, cortisol and aldosterone, are essential for life. Their technical description is steroid hormones. Your steroid medication aims to replace them in a manner that approximates the natural rhythms of the body.

Most Addisonians are prescribed a combination of hydrocortisone and fludrocortisone to replace the cortisol and aldosterone hormones. The exact dose depends on the size and metabolism of the individual, as well as how advanced their condition is. In the early stages of the disease many individuals are still able to produce some cortisol and enough aldosterone. Getting it right depends on collaboration between you and your medical practitioners; they must monitor your blood tests and you must monitor your well-being in response to different levels of medication. In the end, a large part of your mutual success will be due to a considered process of experimentation.

Over time, an individual's need for each medication may alter slightly. If you feel a return of any of your former Addison's symptoms, or start to identify the symptoms of being over-medicated, you will need to review this with your doctor.

Cortisol replacement

Hydrocortisone is usually taken in three or two small doses over the course of the day. You may find it helpful to adjust the timing and amount of each divided dose to match changes in your lifestyle.

Hydrocortisone is the preferred drug because it is quickly absorbed and the closest mimic of what the body would naturally produce. A hydrocortisone tablet has been almost totally absorbed by the stomach and is active in the bloodstream within 30 minutes of being swallowed on an empty stomach.

Hydrocortisone has a further advantage over other types of steroid: the amount of it in your bloodstream can be accurately measured. This makes it easier to monitor your dosage and adjust it up or down if necessary.

3 MEDICATION

Prednisone and dexamethasone are longer-acting steroids and take longer to become active in the bloodstream after being swallowed. Prednisone and dexamethasone are normally taken less often and in larger single doses. Individuals who switch to these drugs from hydrocortisone may find they need to increase their fludrocortisone dose.

Cortisone acetate is an older version of hydrocortisone and less readily absorbed. It is also slower acting, as it must first be processed by the liver into a form the body can use.

Each of these other drugs has a different strength to hydrocortisone; roughly equivalent doses are shown in the table below.

ROUGHLY EQUIVALENT DOSES OF STEROIDS*	
*Hydrocortisone	20mg
*Prednisone	5mg
*Dexamethasone	0.75mg**
*Cortisone acetate	25mg

* Each of these drugs has slightly different properties, which should be discussed with your endocrinologist.
 ** See Glossary for a more thorough discussion of the potency of dexamethasone.

HYDROCORTISONE is prescribed to replace the cortisol steroid normally produced by your adrenal glands. Lack of cortisol is what causes the muscle weakness, low blood sugar and loss of concentration commonly experienced by Addisonians. There are several substitutes for hydrocortisone, the most common ones being prednisone, dexamethasone and cortisone acetate.

FLUDROCORTISONE is prescribed to replace the aldosterone steroid normally produced by your adrenal glands. Lack of aldosterone causes the electrolyte balance in the body to become destabilised as the kidneys retain potassium, the body loses salt and fluids, leading to low blood pressure and dehydration.

“I was on cortisone acetate for a few years and didn't think too much about it. When I switched to hydrocortisone I felt 100% better. It made me feel more 'normal' also when my endocrinologist added fludrocortisone. I got rid of the lower back pain I had had for so many years.” SUE

Why is hydrocortisone the preferred drug?

Although it is a synthetic drug, hydrocortisone is indistinguishable from the cortisol naturally produced by the adrenals. It is also fast acting and can be measured in the blood. For these reasons, hydrocortisone is the preferred drug for most Addisonians. But it may not suit everyone. Some individuals find they do better on a longer-acting steroid.

In the UK, the 1996 survey found that around 89% of all Addisonians were taking hydrocortisone. Of the rest, 6% were taking prednisone and 5% were taking cortisone acetate.

Medical prescribing patterns do vary by country. In the US, the 1997 survey found that 65% of Addisonians were taking hydrocortisone. The remainder were taking either prednisone (20%) or cortisone acetate (15%) while just 1% was taking dexamethasone.

“I did terribly on hydrocortisone (roller coaster days) and terribly on dexamethasone (too potent and I became Cushingoid). I felt pretty good on prednisone. That was the best until my latest endocrinologist put me on cortisone acetate. I’m happy to say it is the one for me! I did find I needed differing amounts of fludrocortisone on the different corticosteroids. I needed more on prednisone than I did on hydrocortisone. And now I need less on cortisone acetate.” KELLY

Aldosterone replacement

Fludrocortisone is usually taken as a single daily dose, first thing in the morning. It is slower acting than cortisol-replacement steroids, so people who only require a small amount can take it every second day. There is a wide variation in how much fludrocortisone individuals need to take. A small number of Addisonians find they do not need fludrocortisone once their hydrocortisone medication is stable.

85% of Addisonians who took part in the 1996 UK survey were taking fludrocortisone. The amounts taken by individuals ranged from 0.025mg every other day up to 0.4mg per day, while most people were taking a standard dose of 0.1mg per day (equal to 100mcg).

Too much fludrocortisone can lead to a potassium deficiency which, in severe cases, causes an irregular heartbeat or other cardiac symptoms. Too little fludrocortisone can lead to a potassium overload, which also causes cardiac symptoms.

You will need to monitor your own response to your fludrocortisone dose and inform your medical practitioner if you experience new symptoms, which you think may be related. It is important not to modify your fludrocortisone medication before checking with your doctor.

People with secondary adrenal failure usually keep their adrenal production of aldosterone so, in almost all cases, do not need to take fludrocortisone. Similarly, individuals who are experiencing steroid-induced adrenal suppression do not usually need any fludrocortisone.

“I recently started taking fludrocortisone after my latest renin test showed that my aldosterone output is waning. I only have to take it every other day so far, and supplement with a little extra salt here and there. I have taken salt tablets when I have been in the heat.” KARLA

“I take fludrocortisone at 0.2mg a day. It helps with my sodium retention and keeps my blood pressure up. But the main reason it’s such a high dose is that my potassium wants to climb to near fatal levels without it.” TRACEY

b) THE NATURAL DAILY CYCLE OF ADRENAL HORMONE PRODUCTION

In a person with healthy adrenals, cortisol levels start to rise around 4am, are at their maximum around the time you wake up in the morning and then gradually taper off during the day. That is why you will be advised to take your largest divided dose first thing in the morning.

Cortisol levels are naturally at their lowest shortly after going to sleep, so it is best to take your last divided dose of the day by early evening. Taking

steroid medication too late in the evening can cause sleeping difficulties. Around 6pm, or at least four hours before going to bed, is best.

Aldosterone production follows a similar pattern to cortisol, with the highest levels naturally occurring around the time you wake up. That is why fludrocortisone is usually taken with the morning medication.

c) HOW DO I KNOW WHAT IS THE RIGHT DOSE FOR ME?

Some medical textbooks still refer to the standard practice of prescribing a single dose of hydrocortisone for both men and women irrespective of size and bodyweight. In practice, most endocrinologists and Addisonians now believe this is inappropriate.

Most women do not need as much as 30mg hydrocortisone while some large, well-built men do need more. Some small, lightly-built women can live comfortably on a dose of less than 20mg. Similarly, lightly-built men need less than 30mg.

■ *A rough rule of thumb is that both women and men should start with a daily dose of 20mg hydrocortisone².*

■ *Then if you still feel noticeably unwell, gradually increase your daily dose in increments of 2.5mg until you feel well enough to live a normal daily life.*

“When I was first diagnosed in 1979 I was put on 15mg hydrocortisone as I am quite petite. This dosage gradually increased to 30mg over many years. Then, about six years ago, there were other health problems. After that I slowly reduced my dose so that, for the past two years, it has been just 12.5mg a day.” SUSAN

Some Addisonians find that adjusting the timing and amount of each divided dose can be just as helpful as taking a larger daily dose.

IT IS IMPORTANT that you do not take any than the smallest possible daily dose of hydrocortisone suitable for your symptoms. Over a period of years too much can lead to damaging side-effects such as glaucoma and osteoporosis.

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. But for your everyday medication, the aim should be to keep your dose as low as possible.

² Some petite women may, in fact, be able to start on a dose of 15mg hydrocortisone per day, particularly if they are also taking contraceptives or other female hormone replacement therapy.

d) TIMING YOUR DIVIDED DOSE

Some medical textbooks still refer to the standard practice of taking hydrocortisone twice a day, early morning and evening. In practice, many endocrinologists and Addisonians agree that taking a smaller, divided dose more frequently has real benefits. For example, if you commonly experience an energy-lag in the afternoon, it may help to switch from taking your medication twice a day to taking it three times a day.

It prevents the energy lag that many Addisonians experience in the afternoon when their cortisone levels are at their lowest. Some people describe this energy lag as a kind of 'brain fog', while others describe it as feeling low on blood sugar, faintly dizzy, or just irritable.

For people taking prednisone or dexamethasone, the timing of divided doses is usually less critical, as the medication stays in the bloodstream for longer. This can be useful for individuals whose lifestyle makes it difficult for them to take medication during the day. However, these drugs also take longer to become active after being swallowed, which can be a disadvantage at times.

"I find it important to split my medication up into three daily doses to avoid those highs and lows. Doing this I am able to continue with my physical and usually stressful work. Don't be afraid to take your medication in smaller, spread-out doses. Taking it with food helps to slow the body's absorption rate."

JAMES

"The morning dose is the most important for me to take on time. But I can tell the difference with the afternoon one if I am more than an hour late. The timing can change with how the day goes, if there are a lot of stresses. Once I got up at 4am and by 7am I was on the way out. I learnt then that once I start my day I need my medication." **ALAN**

NICK is in his mid-thirties and a keen runner. He used to take his 30mg hydrocortisone in two divided doses, 20mg before breakfast and 10mg at 6pm. Following a day curve analysis he now takes 40mg hydrocortisone in three divided doses:

- 20mg on waking
- 10mg with lunch
- 10mg at 6pm

CATHARYN is in her early forties and a full-time mother. She used to take 30mg hydrocortisone in two divided doses. After switching to three divided doses she found she could cut down her total hydrocortisone dose to 25mg:

- 12.5mg on waking
- 7.5mg with lunch
- 5mg at 6pm

MIKE is a senior civil servant. For many years he took 15mg hydrocortisone in two divided doses. Since introducing a low dose of prednisone in the evening, he has been able to reduce his total daily dose to the equivalent of just 12mg hydrocortisone. He takes:

- 7.5mg hydrocortisone on waking
- 1mg prednisone at 6pm

KAREN is in her late thirties and runs her own business as well as bringing up a young family. Before her day curve she was taking 20mg hydrocortisone a day in two doses: 15mg on waking and 5mg with dinner. Following her day curve, she now takes it three times a day:

- 10mg on waking
- 5mg with lunch
- 5mg with dinner

e) MONITORING YOUR MEDICATION: BLOOD TESTS

Thorough blood tests are available in the UK to help you determine the right daily dose of medication for you. Remember that these are tests to monitor your ongoing medication, not tests to determine whether you still have the disease. Some of these tests can be done by your GP while others need to be conducted at a hospital laboratory.

TESTS FOR HYDROCORTISONE

- 8am plasma cortisol
- 8am plasma ACTH
- Day curve analysis

TESTS FOR FLUDROCORTISONE

- Plasma renin
- Electrolytes (potassium and sodium)
- Blood pressure

The most comprehensive test of hydrocortisone medication is known as a day curve and is usually done through the endocrinology department of a major hospital. If you have not had a day curve done, you may wish to ask your GP if they could refer you to a suitable hospital.

The most widely used test of how well the adrenals themselves are functioning is the *ACTH stimulation test*. Most Addisonians will have undergone an ACTH stimulation test to confirm their diagnosis with the disease. A few Addisonians are asked to repeat this test over time to assess any further deterioration in their adrenal function.

Some endocrinologists do not offer a full day curve analysis, preferring to use a combination of 24 hour urine analysis and mid morning/mid-afternoon blood samples recording cortisol and ACTH levels.

If you cannot travel to a major hospital for a day curve, it is still possible for your GP to monitor the adequacy of your hydrocortisone dose in consultation with an endocrinologist. The 24 hour urine sample and the mid morning/mid afternoon samples of cortisol and ACTH can be administered by a GP.

Day curve analysis

Different hospitals have different procedures for a day curve, although all follow the same general principles. Most are done on an out-patient basis.

The most comprehensive day curve analysis involves taking blood samples over an 11 hour day starting early morning before you take your first divided dose of medication for the day. Repeated blood tests during the day track the medication entering your blood stream when you take each dose and record how long it lasts. The amount of hydrocortisone in your bloodstream during the day can then be compared to the ideal for a healthy individual, and the amount and timing of each dose adjusted as necessary.

“It took me a while to get the medication into a good balance after my diagnosis. It really took me a couple of years to get it right. For a while I was still so exhausted I couldn’t even walk to the letterbox and back.” JAMES

When you go for a day curve do remember to check if you are allowed to eat breakfast before you arrive. Make sure you bring all your normal daily medication with you including any medication you take for other conditions.

Following a day curve analysis, your specialist may advise you to reduce or increase your current daily dose or to try taking it in smaller, more frequent amounts. A specialist should advise you to reduce your dose if the day curve showed that your

blood levels of cortisol were significantly higher than those of a healthy person for long periods of time.

Most specialists agree that Addisonians do not need to have a repeat day curve done unless there are major changes in their general health or weight. For example, if you were to develop a further endocrine problem - such as a thyroid disorder - you should probably request a repeat day curve, once your thyroid medication had been stabilised.

f) DRUG INTERACTIONS

Most prescription and over the counter drugs will not affect your steroid medication, although it is always wise to check this with your doctor or pharmacist. However, there are a few prescription drugs that influence the way hydrocortisone and other steroid medications are metabolised by the body, either slowing down or speeding up the rate at which the steroids are metabolised. This means that anyone taking these drugs will probably need to adjust their steroid medication.

Contraceptive pills and Hormone Replacement Therapy (oral oestrogens) slow down the rate at which the body metabolises hydrocortisone by around one-third. This means a woman taking the pill or HRT is usually comfortable on a hydrocortisone dose that is one-third less than she would otherwise need. If she continues taking her previous hydrocortisone dose, she may develop mild signs of over-medication, such as a puffy face and little fat deposits on the tummy, above the collar bones and on the back of the neck.

Anyone who is taking drugs to treat tuberculosis (such as Rifampicin and Rifabutin) will typically need to increase their dose of both hydrocortisone and fludrocortisone by up to double the normal dose, as anti-tuberculosis drugs speed up the rate at which the body metabolises its steroid medication. These anti-tuberculosis drugs are highly potent, so you will need to make sure your condition is being thoroughly monitored by your doctors throughout your treatment.

Anti-epileptic medications (such as phenytoin) can also speed up the metabolism of steroids. Growth hormone treatment can lower blood levels of cortisol, by reducing the amount of cortisol-binding globulin available.

If you should need ulcer-healing drugs (such as carbenoxolone) you may need to reduce your fludrocortisone, as these drugs can lower potassium levels. If you should need to take anti-depressants, you may need to ask your doctor to monitor your electrolytes, as some types can cause the body to lose more sodium than usual.

g) MEDICATION AND FOOD

Hydrocortisone prescriptions are usually issued with instructions to take this medication with food. In practice, most Addisonians can take their tablets with just water without experiencing indigestion. This is because the amounts of steroid we swallow are smaller than those taken by individuals with medical conditions needing supplementary steroids (pharmacological doses).

If you find you do experience indigestion taking your steroid medication with water, a glass of milk (or milk substitutes such as soy, rice milk) is usually all that is needed to prevent indigestion. As a general rule, individual doses of 20mg hydrocortisone or less do not need to be taken with food.

Swallowing your tablets just with water makes it easier to take your first dose for the day as soon as you wake up. Waiting to take your first tablet with breakfast means a delay in getting the steroid into your bloodstream, during which time you will feel less well with no hydrocortisone in your blood.

It is also advisable that, if you wear contact lenses, you should put these in before taking hydrocortisone tablets, so that there is no risk of getting any traces of the medication in your eyes.

h) REPLACEMENT OF OTHER ADRENAL HORMONES?

Addisonians are usually lacking in one other major group of adrenal hormones: DHEA and its related compounds. DHEA is not available in the UK at present. However, during the late 1990s several clinical trials in the UK and internationally concluded that there were moderate benefits for most Addisonians from taking small quantities of DHEA. These benefits included protection against osteoporosis, greater energy, enhanced levels of libido and lean muscle, and relief from dry skin. Side effects noted by some individuals were acne, greasy skin and moderate weight gain. Two of the three clinical trials involved very small numbers of women

only, and none of the trials lasted longer than four months. A follow-up study in the UK is taking place during 2001/2 and this will last a full 12 months.

Anyone who is considering taking DHEA is advised to discuss this with their doctor and have their blood level measured before they do. Although clinical trials to date have recommended a daily dose of 25-50mg, not all Addisonians will need as much as this. A few individuals have reported that doses as low as 10mg brought their levels up to the normal range. Bear in mind that some endocrinologists are cautious about prescribing DHEA in advance of the results from longer-term clinical trials, while others are already doing so.

“I have been taking part in trials of DHEA over the past two years. The first study was a double blind study taking the real McCoy for three months and a dummy for three months. I knew when I was on the real tablets as I had more energy but the side effects were acne on my face, chest and scalp. I lost weight but that may have been down to more horse riding.” JANE

i) ALTERNATIVE AND COMPLEMENTARY THERAPIES

Liquorice root has stimulatory properties similar to aldosterone. If you eat real liquorice in addition to your fludrocortisone medication, you risk over-medication. Liquorice root is not recommended as a substitute for fludrocortisone as it is not possible to predict the strength or consistency of the product. Most liquorice sweets are, in fact, only liquorice-flavoured and can safely be eaten - but do check the list of ingredients. Some cough mixtures also contain real liquorice.

There are no herbal therapies available which can mimic the effects of natural human cortisol. Until steroid medication became available in the 1950s, Addison's disease inevitably resulted in death over a period of time. Addisonians who experiment with 'natural' alternatives to their normal medication risk the same fate.

Complementary therapies such as meditation, massage and yoga can offer benefits. Meditation can be a useful way of dealing with a build-up of fatigue, tension or irritability during the day. Massage and yoga can help with the joint and muscle aches which a number of Addisonians experience. Regular physiotherapy can also bring real relief for joint and muscle aches.

"I know of another male Addisonian who lived 20 miles north of here, but he was 'cured by faith' and threw away his medication. He dropped dead in his house three days later." JAMES

j) STORING YOUR MEDICATION

Most prescription medication is stable when stored between 0° and 25° Celsius (32° and 77° Fahrenheit). Always check the expiry date on your medications, especially your injection kit, and dispose of any medication that reaches its expiry date. Many medications have a shelf-life of around two years, so it is seldom worth keeping more than six months' supply at home.

At temperatures higher than 30° Celsius (85° Fahrenheit), most medication will reach the end of its shelf life more rapidly than the stated expiry date and may start to deteriorate (lose its effectiveness). For this reason, any medication that you carry close to the body in a pocket or bag should be used within a few days, unless you are storing it inside an insulated travelling container. Insulated travelling containers can be ordered through most pharmacists. If you are carrying an injection kit with you in this kind of hot weather, it should be discarded after 12 months, or sooner if

the contents become cloudy or coloured. Injection kits are discussed more fully in sections 9 and 10: *Travel and Crisis Management*.

MEDICATION: A SUMMARY

A combination of hydrocortisone and fludrocortisone is the most usual treatment for Addison's disease.

A variety of blood tests are available to help you and your doctors establish your correct baseline dosage.

You may feel better splitting your daily hydrocortisone medication into three divided doses.

Taking the first dose of the day on waking is recommended.

Individual doses of 20mg or less do not usually need to be taken with food.

The last dose should be taken no later than 4 hours before bedtime.

4 SOME COMMON CONCERNS

Over the course of a lifetime living with Addison's disease, many people find themselves addressing one or more of the following areas of concern. For simplicity, these have been grouped into three topic areas: experiences common to many Addisonians; experiences associated with low cortisol levels; experiences associated with high cortisol levels.

Please remember that, because most Addisonians take several divided doses of medication a day, it is possible to have experiences associated with both high and low cortisol levels over the course of a day. Don't be alarmed if you do have any of the over or under-medication symptoms mentioned below; with experience you will learn to adjust for them. Talking it through with your doctor, reaching your optimum daily dose and (from time to time) self-medicating as outlined in the sections below, will all minimise the frequency and severity of these symptoms.

a) EXPERIENCES COMMON TO MANY ADDISONIANS

i. Osteoporosis

Osteoporosis is more common among Addisonians than among the population at large.

In this manual, osteoporosis is used as a general term to cover the various types of bone disorder that can be detected by a bone scan (the medical terms used are often *osteopenia*, *osteomalacia* and *osteoporosis*).

Post-menopausal women are most vulnerable, but medical studies of bone density among Addisonians have found some evidence of bone thinning even among younger men. Low levels of DHEA are thought to be partly to blame.

Long-term use of higher doses of steroids is also commonly associated with osteoporosis. By taking the minimum daily dose of hydrocortisone needed, you should not be at increased risk of bone thinning from your steroid medication. But anyone who is consistently over-medicated is increasing the chances of osteoporosis.

Because of these associations, every Addisonian is recommended to take preventative steps to reduce their risks of bone thinning. It is never too early, or too late, to start working on your bone density.

"I have had Addison's for 26 years. I used to be on 40mg hydrocortisone and had done pretty well. Then eleven years ago I was diagnosed with insulin dependent diabetes and found that the insulin was messing up the medication for Addison's, so they switched me to prednisone and fludrocortisone. Now a month ago I had a hypoglycaemic seizure and ended up with several compression fractures in my spine. After the x-ray they also found four old compression fractures which had already healed. At which point they tell me I have osteoporosis and I am 40 years old. The bone loss has wiggled me out because I am a landscape gardener by trade and do a lot of heavy lifting." BRIAN

ii. What can I do to prevent osteoporosis?

■ You should make sure your diet includes plenty of calcium and vitamin D. The UK climate makes it easy to run low on vitamin D. Supplements containing calcium and vitamin D are available over the counter for those who do not like dairy products, and are also available on prescription from your GP.

■ Exercise helps to build your bone density. This must be weight-bearing exercise (so swimming will have no effect) but walking is excellent. A daily brisk walk of 20 minutes is sufficient, while a gym workout of 30 minutes, three times a week, is even better.

■ Remember that low-calorie diets that encourage the body to burn ketones also cause calcium loss from the bones, and so can advance osteoporosis. A well-balanced diet, as discussed in section 5: Diet, is best.

■ Post-menopausal women are recommended to use hormone replacement therapy as this helps to give protection against osteoporosis.

■ Bone density scans are available. You are recommended to request a bone density scan every five to ten years. Women should make sure they have a bone density scan at the time of menopause, if they have not already had one. You may wish to request a bone density scan soon after diagnosis to establish a baseline for the condition of your bones, although these are not necessarily always readily available in the UK through the NHS.

■ Prescription medications which act to rebuild bone density are available and have been proven to have positive results for those who are already experiencing noticeable levels of bone thinning.

“I take Fosamax daily for my osteoporosis, which is due to prednisone use. I also take calcium with vitamin D. With the help of these, my last bone density study was normal. Previously I had one compression fracture. So Fosamax has worked for me. I just have to keep taking it every day.” SALLY

iii. Weight gain: are steroids to blame?

Before diagnosis, many Addisonians lose a lot of weight. Some of this is because the disease causes them to lose their appetite and they are taking in fewer calories than a healthy person. An important part of this weight loss is also a loss of normal body fluids, as the electrolyte balance becomes destabilised.

Rapid weight gain in the first few weeks of starting medication is usually due to a restoration of the fluid balance. Following this, however, it is easy to put on stored fat. This is partly because most of us are cautious about taking up strenuous exercise after a period of severe illness.

And steroids generally do increase the appetite. The difference in appetite can be subtle so that the weight gain can be slow but sure. In addition, people taking steroid medication can find it harder to mobilise stored fat, so weight once gained is not so easy to lose.

iv. What helps to shed those excess pounds?

If you feel hungry frequently and are eating in quantities such that your weight concerns you, then gradually reducing your daily steroid dose may help you to shed some of this weight.

If you are experiencing symptoms of energy-lag and low blood sugar without necessarily feeling hungry - and you are snacking on food high in sugars and fats to keep you going through the day - then reducing your total daily dose may not be the most helpful way to lose weight. Instead you may need to review the timing of your divided doses so you can maintain your energy levels and cut back on your intake of snack foods.

Like the general population, most Addisonians would find it easier to maintain their ideal body weight if they exercised more regularly.

“I’ve put on 30 pounds with Addison’s. I’ve become aware of how much I was trying to self-medicate with food - I’d nibble on stuff constantly and lately lots of candy in an attempt to get a pick-me-up from the exhaustion I was feeling. It was the only way I knew besides drinking tons of coffee to try to fix feeling tired.” GLENNA

“When I was first diagnosed I was put on too high a dose of prednisone and started gaining weight quickly. Then I changed to 20mg hydrocortisone daily. By being careful to eat minimal carbohydrates I am slowly getting some of the extra off.” CATHY

“I gained well over 10 kilos while I was pregnant and breastfeeding. Then I lost it all again over 12 months as the children got older. I was getting more exercise just pushing the buggy and chasing after them in the playground. I also realised how much dried fruit, chocolate and crisps I had been eating to keep my energy up. I started eating almonds, fresh apples and carrots as my snack foods instead, and broke up my hydrocortisone dose so I was taking smaller amounts more often through the day to keep my energy up. Then I got rid of around one kilo a month.” CATHARYN

b) EXPERIENCES ASSOCIATED WITH LOW CORTISOL LEVELS

v. Why do I feel lousy at certain times of the day?

The most common reason for feeling lousy at certain times in the day is because your cortisol levels are lower than your body needs. Typically this happens when the gap between one divided dose of hydrocortisone and the next is too long. The majority of UK Addisonians who took part in the 1996 survey were taking their hydrocortisone in just two daily doses. More than two-thirds also said they experienced difficulties with low blood sugar at least some of the time.

If you are experiencing any of the symptoms described right, it may mean you need to adjust your hydrocortisone medication so you take it in smaller, more frequent amounts.

■ Low blood sugar (hypoglycaemia) is a common response to low cortisol levels. This usually means feelings of shakiness, wobbliness, irritability, faintness and hunger. Some people simply describe it as ‘brain fog’.

■ Recurrent headaches or joint aches can also be a response to low cortisol levels.

■ Loss of appetite and feelings of mild nausea are a common response to low cortisol levels and will be familiar to many Addisonians from their illness prior to diagnosis.

■ Severe nausea and vertigo are usually more serious indicators of low cortisol levels and are discussed further below.

The point in the day when Addisonians most commonly experience low cortisol levels is mid-afternoon, but some people have similar symptoms when they first wake up.

If you are experiencing symptoms which do not ease once you have taken your next dose of hydrocortisone then you need to raise them with your medical practitioner.

vi. Low blood sugar and the insulin response

Low blood sugar can be accompanied by snack food cravings as the body seeks instant energy sources from high-sugar and high-fat foods. Sadly, a high sugar/carbohydrate/fat loading tends to perpetuate the cycle of energy-lows. This is because these types of snack foods trigger the pancreas to produce large amounts of insulin. The insulin rapidly converts the available blood sugars to stored fat, leaving you with low blood sugar levels again. This process happens more rapidly in overweight people, which can seem particularly unfair.

Some Addisonians swear by a protein-rich snack as the best way to combat low blood sugar or 'brain fog'. Scrambled eggs and nuts are among the pick-me-ups recommended.

vii. Salt and other cravings

■ *Salt cravings are common among Addisonians. We tend to lose salt faster than people with healthy adrenals, so a moderate intake of salty foods is sensible. If you find your desire for salt becomes unusual and you are also experiencing a recurrence of some of your pre-diagnosis symptoms - like dizziness on standing - then you need to raise this with your doctor and have your medication levels checked.*

■ *Cravings for acid tastes such as lemon juice are also common. Some Addisonians have confessed to a liking for pickle juices and other less common acidic flavours.*

■ *Liquorice cravings are unusual and may indicate that your fludrocortisone medication needs increasing. It is important to have this checked with the relevant blood tests before adjusting your fludrocortisone medication.*

■ *Sugar cravings are common in the population at large. However, if you are getting sugar cravings at the same time each day, this may indicate that the timing between your divided doses is too long for you.*

viii. Feeling very thirsty

If you are feeling thirsty all the time and are passing a lot of urine (as many Addisonians experienced prior to diagnosis) this may indicate that your fludrocortisone medication needs increasing. However, these symptoms may also indicate diabetes or a kidney problem, so it is important that you raise them with your doctor and have the relevant blood tests done before adjusting your fludrocortisone medication.

Some people find that occasional symptoms of unusual thirstiness are a useful early warning sign that their hydrocortisone cover is running low and they need to bring the timing of their next dose forward. Large amounts of alcohol, coffee or tea can cause dehydration and Addisonians are especially vulnerable to this.

iv. Dry skin

Dry, cracked skin is a common problem. Low DHEA levels are thought to be partly to blame. Excessively dry skin can also be associated with hypothyroidism, so you may wish to ask for a blood test to check your thyroid function if this is a persistent problem. Most chemists can offer a good range of soothing emollient creams.

"For three years I had a terrible time with headaches.

I saw a parade of doctors of various stripes, had CAT scans, MRI scans and all kinds of tests and drugs. Finally a glucose tolerance test showed that I overproduce insulin in response to a glucose load. As a result, two or three hours after a big carbohydrate meal my glucose level is way too low, I feel terrible and have a mammoth headache. The headaches are due to this hypoglycaemia and the switch in metabolism, which produces ketones. Since eating more protein and less carbohydrate I have no headaches!" DICK

x. Extra pigmentation

Extra pigmentation - an unusual tan or new growth of freckles and moles - is a common feature of Addison's disease prior to diagnosis. For most, this extra pigmentation starts to fade within weeks of beginning medication.

The physiological cause of the pigmentation is high levels of the pituitary hormone ACTH, which stimulates the adrenals. For some individuals who have had long-standing severe Addison's symptoms, the pituitary loses its ability to reset the ACTH levels back down again. In these cases, the extra pigmentation may not fade over time as completely as it would for others.

If you find you are developing new pigmentation while you are taking your normal medication, this could be a sign that your ACTH levels have risen again in response to low blood cortisol. Your doctor may suggest switching to a longer-acting form of medication, or taking your last divided dose later in the evening to damp down your pituitary ACTH levels.

xi. Exhaustion and 'hitting the wall'

Addisonians tend to rely on stored adrenaline to get them through events which are more stressful or physically demanding than usual, to compensate for the fact that our bodies do not produce the boost in cortisol levels that a healthy person could draw on. Then, when the adrenaline rush finishes, we 'hit the wall'. If you are exhausted enough to need bed rest to recover, then a top-up to your normal daily dosage of hydrocortisone is likely to be justified. A little extra, equivalent to one-quarter of your normal daily dose (5mg for someone on 20mg hydrocortisone), may be all that is needed, depending on the severity of your symptoms.

Over time, most Addisonians come to recognise the kinds of stressful and demanding situations that cause them to reach a state of exhaustion. For some people, it may be giving a conference presentation, for others, it may be big family events such as weddings.

If you are one of those people who find that they reach a state of exhaustion on a regular basis, it is worth keeping a record of the events which lead up to this and reviewing it with your doctor. You may need to adjust your daily medication dose, or you may simply need to be aware of the circumstances in which you have to 'top up' your regular medication. Bear in mind that top-ups should be used sparingly, as taking too much steroid makes you more likely to develop osteoporosis.

xii. Nausea and vertigo

These are the two symptoms most commonly reported by Addisonians who are close to exhaustion or who are running very low on cortisol. For many people these symptoms are preceded by a headache, joint aches or other tell-tale signs. If you experience strong feelings of nausea and vertigo, you need to take your next dose of hydrocortisone immediately. You are likely to need a top-up on your normal daily medication of anywhere between one-third to double your normal daily dose, depending on the severity of your symptoms. Do not be afraid to ring your doctor and ask their advice on how long you should increase your hydrocortisone dose. Experience will teach you what additional dose you need. Make sure you let your doctor know of your symptoms.

"I've started to figure out my symptoms when I am running low. First the fatigue gets worse then I am painfully cold, then come the muscle cramps, then nausea, then vomiting. If I take extra medication in the beginning I seem to break the chain of events. Sometimes too I find that when I am extra tired, feel like I am moving through mud and it hurts to move, then I get dizzy. If I take a bunch of salt, in about half an hour I can feel better!" SHELLEY

c) EXPERIENCES ASSOCIATED WITH HIGH CORTISOL LEVELS

xiii. Insomnia and other sleeping difficulties

Difficulty getting to sleep or getting back to sleep after a middle of the night waking is very common in the population at large and is usually associated with anxieties about work or family life. A small number of Addisonians report that they had difficulty sleeping prior to diagnosis but most report that they were sleeping more than usual as part of their fatigue.

After diagnosis you may find it difficult to get to sleep if you take your last divided dose of the day too late in the evening. The recommended time for the last daily dose is around 6pm, or at least four hours before going to bed.

People with Cushing's disease (who suffer from abnormally high cortisol levels through the night) typically report problems in sleeping. If you are repeatedly having sleep difficulties and are confident there is no other cause, it may be a sign that your total daily dose is higher than you need.

xiv. Anger, mood swings and depression

These feelings are very common in the population at large, so need to be interpreted with caution. However, anger, mood swings and depression can be associated with high levels of cortisol and are noted among people with Cushing's disease. If you feel troubled by these feelings, it may be worth reviewing your medication levels with your doctor and trying out a gradual reduction in your hydrocortisone dose.

A number of Addisonians report that their adrenal symptoms were mistakenly attributed to depression prior to diagnosis. However, a small proportion of Addisonians do also experience clinical depression, and report that they have benefited enormously from seeking medical help and taking anti-depressant medication.

xv. Fluid retention

Puffy ankles and other signs of fluid retention can have a wide variety of causes, including kidney problems and *hypothyroidism*. However, for Addisonians, the first thing to check is your medication levels, as both fludrocortisone and hydrocortisone can affect the fluid balance. A common cause of fluid retention is more fludrocortisone than your body needs. As mentioned earlier, it is important to have the relevant blood tests done before adjusting your fludrocortisone medication.

xvi. Easy bruising

In theory, if you are on the correct steroid dose, you should not experience any of the side effects of steroid medication. In practice, some of the minor side-effects are hard to avoid even on modest doses. Easy bruising is one of these. It results from the effects of hydrocortisone on the capillaries, making them more fragile.

xvii. Thinning skin

A slight amount of thinning skin occurs gradually as we age. However, a sudden and noticeable thinning of the skin can be a side-effect of steroid medication. It results from excess levels of hydrocortisone, which cause protein loss from the body. If you are experiencing this at the same time as some of the other symptoms of excess cortisol described below, this may be as a warning sign that you need to review your medication levels with your doctor and gradually reduce your hydrocortisone dose.

xviii. Muscle weakness/wastage

Some Addisonians lose a lot of muscle strength during their illness prior to diagnosis. It takes hard work and determination to regain this once your medication regime is stabilised, but the experience of other Addisonians shows that it can be done. Also, it is worth remembering that a slight loss of muscle strength occurs gradually as we age.

However, muscle weakness can be one of the few signs of a potassium imbalance and may suggest that your renin and electrolyte levels need to be checked by your doctor.

Prominent muscle wastage can be a sign that you are over-medicated and your hydrocortisone levels are too high. People with Cushing's disease typically experience a thinning of their upper arms and thighs and a sense of muscle weakness. If you are experiencing this at the same time as some of the other symptoms of excess cortisol, you should probably treat this as a warning sign that you need to review your medication levels with your doctor and gradually reduce your hydrocortisone dose.

xix. Puffy/moon face

This can have a wide variety of causes, including hypothyroidism. However, it is also typically found among people with Cushing's disease. If you are experiencing this at the same time as some of the other symptoms of excess cortisol described above, you should probably treat this as a warning sign that you need to review your medication levels with your doctor and gradually reduce your hydrocortisone dose.

5 DIET

Like most people, Addisonians can burn the candle at both ends, eat badly, drink too much alcohol, tea and coffee, and survive. But we are particularly susceptible to the effects of dehydration and have a tendency to lose salt faster than other people. This means that we need to pay attention to our diet to ensure that our mineral balance is stable. And because we tend to have difficulty mobilising stored fat, we are more susceptible to the problems of weight gain when our diet includes too much sugar and fat.

i. Plenty of fluids, protein, vitamins and minerals

You do not need to follow a special diet, unless you have related health conditions like diabetes, coeliac disease or osteoporosis. But it is important to follow a healthy, balanced diet, which includes plenty of fluids, protein, vitamins and minerals without too many sugars or fats.

ii. Tastes and cravings

It is quite common for Addisonians to develop a taste for acid and salty flavours in combination. (The salt and lemon juice chaser that traditionally accompanies tequila must have been invented by an Addisonian!) Mediterranean foods are often a healthy way to satisfy these tastes: fresh salads with olives, pickles, salty cheeses, tuna or anchovies provide a good balance of protein, vitamins and minerals.

iii. Multi-vitamins and other supplements

Multi-vitamin and mineral supplements can be reassuring, so that your body is getting the best balance of nutrients at all times. However, if you are eating well they should not be necessary unless you have other specific health conditions.

Addisonians tend to retain potassium as well as lose sodium, so it is best to avoid supplements that are high in potassium. Fruits that are high in potassium, such as bananas, apricots (especially dried), dates and raisins, should not be eaten in copious amounts.

If you have a family history of osteoporosis or you have had a bone scan showing some loss of bone density, your doctor may recommend calcium supplements and vitamin D as preventative measures.

Some general guidelines for healthy eating are as follows:

DIET: A SUMMARY

You need to maintain a healthy diet to ensure you do not experience dehydration or a mineral imbalance.

You will tend to put on weight easily if you eat too much sugar, refined carbohydrate or fat.

A healthy, balanced diet means plenty of fluids, protein, vitamins and minerals with not too much sugar or fat.

Multi-vitamins and other supplements can be reassuring but should not be necessary if your diet is healthy.

PLENTY OF FLUIDS

Aim to drink at least one and a half litres of water a day.

Try to drink a glass of water for every glass of alcohol, cola drink, or cup of tea/coffee you have.

Avoid sports drinks with added potassium.

Remember to drink more fluids in hot weather.

PLENTY OF PROTEIN

Nuts are a good snack food.

Dairy foods (cheese/milk/yoghurt) provide protein and calcium.

Oily fish (salmon/sardines/mackerel) are a good source of calcium and protein.

Peas, beans and other legumes are a valuable protein source for vegetarians.

PLENTY OF VITAMINS AND MINERALS

Keep your calcium intake up to prevent osteoporosis.

Aim to eat more salty foods in hot weather.

Stick to the rule about at least five servings of fresh fruit and vegetables a day.

Take a multi-vitamin supplement if you wish, but it should not be necessary.

Avoid supplements which are high in potassium.

NOT TOO MUCH SUGAR

Refined carbohydrates (white bread and most bakery goods) are quickly converted to sugar by the body. They need to be balanced in a meal with protein and fibre to avoid a 'sugar crash'.

Complex carbohydrates (whole grain cereals) provide a more sustained source of energy and are less likely to produce a 'sugar crash'.

Fresh fruit is a good source of sugar because it is balanced with fibre.

Treat chocolate biscuits and the like as an occasional luxury.

The best time to eat sugar is at the end of a meal - so the occasional pudding is a treat you deserve!

NOT TOO MUCH FAT

Watch for the 'hidden' fats in takeaway foods and ready-made meals.

Try grilled foods rather than fried foods.

Butter is okay when applied sparingly rather than lavishly!

6 EXERCISE

a) EXERCISE IS GOOD FOR US (SO WHY ARE WE COUCH POTATOES?)

It keeps our hearts and lungs healthy, it keeps our bones and muscles strong. It keeps us from stiffening up as we get older, so that we enjoy middle age and old age more. Walking is an excellent form of exercise for Addisonians; it is gently invigorating and helps to prevent osteoporosis. More strenuous exercise is even better. Naturally, in this age of the car, very few of us exercise as much as we should.

Some Addisonians run marathons and compete in endurance sports. Others climb mountains and travel to remote places. Some of us are couch potatoes who feel exhausted after changing into our gym gear. In this respect we have the same broad range of fitness levels as the general population.

In short, being Addisonian does not stop you being a fit, athletic individual. However it does mean you have to plan your fitness regime carefully, to ensure your medication levels remain adequate for the physical demands of your particular sport or recreation.

“For the past five months I’ve been taking karate and I really enjoy it. For a person of 46 with primary Addison’s, hypothyroidism, a spastic bladder and irritable bowel I guess I’m doing pretty well. Since I’ve been taking karate my energy level has improved and most of the pains in my joints have left me. Now if I could just get this weight off!” KAREN

b) ADJUSTING YOUR MEDICATION TO YOUR FITNESS REGIME

■ *When you undertake a form of exercise that is far more physically demanding than usual, you may need to take a little extra medication beforehand.*

■ *When you undertake a form of exercise which you are used to and which your fitness level means they do not find especially demanding, you do not need to take extra medication beforehand, unless it is of long endurance.*

■ *Remember to drink plenty of fluids during and after exercise, to ensure that your mineral balance remains stable.*

■ *Avoid sports drinks with added potassium.*

Most people establish by trial and error when they need to ‘top up’ their medication and when they do not. The general aim is to keep any extra medication to a minimum while gradually building up your level of fitness. For physically challenging sports, such as competitive running, it is best to plan your training regime and the amount of steroid cover you will require beforehand, with advice from an endocrinologist.

Some approximate guidelines are described right. If you are in any doubt, consult your doctor about your medication requirements before you start on a new fitness programme.

EXERCISE: A SUMMARY

Exercise in all weight-bearing forms - including walking - is an important way to help prevent osteoporosis.

Gentle exercise does not need any extra medication.

Physically challenging exercise may need a ‘top up’ to your normal medication.

For competitive sports, such as marathon running, it is best to consult an endocrinologist about your medication regime.

For the Addisonian, it can also seem like harder work to increase your fitness level than it is for someone with healthy adrenals. This is because your body finds it more physically demanding to do something to which it is unaccustomed. So, the first time you go to a gym class will feel like much harder work than it will once you have done the same class a few times.

Starting a new sport or fitness programme, such as:

■ 60 minute ‘cardio’ workout gym class

Try taking an extra quarter of your normal daily dose (eg. 5mg hydrocortisone for someone on 20mg), 60 minutes before the first class starts.

Then, on the next occasion, try a ‘top-up’ of half that amount before the class.

Try the third class with no ‘top up’.

Remember to drink extra fluids after the class.

Returning to a sport or fitness programme you used to do before your diagnosis, such as:

■ Running, swimming or lifting weights

Start out gradually, eg. run just for 5 minutes the first day, so that you are not over-stretching yourself.

Aim to do a little more each time you exercise without needing to take extra medication.

Training for and taking part in a challenging sporting competition, such as:

■ Marathon

■ Mountain marathons

■ Other competitive mountain sports

Consult your endocrinologist first.

Some runners have successfully completed a marathon on less than double their normal daily dose, others may need more than this.

Mountain sports may need considerably more due to the altitude, cold and other physical demands. (Anywhere up to ten times normal daily dose).

Arrange to have plenty of rehydrating fluids and an emergency medical kit to hand along with an escort who can use the kit if absolutely necessary.

A MARATHON RUNNER’S EXPERIENCE:

“I was diagnosed with Addison’s two years ago at the age of 45. I struggled to find good advice as to whether I would be able to get fit again and run another marathon. After a year I decided to start serious training. For long runs of 17 miles plus I took an extra 10mg of hydrocortisone and doubled my fludrocortisone in the morning. I also took electrolyte stamina tablets and made sure that I was well hydrated before and during running. This worked well for me during training runs and the marathon itself. I had absolutely no problems other than the expected sore muscles. The run itself went extremely well, no ‘wall’ at all and I managed a strong finish to creep in just under four hours. I do not believe that being an Addisonian increases the challenge of marathon training by too much. You just have to be more aware of how you are feeling and take the necessary precautions if you start to feel bad. There is no magic formula, just trial and error. You need to remember you will have good running days and bad running days, just like non-Addisonians. After all, training for and running a marathon is tough on the body.” BRIAN

7 PREGNANCY

Many women who develop primary Addison's disease are diagnosed around the age of 40, by which time most have had their families already. For younger women, it is possible to have a healthy pregnancy and normal childbirth with Addison's, provided that you ensure you receive good medical care throughout the pregnancy and maintain your medication regime carefully.

a) BECOMING PREGNANT

In general, Addison's disease does not affect a woman's fertility. A few women have become pregnant despite advanced, untreated symptoms of the disease. Women with Addison's disease have also successfully completed IVF treatment to become pregnant.

However, Addisonian women may find that they become temporarily infertile if their medication regime is severely inadequate and they are experiencing pronounced symptoms of steroid insufficiency. A small proportion of women with Addison's also develop premature ovarian failure.

"I delivered my firstborn daughter just six months ago. I was lucky enough to conceive straight away and had a fairly uneventful pregnancy until about 7 and a half months. I began having 'sinking' spells in the morning where I felt very weak and had trouble breathing. After initially being told this was 'normal' in late pregnancy my endocrinologist increased my steroids and I felt much better. I delivered vaginally with a 100mg dose of intravenous hydrocortisone prior to delivery. My baby is healthy; she's a big girl (95th percentile) and weighed 9 pounds 3 ounces at birth." WENDI

b) MANAGING YOUR PREGNANCY

During pregnancy most women simply need to continue with their pre-pregnancy medication levels. However, some women find that their medication requirements go either up or down. So it is important to arrange for specialist monitoring of your pregnancy from the first months. It is most common for women to need a slight increase in their medication during the last three months, due to the gain in weight and body fluids.

c) CHILDBIRTH

Giving birth is a highly demanding physical activity, so it needs extra steroid cover. This is usually provided as a hydrocortisone injection at the onset of the second stage of labour. If the first stage of labour is prolonged, you may also need a steroid injection then. After the birth your steroid cover should be returned to your normal medication levels as rapidly as possible. Usually, you can go back to your normal dose immediately after the birth.

If you are having a caesarean this will also need extra steroid cover, the same as for any other form of major surgery.

A caesarean requires extra steroid cover during the operation and for 48 hours afterwards. This can either be provided in the form of steroid injections every 8 hours or as a drip with intravenous hydrocortisone. Major surgery such as a caesarean usually requires that you have no food or drink from 8 hours before the operation. A drip with saline solution can prevent any risk of dehydration occurring during this period so ask your anaesthetist if this can be provided. After 48 hours your steroid cover should be returned to your normal medication dose as rapidly as possible. Usually, you can go back to your normal dose immediately.

Needless to say, home birth is not recommended.

"I have had my three children since I was diagnosed with Addison's. I did fine on my normal medication until the end when I got pretty big. I had to be induced with all three and found that it was important to get a drip with intravenous hydrocortisone going as soon as labour started. I also had to stay on increased steroids for about ten days after delivery. My kids were very healthy and weighed 7 pounds 3 ounces, 9 pounds 5 ounces and 10 pounds 1 ounce at birth. I felt better when I was pregnant than I do when I'm not. My kids are now seven, five and two." LINDA

d) CARING FOR A NEWBORN BABY

During the early months of a newborn's life they require round the clock care. If you are spending long periods of time up in the middle of the night caring for your newborn(s), you may need to adjust your medication regime. Try taking your medication

more frequently, in smaller amounts to match the periods when you and your baby are awake and asleep. You can safely take your first dose of the day as early as 3am, if that is when your baby wakes you.

e) INHERITANCE

In most cases, Addison's disease is not inherited. In a few cases it can be, as mentioned in section 2: *Related autoimmune conditions*. Ask yourself if any of your extended family has Addison's or a related autoimmune condition? If so, there is a small

possibility that your child might develop an autoimmune endocrine condition. In those rare cases where children develop Addison's disease, the symptoms usually become apparent during their teenage years, rather than during early childhood.

8 BEING THE PARENT OF A CHILD WITH ADDISON'S

a) MONITORING YOUR CHILD'S CONDITION

As the parent of a child with Addison's, there are practical and emotional issues surrounding their care, which you will need to address. At the practical level, you need to be able to monitor their condition and be alert to any need to adjust their medication. This means becoming as well informed as you can about the disease, its signs and symptoms, the circumstances which might trigger the need for an increased dosage of medication and how much extra might be needed. It also means being trained so you can give an emergency injection if you have to.

The more you can discuss these practical issues with your child's endocrinologist, the better equipped you will be. You may find it helpful to give them a written list of your questions and the issues you would like to be educated about in advance of your appointment, so they have time to collect the appropriate information.

The amount of steroid medication to prescribe for children and teenagers is a fine balance and is likely to require some experimentation to get it right. Too much can stunt their growth. Too little can leave them lethargic and unable to concentrate on their schoolwork. In general, hydrocortisone is preferable to prednisone or dexamethasone, as it is less likely to interfere with their growth.

In turn, you will have to help your child learn to manage their health as far as possible. Some of the things they will need to be aware of are:

- *Remembering to drink plenty of fluids so they stay hydrated (especially on school trips and sports days).*
- *Remembering to take their pills at the right time of day, so they do not run low on steroid cover.*
- *Being alert to the early warning signs that they are running low on steroid cover, such as a headache, stomach ache or wobbly knees.*
- *Remembering to carry their 'emergency' pack with extra hydrocortisone and their injection kit when they go on school trips.*

Some schools have a trained nurse who is able to give injections. You will need to talk to the staff at your child's school and establish what level of first aid care they are able to give for your child's condition, should it ever be necessary.

At the emotional level, you need to be aware of your own emotional needs and the anxieties that surround being the parent of a child with a chronic illness. You need to find ways to get personal and practical support from your circle of family and friends, so that you can stay calm and caring towards your Addisonian child and their siblings.

There is a helpful publication on caring for your Addisonian child, "Our Addisonian Kids" listed in section 11.

"It's hard for a parent to see their child go through this disease. We can take them to the doctor, do what they say, but we can't make it go away. Since my son has been sick with Addison's my life has been so stressful: going to all different kinds of doctors, holding down a job, and a household with three other kids who need me as well. I feel like I am running all the time. I am so tired. With my son, I do a lot of joking around to make him smile. When he smiles, it makes me feel good." **KAREN**

b) SUPPORTING YOUR TEENAGER

Developing a chronic illness during the teenage years can be doubly challenging for both the teenager and their parents. Particularly where the teenager has been severely ill before diagnosis, they will have experienced anxieties and emotions, which are difficult for them to describe or resolve. Recognising this fact can make it easier to move on. It may help your teenager if you are able to explain your own emotions in a straightforward way.

A teenager is preparing to become an adult and take full responsibility for managing their own health, but they have not yet become that adult person. As a parent, you need to be able to step back where appropriate and move in again when they need more practical help from you.

- *Encourage your teenager to educate themselves as fully as possible about the disease.*
- *Make sure they understand the potential consequences of skipping doses.*
- *Help them become competent to give themselves an emergency injection.*

During their growth spurt, your teenager's medication may need to be adjusted more frequently. Again, you need to be alert to the early warning signs that they are running low on steroid cover and raise any important changes in the general health with their endocrinologist.

"I am a 17 year old girl who has recently been diagnosed with Addison's. I also have several other autoimmune complications. I have some advice for you on how to handle your children's illnesses. Please be as strong as you can for them and make sure you talk about it. My mother tried hard but I ended up having to be the strong one and 'protect' my parents when I was very ill. I never felt I could talk about it with my family because they would cry uncontrollably. And please make sure your children know that you don't think they're doing it to themselves when they are ill. Because after a while of people telling you it's all in your head you start to believe them." **KATIE**

9 TRAVELLING AWAY FROM HOME

a) TAKE ENOUGH STEROID COVER TO ALLOW FOR EMERGENCIES

Whenever you are travelling away from home you should carry a medical kit to allow you to manage an emergency situation. The more remote from medical supplies your travels will take you, the more extensive that medical kit needs to be. The situations in which you might need to use your emergency medical kit are discussed in the next section, *Crisis management*. The injection kit and how to use it are also discussed in the next section.

- For an overnight stay out of town, it is probably sufficient to have a couple of days extra medication on hand plus your injection kit
- It is recommended practice to keep a second injection kit in the car as well, to protect you from the effects of shock in the event of a serious motor accident. Where possible this should be stored inside an insulated container so that the temperature changes in the car do not cause it to deteriorate. An insulated container can be ordered through most pharmacists, to protect against heating to more than 25° Celsius (77° Fahrenheit). If it is not possible to store your car injection kit in an insulated container, the best approach is to discard it after 12 months, or sooner if the contents become cloudy or coloured.

■ For an overseas trip, you need to take enough extra medication to allow for a period of doubling your normal dose should you develop a fever, plus your injection kit and some anti-nausea suppositories or tablets, should you develop a stomach bug.

■ Always carry your medication - or at least enough to cover you for a week - in your hand luggage when flying abroad. Where a suitcase goes astray, it can often take some time for the airline to recover and return it.

■ For a trip to remote areas where medical supplies are unsure, you may need to take some intravenous saline and spare sterile needles as well as plenty of medication, anti-nausea tablets or suppositories and your injection kit.

■ A letter from your doctor explaining why you are carrying syringes and what medical treatment you require in an emergency is also a good idea. If you can get this translated into the language/s of the places you are visiting, this will be even more valuable in an emergency.

Of course, the chances are that you will never need to use the extra medical supplies you carry around the world with you. However, with Addison's, it is always best to be prepared.

"I have had Addison's for eleven years. I am now 65. I have travelled to 18 different countries; some trips were three weeks long. I have even been travel leader for some groups. Life is too short to stay at home and do nothing. Go for it!" PHYLLIS

b) JET LAG

"As a frequent traveller with my work, I go on many business trips around Europe.

Coupled with my love of exotic trekking holidays (Thailand) and the fact that my sister lives in New York, I tend to travel a number of short haul and long haul flights a year. What works for me is to adjust my body clock and the time I take my medication during the flight. You have to remember that flying is exhausting, but after the flight I usually stick to the local time to go to bed." NICK

Unless you find air travel extremely stressful, you should not need to increase your normal medication. However, you do need to calculate your steroid cover so that you have enough to last you through the time shifts while you are travelling on a long distance flight.

The simplest approach to air travel is to keep taking your next hydrocortisone dose five to six hours after the last one, throughout the time you are travelling.

Good general advice for long distance air travel includes:

- Remember that air travel is dehydrating so you will need to drink more fluids than usual in the air. Drink alcohol, cola drinks, coffee and tea sparingly as these dehydrate the body further. If possible, carry a large bottle of water in your hand luggage. If you forget to bring your own water, be assertive about requesting extra refreshments from the cabin crew.

■ Walk around the plane as much as possible. Try to get up out of your seat every two hours to stretch your legs and keep the blood flowing.

■ Many chemist shops now stock knee-length support socks, which can help prevent the formation of blood clots that might lead to a deep-vein thrombosis ('stroke').

■ Adjust your watch to the time of your destination as the flight begins, and adjust your in-flight activities to that new time zone as well. Sleep through the in-flight meals, if necessary, to get attuned to the new time zone.

■ Try to book flights that allow you to arrive at your destination in the late afternoon or early evening local time, so that you get a night's sleep at the end of your travelling. Flights which arrive in the early morning local time will leave you tired after travelling but having to stay up all the day before you get a proper night's sleep.

c) HOT WEATHER

Some Addisonians need to increase their fludrocortisone dose a little once the temperature climbs to around 30° Celsius (85° Fahrenheit) and make sure they eat more salty foods. For example, some people raise their fludrocortisone dose from 0.05mg during the winter months to 0.1mg during the summer, or even 0.2mg during very hot weather.

Eating more salty foods than usual and drinking extra fluids help to keep the body's mineral balance stable in the heat. Carrying a bottle of water with you in your hand luggage is prudent where you are in unusually hot conditions.

Remember that in warmer weather (over 30° Celsius, 85° Fahrenheit) your medication may have a shorter shelf-life than its stated expiry date. Any medication that you carry with you in a bag or pocket should be used within a few days, unless you are storing it inside a fully insulated travelling container. If you will be travelling in areas where the temperature is consistently higher than 40° Celsius (105° Fahrenheit), we suggest you contact the manufacturers of your medication to check how its shelf-life is likely to be affected.

"I have done well, in that I work full time, have continued with my sports and activities and generally lived life as I did before. There are precautions I take with heat and travelling but changing my lifestyle never entered my mind." GWEN

10 CRISIS MANAGEMENT

“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**

“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**

a) WHAT IS AN ADDISONIAN CRISIS?

An Addisonian crisis is what happens when an Addisonian faces extreme physical stress and does not get the extra steroid cover 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**

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:

“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**

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.

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.

11 SELF HELP

Telecommunications technology and the internet have opened up good sources of information and support everywhere in the world. Below is a list of helpful contacts and sources of information:

■ **Addison's Disease Self-Help Group (ADSHG), UK organisation**

<http://www.adshg.org.uk>

Postal address:

Mrs Deana Kenward, ADSHG, 21 George Road, Guildford, Surrey, GU1 4NP, UK
Email: deana@adshg.org.uk

■ **Australian Addison's Disease Association**

<http://addisons.org.au>

■ **Canadian Addison Society**

<http://members.rogers.com/jsoutham>

■ **Joan Hoffman's site, for copies of "Our Addisonian Kids"**

<http://www2.dmci.net/users/hoffmanrj>

■ **National Adrenal Diseases Foundation (US organisation)**

<http://www.medhelp.org>

■ **Aspen Conner's adrenal insufficiency bulletin board**

<http://www.healinglight.com>

■ **Cyndi Mullet's Addison's Discussion Board**

<http://pub21.bravenet.com/forum/show.asp?usernum=1800203370>

■ **Yahoo Addison's bulletin board**

<http://clubs.yahoo.com/clubs/addisonsdisease>

■ **Boston Medical Publishing Corp.**

Online, medical outline of Addison's:

<http://www.emedicine.com/med/topic42.htm>

■ **British Medical Journal online**

<http://www.bmj.com>

■ **PubMed (medical research journal extracts)**

<http://www.ncbi.nlm.nih.gov/entrez>

■ **MedicAlert Foundation**

<http://www.medicalert.org.uk/>

Postal address:

1 Bridge Wharf, 156 Caledonian Road, London N1 9UU, UK

Tel: 020 7833 3034

E-mail: membership@medicalert.co.uk

12 GLOSSARY OF TERMS AND REFERENCES

Glossary

ACTH

Adrenocorticotrophic hormone, the messenger hormone produced by the pituitary gland which regulates adrenal production of cortisol.

ACTH stimulation test

A blood test in which the amount of cortisol in the blood is measured before and after an injection of ACTH, usually over a period of 60 minutes. Individuals with adrenal disease produce a comparatively small amount of extra cortisol in response to the injection. Different hospitals apply different thresholds to define how small an increase in blood cortisol means adrenal failure.

Addisonian crisis

Symptoms of a serious drop in blood pressure, mental confusion and extreme weakness. A potentially life-threatening situation, which requires immediate emergency treatment.

Adrenal glands:

The two small glands located just above each kidney. There are two components to each adrenal gland: the inner core (the medulla) and the outer shell (the cortex).

Adrenaline

The 'fight or flight' hormone produced by the adrenal medulla in response to stress, which raises blood pressure. Adrenaline is also known as epinephrine.

Adrenoleukodystrophy

A rare hereditary condition triggered by the abnormal metabolism of fatty-acids, which includes neurological degeneration and adrenal insufficiency.

Aldosterone

A hormone produced by the adrenal cortex which plays an important role in controlling the body's blood pressure, sodium and potassium levels and water balance.

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.

Autoimmune adrenalitis

The most common cause of Addison's disease in developed countries, where an over-active immune system attacks the adrenal tissues. The cause of autoimmune adrenalitis is not known.

Autoimmune conditions

A general term to describe the wide variety of medical conditions, which can be caused by an over-active immune system. More common autoimmune conditions include hayfever and some forms of arthritis.

Blood sugar

An important component of the blood, which is used by the body for energy. Cortisol helps to maintain blood sugar at a stable level. More technically, blood sugar can be referred to as serum glucose.

Chronic

The medical term for a long-lasting disease involving slow changes. It does not imply anything about the severity of the disease.

Coeliac disease

A sensitivity to gluten (found in wheat, barley, oats and rye) which leads the small intestine to become diseased and fail to absorb nutrients properly. Avoiding all foods containing gluten usually leads to a marked improvement.

Compression fracture

A type of bone fracture often seen with osteoporosis, where the bone crumbles.

Congenital adrenal hyperplasia

A rare hereditary condition where a recessive genetic defect causes variable enzyme defects and blocks the production of cortisol and aldosterone. It can also cause signs of virilization through over-production of male hormones (androgens).

Corticosteroids

A general term to describe cortisol hormone and the synthetic steroids which are used to replace it: hydrocortisone, prednisone and the like.

Cortisol

One of the main hormones produced by the adrenal cortex. Cortisol is essential for life; it stimulates the liver

to raise the blood sugar, mobilizes nutrients, modifies the body's response to pain and inflammation and helps to control blood pressure and sodium levels.

Cortisone acetate

One of the types of synthetic steroid, which is available to replace cortisol in Addison's disease. Cortisone acetate has largely been replaced by the use of hydrocortisone, which is more readily absorbed by the body.

Cushing's syndrome

A serious condition caused by excessive production of cortisol by the adrenals. Symptoms include weight gain (especially around the trunk), reddening of the face and neck, raised blood pressure, raised glucose levels and osteoporosis. Cushing's syndrome can be caused by an overactive pituitary gland or by an adrenal tumour.

Day curve

The most comprehensive test of hydrocortisone medication, where blood samples are taken over a number of hours before and after the normal dose of hydrocortisone medication has been swallowed. Other types of steroid medication (prednisone and dexamethasone) cannot be measured in the bloodstream in the same way.

Depression

A mental state of pessimistic or despairing beliefs where sleep, appetite and concentration are disturbed. A medically-defined state of depression is often associated with excessive levels of cortisol, especially at night.

Dexamethasone

One of the types of synthetic steroid that is available to replace cortisol in Addison's disease. Dexamethasone is more than 30 times as potent as hydrocortisone. Because it remains active within the bloodstream over a much longer period, emerging best practice for adrenal insufficiency is to use doses which are 50 to 80 times lower than with hydrocortisone. (See, for example, *SA Rivkees et al, "Dexamethasone treatment of virilizing congenital adrenal*

hyperplasia: the ability to achieve normal growth", *Pediatrics 106 (4), Oct 2000*). Over-medication with dexamethasone can easily occur with adult doses at or above 0.5mg per day.

DHEA

Dehydroepiandrosterone, the third major hormone produced by the adrenal glands along with cortisol and aldosterone. It is converted by the body to both male and female sex hormones (androgens and oestrogens). Numerous medical studies are now taking place to better understand its uses and influences in human health.

Diabetes

A metabolic disorder causing excessive thirst and large volumes of urine. There are several types of diabetes, the most common being diabetes mellitus types 1 and 2, where blood sugar and insulin levels are affected.

Diabetes insipidus

A rare metabolic disorder where the pituitary gland fails to produce enough of the hormone vasopressin. This causes the body to produce large quantities of dilute urine, leading to the need to drink large quantities of liquid and pass urine frequently.

Electrolyte

In medical terms, this refers to the concentrations of sodium, potassium and other common chemicals found in the blood. The adrenal hormones cortisol and aldosterone play an important part in maintaining normal sodium and potassium levels in the blood.

Endocrine

A term to describe the major glands in the body: the pituitary, thyroid, parathyroid, adrenals, ovaries, testes and part of the pancreas.

Endocrinologist

A professionally qualified medical specialist in the field of the endocrine glands.

Fludrocortisone

The only synthetic steroid that is available to replace the adrenal hormone aldosterone in the treatment of Addison's disease.

Glaucoma

A condition affecting the eye, where a build up of pressure inside the eyeball leads to a loss of vision.

Hydrocortisone

One of the types of synthetic steroid that is available to replace the adrenal hormone cortisol. In most cases hydrocortisone is the preferred drug for the treatment of Addison's disease.

Hyperthyroidism

Over-activity of the thyroid gland, either due to a tumour, overgrowth of the gland or Graves disease, leading to weight loss, increased appetite, rapid pulse and intolerance to heat.

Hypoglycaemia

The medical term for low blood sugar. Very low levels cause muscle weakness and inco-ordination, sweating and mental confusion.

Hypoglycaemic seizure

A reaction to very severe low blood sugar. This is often known as hypoglycaemic coma, where the individual loses consciousness following symptoms of extreme weakness, dizziness, and mental confusion.

Hypoparathyroidism

A deficiency of the parathyroid hormone, causing a fall in the blood concentrations of calcium and muscular spasms.

Hypothyroidism

A deficiency of the thyroid hormones, causing the metabolic rate to slow and leading to a slow pulse, weight gain, sensitivity to cold, delayed reactions, loss of concentration and a coarsening of the skin.

Insulin

The hormone, produced by the pancreas, which plays the main role in regulating blood sugar. Lack of this hormone gives rise to diabetes mellitus.

IVF

In Vitro Fertilisation.

Ketones

The chemical compounds that are formed when the body metabolises fat. Raised levels of ketones occur when there is an imbalance in the body's metabolism and are associated with several conditions, which may damage overall health.

Metabolism

The rate at which your body consumes energy and utilises medication. This rate can vary quite widely between individuals.

Mineralcorticoid

A general term to describe aldosterone hormone and the synthetic steroid fludrocortisone, which is used to replace it.

Normal values

Normal levels of hormones and other chemical components of the blood. For most components of the blood there is an upper and a lower limit to these normal values. The exact numbers attached to the upper and lower limit vary slightly from one laboratory to another.

Osteomalacia

Softening of the bones caused by a progressive loss of calcium. This can often be reversed by treatment with vitamin D.

Osteopenia

The medical term for a general deficiency of bony tissue.

Osteoporosis

In medical terms, the loss of bony tissue leading to bones which are brittle and liable to fracture. In this manual, used as a general term to cover the various types of bone disorder, which can be detected by a bone scan.

Pernicious anaemia

The type of anaemia that results from vitamin B12 deficiency. This can have several causes, including coeliac disease and another autoimmune condition, lack of intrinsic factor, in which the stomach does not secrete the substance needed for the intestine to absorb B12. Treatment is by injection of vitamin B12.

Pituitary gland

The most important of the endocrine glands, the pituitary regulates and controls the activity of other endocrine glands and many body processes. It is located inside the skull, just behind the eyes and tucked below the grey matter of the brain.

Plasma

Blood plasma, the straw coloured fluid that surrounds the blood cells.

Plasma cortisol

The technical term for blood levels of the adrenal hormone cortisol.

Plasma renin

The technical term for blood levels of renin, which is an enzyme released by the kidneys. Renin is important in maintaining blood pressure and renin levels rise to compensate when aldosterone levels are low.

Polyglandular autoimmune syndrome Types 1 and 2

Type 1 of this polyglandular syndrome usually appears in childhood or early adolescence, and often includes failure of the parathyroid gland and thrush (candidiasis) along with adrenal insufficiency. Type 2 of this polyglandular syndrome usually appears in adulthood and may include thyroid disease, insulin-dependent diabetes, failure of the ovaries or testes, or in a few cases diabetes insipidus, along with adrenal insufficiency.

Potassium

An essential mineral in the blood, potassium helps to maintain normal functioning of muscles and nerves. Levels are partly controlled by the adrenal hormone aldosterone.

Prednisone

One of the types of synthetic steroid that is available to replace cortisol in Addison's disease. Prednisone is roughly four times as potent as hydrocortisone and remains active within the bloodstream over a longer period.

Primary adrenal insufficiency

Lack of adrenal hormones, caused by a disease process that has directly affected the adrenal glands. The main form of primary adrenal insufficiency is Addison's disease.

Renin

An enzyme released into the blood by the kidneys which is important in maintaining blood pressure. Renin levels rise to compensate when aldosterone levels are low. Most endocrinologists use plasma renin as the indicator of when fludrocortisone medication is required.

Secondary adrenal insufficiency

Lack of adrenal hormones, caused by a disease process that has impaired the pituitary gland. When the pituitary no longer produces the messenger hormone ACTH, the adrenal glands do not produce sufficient cortisol.

Secondary adrenal suppression

Lack of adrenal hormones, caused by a reaction to high doses of steroid medication, which has impaired the pituitary gland. Because the pituitary gland's normal production of ACTH has been suppressed, the adrenal glands do not produce sufficient cortisol. In most cases, it takes months or years to reverse secondary adrenal suppression; in a few cases the suppression is irreversible.

Sodium

An essential mineral in the blood, sodium helps to maintain normal functioning of muscles and nerves. Levels are partly controlled by the adrenal hormones cortisol and aldosterone.

Steroid

A general term which can refer to the naturally-occurring steroid hormones of the body, (including oestrogen, testosterone, cortisol and aldosterone), and to the synthetic steroids used as medication.

Steroid replacement therapy

The prescription of synthetic steroids to replace a deficiency in the body's production of those hormones.

Thyroid

One of the endocrine glands, located in the neck just below the Adam's apple.

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Finally, the authors accept full responsibility for any errors which this guide may contain, and would welcome comments on its usefulness. Correspondence may be addressed to:

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QUICK SUMMARY PAGE



MEDICATION

- A combination of hydrocortisone and fludrocortisone is the most usual treatment for Addison's disease.
- A variety of blood tests are available to help you and your doctors establish your correct baseline dosage.
- You may feel better splitting your daily hydrocortisone medication into three divided doses.
- Taking the first dose of the day on waking is recommended.
- Individual doses of 20mg or less do not usually need to be taken with food.
- The last dose should be taken no later than 4 hours before bedtime.

CRISIS MANAGEMENT

- 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.

“I have lived with Addison's disease for 17 years. There is no question that the illness has its drawbacks: the need to take daily medication, the loss of energy and vitality and infrequent bouts of symptoms tantamount to crisis. In my case, the worst symptoms usually occur by my own lack of discipline in taking the medication when I should. But the illness has also brought about some very positive things. I look at life differently. I have more tolerance for people's infirmities than I did when I was 'well'. I have accepted the fact that life is short and that we are to make the most of every day - every moment. Life is better than when I was well, because I appreciate it more.”

LUKE