



The Canadian Addison Society

La Société canadienne d'Addison

ISSUE NO.6 (abridged)

Addison Info

August 1996

Are You Feeling the Heat?

From the Australian support group newsletter

One of our members in Western Australia says she finds the Amcal Rehydration Formula in sachet form very helpful to get her through the hot days or after physical exertion. You should feel relief in about a half an hour. Dr Ladliani advised this formula for anyone with Addison's disease) including those of you with diabetes, as long as it was used as a supplement to your ordinary medication and not as a replacement. If the condition persisted see your doctor.

Greta dismissed this item with her pharmacist and asked if it was available in Canada? There is a product called Gastrolyte which he recommended for Canada, and a similar liquid product for infants and children. Discuss it with your doctor.

Addison's Disease

The Adrenal gland is a small gland of approximately 1 1/2 inches lying on top of each Kidney. During embryonic development two different types of tissue come together to form this gland. There is an outer portion called the Adrenal Medulla', and an inner portion called the Adrenal Cortex. The Adrenal Medulla is essentially part of the nervous system and produces chemicals called Catecholamines (Adrenaline is the most important) which are largely concerned with what is known as the "fear, fight or flight" reaction. This is the nervous response which allows the body to react quickly to some stressful event. The adrenal medulla is not, however, essential for survival, since the rest of the nervous system can adequately compensate for its absence.

The Adrenal Cortex produces chemicals called steroids (or adrenocorticosteroids). There are over fifty different steroids but these can be divided into glucocorticosteroids, mineralocorticosteroids and some of the sex hormones.

Glucocorticosteroids are named because of their sugar regulating properties but they have many other metabolic functions and are especially important during stress (such as an injury, illness or infection). They are also important during growth and development. Cortisol (also called Hydrocortisone) is the most important member of this group.

Mineralocorticosteroids are named because of their salt regulating properties and maintain a balance between sodium and potassium, the two most abundant salts in the body. Aldosterone is the most important of this group.

The sex hormones (collectively called Androgens) are produced in small quantities by the Adrenal Cortex in both sexes and in the female this is essentially the only source for such hormones and these have a role in sexual libido and also in the distribution of the female body hair. In females, estrogens are also produced by the adrenal cortex. In premenopausal women adrenal estrogens account for only 4% of the estrogen supply, but in postmenopausal women the Adrenal Cortex is the major source of estrogens. Estrogens may also play a part in the distribution of female body hair since they can be converted to steroids with androgen activity in hair follicles.

The Adrenal gland is controlled by several mechanisms, in particular the glucocorticosteroid production is controlled by a small gland at the base of the brain called the Pituitary Gland. "This gland produces a compound called Adrenocorticotrophic Hormone (ACTH) which circulates in the bloodstream and stimulates the production of glucocorticosteroids.

The Pituitary gland itself is under the control of part of the brain called the Hypothalamus and this in turn is influenced by signals received from other parts of the brain. An excess of glucocorticosteroid circulating in the blood can signal the hypothalamus and Pituitary gland to reduce ACTH. Conversely, too low an amount of glucocorticosteroid signals the hypothalamus and the pituitary to produce more ACTH. On top of all this, the body has certain natural rhythms that control metabolic function so that the amount of ACTH and hence glucocorticosteroid is maximal about 1 - 3am.

ACTH has some effect on the release of mineralocorticosteroids but other regulators are believed to be involved, and there is also more immediate and direct control on these by changes in the levels of body salts. ACTH also has a pigment stimulation activity.

Certain illnesses require the patient to take relatively high doses of steroid e.g. Cortisol, Cortisone, Prednisone or Prednisolone for long periods. These can unfortunately disrupt the normal balance so that ACTH is permanently suppressed. In the absence of this natural stimulation, the adrenal gland will become wasted (or atrophied) so that if the tablets are stopped abruptly for any reason, the adrenal cortex will not be able to produce enough natural steroids to meet the body's requirements. The adrenal cortex can, however, usually compensate, provided the steroid tablets are reduced very slowly. When the adrenal cortex cannot produce enough of its own steroids, the condition is called: adrenocortical insufficiency.

The commonest symptoms are, however, tiredness, weakness, loss of appetite, weight loss and nausea or vomiting.

Occasionally adrenal destruction is rapid in its progress and in these circumstances, all the symptoms tend to be intensified and the sufferer is likely to be admitted to hospital in a crisis with symptoms such as nausea and vomiting, dehydration, abdominal pain, severe muscle weakness, profoundly low blood pressure and often a marked fever. A similar state can arise in known sufferers of Addison's disease who are exposed to stress. In these cases, the need for extra glucocorticosteroids is essential and a patient who does not

receive increased steroid at these times of stress may develop these signs of acute adrenal insufficiency.

Before about 1930, patients with adrenal insufficiency usually died within two years. By 1930s, mineralocorticosteroids could be made in the form of deoxycorticosterone (DOC) and this led to a considerable improvement but sufferers were still subject to the severe consequence of stressful events. By the late 1940s, glucocorticosteroids were available and these have made a profound difference to the life-style of sufferers and the outlook is excellent when appropriate treatment including education and increased cover during stress is carried out.

The patient with undiagnosed established Addison's disease is likely to be dehydrated and salt depleted; these should be corrected. In addition (and especially in acute adrenal insufficiency), there may be low blood sugar level. Glucocorticosteroid replacement needs to be instituted. In established stable adrenocortical insufficiency, some form of glucocorticosteroid replacement is also required. This is usually given as Cortisol (hydrocortisone) or cortisone acetate, but other synthetic steroids are equally effective. To meet the normal rhythms of the body, these are usually given 2/3 the total daily dose in the morning and 1/3 the total daily dosage in the evening. Most patients (but not all) will also need some form of mineralocorticosteroid replacement and this is usually given as Fludrocortisone. The natural mineralocorticosteroid, aldosterone, is not used because it is degraded too quickly by the body.

Generally, a feeling of well being returns quickly but a somewhat longer period is required for full strength to return and many weeks may be required before abnormal pigmentation subsides. Too high doses of these drugs may lead to inappropriate weight gain.

Patients with adrenal Insufficiency suffering injury (Including surgery, illness or infection need extra amounts of glucocorticosteroid and if, for any reason, this cannot be taken by mouth (as for example the patient is vomiting or has diarrhea), then it must be given into the muscle or into a vein by a doctor.

Sufferers of Addison's disease must never stop taking their replacement steroids because they have some illness. It is in precisely these situations that the dose should be increased.

For illness, such as mild infection, it is usual for one or two days to temporarily double or triple the usual maintenance dose of glucocorticosteroid. For more serious illness, much larger doses can be given but this must be supervised by a medical practitioner. Increased dosage of mineralocorticosteroid is, rarely required. Each person with Addison's disease or any other form of adrenocortical insufficiency should at all times carry an identification card or bracelet as notification of their requirements for glucocorticosteroid replacement in the event of emergency.

A Letter from a Very Helpful Doctor

Addison's disease is, as you probably know, a rare disease caused by destruction of the outer layer of the two adrenal glands situated on the two kidneys. This may be caused by T.B., fungi or unknown causes, tumours, etc. Because it is rare as a disease and often exists in a mild form and mimics other diseases, it is difficult to diagnose. For instance, loss of appetite, fatigue, etc are found in many other illnesses also. Diagnosis usually depends upon blood tests and patients do not usually like having batteries of blood tests and intravenous tests. A further confusing factor is that this disease is often found along with other diseases such as diabetes and hypothyroidism and tuberculosis. If pigmentation of the skin (brownish patches or sun-tanned appearance or even blemish spots) occurs, the diagnosis is made easier. This may occur only at a late stage. Tests depend on the ability of the adrenal glands to respond to ACTH (a hormone which can be used as a medicine but comes from the pituitary gland in the brain & directs the adrenal cortex to "start working").

Patients often have a small pulse, lack vigour and have biochemical disturbances in the blood (too little sodium and too much potassium), they lose weight and salt (sodium chloride). They also have low blood pressure and may have diarrhea or constipation and females may have period problems. The two important hormones produced by the adrenal glands are cortisol and aldosterone, they are produced in different amounts at different times by a healthy person.

These patients respond badly to stress and injuries and operations. Operations, accidents, fasting, infections etc may all cause an "Addisonian crisis", in which the patient goes into shock, collapses and may die if untreated.

Usually [a patient in crisis must be have] treatment etc in the hospital. Fortunately, much research has been done on the disease and patients are able to lead normal lives nowadays.

Treatment is aimed at simulating normal rates of secretion of cortisol (hydrocortis and aldosterone. Fludrocortisone, which is a synthetic substitute for aldosterone, has been developed in laboratories and works well. They are also given hydrocortisone. Patients have to take the above in the same way as many diabetics need insulin daily. Extra care is needed during stress, such as after an accident. They may need salt solutions to drink and extra medicines at such times.

However, patients with Addison's disease are advised to follow the treatment of their GPs and Specialists, as individual cases do vary. Infections, fever, etc. need special prompt treatment.

Patients are advised to wear special bracelets in case they faint or are injured in any way. These bracelets should give details of name, age, the disease and other details and patients should carry small syringes of hydrocortisone phosphate with them. The dentist should be informed of the disease also, well before any dental surgery is carried out. A well treated patient should be able to lead a normal, happy life. In the past this was not so, and patients often died within months.

I believe that diet is important for the prevention and treatment of diseases of the adrenal gland. Green leafy vegetables (such as spinach) which have also been found to help combat stress, wheat germ, all types of liver, fish, cheese, fruit, milk and yogurt are all good. A good breakfast is important. The evening meal should not be too heavy. Cod liver oil capsules and moderate amounts of vitamin C and vitamin E probably help. Women may take iron supplements during the monthly periods. Patients should try to obtain all the 40 or 50 nutrients needed by the body. They should try not to eat much "junk food" eg crisps and chocolates and not smoke. Since the adrenal cortex hormones prepare the body to meet all types of stress, they are very important. They increase the blood sugar for energy, they increase the blood pressure, fat is mobilized from the stores, and other changes occur. Nutritional needs are increased. Therefore to keep the adrenal cortex healthy one must eat sensibly and well, as must those who already have Addison's disease. They may also eat foods with B vitamins like yeast, marmite and whole meal bread, enough fiber and of course, the things mentioned previously.