



Caring for the Addison's patient:

Information for GPs

Addison's disease is a rare, potentially fatal, condition where the adrenal glands cease to function. Life-long treatment with replacement steroid hormones is required.

With the right balance of daily medication, people with Addison's can expect to have a normal life span and to lead full and productive lives. It is not unknown for people with Addison's to live into their 90s.

This leaflet outlines the role of the GP in managing Addison's disease at each stage in the patient lifecycle from diagnosis to old age.

1 DIAGNOSIS

The GP can play an important role in the early detection of Addison's when the non-specific nature of the symptoms can make it difficult to distinguish from depression. Postural hypotension, salt craving, hyperpigmentation, unexplained weight loss, nausea/abdominal pain are grounds for suspicion of Addison's in cases of persistent fatigue.

Diagnosis is confirmed in hospital by a physical examination and blood tests, principally:

- Electrolytes (low Na, high K)
- Synacthen (ACTH stimulation) test
- Plasma renin

Diagnosis can occur at any age, from 5 to 80. Almost half of all diagnoses occur outside the most common age of onset (40 - 60 years).

2 MEDICATION MANAGEMENT

In the UK, the normal adult requirement is:

■ Hydrocortisone 15mg - 25mg per day

This is a cortisol tablet and is usually taken in two or three divided doses. Dosage is dependent on bodyweight, metabolism and absorption; this is best confirmed by clinical assessment and in some cases by a hospital cortisol day curve.

■ Fludrocortisone 50mcg- 200mcg daily

This replaces aldosterone and is usually taken in a single morning dose. Dosage is dependent on metabolism and exercise levels and is monitored by measuring blood pressure sitting and standing (lack of postural drop) and plasma renin levels.

■ Possibly, DHEA 25mg - 50mg per day.

This is usually taken as a single morning dose. Although an unlicensed treatment, its use is supported by a number of clinical trials. An endocrinologist is normally best-placed to monitor DHEA replacement.

■ Hydrocortisone injection

Efcortesol 100mg (liquid) or Solu-Cortef 100mg (powder) plus vial of water, a syringe and needle, as an emergency kit for intra-muscular injection in case of vomiting, accident or other severe injury.

It is advisable to issue the patient with 3 - 5 vials injectable hydrocortisone in case of breakages.

See www.addisons.org.uk/emergency

The Addison's Clinical Advisory Panel (ACAP) recommends that all steroid-dependent patients be issued with six-monthly repeat prescriptions of their essential steroid medication to minimise the risk of running out, especially during periodic supply disruptions.

Addison's patients are entitled to receive their medication free of charge. The GP is required to complete a medical exemption form. See www.ppa.org.uk/ppa/medex.htm

ACAP recommends that the GP should check for potential drug interactions on each occasion that they issue a new prescription.

The major steroid interactions are:

- Diuretics, acetazolamide

- Avoid unless clearly indicated

- Drugs influencing electrolytes and blood pressure - NSAIDs, some anti-depressants, some antibiotics

- Can be used but fludrocortisone dose may need adjusting

- Drugs affecting metabolism of hydrocortisone - Anti-epilepsy/neuralgia (eg. phenytoin, carbamazepine), anti-tuberculosis (eg. rifampicin), anti-fungals (eg. ketoconazole)

- Can be used but glucocorticoid dose needs to be adjusted.

3 PATIENT EDUCATION FOR EMERGENCY PREVENTION

Self-medication requires every Addison's patient to be an 'expert patient'. To do this, they require coaching and support from their GP and broader medical team.

Patients must understand the potentially life-threatening consequences of inadequate glucocorticoid replacement, especially during intercurrent illness, surgery or severe injury.

The 'sick day rules' that patients must follow are:

- Double the normal dose of hydrocortisone for a fever of more than 37.5 C

- Take 20mg hydrocortisone immediately after vomiting and sip rehydration/electrolyte fluids (Dioralyte)

- On vomiting twice, use the emergency injection. Then call a doctor, saying Addison's emergency

- Take 20mg hydrocortisone orally immediately after injury to avoid shock ▶

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WHERE CAN I GET FURTHER INFORMATION?

Information and support are available from the Addison's Disease Self-Help Group (ADSHG), a registered charity run by and for people with Addison's. For a small annual subscription, the group offers: an information manual, emergency card, print newsletter, electronic discussion group, website, seminars and meetings.

Please contact: **ADSHG information,**
PO Box 45 445, London SE19 6YR.
Email: info@addisons.org.uk
Website: www.addisons.org.uk

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ACAP has also issued clinical guidelines for:

- **Emergency treatment of hypoadrenalism**
- **Glucocorticoid medication for surgery and dentistry**
- **Managing Addison's disease, an information leaflet for patient**

These are available from the ADSHG at www.addisons.org.uk/publications



The ADSHG works to support people with adrenal failure and to promote better understanding of this rare condition.

The Addison's Clinical Advisory Panel is a group of endocrinologists with an interest in adrenal medicine. It advises the Addison's Disease Self-Help Group on medical matters.

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■ **Ensure the surgical team, dentist or endoscopist are aware of the need for extra oral medication and that they have checked the ACAP surgical guidelines for the correct level of steroid cover.**

All patients requiring antibiotic treatment for infection/sepsis should be reminded of the need to double their normal glucocorticoid medication.

Patients must be educated to renew their repeat prescription in good time, so as to always retain a month's reserve supply.

When travelling away from home, patients must take an extra supply of medication (ie double what they normally need) plus their injection materials.

Airport security will require a doctor's note explaining why they are carrying essential medication, needles and syringes in their hand luggage. This is usually supplied by the GP.

Patients are recommended to purchase a MedicAlert bracelet. MedicAlert keep emergency treatment instructions on record for all Addison's patients. See www.medicalert.org.uk

4 EMERGENCY TREATMENT

■ The GP plays an important role in emergency treatment, through their role in home visits and arranging hospital admissions.

■ Around 6% of Addison's patients per annum require hospital treatment for an adrenal emergency. Vomiting and diarrhoea are the most frequent causes of adrenal emergency.

■ Early self-administration by the patient of 100mg injected hydrocortisone IM will often stabilise an acute episode of vomiting or diarrhoea without the need for further medical intervention.

■ However, some patients deteriorate rapidly and may require hospital treatment for 24 - 72 hours with intravenous hydrocortisone and saline. Postural dizziness is a key indicator that IV fluids are necessary.

■ Circulatory complications ranging from hypotension to hypovolaemic shock may occur during acute hypocortisolaemia. The acutely unwell patient should be stabilised by a saline infusion (for volume repletion) and a 100mg hydrocortisone injection before transportation to hospital.

5 MONITORING FOR 'ADDISON'S PLUS'

Most patients with autoimmune Addison's will already have or will develop in the future at least one associated endocrine or autoimmune condition. The GP must remain alert to the development of new symptoms and be ready to refer back to the endocrinologist or other specialists.

Most common are thyroid disorders but patients may also develop premature ovarian failure, testicular failure, diabetes, Vitamin B12 deficiency, vitiligo or coeliac disease. Asthma also occurs more frequently among autoimmune (primary) Addison's patients.

Osteopenia and type 2 diabetes may result from over-replacement of glucocorticoids. Patients with ovarian or testicular failure are at particular risk of osteoporosis.

6 CARE IN SPECIAL CIRCUMSTANCES

■ Patients who undertake strenuous physical exercise (eg. a marathon) will need extra medication, up to double the dose of glucocorticoid and mineralcorticoid, as well as sufficient fluids. For sports with a risk of injury, eg horseriding, skiing, the patient must ensure that a team-mate has been trained to administer an emergency injection.

■ Addison's patients who work nocturnal shifts will require their dose schedule to be altered according to the work pattern.

■ Essential hypertension is reported in around 10% of treated Addison's patients. This is best managed through ACE inhibitors or calcium blockers. Diuretics should be avoided. A reduction in fludrocortisone dose may be necessary, requiring monitoring of electrolytes, although it is inadvisable for fludrocortisone to be completely stopped.

■ Post-diagnosis pregnancies occur in around 20% of female Addison's patients. The pregnant patient will require hospital-based obstetric monitoring; steroid medication increases may be needed in the latter trimesters and hospital treatment may be required in cases of severe hyperemesis gravidarum. Steroid requirements during labour or caesarean section are addressed by ACAP's surgical guidelines, available at www.addisons.org.uk/publications

Further reading:

On-line endocrinology textbook: www.endotext.org/adrenal
Artl W, Allolio B. Adrenal insufficiency. **Lancet 2003;**
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