Addison's Disease Self-Help Group (ADSHG)
Registered charity 1179825

ADDISON'S Self Help Group

ADDISON'S Clinical Advisory Panel

Addison's Disease and Adrenal Insufficiency:

Notes for Primary Care and A&E Nurses

Managing and caring for a patient with Addison's disease or adrenal insufficiency.

As a Primary Care or A&E nurse, it is important to recognise the signs and symptoms of Addison's disease (AD) and, more broadly, adrenal insufficiency (AI). In particular it is important to recognise an adrenal crisis, the potentially life-threatening emergency faced by Addison's and adrenal insufficiency patients; which may lead to their presentation in the GP practice or A&E.

WHAT ARE ADDISON'S DISEASE AND ADRENAL INSUFFICIENCY?

Addison's disease, also known as primary adrenal insufficiency, is a rare (approx. 1:10,000), potentially fatal endocrine condition, resulting from the adrenal glands ceasing to function. The body is unable to make enough essential steroid hormones so lifelong treatment with replacement corticosteroids is required. With the right daily medication, including additional medication during times of physiological and psychological stress, people with Addison's and adrenal insufficiency can expect a normal lifespan.

Affected hormones include:

	Relevant hormones affected	Cause of adrenal insufficiency
Primary adrenal insufficiency	• Cortisol • Aldosterone • DHEA	Failure of the adrenal glands to produce glucocorticoids. Most commonly an autoimmune condition.
Secondary and tertiary adrenal insufficiency	• Cortisol • Adrenocorticotropin hormone (ACTH)	Low adrenal function from a lack of pituitary or hypothalamus hormone stimulation. Tertiary AI is a potential result of long-term glucocorticoid use.

Cortisol is a key hormone. It controls blood pressure, blood sugar, and muscle strength. Aldosterone manages sodium and fluid balance, while DHEA affects stamina and libido.

Comorbidities that result in patients requiring other time critical medications:

- About 50% of Addison's patients have a thyroid condition.
- 10% of Addison's patients have diabetes mellitus and adrenal failure.

TREATMENT OF ADDISON'S DISEASE AND ADRENAL INSUFFICIENCY

People with AD and AI are steroid-dependent, requiring continuous steroid cover. They can feel unwell within hours of a missed dose.

In the UK, doctors are likely to prescribe:

- Hydrocortisone: 15mg-25mg per day to replace cortisol. Usually taken in two to four divided doses. Hydrocortisone is quickly metabolised, with a single dose usually lasting up to 6 hours, or
- Prednisolone: a longer acting steroid. Usual total daily dose 3mg-5mg orally
- Fludrocortisone: 50mcg-200mcg / day for AD patients. Replaces aldosterone and is usually taken in a single morning dose
- Hydrocortisone sodium succinate or hydrocortisone sodium phosphate 100mg injection is needed in times of acute physiological or psychological stress (e.g. vomiting, severe injury or accident)

Most people take their replacement steroid medication three times a day, to mimic the production of cortisol within a body with fully functioning adrenal glands.

The first dose is taken on waking and then at five to six hourly intervals through the day.

If a patient can't take their normal oral medication (e.g. being nil by mouth or being unable to keep oral medication down due to vomiting), healthcare providers must prescribe and administer other forms of steroid cover. This should be given intramuscularly or intravenously every six hours, or by continuous infusion.

Other endocrine medications

- A few patients may also need other endocrine medications that will be critical to their treatment timeline, such as insulin for Type I Diabetes mellitus or desmopressin for Arginine Vasopressin Deficiency (AVP-D), formerly known as diabetes insipidus. Insulin and hydrocortisone are counter-regulatory drugs, so this combination is acutely time-critical.
- Certain replacement hormones, such as DHEA (25mg-50mg per day) are not time-critical and are not required by all people with adrenal insufficiency.

SICK DAY RULES

As both physical and extreme emotional stress affect the body's need for cortisol, there are a number of scenarios where a patient's prescribed replacement medication will be insufficient for their needs, putting them at risk of adrenal crisis. The 'Sick Day Rules' offer guidance on how and when to increase a patients usual steroid medication.

E.g. vomiting and diarrhoea, virus, infection, injury, some investigative procedures such as colonoscopy, and minor or major surgery, including dental procedures. See the ADSHG Surgical Guidelines.

The following guidance is relevant for anyone over 16 years of age. Separate detailed guidelines are available for babies, children and young people under 16 years: see BSPED consensus guidelines on adrenal insufficiency.

Sick Day Rule 1

Daily oral hydrocortisone should be increased to at least 40mg, divided into 2-4 doses or oral Prednisolone increased to 10mg daily as 1-2 doses if a patient is experiencing:

- a fever
- an illness requiring bed rest
- an illness requiring treatment with antibiotics
- vomiting or diarrhoea but are still able to tolerate food and drink. Sip rehydration/ electrolyte fluids alongside the increased dose
- a fracture or any similar significant injury. This should be continued until the acute illness or trauma has resolved
- intense psychological and emotional stress e.g. bereavement (increase dose for 1-2 days)

Ensure the surgical team, dentist or endoscopist are aware of the need for extra steroid medication for any surgical procedure, and that they have checked the ADSHG Surgical Guidelines for the correct level of steroid cover.

Sick Day Rule 2

Inject hydrocortisone 100mg intramuscularly for one or more of the following:

- They have vomited (within 30 mins of taking their steroid medication)
- They have persistent, severe diarrhoea
- They collapse, experience lightheadness or dizziness on standing, confusion, severe lethargy or are unable to get out of bed
- They are unable to take oral steroid medications e.g. before a surgical procedure or investigation (see Surgical Guidelines)
- Loss of consciousness

Scan for BSPED guidelines on adrenal insufficiency

Scan for Surgical Guidelines

PATIENT CARE IN PRIMARY CARE AND A&E

1. When should I suspect adrenal insufficiency?

You should suspect a person may have adrenal insufficiency if they present with one or more of the following;

- Unexplained hyperpigmentation (including in the mouth and scars present in undiagnosed AD)
- Weight loss
- Salt craving
- Nausea or vomiting
- Hyponatremia
- Hyperkalaemia
- Lethargy
- Muscle weakness

Other signs in children can include hypoglycaemia, problems growing, hypotensive crises, and prolonged neonatal jaundice.

Anyone can develop adrenal insufficiency, but you should be aware of the certain groups of people at higher risk.

A. Adults who have been on steroids for more than 4 weeks or children for more than 3 weeks.

B. People who are on any form of steroids and have an episode of physiological stress.

C. Certain medications and conditions can also affect cortisol levels.

2. What test should I do?

If you suspect the person has adrenal insufficiency, a serum cortisol level between 8-9am should be measured.

People who are taking steroid doses as above or higher do not need a cortisol level measuring. Medications can interfere with cortisol measurements and should be avoided at certain times (NICE guidelines p12, www.nice.org.uk/guidance/NG243.)

Please refer to interpretation of serum cortisol levels for advice (NICE table p13).

3. What should I do if the person is acutely unwell?

If the person is acutely unwell with any of the below, adrenal crisis should be suspected and emergency treatment given. Consider adrenal crisis in people with, or at high risk of, AI who are unwell with milder symptoms.

Seek **urgent** clinical advice and support from medical colleagues and the specialist endocrine team.

Acute Symptoms of Adrenal Crisis	Milder symptoms in people with or at high risk of AI
Low blood pressure, abdominal pain, vomiting and muscle spasms	Weakness and lethargy
Hyperpigmentation in patients with undiagnosed AD	Shakiness / shivering
Hyponatremia	Pallor
Hypoglycaemia (particularly in children)	Feeling cold or feverish
Circulatory shock or collapse	Clamminess
A condition failing to respond to initial treatment	Confusion

ADRENAL CRISIS

An adrenal crisis (also called Addisonian crisis or acute adrenal insufficiency) is a life-threatening emergency that can occur when cortisol levels are insufficient to support life essential functioning.

It requires time critical treatment with parenteral steroids and intravenous (IV) fluids.

ADRENAL CRISIS TREATMENT

Emergency treatment includes 200mg HC IV over 24hrs or 50mg Hydrocortisone IM 4 times a day and in both cases, 1 litre of 0.9% sodium chloride intravenous infusion over 30 minutes.

There is no risk of overdose from hydrocortisone in an emergency.

A. It is important that the following are monitored frequently during an adrenal crisis;

- Blood pressure
- Electrolytes
- Heart rate
- Glucose

B. Continue with the hydrocortisone regimen as above until stable.

C. Sick Day Rules (increasing of steroid replacement to at least 40mg daily hydrocortisone) should be followed when the patient is able to absorb oral hydrocortisone. Further fluids may be required depending on the patient's condition.
Admit to hospital if unable to absorb steroids (e.g. vomiting) and until stable.

D. The underlying cause of adrenal crisis should be treated e.g. infection.

4. What is the medication regime?

The Addison's patient is steroid-dependent and it is essential that they do not miss, or receive a delayed dose.

Adherence to their steroid medication regimen is vital to aid their recovery. Do not withdraw steroids without the advice of the endocrine medical team.

The first steroid dose of the day should be taken with water on waking (empty stomach), to maximise absorption. Patients with digestive issues may need a small snack, milk, or a lactose-free substitute. Patients will know what their own symptoms of steroid undermedication are - they are a valuable source of information.

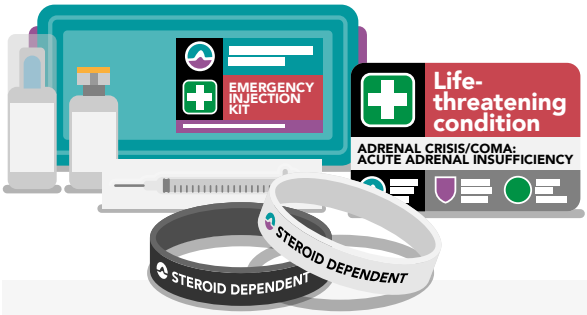
- Many AD and AI patients will have been encouraged to be an "expert patient" and will be competent to self-medicate for their replacement endocrine drugs while in hospital. However, depending on their state of health or mental capacity, self-medication with their regular replacement endocrine drugs may not always be appropriate. Where possible, adopt supervised self-medication for the psychiatric inpatient to help maintain a sense of stability.
- Diet and Hydration: Patients with AD are at risk of dehydration and should drink 1.5 litres of water per day. They can lose salt faster than other people.

NEWLY DIAGNOSED PATIENTS

Ensure that the patient:

1. Has advice on Addison's disease and adrenal insufficiency, the importance of steroid replacements and why they have been prescribed steroids (and fludrocortisone if indicated).
2. Understands the symptoms to look out for, of under or over replacement of medication.
3. Knows how and when to access clinical advice or emergency services.
4. Has the NHS Steroid emergency card.
5. Knows how to set up medic alerts, medical IDs, and apps on mobile phones.
6. Is signposted to relevant support groups and charities (ADSHG). www.addisonsdisease.org.uk
7. Knows how to access free NHS prescriptions.
8. Is provided with information on how to discuss their diagnosis and treatment with employers, educational settings, and with friends and family.
9. Has an emergency hydrocortisone injection kit for intra-muscular injection in case of vomiting, accident or other severe injury.
10. Understands that if they need to give an emergency injection, they then should attend the emergency department.
11. Is advised to always maintain a good supply of oral medicines, and enough for sick day dosing and travelling.
12. Understands how to adjust the timing of medicine dosing when travelling, fasting or doing shift work.
13. Understands the importance of not stopping medicines except on medical advice.
14. Is aware what they should do with their medication when unwell ('Sick Day Rules').

Scan for NICE guidelines



Emergency injection kit, emergency steroid card and medic alert wristbands

PRE-DISCHARGE CHECKS

Sick Day Rules	Is the patient aware of how to apply the Rules?
NHS steroid emergency card	Do they have one? Provide if not.
Surgical Guidelines	Reminder for the patient to inform any healthcare professionals including dentists, of their need for extra steroids for procedures.
2 or 3 Emergency Injection Kits (primary & secondary AI)	• Hydrocortisone Sodium Phosphate 100mg (liquid) or Hydrocortisone Sodium Succinate 100mg (powder), • a vial of water (if prescribed hydrocortisone sodium succinate), • a syringe and blue needle • written instructions on preparation.
Emergency Kit checks	Are they aware they need to check expiry dates on meds and equipment and replace as necessary.
Injection Training	Do they know how to prepare and administer the injection?
Monitoring	Ensure follow up with specialist team.

OTHER CONSIDERATIONS

- Ensure the patient is aware of telling their GP they have Addison's disease/adrenal insufficiency as soon as possible.
- Pregnancy: care should be done by a team experienced in managing adrenal insufficiency.
- Patients should be offered ongoing reviews as per clinical and individual needs, with an appropriate specialist team for people with Addison's and adrenal insufficiency.
- It is the patient's right to be involved in making choices about their care. This includes knowing what their treatment options are, risks and benefits.

For further advice please contact:
Email: enquiries@addisons.org.uk
Website: addisonsdisease.org.uk

The Addison's Disease Self-Help Group works to support people with Addison's and adrenal insufficiency and to promote better medical understanding of this rare condition.

The ADSHG is supported and advised by a Clinical Advisory Panel: a group of medical professionals with an interest in adrenal medicine. Registered charity 1179825, established 1984.

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This leaflet has been authored by Lisa Shepherd, RN, DipHE, BSc, MSc and the ADSHG and reviewed by the ADSHG Clinical Advisory Panel.

Last Reviewed: **February 2025**