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

ADDISON'S Self Help Group

ADDISON'S Clinical Advisory Panel

Diagnosis and Care of a Patient with Adrenal Insufficiency, including Addison's Disease

Guide for GPs and ACPs

Healthcare Professionals

A guide to inform and support GPs and Advanced Care Practitioners

Contents

Introduction

If you are working with a patient with adrenal insufficiency, including Addison's disease, you are in a position of critical influence in terms of helping them to optimally manage their condition, to avoid life-threatening adrenal crises, build confidence and improve their quality of life.

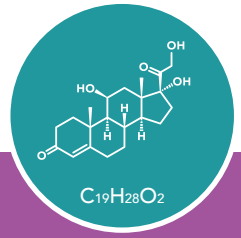
The GP plays an important role in the early detection of Addison's and adrenal insufficiency and the life-saving management of adrenal crisis, which can be rapidly fatal. Without prompt recognition and treatment, there is a high risk of death from circulatory collapse, "shock" or respiratory arrest, or risk of irreversible complications such as cardiac arrhythmias, cardiac arrest, hypoglycaemia and hypoglycaemic coma.

1 Adrenal Insufficiency

WHAT IS ADRENAL INSUFFICIENCY?

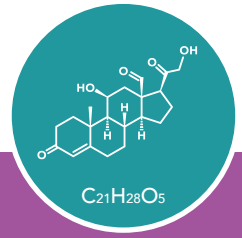
Adrenal insufficiency affects either some or all adrenal cortex function, with, critically, a reduction in the production of some glucocorticoids, such as cortisol, which regulates the stress response, metabolism and immune response.

The pattern of hormonal insufficiency varies depending on the type of adrenal insufficiency: primary (also known as Addison's disease), secondary or tertiary, which in turn relates to the cause of the disruption to production.



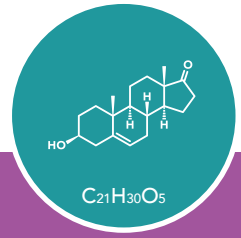
Cortisol

Metabolic control, blood sugar, energy levels, appetite and stress.



Aldosterone

Blood pressure and salt/fluid balance.



DHEA

Stamina and libido.

Significance

Patients with undiagnosed or established primary or secondary adrenal insufficiency as well as those receiving chronic exogenous supraphysiological glucocorticoid treatment (e.g. for asthma or autoimmune disease) and topical or injected steroids, who are at risk of developing tertiary adrenal insufficiency, are all at risk of adrenal crisis. This is a life-threatening emergency resulting from a lack of cortisol, the major glucocorticoid.[1] It can affect patients with any type of adrenal insufficiency.

Adrenal insufficiency-related hospital admissions and adrenal crises (with their associated morbidity and mortality) have increased considerably in the last two decades,[2] with one in 200 adrenal crises being fatal.[1,3] The standardised mortality rate in patients with adrenal insufficiency is twofold compared to the general population.[3,4]

About one in ten patients with adrenal insufficiency are hospitalised at least once a year following an adrenal crisis episode,[5,6] with vomiting and diarrhoea being the most frequent causes.[8]

TYPES OF ADRENAL INSUFFICIENCY

Primary adrenal insufficiency (Addison's disease)

The adrenal cortex itself is damaged, reducing production of cortisol, mineralocorticoids such as aldosterone and adrenal androgens, e.g. dehydroepiandrosterone (DHEA).

A rare endocrine condition, affecting about 1:14,000 with an upper estimate of around 9,000 diagnosed cases in the UK, where around 320 new cases will be diagnosed each year for the UK.[7,8]

Whilst the most common age of onset is 30–50 years, almost half of all diagnoses occur outside of this range and can be at any age from birth to 80 years.

This category also includes patients who have had an adrenalectomy.

Secondary adrenal insufficiency

The production of adrenocorticotrophic hormone (ACTH) in the pituitary gland is reduced, causing reduced production of cortisol.

This occurs if the regulation of adrenal cortisol production by the pituitary or hypothalamus is compromised, as a consequence of a hypothalamic–pituitary condition, such as:

- Pituitary tumours or hypophysectomy
- Traumatic brain injury
- Pituitary Infarction (e.g. Sheehan syndrome)
- Tumours of the hypothalamus
- Hypothalamic surgery
- Subarachnoid haemorrhage
- As a result of immunotherapy treatment

Tertiary adrenal insufficiency

The hypothalamus and pituitary production of CRH and ACTH are suppressed by exogenous medication/ drug use.

Causes include:

- Long-term oral corticosteroid use
- Long-term inhaled corticosteroid use
- Long-term topical corticosteroid use (particularly on mucosal membranes)
- Depot long-acting corticosteroid use (i.e. intramuscular/intra-articular)
- Long-term, high-dose opioid use

Patients on long-term steroids (especially oral steroids taken at high doses for some time), patients using high doses of inhaled steroids, and patients receiving frequent intra-articular steroids are at particularly high risk of developing adrenal insufficiency.

Adrenal insufficiency and primary care

The GP role includes:

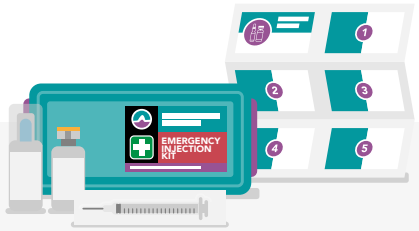
- Early recognition of symptoms suggestive of adrenal insufficiency e.g. unintentional weight loss, postural hypotension, fatigue, hyponatraemia, unexplained increased skin pigmentation (Addison's only).
- Support with effective management to reduce the incidence of adrenal crisis.
- Digital alerts on GP computer system to support remote monitoring for prompt prescriptions (time-critical medication) and priority appointments.
- Sufficient glucocorticoid medication to allow patient to follow Sick Day Rules (regular 3 month prescription plus one-off prescription to provide supplementary supply).
- Provision of emergency injection kits as defined by the NICE Guidelines (see Treatment section for details).
- Provision of NHS Steroid Emergency Card to highlight the time-critical, steroid-dependent nature of a patient's medication regimen.
- Ensuring accurate Summary Care Records to document a patient's steroid dependency (accessed by emergency call handler teams during an emergency ambulance call out).
- Home visits to unwell adrenal insufficiency patients with potential requirements for emergency management of adrenal crisis.
- Supporting patients and carers in an adrenal crisis to self-administer emergency treatment and to access critical care services.
- Arranging hospital admissions from patients presenting at the surgery in an adrenal crisis.
- Provide timely seasonal vaccinations and updated COVID vaccination to eligible patients with adrenal insufficiency.

2 Treatment

EMERGENCY MANAGEMENT OF ADRENAL CRISIS

Adrenal crisis (also known as acute adrenal insufficiency or Addisonian Crisis) is a life-threatening emergency that **requires the time-critical emergency injection of hydrocortisone 100mg IM followed up by a hospital visit for intravenous fluids** and potentially further treatment with hydrocortisone until the patient has stabilised.

In line with the NICE Guidelines for the Identification and Management of Adrenal Insufficiency (NG243), all patients with primary and secondary adrenal insufficiency, and those with a diagnosis of tertiary adrenal insufficiency who have a history of, or are deemed to be at risk of adrenal crisis, should be provided with a minimum of 2 emergency management kits by their endocrinology or primary care team.



ADSHG emergency injection kit

This kit consists of:

- Hydrocortisone sodium phosphate 100mg 1ml vial of liquid OR Hydrocortisone sodium succinate 100mg (powder) plus 2ml vial of water
- 2x Intramuscular (IM, blue) needles and 2x 2ml syringes
- 1 x instruction leaflet in an easy to understand format (available from the ADSHG online shop)
- 1 x NHS Steroid Emergency Card

3-5 vials of injectable hydrocortisone are recommended in case of breakages and for a second emergency injection kit (for administration in case of vomiting, illness, accident or other severe injury).

NB Hydrocortisone acetate (Hydrocortistab) is slow acting so should not be used.

The form of hydrocortisone prescribed for use in an emergency injection may be influenced by national availability/ drug shortages, regional tendencies to favour one type or another, or other factors relating to the endocrinologist prescribing, or the patient requirements and preferences.

Please prescribe Hydrocortisone sodium phosphate 100mg 1ml vial of liquid (PIP code: 1200286) when and where it is available as it removes the need to complete the additional step of mixing powder and liquid before injecting which is challenging whilst experiencing symptoms of adrenal crisis.

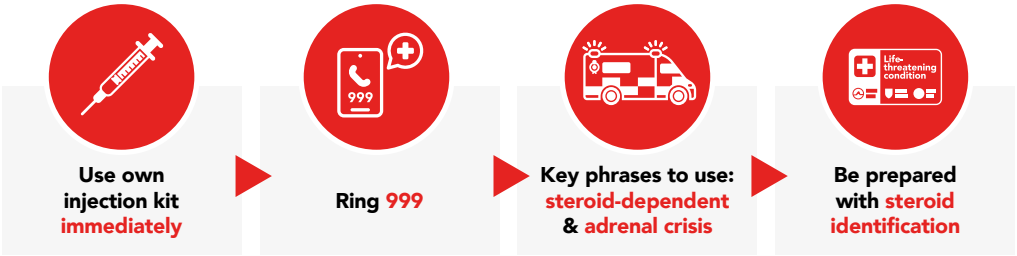
Replacing emergency management kit contents

After use in an emergency, the kit needs replenishing with hydrocortisone and replacement needles and syringes. These are currently not available on drug tariff. Please do what you can to support your patient in getting hold of replacement syringes and needles. Patients may only see their endocrine team once every 12 months and may not have access to an endocrine nurse. We are heartened, as a charity, when we hear of GPs being supportive in providing these essential items to their patients, whilst in the background we build the case for having them added to drug tariff.

URGENT ADMISSIONS TO HOSPITAL

The GP plays an important role in emergency treatment, through home visits and arranging hospital admissions. Untreated adrenal insufficiency, including Addison's disease is universally fatal, and the deterioration may be fast.

Emergency Management of Adrenal Crisis



Common causes of adrenal crisis in people with adrenal insufficiency are gastrointestinal illness (23%), other infections (25%), surgery (including dental procedures 10%) and physiological stress (9%). **An adrenal crisis is a medical emergency and can be fatal.** [9]

Signs and symptoms of adrenal crisis can include:

- nausea or vomiting
- abdominal pain
- low grade fever
- muscle pain/shaking
- headache
- hypoglycaemia (especially in children)

Later signs and symptoms include:

- reduced consciousness
- confusion
- hypotension/tachycardia/bradycardia
- hypovolemic shock
- cold peripheries

In line with Joint Royal Colleges Ambulance Liaison Committee (JRCALC) guidelines, paramedics attending to the patient must administer an IM 100mg hydrocortisone injection and ensure they are stabilised by a saline infusion (for volume repletion) prior to transfer.

In adrenal crisis, hypovolaemic shock, cardiac arrest, stroke or other circulatory complications can occur even in young, fit patients, and complications from hypoxia may leave the patient permanently disabled.

Children with adrenal crisis are particularly susceptible to hypoglycaemia which can cause permanent brain damage if not quickly reversed.

Further information about adrenal crisis:

Society for Endocrinology (SfE)

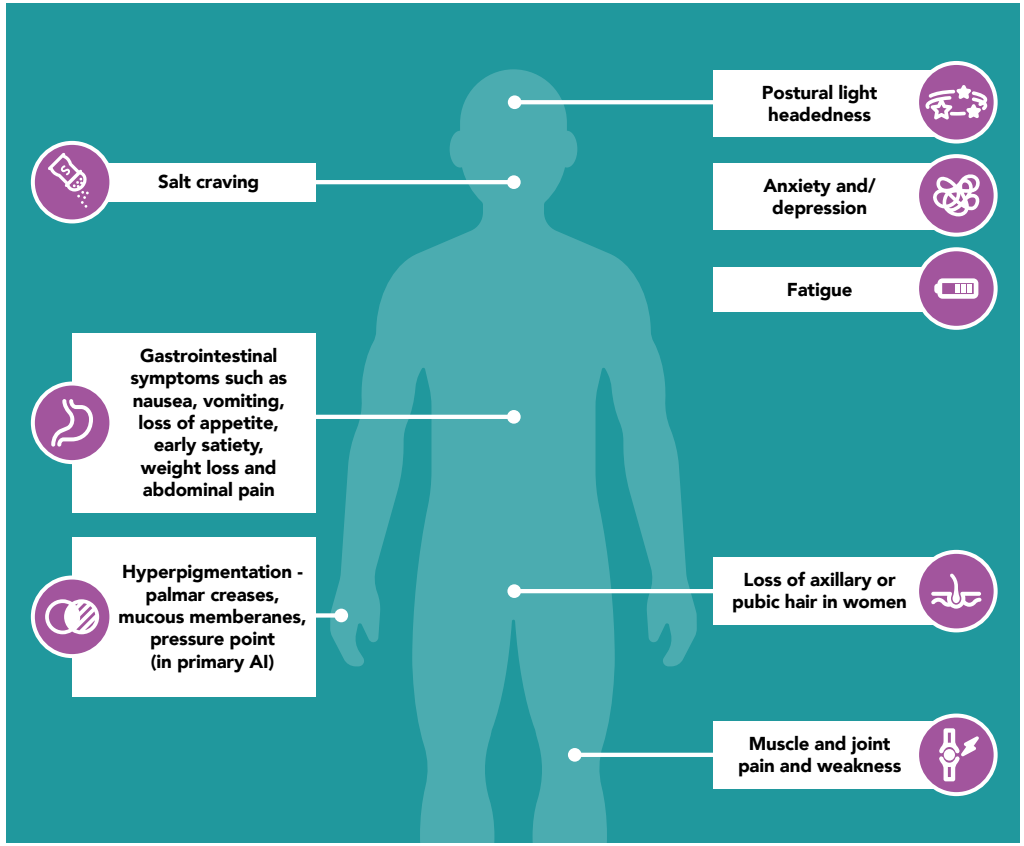
British Society for Paediatric Endocrinology & Diabetes (BSPED)

DIAGNOSIS OF ADRENAL INSUFFICIENCY

At times the non-specific nature of the symptoms can make it difficult to distinguish adrenal insufficiency from other conditions (anorexia nervosa, depression, chronic fatigue).

Consider primary adrenal insufficiency (Addison's disease) in people with unexplained hyperpigmentation. Be aware that hyperpigmentation may not be seen on black or brown skin. Ask the person if they have noticed a change in their skin colour and assess the buccal mucosa, pressure points (knuckles, elbows) or any surgical scars, or when there is no other clinical explanation for the presence of 1 or more of the following persistent symptoms, signs or features:[10]

Symptoms at Diagnosis (Adrenal Insufficiency)



- weight loss
- salt craving
- nausea or vomiting
- lack of appetite or unable to eat a full meal
- diarrhoea
- dizziness or light-headedness on standing
- hyponatraemia
- hyperkalaemia
- lethargy
- feeling of muscle weakness
- hypoglycaemia (particularly in children)
- faltering growth (in children)
- hypotensive crisis (particularly in children)
- prolonged neonatal jaundice

Key questions to ask;

- Has this person had prolonged steroids in the last 6 months?
- Does this patient have postural hypotension?
- Are they losing weight unintentionally?
- Do they have low/borderline blood sodium?
- Do they have salt, soy sauce or liquorice cravings, or increased thirst and urination?
- Do they have appropriate pigmentation or has there been a change in skin colour?
- Is there a history of endocrine or autoimmune conditions such as thyroid disorders, Type 1 diabetes, B12 deficiency or coeliac disease in the extended family?

Primary Adrenal Insufficiency

Possible causes	Signs and symptoms
• Autoimmune (90%) • Infections (e.g. TB – a leading cause in developing countries, HIV, fungal infection or histoplasmosis) • Bilateral adrenalectomy • Bilateral adrenal metastases • Bilateral adrenal haemorrhage • Congenital adrenal hyperplasia – infancy	• Glucocorticoid deficiency, notably anorexia, weight loss and muscle weakness • Mineralocorticoid deficiency (postural light-headedness, increasing thirst and urination, salt cravings) • Excessive fatigue and sleepiness • Mucocutaneous hyperpigmentation due to elevated pituitary ACTH • Unexplained weight loss • Family history may identify endocrine or autoimmune conditions linked with AD among extended family (e.g. thyroid disorders, diabetes, B12 deficiency, vitiligo and coeliac disease) • Hyponatraemia • Hyperkalaemia • Normal or low blood glucose

Hyponatraemia is present in 90% of patients with undiagnosed/newly diagnosed Addison's disease, and hyperkalaemia in 50%.[9] If these features are present, the clinical suspicion becomes high, and you should discuss your patient with endocrinology secondary care or arrange synacthen testing the same day.

ACTH: adrenocorticotropic hormone; AD: Addison's disease; TB: tuberculosis.

Secondary Adrenal Insufficiency

Possible causes	Signs and symptoms
• Pituitary tumours or hypophysectomy • Subarachnoid haemorrhage • Infarction (e.g. Sheehan's syndrome) • Traumatic brain injury	• Glucocorticoid deficiency, notably anorexia, weight loss and muscle weakness • Excessive fatigue and sleepiness • Skin pigmentation usually remains unchanged or displays an alabaster-like pallor • Unexplained weight loss • Hyponatraemia • Hyperkalaemia • Hypoglycaemia may be present in children

Tertiary Adrenal Insufficiency

Possible causes	Signs and symptoms
• Long-term corticosteroid use • Long-term high-dose opioid use	• Glucocorticoid deficiency, notably anorexia, weight loss and muscle weakness • Manifestation of symptoms when cortisol demand increases, or when a long-term steroid treatment is stopped abruptly • Excessive fatigue and sleepiness • Hyponatraemia • Hyperkalaemia • Unexplained weight loss

Table 1: Causes, Signs & Symptoms of the three types of adrenal insufficiency

Associated Conditions

Addison's disease is often associated with conditions such as:

- Hypothyroidism (range 43-44%)
- Asthma (13-15%)
- Vitiligo (9-10%)
- Vitamin B12 deficiency
- Type 1 diabetes (5-6%)
- Premature ovarian insufficiency
- Coeliac (3-6%)
- Hyperthyroidism (4-5%)

Considerations

1. Addison's disease may be the first autoimmune condition to manifest.
2. Pre-existing associated conditions are more likely in women. In established diabetes, a marked reduction in the insulin requirement or increasing hypoglycaemia can be a warning sign of developing hypoadrenalism.
3. Commencing thyroid replacement in early-stage adrenal failure may precipitate hypo adrenal symptoms/crisis, as thyroid hormones increase the metabolic rate of hydrocortisone. Elevated TSH in isolation may be an indicator of hypoadrenalism in an ill patient with extreme fatigue but without the typical features of hypothyroidism and may return to normal with steroid replacement.
4. Osteopenia and type 2 diabetes may result from over-replacement of glucocorticoids.
5. Patients with ovarian or testicular failure are at particular risk of osteoporosis.
6. Think about the possibility of adrenal insufficiency in babies and children with differences in sex development, such as ambiguous genitalia or bilateral undescended testes.

SCREENING

Where the patient is stable, the GP should assess for clinical signs, especially:

- Blood pressure sitting and standing. A 20mmHg drop on standing indicates clear postural hypotension.
- Proximal myopathy or global muscle weakness (difficulty climbing stairs, getting up from a sitting/squatting position, or carrying weights such as grocery shopping).
- Patchy hyperpigmentation or, in Caucasians, unusual pigmentation in the palmar/wrist creases (not present in 20% of cases, depending on the patient's natural level of melanocytes). Hyperpigmentation is typically most obvious in areas of increased friction such as oral mucosa, knuckles, elbows or waistline.

NB. If adrenal crisis is suspected, NEVER delay emergency treatment to perform investigations.

Primary Care Investigations

Initial laboratory investigations that can be conducted in primary care are:

1. Electrolytes (low Na, high K)
   Electrolytes may be borderline/normal where the patient is not in crisis
2. 9am cortisol

9am Serum Cortisol level	People aged 16 years and over	Children and young people between 1 year and over, and under 16 years
Below 150 nmol/L	• Recognise that the person may have adrenal insufficiency. • Refer the person to endocrinology. • Consider starting management for adrenal insufficiency (see the section on routine pharmacological management). • If the person is acutely unwell, follow recommendations for people aged 16 and over in the section on emergency management of adrenal crisis.	• Recognise that the person may have adrenal insufficiency. • Refer the person urgently to paediatrics or paediatric endocrinology. • If the person is acutely unwell, follow recommendations for babies, children, and young people under 16 years in the section on emergency management of adrenal crisis.
150 nmol/L to 300 nmol/L	• Recognise that the probability of adrenal insufficiency is uncertain. • Consider repeating the serum cortisol test. • If it remains at this level, seek endocrinology advice or referral.	• Recognise that the probability of adrenal insufficiency is uncertain. • Consider repeating the serum cortisol test. • If it remains at this level, seek paediatric or paediatric endocrinology advice or referral.
Above 300 nmol/L	Recognise that adrenal insufficiency is very unlikely.	Recognise that adrenal insufficiency is very unlikely.

Table 2: Interpretation of serum cortisol levels from an 8am to 9am test [10]

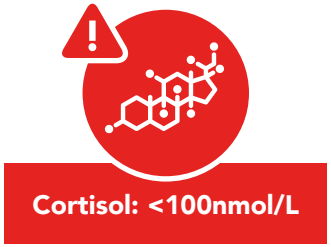
Potential Pitfalls

1. Random (untimed) serum cortisol has a low sensitivity for adrenal insufficiency, especially where the patient is in the early stages of adrenal disease. If you suspect the diagnosis on clinical grounds, then it is worth referring for a Synacthen test, unless a 9am cortisol is over 400nmol/L.
2. The slowly progressive nature of adrenal failure in Addison's disease means many patients are wrongly identified as depressive or anorexic until intercurrent infection precipitates an adrenal crisis. Some anti-depressants are sodium depleting and may precipitate adrenal crisis in patients with undiagnosed adrenal insufficiency.
3. Symptoms of hypocortisolemia or adrenal crisis, in which nausea and vomiting are predominant features, are frequently attributable to gastroenteritis or food poisoning. One third of Addison's patients report receiving hospital treatment for adrenal crisis on one or more occasions prior to their diagnosis without their condition being identified.
4. In pregnancy, Addison's symptoms may be mistaken for hyperemesis and chloasma. Pregnancy, oral contraceptive or HRT usage make interpretation of serum cortisol difficult due to elevation of background CBG and delayed hepatic clearance (i.e. false negative test).
5. Exogenous steroid use for conditions such as mouth ulcers has been documented to mask underlying adrenal insufficiency.

REFERRAL TO A SECONDARY SPECIALIST

Where there is a real suspicion of adrenal insufficiency refer to a specialist endocrine unit for a Synacthen (ACTH stimulation) test.

If the patient is hypotensive and vomiting, or where the 9am serum cortisol is less than 100nmol/L, an immediate and urgent referral is required.



PATIENT SUPPORT DURING THE DIAGNOSIS PHASE

Due to the often lengthy periods waiting for diagnosis, and the reality that many patients experience a delay, sometimes lengthy, in seeing an endocrinologist for the first time, after referral. It is important that they are supported during this time, whilst waiting for a confirmed diagnosis.

The ADSHG, established over 40 years ago, is a fantastic resource to signpost your patient to at this time.

THE ADDISON'S DISEASE SELF-HELP GROUP

The Addison's Disease Self-Help Group (ADSHG) is a UK based charity working to;

Support

Providing resources and building a community that helps people self-manage their Addison's disease and adrenal insufficiency and improve their quality of life.

Connect

Making it possible for the voice of people with Addison's disease and adrenal insufficiency to influence, inform and improve the way the healthcare service delivers their care.

Advance

Funding, contributing to and promoting the development of new innovations and research to improve quality of life.

Healthcare Professionals Newsletter

Sign up to the ADSHG newsletter

We publish a twice yearly newsletter, the ADSHG Connection, to keep healthcare professionals up to date with adrenal insufficiency news and best practice guidance.

Sign up using the QR code to stay in touch.

Useful patient information:

Scan for Newly Diagnosed

Scan for ADSHG Sanctuary of Support

TREATMENT

MEDICATION MANAGEMENT

In line with the NICE Guidelines for the Identification and Management of Adrenal Insufficiency [10], the recommended corticosteroid replacement for adrenal insufficiency in patients of 16 and over is as follows (to be prescribed by endocrinology team):

Treatment	Primary adrenal insufficiency	Congenital adrenal hyperplasia (CAH)	Secondary and tertiary adrenal insufficiency
First-choice glucocorticoid	**Hydrocortisone** total daily dose 15 mg to 25 mg orally in 2 to 4 divided doses.	**Hydrocortisone** total daily dose 15 mg to 25 mg orally in 2 to 4 divided doses. Consider higher doses with specialist advice if needed for control of CAH.	**Hydrocortisone** total daily dose 15 mg to 25 mg orally in 2 to 3 divided doses.
Alternative glucocorticoid (for example, if multiple daily doses are not appropriate)	**Prednisolone** (if they have stopped growing) total daily dose 3 mg to 5 mg orally.	**Prednisolone** (if they have stopped growing) total daily dose 3 mg to 5 mg orally. Consider higher doses with specialist advice if needed for control of CAH.	**Prednisolone** (if they have stopped growing) total daily dose 3 mg to 5 mg orally.
Alternative glucocorticoid (for example, if multiple daily doses are not appropriate)	**Modified-release hydrocortisone tablets** (if they have stopped growing) orally. In August 2024, modified-release hydrocortisone tablets were off-label for under 18s. See NICE's information on prescribing medicines.	**Modified-release hydrocortisone capsules** (if they have stopped growing) orally. Or **Dexamethasone** (under specialist advice only) total daily dose 300 micrograms to 500 micrograms orally.	**Modified-release hydrocortisone tablets** (if they have stopped growing) orally. In August 2024, modified-release hydrocortisone tablets were off-label for under 18s. See NICE's information on prescribing medicines.
Mineralocorticoid if needed (to normalise serum electrolytes and plasma renin, and reduce postural symptoms and salt craving)	**Fludrocortisone** total daily dose initially 100 micrograms and adjusted according to response up to 300 micrograms orally. Consider a higher daily dose orally for young and physically active people. In August 2024, doses of fludrocortisone above 300 micrograms daily were off-label. See NICE's information on prescribing medicines.	**Fludrocortisone** total daily dose initially 100 micrograms and adjusted according to response up to 300 micrograms orally. Consider a higher daily dose orally for young and physically active people. In August 2024, doses of fludrocortisone above 300 micrograms daily were off-label. See NICE's information on prescribing medicines.	Do not offer a mineralocorticoid.

Table 3: Corticosteroid Replacement for adrenal insufficiency in people aged 16 years and over [10]

Check for potential drug interactions on each occasion that you issue a new prescription. Major known interactions are:

1. Diuretics, acetazolamide, NSAIDs. Avoid unless clearly indicated.
2. Drugs influencing electrolytes and blood pressure, e.g. some anti-depressants, some antibiotics, carbenoloxone, drospirenone-containing contraceptive can be used but fludrocortisone dose may need adjusting.
3. Drugs affecting metabolism of hydrocortisone e.g. anti-epilepsy/neuralgia (phenytoin, carbamazepine), anti-tuberculosis (rifampicin), anti-fungals (ketoconazole, itraconazole), barbiturates and etomidate. Can be used but glucocorticoid dose needs to be increased.
4. Patients should be advised that grapefruit and liquorice delay the hepatic clearance of glucocorticoids and are best avoided or consumed sparingly.
5. Ritonavir, a drug used to treat HIV, also prevents clearance of hydrocortisone and other steroids, so expert advice is needed for patients combining this with any steroid.

Prescription Frequency

Patients should be reminded to renew their prescription in good time.

The NHS, Society for Endocrinology and ADSHG Clinical Advisory Panel recommends that all steroid-dependent patients be issued with a minimum of a 3 monthly repeat prescription for their essential steroid medication. It is also advised to offer a one-off 3 month prescription to provide supplementary medication to allow them to follow Sick Day Rules where they will need to increase their glucocorticoid dosage, and to minimise the risk of running out, especially during periodic supply disruptions.

3 monthly repeat prescription recommended

PREVENTING ADRENAL CRISIS THROUGH PATIENT EDUCATION

The responsibility that comes with self-medication and the significance of ensuring sufficient glucocorticoid medication to decrease the chances of an adrenal crisis, means thorough patient education and training is required, to support the patient in becoming confident in managing their condition.

The GP is ideally placed to assist in coaching and supporting the patient, and signposting to other support services as required.

Sick Day Rules

All patients need to know how to adjust their replacement steroid medication for periods of physical stress (illness and injury), short-term periods of psychological stress or strenuous exercise, when the bodies need for cortisol will outstrip the daily prescribed replacement glucocorticoid medication they are taking.

This extra medication is known as stress dosing or updosing. Clinicians have developed guidelines to support patients in managing their medication regime during these times, known as the **Sick Day Rules.**

Additional supplies of oral glucocorticoids will be needed to cover increased dosing. Patients on modified hydrocortisone will also need supplies of immediate-release hydrocortisone to follow Sick Day Rules.

When patients need to follow the rules:

- During episodes of vomiting and/or diarrhoea (but they are still able to tolerate food and drink).
- For an illness needing rest or time in bed due to weakness, or when on a course of antibiotics.
- For a fracture or other major injury; take a minimum of 40mg hydrocortisone to avoid shock.
- For a fever of over 37.5°C but less than 39°C.
- They are also likely to need to updose to some degree with common ailments such as colds, coughs and headaches, even if they are not ill enough to be off work, or in bed.
- During episodes of extreme psychological or emotional stress e.g. exams/tests, bereavement etc.

Scan for NICE Guidelines

Scan for Sick Day Rules

Scan for Adrenal Crisis Guidelines

The Sick Day Rules

40mg in 2-4 doses

Hydrocortisone users
Increase the daily oral hydrocortisone to at least 40mg, divided into 2-4 doses. **If their usual dose is more than 20mg, then double it, rather than increasing it to 40mg.**

10mg in 1-2 doses

Prednisolone users
Increase the daily oral prednisolone or prednisone to 10mg daily as 1-2 doses. If they are feeling severely unwell (e.g. with Covid or flu) and **still feel unwell on an increased dose of 10mg, increase it further to 15mg in 24 hours.**

40mg in 4 doses

Plenadren and Efmody (previously Chronocort) users
Switch to the regular, immediate release hydrocortisone preparation and take 10mg orally every 6 hours.

Additional considerations

- **There is no toxic dose of hydrocortisone for short term use**, however there are side effects of long-term over replacement (see later section).
- If the person vomits within 30 minutes of taking an oral dose, advise them to take a further dose once vomiting subsides, at double the original dose. If vomiting recurs within 30 minutes, 100mg hydrocortisone should be administered by injection and the person should attend the emergency department.
- In the case of major injury (e.g. a fracture), it may be more appropriate to administer 100mg IM injection of hydrocortisone rather than taking 40mg orally. It will depend on the individual and the circumstances as to which action is appropriate.
- Admit the person to hospital during periods of physiological stress if they are unable to absorb oral glucocorticoids, for example, during prolonged diarrhoea and vomiting. Facilitate administration of 100mg hydrocortisone IM.

Surgical Guidelines

For people having planned or emergency surgery or invasive medical procedures, remind them to ensure that they have informed their healthcare team of their diagnosis and provided them with the Surgical Guidelines. This includes procedures such as dental work and endoscopy or colonoscopy.

Scan for Surgical Guidelines

Other considerations

Sport and Exercise: Patients undertaking extended strenuous physical exercise and competitive sports, will require up to double the dose of glucocorticoid and mineralocorticoid as well as sufficient fluids.

For sports or outdoor activities with a risk of injury (skiing, cycling etc.) the patient must ensure that a team-mate has been trained to administer an emergency injection and knows where the patient keeps their emergency injection kit.

Night Shifts: Patients working night shifts will require their dose schedule to be altered according to their work pattern.

Pregnancy: Post diagnosis pregnancies occur in around 20% of female patients with Addison's. 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 caesarian section are addressed by the ADSHG Clincal Advisory Panel's Surgical Guidelines.

Travel: When travelling away from home, patients must take an extra supply of medication (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. When travelling across time zones, patients should take their morning replacement dose every 6 hours until they arrive at their destination, and should then return to their usual regimen, in line with their new time zone.

3 Follow Up and Care

GP FOLLOW UP AND CARE IN SPECIAL CIRCUMSTANCES

In addition, GPs and Advanced Care Practitioners should ensure that;

1. They listen to, and respect the expertise that patients with adrenal insufficiency develop around their individual signs and symptoms of low cortisol.
2. All steroid-dependent patients are invited to have an annual flu vaccine.
3. For patients in England, they sign a Medical Exemption form for their patients to receive their medication free of charge: www.hnsbsa.nhs.uk/exemption-certificates/medical-exemption-certificates
4. The patient's prescription of hydrocortisone for use in an emergency IM injection should be prescribed on their current repeat medication list (as opposed to past medication or one-off prescriptions), to alert anyone accessing their Summary Care Record (SCR) of their current and ongoing need for emergency steroid therapy in the event of illness or injury.
5. The patient and any family, friends or carers are competent to administer an IM injection of 100mg hydrocortisone in an emergency and are supplied with a minimum of 2 emergency injection kits, and injectable hydrocortisone sodium phosphate 100mg or hydrocortisone sodium succinate 100mg (powder plus 2ml water vial), as referenced in the NICE Guidelines for the Identification and Management of Adrenal Insufficiency.
6. The patient's SCR has been updated to ensure that their Major Problems list contains the appropriate code for their condition e.g. Adrenal insufficiency or Addison's disease. Avoid using codes such as 'Oral Steroid Therapy' in isolation as this may not immediately convey the urgent need for IM injection of hydrocortisone in an emergency.
7. Steroid-dependent patients are given high priority for after hours or home visits when unwell.
8. The ADSHG Adrenal Crisis Guidelines and Surgical Guidelines are scanned into the patient's medical notes.
9. The patient understands the need to wear a medical ID bracelet such as Medi-tag (which offers a 24hr emergency phone service for members (www.medi-tag.co.uk)), and to carry spare medication with them at all times.
10. The patient has been signposted to the ADSHG for access to support and further resources.
11. The patient has been issued with an NHS Steroid Emergency Card.

FURTHER LEARNING

BMJ Learning: Adrenal Crisis Management

Medical emergencies in primary care: Addisonian (adrenal) crisis Online course | BMJ Learning

RCGP Learning: Adrenal insufficiency

Adrenal insufficiency with a focus on Addison's disease: Summary of Adrenal insufficiency with a focus on Addison's Disease | RCGP Learning

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For further advice please contact:
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Website: addisonsdisease.org.uk

The Addison's Disease Self-Help Group works to support people with adrenal insufficiency, including Addison's disease, 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.

This leaflet has been authored by the ADSHG and reviewed by the ADSHG Clinical Advisory Panel.

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