

Consultation reference guide for adult patients with Addison's Disease (AD)

The following two tables can be useful as consultation guides for endocrinologists and endocrine nurses to support patients with AD at diagnosis, their follow up reviews, treatment monitoring and shared-decision making. These are intended for general guidance and may not apply to all patients or local services.

Table 1: Newly diagnosed patient with Addison's Disease (AD)

What to do	What to look for and investigations										
Cause of AD	- Positive adrenal antibodies (autoantibodies against 21-hydroxylase) - If antibodies are negative, CT adrenals to rule out infiltration or haemorrhage, 17OHP to rule out congenital adrenal hyperplasia, and in males, very long chain fatty acids to rule out adrenoleukodystrophy/adrenomyeloneuropathy										
Clinical history and presenting symptoms	- Decreased appetite and unintentional weight loss - Nausea, vomiting, abdominal pain - Fatigue, lethargy, low energy, reduced strength - Myalgia, joint pain - Low blood pressure, orthostatic hypotension, and dizziness with salt craving - Hyperpigmentation of the skin, mucous membranes										
Signs and biochemistry	<table border="0"> <tr> <td>Hyponatraemia</td><td>Increased thyroid stimulating hormone</td></tr> <tr> <td>Hypoglycaemia (in children)</td><td>Raised serum creatinine</td></tr> <tr> <td>Hyperkalaemia</td><td>Hypercalcaemia (rare)</td></tr> <tr> <td>Low aldosterone</td><td>Low DHEAS (Dehydroepiandrosterone Sulfate)</td></tr> <tr> <td>High renin concentrations</td><td></td></tr> </table>	Hyponatraemia	Increased thyroid stimulating hormone	Hypoglycaemia (in children)	Raised serum creatinine	Hyperkalaemia	Hypercalcaemia (rare)	Low aldosterone	Low DHEAS (Dehydroepiandrosterone Sulfate)	High renin concentrations	
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Screening tests	Paired morning assay of serum cortisol and ACTH: low cortisol concentration (assay-dependent, often less than 100 nmol/L) and a plasma ACTH >2-fold the upper limit of the reference range (for primary adrenal insufficiency).										
Confirmatory diagnostic test	Standard dose 250 µg Cosyntropin (Synacthen) test, administered IV or IM, measuring cortisol samples at 0 and 30 min (some centres also measure at 60 min). Cortisol cut-off values must be interpreted according to local protocol and the assay for analysis.										
Associated autoimmune conditions	- Autoimmune thyroid disease (Hashimoto thyroiditis, Graves' disease) - Type 1 diabetes - Coeliac disease - Autoimmune gastritis with vitamin B12 deficiency - Primary ovarian insufficiency - Hypoparathyroidism - Vitiligo - Family history of autoimmune diseases										
Prescribe	- Glucocorticoid replacement (higher doses at diagnosis) with Hydrocortisone 15-25 mg (depending on weight) as a total daily dose twice or thrice daily (e.g. 10mg on waking + 5mg at 12pm + 5mg at 5-6pm). Where problems with short-acting Hydrocortisone occur, prescribe Prednisolone 3-5 mg once daily or modified release hydrocortisone (Plenadren® 20 mg once daily) - Mineralocorticoid replacement: fludrocortisone at a starting dose of 50-100 µg orally without restricting salt intake. - Trial of DHEA 25-50 mg could be considered for women with low libido and persistent fatigue. DHEA is not licenced in the UK but can be prescribed by specialists.										
Engage, empower, educate and equip	Support self-management as below, refer to the endocrine specialist nurse for patient education, and inform about resources via the Addison's Disease Self Help Group. - Daily glucocorticoid/mineralocorticoid replacement based on individual needs Sick Day Rule 1: appropriate increase in the dose of glucocorticoid tablets dose during physical illness, e.g. flu and infections, or major emotional stress, e.g. bereavement. Sick Day Rule 2: timely administration of parenteral hydrocortisone (100mg IM or IV; SC is an alternative if the only route available) to prevent or treat an adrenal crisis - Preventive strategies for adrenal crisis: symptom awareness, extra supply of tablets, Medic Alert ID, NHS Steroid Card, prescribe Hydrocortisone injection kit, hands-on education for patient and family/friends for hydrocortisone self-administration.										

Table 2: Review for patients with Addison's Disease (AD)

An annual review is suggested for patients with AD, but the frequency should be adjusted based on the patient's needs and control of their condition, also offering [Patient Initiated Follow-Up \(PIFU\)](#) where possible.

What to do	Checks and investigations
Know and record	- Review of cause of AD (? Antibody positive) - Change in symptoms and health status since last review - Current treatment for AD and concomitant medication/over the counter agents, possible drug interactions - Sick days and hospital admissions - Adrenal crises episodes and how was these were managed
Check that patients have	- Up to date Hydrocortisone injection kit and know how to self-administer - Extra supply of tablets for sick days and emergency Hydrocortisone injection kit - The NHS Steroid Emergency Card, wear a Medic Alert ID and additional information such as surgical guidelines, travel letter to carry injection/needles, etc. - Knowledge of sick day rules, symptom awareness/what to do in an adrenal crisis. - Contact numbers for endocrine team (consultant and endocrine specialist nurse) and out of hours emergencies, other support services e.g. patient support groups. - Considered authorising their Summary Care Record to be available for Ambulance Services in replacement of 'Red Flagging' (phased out August 2024).
Review and identify	- Weight, blood pressure - Clinical symptoms suggestive of glucocorticoid over- or under- replacement such as weight, general well-being, signs of frank glucocorticoid excess - Medication side effects, nonadherence to treatment - Ask and advise about glucocorticoid dose and timing adjustment in situations such as shift work, activity pattern, overseas travel, emotional shock - Monitor mineralocorticoid replacement: blood electrolytes and clinical symptoms such as salt craving, postural hypotension, oedema. - Reduce dose of fludrocortisone if hypertension or oedema develops; renin measurement may also be indicated aiming for low end of reference range level. - Monitor dose of DHEA if applicable (benefits vs side effects of hyperandrogenism) - Clinical symptoms suggestive of other autoimmune disorders such as thyroid disease, type 1 diabetes, premature ovarian failure, coeliac disease. , - Plans for pregnancy - advise on pre-conception and pregnancy management. - Need for patient education (sick day rules, management of adrenal crisis), review of episodes of adrenal crisis, medical up dosing or vomiting. - Assess psychosocial needs and wellbeing and refer appropriately for support.
Measure and Screen	- Electrolytes - Full blood count - Calcium - Where other autoimmune conditions are suspected: Blood glucose and HbA1c, B12, TSH, diabetic autoantibodies and coeliac screen in younger adults - Bone mineral density scan as needed
Explain	- Results of tests and investigations - Changes in treatment and how to manage/minimise potential side effects - Possible symptoms related to other autoimmune disorders - How to prepare for their next review: keep a diary of symptoms, adrenal crises and sick days, come with a list of questions, etc.
Refer to	- Endocrine specialist nurses for patient education and treatment monitoring - Other specialists as needed or independent psychology support services such as Health in Mind, Samaritans, RareMinds, or Mental Health Innovations (MHI) where appropriate. - The Addison's Disease Self Help Group for peer and emotional support, educational resources and other support services/materials.

Bibliography & Useful links

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Resources and useful links:

- **NICE Guidance for Adrenal Insufficiency published 28 August 2024:** <https://www.nice.org.uk/guidance/NG243>
- <https://www.addisonsdisease.org.uk/newly-diagnosed-sick-day-rules>
- <https://www.addisonsdisease.org.uk/surgery>
- <https://www.addisonsdisease.org.uk/emergency>
- <https://www.endocrinology.org/adrenal-crisis>
- <https://www.endocrinology.org/media/3717/sick-day-rules.pdf>
- <https://www.endocrinology.org/media/3873/steroid-card.pdf>
- <https://ec.bioscientifica.com/view/journals/ec/5/5/G1.xml>

Resources and videos on managing an Adrenal Crisis

- <https://www.addisonsdisease.org.uk/emergency>
- <https://www.youtube.com/watch?v=oucBVQ0Whq8>

Patient Support Groups - for peer and emotional support, educational resources and other support services/materials.



The [Addison's Disease Self Help Group](https://www.addisonsdisease.org.uk) (ADSHG): www.addisonsdisease.org.uk



[Alex The Leukodystrophy Charity](https://www.alextlc.org) (Alex TLC): www.alextlc.org



The Pituitary Foundation: www.pituitary.org.uk