Patient Safety Notice

PSN057 / June 2021

GIG CYMRU NHS WALES

Llywodraeth Cymru
Welsh Government

Emergency Steroid Therapy Cards: Supporting Early Recognition & Management of Adrenal Crisis in Adults and Children

To: All NHS Wales chief executives, medical directors, directors of nursing, chief pharmacists, medication safety officers, general practitioners, community, district and school nurses, community pharmacists, dentists, paramedics, Digital Health and Care Wales and registered providers of independent hospitals, clinics and medical agencies.

Adrenal insufficiency is a rare disorder, which can lead to adrenal crisis and death if not identified and treated immediately. Adrenal insufficiency can be primary, secondary or tertiary as defined in Table 1.

Table 1: Causes of adrenal insufficiency

	Primary Adrenal Insufficiency	Secondary Adrenal Insufficiency	Tertiary
Cause	Failure of damaged adrenal glands to produce cortisol.	Failure of damaged pituitary gland to produce enough AdrenoCorTicotrophin Hormone (ACTH), resulting in inadequate cortisol secretion.	Suppression of hypothalamic pituitary axis
Conditions	• Addison's disease (autoimmune). • Congenital adrenal hyperplasia. • Congenital adrenal hypoplasia. • Bilateral adrenalectomy. • Autoimmune polyglandular syndrome type 1. • Autoimmune polyglandular syndrome type 2.	• Congenital hypopituitarism. • Pituitary tumours. • Pituitary surgery/radiation. • Pituitary apoplexy • Sheehan's syndrome.	• Exogenous steroid-induced adrenal failure.

Actions

When: Actions to be implemented by 31st January 2022.

Who: All organisations providing NHS-funded care.

1. Disseminate this alert to all staff responsible for the care of patients receiving steroid therapy.
2. Consider if immediate action needs to be taken locally and ensure that an action plan is underway, if required, to ensure compliance with this alert.
3. All healthcare professionals must be aware of the signs and symptoms, management of adrenal insufficiency and crisis and be able to provide treatment in an emergency.
4. All organisations must review their policies/procedures, clinical documentation and digital systems/software to ensure all patients prescribed steroid replacement therapy for known adrenal insufficiency are identified as at risk of adrenal crisis.
5. Healthcare professionals diagnosing and initiating steroid replacement therapy in patients with known adrenal insufficiency (Table 1) must ensure patients are reviewed by the endocrinology service.

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	Primary Adrenal Insufficiency	Secondary Adrenal Insufficiency	Tertiary
Conditions (Cont'd)	• Adrenal haemorrhage, metastasis, medications (cytochrome P450 inducers/inhibitors, monoclonal antibodies etc.), infections, amyloidosis. • Rare causes e.g. Smith-Lemli-Opitz syndrome, ACTH resistance, adrenoleukodystrophy (ALD), mitochondrial disorders.		

All patients with conditions that cause primary or secondary adrenal insufficiency (Table 1) are physically dependent on daily steroid therapy as a critical medicine. Omission of steroid therapy in these patients can lead to adrenal crisis – a medical emergency which untreated can be fatal. Patients with adrenal insufficiency are at an increased risk of experiencing an adrenal crisis during periods of physiological stress such as inter-current illness, trauma or surgery, when increased doses of steroids are necessary.[1] Incidents of adrenal crisis have also occurred on changing the formulation or route of administration for the steroid therapy. Prescribers are advised to seek advice from endocrinology services before changing the formulation or route of administration of steroid therapy to ensure dose equivalence.

Steroid-induced (tertiary) adrenal insufficiency, arising from suppression of the hypothalamic pituitary axis, may occur in patients' prescribed high dose and/or prolonged courses of steroids for inflammatory and other conditions. Abrupt cessation of steroid therapy is a common cause.[2] However, these patients may require additional doses of steroids, even after discontinuation of steroid therapy, during periods of physiological stress (acute illness, trauma or surgery) until full recovery of adrenal function.

Healthcare professionals involved in the care of patients taking steroids for inflammatory and other conditions, must advise patients to seek medical attention during illness and to inform healthcare staff involved in their care of their use of steroids. Healthcare professionals caring for patients prescribed steroids, regardless of indication and/or route of administration, must be vigilant for signs and symptoms of adrenal insufficiency and crisis and manage the patient appropriately for these risks. Table 2 summarises the characteristic signs and symptoms of adrenal insufficiency and crisis signified by the mnemonic Low STEROIDS and 5Ss respectively.

6. Endocrinology services will assess and provide known adrenal insufficiency patients (Table 1) counselling and resources to support the safe self-management of adrenal insufficiency and crisis including:
   - A NHS Wales Emergency Steroid Therapy Card.
   - Signs and symptoms of adrenal insufficiency and crisis.
   - Care plan for the management of adrenal insufficiency during inter-current illness – "sick day rules."
   - Management plan for adrenal crisis.
   - Where clinically appropriate, patient and/or relative training in the administration of emergency intramuscular hydrocortisone and provision of an emergency hydrocortisone injection kit.
   - Emergency contact telephone numbers for their local hospital and specialist endocrinology service.
   - In addition, for children and young people:
     - A school healthcare plan for school aged children;
     - Open access arrangements to the local hospital.
7. Prescribers reviewing and/or treating (including authorising repeat steroid prescriptions) patients with known adrenal insufficiency (Table 1) must ensure the patient has:
   - Routine reviews by endocrinology services.
   - A care plan for the management of adrenal insufficiency and crisis.

Table 2: Signs and symptoms of adrenal insufficiency and crisis

Adrenal Insufficiency Low STEROIDS:	Adrenal Crisis 5 Ss
Sodium, **S**ugar & **S**alt cravings. **T**iredness & muscle weakness. **E**lectrolyte imbalance – Hyperkalaemia – Hypercalcaemia. **R**eproductive changes – Irregular menstrual cycle – Erectile dysfunction. L**O**w blood pressure. **I**ncreased pigmentation of skin. **D**iarrhoea, **D**epression. **S**hedding weight.	**S**udden pain – stomach, back, legs. **S**yncope. **S**hock. **S**uper low blood pressure. **S**evere vomiting, diarrhoea & headache.

Analysis of all incident data submitted to the National Reporting and Learning System (2018-2020) identified four patient deaths, four patient admissions to critical care and another 320 incidents associated with steroid replacement therapy in patients with adrenal insufficiency or emergency management of adrenal crisis.[3] It is reported that one in every 6-12 adult adrenal insufficiency patients will experience adrenal crisis each year (5-10 crises per 100 patient years), with one in 200 patients dying from adrenal crisis.[4,5] The reported incidence of adrenal crisis in children is 3.4-7.2 events per 100 patient years.[6] Paediatric mortality data is scarce but there are sporadic reports of fatalities in patients with undiagnosed adrenal insufficiency.[5,7] A surveillance study of 120 children diagnosed with adrenal insufficiency between 1990 and 2017 reported that 52% of incidents of adrenal crisis occurred at the initial presentation that led to the diagnosis.[6] This highlights the difficulties of symptom recognition and early diagnosis of adrenal insufficiency. There were no deaths from adrenal crisis in this cohort of paediatric patients[6] with research suggesting that deaths in hospital from adrenal crisis is uncommon (<1%).[7] The most common causes of adrenal crisis in both adults and children are gastrointestinal upset, other infection, surgery and physiological stress/pain.[7,8]

Patient and/or relative education is an essential component of the management of AI, enabling patients to maintain an independent fulfilling life whilst protecting them against the life-threatening risk of adrenal crisis.[9] However, research in both adults and children has shown that the impact of patient education on the occurrence of adrenal crisis is negligible. Only 87% of patients reported carrying their emergency steroid cards.[6] Furthermore only 12% and 22% of adult and paediatric patients respectively issued an emergency hydrocortisone kit reported using the kit during acute illness, with patient/relative confidence in using the kit cited as a barrier.[6]

- Received counselling and is comfortable and confident in recognising and managing adrenal insufficiency and crisis.
- A NHS Wales Emergency Steroid Therapy Card and issue a replacement if lost or damaged.
- Adequate supplies of oral steroids for dose escalation during acute illness.
- Where applicable, an in-date emergency hydrocortisone injection kit and issue a prescription for replacement stock when nearing expiration or used.

8. Emergency Department practitioners should inform and refer patients with known adrenal insufficiency (Table 1) and/or crisis to endocrinology services. Replacement stock for patient held emergency hydrocortisone injection kits should be provided prior to discharge from hospital.
9. All organisations that provide surgical services and/or invasive procedures (elective/emergency/day case) must have policies and procedure in place for identifying and managing patients prescribed steroids during the peri-operative period which all healthcare professionals must follow.
10. All healthcare professionals treating, prescribing, dispensing and administering steroids to patients, regardless of indication and/or route of administration, must adhere to the requirements of Welsh Health Circular WHC 2021/008, provide a National Steroid Treatment

Therefore, patient education is not limited to providing information but also requires healthcare professionals confirming patient competence and confidence in the management of adrenal insufficiency and/or crisis during acute illness. Healthcare practitioners treating, prescribing and supplying steroids to patients have a pivotal role in ensuring these patients have received the appropriate counselling and advice on the use of the prescribed steroids and where applicable the management of adrenal insufficiency.

Guidance has recently been issued by the Society for Endocrinology UK, Royal College of Physicians and Association of Anaesthetists on the prevention and emergency management of adult patients with adrenal insufficiency[1] and management of glucocorticoids during the peri-operative period.[10] All healthcare professionals involved in the care of adult patients prescribed steroids should review these guidelines. It is the responsibility of individual healthcare professionals to ensure maintenance of their knowledge, skills and competence to diagnose and manage adrenal insufficiency and crisis through continuous professional development and revalidation.

National Steroid Treatment Card – "Blue Steroid Card"

Healthcare professionals are reminded of the need to adhere to the requirements of Welsh Health Circular **WHC 2021/008** – *Revised National Steroid Treatment Card*,[11] to provide a National Steroid Treatment Card ("Blue Steroid Card") to all patients prescribed steroids for greater than three weeks and considered at risk of adrenal insufficiency from exogenous steroid use.

NHS Wales Emergency Steroid Therapy Cards for Adults and Paediatrics with Primary and Secondary Adrenal Insufficiency

Endocrinology services are responsible for providing adult and paediatric patients with known adrenal insufficiency with a patient-held Emergency Steroid Therapy card. Endocrinology services are responsible for counselling these patients on the management of adrenal insufficiency and crisis. Patients and/or relatives will be counselled on the importance of carrying the card on their person at all times and to show the card to all healthcare professionals they approach for routine and urgent advice/care related to their health, dental problems, medicines etc. These cards will assist healthcare professionals in the prompt recognition of adrenal insufficiency and provides important information on the emergency treatment to be provided if the patient is acutely unwell, experienced trauma, surgery or other major physiological stressors. For children and young people attending school, an agreed school health plan must be in place for emergencies. Primary care practitioners should reinforce to known adrenal insufficiency patients the importance of carrying the Emergency Steroid Therapy Card and can provide replacements if lost or damaged.

Card, advise patients to seek medical attention during illness and to inform healthcare staff involved in their care of their use of steroids.

11. All healthcare professionals caring for patients prescribed steroids, regardless of indication and/or route of administration, must be vigilant for signs and symptoms of adrenal insufficiency and crisis and manage the patient appropriately for these risks.
12. Share any locally developed good practice resources by emailing: ImprovingPatientSafety@Gov.Wales

Notes

National Steroid Treatment card 'Blue Steroid Card' can be ordered by Health Boards from Shared Services via central procurement ('Oracle').

NHS Wales Emergency Steroid Therapy Cards (red card) can be obtained by emailing print.cav@wales.nhs.uk with the order quantity, delivery address and full invoice address. Electronic copies of both the adult and paediatric steroid therapy replacement cards are available on the Welsh Endocrine and Diabetes Society website and the All Wales Network for Paediatric Endocrinology section of the Children and Young Peoples Diabetes Wales Network website.

The content of the adult emergency cards have been reproduced and translated with the permission of the Society for Endocrinology. The NHS Wales Paediatric Emergency Steroid Card is an update to the previous version.

References

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Stakeholder engagement

This alert was developed in consultation with the Welsh Endocrine and Diabetes Society, Welsh Ambulance Service Trust, All Wales Medication Safety Network and NHS Wales Informatics Service, the Brecon Group (All Wales network for Paediatric Endocrinology).