Adrenal insufficiency: improving paramedic practice

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Abstract

Acute adrenal insufficiency, which includes Addisonian crisis, can lead to severe morbidity and even death if ineffectively managed. Unfortunately in the pre-hospital setting patients with acute adrenal insufficiency often receive sub-optimal care. The early administration of hydrocortisone in these cases is critical and significantly improves outcomes to the extent it can be life saving. Such therapy is part of current paramedic practice; however, there is evidence that hydrocortisone is rarely used in the pre-hospital setting. Ultimately, patients with acute adrenal insufficiency may currently be sub-optimally managed by paramedics. To combat this, this article will define the current optimal practice in this area and explain how an e-learning package will be used within North West Ambulance Service NHS Trust to educate paramedics in best practice in this area.

Key words
- Addisonian crisis
- Adrenal insufficiency
- Hydrocortisone
- Life saving

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Adrenal Insufficiency in context

The prevalence of adrenal insufficiency in the UK is 110 to 120 per million and rising (Simmons-Holcomb, 2006). Chakera and Vaidya (2009) observe that Addison's disease has an incidence of 4.7–6.2 per million people in western populations. While the total number of individuals with adrenal insufficiency is low, all of these patients are at risk of adrenal crisis, with 42% of patients experiencing at least one crisis per year (Hahner and Alloio, 2009). Preuss (2001) specifically considered the prevalence of adrenal emergencies as a presenting condition to an emergency department and describes this as a rare event. To date no data was sourced that identified the level of exposure paramedics have faced in terms of this condition. Difficulties in diagnosis are well documented with 60% of patients having seen two or more clinicians before the diagnosis was made (Postiglione, 2013). This diagnostic complexity is likely compounded by the relative rarity of this condition and also the varying signs and symptoms that this condition presents with.

The particular lack of knowledge and awareness concerning patients with adrenal insufficiency in the pre-hospital field, coupled with the low level of exposure that paramedics receive to this condition, signify that it is difficult to deliver the optimal treatment package. This is true even when patients or their care givers communicate the problem to paramedics. McBride (2012) identified an incident where the ambulance crew were told by the patient that they were having an adrenal crisis and urgent hydrocortisone therapy was needed but because the ambulance crew were unaware of the condition they were reluctant to administer the injection. The patient was transported to hospital but only received the essential hydrocortisone and fluid...
therapy after a long period in hospital where the duty doctor had apparently taken senior advice. Baker and Waas (2009) highlight that healthcare workers do not always realize the urgency of which treatment for adrenal crisis must be given and sometimes even fail to heed to the requests of well informed patients for hydrocortisone. This point is illustrated in Case Study 1.

It is apparent that early treatment with hydrocortisone and intravenous rehydration with fluids are essential measures to avoid mortality. It is assumed that pre-hospital clinicians do not currently possess the required education and skill level to safely manage a patient in adrenal crisis. Baker and Waas (2009) state that hydrocortisone should be given within 6–8 hours of presenting symptoms and, as such, ambulance clinicians have an important role to play in delivering this care. The Pituitary Foundation have developed a decision support tool to guide clinicians in identifying the criticality of patient's need (Figure 1). Paramedics should be advised that when patients present in the pre-hospital setting with adrenal insufficiency they may have been in crisis for some time and the clock should start from the onset of illness and not the time of call.

Aetiology
Adrenocortical insufficiency was first recognised by Thomas Addison in 1849 (Preuss, 2001) and is an uncommon life-threatening condition that is readily treated if recognised. The hallmark of the condition is inadequate production of glucocorticoids (Wu et al, 2008). Klauker (2012) observes that this condition was once fatal, but effective treatment with corticosteroid therapy was developed in the 1950s and for that reason a patient's outlook has vastly improved (Margolis, 2012). Adrenal insufficiency may be categorised as primary or secondary, congenital or acquired (Simmons-Holcomb, 2006). In primary adrenal insufficiency (Addison's disease) glucocorticoid and frequently mineral corticoid hormones are lost. Postglione (2013) identifies that in the United States 80% of cases are caused by autoimmune destruction. Glandular infiltration by tuberculosis is the second most frequent aetiology (Schulman et al, 2007). Reisch and Arlt (2009) point out that in western countries autoimmune destruction accounts for 85% of causes but in developing countries tuberculosis is the most common cause.

In primary adrenal insufficiency the adrenal gland cannot produce cortisol or aldosterone or both because of destruction of adrenal gland tissue (Preuss, 2001).

In secondary adrenal insufficiency there is a lack of corticotropin releasing hormone (CRH) from the hypothalamus and/or adrenocorticotropic hormone (ACTH) secretion from the pituitary gland, which results in a malfunction of the adrenal cortex and therefore an insufficiency and clinical presentation (Margolis, 2012). The two adrenal glands are located on top of the kidneys. They consist of the outer portion, the cortex and the inner, which is called the medulla. The cortex produces three types of hormones, all of which are called corticosteroids (Stewart, 2011). While there are

Case Study 1

South Wales HM coroner's report in February 2013 into the death of a 23-year-old nurse. The patient had developed symptoms of gastroenteritis and rang her general practitioner for advice, who had prescribed antibiotics and an antemetic therapy. Later that day the patient deteriorated and the family on scene stated that the patient could not control her breathing. The patient had had her pituitary gland removed some years earlier post brain tumour surgery which had left the patient with secondary adrenal insufficiency. The ambulance crew failed to take the patient to the hospital and subsequently the patient died several hours later. It is worth noting at this point that the HM coroner's report states that the family and also the ambulance service did not have any prior knowledge of an adrenal crisis and that it did not seem to be a part of paramedic training. The coroner summed up by stating that the ambulance crew had serious failings but because of the rarity of the condition and the crew's lack of knowledge the failure cannot be labelled as gross failings (HM Coroner for Neath Port Talbot, 2013).

A = period of first feeling unwell (within, or up to, one hour)
B = period of increasing illness (failure to retain oral cortisol) (during second hour)
C = DANGER ZONE emergency cortisol injection needed (by the third hour)

Figure 1. Cortisol clock
more than 50 steroids that are produced within the cortex, cortisol and aldosterone are by far the most physiologically active.

Cortisol is a glucocorticoid, a type of corticosteroid that helps regulate blood glucose, suppresses the immune response and is released as part of the body’s response to stress; as such cortisol is essential for life (Schulman et al, 2007). Its production is regulated by the pituitary gland, which releases ACTH, causing the adrenal glands to release cortisol. This physiological pathway is illustrated in Figure 2. Glucocorticoids are non-specific cardiac stimulants that activate the release of vasoactive substances. In the absence of corticosteroids stress results in hypotension, shock and even death (Schulman et al, 2007). Cortisol prevents life-threatening hypotension by decreasing capillary permeability and decreasing the production and activity of nitrous oxide during stress (Wu et al, 2008).

This complexity of conditions, each of which produce adrenal insufficiency, could lead the clinician to a range of potential diagnostics labels which are often used interchangeably and loosely by health professionals and patients despite these having specific meanings. Some of the terms that you may encounter are listed in Box 1.

Box 1. Adrenal insufficient patients may state:

- I am all have:
- Cortisol dependant/steroid dependant
- Pituitary patient
- Addison’s disease
- Adrenal deficient
- Hypocortisolism

Defining and exploring the problem
It is clear that education is needed to develop pre-hospital clinicians with their understanding and management of patients in an apparent adrenal crisis (McBride, 2012). On researching the critical incident database for a large NHS ambulance Trust from 2008–2013, only one incident involving an adrenal insufficient crisis was apparent. This figure is minute when you take into consideration the estimated 1.4 million patient journeys per year performed by that ambulance Trust.

This said, critical incident reporting relies on the staff awareness of the situation and
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Table 1. Clinical presentation of adrenal insufficiency

<table>
<thead>
<tr>
<th>Signs and symptoms</th>
<th>% of patients with this sign or symptom on presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weakness</td>
<td>99</td>
</tr>
<tr>
<td>Pigmentation of skin</td>
<td>98</td>
</tr>
<tr>
<td>Weight loss</td>
<td>97</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>34</td>
</tr>
<tr>
<td>Salt craving</td>
<td>22</td>
</tr>
<tr>
<td>Diarrhoea</td>
<td>20</td>
</tr>
<tr>
<td>Constipation</td>
<td>19</td>
</tr>
<tr>
<td>Syncope</td>
<td>16</td>
</tr>
</tbody>
</table>

Patients with acute adrenal insufficiency generally present with acute dehydration, hypotension, hypoglycaemia or altered mental status (Postiglione, 2013)

Box 2. Guidelines for administration of hydrocortisone—adrenal crisis (including Addisonian crisis)

Sudden severe deficiency of steroids (occurs in patients on long-term steroid therapy for whatever reason) producing circulatory collapse with or without hypoglycaemia.

Administer hydrocortisone to:
- Patients in established adrenal crisis
- Steroid-dependent patients who have become unwell to prevent them having an adrenal crisis.

If in doubt it is better to administer hydrocortisone.

From: AACE, 2013

their understanding that the incident will be handled in a non punitive way. This is observed by Vincent et al (1999), who argue that there are significant problems in local and national reporting systems. A study by Lawton and Parker (2002) found that reporting is inhibited by the specific hierarchies of health care where professionals are typically reluctant to report their experiences of error or rule infringement or poor performance because of taboos of whistle blowing and the association that it could hinder career development. Local discussions within an ambulance Trust agreed with this observation, confirming that reporting figures may not be a true reflection of the potential problem. Thus it could be hypothesised that there is a high likelihood of under reporting adrenal insufficiency incidents. The prevalence of adrenal insufficiency is rare; however, it can be approximated based upon epidemiological data that 800 patients in the North West Ambulance Service NHS Trust (NWAS) area of responsibility are likely to have adrenal insufficiency of some cause. Hahner and Allolio (2009) report that 42% of these patients will have at least one adrenal crisis per year. This would equate to approximately 336 possible pre-hospital presentations. Taking this into consideration, to arrange an adequate audit system for adrenal insufficiency would be extremely difficult because of the low expected numbers in comparison to the 1.4 million patient journeys made by NWAS annually.

Anecdotal evidence suggests that there have been instances within ambulance services where paramedics did not recognise and effectively manage an adrenal insufficient patient. A paramedic peer-review session identified a lack of confidence in clinical capabilities to deliver effective care to a patient in an apparent adrenal crisis. This reinforces the belief that education and awareness is required to support clinical development in the area.

Recognising adrenal insufficiency

Acute onset of adrenal crisis is a difficult diagnosis to make as many of the presenting signs and symptoms are non specific (Arlt, 2009). The first symptom listed by the Joint Royal Colleges Ambulance Liaison Committee (JRCALC) (2006) is sudden penetrating pain in the legs, back or abdomen. However, neither Baker and Wass (2009) nor Preuss (2001) reported on penetrating pain in the back, legs or abdomen. This further reinforces the fact that diagnosis is difficult to make and supports the understanding that this condition is very often misdiagnosed and a challenge for primary care medical teams to manage. With this in mind and given the support network available within the hospital environment, there is even greater challenge for the paramedic to come to the same conclusion in an uncontrolled pre-hospital environment.

Wass and Arlt (2012) claim that because adrenal crisis is so rare, it is often not diagnosed on initial presentation. They highlight that the most important symptoms are weight loss, headaches and dizziness, and vomiting. Preuss (2001) quantifies this to state that 97%-100% of adrenal insufficient patients will have weight loss and 87% will have nausea and vomiting. While these are common presenting complaints that can indicate a broad spectrum of differential diagnoses, where other features of adrenal insufficiency are present or a patient reports a secondary cause such
as pituitary surgery, paramedics should have a high index of suspicion of adrenal insufficiency in this group. The signs and symptoms that are commonly observed in the adrenal insufficient patient in crisis are listed in Table 1.

Adrenal Insufficiency and diabetes
Adrenal insufficiency is at least five times more common in the diabetic population (MacCuish and Irvine, 1975). In patients with type 1 diabetes consideration should be given to the development of an associated endocrinopathy such as adrenal insufficiency (McAulay and Frier, 2007). Hypoglycaemia is a relatively common emergency call for pre-hospital clinicians in the United Kingdom. NWAS has developed a pathway to not convey patients to hospital following an apparent hypoglycaemic episode. While this is a valid form of treatment in the majority of patients, for patients with diabetes with signs and symptoms that may indicate adrenal insufficiency, the paramedic should exercise caution and at least consider the possibility of adrenal insufficiency. McAulay and Frier (2007) make the point that clinicians should have an awareness that patients with established autoimmune disorders such as latent autoimmune diabetes are at risk of developing other endocrinopathies.

Treating the patient with acute adrenal insufficiency in crisis
As described in Table 1, the patient who presents in the pre-hospital setting with adrenal insufficiency may use terms that the attending paramedic may not associate with that condition. The paramedic should focus their attention on whether the condition indicates the patient is steroid dependent. If this is the case and the patient is unwell, the paramedic should intervene and administer hydrocortisone 100 mg, ideally intravenously; however, the intramuscular route is an acceptable alternative if venous access cannot be readily established. This said, intravenous fluid therapy may be required to maintain blood pressure in this group of patients; this can be administered according to standard fluid therapy guidelines.

The hydrocortisone therapy aims to prevent the patient having an adrenal crisis and the fluid is supportive. If in doubt it is better to administer hydrocortisone (Association of Ambulance Chief Executives (AACE), 2013). Wass and Arlt (2012) comment that as a rule, taking supraphysiological amounts of steroid over 24–48 hours is not harmful, but too little in the event of a crisis may be fatal. The AACE (2013) guidelines are summarised in Box 2.
Key points

- Rapid intervention with hydrocortisone is essential in a patient with acute adrenal insufficiency/Addisonian crisis.

- Adrenal insufficient patients in crisis may be severely dehydrated and fluid therapy may be necessary.

- Pre-hospital clinicians may be presented with a patient who states that they are 'pituitary patients' or 'cortisol dependent'. Many patients use the term 'cortisol crisis'. This is the same as acute adrenal insufficiency/Addisonian crisis.

- As a paramedic you can help save the life of a patient who takes hydrocortisone but is unable to, by parenteral injection of 100 mg hydrocortisone.

- The most common reason for admission to hospital for adrenal insufficient patients is that they do not take sufficient extra hydrocortisone early enough when they become ill.

Quality-enhancing solutions

While paramedics have had a drug therapy and guideline for adrenal insufficient crisis for over 10 years, it is rarely recognised and treated appropriately. As such it became apparent that a learning resource is needed to facilitate the paramedic to recognise and deliver effective care to the patient in adrenal insufficient crisis.

Cabana et al (1999) observed that there are three barriers to adherence to guidelines: lack of awareness, lack of familiarity and lack of agreement. Moulding et al (1999) point out that a central tenet in health education theory is that behaviour change cannot take place without attention to knowledge and skills gaps. Green et al (1980) argue that a careful assessment of an individual's education needs is important before this can take place. To explore this further, Welsh et al (2006) point out that some students lack the self-awareness needed to act on professional values and this prevents them from enhancing their clinical education. This brings into discussion how paramedics learn and how they maintain their continuous professional development. Reflective practice is taught on many modules which encompass the majority of higher education learning that paramedics are involved in. Dolphin (2013) highlights this and points out reflective practice is taught at university for healthcare students and is crucial to being a good practitioner, allowing the individual to use the experience to improve care. In contrast, Benner (1982) notes that beginners have no experience with the situations in which they are expected to perform tasks. This is also mentioned by Welsh et al (2005), who also note that novices have no experiences to relate to so they cannot fully engage in the reflective process. An argument can be made in the discussion around adrenal insufficiency and sub-optimal treatment of this rare condition that because there is low exposure to this pre-hospital clinicians have no experience to reflect on. Spencer (2003) agrees and states that paramedic training concentrates on life-threatening conditions with protocol-driven practice based on limited underpinning knowledge.

To bridge this education and awareness gap an online learning resource was produced and made available to all paramedics within NWAS. Short et al (2010) claims that continuing medical education (CME) both live and online is an important quality enhancement tool for health care. By producing an online module, Aminz (2000) argues that this allows the student to learn at their own pace and fits in with their own preferred way to learn.

The package will be accessible on the NWAS intranet e-learning zone and available to all 5000 staff in the NWAS. Once completed, the clinicians will be able to download a certificate stating that they have completed the module and be encouraged to self-reflect on the new education gained. This package has been peer-reviewed by the Pituitary Foundation with a hope to convert this into national resources, which can be accessed by all ambulance clinicians.

Patient and public involvement

Patient involvement is highlighted by the Royal College of General Practitioners (2010) statement on safety and quality of care. The health foundation which is an independent charity working to improve health care in the United Kingdom emphasise the importance of this patient-centred approach by reporting that if health staff can engage patients and help them feel involved, patients are likely to hear the advice they are given and, more importantly, act on it (Goose, 2012). For this reason a patient with adrenal insufficiency was approached to tell her story as a part of the learning material. The patient representative was heavily involved in the development of the training package and a voice video displaying a patient's journey through an adrenal crisis will be reviewed to an adrenal insufficient patient group at their bi-monthly support group meetings.

By direct patient involvement with the patient group and by continuous interaction with the
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patient group representative, a level of achievement can be claimed. This can be evidenced on production of the accompanying film of a patient’s journey through adrenal crisis to be displayed on the package. The patient states that she: ‘finally feels like someone is listening and more importantly doing something about it.’

Conclusions

There is evidence to suggest that adrenal insufficiency, which can lead to an acute adrenal insufficiency crisis, is poorly recognised by prehospital based clinicians and therefore not optimally treated. Paramedics and other prehospital clinicians need to be aware that rapid early intervention with the life-saving therapy of hydrocortisone and fluids are essential in the patient in an adrenal crisis.

The prehospital clinician who is presented with a patient in an apparent adrenal insufficiency crisis will have to make a difficult decision based on vague clinical signs and symptoms. Where there is a probability that the signs and symptoms can be explained by adrenal insufficiency, hydrocortisone should be administered to avert adrenal insufficiency crisis. Even if the clinician is presented with a patient displaying similar clinical signs but who is not adrenal insufficient the administration of hydrocortisone will have no detrimental effects.

Conflict of interest: none declared

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