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### **Abstract**

Adrenal insufficiency is rare in childhood. However, it carries the risk of adrenal crisis in the event of illness, stress or injury. An increased risk of morbidity and mortality is associated with adrenal crisis, therefore, parental education and prompt sick day intervention is paramount to the care of a child with this condition. This paper addresses the management of adrenal insufficiency in childhood and provides direction on parental education and prompt sick day interventions. On-going parental support, education and empowerment are vital to ensure appropriate management at home.

### **Keywords**

Adrenal insufficiency, adrenal crisis, hydrocortisone, parental education, parental knowledge.

## **Background**

Adrenal insufficiency is rare in childhood, but in the event of illness, stress or injury, it carries the risk of adrenal crisis. An increased risk of morbidity and mortality is associated with adrenal crisis, therefore, parental education and prompt sick day intervention is paramount to the care of a child with this condition.

The adrenal glands produce several hormones including cortisol which are essential for general well-being. Cortisol, a glucocorticoid, is produced by the adrenal cortex. It is a vital stress hormone involved in maintaining normal physiological functions such as blood glucose levels, blood pressure and blood volume. Cortisol production has a diurnal pattern (Coursin and Wood 2002) which usually peaks in the morning on waking, declines during the day and is at its lowest level at midnight. The secretory rate increases during physiological stress resulting from the feedback mechanism of the hypothalamic-pituitary-adrenal (HPA) axis (Arlt *et al* 2003). In response to stress, the production of cortisol increases up to six-fold (Cooper 2003).

Adrenal insufficiency describes the inability to produce adequate amounts of cortisol, which can result in a failure to generate a sufficient response to illness, trauma and injury. In these circumstances adrenal insufficiency carries the risk of adrenal crisis. Adrenal crisis may result due to hypoglycaemia, shock, and electrolyte abnormalities (Fischer *et al* 2000). Consequences can be severe including coma, organ damage and death. Adrenal insufficiency may result in severe morbidity and mortality (Osuwannaratana *et al* 2008). Mortality can be three times greater in children with adrenal insufficiency than that of the general population (Swerdlow 1998).

## **Aetiology**

Adrenal Insufficiency has a multi-factorial aetiology in paediatrics. It can be characterised as primary or secondary. It can also be congenital or acquired. Therefore, children can have differing presentations with acute or chronic clinical features (Hsieh and White 2011). Primary adrenal insufficiency involves the adrenal gland whereas secondary adrenal insufficiency is a result of hypothalamic/pituitary dysfunction. Children with primary adrenal failure can have both glucocorticoid and mineralocorticoid deficiencies. In contrast, children with secondary adrenal failure do not.

The most common cause of primary adrenal insufficiency in young children is congenital adrenal hyperplasia (Akin *et al* 2010). An incidence of approximately 1:10,000 – 18,000 live births has been reported (White and Speiser 2000). Other causes of primary adrenal insufficiency include autoimmune adrenalitis, autoimmune polyendocrinopathy syndrome, adrenoleukodystrophy and familial glucocorticoid deficiency. Addison's disease is the term used to describe forms of acquired primary adrenal insufficiency. Causes of secondary adrenal insufficiency include hypopituitarism, resulting from chronic suppression due to long term glucocorticoid therapy, and brain tumours.

## **Management of adrenal insufficiency**

The aim of physiologic glucocorticoid dosing is to mimic what the body normally produces (Crown and Lightman 2005). It is important to strike a balance to avoid both under-treatment and over-treatment (Shulman *et al* 2007). Under-treatment can result in increased

vulnerability to illness and poses a potential risk of adrenal crisis. Over-treatment can precipitate side effects of high dose steroid therapy which include weight gain, poor growth velocity and cushingoid features. Hydrocortisone is usually the medication of choice in adrenal insufficiency (Mah *et al* 2004). Prednisolone and dexamethasone are more potent medications and are not recommended for use in the paediatric population for glucocorticoid replacement therapy (Hindmarsh 2009).

A lack of consensus exists between monitoring of therapy and twice versus thrice daily doses (Howlett 1997). However, this must be determined on an individualised basis. Indeed, children with congenital adrenal hyperplasia require higher doses to suppress adrenocorticotrophic (ACTH) production (Hindmarsh 2009). In contrast, children with secondary adrenal insufficiency require a lesser dose. Dose adjustment is based on the child's body surface area, in addition to clinical examination. Auxological measurements also assist in monitoring growth velocity and effectiveness of exogenous steroid therapy (Dauber *et al* 2010).

Health care professionals must be continually aware that prompt intervention is necessary for children with adrenal insufficiency particularly during inter-current illness when glucocorticoid doses may not have been increased or adequately ingested due to vomiting and or diarrhoea prior to presentation to hospital. In addition, parents may have been unable to administer parenteral hydrocortisone at home.

Children who require long-term exogenous glucocorticoid therapy need to be weaned off doses gradually due to suppression of the HPA axis. A tapering regime is recommended as abrupt withdrawal can lead to acute adrenal insufficiency, hypotension and death. However, there is limited data on safe withdrawal in children (Shulman *et al* 2007). A child is at risk of adrenal insufficiency and crisis when doses are weaned to less than the body's normal

physiologic requirements. Parents must be aware of the risks of adrenal crisis, signs and symptoms of adrenal insufficiency and that sick day rules must apply until HPA axis recovery is demonstrated.

### **Sick Day Rules**

Normal maintenance doses of glucocorticoids are insufficient during illness, trauma or injury. To minimise the risk of adrenal crisis, hydrocortisone doses need to be increased. This is known as stress dosing and is crucial in situations such as inter-current infections or trauma (Weise *et al* 2004).

The following principles to stress dosing of hydrocortisone are recommended in Ireland.

1. Doubling or tripling the oral maintenance dose in the event of illness, stress or injury, including febrile illness. This is usually continued for twenty four hours until symptoms have resolved.
2. When a child is vomiting, a hydrocortisone suppository can be administered. If the child's condition does not improve within one hour or if vomiting persists, parenteral administration of hydrocortisone is recommended. Parents need to be aware that young children can extrude the suppository which can compromise effectiveness.
3. If a child has vomiting and diarrhoea, intramuscular hydrocortisone is required. This necessitates prompt intervention and parents should be advised of this. Parents are then advised to seek immediate medical attention.

Debate still exists around the use and reliability of hydrocortisone suppositories at home. Two studies have shown that rectal hydrocortisone is an effective way to administer hydrocortisone if a child is vomiting (De Vroede *et al* 1998, Ní Chróinín *et al* 2003).

Moreover, parents have expressed their preference for rectal hydrocortisone as an emergency glucocorticoid replacement therapy in adrenal insufficient therapy (Ní Chróinín *et al* 2003). Both De Vroede *et al* (1998) and Ní Chróinín *et al* (2003) advise that an initial test dose should be performed to demonstrate reliability. Parenteral hydrocortisone is still advocated when rectal administration is not feasible. Sick day management should be reviewed at every clinic visit; including circumstances that stress dosing has applied (if any) since their last review.

In general, stress dosing is usually required for any procedures under general anaesthetic including minor surgery. Pre-operative fasting times also require close consideration and intravenous fluids may be indicated, especially in younger children. A stat dose of hydrocortisone is given intravenously at induction and repeated if surgery exceeds four hours (Hindmarsh 2009). Stress doses post-operatively are generally determined by the child's speed of recovery.

### **Parental Education**

Once a diagnosis has been established and the child's condition is stabilised, the focus must then shift to the sharing of information with parents to empower them to facilitate self-care for the child and family upon discharge home. The importance of developing a positive working relationship between families and healthcare professionals has been well documented in the literature (e.g. McKlindon and Schlucter 2004; Fisher and Broome 2011). Information provision and education remain central to an effective prevention approach (Hahner and Allolio 2009). Optimal management relies on parents' abilities to identify circumstances that can trigger the risk of adrenal crisis and the significance of early intervention. These are both crucial components in the management of adrenal insufficiency and can predict a safer outcome for the child. Emphasis must be placed on crisis prevention,

particularly during episodes of inter-current illness. Parents must clearly understand when and how to give stress doses, especially in vomiting and diarrhoea. Parents must also clearly appreciate indications for both rectal and intramuscular hydrocortisone and when to bring their child to their local Emergency department.

Knowing how parents may wish to receive this knowledge can help health care professionals. Although related to an adult sample (n=338) with adrenal insufficiency, the findings reported by Harsch *et al* (2010) are relevant to this topic because they illustrate how patients obtain information relating to self-care. Harsch *et al* (2010) report that patients' main source of information was verbal communication by a doctor (89%), and this was the preferred choice of knowledge transfer. Additional sources included support groups (66%) and booklets (60%). The internet (45%) was also identified, particularly in younger participants. Disappointingly, only 52% of questions relating to steroid replacement therapy were answered correctly and 24% of participants reported to have required hospitalisations since diagnosis for management of adrenal crisis (Harsch *et al* 2010). A knowledge gap clearly exists and requires further exploration to identify deficits in knowledge transfer and utilisation.

Addressing the issue of knowledge deficits specific to congenital adrenal hyperplasia, King *et al* (2008) developed the congenital adrenal knowledge assessment questionnaire (CAHKAQ). The CAHKAQ was tested by administering it to 98 individuals; 16 patients with CAH, 58 parents, 14 grandparents, 2 siblings and eight extended family members. Following testing, the questionnaire showed content validity and was deemed internally consistent with a Cronbach's coefficient alpha of 0.67. The final questionnaire contains 22 multiple-choice



questions related to definition, diagnosis, treatment, medication and management of illness. Interestingly, the study revealed that parents had a significantly higher score than patients, prompting the authors to conclude that this result may be because health care professionals normally target their discussions and education towards parents rather than the child (King *et al* 2008). Also interesting was the finding that family members of children diagnosed longer than 10 years had lower knowledge scores which highlights the need for ongoing education as the child negotiates the many stages of childhood development (King *et al* 2008). While the CAHKAQ will be a useful tool for health care professionals, it is limited in that it does not identify deficits in the application of knowledge into behaviour.

An extensive literature search failed to identify any outcome studies directly relating to parental knowledge and competencies in the management of paediatric adrenal insufficiency. This transferability of knowledge to competence warrants further exploration. In addition, the impact of the paediatric endocrine nurse specialist on parental education has not been investigated. Both topics would make interesting studies to allow professionals refine best practice and may have implications on existing practice.

### **Discharge Planning**

The role of the endocrine nurse specialist involves offering support and guidance to families to promote a better understanding of the child's condition and management. Nursing care should directly focus on education and sick day management. Parents must be well-informed and prepared to take the necessary action in the event of an illness, injury or trauma. A suggested checklist prior to hospital discharge is outlined in Box 1. Fostering age-appropriate

effective communication techniques are imperative; children need both reassurance and inclusion in care. In addition, teenagers need to learn how to take responsibility for their own well-being. In particular, they need to be aware of living with a chronic illness and the consequences of non-compliance (Lindsay Waters 2008; Kakleas et al 2009).

**Box 1: Checklist prior to hospital discharge**

Verbal and written information on the specific condition.

Maintain non-hurried approach. Allow time for discussion and address any questions that a family might have.

Steroid therapy card, with both verbal explanation and written instructions on how and when to adjust hydrocortisone doses, particularly if vomiting and diarrhoea occur and when to bring child to hospital

Prescription and clear instructions regarding medication, dosage and safe storage.

Provision of an emergency intramuscular hydrocortisone pack.

Education and instruction on when and how to administer both rectal and intramuscular hydrocortisone.

Advice relating to importance of wearing a medical alert bracelet (Note: chains are not recommended in young children due to the risk of accidental choking).

Contact information for relevant community agencies and support groups (if required)

Provide contact details and telephone number of nurse specialist.

Out-patient's appointment date, a nurse-led clinic appointment may be offered in the interim to review education and to see how families are coping at home.

Specific instructions relating to travel (if applicable)

Nurse-led telephone follow-up, within first week of discharge

## **Steroid therapy card**

Steroid therapy cards in general play an important role in minimising risk for patients (Cheung 2004; Rusby 2010). However, these type of cards can often have deficiencies in terms of limited space for dosing and highlighting important health issues (Cheung 2004). There are many cards available for specific conditions; however, a local audit of steroid cards revealed that parents of children attending our centre wanted written information appropriate to the specialist centre their child attended. A steroid therapy card for adrenal insufficiency was therefore developed following cross-site collaboration between the three Dublin children's hospitals and Cork University Hospital. The card is the size of credit card size with 8 pages of information.

A literature search and review was performed to ensure that information on the steroid card would be evidence based. While the literature revealed no universal agreement relating to optimal hydrocortisone replacement therapy, sick day regimes are, in general, recommended as accepted best practice (Speiser *et al* 2010). The new steroid card includes information relating to personal, next of kin and hospital contact details, medication regimes, sick day management instructions and oral, rectal and intramuscular dosing in the event of illness, stress or injury. Guidelines for healthcare professional are also included on the card on how to treat a child if they present to a local hospital that may be unfamiliar with the management of adrenal crisis

The driving force behind the development of this new steroid card was to empower parents in the care of their child. Although the new steroid card has not been formally evaluated to date, informal feedback from parents reveals positive feedback. Comments offered by parents

include, the card is ‘easy to read’, and they feel ‘more confident now’, ‘feel safer’, they ‘can update medicine’, it ‘avoids confusion, especially on holidays’, and ‘fits in wallet’. A plastic pocket to hold the card was also been introduced as a result of one parent’s suggestion. The steroid therapy card is currently in use between four paediatric endocrinology sites (three in Dublin and one in Cork). It is planned to formally evaluate the new steroid care in the coming months to assess the benefits and relevance of the steroid therapy card to the care of a child at home.

## **Conclusion**

While a rare disorder, adrenal insufficiency can have devastating consequences if not treated promptly. Parents must be well informed and confident in how to manage their child safely at home. Education initiatives can help achieve this objective. Awareness of how to avoid preventable episodes of adrenal crisis may reduce hospital admissions and prevent fatal consequences. This awareness can be improved through effective use of steroid therapy cards for adrenal insufficiency.

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## **Conflict of Interest**

The authors declare that they have no conflict of interest.

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