

BRITISH PAEDIATRIC SURVEILLANCE UNIT

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Autoimmune Addison's Disease in Children

Abstract

The most common cause of severe adrenal insufficiency in children, now known as Addison's disease, is autoimmune. In this condition the body's own immune system attacks the adrenal glands and destroys them. People suffering from Addison's disease are also at increased risk of other organ-specific autoimmune diseases, e.g. of the thyroid gland. If untreated the condition is life threatening and unfortunately it is not always spotted early enough. Though it is now more than 150 years since first described, the disease remains under diagnosed, leading to unnecessary morbidity and mortality. Autoimmune Addison's disease in children is an uncommon but potentially lethal condition.

We intend to undertake a 13 month study to evaluate a rare condition with important clinical relevance. The current incidence of autoimmune Addison's disease in UK (under 16 yrs) will be identified. Clinical presentation and diagnostic strategies will be analysed and reported in the scientific literature. Variations in emergency management will be highlighted (for instance provision of emergency injectable treatment, steroid card and ambulance cover).

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Background

Addison's disease is a rare, life threatening chronic condition which unfortunately is not always spotted very quickly. The incidence of this condition in children is unknown. Further knowledge about the incidence, clinical presentation, diagnostic strategies and variations in emergency management would increase awareness of this condition and help us mitigate the morbidity and mortality in children with this condition.

Coverage

United Kingdom and Republic of Ireland

Duration

July 2011 - July 2012 (13 months)

Research Questions

We intend to undertake a 13 month study to identify the following aspects of autoimmune Addison's disease in the paediatric population in the UK

- 1) How common is Addison's disease in children in the UK and Ireland under the age of 16 years? This would help us define the incidence of this condition in the UK and Ireland
- 2) What are the clinical patterns of presentation, particularly the interval between the onset of symptoms and diagnosis and associated endocrine and non-endocrine conditions?

Answering these two questions would help us understand the clinical burden of this disease in UK, and the level of awareness of this condition amongst health professionals.

- 3) What are the variations in the emergency management of children with autoimmune Addison's disease (for instance provision of emergency treatment, steroid card, ambulance cover)?

This question will inform us about various aspects of emergency management of Addisonian crisis.

Case definition

Any child from birth up to but not including 16 years of age with newly presenting suspected or proven auto immune Addison's disease should be reported on the BPSU orange card. The diagnosis of autoimmune Addison's disease can be made following clinical presentation of adrenal insufficiency or Addisonian crisis.

Analytic Case definition:

A child will be considered to have a diagnosis of autoimmune Addison's disease if the following criteria are met:

Presence of adrenal cortical antibodies at diagnosis or confirmed subsequently

AND

One or more of the following signs and symptoms

- Hyperpigmentation
- Poor growth in weight and height
- Electrolyte abnormalities (Hyperkalaemia, hyponaetremia, hypoglycaemia)
- Addisonian crisis

AND

Low cortisol levels with high ACTH levels

"Addisonian crisis" or "adrenal crisis" indicates severe adrenal insufficiency.

Characteristic symptoms are:

- Severe vomiting and diarrhea, resulting in dehydration
- Low blood pressure
- Syncope (loss of consciousness)
- Hypoglycemia, severe hyponatremia and hyperkalaemia
- Confusion, psychosis, slurred speech, convulsions

Exclusion Criteria: children on steroid medication for other causes.

Reporting instructions

Please report any child seen for the first time in the UK or the Republic of Ireland during the study period who satisfies the case definition regardless of country of birth

If the diagnosis is awaiting confirmation, (adrenal cortical antibodies not done or awaited), the child should still be reported.

Methods

Paediatricians reporting a case of autoimmune Addison's disease through the orange card system will be sent a questionnaire which explores epidemiological and clinical information about the affected child. They will also be sent a follow up questionnaire one year later which explores the research questions. Postage paid return envelopes will be enclosed with questionnaires.

Ethics approval

This study has been approved by West London REC (Ref: 11/LO/0581) and has been granted Section 251 NIGB permission under reference: ECC 6-02 (FT4/BPSU)/2011

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