

Autoimmune Addison's Disease of Childhood

Name

Address

.....

.....

Date

Dear

Thank you for notifying a case(s) for this study, which is being undertaken by the Royal College of Paediatrics and Child Health Surveillance Unit.

This study, covering the whole of the UK and Ireland, aims to provide comprehensive national data on the incidence, pattern of presentation, initial management and variations in emergency management of Auto immune Addison's disease and subtypes in children (<16yrs of age). We hope that the data gathered may allow greater understanding of how this condition affects children, and that it may become a reference for the future.

We will not be contacting your patient or his/her family at any time. Some patient identifiable data are needed to avoid duplication and to allow an estimation of the completeness of reporting. These will be removed once the case has been confirmed to be a unique case and all information you provide will be treated in strict confidence.

The study is funded by the BPSU and has been approved by the MREC and by the Ethics and Confidentiality Committee.

Please do not hesitate to contact Dr Bindu Avatapalle if you have any queries about the questionnaire, or any aspect of the study. If you need any clinical advice regarding the eligibility of a particular case



Autoimmune Addison's disease

Please report any child from birth up to but not including 16 years of age with newly presenting cases of suspected or proven autoimmune Addison's disease. If the diagnosis is awaiting confirmation, (adrenal cortical antibodies not done or awaited), the child should still be reported.

The diagnosis of autoimmune Addison's disease can be made following clinical presentation of adrenal insufficiency or Addisonian crisis.

Surveillance Case definition:

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

One or more of the following signs and symptoms

- Hyperpigmentation
- Poor growth in weight and height
- Electrolyte abnormalities (Hyperkalemia, hyponatremia, hypoglycemia)
- Addisonian crisis

AND

- Low cortisol levels with high ACTH levels

With or without

- Positive adrenal antibodies at the time of notification.

"Addisonian crisis" or "adrenal crisis" indicates severe adrenal insufficiency. Characteristic symptoms are:

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

Exclusion Criteria: children on steroid medication for other causes.

BPSU ID

Consultant name

Name of person completing form (if different)

Date form completed/...../.....

Section 1: Patient Details

- 1.1 Date of Birth
- 1.2 Hospital reference number
- 1.3 NHS number or CHI equivalent
- 1.4 Home post code (first half only)
- 1.5 Ethnic Origin:

A White

British Irish

Any other white background, please specify

White and Black Carribean

Any other mixed background, please specify

1.6 Gender/...0shni42Male 13.7Female re553.47844 0 6 mSBT/TT2 1 Tf1814.7 13fo.26 19D0.027

Section 2: Family History

2.1 Are any first degree relatives (parents or siblings only) affected by Addison's disease?

Yes Please specify the diagnosis and their relationship to the child

.....

.....

No Not known

2.2 Are any other relatives affected by Addison's disease?

Yes Please specify the diagnosis and their relationship to the child

.....

.....

No Not known

2.3 Is there a family history of (first degree or second degree) other autoimmune disease?

Autoimmune disease	Please circle			Relationship to child
Hypoparathyroidism	Yes	No	Don't know	
Mucocutaneous candidiasis	Yes	No	Don't know	
Pernicious anaemia	Yes	No	Don't know	
Vitiligo	Yes	No	Don't know	
Alopecia	Yes	No	Don't know	
Type 1 diabetes	Yes	No	Don't know	
Autoimmune thyroid disease	Yes	No	Don't know	
Other, please specify	Yes	No	Don't know	

Please now complete the following sections about the child's clinical management.

3.3 Which of the features of Addisonian crisis were present?
Please tick.

	Yes	No	Don't Know
Diarrhoea			
Vomiting			
Dehydration			
Hypotension			
Syncope			
Severe Hyponatraemia (Plasma sodium <120mmol/L)			
Hyperkalaemia (Plasma potassium >7mmol/L)			
Hypoglycaemia (Plasma glucose <2.6mmol/L)			
Altered consciousness			
Convulsions			

3.4 Was emergency treatment given during first presentation?

Yes No Don't know

3.5 If yes, details of treatment given in the hospital/ambulance:

Fluid resuscitation Type of fluid used

Corticosteroids Name of drug used

Route of administration

Section 4:

4.1 If first presentation was not an Addisonian crisis, what symptoms and signs were present at diagnosis? Please select and tick an answer for each one

Symptoms	Yes	No	Don't Know
Weight loss			
Diarrhoea			
Vomiting			

Signs	Yes	No	Don't Know
Adrenal crisis			
Short stature			
Hyper-pigmentation of skin			
Postural hypotension			
Electrolyte abnormalities found on incidental investigations. If yes, please specify.			

4.2 Were there any other symptoms at diagnosis?

Yes No

If yes, please specify

.....

.....

4.3 Were there any other signs at diagnosis?

Yes No

If yes, please specify

.....

.....

4.4 Estimated duration of symptoms before diagnosis:

..... years months days

4.5 Were these noted by:

GP Family Other?

Please tick one and provide details if 'Other'

Section 5:

5.1 Does this child have any other autoimmune disorders?

Yes No Don't know

If yes, which disorder?.....

How long have they had this condition? years months

Section 6: Pubertal status at diagnosis

Please indicate the child's pubertal stage at first diagnosis using the Tanner staging given at the end of the questionnaire:

Pre pubertal = Tanner stage 1 girls and boys;

Adult = Tanner Stage 5 boys or shaving or girls post menarche Adult

Pubertal (Stage 2 - 4) Pre pubertal Don't know

Section 7:

7.1 Weight at presentationkg

7.2 Height at presentationcm

Section 8: Investigations at the time of diagnosis

Please provide results of the following investigations at the time of diagnosis

	Results and units	Not done (tick)	Don't Know (tick)
Cortisol			
ACTH			
Adrenal auto antibodies			

8.1 Was Synacthen stimulation test performed? Yes No

Dose of Synacthen

Test result

Normal Abnormal Not done Don't know

Date of report/...../.....

Section 9:

Section 11:

Does this child have significant health problems?

If yes, please provide details (name of condition, treatment)

.....
.....

Section 12:

12.1 If the child has died, date of death/...../.....

Cause(s) of death

12.2 Was the death related to the adrenal pathology?

Yes No Don't know

If yes, please provide details

Section 13: Contact details of person completing this questionnaire:

Your Name (Please print):

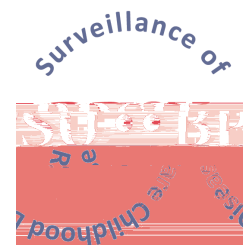
Address:

.....
.....

Telephone: Email:

Date:/...../.....

Thank you for taking the time to complete the questionnaire.



Autoimmune Addison's Disease of Childhood

Name

Address

.....

.....

Date

Dear

**RE: BPSU Autoimmune Addison's disease of childhood
1 year follow up questionnaire.**

In month/year you kindly provided us with details of a case (case NHS number/DOB) of Autoimmune Addison's disease of childhood.

It is now one year since this child was diagnosed with Autoimmune Addison's disease and we would now like to collect further information on each case regarding their treatment and progress during the year since diagnosis.

We would be grateful if you could now complete the attached questionnaire to provide further clinical details of the case.

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Thank you for your contribution to this study.

Dr Bindu Avatapalle

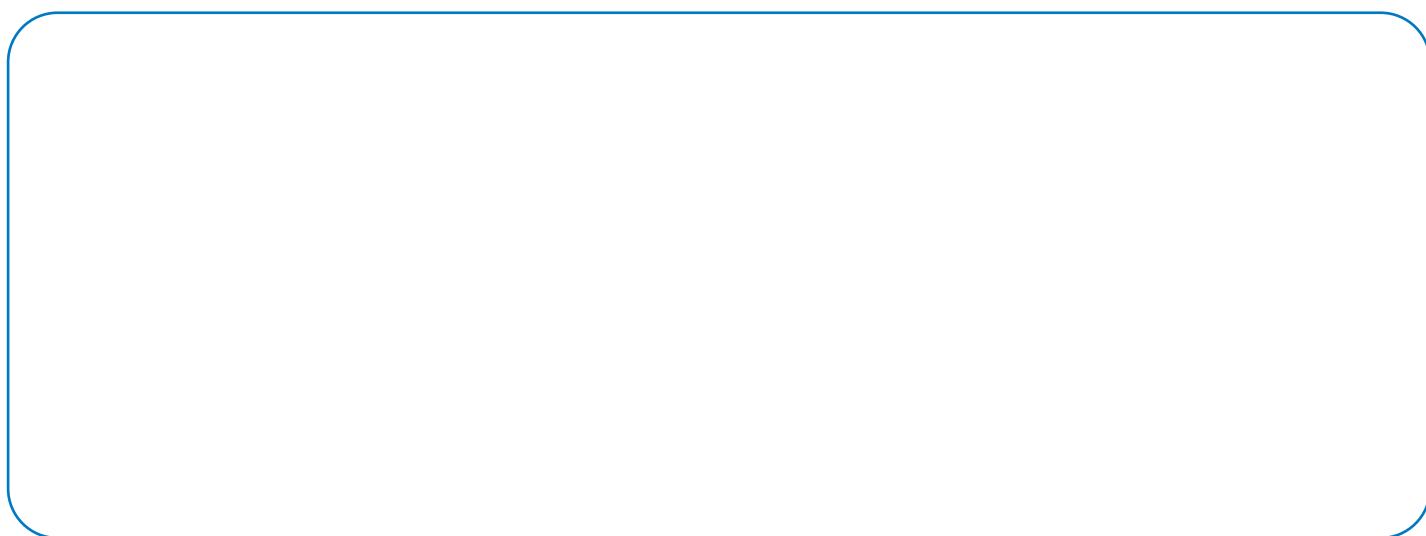
Dr Jerry Wales

Please return the completed questionnaire in the pre-paid envelope to:

Dr Bindu Avatapalle
Department of Endocrinology,
Sheffield Children's Hospital,
Sheffield S10 2TH.

Thank you for filling out our initial case-notification form.
This questionnaire asks you about this child in the year following initial notification.
We would be grateful if you could complete the following questions on clinical outcomes
for this child.

Please provide information for this child for the period from, to the most
recent clinical appointment/admission.

A large, empty rounded rectangular box with a blue border, intended for providing information.

Please now complete the following sections about the child's clinical management. If the child is alive, please complete the questions for the period between and



5.3 Details of treatment given in the hospital/ambulance:

Section 6:

Please note here any further details about the child's progress or condition in the last year that you consider are relevant:

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Thank you for taking the time to complete the questionnaire.

Please return the completed questionnaire in the pre-paid envelope to:

Dr Bindu Avatapalle
Department of Endocrinology,
Sheffield Children's Hospital,
Sheffield S10 2TH.

Boys - development of external genitalia