



The British Paediatric Surveillance Unit (BPSU) is part of the Research Division of the Royal College of Paediatrics and Child Health

**Editor**  
Helen Friend  
Research Facilitator

Tel: 020 7092 6174  
Fax: 020 7092 6001  
Email: [bpsu@rcpch.ac.uk](mailto:bpsu@rcpch.ac.uk)  
Website: [www.bpsu.inopsu.com](http://www.bpsu.inopsu.com)

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## BPSU Celebrates 25 Years of Surveillance



First BPSU Executive Committee

This month the BPSU completes its 25th year of facilitating rare disease surveillance in the UK and Ireland.

Since the BPSU's inception in 1986 over 6,000 paediatricians have participated, returning 90% of orange cards and reporting more than 25,000 cases. Ninety conditions have been studied with the data collected leading to the publication of over 400 papers and presentations both nationally and internationally. BPSU studies have informed a range of

public health policies to control or mitigate the effects of infectious diseases, monitor their effectiveness, and determined the impact of emerging transmissible diseases on children. It has provided evidence for decisions over national screening and contributed evidence for guideline development. The BPSU methodology has been adopted by other specialties in the UK and by national paediatric associations across the world.

*"It is a matter of pride for this country that the innovative and scrupulous epidemiology of the BPSU has been emulated by several other countries in Europe and beyond."*

*Sir Liam Donaldson in the opening to the 15th Annual report*

Without the willing involvement of the clinicians, reporting cases and completing questionnaires, the impact on public health could not have been achieved. The success of the BPSU has also been greatly dependent on the hard work of over 70 clinicians who have worked on the BPSU Executive over the past 25 years, 2 medical coordinators, 8 medical advisors, representatives of the various national bodies involved in the unit and BPSU office staff.



## E-card reporting Trial Commences



In July 2011 a study on Autoimmune Addison's Disease of Childhood will commence. The study will be undertaken by Dr Bindu Avatapalle (inset), the 2009 Sir Tizard Bursary winner and Dr Jerry Wales.

Although first described by Thomas Addison in relation to tuberculosis, currently the most common cause of severe adrenal insufficiency, 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 an increased risk of other organ-specific autoimmune diseases, e.g. of the thyroid gland. It is also known that autoimmune diseases are hereditary.

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.

A 13 month study will be undertaken to evaluate this rare condition with important clinical relevance. The current incidence of autoimmune Addison's disease in UK (under 16 years) will be identified. Clinical presentation and diagnostic strategies will be analysed and reported in the scientific literature. Variations in emergency management will also be highlighted.

**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.

If you would like advice on the eligibility of a case for inclusion in the study please contact: Dr Jerry Wales, Sheffield Children's Hospital, 0114 2717508 [j.k.wales@sheffield.ac.uk](mailto:j.k.wales@sheffield.ac.uk)

## Surveillance of Primary Congenital Hypothyroidism in Children Aged Five and Under

Congenital hypothyroidism (CHT) will be added to the 'Orange Card' for one year from June 2011.

A newborn screening programme for congenital hypothyroidism was first introduced in the early 1980's so many paediatricians might assume that there is little more to learn. However, several important questions remain unanswered and this collaborative<sup>1</sup> BPSU study has been designed to make a major contribution to answering them and, in the process, provide valuable insights into ways to improve patient outcome. This is an important initiative and strongly supported by the BSPED and ESPE, as well as parent organisations, because of its benefits for patient care.

A key aim of the study is to determine how many babies and children are diagnosed each year with CHT, and whether this follows a positive newborn screening result or clinical presentation. Of particular interest are the children who turn out to have transient fluctuations in hormone levels.

In the UK, around 470 children are referred annually for further investigations after a positive screening result for CHT. However, the programme currently has no reliable information about individual diagnostic outcomes and how these relate to screen positive results or screening laboratory cut-off levels (which vary throughout the UK). This study will provide valuable information about the performance of the current UK screening programme for children with suspected CHT, patterns of clinical service provision, management and care, as well as outcomes in the first two years after diagnosis.

A novel aspect of this BPSU study is that we will be using an online questionnaire system, designed to be easier to complete and submit than paper questionnaires, especially for clinicians who regularly see children with suspected CHT. Any paediatrician who reports a case will receive a link by email to the online questionnaire shortly after notification, making the reporting system much more efficient. The National Information Governance Board and Research Ethics Committee have approved the high-level security in this new data collection system and the BPSU and study investigators will also want to hear your views on how well this new system is working.

<sup>1</sup> This UK study will be based at the MRC Centre of Epidemiology for Child Health at UCL Institute of Child Health and involves a team of investigators from the British Society of Paediatric Endocrinology and Diabetes (BSPED), MRC Centre of Epidemiology for Child Health, UK Newborn Screening Programme Centre, UK Newborn Screening Laboratory Network and European Society for Paediatric Endocrinology (ESPE). The study is supported by the British Thyroid Foundation and funded by the UK NHS Newborn Screening Programme. Contact: Rachel Knowles [r.knowles@ich.ucl.ac.uk](mailto:r.knowles@ich.ucl.ac.uk); Juliet Oerton [j.oerton@ich.ucl.ac.uk](mailto:j.oerton@ich.ucl.ac.uk)



## Child and Adolescent Psychiatry Surveillance System

### Surveillance of Raised Blood Lead Levels in Children

The Health Protection Agency's Surveillance of raised blood lead in children (SLIC) study has increased awareness of childhood lead poisoning in all professional groups involved in case management. Information for clinicians now joins the existing resources for the public, environmental health and health protection professionals on the study website. Developed by toxicology experts in the National Poisons Information Service, the Frequently Asked Questions for Paediatricians covers signs and symptoms, diagnosis and treatment options for children with lead poisoning. These are available at [www.hpa.org.uk/chemicals/slic](http://www.hpa.org.uk/chemicals/slic)

### Chylothorax in Infants and Children - Study Ending

After 13 months of surveillance the Chylothorax study will end on 30th June 2011. To date the number of reported cases has been greater than predicted with the infants and children presenting, being treated and managed, in 53 different hospitals across the UK and Republic of Ireland. The response rate to questionnaires is currently 55%, with results awaited from three centres which account for a further 34% of reported cases. Interim results currently indicate that the majority of cases have either a neonatal or post cardiac surgical focus.

To increase ascertainment, data collected by the BPSU will be supported and cross referenced with data obtained through the Paediatric Intensive Care Audit Network (PICANet), Hospital Episode Statistics (HES), and the Central Cardiac Audit Database (CCAD).

Please continue to report any suspected or confirmed chylothorax case in infants or children from 24 weeks gestation to 16 years of age up until the end of June 2011 through the BPSU system. For those who have received a questionnaire, we would be grateful if it could be completed and returned to us as soon as possible. Please do not worry if you feel you are providing duplicate case information, it is much better for us to have duplicate reports or to have reports subsequently withdrawn than to miss cases!

We are extremely grateful to all who have supported this study and if you would like any further information please do not hesitate to contact Dr Peter Davis, e-mail: [peter.davis@uhbristol.nhs.uk](mailto:peter.davis@uhbristol.nhs.uk)

## HPA celebrates BPSU's 25 years of contributions to Health Protection

The BPSU has been making important contributions to paediatric health protection over the past 25 years. For newly emerging infection-related issues such as HIV in children, Haemolytic-Uraemic-Syndrome and vCJD; for the childhood vaccine programme with congenital rubella syndrome and pandemic influenza vaccination and for non-infection related problems such as lead poisoning.

To celebrate the work of the BPSU the HPA is to hold a morning symposium as part of its annual conference on September 15th 2011 at Warwick University.

This session will:

- provide an overview of the achievements of the BPSU in relation to health protection over the past 25 years
- provide insights into specific past and present BPSU studies with a focus on health protection
- provide an opportunity to discuss new developments within the BPSU with both national and international partners.

The session will include presentations from key national and international experts with direct links to the BPSU past and present and be of interest to public health professionals and trainees, clinicians and the wider scientific community.

Speakers already confirmed include Professor Angus Nicoll (ECDC); Dr Chris Verity discussing surveillance of emerging diseases vCJD and H1N1; Dr Bob Adak on the rise of HUS in the UK and the lessons learnt; Dr Pat Tookey on paediatric HIV surveillance and impact on clinical practice and Dr Ruth Ruggles on lead providing an example of environmental paediatric surveillance.

More details from [www.healthprotectionconference.co.uk](http://www.healthprotectionconference.co.uk) or Email: [hpaconference@hpa.org.uk](mailto:hpaconference@hpa.org.uk)

## UK Orphanet site launched

The University of Manchester is hosting the UK arm of the Orphanet database which aims to improve the diagnosis, care and treatment of people suffering from rare diseases across Europe, including a directory of research projects.

The Orphanet website gives researchers, health professionals and patients free access to information on 3,600 genetic, auto-immune or infectious rare diseases, some of which are not yet classified. It also holds information on 'orphan drugs,' intended to treat very rare diseases which sponsors are sometimes reluctant to develop under usual market conditions.

The new UK site [www.orpha.net/national/GB-EN/index/homepage/](http://www.orpha.net/national/GB-EN/index/homepage/) has been recently launched and unlike other Orphanet sites has a section dedicated to paediatrics. Currently populated with BPSU information Orphanet are looking to develop this section. Please send us suggestions on the content you would like to see included to [bpsu@rcpch.ac.uk](mailto:bpsu@rcpch.ac.uk) or [contact@orphanet.co.uk](mailto:contact@orphanet.co.uk)



## UK Orphanet site launched



### BPSU Symposium at the 2011 RCPCH Annual Conference in Warwick

The BPSU held a successful symposium in April to celebrate 25 years of national surveillance at the RCPCH Annual Conference. A guest lecture was given by Professor Elizabeth Elliott, chair of the Australian Paediatric Surveillance Unit.

### International Network of Paediatric Surveillance Units (INoPSU) Conference

Following the success of the INO PSU Dublin meeting a 7th INO PSU meeting will be held in Montreux, Switzerland on 2nd September 2011. The meeting is to be part of the Swiss Paediatric Society Scientific Conference. For further details contact [bpsu@rcpch.ac.uk](mailto:bpsu@rcpch.ac.uk)

### BPSU website

In April the BPSU launched its new website. We welcome comments on design and content to be sent to [helen.friend@rcpch.ac.uk](mailto:helen.friend@rcpch.ac.uk)

### Recently published

DE Nicholls, R, Lynn RM Viner. Childhood eating disorders: British national surveillance study. British Journal of Psychiatry Apr 1, 2011; 198 (4) 295-301

## Analysis

The 2010/11 Annual report is currently being prepared. Analysis shows that the yearly response rate for returned cards and questionnaires remains high at 93%. Nearly 1500 case reports were made in 2010 and to date over 800 have been confirmed. 21% of clinicians have reported a case, with only a third having had more than one case to report.

These figures confirm that after 25 years the BPSU is still being supported by clinicians and we thank you for this support.

**TABLE 1 - % RESPONSE RATE  
(for 6 months)**

Region	% rtd	Rank
North	91.5%	14
Yorks	91.7%	12
Trent	91.9%	11
EAnGl	92.4%	9
NWT	89.7%	16
NET	85.8%	20
SET	89.0%	17
SWT	91.7%	13
Wessx	94.1%	3
Oxfrd	92.0%	10
SWest	92.6%	7
WMids	91.5%	15
Mersy	92.8%	6
NWest	92.6%	8
Wales	97.5%	2
NScot	99.2%	1
SScot	94.0%	4
WScot	87.9%	18
Nlre	93.3%	5
Rlre	87.4%	19
<b>Average</b>	<b>91.9%</b>	

ALL DATA IS PROVISIONAL &  
CONTINUALLY BEING UPDATED

**TABLE 2: Cases followed up to 31.5.2011**

Condition	Started	VALID			INVALID			C&R	D&E	X
		C/R	D	E	X	Total	(as % of Total)			
HIV/AIDS	1986	6,744	757	692	514	8,707	77	17	6	
CR	1990	85	35	56	6	182	47	50	3	
PIND	1997	1,772	372	824	36	3,004	59	40	1	
CD	2008	125	15	44	70	254	49	23	28	
SUPC	2008	45	7	35	4	91	49	46	4	
CNS	2009	114	28	25	30	197	58	27	15	
GBS	2009	79	19	6	35	139	57	18	25	
CSYP	2010	24	5	6	5	40	60	28	13	
GSCT	2010	7	2	18	4	31	23	65	13	
Chylo	2010	57	6	8	38	109	52	13	35	
GA1	2010	9	3	3	2	17	53	35	12	
Lead	2010	7	2	9	3	21	33	52	14	
Neomen	2010	108	14	21	77	220	49	16	35	
<b>Total</b>		<b>9,176</b>	<b>1,265</b>	<b>1,747</b>	<b>824</b>	<b>13,012</b>	<b>69</b>	<b>22</b>	<b>8</b>	

**C** confirmed/already known  
**D** duplicate  
**E** reporting error or revised diagnosis  
**X** status not yet reported to BPSU by investigator