VOL 25 NO 3 OCTOBER 2011

BPSU Bulletin

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

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Inside this issue

Surveillance of Gender Identity Disorder to commence in November

Gender Identity Disorder (GID) will be added to the Orange Card for one year from November 2011. This will be the 3rd joint study between the British Paediatric Surveillance Unit and the Child and Adolescent Psychiatry Surveillance System. The study will be undertaken by Dr. Sophie Khadr, a Walport Lecturer in Paediatrics and Child Health (inset) and involves a team of investigators from UCL Institute of Child Health, the Gender Identity Development Service at the London Tavistock Centre, and the National Children's Hospital, Dublin.

GID is an important condition where a person's gender identity differs from their biological sex. Some describe this as feeling they are in the wrong body or that their gender and body do not match. Children and adolescents with GID experience significant distress, particularly with the physical changes of puberty. There are increased risks of self-harm, suicide and eating disorders. While a psychological condition, concerns about possible hormone abnormalities and inter-sex conditions can prompt initial referral to Paediatricians. After diagnosis, Paediatric Endocrinologists are involved in the medical management of GID, including pubertal suppression and treatment with cross-sex hormones.

Population-level data about the epidemiology, characteristics and outcomes of GID are lacking - information that is critical for optimising service provision and treatment options for individuals with GID. These debates are occurring without reliable data on the incidence, burden and natural history of childhood/adolescent GID, including whether GID in early adolescence is likely to be transitory or persistent.

This study will provide important population-level data about the incidence, clinical presentation, co-morbidities and stability/persistence of gender dysphoria at one and two years of age. These data will inform service provision for and clinical management of this vulnerable group of patients.

The study is funded by a Tavistock & Portman grant via the Advisory Group for National Specialised Services.
Contact: Sophie Khadr s.khadr@ich.ucl.ac.uk; Faye Sweeney f.sweeney@ich.ucl.ac.uk
Further details are available at www.rcpch.ac.uk/what-we-do/bpsu/current-studies/GID

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Review of Patient and Public Involvement in the BPSU

The BPSU has recently published two reports reviewing patient and public involvement (PPI).

Since 2006 the processes for PPI in the research facilitated by the BPSU have been strengthened. These reports provide a review of these developments and how PPI has been adopted in studies currently on the BPSU Orange Card.

The Review has been split into two stages using two different methods:

- The first was undertaken by a working group comprised of members from the BPSU Executive committee. A summary of documentary evidence from the unit, Executive Committee and individual studies record decisions, discussion and use of new PPI processes were collated.
- TwoCan Associates and a steering group were responsible for the second stage. Qualitative research in the form of semi-structured interviews with a range of BPSU stakeholders was conducted. The aim was to understand perception and views of PPI within the activities of the BPSU.

A Summary and the Full Reports can be downloaded from the BPSU website www.rcpch.ac.uk/bpsu/ppi

Supported by the Department of Health

Vitamin D Deficiency - Study Started

It is widely recognised that vitamin D deficiency has re-emerged as a significant health problem for children in the UK, particularly among high risk ethnic groups. However, there is extremely limited epidemiological data regarding the incidence of its various clinical manifestations, of which hypocalcaemic seizures are one of the most dramatic and severe. The majority of such seizures occur in infancy, and are likely to be preventable by dietary supplementation (for the mother during pregnancy, and for at risk infants following delivery).

Despite concerns regarding the increasing prevalence of vitamin D deficiency, there is no consensus on advice regarding preventative vitamin D supplementation for pregnant women and children in the UK. The Scientific Advisory Committee on Nutrition (SACN) recommends that vitamin D supplements should be taken by all pregnant and breastfeeding women, and by children aged 6 months to 5 years who are at high risk of vitamin D deficiency. NICE antenatal care guidelines recommend that 'all women should be informed at the booking appointment about the importance for their own and their baby's health of maintaining adequate vitamin D stores during pregnancy', however, they do not recommend that supplementation should be provided, even for at risk groups. Uptake of vitamin D supplements among pregnant women and children is low.

BPSU surveillance of hypocalcaemic seizures secondary to VDD will be undertaken for 13 months. In addition to the initial case notification questionnaire, reporting clinicians will be asked to complete a 12-month follow-up questionnaire, which will be used to collate information on the management and outcome of cases.

The results of the study will be used to calculate incidence figures for this condition, and provide information regarding the demographic characteristics of affected children and medium term outcomes. This will contribute to our understanding of the effectiveness of current public health policy in preventing clinically significant vitamin D deficiency in UK children.

The study is being undertaken by Dr Emre Basatemur (inset) and Dr Alastair Sutcliffe at the UCL Institute of Child Health. The study is funded by the Sir Peter Tizard Research Bursary from the RCPCH, and has REC (Ref: 11/LO/0838) and NIGB (ECC/BPSU 6-02(FT7)/2011) approval.

Please contact Dr Emre Basatemur if you would like any advice regarding the study, or regarding the eligibility of a particular case for inclusion in the study: emre.basatemur@ucl.ac.uk, Tel: 07585 227 463.

Surveillance of Haemolytic Uraemic Syndrome

In October 2011, a BPSU study on haemolytic uraemic syndrome (HUS) in children will be launched. The study, lead by Dr G K Adak (inset) of the Health Protection Agency (HPA), aims to better define the incidence of HUS in children in the UK and Ireland. The study will provide valuable data on the incidence of HUS, clinical progression and management of cases, and outcomes of disease one year after presentation. By linking to laboratory reporting we will be able to determine which strains of VTEC are causing HUS and through making comparisons to the previous BPSU-HUS study (1997-2001) will allow us to assess changes in epidemiology and clinical management.

HUS is a rare but serious condition that can develop following diarrhoeal illness caused by vero cytotoxin producing Escherichia coli (VTEC). The peak incidence of HUS is in children under five years of age. Patients generally develop HUS a week or more after their symptoms have disappeared; at which time they present a minimal risk to other people, which means the previous regular connection with the HPA may have ceased. Cases of HUS are therefore not often identified through the existing VTEC enhanced surveillance system for England. HUS has been added to the notifiable disease list Schedule 1 of the Health Protection (Notification) Regulations (2010), however, it is known that data collected through the notifications of Infectious Disease system is incomplete. This study will therefore allow us to continue to build a complete understanding of the epidemiology of HUS in the UK and Ireland.

Clinicians in England, Wales, Northern Ireland and the Republic of Ireland will be asked to report cases of HUS under the age of 16 through the BPSU. Scottish clinicians will continue to report through the existing HUS surveillance system run by Health Protection Scotland, and data from this system will be included in the study. The study will run for 13 months with a one year follow-up questionnaire.

Together with the national surveillance system for VTEC, data collected will help identify factors associated with an increased risk of developing HUS, in the hope that we might, in the future, be in a position to prevent at risk children from developing HUS after a VTEC infection.

For further information please contact: VTEC@hpa.org.uk or visit www.rcpch.ac.uk/what-we-do/bpsu/hus

Study Ending

Bacterial meningitis in babies <90 days of age: The current burden of disease

We would like to thank all respondents to this very important study of bacterial meningitis in babies 0-90 days of age. The study is no longer on the orange card as last case reports were cases seen in July 2011.

We had excellent response via the BPSU Orange card reporting system receiving 481 reports of which 349 (72.6%) have been completed and returned and we are very grateful to all the contributing Paediatricians, Pathologists and Microbiologists.

At the moment, 132 (27.4%) questionnaires are outstanding and we would encourage you to return your completed questionnaires if you have not done so. Please do not hesitate to contact the study team at meningitis@sgul.ac.uk if you need a replacement questionnaire or have any questions regarding the study. Our analyses will begin once all case reports are in and we hope to present our study findings in the coming year.

Please send all completed questionnaires to: Dr I O Okike. Vaccine Institute, St George's University of London. 2nd Floor Ingleby House. Blackshaw Road, London SW17 0QT.

Montreux hosts 7th INoPSU Scientific Meeting

In September the Swiss Paediatric Surveillance Unit sponsored the 7th INoPSU scientific meeting. The meeting was part of a wider Swiss Paediatric Society scientific meeting held in the lovely setting of Montreux by Lake Leman. In addition to the Swiss, there were representatives from seven other national units, Australia, Canada, UK, the Netherlands, Portugal and the newest unit Belgium.

The meeting provided an excellent opportunity for representatives from each of the national units to meet and exchange views on rare disease surveillance and discuss issues that currently pose challenges to the units. Funding and processes for ethical approval of surveillance studies were a particular focus.

The meeting comprised of a half day INoPSU business meeting followed by a half day scientific meeting.

The scientific meeting was attended by around 80 conference delegates consisting of paediatricians, epidemiologists and health workers, and enabled INoPSU to showcase its work. Talks included those highlighting the Swiss unit's activities work on HUS, shaken baby syndrome and hyperbilirubinaemia. In addition there were presentations on; alcohol induced coma admissions, coeliac disease (Netherlands) Guillan-Barré syndrome, 25 years of vaccination policy (UK), vitamin D deficiency rickets, patient participation in research (Australia), adverse drug reactions and injuries associated with baby products (Canada). The Belgium surveillance unit also presented data on their activities for the first time in this forum. The presentations can be downloaded from www.INoPSU.com/publications/index.html

The business meeting focussed on the roles of INoPSU, its benefits to members and the importance of good governance and financial accountability. The request for membership by the Belgium Paediatric Surveillance Unit was approved and the unit was formally welcomed to INoPSU. INoPSU has now added a third level of membership "associate" to the two current levels full and affiliate. The new level will allow individual and groupings who have an interest in rare paediatric conditions to join INoPSU. Other decisions made include affirming BPSU as the INoPSU administrative centre, re-developing the INoPSU website, raising the profile of INoPSU activities, and production of collaborative international publications. More detail on all these will be available on the INoPSU website.

The 8th INOPSU meeting celebrating its 15th anniversary will be held in conjunction with the International Paediatric Association conference (www.ipa-world.org/) in Melbourne, 24-29th August 2013.

News in Brief

BPSU – HPA anniversary symposium

The BPSU in conjunction with the HPA led at successful half day symposium at the HPA conference in Warwick in September. The session marked the BPSU's important contributions to health protection in children over the past 25 years in conditions including; newly emerging infection-related issues such as HIV in children, Haemolytic-Uraemic-Syndrome and vCJD; childhood vaccine programmes for rubella syndrome and pandemic influenza and for non-infection related problems such as lead poisoning. Details on the abstracts including presentations are available at www.healthprotectionconference.org.uk

Recently published

Knowles, R; Friend, H; Lynn, R; Mitchell, S; Michie, C; Ihekweazu, C; on behalf of the British Paediatric Surveillance Unit (BPSU) Surveillance of rare diseases: a public health evaluation of the British Paediatric Surveillance Unit Journal of Public Health 2011; doi: 10.1093/pubmed/fdr058

British Paediatric Surveillance Unit 25th anniversary report

This report was recently published and is available online at www.rcpch.ac.uk/bpsu A few hardcopies are available; please contact the BPSU office bpsu@rcpch.ac.uk

And Finally

After four productive years working for the BPSU our research facilitator, Helen Friend, is moving on to a post at the UCL's Institute of Global Health. We wish her well in her new venture.

Analysis

E-card reporting

September saw the end of the e-card reporting pilot. Response rate has averaged 76%, slightly lower than our normal card response. We are currently evaluating the data and will be making a decisions on rolling out the E-card to all BPSU responders.

TABLE 1 - % RESPONSE RATE (for 6 months)

Region	% rtnd	Rank
North	91%	8
Yorks	92%	5
Trent	88%	14
EAngl	87%	16
NWT	88%	15
NET	83%	20
SET	88%	13
SWT	86%	18
Wessx	91%	6
Oxfrd	90%	10
SWest	90%	9
WMids	89%	12
Mersy	91%	7
NWest	94%	2
Wales	94%	3
NScot	95%	1
SScot	94%	4
WScot	86%	17
NIre	90%	11
RIre	86%	19
Average	**89.9%**	

ALL DATA IS PROVISIONAL & CONTINUALLY BEING UPDATED

TABLE 2: Cases followed up to 10.10.2011

		VALID			INVALID		C&R	D&E	X
Condition	**Started**	**C/R**	**D**	**E**	**X**	**Total**	(as % of Total)		
AIDS/HIV	1986	6758	759	711	831	9,059	75	16	9
CR	1990	85	35	31	4	155	55	43	3
PIND	1997	1789	376	825	130	3,120	57	38	4
GBS	2009	104	32	7	59	202	51	19	29
GSCT	2010	7	2	18	18	45	16	44	40
Lead	2010	8	2	10	9	29	28	41	31
SYP	2010	30	5	6	12	53	57	21	23
Chylo	2010	96	9	10	97	212	45	9	46
GA1	2010	9	6	7	17	39	23	33	44
NeoMen	2010	193	33	53	204	483	40	18	42
ESRD	2011	0	0	0	41	41	0	0	100
CHT	2011	5	0	0	136	141	4	0	96
AAD	2011	0	0	0	12	12	0	0	100
VITD	2011	0	0	0	3	3	0	0	100
Total		**9,084**	**1,259**	**1,678**	**1,573**	**13,594**	**67**	**22**	**12**

AIDS/HIV ..Human immunodeficiency virus in childhood
CR.....Congenital rubella
PIND .....Progressive intellectual neurological degeneration
GBS .....Guillain-Barré syndrome / Fisher syndrome
GSCT.....Gonorrhoea, Syphilis, Chlamydia, and Trichomonas infections
Lead.....Raised Blood Lead Levels in Children
SYP.....Congenital syphilis
Chylo.....Chylothorax in Infants and Children
GA1.....Glutaric Aciduria 1
NeoMen ....Bacterial meningitis in babies <90 days of age
ESRD.....End-Stage Renal Disease
CHT.....Primary Congenital Hypothyroidism
AAD .....Autoimmune Addison's Disease in Children
VITD.....Vitamin D Deficiency

C confirmed/ already known

D duplicate

E reporting error or revised diagnosis

X status not yet reported to BPSU by investigator