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BPSU

BPSU Bulletin

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

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Inside this issue

End of term report from our out-going Chair

Alan Emond

The past year has been one of change and transition at the BPSU, but surveillance activity has continued, and the service provided by the unit to investigators continues to be of a high standard.

In August 2012 the core funding for the BPSU from the Department of Health came to an end, and since that time the unit has been supported by contributions from each of the parent bodies of the BPSU (RCPCH, PHE, / UCL-ICH), the Scottish Executive and income from investigators' fees.

We have reduced costs associated with running the unit and continue to provide a very cost-effective service. A new website has been launched and much of our output is now electronic. Thanks are due to our scientific coordinator Richard Lynn and Rachel Winch for their hard work in keeping the BPSU functioning in difficult circumstances.

During the last year, we have continued to move reporting clinicians from paper orange cards onto web based e-cards, and the proportion now returning electronically has increased to 70%. We have realised that clinicians opting for e-reporting need to be chased up quickly if they don't respond, as once the e-mail drops off the screen and into the inbox, it disappears from the clinician's consciousness! However, with these regular reminders, the overall response rate has been kept above 90%. We have also piloted the use of on-line data collection questionnaires. Its use will be evaluated but we expect it to be quicker and easier for the clinician and for the researcher, but this methodology requires high standards for data security and data management.

The work on public and patient involvement (PPI) in BPSU has continued in 2012-13, implementing the recommendations of the external review undertaken the year before. PPI is so important in research which does not involve individual consent, to ensure openness, transparency and accountability to the public. A guide for researchers on how to involve the public in their research has been produced by TwoCan Associates and the BPSU and is available on the website: www.rcpch.ac.uk/bpsu/ppi. I especially wish to thank the two lay representatives on the BPSU Scientific Committee, Ann Seymour and Sue Banton, for their hard work over many years in developing PPI at the BPSU.

Support available for investigators has also been enhanced, with clearer guidance, supporting documentation and questionnaire templates available on the website. The two medical advisors, Rachel Knowles and Dominik Zenner and our scientific coordinator put in a lot of time and effort into helping researchers get their ideas into clear scientific proposals, and taking them through the process of gaining the necessary permissions. A clearer contract has now been developed to clarify the expectations of the BPSU of researchers, and what investigators can expect from the BPSU. I am very pleased to announce that the Tizard bursary for young investigators will now be re-established – further details on page 3.

In the autumn, I will be stepping down as chair of the BPSU Scientific Committee, and I will miss the very stimulating science and discussing the wide variety of conditions researched by the BPSU. Looking back over the last four years the funding environment has become much harsher for research, but the BPSU has continued to develop and modernise. We have managed to move to electronic reporting and online questionnaires, expanded the range of topics investigated with joint projects with psychiatrists and cardiologists, initiated a fast-track process to respond to urgent public health issues, and developed good practice in public involvement in research. The BPSU can offer a unique and cost-effective way of undertaking active surveillance across the whole of the UK and Ireland, and it continues to maintain high coverage rates only because of the reporting clinicians who believe the work to be important – thanks very much to you all!

Prof Alan Emond
Chair, BPSU Scientific Committee

New Committee Members

Dr Kathryn Johnson, Dr Sam Oddie, and Dr Alastair Sutcliffe have now joined the BPSU scientific committee. Kathryn is a neonatal paediatrician and is responsible for promoting research within the Leeds Neonatal Service. Alastair is a reader in General paediatrics at the UCL- Institute of Child Health having previously been trained as a paediatric epidemiologist and is supervising the Sir Peter Tizard project on hypocalcaemic seizures secondary to vitamin d seizures. Sam is a Consultant Neonatologist at Bradford Teaching Hospitals and was the principal investigator on the BPSU hypernatremia study.

Two new lay members have been nominated to the BPSU scientific committee – Mrs Madeline Wang and Dr Jane Sutton. Madeleine has, for many years, been actively involved in health related community work with children and young people. She has advised NRES, MHRA, and Royal College of Anaesthetists on the development of information materials for children and young people. At present she is working as a patient advocate helping patients get the information from healthcare providers they want. Jane has wide ranging experience as an NHS professional. For 13 years she worked as a Public Health Specialist in Leicestershire. In 1991 she completed her PhD on 'Accidents to Patients in Hospital' and is a Member of Medicines for Children Research Network.

We say goodbye to Colin Michie and Simon Mitchel and lay member representatives Sue Banton and Ann Seymour. Thanks very much to all of them for many years' support for the BPSU!

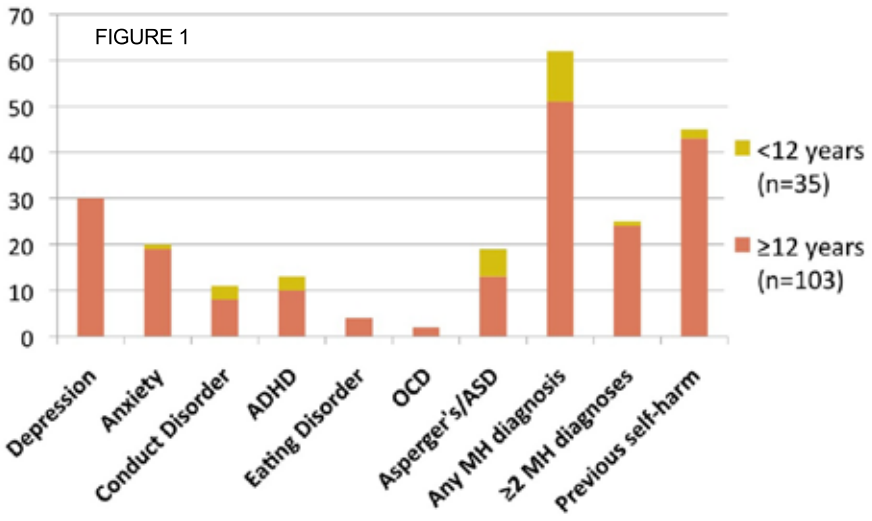
Study News

Dr Sophie Khadr

Surveillance for the **gender identity disorder (GID)** study recently came to an end, though the 2 year follow-up to May 2015 continues. Please do return any outstanding questionnaires. Here Dr Sophie Khadr (inset) updates some of the analysis. In the first 15 months of surveillance (November 2011 to January 2013), there were 410 notifications.

138 cases have been confirmed (taking into account duplicates from more than one source). A number of other cases are under review due to missing data and have not yet been confirmed or excluded. Cases of children who did not meet the case definition, such as prevalent cases, children over 16 years at diagnosis, those with a disorder of sexual differentiation (DSD), or reporting errors have been excluded. Median age at diagnosis is 14.53 years. Only 25% of cases presented at less than 12 years of age. Fifty per cent of cases reported by paediatricians were under 12 years at notification and paediatricians reported 9/35 cases aged less than 12 years (26%). Median age at onset of symptoms is seven (IQ4-12) years. 91% of cases to date are white ethnicity and 50% of cases (n=69) are male. Significantly more (75%) of those who present early (at less than 12 years) are male, with more girls than boys (60% vs. 43%) diagnosed later in puberty (at ≥ 12 years).

Psychiatric co-morbidity is common at diagnosis, particularly in those aged ≥ 12 years (Fig 1). Males and females appear to be affected in equal measure. 45% of the whole sample had at least one other psychiatric diagnosis at notification: 51/103 of those aged ≥ 12 years and 11/35 under 12s. Nearly half of the sample has experienced bullying requiring school action, with reduced school attendance reported in 25%. 10% of our sample was home schooled or receiving hospital schooling.



A key preliminary finding from this study is that similar numbers of male and female children appear to be affected by GID, although more of those who present early (at less than 12 years of age) are male. A significant proportion of children and young people with GID have associated mental health problems at diagnosis, particularly the adolescent group. Asperger syndrome and ASD, observed in about 1% of the general population, were reported in 14% of GID cases. The nature of this apparent association is unclear. Reduced school attendance in a quarter of cases (with 10% home schooled or hospital schooled) has the potential for significant impact on schooling.

Public patient engagement: Mermaids – www.mermaidsuk.org.uk
Gendered Intelligence - genderedintelligence.co.uk

Researcher contacts: Dr Sophie Khadr, Email: s.khadr@ucl.ac.uk

Study News

Surveillance of **Progressive Intellectual and Neurological Deterioration (PIND)** is to continue. PIND has agreement from the BPSU to continue surveillance until end of April 2014 and the Department of Health has given the study further funding.

Cases of variant Creutzfeldt-Jakob disease (vCJD) in children could still occur in the future. In the absence of a diagnostic test the PIND study is the only practical way to search for vCJD cases in the complex group of children/young people with progressive neurological disease under the age of 16 years. The PIND Expert Group reviews the clinical data on diagnosed and undiagnosed children with progressive neurological disease and thus search for possible cases of vCJD in children, who may have a clinical presentation different from that in adults. If the PIND study finds no cases in this age group it provides supportive evidence that public health measures have been effective.

There is still no evidence of vertical transmission but we cannot be confident that this may not happen. Similarly there is increasing concern about secondary transmission to children via surgical instruments and through dental procedures; it is also vital to exclude the possibility of vCJD cases infected through blood transfusion. The PIND study additionally provides unique epidemiological data on the many different causes of progressive neurological disease in children and the variation in the incidence of these disorders in different ethnic groups. Figure 2 outlines the 6 most common diagnoses to date. These findings can contribute to the appropriate planning of diagnosis, clinical management and the provision of services.

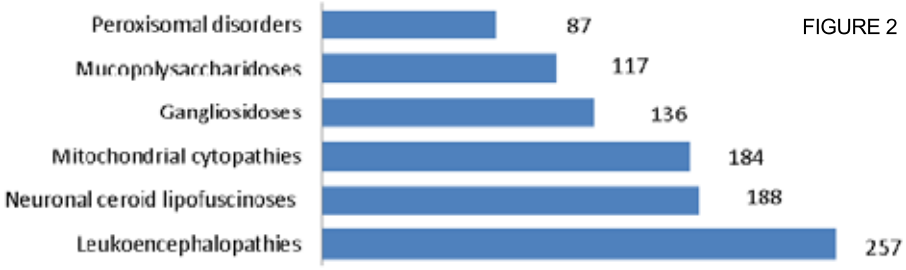


FIGURE 2

We continue to receive positive feedback from paediatricians telling us the PIND study is very useful to them in their practice and the study also attracts international interest due to the diversity of data we regularly publish and present at scientific meetings.

Researcher contacts: Dr C. Verity, L. Stellitano & A. M. Winstone. Email: lesley.stellitano@addenbrookes.nhs.uk

In the News

New studies

One commenced in 2012, the Tizard bursary funded study into surgical ligation of the Patent Ductus Arteriosus in premature babies (investigator Dr Lleona Lee – Nottingham City Hospital) in October. Two studies have started in 2013, a study on Kawasaki Disease (investigator Dr Robert Tulloh, RHSC Bristol) and acute pancreatitis (investigator Dr Julian H Shield, RHSC Bristol). Studies on acute rheumatic fever, self-harm, hepatitis and exchange blood transfusion have been approved and are awaiting funding or ethics review before commencement.

Extensions

Six studies had their period of surveillance extended: HIV, congenital rubella, progressive intellectual and neurological deterioration (PIND), lead in children and congenital syphilis and gender identity disorder.

International activities

The BPSU continues to take an important role in the development of INoPSU. This year saw the BPSU hand over the administration of INoPSU to the Australian Unit, who will be hosting the 8th INoPSU conference in Melbourne in conjunction with the International Paediatric Association conference.

Recent publications

During 2012/13, there were 19 publications relating to BPSU studies. The most recent has been the joint BPSU/psychiatry study on conversion disorder: Incidence and 12-month outcome of non-transient childhood conversion disorder in the UK and Ireland C Ani, R Reading, R Lynn, S Forlee, E Garralda. *BJP* published online April 25, 2013 Access the most recent version at DOI: 10.1192/bjp.bp.112.116707.

RCPCH/EPA conference Glasgow

The BPSU scientific coordinator presented two papers at the European Paediatric Association conference in Glasgow, one on INoPSU and the other on multi-national comparison of early onset eating disorders. Whilst in RCPCH plenary sessions there were presentations on Gender identity disorder (Dr Sophie Khadr), Bacterial meningitis in neonates (Dr Ifeanyichukwu Okike), Vitamin D dependent seizures (Dr Emre Basatemur). Abstracts are available http://adc.bmj.com/content/98/Suppl_1.toc

Tizard Bursary

We are pleased to announce that the Tizard bursary is to be re-launched. On offer is a free slot on the orange card, worth £10,000 plus upto a £1,000 stipend. Look out for details in the autumn on how to apply. In the meantime visit www.rcpch.ac.uk/bpsu/bursary

2012 Data Review

Participation in the orange card scheme remained high during the year 2012

The overall card return compliance rate for the year 2012, calculated as a proportion of orange cards returned, was 93.3% (39,319/36681) a rise of 1.9% from 2011. Monthly response rates ranged from 91.6% in December to 95.1% in July with a median of 93.3%. Wessex is the highest reporting region with an average monthly response of 96.9%. Ireland is the lowest 88.4%. Scotland and Wales rankings have fallen the greatest and this may reflect problems with the e-card system and local NHS firewalls.

Workload of those reporting in the scheme

81% (3043) of participants had no cases to report in 2012, 11% (382) reported a single case, 5.5% (186) reported between two and four cases and 1.7% (61) reported five or more cases. The greatest number of cases reported was by HIV/AIDS specialists, one of whom reported 90 cases!

E-reporting

We now have over 2200 (70%) respondents reporting via our E-web service. Of these we receive 1500 (70%) within a week. But we have to send out weekly reminders to get the response up to 90%. For those who wish to change there is now a box on the postal card which you can tick and we will switch you ASAP or you can contact the office bpsu@rcpch.ac.uk

TABLE 1 – Response rate 2012

Region	Return %	Rank 2012
East Anglia	96.3	3
Mersey	95.6	4
NET	91.0	17
North Scotland	94.5	7
North Western	91.9	14
Northern	94.6	6
Northern Ireland	90.0	18
NWT	91.2	16
Oxford	92.7	13
Rep of Ireland	88.4	20
SET	94.4	9
South Scotland	92.9	12
South Western	96.4	2
SWT	91.3	15
Trent	93.5	10
Wales	93.1	11
Wessex	96.9	1
West Midlands	94.4	8
West Scotland	89.5	19
Yorkshire	95.3	5

DATA IS PROVISIONAL AND SUBJECT TO CHANGE

TABLE 2: All cases reported and follow ups to 02.07.2013

		VALID			INVALID		C&R	D&E	X
Condition	Started	C/R	D	E	X	TOTAL	(as % of total)		
AIDS/HIV	1986	7501	788	714	1,094	10,097	74	15	11
CR	1990	85	35	61	10	191	45	50	5
PIND	1997	1960	415	902	116	3,393	58	39	3
Lead	2010	22	3	10	23	58	38	22	40
SYP	2010	53	9	9	25	96	55	19	26
VITD	2011	42	4	18	67	131	32	17	51
HUS	2011	87	63	32	100	282	31	34	35
GID	2011	20	1	76	41	138	0	185	30
PDA	2012	94	51	20	190	355	1	11	54
Kawasaki	2013	4	0	4	159	167	2	3	95
Apan	2013	0	0	0	23	23	3	0	100
Total		**9,868**	**1,369**	**1,846**	**1,848**	**14,931**	**66**	**22**	**12**

AIDS/HIV ... Human immunodeficiency virus in childhood
CR ..... Congenital rubella
PIND ..... Progressive intellectual & neurological deterioration
Lead..... Raised Blood Lead Levels in Children
SYP ..... Congenital syphilis
VITD ..... Seizures Vitamin D Deficiency
HUS ..... Haemolytic uraemic syndrome
GID ..... Gender identity disorder. Excludes psychiatry reports
PDA ..... Surgical ligation of patent ductus arteriousus
Apan ..... Acute pancreatitis

C confirmed/ already known

D duplicate

E reporting error or revised diagnosis

X status not yet reported to BPSU by investigator

BPSU Surveillance of Rare Childhood Disease
Public Health England
RCPCH Royal College of Paediatrics and Child Health
UCL INSTITUTE OF CHILD HEALTH