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BPSU

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

Editor
Richard Lynn
BPSU Scientific Coordinator

Tel: 020 7092 6173/4
Fax: 020 7092 6001
Email: bpsu@rcpch.ac.uk
Website: www.rcpch.ac.uk/bpsu

Inside this issue

BPSU Bulletin

Surveillance of deaths in children with epilepsy commences this October

Dr Omar Abdel-Mannan

This October saw the commencement of a 13 month study into sudden unexpected death in children with epilepsy. The study is being led by Dr Omar Abdel Mannan (inset) and Professor Alastair Sutcliffe from UCL Great Ormond Street Institute of Child Health.

It is widely recognised that epilepsy is the most common long-term condition of the nervous system, affecting over 600,000 people in the UK. It is associated with significant NHS burden, economic and personal costs and suffering. People with epilepsy are 2 to 3 times more likely to die early than the general population, from a number of causes including sudden unexplained death in epilepsy (SUDEP), associated with co-morbidities (e.g. pneumonia complicating cerebral palsy as a terminal event), drowning and status epilepticus.

SUDEP is the subject of intense on-going study in adult patients, but there are few studies of SUDEP in childhood, with the incidence in children in addition to associated risk factors and underlying mechanisms are largely unknown. A prospective pilot study is much needed, to determine the size of the problem of mortality in children with epilepsy, to compare the differences between SUDEP and other causes of epilepsy deaths, and to allow future studies in this field.

The study will contribute to our understanding of the epidemiology of mortality in epilepsy in children in the UK and Ireland, and help us to evaluate the effectiveness of current public health measures in preventing these premature deaths. This research will be of benefit to children with epilepsy, their families and the clinicians who manage them in that they will be able to provide more accurate evidence-based information regarding SUDEP in children. This research will help healthcare providers, advocacy groups, and others interested in improving outcomes for children with epilepsy to work together to prevent complications associated with epilepsy and reduce the risk of premature death amongst this group.

Case definition: Please report any new cases of a child who dies aged between 1 month and 16 years of age who also has the simultaneous diagnosis of epilepsy. The child must have had seizures or been treated with antiepileptic medication within the last 5 years.

Also please report all suspected cases, even if the results of investigations are pending.

The study is funded by the charity SUDEP Action and has been approved by the London Central REC (Ref: 16/LO/1265) and granted Section 251 CAG permission (Ref: 16/CAG/0093).

Patient support details: Epilepsy Action (www.epilepsy.org.uk) and Epilepsy Research UK (www.epilepsyresearch.org.uk). A public information leaflet on the study is available at www.rcpch.ac.uk/bpsu/epilepsy and you are encouraged to display this within the hospital.

Please contact Dr Omar Abdel-Mannan if you would like any information on the study, or regarding the eligibility of a particular case for inclusion in the study:
E-mail: ich.bpsu.sudep@ucl.ac.uk Tel: 07717 74 7012

Rare Disease Day conference February 27th 2017

The BPSU in collaboration with Birmingham Children's Hospital are holding a second Rare disease day conference on 27th February 2017. The theme is "From bench to bedside new treatments for children with rare disease". A key note speech will be received from Dr Gina Radford, Deputy CMO for England. Presentations will also be received from, Dr Larissa Kerecuk, Rare Disease Lead Birmingham Children's Hospital; Professor Timothy Barrett, Rare Disease - Translational Research Consortium, Paediatric Cross Cutting Theme Lead; Sheela Upadhyaya, Associate Director of the Highly Specialised Technology program, NICE.

For further information on registration please visit www.rcpch.ac.uk/bpsu/rdc17

Supported by Public Health England, Royal College of Paediatrics and Child Health, and UCL-Institute of Child Health with support from GOSH Children's Charity

Study updates

Dr Michael Absoud

A 13 month joint BPSU – CAPSS study on **childhood disintegrative disorder (CDD)** will commence in November. The principle investigator, Dr Michael Absoud from Evelina Children's Hospital, reports "CDD is a rare condition where a previously normal child very rapidly, sometimes even over a few days, loses intellectual and developmental skills. Children then stop communicating and playing with other children, and cannot look after themselves, often resembling a severe form of autism.

At present, we do not know what causes this devastating condition. Approaches to investigate and manage this condition vary between clinicians, leading to confused messages to parents and carers, adding to the distress already encountered.

To begin to unravel such rare conditions, we first require knowledge on the incidence of CDD, the true spectrum of children with such presentation, how they are investigated and cared for, and crucially, their outcome. With a one and two year follow–up we will analyse this information to help inform on the optimal delivery of appropriate services, support, and interventions for affected children and families. Following this and with consent, we hope to undertake a long term follow-up of the cohort.

Please report any child younger than 16 years of age presenting to you for the first time and seen in the last month, who has had a significant regression in language, play, adaptive behaviour, or functional skills, resulting in impairments similar to Autism Spectrum Disorder who apparently had normal development for at least the first 2 years of life. Please report children even if currently they are undergoing medical investigation for exclusion purposes. Please report a child whose regression may have occurred earlier, but is presenting to you for assessment for the first time."

Do not report children with classic autistic stasis/regression of language and social skills occurring before the age of 1 year 11 months. Do not report children with a static brain lesion such as acquired brain injury or those meeting the criteria for the BPSU PIND study. [See analytic case definition http://www.rcpch.ac.uk/pind]

We appreciate the case definition is complex and details are available from http://www.rcpch.ac.uk/bpsu/cdd

The study is funded by the charity The Shirley Foundation and has been approved by the London Bloomsbury REC (Ref: 16/LO/0799) and granted Section 251 CAG permission (Ref: 16/CAG/0061).

Patient support details available from Autistica UK (www.autistica.org.uk) and a public information leaflet on the study is available at http://www.rcpch.ac.uk/bpsu/cdd and you are encouraged to display this within the hospital.

Please contact Dr Michael Absoud if you would like any advice regarding the study, or regarding the eligibility of a particular case for inclusion in the study: E-mail: michael.absoud@gstt.nhs.uk Tel: 020 7188 4665

Study update

The **Female Genital Mutilation** (FGM) study aims to review the incidence and clinical presentation of FGM in the UK and Republic of Ireland in children aged 0-16 years. The study is being led by Dr Deborah Hodes from University College Hospital in conjunction with the RCPCH Research and Policy division. The project went live on the 1st November 2015 and unlike the traditional method of paper based BPSU data collection methodology, this project is the first to use of an online clinical questionnaire and reporting system.

A total of 54 cases were notified in the first ten months (to September 2016) of which 20 (37%) are confirmed cases, 13 (24%) are reporting errors and data on 16 (29%) is still awaited, 5 cases are duplicates (9%). Given the concern of the condition and the need for the continued collection of reliable data the BPSU have agreed to continue surveillance on the basis that further funding can be secured.

Please do report any cases of children 0-16 years arising since November 1st 2015 presenting with either FGM or labioplasty defined by the case definition.

Also **please** do remember that there is a statutory requirement to report cases to the appropriate authorities. A recent Home office committee report on FGM questioned the willingness of medical profession to report cases. On behalf of its members the RCPCH robustly responded. http://www.rcpch.ac.uk/news/rcpch-responds-home-affairs-committee-report-fgm

The study protocol information and a lay public information guide which can be distributed in your ward/clinics is available from http://www.rcpch.ac.uk/bpsu/fgm

For further information on the study contact karina.pall@rcpch.ac.uk

Severe visual impairment and blindness

So far 260 cases have been reported and 78 have been confirmed, 135 report forms awaited. If you have an outstanding questionnaire to return could you please do so as each is important. The last surveillance month will be November. Details on the study are available from http://www.rcpch.ac.uk/bpsu/bcvis2

Study updates continued

ADHD Transition study – regional ascertainment issues!

The study, led by Professor Tamsin Ford of Exeter University, commenced in November 2016 was initially planned to run for 7 months has now been extended to 13 months due to the fewer than expected case reports. This may reflect the true incidence of the condition or under ascertainment; the extension will allow investigation of both.

Whilst undertaking cross comparative analysis with the child psychiatry reporting, potential holes in regional reporting have been noted. This may be due to how and where the cases are being seen and treated. Areas for which there are particularly low case reporting include North and Central Scotland, North West, Welsh borders, N&W Wales, South Yorkshire. If you have seen a case this past year there is still time to report

To date we have had 265 case notifications from BPSU (that is reported cases using the "orange card system"). Of those notifications, 143 are confirmed case, 60 forms awaited. The psychiatrists have reported 111 cases, returned 45 questionnaires of which 38 are eligible cases.

We appreciate that many of you reporting these cases have a high caseload and are snowed under with all sorts of requests. No question this study adds to that pressure. However, this study will shed a light on the woeful situation that leads to the pressure that you work under. Without the data, we cannot be nearly as effective; so, please contact us to discuss a more manageable strategy to collect the necessary data.

The study protocol information and a lay public information guide which can be distributed in your ward/clinics is available from http://www.rcpch.ac.uk/bpsu/ADHD

For further information please contact; Professor Tamsin Ford: t.j.ford@exeter.ac.uk
Website: http://www.medicine.exeter.ac.uk/catchus/

New studies coming on line

The BPSU scientific committee has approved applications on severe enterovirus, accidental poisoning, listeria, bronchopulmonary dysplasia, and resuscitated term babies with no heart rate detected at 10 mins. Start dates subject to funding and ethics approval.

BPSU on the road

Since the last newsletter the BPSU continued on its travels. Apart from attending the RCPCH conference, the Unit also had a presence at the EURORDIS and the ESPID conferences, where we had a stand and several abstracts and posters were presented.

This December also sees the BPSU hosting a session on rare metabolic disease at the Excellence in Paediatrics conference in London. Speakers include Dr Rachel Knowles (metabolic screening), Prof Tim Cox (lysosomal storage disorders) and Dr Sue Laurent (family support for those with metabolic disorders).

BPSU was also represented at the International network of Paediatric Surveillance Unit (INoPSU) conference in Vancouver by Dr Clarisser Oeser who presented data on the congenital Zika syndrome study. The protocol has now been used by several of the INoPSU and will allow international comparison. Following the conference, the INoPSU business meeting outlined the future direction of the network. Our scientific coordinator attended by skype – at 2am in the morning! The Swiss Unit will be taking the lead as convenor whilst the BPSU will continue to maintain the website— http://www.inopsu.com

Publications

Over the past year the BPSU has been celebrating 30 years of surveillance. To mark this milestone we have published an anniversary report highlighting the work of the unit and its contribution to improving the health of children with rare disease. There is an introduction from HRH Princes Royal and a Foreword from Dame Sally Davies, Chief Medical Officer for England, as well as contributions from clinicians and patient groups. A copy of the report can be downloaded at http://www.rcpch.ac.uk/bpsu/30yearreportIf you would like a hardcopy of the report please contact the office.

The HIV, PIND, congenital syphilis and acute pancreatitis research teams have recently published results from their studies, details below:

- . Peters H, Byrne L, de Ruiter A, Francis K, Harding K, Taylor GP, Tookey PA, Townsend CL. Duration of ruptured membranes and mother-to-child HIV transmission: a prospective population-based surveillance study. BJOG 2016 May;123(6):975-81
- Majbar AA, Cusick E, Johnson P, Lynn RM, Hunt LP, Shield JP. Incidence and Clinical Associations of Childhood Acute Pancreatitis. Pediatrics. 2016 Sep;138(3). pii: e20161198.
- Stellitano LA, Winstone AM, van der Knaap MS, Verity C. Leukodystrophies and genetic leukoencephalopathies in childhood - a national epidemiological study. Dev Med Child Neurol. July 2016; Vol. 58 (7):680-9
- Simms I, Tookey PA, Goh BT, Lyall H, Evans B, Townsend CL, Fifer H, Ison C. The incidence of congenital syphilis in the United Kingdom: February 2010 to January 2015. BJOG. 2016 Mar 2. doi: 10.1111/1471-0528.13950

Reports and Analysis

Analysis: For the period December 2015 to May 2016 Orange Card return rates stand at 89.1% (Table 1). North Scotland are once again the top reporting region in UK - congratulations!!! Response rates in Ireland are lower than we would like. The BPSU Scientific Committee is also looking to appoint a Irish clinical representative to its Scientific Committee. Please contact the BPSU for further information.

As always, if you are experiencing any problems with the electronic orange card, do get in touch with the BPSU team.

Table 1 - % Regional Response Rates December 2015-May 2016

Region	% retd	rank
EAngl	92.4%	5
Mersey	88.9%	13
NET	85.8%	19
NScot	96.1%	1
NWest	86.4%	17
North	88.4%	14
NIre	86.0%	18
NWT	88.9%	11
Oxfrd	91.2%	7
RIre	81.8%	20
SET	87.2%	16
SScot	90.0%	9
SWest	91.9%	6
SWT	93.6%	2
Trent	88.9%	12
Wales	90.8%	8
Wessx	89.7%	10
WMids	87.8%	15
WScot	93.2%	3
Yorks	92.6%	4
Average	**89.1%**	

Table 2: All cases reported and follow ups to 21.09.2016

		VALID		IN-VALID				(as % of total)		
Condition	Start	C	R	D	E	X	TOTAL	C&R	D&E	X
HIV	1986	8,493	115	859	781	1,508	11,756	73	14	13
CR	1990	81	12	39	65	2	199	47	52	1
PIND	1997	1,699	0	521	1,298	65	3,583	47	51	2
EBT	2014	116	0	26	6	25	173	67	18	14
RKT	2015	74	0	5	69	90	238	31	31	38
T2D	2015	104	0	11	17	23	155	67	18	15
BEH	2015	33	0	13	10	43	99	33	23	43
ARF	2015	17	0	5	6	31	59	29	19	53
VIB	2015	78	0	1	46	135	260	30	18	52
FGM	2015	13	0	2	12	23	50	26	28	46
ADHD	2015	88	0	0	45	71	204	43	22	35
PRS	2016	41	12	7	5	130	195	27	6	67
ZIKA	2016	0	0	0	1	3	4	0	25	75
Total		**10,837**	**139**	**1,489**	**2,361**	**2,149**	**16,975**	**65**	**23**	**13**

HIV	Human immunodeficiency virus in childhood
CR	Congenital rubella
PIND	Progressive intellectual & neurological deterioration
EBT	Exchange blood transfusion
RKT	Nutritional Rickets
T2D	Type 2 Diabetes
BEH	Behcet's syndrome
ARF	Acute rheumatic fever
VIB	Visual impairment & blindness
FGM	Female genital mutilation
ADHD	ADHD transition
PRS	Pierre Robin sequence
ZIKA	Congenital Zika syndrome

C/R = confirmed/already known

D = duplicate

E = reporting error or revised diagnosis

X = status not yet reported to BPSU by investigator

ALL DATA IS PROVISIONAL & CONTINUALLY BEING UPDATED