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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

Editor
Richard Lynn
BPSU Scientific Coordinator

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

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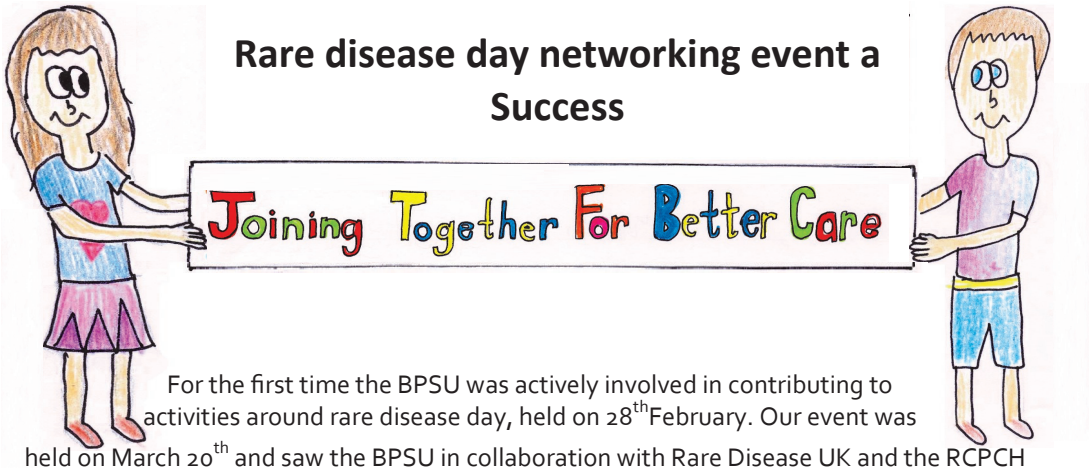


For the first time the BPSU was actively involved in contributing to activities around rare disease day, held on 28th February. Our event was held on March 20th and saw the BPSU in collaboration with Rare Disease UK and the RCPCH Youth Advisory Panel hold a rare disease tea party.

Under the theme of Joining together for better care the event, by invitation, allowed an opportunity for informal networking, bringing together patients and carers, healthcare professionals, industry, researchers and policy makers to discuss rare disease. Over 80 guests joined us at the RCPCH to hear presentations from the College President Dr Hilary Cass, Alastair Kent of RDUK, Dr Chris Verity and Miriam Evans, project coordinator of the Niemann-Pick Disease Support Group. And most importantly to view a moving podcast introduced d by Earl Howe and produced by the young people living with rare disease.

The podcast, photos and details of the event can be viewed online at www.rcpch.ac.uk/bpsu/rarediseaseday

We also asked guests to make a pledge in relation to rare disease to be carried out over the next year; and we will be checking – examples included strengthening patient and carer involvement in rare disease research. Given the positive response form the meeting the BPSU are looking to hold similar events in the future.

UK Rare Disease Strategy

The BPSU also had representation at the RDUK event at Westminster where Earl Howe introduced the launching of the implementation strategy. It was also announced that the deputy CMO for England John Watson would be responsible for implementing the UK rare disease strategy. The Rare disease Forum will be reconvened to see that the implementation strategy was being followed and this will be chaired by Alastair Kent.

Supported by Public Health England, Royal College of Paediatrics and Child Health, and University College London—Institute of Child Health

New Studies - Group B Streptococcal Disease

Prof Paul Heath

In April 2014, the BPSU will launch a new surveillance study to capture the incidence of invasive group B streptococcal disease in infants less than 90 days of age in the UK and Ireland. This will be for a period of 13 months. The study is led by Professor Paul Heath at St George's, University of London in collaboration with Public Health England, Public Health Scotland, Public Health Wales, Public Health Agency Northern Ireland and Health Protection Surveillance Republic of Ireland.

Group B Streptococcus (GBS) is the most common cause of serious bacterial infections in the first week of life and of meningitis in the first three months of life. Approximately 10% of babies with GBS disease will die and neurodisability occurs in up to 50% of survivors of GBS meningitis.

Early onset GBS disease (in first six days of life) may be prevented by antibiotics given intravenously to the mother during labour; national guidelines introduced in 2003 and updated in 2012 currently recommend this for women with certain risk factors. There are no prevention strategies targeted at late onset disease. A vaccine against GBS has been developed and is currently being tested.

The last national surveillance for GBS was 13 years ago (2000-2001). Comparison with this study will indicate whether current prevention strategies (including recently updated RCOG and NICE guidelines) have had an impact on incidence, morbidity and mortality. Knowledge of the current burden and serotype distribution of GBS isolates will also be important for the implementation of a GBS vaccine programme.

The case definition for this surveillance study is any case of invasive group B streptococcus (GBS grown from a sterile site) in an infant of less than 90 days of age with early onset disease being where the infant is less than seven days of age at the onset of infection and late onset disease being where the infant is between 7-89 days of age (inclusive). Clinicians and microbiologists are asked to report cases diagnosed between 1st April 2014 to 30th April 2015.

Clinicians in the UK and Republic of Ireland (ROI) will be asked to report cases via the BPSU orange card system and to complete case details using a secure web-based data capture system hosted by Public Health England. Clinicians in the ROI will also report cases to the Health Protection Surveillance Centre (HPSC) as invasive GBS is a notifiable disease in the ROI. Clinicians in Northern Ireland will also report cases to the Public Health Agency.

Microbiologists and laboratory staff will be requested to report invasive GBS cases through existing surveillance mechanisms and to submit isolates to their national reference laboratory.

Funding: This study is funded by a grant from Meningitis Now.

For further information about the study please contact: Dr Catherine O'Sullivan, Paediatric Infectious Diseases Research Group, St George's, University of London, Cranmer Terrace, London SW17 0RE **Email:** cosulliv@sgul.ac.uk

New Studies - Enterovirus and Parechovirus Meningitis

Enterovirus and Parechovirus meningitis in infants <90 days of age surveillance commences in May. This will be for a period of 13 months. The study is led by Dr Shamez Ladhani (inset) at St George's, University of London in collaboration with Public Health England, who writes.

"Approximately 85% of childhood meningitis in the conjugate vaccine era are due to enteroviruses and Human Parechoviruses (HPeV). Young infants are particularly susceptible to enterovirus and HPeV meningitis and often present with non-specific symptoms which are difficult to differentiate from serious bacterial infections. Real-time PCR is becoming increasingly available and it is anticipated that more cases will be diagnosed in the coming years.

Dr Shamez Ladhani

There is however very limited data assessing the incidence, clinical features, sequale and outcome of infants with meningitis as a result of these viruses. The study will aim to improve our understanding of enterovirus and parechovirus meningitis and also the outcomes. Furthermore, there are no data linking the molecular subtypes of enteroviruses and HPeV currently circulating in the UK and Ireland with clinical severity, laboratory markers or outcomes. No specific antiviral treatments are licenced or in the immediate pipeline to treat these important viruses. In part the lack of good clinical data defining the burden of disease is a barrier to the development of novel antiviral therapy."

Case definition: Any case where the clinician has made a clinical diagnosis of enterovirus or parechovirus meningitis in infants less than 90 days of age.

For further information about the study please contact: Dr Shamez Ladhani, Public Health England, 61 Colindale Avenue London NW9 5EQ **Email:** Shamez.Ladhani@phe.gov.uk. Web: www.rcpch.ac.uk/bpsu/neoentero

Study Extensions - Congenital Rubella

Extensions: Congenital rubella - Although there have been very few confirmed reports in recent years, active surveillance through the BPSU has been invaluable. The last significant rubella outbreak was in 1996, when there were 14 associated congenital rubella cases, 12 of which were notified through the BPSU within two months of birth. Since then there have been about 20 cases in total, with about two-thirds of the maternal infections being acquired in the UK. Although prospectively recognised maternal infections in pregnancy are reported through the PHE, maternal infections resulting in live births of congenitally infected infants are generally not recognised antenatally, and are only retrospectively diagnosed; comprehensive paediatric reporting is therefore essential.

Rubella infection continues to be rare in Britain, but we still have a cohort of young people who missed out on MMR vaccine in 1998-2007. In addition, several other European countries have low MMR uptake, resulting in periodic rubella epidemics (recently in Holland, Romania and Poland). A substantial rubella epidemic, with consequent congenital rubella cases, was also reported from Japan in 2012/2013. Women who travel abroad during early pregnancy may come into contact with the infection, and the potential for imported infection astill exists. Awareness of rubella infection and congenital rubella among paediatricians, and health professionals looking after pregnant women must be maintained.

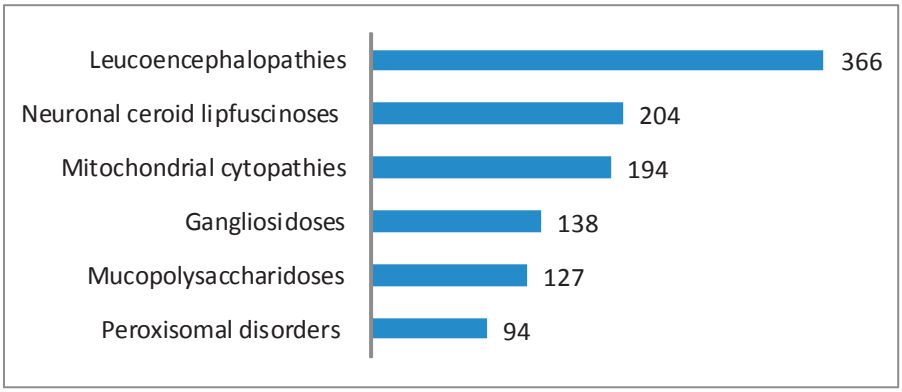
The National Screening Committee (NSC) is planning to stop the antenatal rubella screening programme, and alternative strategies to monitor on-going susceptibility in women of child-bearing age, and ensure timely immunisation of susceptible individuals, are currently under discussion. In the light of the change in national screening policy, and the WHO Europe goal to eliminate rubella and congenital rubella from the European Region by 2015, it has been agreed that monitoring of congenital rubella through the BPSU will continue, with support from the NSC.

For further information visit www.rcpch.ac.uk/bpsu/congenitalrubella

Study Extensions - PIND

The Progressive Intellectual and Neurological Deterioration (PIND) study has agreement from the BPSU to continue surveillance until end of April 2015 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 review 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* children being infected with vCJD *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 1 above 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.

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.

For further information visit www.rcpch.ac.uk/bpsu/PIND

Reports and Analysis

BPSU publications: Knowles RL, Khalid JM, Oerton JM, et al. Late clinical presentation of congenital adrenal hyperplasia in older children: findings from national paediatric surveillance. Arch Dis Child 2014;99:30–34.

Verity C, Stellitano L, Winstone AM, et al. Pandemic A/H1N1 2009 influenza vaccination, preceding infections and clinical findings in UK children with Guillain–Barré syndrome. Arch Dis Child Published Online First March 2014: doi:10.1136/archdischild-2013-304475

E-reporting: We now have over 2500 (75%) respondents reporting via our E-web service. Of these we receive 1500 (70%) within a week. To improve responsiveness, our E-web service is now SMARTPHONE responsive. So if you open the site using the phone it will be just as easy to reply. For those who wish to change to e-card reporting 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. We aim to go all electronic by 2016.

Hospital Analysis: We regularly report the response rate for the return of cards by region. In this way it helps us identifying potential areas of under-reporting. For the first time we have looked at the response rates for the main paediatric centres. Taking the average response for January to October at 93% five hospital had a 100% returns for this period Royal Free Hospital, Royal United Hospital- Bath; Singleton Hospital – Swansea; University Hospital Lewisham and the Royal Blackburn hospital. We found that 11 centres fell below the average. St George's Hospital Medical School (93%); Royal Aberdeen Children's Hospital(93%); Royal Manchester Children's Hospital (92%); Sheffield Children's Hospital (92%); St James's University Hospital – Leeds (91%); Royal Liverpool Children's Hospital(91%); Royal Brompton Hospital & NHLH (90%); Whittington Hospital (90%); Northwick Park Hospital (89%); Great Ormond Street Hospital (87%) and Chelsea & Westminster Hospital (86%). However this does not mean their case return rate was any less.

Table 1 - % Regional Response Rates March-Oct 2013

Region	Return %	Rank
Eangl	96.50%	4
Mersey	92.10%	18
NET	90.60%	20
NScot	96.60%	3
Nwest	94.5%	10
Northern	95.00%	9
Nire	93.60%	15
NWT	90.60%	19
Oxfrd	97.80%	1
Rire	94.00%	14
SET	93.50%	10
SScot	96.00%	5
Swest	94.90%	10
SWT	94.6%	12
Trent	95.90%	6
Wales	95.90%	7
Wessex	95.10%	8
WMids	94.60%	11
WScot	93.30%	17
Yorks	97.10%	2
Average	**94.30%**	

TABLE 2: All cases reported and follow ups to 08.03.2014

		VALID			INVALID		C&R	D&E	X
Condition	Start	C/R	D	E	X	TOTAL	(as % of total)		
AIDS/HIV	1986	7,807	809	731	1,131	10,478	75	15	11
CR	1990	89	37	62	3	191	47	52	2
PIND	1997	1,902	426	1,017	237	3,582	53	40	7
SYP	2010	50	25	15	16	106	47	38	15
VITD	2011	88	15	19	16	138	64	25	12
HUS	2011	146	114	66	111	437	33	41	25
GID	2011	20	1	76	41	138	0	185	30
PDA	2012	152	125	45	191	513	1	24	37
KAW	2013	151	25	61	199	436	2	31	46
APAN	2013	38	10	29	105	182	3	28	58
HEP	2014	0	0	0	0	28	0	0	100
Total		**10,443**	**1,587**	**2,121**	**2,050**	**16,229**	**64**	**23**	**13**

AIDS/HIV	Human immunodeficiency virus in childhood
CR	Congenital rubella
PIND	Progressive intellectual & neurological deterioration
HEP	Acute Symptomatic Hepatitis
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
KAW	Kawasaki Disease

C = confirmed/already known

D = duplicate

E = reporting error or revised diagnosis

X = status not yet reported to BPSU by investigator

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Public Health England

RCPCH Royal College of Paediatrics and Child Health

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