



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



Royal College of Paediatrics
and Child Health

Editor
Richard Lynn
Tel: 020 7323 7911
Fax: 020 7323 7901
Email: bpsu@rcpch.ac.uk
Website: <http://bpsu.inopsu.com>

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BPSU system to undergo evaluation – Please help

As the BPSU completes its 21st year of surveillance it is most appropriate to take stock of what has been achieved and consider our future direction. To help with this assessment the BPSU office has commenced an evaluation, an audit if you will, of its systems. An evaluation sub-group led Colin Mitche and Simon Mitchell has been set up to coordinate the activity, it will be using Center for Disease Control and Prevention criteria on evaluating surveillance systems. Areas to be evaluated include the BPSU operational model, its efficiency, flexibility and responsiveness; the BPSU's contribution to the advancement of knowledge and impact on public health policy; and to determine the BPSU's engagement with the public.

The first Phase, evaluation of the operational model, is currently underway. One major part of this phase is to seek opinions on the BPSU and its work from those who use the system (investigators) and those who respond (clinicians). To this end we are sending out a questionnaire to a randomly chosen number of clinicians and to all investigators who have used the system in the past 10 years. If you are a recipient of the questionnaire we would be most grateful if you could take time to complete and return the form.

Surveillance of Intussusception to commence in March

As you may recall in the last bulletin we advertised the start of a study into intussusception (IS) in children less than 12 months. However, this has had to be delayed until March. Dr Haitham El Bashir, whilst remaining on the project, will no longer be over-seeing the day to day management due to overseas commitments, instead, Professor Brent Taylor at ICH, UCL will now lead the project. Mr Sean Marven, Consultant paediatric surgeon (Sheffield Children's Hospital) and Dr J Claire Cameron, Epidemiologist, Health Protection Scotland are also co-investigators on the study.

Case definition: Any child less than 12 months of age, who has suspected or confirmed intussusception based on clinical, radiological and / or surgical findings.

This study, which has MREC approval through Wandsworth Research Ethics Committee (07/Q0803/62) and PIAG approval (PIAG/BPSU 2-05(FT1)/2007), is being run by the General and Adolescent Paediatric Unit, UCL Institute of Child Health, London, in collaboration with the BPSU and the British Association of Paediatric Surgeons (BAPS). Funding is via an educational grant from GlaxoSmithKline Biologicals.

For further information visit <http://bpsu.inopsu.com/> or contact Brent Taylor
E-mail: b.taylor@ich.ucl.ac.uk

Office Re-location

Along with the rest of the RCPCH the BPSU office is to re-locate at the end of March to 5-11 Theobalds Road, Holborn, London WC1X 8SH. Please continue to return the orange cards; we do have a redirect mailing system set up so they will reach us but it would be helpful if you could send any outstanding cards you may have back to us as quickly as possible.

Study Extensions

Two studies, PIND and FMAIT have had their period of surveillance extended for a further year.



Dr Marion Knight

The **Fetomaternal Alloimmune Thrombocytopenia (FMAIT)** study, which commenced in October 2006, aims to describe the current obstetric and paediatric management of FMAIT in the UK; describe outcomes and to use the information gained to inform ongoing review of the case for antenatal screening for this condition. Cases have been ascertained through obstetric, paediatric and National Blood Service reporting systems to assess the incidence of FMAIT in the UK. For the 6 month period to March 2007, 38 cases have been identified, giving an estimated incidence 1 case in 9600 births (95% Confidence interval 1/7000 to 1/13,500). The incidence of FMAIT as estimated by the study so far is less than one third of the incidence predicted from the published literature.

From the preliminary capture-recapture analysis used, there does not appear to be a significant under-reporting of cases. Thus this is likely to be an accurate estimate of the true incidence of clinically detected disease.

However, this low incidence, impacts on the researchers ability to answer their secondary research questions. They plan to describe the outcomes of affected infants and relate these to antenatal and postnatal management. However, with only one third of the predicted cases, the study currently has insufficient power to be able to demonstrate significantly even relatively large differences. In order to make this possible the BPSU Executive has agreed for a further year's surveillance, to September 2008 to be undertaken.

Contact: Dr Marian Knight, UKOSS Clinical Co-ordinator, National Perinatal Epidemiology Unit (NPEU), University of Oxford Old Road Campus, Oxford OX3 7LF. E-mail: marian.knight@perinatal-epidemiology.oxford.ac.uk

The study of **Progressive Intellectual and Neurological Deterioration (PIND)** has now completed its 10th year of surveillance. The study set up to identify paediatric vCJD cases and has to date been notified about 2345 cases of suspected PIND. 996 have a confirmed diagnosis including 116 different neurodegenerative disorders. 6 have been identified as vCJD with 4 cases being confirmed by autopsy. Principal investigator **Dr Chris Verity** told the Bulletin that reasons for seeking an extension were as follows;

1) *"We still have not observed any cases of vCJD with an MV or VV genotype at codon 129 of PRNP. Observational studies like the appendix study and the reported transfusion recipient case suggest that both these genotypes are susceptible to infection with vCJD/BSE and this is supported by studies in human transgenic mice. It is too early to be certain that we could not see cases with an extended incubation period and this could involve those aged less than 16 years at clinical onset"*

2) *"We still have no evidence of vertical transmission but it will be many years of observation before we can be confident that this cannot happen. There is an estimated population of around 4000 individuals infected with vCJD who presently are pre- or sub-clinical and it will be essential to continue to look for evidence of vertical transmission for some years. The fact that we have not identified any case of vCJD born after 1989 is potentially of great importance for public health as the SBO ban was introduced in late 1989 in the UK.*

3) *There is increasing concern about the possibility of secondary transmission via surgical and dental instruments*

4) *There are three known cases of vCJD in European countries born after 1990 and these countries did not introduce equivalent measures to protect public health until many years later than the UK. It is likely that significant dietary exposure of the UK population to BSE continued until 1996 and there is still the possibility of identifying children aged 11-16 years infected in the period 1990-1996. I personally think this is of some importance as the failure to identify cases born in this period through PIND would provide supportive evidence that the public health measures introduced were effective. This has implications for public health policy measures in the UK and other countries.*

Dr. John Stephenson, Chief Research Officer - New and Emerging Infections and Vaccines, at the Department of Health has stated that *"The latest advice we have from all the DH independent advisory committees is that surveillance is absolutely key to managing the vCJD epidemic"*. It is for all these reasons the BPSU has agreed to continue surveillance until 2010.

Contact: Dr. C. Verity, Consultant Paediatric Neurologist (Principal Investigator), Ms A.M.Winstone and Ms L. Stellitano Addenbrooke's Hospital, Cambridge, CB2 2QQ. Tel. 01223-216299. E-mail: annemarie.winstone@addenbrookes.nhs.uk



Dr Chris Verity

Study News



Dr Ariane Herrick

Scleroderma in childhood can cause major disability and (in the case of systemic sclerosis) mortality. At present, there is very little information available on the occurrence of childhood scleroderma. Our main aim was to assess the incidence of different forms of childhood scleroderma. Information was also obtained on the interval between symptom onset and diagnosis, and the pattern of care received by children before and after diagnosis.

The study recruited for 25 months from July 2005 until July 2007. To date, 170 cases have been notified, and 100 questionnaires have been returned. 53 questionnaires are currently outstanding. The remaining 17 cases were excluded for not meeting the study criteria or as a result of reporting errors. Of the 100 questionnaires returned, 78 valid cases were identified. As anticipated, most of the notifications for the study came via the BPSU (60%). Members of the UK Systemic Sclerosis Study Group, of the British Association of Dermatologists and the British Society for Paediatric and Adolescent Rheumatology were also mailed and 68 notifications (40%) came from these organisations.

Of the 78 valid cases, 73 (94 %) are of localised scleroderma and 5 (6 %) are of systemic sclerosis. 30 have been male and 48 have been female. The median age of valid cases so far has been 11 years (range four to sixteen years).

An important part of the study is the 12 month questionnaire, which asks for follow-up data and (if appropriate) whether the diagnosis has been confirmed by a rheumatologist or dermatologist. These questionnaires will continue to be collected until the end of July 2008. From the data obtained from the initial and 12 month questionnaires we shall obtain information not only on incidence, but also on the nature of presenting symptoms, the delay between symptom onset and diagnosis and the type of care received.

The low number of notifications has been disappointing, and suggests that childhood scleroderma, at least linear and systemic sclerosis, is even rarer than previously believed. If anyone reading this article is aware of a case presenting between July 2005 and July 2007 but not yet notified, please contact us as we are very keen to ensure that we receive as many notifications as possible. We are extremely grateful to all those who have already notified cases.

Contact: Dr Ariane Herrick, Senior Lecturer in Rheumatology, ARC Epidemiology Unit, University of Manchester. Tel: 0161 275 5993. E-mail: ariane.herrick@man.ac.uk

Genital Herpes in children under 11 years of age presenting to secondary care: Genital herpes in a young child presents a great clinical and child protection dilemma for paediatricians. How was it contracted? How likely is it to be the result of sexual contact? Can it be contracted in other ways? Do the implications depend on age, virus type, or child care contact such as nappy changing? We ask ourselves these questions, realising how little we know, being palpably aware that parents, social workers, police and others will shortly be asking us the same questions and expecting some type of authoritative answer.

The BPSU Genital Herpes study will not be able to provide the final definitive answers to these questions but will give us useful information about the clinical epidemiology of this condition, and a description of the actions taken in these cases and the results of any child protection investigations. This type of study is highly relevant at a time of intense public, judicial and professional scrutiny of the quality of medical evidence in the field of child protection.



Dr Richard Reading

The study commenced in April 2007 and to December 12 reports have so far been received. Of those that we have collected full information on, around half are confirmed cases. It was initially felt that the condition was likely to be very uncommon, even by the standards of the BPSU which "specialises" in rare conditions, and this seems to be confirmed. If anyone reading this feels they may have missed reporting a case which first presented to them after April 1st 2007 then please let us know – either by putting a tick on the next orange card you receive, or by contacting the BPSU directly. It is not too late. Information about the study has been circulated to paediatric dermatologists and genito-urinary physicians but you may be aware of cases seen by non-paediatric colleagues, in which case again let us know.

No formal analysis of the data so far but one clear message is the importance of seeking a confirmation of the diagnosis. The most useful test is viral culture and PCR testing if culture is negative. If this does not provide a diagnosis, paired herpes simplex antibodies can exclude primary genital herpes if there is no change in titres. Although the lesions of genital herpes are fairly characteristic, two of our case reports have been identified as having other conditions because their clinicians were thorough in pursuing investigations. The importance of this is that it removes any speculation about child abuse (quite apart from making our study more accurate!).

For further information contact Richard Reading: Consultant Paediatrician, Jenny Lind Department, Norfolk and Norwich University Hospital, E-mail: Richard.reading@nnuh.nhs.uk or visit BPSU website on <http://bpsu.inopsu.com>

Study News

National Study of HIV in Pregnancy and Childhood: Grateful thanks to everyone who reports cases to the National Study of HIV in Pregnancy and Childhood (NSHPC) through the BPSU. Can we just remind you that all infants born to HIV-infected women should be reported, regardless of their own infection status, as well as any infants and older children with confirmed HIV infection. Diagnosed pregnant women are reported through the parallel obstetric reporting system, but we rely on the paediatric reports to establish the infection status of the children born to infected women.

The NSHPC now has its own website (www.nshpc.ucl.ac.uk), with features including background to the surveillance programme, details of ethics approval, reporting instructions and pdf versions of the data collection forms. Quarterly updated results are available in slides and tables that can be downloaded and used in presentations. Publications and conference abstracts that make use of the surveillance data are also listed. Please visit the website! We would appreciate feedback, so please let us know what you think.

Contact: Dr Pat Tookey, Institute of Child Health, London WC1N 5EH. E-mail: p.tookey@ich.ucl.ac.uk

Recent Publications & Analysis

BPSU bulletin: This will now be produced 3 times a year in January, May and September. Circulation of hardcopies will also be reduced. Copies of the bulletin as all our publications will be placed on the website at http://bpsu.inopsu.com/publications/quarterly_bulletin.html. However, to keep all up to date with our activities we intend to use the 'President's email', circulated to all College members, more regularly with lead headlines being hyperlinked to articles on the BPSU website.

Recent paper: Cameron JC, Allan G, Johnston F, Finn A, Heath PT, Booy R. Severe complications of chickenpox in hospitalised children in the UK and Ireland. *Arch Dis Child.* 2007 Dec;92(12):1062-6. Epub 2007 Nov 8.

TABLE 2 - % RESPONSE RATE

Region	% rtd April 07 – Sept 07	Rank (Jan-June 07)
North	93.2	5 (8)
Yorks	93.1	6 (4)
Trent	92.9	7 (5)
EAngl	92.9	8 (2)
NWT	89.6	18 (18)
NET	87.3	19 (19)
SET	91.1	14 (13)
SWT	84.1	20 (15)
Wessex	94.2	3 (3)
Oxford	91.5	13(7)
SWest	91.8	12 (14)
WMids	92.6	9 (9)
Mersey	92.4	10 (16)
NWest	89.7	17 (11)
Wales	96.5	1 (1)
NScot	95.8	2 (10)
SScot	90.0	15 (20)
WScot	92.0	11 (12)
Nlre	93.6	4 (6)
Rlre	89.9	16 (17)
Total	91.4%	

TABLE 3 - ALL CASES REPORTED AND FOLLOW-UPS TO DECEMBER 2007

Condition	Started	VALID				Total	as % of total		
		C/R	D	E	X		C&R	D&E	X
HIV	1986	4994	595	623	373	6585	76	18	6
CR	1990	75	38	56	43	173	43	54	2
PIND	1997	1339	292	672	30	2333	57	41	1
MCADD	2004	171	47	31	78	327	52	24	24
MRSA	2005	88	18	26	32	164	54	27	20
Scleroderma	2005	48	4	22	29	103	47	25	28
VKDB	2006	4	2	6	6	18	22	44	33
FMAIT	2006	39	1	9	18	67	58	15	27
Genital Herpes	2007	3		4	52	12	25	33	42
IIH	2007	0	0	1	82	83	0	10	99
CAH	2007	0	0	0	55	55	0	0	100
Total		6761	997	1450	7127	9920	68	25	7

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

HIV Human immunodeficiency virus in childhood
 CR Congenital rubella
 PIND Progressive intellectual neurological degeneration
 MCADD Medium chain acyl CoA dehydrogenase deficiency
 MRSA Methicillin-resistant Staphylococcus aureus
 VKDB Vitamin K deficiency bleeding
 FMAIT Fetomaternal alloimmune thrombocytopenia
 IIH Idiopathic intracranial hypertension
 CAH Congenital adrenal hyperplasia

ALL DATA IS PROVISIONAL & CONTINUALLY BEING UPDATED

