



## Royal College of Paediatrics and Child Health

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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## Survey on severe complications of varicella to commence

Due to commence in November is a study on severe complications of varicella. Varicella is generally a mild disease in healthy children. Nevertheless, some severe complications may still occur, including secondary bacterial infections, central nervous system manifestations, and death. Groups at increased risk of complications include newborns with maternal rash onset temporarily close to birth and immunocompromised individuals of any age. There are few data on complicated varicella cases in the UK. Routine hospital discharge records and mortality data in the UK have been analysed previously, but cannot provide data with sufficient detail or accuracy.

The main objective of the study is to estimate the annual incidence of complicated varicella in hospitalised children, including deaths. It will also characterise these complications, describe the children affected, and estimate the annual financial costs to the health service.

Currently, there is no immunisation programme against varicella in the UK, or Ireland. The data provided will therefore help to determine its advisability and clarify high-risk groups. The data will also provide one baseline against which the impact of vaccination may be evaluated. The study will run for 13 months.

### Case definition

Any child less than 16 years hospitalised with complicated varicella, as defined by list of clinical conditions\*, or admitted to a paediatric ICU or HDU with varicella or one of its complications.

\*Varicella with: Bacteraemia / septic shock; toxic shock syndrome / toxin-mediated disease; necrotising fasciitis; encephalitis; purpura fulminans / disseminated coagulopathy; pneumonia (abnormal x-ray); neonatal varicella; fulminant varicella; Reye's syndrome; ataxia; admitted into ICU / HDU or death due to varicella.

### Virological testing

All reporting clinicians wishing virological testing for severe cases of varicella are encouraged to submit throat swabs and/or vesicle fluid samples for molecular analysis. Please contact Dr Judy Breuer, Consultant in Virology, St Barts and The London. Tel: 020 7 377 7141.

An information flyer will be included in the **October** mailing and the protocol card will be distributed in the **November** mailing.

For further information contact: Mrs Geraldine Allan or Dr Claire Bramley, Scottish Centre for Infection and Environmental Health, Clifton House, Clifton Place, Glasgow G3 7LN  
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Email: [gerry.allan@scieh.csa.scot.nhs.uk](mailto:gerry.allan@scieh.csa.scot.nhs.uk); [claire.bramley@scieh.csa.scot.nhs.uk](mailto:claire.bramley@scieh.csa.scot.nhs.uk)

## BPSU 16th Annual Report Published

The BPSU 16th Annual Report 2001-2002 has recently been published. College members will receive their copies with the College Newsletter that should be reaching you shortly. A limited number of additional copies are available from the BPSU office, alternatively the report can be viewed on the College's website at <<http://www.rcpch.ac.uk/publications/bpsu.html>>. We hope you will find this an interesting read and worthy of storage on your overcrowded bookshelves. Alternatively do feel free to circulate it within the department or pass it on to the hospital library.

The report contains feedback on the nine current projects underway, includes a detailed international section and contact information for parent organisation as well as the yearly Unit analysis. The willingness of paediatricians to continue to contribute to the system is reflected in an average monthly response rate of 93% which has led to over 1300 case reports, one of the highest ever for a single year. Even so, case ascertainment is an area the BPSU is acutely concerned with and we encourage all to report cases even if they are not sure they fit the case definition or even if they feel a colleague may have already done so. This, with the increased use of alternate sources of ascertainment, will improve still further the number of cases reported. On behalf of the Unit and the investigators we thank you for this magnificent response.

## PIND survey update

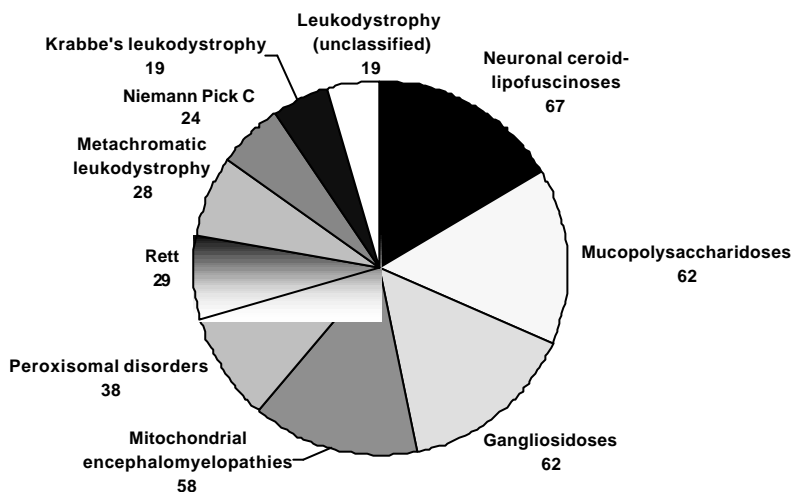
Surveillance for progressive intellectual and neurological deterioration (PIND) in UK children has been in progress for over five years now, having commenced in May 1997. The study has recently been granted an extension by the Department of Health and is now expected to continue until April 2004.

By 9 August 2002, **1369** children had been reported to the study at a fairly steady rate of around 15 per month. There still appears to be a lot of interest in the study and we remain very grateful to UK paediatricians for their continued support in reporting cases and supplying information for both new and follow-up cases.

The PIND Expert Group of six paediatric neurologists meets quarterly in London and has now discussed **1000** anonymised clinical summaries. Of these, **576** have a confirmed PIND diagnosis (99 specific neurodegenerative conditions have been identified). **6** have vCJD (see later).

### **Figure 1: The ten most commonly occurring PIND diagnoses**

It is hoped that ultimately the analysis of PIND data will prove helpful and informative to paediatricians in terms of diagnosis, protocol and service provision. Preliminary analysis has been carried out as follows:



- 638 children with PIND were analysed to describe variations according to district of residence, age at presentation, sex, ethnicity and consanguinity. A wide variation in neurodegenerative disease across the UK is apparent. Numbers reported by district of residence are very variable and ethnicity and consanguinity are contributory factors.
- It has been suggested that Alpers' disease is an important differential diagnosis of variant Creutzfeldt-Jacob disease (vCJD) because spongy brain degeneration is found in both. Nine children with a provisional diagnosis of Alpers' syndrome were reviewed. Despite initial fears, we have found no evidence that Alpers' syndrome is likely to be confused with vCJD.
- The 10 children with infantile neuronal ceroid-lipofuscinosis reported to the study in the first four years were described in terms of clinical presentation and diagnosis. Despite genetic advances in the diagnosis of INCL, early clinical recognition is paramount. Presenting features are more varied than the literature suggests.
- The group of 51 children with "idiopathic" PIND who form a clinically heterogeneous group and have been thoroughly investigated or have died without reaching a diagnosis, have been further analysed by the Expert Group. On close examination, none of this group has evidence of vCJD or a disease resembling vCJD.

#### **In conclusion:**

- *The incidence of vCJD has increased by about 20% annually for both onsets and deaths and there is still uncertainty about the incubation period - continuing surveillance is therefore very important because more childhood cases may appear.*
- *It is hoped that the large amount of data which has been gathered on the variety of conditions causing PIND will ultimately be of interest and help to UK paediatricians in terms of practice, diagnosis and service provision.*
- ***Please continue telling us about all your diagnosed and undiagnosed cases of PIND - even if you are not sure that they strictly fulfil the criteria.***

First contact: Ms G. Devereux, Research Nurse; Mrs L. Stellitano, Research Administrator, c/o Paediatric Administration Office, Box 45, Addenbrooke's NHS Trust, Hills Road, Cambridge, CB2 2QQ Tel: 01223-216299 Fax: 01223-586508 Email: gilliandev@yahoo.com

## Study Extensions

### The national register of hepatitis C – Call for study proposals

**The national register of hepatitis C with a known acquisition date:** In 1998, a national register of hepatitis C (HCV) infections with a known date of acquisition was established.<sup>1</sup> The register was set-up to help inform the natural history of HCV-related disease in the UK,<sup>2</sup> and now contains anonymous data for one of the largest cohorts of individuals with known-date HCV infections. The majority of infections in the register are those that were acquired following transfusion of HCV-infected blood that was issued before the introduction of routine screening of the blood supply for HCV.<sup>3</sup>

In order to get maximum benefit from this national resource, the register steering group would like to invite clinical and epidemiological researchers to submit proposals to access data held in the register. It is envisaged that a variety of studies might benefit from linkage with or access to the register, and proposals from all specialties and institutions are welcomed. Such studies are urgently needed to help determine the current and future burden of HCV-related disease on health care services, and to assess the impact of currently available treatments as well as those that may become available in the future.

**Any researchers interested in applying for access to information held within the national register should contact the register co-ordinator (details below) for a list of available data, and an application form. No data will be released that could identify individual patients directly or via linkage to other data. Any study proposals should then be submitted to the register co-ordinator for consideration by the steering group by Monday, 16<sup>th</sup> December 2002 (deadline).**

1. Harris HE, Ramsay ME, Heptonstall J, Soldan K, Eldridge KP. The HCV National Register: towards informing the natural history of hepatitis C infection in the UK. *Journal of Viral Hepatitis* 2000;7:420-7.
2. Harris HE, Ramsay ME, Andrews N, Eldridge KP. Clinical course of hepatitis C virus during the first decade of infection: cohort study. *British Medical Journal* 2002;324:450-3
3. Chief Medical Officer. Hepatitis C and blood transfusion lookback. London: HMSO, 1995 [PL CMO (95) 1]

*Dr Helen Harris (Register Co-ordinator) or Ms Lisa Beck (Research Assistant): Immunisation Division, Communicable Disease Surveillance Centre, Public Health Laboratory Service, 61 Colindale Avenue, London, NW9 6EQ. Tel: 020 8200 6868 ext. 4676 (Mon-Wed) or ext. 4496 (Mon-Fri); Fax: 020 8200 7868; Email: [hharris@phls.nhs.uk](mailto:hharris@phls.nhs.uk); [lbeck@phls.nhs.uk](mailto:lbeck@phls.nhs.uk).*

### MECP2 mutation aids detection of Rett syndrome

Dr Alison Kerr updates us on recent developments: “The British Isles Survey for Rett Syndrome commenced in 1990 when the diagnosis relied entirely on the characteristic clinical signs. To date 1093 cases have been reported. Since 2000 it has been possible to confirm the diagnosis in over 80% by detecting mutations on *MECP2* (Xq28). It is clear that the range of severity in the disorder is wider than the ‘classic’ phenotype and that males as well as females may be affected. A few cases have been reported to present a different phenotype. The prevalence of disorder due to *MECP2* mutations is therefore expected to be considerably higher than the present UK estimate of 1 in 10,000 females and the male prevalence is still unknown. Recurrence is generally rare but there is a high risk to a minority of families so that mutation testing is recommended in all cases. Physicians are requested to continue reporting to the survey all previously unreported cases with clinical Rett, with or without genetic confirmation and all cases with pathological *MECP2* mutations, regardless of clinical presentation in order to achieve more accurate estimates of prevalence and insight into the range of associated *MECP* presentations.

The main Rett characteristics are non-dysmorphic appearance, initial developmental progress (frequently poor), a late infancy period of regression in hand use, speech and contact and thereafter severe to profound learning disability but with preserved personal contact, poor cardio-respiratory regulation, hypotonia evolving into dystonia/ hypertonia, involuntary movements including marked hand stereotypy and dyspraxia. Epilepsy is common. Head growth is usually suboptimal from within the first year.”

Ref: Kerr et al (2001). Guidelines for reporting clinical features in cases with *MECP2* mutations. *Brain and Development* 23:208-211.

*Dr Alison M Kerr FRCP FRCP&CH, Senior lecturer, honorary consultant in paediatrics and learning disability, Academic Centre, Glasgow University, Department of Psychological Medicine, Gartnavel Royal Hospital, 1055 Great, Western Road, Glasgow G12 0XH, Tel/ans: 0141 211 0281, Fax: 357 4899, Email: [amk5m@clinmed.gla.ac.uk](mailto:amk5m@clinmed.gla.ac.uk)*

## News in brief

**IMPORTANT BPSU Database news:** With the help from programmers the BPSU office is in the process of redeveloping its epidemiology software. Testing of the software begins this autumn and if successful it should go live next spring. This will be a crucial period for the BPSU office as the methodology is very much reliant on the epidemiology software. Consistent and accurate data is obviously of importance and to this end it would be helpful if you could check the hospital mailing address your card is being directed to, letting us have any amendments, of particular importance is the hospital name, road and postcode. These will be standardised so analysis of local as well as regional returns will be possible in the future. Please note because the cards are printed in advance any amendments may not appear on your card immediately.

**BPSU on the web:** Following recent contact with the National Electronic Library for Child Health (NELCH), the BPSU is to have a small but what we hope will be a useful link on the NELCH site at <<http://www.nelh.nhs.uk>>. The BPSU now also has a prominent link on the re-designed Contact a Family website <[www.cafamily.org.uk](http://www.cafamily.org.uk)>.

**HIV/AIDS information on the web:** A regularly updated Powerpoint presentation of the obstetric and paediatric surveillance data is now available. It includes some information about how the surveillance is carried out, and how the data are used, a basic set of tables, for example the unlinked anonymous surveys and DH targets. Email copies are available from the HIV/AIDS administrative ~~assistant, Ms Tina Stokes, at [Tina.Stokes@ich.ucl.ac.uk](mailto:Tina.Stokes@ich.ucl.ac.uk)~~. The PHLS' HIV & AIDS Report Section & Unlinked Anonymous Prevalence Monitoring Programme is available at <<http://www.phls.co.uk/facts/HIV/hiv.htm>>. Guidelines for the Management of HIV infection in pregnant women and the prevention of mother to child transmission, 2001 is accessible from <<http://www.aidsmap.com/about/bhiva/index.asp>>.

## Monthly Analysis

**TABLE 1 –  
% RESPONSE RATE  
Jan-June 2002**

Region	% retd	Rank (Dec-May 2002)
North	88.5	14(13)
Yorks	91.8	9 (7)
Trent	88.3	16 (18)
EAngl	94.8	1 (6)
NWT	85.1	19 (19)
NET	80.0	20 (20)
SET	87.5	18 (15)
SWT	88.0	17 (14)
Wessex	91.1	12 (8)
Oxford	94.6	2 (2)
SWest	91.4	10 (11)
WMids	88.3	15 (5)
Mersey	92.3	7 (4)
NWest	92.4	6 (16)
Welsh	93.7	5 (10)
NScot	94.4	3 (3)
SScot	88.6	13 (1)
WScot	93.8	4 (9)
Nlre	92.1	8 (12)
Rlre	91.2	11 (5)
<b>Total</b>	<b>89.5</b>	

**TABLE 2 - ALL CASES REPORTED AND FOLLOW-UP at 25/5/02**

Condition	Started	I		II		NYK	Ttl	as % of total		
		VALID	INVALID	Ia	Ib			I	II	III
HIV/AIDS	1986	1835	323	413	228	2799	66	26	8	
CR	1990	66	24	43	6	139	47	48	4	
PIND	1997	806	142	336	90	1374	59	35	7	
CVD/S	2001	188	12	46	68	314	60	18	22	
VKDB	2001	4	2	11	17	34	12	38	50	
Thrombosis	2001	102	17	42	50	211	48	28	24	
CMV	2001	103	17	35	50	205	50	25	24	
IAI	2001	17	17	19	9	61	28	57	15	
SFADR	2002	0	0	0	0	3	0	0	0	
Con Toxo	2002	0	0	0	0	3	0	0	0	
<b>Total*</b>		<b>4021</b>	<b>553</b>	<b>945</b>	<b>525</b>	<b>5144</b>	<b>61</b>	<b>29</b>	<b>10</b>	

\* All data is provisional & continually being updated

### Key to table / abbreviations

I	= confirmed/already known	Ia	= duplicate
Iib	= reporting error or revised diagnosis	III	= status not yet reported to BPSU by investigator
AIDS/HIV	Acquired Immunodeficiency Syndrome/ Human Immunodeficiency Virus	GBS	Group B streptococcus disease
CR	Congenital Rubella	CV/S	Cerebrovascular disease/stroke & like illness
PIND	Progressive Intellectual Neurological Degeneration	CMV	Congenital Cytomegalovirus
VKDB	Vitamin k Deficiency Bleeding	IAI	Internal abdominal injuries due to child abuse in children under 14 yrs
Enceph	Encephalitis in children (2-36months)		