



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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End Stage Renal Disease Study Commences Soon

We are pleased to announce that a study on End Stage Renal Disease (ESRD) will commence in early summer 2011. The study is to be undertaken by Dr Karl McKeever (RHSC Belfast).

End Stage Renal Disease (ESRD) in early infancy is rare with an incidence quoted of 0.31 per million in the UK population per year. There are reports of poor outcomes and significant morbidity in this group and there is controversy as to how best care for these children. The condition also presents complex clinical and ethical problems.

It is now possible to support infants with ESRD through Renal Replacement Therapy (RRT). However, this population differs from older children receiving dialysis in terms of their primary renal diagnosis, and are more likely to be diagnosed with renal dysplasia or obstructive uropathy. They may also have more co-morbidities which impact upon long-term outcome.

There is anecdotal evidence in the UK of increasing numbers of young infants being treated with dialysis but at present there is no data available on the outcome of ESRD during the first 6 months of life. Therefore, national ascertainment is required to establish a sufficient cohort to provide accurate data on this rare, but important, condition.

Karl McKeever: "The provision of long term dialysis for neonates with ESRD presents major public health issues with complex clinical, ethical and health-care resource issues. Improvements in neonatal survival and advances in RRT have resulted in higher numbers of infants presenting with ESRD for whom a decision has to be made to dialyse or treat palliatively, yet the experience of the infant population has not been reported."

Case definition: Any infant from age 4 weeks to 6 months with presumed ESRD (CKD5) who has a serum Creatinine of equal to, or greater than 120 micromols/l.

ESRD (CKD5) is defined as either a Glomerular Filtration rate (GFR) of less than 15 mL/min/1.73 m², which is accompanied in most cases by signs and symptoms of uraemia, or a need for initiation of kidney replacement therapy (dialysis or transplantation) for treatment for complications of decreased GFR, which would otherwise result in an increased risk of mortality and morbidity.

A serum creatinine of equal to, or greater than 120 micromols/l will give an estimated GFR of less than 15 mL/min/1.73 m² for infants in this population.

Funding and ethics: Northern Ireland Childrens' Renal Charity and has ethics approval from Belfast REC (Ref 10/NIR03/32) and NIGB

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E-card reporting Trial Commences

In January 2011 the BPSU commenced a 9 month trial of using an electronic orange card. During the trial we hope to establish whether the high response rate, which is vital to the robustness of the BPSU, can be maintained through moving to an email-based system.

A stratified random sample of 500 clinicians, 15% of all those receiving the card, has been selected. This means that from January until September 2011 these clinicians will receive an electronic orange card via email and will no longer be sent the paper version. We intend to contact this group for feedback on the system at the end of the trial and will feedback our findings more widely by the end of the year.

BPSU Contribute to Rare Disease Day

The 4th Rare Disease Day took place on the 28th February. Rare Disease Day began as a European initiative and is now marked across the globe as an annual, awareness-raising event coordinated by EURORDIS at the international level and by the National Alliances of Patient Organisations at the national level. Hundreds of patient organisations from more than 40 countries worldwide organised awareness-raising activities and converging around this year's theme of "Rare but Equal".



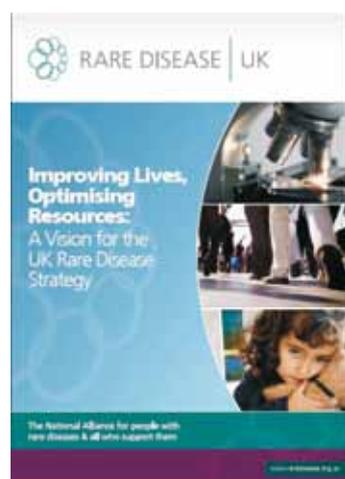
Rare Disease day aims to draw attention to:

- Gaps in health that exist for rare disease patients between and within countries in the EU.
- Gaps in health that exist for rare disease patients compared to other sectors of society.

The campaign will serve to advocate for:

- Equal access for rare disease patients to health care and social services.
- Equal access to basic social rights: health, education, employment, housing.
- Equal access to orphan drugs and treatments.

In the UK Rare Disease UK (RDUK) www.raredisease.org.uk/index.html has over 650 registered members including academics, clinicians, industry, individual members and patient organisations coordinated activities. To raise awareness of the importance of rare diseases lobby meetings were held at Westminster, the Scottish Parliament and the Welsh and Northern Ireland Assemblies'. The BPSU Chair and scientific coordinator were pleased to be able to attend the events at The Welsh Assembly and Westminster.



To coincide with Rare Disease Day, RDUK released a new report outlining recommendations for a strategy for rare diseases – **'Improving Lives, Optimising Resources: A Vision for the UK Rare Disease Strategy'**. The report is a product of a year and a half of work in collaboration with members and the broad rare disease community. The BPSU contributed, through the involvement of Richard Lynn, scientific coordinator, in addition to contributions from BPSU study investigators. This report was presented to Earl Howe, who is part of the Department of Health Ministerial team at a reception at the House of Commons.

The BPSU is now pleased to announce that it has linked with RDUK and we hope to work closely with them to promote each others activities. To this end Alistair Kent, Chair of RDUK will be giving a keynote speech at the BPSU symposium during the RCPCH annual meeting on 5th April.

In Australia, The Australian Paediatric Surveillance Unit (APSU), a BPSU counterpart and fellow INoPSU member, has been deeply involved in Rare Disease Day and has undertaken a number of associated activities. This has included running a brief survey via their website www.apsu.org.au to enable the community to express their support for a national plan for rare diseases in Australia. A national conference "Awakening Australia to rare Diseases" is to be held in April 2011 in Fremantle Western Australia www.rarediseases.org.au. The APSU has also partnered with the Steve Waugh Foundation, The Smile Foundation, Association of Genetic Supports Australasia and the Sydney Children's Hospitals Network (Randwick and Westmead) to plan a research study on the impacts of rare diseases on families, on health professionals and on health services.

Child and Adolescent Psychiatry Surveillance System

The Child and Adolescent Psychiatry Surveillance System (CAPSS), is a newly established surveillance unit run in conjunction with the Royal College of Psychiatry. It facilitates surveillance of psychiatric conditions, many which have paediatric elements such as early onset eating disorders and conversion disorders.

The CAPSS committee is currently looking for an additional paediatric member for its executive. If you are interested in joining this newly established unit please contact Alan Quirk (aquirk@cru.rcpsych.ac.uk) CAPSS coordinator, Dasha Nicholls (d.nicholl@ich.ucl.ac.uk) CAPSS chair or Richard Reading (richard.reading@nnuh.nhs.uk) a paediatrician who sits on the CAPSS committee. An interest in child mental health and / or epidemiology with some experience in study protocol review is desirable. Closing date April 21st.

CAPSS is also looking for investigators to undertake potential studies on conditions including early onset OCD and auto-immune disorders and severe psychopathology. If you would like to learn more about the unit please visit www.rcpsych.ac.uk/quality/research/capss.aspx.

Sir Peter Tizard Research Bursary – Application Call

The RCPCH is inviting applications for the 2011-12 Sir Peter Tizard Research Bursary from paediatricians wishing to undertake an epidemiological surveillance study through the BPSU. The successful applicant will receive up to £15,000 towards the costs of a surveillance study.

The principle purpose of the bursary award is to encourage paediatricians to develop skills and experience in epidemiological research by undertaking a study of a rare disease or condition. Studies conducted should add to the body of knowledge of rare childhood diseases and conditions. They should also support the RCPCH's objective of building and strengthening research and research capacity in paediatrics.

Applicants must be members of the RCPCH, with an NHS contract working at ST6 level or above, SASGs, or a consultant for less than five years. Applicants cannot have previously undertaken a BPSU study.

Studies considered will be required to meet the criteria for a surveillance study through the BPSU. Visit www.bpsu.inopsu.com for further details.

Applications will be judged on: the scientific quality of the application, justification for why the study should be carried out through BPSU, the quality of the candidate and the likely benefits to the candidate in terms of developing their research knowledge and skills. The clinical and public health importance of the research question posed is an important but not sole criterion.

For further details and an application form visit the BPSU website http://bpsu.inopsu.com/home/tizard_bursary.html. The closing date for applications is **Friday June 3rd 2011**

Study News

Congenital rubella: The BPSU Executive has approved an extension of the congenital rubella study until April 2014. Dr Pat Tookey (inset) reports on the reasons for the continued need for surveillance

“Although there have been few confirmed reports of congenital rubella in recent years, active surveillance through the BPSU remains invaluable. The last significant upturn in cases, in 1996, was rapidly recognised when most of the 14 reported cases were notified through the BPSU within two months of birth.

Since 2000 there have been 18 confirmed CR births in the UK and Ireland, of which 15 were reported on the orange card. Very few maternal infections resulting in live births of congenitally infected infants are recognised antenatally, so paediatric identification and reporting of cases is absolutely essential.

While rubella infection is currently rare in Britain, 15 years of sub-optimal MMR vaccine uptake leaves us vulnerable to renewed circulation of infection. Furthermore the first cohorts of children who experienced lower vaccine uptake are now entering early adulthood.

Measles and mumps control has proved challenging, localised outbreaks have occurred, and the annual number of confirmed cases continues to increase. Several other European countries also have low MMR uptake, resulting in periodic rubella epidemics, and in recent years clusters of congenital rubella cases have been reported from several European countries, including Italy, Romania, and Holland.

Areas of low MMR uptake in Britain are often the same areas which have substantial numbers of first generation immigrants. The combination of higher rubella susceptibility in immigrant groups and higher numbers of unvaccinated children provides the opportunity for the introduction and transmission of rubella, putting susceptible pregnant women at risk. Rubella susceptibility rates are higher among ethnic minority pregnant women, especially those who were born abroad; a disproportionate number of infected infants have mothers who were born abroad and could not take advantage of the UK's childhood immunisation programme. Susceptible women who travel abroad during early pregnancy may also come into contact with infection. Awareness of rubella infection and congenital rubella among paediatricians, and health professionals looking after pregnant women, must be maintained.

The European Region of the WHO established a goal of elimination of measles and rubella, and the prevention of congenital rubella infection (less than 1 case of congenital rubella per 100,000 live births) in Europe by 2010; the UK's surveillance for rubella, and particularly the active surveillance of congenital rubella, is regarded as a model system. We continue to be most grateful to all those who have reported a case and completed the proformas.”

Further information: Tel: 020-7905 2604 E-mail: p.tookey@ich.ucl.ac.uk



Upcoming Events

BPSU website: In April the BPSU plan to launch its new website when it migrates onto the new RCPCH site. We welcome comments on design and content. Our address remains the same at www.bpsu.inopsu.com

Study delays: The surveillance of acute pancreatitis to be led by Professor Julian Hamilton–Shield, Professor Paul Johnson and Richard Lynn has been delayed due to issues surrounding funding. The investigators hope this study will commence before the end of the year subject to funds being secured.

Published papers: The Congenital cytomegalovirus paper “Surveillance of congenital cytomegalovirus in the UK and Ireland.” Claire L Townsend, Catherine S Peckham, Pat A Tookey ADC-FNN 2011 has recently been published on-line and can be viewed at <http://fn.bmj.com/content/early/2011/02/01/adc.2010.199901.full.pdf>

International Network of Paediatric Surveillance Units (INoPSU) Conference: Following the success of the INOPSU Dublin meeting a 7th INOPSU meeting will be held in Montreux, Switzerland on 2nd September 2011. The meeting is to be part of the Swiss Paediatric Society Scientific Conference. For further details contact bpsu@rcpch.ac.uk

Analysis

**TABLE 1 - % RESPONSE RATE
(for 6 months-
May – October 2010)**

Region	% rtnd	Rank
North	91.4%	15
Yorks	92.6%	11
Trent	92.1%	12
EAnGl	95.4%	5
NWT	90.6%	16
NET	86.1%	20
SET	90.1%	17
SWT	91.6%	14
Wessx	95.7%	3
Oxfrd	95.1%	6
SWest	92.9%	8
WMids	92.6%	9
Mersy	91.9%	13
NWest	92.6%	10
Wales	97.5%	1
NScot	96.6%	2
SScot	94.3%	7
WScot	88.5%	19
Nlre	95.6%	4
Rlre	89.0%	18
Average	92.6%	

TABLE 2: Cases Followed up to 25.1.2011

Condition	Started	VALID			INVALID		C&R	D&E	X
		C/R	D	E	X	Total			
HIV/AIDS	1986	6,654	751	703	542	8650	77	17	6
CR	1990	84	34	28	5	151	56	41	3
PIND	1997	1,736	365	791	96	2,988	58	39	3
CD	2008	125	12	31	86	254	49	17	34
SUPC	2008	39	7	25	20	91	43	35	22
SNH	2009	61	1	28	5	95	64	31	5
GBS	2009	53	13	6	65	137	39	14	47
CNS	2009	90	14	6	65	175	51	11	37
SYP	2010	24	5	6	3	38	63	29	8
GSCT	2010	6	1	0	7	14	43	7	50
Lead	2010	5	2	4	10	21	24	29	48
Chylo	2010	44	3	5	48	100	44	8	48
GA1	2010	1	1	1	13	16	6	13	81
BacMen	2010	98	9	15	84	206	48	12	41
Total		9020	1,218	1,649	1049	12936	70	22	8

HIV.....Human immunodeficiency virus in childhood
 CR.....Congenital rubella
 PIND.....Progressive intellectual neurological degeneration
 CD.....Conversion Disorder -
 Excludes reports from psychiatrist
 SUPC.....Sudden unexpected early postnatal collapse
 CNS.....CNS Inflammatory Demyelinating Disease
 SNH.....Severe Neonatal Hyponatraemia
 GBS.....Guillain-Barré syndrome / Fisher syndrome
 SYP.....Congenital syphilis
 GSCT.....Gonorrhoea, Syphilis, Chlamydia, and
 Trichomonas infections
 Lead.....Raised Blood Lead Levels in Children
 Chylo.....Chylothorax in Infants and Children
 GA1.....Glutaric Aciduria 1 Paediatric Surveillance Study
 BACMen ..Bacterial meningitis in babies <90 days of age

C 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