

British Paediatric
Surveillance Unit

Annual Report 2013-2014



Aims of the British Paediatric Surveillance Unit

To:

- Facilitate research into uncommon childhood infections and disorders for the advancement of knowledge and to effect practical improvement in prevention, treatment and service planning
- Allow paediatricians to participate in surveillance of uncommon disorders and to lessen the burden on reporting doctors of such requests arising from numerous different sources
- Increase awareness within the medical profession of the less common disorders studied and respond rapidly to public health emergencies.

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Chair, BPSU

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Foreword

I became chair of the Scientific Committee of the BPSU in autumn of last year. It is a huge privilege and a daunting responsibility. Alan Emond led the BPSU through a testing time and left it a much stronger, more robust organisation when I took over.

There is a new governance structure for the BPSU, with an overseeing governance committee chaired by John Newton of Public Health England, with representation of the three parent bodies: Public Health England, RCPCH and the UCL-Institute of Child Health. They are responsible for the strategic direction of the BPSU and ensuring the financial viability of the organisation. Our funding is still tight, in common with many other public bodies during these times, but a lot more secure in the longer term.



Dr Richard Reading
Chair, BPSU

The work the BPSU is set up to do continues apace. Inside you will read reports of current studies, but there are several more upcoming studies of public health and clinical importance in preparation which we are excited about. There have also been publications of a number of completed studies and we continue to encourage investigators to disseminate their findings in high quality journals, conferences and in ways accessible to the public.

One of these routes is through activities associated with the Rare Disease movement. Rare diseases are becoming important professionally, politically and commercially. The UK advocacy group is Rare Disease UK, and Richard Lynn, our scientific coordinator, who sits on their management board, has been very active in promoting the BPSU activities to this group. Richard organised a very successful Rare Disease Day "tea party" earlier this year hosted at the college and attended by children, families, advocacy groups, professionals and academics, and industry. We were delighted that Health Minister Earl Howe was able to attend and participate in the afternoon. A repeat party is planned for next March.

Most of you will have changed over to e-card reporting, and we are working to ensure as many as possible are able to complete orange card returns electronically. While some will regret the passing of the iconic orange post-card, e-card reporting has been associated with our highest ever response rate at 95%. Thanks to everyone for this. Our investigators depend on the "Nothing to Report" responses for reliable incidence measurement. We are a little concerned that questionnaire returns on reported cases are not so reliably completed. We try to ensure the information requested is reasonable and easily accessible in the notes. We are also piloting electronic data collection in some studies to see whether this helps, but please do return questionnaires as soon as possible on any cases you report.

We encourage potential investigators to submit applications for new studies all the time. The BPSU is an internationally recognised surveillance system which continues to be spectacularly successful. There are changes in the research governance climate in the UK which may make some aspects of BPSU studies easier. For instance there is the opportunity for longer follow-up periods, the possibility of record linkage with appropriate safeguards, and the possibility of contributing to existing disease registers. Please contact Richard Lynn for more information. We are also working more closely with CAPSS (Child and Adolescent Psychiatry Surveillance System) on joint studies and protocols. We have been able to re-instate the Sir Peter Tizard Bursary for up-coming investigators. This year the bursary winner is Marie Wright from Edinburgh who will undertake surveillance on Pierre Robin Sequence.

I would like to record comings and goings on the scientific committee. Over the last year or so Mrs Ann Seymour, Mrs Sue Banton, Professor Simon Mitchell, Dr Colin Michie, Dr Piers Daubeney, Dr Delane Shingadia, Professor Carol Dezateux, Dr Sam Oddie, Dr Richard Pebody have all stepped down. Many thanks to them all, some whom had served the BPSU over many years.

Finally, June 2015 marks the start of the thirtieth year of the BPSU. I hope the summary above reflects a vibrant and forward-looking organisation. While we will be celebrating the achievements over the past thirty years, our emphasis will be on the future. New technology, developments in public health, changes in public understanding and involvement in research all offer great opportunities. Emerging public health and clinical problems will continue to pose challenges the BPSU is ideally suited to responding to. With your support, the BPSU will thrive over the next thirty years as it has in the past thirty. Thanks to you all.

A handwritten signature in dark ink, appearing to read 'Richard Reading'.

Richard Reading. Chair BPSU,
September 2014

1 How the Surveillance System Works

Background

Rare diseases and infections are a numerically important cause of morbidity and mortality in childhood. Individually uncommon, together they number thousands, and many result in severe sequelae. Many are characterised by chronicity, high rates of disability or death. These conditions pose a large financial and emotional burden for affected children, their families and health systems.

To address this problem in the UK and Ireland, the British Paediatric Surveillance Unit (BPSU) was set up in July 1986, enabling paediatricians to participate in the surveillance and further study of rare disorders affecting children.

Several agencies founded and continue collaborating to support the work of the BPSU: the Royal College of Paediatrics and Child Health (RCPCH), Public Health England, University College London - Institute of Child Health (ICH), Health Protection Scotland (HPS) and the Faculty of Paediatrics of the Royal College of Physicians of Ireland. The BPSU's Scientific Committee meets every ten weeks to consider individual applications and the progress of studies.

Selection of studies for inclusion in the scheme

Details on the selection process and application process for the BPSU is now available online at www.rcpch.ac.uk/bpsu

Each application requires approval from the BPSU Scientific committee and approval from the Research Ethics Committee (REC) and Confidentiality Advisory Group of the Health Research Authority before commencement.

The reporting system

Surveillance is 'active' in that the BPSU office actively sends out cards to consultant paediatricians in the UK and Ireland asking for cases to be reported on the BPSU orange card (Figure 1). Each month, all clinicians participating in the surveillance scheme are sent either a postal or electronic orange card, listing the conditions currently under surveillance; follow-up reminders are sent to those who have not returned their card. A set of instructions for completing the card, including case definition of the conditions listed on the card is also circulated. When a new study begins, the mailing includes a specially produced protocol card and other information about the study.

Participants are expected to return **cards even if they have no cases to report** - there is a 'nothing to report' box on the card for them to tick. This is an important feature of the surveillance scheme as it allows us to measure compliance, which is continually monitored, to the reporting system.

Figure 1: Orange Card

British Paediatric Surveillance Unit Report Card
November 2013 [1311]
CODE No []

NOTHING TO REPORT ☐

Specify in box the number of cases seen

<input type="checkbox"/>	HIV & AIDS
<input type="checkbox"/>	Progressive Intellectual & Neurological Deterioration
<input type="checkbox"/>	Congenital Rubella
<input type="checkbox"/>	Congenital Syphilis
<input type="checkbox"/>	Acute Pancreatitis
<input type="checkbox"/>	Haemolytic Uraemic Syndrome (excl Scotland)
<input type="checkbox"/>	Kawasaki Disease

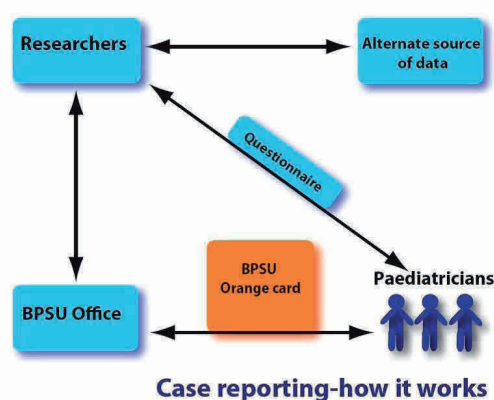
☐ E- CARD I would prefer to receive this now

Follow-up and confirmation of case reports

On receiving a case report the BPSU informs the relevant study team who send a short questionnaire to the reporting clinician to gather further information. As the questionnaire cannot be fully anonymised, the amount of patient identifiable data collected is strictly limited to preserve patient confidentiality. The study investigators report back to the BPSU, indicating when cases have been confirmed or are duplicate case reports (Figure 2). Duplication of reporting is most likely to occur when the condition requires referral to another clinician, but this is encouraged, as it is better to receive duplicate reports than to miss a case.

To improve case ascertainment for specific studies where a child may see specialist clinicians, consultants working in a number of other specialties have been invited to participate in the scheme. Apart from helping to improve ascertainment such complementary data sources help to validate the surveillance system.

Figure 2: Surveillance mechanism



2 Scientific Coordinator's Yearly Review of Activities

Two studies started in 2013, Kawasaki Disease (investigator Prof Robert Tulloh, RHSC Bristol) and acute pancreatitis (investigator Prof J H Shield, RHSC Bristol).



Richard Lynn,
Scientific Coordinator

Four studies had their period of surveillance extended: HIV, congenital rubella, progressive intellectual and neurological deterioration (PIND), and congenital syphilis.

Three new studies have commenced so far in 2014: acute hepatitis (Dr Shamez Ladhani, Public Health England), Group B streptococcal infection (Prof Paul Heath, St George's Hospital London), enterovirus and human parechovirus meningitis (Prof Mike Sharland, St George's Hospital, London).

During 2013/14, there were 16 publications relating to BPSU studies (see Appendix, p.24).

Participation in the scheme during the year 2013

Reporting rates for returning the orange cards remain high - the overall card return compliance rate for the year 2013, calculated as a proportion of orange cards returned, was 95.3% (38,436 / 40,321) a rise of 2% from 2012. Monthly response rates ranged from 96.7% in January to 93.9% in April with a median of 95.3%. Details of regional response rates are provided in Table 1 (overleaf).

The electronic reporting system introduced in 2012 now has over 75% of respondents receiving E-cards. By the end of 2014 we wish to see this increase to 90%.

Table 2 (overleaf) summarises the outcome of the follow-up of cases and provides evidence for their level of accuracy of reporting by clinician. By the end of a study 80-95% of the questionnaires will have been returned. The time taken to follow-up cases notifications varies between conditions and may be longer if microbiological/pathological details are required, or if a specialist committee has to convene to adjudicate on the case data.

Workload of those reporting in the scheme: 78% (3630) of participants had no cases to report in 2013, 14% (534) reported a single case, 7% (250) reported between two and four cases and 1% (49) reported five or more cases. The greatest number of cases reported was by HIV specialists, one of whom reported over 90 cases.

Public Patient Engagement

The BPSU is committed to wider public patient involvement in the development and dissemination of our work and that of the studies. To support clinicians when preparing their protocols several resource packs have been developed. These are available on our website at www.rcpch.ac.uk/bpsu/ppi

In March 2014 the BPSU in collaboration with Rare Disease UK and the RCPCH Youth Advisory Panel (YAP) hosted its first Rare Disease Day event. An invited audience of nearly 100 individuals, including patient support groups, researchers, clinicians, pharma, healthcare professionals and policy makers attended. Earl Howe spoke on addressing the complex needs of young people living with rare disease and how the UK rare disease strategy aims to help. He then presented a podcast made by the YAP, which presented the views of young people living with rare diseases. More details on the event including photos, presentation slides and the YAP living with rare disease podcast, can be found at www.rcpch.ac.uk/bpsu/rarediseaseday.

International activities

The BPSU continues to take an important role in the development of International Network of Paediatric Surveillance Units (www.inopsu.com). This year saw the BPSU attend the INoPSU conference facilitated by the Australian Unit in Melbourne in conjunction with the International Paediatric Association conference. The conference was well attended by delegates from around the world.

There were sessions to showcase the achievements of INoPSU over the last 15 years including impacts on clinical practice and policy. Examples include: changes in legislation for child restraints and seatbelts to keep young children safe from injury; supporting vaccination programmes including polio, rubella, and varicella; supporting a review of diagnostic criteria for early onset eating disorders and fetal alcohol syndrome; the value of long term surveillance for very rare groups of conditions such as progressive intellectual and neurological deterioration in children.

Funding

From August 2012 the BPSU has been funded through grants from the Scottish Executive; Great Ormond Street Charity, UCL, RCPCH, PHE, along with the contributions from researchers.

Table 1: Regional Response rate 2012 and 2013

Region	% return	Rank 2013	Rank 2012
East Anglia	97.1%	5	3
Mersey	92.9%	19	4
NET	93.8%	18	17
North Scotland	97.2%	3	7
North Western	94.8%	15	14
Northern	95.5%	10	6
Northern Ireland	93.9%	17	18
NWT	91.0%	20	16
Oxford	97.8%	1	13
Republic of Ireland	94.7%	16	20
SET	95.2%	13	9
South Scotland	95.5%	9	12
South Western	95.1%	14	2
SWT	95.4%	12	15
Trent	97.1%	4	10
Wales	97.2%	2	11
Wessex	96.7%	6	1
West Midlands	95.4%	11	8
West Scotland	96.3%	8	19
Yorkshire	96.5%	7	5

Figure 3: Regional Response rate 2013



Table 2: Outcome of follow-up of the cases reported in 2013 for conditions under surveillance at May 2014

Condition under surveillance	Date when reporting began	Valid reports	%	Duplicates	Errors	(D&E) %	Not yet known	%	Total
HIV	Jun-86	7,807	74	809	754	15	1190	11	10,560
CRU	Jun-91	89	47	37	65	53	0	0	191
PIND	May-97	1,963	54	444	1,053	41	158	4	3,618
CSY	Feb-10	50	47	25	15	38	16	15	106
VITD	Sep-11	88	64	15	19	25	16	12	138
HUS	Oct-11	146	32	114	67	40	128	28	455
GID	Oct-11	20	14	1	78	57	40	29	139
PDA	Oct-12	177	33	196	60	48	97	18	530
KAW	Jan-13	186	37	26	71	19	220	44	503
APAN	Apr-13	60	29	21	74	46	50	24	205
HEP	Jan-14	0	0	0	0	0	38	100	38
Total		10,586	64	1,688	2,256	24	1,953	12	16,483

HIV	Human immunodeficiency virus infection and perinatal human immunodeficiency virus exposure
CRU	Congenital rubella
PIND	Progressive intellectual and neurological deterioration
CSY	Congenital syphilis
VITD	Hypocalcaemic seizures due to vitamin d deficiency
HUS	Haemolytic uraemic syndrome
GID	Gender identity disorder
PDA	Patent ductus arteriosus
KAW	Kawasaki disease
APAN	Acute Pancreatitis
HEP	Acute infectious hepatitis

3 Surveillance Studies Undertaken in 2013

This year the individual reports have been shortened, concentrating on the background to the condition and on the analysis. General methodology information is contained in the study protocols and can be found at www.rcpch.ac.uk/bpsu/studies. Please take into consideration that the analysis presented here is provisional and has yet to be peer reviewed.

The investigators would like to acknowledge all those who are involved in their projects but are not mentioned and also the BPSU would like to thank all those who have returned cards, reported cases and completed the questionnaires.

Acute pancreatitis

Key points

- Acute pancreatitis is characterised by a sudden inflammation of the pancreas that can lead to multi-organ failure, pseudo-cyst formation and diabetes.
- To date, 197 case notifications have been received through the BPSU and clinicians have returned 134 (68%) questionnaires providing data on these cases; 78 cases have been confirmed as meeting the case definition.
- In an analysis of these initial cases, Acute pancreatitis appears to be more common than in previous studies and is associated with known factors, such as gallstones, anatomical abnormalities, infection and medication.

Summary

Acute pancreatitis (AP) is a rare disease in childhood, developing as a sudden inflammation of the pancreas with variable involvement of other regional tissues or remote organs.¹ It can lead to significant morbidity in the short, medium and long-term such as multi-organ failure, pseudo-cyst formation and diabetes. In adults, the most common causes of AP are alcohol and gallstones, but in children it can result from a wide variety of aetiologies, including trauma, drugs, infections, systemic diseases and congenital anomalies.² There is limited data on AP in childhood with no estimate of incidence in the UK and no guidelines for optimal management. A number of changes in population characteristics are likely to have altered the incidence and causative agents responsible for this condition in childhood, including mumps vaccination,³ obesity-associated gallstones and alcohol misuse.

This study will allow us to characterise the disease across the severity spectrum, the associated factors, such as obesity, and likelihood of death or long term complications. With this information we would hope to better understand how we might prevent the condition and perhaps treat patients more effectively in the future.

Collection of new case notifications. Follow-up of all cases, to determine outcomes at one year after diagnosis, is underway.



Dr Abdalmonem Majbar



Prof Julian Hamilton-Shield

Methodology

Data capture uses standard BPSU methodology. Details of the study protocol are available at www.rcpch.ac.uk/bpsu/apan. The British Association of Paediatric Surgeons (BAPS) is supporting this study and consultant paediatric surgeons who are members of BAPS received the Orange Card and reported cases.

Surveillance period

April 2013 – April 2014 (inclusive). Follow-up period: For a twelve month period after initial diagnosis (ending in April 2015).

Analysis

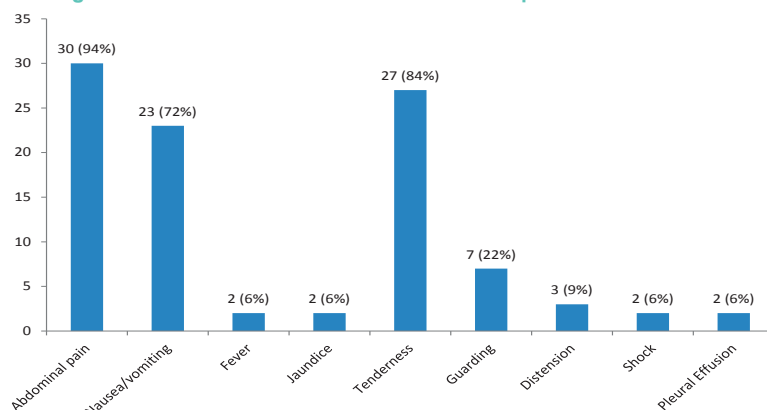
As of April 2014, there have been 197 case notifications to the BPSU. Of these, 134 (68%) responses to the study questionnaire have been received, either by completing it (119 cases) or by sending an email saying that the case was ineligible for the study (15 cases), and there has been no response or responses are still pending for the remaining 63 cases. The case status for the 134 responses was as follows: 78 cases were confirmed and valid, six require clinical discussion and 50 were either errors or duplicate cases.

Initial analysis of the first 32 valid cases: The median age of diagnosis was 7.12 years, range (2.3 to 14.9 years). Ten (69%) of the 32 patients were boys; the female to male ratio was 2:1.

As shown in Figure 4, abdominal pain was the most common presenting symptom (94%), followed by nausea and vomiting (72%). Abdominal tenderness was the most common clinical sign (84%).

Serum amylase was available for all cases and it was increased to the diagnostic level (≥ 3 times of the upper normal limit) in 30/32 (94%) cases,

Figure 4: Presenting clinical features for 32 children with acute pancreatitis



whereas serum lipase was requested in only 7/32 cases and it was raised to diagnostic level in six out of seven cases (86%).

At least one imaging study was performed for 30/32 (94%) cases. Abdominal ultrasound (US) was undertaken in 28 cases (88%), Endoscopic US in one case (3%), CT scan in five cases (16%), MRI in two cases (6%) and Magnetic resonance cholangiopancreatography (MRCP) in seven cases (22%). Endoscopic retrograde cholangiopancreatography (ERCP) was not done in any case. Abnormal findings were detected in 21/30 (70%) cases.

The frequency with which known associated factors were identified in this sample are shown in Table 3.

The major comorbidities reported were, hyperglycaemia in eight children (two were

discharged on regular subcutaneous insulin), respiratory failure in three (9.5%), pancreatic necrosis in three (9.5%), pseudo-cyst formation in two (6%) and circulatory collapse in two (6%).

Discussion

Interim analysis appears to confirm the original impression that although rare, AP is a condition that is seen in paediatric practice and has significant morbidity. Gallstones are now one of the commonest associations. Further analysis will reveal to what extent obesity may also contribute to this association.

Public and patient engagement

Pancreatitis Support Network.

Web: www.pancreatitis.org.uk.

Funding

University of Bristol.

References

1. Sarles H. Definitions and classifications of pancreatitis. *Pancreas* 1991; 6: 470–4.
2. Nydegger A, Couper RTL, Oliver MR. Childhood pancreatitis. *J Gastroenterol Hepatol* 2006; 21:499–509.
3. Savage E, Ramsay M, White J et al. Mumps outbreaks across England and Wales in 2004: observational study. *BMJ* 2005; 330 (7500):1119–20

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John Radcliffe Hospital: Prof Paul Johnson.
University of Bristol: Dr Linda Hunt,
Miss Eleri Cusick.
British Paediatric Surveillance Unit:
Mr Richard Lynn.

Table 3: Associated factors of acute pancreatitis*

Causes	No of cases (%)
Idiopathic (no cause report)	8 (25%)
Gallstone	5 (17%)
Anatomical anomaly	5 (17%)
Pancreatic divisum	2
Choledochal malformation	1
Others	2
Infection	4 (13%)
HSV 1	2
Non specified virus	1
Mycoplasma	1
Medication	4 (13%)
Sodium Valporate	2
Azathioprine	1
Asparaginase	1
Hereditary	3 (9%)
Others	6 (19%)
Cystic Fibrosis	2
Propionic acidemia	2
Methylmalonic acidemia	1
Isovaleric acidemia	1
*some cases have more than one associated factor	

Congenital hypothyroidism: UK collaborative study (UKCS-CHT)

Key points

- Congenital hypothyroidism affects around 200 babies in the UK each year and has been included in the UK newborn screening programme since 1981.
- During 13 months of surveillance, 726 children were notified to the study, including 634 notifications through both the BPSU and screening laboratories, 70 screen positive babies reported through UK screening laboratories only and 22 screen negative children reported through the BPSU only.
- The study has achieved a response rate of over 90% to the notification and one year follow-up questionnaires and has now commenced the second year of follow-up.

Summary

Primary congenital hypothyroidism (CHT) is a disorder involving decreased thyroid hormone production by the thyroid gland. Babies with CHT may have feeding difficulties, sleepiness, constipation and jaundice. Thyroid hormone can be replaced by giving babies oral thyroxine and, if treatment is started soon after birth, mental development and growth are improved.^{1,2} Around 200 babies with CHT are born in the UK each year and it is more common in girls than boys (approximately 2:1). Most babies with CHT are detected by newborn bloodspot screening (NBS).³ Although NBS for CHT started in 1981, we do not know how successful it is in identifying babies who require lifelong therapy.

This study will provide better information about how many children in the UK have CHT and how many of those are detected by NBS. It will also allow us to describe variations in clinical management and care. The surveillance period ended in June 2012 and we will follow-up every child for two years to find out about diagnosis and treatment and also to see if their CHT is temporary (transient). Importantly, we will also follow-up babies in whom the screening tests suggested CHT but the diagnosis was not confirmed, to find out if these babies remain well or develop any late problems.

This study will also provide a comprehensive picture of the investigation and management of CHT referrals after screening around the UK. Importantly, it will also tell us more about the effectiveness of the current NBS programme which will help to improve screening and underpin future clinical guidelines for care.

Surveillance period

June 2011 – June 2012 (inclusive). Follow-up period: For a 24 month period after initial diagnosis (period ending in June 2014).



Ms Juliet Oerton

Dr Rachel Knowles

Methodology

Data capture uses standard BPSU methodology; details of the study protocol are available at www.rcpch.ac.uk/bpsu/cht.

Analysis

Cases reported through the BPSU: Surveillance through the BPSU resulted in 428 valid notifications of which 360 (84%) were reported through the secure online questionnaire. Figure 5 shows a breakdown of case reports that were made by BPSU clinicians during 13 months of BPSU surveillance. (This does not include cases that were notified through laboratories which then provided contact details of the clinician who could complete the notification questionnaire).

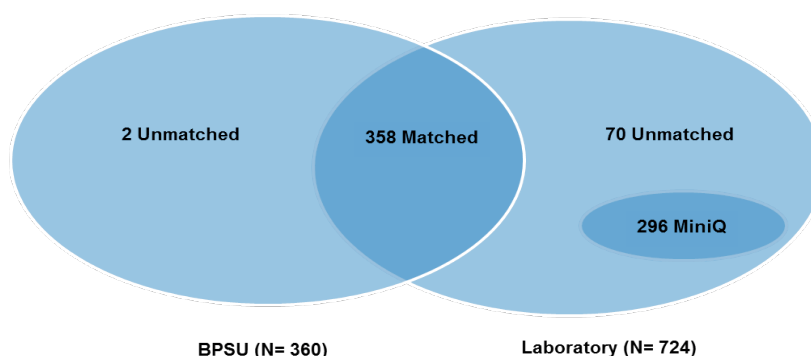
Newborn screening laboratories concurrently reported information about 704 babies with suspected CHT who were referred for further tests following a positive NBS result. Of the 360 cases notified through the BPSU, 338 were also notified as screen positive results by the screening laboratories. Information was sought from laboratories on a further 22 cases which were reported through the BPSU but had not been referred by laboratories as NBS positive; 20 of these were matched to a negative screening result and, in two cases, the screening result could not be traced. Finally for 366 cases reported as having positive screening results by laboratories but not reported through the BPSU, short questionnaires (miniQ) were sent to BPSU clinicians; 296 of these were completed and 70 were not.

Overall the response rate to questionnaires for valid case notifications was over 90%.

Discussion

Surveillance through the BPSU resulted in 360 valid cases reported through the secure online questionnaire. Laboratories referred 704 babies with 'suspected CHT' following a positive routine newborn screening test and traced screening results for a further 20 babies following a clinical notification through the BPSU. Full data from both clinician and laboratory were obtained for a total of 634 cases.

Figure 5: Matching of cases reported through the BPSU and laboratory surveillance schemes



A template for reviewing and classifying matched cases has been developed by the study expert review panel. Of 655 cases reviewed at diagnosis by the expert panel, 64% were definite CHT cases and 17% possible/probable CHT cases. Cases will be reviewed again once the additional one and two year follow-up data is complete.

There were a high number of 'unmatched' laboratory reports (n=366) without a corresponding paediatrician notification through the BPSU. Preliminary analysis of the expert panel classification suggests that this group may include a higher proportion of cases assigned as not CHT (30% vs. 9%). As much information as possible will be collected on these babies to ensure as complete a dataset as possible for analysis and to account for any reporting bias.

This study is providing important information about one of the major conditions that the UK NBS programme seeks to identify and, importantly, about the effectiveness of the current programme in both detecting CHT early in life and preventing adverse outcomes. Results from the first and second year of follow-up are required to confirm the permanence of the CHT diagnosis and outcomes up to two years of age.

Acknowledgements

We are very grateful to reporting paediatricians, staff in the participating laboratories for their support in submitting monthly data and for assisting in tracing clinical information for initially unmatched cases.

We would like to acknowledge the help of the current epiLab staff – Rich Hutchinson and Luke Romanowski – for their help in establishing and supporting the online questionnaire system and Jamili Miah (NHS Newborn Bloodspot Screening Programme, Public Health England) for providing administrative support to study team meetings.

Funding

Department of Health through the UK Newborn Screening Programme Centre (now based within Public Health England).

Public patient engagement

British Thyroid Foundation.
Web: www.btf-thyroid.org.

NHS Newborn Bloodspot Screening Programme.
Web: newbornbloodspot.screening.nhs.uk.

References

1. Klein AH, Meltzer S, Kenny FM. Improved prognosis in congenital hypothyroidism treated before age three months. *J Pediatr*. 1972; 81:912-915
2. Selva K, Harper A, Downs A, Blasco PA, LaFranchi SH. Neurodevelopmental outcomes in congenital hypothyroidism: comparison of initial T4 dose and time to reach target T4 and TSH. *J Pediatr* 2005; 147: 775-780.
3. Department of Health and Social Security. Screening for congenital hypothyroidism. London: DHSS Joint Standing Sub-Committee on Screening in Medical Care, 1980; HN(81)20, Appendix 1

Researcher contacts

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Birmingham Children's Hospital: Mr Paul Griffiths.

Congenital rubella

Key points

- Congenital rubella syndrome can occur when a woman contracts rubella during the first trimester of pregnancy. It can cause deafness, blindness and heart defects in the fetus, amongst other symptoms.
- No congenital rubella births were reported in the UK or Ireland in 2013.

Summary

Surveillance of congenital rubella (CR) has been in place since 1971, when the National Congenital Rubella Surveillance Programme (NCRSP) was set up to monitor the impact of rubella vaccine; there has been active surveillance through the BPSU since 1990.¹

Antenatal screening for rubella susceptibility, with post-partum vaccination of susceptible women, has also formed part of the strategy for preventing CR infection. The National Screening Committee and Public Health England are currently considering alternatives to the antenatal rubella screening programme, and in the meantime, the National Screening Committee has agreed to support the continuation of CR surveillance through the orange card.

There have been no confirmed cases of CR reported in infants born in the UK or Ireland since 2011. However, rubella is still endemic in many parts of the world and national borders are no barrier to infection. In 2012 and 2013 there were substantial outbreaks of rubella in Romania, Poland and Japan, all of which resulted in the births of infants with CR. The World Health Organisation has a target date of 2015 for the elimination of measles and rubella, and prevention of congenital rubella infection (<1 case of congenital rubella syndrome per 100,000 births) in Europe.^{2,3}

National surveillance of CR through the BPSU continues to be extremely valuable. It provides a mechanism for timely reporting of any cases which do occur, and establishing whether maternal infections were acquired abroad or at home, and it helps to maintain awareness of this now rare but potentially devastating infection. Congenitally infected infants can excrete rubella virus for an extended period of time, and every infected infant must be diagnosed and managed appropriately to avoid the risk of contributing to further community transmission.

Methodology

Data capture uses standard BPSU methodology; details of the study protocol are available at www.rcpch.ac.uk/bpsu/cr.



Dr Pat Tookey

Surveillance period

January 1990 and is reviewed yearly.

Analysis

There were no reports of CR to the BPSU in 2013, and the only confirmed report in 2012 related to an older child who was born abroad. The surveillance case definition was revised in 2005 to include newly diagnosed children who were born abroad, in order not to miss any cases, and to contribute to European surveillance data.

CR births in the UK or Ireland 1990-2013: 64 children and four stillborn infants with confirmed or compatible congenital rubella have been born and reported since active surveillance was established in 1990; 51 of these (75%) were first reported through the BPSU (Table 4). Twenty infants were born since 2000, including two born in Ireland or Northern Ireland, and two who were stillborn. Although 12 were imported cases with maternal infection acquired abroad (seven in Southern or South Eastern Asia, five in Africa), eight infants were born to women whose infection occurred in the UK or Ireland.

Table 4: Confirmed and compatible congenital rubella births in the UK and Ireland 1990-2013

Year of birth	Primary Source of notification		
	BPSU	Other	Total
1990-94 * ^	22	10	32
1995-99	12	4	16
2000-04 *	10	1	11
2005-09 *	4	2	6
2010-13	3	0	3
Total	51	17	68
* includes a stillborn infant			
^ includes a set of triplets, one of whom was stillborn			

Discussion

Very few cases of CR have been reported in the last decade, with none reported in 2012 or 2013. Most reports concern infants with serious rubella-associated defects present at birth, and it is possible that some infants with less obvious signs of CR are not diagnosed and reported.

Even while rubella infection is rare in the UK and Ireland, susceptible women who travel abroad during early pregnancy may come into contact

with infection. Health professionals, particularly paediatricians and those working in primary care and antenatal care, must continue to be aware of the potential serious implications of rash illness in early pregnancy, the guidelines for the management of rash illness in pregnancy,⁴ and also of the early signs of congenital rubella.

Funding

UCL – Institute of Child Health: National Screening Committee (2014).

Public patient engagement

Sense. Web: www.sense.org.uk

References

1. Tookey PA, Peckham CS. Surveillance of congenital rubella in Great Britain, 1971-96. *BMJ* 1999; 318:769-70.

2. Paradowska-Stankiewicz I, Czarkowski MP, Derrough T, Stefanoff P. Ongoing outbreak of rubella among young male adults in Poland: increased risk of congenital rubella infections. *Euro Surveill.* 2013;18(21) pii: 20485

3. CDC. Nationwide rubella epidemic-Japan, 2013. *MMWR* 2013;62:457–62

4. HPA Rash Guidance Working Group. Guidance on viral rash in pregnancy. Available at www.gov.uk/government/uploads/system/uploads/attachment_data/file/322688/Viral_rash_in_pregnancy_guidance.pdf

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Congenital syphilis

Key points

- Congenital syphilis (syphilis infection transferred from mother to baby in the womb) can cause miscarriage, neonatal death, deafness and bone deformities.
- Between February 2010 and December 2012, 15 confirmed cases and one possible case of congenital syphilis were found.
- Since 2001, rates of infectious syphilis have increased amongst reproductive age women.

Summary

Congenital syphilis (CS) occurs when syphilis is transmitted from a woman to her unborn baby during pregnancy. This can lead to miscarriage, stillbirth, neonatal death, or disorders such as deafness and bone deformities. As such, CS is a distressing and costly condition. Since 2001, rates of infectious syphilis have increased amongst women of reproductive age. Cases of CS can be prevented through antenatal screening and appropriate treatment. These control methods are highly cost effective but are dependent on well-structured healthcare pathways. Despite the availability of antenatal screening, cases of CS have been reported over the past decade. The re-emergence of CS reflects a failure of prenatal care and syphilis control programmes, and concerns have been raised about the effectiveness of the present control strategies. In particular, the development of control efforts has been limited by the absence of comprehensive systematic national surveillance of congenital syphilis.

This study started in 2010 and its main aim is to compare the incidence, management and presentation of cases with that observed in the similar UK study carried out in the 1990s which used a similar methodology.¹ The



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information will be used as an evidence base to improve clinical pathways and patient management systems.

Surveillance period

January 2010 - December 2014 (inclusive).

Methodology

Details of the study protocol are available at www.rcpch.ac.uk/bpsu/cs. The methodology was based on that described by Hurtig et al. (1998) in the previous study of congenital syphilis undertaken between 1994 and 1997.¹

Analysis

Children born between February 2010 and December 2014 with a suspected CS diagnosis were investigated. For all years reported CS incidence was below the WHO threshold for elimination (<0.5/1000 live births). 15 confirmed and one possible case of CS were identified, 12 of which were male and four female. Of the confirmed cases, 13 were reported through the BPSU. Two were notified by microbiologists outwith the BPSU and PHE surveillance systems. Median birth weight was 2822g (range: 1340g to 3686g). All cases of CS were reported for English Strategic Health Authorities except South West, and Yorkshire and The Humber. Five cases were born preterm (<37 weeks

gestation). Clinical presentation varied from asymptomatic to acute, including severe anaemia, mucosal lesion, hepatosplenomegaly, rhinitis, oedema, thrombocytopaenia, skeletal damage, and neurosyphilis. One infant was deaf and blind. Median maternal age was 23 (range: 17 to 35). Where maternal stage of infection was recorded, 7/11 presented with primary and 4/11 secondary syphilis. Most mothers were white (12/15) and three were born in Eastern Europe or the Middle East. In general, CS was seen in infants born to women who had difficulty accessing healthcare due to cultural barriers and/or high levels of socio-economic deprivation.

Discussion

WHO Europe seeks to eliminate CS by 2015 using a highly cost effective three step strategy; universal access to antenatal care: access to care in early pregnancy and on-site testing and treatment supported by clearly structured healthcare pathways (WHO, 2013).² In the UK, this well established strategy is supported by open access, free and confidential GUM services, including partner notification and consequently the incidence of CS is below the WHO elimination threshold. Although few cases have been detected the results show that CS still presents complex clinical, social and public health problems across England. Reducing the public health impact of this avoidable disease is highly dependent on the successful implementation of WHO standards for the elimination of mother to child transmission of syphilis across Europe.

Funding

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References

1. Hurtig A-K, Nicoll A, Carne C, et al. (1998) Syphilis in pregnant women and their children in the United Kingdom: Results from national clinician reporting surveys 1994–7. *BMJ* 317:1617–1619
2. WHO. Global guidance on criteria and processes for validation: elimination of mother-to-child transmission (EMTCT) of HIV and syphilis. Geneva, World Health Organisation, 2013. Available at: www.who.int/reproductivehealth/publications/rtis/9789241505888/en/index.html.

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Imperial College: Dr Hermione G Lyall.

Haemolytic uraemic syndrome (HUS)

Key points

- HUS is a syndrome that can develop as a result of E. Coli VTEC infection. It is characterised by breakdown of red blood cells. It can lead to acute kidney failure and, occasionally death.
- The HUS study launched in October 2011, with 213 patients reported through the BPSU orange card to December 2013.
- The majority of cases for which questionnaires were returned were also identified as VTEC cases in other surveillance systems. However, many of these cases were not noted as having HUS on these systems, demonstrating the added value of this surveillance study.

Summary

Haemolytic Uraemic Syndrome (HUS) is a rare, but occasionally fatal, condition that can develop following gastrointestinal infection with a Verocytotoxin-producing strain of Escherichia coli (VTEC). The peak of incidence is in children under five years of age who are the most susceptible age group. A previous BPSU study¹ was conducted between 1997 and 2001 when 413 cases were reported, 330 of whom were VTEC related.

This study aims to measure the incidence of HUS in England, Northern Ireland, Wales and the Republic of Ireland, and describe clinical and demographic features. Scottish data is collected through a separate enhanced surveillance program set up in 2003. By comparing the results from this study to that of the previous BPSU study any changes in epidemiology will be apparent. The one year follow-up will provide useful information on the outcomes of illness. By linking cases reported through this study to national surveillance systems for VTEC, we will seek to identify factors associated with an increased risk of developing HUS, in the hope that we might, in the future, be in a position to prevent at risk children from developing HUS following VTEC infection.



Dr Bob Adak

Surveillance period

October 2011 – October 2014 (inclusive). Follow-up period: For a twelve month period ending in October 2015.

Methodology

Data capture uses standard BPSU methodology; details of the study protocol are available at www.rcpch.ac.uk/bpsu/hus.

Analysis

Between 1st October 2011 and 31st December 2013, 426 orange card notifications of HUS were made to the BPSU from clinicians in England, Wales, Northern Ireland and the Republic of Ireland. Clinicians responded that 73 (17.1%) notifications were made in error (i.e. duplicate notification, ticked the wrong box etc). In total, 274 completed study questionnaires were received, two were incomplete and 61 were duplicates. Thus, a total of 213 patients have been reported through the BPSU.

One year follow-up questionnaires are sent to reporting clinicians 12 months after the original hospital admission date of patients. Of the 126 cases eligible for follow-up during the study period, 105 (83.3%) have been received and 21 are awaited.

Where ethnicity was reported, 97% of confirmed cases reported through the BPSU were of white ethnicity and slightly more females (55%) were reported than males. This is comparable to the demographic composition of VTEC cases.

Figure 6: Venn diagram of HUS notifications

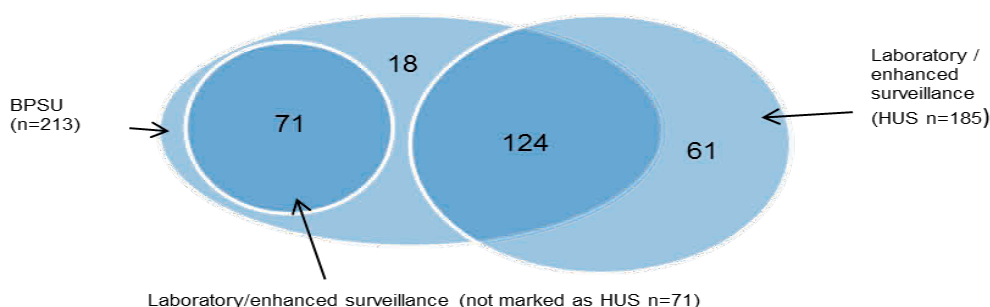


Table 5: Case status and incidence by country

Place of birth / report	Confirmed or compatible cases*	Incidence**
England	154	8.4
Wales	10	7.2
N Ireland	8	17.5
Republic of Ireland	42	19.3
Total***	214	

* The case definition for inclusion in the study have not yet been applied to reported cases of HUS so confirmed/compatible cases include all those reported as HUS by the reporting clinician.

** Incidence is per million population aged under 16 years of age per year. Denominator data is taken from the 2011 census for both the UK countries and Republic of Ireland.

*** Scottish data available from Health Protection Scotland.

Discussion

The response rate in the first 26 months of the study was 81% overall. Reconciliation against existing surveillance systems demonstrates the value of the surveillance study, capturing an additional 71 cases captured by existing surveillance systems for VTEC, but not marked by HUS, and 18 cases only captured by the BPSU system. HUS often develops weeks after VTEC infection, by which time, exposure questionnaires and laboratory forms may already have been completed and we remain unaware of the development of HUS.

61 cases have been identified as HUS through national surveillance systems of VTEC which are not captured through the BPSU study and study investigators are working to obtain questionnaires for these cases and improve ascertainment overall.

In England, examination of laboratory data for HUS cases reported through the BPSU indicated that 13% of cases had no specimen submitted to Gastrointestinal Bacteria Reference Unit. 20 cases had specimens submitted but there was no evidence of VTEC infection.

Funding

Public Health England.

Public patient engagement

Haemolytic Uraemic Syndrome Help (HUSH).
Web: www.ecoli-uk.com

References

1. Lynn R.M, O'Brien S.J, Taylor C.M, Adak G.K, Chart H, Cheasty T, et al. Childhood Haemolytic Uraemic Syndrome, United Kingdom and Ireland. *Emerg Infect Dis* 2005; 11:590-96

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Health Protection Scotland: Mary Locking
Birmingham Children's Hospital: Mark Taylor
Royal Manchester Children's Hospital: Nick Webb
Public Health Wales: Robert Smith

HIV infection & perinatal HIV exposure

Key points

- The number of children born each year in the UK and Ireland to women with diagnosed HIV infection is currently stable at about 1300-1400; the mother to child transmission rate from diagnosed women is now under 0.5%.
- Median age at diagnosis of children reported 2008-2013 was eight years; about two-thirds were born abroad.
- In 2013 the NSHPC instituted an audit to explore the circumstances of all perinatal infections reported in the UK since 2006 (109 by end 2013).

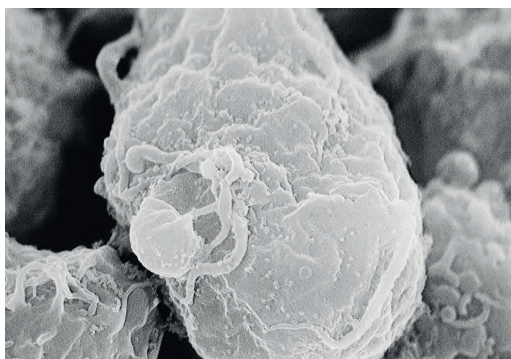
Summary

National surveillance of paediatric AIDS began in 1986 and was extended to include HIV infection and perinatal HIV exposure in 1989; it is based on comprehensive, anonymised, paediatric and obstetric reporting. Data from all sources are combined as the National Study of HIV in Pregnancy and Childhood (NSHPC) at the UCL Institute of Child Health.¹

Out of approximately 2,500 children diagnosed with HIV infection in the UK and Ireland to date (Figure 7), 83% acquired their infection through mother-to-child transmission (MTCT). About 270 young people with haemophilia were reported, all before 1995, and some children (fewer than 100) acquired their infection through other routes e.g. blood transfusion abroad. The majority of recently diagnosed children were born abroad, mostly in sub-Saharan Africa.

Antenatal screening for HIV has been offered and recommended to all pregnant women in the UK and Ireland for more than a decade; uptake of screening is very high (estimated 95% or higher nationally since 2008, see www.screening.nhs.uk/news.php?id=15686). Diagnosis rates are also extremely high, with very few HIV-positive women remain undiagnosed by the time of delivery. The number of births to diagnosed women increased substantially from about 100 in 1997 to 1,400 in 2007, and stabilized thereafter. By 2009 four in every ten pregnancies reported were in women having a second or subsequent pregnancy

Figure 7: Scanning EM of HIV, grown in cultured lymphocytes. Virions are seen as small spheres on the surface of the cell



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since their HIV diagnosis,² and since 2010, 75% of all HIV-positive pregnant women were diagnosed before their current pregnancy. MTCT rates from diagnosed women are now under 1% compared with an estimated 20-30% from undiagnosed women.³ Despite this, up to 20 children a year probably still acquire HIV infection perinatally, or through breastfeeding; most of these infections occur in children born to women who are not aware of their HIV infection at the time of delivery. Additional information on antenatal screening and management, and infant presentation and diagnosis, is currently being sought in an NSHPC audit of perinatal HIV infections occurring in the UK since 2006.

Methodology

Details of the study protocol are available at: www.rcpch.ac.uk/bpsu/hiv.

Surveillance period

Surveillance began in June 1986 and is reviewed yearly. Follow-up period: Infection status of perinatally-exposed infants is usually reported during the child's first year of life. Long-term follow-up information on children with confirmed HIV infection is collected through the Collaborative HIV Paediatric Study (CHIPS), a collaboration between the NSHPC, the MRC Clinical Trials Unit, and clinicians (www.chipscohort.ac.uk).

Analysis

During 2013 there were 345 reports from the orange card, bringing the total number of BPSU reports since surveillance began to 10,238. 7,811 were confirmed cases of HIV infection or exposed infants at risk of vertical transmission; 836 reports were still being investigated and 1,591 reports were duplicate or error reports. A further 10,611 confirmed cases and exposed infants were reported through other sources (see study protocol). The rest of this report includes data from all reporting sources.

Altogether 18,830 HIV-infected or exposed children have been reported, 16,823 (89%) since 2000 (see Table 6 overleaf). In England, the proportion of reports made from outside the London regions has increased from 29% (453/1,573) for the reporting period from 1986 to 1999, to 50% for the period 2000 to 2013.

Table 6: HIV infection and infants born to HIV-positive women (all reporting sources) Region and time period of report (notified by 31 December 2013)

Region of first report	1986-1999	2000-2013	Total
England total	1,573	1,464	16,037
London	1,120 (71%)	7,302 (50%)	8,422
North	181 (11%)	2,184 (15%)	2,365
Midlands & East	128 (8%)	3,145 (22%)	3,273
South	144 (9%)	1,833 (13%)	1,977
Wales	26	237	263
Northern Ireland	4	97	101
Scotland	232	514	746
Ireland	170	1,513	1,683
Total	2,005	16,823	18,830

Children born to HIV-positive women: Most paediatric reports (18,403/18,830, 98%) were of children born to HIV-positive women. Of these children, 2,104 (12%) were known to be infected, and 14,036 (76%) uninfected by the end of 2013; infection status for the remaining 2,263 (12%) had not yet been reported, but the majority were recent reports and very few are likely to be infected children.

Between 2006 and 2012 the annual number of births to diagnosed women in the UK and Ireland stabilized at about 1,300-1,400 (reports for recent years are still incomplete) (Table 7). We recently reported an overall transmission rate of 0.5% for births to diagnosed women 2010-2011 (9/1975, 95% CI: 0.2-0.9%),² declining from 2.1% in 2001-2002. This is primarily due to an increase in the proportion of women on combination antiretroviral therapy (cART) in early pregnancy and fewer transmissions related to late initiation or non-receipt of antenatal cART. An increasing proportion of pregnancies reported are second or subsequent pregnancies since maternal HIV diagnosis, reaching almost 40% in 2009.³ About three-quarters of infants born since 2010 had mothers who were aware of their HIV status at conception, with over 50% on combination antiretroviral therapy at conception in 2013 (regularly updated NSHPC slides available at www.ucl.ac.uk/nshpc/slides).

Infected children: Since surveillance started in 1986, 2,527 children with HIV infection have been reported; this includes 267 probably infected in the course of treatment for haemophilia, all born before 1985 and reported before 1995. Among the remaining 2,260 mostly perinatally infected children, 281 (12%) are known to have died, 120 (5%) to have gone abroad and 408 (18%) to have transferred to adult services; a further 230 (10%) are lost to follow up. More than half (56%) of all children reported with HIV infection were born abroad, the majority in sub-Saharan Africa; this proportion increased from about one third of those diagnosed before 2000 to about two-thirds of those diagnosed since.

Of the children diagnosed 2006-2013, median age at diagnosis for those born abroad was 10 years (IQR 7-13), compared with 1 year (IQR 0-5) for UK and Irish born children; overall median age at diagnosis was eight years (IQR 3-12). Of the 1,301 children and young people still being seen in paediatric or transition clinics in 2013, over 40% were aged 16 or older and another 37% were 11-15 years. Over 70 pregnancies have been reported in perinatally infected young women notified as paediatric cases.

Of the 1,007 children known to have acquired infection perinatally in the UK or Ireland, most (76%) had mothers who were not diagnosed by

Table 7: Year of birth and infection status of children born in the UK or Ireland to women diagnosed by the time of delivery. (notified by 31 December 2013)

Year of Birth	Infected	Indeterminate	Not infected	Total
1984-1999	111	141	899	1,151
2000-2005	62	308	4,627	4,997
2006-2008	27	150	3,883	4,060
2009-2011	17	443	3,562	4,022
2012-2013*	4	1,141	748	1,893
Total	221	2,183	13,719	16,123

*reports for recent years are subject to reporting delay

the time of delivery. There were 109 children born since 2006 confirmed infected by the end of 2013 (39% born to diagnosed and 61% to undiagnosed women). The number of reports of perinatally infected children born to undiagnosed women during this period will increase as later diagnoses are made.

Discussion

The number of births to HIV-positive women in the UK and Ireland increased substantially each year until 2007, and then stabilised. An increasing number of women are embarking on second and third pregnancies after their initial HIV diagnosis, and by 2012 more than half of all infants born to diagnosed women were exposed to cART at conception.

MTCT rates from diagnosed women have declined from about 2% in 2000 to 0.5% in 2010/11, with extremely low rates among women who received optimal treatment according to the British HIV Association pregnancy management guidelines (www.bhiva.org/guidelines.aspx). However, despite high uptake of antenatal testing and interventions to prevent MTCT, some infants are still acquiring HIV infection perinatally or through breastfeeding. An NSHPC audit of perinatal HIV infection among children born in the UK since 2006 was established in 2013; findings are expected to be available late in 2014.

Almost two-thirds of infected children diagnosed since 2000 in the UK or Ireland were born abroad, mostly in sub-Saharan Africa, and the median age of newly diagnosed children is likely to remain high. As young people leave paediatric care, it becomes more important to ensure that it is possible to monitor their health and well-being in adult life. Following on from the NSHPC and CHIPS surveillance studies, a prospective cohort study of perinatally infected young people and HIV negative controls started recruitment at the end of 2012. AALPHI (Adolescents and Adults Living with Perinatal HIV) aims to explore the impact of life-long HIV and long-term antiretroviral therapy on neurocognitive, cardiac and metabolic function, growth, and sexual and reproductive health (more details at www.ctu.mrc.uk).

Children affected by HIV are reported to the NSHPC from all parts of the UK and Ireland.¹ The wide geographical distribution of newly reported cases highlights the valuable role of the BPSU in identifying infected children diagnosed outside the specialist paediatric HIV centres, as well as exposed infants born to infected women in lower prevalence areas throughout the British Isles.

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Public patient engagement

Positively UK.

Web: www.positivelyuk.org

Body and Soul.

Web: www.bodyandsoulcharity.org

References

1. Townsend CL, Cortina-Borja M, Peckham CS, Tookey PA. Trends in management and outcome of pregnancies in HIV-infected women in the United Kingdom and Ireland, 1990-2006. *BJOG* 2008; 115:1078-86
2. French CE, Cortina-Borja M, Thorne C, Tookey PA. Incidence, patterns, and predictors of repeat pregnancies among HIV-infected women in the United Kingdom and Ireland, 1990-2009. *J AIDS* 2012; 59:287-93
3. Townsend CL, Byrne L, Cortina-Borja M, Thorne C, de Ruiter A, Lyall H, et al. Earlier initiation of ART and further decline in mother-to-child HIV transmission rates, 2000-2011. *AIDS* 2014;28(7):1049-1057.

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Hypocalcaemic seizures secondary to vitamin D deficiency

Key points

- Surveillance for this study commenced in September 2011 and finished in September 2013.
- There were 81 confirmed cases and ten probable cases notified; incidence of 3.49 per million children age 0-15 years.
- Incidence was greatest in infants (aged less than 12 months), and in children from South Asian and Black ethnic backgrounds.
- An unexpected finding was a marked gender imbalance, with incidence higher in males.

Summary

Vitamin D deficiency (VDD) can cause fits (seizures) in children by lowering blood calcium levels. Young babies are at particularly high risk of this complication, and at this age the cause is often VDD in the mother during pregnancy. There is concern that VDD, once rare in the UK, is increasing in prevalence due to inadequate sunlight exposure and a diet low in vitamin D. Certain ethnic groups appear to be at particularly high risk, e.g. South Asian and African-Caribbean. However, there is very little information regarding the extent of the problem.

VDD is preventable by dietary supplementation. Current UK recommendations are that vitamin D supplements are taken by all pregnant and breastfeeding women, and by children aged six months to five years who drink less than 500ml of formula milk per day. However, these supplements are currently only offered to low income families by the NHS (as part of the Healthy Start Scheme), and uptake of the supplements is low. It is argued by some professionals that vitamin D supplements should be routinely available and encouraged for all at risk groups. The study will estimate the incidence, demographic distribution and prevalence of known risk factors and outcomes of hypocalcaemic fits resulting from VDD in children (aged 0-15 years) in the UK and identify which sub-groups of the population are most affected. It will also help to identify whether or not current public health measures are adequate at preventing fits due to VDD in children.

Methodology

Data capture uses standard BPSU methodology; details of the study protocol are available at www.rcpch.ac.uk/bpsu/vitd

Surveillance period

September 2011 - September 2013 (inclusive).



Dr Emre Basatemur

Analysis

Of 137 notifications in total, there were 81 confirmed cases and ten probable cases. It was not possible to ascertain case status for ten notifications (7%), either because the reporting consultant was unable to recall the patient's identifiers and locate the case notes (n=5), or because the reporting consultant was not contactable or did not return the data collection form despite several reminders (n=5). There were 18 duplicate notifications, and 18 cases were reported in error or did not meet the case criteria. The 91 confirmed and probable cases were included for analysis.

There was a marked difference in the number of cases by sex, with 82% being male. eighty-six (95%) of cases were young children aged 0-2 years, and the remaining five children were adolescents aged between 11-15 years. The majority of cases came from high-risk ethnic groups for VDD; 54 (59%) were of South Asian ethnicity and 22 (24%) were black, whilst 11 (12%) came from a white background and four (4%) were of mixed or other ethnicity.

The estimated annual incidence of hypocalcaemic seizures secondary to VDD was 3.49 per million children age 0-15 year in the UK and Ireland (95% CI: 2.81 to 4.26). Incidence was significantly greater in males compared to females, in infants compared to older children, and in children of South Asian or Black ethnicity compared to children from white ethnic backgrounds.

61% of cases had multiple seizures. In the majority of children, seizure duration was less than five minutes (69%), with more prolonged seizures lasting ≥ 10 minutes in 19% of cases. 80% of children did not have any other clinical features of VDD, whilst 15% exhibited clinical features of rickets.

Discussion

Our results are consistent with previous studies in identifying children from South Asian and Black ethnic groups as being at highest risk of symptomatic VDD in the UK. The age distribution of cases is also consistent with previous reports, which suggest that VDD presents with hypocalcaemic symptoms in two distinct age groups; young children under

three years of age (the majority of cases) and older children above ten years. These age groups have been shown to correlate with periods of rapid growth, and it has been suggested that higher metabolic demand for calcium during periods of rapid bone growth increases susceptibility for the development of hypocalcaemia. An unexpected male predominance in cases, which has been reported in several recent cases series was observed.^{3,4}

In the majority of cases, seizures were brief and self-terminating. Clinical outcomes were good, with no deaths reported, and only one child reported to have sequelae at the time of discharge (an iatrogenic burn related to intravenous treatment). Guidelines for treatment of VDD have recently been published by the Royal College of Paediatrics and Child Health.⁵

The full report has now been published.⁶

Funding

Sir Peter Tizard Bursary.

References

1. Callaghan AL, Moy RJD, Booth IW, Debelle G, Shaw NJ. Incidence of symptomatic vitamin D deficiency. *Archives of disease in childhood*. 2006;91(7):606–7.
2. Ladhani S, Srinivasan L, Buchanan C, Allgrove J. Presentation of vitamin D deficiency. *Archives of*

Disease in Childhood. 2004;89(8):781-4.

3. Bicakci Z. The relationship of hypocalcemic convulsions related to nutritional rickets with age, gender, season, and serum phosphorus levels. *Neurosciences (Riyadh)*. 2007;12(4):302–305.

4. Thomas TC, Smith JM, White PC, Adhikari S. Transient neonatal hypocalcemia: presentation and outcomes. *Pediatrics*. 2012;129(6):e1461–e1467.

5. Guide for vitamin D in childhood. October 2013. RCPCH www.rcpch.ac.uk/improving-child-health/public-health/vitamin-d/vitamin-d

6. Basatemur E, Sutcliffe A. Incidence of Hypocalcaemic seizures due to vitamin D deficiency in children in the United Kingdom & Ireland. *J Clin Endocrinol Metab*. 2014; Oct 3: jc20142773

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Kawasaki disease: complete & incomplete

Key points

- Kawasaki disease is the commonest cause of acquired heart disease in children in the UK and USA. The serious health impacts of Kawasaki disease make it important to diagnose this condition early in order to treat it effectively and therefore minimise complications and long-term ill health within the paediatric population.
- There have now been 398 cases registered with BPSU for the study – this figure includes duplicate registrations. To date, completed questionnaires have been returned for 253 cases, of which 186 have the complete form of Kawasaki disease.

Summary

Kawasaki disease (KD) is the commonest cause of acquired heart disease in children in the UK and USA but the cause is unknown. The serious health impacts of KD make it important to diagnose this condition early in order to treat it effectively and therefore minimise complications and long-term ill health within the paediatric population. A review of KD in 2009 reported that the incidence varies from 184 per 100,000 children under five years in Japan to about 8-9 per 100,000 in England. Seasonal variation, with peaks in winter and spring, and reported epidemics have suggested that there are environmental factors, for example viral infections, which trigger the condition. Although most children are diagnosed under the age of five years, KD also occurs at older ages and the incidence, presentation and course of the disease in older children in Western Europe will be of scientific interest.

This study will describe the incidence and characteristics of children diagnosed with KD and describe the clinical presentation and management and to describe the cardiac and non-cardiac sequelae associated with KD within 30 days following diagnosis.

A change in guidelines during this year has resulted in significant changes in the way that clinicians manage and report cases of KD.¹ As a result, ethical and BPSU approval was given to extend surveillance for another year to take account of these changes. In addition, it became clear that the restriction of reporting to *complete* KD only, limited the number of cases, and may have caused some confusion amongst reporting clinicians. Hence, it was agreed to extend the case definition to *complete and incomplete* cases of Kawasaki disease in the second year of surveillance in order to ensure case ascertainment would be complete.



Prof Rob Tulloh

Surveillance period

January 2013 - January 2015 (inclusive).

Methodology

Data capture uses standard BPSU methodology; details of the study protocol are available at www.rcpch.ac.uk/bpsu/kd. The case definition for the period January 2014 to January 2015 has been extended to include “incomplete” KD as well as “complete.”

Analysis

As of March 2014 there have been 253 completed questionnaires returned to the study team.

Of these:

186 cases of complete KD.

21 cases had sufficient data to class as symptomatically incomplete KD treated with Intravenous immunoglobulin (IVIG) (this category includes children with abnormal ECHO results).

46 cases did not fulfill the criteria for KD, were duplicate reports or reported in error.

Interim analysis of 186 complete cases reported to date was undertaken.

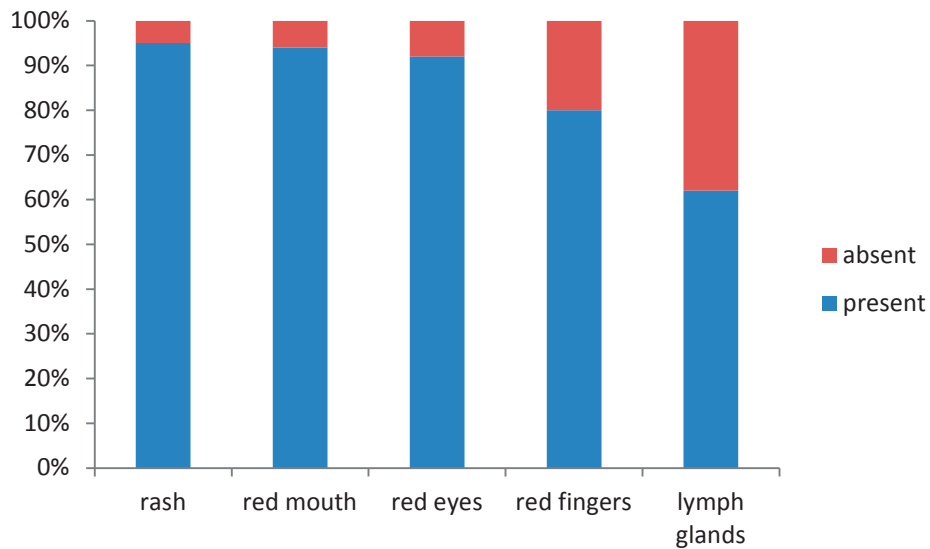
Gender: Complete KD: 66 girls, 120 boys (sex ratio 1:1.8).

Age range: Sixty-four (88%) of the complete cases were aged between one and five years (age range 2 months to 15 years).

Table 8: Ethnicity of cases of complete Kawasaki disease notified to the study

Ethnic Group	No of children (%)
White	120 (65%)
Black (African & Caribbean)	20 (11%)
Asian (excluding Chinese)	15 (8%)
Mixed (White/Black or White/Indian)	13 (7%)
Chinese/Japanese	17 (9%)
Not reported	1
Total	186 (100%)

Figure 8: Percentage of complete cases with features in addition to fever



Ethnicity: The percentage of Chinese/Japanese/Korean children (n=17, 9%) within the study sample is relatively high, in comparison with the UK child population, and the number of Black children (n=20, 11%) is also higher than expected.

All children presented with a history of fever, which was five or more days in all but one case (185/186). A rash was the second most common symptom, present in 176/186 cases. It was usually the second symptom to appear.

174/184 complete cases were given IVIG (missing data: n=2).

The period from onset of KD (i.e. date of first symptom) to start of IVIG was a median of seven days (range three to 32 days).

Clinicians reported clinical sequelae at discharge for 23 of the complete cases (two cases missing data). No deaths were reported in children with onsets since November 2012.

Discussion

The project has produced interesting data already from the first year, although we are still waiting for some cases to be completely reported and full analysis will only be possible when this has been submitted.

Funding

Kawasaki disease patient support group and investigators.

Public patient engagement

Kawasaki Disease Support Group.
Web: www.kssg.org.uk

References

1. Eleftheriou D, Levin M, Shingadia D, Tulloh R, Klein N, Brogan P. Management of Kawasaki disease Arch Dis Child 2014 Jan;99(1):74-83. PMID:24162006.

Researcher contacts

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Progressive intellectual and neurological deterioration in children (including Creutzfeldt - Jakob disease)

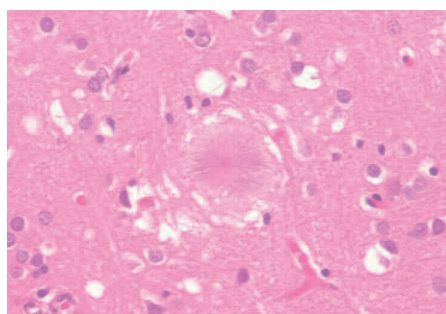
Key points

- Continuing surveillance of UK children with progressive intellectual and neurological deterioration (PIND) is important to ensure that new cases of variant Creutzfeldt-Jakob disease (vCJD) are not being missed among the numerous rare neurodegenerative disorders of childhood.
- The study provides unique information about the epidemiology of neurodegenerative diseases in UK children. Since May 1997, 3,583 children have been notified; 1,520 children have a known diagnosis other than vCJD, with over 170 different neurodegenerative conditions in this diagnosed group.
- Six cases of vCJD have been reported to the study since December 1998; four have been classified as “definite” and two “probable”; all have now died.

Summary

Active prospective surveillance of UK children with progressive intellectual and neurological deterioration (PIND) commenced in May 1997.¹ Funded by the Department of Health (England) [121/6443] it is being carried out via the BPSU in conjunction with the National Creutzfeldt-Jakob Disease Surveillance Unit in Edinburgh (NCJDSU) and Public Health England. The study strategy is to look at the broad group of rare neurodegenerative disorders affecting children, carefully examine the clinical details and determine whether there are cases of vCJD (Figure 9) amongst these PIND cases. This unique dataset provides the opportunity to detect vCJD cases and highlight the variety of PIND conditions in the UK.²

Figure 9: Florid plaque vCJD x 400 haematoxylin eosin stain



The study was extended for another year because the risk of vCJD has not gone away and, in the absence of a diagnostic test, the PIND study remains the only way to search for vCJD in children. The second national appendix survey showed high prevalence of infection with abnormal prion protein.³ Vertical transmission from mother to infant remains a possibility; there is also the possibility of transmission through dental procedures, operations and via plasma products.



PIND Research group

Methodology

Data capture uses standard BPSU methodology; details of the study protocol are available at www.rcpch.ac.uk/bpsu/pind

Surveillance period

May 1997 – April 2015 (inclusive) and is reviewed yearly.

Analysis

By March 2014, 3,583 children had been notified; 217 are still “under investigation” by their paediatricians; 1,524 did not meet the PIND definition, were duplicate or error notifications, and 139 cases remained outstanding; the majority of these were being worked-up e.g. cases prepared for the next Expert Group meeting or awaiting a visit from one of the researchers.

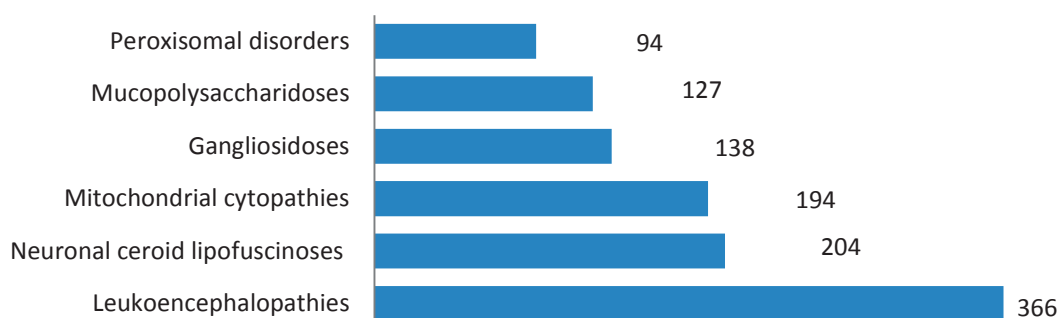
The remaining 1,703 cases were classified as follows:

Definite and probable cases of vCJD: Six cases of vCJD (four definite and two probable) have been notified - the youngest was a girl aged 12 years at onset. There were three other girls (two aged 14 years and one aged 13 years at age of onset) and two boys aged 15 years at onset. The last child who developed symptoms did so in 2000. All have now died and neuropathology has confirmed vCJD in four cases; a post-mortem was not carried out on the remaining two cases.

Children with PIND who have definite diagnoses other than vCJD: More than 170 different neurodegenerative conditions were diagnosed in these 1,520 children. In the diagnosed cases the six commonest groups are outlined in Figure 10 overleaf.

Children with PIND and no underlying diagnosis (idiopathic group): The Expert Group meet regularly to discuss this group of children, currently 177. If a “new” variant of vCJD should arise or if the paediatric presentation differed from the adult presentation, this group could include such a phenotype. However, there is currently no evidence of a “new” unrecognised disorder in this group.

Figure 10: six most commonly reported PIND diagnostic groups



Discussion

During almost 17 years of surveillance, six children presenting with vCJD under 16 years of age have been notified to the study, including four with definite vCJD and two with probable vCJD. There remains concern that more childhood cases may appear, perhaps related to underlying genotype, and children within the 'idiopathic' PIND group are under regular review. Children are still at risk of vCJD infection by blood, plasma products, surgical and dental instruments and theoretically via vertical transmission. Continued surveillance is essential as there are still many unanswered questions about this relatively new disorder – in particular, the number of children who may be incubating vCJD, the length of the incubation period and the exact nature of transmission. Meanwhile the study continues to yield unique information about the epidemiology of childhood neurodegenerative disorders in the UK.⁴⁻⁶

References

1. Verity CM, Nicoll A, Will RG, Devereux G, Stellitano L. Variant Creutzfeldt-Jakob disease in UK children: a national surveillance study. *Lancet* 2000;356:1224-7.
2. Devereux G, Stellitano L, Verity CM, Nicoll A, Rogers P. Variations in neurodegenerative disease across the UK: findings from the national study of Progressive Intellectual and Neurological Deterioration (PIND). *Arch Dis Child* 2004; 89:8-12.
3. Gill ON, Spencer Y, Richard-Loendt A, Kelly C, Dabaghian R, Boyes L, Linehan J, Simmons M, Webb P, Bellerby P, Andrews N, Hilton DA, Ironside JW, Beck J, Poulter M, Mead S, Brandner S. Prevalent abnormal prion protein in human appendixes after bovine spongiform encephalopathy epizootic: large scale survey. *BMJ* 2013; 347:f5675. doi: 10.1136/bmj.f5675

4. Verity C, Winstone AM, Stellitano L, Krishnakumar D, Will R, McFarland R. The clinical presentation of mitochondrial diseases in children with progressive intellectual and neurological deterioration: a national, prospective, population-based study. *Dev Med Child Neurol* 2010;52(5): 434-440

5. Verity CM, Winstone AM., Stellitano L., Will RG., Nicoll A. The epidemiology of progressive intellectual and neurological deterioration in childhood. *Arch Dis Child* 2010; 95:361-364

6. Smith NJ, Winstone AM, Stellitano L, Cox T, Verity C. GM2 gangliosidosis in a United Kingdom study of children with progressive neurodegeneration: 73 cases reviewed. *Dev Med Child Neurol* 2012; 54 (2): 176-182

Funding

Department of Health England.

Public patient engagement

Creutzfeldt-Jakob disease support network. Web: www.cjdsupport.net

Batten disease family association. Web: www.bdfa-uk.org.uk

Society for mucopolysaccharide diseases. Web: www.mpsociety.co.uk

ALDLife (Adrenoleukodystrophy). Web: www.aldlife.org

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APPENDIX - Publications 2013 -2014

Bacterial meningitis in Infants aged <90 days

1. IO Okike, AP Johnson, KL Henderson, RM Blackburn, B Muller-Pebody, SN Ladhani, M Anthony, N Ninis, PT Heath; the neoMen study group; Incidence, Aetiology and Outcome of Bacterial Meningitis in Infants Aged <90 days in the UK and Republic of Ireland: prospective, enhanced, national population-based surveillance; Clin Infect Dis. 2014 Jul 4. pii: ciu514 [Epub ahead of print] www.ncbi.nlm.nih.gov/pubmed/24997051

Chylothorax

2. C Haines, B Walsh, M Fletcher, P J Davis. Chylothorax development in infants and children in the UK. Arch Dis Child. 2014 Aug;99(8):724-30. www.ncbi.nlm.nih.gov/pubmed/24704707

Congenital hypothyroidism

3. R Knowles, J Oerton. An update on the British Paediatric Surveillance Unit study on congenital hypothyroidism. FootPrint - Issue 9 - March 2013. <http://newbornbloodspot.screening.nhs.uk/footprint>

Congenital adrenal hyperplasia

4. JM Khalid, JM Oerton, C Dezateux, PC Hindmarsh, CJ Kelnar, RL Knowles. Late clinical presentation of congenital adrenal hyperplasia in older children: findings from national paediatric surveillance. Arch Dis Child 2014 Jan;99(1):30-4. www.ncbi.nlm.nih.gov/pubmed/24043550

Conversion disorder

5. C Ani, R Reading, R Lynn, S Forlee, E Garralda. Incidence and 12-month outcome of non-transient childhood conversion disorder in the UK and Ireland. Br J Psychiatry. 2013 Jun;202:413-8. www.ncbi.nlm.nih.gov/pubmed/23620449

Gonorrhoea, chlamydia, syphilis and trichomonas

6. R. Reading, K Rogstad, G Hughes, G Debelle. Gonorrhoea, chlamydia, syphilis and trichomonas in children under 13 years of age: national surveillance in the UK and Republic of Ireland. Arch Dis Child. 2014 Aug; 99(8):712-6. www.ncbi.nlm.nih.gov/pubmed/24771307

Guillain-Barre Syndrome

7. C. Verity, L. Stellitano, AM Winstone et al Pandemic A/H1N1 2009 influenza vaccination, preceding infections and clinical findings in UK children with Guillain-Barre Syndrome". Arch Dis Child, doi: 10.1136/archdischild-2013. www.ncbi.nlm.nih.gov/pubmed/24585755

HIV infection & perinatal HIV exposure

8. CL Townsend, L Byrne, M Cortina-Borja, C Thorne, A de Ruiter, Lyall H, et al. Earlier initiation of ART and further decline in mother-to-child HIV transmission rates, 2000-2011. AIDS. 2014;28(7):1049-1057.
9. E Chiappini, L Galli, C Giaquinto, L Ene, T Goetghebuer, A Judd, et al. Use of combination neonatal prophylaxis for the prevention of mother-to-child transmission of HIV infection in European high-risk infants. AIDS. 2013;27(6):991-1000.
10. K Donegan, K Doerholt, A Judd, H Lyall, E Menson, K Butler, et al. Lopinavir dosing in HIV-infected children in the United Kingdom and Ireland. Pediatr Infect Dis J. 2013;32(1):45-50.
11. CE French, PA Tookey, M Cortina-Borja, A de Ruiter, CL Townsend, C Thorne. Influence of short-course antenatal antiretroviral therapy on viral load and mother-to-child transmission in subsequent pregnancies among HIV-infected women. Antivir Ther. 2013;18(2):183-92.
12. SE Huntington, C Thorne, LK Bansil, J Anderson, ML Newell, GP Taylor, et al. Predictors of pregnancy and changes in pregnancy incidence among HIV-positive women accessing HIV clinical care. AIDS. 2013;27(1):95-103.

Intussusception

13. L Samada, M Cortina-Borja, H El Bashira, A G. Sutcliffe, S Marven, J. Claire Cameron, R Lynn, B Taylor. Intussusception incidence among infants in the UK and Republic of Ireland: A pre-rotavirus vaccine prospective surveillance study. Vaccine 31 (2013) 4098- 4102

Neonatal Hypernatraemia

14. SJ Oddie, V Craven, K Deakin, J Westman, A Scally. Severe neonatal hypernatraemia: a population based study. Arch Dis Child Fetal Neonatal Ed 2013 Sep;98(5):F384-7

Toxic Shock Syndrome

15. S Adalat, T Dawson, SJ Hackett, et al. Toxic shock syndrome surveillance in UK children. Arch Dis Child 2014 May 1. doi: 10.1136/archdischild-2013. www.ncbi.nlm.nih.gov/pubmed/24790135

Vitamin K deficiency bleeding

16. A Busfield, R Samuel, A McNinch, et al. Vitamin K deficiency bleeding after NICE guidance and withdrawal of Konakion Neonatal: British Paediatric Surveillance Unit study, 2006-2008. Arch Dis Child 2013; 98: 41-47

Membership of Scientific Committee 2013

Dr Richard Reading	Chair
Dr Rachel Knowles	Medical Advisor (non-infectious disease)
Dr Iain Kennedy	Medical Advisor (infectious disease)
Mr Richard Lynn	Scientific Coordinator
Dr Robert Cunney	Royal College of Physicians of Ireland
Dr Piers Daubeney	Consultant Paediatric and Foetal Cardiologist
Ms Jacqueline Fitzgerald	Director of Research and Policy Royal College of Paediatrics & Child Health
Prof Anne Greenough	Vice President Science & Research; Royal College of Paediatrics & Child Health
Dr Kathryn Johnson	Consultant in Neonatology
Dr Simon Lenton	Consultant Paediatrician
Dr Simon Nadel	Consultant in Intensive Care
Dr Kevin Pollock	Health Protection Scotland
Dr Delane Shingadia	Consultant in Paediatric Infectious Diseases
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Dr Sam Oddie*	Consultant Neonatologist
Dr Richard Pebody*	Consultant Epidemiologist
Mrs Anne Seymour*	Patient and Carers Advisory Group

* Stepped down in 2013*



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