BPSU

British Paediatric Surveillance Unit

Annual Report 2019-2020

Surveillance of Rare Childhood Disease • BPSU •

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 the 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.

http://www.bpsu.org.uk

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Foreword

Dr Shamez Ladhini
Chair, BPSU

Normally a foreword would précis the report year, pick up highlights and thank all. This I will do, but first I think it is important to address what is becoming the greatest threat to public health in the past century. In January this year reports were coming out of China of new Coronavirus (Sars-Cov2) which had the potential to be easily spread with a mortality rate significantly higher than that of flu. By February, the World Health Organization had declared the spread of the virus, now commonly referred to as COVID-19, as a pandemic. Since then worldwide there have been over 46 million confirmed reports with over a million deaths.

In the UK public health bodies have taken a lead in monitoring the spread of the virus across the nation. The BPSU has also played a small but significant part in monitoring the disease through its surveillance of the virus in neonates. As early as the second week in March the BPSU was receiving enquiries from clinicians wanting to report paediatric cases. The BPSU along with its sister unit, the UK Obstetric Surveillance System, and Imperial College responded swiftly, setting up weekly surveillance of neonatal COVID-19 and positive expectant mothers. Public Health England (PHE) itself commenced surveillance of all children excluding neonates at the same time. Thankfully the numbers of children admitted to hospital and placed on ventilators has been low, unfortunately this is not the case of those more elderly or those who have underlying conditions.

Then in April reports started to come in of a rare presentation in children of an inflammatory syndrome, possibly as a result of COVID-19 infection. Some of the presenting features mirrored that of Kawasaki disease and Toxic Shock Syndrome - two conditions very familiar to the BPSU. A review of the known cases led to the fast tracking of a BPSU-PHE facilitated national surveillance study of paediatric multisystem inflammatory syndrome (p.25).

One other concern had been raised by clinicians and this related to the fall in numbers of children and young people presenting to Emergency Departments. The BPSU instigated a snap shot survey to identify whether there was evidence of delayed presentations of children presenting to hospital. Over a one-week period the BPSU asked all its respondents if they thought that any children presenting to hospital setting had done so due to the COVID-19 situation. Over 2,500 responses were received, many reporting what they felt to be a delayed presentation. The results included reports of nine deaths (p.12).

Over a short period the BPSU has shown its adapatabilty, its developed its methodology to undertake weekly surveillance of a particular disease and a snap-shot survey. The BPSU has worked with many agencies to fast track paperwork at unheard of pace and dealt with a huge number of case reports. We will take the knowledge learnt over this period to consider new ways to facilitate disease monitoring.

With this in mind there was concern as to how this may affect the monthly reporting system. With the research teams we are monitoring report numbers and considering any barriers to data collection. None of this work could have been undertaken without the support of the clinicians and others who report into the BPSU. With the prospect of a second wave of infection and the introduction of a vaccine, which will require monitoring, I have no doubt the BPSU will remain at the forefront of evidence gathering.

June 2019 saw the start of the BPSU 35th year of surveillance, an amazing feat! With a continued orange eCard response rate of over 90% - 'buy-in' from you the clinicians remains high. In saying that we have noted a number of respondents not returning any card over the year and we are not sure why this is. So, we will be reaching out to those clinicians to see if there are particular issues that are preventing them from completing their reporting card. We encourage all paediatricians to complete the eCard and return questionnaires. In recognition, clinicians receive a biannual revalidation certificate and, for those who report a case, they now receive a questionnaires completion certificate for inclusion in their revalidation portfolios. Our two scientific committee trainee representatives, Drs Chenqu Suo and Sarah Clarke, are also putting in motion plans to raise the awareness of the BPSU within the trainee community.

Speaking of trainees, the Sir Peter Tizard Bursary 2019 was awarded to Dr Tng Kwok from Nottingham. Tng will be undertaking surveillance of neonatal stroke. This study will also act as the pilot for the online integrated case notification and data collection platform - for the first-time reporting clinicians will be able to report cases and return clinical information in one simple unobtrusive process. The aim now is to launch the platform in Spring 2021.

The BPSU increasingly has an important role to play in the education and training of healthcare professionals and the public. Not just through the Tizard bursary but also through publications (p.33), running workshops, seminars, and now the BPSU-RCPCH webinar series. Thus far we have run webinars on Kawasaki disease, ADHD transition and most recently on nutritional rickets. Many more are planned

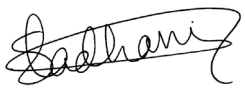
for 2021. We are also developing plans to develop the BPSU website. The launch of the Rare Disease Development Fund has got off to a good start with an educational grant from Novo Nordisk to deliver a webinar on type 2 diabetes.

Of all the surveillance undertaken last year, I wish to highlight the HIV surveillance. In 1986 a new disease was presenting itself to the world, much as the case now. The BPSU and UCL GOS Institute of Child Health responded by launching surveillance of this new disease on the very first “orange card”. Over the following 34 years the collection of data of HIV-exposed infants and children diagnosed with HIV has given the UK the most complete record of the disease epidemiology. The information helped with the development of clinical management and treatment. Such, that the transmission rate to infants born to diagnosed women is a fraction of what it was. Paediatric HIV surveillance on the orange eCard has now ceased but continues through the Integrated Screening Outcomes Surveillance Service (**https://www.ucl.ac.uk/isoss**) based at UCL GOS Institute of Child Health and funded by Public Health England’s Infectious Diseases in Pregnancy Screening Programme (**https://www.gov.uk/government/publications/integrated-screening-outcomes-surveillance-service-isoss**).

Internationally there is continued collaboration between the national surveillance units which make up the International Network of Paediatric Surveillance Units (INoPSU). Richard Lynn, our scientific coordinator, is co-chair of INoPSU and has encouraged the sharing of protocols studies to allow comparative analysis. Close co-operation over COVID-19 surveillance is taking place. Sadly,the planned INoPSU 2021 conference has been cancelled - it is hoped a virtual session will be organised.

This work is only possible because of a successful team and I would like to thank the members of the BPSU scientific committee for contributing their personal time. This year I have been working heavily on COVID-19 here at the PHE and have left Richard and Jacob to lead the Unit from their respective home offices. I think you will all agree they have done an amazing job in taking the Unit forward and supporting the research teams.

But most of all I would like to thank you the clinicians who by return the eCard and complete the questionnaires and particularly trying circumstances, without your support the BPSU would not have the impact it has.



Shamez Ladhani. Chair BPSU,
November 2020

1 How the Surveillance System Works

Background

Rare diseases and infections are a numerically important cause of illness and death and mortality in childhood. There are upwards of 8,000 rare diseases and though individually uncommon, together they affect thousands. 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 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 (PHE), University College London GOS Institute of Child Health (UCL GOS ICH) and GOSH Children's Charity. The BPSU's Scientific Committee meets five or six times a year 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 available at **http://www.rcpch.ac.uk/bpsu/apply**.

Each application requires approval from the BPSU Scientific committee, a Research Ethics Committee (REC), the Confidentiality Advisory Group (CAG) of the Health Research Authority and the Scottish Public Benefits and Privacy Panel (PBPP).

The reporting system

Surveillance is 'active' in that the BPSU office actively sends out each month an email to consultant paediatricians in the UK and Ireland. A link directs the recipient to the BPSU electronic-orange card ('eCard') (Figure 1); which is hosted securely on the UCL server. The eCard lists the conditions currently under surveillance. A set of instructions for completing the card, including case definitions of the conditions listed on the card is also circulated. When a new study begins, the mailing also includes a link to a specially produced study protocol card and other information about the study.

Participants are expected to return eCards **even if they have no cases to report** - there is a 'nothing to report' box 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.

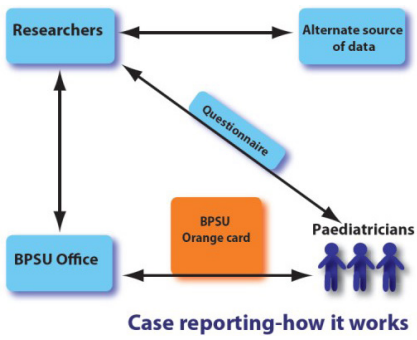
Figure 1: Orange eCard

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. Due to the need to discount duplicates a limited amount of patient identifiable data is collected. 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 other specialties have been invited to participate in the scheme. Aside from helping to improve ascertainment, such complementary data sources help to validate the surveillance system.

Figure 2: Surveillance mechanism



2 Scientific Coodinator's Yearly Review of Activities

Three studies commenced surveillance in 2019, Severe chronic fatigue - Dr Esther Crawley, Bristol; Food protein-induced enterocolitis syndrome – Dr Gary Stiefel, Leicester; and Herpes simplex disease - Dr Katy Fidler, Brighton.

Richard Lynn, Scientific Coordinator

Two studies had their period of surveillance extended: congenital rubella and progressive intellectual & neurological deterioration.

Surveillance of Juvenile-onset Systemic Lupus Erythematosus, accidental poisoning, fetal alcohol syndrome and Listeria ended in 2019. After 34 years of continuous surveillance reports of HIV-exposed and diagnosed children no longer come through the BPSU but via the Integrated Screening Outcomes Surveillance Service (ISOSS), a bespoke secure online reporting portal funded by Public Health England's Infectious Diseases in Pregnancy Screening Programme. The ISOSS team are based at UCL's GOS Institute of Child Health, London - **https://www.ucl.ac.uk/isoss**.

During 2019-20, there were 18 known publications relating to BPSU studies and nine conference oral and poster presentations (see Appendices, p.33).

Participation in the scheme during the year 2019

The overall card return compliance rate for the year 2019, calculated as a proportion of orange eCards returned, was 90.8% (44,421/48,917) a fall of 0.7% from 2018. Monthly response rates ranged from 93.3% in January to 89.2% in December with a median of 91.6%. Details of regional response rates are provided in Table 1 (overleaf). The rate has again fallen and, though still an acceptably high rate for the type of methodology, action is being taken to address this fall. Of concern is the increased in those for whom no card report was reported for over the year. We are making an even greater effort to raise awareness of the activity amongst trainees and are identifying regional champions to help promote the activity.

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 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: 712 of 4,190 (17%) receiving an eCard reported a case in 2019. 520 (12.4%) reported a single case, 4.4% (176) reported between two and four cases and 16 (<1%) reported five or more cases. The greatest number of cases reported by an individual was 50 cases.

Public and patient engagement

The BPSU is committed to wider public patient engagement (PPE) 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 introduced. These are available at **http://www.rcpch.ac.uk/bpsu/apply**. The BPSU held a success tea party led by young people. This was attended by Baroness Nicola Blackwood, then Minister of Health, and Deputy Chief Meidcal Officer, Dr Jenny Harries. Unfortunately, the 2020 tea party had to be cancelled due to the COVID-19 pandemic, as was a workshop that we had planned for the RCPCH 2020 conference.

The BPSU continues to contribute to work of patient advocacy groups such as Rare Disease UK, Cambridge Rare Disease Network, Rare Revolution Magazine and Findacure. The BPSU also works with Medics 4 Rare Diseases helping to raise awareness of rare disease research amongst student medics and trainees. More recently it has promoted the launch of the RareYouthRevolution: **http://www.rareyouth.revolution.com**

Education

With the RCPCH the BPSU has launched a rare disease webinar series. Thus far we have held webinars on Kawasaki disease (**http://www.rcpch.ac.uk/bpsu/kawasaki**), ADHD transition to adult services (**http://rcpch.ac.uk/bpsu/adhd**) and nutritional rickets (**http://www.rcpch.ac.uk/bpsu/rkt**). More will follow in 2021.

International activities

The BPSU continues to take an important role in the activities of the International Network of Paediatric Surveillance Units (INoPSU). The BPSU developed a searchable database of over 200 rare paediatric conditions surveyed by units within INoPSU (**http://www.inopsu.com**). Here you will also find information on affiliated national surveillance units, studies currently being undertaken; published papers; study protocols; and questionnaires.

Funding

BPSU is funded through grants from UCL GOS Institute of Child Health, Great Ormond Street Hospital Children's Charity, RCPCH, and Public Health England along with contributions from researchers.

Table 1: Regional Response rate 2019 and 2018

Region	% return	Rank 2019	Rank 2018
East Anglia	92.1%	8	2
Mersey	89.1%	15	17
NET	85.3%	20	20
North Scotland	96.7%	2	1
North Western	89.0%	16	15
Northern	93.6%	5	5
Northern Ireland	92.5%	6	4
NWT	93.4%	3	19
Oxford	86.4%	18	8
Republic of Ireland	86.0%	19	14
SET	88.4%	17	18
South Scotland	91.2%	12	11
South Western	91.9%	9	7
SWT	91.9%	10	13
Trent	90.1%	14	16
Wales	97.5%	1	6
Wessex	90.6%	13	10
West Midlands	92.5%	7	9
West Scotland	91.7%	11	12
Yorkshire	93.0%	4	3

Figure 3: Regional Response rate 2019

Table 2: Outcome of follow-up of the cases reported in 2019 for conditions under surveillance

Condition under surveillance	Date when reporting began	Valid reports	%	Duplicates	Errors	(D&E) %	Not yet known	%	Total
CRU	Jun-91	93	45	42	67	53	4	2	206
PIND	May-97	2,316	53	566	1,135	39	389	9	4,406
BPD	Jul-17	154	47	18	67	26	90	27	329
LUP	Sep-17	155	67	24	38	27	16	7	233
LIS	Oct-17	18	64	7	2	32	1	4	28
POI	Jul-18	32	28	4	50	47	30	26	116
FAS	Oct-18	63	43	6	32	26	47	32	148
ICH	Nov-18	19	35	12	21	60	3	5	55
SC	Nov-18	23	52	6	3	20	12	27	44
FPIES	Jan-19	91	47	7	74	42	22	11	194
CFS	Feb-19	100	50	1	21	11	80	40	202
HSV	Jul-19	30	**35**	**18**	**7**	**29**	**31**	**36**	**86**
Total		**2,651**	**50**	**632**	**1,308**	**36**	**753**	**14**	**5,344**

CRU Congenital rubella
PIND Progressive intellectual and neurological deterioration
BPD Bronchopulmonary dysplasia
LUP Juvenile-onset systemic lupus erythematosus
LIS Invasive listeria infection
POI Accidental poisoning
FAS Fetal alcohol syndrome
ICH Ichthyosis
SC Sydenham's chorea
FPIES Food protein induced enterocolitis syndrome
CFS Severe chronic fatigue syndrome
HSV Neonatal herpes simplex virus

3 Surveillance Studies Undertaken in 2019-20

Once again individual reports have concentrated on the summary of the condition and on the analysis. General methodology information is contained in the study protocols and can be found at https://www.bpsu.org.uk. 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. The BPSU would like to thank all those paediatricians who have returned cards, reported cases and completed the questionnaires.

Accidental poisoning

Key points

- Completion of surveillance in July 2019 with 116 reported cases, 32 confirmed cases meeting the surveillance definition with one excluded as not meeting the analytic case definition.
- One reported death related to an accidental poisoning event.

Summary

Accidental or unintentional poisonings involve people poisoning themselves or others without wanting to cause harm. In children it is a common reason for attendance at healthcare providers. Children, especially under the age of five, have an inquisitive nature, and frequently put things found in their environment into their mouths. Teenagers involved in risk-taking behaviour by taking illicit drugs or alcohol are also in danger of unintentionally poisoning themselves.

Serious consequences following accidental poisoning are rare in young children. The World Health Organization and UNICEF state that child resistant packaging is one of the best-documented successes in preventing accidental poisoning of children. A child resistant package requires a special 'trick' to open it which is deemed too complicated for most young children to work out. Within UK legalisation, certain household substances such as household bleach, some toiletries and gardening products are poisonous when swallowed or inhaled, or can cause skin corrosion, must be sold in child resistant packaging. At present only medicines that contain aspirin, paracetamol or more than 24mg of iron must legally be in child resistant packaging. Despite this a number of children continue to suffer significant harm from accidental poisoning, with five to ten deaths each year and approximately 50 children admitted to intensive care within the UK.[1,2] In particular, a single dose unit of certain adult medications can be fatal to a toddler.[3] Most of these are in blister packs, which are not subject to current child resistant closure legislation. Accidental poisoning is essentially an avoidable problem and, as a result, remains an important public health issue.

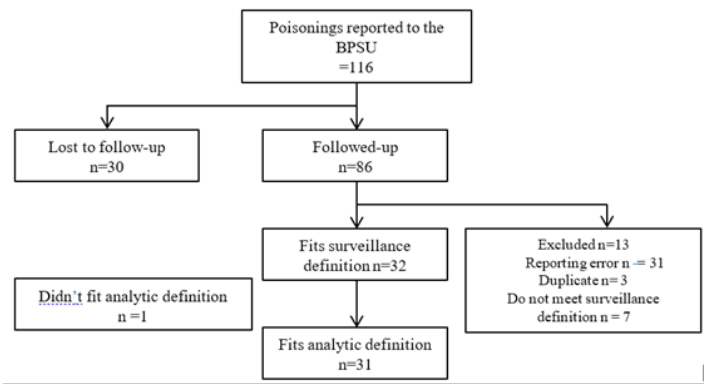
This is the first study in the UK and Republic of Ireland to estimate the incidence and identify the circumstances surrounding serious accidental poisonings in children. The study will look at poisonings resulting in death, or signs and symptoms needing significant monitoring or support. By identifying predisposing factors, in particular for specific substances that frequently cause significant harm, it is hoped that these can help improve medication safety. For example, reducing harm by legislation around more robust child-resistant packaging.

Dr Elizabeth Starkey

Surveillance period

July 2018 - July 2019 (inclusive).

Figure 4: Cases reported between July 2018 to July 2019 in the UK and Republic of Ireland



Methodology

Data capture uses standard BPSU methodology. Details of the study protocol are available at **http://www.rcpch.ac.uk/bpsu/poisoning**

Following data collection, further information on number of deaths by accidental poisoning during the study time-period is being obtained from the Office of National Statistics. Data from the child death overview panels has yet be sourced due to the changes in child death reporting from April 2019.

Further information on admissions to PICU with accidental poisoning via the PICAnet database during the study period is also being obtained. These sources are not to provide additional case reports but to check the accuracy of case reporting.

Analysis

Over the surveillance study period, 116 cases have been reported to the study group (see Figure 4). Thirty-two cases met the surveillance case definition but one case was excluded after review of the questionnaire as it did not meet the analytic case definition of a poisoning severity score of greater than two. One case resulted in death during their hospital admission.

The European Association of Poisons Centres and Clinical Toxicologists (EAPCCT) poisoning severity score (PSS) is a classification scheme for cases of poisoning in adults and children. This scheme is used for the classification of acute poisonings regardless of the type and number of agents involved. It takes into account the overall clinical course following a a poisoning event. One case was excluded from the analytic case definition as the child had a poisoning score of one. Table 3 shows the PSS scores of the included cases.

Table 3: European Association of Poisons Centres and Clinical Toxicologists (EAPCCT) poisoning severity score (PSS) of cases reported, n=31

Severity grades	N
None: No symptoms or signs related to poisoning	0
Minor: Mild, transient and spontaneously resolving symptoms	0
Moderate: Pronouced or prolonged symptoms	14
Severe: Severe or life-threatening symptoms	16
Fatal: Death	1

Of thirty-one cases, 21 cases (68%) were male and 10 cases (32%) were female. Twenty-seven cases (87%) were white, with three from other ethnic groups. In one case the ethnicity was not reported.

From the substances involved, 13 (42%) were medications, 12 (39%) were illicit drugs or alcohol, five (16%) were household or industrial products, and one (3%) was carbon monoxide poisoning. Of the thirteen medication poisonings, five were opioid related, five were psychiatric medications and one an ophthalmology treatment. One was an over the counter medication and one including multiple prescription medication. Of the illicit substances, eight (25%) were from cannabis or synthetic equivalent and three were related to the amphetamine class. Table 4 highlights where these accidental poisonings took place.

Table 4: Location of the accidental poisoning, n=31

Location of accidental poisoning	N
Home	17
Other home, family	10
Other home, non-family	2
Other	1
Unknown	1

Of the 25 substances where medications, illicit drugs or alcohol were involved, nine were the parents', seven it was unclear, and five were relatives. In three cases the information was missing and one case was another carer. With regards to the 13 medication cases, six were in their original packaging (blister pack or plastic bottle), two were loose and one in a pill box. and in four it was unknown.

Symptoms based on the poisoning event were categorised depending on how they presented in children and young people. The most common were neurological (n=28) and respiratory (n=18) symptoms, followed by cardiology (n=13), metabolic (n=10) and gastrointestinal (n=7). Other symptoms were collectively noted in eye, renal, muscle, skin liver and blood systems (n=10).

Discussion

Our results, despite relatively small numbers, highlight that accidental poisoning is a preventable condition, with most having no long term sequelae. Significant poisoning can cause serious symptoms requiring various treatments and interventions and rarely result in an death.

Despite the fact that full data analysis has yet to be finalised, none of the medications reported in this study were in child resistant packaging. The most common medication causing significant harm were narcotic or psychiatric medications with most being in their original blister packaging. Despite small numbers, this study demonstrates that further legislation around child resistance packaging with more medications is required to prevent harm and even death in younger children.

A large number of poisonings occurred within the family or a close relatives home highlighting a key public health message around medicine and drug safety within family homes.

Public and patient engagement

Child Accident Prevention Trust
Web: **https://www.capt.org.uk**

References

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Acknowledgements

The study team are grateful to the Research and development department at Derby hospital, in particular Rachelle Sherman, for her guidance and support.

Funding

This study is funded by Sir Peter Tizard Research Bursary.

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Congenital rubella

Key points

- Between April 2019 and March 2020 there have been no reports of congenital rubella syndrome to the BPSU.
- Since 2005, 13 congenital rubella births were reported in the UK.
- Congenital rubella syndrome can occur when a women contracts rubella during the first trimester of pregnancy. It can cause deafness, blindness and heart defects in the fetus, amongst other symptoms.
- The most effective intervention to prevent congenital rubella syndrome is Measles, Mumps and Rubella (MMR) vacinations prior to conception.
- Antenatal screening for rubella susceptibility was discontinued in England in April 2016.
- Rubella is a notifiable disease and is monitored by Public Health England.

Summary

Rubella has been a notifiable disease[1] since 1988 and is monitored by Public Health England (PHE) rubella surveillance programme team, part of the National Infection Service based at PHE Colindale. The Integrated Screening Outcomes Surveillance Service (ISOSS), part of Public Health England's Infectious Diseases in Pregnancy Screening (IDPS) Programme,[2] maintains national surveillance of congenital rubella (CR) cases in the UK. This remains important following the discontinuation of screening for rubella susceptibility in pregnant women in England in April 2016.[3]

The IDPS programme commission the BPSU to provide case notifications on CR in the UK to the ISOSS team based at UCL's Great Ormond Street Institute of Child Health. The team liaise with the IDPS programme and the rubella surveillance team on any notifications. Case data are then collected directly from paediatricians in by the ISOSS team. ISOSS collects patient data under legal permissions granted to PHE under regulation 3 of the Health Service (Control of Patient Information) Regulations 2002. The ISOSS team then conduct enhanced data collection on any confirmed case to review with the IDPS and NIS team to ascertain contributory factors and inform and evidence any potential policy or programme reviews.

Surveillance period

January 1990 and is reviewed yearly.

Methodology

Data capture uses standard BPSU methodology; details of the protocol are available at **http://www.rcpch.ac.uk/bpsu/congenitalrubella**

ISOSS team

Analysis

Between April 2019 and March 2020 there have been no reports of CR to the BPSU.

Since active surveillance was established 30 years ago, 68 children and four stillborn infants with confirmed or compatible CR have been born and reported; 54 (76%) reported through the BPSU notifications (Table 5).

Table 5: Confirmed and compatible congenital rubella births in the UK and Ireland 1990 to March 2020

	Primary Source of notification		
Year of birth	**BPSU**	**Other**	**Total**
1990-94 * ^	22	10	32
1995-99	12	4	16
2000-04 *	10	1	11
2005-09 *	4	2	6
2010-20 *	6	1	7
Total	**54**	**18**	**72**

*** includes a stillborn infant**
^ includes a set of triplets, one of whom was stillborn

Since 2005 there have been 13 confirmed reports of CR. None of the mothers was UK-born, and none had a previous pregnancy in the UK. Seven of the women acquired their infection abroad in early pregnancy and six were exposed to rubella in the UK.

Discussion

Very few cases of CR have been reported in the last decade, with none since 2018. Most reports concern infants with neonatal symptoms who also had serious rubella-associated defects identified at birth or soon afterwards. In the last 15 years, only half of the maternal infections were acquired in the UK. Pregnant women may enter the UK having acquired infection in early pregnancy elsewhere, and susceptible women resident in the UK who travel abroad during early pregnancy may also come into contact with rubella.

All health professionals, particularly paediatricians, those working in primary care and antenatal care, or with refugees or other recent migrants, must continue to be aware of the potential serious implications of rash or rash illness in early pregnancy and promote the Measles, Mumps and Rubella (MMR) vaccine as the most

effective intervention to prevent CR.[4] Updated PHE guidelines for the management of viral rash in pregnancy and a quick reference guide are available at **https://www.gov.uk/government/publications/viral-rash-in-pregnancy**

Since cessation of rubella susceptibility screening in pregnancy in 2016 there have been no cases of congenital rubella reported where the mother acquired rubella in the UK. To achieve continued population level control of rubella, the key action is still MMR vaccination. The measles and rubella elimination UK strategy 2019 (**https://www.gov.uk/government/publications/measles-and-rubella-elimination-uk-strategy**) focuses on four core components required to maintain elimination of measles and rubella:

1. Achieve and sustain ≥ 95% coverage in the routine childhood programme.
2. Achieve ≥ 95% coverage with 2 doses of MMR vaccine in older age cohorts through opportunistic and targeted catch-up.
3. Strengthen measles and rubella surveillance.
4. Ensure easy access to high-quality, evidence-based information

Integrated Screening Outcomes Surveillance Service

ISOSS builds on the methodology of the National Surveillance of HIV in Pregnancy and Childhood (NSHPC).[6] ISOSS is commissioned to conduct surveillance of the three infections screened for in pregnancy: HIV, syphilis and hepatitis B, as well as to monitor any cases of congenital rubella (**https://www.ucl.ac.uk/isoss**).

HIV surveillance

All pregnancies to women living with HIV, their infants and any children diagnosed with HIV (<16 years) are reported to ISOSS using a bespoke secure online portal. This data has been collected for over 30 years and provides a unique comprehensive population-level surveillance that informs national guidelines and policy, as well as supporting collaborations more widely. Bi-annual slides produced by ISOSS are made freely available providing an up-to-date picture of HIV in pregnancy and childhood in the UK. Currently nearly 90% of women living with HIV who become pregnant are already aware of their status, the majority of women deliver with undetectable viral load and the 2015-16 vertical transmission rate stands at under 0.3%.[7] ISOSS also conducts an enhanced data collection of all HIV vertical transmissions in UK-born children; all cases are included in a national review by a Clinical Expert Review Panel (CERP). The findings are fed back to the IDPS Programme to evidence and inform future service provision.

Syphilis surveillance

Active surveillance of congenital syphilis in the UK by the ISOSS Team commenced in 2019. All cases of congenital syphilis (confirmed or suspected) born in the UK are reported to ISOSS and have been included in a review by a CERP in order to inform national guidelines and policy. Since 2020 data has been collected on infants born to syphilis screen positive mothers who required paediatric follow-up in line with British Association for Sexual Health (BASHH) guidelines and birthplan. Data collection forms are designed to support the HIV (BASHH) Syphilis Birth Plan (**https://www.bashhguidelines.org/media/1196/syphillis-bp_print_2016_p3.pdf**)

Hepatitis B Surveillance

Surveillance of infants exposed to hepatitis B in pregnancy and children with hepatitis B will commence in 2021.

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7. **https://www.ucl.ac.uk/integrated-screening-outcomes-surveillance/resources/biannual-data-update-slides**

Funding

Public Health England's Infectious Diseases in Pregnancy Screening (IDPS) Programme.

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Delayed presentations to hospitals - snap-shot survey

Summary

The COVID-19 pandemic led to concerns that children are, for various reasons, presenting late to emergency departments. This has led to delayed diagnosis and hence a delay in treatment. Such presentation delays are well known, but they appear to have increased during this pandemic. Unfortunately, there have been reports of increased morbidity and mortality as a result. This increase in delay may be due to the warnings with regards staying home and also nervousness on behalf of parents who may worry about their child catching COVID-19 whilst at hospital. This survey aimed to give a snap-shot of the frequency of such a delay.

Based on the results of the survey, a letter was published in Archives of Disease in Childhood and an opinion piece was published in the British Medical Journal - links to which can be found below.[1,2]

Surveillance period

April 2020

Methodology

The BPSU received several queries from paediatricians who were concerned about late presentations of children because of lockdown and public messages to “stay at home”. To assess the extent of the problem, a one-off email was sent in April 2020 covering the ‘last 14-days’ asking three questions:

1. Are you working on the front line?

2. Have you seen any children with delayed presentations?

3. In your opinion, do you think they are COVID response related?

No details on individual children were collected.

The findings from the survey helped to raise awareness of the issue of delayed diagnosis and assess whether a more in-depth study was required to help address the issue.

Dr Shamez Ladhani

Details of the study protocol are available at **https://www.rcpch.ac.uk/bpsu/delayed-presentations**

References

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2. Ladhani S, Viner RM, Lynn RM, Baawuah F, Saliba V, Ramsay M. The unintended consequences of COVID-19 in children. *BMJ* In press 2020

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Fetal alcohol syndrome

Key points

- 13 month surveillance period has ended. To date 148 cases have been reported and 48 classified by the expert group as probable / confirmed.
- The results suggest that many paediatricians in the UK are not familiar with the diagnostic criteria for fetal alchohol syndrome.
- Vulnerable children in the UK are being failed as a result. Education across all professional groups is vital if we are to improve the lives of affected children and families.

Summary

Fetal Alcohol Syndrome (FAS) is a rare condition which occurs when a developing baby is exposed to alcohol in the womb.

The exposure of an unborn baby to alcohol can then affect the development of the brain, leading to challenges in learning and development. Alcohol can also affect the development of other parts of the unborn baby's body, particularly the face. The exact amount of alcohol that cause FAS is unknown but FAS can be completely prevented by avoiding the use of alcohol in pregnancy.

In the UK and Republic of Ireland little is known about the exact numbers of babies and children affected by FAS and the services they require to support them throughout their childhood. The aim of the study is to answer both of these questions and ultimately improve the treatment and support we provided for babies, children and families affected by FAS.

A significant study outcome alongside the incidence data is the evidence that there is a need to educate paediatricians about the diagnostic criteria for FAS. Due to a lack of awareness and understanding of the condition the diagnosis of FAS is frequently missed and children can remain undiagnosed or wrongly diagnosed well into their teenage years hence we have included children aged 0-16.

Earlier diagnosis in affected babies and children will allow individual care pathways to be created for each affected child with the appropriate services to support them.

This study has not investigated the wider condition known as Fetal Alcohol Spectrum Disorder (FASD). FASD is an umbrella term for several diagnoses that are all related to prenatal exposure to alcohol. These are: Fetal Alcohol Syndrome (FAS); Partial Fetal Alcohol Syndrome (PFAS); Alcohol Related Neuro-developmental Disorder (ARND); and Alcohol Related Birth Defects (ARBD).

FAS team

Surveillance period

October 2018 - October 2019 (inclusive).

Methodology

Data capture uses standard BPSU methodology. Details of the study protocol are available at **http://www.rcpch.ac.uk/bpsu/fas**

Analysis

As of May 2020 there have been 148 notification of which nine were duplicates. Follow-up contact with reporting clinicians has been made for 113 (>80%). Contact with the remaining 26 clinicians has not been possible. Only two cases were reported from the Republic of Ireland and in neither case has it been possible to contact the referring paediatrician.

Of the 113 reports for which further communication with the reporting clinician has been possible, reports of 46 children were withdrawn. The reasons for withdrawal were: data for 10 cases could no longer be provided, 34 cases were thought not to fit the case definition after reconsideration by the reporting clinician, and in two cases the clinician could not be traced after initial contact. This left 67 submitted data forms and of these 19 more were excluded as they did not fit the surveillance definition, leaving 38 probable cases and 10 confirmed cases.

Of these probable and confirmed cases, 21 were female and 27 male. Only four of these 48 were living with their birth mother at the time of diagnosis. Three of the four cases living with their birth mother had social services involvement.

Of the 44 cases not with their birth mother, 13 had been adopted, 14 were in foster care, 14 with family members and for three cases information was not provided.

Discussion

The response following receipt of the questionnaire has been slow but ultimately satisfactory.

The study has faced a number of significant challenges; the number of withdrawn cases upon receipt by clinicians of the full questionnaire and a reminder of the diagnostic criteria has been high. This reflects a broad lack of understanding of the case definition of FAS, resulting perhaps from insufficient education and confusion with the broader term; FASD.

The BPSU committee and study team anticipated the issues around facial features, and the measurement and centile plotting of these (required for a formal, internationally recognised diagnosis of FAS) and hence created both a probable and confirmed case definition; where in the probable cases facial features were present but where centiles had not been recorded.

Upon further discussion at the study's independent expert group meeting in March 2020 it became clear the measurement of palpebral fissures was frequently inaccurate and hence within the probable group a sub-group was created. This group included cases where facial features were present & centiles were available but where the numeric measurement of palpebral fissures was inaccurate and inconsistent with the description given.

Clarity about diagnostic criteria has been provided in Scotland by the SIGN (Scottish Intercollegiate Guideline Network) Guideline: Children and young people exposed prenatally to alcohol. NICE (National Institute of Clinical Excellence) guidance is currently being created and will provide clarity for clinicians.

There is a need for widespread, effective education of clinicians across the UK in order to address this and will form a key part of the outcomes and dissemination of the study.

Better education and understanding of FAS and associated diagnoses by clinicians will lead to earlier diagnoses and better packages of care for this incredibly vulnerable group of babies and children where >90% are not living with their biological mother at the time of diagnosis.

It is the intention of the study group to collaborate with the International Network of Paediatric Surveillance Units (INoPSU) in comparing data on FAS across national surveillance networks, and perhaps, following clarity about terminology and the publication of national guidance; undertake national surveillance for FASD.

Public and patient engagement

FAS Aware UK
Web: **https://www.fasaware.co.uk**

Funding

This study is jointly funded by The Halley Stewart Trust, Public Health England and the Seedbed Community Trust.

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Food Protein Induced Enterocolitis Syndrome

Key points

- Two hundred and four case reports were received of which 98 met the surveillance case definition
- We estimate an incidence for England of 0.006%, significantly below estimates from other countries.
- Mean diagnostic delay was 14 months and cases were clustered around centres with paediatricians with allergy training.
- Preliminary analysis suggests significant under reporting and under recognition by non-specialist paediatricians.

Summary

Food Protein Induced Enterocolitis Syndrome (FPIES) is a delayed type of food allergy which leads to repeated vomiting and other gastrointestinal symptoms up to several hours after a problem food (or baby formula) is consumed.[1] It can have serious consequences leading to shock and metabolic acidosis.

Delays in diagnosis are frequent, since many frontline healthcare providers are not aware of the condition and presenting clinical features can mimic more common paediatric presentations, such as sepsis and surgical abdominal emergencies. Consequently, children with FPIES often have multiple episodes, additional investigations and hospital admissions before a diagnosis of FPIES is made.[2-4]

There is very limited data on FPIES in the UK and the Republic of Ireland.[5] International studies suggest the incidence is between 0.015 – 0.7%.[1] This study is intended to improve our knowledge of the incidence, clinical presentation and management as well as improve awareness amongst paediatricians. This has the potential to benefit patients and families through quicker diagnosis and instigation of effective management of FPIES.

Surveillance period

January 2019 - January 2020 (inclusive).

Methodology

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

In addition, case ascertainment was maximised by:

1. Contact with regional children's allergy clinics to ensure completeness of reporting half-way through the surveillance at 6 month and at the end of reporting after 13 months of BPSU surveillance to ensure that all cases had been included from larger centres.

Dr Mich Lajeuensse & Dr Gary Stiefel

2. Asking patient support groups FPIES UK and Allergy UK to advertise the study on their website and to provide a link to the public information leaflet.

Analysis

Two hundred and four cases were reported, of which 98 (48%) met the final epidemiological case definition, giving an incidence in England (93 cases) of 0.006%. We were unable to calculate incidence in other participating countries, as only five further cases were reported. 53.1% of reported cases were male, 73.5% were single food triggered FPIES, and 26.5% had multiple food triggers. Multiple food trigger FPIES cases were more likely to be female (65.4% female versus 34.6% male, p =0.03). The most common primary food trigger was milk (32.7%), followed by fish (15.3%) and egg (14.3%).

Diagnosis and management: Mean age at first reaction was 6.6 months (range 1 to 20 months). All cases of multiple food triggered FPIES reacted before nine months, as opposed to 70.6% of single food trigger FPIES (p =0.003). 51% of cases were admitted or observed during an FPIES reaction. In 42.2% of cases the implicated food had been ingested three or more times prior to diagnosis. Following diagnosis food exclusion was advised to all patients, this was exclusion of causative foods 81.6%, with wider exclusion recommended in the other 18.4%.

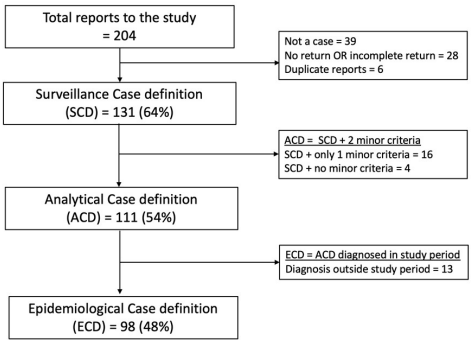
Under-recognition and under-reporting: In 74.2% of cases the diagnosis of FPIES was made by a health-care professional with expertise in allergy.

- Mean diagnostic delay from first reaction of 14.2 months (range 0 to 144 months). Diagnostic delay was greater for multiple trigger FPIES compared to single-trigger FPIES (p=0.02).
- Average distance from home to diagnosing hospital was 10.9 miles. Patients in higher socioeconomic quintiles travelled further for review (p=0.006)

Discussion

The incidence in England was much lower than expected at 0.006%. The Australian experience

Figure 5: Cases reported between January 2019 to January 2020 in the UK and Republic of Ireland



identified 230 children over a 29-month period. Therefore, assuming a similar incidence of FPIES in the UK population, there would be 239 cases in 13 months as opposed to the 128 meeting our case definition.[6] Cases were clustered around centres with specialist allergy services suggesting under reporting elsewhere. Families travelled less than 20 miles for a diagnosis and although longer distances were travelled according to socioeconomic status, this was still within the regional area. There was a significant delay in diagnosis of 14 months on average. In consequence most children suffered several reactions before the diagnosis was made.

The primary food triggers were milk, egg and fish common weaning foods in the UK. Most FPIES is related to local weaning foods a triggers.[7] A quarter of cases reacting to multiple foods in keeping with rates of multiple FPIES reported elsewhere.[8]

The BPSU has provided the first prospective epidemiological study of this rare but serious type of food allergy. We hope that the national coverage that the survey provided would have increased awareness amongst paediatricians. Our results suggest that better recognition of FPIES is needed in primary and emergency care to ensure prompt diagnosis and referral for specialist advice.

Funding

This study has been funded by Midlands Asthma and Allergy Association (MAARA).

Public patient engagement

Allergy UK
Web: **https://www.allergyuk.org**

FPIES UK
Web: **https://www.fpiesuk.org**

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Ichthyosis in neonates

Key points

- A total of 21 confirmed cases of ichthyosis were reported from November 2018 to May 2020.
- The estimated incidence is 2.65 per 100,000 live births in the United Kingdom and Northern Ireland.
- Two deaths have been seen.
- Overall, the number of reported cases of collodion membrane were higher than expected (approximated to be 12 every year so 18 in one and a half years) but there were no reports of harlequin ichthyosis.

Summary

Ichthyosis is a group of incurable genetic conditions with abnormally thick, scaly skin. The severe types, collodion membrane (CM) and harlequin ichthyosis (HI), are present at birth and can cause problems with breathing, feeding, movement, eye closure and temperature control. Some babies do not survive, most deaths occurring at or within days of birth. Babies with HI and CM are very rare. There is no proven correct treatment so practice varies, for example some babies remain in the neonatal intensive care unit for weeks whilst others are nursed within a ward or home setting.

Unlike for several rare diseases (for example the skin-fragility disorder epidermolysis bullosa) there is no national specialised service, even though the morbidity and mortality are similar. Families often rely on each other for advice, for example via the Ichthyosis Support Group (ISG). Our study objectives are to estimate the number of new cases per year in the UK, management of these babies including age on discharge from hospital, factors relating to the health of mother and baby that might affect the outcome and death rate and significant problems (such as growth and skin care) within the first year. Information will be made available to medical professionals and to parents via the ISG, hoping to improve the care of babies with ichthyosis. Our data will also be used to support an application to NHS England for a Highly Specialised Service for ichthyosis.

Surveillance period

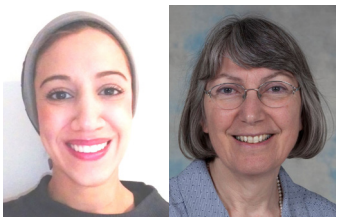
November 2018 - November 2020 (inclusive).

Follow-up period: Until November 2021.

Methodology

Data capture uses standard BPSU methodology. Details of the study protocol are available at **http://www.rcpch.ac.uk/bpsu/ichthyosis**

Some cases may not be reported if they have not been seen by a paediatrician after birth e.g. those who are still born or die immediately after birth. Therefore, data will be cross checked with National Congenital Anomaly and Rare Disease Registration Service (NCARDRS) once appropriate approvals are received.



Dr Fozia Roked & Professor Celia Moss

Though not collecting data from the Republic of Ireland, a study using similar methodology is being run through the Irish Paediatric Surveillance Unit. This will allow data comparison.

We have representative photographs of collodion membrane and congenital ichthyosis available for reference online at **https://www.rcpch.ac.uk/sites/default/files/2018-12/representative_photographs_of_collodion_membrane_and_harlequin_ichthyosis_0.pdf**

Analysis

The data reported in this study includes cases reported from November 2018 to May 2020. Twelve month follow-up questionnaires yet to be analysed. To date 67 case reports have been received, 22 were errors, two were duplicates leaving 21 babies that were confirmed cases. Data on 12 is awaited. These numbers will change as further reports are received.

The following data is based on the 21 confirmed cases, unless stated otherwise. Five babies were preterm which means being born before 37 weeks. The median gestation was 37+3 weeks. Nine cases were girls. In terms of ethnicity 28% (n=6) of babies were from ethnic groups other than white. There was a family history of consanguinity in only one case. There was a family history of ichthyosis in four cases.

In terms of neonatal intervention: 71% (n=15) babies were placed in an incubator, 52% (n=11) babies had artificial feeding initially (either nasogastric feeds or total parental nutrition), three babies were intubated and four received another form of respiratory support. Seven babies required central venous access. 52% (n=11) babies received antibiotics, all babies received topical emollients and 76% (n=16) eye lubricants.

Discussion

The data shows that babies with ichthyosis tend to be preterm and are there was a high percentage of BAME ethnicity cases. The neonatal outcome was variable; half of patients were treated for sepsis and there were two deaths. The remainder were discharged within 30 days. Most babies were placed in an incubator and more than half required feeding support.

The reported number of cases of collodion membrane is above expected figure but there have been no reports of harlequin ichthyosis. We are uncertain whether zero cases in one year truly represents the rarity of this condition or whether we are missing cases. Therefore, we raised awareness by presenting our interim data at the British Society of Dermatologists annual conference and circulating messages on twitter. However, there have still been no reports of harlequin ichthyosis in UK since 1st November 2018. It may be that some affected babies have been still-born or died soon after birth: we are hoping that the NCARDRS data will reveal any such cases.

Public and patient engagement

Ichthyosis Support Group
Web: **https://www.ichthyosis.org.uk**

References

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Acknowledgements

The ichthyosis team would like to thank dermatology interns Kerry Dewsbery (September 2018-19), Helen Light (September 2019-20) and Sasha Reed (September 2020-21). Lastly, the study team would like to acknowledge Dr Murtaza Khan for designing the electronic questionnaires.

Funding

This study is funded by Great Ormond Street Children's Hospital Charity-SPARKS.

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Invasive listeria infection

Key points

- Thirty-nine patients have been notified through BPSU and Public Heath laboritories over a two year period, of which 25 were confirmed cases.
- Of the 25 cases for which full data are available all were infected within the first two weeks of life. 24 in the first day and one on the fourteenth day.
- Case fatality rate was 8%.

Summary

Listeriosis is a rare bacterial infection that can cause severe disease in young babies, pregnant women, people with weakened immune systems and the elderly. Pregnant women can become infected by eating contaminated food, such as fresh cheese and unpasteurised milk, and may then pass on the infection to their unborn babies. This can cause miscarriage, premature birth, death or severe disease in babies, often leaving the baby with long-term disabilities. Listeria may be becoming more common, particularly in ethnic minority groups.

This study aims to establish how common this infection in babies and at what age babies are getting the infection. It will also establish how the babies are treated and what is the mortality and the long term consequences of the disease.

The importance of the study is to inform the national policy on antibiotics: current national guidelines (e.g. the NICE guidelines), advising doctors on antibiotic treatment for babies younger than three months, recommend an antibiotic combination that will treat listeria infection. Unfortunately, this does not happen everywhere and some babies with possible listeria infection do not get the right antibiotics. Conversely, we know from a recent national study of meningitis that only babies younger than one-month of age had listeria meningitis. This raises the possibility that thousands of babies between two and three months of age may be receiving antibiotics that are not needed. This study will provide data to establish whether a more targeted policy is required.

Dr Stefania Vergnano

Surveillance period

September 2017 - September 2019 (inclusive).

Follow-up period: Follow-up questionnaire at 12 months after initial diagnosis, ending in September 2020.

Methodology

Data capture uses standard BPSU methodology. In addition, microbiologists in each country will continue reporting cases through established routine public health laboratory reporting systems. The reference public health laboratories are regularly contacted, and the cases reported will be compared with the cases detected through the BPSU orange eCard system to ensure all cases are detected.

In England, the Office for National Statistics (ONS) and the Hospital Episode Statistics (HES) will also be contacted to ensure all admissions and deaths due to listeria in infants less than 90 days are captured.

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

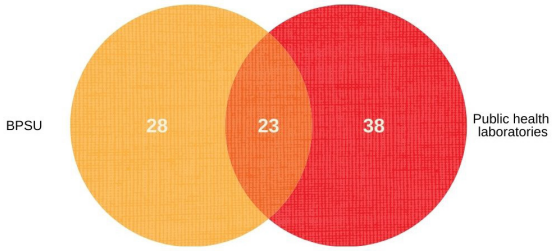
Analysis

Over the surveillance period there have been 28 BPSU, 38 public health laboritories' notifications, and two cases identified through the UK Neonatal Infection Network (neonIN), a voluntary UK neonatal surveillance network - see Figure 6.

After deduplication and verification of the BPSU cases, eight were excluded as duplicate, three did not meet the study definition.

From the neonIN database, one was a duplicate of the BPSU reports. Data from public health laboritories included five miscarriages/stillbirths, one older infant and 15 duplicates with the neonIN and/or BPSU data. Of the remaining 25 cases were

Figure 6: All Cases reported by BPSU and Public health laboratories



verified and complete information were collected, for two information is pending.

The study ran for 25 months and will continue collecting follow up data until September 2020. To this date 25 cases have been verified and full information are available. The incidence of Listeria in young infant was 1.5 per 100,000 live births, 17 infants were preterm (<37 weeks gestation). The median Birth weight was 2300g (range 605-3742g), gestational age was 34 (range 24-40) and 14 were girls. All but one infant presented within the first 24 hours from birth, one on day 14. Two babies died giving a case fatality rate of 8% (n=2/25). Of the affected babies, 10 were white, three were African, seven Asian, two of mixed race: white and black African, and three unknown.

Discussion

The survey is showing a low incidence of listeria (1.5 per 100,000 LB). Case fatality was 8%. All but one case presented within the first 24 hours of birth and at two weeks of age making a very compelling case for changing the national empiric antibiotic therapy for infants under 90 days to include amoxicillin only for the first month. Of the babies with known ethnic background 50% were from either Asian or African or mixed background. Data about medium term follow up are awaited.

Following from this study it is realistic to expect that the current NICE guidelines would need amending.

It is also hoped that an international comparison with other surveillance units will be undertaken. – A Canadian and a Swiss group are collecting very similar data and the aim is to write a joint publication to compare incidence, age of infection and outcome of Listeria infection in young infants.

Funding

This study has been funded by St. George's University of London.

Public and patient engagement

Bliss.
Web: **https://www.bliss.org.uk**

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Temple Street Children's Hospital, Dublin:
Dr Robert Cunney
University Hospital Galway:
Professor Martin Cormican

Juvenile-onset Systemic Lupus Erythematosus

Key points

- Surveillance of juvenile-onset systemic lupus erythematosus through the BPSU with additional reporting by adult clinicians including dermatologists, nephrologists and rheumatologists commenced September 2017 and completed September 2019
- Collection of one year follow-up data will complete September 2020
- Over the surveillance period 253 cases have been reported. After exclusions 133 cases are included in the analysis to date. A further 17 cases from year two still have clinical data pending.

Summary

Juvenile-onset systemic lupus erythematosus (JSLE) or 'childhood lupus' is a rare disease where the immune system attacks many parts of the body. JSLE can be variable in how it presents, with some children having a mild disease and others having very severe disease (e.g. developing kidney failure or brain abnormalities). It is not known exactly why JSLE develops. It is likely to be a complicated combination of genetic and environmental factors. It is also not known how many children and young people in the UK and Ireland develop JSLE. This study will help us understand how many children are affected by JSLE and which medical teams look after them. Children with JSLE present with different features and it can be difficult to diagnose children with JSLE. Classification criteria are important to help doctors diagnose JSLE and to help develop better treatments for JSLE. The study aims to estimate disease incidence as well as determine how JSLE affects children when they first present, management, diagnostic delay and how new classification criteria used for adults perform in children being treated as JSLE.

Surveillance period

September 2017 - September 2019 (inclusive).

Follow-up period: Follow-up questionnaire at 12 months after initial diagnosis, ending in September 2020.

Methodology

Data capture uses standard BPSU methodology.

In addition, adult clinicians including dermatologists, nephrologists and rheumatologists who may see young people with a new diagnosis of JSLE were also asked to report all new cases they might have seen on a monthly basis in parallel to the BPSU reporting system. Details of the study protocol are available at **http://www.rcpch.ac.uk/bpsu/lupus**

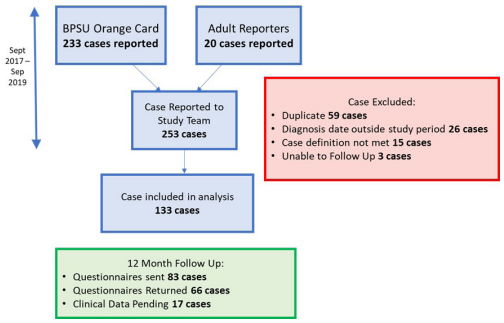
Dr Hanna Lythgoe

Analysis

This preliminary analysis is performed on clinical data obtained at the time of diagnosis for cases reported up until 1st June 2020. Follow-up data, inclusion of outstanding clinical data and detailed analysis are still pending so these figures are subject to change.

Of the 133 confirmed cases, 74% (n=99) met the American College of Rheumatology classification criteria for SLE (ACR-1997), 85% (n=113) met Systemic Lupus International Collaborating Clinics classification criteria (SLICC-2012) and 86% (n=114) met at least one of the two different classification criteria (ACR-1997 or SLICC-2012).

Figure 7: Cases reported to the BPSU JSLE study over the surveillance period



Of the 99 patients meeting the ACR-1997 classification criteria, median age at diagnosis was 13.6 years (interquartile range (IQR) 11.7–15.2 years) with a female:male ratio of 5.9:1 respectively. Excluding two patients of unkown ethnicity, 64% (n=62) were non-Caucasian. Median time from symptom onset to diagnosis was four months (IQR 2–10.5 months). The longest delay was 106 months (child initially diagnosed with Henoch Schnolein Purpura. Of the 99 patients, 17% (n=17) patients had been reviewed by ≥1 paediatric sub-specialist prior to achieving a diagnosis of JSLE, and 15% (n=15) had not been referred to a paediatric rheumatology specialist despite seeking medical review. Further analysis looking at predictors of diagnostic delay is planned.

No patients had died within one month of diagnosis.

Discussion

Early analysis suggests that of the 133 reported cases 14% (n=19) being treated as JSLE do not meet either of the currently widely used classification criteria (ACR-1997 or SLICC-2012). The varied presentation and heterogenic nature of JSLE makes diagnosis and classification challenging; we are currently looking at these patients in more detail as one-year follow-up data are available to see if they meet the case definition of probable lupus or not.

Table 6: Treatment started/planned within 3 months of diagnosis of the 99 cases

Treatment	%	N
Oral and/or intravenous steroids	93	92
Hydroxychloroquine	98	97
Mycophenolate mofetil	57	96
Rituximab	13	13
Azathioprine	16	16
Methotrexate	18	18
Cyclophosphamide	13	13
Intravenous immunoglobulin	4	4

The study was designed with a primary aim of estimating incidence of JSLE in patients up to 16 years of age in line with traditional and proven BPSU methodology. However, we also aimed to capture data on patients aged 16 and 17 years presenting with JSLE, acknowledging that most of these patients may be diagnosed within adult services, through extending our case reporting to relevant adult clinicians. Unfortunately, very low numbers of reported cases from adult clinicians and low numbers of cases reported in patients aged 16–17 years make it clear that it is probable we have not been able to accurately capture incidence data in this subgroup.

Acknowledgements

The Lupus team would like to thank Carla Roberts for the support she has lent to the study,

Funding

This study has been funded through the Sir Peter Tizard Bursary and LUPUS UK.

Public and patient engagement

LUPUS UK.
Web: **https://www.lupusuk.org.uk**

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Dr Ruth Murphy

Life-threatening bronchopulmonary dysplasia

Key points

- 329 notifications of cases have been received during the 13 month surveillance period, of which 153 were confirmed cases.
- Data collection is across a series of three questionnaires: 153 have provided demographic and delivery details (questionnaire 1), 94/153 (61%) have provided data to the point of discharge from the neonatal unit (questionnaire 2), and 75/94 (80%) have provided data to 1 year or death (questionnaire 3). Final reminders are in progress.
- 62% of eligible cases are male, with a median gestational age 26^{+3} weeks and birth weight of 730g.

Summary

Many babies born more than eight weeks early will have some problems with their breathing and need oxygen for many weeks. This is known as chronic lung disease or Bronchopulmonary dysplasia (BPD) and happens because the lungs were immature at birth. Most babies recover well, but some may go home in oxygen. A small number have such severe lung problems that they need to stay in hospital on breathing machines for many weeks or months, and may even die. Because very small numbers of babies have such severe problems, little is known about this important group of preterm babies and how they are cared for or what might make their outcome better.

This study aims to collect data on these babies and how they are looked after, including what sort of breathing support and medicines they receive, as a step to better understanding this serious problem.

Surveillance period

July 2017 - July 2018 (inclusive).

Follow-up period: Follow-up questionnaire at 12 months of age ending in July 2019.

Methodology

Data capture uses standard BPSU methodology. Details of the study protocol are available at **http://www.rcpch.ac.uk/bpsu/bpd**

Life threatening BPD is defined as any infant born at <32 weeks gestation, without significant congenital anomaly, requiring positive pressure support (ventilation, nCPAP, BiPAP or high flow >2L/min) or pulmonary vasodilators at, or beyond, 38 weeks gestation, without intercurrent illness to explain this need.

Analysis

Data were collected in a series of three questionnaires:

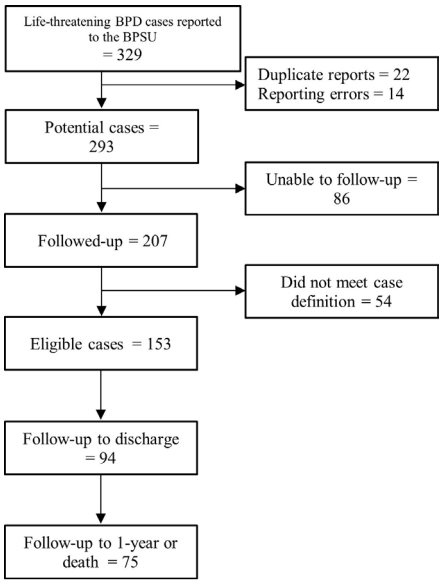
Q1: Demographic, maternal and delivery details;

Drs S Harigopal, J Berrington & S Ramaiah

Q2: Data regarding neonatal unit admission; Q3: Post-discharge information up to one year of age.

In total, 329 case notifications were received by the BPSU, resulting in 153 confirmed cases. Figure 8 outlines the data collection process to date. Final reminders and post-discharge data collection are ongoing.

Figure 8: Cases reported to BPSU between July 2017 and July 2018.



Using the UK Office for National Statistics population estimates, the estimated incidence of life-threatening BPD in infants born at <32 weeks gestation during the surveillance period is 1,681/100,000 (1.68%). Median gestational age was 26.4 (IQR: 24.6-28) weeks, and birth weight 730g (620-910g). 62% (95/153) of infants reported were male and 79.7% (122/153) were White British.

Ninety-one percent (139/153) of infants received antenatal steroids, and all except one received surfactant. Median duration of invasive ventilation was 29 days (IQR: 18-51), and total duration of respiratory support on the neonatal unit 110 days (91.3-141). 56% (53/94) of infants received high frequency oscillation, and 32% (30/94) received inhaled nitric oxide. Postnatal steroids were used for BPD in 57/94 (61%) infants, and 82/94 (87%) received diuretics. To date, 15 deaths have been reported in the 94 infants followed up to, or post-, discharge.

Further data regarding outcomes will be obtained from Q3, which covers post-discharge from the neonatal unit up to one year of age. All eligible cases have been sent this questionnaire at least once, and further analysis will be completed when final reminders are complete.

Discussion

During the study period, the estimated incidence of life-threatening BPD in infants born <32 weeks gestation in the UK was 1,681/100,000 (1.68%). As anticipated, the infants in this study are extremely premature (average gestational age 26.4 weeks), and of extremely low birth weight (average 730g). These infants by definition receive a protracted period of respiratory support during a prolonged NICU admission, highlighting the significant healthcare resource use, and a high morbidity associated with this condition.

There is significant variation in practice in the management of infants with BPD and many of the interventions and medications used have a very limited evidence base. This uncertainty is particularly challenging in the subgroup of infants with life-threatening BPD, as the trajectory of the condition and current approaches to management are not well described. This study is providing a detailed description of these infants and will identify areas for focus of future research.

Data collection regarding one year outcomes is still ongoing, and once complete further analysis will be undertaken to assess the short-term outcomes of this condition and identify factors related to key outcomes, particularly death and long-term ventilation.

Public and patient engagement

The Tiny Lives Trust
Web: **https://www.tinylives.org.uk**

Acknowledgements

The BPD team would like to thank Mrs Sue Morrison for the administrative support she has lent to the study, Tiny Lives for funding and supporting the study and Bliss Charity for their endorsement. The BPD would also like to acknowledge the RCPCH members and parents helping with the study. Lastly, the study team would like to acknowledge Newcastle Upon Tyne Hospitals NHS Foundation Trust for sponosring the study.

Funding

This study is funded by The Tiny Lives Trust.

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Multisystem inflammatory syndrome, Kawasaki disease and toxic shock syndrome


Dr Shamez Ladhani

Summary

During April 2020, a number of children became very unwell with symptoms and signs of hyperinflammation and were admitted to the paediatric intensive care units. Many children had symptoms common to both Kawasaki disease, which causes inflammation of the arteries, and toxic shock syndrome, a rare life-threatening immune reaction to certain infections. Some children had predominantly gastrointestinal presentations, with abdominal pain, vomiting and diarrhoea. Around half the children with this condition in London were also positive for SARS-CoV-2. Although such cases are rare, they have been reported in different regions of the UK and in cities across Europe and the United States that have been affected by COVID-19. At present, we do not understand the relationship between SARS-CoV-2 and this new condition, or even if there is one. We ask paediatricians to report all cases which have features of this multisystem inflammation.

We also ask paediatricians to report cases of Kawasaki disease and toxic shock syndrome to assess whether their incidence has increased compared to recent BPSU estimates for both conditions – and to determine any role of COVID-19. Collecting data on these two conditions may also help assess any differences in clinical disease, course and outcomes compared to the hyperinflammatory syndrome which is temporally associated with COVID-19, but may or may not be caused by it.

Surveillance period

March 2020 - March 2021 (inclusive).

Methodology

Data capture uses standard BPSU methodology; details of the study protocol are available at **https://www.rcpch.ac.uk/bpsu/multisystem-inflammatory-syndrome**

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Funding

This study is funded through a grant from Public Health England.

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Neonatal complications of coronavirus disease (COVID-19)


Dr Chris Gale

Summary

Surveillance of the neonatal complications of coronavirus disease (COVID-19) commenced in March 2020. The study aims to find out how many babies develop coronavirus infection in the first month after birth, and how many babies born to women with coronavirus need neonatal care; describe which babies develop COVID-19 infection and what symptoms or signs they have; and describe how COVID-19 in babies is identified and treated.

The SARS-CoV-2 coronavirus is a new virus that originated in China, where it was first recognised as a causing a new infection in late 2019. It has crossed the world and we started to see people affected by the virus in the United Kingdom in February 2020. Since this coronavirus is a new virus, no one has immunity to it.

We have very little information about how often babies get this coronavirus infection, whether it transmits from mothers to their babies while they are still pregnant, during labour and birth, or whether the infection occurs following birth. Carrying out research now will mean we can give the best care to mothers and babies and the best advice to pregnant women about the effects of coronavirus on them and their baby.

We are undertaking a national study collecting information about newborn babies who have coronavirus and need medical care or who are born to mothers who have coronavirus. We will be asking all paediatricians and neonatologists in the UK to report any babies that meet the surveillance case definition to the BPSU every week.

Data from the first two months of surveillance is due to be published shortly in the *Lancet Child and Adolescent Health*.

Surveillance period

March 2020 - March 2021 (inclusive).

Methodology

Data capture uses standard BPSU methodology; details of the study protocol are available at **https://www.rcpch.ac.uk/bpsu/covid-19**

Funding

This study is funded through a grant from the Department of Health and Social Care (reference: PR-PRU-1217-21202)

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Professor Elizabeth Draper
University of Nottingham: Professor Don Sharkey
University of Oxford: Professor Marian Knight

Neonatal herpes simplex disease

Key points

- In the first 11 months of surveillance 107 case notifications were received of which 47 cases have been confirmed to date.
- This would equate to an incidence of 7.2/100,000 at this stage.[1]
- 43% (n=20/47) of cases were classified as disseminated neonatal herpes simplex (HSV) disease. The mortality rate amongst these patients was 65% (n=13/20)

Summary

Neonatal herpes simplex virus (HSV) disease is a potentially devastating condition which can lead to significant morbidity and death.[1] Transmission typically occurs during delivery through an infected birth canal, or after delivery, following exposure to HSV infections such as cold sores. Transplacental transmission can also occur, although this is rare.

It is known that sexually transmitted herpes infections in adults have increased in the last decade and it is suspected that the number of cases of neonatal HSV is therefore also rising.

It is also known that the UK incidence increased from 1.65 to 3.58/100,000 live births between the first national BPSU study (1986-1991) and the second (2004-2006).[2] A recent local study from Nottingham[3] showed rates of 17.5/100,000 live births, ten times higher than the first BPSU study, and served as a reminder of the devastating consequences of the disease.

Currently, there is insufficient information about the number of UK cases of this disease, which babies are most at risk, ways we might be able to reduce those risks and whether the treatment and prophylaxis we are using is reducing long term problems and later relapses. There is a lack of clarity as to the optimum management of mothers and babies who are at risk: a number of different guidelines are available for clinicians to follow, resulting in variation in practice across sites. All objectives this study will seek to address.

This is of specific relevance to neonates presenting to the emergency department with non-specific signs of sepsis. Currently not all babies are treated for HSV; however, if the prevalence of HSV has increased significantly nationwide to those approaching levels in Nottingham, this may need to be re-considered.

Surveillance period

July 2019 - July 2021 (inclusive).

Follow-up period: Follow-up questionnaire at 12 months of age ending in July 2022.

Methodology

Data capture uses standard BPSU methodology. Details of the study protocol are available at **http://www.rcpch.ac.uk/bpsu/hsv**

Dr Katy Fidler

Further information on the number of deaths from neonatal HSV disease will be requested from the National Child Mortality Database and the Office of National Statistics. Information on the number of admissions to PICU with neonatal HSV disease will be requested from the PICANet database. This information will be used to ensure that the reporting methods used are accurately capturing cases of neonatal HSV.

Analysis

This is an interim analysis based on data collected during the first 11 months of the surveillance period. A more detailed analysis will be conducted on completion of the study.

One-hundred-and-seven case notifications were received during this period. Six were identified as reporting errors prior to questionnaires being returned (two were unintentional notifications and four did not meet the case criteria). The response rate for the remaining cases was 65% (n=66/101). Three clinicians reported that their cases were duplicates and did not complete the clinical questionnaire. After removing the 16 duplicate report, 47 confirmed cases remained giving an incidence of 7.2/100,000.

Of the 47 cases, 23 infants were female and 24 male. The mean birthweight was 2.69kg (range 0.56kg–4.14kg). Thirty-nine were white, four were mixed ethnicity (White and Asian; Black Caribbean, Asian and White; Black African and White); ethnicity was not recorded for four cases. Twenty-seven infants (57%) were born at term, 11 were moderate to late preterm (32-37 weeks) and nine were either very or extremely preterm (<32 weeks).

Maternal age was known in 35 of the 47 cases. The median age was 24.5 years (range 14 to 37 years). The mother was reported as the source of infection in 16 cases of whom seven had known genital infections.

Clinical presentation varied between infants. Only 13% (6/47) infants had a fever at presentation, of which five had meningoencephalitis and one had disseminated disease. Admission CRP was normal (<5) in 56% (23/41) infants with an available result; 22% (9/41) had a CRP >20 at presentation. Only 27% (3/11) of infants with meningoencephalitis had skin lesions at presentation and 45% (9/20) of those with disseminated disease had lesions. In 66% (13/20) cases of disseminated disease, the infant presented with “sepsis” clinically.

Final diagnosis was isolated skin, eye, mouth (SEM) infection in 34% (16/47); meningoencephalitis/ CNS disease in 11/47 (23%) (two with concomitant SEM infections) and disseminated disease in 20/47 (41%), of whom seven had concomitant SEM infection and three had both SEM and CNS intercurrent infections.

Among the nine babies born before 32 weeks gestation, five died, three were still admitted at the time of data collection and one was discharged home healthy. Mortality for disseminated disease was 65% (13/20). Of the 13 who died 62% had HSV-2 and 31% had HSV-1.

Ninety-four per cent (44/47) of the infants received antiviral medication. In the remaining three cases, the infants sadly died on the day of admission before treatment was started. Intravenous aciclovir was commenced in 93% (43/44) of cases; one infant was given topical aciclovir to treat an isolated HSV-1 conjunctivitis. Foscarnet was used in addition to aciclovir for one infant. Treatment day of illness compared to mortality has not yet been analysed.

Discussion

Analysis of preliminary data demonstrates an increased incidence of neonatal HSV disease compared to findings of the previous BPSU studies. Incidence is estimated using incomplete data and is likely to increase further once the data from the outstanding questionnaires are included in calculations. Prognosis for infants with disseminated infection remains poor, even in those receiving antiviral treatment.

These results remind us of the challenges of detecting disseminated HSV infection in unwell infants. Typical markers of serious infection such as fever and abnormalities in CRP at presentation are absent from the majority of cases. Characteristic skin lesions are also not consistently present at the onset of illness.

The service disruptions caused by the COVID-19 pandemic may have caused reductions or delays in clinician case reporting between March and May 2020. Analysis of monthly incidence will also be completed at the end of the surveillance period and the COVID period compared to non-COVID period to determine if social distancing measures could have influenced postnatal acquisition of this virus.

Public and patient engagement

Kit Tarka Foundation
Web: **https://www.kittarkafoundation.org**

The Herpes Viruses Association
Web: **http://www.herpes.org.uk**

Further information on the NHS choices website:
Web: **https://www.nhs.uk/conditions/neonatal-herpes**

Acknowledgements

We would like to thank Dr Catherine O'Sullivan for her help in setting up the study.

Funding

This study has been funded by the Rockinghorse Charity, Brighton and the Kit Tarka Foundation, Brighton.

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

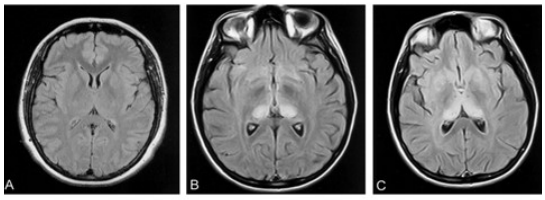
Key points

- 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. From May 1997 until 31st March 2020 4,675 children have been notified; 2,032 children have a known diagnosis other than vCJD, with over 220 different neurodegenerative disorders 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 National Institute for Health Research (NIHR) Policy Research Programme (PR- ST-1216-10001) it is being carried out via the BPSU in conjunction with the National Creutzfeldt-Jakob Disease Research and Surveillance Unit in Edinburgh (NCJDRSU). 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 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: MRI brain scan findings in vCJD[3]



A, Normal FLAIR image at the level of the basal ganglia shows the thalamus is normally isointense or slightly hypointense relative to the putamen. This appearance is depicted with most sequences, particularly the FLAIR sequence.

B, Pulvinar sign of vCJD. FLAIR image shows marked, symmetrical hyperintensity of the pulvinar (posterior) thalamic nuclei. In this case, the pulvinar signal intensity was scored as grade 4 by both observers.

C, "Hockey-stick" sign of vCJD. FLAIR image shows symmetrical pulvinar and dorsomedial thalamic nuclear hyperintensity. This combination gives a characteristic "hockey-stick" appearance and was present in 93% of cases with FLAIR imaging.

The PIND team

The main objective of this study is to provide prospective on-going surveillance for vCJD in children. This is done by identifying all diseases that meet the case definition for PIND – see below. The study provides the only means of searching for vCJD in children. In addition the study yields a UK-wide overview of childhood neurodegenerative diseases.

Methodology

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

An Expert Group of specialists in paediatric neurology, neurogenetics and metabolic disease, plus a NCJDRSU representative, meets quarterly to review anonymised clinical information and classify cases, looking for cases of vCJD. The characteristic clinical features of vCJD have been published and provide a basis for that discussion. For instance there are characteristic features on brain MRI Scans – see Figure 9.

Surveillance period

May 1997 – April 2021 (inclusive)

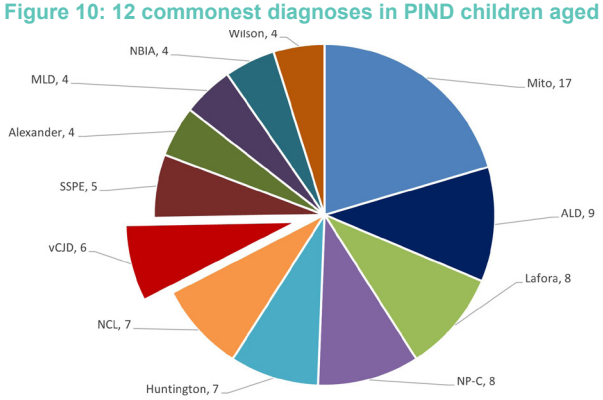
Analysis

Between May 1997 and April 2020 4,675 cases had been notified.

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: A recent analysis showed that between May 1997 and October 2019 2,255 cases meeting the PIND criteria had been notified and had been extensively investigated. Of these 2,008 (male 1,085, female 923) had an underlying diagnosis to explain their deterioration. The numbers of diagnosed cases presenting by age were: < 1year: 40% (n=805), 1-4 years inclusive: 41% (n=825), 5-9 years inclusive

Figure 10: 12 commonest diagnoses in PIND children aged 10-15 years)



Key -
Mito: *mitochondrial disorders*
ALD: *adrenoleukodystrophy*
Lafora: *Lafora body disease*
NP-C: *Niemann-Pick type C*
Huntington: *Huntington disease*
NCL: *neuronal ceroid lipofuscinosis*
vCJD: *variant Creutzfeldt-Jakob disease*
SSPE: *subacute sclerosing panencephalitis*
Alexander: *juvenile Alexander disease*
MLD: *metachromatic leukodystrophy*
NBIA: *neurodegeneration with brain iron accumulation*
Wilson: *Wilson disease.*

13% (n=264), 10-15 years inclusive 6% (n=114), with over 220 different disorders. The distribution by ethnic origin was: White 58.2%, Black 1.9%, Indian 1.3%, Pakistani 17 %, Bangladeshi 2.2%, Chinese 0.25%, Other (other ethnic groups, mixed race, unknown etc.) 19%. Figure 10 shows the 12 commonest diagnoses in the 10-15 year old children, showing the differential diagnosis of PIND in this age group – which includes the six cases of vCJD.

Children with PIND and no underlying diagnosis (idiopathic group): The Expert Group has met regularly and discussed this group of children, currently 258. If a "new" variant of vCJD should arise or if the paediatric presentation differed from the adult presentation, this group could include such a case. However, there is currently no evidence of a "new" unrecognised disorder in this group.

Discussion

The National Creutzfeldt-Jakob Disease Research and Surveillance Unit in Edinburgh reports that there have been 178 deaths from definite or probable vCJD in UK patients of all ages. Until 2016 all these cases were methionine homozygous (MM) at codon 129 of the prion protein gene (PRNP). The first and only confirmed methionine/valine (MV) heterozygous vCJD adult case was identified four years ago (2016).[4] There remains concern that more childhood cases may appear, perhaps with the MV genotype. Children are still at risk of vCJD infection by blood, plasma products, surgical and dental instruments and theoretically via vertical transmission.

Continued surveillance for vCJD is essential and the PIND Study continues to yield unique information about the epidemiology of childhood neurodegenerative disorders in the UK.

Acknowledgements

The PIND team are very grateful for the support of the PIND Expert Group: Dr Peter Baxter, Dr Carlos de Sousa, Professor Paul Gissen, Professor Manju Kurian, Dr Ming Lim, Professor John Livingston, Professor Robert McFarland, Dr Helen Mundy, Dr Suvankar Pal, Professor Richard Robinson, Dr Evangeline Wassmer, and Professor Sameer Zuberi. Our thanks to Dr Michael Pike who has just left the Group.

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3. Collie DA, Summers DM, Sellar RY et al. Diagnosing Variant Creutzfeldt-Jakob Disease with the Pulvinar Sign: MR Imaging Findings in 86 Neuropathologically Confirmed Cases. Am J Neuroradiol 2003;24:1560–1569.

4. Peden AH, Head MW, Ritchie DL, Bell JE, Ironside JW. Preclinical vCJD after blood transfusion in a PRNP codon 129 heterozygous patient. Lancet 2004; 264:527-29.

Funding

National Institute for Health Research Policy Research Programme [PR-ST-1216-10001].

Public and patient engagement

Creutzfeldt-Jakob Disease Support Network. Web: **http://www.cjdsupport.net**

Batten Disease Family Association. Web: **http://www.bdfa-uk.org.uk**

Society for Mucopolysaccharide Diseases. Web: **http://www.mpsociety.co.uk**

Alex TLC (Adrenoleukodystrophy). Web: **http://www.alextlc.org**

The Cure & Action for Tay-Sachs (CATS) Foundation. Web: **http://www.cats-foundation.org**

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Sydenham's chorea

Key points

- The first year of surveillance has been completed, with 23 new confirmed or suspected cases of Sydenham's chorea (SC), from 42 reports to BPSU.
- No cases have been reported through the Child and Adolescent Psychiatry Surveillance System (CAPSS).
- The second year of surveillance will be completed in November 2020, at which point we will be ready to complete analysis of the surveillance data and report incidence estimates.

Summary

Sydenham's chorea (SC) is a rare condition which can affect the brain following infection with a type of bacteria (*Streptococcus*). It largely affects children and young people. Symptoms include abnormal body movements and muscle weakness, which range from mild to severe and may disrupt a child's ability to carry out activities of daily living like writing and walking. Whilst SC may get better within six months, symptoms can return repeatedly over the course of two years or longer. SC is often accompanied by emotional and behavioural symptoms such as hyperactivity, obsessions and compulsions, and anxiety. In the UK and the Republic of Ireland (ROI), SC is currently considered a 'rare disease'; but little is known about how many children nowadays are affected by the disorder, what happens to them after diagnosis, or about their needs. Through this research we are studying the numbers, characteristics, management and outcomes of new cases of SC aged between 0 and 16 years in the UK and ROI. Information on investigations, management, recovery and outcomes such as educational difficulties occurring as a result of SC is being sought. As well as describing the current pattern of SC, it is hoped findings will raise awareness amongst clinicians to improve diagnosis, contribute to planning effective services, and assist in designing research trials to test treatments.

Surveillance period

November 2018 - November 2020 (inclusive).

Follow-up period: Until November 2022 at 12-months and 24-months.

Methodology

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

A parallel Child and Adolescent Psychiatry Surveillance System (CAPSS) study is running from May 2019 to December 2020. The gathered data will focus on the psychiatric presentation and the course of those associated symptoms. Analysis will therefore be conducted separately for each study. However, we will also conduct analysis on the crossover between cases reported by paediatricians and by child psychiatrists; and compare characteristics between these two groups.

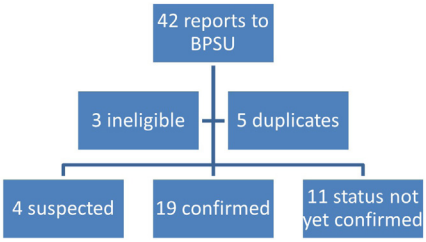
Dr Tamsin Newlove-Delgado

Analysis

Surveillance is still running, and so data analysis has yet to be completed. Interim findings from the first year of the study (November 2018-December 2019) are presented. Cases meeting the surveillance case definition of suspected or confirmed SC, will be finally confirmed as the diagnosis after the 12 month follow-up.

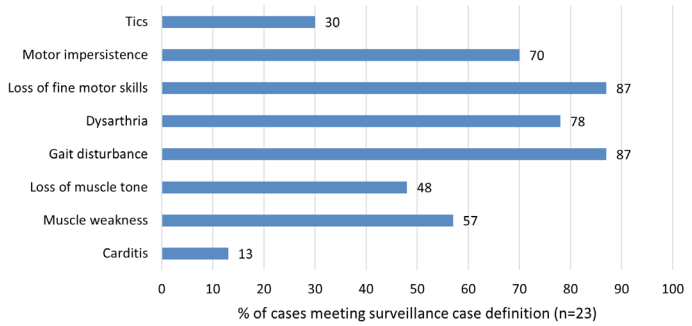
During this time period, there have been 42 reports of which there have been 23 new confirmed or suspected cases of Sydenham's chorea (SC), to the BPSU. Of the 23 cases meeting our surveillance case definition, 60% were female, with an age range of four to 15 years (mean age = nine years). The majority (65% n=15) presented with 'moderate' severity of chorea. There were no reports of antecedents other than sore throat. (Figure 11).

Figure 11: Breakdown of case reports November 2018-December 2019



The most common neurological presenting features (apart from chorea which was present in all and needed to meet the case definition), were loss of fine motor skills (87% n=20) and gait disturbance (87% n=20), followed by dysarthria (78% n=18) and motor impersistence (70% n=16) (Figure 12). Carditis was the least common, reported at presentation in only 13% (n=3). The most common emotional and behavioural symptoms on presentation were emotional lability (70% n=16), anxiety (43% n=10) and inattention/attention deficit (35% n=8).

Figure 12: Presenting neurological features



Almost all cases had evidence of prior streptococcal infection, most commonly confirmed by ASOT (Antistreptolysin O Titre). All except one were prescribed a course of antibiotics. The duration of the prescription was variable, ranging from ten days to expected lifelong prophylaxis in cases that had carditis. Steroids as an immunomodulatory treatment were also used, and around half received symptomatic treatment of chorea with anticonvulsants and neuroleptics. Referral to CAMHS for emotional or behavioural symptoms was uncommon.

Discussion

Over the first 12 months of our surveillance through BPSU, there have been 23 new confirmed or suspected cases of SC reported to us by paediatricians, a slightly lower number than expected based on the study by Crealey *et al.*,[2] but surveillance continues until November 2020, incidence to be calculated over a two-year period. From the limited analysis undertaken we have been able to characterise some of the most common presenting features, and also note that emotional and behavioural symptoms were common in the cases reported by paediatricians. Almost 80% had emotional lability on presentation, and anxiety and/or inattention/attention deficit were also reported in approximately a third of cases. However, despite this presentation, no cases reports have so far been reported by child and adolescent psychiatrists in our parallel surveillance study through CAPSS; which is an interesting finding. Explanations are being sought for this, but one reason may be that children with these symptoms are being seen by psychologists within paediatric services, rather than CAMHS. Early findings also suggest that practice in investigation and management may be variable, particularly with regards to antibiotic prophylaxis prescribing duration.

Acknowledgements

We would like to thank Dr Eva Wooding, Academic Clinical Fellow in Paediatrics and Ms Lucy Clark, Medical Sciences graduate, for assistance with data extraction.

Funding

This study has been funded by the Paul Polani Award from the Royal College of Paediatrics and Child Health/British Academy of Childhood Disability, and by the Sydenham's Chorea Association.

Public patient engagement

The Sydenham's Chorea Association

Web: **http://www.sydenhamschorea.org.uk**

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2. Crealey M., Allen N.M., Webb D., Bouldin A., Mc Sweeney N., Peake D., Tirupathy S., Butler K., King M.D. Sydenham's chorea: not gone but perhaps forgotten. Archives of Disease in Childhood 2015, 100(12):1160-2.

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Sydenham's Chorea Association:
Andrew Samuel, Catriona Clark
University of Cambridge:
Professor Tamsin Ford

Appendix - Publications 2019 -2020

Attention deficit hyperactivity disorder

1. Price, A., Janssens, A., Woodley, A.L., Allwood, M and Ford, T. (2019) Experiences of healthcare transitions for young people with attention deficit hyperactivity disorder: a systematic review of qualitative research *Child and Adolescent Mental Health* 24, No. 2, pp. 113–122 doi:10.1111/camh.12297

2. Eke, H., Janssens, A and Ford, T. (2019) Transition from children's to adult services: a review of guidelines and protocols for young people with attention deficit hyperactivity disorder in England *Child and Adolescent Mental Health* 24, No. 2, pp. 123–132

3. Eke, H., Ford, T., Newlove-Delgado, T., Price, A., Young, S., Ani, C., Sayal, K., Lynn, R. M., Paul, M. and Janssens, A. (2019).Transition between child and adult services for young people with Attention Deficit Hyperactivity Disorder (ADHD): findings from a British national surveillance study *The British Journal of Psychiatry*. pp. 1–7. doi: 10.1192/bjp.2019.131

4. Price A, Janssens A, Dunn-Morua S, Eke H, Asherson P, Lloyd T, Ford T: Seven steps to mapping health service provision: lessons learned from mapping services for adults with Attention-Deficit/Hyperactivity Disorder (ADHD) in the UK. *BMC Health Services Research* 2019, 19(1):468

5. Eke H, Janssens A, Downs J, Lynn RM, Ani C, Ford T. (2019) How to measure the need for transition to adult services among young people with Attention Deficit Hyperactivity Disorder (ADHD): a comparison of surveillance versus case note review methods. *BMC Medical Research Methodology*, 19:179

BPSU

6. Lynn RM, Avis JL, Lenton S, Amin-Chowdhury Z, Ladhani SN. Delayed access to care and late presentations in children during the COVID-19 pandemic: a snapshot survey of 4075 paediatricians in the UK and Ireland [published online ahead of print, 2020 Jun 25]. *Arch Dis Child*. 2020;archdischild-2020-319848. doi:10.1136/archdischild-2020-319848

7. Ladhani S, Viner RM, Lynn RM, Baawuah F, Saliba V, Ramsay M. The unintended consequences of COVID-19 in children. *BMJ* In press 2020

8. Lynn RM, Reading R; BPSU Ascertainment Group. Case ascertainment in active paediatric surveillance systems: a report from the British Paediatric Surveillance Unit Ascertainment Group. *Arch Dis Child*. 2020;105(1):62-68. doi:10.1136/archdischild-2019-317401

Death in epilepsy

9. Abdel-Mannan O, Sutcliffe AG. A national surveillance study of childhood epilepsy mortality in the UK and Ireland. Eur J Neurol. 2020;27(2):327-333. doi:10.1111/ene.14081

Enterovirus and parechovirus meningitis

10. Kadambari S, Braccio S, Ribeiro S, et al. Enterovirus and parechovirus meningitis in infants younger than 90 days old in the UK and Republic of Ireland: a British Paediatric Surveillance Unit study. *Arch Dis Child*. 2019;104(6):552-557. doi:10.1136/archdischild-2018-315643

Female Genital Mutilation

11. Hodes D, Ayadi O'Donnell N, Pall K, Leoni M , Lok W, Debelle G, Armitage A, Creighton SM Lynn RM. Female Genital Mutilation in children and young people under 16 years of age; results of an epidemiological surveillance study. *Arch Dis Child* in press 2020

Enterovirus and parechovirus meningitis

12. Kadambari S, Braccio S, Ribeiro S, et al Enterovirus and parechovirus meningitis in infants younger than 90 days old in the UK and Republic of Ireland: a British Paediatric Surveillance Unit study Archives of Disease in Childhood 2019;104:552-557.

Kawasaki disease

13. Tulloh RMR, Mayon-White R, Harnden A, Ramanan AV, Tizard EJ, Shingadia D, Michie CA, Lynn RM, Levin M, Franklin OD, Craggs P, Davidson S, Stirzaker R, Danson M, Brogan PA. Kawasaki disease: a prospective population survey in the UK and Ireland from 2013 to 2015 [published correction appears in Arch Dis Child. 2020 Mar;105(3):e5]. *Arch Dis Child*. 2019;104(7):640-646. doi:10.1136/archdischild-2018-315087

Haemolytic Uraemic Syndrome

14. Adams N, Byrne L, Rose T, Adak B, Jenkins C, Charlett A, Violato M, O'Brien S, Whitehead M, Barr B, Taylor-Robinson D, Hawker J. Sociodemographic and clinical risk factors for paediatric typical haemolytic uraemic syndrome: retrospective cohort study. *BMJ Paediatr Open*. 2019 Dec 17;3(1):e000465. doi: 10.1136/bmjpo-2019-000465. PMID: 31909217; PMCID: PMC6936988.

Lead ingestion

15. Annual Report of the Lead Exposure in Children Surveillance in England, detailing the cases captured through the surveillance in 2018: **https://www.gov.uk/government/publications/lead-exposure-in-children-surveillance-reports**

Appendix - Publications 2019 -2020

Progressive intellectual and neurological deterioration

16. Verity C, Winstone AM, Will R, Powell A, Baxter P, de Sousa C, Gissen P, Kurian M, Livingston J, McFarland R, Pal S, Pike M, Robinson R, Wassmer E, Zuberi S.. Surveillance for variant CJD: should more children with neurodegenerative diseases have autopsies?. *Arch Dis Child*. 2019;104(4):360-365. doi:10.1136/archdischild-2018-315458

Rickets

17. Julies P, Lynn RM, Pall K, et al. Nutritional rickets under 16 years: UK surveillance results. *Arch Dis Child*. 2020;105(6):587-592. doi:10.1136/archdischild-2019-317934

Zika virus

18. Oeser C, Aarons E, Heath PT, Johnson K, Khalil A, Knight M, Lynn RM, Morgan D, Pebody R Surveillance of congenital Zika syndrome in England and Wales: methods and results of laboratory, obstetric and paediatric surveillance. *Epidemiol Infect*. 2019;147:e262. Published 2019 Sep 4. doi:10.1017/S0950268819001535 https://pubmed.ncbi.nlm.nih.gov/31481135/

Appendix - Presentations 2019-2020

Attention deficit hyperactivity disorder

1. Eke H, *et al.* In transition from children's services to adult services: the case of ADHD World Congress of the International Association for Child and Adolescent Psychiatry and Allied Professions (IACAPAP) Prague Czech Rep. .July 2018

HIV in pregnancy and childhood

2. Francis K, Thorne C, Horn A, Peters H. Peters H. Successes and emerging challenges in prevention of vertical HIV transmission in the UK and Ireland. British HIV Association (BHIVA). Bournemouth, 2019.

Food Protein Induced Enterocolitis Syndrome

3. Stiefel, G et al. "BPSU Surveillance of Food Protein Induced Enterocolitis Syndrome (FPIES) in UK and Republic of Ireland: First 4 months". *Poster*. BSACI conference 2019.

Nutritional rickets

4. Shaw N, Pall K, Leoni M, Lynn R et al. Vitamin D deficiency nutritional rickets presenting to secondary care in children (<16 Years) – A United Kingdom surveillance study. 9th International Conference on Child Bone Health, 22-25 June; Salzburg, Austria 2019.

Progressive intellectual and neurological deterioration

5. Baker E, Maunder P, Winstone AM, Verity C. "Children with progressive intellectual and neurological deterioration in Northern Ireland 1997-2019". *Poster* Annual Meeting of the British Paediatric Neurology Association, Belfast, January 2020.

6. Baker E, Maunder P, Winstone AM, Verity C. "The differential diagnosis of variant CJD in UK children 1997-2019". *Poster* Annual Meeting of the British Paediatric Neurology Association, Belfast, January 2020.

7. Verity C, Powell A, Winstone AM. The changing clinical spectrum of Aicardi-Goutières syndrome in UK children (poster presentation); 45th BPNA Annual Scientific Meeting; 23 - 25 January 2019; Liverpool, UK.

8. Powell AE. Has SSPE disappeared from the UK? Findings from the PIND Study. 45th BPNA Annual Scientific Meeting; 23 - 25 January 2019; Liverpool, UK.

Notes

Membership of Scientific Committee 2019

Dr Shamez Ladhani	Chair
Dr Lamiya Samad	Medical Advisor (non-infectious disease), UCL Great Ormond Street Institute of Child Health
Dr Ellen Pringle	Medical Advisor (infectious disease), Public Health England
Mr Richard Lynn	Scientific Coordinator
Ms Emily Arkell	Director of Research and Policy. Royal College of Paediatrics & Child Health
Professor Nick Bishop	Vice President Science & Research, Royal College of Paediatrics & Child Health
Dr Hani Ayyash	Consultant in Neurodevelopment
Dr Sarah Clarke	Trainee representative
Dr Gavin Dabrera	Public Health England
Dr Peter Davis	Consultant in Intensive Care
Dr David Elliman	UCL Great Ormond Street Institute of Child Health
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Dr Simon Nadel	Consultant in Intensive Care
Dr Robert Negrine	Consultant Neonatologist
Dr Ifeanyichukwu Okike	Consultant paediatrician
Dr Arlene Reynolds	Senior Epidemiologist, Health Protection Scotland
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Dr Jane Sutton	Patient and Carers Representative
Dr Marc Tebruegge	Consultant Paediatric Infectious Diseases & Immunology
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Published November 2020 by the **British Paediatric Surveillance Unit:**
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