What is the British Paediatric Surveillance Unit (BPSU)?

The aim of the BPSU is to encourage the study of rare conditions in children. It was founded in 1986 by the Royal College of Paediatrics and Child Health, Public Health England and Great Ormond Street Institute of Child Health (University College London).

What does the BPSU do?

It allows doctors and researchers to find out how many children in the UK and the Republic of Ireland are affected by the particular disease or condition each year - this is called epidemiological surveillance. Doctors can also gather information about all the cases of a particular rare condition so they can begin to understand what might have caused it and how to diagnose and treat it. BPSU studies can benefit future patients with rare conditions.

How does the BPSU work?

Each month the BPSU sends an e-card to over 4000 consultant paediatricians and specialists; the card lists the rare conditions currently being studied. If any doctor has seen a child affected by one of these conditions they tick a box on the card and send it back. The BPSU informs the research team who send the doctor a short confidential questionnaire asking for more information. Researchers are not usually told the names of patients, and families are not contacted.

What has the BPSU achieved?

The BPSU has now helped to undertake surveys of over 110 rare conditions which may affect children. These have helped to increase understanding of why the conditions occur and can help to provide better diagnoses and treatments.

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Cambridge University Hospitals NHS
NHS Foundation Trust

BRITISH PAEDIATRIC SURVEILLANCE UNIT

Public Information Leaflet

Progressive Intellectual and Neurological Deterioration in Children (the PIND Study)

A study of rare neurodegenerative diseases of childhood

This leaflet provides information about the PIND Study, which is designed to detect any cases of variant Creutzfeldt-Jakob disease in UK children. It gives the background to the Study, which has been running since 1997, and explains why it is so important. It also provides the contact details of the researchers undertaking the study and a link to the website where the results will be published.

What is variant Creutzfeldt –Jakob disease (vCJD)?

vCJD is a rare disorder that was first described in print in 1996. It is caused by a small protein particle called a prion which leads to deterioration in brain function over the course of months and eventually to death. The disease was originally transmitted to humans via food that came from cattle suffering from bovine spongiform encephalopathy (BSE or “mad cow disease”). By October 2021, there had been 178 deaths in the UK due to vCJD in patients of all ages (including 6 children).

How do children get vCJD?

Measures have been taken to remove the risk of transmission of vCJD by eating meat or meat products. However there is still a risk that vCJD could infect humans via surgical or dental instruments, by blood transfusions or pass directly from mothers to children around the time of pregnancy.

Why is this study happening?

Recently an adult was diagnosed with vCJD and was found to have a different genetic type from all the previous cases. Possibly more cases with that genetic type will now appear so it is important to continue to search for cases of vCJD.

The PIND Study provides the only means of searching for vCJD among the many neurodegenerative diseases of childhood. We ask paediatricians to report all children that they see with worsening neurological problems. A group of experts then reviews the anonymised clinical details for each child and decides whether or not that child has vCJD or has another diagnosis. Although the members of the PIND Expert Group cannot identify the children that they discuss, the PIND Study team provides feed-back from the experts to the paediatricians who report children to us.

Because the PIND study provides unique data about many rare neurological diseases the findings are of great interest to health professionals and to those who are caring for the children. Results from the study are presented in medical journals and on various websites for parents and families. It is not possible to identify individual patients in these reports.

How will the information be collected and used?

Cambridge University Hospitals NHS Foundation Trust Research and Development Department is the sponsor for this surveillance study. Dr Christopher Verity the Principal Investigator is the data custodian so is responsible for looking after the information and using it properly.

Information about notified children is collected via a telephone interview with their doctors, by sending questionnaires or by visiting hospitals to review the hospital notes. The PIND Study needs to collect patients' names and addresses, dates of birth and home postcodes in order to obtain their investigation results and follow their progress. Because of the public health importance of the study the PIND team has approval to gather this information about patients without consent. The approval was given by the Public Health England Caldicott Advisory Panel and the Scottish Public Benefit and Privacy Panel for Health and Social Care. After the PIND Study finishes, Cambridge University Hospitals NHS Foundation Trust will store the clinical information securely for 30 years.

How long will the study last and where is it happening?

The study will last until 31st March 2024 and will be collecting clinical information from consultant paediatricians, paediatric neurologists and other paediatric sub-specialists across the United Kingdom.

Who is funding this study?

The Department of Health and Social Care Policy Research Programme (PR-ST-1216-10001).

Where will the results be published?

Information about the PIND Study can be found in the BPSU Annual Report on the BPSU website: www.rcpch.ac.uk/bpsu

Who has reviewed this study?

The PIND Study was approved by the NRES Committee East of England Cambridge Central: REC ref 97/010.
The Public Health England (PHE) Caldicott Advisory Panel has agreed that patient identifiable information can be processed without consent under Regulation 3 of the Health Service (Control of Patient Information) Regulations 2002.
The Public Benefit and Privacy Panel for Health and Social Care (Scotland) has approved the PIND Study.

Who to contact if you have any questions

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Public and patient engagement: Creutzfeldt-Jakob Disease Support Network. http://www.cjdsupport.net
Batten Disease Family Association http://www.bdfa-uk.org.uk
Society for Mucopolysaccharide diseases http://www.mpssociety.org.uk
ALD Life (Adrenoleukodystrophy) http://www.aldlife.org
The Cure & Action for Tay-Sachs (CATS) Foundation www.cats-foundation.org/