

BRITISH PAEDIATRIC SURVEILLANCE UNIT

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**PROGRESSIVE INTELLECTUAL AND NEUROLOGICAL DETERIORATION
IN CHILDREN (INCLUDING CREUTZFELDT-JAKOB DISEASE)**

Abstract

Active prospective surveillance of UK children with progressive intellectual and neurological deterioration (PIND) commenced in May 1997. The main aim is to determine whether or not any child has developed variant Creutzfeldt Jakob disease (vCJD). vCJD has appeared in patients as young as 12 years of age and could occur in younger children. Although paediatric cases have not appeared since 2001, vCJD has not gone away. As the clinical presentation of vCJD is not typical of classical CJD and could be different in children, the aim is to detect suspected cases by looking at a broader group of conditions. This group needs to be large enough to include all possible cases of CJD hence the need to perform surveillance for all children with PIND.

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Background:

The recent appearance of **variant CJD (vCJD)** in patients as young as age 16¹ has raised the question as to whether the condition is occurring in children. Either the detection of vCJD in UK children, or the demonstration that it is not occurring, would be an important finding for paediatrics and public health. As the presentation of vCJD is not typical of classical CJD¹, the aim is to detect suspected cases by looking at a broader group of conditions. The group needs to be large enough to include all possible cases of CJD, hence surveillance for **Progressive Intellectual and Neurological Deterioration (PIND)**.

Coverage:

United Kingdom and the Republic of Ireland.

Duration:

May 1997 to January 2007.

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Objective: To carry out active prospective surveillance of UK children with paediatric neurological conditions (including those with specific diagnoses) defined by their **common** presentation - **Progressive Intellectual and Neurological Deterioration (PIND)** - to determine the incidence and distribution of PIND. Cases presenting with PIND will be evaluated critically in order to classify them and investigate the possibility that Creutzfeldt-Jakob Disease (CJD) is occurring in children.

Case Definition

Any child under 16 years of age at onset of symptoms who fulfils **all** of the following three criteria:

- Progressive deterioration for more than three months ***with***
- Loss of already attained intellectual/developmental abilities ***and***
- Development of abnormal neurological signs.

Excluding: Static intellectual loss e.g. after encephalitis, head injury or near drowning

Including: Children who meet the case definition even if specific neurological diagnoses have been made.
Metabolic disorders leading to neurological deterioration.
Seizure disorders if associated with ***progressive*** deterioration.
Children that have been diagnosed as having neurodegenerative conditions but who have not yet developed symptoms

Reporting restricted to: cases seen in the last month but including those whose conditions began earlier (i.e. including “old cases” of children in follow-up if seen in that month).

Reporting Instructions: Please report any child seen in the last month who meets the case definition, **including** those who have already been given a specific diagnosis.

Methods: Paediatricians reporting a child who presents with PIND will be sent an initial short (one page) contact form and asked to provide a telephone number (and fax number if available). They will then be contacted by the research nurse or research administrator to arrange a more detailed telephone discussion to gather further information. Thereafter, the research nurse may request a visit to the reporting paediatrician to review the case notes and discuss the case further. The PIND Research Group will not expect the referring paediatrician to discuss the notification of PIND with the child’s family. The aim of the PIND Research Group is to classify all cases of PIND and to identify any child with clinical features suggestive of CJD. If such a child is identified, the PIND Research Group will discuss that child with the referring paediatrician. If the referring paediatrician is in agreement, the child with suspected CJD would then be notified to the National CJD Surveillance Unit. Throughout, all patient data will be dealt with in strict confidence and the paediatrician managing the case will remain in control of patient referral. The PIND Research Group will not be contacting families directly.

Ethics Approval

Approved by Addenbrooke’s NHS Trust and the HPA.

Funding

Department of Health.

Reference

Will RG, Ironside JW, Zeidler M et al. A new variant of Creutzfeldt-Jakob Disease in the UK. Lancet 1996; 347: 921-5.