UKCP: a collaborative network of cerebral palsy registers in the United Kingdom

Geraldine Surman\textsuperscript{1}, Sandra Bonellie\textsuperscript{2}, James Chalmers\textsuperscript{3}, Allan Colver\textsuperscript{4}, Helen Dolk\textsuperscript{5}, Karla Hemming\textsuperscript{6}, Andy King\textsuperscript{1}, Jennifer J. Kurinczuk\textsuperscript{1}, Jackie Parkes\textsuperscript{7}, Mary Jane Platt\textsuperscript{8}

\textsuperscript{1}National Perinatal Epidemiology Unit, University of Oxford, Old Road Campus, Headington, Oxford OX3 7LF, UK
\textsuperscript{2}Applied Statistics Group, Dept of Mathematics, Merchiston, Napier University, 10 Colinton Rd, Edinburgh EH10 5DT, UK
\textsuperscript{3}Information and Statistics Division, Gyle Square, South Gyle Crescent, Edinburgh EH12 9ED, UK
\textsuperscript{4}University of Newcastle upon Tyne, Sir James Spence Institute, Royal Victoria Infirmary, Queen Victoria Rd, Newcastle-upon-Tyne NE1 4LP, UK
\textsuperscript{5}Room 1F08 Faculty of Life and Health Sciences, University of Ulster at Jordanstown, Shore Rd, Newtownabbey, Co. Antrim BT37 0QB, UK
\textsuperscript{6}University of Warwick, Coventry, UK
\textsuperscript{7}NICPR, Room 1.36 Mulhouse Bldg, The Queen's University of Belfast, Institute of Clinical Sciences, Belfast BT12 6BJ, UK
\textsuperscript{8}Division of Public Health, School of Population, Community and Behavioural Sciences, University of Liverpool, Liverpool, UK

Address correspondence to Geraldine Surman, E-mail: geraldine.surman@npeu.ox.ac.uk

ABSTRACT

Cerebral palsy (CP) is a relatively rare condition with enormous social and financial impact. Information about CP is not routinely collected in the United Kingdom. We have pooled non-identifiable data from the five currently active UK CP registers to form the UKCP database: birth years 1960–1997. This article describes the rationale behind this collaboration and the creation of the database. Data about 6910 children with CP are currently held. The mean annual prevalence rate was 2.0 per 1000 live births for birth years 1986–1996. Where type is known, 91 per cent have spastic CP. Where data are available, nearly one-third of children have severely impaired lower limb function, and nearly a quarter have severely impaired upper limb function. As well as describing the range and complexity of motor and associated impairments, the pooled data from the UKCP database provide a platform for studies of aetiology, long-term outcomes, participation and service needs. The UKCP database is an important national resource for the surveillance of CP and the study of its epidemiology in the United Kingdom.

Keywords cerebral palsy, collaborative research, epidemiology, UKCP

Background

In the United Kingdom, as in other developed countries, there have been increasing rates of preterm births and increasing survival of these early births over recent decades.\textsuperscript{1} Such preterm survivors, and especially those from multiple pregnancies, have a significantly greater risk of cerebral palsy (CP).\textsuperscript{2}

Infants with CP have an impact on families and how they cope with what may be a considerable change in their lifestyle and financial circumstances, on services necessary to support the children and their families and on the National Health Service (NHS) through claims for compensation. The rate of such claims across the United Kingdom is variable,\textsuperscript{3} and the overall cost to the NHS is increasing.\textsuperscript{3} Obstetrics accounts for some 60 per cent of the NHS litigation bill,\textsuperscript{4} £446 million in the year 2001/2002.\textsuperscript{5} Recent individual CP-related settlements have been as high as £6 million, and even unsuccessful claims can cost the NHS up to £0.5 million in legal costs. The majority of settlements relate to adverse events around birth and to children with CP.

With the increasing survival and a shift in emphasis from mortality to morbidity, concern has focused on the quality of life of children with CP. The need to participate in the activities of daily life for children with CP and their families is as important as for all families.\textsuperscript{6} Increased dialogue between families and those involved in research will enhance knowledge in this area.

Geraldine Surman, Researcher
Sandra Bonellie, Statistics Lecturer
James Chalmers, Consultant in Public Health Medicine
Allan Colver, Reader in Community Child Health
Helen Dolk, Professor of Epidemiology and Health Services Research
Karla Hemming, Statistics Research Fellow
Andy King, Computer programmer
Jennifer J. Kurinczuk, Consultant Clinical Epidemiologist
Jackie Parkes, Senior Lecturer
Mary Jane Platt, Senior Lecturer in Public Health Medicine
The UK Collaborative Network of Cerebral Palsy Registers and Surveys (UKCP) was formed in 1999. This article describes the establishment, characteristics and potential of the UKCP database as a national information and research resource for CP. The data analysis presented is for the purpose of describing the data held by UKCP, rather than describing the epidemiology of CP that will be reported elsewhere.

The cerebral palsy registers, databases and surveys

The collaboration comprises five active CP registers, databases and surveys in the United Kingdom, which will all be referred to as registers. The registers cover the birth populations of Northern Ireland and Scotland and the three former English health regions of Mersey, Northern and Oxford, around 15 per cent of England and Wales (Fig. 1). Each register has ethics committee approval.

All registers use multiple sources of ascertainment including child health systems, assessment centres, other registers, paediatricians, physiotherapists, death registration and periodic requests for notification. Capture–recapture analysis has been used by one register to assess ascertainment.7 Methods of monitoring data quality include validation carried out by Surveillance of Cerebral Palsy in Europe (SCPE)8 and by UKCP, double data entry and resolution of differences, inbuilt computer algorithms to identify values outside the expected range and data checking for reports and research studies.

As the registers were set up at different times (Table 1), with some collecting data retrospectively and some collecting data about newly diagnosed children, there is a variability in the completeness of data over time. Records of children in all registers are ‘flagged’ for death notification with the National Health Service Central Records System (NHSCR), the Registrar General for Scotland or the Central Services Agency for Northern Ireland as appropriate.

Merseyside and Cheshire Cerebral Palsy Register

Merseyside and Cheshire Cerebral Palsy Register (MCCPR) has been largely concerned with monitoring trends in the birth prevalence of CP, life expectancy and cause of death and the aetiological implications of those trends, with particular interest in multiple births.9,10 Births during 1966–1977 formed a retrospective cohort, and prospective data collection was from 1978. Having identified the cases, clinical information is then abstracted from obstetric and paediatric case notes. The original ethics approval specifically precluded any contact with parents, but clinicians’ concerns about providing access to information without informed consent (following the Alder Hey enquiry11) led to reduced ascertainment and data collection ceasing in 2000. Consent-based data collection has restarted with Department of Health funding to 2008.

North of England Collaborative Cerebral Palsy Survey

North of England Collaborative Cerebral Palsy Survey (NECCPS) focused on the development of the assessment6 of lifestyle and the quality of life. It has also studied differences over time in the pattern of severity and demonstrated how severity thresholds influence the completeness of ascertainment.12 Retrospective searches were carried out in 1980, 1985 and 1995 for the survey of births between 1960 and 1990 for three districts. Consent was sought when children...
were 4–5 years of age. From 1991, the survey was extended to the whole of the Northern Health Region, and data were collected prospectively from local convenors in each of the 16 former health districts. Data collection from 1991 births onwards has been continuous and consent obtained before notification to the survey.

**Northern Ireland Cerebral Palsy Register**

In 1991, the Northern Ireland Cerebral Palsy Register (NICPR)\(^{13,14}\) retrospectively identified cases of CP in children up to 14 years of age and then in all newly diagnosed cases. Preliminary notifications are obtained by sending cards every month to clinicians and the gait laboratory, supplemented by regular surveys of paediatric physiotherapists, interrogation of the child health system, outpatient lists and death certificates, resulting in multiple notification for 73 per cent of cases. Follow-up clinical information is sought from the child’s clinician; up to 1997, such information was available from 97 per cent of the cases.\(^{14}\) When the register had required parental consent, this was gained in 60 per cent of the cases, although only 2 per cent of parents actually refused. In 1997, the advisory and ethics committee lifted the requirement for consent, allowing retrospective data completion, but the situation is again under review.

**4Child—Four Counties Database of Cerebral Palsy, Vision Loss and Hearing Loss in Children**

4Child—Four Counties Database of Cerebral Palsy, Vision Loss and Hearing Loss in Children (formerly the Oxford Register of Early Childhood Impairments (ORECI)). This register began in 1984 following a pilot study in 1983.\(^{15}\) 4Child is unique in the United Kingdom in also collecting data about children with vision loss and hearing loss, irrespective of whether they have CP. Despite several NHS administrative boundary changes and now being covered by two Strategic Health Authorities, the 4Child catchment area remains Oxfordshire, Berkshire, Buckinghamshire and Northamptonshire. Paediatricians are contacted for clinical information when children are aged 3 and 5 years. 4Child is currently piloting a consent-based procedure. Information about 4Child is made publicly available through annual reports and on the internet.\(^{16}\)

**Cerebral Palsy Register for Scotland**

Cerebral Palsy Register for Scotland (CPRS) began as the Scottish Register of Children with a Motor Deficit of Central Origin. It was established by the Public Health Research Unit in Glasgow in 1990, retrospectively ascertaining cases from 1984.\(^{17}\) Administration of the original register was transferred to the Information Statistics Division of NHS Scotland when
funding ceased. Data for birth years 1984–1990 are held currently by UKCP.

CPRS seeks to identify cases from 1990 births, and the register was re-launched with charity funding in 2002 as a joint venture between the NHS and Napier University, from where it is now administered. A designated paediatrician in each Health Board helps co-ordinate the local registration of cases. Parents can also register their child directly via the register’s website.18

Parental consent is obtained, and parents of children on the former register are also contacted for permission to re-register the child. Consent includes entry into the register, obtaining clinical information from existing NHS systems and linking the child’s data with data from the mother’s pregnancy record. A particular strength of the Scottish register is the ability to link to routinely collected pregnancy data.

### The UKCP database

The common UKCP database collates core data items from each of the five collaborating registers (Table 2). No personal identifiable information is included in the common database that is held at the National Perinatal Epidemiology Unit.

#### Table 2 Core variables held on the UKCP common database

<table>
<thead>
<tr>
<th>Demographics and background</th>
<th>Clinical information</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td>Limb function</td>
</tr>
<tr>
<td>Year of birth</td>
<td>Which/number of limbs affected</td>
</tr>
<tr>
<td>Month of birth</td>
<td>Lower limb function</td>
</tr>
<tr>
<td>Birthweight</td>
<td>Severe impairment of lower limb function</td>
</tr>
<tr>
<td>Gestational age</td>
<td>Upper limb function</td>
</tr>
<tr>
<td>Residence in area at birth</td>
<td>Severe impairment of upper limb function</td>
</tr>
<tr>
<td>Number of babies at delivery</td>
<td>Type of cerebral palsy (CP) (Surveillance of Cerebral Palsy in Europe classification17)</td>
</tr>
<tr>
<td>Order of birth</td>
<td>Type of spastic CP</td>
</tr>
<tr>
<td>Mother’s age</td>
<td>Seizures</td>
</tr>
<tr>
<td>Subject alive or dead</td>
<td>Congenital malformation</td>
</tr>
<tr>
<td>Year of death</td>
<td>Postneonatal insult</td>
</tr>
<tr>
<td>Carstairs score</td>
<td>Age at postneonatal insult</td>
</tr>
<tr>
<td>Vision</td>
<td>Vision impairment</td>
</tr>
<tr>
<td>Hearing</td>
<td>Severe vision impairment</td>
</tr>
<tr>
<td>Hearing Impairment</td>
<td>Severe hearing impairment</td>
</tr>
<tr>
<td>Intellectual</td>
<td>Intellectual impairment (IQ &lt; 70)</td>
</tr>
<tr>
<td>Intellectual Level</td>
<td>Severe intellectual impairment (IQ &lt; 50)</td>
</tr>
</tbody>
</table>

### Classification and definitions

The classification of CP agreed by SCPE is used, and the SCPE core data items are included in the UKCP database in addition to items unique to UK registers. Categories of definition and severity of the impairments were agreed, and these are summarized in Table 3. Most registers do not have a stated upper age limit for postneonatal cases, but in practice this age is 5 years.

All children with CP from the individual registers are included in the UKCP database, including those with CP related to a defined event occurring after the first 28 days of life associated with subsequent clinical signs of CP (postneonatal or late impairment CP).

### Preliminary analyses

Data, abstracted in July 2004 about all children and held on the UKCP database for birth years 1960–1997, are used here to illustrate the range of data available. Information about all live births for the appropriate geographical areas was available only for 1976–1996, and therefore, rates per 1000 live births are presented for only those years. Proportions are used where denominator data were not available.
Characteristics of the children
There are 6910 records of children born 1960–1997 inclusive. Some children with CP die before a diagnosis can be confirmed, and historically, registers have taken different decisions about whether to record these early deaths. For consistency, the data about children who died before their first birthday are excluded from this analysis, leaving 6855 records. There were 6257 (91 per cent) children born to mothers resident at the time of delivery in the areas covered by the registers. Area of residence is unknown for 33 children (0.5 per cent), and 565 children (8 per cent) were born to mothers resident outside the register areas. The latter two groups of children were included for the purposes of the general discussion but excluded where rates have been calculated. CP rates were generally lower in the 1990s following a peak in the 1980s (Fig. 2), although mean prevalence of CP was 2.0 per 1000 live births for birth years 1986–1996 compared with 1.9 per 1000 live births for 1976–1985. Data about intellectual impairment for Scotland and vision impairment before 1975 for North of England were not systematically collected, and these centres’ data are excluded from the analyses of those items.

Postneonatal causes of cerebral palsy
Five hundred and thirty-seven children (8 per cent) are known to have had a postneonatal cause for their CP. The aetiology for these cases is different to those with pre- and perinatal causes, so although their data are included for the general discussion about CP and the range of impairments, their data are excluded from the analysis of rates.

Cerebral palsy subtypes
The number of children ascertained as having CP, of any type, by each register is listed in Table 1. The type of CP is dominated by spastic CP, with bilateral and unilateral spastic CP making up 91 per cent of cases on the database (Table 4). Rates per 1000 live births, between 1976 and 1996, for each of the registers, range from 0.8 to 2.0 for spastic CP and from 0.1 to 0.3 for non-spastic CP.

Impairments
The children have a wide range of impairments and varying severity of impairments. Registers’ different methods of data collection mean that some items of data are incomplete in the earlier years, for example, intellectual impairment and the presence of seizures. Where information was available, almost one-third of children had severely impaired lower limb function and nearly a quarter had severely impaired upper limb function (Table 4).

Migration
Some children will be lost to follow-up when they move out before diagnosis. UKCP does not currently hold information to analyse the proportions.

Deaths
The proportion of deaths recorded ranged from 7 to 14 per cent, although it should be noted that some registers hold data from as early as the 1960s. Over the whole database, 739 children (11 per cent) are known to have died by July 2004. Twenty-seven per cent (184/684) of deaths occurred between the ages of 1 and 4 years. Long-term survival has been extensively analysed and reported elsewhere.19

Discussion
Information about CP is not routinely collected in the United Kingdom. Child Health Information Systems were expected
to deliver this important service, but despite many years of discussion,20 little has yet been done to implement improvements to the range and quality of the data collected. For example, 52 per cent of children on the Northern Ireland CP register were not recorded as having CP on the Child Health System of the Health Board in Northern Ireland. 21 A further study,22 showing failures of local systems, strengthened the case for specialized registers. The new NHS National Programme for Information Technology seems to promise only integration of existing systems but not improvement.23

The CP rate reported here of around two per 1000 live births in the United Kingdom is in agreement with earlier analyses,24 and although CP is the commonest cause of serious motor impairment in young children, the number of children affected is not large. Although rates in particular subgroups, such as low birthweight, may be much higher, absolute numbers remain small, limiting investigation into the causes. To improve the reliability of estimated CP prevalence, rates need to be calculated from large numbers of births. Each of the registers is based in a different geographical area, and so caution is needed in interpreting the pooled data. Differing time periods, encompassing changing technologies and patterns of care, are also covered by the different registers. Pooling data can enable both differences and similarities to be investigated,25 and currently, the only means of describing the UK picture is to refer to this pooled database.

The variable nature of CP and associated impairments means that where only small numbers of cases exist, it is difficult to adequately describe the different patterns of impairments, either in their range or in their severity. Despite decades of CP research, debate about how best to describe the severity of disability continues. Subjective terms such as mild, moderate and severe or profound are in common usage in publications of the last 20 years but rarely clearly defined. Working closely together enables researchers in different centres to arrive at a common language to describe children with CP and to compare their lives with those without CP. Other disease registers have found such collaboration to be of great benefit, for example, the United Kingdom Association of Cancer Registries (UKACR)26 and

At the time that the collaboration began, the environment in which disease registers operate was changing. The Data Protection Act, 1998, requires that database custodians in the United Kingdom must do whatever is reasonable to ensure that subjects are aware of the likelihood of inclusion of their personal data on that database. The requirement that no personal information could be released by clinicians to registers without the specific consent of the data subject prompted cancer registry custodians to suggest that data collection would become unworkable. An additional clause, Section 60, was added to the Health & Social Care Act, 2001, to help address this concern and led to the setting up of the Patient Information Advisory Group (PIAG). All studies, research projects and registers intending to use personal identifiable information without specific consent were required to apply to PIAG for ‘Section 60 support’ which would allow them to carry on their work without either specific informed consent or anonymization of data for a transitional period. CP registers cannot anonymize their data, because they need to check for duplicate notifications and to follow children’s development over a period of years. Register staff are unable to obtain consent directly because they do not have a direct clinical relationship with children and their families, so consent must be sought by children’s clinicians. Although few parents refuse consent when approached, difficulties arise in the process. There may be uncertainty about who should ask for consent, consent may be sought repeatedly, or where a child is seen by several clinicians, the assumption may be made that consent will have been sought earlier. The situation for UKCP registers is complicated further as the Health & Social Care Act applies to only England and Wales, and this aspect of governance is still uncertain for Scotland and Northern Ireland.

Many registers are moving towards a fully consent-based system of ascertainment, although Newton asserts that it is sometimes impractical, or even impossible, for a disease register to use this system in its entirety, as it would be too onerous for both patients and clinicians. The British Isles Network of Congenital Anomaly Registers (BINOCAR) has tried to address this issue by developing a system of ‘section 60 support’ which allows registers to continue their work without either specific informed consent or anonymization of data for a transitional period.

Table 4  Cerebral palsy (CP) subtype, indicators of severity and associated impairments

<table>
<thead>
<tr>
<th>CP subtype</th>
<th>Total with available data</th>
<th>With this impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Number</td>
<td>% Range*</td>
</tr>
<tr>
<td>Spastic bilateral</td>
<td>3666</td>
<td>54–56</td>
</tr>
<tr>
<td>Spastic unilateral</td>
<td>2296</td>
<td>33–35</td>
</tr>
<tr>
<td>Spastic, laterality not known</td>
<td>28</td>
<td>0.4–0.4</td>
</tr>
<tr>
<td>Dyskinetic</td>
<td>362</td>
<td>5.3–5.5</td>
</tr>
<tr>
<td>Ataxic</td>
<td>243</td>
<td>3.5–3.7</td>
</tr>
<tr>
<td>Motor impairment</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Severe impaired lower limb function</td>
<td>6304</td>
<td>29–31</td>
</tr>
<tr>
<td>Severe impaired upper limb function</td>
<td>6178</td>
<td>21–23</td>
</tr>
<tr>
<td>Associated sensory impairment</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vision impairment†</td>
<td>5748</td>
<td>34–40</td>
</tr>
<tr>
<td>Severe vision impairment</td>
<td>5445</td>
<td>9–11</td>
</tr>
<tr>
<td>Hearing impairment</td>
<td>6026</td>
<td>7–8</td>
</tr>
<tr>
<td>Severe hearing impairment</td>
<td>6216</td>
<td>2.2–2.4</td>
</tr>
<tr>
<td>Associated intellectual impairment</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intellectual impairment‡</td>
<td>5229</td>
<td>39–51</td>
</tr>
<tr>
<td>Severe intellectual impairment</td>
<td>1612</td>
<td>24–31</td>
</tr>
<tr>
<td>Other associated impairments</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Seizures</td>
<td>3620</td>
<td>18–33</td>
</tr>
<tr>
<td>Congenital malformations</td>
<td>2757</td>
<td>7–18</td>
</tr>
</tbody>
</table>

Includes data from all UKCP registers, all years (as detailed in Table 1) and postneonatal CP.

*The range of proportions are shown where there are missing data, for example, for severely impaired lower limb function, 1968/6855 = 29%, 1968/6304 = 31%.

†Cases from the North of England before 1975 were excluded because information about vision impairment was not regularly collected up to that time.

‡Cases from Scotland for all years were excluded because information about intellectual impairment was not systematically collected.
The work of the UKCP owes much to Ann Johnson (Acknowledgements), whose research not possible with smaller databases and a sup-
port from Department of Health funding aimed specifically at research-active disease registers. Disease research registers in England alone were estimated to number nearer to 400 than the 250 identified, and competition for funding was fierce. The UKCP collaboration receives no direct funding. Whilst registers in Merseyside and Scotland continued passive accrual of new cases, ascertainment in the 1990s declined substantially because of underfunding. Long-term planning is essential for CP registers, as trends in CP rates can only be discerned when sufficient time has elapsed to allow a firm diagnosis to be made, typically around the age of 4 or 5 years. The quality of a disease register, its research output and the data collected are functions of continuity. Continuity of funding is among the key characteristics required to run a successful register. Following the Newton and Garner report, two of the three English CP registers received support from Department of Health funding aimed specifically at research-active disease registers. Disease research registers in England alone were estimated to number nearer to 400 than the 250 identified, and competition for funding was fierce. The UKCP collaboration receives no direct funding.

Initial UKCP collaborative work has successfully exploited facilities not available in many other areas of the world. Linkage to death notification has enabled the generation of survival data and the investigation of the relationship of survival to severity of disability. Linkage to census-generated area deprivation data using postcode is currently underway and will examine the relationship between CP and socio-economic factors. Future work is being planned to investigate trends in the type of CP, the effect of CP on the participation of adolescents and the frequency and diversity of multiple impairment.

Over the past 20 years, population-based CP registers in the United Kingdom have played a key role in monitoring rates of CP over time. They have provided a framework for epidemiological and clinical research and have been a source of information to guide the planning of services for children and adults with CP. Closer collaboration should bring benefits from the pooling of expertise, a larger database enabling research not possible with smaller databases and a supportive network of colleagues familiar with similar issues. The UKCP database is an important developing national resource for surveillance and research into the rates, causes and consequences of CP.

Acknowledgements

The work of the UKCP owes much to Ann Johnson (4Child), Peter Pharoah (MCCPR) and Steve Jarvis (NECCPS) whose energy, drive and commitment kept their respective registers running effectively for many years and contributed to the formation of the UKCP collaborative network and also to the statistical support provided by Jane Hutton, University of Warwick.

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