Surveillance of cerebral palsy in Europe: a collaboration of cerebral palsy surveys and registers

Surveillance of Cerebral Palsy in Europe (SCPE)

Correspondence to **Christine Cans**, RHEOP, 23, Av Albert 1er de Belgique 38000 Grenoble, France.
E-mail: christine.cans@imag.fr

Although cerebral palsy (CP) is the most common cause of motor deficiency in young children, it occurs in only 2 to 3 per 1000 live births. In order to monitor prevalence rates, especially within subgroups (birthweight, clinical type), it is necessary to study large populations. A network of CP surveys and registers was formed in 14 centres in eight countries across Europe. Differences in prevalence rates of CP in the centres prior to any work on harmonization of data are reported. The subsequent process to standardize the definition of CP, inclusion/exclusion criteria, classification, and description of children with CP is outlined. The consensus that was reached on these issues will make it possible to monitor trends in CP rate, to provide a framework for collaborative research, and a basis for services planning among European countries.

Cerebral palsy (CP) is one of the most severe disabilities in childhood and makes heavy demands on health, educational, and social services as well as on families and children themselves. In recent years, the probability of survival has increased even among children with a severe level of disability (Hutton et al. 1994). This means that appropriate services will need to be provided for children with CP through adolescence and into adulthood.

Research into the origins and management of CP must remain a high priority. During the past 20 years, much of the research interest has been focused in two areas. First, it is known that babies born very preterm or with a very low birthweight are at a higher risk of CP than babies born near term (Stanley and Alberman 1984). With the development and use of neonatal intensive care units throughout Europe and the associated decrease in neonatal mortality in most countries during the 1970s, 1980s, and 1990s, more of these vulnerable babies are surviving (McCormick 1993). There was some evidence of a rise in CP rates among low-birthweight babies during the 1970s and 1980s (Hagberg et al. 1989, Pharoah et al. 1990, Topp et al. 1997a) but the trends in recent years have been less clear (Meberg and Broch 1995, Hagberg et al. 1996, Pharoah et al. 1998). There has also been concern that the level of severity of disability may be increasing among children with CP (Stanley 1992, Uldall et al. 1995, Hagberg et al. 1996, Pharoah et al. 1996). Continued monitoring of the characteristics of children with CP is clearly necessary.

The second research area has focused on the relation between care given to mothers and babies around the time of birth and CP. Although it is widely agreed that only a very small proportion of children with CP have had a major adverse intrapartum event (Nelson et al. 1986, Blair and Stanley 1988, Gaffney et al. 1994), the inadequacy of markers of perinatal asphyxia result in continuing uncertainty and confusion in this area (Blair 1993). Many cases continue to be attributed to negligent care, therefore, better ways of identifying timing of brain insults are needed. Attention is now switching to the antenatal period with maternal infection and preeclampsia as possible causal factors (Grether and Nelson 1997).

Much of the research on CP has been based on registers of children with CP. Such registers exist in several places around the world: in Australia (Stanley and Watson 1992) and in Europe. Some European centres have been established for a long time, e.g. Ireland (Cussen et al. 1978), Denmark (Glenting 1982), and Sweden (Hagberg et al. 1975), as well as in Norway (Meberg 1990), and the UK (Jarvis et al. 1985, Pharoah 1987). In other places, such as France and Italy, the registers started more recently (Johnson 1989, Cans et al. 1996, Di Lallo et al. 1996). One of the difficulties however, is that although CP is the most common childhood disabilities, occurring twice as frequently as Down syndrome, CP occurs in only 2 to 3 per 1000 live births. Large populations are needed to amass sufficient numbers of participants to answer research questions, particularly when studying subgroups of children with CP, such as those born very preterm or with a very-low birthweight. To address these issues a network was formed across Europe of CP surveys and registers to monitor trends in the CP rate, to provide a framework for collaborative research, and as a basis for service planning. The collaboration was funded by the European Commission (Biomed2). It was recognized that difficulties might arise because of differences in the definition of CP,

inclusion and exclusion criteria used for surveys and registers, and the classification systems used. This paper begins by describing how the network was established and reports the prevalence rates of CP in the centres before any harmonization of data across centres. Once areas of difference were identified, a multidisciplinary approach was used to achieve a consensus on issues of definition and classification; the decisions reached are reported.

Method

Fourteen centres in eight countries within Europe were contacted in 1998 and invited to participate in an European network of CP surveys and registers. The network was called Surveillance of Cerebral Palsy in Europe (SCPE). All centres registered data on children with CP from geographically defined areas over varying periods of time. The location of centres with the number of live births each year within their geographic area are shown in Table I. Centres are ordered by size of the live birth population.

BIRTH COHORT PREVALENCE RATES BEFORE DATA HARMONIZATION

Each centre provided information on their most recent prevalence rate data from annual reports (five centres) or published articles (eight centres). The report or publication containing the most recent comparable data was chosen. Information was available from 13 of the 14 centres.

At this stage there was no attempt to examine differences between centres in CP definition or inclusion/exclusion criteria although, generally, children with CP were included only if ≥ 5 years old. Rates of CP based on at least 3 consecutive birth years were used. It was noted when children with postneonatal CP (i.e. when the event causing brain injury occurs some time after birth) were excluded. Apart from overall CP prevalence rate, birthweight-specific rates were retrieved where available. In other centres that could not produce birthweight-specific rates of CP, it was possible to examine the proportion of all children with CP within each birthweight group. Centres also provided information on the proportion of children in each CP subtype group and the proportion of children with CP who had intellectual impairment, based on IQ level. The ratio between the highest and lowest rates was used to describe the extent of the variation in prevalence rates.

QUESTIONNAIRES ON REGISTER/SURVEY METHODOLOGY

Between April and September 1998, detailed information was obtained about each register or survey by asking those responsible for the data in each of the 14 centres, to complete a detailed questionnaire. The questionnaire contained 93 items, including questions about the definition of CP used, methods of determining inclusion on the register, ways of describing children with CP, and the administrative and technical aspects of data collection and storage. In addition, demographic data relating to the area from which cases were drawn were also requested, including information on numbers of live births and neonatal deaths occurring between 1980 and 1990.

WORKING GROUPS

Five working groups were set up to address different aspects of the collaboration. One of the five groups was concerned with obtaining an agreed definition of CP, and a description and classification system for CP which could be used by the network. Representatives from 10 of the 14 centres participated in this working group which met on three occasions for 2 to 3 days in the first year of the project. The objective of this group was to obtain agreement on a definition for CP, to define inclusion and exclusion criteria, agree on a classification system, and define ways of describing levels of disability. Conclusions from this group were then presented to all participants at a plenary meeting and an agreed consensus document emerged.

Results

BIRTH COHORT CP PREVALENCE RATES BEFORE HARMONIZATION

Overall CP prevalence rate

The reported overall CP prevalence rates for 13 of the 14 centres are shown in Table II. The centres are ordered by CP prevalence rate, the highest rate reported first. Of the 12 centres with data on all CP cases, the prevalence rate varied from 1.5 to 3 per 1000 live births. This difference does not seem to be accounted for by differing time periods or by inclusion or exclusion of postneonatal cases. One centre included children with bilateral spastic CP only (Germany); if the assumption is made that bilateral spastic CP accounts for 50 to 60% of all cases of CP, then the estimated overall rate for this centre would be 2.2 per 1000 live births.

Birthweight-specific CP rates

Half of the centres could provide information on birthweight-specific rates of CP. These are shown in Table III, in order of birth years included in each centre. The widest variation between rates is in the group weighing less than 1500 g; the ratio of the highest to lowest reported rate is 3.4. In the group weighing 1500 g to 2499 g, the ratio is 2.2, and in those weighing 2500 g and more the ratio is 1.5. In the latter group the rates are similar, if the centre collecting data only on children bilateral spastic CP is excluded. Rates are consistently higher in the lower birthweight groups as expected, and seem to increase over time. There is no clear trend over time in the rates for the two groups above 1500 g, although this could have been obscured by the considerable overlap of the birth years included within each cohort.

Table I: Centres included in SCPE with number of live births per year and birth years covered by the survey/register

Location of centre	*Birth years in survey/register*	*Register or survey*	*Nr live births per year*
Scotland, UK	1984–1996	R	60 000
Mersey region, UK	1966–1996	R	35 000
Oxford, UK	1984–1993	R	35 000
East Denmark, Denmark	1950–1990	R	30 000
Northern Ireland, UK	1977–1998	R	25 000
Göteborg, Sweden	1954–1990	R	20 000
East Ireland, Ireland	1976–1994	R	20 000
Tübingen, Germany	1973–1986	S	17 000
Gelderland, Netherlands	1977–1988	S	15 000
Isère, France	1980–1989	R	14 000
Haute Garonne, France	1976–1985	S	10 000
Cork and Kerry, Ireland	1966–1989	R	10 000
Northern region, UK	1966–1999	R	10 000
Viterbo province, Italy	1977–1992	R	3 000
Total		11 (R), 3 (S)	304 000

Subtypes of CP

Twelve centres had information on CP subtypes. Methods of description and classification varied; two centres described the number of limbs involved and the neurological findings rather than using the traditional clinical terminology (e.g. diplegia, quadriplegia, hemiplegia). Two centres reported their data on subtypes in a pooled form; the centre which had included only children with bilateral spastic CP had no children with hemiplegia in their cohort. Findings are summarized in Table IV. The proportion of all CP children described as having hemiplegic CP varied from 18% to 36% and there was a similar wide variation in the group with diplegia: 13% to 55%. Six centres identified a group with ataxia (included within the 'other' group on Table IV). The proportion of children with CP in this group varied between 1% and 7%.

Proportion of those with CP with learning disabilities

In 10 centres the proportion of children with learning disabilities was estimated. Definitions differed between centres and this was reflected in the proportion reported, from 23% to 56%. In two centres the reported rate of children with severe intellectual impairment (IQ less than 50) was 30% and 41% respectively. In the other centres the reported rates of children with any learning disability (IQ < 70) varied from 23% to 44%.

ISSUES ARISING FROM PARTICIPANT QUESTIONNAIRES

Participants' responses revealed four areas where there were important differences between centres: definition and inclusion/exclusion criteria, case ascertainment, interobserver error, and method of classification and recording.

Definition

All centres were asked for their definition of CP. Eight centres used the definition compiled by Bax (1964), and three centres used the definition by Mutch and colleagues (1992). The three remaining centres used different definitions (Mac Keith 1959, Ingram 1984, Rumeau-Rouquette et al. 1997).

In the first meeting of the working group it was decided that rather than compile another definition of CP, participants should continue to use the definition of their own choice, providing that it included the following five key elements: CP is a group of disorders i.e. it is an umbrella term; it

Table II: Overall CP prevalence rate in centres in Europe before harmonization of data

Birth years	*Overall CP/1000 rate (95% CI)*	*Source of information*
1983–86	3.00[b] (2.69–3.31)	Topp et al. 1997a, East Denmark
1966–70	2.4 (2.02–2.88)	Cussen et al. 1978, Cork and Kerry, Ireland
1987–91	2.37[b] (2.11–2.63)	Annual report 1997, Northern Ireland, UK
1987–90	2.36[a] (2.05–2.67)	Hagberg et al. 1996, Göteborg, Sweden
1984–91	2.30[b] (2.15–2.51)	Annual report 1996, Oxford, UK
1985–88	2.03 (1.22–2.84)	Di Lallo et al. 1996, Göteborg, Sweden
1982–84	2.00 (1.72–2.28)	Pharoah et al. 1990, Mersey, UK
1976–81	1.95 (1.73–2.17)	Dowding et al. 1988, East Ireland
1991–93	1.90[a, b] (1.65–2.15)	Annual report 1997, Northern region, UK
1984–86	1.50 (1.32–1.68)	Annual report 1992, Scotland, UK
Spastic bilateral CP only		
1975–1986	1.22[b] (1.07–1.37)	Krageloh-Mann et al. 1994, Tübingen, Germany
Overall CP rate/1000 resident children		
1982–84	2.61[b] (2.03–3.19)	Rumeau-Rouquette et al. 1997, Haute Garonne, France
1980–87	1.70 (1.46–1.94)	Annual report 1995, Isère, France

[a]Includes some children under the age of 5 years.
[b]Post-neonatal cases excluded.
Birth years included for estimate of prevalence rate.

Table III: Birthweight specific rates of CP in seven European centres

Birth years included	*Centre*	*Nr children with CP*	*Rate /1000 live birth for birthweight groups*		
			<1500 g	*1500–2499 g*	*≥ 2500 g*
1975–86[a]	Tübingen, Germany	249	39.4	10.9	0.5
1976–81[b]	East Ireland,Ireland	258	28.2	6.4	1.2
1977–92	Viterbo, Italy	89	29.2	14.1	1.0
1982–84	Mersey, UK	190	59.2	9.7	1.1
1984–91	Oxford, UK	644	55.9	10.3	1.4
1987–90	Göteborg, Sweden	206	62.2	13.9	1.4
1987–91[c]	Northern Ireland, UK	319	95.5	13.1	1.2

[a] Includes only children with bilateral spastic CP.
[b] Rate per 1000 total births.
[c] Children with unknown birthweight were assumed to have the same distribution as known birthweight.

is permanent but not unchanging; it involves a disorder of movement and/or posture and of motor function; it is due to a non-progressive interference/lesion/abnormality; this interference/lesion/abnormality is in the developing/immature brain.

Inclusion and exclusion criteria

Participants were then asked on the questionnaire about the inclusion and exclusion criteria they used including the age at which cases were included in analyses.

Age of registration. All centres recognized the changing clinical picture in young children with motor disorders; the lowest age at registration was 3 years. Three years was the lower limit for four centres, two of which rechecked the diagnosis at age 5 years. In two centres the lowest age was 4 years and in the remainder, the children were at least 5 years old when registered. Participants agreed that for the purposes of the collaboration, children should be at least 4 years old before being included on the European database.

Postneonatal cases. Two centres did not register children with CP of postneonatal origin; among the remainder, there were differences on the age cut-off used to define children with 'postneonatal' CP. Ten of the 12 centres agreed that a lower age limit of 28 days should be used. The other two used a 7 days cut off period. However, there was wide variation in the upper age limit, ranging from 1 year to 7 years. Participants agreed that, as the time of the aetiological event causing the CP can be precisely timed in these children, differences in the upper age limit could be taken into account in the analysis.

Other inclusion and exclusion criteria. These differed from centre to centre: in nine centres written guidelines were used and two registers used the recently published guidelines of Badawi and colleagues (1998). Two areas of particular concern were identified by participants. First there was uncertainty about the inclusion or exclusion of children with a recognized syndrome or chromosome anomaly. Secondly, there were different views about whether to include children with severe hypotonia. After considerable discussion it was agreed that if the 'rules' of the definition were adhered to and the neurological signs of one of the subtypes of CP were present (Table V) children with recognized syndromes, brain malformations, or chromosome anomalies should be included, but clearly identified as such on the database. It was also agreed that children with hypotonia but no other neurological signs should not be included. However, if ataxia was also present, children were to be included as having an ataxic CP subtype.

A decision tree which incorporated all these consensus views was devised as a guide for centres to assist with determining inclusion or exclusion of children in the European database (Fig. 1). It was also considered to be potentially useful for centres establishing new registers and surveys.

Case ascertainment

Centres used a range of sources for case finding. In all centres, except one, hospital sources were used for ascertaining cases of CP, mainly from paediatric, neonatology, child neurology, and rehabilitation departments. Community sources, such as community paediatricians, general practitioners, health visitors, and physiotherapists, quite often report children with CP to registers in Ireland, UK, and Italy, but not in the other centres. Social sources, such as financial support services and parents associations, were used as sources in only a few centres. The same heterogeneity between centres can be observed regarding the use of educational sources, such as special schools and education centres, for reporting cases of CP. All the centres recognised the importance of multiple sources of ascertainment and used more than one source to compile their databases. Many centres did not record the number of reporting sources per case, and it was not possible to precisely measure the extent of multiple

Table IV: Proportion of children with CP within subtype groups

Birth years included	*Centres*	*Nr of children with CP*	*CP subtype (% of all CP)*			
			Hemiplegia	*Diplegia*	*Quadriplegia*	*Other*
1982–1984	Isere France	186[a]	22	14	36	28
1984–1991	Scotland, UK	502	21	22	33	24
1966–1975	Cork and Kerry, Ireland[b]	254	33	25	20	22
1987–1991	Northern Ireland, UK	319	34	19	30	17
1987–1990	Göteborg, Sweden	206	34	45	9	12
1976–1981	East Ireland	258[a]	25	44	17	14
1991–1993	Northern region, UK	222[a]	35	13	43	9
1984–1991	Oxford, UK	697	24	13 (2[c]) 4 (3[c]) 44 (4[c])	–	15
1975–1986	Tübingen, Germany	249[a]	n/a	59 (2[d]) 14 (3[d]) 20 (4[d])	–	7
1984–1989	Mersey, UK	497	36	22	35	7
1983–1986	East Denmark	324[a]	18	55	10	17

[a] Postneonatal cases excluded.

[b] Cussen et al. (1979).

[c] Number of affected limbs.

[d] Number of affected limbs taking into account predominant localization of the disorder.

reporting. Active and passive search techniques for identifying cases were used, either alone or in combination; some registers/surveys used only active searching through clinical records or other listings, while a number relied completely on passive reporting. This involved the cooperation of professionals in health and educational areas who were asked to report new cases to the register.

The ways in which data on each child were collected also differed. In some centres, information was entered onto a data collection form by a clinician, while in others, the information on each child was abstracted from medical records. A third area where there were differences among centres was in the procedures used for coding and classifying the data. In three centres a clinician coded the data, in five centres, an epidemiologist, and in the remaining five, different staff including a research nurse, a research assistant, a midwife, and a technician coded data. In six centres there was an intermittent check on coding consistency.

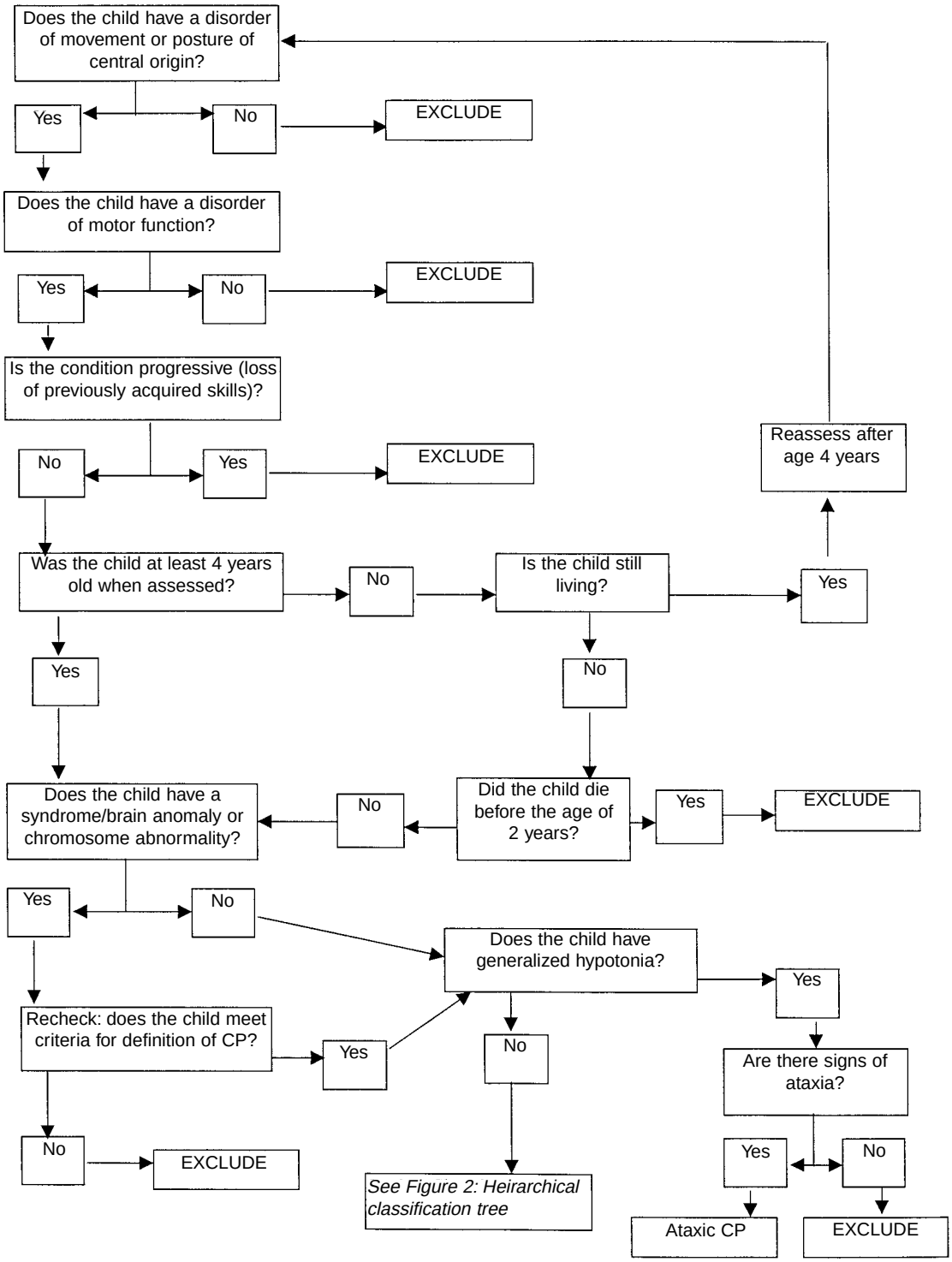

Figure 1: *Decision tree for inclusion/exclusion of cases of cerebral palsy on SCPE register.*

Three areas of case ascertainment gave rise to particular concern. Children with CP who die in the early years of life may not be recorded on registers and surveys, but such children need to be included when estimating birth-cohort prevalence rates. Although half the centres regularly scrutinized death certificates, it is widely recognized that the term CP often does not appear on death certificates. A second group of children who tend to be overlooked were identified as those with very severe disabilities and in long-term residential care. Children with mild disease where a diagnosis of CP is not consistently made are also less likely to be recorded. It was agreed that by providing information on level of severity of motor impairment or disability on the data base, these children could, when necessary, be excluded when estimating birth-cohort prevalence rates.

Interobserver error

A number of reasons for interobserver differences were identified. First, the experience of the person examining the child differed considerably from centre to centre, and possibly within centres. In some centres all children were seen by a single experienced observer, while in others, children were reported to the register by a range of professionals, including health personnel involved in primary care, paediatricians, neurologists, and physiotherapists. These may change over time within one register or survey. In other centres, information was abstracted from clinical notes. Here, further differences may arise because of the quality of information in the notes, the experience of the person examining the child, and the experience of the person abstracting the notes. Participants recognized the problems of dealing with errors arising in these variations. As the method of case identification varied so much from centre to centre it did not seem possible to develop a single way of checking data reliability. However, each centre was asked to devise an appropriate quality check on data reliability, to

Table V: Definitions adopted for European classification of cerebral palsy

Spastic CP is characterized by at least two of:
Abnormal pattern of posture and/or movement
Increased tone (not necessarily constant)
Pathological reflexes (increased reflexes: hyperreflexia and/or pyramidal signs e.g. Babinski response)
Spastic CP may be either bilateral or unilateral
Spastic bilateral CP is diagnosed if:
Limbs on both sides of the body are involved
Spastic unilateral CP is diagnosed if:
Limbs on one side of the body are involved
Ataxic CP is characterized by both:
Abnormal pattern of posture and/or movement
Loss of orderly muscular coordination so that movements are performed with abnormal force, rhythm, and accuracy
Dyskinetic CP is dominated by both:
Abnormal pattern of posture and/or movement
Involuntary, uncontrolled, recurring, occasionally stereotyped movements
Dyskinetic CP may be either dystonic or choreo-athetotic
Dystonic CP is dominated by both:
Hypokinesia (reduced activity, i.e. stiff movement)
Hypertonia (tone usually increased)
Choreo-athetotic CP is dominated by both:
Hyperkinesia (increased activity, i.e. stormy movement)
Hypotonia (tone usually decreased)

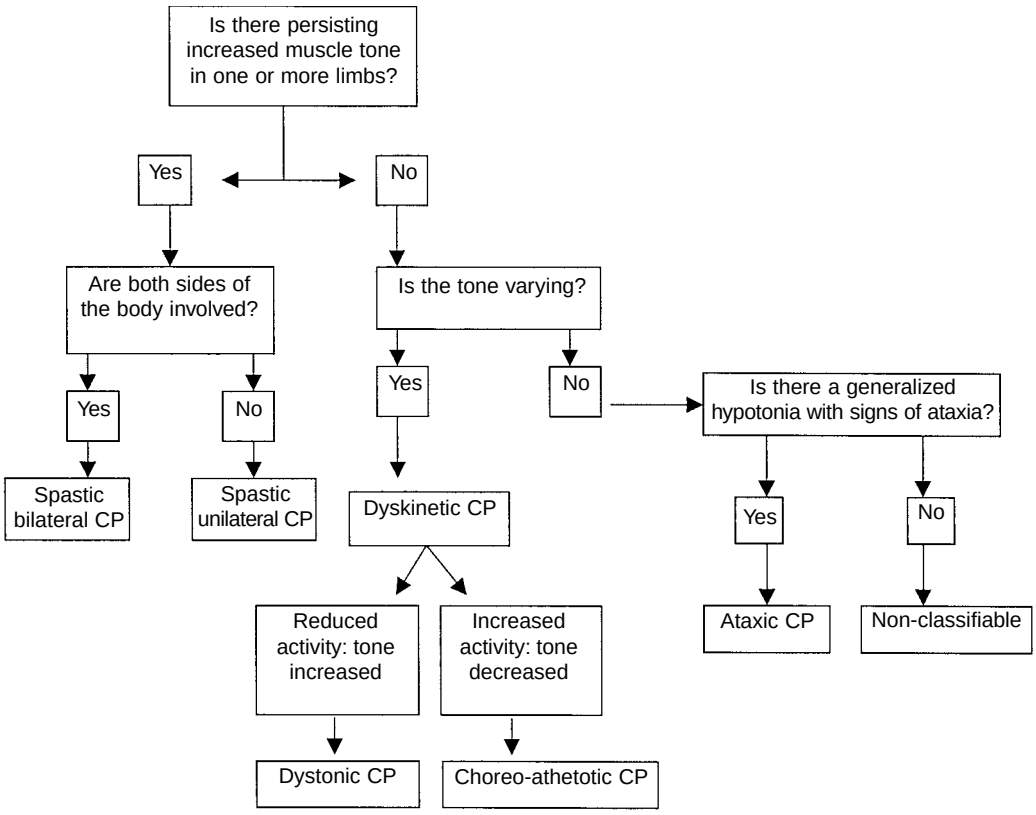

Figure 2: *Hierarchical classification tree of cerebral palsy sub-types.*

include a review random checks by a second observer and a review of children on registers with an atypical clinical picture or with an 'unclassifiable' subtype of CP.

Method of classification and recording.
The questionnaire revealed that there was a wide variation in the forms used and in the methods of classifying children with CP. Four centres were using the Standard Form for Describing Children with a Central Motor Deficit, devised by Evans and coworkers (1989), and a further two used a modified version of this form. The remaining centres were using forms of their own design. It became apparent to participants that there were wide variations in the use of terms such as spastic diplegia and spastic quadriplegia in classifying children. A simple, reliable system of classification was needed for SCPE; the hierarchical classification agreed is shown in Figure 2. Definitions for each subtype are shown in Table V. It was recognized that as surveys and registers had established their own recording systems over many years it would not be feasible to suggest the use of a standard form across European centres. Rather the participants agreed that each centre would reclassify the children on their own database to whatever level was feasible on the agreed SCPE classification. The classification system was tested in a validation exercise in which all centres participated. This will be reported separately, but it reassured participants that, in general, similar children were being classified in a similar way.

Discussion

This review of existing information on CP prevalence rates in 13 centres included in the SCPE collaboration showed that before harmonization of data, there were differences in overall CP rates, and in particular there were large and important variations in birthweight-specific CP rates and in the proportion of children in the different subtypes. From the questionnaire responses, it seemed that these differences might be explained in a number of ways.

First, differences in rates might well be due to the differences in the definition of CP used by centres. CP is an umbrella term (Mutch et al. 1992) and the criteria for including and excluding a child from a survey or register may differ from centre to centre. Consensus on definition of CP and agreement about the criteria by which children should be included or excluded on CP registers was an important first step for the European collaboration. Over the years there have been many attempts to obtain an internationally agreed definition of CP and to standardize the way in which children with CP are described (Bax 1964). There have been recent attempts to compile inclusion and exclusion criteria (Badawi et al. 1998, Williams and Alberman 1998) although the concepts used are not universally accepted. SCPE has adopted a pragmatic approach, and without major changes in existing local data systems, we have reached a consensus on definition, inclusion, and exclusion criteria, and a simple classification system using five key points. Input from different disciplines means that the decisions made in reaching a consensus were well informed at both a clinical and epidemiological level.

The 'five-key points' approach to the definition of CP circumvented lengthy debate on the exact wording of a definition, as did the clear guidelines which emerged on age for inclusion. Other inclusion and exclusion criteria were less straightforward. With the increasing use of brain imaging diagnostic procedures and genetic studies, syndromes, chromosomal abnormalities, and brain lesions are now detected more frequently. Exclusion or inclusion of these based on aetiological criteria alone will alter prevalence rates and make it difficult to interpret trends over time. Our approach to this issue has been to agree that children with syndromes, chromosome anomalies, or developmental brain anomalies will be included on the database if they meet the clinical criteria of the agreed definition of CP, and excluded if they do not. The agreement reached will need to be reviewed as work on the database progresses. Reliability may be improved by developing a reference manual and possibly an interactive video. The list of diagnoses and the clinical presentation of both the included and excluded children will be reviewed from time to time for consistency, as we recognize that this as an area where inconsistency may lead to spurious differences in prevalence rate (Williams and Alberman 1998). Second, there are likely to be differences in case ascertainment. The principle of multiple sources of ascertainment is widely accepted and the importance of this has been highlighted by work in Denmark (Topp et al. 1997b), in the UK (Parkes et al. 1998, Johnson and King 1999), and in France (Guillem et al. 2000). In all these reports, comparisons between different sources of children with CP demonstrated that serious under-ascertainment occurs if only one source is used. Comparison of rates of children with more severe levels of functional loss may help to detect differences in level of ascertainment of milder cases. The age at which children with CP are registered is also crucial; ascertainment which is too early may be unreliable as children with milder CP are missed or over reported and the clinical picture may also change over time. Population migration can result in 'loss' of children with CP from the birth population. Population movement varies from area to area and some centres report a net gain of children with CP into their area. Prevalence rates will differ, therefore, if current residence is a criterion for defining either the numerator (CP children currently resident) or the denominator (number of children currently resident in the area; Baille et al. 1996).

Differences in proportion of subtypes of CP reflect a long-standing problem in terminology. The traditional clinical terminology used to describe the subtypes of CP has been confusing (Ingram 1984), and the standard form for describing children with a central motor deficit is an attempt to avoid these terms (Evans et al. 1989). A similar but more precise approach was used in the collaborative work done by groups in Göteborg and Tübingen (Krageloh-Mann et al. 1993).

The classification system adopted by the participants of SCPE is a simple and practical one. It is recognized that the clinical implications of different clusters of neurological signs will differ from child to child. However, for epidemiological purposes, we chose to impose a hierarchical system which would be more likely to place similar children in a subtype group. By this system, the presence of predominantly increased tone places a child in one of the spastic subtypes even when additional signs of dyskinesia are present. The spastic subtypes can be defined by the number of limbs involved and whether the involvement is unilateral or bilateral. This approach is similar to the one used in the collaborative work between Sweden and Germany (Krägeloh-Mann et al. 1993), and avoids the use of confusing terminology. It also

allows participants to classify the children from their own database to whatever level is feasible, depending on the detail and quality of the available data. A final problem of reaching a consensus about level of severity of functional loss is still ongoing. There appeared to be wide differences between centres and, at this stage, the collaboration has agreed to a simple system for describing walking ability and defined appropriate cut-off points to describe learning disability, vision, and hearing loss.

The overall consensus reached on definition and classification reflects a remarkable collaboration across several disciplines including obstetrics, neonatology, paediatrics, neuropaediatrics, rehabilitation medicine, and epidemiology. Participants from these different specialities inevitably have differing perspectives on information about children with CP. Epidemiologists administering CP registers generally base decisions about inclusion and exclusion on 'second-hand' reports of neurological signs and diagnostic labels, and focus on classification and grouping of children with a similar clinical picture. Clinicians, on the other hand, are able to be more precise in the way they describe children and will include only forms of CP with clear neurological signs (i.e. spasticity, dyskinesia, ataxia) whatever the diagnostic label. Although in the past this has given rise to differing views of the purpose and value of surveys and registers, SCPE has provided a unique opportunity for a number of disciplines in different CP registers to openly discuss these issues and reach a consensus. A model for this type of cooperation had been given by the collaborative work done by two groups, one in Sweden and one in Germany. These two centres decided to pool and compare data on children with CP; after discussion a remarkable consensus in description and classification of children emerged, and close prevalence rates of bilateral spastic CP and similar distributions of disabilities were observed (Krägeloh-Mann et al. 1994). With case definition and classification agreed among participants from the 14 centres, it becomes possible to detect 'true' differences in prevalence rates. Caution will be given as two countries, UK and Ireland, provided two thirds of the whole number of live births per year. The differences could be related to variations in demographic factors, for example, socioeconomic level (Dowding and Barry 1990), to maternal or neonatal risk factors, or to differences in perinatal practice and health policy. Examining neonatal mortality rates in relation to CP rates may be helpful in understanding such differences (Ens-Dokkum et al. 1994), and information on mortality as well as on perinatal care organization will be collected as a result of this collaboration.

With the work of harmonization and agreement over classification now almost complete, SCPE is developing a database of children with CP who were born between 1980 and 1990 from a total live-born population of over 300 000 per year. Once established, it will be possible to monitor trends and variations in birthweight-specific rates of CP and examine the changing patterns of CP subtypes. The next step will then be to use the database as a framework for research. It will be a powerful tool which can be used to explore aetiological questions, the impact of changes in care on prevalence of CP, and address other issues of importance to service providers, parents, and the children themselves.

Accepted for publication 23rd March 2000.

Acknowledgements

This work was supported by European Commission funds: DGXII-BIOMED2, Contract N°BMH4-983701.

List of SCPE participants: C Cans and P Guillem (RHEOP, Grenoble, FR), F Baille and C Arnaud (INSERM U518, Toulouse, FR), J Chalmers (ISDSHS, Edinburgh, UK), G Cussen and V McManus (Lavanagh Centre, Cork, IE), J Parkes and H Dolk (Belfast University, UK), G Hagberg and B Hagberg (Göteborg University, Göteborg, SW), O Hensey and V Dowding (Central Remedial Clinic, Dublin, IE), S Jarvis and A Colver, (University of Newcastle, Newcastle, UK), A Johnson and G Surmann (NPEU, Oxford, UK), I Krägeloh-Mann and R Michaelis (Tübingen University, Tübingen, DE), P Pharoah, MJ Platt (University of Liverpool, Liverpool, UK), M Topp and P Udall (DIKE, Copenhagen, DK), MG Torrioli and M Miceli (Lazio Cerebral Palsy register, Rome, IT), M Wichers (Groot Klimmendal, Arnhem, NL), O van Nieuwenhuizen (Utrecht, NL).

References

Badawi N, Watson L, Petterson B, Blair E, Slee J, Haan E, Stanley F. (1998) What constitutes cerebral palsy? *Developmental Medicine & Child Neurology* **40:** 520–7.

Baille MF, Arnaud C, Grandjean H, Cans C, Rumeau-Rouquette C. (1996) Prevalence des déficiences de l'enfant: aspectsméthodologiques. *Archives of Public Health* **53**(Suppl.1): 13. (Abstract).

Bax MCO. (1964) Terminology and classification of cerebral palsy. *Developmental Medicine & Child Neurology* **6:** 295–307.

Blair E. (1993) A research definition for 'birth asphyxia'. *Developmental Medicine & Child Neurology* **35:** 449–55.

— Stanley FJ. (1988) Intrapartum asphyxia: a rare cause of cerebral palsy. *Journal of Pediatrics* **112:** 515–9.

Cans C, Billette A, Fauconnier J. (1996) Un registre des handicaps de l'enfant en Isère. *Revue d'Epidémiologie et de Santé Publique* **44:** 287–90.

Cussen GH, Barry JE, Moloney AM, Buckley NM, Crowley M, Daly C. (1978) Cerebral palsy: a regional study. *Irish Medical Journal* **71:** 568–72.

— — — — — — (1979) Cerebral palsy–a regional study: part II. *Irish Medical Journal* **72:** 14–8.

Di Lallo D, Miceli M, Mastroiacovo P, Schiripa G, Perucci CA. (1996) Cerebral palsy register in central Italy: methodology and preliminary results. *Paediatric and Perinatal Epidemiology* **10:** A1. (Abstract).

Dowding VM, Barry C. (1988) Cerebral palsy: changing patterns of birthweight and gestational age (1976/81). *Irish Medical Journal* **81:** 25–9.

— Barry C. (1990) Cerebral palsy: social class differences in prevalence in relation to birthweight and severity of disability. *Journal of Epidemiology and Community Health* **44:** 191–5.

Ens-Dokkum MH, Johnson A, Schreuder AM, Veen S, Wilkinson AR, Brand R, Ruys JH, Verloove-Vanhorick SP. (1994) Comparison of mortality and rates of cerebral palsy in two populations of very low birthweight infants. *Archives Disease in Childhood, Fetal Neonatal* **70:** F96–100.

Evans P, Johnson A, Mutch L, Alberman E. (1989) A standard form for recording clinical findings in children with a motor deficit of central origin. *Developmental Medicine & Child Neurology* **31:** 119–27.

Gaffney G, Sellers S, Flavell V, Squier M, Johnson A. (1994) Case control study of intrapartum care, cerebral palsy and perinatal death. *British Medical Journal* **308:** 743–50.

Glenting P. (1982) Cerebral palsy in Eastern Denmark 1965–1974. I: Decreased frequency of congenital cases. Cerebral palsy register of Denmark. Report no VII. *Neuropediatrics* **13:** 72–6.

Grether JK, Cummins SK, Nelson KB. (1992) The California Cerebral Palsy Project. *Paediatric and Perinatal Epidemiology* **6:** 339–51.

— Nelson KB. (1997) Maternal infection and cerebral palsy in infants of normal birthweight. *Journal of the American Medical Association* **278:** 207–11

Guillem P, Lavergne C, Cans C. (2000) How to calculate the completeness of ascertainment of a morbidity register? Example of the Isere childhood handicap register in Grenoble, France. *Revue d'Epidémiologie et de Santé Publique* **48:** 41–51.

Hagberg B, Hagberg G, Olow I. (1975) The changing panorama of cerebral palsy in Sweden 1954-1970. II. Analysis of the various syndromes. *Acta Paediatrica Scandinavica* **64:** 193–200.

Hagberg B, Hagberg G, Olow I, von Wendt L (1989) The changing panorama of cerebral palsy in Sweden. V: The birth year period 1979–1982. *Acta Paediatrica Scandinavica* **78:** 283–90.
— — — — (1996) The changing panorama of cerebral palsy in Sweden. VII. Prevalence and origin in the birth year period 1987–90. *Acta Paediatrica* **85:** 954–60.
Hutton JL, Cooke T, Pharoah PO. (1994) Life expectancy in children with cerebral palsy. *BMJ* **309**: 431–5
Ingram TTS. (1984) A historical review of the definition and classification of the cerebral palsies. In: Fiona Stanley and Eva Alberman, editors. *The Epidemiology of the Cerebral Palsies.* Oxford: Blackwell Scientific. p 1–11.
Jarvis SN, Holloway JS, Hey EN. (1985) Increase in cerebral palsy in normal birthweight babies. *Archives of Disease in Childhood* **60:** 1113–21.
Johnson A, King R. (1989) A regional register of early childhood impairments: a discussion paper. *Community Medicine* **11:** 352–63.
— — (1999) Can routine information systems be used to monitor serious disability? *Archives Disease in Childhood* **80:** 63–6.
Krageloh-Mann I, Hagberg G, Meisner C, Schelp B, Haas G, Eeg-Olofsson KE, Selbmann HK, Hagberg B, Michaelis R. (1993) Bilateral spastic cerebral palsy: a comparative study between south-west Germany and western Sweden. I: Clinical patterns and disabilities *Developmental Medicine & Child Neurology* **35:** 1037–47.
— — — — — — — — — (1994) Bilateral spastic cerebral palsy – a comparative study between south-west Germany and western Sweden. II: Epidemiology *Developmental Medicine and Child Neurology* **36:** 473–83.
McCormick MC. (1993) Has the prevalence of handicapped infants increased with improved survival of the very low birthweight infant? *Clinics in Perinatology* **1:** 263–77
Mac Keith RC, Mackenzie ICK, Polani PE. (1959)Memorandum on terminology and classification of 'cerebral palsy' *Cerebral Palsy Bulletin (Developmental Medicine & Child Neurology)* **1:** 27–35.
Meberg A (1990) Declining incidence of low birth weight – impact on perinatal mortality and incidence of cerebral palsy. *Journal of Perinatal Medicine* **18:** 195–200.
— Broch H. (1995) A changing pattern of cerebral palsy: declining trend for incidence of cerebral palsy in the 20-year period 1970–89. *Journal of Perinatal Medicine* **23:** 395–402.
Mutch L, Alberman E, Hagberg B, Kodama K, Perat MV. (1992) Cerebral palsy epidemiology: where are we now and where are we going? *Developmental Medicine & Child Neurology* **34:** 547–51.
Nelson KB, Ellenberg JH. (1986) Antecedents of cerebral palsy: multivariate analysis of risk. *New England Journal of Medicine* **315:** 81–6.
North of England Collaborative Cerebral Palsy Survey. (1997) *Annual report.*
Northern Ireland Cerebral Palsy Project. (1997) *Annual Report.*
Oxford Register of Early Childhood Impairment. (1996) *Annual Report.*
Parkes J, Dolk H, Hill N. (1998) Does the child health computing system adequately identify children with cerebral palsy? *Journal of Public Health Medicine* **20:** 102–4.
Pharoah PO, Cooke T, Cooke RW, Rosenbloom L. (1990) Birthweight specific trends in cerebral palsy. *Archives Disease in Childhood* **65:** 602–6.
— Platt MJ, Cooke T. (1996) The changing epidemiology of cerebral palsy. *Archives Disease in Childhood, Fetal Neonatal* **75:** F169–73.
Pharoah POD, Cooke T, Rosenbloom I, Cooke RWI. (1987) Trends in birth prevalence of cerebral palsy. *Archives of Disease in Childhood* **62:** 379–84.
— — Johnson MA, King R, Mutch L. (1998) Epidemiology of cerebral palsy in England and Scotland, 1984–9. *Archives Disease in Childhood, Fetal Neonatal* **79:** F21–5.
Registre des Handicaps de l'Enfant et Observatoire Perinatal de l'Isere. (1995) *Annual report.*
Rumeau-Rouquette C, Grandjean H, Cans C, du Mazaubrun C, Verrier A. (1997) Prevalence and time trends of disabilities in school-age children. *International Journal of Epidemiology* **26:** 137–45.
Scottish Register of Children with Central Motor Deficit. (1992) *Annual Report.*
Stanley F, Alberman E. (1984) Birthweight, gestational age and the cerebral palsies. In: Stanley F, Alberman E, editors. *The Epidemiology of the Cerebral Palsies. Clinics in Developmental Medicine. No. 87.* London: Spastics International Medical Publications. p 57–68.
Stanley FJ. (1992) Survival and cerebral palsy in low birthweight infants: implications for perinatal care. *Paediatric and Perinatal Epidemiology* **6:** 298–310.
— Watson L (1992) Trends in perinatal mortality and cerebral palsy in Western Australia,1967 to 1985. *British Medical Journal* **304:** 1658–63.
Takeshita K, Ando Y, Ohtani K, Takashima S. (1989) Cerebral palsy in Tottori, Japan: benefits and risks of progress in perinatal medicine. *Neuroepidemiology* **8:** 184–92.
Topp M, Uldall P, Langhoff-Roos J. (1997a) Trend in cerebral palsy birth prevalence in eastern Denmark: birth-year period 1979–86. *Paediatric and Perinatal Epidemiology* **11:** 451–60.
— Langhoff-Roos J, Uldall P (1997b) Validation of a Cerebral Palsy Register. *Journal of Clinical Epidemiology* **50:** 1017–23.
Uldall PV, Topp MW, Madsen M. (1995) Congenital cerebral paresis in Eastern Denmark, birth years 1971–1982. *Ugeskr Laeger* **157:** 740–2.
Williams K, Alberman E. (1998) The impact of diagnostic labelling in population-based research into cerebral palsy. *Developmental Medicine & Child Neurology* **40:** 182–5.