

## Recommendations from the SCPE collaborative group for defining and classifying cerebral palsy

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**Introduction:** Several aims of classification can be argued: (1) monitoring subgroups of cerebral palsy (CP) whose prevalence rate is expected to change over time; (2) aetiological research on CP, according to the neurological subtypes and to the birth circumstances; (3) evaluation of interventions in children with CP, according to the severity of the motor impairment; and (4) comparisons with other studies. The act of classifying requires to have some properties, and among them the three most important are the reliability, the validity, and the simplicity.

Reliability is the consistency or repeatability of classifying, i.e. different persons at different time periods will classify a group of CP children in the same way. Accuracy of the reliability (reproducibility) can be assessed through statistical measurement (kappa coefficient), and has to be tested between professionals (observer) and within the same observer at different time periods (intraobserver).

Validity means that we are measuring what we are supposed to measure, i.e. the quality of its effectiveness. In deciding if a classification is valid or not, we come back to the aims of the classification, since its relevance may vary according to its use, e.g. a classification based on etiological circumstances might not be relevant for evaluating intervention therapies.

Simplicity is the quality of being simple or uncompounded or, in other words, 'easy to use' by anybody. It would not be practical or easy to require an expert child neurologist to examine a child for an hour in order to classify that child as having CP and this would, therefore, be difficult to recommend on a large scale.

It has always been difficult to compare data from CP registers and thus be sure that the differences observed reflect actual 'true' differences, or are due to differences in definition, criteria, and classification. This may concern CP prevalence rates, trends in CP prevalence rates, or comparison of characteristics between CP subtypes. For instance, one study (Meberg and Broch 1995) showed a decrease in occurrences of CP during the 1980s whilst all other studies had shown an increase (Hagberg and Hagberg 1996, Pharoah and Cooke 1997, Topp et al. 1997). More recently, there have also been some differences between authors in the one country regarding the changing trends in the severity of CP (Colver et al. 2000, Surman et al. 2003).

In 1998, a collaborative network of CP registers and population-based surveys was established with the help of funds

from the European Commission. The reasons for this collaborative effort were: (1) the need for standardization and harmonization of the definition, inclusion/exclusion criteria, and the characteristics used currently for describing children with CP, and (2) the need to get large numbers in order to be able to analyze distinct subgroups of CP and, in particular, their trends over time. The Surveillance of Cerebral Palsy in Europe (SCPE) network started in 1998 with 14 centres from eight countries, and at present there are 22 centres from 15 countries. The aim of this network was to: (1) harmonize CP data collection using a standard CP definition and an agreed minimum data set; (2) to develop a central database of children with CP in order to monitor trends in birthweight specific rates, and to provide information for service planning; and (3) to provide a framework for the development of collaborative research projects in CP field (SCPE 2000).

**Methods:** Agreeing on classification: During the first 3 years of the network, a subgroup of professionals, comprising child neuro-paediatricians, child rehabilitation doctors, and epidemiologists, representatives from different centres, met together several times before being able to propose a consensus on definition, criteria, and classification of CP. This consensus was presented to all participants of the SCPE network during a plenary meeting held in Oxford in 1999. Besides the skills already described above, other professional skills were present at this meeting, i.e. neonatologists, obstetricians, paediatricians, geneticists, and public health doctors. There was more discussion but also a strong desire to reach a consensus, and thus the SCPE criteria and classification system for CP were agreed.

Implementing the classification: A few months after this meeting it appears that difficulties remained when pooling and comparing information from different sources. The persisting problems were mainly due to the matter of language since not all partners from the different countries were English native speakers. Not everyone had derived the same meaning from terms such as 'increased tone' and 'walking fluently'. Thus, during the next 3 years, collaborative efforts were put together, mainly between child neuro-paediatricians, in order to develop a video-based tool, the SCPE Reference and Training Manual (SCPE R&TM). The aim of this tool was to promote a shared understanding of the words and phrases used to describe the clinical, functional, and neurological features of CP. Text and video material were first discussed within the small group of child neuro-paediatricians and then proposed to illustrate these features and to discuss pitfalls in diagnosis and classification. Interobserver exercise has been performed before spreading widely the use of this SCPE R&TM. After a few years of use, the hope is that it will help to improve the harmonization and standardization level between different CP registers/studies, and that it will encourage new registers in new countries to join the SCPE network.

During the latest years of the SCPE, we have been still working on the data quality and also toward the improvement of available information on denominators within the EURO-PERISTAT project.

**Results:** SCPE CP definition and criteria: Several definitions of CP already exist in the literature. However although these may vary in wording, they are broadly similar, and can be summarized as follows:

*Cerebral Palsy is a group of permanent, but not*

*unchanging, disorders of movement and/or posture and of motor function, which are due to a non-progressive interference, lesion, or abnormality of the developing/immature brain.* This definition specifically excludes progressive disorders of motor function, defined as loss of skills previously acquired in the first 5 years of life.

For any study of CP to be valid, there must be agreement on the 'similar characteristics' of the cases eligible for inclusion. SCPE has spent time agreeing on inclusion and exclusion criteria that should accompany CP definition (SCPE 2000).

Since there are great variations in ability of performing diagnosis in different places, and techniques for these diagnosis are improving over time, the CP definition must be simple and rely on phenomenology (clinical picture and history) criteria and not on aetiology criteria. The CP definition must be valuable and logical for both epidemiologists and clinicians, and, by implication, must be independent of the country in which the child lives.

**Inclusion criteria: Optimal age:** CP is not an easy diagnosis. It needs time to be confirmed. Premature diagnosis might lead to over-ascertainment (because of transient anomalies in preterm babies) or under-ascertainment, e.g. in mild unilateral spastic cases or ataxic cases. CP, as stated above, is not an unchanging condition, with the clinical picture in some cases altering as a child develops. It was agreed that age 5 years was the optimal age for confirmation of diagnosis.

**What about children who die early?:** It is recognized that some children with severe CP are correctly diagnosed at a young age, but die before their 5th birthday. Exclusion of these children could result in under-estimation of the prevalence of CP in Europe. Also when studying the aetiology, it would be better to include these cases, for instance cases of hypoxic-ischemic encephalopathy who die early. In fact a compromise was needed, and as a group, SCPE had followed the recommendation from Hagberg that we should not include children with CP who die too early, i.e. before the age of 2 years, and that children with clear signs of CP who die between the ages of 2 and 5 years must be included.

**No upper age limit** of onset of CP (in children with a post-neonatal cause) was identified. But it is useful to isolate CP cases of post-neonatal origin, defined as cases arising from an aetiological event 27 completed days after birth.

**Exclusion criteria:** All progressive *conditions* resulting in loss of acquired skills are excluded. However, we recognize that some progressive disorders might be registered wrongly as CP, due to the delay required, in some circumstances, to confirm a diagnosis of progressive disorder. However, the proportion of these misdiagnosed CP cases does not represent more than a few per cent of all CP cases, at least in the SCPE data.

Children with hypotonia as the sole clinical feature and children with isolated spinal neural tube defects should also be excluded from the CP cases.

An interesting paper had suggested a list of conditions that should or should not be considered 'cerebral palsy' (Badawi et al. 1998). This led to numerous discussions between epidemiologists and clinicians, but finally SCPE has agreed not to adopt this classification system but to rely solely on the clinical features of a case to determine eligibility.

**SCPE CP classification scheme:** Classification means 'the basic cognitive process of distributing children with CP into classes or categories of the same type'. Different classification systems for CP serve different functions, but

for epidemiological purposes, classifications systems based on clinical findings are currently the most widely used.

Drawing on published work, SCPE has classified CP into three main groups, which are based on clear neurological signs indicating pathology in the cerebral motor systems, e.g. spastic, ataxic, and dyskinetic CP.

All CP subtypes have an abnormal pattern of movement and posture in common.

**Spastic CP cases** have increased tone and pathological reflexes, either increased reflexes, e.g. hyper-reflexia or pyramidal signs, such as Babinski response. Increased tone in spasticity is characterized by an increased resistance which is velocity dependent (Sanger et al. 2003). A spastic catch is felt some time after onset of movement. Clonus is often associated with hyper-reflexia. It is considered pathological when it is prolonged or does not stop spontaneously. Pathological posturing of lower limbs is characterized by: (1) internal rotation of the hip; (2) hip adduction; and (3) equinus foot, resulting in a 'scissored' position.

**Dyskinetic CP cases** present involuntary, uncontrolled, recurring, and occasionally stereotyped movements. The primitive reflex patterns predominate, and the muscle tone is varying. SCPE uses dystonic and choreo-athetotic CP subtypes for subgrouping.

Dystonic CP is dominated by abnormal postures (may give the impression of hypokinesia) and hypertonia (tone fluctuating, but easily elicitable tone increase).

Characteristics are involuntary movements, distorted voluntary movements, and abnormal postures due to sustained muscle contractions (slow rotation, extension, flexion of body parts). Choreo-athetotic CP is dominated by: hyperkinesia and hypotonia (tone fluctuating, but mainly decreased).

Chorea means rapid involuntary, jerky, often fragmented movements. Athetosis means slower, constantly changing, writhing, or contorting movements.

In some cases, however, it may be difficult to delineate these subgroups when features are present from both. Then the term dyskinetic CP should be used.

**Ataxic CP cases** present loss of orderly muscular coordination, so that movements are performed with abnormal force, rhythm, and accuracy. Abnormal pattern of movement in ataxic CP is characterized by: (1) Loss of orderly muscular coordination, so that movements are performed with abnormal force, rhythm, and accuracy. Typical features are trunk and gait ataxia (disturbed balance) and past pointing (over- or undershooting of goal directed movements). (2) Tremor is another common sign (mainly a slow intention tremor). (3) Low tone is also a prominent feature.

**Mixed CP forms:** When it is a mixed CP form, i.e. spasticity with ataxia and/or dyskinesia, the child should be classified according to the dominant clinical feature.

Pure dyskinetic movement disorder does not show hyper-reflexia with clonus nor pyramidal signs. But in dyskinetic CP, these signs of spastic disorder may be present. The dominating features should determine subtype classification. Also, in spastic CP, some dystonic features are often present, especially when the upper extremities are involved. A dystonic posturing of the hand would, however, not be sufficient to classify a child as having the dystonic form of dyskinetic CP. The dystonic posturing of the trunk, arms, and face in the presence of lower-limb spasticity would qualify, however, as predominant dyskinetic features, thus, dystonic

CP (Krägeloh-Mann et al. 1993).

Motor function impairment in CP children: SCPE choice was to recommend the scoring of motor function according to: (1) the Gross Motor Function Classification System (GMFCS) for the lower limbs function (Palisano et al. 1997), <http://www.can-child.ca>; and (2) Bimanual Fine Motor Function (BFMF) for the upper limbs function. This last choice was achieved only very recently, and in order to conform with the S property (simplicity). A study has now shown the possibility to use this BFMF scoring through medical notes (Beckung and Hagberg 2002). However it has not yet been validated.

Since SCPE does not recommend the use of diplegia/quadruplegia terms, and recommends using instead the term bilateral spastic CP subtypes, the two motor function scales can then be used for describing children with CP according to the functional grading given. For instance, a child with bilateral spastic CP may be 'scored' as GMFCS Level IV and BFMF Level II – which for the clinician involved would give the feature of a diplegia – and another child with unilateral spastic CP may be scored as GMFCS Level II and BFMF Level I.

Associated impairments in CP children: The SCPE collaborative group recommends collecting information on four associated impairments. These recommendations are the minimum information that should be collected for those wishing to pool data or to compare it with data from other centres/countries.

**Intellectual impairment:** The cognitive impairment should be classified according to the thresholds recommended by the World Health Organization. These thresholds are shown in Table I.

**Table I: Thresholds of cognitive impairment as classified by the World Health Organization**

Normal	IQ > 85, attendance of regular school without support
Borderline	IQ 70–84
Mild impairment	IQ 50–69, some basic literacy and numeracy achieved
Moderate to severe impairment	IQ 20–49
Profound impairment	IQ < 20

For **visual and hearing impairment**, the recommendation is to determine the presence or absence of such impairment, and then to classify the impairment as severe or not, according to the visual acuity (<0.1 in both eyes after correction) or hearing loss (more than 70 dB in the better ear before correction).

**Epilepsy** can be defined as two unprovoked seizures, neonatal seizures being excluded. Firstly it must be known if the child 'had ever' or 'never had' epilepsy. Then it will be grouped as severe epilepsy if the epilepsy is still active.

**Discussion:** SCPE trees (SCPE 2000) are used for categorizing children with CP. Firstly, the decision tree is based on the presence of disorder of 'movement and/or posture' and of motor function. Secondly, the classification tree relies on neurological signs and topography for distinction between CP subtypes. By doing so, CP cases that are difficult to classify are not so numerous and less than 5% are observed in data from European centres (SCPE 2002).

At the beginning of the SCPE network, it was decided to

use the words 'bilateral/unilateral spastic', with, in addition, the numbers of limbs involved, instead of using the words 'diplegia, tetraplegia'. After a while, the disappointment was great when we observed persisting important differences between centres on the 'theoretically' harmonized data. The overlap between the 'diplegia/quadruplegia' groups in CP classifications has been well described in a recent paper (Colver and Sethumadhavan 2003). These differences, between two and four limbs for example, could not be explained by anything else than by coding differences. Despite having agreed on a text definition and classification categories, large variations in classifying CP cases were still shown in a cross-validation exercise. The distinction between the number of limbs affected, used by several centres, in opposition to the number of limbs predominantly affected used by other centres, was the main reason responsible for these differences.

Thus SCPE's recommendation moved to a more simple categorization, i.e. classifying spastic CP cases in unilateral versus bilateral CP cases. Bilateral spastic CP was not further subdivided into arm/leg-dominated, diplegia/quadruplegia, nor 2-limb/3-limb/4-limb dominated, due to the great inter-rater variability when these terms are not defined using functional scores respectively for upper and lower limbs.

In a different way the Australian group gives an example that harmonization within one country may authorize more detailed description and classification than what is possible when dealing with several different countries. They are using four levels (minimal, mild, moderate, severe) to describe severity of neurological signs in each limb. However, there is still discussion in Australia about the overlap between triplegia, diplegia, and quadruplegia CP subtypes, and the need for an international consultation was expressed (Blair and Watson 2005).

We agree with Eve Blair's recent comment (2005) that, presently, no satisfactory scale is able to classify multiple deficits, i.e. the GMFCS score and, even more so, the BFMF score, are certainly influenced by the presence/absence of associated intellectual impairment, and thus they do not describe only the motor function. However, these scoring systems are very helpful for epidemiological purposes and evaluation of care.

The reasons for SCPE choosing the BFMF scoring system rather than the MACS (Eliasson et al. 2006) are that: (1) BFMF takes into account possible asymmetry in the hand functions, whilst MACS does not; and that (2) BFMF can be retrieved from written medical records whilst MACS cannot. When collecting data on children with CP for CP registers or surveys, the situation of not directly examining the child is quite common.

In the US there was an attempt to classify children with CP according to severity criteria based on the functional ability of the most affected limbs, i.e. severe involvement meaning no useful function, and moderate involvement meaning the preservation of some function with or without the use of assistive devices. In most studies using data from this California survey, the children with mild involvement or pure hypotonia were excluded (Grether et al. 1992).

The results of the workshop in Washington was an agreement on a more accurate CP definition, with details given for each word of the definition (Bax et al. 2005). This constitutes a great step forward, although this is probably not sufficient. There is a need for precision on inclusion/exclusion criteria

so that the definition can be used, not only as a concept, but also in a pragmatic approach. In the same manner that the GMFCS scoring system is proposed with video pieces and written explanations for training, CP definition should be provided with tools that help in applying the definition criteria. For instance within the SCPE network it was clear that there was a need for further training tools to clarify SCPE written classification guidelines with illustrations, i.e. using video description of children with CP, such as in the SCPE R&TM. Now this manual has been already translated into eight different European languages: French, German, Italian, Lithuanian, Swedish, Spanish, Slovenian, and Dutch, and it will soon be available in Portuguese. Also, alongside the decision and classification trees proposed by SCPE, an ideal data collection form might be very useful for harmonizing the available data used to classify the CP cases. The SCPE Data Collection Form (SCPE-DCF) is one of such possible standardized forms. It comprised the minimum common items that normally should be available for registration everywhere in each country. This form is available on the SCPE website (<http://www-rheop.ujf-grenoble.fr/scpe2>).

A very positive result of the SCPE harmonization work was to highlight interesting characteristics or trends in some subgroups of CP that needed large numbers before any analysis. After application of the inclusion/exclusion/classification criteria for CP cases, pooling data from several centres allows SCPE to show a four-fold increased risk of CP in multiple birth mainly explained by gestational age distribution (Topp et al. 2004), a decreasing trend of infection as cause of post-neonatal CP case (Cans et al. 2004), an optimal birthweight associated with a lower risk of CP (Jarvis et al. 2003), and a decreasing CP prevalence in children with a birthweight between 1000 and 1500g (Platt et al. Forthcoming). Pooling data on CP cases already 'harmonized' was also very useful in a research project on quality of life and participation of children with CP (Colver 2006).

With its CP definition, criteria, and classification, SCPE has got agreement on a 'minimum data set or minimum description' of a child with CP, i.e. a common language which enabled us to build up a reliable database throughout Europe. In addition to this basic description, it is of course possible to go into more detail in some countries, and/or for specific substudies, according to different interests. This could concern more detailed description of the disability profile with respect to cognitive functions, or of the quality of life and participation of CP children and their family, or of additional orthopaedic problems or of information on aetiology/pathogenesis (neuroimaging, genetic findings).

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