Children with cerebral palsy in Europe: figures and disability

on behalf of SCPE Collaborative Group
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Surveillance of Cerebral Palsy in Europe

SCPE is a European network with two aims:

i) monitoring CP rates and trends using harmonised and standardised tools, and

ii) studying disability characteristics of CP children, causal pathways and impact on families.
Surveillance of Cerebral Palsy in Europe

• Why SCPE network?
• How does SCPE collaboration work?
• What does SCPE network achieve?
Why SCPE network?

- Cerebral Palsy is an umbrella term
- Cerebral Palsy is the most common cause of motor deficiency in children
- Cerebral Palsy has been related to perinatal events
- Cerebral Palsy has been ascertained in population based studies for >50y
Severe mental retardation (IQ<50)

Mean 29.5%  Min 22.0%  Max 44.5%
### Table II: Overall CP prevalence rate in centres in Europe before harmonization of data

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

*aIncludes some children under the age of 5 years.

*bPost-neonatal cases excluded.

Birth years included for estimate of prevalence rate.
Why SCPE network? 1/3

- Are the observed differences in prevalence rate «true» differences?

- How can we explain differences in the proportion of CP children with severe intellectual impairment?

→ Are we talking about the same CP?

HARMONISATION & STANDARDISATION
Why SCPE network?

• Are we all talking about the same CP?
  Little 1862, Freud 1893, Mc Keith 1958, Bax 1964, Ingram 1984, Mutch 1992
  more recently Washington workshop Bax 2005

• Terms in use:
  Motor deficiency of central origin, encephalopathy, Hemiplegia, neurosensory impairment,
  Brain disability, congenital ataxia, Little syndrome, diplegia

Colver AF, Sethumadhavan T. The term diplegia should be abandoned
POOLING DATA IN ORDER TO MONITOR RARE EVENTS

Distribution of 'Walking ability' by 'Type of CP'

- **Total without unknown walking ability (n=5908)**
  - Unaided walking: 52.7%
  - Walking with aids: 16.7%
  - Unable to walk even with aids: 30.7%

- **Ataxic (n=245)**
  - Unaided walking: 71.8%
  - Walking with aids: 18%
  - Unable to walk even with aids: 10.2%

- **Dyskinetic (n=374)**
  - Unaided walking: 19%
  - Walking with aids: 23.8%
  - Unable to walk even with aids: 57.2%

- **Spastic hemiplegic (n=1827)**
  - Unaided walking: 90.7%
  - Walking with aids: 5.8%
  - Unable to walk even with aids: 3.4%

- **Spastic bilateral 3 or 4 limbs (n=1822)**
  - Unaided walking: 11.5%
  - Walking with aids: 18.2%
  - Unable to walk even with aids: 70.4%

- **Spastic bilateral 2 limbs (n=1640)**
  - Unaided walking: 60.8%
  - Walking with aids: 25.2%
  - Unable to walk even with aids: 14%
POOLING DATA IN ORDER TO MONITOR RARE EVENTS

- Important increase in the rate of VLBW/multiple live births but few VLBW/multiple CP cases in each centre
  - 60 CP cases per year if area of 30 000 live births is covered
  - Among them only 12 below 32 weeks GA, and only 6 multiple

- Several CP sub-types

→ We need large numbers to analyse and interpret changes
Why this network?

COLLABORATIVE EFFORTS IN RESEARCH ON CP

- Few researchers in CP field, in childhood disability field
- Multi-centre studies need preliminary harmonisation between centres

→ Development of first SCPE joint proposal through a number of meetings

SURVEILLANCE OF CEREbral PALSY IN EUROPE
CONSENSUS ON TOOLS

- Guidelines for inclusion/exclusion
- Decision and classification trees
- Standardised Data collection Form
- Reference and training Manual

→ Sharing, translating and disseminating these tools
How does SCPE collaboration work?

CONSENSUS ON TOOLS

- Guidelines for inclusion/exclusion
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Sharing, translating and disseminating these tools
Cerebral Palsy is a group of disorders; it is permanent but not unchanging; it involves a disorder of movement and/or of posture and of motor function, it is due to a non progressive interference/lesion/abnormality in the developing/immature brain.

How do we decide if it is a CP case?

- INCLUSION criteria according to the clinical features, and not based on etiology (historically included syndromes)
  → Phenomenological approach

  Optimal age at registration 4-5 years old
  CP diagnosis made after the age of 2

  Disorder of movement/posture
  AND Disorder of motor function

  *e.g. a clumsy child is not a CP*
  *or neurological signs alone are not CP*
Figure 1: Decision tree for inclusion/exclusion of cases of cerebral palsy on SCPE register.

1. Does the child have a disorder of movement or posture of central origin?
   - Yes: EXCLUDE
   - No: REASSESS after age 4 years

2. Does the child have a disorder of motor function?
   - Yes: EXCLUDE
   - No: REASSESS after age 4 years

3. Is the condition progressive (loss of previously acquired skills)?
   - No: REASSESS after age 4 years
   - Yes: EXCLUDE

4. Was the child at least 4 years old when assessed?
   - No: Is the child still living?
   - Yes: REASSESS after age 4 years

5. Is the child still living?
   - Yes: EXCLUDE
   - No: Did the child die before the age of 2 years?

6. Did the child die before the age of 2 years?
   - Yes: EXCLUDE
   - No: Does the child have a syndrome/brain anomaly or chromosome abnormality?

7. Does the child have a syndrome/brain anomaly or chromosome abnormality?
   - No: Recheck: does the child meet criteria for definition of CP?
   - Yes: EXCLUDE

8. Recheck: does the child meet criteria for definition of CP?
   - Yes: Are there signs of ataxia?
   - No: EXCLUDE

9. Are there signs of ataxia?
   - Yes: Ataxic CP
   - No: EXCLUDE

See Figure 2: Hierarchical classification tree
How do we classify a CP case?

- Different classifications historically based on neurological signs/severity: Hagberg, Ingram, Australia on the topography of the altered motor function: diplegia/quadriplegia on the associated impairments: intellectual, sensorial on limb function assessment and other activity limitations: ability to walk, to dress, to eat, ..... on the (presumed) origin of the CP: prenatal, perinatal, postnatal on ....

- Classifications need to be simple to be as reproducible as possible
SCPE Recommendation for Neurological signs

Abnormal pattern of posture/movement

- **Spasticity**
  - Increased tone
  - Hyperreflexia / pyramidal signs

- **Dyskinesia**
  - Involuntary, uncontrolled, recurring stereotyped movements
  - Primitive reflex patterns predominate
  - Muscle tone is varying

- **Ataxia**
  - Movements are performed with abnormal force, rhythm and accuracy


+ Decision for mixed forms
SCPE Recommendation for the associated impairments

- **Intellectual impairment**
  - IQ < 50
  - IQ 50 to 69 or clinical estimate
  - IQ >= 70

- **Visual impairment**
  - Severe visual impairment defined by vision <0.1 in both eyes after correction

- **Hearing impairment**
  - Severe hearing impairment defined by loss >70dB before correction on the better ear

- **Epilepsia (2 unprovoked seizures)**
  - Never/ever
  - Is it still on medication?
SCPE Recommendation for activity limitation

- GMFCS: between the 4th and 6th birthdays
  5 levels, up to age 12, → CanChild website
SCPE current recommendations

- **STEP 1**  CP case YES/NO
- **STEP 2**  CP TYPE based on neurological signs and topography
- **STEP 3**  Assessing severity of the motor function loss
  - GMFCS for lower limbs
  - BFMF for upper limbs
- **STEP 4**  Assessing presence and severity level of associated impairments
How does SCPE collaboration work? 2/3

• COMMON DATABASE ON CP CHILDREN
  - Data on CP cases and denominators from each centre
  - Birth cohort, years 1976-1998
  - 11 300 CP cases from 18 centres in 10 countries

• SCPE WEB SITE
  http://www-rheop.ujf-grenoble.fr/scpe2/site_scpe
How does SCPE collaboration work?

FUNDED BY THE EUROPEAN COMMISSION

- Concerted action, BIOMED4, 1999-2001
- Accompanying measure, RFP5, 2002-2004
- DG SANCO, EURO-PERISTAT, 2005-2007

- Funded activities: workshops, visits, meetings, standardised tools & common database development, coordination work
How does SCPE collaboration work?

- **SCPE1 1999-2001**
  - Harmonisation of definition and inclusion/exclusion criteria
  - Common database (children born 1975-1990)
  - Support to research projects

- **SCPE2 2002-2004**
  - Reference and Training Manual on CP, using video pieces
  - QoL- SPARCILE study on CP children

- **SCPE3 2005-2007 - EUROPERISTAT**
  - Improving information on denominators, and quality of the data
  - Encouraging the implementation of new registers

*SCPE Collaborative Group. Dev Med Child Neurol 2000; 44:633-40*
EURO-PERISTAT

• Public Health programme leaded by INSERM U 149 (Paris) G Breart / J Zeitlin
• AIM : defining and pooling common perinatal indicators across Europe, perinatal health and its determinants
• CP is one of the selected indicator for future developpement
• SCPE is part (2 WP out of 10) of the whole project

if perinatal conditions are different between
the 25 EU countries, shall we expect some differences
in CP prevalence rates ??
What does SCPE network achieve?

RELEVANT INFORMATION ON CP SUBGROUPS

– CP PREVALENCE: 2 / 1000 live births
– CP rate in >2500 g: 1 / 1000 live births
– CP rate in 1500-2500 g: 10 / 1000 live births
– CP rate in <1500 g: 100 / 1000 live births
Figure 5: Trends in rate of CP from 1976 to 1989.
SCPE data 1980-1996
N=8979 CP cases

- 26 % < 32 weeks GA
- 19 % 32-36 weeks GA
- 54 % >= 37 weeks GA

- 30 % were unable to walk
- 30 % had a severe intellectual impairment
- 12 % had a severe visual impairment

What does SCPE network achieve?

- CP in children from multiple births
  

- CP cases of postneonatal origin
  

- CP in VLBW children
  

Rates of one and two sided spastic Cerebral Palsy from 9 European countries 1980-1996 (3 yr moving average)
NEW INSIGHTS

• on the consequences of CP, e.g. SPARCLE study

• on some risk factors of CP, e.g. Growth retardation
NEW INSIGHTS

- on the consequences of CP, e.g. SPARCLE study
- on some risk factors of CP, e.g. Cerebral Palsy and intra-uterine growth retardation

Cerebral Palsy and intra-uterine growth retardation

Cerebral Palsy and intra-uterine growth retardation

Rates of CP by growth Z score with denominators adjusted for neonatal mortality
Figure: Box and whisker plots of KIDSCREEN quality of life scores by domain for children with cerebral palsy and children in the general population.

The scales for each domain have mean=50 and SD=10 in the reference population.²⁶
Description of children with cerebral palsy

- **STEP 1** CP case YES/NO
- **STEP 2** CP TYPE based on neurological signs and topography
- **STEP 3** To assess severity of the motor function loss
  - GMFCS for lower limbs
  - BFMF for upper limbs
- **STEP 4** To assess presence and severity level of associated impairments
- **STEP 5** To assess QoL and Participation
What does SCPE network achieve? 3/3

ENCOURAGING OTHER COUNTRIES TO MONITOR CP RATES

- New Cerebral palsy / childhood disability registers or population based studies
- Data collection and training tools are now available

DEVELOPING NEW ACTIONS / RESEARCH PROJECTS ....
• SCPE WEB SITE

http://www-rheop.ujf-grenoble.fr/scpe2/site_scpe

• Coordinator: Christine Cans, Grenoble