

Editorial

The Cerebral Palsies—Using a Common Language in Research Allows New Insights

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This issue of *Neuropediatrics* is dedicated to cerebral palsy (CP). It has long been recognized that the term does not describe a single disease but covers a group of diseases with different etiologies, therefore also referred to as the CPs. This group is characterized by the following common characteristics: (1) CP is permanent but not unchanging, (2) it involves a disorder of movement and/or posture and of motor function, (3) it is due to a nonprogressive interference/lesion/abnormality, and (4) this interference/lesion/abnormality is in the developing/immature brain.¹ CP is considered as the commonest cause of physical disability in early childhood and makes heavy demands on health, educational, and social services, as well as on families and children themselves. Many countries run registers to monitor CP rates as a measure for pre-, peri-, and neonatal cares. But data are often difficult to compare and often inconsistent. Therefore, CP prevalence is often referred to as unchanging despite the continuous effort to improve the peri- and neonatal condition of pre- and full-term born children.²

This issue assembles five papers that address several aspects of CP related to origin, prevalence, clinical consequences, and functional needs. Authors have in common that they are partners of a European network and most data presented here come from this network. SCPE (surveillance of CP in Europe), a network of population-based CP surveys and registers, extends across Europe and was set up in 1998 to monitor trends in CP rate, provides a framework for collaborative research.¹ The prerequisite for a common database was a “common language” on CP and its subtypes, based on clear neurological criteria (spastic, dyskinetic, and ataxic), and on functional severity. A common language has also been agreed on with respect to neuroimaging.

One of the seminal results of SCPE research has been the declining prevalence in CP in preterm born children.^{3,4} And here, for the first time, two continents with networks having sustained coverage of large populations over several decades

join their CP databases and can show a similar reduction in birth prevalence of CP in these children in Europe and Australia.⁵ In addition, this paper demonstrates the internal validity of pooling data across the two networks providing additional strength to the observation and opening the path for additional comparative studies in the future.

Research on CP origin has always been of particular interest.⁶ In this issue, the first analysis of magnetic resonance imaging (MRI) in the European CP database confirms on a large population basis, that CP is mainly due to lesions which are considered acquired during second or third trimester of pregnancy or around birth, and that maldevelopments are arising during embryonic and early foetal life are relatively uncommon.⁷ In preterm-born children, lesions typical for their gestational age at birth were by far predominant. This is especially interesting with respect to the epidemiological evidence of a decrease in CP morbidity. Future work within the European database will hopefully be able to document the decrease over time of these lesions in preterm born children. The data on MR patterns which cannot be clearly allocated to an acquired lesional pathology, for example, malformations, miscellaneous, and normal findings, which are more often found in term than in preterm born children, give an estimate of the importance of non-acquired, thus genetic origin of CP. Taken together, they accounted for around 30% of CP cases, which is somewhat higher to what is reported in studies on findings of pathogenic and likely pathogenic variants, when it concerns unselected individuals with CP.⁸

A third paper addresses the impact of associated impairments in CP and suggests an “impairment index” characterizing severity of impairments and their combinations.⁹ An important message is that on a population basis, CP is not always a severe condition: 30% of children with CP can walk unaided and have a normal or near-normal intellect without additional problems (concerning vision, hearing, or epilepsy)

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which is defined as low impairment index. Another important finding is the strong association between severity of gross motor deficit and occurrence of associated impairments in CP, with ataxic CP as an exception. This indicates that additional impairments do not only cooccur but are associated. The authors discuss this in the light of neuroimaging findings, which have shown a morphology–function relationship (the more severe the lesion the more severe the functional deficit, not only for motor but also for other functions) and argue that their findings contribute to the understanding of CP as a mainly lesional condition.

The report on inequality, also gender specific, in access to intrathecal baclofen treatment (ITB) in children with CP across Europe is intriguing.¹⁰ Population-based data from SCPE centers (Sweden, Norway, England, Portugal, Slovenia, and Denmark) show a great variation in treatment between centers (varying from 0.4–4.7%). Access to ITB correlated with a higher gross domestic product (GDP) and percentage of GDP spent on health and, probably most intriguingly, boys were more often treated than girls. Indications also differed, while in most centers, mainly children with severe bilateral spastic CP were treated, in Sweden dyskinetic CP was the most commonly treated subtype.

Tone reduction and physical therapy are equally important in the treatment of children with CP, especially spastic CP, by far the most frequent subtype of CP accounting for around 90% of cases. Here, Franki and colleagues¹¹ address these two aspects in combination and give an overview of different tone-reducing modalities and how they facilitate the different techniques applied in PT. As specific guidelines are not available concerning the appropriate combination of both interventions, the authors discuss the scientific evidence in the context of their clinical expertise. They deal with botulinum toxin, oral medication, intrathecal baclofen, and selective dorsal rhizotomy together with physical therapy, and discuss clinical reasoning strategies for appropriate treatment goals.

Taken together, these papers give new insights into the epidemiology and origin of CP, as well as the complex clinical features, and needs of patients suffering from CP. They also stress the importance of a common language as a prerequisite for research on this disease group. This seems more

important, among all, in the light of the documented successes in pre-, peri-, and neonatal cares on the one hand, and rising possibilities for genetic diagnostics on the other.

Conflict of Interest

None declared.

References

- 1 Surveillance of Cerebral Palsy in Europe. Surveillance of cerebral palsy in Europe: a collaboration of cerebral palsy surveys and registers. *Surveillance of Cerebral Palsy in Europe (SCPE)*. *Dev Med Child Neurol* 2000;42(12):816–824
- 2 Moreno-De-Luca A, Ledbetter DH, Martin CL. Genetic [corrected] insights into the causes and classification of [corrected] cerebral palsies. *Lancet Neurol* 2012;11(03):283–292
- 3 Platt MJ, Cans C, Johnson A, et al. Trends in cerebral palsy among infants of very low birthweight (<1500 g) or born prematurely (<32 weeks) in 16 European centres: a database study. *Lancet* 2007;369(9555):43–50
- 4 Sellier E, Platt MJ, Andersen GL, Krägeloh-Mann I, De La Cruz J, Cans C; Surveillance of Cerebral Palsy Network. Decreasing prevalence in cerebral palsy: a multi-site European population-based study, 1980 to 2003. *Dev Med Child Neurol* 2016;58(01):85–92
- 5 Sellier E, McIntyre S, Smithers-Sheedy H, Platt MJ on behalf of SCPE and the ACPR Group. European and Australian cerebral palsy surveillance networks working together for collaborative research. *Neuropediatrics* 2020;51(02):105–112
- 6 Korzeniewski SJ, Slaughter J, Lenski M, Haak P, Paneth N. The complex aetiology of cerebral palsy. *Nat Rev Neurol* 2018;14(09):528–543
- 7 Horber V, Sellier E, Horridgen K, et al. The origin of the cerebral palsies: contribution of population-based neuroimaging data. *Neuropediatrics* 2020;51(02):113–119
- 8 Corbett MA, van Eyk CL, Webber DL, et al. Pathogenic copy number variants that affect gene expression contribute to genomic burden in cerebral palsy. *NPJ Genom Med* 2018;3:33
- 9 Horber V, Fares A, Platt MJ, Arnaud C, Krägeloh-Mann I, Sellier E on behalf of the SCPE. Severity of cerebral palsy—the impact of associated impairments. *Neuropediatrics* 2020;51(02):120–128
- 10 Himmelmann K, Pählman M, Andersen GL, et al. Access to intrathecal baclofen treatment for children with cerebral palsy in European countries: an SCPE survey reveals important differences. *Neuropediatrics* 2020;51(02):129–134
- 11 Franki I, Bar-On L, Molenaers G, et al. Tone reduction and physical therapy: strengthening partners in treatment of children with spastic cerebral palsy. *Neuropediatrics* 2020;51(02):89–104