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Review article

Cerebral palsy and congenital malformations

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ABSTRACT

Aim: To determine the proportion of children with cerebral palsy (CP) who have cerebral and non-cerebral congenital malformations.
Methods: Data from 11 CP registries contributing to the European Cerebral Palsy Database (SCPE), for children born in the period 1976–1996. The malformations were classified as recognized syndromes, chromosomal anomalies, cerebral malformations or non-cerebral malformations. Prevalence of malformations was compared to published data on livebirths from a European database of congenital malformations (EUROCAT).
Results: Overall 547 out of 4584 children (11.9%) with CP were reported to have a congenital malformation. The majority (8.6% of all children) were diagnosed with a cerebral malformation. The most frequent types of cerebral malformations were microcephaly and hydrocephaly. Non-cerebral malformations were present in 97 CP children and in further 14 CP children with cerebral malformations. The most frequent groups of non-cerebral malformations were cardiac, facial clefts and limb and skeleton malformations. Children born at term had a significantly higher prevalence of cerebral malformations compared to children born before 32 weeks (12.1% versus 2.1%, $p<0.001$).
Conclusion: Cerebral malformations were much more frequent among children with CP than among all livebirths in the population. Malformations in organ systems close to the brain (eye, facial clefts) were more frequent in the CP population while malformations in organ systems further from the brain (renal, genital) were more frequent in the general population.

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1. Introduction

Cerebral palsy (CP) is a chronic disabling disease diagnosed in early childhood. CP is defined as "a disorder of movement and/or posture and of motor function, permanent but not unchanging, due to a non-progressive lesion or abnormality of the immature brain".[1,2]

Antenatal risk factors are thought to be important elements of the "causal pathway" to CP. Evidence implicating antenatal factors includes the high risk of CP among growth-retarded babies[3,4] and evidence implicating maternal infection,[5–7] as well as evidence that a higher than normal proportion of children with CP have congenital malformations.[8] Some of these congenital malformations are cerebral malformations originating in early pregnancy which MRI studies now directly implicate in the pathogenesis of CP.[9,10] With more MRI being used for diagnosis of children with CP,[11] the frequency of reported associated cerebral malformations is likely to increase. Non-cerebral malformations may be related etiologically through common risk factors acting both early and late in pregnancy. There is currently little evidence to allow evaluation of non-cerebral malformations as most published studies have been too small to specify malformation by type.[8] Large population-based studies are needed in order to evaluate the relation between CP and non-cerebral malformations. Finally, CP can also be an indirect result of the malformations, such as the malformation predisposing to a difficult birth with birth asphyxia or CP resulting from surgery to correct a malformation. The causal direction may not always be clear as the signs of CP become evident later in infancy.

The aim of this study is to determine the proportion of children with CP who have congenital malformations (cerebral and non-cerebral) based on data from a large European Cerebral Palsy Database (SCPE) and compare this with expected proportions in the population, based on data from a European network of population-based registries of congenital malformations (EUROCAT).

2. Methods

SCPE is a collaborative network of CP registries across Europe. The aim of the network was to develop a central database of children with CP in order to monitor trends in birthweight-specific rates, to provide information for service planning and to provide a framework for collaborative research.[1,12] Before establishing the common database, a common CP definition was agreed on. This definition is based on phenomenology not on etiology in order to account for different levels of diagnostic facilities and knowledge in different time periods and different countries.[13]

SCPE network elaborated decision trees in order to ensure harmonization in the criteria used to define and classify a CP case. All cases included into the SCPE database were first checked with these criteria.

Children were included in the registry at the age of at least 4 years, but children dying between 2 and 4 years old were included if they had clear signs of CP.

The SCPE common database contains data issuing from 16 different CP registries or population-based surveys, located in nine different countries. Overall, the SCPE network covers around 200,000 livebirths per year, and more than 10,000 CP cases are registered in the database at present. This report is based on children with CP born in 1976–1996.

The common data set include 45 variables and a standard minimum perinatal dataset was agreed. Children with isolated neural tube defects are not included in the SCPE database. Cases with acquired CP after the neonatal period were excluded from this study.

The database has four variables for coding syndrome, congenital anomaly, brain malformation and chromosomal anomaly. Each variable is coded 1 = yes, 2 = no or 0 = unknown. Further the ICD code and written text are given. Congenital brain malformation was defined as an antenatal developmental abnormality of the brain including developmental abnormality due to infectious agents and excluding postnatal developmental anomaly (acquired hydrocephaly and microcephaly). In the ICD coding system, there are codes for both congenital and acquired hydrocephalus. We have only included cases coded with congenital hydrocephalus. Non-cerebral malformation was reported if mentioned in the Smith book: Smith's Recognizable Patterns of Human Malformation (5th edition).

All cases coded 1 = yes for at least one of the four variables syndrome, congenital anomaly, congenital brain anomaly and chromosomal anomaly were extracted from the database. All cases were then manually reviewed. Cases with ICD codes or written text for congenital infections (without malformations), metabolic, neonatal events, other diseases and no or uncertain information were excluded.

Eleven SCPE registries which recorded information on congenital malformations were included in the study. One registry (East Denmark) had a period without ICD codes and written text and some of the years from this registry were excluded from the analysis.

After inclusion to the study, all cases were classified into four groups according to the following hierarchical criteria.