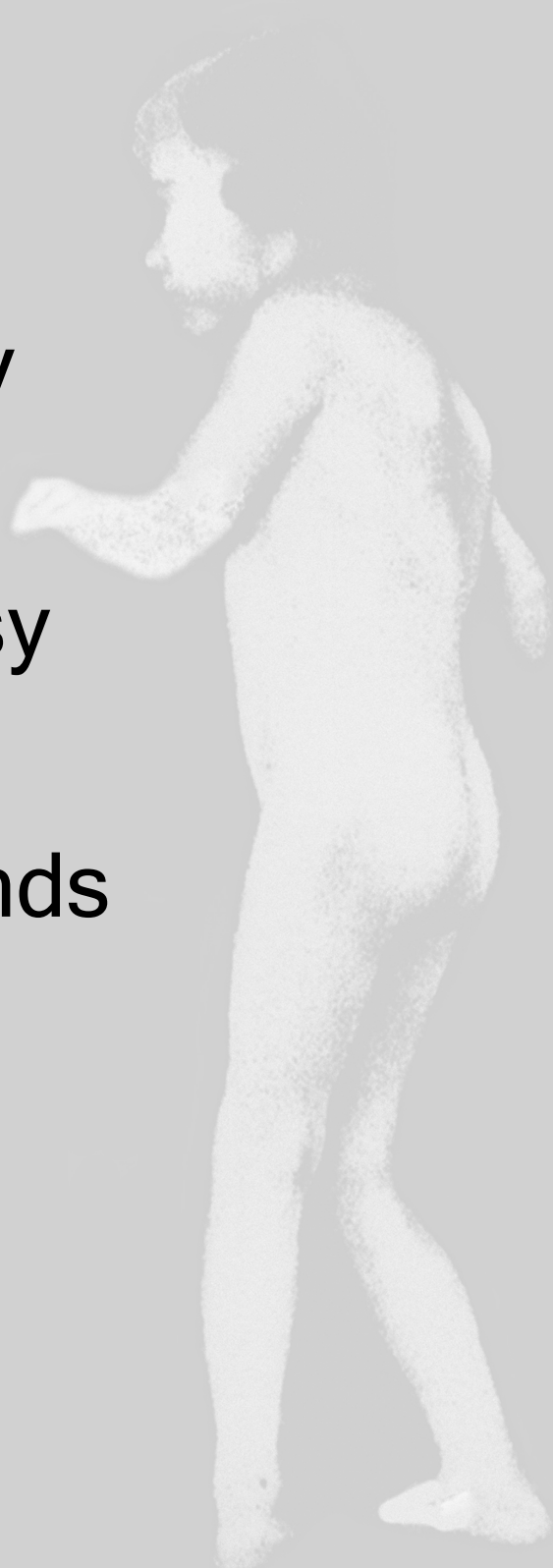
epidemiology of cerebral palsy in the netherlands



marc wichers

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Epidemiology of Cerebral Palsy in the Netherlands

Epidemiologie van Cerebrale Parese in Nederland

Proefschrift

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voor José, Marije, Otto en Leonie

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An die Musik

Du holde Kunst, in wieviel grauen Stunden,
Wo mich des Lebens wilder Kreis umstrickt,
Hast du mein Herz zu warmer Lieb entzunden,
Hast mich in eine beßre Welt entrückt!

Oft hat ein Seufzer, deiner Harf' entflossen,
Ein süßer, heiliger Akkord von dir
Den Himmel beßrer Zeiten mir erschlossen,
Du holde Kunst, ich danke dir dafür!

Tekst: Franz von Schober
Muziek: Franz Schubert (1817)

Contents

1

Introduction

Background of the thesis

Cerebral palsy (CP) is the most prevalent cause of primary motor disease in children in the world. Children with CP make strong demands on services, treatment and technical adaptations [1].

The definition of CP and its inclusion- and exclusion criteria have been the object of longtime discussion amongst experts in the field [2-4]. In these expert discussions attention has been drawn to obviously important features of encephalopathy other than the motor problems. These features such as learning problems, sensory problems, seizures may eventually be crucial to the level of functioning. However the common agreed essential element in the definition of CP stays the "palsy" part – being the disorder of movement / motor function, being the manifestation of a non-progressive brain lesion that occurred to the developing brain [5]. As such the presence of white matter lesions on the MRI does not mean a child has CP unless there is a disorder of movement or motor function, and if it can be assumed that the brain lesion is non-progressive and dates from an early phase in the child's development.

The definition cited in the separate papers in this thesis is based on the one described by Mutch et al. [3]. CP can present with a range of clinical manifestations caused by a range of causes, all being an injury to the immature brain [6, 7]. It is relevant to not only have information on the prevalence of CP per se but also on the severity of impairments, associated disabilities and limitation of activities and participation as they are present in the population. Different clinical or functional problems obviously ask for distinct therapeutic or rehabilitation approaches.

Alarming prevalence rate in the 1980s in other countries

Dutch representative figures and details on prevalence of CP and associated aspects were scarce until the mid-nineties. In that period population-based studies from other countries in Europe and the USA [8-10], indicated a strong rise in CP prevalence (looking back at the eighties as birth years). This prevalence rise was thought to be related to higher survival rates of dysmature and/or premature babies.

The rise in CP prevalence, reported elsewhere and the unknown proportion of children with severe CP, the children who need relatively more services, made it relevant to perform the Dutch population-based study.

The Dutch study, initiated by Utrecht child neurologist Onno van Nieuwenhuizen, adopted the study protocol from the co-operative population-based German-Swedish studies by Kraegeloh-Mann (Tubingen, Germany) and Hagberg, Goteborg, Sweden

[11-13]. Scoring of clinical items and associated features was done according to their criteria. Visits were made to Tubingen to get acquainted with the German survey protocol and Prof Hagberg visited us to see a group of children with CP, sharing clinical judgement.

As the Dutch study was carried out from a rehabilitation medical specialist point of view as well, we gathered additional details on functional aspects, treatment by medical specialist and allied health, in the broad sense, day-time situation, and psychosocial aspects.

International Classification of Functioning

Since the seventies and eighties of the 20th century the Dutch rehabilitation physicians (a medical specialism of its own) emphasized that there is more to the consequence of disease than labelling the original diagnosis, trauma or intervention [14]. In the twenty-first century, the framework of the International Classification of Functioning, Disability and Health ICF recognises the sequelae of disease, trauma or congenital disorder at the level of impairment of body function or structure, of activity (limitations) and of participation (restrictions) [15]. These three aspects were basically already there in ICF's predecessor, the International Classification of Impairments, Disabilities and Handicaps ICIDH [16]. However in the ICF these three are described in the context of two other aspects of a different order, being environmental factors and personal factors. Both of these two are crucial to the potential of the patient and his family of dealing with the three first mentioned.

ICF terminology nowadays provides a common language for experts to exchange data and views. In ICF terms, attention in the CP survey needed to be paid to "body functions" as well as "activity limitations" and "participation restrictions". Although at the time of the study design ICF was not there the crucial ICF elements were included.

Aim of the study

The study intends to give a broad picture of the general situation of Dutch children with CP. How prevalent are children with CP, what is the distribution and inter-relationship of clinical features and associated disorders, what are impairments of body function and structure, what about activity limitations. Are trends present in studied items? Where and by whom are children with CP treated, what interventions do they undergo and where do they go to school? Are behaviour and communication problems recognizably related to manifestations of CP?

Method of the survey

A cross-sectional population-based study was performed between 1995 and 1997 within the province of Gelderland. The study area has 1.2 million inhabitants (about 8% of the Dutch population of 15.4 million in 1995), comprises rural and urban areas and is considered to be representative for the Dutch situation regarding health care and demography [17]. Written approval was obtained from ethical committees of participating hospitals and institutions.

Children were 'supposed' CP cases for this study when they fulfilled the following criteria: (1) a diagnosis of CP recorded at any time (in the files of the particular source), (2) date of birth between January 1, 1977 and December 31, 1988, and (3) parents living – at the time of the study – in the western, central or eastern part of Gelderland.

CP was defined as: a disorder of movement and posture due to a non-progressive brain lesion with a proven or assumed onset no later than 1 year after birth. Positive neurological signs such as spasticity, dyskinesia or ataxia were obligatory. This definition is in line with frequently quoted definitions [3, 4]. Excluded from the definition of CP were minor sensory and motor problems (e.g. clumsiness due to minimal neurological dysfunction or developmental coordination disorder), as well as clumsy movements without genuine neurological pathology as seen related to mental retardation.

Genetic syndromes due to chromosomal anomalies – when recognised – were included only if an impairment of movement and posture, unrelated to the chromosomal anomaly itself, was present.

Motor disorders were classified as follows. Spastic syndromes were either unilateral (hemiplegia) or bilateral (diplegia, three-limb dominated BSCP and tetraplegia). Non-spastic syndromes were classified as either dyskinesia (involuntary movements, paroxysmal muscle tone change) or ataxia (impaired coordination). Patients that presented both spastic and dyskinetic or ataxic features were classified according to the predominant feature. This classification is adapted from Krägeloh-Mann et al. [12].

In order to make case ascertainment from the population as complete as possible, four consecutive ways were used that are described in detail in Chapter 2.

After parents of the children with supposed CP gave their informed consent, a visit was made by one of two examiners (both consultants in rehabilitation medicine). A questionnaire on maternal, obstetric, peri- and post-natal data, development and current functioning of the child was sent in advance. At the examiner's visit (usually at the child's home) answers to the questionnaire were obtained and the child was

examined. This expert examination was decisive as to whether or not the child was classified as having definite CP. These visits were made from spring 1995 through summer 1997.

Outline of this thesis

After this Introduction Section – Chapter 1 – five chapters are in fact separate papers with a distinct focus.

The first and very essential matter is the calculation of CP prevalence and trends in time through birth years 1977–1988. Dutch figures on CP prevalence were scarce. De Vries reported on several regional studies in the fifties and sixties without background information[18]. In 1982 the Phelps Foundation for the Spastics [19] published results of a telephone survey via special schools to estimate the CP prevalence. So that information was based on small-scale inquiries.

Our findings on (trends in) prevalence of CP in the Netherlands are presented in Chapter 2. This chapter also elaborates on ascertainment and the check to warrant representative data.

Chapter 3 gives insight in the distribution of major clinical presentations of children and important co-morbidity such as epilepsy, mental retardation and visual impairments [20]. The questions addressed were the following: a) what are the clinical presentations, major associated disorders and aetiological moments within the CP syndrome? b) do these phenomena change in time? and c) are the findings in the Netherlands comparable to those found in studies from other countries?

Chapter 4 looks more closely into impairments of body function and activity limitations. Activity limitations (ICF terminology) refer to difficulties in executing functional activities [15]. This paper focuses on limitations in mobility, addressing walking, lifting and arm/hand use, and in self-care activities. The objectives of this paper are to provide prevalence data on specific impairments and limitations of functional activities in the Dutch population of children with spastic CP and to get insight into the relationship between impairments and activities.

Chapter 5 reports on major aspects of cognition, behaviour and communication in the group of children with CP as encountered in the Dutch cohort. More specifically, general cognitive functioning (i.e., orientation, memory, learning capacity, contactual proficiency), personality descriptors (i.e., self-esteem, control, agreeableness, mood) and problem behaviour are explored. Furthermore, associations between these outcome measures and CP characteristics (gross motor functioning, limb distribution, intellectual level) are analysed.

Chapter 6 reports on types of treatment – in the broad sense – and types of school or daytime situation that are relevant to Dutch children with CP. In the Netherlands co-operation of multi-disciplinary (rehabilitation) teams with special schools is well-established. Several aspects of this cooperation have been the object in Dutch rehabilitation-based studies, both from the point of view of teamwork, the team communication process itself, and from the parent's or family point of view [7-9].

In addition to a comprehensive description of the current utilization of health care and educational services, we assessed the children's history regarding these items in special or mainstream facilities. Furthermore, to get insight whether the provided care and educational services were typical for subgroups of children with CP the associations with CP-related characteristics are explored as well as the parents' general opinion on services delivered for their child.

Chapter 7 is the general discussion which concludes this thesis. The framework of CP epidemiology in general and the Dutch survey in particular will be reconsidered; results as presented in the separate papers are summarized and if needed are highlighted again. On several aspects elapsed time and possible developments since the time of the field work may make our data seem "out-dated". They are in fact from the 20th century. New aspects are added in this chapter to the remarks that have already been made in the Discussion sections in the separate papers.

Furthermore in the General Discussion attention is given to CP – epidemiology in the Netherlands as such.

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2

Prevalence of cerebral palsy in the Netherlands (1977–1988)

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H.J. Stam, O. van Nieuwenhuizen

European Journal of Epidemiology 2001; 17: 527–532

Abstract

Children with cerebral palsy (CP) and their families often make strong demands on diagnostic, therapeutic, technical and social facilities. Prevalence estimates are needed to improve treatment and services. As recent Dutch data are not available, the present study aimed to assess the population prevalence of CP in the Netherlands. A representative Dutch area with 1.2 million inhabitants of which 172,000 were born between 1977 and 1988 was studied. To ascertain the children with CP from these birth years, medical practices (such as rehabilitation centres, paediatric and child neurological departments) were consecutively asked to contact their (supposed) CP cases. Next, a parents' organisation and finally regional news media assisted in the ascertainment. In total, 170 'supposed' CP cases underwent an expert examination. Of these 170, 127 children proved to be definite CP-cases, yielding a 'crude' average prevalence of 0.74 per 1000 inhabitants (95% CI: 0.61–0.87). Under-ascertainment was recognised and quantified. Accordingly, the population prevalence of CP over the birth year period 1977–1988 was calculated as 1.51 per 1000 inhabitants (average over the 12 birth years). The calculated CP prevalence rose significantly over time: from 0.77 (1977–1979) to 2.44 (1986–1988). This trend is in accordance with other studies.

Introduction

Cerebral palsy (CP) is an umbrella term comprising multiple aetiologies and clinical manifestations. It is not a nosological entity, but it is 'a useful framework for certain motor-disabled children with special needs' [1]. The usual definition of CP comprises a group of motor disorders caused by a non-progressive lesion of the immature brain [2]. Many aetiologies of the nonprogressive lesions are considered to be prenatal (from conception up to the delivery), others are considered to be perinatal (from the start of delivery to the first week of life) or neonatal (from the first week of life up to 27 days after birth). There are few non-progressive brain lesions (e.g. head trauma or infection) after the first month of life and because they are all well monitored the aetiology is rarely unknown [3].

Many children with CP have limited capabilities in mobility and activities of daily living, mostly due to spasticity. Associated mental and/or communicative impairments are frequently present. Children with CP and their families often make strong demands on diagnostic, therapeutic, technical and social facilities. Therefore, epidemiological studies including prevalence figures are needed both for further research on prevention and treatment, as for adequate planning of services for CP children and their families.

Trends in CP prevalence over 40 years have been reported for western Sweden where the prevalence rose from around 1.3 per 1000 inhabitants in the 1970s to 2 per 1000 in the 1980s [4–6]. In southwest Germany a population survey on bilateral spastic cerebral palsy (BSCP; constituting the major part of the CP population) was performed in the early 1990s yielding a prevalence of BSCP between 1.1 and 1.5 per 1000 inhabitants [7].

Other prevalence studies, with various designs, have been conducted in Europe and elsewhere (Table 2.1). Some of these studies are population-based [5, 7, 8–9] whereas others are based on permanent registers on CP or on chronic childhood disease [10–12]. Several studies rely on case reporting by third parties, such as treating physicians or public health workers. Also, different criteria for inclusion and exclusion are used, which complicates comparison of prevalence data. However, the general impression for the 1980s is that there is a rising tendency in the prevalence of CP.

Recent Dutch data are not available and it is not appropriate to extrapolate CP prevalence figures from other countries to the Netherlands.

Cross-sectional studies have been conducted in the Netherlands in the 1960s and 1980 [13, 14]. At both time points the reported CP prevalence was 1.5 per 1000, despite differences in study design.

The aim of the study presented in this paper was to assess the population prevalence of CP in the Netherlands.

Table 2.1 Data on CP prevalence from other studies

Country and reference	Births years studied	Prevalence N per 1000	Trend
Germany [7]*	1984–1986	1.1	Decrease (low birth weight)
Western Sweden [5]	1983–1986	2.5	Increase (preterm)
France [19]	72 / 76 / 81	1.8	Stable
England [9]	1967–1984	1.7	Increase (low birth weight)
Northern Ireland [20]	1981–ongoing	2.2	Increase
Denmark [10]	1979–1986	2.8	Increase
Norway [8]	1970–1989	70s 2.8 / 80s 2.0	Decrease
USA [11]	1981–ongoing	1.79 (80s)	Increase
Western Australia [12]	1956–ongoing	2.15 (75–92)	Increase

* This study focused on BSCP only (it excluded hemiparetic and non-spastic forms of CP).

Patients and methods

To estimate the prevalence of CP in the Netherlands, a cross-sectional population-based study was performed between 1995 and 1997 within the province of Gelderland. The study area has 1.2 million inhabitants (about 8% of the Dutch population of 15.4 million in 1995), comprises rural and urban areas and is considered to be representative for the Dutch situation regarding health care and demography [15]. Written approval was obtained from ethical committees of participating hospitals and institutions.

Children were 'supposed' CP cases for this study when they fulfilled the following criteria:

1. a diagnosis of 'cerebral palsy' recorded at any time (in the files of the particular source),
2. date of birth between 1 January 1977 and 31 December 1988, and
3. parents living – at the time of the study – in the western, central or eastern part of Gelderland.

In the present study CP was defined as: a disorder of movement and posture due to a non-progressive brain lesion with a proven or assumed onset no later than 1 year after birth. Positive neurological signs such as spasticity, dyskinesia or ataxia were obligatory. This definition is in line with frequently quoted definitions [2, 16, 17]. Excluded from the definition of CP were minor sensory and motor problems (e.g. clumsiness due to minimal neurological dysfunction), as well as clumsy movements without genuine neurological pathology as seen related to mental retardation. Genetic syndromes due to chromosomal anomalies – when recognised – were included only if an impairment of movement and posture, unrelated to the chromosomal anomaly itself, was present.

Motor disorders were classified as follows. Spastic syndromes were either one-sided (hemiplegia) or bilateral (diplegia, three-limb dominated BSCP and tetraplegia). Non-spastic syndromes were classified as either dyskinesia (involuntary movements, paroxysmal muscle tone change) or ataxia (impaired coordination). Patients that presented both spastic and dyskinetic or ataxic features were classified according to the predominant feature, usually the spastic syndrome. This classification is adapted from Krägeloh-Mann et al. [1].

In order to make case ascertainment as complete as possible, four consecutive ways were used.

In the Dutch medical practice consultants in rehabilitation medicine play very much a co-ordinating role in the integral medical care of CP children. So, as first ascertainment method, these consultants of the rehabilitation centres in the study area (referred to as centres 1–4 in Table 2.2) were contacted. Two centres with a small adherence in the area were mentioned together as one centre number 4. They were asked to inform the parents of children supposedly fitting the inclusion criteria about the study by letter, asking them to participate and refer directly to the principal investigator (MJW).

As a substantial number of parents of 'supposed' CP cases – some 50% – either refused to participate or did not respond a major under-ascertainment by this first method was recognised (Table 2.2). Hence, a second method was chosen to contact 'supposed CP cases'. Physicians of the departments of Paediatrics and Child Neurology of the University Hospital Nijmegen (the only University Hospital in the study area), and physicians working in institutions for the mentally handicapped were contacted similarly as described above and sent similar letters to their 'supposed' CP cases according to their registration.

Table 2.2 Ascertainment of CP cases via four rehabilitation centres

Centre number	1	2	3	4	Total
Supposed CP – contacted[a]	130	143	10	4	287
Refusal or non-response[a]	67	78	5	1	151
Examined[a]	63	65	5	3	136
Definite non-CP[a]	9	20	0	1	30
Definite CP[a]	54	45	5	2	106
Individual definite CP-cases ascertained primarily through this centre[b]	52	40	5	1	98

[a] Including multiple ascertainments (shared with consecutive sources other than the rehabilitation centres).
[b] Multiple ascertainments excluded.

Third, in order not to be dependent only on ascertainment sources in the medical field; the 'BOSK', the Dutch parents' organisation for children with primary motor deficits sent similar letters to all parents of children known to them as having 'spasticity'.

Fourth, as some CP cases might still have been missed by the previous efforts, finally the regional news media were informed (radio stations, newspapers) which resulted in articles and interviews with a 'call for patients' in most regional newspapers and regional radio stations. All general practitioners in the study area were informed about the study and the media covering in that stage.

After parents of the children with supposed CP gave their informed consent, a visit was made by one of two examiners (both consultants in rehabilitation medicine). A questionnaire on maternal, obstetric, peri- and post-natal data, development and current functioning of the child was sent in advance. At the examiner's visit (usually at the child's home) answers to the questionnaire were obtained and the child was examined. This expert examination was decisive as to whether or not the child was classified as a definite CP case. These visits were made from spring 1995 through summer 1997.

Data analysis included the estimation of CP prevalence with 95% confidence intervals (95% CI) per birth year as well as over the study period (1977–1988). Subsequently we evaluated whether there was a significant time trend of CP prevalence over the 12 birth years using linear regression analysis.

Results

Table 2.2 shows the numbers of letters sent by the rehabilitation centres (centre 1–4), being the first ascertainment method. In the rows of Table 2.2 per centre numbers are given on non-response or refusal, the number of children actually examined, the result of that examination and the number of definite CP-cases that were ascertained primarily through that centre.

Ninety-eight definite CP-cases were ascertained primarily by this first method. The second ascertainment method (the University Hospital departments) yielded 55 definite CP children. Among these, 36 individual children had already been identified and visited via the rehabilitation centres, so actually 19 extra definite CP-cases were added via this method.

The third ascertainment method through the parent's organisation yielded 18 definite CP-cases, but only two of them were new definite CP-cases not ascertained before.

Finally, the fourth ascertainment via de news media yielded nine definite CP-cases of which one was already previously ascertained.

After examination of 170 individual supposed CP cases, ascertained through the efforts described, 127 of these children proved to be definite CP-cases.

At 1 January 1995, 172,000 children that were born between 01-01-1977 and 31-12-1988 were living in the study area (Table 2.3). Hence the crude' CP prevalence based on children examined was the following fraction: 127 per 172,000 or 0.74 per 1000 children (95% CI: 0.61–0.87) as an average in the 12-year birth period.

In spite of the different consecutive ascertainment methods, we found that a large number of parents of supposed CP cases did not participate in the study. In the rehabilitation centre 1 it was established that 67 parents had either refused or not responded at all. The principal investigator (MJW) was also treating physician at that centre and had as such access to the medical files of the non-responders. After a thorough check of these files containing comprehensive information 59 from these non-respondent 67 children proved to be definite CP-cases.

Hence, the above-calculated prevalence was an under-estimation and the numerator of the 'crude prevalence' fraction had to be supplemented with at least 59 definite CP-cases.

As centre 2 was a rehabilitation centre was comparable to centre 1, the 59 to 67 or 88% ratio described above was applied to the 78 non-responders from that centre, yielding 69 children that most likely are definite CP-cases. For the centres 3 and 4 together, similar centres, but with a small patient adherence in the area, adjustment on an 88% basis of a total of six non-responders yielded five more 'definite CP-cases'.

Taking the estimated 'definite CP-cases' among the non-responders from the rehabilitation centres into account, the adjusted prevalence fraction (for an over-all value for the 12 birth years studied) is (127+59+69+5) = 260 over 172,000 children or 1.51/1000 (95% CI: 0.00133–0.00170, see Table 2.3).

Table 2.3 Prevalence per 3-year period and as a total during 1977–1988

From birth year period	1977–1979	1980–1982	1983–1985	1986–1988	Total
Number of definitive CP-cases found	17	21	37	52	127
Number of children living in the area on 1/1/1995	44,364	42,886	40,981	43,645	172,376
'Crude' population prevalence in N per 1000	0.38	0.49	0.90	1.19	0.74
Estimated population prevalence in N per 1000	0.77	1.00	1.84	2.44	1.51

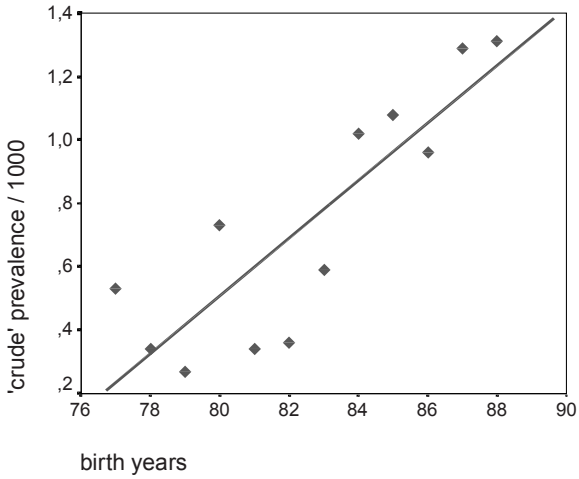

Figure 2.1 Crude CP prevalence per birth year.

Figure 2.1 shows the crude prevalence (i.e. not adjusted for under-ascertainment) per birth year over the studied period. There is a significant increase (p value < 0.0001) in CP prevalence over the years. As the number of CP cases seen per birth year was rather small, especially in the first birth years studied, we calculated the average prevalence per 3-year period (Table 2.3). The overall population in the area changed little during these years. In the bottom row again the adjusted prevalence figures according to the same methods as described above for the overall prevalence) are presented. These figures show that the population prevalence of CP rose from 0.77 in birth years 1977–1979 to 2.44 per 1000 children from the birth years 1986–1988.

Discussion

In a cross-sectional study in a representative area of the Netherlands, we found a CP prevalence of over 2 per 1000 inhabitants by the mid-80s. The CP prevalence has been rising since the mid-70s. Before interpreting the results, some issues need to be addressed.

Due to the anonymity of 'supposed' CP cases to the investigators, no reminder mailing could be sent to non-responders. However, the positive answers – or refusals with name mentioned – from the mailings through the university sources or the parents' organisation showed many multiple ascertainments with the mailings from the rehabilitation centres. So these mailings did reach (some of the) non-responders of the first ascertainment method.

As an extra check an effort was made to assess whether the non-responding definite CP-cases from rehabilitation centre 1 were a representative group. Several characteristics of these non-responders (birth weight, gestation age, birth year distribution, bilateral spastic or other type of CP) were compared to the examined definite CP-cases from all sources. No significant differences were found (Table 2.4). This justified adding these cases – being a representative CP-group, to the calculation of prevalent CP cases in the area and doing so for non-responders from other rehabilitation centres.

Since 35 definite CP-cases were ascertained by two methods and 13 by three methods, no further proportion of the non-responders from the second and third method were added to the numerator of the prevalence fraction, as the same non-responder might thus be counted twice. Besides there is no clue to estimate the proportion of definite CP-cases in these groups.

In the present study the birth cohort 1977–1988 was chosen to make sure that the children examined would be 6 years or older at the start of the study visits in spring 1995. Under the age of 5 years the clinical picture of a CP case may change; some children labelled early in life as 'CP' for a clinical reason eventually have no disorder of movement and posture, but mental retardation, or clumsiness, or they may present 'a transient CP picture' [18].

Notwithstanding the adjustments made, the calculated population prevalence might still be an under-estimation. Some prevalent CP cases might not or not any longer be receiving medical attention, or might not have been reached by or responded to the letter from the parents' organisation or to media coverage. There is no way however to quantify these CP children. There is no official Dutch register with compulsory reporting on CP cases.

Table 2.4 Comparison between all responders from all sources with CP (N = 127) and non-responders with CP from centre 1 (N = 59)

Patient characteristics	Resp. CP (N = 127)	Non-resp. from centre 1 (N = 59)	*p*-value
Birthweight (g)	2493 (941)	2408 (941)	0.6[a]
Gestation age (weeks)	36 (9)	35 (5)	0.5[a]
Birth year average 19xx (all children born 1977–1988)	83.9 (3.4)	83.3 (3.2)	0.2[a]
Clinical picture: ratio (N) BSCP / (N) all cases	71/127 = 56%	37/59 = 62%	0.4[b]

Values are means (standard deviation between paranthesis). BSCP, bilateral spastic cerebral palsy.
N denotes number of children.
[a] Student T-test was used.
[b] χ^2 test was used.

A striking finding is that 43 of 170 children at any time classified as 'CP', after expert examination proved to be non-CP cases. If a prevalence count had been performed on the basis of cases reported by third parties and the original classification 'CP' from the file was taken for correct this would greatly influence the calculated prevalence. This underlines both the value of expert examination and having this done at a sufficient age – in assessment of CP cases as an essential feature of this Dutch study.

As the province of Gelderland is considered 'large and average' [15] it is reasonable to apply the CP prevalence data acquired from this study to the entire country. In that case the Dutch population (15,400,000 in 1995) is estimated to have some 3300 cases of CP (13 × 260) born in the birth year period 1977–1988, with numbers rising towards more birth recent years.

'Loss of cases' due to prior death of children with CP was not established in this study as population prevalence obviously deals with individuals currently present requiring treatment and services. CP itself is not a recorded cause of death in the Netherlands (some complications may be, such as respiratory infections). If the onset of the brain damage would be taken no later than 1 month after birth, the prevalence count would change little: it would exclude six CP cases who had their brain damage in the first year of life; but after the first month.

Figure 2.1 shows the rise of 'crude' prevalence of CP in the Netherlands. This rise could be 'genuine' or an artefact. Selection bias as the cause of a possible artefact must be considered. It is possible that the children from earlier birth years had a higher proportion of non-responders/refusals, because the children are adolescents and tired of examinations. This may not be fully resolvable. However, in centre 1, the practice of the first author, non-response in a large number of children could be studied in detail and it was not higher in older children. Even if there is some relative under-ascertainment of older CP cases, this is insufficient to counteract the significant rise in prevalence (calculated from our examined cases). Thus, the rise in prevalence in our study can be considered genuine. Comparing the Dutch results with other studies, (with the western-Swedish project as a prototype because in this project the entire CP group, not only bilateral spastic CP, was studied by expert examination) the common finding is a rising trend in CP prevalence in the 1980s (Table 2.1).

Conclusion

Estimated prevalence of CP in the Netherlands has reached well over two per 1000 by the birth years of the mid-1980s. Moreover, CP prevalence has been rising since the mid-1970s. This trend is in accordance with findings from several studies performed elsewhere.

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3

Clinical presentation, associated disorders and aetiological moments in Cerebral Palsy: A Dutch population-based study

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Abstract

Purpose: Cerebral Palsy (CP) contains varying clinical presentations, associated disorders and aetiological moments. Quantitative data and trends on these aspects were lacking in The Netherlands.

Method: Within a population-based study on prevalence, presentation and functioning of Dutch children with CP born inthe years 1977–1988, individual history taking, examination and medical file checking was done by experienced clinicians. Clinical subtypes, motor disability, important co-morbidity (mental retardation, visual disability and epilepsy) were recorded, aetiological moments identified if possible. By comparing the four most recent years with the earlier years possible trends were studied.

Results: A quarter of children beforehand recorded as CP did not meet inclusion criteria after individual examination. Spastic subtypes accounted for over 90% of all CP cases: bilateral spastic cerebral palsy as a group are the majority although spastic hemiplegia is percentage-wise the largest individual clinical subtype. Epilepsy and mental retardation are common. Clinical patterns and associated disorders remained rather constant comparing earlier to more recent birth years.

Conclusions: An early diagnosis of CP may be challenged. General clinical patterns remained rather constant in following years, as did most studied items. Even if this study revealed a prevalence rise, no aspect stood out as a possible explanation for this prevalence rise. Comparable studies performed elsewhere showed similar findings.

Introduction

In epidemiological studies on cerebral palsy (CP) it is important to report on clinical presentation, associated disorders, possible risk factors and aetiological moments in order to determine treatment policy and provide and anticipate on services needed [1–4].

In our Dutch population-based study on children born in the years 1977–1988, the prevalence of CP was found to be rising from 0.77 per 1000 inhabitants in 1977–1979 to 2.44 per 1000 in 1986–1988 [5].

In the present paper the general content of the CP syndrome in The Netherlands is described. The questions to be addressed were the following: (a) What are the clinical presentations, major associated disorders and aetiological moments within the cerebral palsy syndrome? (b) Do these phenomena change in time? (c) Are the findings in The Netherlands comparable to those found in studies from other countries?

Method

To accomplish comparability of the data the study design followed previous European studies, sharing definitions and classifications of associated disorders [3, 4, 6].

In the study CP was defined as a disorder of movement and posture due to a non-progressive brain lesion with an onset no later than 1 year after birth. Positive neurological signs such as spasticity, dyskinesia or ataxia were obligatory.

Approval from ethical committees of the University Hospital serving the region and of other institutions involved was obtained to perform the study. To comply with privacy laws involved, ascertainment of the children was achieved through medical practices such as rehabilitation centres, and the departments of Paediatrics and Child Neurology of the University Hospital serving the region. Staff at these practices asked the parents or caretakers of (supposed) CP cases within the area and age group to participate in the survey. A national parents' organisation and finally the regional media assisted in the ascertainment. Details on ascertainment and the resulting prevalence figures have been reported elsewhere [5].

One hundred and seventy children, who reportedly had CP and were born between 1977 and 1988, identified by the sources mentioned in a representative area with 1.2 million inhabitants and about 14,000 live births/year were volunteered by their parents to participate. From 1995 to 1997 these 170 children were visited, usually in their home. During the visit the parents or caretakers were interviewed about the medical and perinatal history, and the child's current functional level. The

interview questions had been sent in advance. After the interview a neurological and orthopaedic physical examination was performed by an experienced clinician. The parents gave permission to supplement or check their answers with data from medical files (including the results of neuro-imaging).

Clinical presentations, associated disorders, risk factors and aetiological moments were classified as follows:

> The clinical presentation was described according to the common major types of CP: (1) bilateral spastic CP (BSCP), (2) unilateral spastic (hemiplegia), and (3) non-spastic types [3, 7]. The BSCP group was subdivided in three groups: two-limb dominated or diplegia (legs more affected than arm), three-limb dominated, or 'triplegia' (one arm not or minimally affected), and four-limb dominated or 'tetraplegia' (both arms and legs equally affected). The non-spastic CP subtypes were ataxia (primary disorder of coordination), and dyskinesia (involuntary movements and paroxysmal muscle tone change either with a choreo-athetotic or a dystonic presentation).
>
> Motor disability was classified as mild or severe, motor disability being defined as mild if the child by the age of 5 years had reached a form of independent walking (with or without walking aids). Severe motor disability was present if the child by the age of five had not done so.

Common disorders associated with CP include epilepsy, mental, and visual impairments. Epilepsy was classified in three subgroups as: (1) never: the child had no signs of epilepsy at any time, (2) ever, or non-active: epilepsy was diagnosed and treated in the past, and (3) active: epilepsy is presently being treated, although the child may be free of fits under medication. Mental capacity was classified in three groups: (1) normal: child attends regular school, no known learning difficulties, IQ estimated at 85 or higher, (2) moderate learning difficulties (specific schooling needs) estimated IQ 70–85, and (3) mental retardation (very specific schools or day centre, either elementary learning, some practical training, or no learning whatsoever) estimated IQ below 70. Visual impairment was also divided into three categories: (1) none: or a correctable refraction anomaly, (2) mild: reduced visual acuity, squint or nystagmus, and (3) severe: minimal visual perception or blind.

Birth weight was classified into three categories: (1) Very Low Birth weight (VLBW): less then 1500 g, (2) Low Birth weight (LBW): 1500–2499 g, and (3) Normal Birth weight (NBW): 2500 g and higher.

The aetiological moment (i.e., not the aetiology in strict sense but the established time of the cerebral insult leading to CP) was decided on by the third author, a child

neurologist (OvN). This was done only on the basis of conclusive evidence from both history and medical files.

Classification of the aetiological moment was done following the guidelines summarised by Krägeloh-Mann et al. using four categories [4]. It required conclusive evidence on the following:

> A *prenatal* aetiological moment in hereditary forms; definite prenatal syndromes – chromosomal abnormalities; verified congenital infections (toxoplasmosis, rubella, cytomegalovirus and herpes); cerebral malformations; and for term babies: signs of periventricular leucomalacia on CT scan in cases not meeting the criteria of a peri-neonatal aetiology.
> *Peri-neonatal* aetiological moment *in term infants*: confirmed cerebral aemorrhage of any grade; confirmed brain oedema or strong evidence for neonatal shock, asystoly, need for resuscitation, organ failure; confirmed sepsis or CNS infection, hypoxic-ischaemic encephalopathy with two of the following three signs: low apgar scores, resuscitation/ventilation, convulsions before day 3.
> *Peri-neonatal* aetiology *in preterm infants*: confirmed cerebral haemorrhage grade III or IV; confirmed brain oedema or strong evidence for neonatal shock (i.e., asystoly, need for resuscitation, organ failure; confirmed bacteriaemia or CNS infection; also in cases of low apgar scores, prolonged mechanical ventilation or pneumothorax.
> A *postneonatal* aetiology: the brain lesion producing the CP picture occurred recognisably after the first week after birth.

The aetiological moment was classified as *unknown* if the data from parents or medical files were inconclusive or lacked positive evidence of any kind.

Results

Of the 170 children that were visited and examined 43 were excluded, because they did not fulfil the criteria for CP. Of the remaining 127 children, 78 were boys (61.4%) and 39 were girls (38.6%) At the time of examination these children's ages varied between 6 and 19 years (mean age 11 years, median age 10 years).

The clinical presentation of CP among the 127 children is given in Table 3.1. This table shows that the majority of cases fall into the BSCP group (55.9%). Among all spastic sub-types, the hemiplegia group is the largest single group (37.8%), while

Table 3.1 Clinical presentation of CP and motor disability, related to subtype in Dutch children

Type	N	% of total	95% CI	% of spastic type	95% CI	% severe motor disability
Spastic	119	93.7	[89.5–97.9]			
BSCP	71	55.9	[47.3–64.5]	59.7	[50.9–68.5]	64.8
Diplegia	30	23.6	[16.2–31.0]	25.2	[17.4–33.0]	40.0
Triplegia	12	9.5	[4.4–14.5]	10.1	[4.8–15.5]	58.3
Tetraplegia	29	22.8	[15.5–30.1]	24.4	[16.6–32.1]	93.1
Hemiplegia	48	37.8	[29.4–46.2]	40.3	[31.5–49.2]	8.3
Non-spastic	8	6.3	[2.1–10.5]			75.0
Ataxia	4	3.2	[0.001–6.2]			50.0
Dyskinesia	4	3.2	[0.001–6.2]			100.0

there are a similar number of children in each of the diplegia and tetraplegia groups (about 23% each). The triplegia group is much smaller (9.5%). The non-spastic types are much less common; only 6.3% of the children can be classified as either ataxic or dyskinetic.

The distribution of the associated disorders related to the type of CP is given in Table 3.2. Epilepsy is a common disorder among these children. Of all children included in the study, 18.9% had active epilepsy at the time of examination, and a further 21.3% had a history of epilepsy. Of the children with tetraplegic CP 44.8% had never had epilepsy, compared with 66.7% of the children with spastic diplegia, triplegia, and hemiplegia, and 37.5% of the children with ataxia and dyskinesia. Among all CP children only the hemiplegic subtype showed a majority with normal mental capacity; severe learning difficulties were most common in tetraplegic children.

The children with spastic hemiplegia also had much less visual impairments, while again the children with spastic tetraplegia had the most severe visual impairments.

In Table 3.3 the distribution of birth weight and the four categories of identified aetiological moments are given according to the type of CP. Only 73 out of 127 children could be classified according to their aetiological moment.

Trends

To examine possible time trends, the frequency of clinical presentation, associated disorders, birth weight categories and aetiological moments were compared in the birth year groups 1977–1984 and 1985–1988. As the number of subjects in the second year

Table 3.2 Associated impairments within the CP subtypes in Dutch children

Subtype of CP	*BSCP dipl*	*BSCP tripl*	*BSCP tetrapl*	Spastic BSCP total	Spastic hemipl	Non-spastic atax-dysk	Total
Number	*30*	*12*	*29*	**71**	**48**	**8**	**127**
Epilepsy*							
Never	*66.7*	*66.7*	*44.8*	57.7	66.7	37.5	59.8
Ever	*10.0*	*16.7*	*31.0*	19.7	22.9	25.0	21.3
Active	*23.3*	*16.7*	*24.1*	22.5	10.4	37.5	18.9
Mental capacity*							
Normal	*40.0*	*25.0*	*6.9*	23.9	54.2	12.5	34.6
Learning difficulty		*26.7*	*25.0*		31.0	37.5	26.8
Mental retardation		*33.3*	*50.0*		62.1	50.0	38.6
Visual impairment*							
None	*66.7*	*50.0*	*44.8*	54.9	83.3	62.5	66.1
Moderate	*26.7*	*50.0*	*27.6*	31.0	10.4	0.0	21.3
Severe	*6.7*	*0.0*	*27.6*	14.1	6.3	37.5	12.6

* Percentages among this type.

Table 3.3 Birth weight categories and identified aetiological moments in percentages according to type of CP in Dutch children

		BSCP dipl	*BSCP tripl*	*BSCP tetrapl*	Spastic BSCP total	Spastic hemipl	Non-spastic atax-dysk	Total
Birth weight*		*N=30*	*N=12*	*N=29*	N=71	N=48	N=8	N=127
VLBW		*16.7*	*33.3*	*31.0*	25.4	10.4	0.0	18.1
LBW		*26.7*	*58.3*	*34.5*	35.2	27.5	12.5	30.7
NBW		*56.7*	*8.3*	*34.5*	39.4	62.5	87.5	51.2
		N=16	*N=10*	*N=18*	N=44	N=25	N=4	N=73
Prenatal		*6.3*	*30.0*	*22.2*	18.2	20.0	0.0	17.8
Perinatal	Preterm	*68.8*	*50.0*	*66.7*	63.4	32.0	25.0	50.7
	Term	*18.8*	*20.0*	*5.6*	13.6	36.0	50.0	23.3
Postnatal		*6.3*	*0.0*	*5.6*	4.5	12.0	25.0	8.2

* Percentages among this type.

group 1985–1988 was comparable to the complete first 8 years due to increasing CP prevalence no effort was made to distinguish three 4-year groups as the first would have been too small for relevant comparison. Table 3.4 shows that there were no significant changes over frequencies in time for type of CP, severity of motor disability, epilepsy, mental capacity, and aetiological moment. There was, however, a significantly larger proportion of children with VLBW in the second birth cohort, (and a significantly

lower proportion with a LBW). Severe visual impairments are significantly more common in the later birth cohort.

Within three birth year groups the proportion of 'unknown' aetiological moment decreased: for 1977–1980 it was 50% (14 of 28); for 1981–1984 46.9% (15 of 32) and for 1985–1988 37.3% (25 of 67). The overall percentage of 'unknown' aetiological moment is 42.5%.

Table 3.4 Trends in time – significant changes in studied items

		Birth years 1977–1984	Birth years 1985–1988	
		N = 60	N = 67	*p*-value if $p < 0.05$
Clinical presentation				
Spastic	Diplegia	23.3	23.9	
	Triplegia	8.3	10.4	
	Tetraplegia	23.3	22.4	
	Hemiplegia	41.7	34.3	
Non-spastic	Ataxia	1.7	4.5	
	Dyskinesia	1.7	4.5	
Motor disability	Mild	61.7	61.2	
	Severe	38.3	38.8	
Associated impairments				
Epilepsy	Never	61.7	58.2	
	Ever	20.0	22.4	
	Active	18.3	19.4	
Mental capacity	Normal	31.7	37.3	
	Learning difficulty	25.0	28.4	
	Mental retardation	43.3	34.3	
Visual impairment	None	75.0	58.2	$p = 0.046$*
	Mild	20.0	22.4	
	Severe	5.0	19.4	$p = 0.015$*
Birth weight and aetiological moment				
Birthweight	VLBW	10.0	25.4	$p = 0.025$*
	LBW	40.0	22.4	$p = 0.032$*
	NBW	50.0	52.2	
Aetiological moment		(N = 31)	(N = 42)	
(identified cases)	Prenatal	16.1	19.0	
	Perinatal preterm	51.6	50.0	
	Perinatal term	19.4	26.2	
	Postnatal	12.9	4.8	

* Chi-square test.

Discussion

In a population-based study of Dutch children with CP, aged 6–19 years at the time of examination, several features of CP were investigated. The definition of CP as used in this study was in line with other frequently quoted definitions [7–9]. Our approach of definition and clinical subtype complies with, and contributed to the 'decision tree' used within SCPE, a European BIOMED cooperative study on CP [10].

Children with CP in the study area that might have been ascertained (and examined) in this study but died previously were not taken into account, as the study assessed prevalence among other aspects. Prevalent cases are the children present with CP who need treatment and provisions. Besides, there is obvious difficulty in verifying the CP picture in deceased cases.

The vast majority (93.7%) of children in the sample had CP of the spastic type, of which 59.7% was of the bilateral spastic sub-type. However spastic hemiplegia was the largest single subtype. Severe motor disability was especially common in children with tetraplegia (93.1%) and those with non-spastic types of CP (75%), but more than half of the children with triplegia also had a severe motor disability. Of the common associated disorders epilepsy either was present or had been present in almost 40% of the children. Epilepsy was particularly common in tetraplegics (more than half had or had had epilepsy). Mental capacity was reduced in 65% of the cases.

Visual impairments were present in a third of all children, again with the children with tetraplegia and non-spastic CP having the most severe impairments. The comparatively good visual performance of children with hemiplegia can be explained by the intactness of the geniculo-striate pathway in the unaffected hemisphere, essential for visual acuity [11]. Significantly more CP children with triplegia had a low or very low birth weight, and significantly more children with hemiplegia had a normal birth weight. Among the other spastic types of CP there were no significant different numbers of normal and low birth weight children.

There were no significant changes or trends identified when comparing the birth year group 1985–1988 to the earlier birth years in clinical subtype, motor disability, epilepsy and mental capacity.

In approximately three-quarters of all children with an established aetiological moment this moment was perinatal; only 17.8% were identified as prenatal and 8.2% as postneonatal. No reliable aetiological moment could be established in 42.5% of cases, a proportion similar to that reported in other series [4, 12]. However a decrease over time in the proportion 'unknown' was seen from 50% in birth years 77–80 to 37% for 85–88. Availability of new neuro-imaging techniques (ultrasonography, CT-

or MRI-scanning) contributed to this decrease. Neuro-imaging was performed in the majority of cases – however if neuro-imaging showed no brain abnormality and no other positive indications were available the 'judgement' on aetiological moment remained 'unknown'. As an example several children were encountered with uneventful pregnancy and delivery, born at term with normal findings of neuro-imaging and presenting a hemiplegia as they develop. As no event perinatal or post-neonatal event could be traced here a case could be made for a prenatal aetiological moment. These cases however were classified as 'unknown'. In cases with an established aetiological moment no specific trends in the distribution of timing of insult could be assessed. So, although a higher percentage of VLBW children with CP (which are usually born pre-term) has been observed in recent years, the rate of perinatal–preterm events identified as aetiological moments does not alter in our study. The rising percentage of VLBW and the falling percentage of LBW children with CP may mirror neonatal trends for all children.

In a population-based study a crucial question is whether the sample can be considered representative of the total population of CP children. We addressed this issue in our previous paper by using both multiple ascertainment approaches, to reduce selection bias and by comparing the examined group of children to large group non-responders of CP children for whom comparable data were available [5]. Information bias can occur in studies when data comprises reports from third parties with different health care professionals reporting on individual CP children. In this study, however, two experienced clinicians examined all children personally, and the children were not younger than 6 years of age. This minimum age is important as in the early years of life the clinical picture of the CP syndrome may change considerably – or it may even vanish [13]. As the age at examination was 6 or higher (average age of 11, mean age of 10), the clinical features of the studied children have stabilised. This also improves the reliability of the classification of subtype of CP, the associated disorders and other variables.

The impact of this consistent approach is illustrated by the fact that after expert clinical examination no less than one-quarter (43) of 170 children, previously reported as having cerebral palsy, were excluded from the study. Of these 43 children 13 showed mental retardation without genuine neurological pathology, 12 had no motor problems whatsoever at the time of examination, and 17 had only minor motor dysfunction (clumsiness). One child, previously labelled as 'hypotonic tetraplegic CP' went on – after our examination – to be diagnosed as a HMSN type 1. We can conclude that the children classified as having CP are as free from selection and information bias as possible and therefore consider our estimates as representative for the Dutch CP population.

Data provided by parents on birth weight, gestational age and results of neuro-imaging showed to be reliable when cross-checked with the medical files. Very few amendments were necessary.

The differentiation between two-limb dominated and three-limb-dominated CP is not generally made [2, 12]. We chose to recognise the category 'three-limb dominated' or 'triplegia' as to clinicians the bilateral spastic CP child with 'one good hand' is familiar.

The comparison of our data with others studies confirmed that, the Dutch CP population is not different from the CP population in other European countries. We found a male–female ratio of 61–39%. Over-representation of boys is the normal finding in CP epidemiology studies [12].

The distribution of clinical subtypes reported here (Table 3.1) are in-keeping with studies summarised in Stanley et al. [12]. However, non-spastic CP presentations (ataxia and dyskinesia) in the Dutch data were lower as a percentage of the whole: 6%, compared with 12 or more reported elsewhere [12]. However, as there are only a few non-spastic cases in the study population, minor differences in study percentages are not conclusive.

We also compared our data with previously published Swedish and German data on the same aspects of CP [1]. After all our study design was made in cooperation with the authors of this cooperative study, sharing definitions and criteria. The distribution of the birth weight categories among BSCP children is similar in both The Netherlands and Germany, while Sweden reported slightly more NBW and less VLBW children in the BSCP as well as in the total group. All three countries report the same distribution of mild and severe motor disability in the total as well as the BSCP group. The Swedish and German researchers reported more children who never had epilepsy, while only the group who had ever had epilepsy is significantly larger in the Dutch study.

The percentage of children with a 'normal' mental capacity in BSCP is similar to that reported in both the German and Swedish studies. The threshold between 'moderate learning disability' and 'mental retardation' however may be hard to standardize. We used the cut-off point of an estimated IQ below 70 for mental retardation (as in the German – Swedish reference article).

Finally, compared with both Germany and Sweden, there were significant more children without any visual disability in our study. In the Dutch group of BSCP children the percentage of children with 'no' visual disability (55%) was high, compared to 37 or 38% for Germany and Sweden. We have no explication for this and believe that this is unlikely to be a threshold issue as the category 'no visual disability' should be easy to

recognize (more so than the threshold between mild and severe visual disability). Our judgement 'no visual disability' was decided on by the combination of answers from parents to the questionnaire and the findings at examination. As the average age of the children at examination was over 10 it is unlikely that the examination for our study would be the occasion to reveal a child's vision problems to the parents. One remark can be made: The data from the Swedish study relate to younger children (mean age 5.8 years at examination) and visual acuity is reported to improve with age [1, 11]. This may account for an overall better visual acuity in the Dutch group.

In cases with a recognised aetiological moment no specific trends in relation to prenatal or perinatal origins were noted. So if the higher percentage of VLBW children (usually born pre-term) in recent years is obvious in the CP group, the percentage of (perinatal–preterm aetiological moments) as part of the whole has not altered. A conclusion may be that the rising percentage of VLBW and the declining percentage of LBW children is a mirror of neonatal trends for all children.

The combination of knowledge of clinical presentations of CP, their association with motor disability, associated disorders and the prevalence in an population will give information on the services needed both for treatment and for technical and social provisions for children with CP in The Netherlands. Forthcoming papers will discuss the consequences for society in greater depth.

Data as provided at the population level for the birth years after 1988 are not presently available for any Dutch cohort. Within The Netherlands there is no agreed policy on reporting all or specific forms of chronic childhood disability such as CP. Privacy and the liberty of parents to co-operate or not in population-based studies is a central issue regarding comprehensive personal medical data. However valuable, these rights act as a persistent block in obtaining high ascertainment rates in population-based studies.

Conclusion

A population-based study (involving comprehensive history taking and individual expert examination) on the profile of the cerebral palsy syndrome in The Netherlands was performed. Clinical subtypes and patterns in time regarding motor disability, epilepsy, mental retardation and aetiological moments (if identified) of cerebral palsy showed a rather stable picture in the birth years 1977–1988. This stable picture occurred during a time when the prevalence of CP had risen significantly; so among the studied features none significantly stood out as a possibly explanation for the prevalence rise. Moreover the general findings from this study are in line with studies done elsewhere. Both the

stable picture and the consistency with studies from other countries suggest that these comprehensive data of the children give a representative picture of children with CP in The Netherlands.

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4

Motor impairments and activity limitations in children with spastic cerebral palsy: A Dutch population-based study

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Abstract

Objective: To determine the prevalence of motor impairments and activity limitations and their inter-relationships in Dutch children with spastic cerebral palsy.

Patients and methods: In a population-based survey 119 children, age range 6–19 years, with spastic cerebral palsy were examined. Anthropometry, muscle tone, abnormal posture, joint range of motion, major orthopaedic impairments and gross motor functioning and manual ability were assessed or classified, in addition to limitations in mobility and self-care activities. Spearman's correlation coefficients, bivariate post hoc analyses and univariate and multivariate logistic regression analyses were used.

Results: Children with spastic cerebral palsy had a lower body height and weight compared with typically developing peers. Forty percent had no range of motion deficits. Hip dislocations were rarely encountered. Motor impairments were associated with gross motor functioning and manual ability levels. Close to sixty-five percent walked independently. Children with diplegia and tetraplegia differed in activity limitations. Motor impairments and limitations in mobility and self-care activities were only modestly related in multivariate analyses.

Conclusion: Distribution of cerebral palsy-related characteristics is consistent with that found in representative studies of other countries. The distinction between diplegia and tetraplegia is relevant from an activity point of view. The child's activity limitations are not a mirror of the motor impairments, which suggests multifactorial influences. An activity-oriented rehabilitation approach goes beyond treating specific impairments.

Introduction

Children with cerebral palsy (CP) present a variety of clinical presentations and a range of motor impairments and activity limitations [1]. Insight into the distribution of these elements within the group of children with CP and into the relationship between specific impairments and activity limitations may be helpful in directing rehabilitation goals. Quantitative data on CP-related motor impairments and activity limitations can identify phenomena appropriate for longitudinal study, thus promoting adequate planning for both research and health services.

Several population-based CP studies in other European countries have explored prevalence and inter-relationships of clinical features, motor impairments, activities and described appropriate methods of classification [2–7]. The inter-relationship of motor impairments and activity limitations is not always straightforward. One issue raised is whether the distinction between leg-dominated and 4-limb dominated spastic CP is relevant in the light of activity limitations or whether describing a Gross Motor Functioning Classification System (GMFCS) level is sufficient to describe the child in this respect [8].

In the Netherlands prevalence data on motor impairments and activity limitations in children with CP has not been available until now. This paper presents representative Dutch data on these issues. We focus on spastic CP, as this form is by far the largest sub-group of CP [1, 9].

According to the World Health Organization's (WHO) International Classification of Functioning, Disability and Health (ICF), impairments are described as significant deviations or loss of body function or body structure [10]. In CP, dysfunction of muscle control prevails, which can lead to spasticity or shifting muscle tone, to associated pathological postures, and to decreased range of joint motions.

Activity limitations refer to difficulties in executing tasks or actions [10]. The current study focused on limitations in mobility, addressing walking, lifting and arm/hand use, and in self-care activities.

The objectives of this paper are to provide prevalence data on neuromusculoskeletal impairments, (i.e. "motor impairments") and activity limitations in the Dutch population of children with spastic CP and to gain insight into the relationship between impairment and activity limitation.

Methods

Subjects

The present study is part of a cross-sectional population-based survey. Previous publications from this study addressed prevalence and clini cal characteristics of CP in the Netherlands [9, 11]. CP was defined as a disorder of movement and/or posture caused by a non-progressive brain lesion with an onset no later than one year after birth [12]. Obligatory neuromotor disorders (spasticity, dyskinesia or ataxia) were present in all patients. Patients were included if they had: (1) a diagnosis of "cerebral palsy" recorded in their patient files; (2) date of birth between 1 January 1977 and 31 December 1988; and (3) parents living, at the time the study was conducted, in Gelderland, a region in the east-central part of the Netherlands. In the present study, we concentrated on children with spastic CP (over 90% of the total group). Children with ataxic or dyskinetic CP were excluded; these non-spastic sub-groups in the cohort each comprised only 4 children. Hence, we present the results of a representative group of 119 children with spastic CP. Informed consent was obtained from the parents of each participant. The study was approved by the ethics committees of the university medical centre and collaborating institutions.

Data collection

Data collection was carried out according to the study protocol of previous German and Swedish studies on the epidemiology of CP [13–16]. The protocol and classification criteria were discussed and trained (in advance) together with the authors of these studies.

An experienced child rehabilitation physician visited each child and his or her parents. A structured interview with the parents or caretakers and a physical examination of the child were performed. The interview covered the child's CP-related and general medical history, current and past treatment, adaptations, milestones in development, present performance of activities of daily living, communication, behaviour, school career, and family situation. In the examination, basic characteristics of the child were recorded, e.g. sex, age, body height and weight recorded with a tape measure and household scales, respectively. Body mass index (BMI) was calculated. Intellectual functioning was classified in 3 major levels: (1) normal; (2) learning disability; or (3) mental retardation, according to the German-Swedish distinctions [15]. The limb distribution of a child's spastic CP was classified as unilateral spastic CP

(hemiplegia) or bilateral spastic CP (BSCP). BSCP was subdivided in the leg-dominated form diplegia, or the 4-limb dominated form tetraplegia [17]. In leg-dominated spastic CP or diplegia the arms still can be involved and often are, but to a lesser degree than the legs. Gross motor functioning and manual ability were classified according to the GMFCS [18] and the Manual Ability Classification System (MACS) [19], respectively.

Examination of the lower extremities was performed with the child in supine position and for trunk and upper extremities in sitting position. Using manual passive or (assisted active) flexion/extension of the entire extremities in the major joints, the physician judged muscle tone as elevated (1) or not (0) in each of the 4 limbs (both at rest and in action), following the German-Swedish protocol. Spontaneous pathological postures were assessed by inspection (addressing equinus foot, hip endo-rotation/flexion, elbow flexion, abnormal posture of the shoulder, and impaired head and trunk control). Range of motion (ROM) was assessed in all flexion-extension-rotation directions normally possible in shoulder/elbow/wrist and hip/knee/ankle and graded by the clinician as not restricted/slightly restricted/obviously restricted. Fixed scoliosis and kyphosis was defined as a persistent spine deformity. Radiographic evidence of complete hip dislocation was always verified. The presence and severity of the motor impairments determined the most affected side of the body. In cases of a left-right symmetric presentation we included – arbitrarily – the right side of the body in further analyses.

Activity limitations were assessed according to the Dutch LIVRE system, a standardized recording system used at the time in all rehabilitation centres in the Netherlands [20]. LIVRE is based on the SAMPC model addressing 5 activity domains, i.e. S: Somatic aspects; A: Activities of daily living; M: maatschappelijk (= social functioning in the community); P: psychological functioning (cognition and behaviour) and C: communication [21]. In this study we focused on the first 2 areas, further indicated as mobility activities and self-care activities, respectively. Mobility activities (10 items) included walking, rising, manipulating and lifting, self-care activities (3 items) refer to eating, toileting and washing (see Table 4.3). The items were scored on a 4-point Likert scale describing difficulty of performance of the activity specified, with (0) indicating "manages without problems", (1) "slight difficulty, but manages", (2) "manages only with obvious difficulty or with help" and (3) "does not manage even with help". Sum scores of a domain were calculated, ranging from 0 to 30 (mobility activities) and 0 to 9 (self-care activities). Factor analyses confirmed unidimensionality of each domain (maximum likelihood, oblique rotation) with good reliability (Cronbach's alpha of 0.98 and 0.94, respectively). For further analyses item scores of ≥ 2 were indicated as an activity limitation; to dichotomize the sum scores we used the median score as cut-off point.

Analyses

Cases of missing data were negligible, since data collection took place by means of face-to-face interviews and physical examination. Anthropometric data (body weight, body height and BMI) were compared using the data-set from the Dutch Growth Foundation [22] on the Dutch child population in 1997 by means of 1-sample *t*-tests.

Associations between basic demographic characteristics (sex and age), CP characteristics (limb distribution of paresis, GMFCS and MACS level), and prevalence of motor impairments and limitations in activities were explored by Spearman's rank correlation coefficient. If less than 10% of the children suffered from a specific impairment, no correlation between impairment and CP characteristics was calculated. Additionally, we tested differences between subgroups of patients regarding limb distribution using the Pearson χ^2-tests (in case of motor impairments) and analysis of variance (ANOVA) Tukey Honestly Significant Different *post hoc* tests (in case of activity limitations).

In order to determine the association between motor impairments and activities, univariate logistic regression models were computed. Subsequently the significant variables from the univariate analyses ($p < 0.05$, 2-tailed) were applied to multivariate logistic regression models. These variables were entered as a single block into the regression equation. Nagelkerke *R*-square values were used to reflect the proportion of declared variance. Analyses were performed with SPSS 14.0.

Results

Child and CP characteristics

Table 4.1 gives an overview of the group characteristics of the 119 children with spastic CP. Nearly two-third (75/119) of the patients were boys. Basic anthropometry revealed that these children had both a lower height than the reference population of Dutch children (height related to age ($t = -7.76$, df = 110, $p < 0.001$)) and a lower weight (weight related to age ($t = -4.62$, df = 110, $p < 0.001$). This deviation from the reference population was larger in higher GMFCS levels. The BMI did not differ significantly from the reference population ($t = -1.3$, df = 110, $p = 0.194$).

Sixty percent of the children had bilateral spastic CP (Table 4.1). Almost two-thirds of the children were independent walkers (GMFCS-levels I–II: 64.8%). The MACS distribution showed that the same proportion of the children handled objects without help (MACS levels–I–II: 65%) In addition, approximately two-thirds of the children had normal intellectual functioning or learning disabilities.

Table 4.1 Characteristics of the children with cerebral palsy (CP)

Variable cohort	(*N* = 119)
Child characteristics	
Sex, male, *N* (%)	75 (63.0)
Age, years, mean (SD)	11.1 (3.6)
Length, m, mean (SD)	1.44 (.19)*
Weight, kg, mean (SD)	39.0 (16.8)*
Body mass index, kg/m^2, mean (SD)	18.1 (4.1)
CP characteristics, N (%)	
Limb distribution	
Unilateral spastic CP	48 (40.3)
Bilateral spastic CP	71 (59.7)
Diplegia	42 (35.3)
Tetraplegia	29 (24.4)
GMFCS levels	31 (26.1)
I	46 (38.7)
II	10 (8.4)
III	10 (8.4)
IV	22 (18.5)
V	
MACS levels	
I	23 (19.3)
II	55 (46.2)
III	23 (19.3)
IV	4 (3.4)
V	14 (11.8)
Intellectual functioning	
Normal	43 (36.1)
Learning disability	31 (26.1)
Mental retardation	45 (37.8)

* Significantly lower compared with age-matched peers.
GMFCS, Gross Motor Functioning Classification System; MACS, Manual Ability Classification System; SD, standard deviation.

Limb distribution by GMFCS level correlated strongly (Spearman's $r = 0.78$, $p < 0.001$). The children with GMFCS level I have mostly unilateral spastic CP (90%), whereas in the higher levels bilateral spastic CP was almost exclusively present (7 out of 10 and 22 out of 22 for levels IV and V respectively). The correlation between GMFCS level and MACS level was Spearman's $r = 0.68$ ($p < 0.001$). The distributions of GMFCS and MACS levels are comparable, especially for the low levels (Tables 4.1 and 4.2).

Table 4.2a Motor impairments in numbers (in total cohort of 119 children with spastic cerebral palsy (CP))

	Elevated muscle tone at rest*		Elevated muscle tone in action*		Spontaneous pathological postures				Head and trunk control impaired	
CP characteristic	Arm	Leg	Arm	Leg	Shoulder retraction	Elbow	Hip	Equinus	Head	Trunk
Limb distribution										
Unilateral (N = 48)	16	16	29	31	3	16	7	13	–	–
Bilateral (N = 71)	29	60	52	67	2	21	35	35	16	41
Diplegia (N = 42)	10	30	24	39	2	7	17	15	2	18
Tetraplegia (N=29)	19	27	28	28	0	14	18	20	14	23
Spearman's *r* †	–	0.52	–	0.38	^	–	0.36	0.22	0.32	0.60
p-value		< 0.001		< 0.001			< 0.001	0.015	< 0.001	< 0.001
GMFCS										
I (N = 31)	9	9	20	21	1	8	2	5	–	–
II (N = 46)	15	29	25	36	3	12	17	20	–	4
III (N = 10)	–	7	5	9	–	2	6	4	–	6
IV (N = 10)	5	10	9	10	1	3	5	5	2	10
V (N = 22)	16	21	22	22	–	12	12	14	14	21
Spearman's *r*	0.24	0.50	0.25	0.32	^	0.17	0.37	0.31	0.56	0.78
p-value	0.008	< 0.001	0.006	< 0.001		0.066	< 0.001	0.001	< 0.001	< 0.001
MACS										
I (N = 23)	2	10	7	15	1	1	3	5	–	5
II (N = 55)	18	29	34	43	2	16	16	18	1	2
III (N = 23)	12	20	22	22	2	9	12	12	3	17
IV (N = 4)	4	4	4	4	–	3	3	2	1	3
V (N =14)	9	13	14	15	–	8	8	11	11	14
Spearman's *r*	0.40	0.39	0.52	0.32	^	0.35	0.34	0.33	0.54	0.61
p-value	< 0.001	< 0.001	< 0.001	< 0.001		< 0.001	< 0.001	< 0.001	< 0.001	< 0.001
Total N of children with this impairment	45	76	81	98	5	37	42	48	16	41

* Most affected side.

† Correlation between impairments by limb distribution (1 = unilateral spastic CP, 2 = bilateral spastic CP).

–, not significant; ^, correlation not computed (low number of prevalent cases); GMFCS, Gross Motor Functioning Classification System; MACS, Manual Ability Classification System.

Motor impairments

Tables 4.2a and 4.2b summarize the occurrence of motor impairments (elevated muscle tone, spontaneous pathological postures, impaired trunk or head stability, ROM deficits of the extremities and spine deformities) in relation to limb distribution, GMFCS level and MACS level. Overall, 91 children (76.5%) had no ROM deficits of the upper extremities. Thirteen children displayed 1, and 15 children 2 or 3 ROM deficits in the most affected arm. Similarly, 58 children had no lower extremity ROM deficits, 21 children had 1, and 40 children 2 or 3 ROM deficits in (the most affected) leg. Limb distribution (unilateral vs bilateral spastic CP), gross motor functioning and manual ability showed moderate to good correlation with elevated muscle tone in the legs at rest, impaired head and trunk control and ROM deficits in the leg ($p < 0.001$). Limb distribution (also unilateral spastic CP vs BSCP), did not correlate with impairments in the upper extremity. The prevalence of other motor impairments correlated only poorly with limb distribution and gross motor functioning. More severely affected gross motor functioning correlated, as could be expected, with elevated muscle tone, spontaneous pathological postures and impaired trunk and head control.

Motor impairments were not related to sex and age group. Sub-group comparison analyses of children with BSCP revealed that children with tetraplegia significantly more often suffered from impairments in the upper extremities, equinus position, problems with head and trunk control compared with children with diplegia ($p < 0.05$). The presence of both elevated muscle tone in the legs and spontaneous pathological posture of the hips did not differ between children with tetraplegia and diplegia.

Activity limitations

Table 4.3 shows the degree of functional activity limitation by limb distribution and GMFCS and MACS levels. The specific activities in which the highest proportion of the children encounters limitations were walking outdoors and climbing stairs (40–41%). Regarding self-care activities, 35% of the children had limitations in toileting and washing or bathing. Bilateral limb involvement correlated to limitations in mobility (Spearman's $r = 0.70$, $p < 0.001$) and self-care activities (Spearman's $r = 0.50$, $p < 0.001$). Correlations with levels of gross motor functioning and manual ability ranged from $r = 0.70$ to $r = 0.88$, see Table 4.3. ANOVA post hoc analyses showed that children with tetraplegia encountered more activity limitations than children with diplegia, who in fact experienced more limitations than children with hemiplegia ($p < 0.001$).

Table 4.2b Motor impairments in numbers (in total cohort of 119 children with spastic cerebral palsy (CP)) (contd)

	ROM deficits (1 or more)			Spine deformities	
CP characteristic	Arm*	Leg*	Dislocated hip	Fixed scoliosis	Fixed kyphosis
Limb distribution					
Unilateral (N = 48)	6	12	–	–	–
Bilateral (N = 71)	22	49	4	9	7
Diplegia (N = 42)	6	29	1	3	2
Tetraplegia (N = 29)	16	20	3	6	5
Spearman's *r* †	0.21	0.43	^	^	^
p-value	0.020	< 0.001			
GMFCS					
I (N = 31)	2	5	–	–	–
II (N = 46)	7	25	–	–	–
III (N = 10)	–	6	–	2	1
IV (N = 10)	6	9	2	1	1
V (N = 22)	13	16	2	6	5
Spearman's *r*	0.42	0.43	^	^	^
p-value	< 0.001	< 0.001			
MACS					
I (N = 23)	–	9	–	1	1
II (N = 55)	8	21	–	–	–
III (N = 23)	7	16	2	2	1
IV (N = 4)	4	4	–	2	1
V (N =14)	9	11	2	4	4
Spearman's *r*	0.48	0.32	^	^	^
p-value	< 0.001	< 0.001			
Total N of children with this impairment	28	61	4	9	7

* Most affected side.

† Correlation between impairments by limb distribution (1 = unilateral spastic CP, 2 = bilateral spastic CP).

^ Correlation not computed (low number of prevalent cases).

GMFCS, Gross Motor Functioning Classification System; MACS, Manual Ability Classification System; CP:,cerebral palsy; ROM, range of motion; –, not significant.

Table 4.3 Distribution of activity limitations, by gross motor functioning classification system (GMFCS), manual ability classification system (MACS) and limb distribution (N = 119)

	GMFCS					MACS					Limb distribution		
												Bilateral	
Functional activity domains	I N = 31	II N = 46	III N = 10	IV N = 10	V N = 22	I N = 23	II N = 55	III N = 23	IV N = 4	V N = 14	Unilateral N = 48	Diplegia N = 42	Tetraplegia N = 29
Mobility*													
From lie to sit	–	1	3	7	22	1	2	12	4	14	–	10	23
From sit to stand	–	3	8	10	22	6	4	15	4	14	–	16	21
Walking indoors	–	4	9	10	22	5	5	17	4	14	–	21	22
Walking outdoors	–	7	9	10	22	5	6	19	4	14	1	19	20
Walking stairs	–	7	10	10	22	6	6	19	4	14	–	21	22
Positioning	–	–	2	4	19	1	–	7	3	14	–	6	16
Manipulating	–	4	2	6	19	1	–	13	3	14	1	9	18
Endurance	1	2	3	2	14	2	2	4	2	12	1	5	9
Bending	–	2	4	7	20	1	1	14	3	14	–	6	13
Lifting	–	5	3	7	21	1	4	13	4	14	1	7	13
Sum score (SD)	1.1	5.6	14.9	21.7	27.5	4.5	4.2	18.1	25.8	29.2	2.1	11.5	23.5
(0-30)†	(1.9)	(4.6)	(4.0)	(4.3)	(3.5)	(6.1)	(4.8)	(6.7)	(5.0)	(1.5)	(3.0)	(7.8)	(8.6)
Spearman's *r*					0.88					0.72			0.70‡
p-value					< 0.001					< 0.001			< 0.001
Self-care*													
Eating / drinking	–	3	1	4	19	1	2	7	3	14	1	8	18
Toileting	–	7	3	9	22	3	4	16	4	14	4	13	24
Washing / bathing	–	10	3	8	22	2	7	16	4	14	4	16	23
Sum score (SD)	0.4	1.6	2.4	5.6	8.2	1.1	0.9	5.1	8.0	8.6	0.9	2.7	6.6
(0-9)†	(0.6)	(2.4)	(2.4)	(2.1)	(1.1)	(1.8)	(1.7)	(2.5)	(1.4)	(0.6)	(1.7)	(3.0)	(2.9)
Spearman's *r*					0.74					0.70			0.50‡
p-value					< 0.001					< 0.001			< 0.001

* Numbers refer to cases with a limitation in activity, i.e. obvious difficulty (requires assistance or major adaptations or completely incapable).

† Mean sum score (SD).

‡ Correlation between domain score by limb distribution (1 = unilateral spastic CP, 2 = bilateral spastic CP).

SD, standard deviation; CP, cerebral palsy.

Associations between motor impairments and activity limitations

As presented in Table 4.4, limitations in mobility activities were associated with deficits in the lower limbs in univariate modelling, while self-care activities were constrained by impairments in both the upper and lower extremities ($p < 0.001$ to $p < 0.05$).

Table 4.4 Determinants of limitations in activities, dichotomized sum scores (cut-off point median score). Univariate and multivariate logistic regression analyses (N = 119)

	Univariate analyses				Multivariate analyses			
	Mobility activities		Self-care activities		Mobility activities		Self-care activities	
Impairments†	OR (95% CI)	R^2	OR (95% CI)	R^2	OR (95% CI)	R^2	OR (95% CI)	R^2
Determinants						0.42***		0.34***
Tonus‡								
Arm at rest	n.s.		3.1** (1.4–6.7)	0.09	–		n.s.	
Arm in action	n.s.		3.0** (1.3–6.8)	0.08	–		n.s.	
Leg at rest	8.9*** (3.6–22.1)	0.27	3.5** (1.6–7.9)	0.11	n.s.		n.s.	
Leg in action	8.0** (2.2–28.9)	0.15	2.9* (1.1–8.2)	0.05	n.s.		n.s.	
Spontaneous pathological postures								
Armδ	n.s.		2.5* (1.2–5.2)	0.07	–		n.s.	
Leg¶	3.2*** (1.8–5.7)	0.20	3.9*** (2.1–7.0)	0.25	n.s.		2.8*** (1.3–5.7)	
ROM deficits								
Arm§	2.2** (1.3–3.8)	0.11	2.4** (1.3–4.3)	0.12	n.s.		n.s.	
Leg°	3.1*** (2.0–4.8)	0.35	1.9*** (1.4–2.8)	0.16	2.0** (1.2–3.4)		n.s.	
Spine deformities♦	n.s		n.s.		–		–	

* $p < 0.05$, ** $p < 0.01$, *** $p < 0.001$.
† In limb of most affected side.
‡ 0 = not elevated; 1 = elevated.
δ Presence of spontaneous pathological posture in elbow and/or shoulder (range 0–2).
¶ Presence of spontaneous pathological posture in ankle and/or hip (range 0–2).
§ ROM deficit of wrist and/or elbow and/or shoulder (range 0–3).
° ROM deficit of ankle and/or knee and/or hip (range 0–3).
♦Presence of fixed scoliosis and/or kyphosis (range 0–1).
OR, odds ratio; CI, confidence interval; R^2, explained variance by Nagelkerke *R*-square test; –, not tested; n.s., not significant.

Multivariate models included only the determinants that were significant in the univariate analysis, and demonstrated that elevated muscle tone, as such, was no longer a determinant of activity limitation, once the other motor impairments were taken into account. We found, however, that children with one or more ROM deficits or a pathological posture in the legs were 2–3 times more frequently limited in both mobility and self-care activities ($p < 0.001$ to $p < 0.01$).

Discussion

The nature and prevalence of motor impairments and activity limitations in spastic CP has been studied in previous publications, originating from population-based studies or pooled populations [2, 4–6]. The results of these studies share elements such as distributions of clinical presentations and GMFCS and MACS levels, but in their conclusions and considerations accents differ. For example Östensjö et al. [4] reported in 2004 that "spasticity and ROM deficits were both stated to be of importance for predicting functional performance along with selectivity of movement"; however, "motor impairments were only one component among many factors that could predict gross motor function and everyday activities". This finding is confirmed in our study.

More recently in the Netherlands, longitudinal studies on the nature and course of motor impairments and activities in children and adolescents with CP have been and are being performed within the PERRIN programme (PEdiatric Rehabilitation Research In the Netherlands) [23, 24]. PERRIN studies recruit their subjects via cooperating rehabilitation centres, thus focusing on a group within the CP population. CP (sub)groups that are followed longitudinally give good intra-subject and intra-group insight. The cross-sectional population-based study presented in this paper covers a wide field of aspects regarding CP children. Thus, in the Netherlands, cross-sectional "population-based" data join longitudinal "focused" data.

Children's characteristics

A general description of a representative group of 119 Dutch children with spastic CP is provided, as well as prevalence data on specific impairments and activities in mobility and self-care. The distribution of boys/girls and of major clinical characteristics is in line with other representative pooled data [1]. Children with spastic CP had both a lower age-adjusted body height and body weight compared with typically developing peers [22]; this was especially the case in children with higher GMFCS levels. This

latter finding is in line with reports from multi-centre studies on growth in American children with moderate to severe CP (GMFCS III–V) [25]. Children with spastic CP did not differ from the general population with respect to their BMI. Although CP is more prevalent in boys [1], we found that the consequences of CP in terms of the nature of specific motor impairments and performance of activities were not related to sex or to age-group.

ROM deficits were encountered in 60% of the children. This means that no less than 40% of the children with spastic CP (especially the children with low GMFCS levels) had no ROM deficits whatsoever. Either the natural course of spastic CP in these cases had not resulted in what professionals indicate as contractures, or preventive treatment had been completely effective. This is interesting, as the need to prevent or treat the unavoidability of "contractures" in CP is stressed frequently. We find that the "need to treat" is probably less present in less severe cases of CP. Children with spastic CP in GMFCS and MACS levels I and II who use their extremities actively perform ROM exercises in a sufficient way.

Severe orthopaedic problems, such as complete hip dislocation or fixed spine deformities, were seen only in GMFCS IV–V levels. However, the prevalence of these severe orthopaedic problems was low; only 4 children had hip dislocation (and this only unilaterally). These low numbers are puzzling as the co-existence of severe CP and hip dislocation is reported frequently. In a recent English study, within a group of children with bilateral spastic CP and GMFCS level V (41 individuals), 50% had a hip dislocation by the age of 15 years [26]. In an older population-based Swedish study, 75% of children with tetraplegic CP had hip dislocation and scoliosis [27]. In recent years Boldingh et al. [28] examined 160 Dutch patients with severe tetraplegic CP, aged 16–84 years, and found "moderate" hip deformity in 41% and "severe" hip deformity in 29%. These last 2 studies focused on subjects with 4-limb involvement and a high GMFCS level (which is of course a selected group) and included (much) older patients who may not have had preventive treatment during their growth. An explanation might be that in the Netherlands the long-standing practice of radiographic monitoring of hip migration including timely conservative or operative measures results in low rates of complete hip dislocation. Another possibility is that the average age of our cohort is lower than the age at which dislocations become manifest. Due to the low number of hip dislocations in our cohort we cannot analyse the relation with age subgroups. The low prevalence of hip dislocation in the Dutch population could be the subject of further research.

As could be expected, more severely affected gross motor functioning (higher GMFCS levels) and total body involvement, such as in tetraplegia, correlated strongly with the degree of impairment present and the presence of limitations in activities. This

finding is not surprising and is frequently reported [2, 4, 6, 24]. GMFCS and MACS are known to correlate, as shown by Eliasson et al. [19] and in this study. However, the difference of distribution in the higher levels between GMFCS and MACS show that they do classify different types of activities.

Over 60% of the children could walk without assistive walking devices (GMFCS levels I–II). Thus, a typical child with spastic CP will be an independent walker rather than a wheelchair-user. This finding is consistent with European population-based CP-studies pooling data from more than 6000 children [29].

Some aspects of the study should be addressed

We assessed the motor impairments and activity limitations, as they were encountered in the group of children with spastic CP who were being treated according to professional standards in the Netherlands. Previous and current treatments and interventions (orthopaedic surgery, orthotics, medication) were known to us from the parents' interview. No causal relationships have been statistically explored in this study between the impairments found and specific previous interventions, such as anti-spastic medication, advanced spasticity treatment (these 2 were hardly present in this cohort) or orthotics and orthopaedic operations (which both were frequently encountered). Impairments can be both present or absent, either in the natural course of CP or when interventions are (repeatedly) undertaken. Exploration causality between interventions and impairments would need a longitudinal study design, such as a Swedish longitudinal study with the focus on prevention of hip dislocation, showing that that implementation of a protocol with radiographic hip development follow-up and timely interventions resulted in less dislocations compared with a control group that lacked this approach [7].

The distinction between diplegia and tetraplegia is supposed to be mainly of clinical importance (clinicians "picture" a child from this type of description). It has been suggested to use only the term "bilateral spastic CP" or BSCP (for CP epidemiology), which has the benefit of avoiding debates where diplegia stops and tetraplegia starts, but also because the GMFCS level by itself describes functional performance and a limb-oriented classification could move to the background [8]. We recognized the important main groups unilateral and bilateral spastic CP, but kept track of the leg-dominated and 4-limb dominated subcategories of BSCP – diplegia and tetraplegia (owing to the fact that we used the German-Swedish protocol). We found that diplegia and tetraplegia differed not only from an impairment point of view (as reported by Östensjö et al. [4]), but also from an activity point of view, as the children

with spastic diplegia had fewer limitations compared with the children with tetraplegia, in self-care activities but also in the broad domain of mobility activities, including positioning and manipulating. We conclude that besides the obvious relevance of the terms diplegia and tetraplegia to physicians treating individual children, these terms do refer to differences in activities, which, for a child with the upper extremities less affected, also seems logical.

Clinical practice often assumes a direct relationship between impairments and activity limitations. Indeed, univariate models showed that increased tonus, pathological postures and ROM deficits were related to limitations of the studied activities. However, multivariate relations between impairments and activities revealed that mainly ROM deficits in the lower extremity were related to mobility activities and spontaneous pathological postures were related to self-care. Because there were only modest associations between the presence of motor impairments and limitations in activities, a treatment that specifically targets motor impairments (such as disorders of muscle tone) may not be sufficient to achieve an enhancement of activities. Other aspects, such as environmental aspects or non-motor impairments, have to be taken into account. Moreover, what the ability to perform a given activity means to a child and his or her parents might be influenced by individual perceptions and expectations. For example, a child might prefer using a wheelchair with ease to walking, if walking is only possible with great effort using walking aids.

Limitations of the study

First there is a possibility of under-reporting of "minor however present" motor impairments and activity limitations as we chose to do the analyses with motor impairments that were more than "slight, or minor" and with activity limitation that were at least valued as "obvious". Including the "slight" or "minor" category for both could give rise to threshold-issues between "no problems" and "very minor problems" and the practical relevance may be limited.

The LIVRE method was designed in the 1990s as a registration tool and has not been validated, in the way the Pediatric Evaluation of Disability Inventory (PEDI) has been, which became available also in Dutch [30]. LIVRE was in use in all Dutch rehabilitation centres at the time of the study and gave a bird's-eye view of the patients' functioning in 5 major domains of functioning. To date in 2008 the 5 SAMPC domains used in LIVRE are still the basis of many systematic medical patient files in rehabilitation medicine in the Netherlands. The 4-step LIVRE grading of none, minor, obvious difficulty up to impossibility to perform the activity resembles the

result-oriented scoring of PEDI. Moreover, the risk that we indicated a limitation erroneously is low, since we started at obvious difficulty or worse as scores to indicate an activity limitation.

GMFCS and MACS were not available at the time the physicians examined the child. The classification was done on a retrospective basis by the first author. Comprehensive information from the parents on the child's performance in daily life, assistive devices, the personal examination and an extra qualitative structured description of both walking and of hand and arm function yielded a vivid picture of the child. Both GMFCS and MACS are known to be rather unequivocal, use descriptions of the levels that also non-professionals can deal with, which contributes to the good inter-observer reliability [18, 19]. So "knowing, examining and observing the child personally" formed a good basis for post-hoc classification. Borderline classification issues will always be present (and will be as well in the observations done today) but the contours of the different levels are clear-cut.

In conclusion, the distribution of CP-related characteristics in this Dutch cohort is consistent with that found in other representative studies. The prevalence of motor impairments and activity limitations has been determined in relation to major CP characteristics. A markedly low rate of hip dislocation was found in comparison to other studies. Distinction between diplegia and tetraplegia is relevant from a clinical point of view but also from an activity point of view. Activity limitations are determined only partly by the mere presence of motor impairments, which confirms the findings of other studies [4, 8]. Individual goal setting in rehabilitation should identify all factors relevant to the child, including environmental factors. An activity-oriented rehabilitation approach goes beyond the treatment of motor impairments that are present.

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5

Psychological functioning, communication and behaviour difficulties in children with cerebral palsy: A Dutch population-based study

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Abstract

Objective: To explore the prevalence and related characteristics of personality descriptors, communication and behaviour difficulties in school-aged children with cerebral palsy.

Patients and methods: A Dutch population based cohort of 127 children with cerebral palsy aged 6-19 years. Personality descriptors (addressing self-esteem, control, prosocial capacity and mood), difficulties in communication, contactual skills and problem behaviour were assessed during a parent-reported interview. Spearman's correlation coefficients were used to explore associations with CP related characteristics (intellectual level, gross motor functioning, and laterality of the paresis).

Results: The degree to have control over one's life and self-esteem appeared to be restricted in 10-20% of the children. The ability to express one-self was impaired in 26%, writing in 50%. About one third suffered from incidental to frequent behaviour problems. Impaired intellectual functioning was an associated factor, while impaired gross motor functioning affected some domains.

Conclusions: The most prevalent psychological and socio-emotional problems in children with CP appeared to be emotional instability, being dependent, lack of confidence and communication limitations (self expression and writing). Children without intellectual disability seem to function without significant problems.

Introduction

The prevalence of cerebral palsy (CP) in the Netherlands is estimated to be 2.44 per 1,000 living births [1]. Children with CP may present a variety of motor impairments that impact their functional activities [2]. Motor impairments resulting from CP have been the subject of many publications. Other publications focused on perceived competence and social issues in children and adolescents with CP, such as relationships with peers and parents and social participation [3-7].

Few studies described psychological functioning and socio-emotional behaviour difficulties of children with CP into more detail [8-11]. A representative British study on children with unilateral CP (aged 6–10 years) reported that 61% of the children who were individually assessed suffered from psychiatric disorders, with emotional disorders – mostly anxiety (25%) and behavioural disorders (24%) – most frequently being observed [9]. A majority of these problems persisted in a four-year period [10]. More recently, a large-scale European study using population-based cohorts of children with CP aged 8–12 years identified peer problems (32%), problems concerning hyperactivity (31%), emotion (29%) and behaviour (17%) as clinically relevant issues [8]. In a Dutch rehabilitation-based cohort among adolescents internalizing behaviour (e.g., being shy, feeling alone, unhappy) correlated negatively with several aspects of self-esteem, but did not correlate with level of gross motor function [11].

Specific CP related characteristics appear to be related to psychological problems. Greater severity of neurological damage and impaired cognitive functioning were bivariately associated with psychiatric problems in unilateral affected children [9]. In addition to impaired intellectual functioning, impaired communication abilities – i.e. having hearing aids, speech impairment – were reported to correlate with emotional and behavioural problems in children with various types of CP [8]. The finding that children with low levels of gross motor functioning showed less emotional and behavioural symptoms compared to children with a high level of gross motor functioning (GMFCS level I) might be explained by them being less able to participate in poor behaviour, or by the impossibility to validly assess these symptoms in severely disabled children CP [8]. Overall, the above studies suggest that CP related characteristics might increase the risk of problems regarding psychological functioning and socio-emotional behaviour.

Aim of the present study was to describe personality descriptors – including self-esteem, control, social competence, mood – and to screen communication and behaviour difficulties in a representative cohort of Dutch children with CP aged 6–19 years. Furthermore, we explored associations of CP related characteristics, addressing both intellectual and motor disability, with these aspects of functioning.

Methods

Subjects

The present study is part of a cross-sectional population-based study on prevalence and clinical presentation of CP in the Netherlands [1, 12, 13]. CP was defined as a disorder of movement and posture caused by a non-progressive brain lesion with an onset no later than one year after birth. Evident neurological symptoms (e.g., spasticity, dyskinesia or ataxia) were obligatory [14]. Patients were included if they had (a) a diagnosis of 'cerebral palsy' recorded in the patient files, (b) date of birth between 1 January 1977 and 31 December 1988, and (c) parents living – at the time of the study – in Gelderland, a region in the east-central part of the Netherlands. Evidence for completeness of case ascertainment and representativeness of the study cohort was given in a previous publication [1]. Informed consent was obtained from the parents of the children. The study was approved by the ethics committees of the region's university medical centre and collaborating institutions.

Data collection

Data collection was carried out according to the study protocol of previous German and Swedish studies on the epidemiology of CP [15-17]. The protocol and classification criteria were discussed and trained (in advance) together with the authors of these studies.

An experienced rehabilitation physician visited each child and his or her parents/care-takers. A structured interview with the parents or caretakers and a physical examination of the child were performed. Basic characteristics of the child were recorded, e.g. sex, and age. Intellectual functioning was classified in 3 major levels, according to the German-Swedish distinctions [16]: (1) normal: child attends regular school, no known learning difficulties; IQ estimated at 85 or higher, (2) learning disability: specific schooling needs; estimated IQ 70–85, and (3) mental retardation: very specific school or day centre (either elementary learning, some practical training, or no learning whatsoever); estimated IQ below 70. Level of gross motor functioning was classified according to the Gross Motor Functioning Classification System (GMFCS) [18]. In spastic chidren laterality was classified as unilateral or bilateral spastic CP [19].

Psychological functioning

We screened personality descriptors using a list of 16 bipolar personality types or emotions (e.g., emotional – stable, bored – interested, unhappy – happy), adapted from

Osgood et al. [20]. On each item respondents choose their position on a five-points scale (0–4). Most items were scored by the parents of the children, although in many cases they had checked the answers with their children if the children were capable of expressing their opinion. From twenty adjective pairs that were assumed relevant to (severe) brain injury [21], we excluded four items as ambiguous for children with CP (i.e., clever vs smart, forgetful vs attentive, distracted vs concentrated, handy vs clumsy). Factor analysis (oblique rotation) revealed five factors (16 items) of which four domains (13 items) were interpretable. Domains addressed (a) self-esteem (3 items, Cronbach's alpha 0.69), (b) control (2 items, Cronbach's alpha 0.67), (c) prosocial capacity (4 items, Cronbach's alpha 0.73), and mood (3 items, Cronbach's alpha 0.71). Single item scores < 1 and a domain-score < *k* (*k* items in domain) indicated a subnormal level.

Communication difficulties

Communication performance was assessed according to the Dutch LIVRE system, a standardized recording system of functioning in several domains, used in rehabilitation centres of the Netherlands at the time of study [22]. The communication domain consisted of four items (i.e., self-expression, hearing, seeing, writing) scored on a four-point Likert scale ((0) complete limitations or impossible, (1) severe limitations, (2) mild limitations or problems, and (3) no problems), calculating a sum-score (Cronbach's alpha 0.72). A single item scores < 1 and a domain-score < 4 indicated a subnormal level.

Behaviour difficulties

Behaviour difficulties were screened addressing the aspects of capacity and performance. We assessed contactual skills of the children in three categories ((0) normal, (1) impaired (stereotype speech, inappropriate emotions in social context), and (2) severely impaired), according to the German-Swedish protocol. Also parent-perceived problem behaviour was assessed, using a single question with 3 categories: (0) no problem behaviour, (1) incidental, and (2) frequent or unbearable problem behaviour (impact on environment). Using parent perceived difficulties in problem behaviour as a one-item screen is known to be a good predictor of clinical status [23].

Analyses

Descriptive statistics are performed for the total cohort, and subgroups of children regarding intellectual functioning (3 subgroups), dichotomized GMFCS levels (GMFCS

levels I/II and III-V) and limb distribution of spastic paresis (unilateral and bilateral, N = 119; not applicable for N = 8 children with non-spastic CP). With chi-square tests (categorical data) or oneway analysis of variance (age) we tested whether three subgroups of intellectual functioning differed regarding sex, age and CP characteristics. Several personality descriptors were not assessable in 17 to 19 children of the subgroup of children with mental retardation; these were indicated as missing values in the analyses on these aspects. Associations with child and CP characteristics were explored by means of Spearman's rho (except for age: Pearson's correlation). Analyses were performed with SPSS 14.0.

Results

Subjects

Table 5.1 presents characteristics of the cohort of 127 children with CP, of which 44 children (34.6%) had normal intelligence (NI), 34 children (26.8%) had a learning disability (LD) and 49 (38.6%) a mental retardation (MR). The total sample consisted of 78 boys, children had a mean age of 11.1 years (SD = 3.6 years). Unilateral CP was seen in 37.8%, bilateral CP in 55.9% of the children and was observed more often in the MR group (Chi-square = 14.3, df = 4, $p = .006$). In total 63.8% of the children were independent walkers (GMFCS-levels I–II), children in the MR group had lower levels of gross motor functioning (Chi-square = 38.8, df = 8, $p < .001$). Distribution of gender and age did not differ between subgroups of intellectual functioning.

Psychological functioning: personality descriptors

Table 5.2 shows the item-scores and domain-scores on the four domains addressing distinct aspects of personality. Data are presented for the total sample, and for three subgroups of intellectual functioning (NI, LD and MR). Higher scores represent better functioning. The domains of prosocial capacity and mood indicated relatively few problems. The degree of experiencing control appeared to be hampered in 21% of the children, and appeared more often in the MR and LD groups compared to the NI group (Spearman $r = -0.45$, $p < .001$). Although self-esteem seemed not to be a major problem (prevalence of 12%), lack of confidence and emotional instability were frequently observed: on these items 25.7% and 41.8% of the sample had subnormal scores. In children with MR emotional instability was a problem in 53%.

Table 5.1 Characteristics of the cohort; children with CP (N = 127)

Variable	Cohort (N = 127)	Children with NI (N = 44)	Children with LD (N = 34)	Children with MR (N = 49)
Child characteristics				
Sex (male) (N (%))	78 (61.4)	26 (59)	21 (62)	31 (63)
Age in years (mean (SD))	11.1 (3.6)	10.6 (3.6)	11.3 (3.6)	11.3 (3.5)
Intellectual functioning (N (%))				
Normal	44 (34.6)	44 (100)		
Learning disability	34 (26.8)		34 (100)	
Mental retardation	49 (38.6)			49 (100)
CP characteristics (N (%))				
Limb distribution				
Unilateral spastic CP	48 (37.8)	26 (59)	11 (32)	11 (22)*
Bilateral spastic CP	71 (55.9)	17 (39)	20 (59)	34 (70)
Non-spastic CP	8 (6.3)	1 (2)	3 (9)	4 (8)
GMFCS				
I	32 (25.2)	21 (48)	8 (23)	3 (6)**
II	49 (38.6)	17 (39)	11 (32)	21 (43)
III	12 (9.4)	1 (2)	7 (21)	4 (8)
IV	10 (7.9)	4 (9)	3 (9)	3 (6)
V	24 (18.9)	1 (2)	5 (15)	18 (37)

NI, normal intelligence; LD, learning disorder; MR, mental retardation.
* $p < 0.01$, ** $p < 0.001$.

Communication and behaviour difficulties

Regarding communication, self-expression and writing were common problems (26% and 50.4% respectively), strongly correlated to level of intellectual functioning (Spearman $r = -0.73$, $p < .001$). Of children with MR 63% and 86% scored subnormally on these aspects.

Difficulties in contactual skills and incidental or frequent behaviour problems according to their parents were prevalent in one out of three children of the sample. Contactual skills as well as parent reported problem behaviour were normal in the NI group (100 tot 93%).

Difficulties in contactual skills were specifically prevalent in children with MR (67%), whereas problem behaviour was both seen in children with LD or MR (41 or 51%), see Table 5.3. Both aspects of behaviour difficulties correlated moderately to level of intellectual functioning (Spearman $r = 0.63$ or 0.42, $p < .001$).

Table 5.2 Personality descriptors by levels of intellectual functioning

Item / Domain	Cohort (N = 127)		NI (N = 44)		LD (N = 34)		MR (N = 49)[a]		Spearman *r* (*p*)
	Mean (SD)	Subnormal N (valid %)	Mean (SD)	Subnormal N (valid %)	Mean (SD)	Subnormal N (valid %)	Mean (SD)	Subnormal N (valid %)	
Personality descriptors[b]									
Lacks confidence – Self confident	2.1 (1.2)	28 (25.7)	2.8 (1.3)	9 (20)	1.8 (1.1)	12 (35)	2.3 (1.3)	7 (23)	
Emotional instable – Stable	1.8 (1.2)	46 (41.8)	2.1 (1.3)	15 (34)	1.7 (1.0)	14 (41)	1.4 (1.0)	17 (53)	
Worthless – Of value	3.1 (1.0))	6 (5.8)	3.1 (1.0)	2 (5)	3.1 (1.2)	3 (9)	3.0 (1.0)	1 (4)	
Domain score Self-esteem	6.8 (2.8)	13 (12.0)	7.4 (3.0)	6 (14)	6.4 (2.8)	4 (12)	6.4 (2.6)	3 (10)	-0.17 (0.08)
Helpless – In control	2.6 (1.0)	12 (11.0))	3.1 (0.8)	1 (2)	2.4 (0.9)	4 (12)	2.1 (1.1)	7 (23)	
Dependent – Independent	2.0 (1.3)	39 (35.4)	2.8 (1.2)	7 (16)	1.7 (1.3)	15 (44)	1.4 (1.3)	17 (53)	
Domain score Control	4.5 (2.1)	23 (21.0)	5.7 (1.7)	3 (7)	4.1 (1.8)	8 (24)	3.4 (2.0)	12 (39)	-0.45 (< 0.001)
Bored – Interested	3.3 (1.0)	6 (5.4)	3.5 (1.0)	3 (7)	3.3 (0.8)	0 (0)	2.9 (1.1)	3 (9)	
Unfeeling – Caring	3.7 (0.7)	2 (1.8)	3.7 (0.7)	1 (2)	3.7 (0.5)	0 (0)	3.5 (1.0)	1 (3)	
Difficult – Co-operative	3.0 (1.0)	6 (5.4)	3.1 (0.9)	1 (2)	2.9 (0.9)	1 (3)	2.8 (1.1)	4 (13)	
Passive – Active	2.9 (1.1)	11 (9.9)	3.2 (1.0)	2 (5)	2.8 (1.1)	5 (15)	2.7 (1.3)	4 (12)	
Domain score Prosocial	12.8 (2.9)	3 (2.7)	13.5 (2.7)	0 (0)	12.7 (1.9)	0 (0)	11.6 (3.8)	3 (9)	-0.26 (0.01)
Unhappy – Happy	3.4 (1.0)	8 (7.2)	3.4 (1.0)	3 (7)	3.2 (1.0)	2 (6)	3.4 (1.1)	3 (9)	
Worried – Relaxed	2.9 (1.2)	16 (14.7)	2.9 (1.3)	8 (18)	2.9 (1.2)	5 (15)	3.1 (1.1)	3 (9)	
Despondent – Hopeful	3.3 (0.9)	5 (4.6)	3.5 (0.9)	1 (2)	3.0 (1.0)	3 (9)	3.3 (0.9)	1 (3)	
Withdrawn – Talkative	3.0 (1.2)	13 (12.0)	3.1 (1.2)	4 (9)	3.2 (1.2)	5 (15)	2.7 (1.1)	4 (13)	
Domain score Mood	12.5 (3.2)	2 (1.8)	12.9 (3.3)	1 (2)	12.2 (3.2)	1 (3)	12.5 (3.0)	0 (0)	-0.10 (0.33)

NI, normal intelligence; LD, learning disability; MR, mental retardation.

[a] In 17–19 children with MR personality descriptors were not assessable (missing values).

[b] Subnormal functioning on items are indicated by item-scores 0 or 1 (range 0–4); Subnormal functioning on domains are indicated by domain-scores ≤ *k* (*k* items in domain; range 0–*k**4).

Table 5.3 Communication and behaviour difficulties by levels of intellectual functioning

Item / Domain	Cohort (N = 127)		NI (N = 44)		LD (N = 34)		MR (N = 49)		Spearman *r* (*p*)
	Mean (SD)	Subnormal N (valid %)	Mean (SD)	Subnormal N (valid %)	Mean (SD)	Subnormal N (valid %)	Mean (SD)	Subnormal N (valid %)	
Communication[a]									
Self-expression	2.1 (1.1)	33 (26.0)	2.9 (0.4)	1 (2)	2.5 (0.7)	1 (3)	1.1 (1.2)	31 (63)	
Hearing	2.8 (0.7)	7 (5.5)	3.0 (0.0)	0 (0)	2.9 (0.6)	2 (6)	2.6 (0.9)	5 (10)	
Seeing	2.4 (0.9)	19 (15.0)	2.9 (0.4)	1 (2)	2.4 (0.7)	4 (12)	2.0 (1.2)	14 (29)	
Writing	1.4 (1.3)	64 (50.4)	2.3 (1.0)	7 (16)	1.5 (1.1)	15 (18)	0.5 (0.9)	42 (86)	
Domain score Communication	8.8 (3.0)	13 (10.3)	11.2 (1.1)	0 (0)	9.3 (2.1)	1 (3)	6.2 (2.9)	12 (25)	-0.73 (< 0.001)
	N (valid %)		N (valid %)		N (valid %)		N (valid %)		
Behaviour difficulties									
Contactual skills									0.63 (< 0.001)
Normal	90 (71.4)		44 (100)		30 (88)		16 (33)		
Limited	20 (15.9)				3 (9)		17 (36)		
Severely limited	16 (12.7)				1 (3)		15 (31)		
Problem behaviour									0.42 (< 0.001)
No	85 (66.9)		41 (93)		20 (59)		24 (49)		
Incidental	32 (25.2)		3 (7)		13 (38)		16 (33)		
Frequent	10 (7.9)				1 (3)		9 (18)		

NI, normal intelligence; LD, learning disability; MR, mental retardation.

[a] Subnormal functioning on these items are indicated by item-scores 0 or 1 (range 0–4). Subnormal functioning on this domain are indicated by domain-scores 0–4 (range 0–16).

Table 5.4 shows that children with higher levels of gross motor functioning (GMFCS I–III) scored better on experiencing control, and had less limitations in communication (including writing, which addresses an aspect of motor functioning) and contactual skills (Spearman r = -0.36 to -0.49, $p < .001$). Except for contactual skills, similar correlations were found with laterality of the spastic paresis (Table 5.4). Other personality descriptors and problem behaviour were not associated with gross motor functioning or laterality of the paresis.

Discussion

The present study described the prevalence of difficulties in psychological functioning, communication and problem behaviour in school-aged children with CP. Compared with previous publications, the present study includes all types of CP and covers a broad age-range. Concerning intellectual functioning, almost 40% of children with CP were mentally retarded, and another quarter had a learning disability.

Addressing personality characteristics, the most prevalent problems (greater than 25%) appeared to be emotional instability, being dependent and lack of confidence. In children with normal intelligence specifically emotional instability appeared to be an issue. Children with mental retardation did not differ from the other children with respect to mood. Although we lack reference data of healthy age-mates on the four personality descriptors, our data suggest that children with CP may face additional problems with their emotional stability and feeling to have control over their lives. Negative mood might be a response to the dependence on others. In a study by Sandström, an adult with CP remarked "I feel like I'm a burden to so many people" [24].

Severe communication difficulties were common, especially in writing and self-expression. A minority of the children encountered severe limitations in contactual skills and frequent problem behaviour. Communication and problem behaviour seemed modertaly related to level of gross motor functioning. The majority of communication and behaviour problems occurred in children with mental retardation. Also other studies reported that these children had increased risk for emotional and behavioural problems [8, 9].

Additionally, even children with moderate learning disability showed an increased prevalence of these problems. For clinical practice this means that apart from the attention for school achievements professionals need to be sensitive to emotional and behaviour problems, to support these children's participation and autonomy later in life.

Table 5.4 Personality descriptors, communication and behaviour difficulties by physical CP characteristics

	GMFCS (N = 127)			Limb distribution (N = 119)		
	I–III	IV–V		Unilateral	Bilateral	
	Mean (SD)	Mean (SD)	Spearman *r* (*p*)	Mean (SD)	Mean (SD)	Spearman *r* (*p*)
Personality descriptors[a]						
Self-esteem	6.8 (2.9)	6.8 (2.6)	-0.02 (0.88)	7.1 (2.8)	6.9 (2.8)	-0.05 (0.61)
Control	4.9 (2.0)	3.1 (1.9)	-0.36 (< 0.001)	5.5 (1.8)	4.1 (1.9)	-0.37 (< 0.001)
Prosocial	12.9 (3.0)	12.0 (2.7)	-0.17 (0.84)	13.2 (2.8	12.5 (2.8)	-0.15 (0.13)
Mood	12.6 (3.3)	12.4 (2.6)	-0.07 (0.46)	12.8 (3.2)	12.4 (3.1)	-0.06 (0.56)
Communication	9.7 (2.2)	6.1 (3.3)	-0.49 (< 0.001)	10.5 (1.8)	7.9 (3.1)	-0.46 (< 0.001)
Behaviour difficulties	N (valid %)	N (valid %)		N (valid %)	N (valid %)	
Contactual skills			0.41 (< 0.001)			0.15 (0.11)
Normal	75 (81)	15 (44)		39 (81)	49 (70)	
Limited	14 (15)	6 (18)		7 (15)	9 (13)	
Severely limited	3 (3)	13 (38)		2 (4)	12 (17)	
Problem behaviour			-0.05 (0.57)			0.04 (0.66)
No	61 (65)	24 (71)		34 (71)	47 (66)	
Incidental	24 (26)	8 (23)		10 (21)	19 (27)	
Frequent	8 (9)	2 (6)		4 (8)	5 (7)	

GMFCS, Gross Motor Function Classification System.

[a] In 17–19 children personality descriptors were not assessable (missing values).

We need to address some limitations of the study. Although we used the assessment methods that were used in other study protocols [16], these methods lack validation data. For instance, for the personality descriptors we used of the semantic differential approach as used by Tyerman in patients with traumatic brain injury [21]. By means of factor analyses and experts opinion we discerned four domains. Although the internal consistency of these domains was sufficient, the method lacks validation to know how these measures correlate with other instruments for these concepts. To screen difficulties in communication we used the domain Communication, adapted from the LIVRE method. The LIVRE method was in use in all Dutch rehabilitation centres at the time of the study. It was designed as a registration tool, giving a bird's eye view of the patients' functioning in five major domains of functioning [12].

In conclusion, in the present study school-aged children with CP showed overall few behaviour problems or severely limited contactual skills. Severe limitations in communication, especially in writing and self-expression, were more prevalent. If there were problems in one of the investigated areas, these were largely related to severely impaired intellectual level. However, children with mental retardation did not differ from the other children with respect to mood.

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6

Health care utilisation and educational career of Dutch children with cerebral palsy

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Abstract

Objective: To describe health care utilisation and school careers of Dutch children with cerebral palsy (CP).

Design: Cross-sectional – population-based survey.

Subjects: 127 children with CP, age 6 through 18.

Methods: Parent interview on the child's previous and current situation in medical and allied health care and school career.

Results: Eighty percent of the Dutch children with CP are seen on a regular basis by a consultant in rehabilitation medicine. Hip and ankle-foot surgery has been performed in 24 and 20 percent of cases respectively, hip surgery mainly in strongly affected children (GMFCS level IV or V). Apart from anti-epileptic medication and laxatives (in 20% and 5% of cases respectively) medication is hardly taken, notably no spasmolytic drugs. Orthotic devices are used by 80%, mobility devices by 50%. 62% of the children are treated in a multidisciplinary setting and 56% attend special schools, treatment and school usually in combination. Less affected children (GMFCS level 1 and normal intelligence) more often had monodisciplinary or no treatment and attended mainstream schools.

Conclusion: Dutch practice regarding CP children, especially those more affected, features special schools combined with multidisciplinary treatment. Adaptive devices and orthopaedic surgery are widely used. Medication use is limited to anti-epileptics or laxatives. Timely hip surgery may contribute to the low rate of hip dislocation in the Netherlands.

Introduction

Cerebral palsy (CP) is an umbrella term for a diversity of aetiologies and clinical presentations. CP is the most common cause of primary motor deficit. Most children with CP have a spastic type of motor disorder, many of them have associated disorders and the degree of activity limitation varies [1, 2].

Many children with CP need additional health care and education facilities [3]. In the Netherlands allied health care facilities exist in community-based mono-disciplinary practices as well as in regional multidisciplinary institutions. The Dutch education system, similarly, features local mainstream schools and regional special schools.

Treatment of children with CP within the Dutch health care system, educational system and related services has until now not been comprehensively described from a representative cohort. Previous Dutch publications on children with CP relied on rehabilitation-based cohorts, e.g. by Voorman et al. [4, 5]. Internationally, surveys on these two aspects in relation to each other and to CP characteristics are scarce as well. In the United Kingdom, a population-based survey by Parkes et al. on allied health care showed that almost all children (with moderate to severe CP) were involved in some form of allied health care treatment , but the availability of these services was found to be limited apart from situations when the child followed special education [6].

In the Netherlands co-operation of multi-disciplinary (rehabilitation) teams with special schools is well-established. Several aspects of this cooperation have been the object in Dutch rehabilitation-based studies, both from the point of view of teamwork, the team communication process itself, and from the parent's or family point of view [7-9].

In addition to a comprehensive description of the current utilization of health care and educational services, we assessed the children's history regarding these items in special or mainstream facilities. Furthermore, to get insight whether the provided care and educational services were typical for subgroups of children with CP the associations with child and CP-related characteristics are explored as well as the parents' general opinion on services delivered for their child.

Methods

Subjects

The present study is part of a cross-sectional population-based study on prevalence, clinical presentation, impairments and activity limitations of children with CP in the

Netherlands [2, 10, 11]. CP was defined as a disorder of movement and posture caused by a non-progressive brain lesion with an onset no later than one year after birth [12]. Evident neurological symptoms (e.g., spasticity, dyskinesia or ataxia) were obligatory. Patients were included if they had (a) a diagnosis of 'cerebral palsy' recorded in their medical files, (b) date of birth between 1 January 1977 and 31 December 1988, and (c) parents living in Gelderland, a region in the east-central part of the Netherlands – at the time of the study. Informed consent from the children's parents and approval by relevant ethical committees were obtained.

Data collection

One of two experienced rehabilitation physicians visited the parents and child. Apart from a physical examination a structured interview on the child's comprehensive medical history and educational career was performed. Basic characteristics of the child were registered (i.e. sex, age). Clinical sub-types of CP were classified in three categories (a) unilateral spastic CP, (b) bilateral spastic CP, and (c) non-spastic CP. Intellectual functioning was classified as (a) normal, (b) learning difficulty, or (c) mental retardation. Classifications for both clinical distribution and intellectual functioning followed the Swedish and German population-based studies [13, 14] as adopted within the Surveillance of Cerebral Palsy in Europe group [15]. Gross motor functioning was retrospectively classified according to the Gross Motor Functioning Classification System (GMFCS), level 1 describing the least and level 5 the most affected children. [16].

The setting in which the child's CP-related medical history started was surveyed ((a) through primary health care, (b) through specialist medical care)). Undergone surgery, distinguishing (a) CP-related orthopaedic procedures, (b) strabismus surgery and (c) other was recorded. The orthopaedic group comprised as main sub-groups hip surgery (adductor tenotomies or osteotomies) and ankle-foot surgery (Achilles Lengthening Procedures) and sporadic hand or spine surgery. Other types of surgery were "miscellaneous" (e.g. ear-nose-throat procedures, inguinal hernioplasty, orchidopexy).

The different medical specialists by whom the child had been seen in the past twelve months were recorded. In addition, the current use of medication was recorded ((a) no medication, or (b) anti-epileptic medication, (c) laxatives, (d) miscellaneous).

Allied health care services at the time of interview were divided in four main categories: (a) no treatment, (b) mono-disciplinary treatment, (c) multidisciplinary treatment in rehabilitation centre, and (d) multidisciplinary treatment in institutions

for mentally retarded children. Box 6.1 describes these allied health care modalities in more detail.

The child's history through allied health care until the current situation was recorded. The number of distinct allied health care services in which the child had ever been treated was summed (0–6).

Finally, the current utilization of "orthotic" devices (e.g. shoes, braces) and of devices for walking and mobility (e.g. walker, wheelchair, three wheel bicycle) was recorded. For each of these groups of devices a sum-score was computed.

Education

The fact that a child made a start at primary school level was considered the inclusion point both for mainstream or special education. We distinguished (a) primary education, (b) secondary education and (c) no school. "No school" was defined by pre-school activities (e.g., crèche, therapeutic toddler group) or a day care facility if the child never enrolled in any school in cases of severe mental retardation. Furthermore we differentiated between (a) mainstream education and (b) special education (see Box 6.2). The child's entire school career was also assessed ((a) mainstream only, (b) special only, (c) switched from mainstream to special, and (d) switched from special to mainstream).

At the end of the interview we asked the parents' opinion on to what extent therapeutic and education possibilities for their child to had been utilized (rating from (1) maximally – (5) not at all). Furthermore they were asked whether they had visited alternative healers with the child ((0) no, (1) yes) and if they were member of an organisation for physically disabled people and their parents ((0) no, (1) yes).

Box 6.1 Allied healthcare services

Allied healthcare service in the Netherlands has several forms. A major distinction can be made between mono-disciplinary and multidisciplinary forms.

Monodisciplinary therapies usually refer to physiotherapy, sometimes speech therapy provided in a community-based practice or in a local hospital. Occasionally monodisciplinary care addresses hydrotherapy, or occupational therapy.

Multidisciplinary care in a team approach supervised by a consultant in rehabilitation medicine (with all options such as physiotherapy, occupational therapy, speech therapy, social work, child psychology, pedagogy and assistive technology) is provided in a regional rehabilitation centre or department.

Within a residential setting or day care facility for the mentally retarded allied health treatment with a multidisciplinary team approach usually is also present.

Box 6.2 School system in the Netherlands – mainstream schools and special schools

Mainstream: local schools for all local children, age 4 to 12 for primary education. For children from age twelve secondary mainstream education has school types at three academic levels, the lowest emphasizing practical and vocational skills, the highest leading to university admission.

Special education: schools for children with both a specific diagnosis and specific educational needs from age 4 upward. Traditionally, the Netherlands have a wide variety of special schools. There are specific schools for children with motor disabilities (often combined with learning difficulty or mental retardation); also there are special schools for children with "only" severe learning difficulty, for children with severe behaviour problems and for children with sensory deficits (low-hearing or deaf, low-vision or blind).

Children with identified disabilities are entitled to go to the mainstream schools if parents prefer so. In that case educational support toward the individual student and the mainstream school is paid for by the government and provided by experts from special schools. This child-dedicated budget is called the "backpack" and intends to keep the child in the local school with its peers. So there can be a parents' choice to let the child be "special" in a normal (mainstream) school or "normal" in a special school.

Day care facility: Children with severe mental retardation and incapability of any classroom learning activities visit child day centres. Adolescents requiring day activities rather than a form of work can make the transition either from a special school or a child day centre to a day centre for adults. Day centres for children that never went to school and day centres for (young adults) are both part of institutions for the mentally retarded.

Analyses

The sample was broken down in two age groups (i.e. 6–12 years and 13–19 years). In the Netherlands this age division usually marks primary or secondary education. Differences between the two age-groups regarding sex and CP characteristics (GMFCS, limb distribution and intellectual functioning) were explored with Chi-square tests. Correlations were explored with Spearman correlation coefficient. In order to adjust for level of gross motor functioning, a partial correlation was computed between the utilization of allied health care and education, controlling for GMFCS level.

Results

Subjects

Table 6.1 describes the cohort of 127 children with CP, divided in age-groups of 6–12 and 13–19 years. Sixty-two percent of them were boys. Their mean age was

Table 6.1 Characteristics of the cohort; children with CP (N = 127)

Variable	6–12 years^ (N = 87)	13–19 years^ (N = 40)	Total^ (N = 127)
Child characteristics			
Sex (male) (%)	54 (62)	24 (60)	78 (61)
Age in years (mean (SD))	9.0 (1.8)	15.6 (1.9)	11.1 (3.6)
CP characteristics (%)			
Limb distribution			
Unilateral spastic CP	33 (38)	15 (38)	48 (38)
Bilateral spastic CP	48 (55)	23 (58)	71 (56)
Non-spastic	6 (7)	2 (5)	8 (6)
GMFCS			
I	23 (26)	9 (23)	32 (25)
II	33 (38)	16 (40)	49 (39)
III	7 (8)	5 (13)	12 (9)
IV	6 (7)	4 (10)	10 (8)
V	18 (21)	6 (15)	24 (19)
Intellectual functioning			
Normal	32 (37)	12 (30)	44 (35)
Learning difficulty	22 (25)	12 (30)	34 (27)
Mental retardation	33 (38)	16 (40)	49 (38)

^ rounded percentages.

11.1 years (SD = 3.6). Thirty-eight per cent had unilateral spastic CP, 56% bilateral spastic CP and 6% had a non-spastic form of CP. Almost two-third of the children walked independently (GMFCS-levels I–II: 64%). About two-third of the children had either normal intellectual functioning or learning difficulty, leaving 38% with mental retardation. Both age groups did not differ significantly in regard to these characteristics.

Specialist medical care, surgical interventions, medication

Eighty-five children (67%) had their first contact with the health care system via a medical specialist, usually the paediatrician. The other 42 children (33%) were referred via primary health care.

Sixty-one children (48%) had orthopaedic or strabismus surgery performed at any time (of whom 20 children the same procedure twice). Table 6.2 shows major groups of surgery performed in the children's history, by GMFCS level. Achilles tendon lengthening procedures totalled 27 in 25 children (i.e. 2 children two times), hip surgery was performed 39 times in 29 children (i.e. 10 children two times). Hip

Table 6.2 CP- related surgical procedures

GMFCS level	N children	Achilles tendon lengthening	Hip surgery	Other orthopaedic	Strabismus
I	32	7(1)	0	1 (hand)	4 (1)
II	49	13	7 (1)		11 (1)
III	12	5(1)	3 (1)		4 (1)
IV	10	0	9 (3)		4 (1)
V	24	2	20 (6)	2 (spine)	4 (2)
Total	127	27	39	3	27

Numbers are historic surgical procedures (in parenthesis the number of children that had a similar operation two times).

surgery correlated with more impaired gross motor functioning (r = .51, p < .001) and had not been performed in any child with unilateral spastic CP. Achilles tendon lengthening procedures did not correlate to GMFCS level. Sporadically CP-related surgery on the hand (one child with unilateral spastic CP) and the spine (2 children both with bilateral spastic CP, GMFCS V) had been performed.

Furthermore 21 children (18%) had undergone strabismus surgery (27 procedures, in 6 children two times), which was not related to GMFCS level.

Among the "other surgery" category ear-nose-throat surgery (tonsillectomy and tympanoplasty) was frequent (25 procedures in 22 children), other types of surgery only sporadic (such as inguinal hernia, orchidopexy).

Current specialist medical health care

Table 6.3 shows medical specialists that had seen the child in the past twelve months before the examination. Paediatricians and orthopaedic surgeons were significantly more involved with more affected children (high GMFCS levels). Consulting a paediatrician significantly related to the younger age-group (r = .25, p = .004). The great majority of children (80%) were seen on a regular basis by the consultant in rehabilitation medicine. Children consulted a median of 1 specialist (range 0–4).

Children who consulted more specialists had higher GMFCS-levels (r = .35, p < .001); age was not related to the number of specialists visited.

Medication

Of the 127 children, 35 (27%) currently used any medication. Thirty of these 35 children used anti-epileptic medication, four of them had also other medication. As anti-epileptic drug carbamazepine was used in 14 cases and valproate in 13 cases

Table 6.3 Current specialist health care and allied health care treatment – by GMFCS

	GMFCS						r^
Medical specialist	Total N = 127	I N = 32	II N = 49	III N = 12	IV N = 10	V N = 24	
Paediatrician	34	4	13	3	3	11	.23 (p = .009)
Child neurologist	31	6	9	5	1	10	---
Rehabilitation physician	105	26	37	11	10	21	---
Orthopaedic surgeon	40	6	12	5	6	11	.25 (p = .004)
Allied health treatment	Total N = 127	I N = 32	II N = 49	III N = 12	IV N = 10	V N = 24	
No treatment	22	11	9	1	0	1	
Monodisciplinary	26	8	10	2	1	5	
Multidisciplinary RC	58	12	23	7	8	8	
Multidisciplinary MR	21	1	7	2	1	10	

Current specialist health care refers to medical specialists consulted –CP-related– in the past year, allied health care to the treatment received at the time of examination.
^, Spearman correlation; ---, not significant.
Monodisciplinary, community based or local hospital; Multidisciplinary RC, multidisciplinary treatment in rehabilitation centre; Multidisciplinary MR, multidisciplinary treatment in institution for mentally retarded children.

(3 children used both), in one child epilepsy was treated with homeopathic drugs. Seven children, all with GMFCS levels IV–V, used laxative drugs. No child used drugs to diminish spasticity (e.g. baclofen or tizanidin), although in some cases these drugs were reported to have been tried earlier on. Other medication taken was sporadic and miscellaneous; this included asthma drugs, antibiotics and homeopathic drugs.

Allied health care

All children had been treated, and most were still treated in some type of allied health care. The second part of Table 6.3 shows the different types of allied health care that the children were treated in at the time of interview, in relation to GMFCS. Almost half of the children (58 children (46%)) were treated in a rehabilitation centre by a multi-disciplinary team; 21 children (17%) had multidisciplinary treatment in an institution for children with mental retardation and 26 children (20%) had mono-disciplinary treatment. Seventeen percent was currently not treated (any more) in a form of allied health care. Younger age, more severely affected gross motor functioning, and impaired intellectual functioning were seen in more intensive treatment modalities (Table 6.4).

Table 6.4 Orthotic and mobility devices by GMFCS-level (N)

		GMFCS					*r*^
	Total N = 127	I N = 32	II N = 49	III N = 12	IV N = 10	V N = 24	
Orthotic devices							
Orthopaedic shoes	55	11	25	8	6	5	---
Leg orthosis	18	4	4	1	3	6	---
Night splints	31	9	14	2	1	5	---
Lying brace	9	–	1	–	1	7	.34 ($p < 0.001$)
Sitting brace	3	–	–	–	–	3	X
Standing brace	20	–	4	4	3	9	.39 ($p < 0.001$)
Other	24	6	8	5	2	3	---
Mobility devices							
Walking devices	13	–	4	9	–	–	X
Mobility devices							
Manual wheelchair	45	2	15	9	7	12	.45 ($p < 0.001$)
Three-wheel bicycle	31	4	19	5	2	1	---
Electric wheelchair	10	–	1	1	1	7	X
Pushing cart	6	–	1	–	–	5	X
Other	8	1	4	1	1	1	X

^, Spearman correlation; X, correlation not computed; ---, not significant.

Orthotic devices, walking aids and mobility devices

Hundred children (79%) currently used one or more type of orthotic device (Table 6.4). Most children had one orthotic device; 13 children (10%) had 3 or 4 types of orthotic devices. A total of 13 children (10%) used a form of walking aid (in 12 cases a walker) . Mobility devices were used frequently. Sixty-five children (51%) used at least one type of mobility device and 35 children (27%) used an additional device. Typically a three-wheel bicycle or a powered wheelchair were combined with a self-propelled or pushed wheelchair.

Compared to children with mono-disciplinary treatment, children treated in a rehabilitation centre or in an institution for the mentally retarded had been in contact with a broader variety of distinct allied health care professionals ($r = .22$, $p = .012$), with more health care specialists in the past year ($r = .30$, $p = .001$) and used more frequently one or more mobility aids ($r = .23$, $p = .009$).

Education

Table 6.5 presents the types of education in the two age-groups. Just over one-quarter (27%) of the children attended a mainstream school. The largest single subgroup however (48%) attended a special school for children with physical disabilities (or physical and learning disabilities), 9% went to other types of special education (e.g. a school for children with severe learning difficulty). Sixteen percent did not attend school but a day care facility.

Thirty children (24%) attended only mainstream schools in their entire school career, 62 children (49%) only special schools; no more than 14 children (11%) switched during their school career, of which 10 children started in "mainstream" and switched to "special". Attending mainstream schools was associated with better gross motor function (r = .58, p < .001), lesser limb involvement (r = .51, p < .001) and normal intellectual level (r = .73, p < .001) (Figure 6.1). Furthermore attending a mainstream school had a strong relation with either not being treated (any more) in allied health care or in mono-disciplinary treatment (r = .49, p < .001) (Figure 6.1). The partial correlation between types of allied health care and school type, corrected for GMFCS, was still significant (r = .34, p < .001).

Parents' opinion

Seventy-nine percent of the parents judged that therapeutic and educational possibilities for their child had been utilized maximally, another 13% regarded this as being predominantly the case. About one quarter (24%) had visited alternative healers

Table 6.5 Current school type (N (%))

School type	Total N = 127	Age 6–12 N = 87	Age 13–19 N = 40
Mainstream			
Primary	20 (16)	20 (23)*	–
Secondary	14 (11)	4 (5)	10 (25)
Special education			
Primary school for children with physical disabilities and/or learning disabilities	47 (37)	39 (45)	8 (20)
Secondary school for children with physical disabilities and/or learning disabilities	14 (11)	1 (1)	13 (33)
Other special school	11 (9)	7 (8)	4 (10)
No school – day care	21 (16)	16 (18)	5 (12)

* Of whom four children with ambulant counselling in mainstream school.

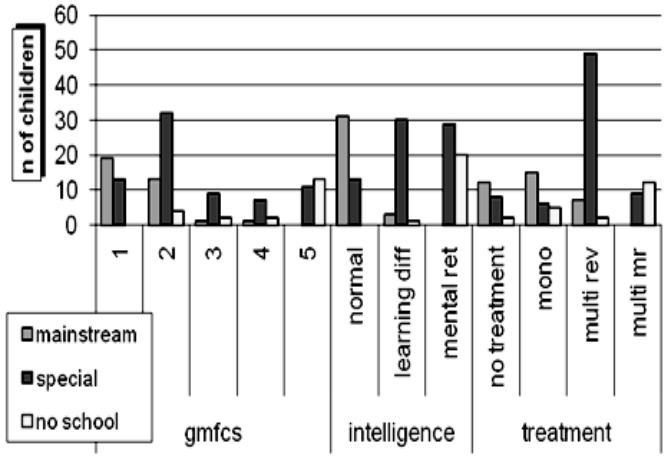

Figure 6.1 School type by CP characteristics and treatment.

with the child at least once. These parents showed lower scores regarding to the extent the care provided was considered optimal ($r = .28$, $p = .002$).

Twenty-seven percent of the parents was a member of an organisation for physically disabled people and their parents – most of them were parents of children with high GMFCS levels ($r = .22$, $p = .015$).

Discussion

Children with cerebral palsy need a variety of services regarding treatment and educational facilities. There is a range from little or no treatment and mainstream schooling to intensive programmes combining major interventions, comprehensive allied health treatment, special education and assistive technology. As services and their availability are obviously influenced by national policies and historic developments, they may differ from country to country.

In the United Kingdom, Parkes et al. described allied health services utilized in a population-based CP study; they emphasized that it is useful to know both the proportion of children treated (and where and how) as the proportion not treated [6, 17]. CP is broader than the need for forms of treatment.

Several rehabilitation-based Dutch studies focused on how professionals working in (special) education and multidisciplinary rehabilitation teams cooperate, as seen

from the parents' point of view or looking at the cooperation and team communication process as such [8, 9, 18]. The present study gives a representative description of how and where Dutch children with CP get the services needed.

The cohort's characteristics are comparable to those of pooled population-based studies such as SCPE [1].

Specialist medical care, past surgery and medication

If a child's medical history started within specialist medical care (two thirds of the cases), this was typically in neonatal care, supervised by the paediatrician. In a minority of cases developmental problems made the general practitioner initiate specialist medical diagnostics.

In time rehabilitation physicians become the coordinating medical specialist for CP, as they see 80% of the children of all ages on a regular basis. Other medical specialists participate to a lesser degree, neurologists usually in children with epilepsy and orthopaedic surgeons in cases of surgery. The dominant role of the rehabilitation physician can probably be seen as a typical Dutch practice. For example, in the United Kingdom the paediatrician and orthopaedic surgeon are most involved as they saw the children in 60% and 45% of cases within the last six months respectively [6].

Surgery in this cohort showed three large groups, two types of orthopaedic surgery and one of strabismus surgery. Achilles tendon lengthening procedures were seen mostly in children with (any) walking abilities, however not significant as that group is most numerous anyway. Hip surgery, aimed at preventing or reducing hip deformity and applied 39 times in 29 children, has a significant relationship with high GMFCS levels. The relationship between severe forms of CP and prevalence of hip deformity and/or dislocation is well known [19, 20]. The low number of hip dislocations in the Netherlands [11] may have a relation with the fact that more than half of the children with severe CP have preventive or corrective hip surgery along the way. In the Netherlands hip monitoring and for preventive hip surgery are well established as part as the integral care for children with CP. This has been confirmed recently in Dutch Treatment guidelines for children with Spastic CP [21]. Hagglund et al. described how implementation of a hip monitoring protocol in Sweden resulted in reduced prevalence of hip dislocations [22].

Strabismus surgery was frequently reported (15% of the children). Prevalence of strabismus in all Dutch children is estimated to be 3 to 5% in the general population [23]. Taking into account that the number of children actually operated for strabismus is obviously lower than that of those with strabismus, it may be inferred from the

number of children operated for strabismus found in this cohort that the prevalence of strabismus in children with CP is at the least three times higher than in all children. The strong relationship of strabismus with brain disorders (of any kind) is reported earlier, e.g. in cases of severe CP or mental retardation [24-26].

Except for anti-epileptic medication and laxative drugs other medication taken was sporadic, miscellaneous and assume to be comparable to what is taken in a general child population. Notably no anti-spasticity medication was taken by any child at the time of interview, although in some children these drugs had been tried but not continued. Side effects were noted, the absence of functional benefit prevailed or even the perception that diminishing the spastic component of the motor disorder actually weakened the child.

Some disadvantages of oral baclofen may be avoided by administration as intrathecal baclofen (ITB) – which was not available as an intervention in the time of the interviews.

Recent options in medical treatment of CP

Some remarks have to be made on evolving medical interventions since the time of the survey.

Intrathecal Baclofen (ITB) has become an available option for treating generalized spasticity [27, 28] but remains a major intervention that needs to be tried and monitored in carefully selected cases. For Selective Dorsal Rhizotomy (SDR) experience is building up in recent years. The indication for SDR in the Netherlands is concentrated by national agreement at one specialized team [27-29]. With SDR a comparable painstaking pre-procedure assessment as with ITB has to be gone through. Multi-level single-event (MLSE) orthopaedic surgery (as opposed to orthopaedic surgery around one major joint) and multi-level injection of botulinum-toxin A are in wider use since the late nineties, often preceded by adjuvant diagnostics such as gait analysis. Dutch reports and case series are available on these interventions [30-32]. However, as yet no Dutch population-based data on actual use of ITB, SDR and MLSE surgery are available.

Assistive devices

A large-scale use of orthotic devices is found, especially of orthopaedic shoes. In the Netherlands a popular option for children is the "semi-orthopaedic" shoe, factory-made but allowing an adequate individual insole or orthosis to fit in the shoe together with the (deformed) foot.

Mobility devices were seen, not surprisingly, most frequently in high GMFCS levels, this was also reported by Ostensjø et al. [33]. In sporadic cases of use of a wheelchair or three-wheel bicycle in children with GMFCS I co-morbidity such as mental retardation influenced the need for a device that from a motor function viewpoint probably was not needed.

Types of allied health care

The high proportion of Dutch children with CP treated in a multidisciplinary approach in a rehabilitation centre probably has a historic background. In the Netherlands rehabilitation of children with motor and other deficits started in the first half of the twentieth century. Large inpatient institutions that provided nursing care (and orthopaedic surgery) for children with physical disabilities developed into modern child rehabilitation centres with specialized professionals of medical, allied health, social, psychological and technical disciplines. These residential institutions' schools for children with motor (and other) disabilities were the basis of school systems that still serve many children. Close co-operation and co-habitation of these schools with the child rehabilitation centres is still present.

In the Netherlands only a quarter of children with CP (27%) attends mainstream schooling. This group has a significant higher proportion of low GMFCS level and normal intellectual level compared to the children in special education. They are treated with monodisciplinary or no allied health care.

In more recent years the Dutch education policy has been stimulated by the UNESCO-treaty of Salamanca [34] towards inclusive education by providing extra assistance in the mainstream school situation. The number of children attending special schools as such however did not decline, but the number of children in mainstream schools with dedicated budgets grew strongly [35]. CP-specific information on this aspect is lacking.

Parents' opinion

Looking back on the utilization of all chances available in the health care and rehabilitation services delivered, the parents were to a very large extent satisfied or predominantly satisfied. Among those parents that were less satisfied a significant larger group had been consulting alternative healers such as clairvoyants and magnetic healers. The proportion of parents that at any time sought alternative treatment seems comparable to the 25% Parkes et al. found in the United Kingdom [6] but it must be

noted that in the UK study alternative treatment was described as any treatment other than regular treatment as available by National Health Service.

Practical impact of having a child with CP for a family

Children with CP and their caretakers meet many practical consequences of the condition. Having to visit medical and allied health professionals for child and parents either at intervals or on a regular basis, to use technical devices that have to maintained and renewed, daily trips to a special school or to specific extra activities within the mainstream school defines the practical impact, or care additional to the care considered normal for the child's peers. This additional care cannot easily be calculated by adding up the separate elements. It becomes part of family life, may even be taken for granted. Without having assessed the emotional burden by CP on families that must not be ignored [36] or having calculated the toll paid in terms of hours' work or money by families (or society) this paper is meant to shed a light on obvious extras that the family has to be deal with – in relation with CP characteristics, as there is no standard CP child.

Additional care – compared to the care for typically developing children – is recently getting the attention it deserves. A "capacity" profile CAP has been developed and evaluated in the Netherlands as a generic tool to help professionals inform and counsel parents in an early phase on the future perspective of their child's needs in all fields [37, 38].

The study shows the variety of educational options and the options for modalities of allied health care. The subgroup of children with normal intelligence, better motor performance and (mostly) unilateral spastic CP frequently attends mainstream school and receive mono-disciplinary allied health care or none. On the other end of the severity spectrum a group of children with mental retardation, half of them with GMFCS level V attends day care facilities. The largest group is in-between, attending schools for children with primarily physical disabilities. These schools host the variety of GMFCS levels and intellectual levels and many of the children have multidisciplinary rehabilitation treatment care within the school context.

In conclusion, a representative group of Dutch children with CP is surveyed regarding their path through specialist and allied health care and education. While Dutch children with CP as a group are comparable to other European population-based cohorts, the Dutch health care and education system bear the traces of the historic child rehabilitation facilities and its interaction with a well-developed special education system. Furthermore the medical approach in the Netherlands,

with a central role of rehabilitation medicine specialists, promotes using no more medication than necessary but an active policy in using orthotics, assistive technology and orthopaedic surgery.

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7

General discussion

Background

Cerebral palsy (CP) is the most prevalent cause of primary motor disease in children in the world. Children with CP make strong demands on services, treatment and technical adaptations [1]. They are a large proportion of the children requiring treatment in child rehabilitation departments.

The umbrella of the CP notion [2, 3] by any definition covers a range of clinical manifestations, caused by a range of non-progressive laesions, all influencing the immature brain. Knowing the prevalence of CP in a population is very relevant, but prevalence rates give no information on the distribution and severity of impairments, associated disorders and limitation of activities and participation in the entire group. Of course distinct clinical or functional features ask for distinct rehabilitation approaches, or services in general.

The Dutch study, initiated by Utrecht child neurologist Onno van Nieuwenhuizen adopted the unchanged basis of its study protocol from the well-known cooperative German-Swedish studies by Krägeloh-Mann and Hagberg [4-6]. Scoring of clinical items and associated features was done by their criteria and the German and Swedish researchers were personally involved in training the clinicians that did the Dutch field work. However as the Dutch study was set up from a rehabilitation medical specialist's point of view as well, we gathered additional details on activities and participation, on treatment, day-time situation, and psychosocial aspects.

Rehabilitation Medicine in the Netherlands, being a distinct medical specialism takes a position where cure and care meet. The concept of impairment, disability and handicap and looking at all three aspects of disease has been the basis of rehabilitation medicine [7]. Worldwide these aspects were described in 1980 in the International Classification of Impairments, Disabilities and Handicaps ICIDH [8]. ICIDH's successor, the International Classification of Functioning, Disability and Health ICF by the World Health Organization in 2001 recognizes sequelae of disease, trauma or congenital disorder at the level of impairment of body structure or body function, activity limations and restriction of participation [9]. An important new aspect in ICF was the description of personal and environmental factors.

The aim of the study

The study intended to give a broad picture of the general situation of Dutch children with CP. How prevalent are children with CP, what is the distribution and inter-relationship of clinical features and associated disorders, what are impairments of body

function and structure, what are activity limitations. Are trends present in studied items? Where and by whom children with CP are treated, what interventions do they undergo and where do they go to school? Are behaviour and communication problems recognizably related to manifestations of CP?

In ICF terms attention had to be paid to "impairments of body function and structure" as well as to "activity limitations" and "participation". Treatment modalities can be seen as environmental factors and the personal factors are self-evident.

The cornerstone work in this survey is the calculation of prevalence of CP, as described in the first paper, published in the European Journal of Epidemiology, Chapter 2 of this thesis [10].

Dutch figures on CP prevalence were scarce, only in the PhD thesis of De Vries [11] mention is made of several regional studies in the fifties and sixties without details on these studies. In 1982 the Phelps Foundation for the Spastics [12] published results of a telephone survey via special schools to estimated the CP prevalence and calculated 1.5 per thousand children.

So Dutch representative figures and details on prevalence of CP and associated aspects were not present on the birth years through the eighties. As prevalence counts by birth year of a condition as CP can only be published years later, the trend of the eighties as studied in other countries in Europe and the USA [13-15] were indicative of a strong rise in CP prevalence. This prevalence rise was thought to be related to higher survival rates of dysmature and/or premature babies, due to improved perinatal and neonatal care.

This rise in CP prevalence as seen in other countries and the unknown proportion of children with severe CP, the children who need relatively more services, made it relevant to perform the Dutch population-based study.

Results discussed, chapter by chapter

Some extra attention will be given to significant findings from chapters 2 through 6. If elapsed time makes it necessary to expand on possible developments later on, more than we did in that paper's own discussion section, we will discuss these developments. In the end of this general discussion some recommendations for future research or possible policy regarding CP epidemiology are made.

In Chapter 2 we reported a rise in CP-prevalence to 2.44 per thousand live births by 1986–1988, which rise was comparable to trends reported elsewhere [16]. Has the rising trend in the Netherlands continued after birth year 1988? Very recently, in 2011, Van Haastert et al. reported on a strong decline of both the rate and severity of

CP in a follow-up cohort of 3000 ex-premature Dutch children born between 1990 and 2005 [17]. This is very interesting, however not a CP prevalence count at the population level.

More recent CP prevalence data come from other countries. Colver et al. described rising CP-prevalentie in 2000 – across the severity spectrum covering a long time span – looking back at birth years 1964–1993, rates rose 1.68 per 1000 neonatal survivors during 1964–1968 to 2.45 during 1989–1993 [16].

In Europe the paramount CP-epidemiology group is Surveillance of Cerebral Palsy in Europe SCPE, a cooperative project of CP-registers and some CP-surveys in many European countries [18, 19]. Our study participated in the first tranche of SCPE from 1998–2001. The partners of SCPE share inclusion criteria and deliver representative data for their area [20].

New registers qualified to join SCPE are rare. One new register, from Norway, reported in 2008 on close to 300 Norwegian children from birth years 1996–1998 and found a prevalence of 2.1 per thousand [21]. An other register from Southern Sweden reported on children born between 1990 and 1997 and the prevalence was 2.4 per thousand [22].

In conclusion regarding CP prevalence: looking at reported prevalence "elsewhere" of CP after 1990, reported rates indicate that the prevalence rise came to an end at rates at or above 2 per thousand but also that so that decline of prevalence has as yet not been proven.

One striking finding in the first paper is the large proportion of children (one quarter) that in their very early years were initially diagnosed with CP but upon personal examination in our study had something else than CP, like mental retardation as such, or developmental coordination disorder and even one child that eventually was re-diagnosed with HMSN. The standard age to confirm a child as having CP was discussed intensively within SCPE and, although in young children with obvious spasticity CP may be allowed as a "working diagnosis" a re-check by a pediatrican or qualified medical specialist is advised by the age of at least five [18, 19].

In Chapter 3 we reported on the distribution of clinical sub-types and major associated disorders [23].

Main findings were: Spastic subtypes accounted for over 90 percent of all CP cases: bilateral spastic CP as a group are the majority although spastic unilateral spastic CP is percentage-wise the largest individual clinical subtype. Epilepsy (40 percent, active and "ever" combined) and mental retardation (almost 40 percent) are common. Clinical patterns and associated disorders remained rather constant comparing earlier to more recent birth years.

As we consider whether the distribution of motor types in the population might have changed after birth year 1988 we again need data from abroad. An Australian population-based study by Reid in 2010 reports that in over 3000 children with CP, born between 1970 and 2003 in Victoria, predominant motor types were spasticity in over 90 percent of cases, ataxia in 5 and dyskinetic disorders in 4 percent. No specific trends in this regard were reported [24]. Similar findings were reported by Howard in 2005 [25] and both reports are similar to what we found in our study [23] and we conclude that probably the overall distribution of motor types within the CP group has changed little. As it is, current clinical practice in rehabilitation medicine still presents us with a vast majority of spastic children among the CP patients.

The rate of epilepsy that we found was around 20 percent "active" and an additional 20 percent "ever", notably in a group with children ages 6 through 19 (those with epilepsy "ever" were previously but not any more treated for seizures). In a Japanese study by Suzuki et al. in 2009 42 percent had epilepsy at age six in a group of 569 children from birth years 1977–2000 [26]. This group may be too young for the "ever" category so the epilepsy rate still seems comparable to ours (active and ever combined).

In Chapter 4 we described motor impairments and activity limitations in relation to types of spastic CP in the birth years 1977–1988. As the non-spastic group was too small for statistic power (4 ataxic and 4 dyskinetic out of 127 children) in this paper analyses could be done purely for the spastic group, 119 children.

Children with spastic CP had a lower body height and weight compared with typically developing peers. Forty percent had no range of motion deficits. Hip dislocations were rarely encountered and only in cases with bilateral spasticity. Motor impairments were associated with gross motor functioning and manual ability levels. Close to sixty-five percent were independent walkers (GMFCS I and II). Children with diplegia and tetraplegia differed in activity limitations. Motor impairments and limitations in mobility and self-care activities were only modestly related in multivariate analyses.

Distribution of CP-related characteristics regarding motor impairment and activity limitation is consistent with that found in representative studies of other countries, apart from the lower rate of hip dislocation in The Netherlands. The distinction between leg-dominated spastic cebrebral palsy (diplegia) and four-limb dominated spastic CP (tetraplegia) is relevant from an activity point of view, in spite of other authors who advocate recognizing the bilateral spastic group as a whole while focusing on GMFCS as the child's description [27]. The child's activity limitations are not a mirror of the motor impairments, which suggests multifactorial influences (here personal and environmental factors probably come in). We concluded

that an activity-oriented rehabilitation approach goes beyond treating specific impairments.

The few cases of hip dislocation were confined to bilateral spastic CP with GMFCS level IV or V, never in unilateral CP. This finding could be related in chapter 6 to the Dutch pro-active attitude towards preventive hip surgery.

There is little chance the distinct clinical manifestations of CP will in later birth years produce a changing set of impairments. Furthermore the activity limitation will continue to be influenced by more factors than the impairments per se.

Chapter 5 reports on communication, behaviour and cognitive aspects and shows that about one third of children wit CP suffered from behaviour problems in some form. Limited intellectual functioning was the most important associative factor, while limited gross motor functioning affected some domains. Overall the most prevalent problems in this field (greater than 25%) appeared to be communication skills (self expression and writing), general cognitive functioning, lack of confidence, emotional instability and being dependent. Children without intellectual limitations seem to function without significant problems.

Our findings on the rate of behaviour problems are in contrast with reported relatively high proportions of behaviour problems in English children with unilateral CP [31, 32]. The children with unilateral spastic CP are actually the group that as a rule has better intellectual possibilities. We have no explanation as to what causes this difference with the English group.

Chapter 6 gives an overview of treatment, interventions and education. The course of the history of this group bears the traces of the Dutch tradition in child rehabilitation institutions in combination with special education as places where children with CP are treated and go to school. Less affected children, especially those with normal intelligence, predominantly use mainstream facilities.

Eighty percent of the children of all age groups had been in contact with a rehabilitation physician within the last twelve months. This illustrates the central position of Dutch specialist rehabiliation medicine regarding the integral care for children with CP.

The practice of medical care, medication and operations may have changed on some specific topics. Interventions such use of multi-level botulinum toxin-A injections or multiple level – single event orthopedic surgery, intrathecal baclofen (ITB) and selective dorsal rhizotomy (SDR) have either started or increased in numbers since the mid-nineties [28-30]. However Dutch "numbers" for the ITB and SDR performed untill now are still limited. These interventions aimed at reducing spasticity have their indications but also contra-indications. Scrutinous pre-intervention screening is important. If weakness or limited range of motion contribute

more to the activity limitation than spasticity as such does, reduced or eliminated spasticity after a major intervention will not enhance, may even impede motor performance.

By January 2011 some fifty SDR procedures on Dutch children had been performed (personal communication Van Schie, VUMC Amsterdam, January 2011).

The large proportion of children with severe bilateral spastic CP that had hip surgery at any time indicates in our opinion how these children are monitored structurally – by the rehabilitation physician – and introduced to the orthopedic surgeon at a time early enough to preserve a non-dislocated hip joint.

Limitations of the study

Research by doing a population-based cross-sectional survey is by nature observational, it is not an experiment or an evaluation of interventions. This is not a limitation in itself, but a matter of fact.

Time has elapsed since the field work (fourteen years) and papers with data from this survey were published at relatively large intervals. The delay could be a problem. Nevertheless we think it is still worthwhile to presents the survey's results once again as a total. After all there has been no Dutch population-based follow-up since. Where current practice might give other information (if the survey were to be performed now) this matter needs discussion as we did in the chapters and in this General Discussion. Apart from "new" major interventions such as ITB and SDR that are still relatively infrequent in the Netherlands but can be counted by the few centres that perform them, we are uninformed on current rates of multi-level operations or injections with botulinum toxin-A. We know these interventions, often preceded by instrumented movement analysis, have gained wide application but their numbers are not known so reports on these interventions are selected case series.

An other limitation of the study is that, at the time of the field work, classification systems now adopted worldwide were "under construction" such as Gross Motor Function Classification System [31]. We used a clinical, qualitative approach rather than an approach of fine or instrumented measurements. Children were described accurately regarding their motor performance, allowing us to classify GMFCS later on. We classified according to the German-Swedish (non-validated) items on the examination form. For an epidemiological survey, classifying impairment or activity limitation at the level of "clearly present" or "not or minimal" suffices for a general picture of the child. This asks for good clinical judgment, provided in our study by individual examination by rehabilitation physicians.

As one example: range of motion (ROM) tests were done with the child situated on a bed, couch or floor mat at the child's home. Possible scores upon examination of ROM limitation were: none, slight, definite and severe. To stick to this example, in order to make no disputable distinctions in the final report (Chapter 3) the categories none and slight ROM limitations were combined and only definite and severe ROM limitations were reported in the paper. This may provide rates of ROM limitations that are somewhat too low by missing the "slight" category, but we at least know that the impairments reported are obvious.

Each child was personally examined by one of two experienced consultants in child rehabilitation medicine. This differs from several other population-based studies (mostly registers) that use third-party reporting. There the keeper of the register depends on the expert judgment of the field worker that fills in the Register's case form. The intention of the partners of Surveillance of Cerebral Palsy in Europe to warrant population-based data not only to be complete but also accurate and comparable led to the production of a Reference and Training Manual on CDROM for workers in the field reporting to regional registers to diminish the above mentioned problem.

General recommendations

First: a young child diagnosed with CP, should be re-checked by looking again at the clinical manifestation at age five or six.

Second: CP is not life-threatening condition, children become adults, so to consider it a matter that concerns children is not sensible. For further research, it is worthwhile to look into CP problems for adults at the population level, as signaled by Andersson et al. in Sweden [32].

Third: in the Netherlands, we keep lacking insight at the population level on the group of children with CP as a whole. The present study, although the field work has been performed in the mid-nineties of the 20th century, gives the last Dutch population-based data on children with CP – the last birth year was 1988. Our study was a partner in SCPE (first tranche) and we had part in the SCPE working group that prepared the SCPE paper on CP-classification with the "decision tree" [17].

Dutch current research on children with CP is being done via the programme PERRIN (Pediatric Rehabilitation Research In the Netherlands [33-41]. This outstanding programme with co-operative research from medical, allied health and psychological disciplines has raised our understanding of many specific aspects: functioning with or without interventions or adaptations, evaluation of interventions, family centeredness and more. PERRIN research relies on well-described cohorts that

were acquired through convenience samples or rehabilitation practices. Conclusions drawn from this research can be generalized only to those children with CP that are comparable with the selected sample studied.

The key to providing "up-to-date" population-based data on CP epidemiology is a regional register in a Dutch area with (indicative) 1.5 million inhabitatants. Registers working at standards set by SCPE warranting a representative and up-to-date picture give the best quantitative basis for optimal services. An optimal framework for prospective research on many aspects is set by a register.

Find out if a register is feasible

Looking at the efforts invested in the 90s for a one-time representative Dutch study it is not advisable to do a single, cross-sectional study again. We recommend that from 2011 a serious attempt be undertaken to identify all conditions that have to be met to set up a sound register. This will need sufficient funding for a potential lifespan of no less than 12 years. Issues of size of the area to be included, methods of recruitment, cooperation of "the field" being the professionals that diagnose and report on the children, data quality and privacy issues are large assignments. Policy-makers will have to be convinced of the value and prepared to provide the means. Then again: if a register for children with CP is feasible, other diagnosis that benefit from being recognized at the group level could be included in the register to make funding the register benificial for more children with chronic disorders.

Stakeholders in a CP and even more in a CP-plus or Childhood Disability register are the children and their families in the first place. They will benefit from recognition of the size and characteristics of their group, which will be a stimulus for optimal services. Workers in medical or allied medical professions will have better insight in the group's need for professional attention, rehabilitation, adaptive devices and sometimes interventions. Other stakeholders are researchers, who can benefit from a representative group for fundamental or evaluative studies, prospective where needed. Important stakeholders of course are government authorities that have the political responsibility to plan ahead for the availability of services in the broad sense (medical, educational, technical and social).

The SCPE group knows problems and possibilities and gives valuable advice [20, 42]. An example: Parkes from the Northern Ireland CP register looked into possible recruitment bias in 2006 [43] and concluded: "Care must be taken in the recruitment of children with CP through clinic-based populations, although these routes may prove more successful in follow-up. Provided they are comprehensive, case registers

have a valuable contribution to make to clinical research by providing a sampling frame including information on baseline characteristics of an affected population" unquote.

A register is something that has to be set up well – there is no way of doing it "half".

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Summary

Cerebral Palsy (CP) is defined by the presence of motor impairment resulting from non-progressive cerebral pathology acquired early in life. CP is not a single disease with one known cause and one manifestation, but it is an umbrella term for several distinct cerebral laesions and motor manifestations. It is the largest cause of primary motor disability among children and consequently the children with CP and their parents are a very relevant group in the field of child rehabilitation medicine.

Children with CP and their families often make demands on diagnostic, therapeutic, technical and social facilities. Prevalence estimates (knowing the presence in numbers of these children in relation to the entire population) are needed to improve treatment and services. Information on the distribution of clinical presentations and associated disorders is needed as well. As 'recent' Dutch data were not available, the study described in this thesis aimed – among other things – to assess the population prevalence of CP in the Netherlands. This study was performed in a representative Dutch area with 1.2 million inhabitants of whom 172,000 were born between 1977 and 1988. The children were visited and examined, usually at their homes, in the period 1995–1997. The time interval from 1988 guaranteed that the children were at least six years and their motor manifestation of CP had stabilized.

The protocol used in this study was adopted from previous well-known cooperative studies from Germany and Sweden. Criteria for classification were shared and trained with the research workers from these studies. The protocol was expanded with items relevant to the Dutch situation and to Dutch rehabilitation medicine.

Chapter 2 describes the ascertainment and prevalence calculation [1]. Care had to be taken to avoid selection bias. Medical practices (such as rehabilitation centres, paediatric and child neurological departments and institutions for mentally handicapped) were consecutively asked to contact their (supposed) CP cases. Next, the BOSK – association of the physically disabled and their parents – and finally regional media (newspapers, radio stations) assisted in the ascertainment. In total, 170 supposed children with CP – as reported by any of the sources – underwent an examination by an experienced clinician (consultant in child rehabilitation medicine). Of these 170 children, 127 proved to be definite cases of cerebral palsy. This 'loss' of 25% by early mis-diagnosis was an important finding and it taught us that a too early CP-diagnosis may be challenged. Preferably this must be rechecked at the child's age of five or six.

Under-ascertainment (due to parents' refusal to cooperate) was present but could be quantified. Looking at major characteristics, the group that refused did not differ significantly from the group that was actually examined.

The population prevalence of CP over the birth year period 1977–1988 was calculated as 1.51 per 1000 inhabitants (average over the 12 birth years). The calculated prevalence however rose significantly over time: from 0.77 ('77–'79) to 2.44 ('86–'88). This rising trend was in accordance with other studies.

Chapter 3 described the distribution of clinical subtypes, motor disability and important co-morbidity (additional signs of cerebral dysfunction such as mental retardation, visual disability and epilepsy) [2]. In-depth studies of medical files in association with the history taken at the visit helped to identify aetiological moments in a majority of cases. Perinatal probems in prematurely born children accounted for fifty percent of identified aetiological moments. By comparing the four most recent birth years with the earlier eight birth years possible trends were studied.

Spastic subtypes accounted for over 90 percent of all CP cases: bilateral spastic cerebral palsy as a group (spasticity on both left and right legs and/or arms) are the majority, although spastic hemiplegia (left or right side affected) is percentage-wise the largest individual clinical subtype. Epilepsy and mental retardation are common. In general the distribution of clinical patterns remained rather constant in following years, as did the other studied items. Comparable studies performed in other countries showed similar findings. Although this study revealed a prevalence rise, no possible explanation for this phenomenon could be found.

Chapter 4 studied the prevalence of impairments of body function and structure (as found at the physical examination) and limitations in functional activities and their inter-relationships in Dutch children with spastic cerebral palsy (CP) [3]. Although their data were present the non-spastic subgroups were too small for valid statistics so they are not included in this chapter. So 119 children with spastic CP, aged 6 to 19 years, were examined. Anthropometry (body measures), gross motor functioning (by Gross Motor Function Classification System, a system to classify gross motor functioning) and manual ability (by Manual Ability Classification System, a similar system), muscle tone, abnormal posture, joint range of motion (ROM) and major orthopaedic impairments were assessed, besides limitations in mobility and self-care activities.

Children with spastic CP on the average had both a lower body height and weight compared to typically developing peers. Over sixty percent walked independently, 40% had no range of motion deficits in the major joints. The rate of impairments was associated with GMFCS and MACS levels. Hip dislocations were rarely encountered and if so only in more severe cases. Children with tetraplegia (legs and arms equally affected) and diplegia (legs more affected than arms) differed in activity limitations. Impairments and limitations in mobility and self-care activities were only modestly

related in multivariate analyses. This chapter showed the distribution of CP-related characteristics to be comparable with representative studies of other countries. The distinction between diplegia and tetraplegia is relevant from a functional point of view. The child's functional limitations are not a straightforward consequence of the neuro-orthopedic impairments, which makes likely that other factors are important as well (such as intra-personal or environmental factors). An activity-oriented approach in rehabilitation medicine goes beyond treating specific impairments.

Chapter 5 explored characteristics of personality descriptors, communication and behaviour difficulties in the representatiev group of children with cerebral palsy. Personality descriptors (addressing self-esteem, control, prosocial capacity and mood), difficulties in communication, contactual skills and problem behaviour were assessed at the interview with the parents. We found that the felt degree to have control over one's life and self-esteem appeared to be restricted in 10–20% of the children. Mood was not a specific problem in children with cerebral palsy. The ability to express one-self was impaired in 26%; writing was impaired in 50%. Approximally one third suffered from incidental to frequent behaviour problems. Impaired intellectual functioning was an associated factor with behaviour problems, impaired gross motor functioning to a lesser extent. This chapter concludes that the most prevalent psychological and socio-emotional problems in children with CP appeared to be emotional instability, being dependent, lack of confidence and communication limitations (self expression and writing). Children without intellectual disability seem to function without significant problems. For daily practice: it is important for professionals to be attentive not only to academic achievement but also to emotinal and behaviour problems, in order to stimulate the children's development towards participation and autonomy.

Chapter 6 describes health care utilisation – in the broad sense – and school careers of Dutch children with cerebral palsy. The interview with the parents focused on the child's previous and current situation in medical and allied health care and school career. It was found that eighty percent of the Dutch children with CP are seen on a regular basis by a consultant in rehabilitation medicine. Hip and ankle-foot surgery has respectively been performed in 24 and 20 percent of children, hip surgery mainly in strongly affected children (GMFCS level IV or V). Apart from anti-epileptic medication and laxatives (in 20% and 5% respectively) hardly any medication is taken, notably no spasmolytic drugs. Orthotic devices (orthopedic shoes or orthoses) are used by 80% of the children, mobility devices (wheelchairs, special bicycles) by 50%.

Sixty-two percent of the children are treated in a multidisciplinary setting and 56% attend special schools, multidisciplinary treatment and school frequently in

combination. Less affected children (with GMFCS level 1 and normal intelligence) more often had monodisciplinary or no treatment at all and attended mainstream schools.

In conclusion: Dutch practice regarding CP children, especially for those more affected, features special schools combined with multidisciplinary treatment. Adaptive devices and orthopaedic surgery are widely used. Medication use is minimal. Timely preventive hip surgery probably contributes to the low rate of hip dislocation in the Netherlands.

Chapter 7, the General discussion starts with a recapitulation of the reason and scope of the study. The separate chapters are summarized and, if applicable and not discussed in the chapter itself, considerations or knowledge known from more recent practice or studies performed abroad are discussed. After all, even though this study's field work is from some time ago, no more recent Dutch data are is available at a population-based level.

More recent prevalence rates as known from studies abroad do not seem to indicate that the strong rise as reported by us in the birth years up to 1988 has continued. Most likely prevalence across Europe stays between 2 and 3 per 1000. If the last Dutch prevalence calculation (2.44 per 1000, calculated for the birth years '86–'88) were still valid in recent years, in the Netherlands in each year group we would see some 400 children with CP.

The General discussion ends with three recommandations. First: check the working diagnosis CP in a young child by looking again to the clinical manifestation at age five or six – as advocated by SCPE.

The second recommandation is that, as CP is not a life-threatening condition, care and services should focus not only on children. The transition of youth to adulthood needs major attention and so do adults with CP. The Dutch system, as described in chapter 6, often provides for children a daily environment of multidisciplinary rehabilitation and school together. After leaving school, the young man or woman with CP (and family or partner) finds the medical, technical and social services no longer at one place. The needs also change with growing into or in adulthood.

The third recommendation in the general discussion is: start a Dutch CP register or at least find out if a Dutch CP register is feasible. In other countries, professionals and policymakers are informed on regional rising or falling rates of CP, possible changes in distribution of the CP picture itself or of important associated disorders. Representative and reliable information is gathered there via regional CP registers that work at the standards set by and cooperate within, the Surveillance of Cerebral Palsy in Europe group. As such they contribute to multi-centre research, an important

example of which is SPARCLE, a European multi-centre project focusing on contextual factors and quality of life.

The Netherlands are outsiders again in this field -after this survey on birth years 1977 through 1988. There is little sense in performing a one-time survey again. A Dutch register should focus on an area of some 1.5 million inhabitants and be set up for no less than twelve or fifteen years in order to truly signal trends. Policymakers have to be informed to cooperate and provide funding. Essential support is needed from the professionals 'in the field', that report on children with CP in the register's area, of course with privacy. Then the register itself needs careful management of people, data and resources.

Then again, a register set up for children with CP could provide information as well on children with other conditions that cause activity limitations (e.g. neuromuscular diseases, reumatoid diseases). This would change the register's scope from disease-oriented to activity-oriented as well and this would increase its benefits to society.

A register is something that has to be set up well – there is no way of doing it "half".

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Samenvatting

Bij Cerebrale Parese (CP) is per definitie sprake van motorische stoornissen veroorzaakt door een niet-progressieve hersenbeschadiging of hersenafwijking, verworven in de vroege fase van het leven. CP is niet één enkel ziektebeeld met één bekende oorzaak en een bekend klinisch beeld of verloop, maar het is een koepelbegrip voor meerdere te onderscheiden vroeg ontstane hersenbeschadigingen en ook meerdere motorische uitingen daarvan. CP is bij kinderen de meest voorkomende oorzaak van primaire (van jongs af aan aanwezig) motorische stoornissen en als zodanig zijn kinderen met CP en hun ouders een grote, wellicht de grootste, groep 'klanten' in de wereld van de kinderrevalidatiegeneeskunde.

Kinderen met CP en hun gezinnen doen een groot beroep op diagnostische, therapeutische, technische en sociale voorzieningen. Prevalentieschattingen – het schatten van de aanwezigheid, in aantal van deze kinderen, in relatie tot de hele bevolking – zijn nodig om de behandeling en voorzieningen te verbeteren. Omdat 'recente' Nederlandse gegevens niet beschikbaar waren, beoogde de studie beschreven in dit proefschrift – onder andere – om de prevalentie van CP in Nederland te berekenen. De studie werd uitgevoerd in een representatief Nederlands gebied met 1,2 miljoen inwoners, met 172.000 kinderen geboren tussen 1977 en 1988. De kinderen werden, meestal thuis, bezocht in de periode 1995–1997. Dit tijdsinterval vanaf 1988 zorgde er voor dat ze tenminste 6 jaar oud waren en dat hun motorische beeld zich had gestabiliseerd.

Het protocol dat werd gebruikt in deze studie was overgenomen uit eerdere, bekende studies uit Duitse en Zweedse samenwerkingsprojecten. Met de onderzoekers van deze studies werden de criteria voor classificatie van te onderzoeken items gedeeld en geoefend. Het protocol werd uitgebreid met items van belang voor de Nederlandse situatie, gezien vanuit de Nederlandse revalidatiegeneeskunde.

Hoofdstuk 2 beschrijft de methode van 'werving' en de prevalentieberekening [1]. Selectiebias diende te worden vermeden. Medische praktijken (zoals revalidatiecentra, universitaire kindergeneeskundige en kinderneurologische afdelingen en instellingen voor kinderen met verstandelijke beperkingen) werden achtereenvolgens gevraagd om hun eerder gediagnosticeerde kinderen met CP te benaderen. Vervolgens hielpen de BOSK – vereniging van motorisch gehandicapten en hun ouders – en de regionale media (kranten, radiostations) bij de werving. In totaal werden 170 'vermeende' kinderen met CP – zoals door de ouders aangemeld na benadering door een van de bronnen – bezocht en onderzocht door een ervaren kinderrevalidatiearts. Van deze 170 kinderen bleken 127 aan de criteria van CP te voldoen. Dit 'verlies' van 25% door een eerdere foutdiagnose was een belangrijke bevinding en leerde ons dat een te vroege

diagnose van CP ter discussie moet worden gesteld. Bij voorkeur moet de diagnose opnieuw klinisch worden geverifieerd als het kind 5 of 6 jaar is.

De uitval door weigering van aangeschreven ouders om mee te doen aan het onderzoek kon worden berekend. De groep die niet meedeed week op de belangrijkste kenmerken niet significant af van de groep die daadwerkelijk werd onderzocht.

De prevalentie van CP in de bevolking over de periode van de geboortejaren 1977 tot en met 1988 werd berekend als 1,51 per 1000 inwoners (gemiddelde over de 12 geboortejaren). De berekende CP-prevalentie steeg echter aanzienlijk in de tijd: van 0,77 in de periode '77–'79 tot 2,44 in de periode '86–'88. Deze stijgende trend is ook gezien in buitenlandse studies.

In hoofdstuk 3 worden de klinische subtypes, de ernst van de motorische beperking en belangrijke comorbiditeit, zoals mentale retardatie, visuele beperkingen en epilepsie, beschreven [2]. Het met toestemming van de ouders doornemen van ziekenhuisdossiers, samen met de opgenomen ziektegeschiedenis, maakte het mogelijk om etiologische momenten (het waarschijnlijke moment van de hersenbeschadiging – voor, rond of na de geboorte) te bepalen in een meerderheid van de gevallen. Als dat lukte bleek dat voor ongeveer vijftig procent te gaan om perinatale problemen bij vroeggeboorte. Door het vergelijken van de vier meest recente geboortejaren met de eerdere acht geboortejaren konden trends worden bestudeerd. Spastische subtypes waren goed voor meer dan 90% van alle gevallen van CP. Bilaterale spastische cerebrale parese (spasticiteit zowel links als rechts aan de benen en/of armen) als groep vormt de meerderheid, hoewel spastische hemiplegie (alleen de linker- of rechterkant aangedaan) procentueel gezien het grootste individuele klinische subtype is. Epilepsie en leerstoornissen en mentale retardatie komen vaak voor. In het algemeen bleef de verdeling van de klinische patronen vrij constant in de volgende jaren, net als de andere onderzochte items. Vergelijkbare onderzoeken uitgevoerd in andere landen toonden soortgelijke bevindingen. Terwijl deze studie door de jaren een duidelijke prevalentiestijging liet zien, werd geen indicatie gevonden voor een verklaring van deze prevalentiestijging.

Hoofdstuk 4 beschrijft de prevalentie van stoornissen in functie en structuur van het lichaam (zoals vastgesteld bij lichamelijk onderzoek), van beperkingen in functionele activiteiten en hun onderlinge relaties in een groep Nederlandse kinderen met spastische CP [3]. Hoewel deze gegevens ook voor de 8 niet-spastische kinderen aanwezig waren, waren de subgroepen hiervan te klein voor valide conclusies, zodat ze in dit hoofdstuk niet zijn meegenomen. In totaal werden 119 kinderen met een spastische vorm van CP in de leeftijd van 6 tot 19 jaar onderzocht. Lengte en gewicht,

grove motoriek (bepaald met de Gross Motor Function Classification System, een systeem om de grove motoriek te classificeren) en manuele vaardigheid (bepaald met de Manual Ability Classification System, een soortgelijk systeem voor de arm-handfunctie), spierspanning, abnormale houding, eventuele bewegingsbeperkingen van de grote gewrichten en de belangrijkste orthopedische afwijkingen van heupen en rug werden onderzocht, naast de beperkingen in mobiliteit en zelfzorgactiviteiten. Kinderen met spastische CP blijken gemiddeld zowel korter als lichter in vergelijking met zich normaal ontwikkelende leeftijdgenoten. Ruim zestig procent liep zelfstandig, 40% had geen bewegingsbeperkingen in de grote gewrichten. De vastgestelde stoornissen was gerelateerd aan GMFCS- en MACS-niveaus. Heupluxaties kwamen weinig voor. Kinderen met tetraplegie (benen en armen in gelijke mate aangedaan) en diplegie (benen meer aangedaan dan armen) verschilden in beperkingen in activiteiten. De gevonden stoornissen en de beperkingen in activiteiten met betrekking tot mobiliteit en zelfzorg hingen slechts beperkt samen in de multivariate analyses. Dit hoofdstuk stelt tenslotte vast dat de verdeling van de gevonden CP-gerelateerde kenmerken vergelijkbaar is met representatieve studies uit andere landen. Het onderscheid tussen diplegie en tetraplegie is relevant vanuit functioneel oogpunt. De functionele beperkingen van het kind waren niet het rechtstreekse gevolg van de stoornissen van het bewegingsapparaat, wat aannemelijk maakt dat andere factoren (zoals intra-persoonlijke factoren of omgevingsfactoren) evenzeer van belang zijn. Revalidatiegeneeskundige behandeling gericht op het verbeteren van activiteiten gaat dus verder dan de behandeling van specifieke stoornissen.

Hoofdstuk 5 onderzocht de kenmerken van persoonlijkheid, communicatie en gedragsproblemen bij deze representatieve groep kinderen met CP. Gegroepeerde persoonlijkheidskenmerken (eigenwaarde, mate van ervaren controle, (pro)sociale vaardigheid, stemming), problemen in de communicatie, contactuele vaardigheden en probleemgedrag werden onderzocht middels het interview met de ouders. De mate van ervaren controle greep op het leven en het gevoel van eigenwaarde bleken bij 10–20% van de kinderen met CP lager te zijn dan in een referentiegroep. Stemmingsproblemen waren niet significant aanwezig. De mogelijkheid om zich te uiten was verminderd bij 26% van de kinderen, het vermogen tot schrijven bij 50%. Bij ongeveer een derde van de kinderen was er in mindere of meerdere mate sprake van gedragsproblemen. Gedragsproblemen hingen samen met lager intellectueel functioneren, in mindere mate gold dat voor de ernst van de beperkingen in grove motoriek. In dit hoofdstuk wordt vastgesteld dat de meest voorkomende psychologische en sociaal-emotionele

problemen bij kinderen met CP emotionele instabiliteit, afhankelijkheid, gebrek aan zelfvertrouwen en communicatiebeperkingen (zelfexpressie en schrijven) bleken te zijn. Kinderen zonder verstandelijke handicap lijken te functioneren zonder noemenswaardige problemen. Voor de praktijk betekent dit, dat naast aandacht voor schoolse prestaties professionals ook dienen te letten op emotionele en gedragsproblemen, om de ontwikkeling van deze kinderen met CP naar participatie en autonomie later in het leven te bevorderen.

Hoofdstuk 6 beschrijft het gebruik van gezondheidszorg – in brede zin – en de schoolloopbaan van Nederlandse kinderen met CP. Het interview met de ouders behandelde de vroegere en huidige situatie van het kind in de medische en paramedische zorg en over de schoolloopbaan. Het bleek dat 80% van de Nederlandse kinderen met CP met regelmaat wordt gezien door een revalidatiearts. Heup- en enkel-voet-operaties zijn uitgevoerd in respectievelijk 24% en 20% van de kinderen, de heupoperaties vooral bij ernstig aangedane kinderen (GMFCS niveau IV of V). Afgezien van anti-epileptische medicatie en laxeermiddelen (in respectievelijk 20% en 5%) werd nauwelijks medicatie ingenomen, met name geen spasticiteitverminderende medicijnen. Orthopedische hulpmiddelen (orthopedische schoenen of orthesen) worden gebruikt door 80%, mobiliteitshulpmiddelen (rolstoelen, speciale fietsen) door 50% van de kinderen. Tweeënzestig procent van de kinderen wordt behandeld in een multidisciplinair team en 56% gaat naar een speciale school, bij veel van deze kinderen in een combinatie van beide. Minder aangedane kinderen (GMFCS niveau I, met een normale intelligentie) kregen vaker een enkelvoudige of helemaal geen behandeling en bezochten reguliere scholen. Samenvattend wordt de Nederlandse praktijk ten aanzien van kinderen met CP, vooral voor hen die ernstiger aangedaan zijn, gekenmerkt door speciale scholen waarin onderwijs en multidisciplinaire revalidatiebehandeling samengaan. Technische hulpmiddelen, evenals orthopedische chirurgie, zijn op grote schaal gebruikt. Medicijngebruik is minimaal. Tijdige preventieve heupchirurgie draagt wellicht bij aan het lage aantal gevallen met heupluxatie in Nederland.

Hoofdstuk 7, de algemene discussie, begint met een recapitulatie van wat leidde tot de studie. De afzonderlijke hoofdstukken worden samengevat en, indien van toepassing, wordt kennis en overwegingen die sindsdien beschikbaar kwamen door recente studies in het buitenland besproken. Voor de situatie in Nederland zijn geen recente gegevens beschikbaar op bevolkingsniveau, omdat het veldwerk van deze studie al van enige tijd geleden is. Recentere prevalentiecijfers uit het buitenland lijken niet aan te geven dat de sterke stijging zoals gerapporteerd tot eind jaren tachtig zich na die jaren heeft voortgezet. Het meest waarschijnlijke prevalentiecijfer in Europa

blijft tussen de 2 en 3 per 1.000. Als onze laatste Nederlandse prevalentieberekening (2,44 per 1.000, berekend voor de geboortejaren '86–'88) nog juist zou zijn, dan zien we de afgelopen jaren in Nederland in elke jaargroep ruim 400 kinderen met CP.

De algemene discussie eindigt met drie aanbevelingen. De eerste is: verifieer de waarschijnlijkheidsdiagnose CP als een kind vijf of zes jaar is – zoals ook door SCPE aanbevolen.

De tweede aanbeveling is: bedenk dat, omdat CP immers geen levensbedreigende aandoening is, behandeling en voorzieningen niet alleen gericht moeten zijn op kinderen. De overgang van jeugd naar volwassenheid heeft grote aandacht nodig evenals de hulpvraag van volwassenen met CP. Het Nederlandse systeem, zoals beschreven in hoofdstuk 6, biedt vaak voor kinderen een dagelijkse omgeving van de multidisciplinaire revalidatie en school samen op één plek. Na het verlaten van school kan de jonge man of vrouw met CP (en ouders of partner) de medische, technische en sociale voorzieningen niet langer vinden op één plek. De hulpvragen veranderen ook bij (de groei naar) volwassenheid.

De derde aanbeveling van dit onderzoek is het starten van een Nederlands CP-register, of tenminste het onderzoek of dit haalbaar is. In andere landen hebben professionals en beleidsmakers kennis van de regionale (stijgende of dalende) prevalentie van de CP, mogelijke veranderingen in de verdeling van de manifestatie van het CP-beeld zelf of van belangrijke bijkomende aandoeningen. Daar wordt representatieve en betrouwbare informatie verzameld via regionale CP-registers, die werken volgens de standaarden van, en samenwerken in, de Surveillance van Cerebral Palsy in Europe groep. Als zodanig dragen zij bij aan multi-centre onderzoek, waarbij een belangrijk voorbeeld is SPARCLE, een Europees project rond contextuele factoren en kwaliteit van leven.

Nederland is na het beschreven onderzoek over de geboorte jaren 1977 tot en met 1988 weer buitenstaander op dit gebied. Het is weinig zinvol om een eenmalig onderzoek in Nederland te herhalen. Er dient een Nederlands register dat zich richt op een gebied van ongeveer 1,5 miljoen inwoners te worden ingesteld voor minstens 12 tot 15 jaar om werkelijk trends te kunnen volgen. Beleidsmakers moeten hierover worden geïnformeerd opdat zij het belang zien van het meewerken aan een dergelijk register en financiering waarborgen. Onmisbare steun is nodig vanuit 'het veld', de professionals die over de kinderen met CP in het gebied van het register rapporteren, uiteraard met borging van privacy. Het register zelf dient met mensen, data en middelen zorgvuldig om te gaan.

Indien een register voor kinderen met CP kan worden opgezet zou dat zich ook kunnen richten op kinderen met andere aandoeningen die leiden tot beperkingen in activiteiten (bijvoorbeeld neuromusculaire ziekten, reumatoïde aandoeningen). Dan wordt de blik verruimd van aandoeninggeoriënteerd tot activiteitgeoriënteerd, wat het maatschappelijk rendement zal vergroten.

Een register moet goed worden opgezet – dat kan niet 'half'.

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Ook de stichting BIO Kinderrevalidatie dank ik voor de verleende subsidie. BIO was in de persoon van directeur Willem van Tuyll van Serooskerken vooral de eerste jaren betrokken. Beste Willem, hartelijk dank voor de belangstelling die je steeds had voor het onderzoek en voor je deelname aan de stuurgroep.

Toen dat wat te doen stond duidelijk werd betreffend veldwerk (misschien wel driehonderd of meer kinderen opzoeken en onderzoeken) besefte ik dat mij dat alleen niet zou lukken. Ik vroeg mijn gewaardeerde "rustende" collega Rob Blankesteijn van de Sint Maartenskliniek om mij te helpen. Beste Rob, na een korte zorgvuldige overweging stonden Aty en jij aan de deur met prachtige zelfgekweekte bloemen – en met het antwoord dat je graag meedeed. Die medewerking is fantastisch verlopen, daar dank ik je heel hartelijk dank voor. We gingen samen met Onno naar de collegae in Tübingen in de voorbereidingsfase. Naast je "productie" aan bezochte kinderen deed je ook enthousiast mee aan overleggen met de methodologen en promotoren. Jouw kritische opmerkingen leidden er mede toe dat we besloten dat prevalentieberekening middels capture-recapture als niet goed genoeg terzijde werd geschoven – en uiteindelijk zelfs niet in de discussie genoemd werd omdat "wat je niet gebruikt je ook niet hoeft te bespreken". Dat was wel wat want daar was heel veel moeite en tijd in gaan zitten! Typerend voor je onbaatzuchtige instelling Rob, is dat je de vergoeding voor je bijdrage naar een project van een bevriende klinisch chemicus in de tropen liet overmaken.

Bea Heybroek was mijn gewaardeerde secretaresse in Groot Klimmendaal. Zij was ook de vriendelijke, nauwgezette spil van het onderzoek, regelde alle huisbezoeken voor ons met de ouders en verwerkte de floppies die we mee terugnamen. Wat schrok ik toen ik, na mijn overstap naar Tilburg, vernam van haar overlijden. Ik kan de dank die ik haar verschuldigd ben nu alleen aan haar Geert overbrengen.

Jan Grootaarts voorzag ons van een programma op de laptop waarmee we de bevindingen bij het onderzoek op locatie konden invoeren. Hartelijk dank Jan, misschien heb je niet vaak meer aan het onderzoek gedacht de laatste jaren, maar met jouw software is het gegevensbestand gebouwd, waardoor we qua data-invoer later geen aanvullend handwerk hoefden te doen.

Het bereiken van ouders en kinderen is eerst en vooral via de kinderrevalidatie-praktijken gebeurd. Ik kreeg alle medewerking van de collegae revalidatieartsen. Ik noem van Groot Klimmendaal in Arnhem Harry Vos, van de Sint Maartenskliniek in Nijmegen Margriet Poelma, van de Hoogstraat in Utrecht Marja van Tol, van de Kastanjehof in Apeldoorn Harry Daudt en van het Roessingh in Enschede Karel Maathuis. Hartelijk dank centra en collegae!

Ook vroegen en kregen we – niet in het minst door de goede relaties van Rob Blankesteijn – royale medewerking van de afdelingen Kindergeneeskunde (prof. dr. R. Sengers) en het Interdisciplinair Kinder Neurologisch Centrum (prof. dr. F. Gabreels) van het Radboud Ziekenhuis. Beide centra schreven voor ons de ouders aan van bij hen gediagnosticeerde kinderen met CP. Hartelijk dank aan genoemde hoogleraren en aan de instituten!

De BOSK, vereniging van motorisch gehandicapten en hun ouders, schreven ook hun leden aan voor zover de kinderen waarschijnlijk CP hadden. Ook langs deze weg kregen we respons. Hartelijk dank voor deze medewerking!

Om als het ware het net van de acquisitie dicht te trekken hebben we met een persbericht contact gelegd met "de media". Dat resulteerde in artikelen of vermeldingen in regionale dagbladen, huis-aan-huis-bladen en in interviews met regionale en lokale radiostations. Het is ondoenlijk om alle bijdragen op dit gebied op naam te vermelden maar deze welwillende medewerking memoreer ik toch. Bijzonder is dan hoeveel mensen je terugmeldden dat ze "ergens" in krant of op de radio over het onderzoek lazen of hoorden.

De ondersteuning op epidemiologisch/methodologisch gebied bij het eerste artikel –eind vorige eeuw dus– kregen we in Utrecht van dr. Yvonne van der Schouw en dr. Carl Moons van het Julius Centrum. Het verbaast me niet dat jullie inmiddels beide hooggeleerd zijn, beste Yvonne en Carl, want jullie scherpe analyses maakten grote indruk op mij, hartelijk dank daarvoor!

In een latere fase kwam de methodologische ondersteuning van Henk's afdeling Revalidatie van de Erasmus Universiteit. Eerst werkte ik samen met dr. Else Odding, later met dr. Marij Roebroeck en met drs. Sander Hilberink. Bij Else en Marij was de belangstelling voor CP-epidemiologie gewekt, zij publiceerden een groot reviewartikel over dit onderwerp.

Beste Else, mijn herinnering aan de samenwerking bji "ons project", het artikel over klinische en bijkomende zaken, is erg prettig, heel hartelijk dank daarvoor.

Beste Marij, de laatste jaren hebben we soms meer, soms minder intensief contact gehad, overigens ook omdat mijn eerste aanspreekpunt vaak Sander was. We gingen elkaar steeds beter begrijpen. Ik ben onder de indruk van wat je voor mij en voor jouw afdeling deed. Het is mij een voorrecht dat je opponeert op mijn promotie. Ook daarvoor mijn hartelijke dank.

Beste Sander, het was altijd een leuk ritje van Nijmegen-Oost naar Arnhem-Zuid om op vrijdagochtend, schouder aan schouder, met de data bezig te zijn. We hebben het lang niet altijd over epidemiologie gehad trouwens en we waren het ook niet altijd eens. Daarnaast ging/gaat het contact per e-mail ook prima. Over Vestdijk zei men dat hij sneller schreef dat Onze Lieve Heer kon lezen, zo zeg ik wel eens dat Sander sneller de mail antwoordt dan Onze Lieve Heer kan mailen. Ik dank je heel hartelijk voor je steun, humor en energie. José en ik kijken uit naar jouw promotie. Ik dank je ook voor je bereidheid om vandaag mijn paranimf te zijn samen met Marije, mijn oudste dochter.

Beste Betty Koehorst, als secretaresse van Henk Stam was je de altijd correcte verbinding. Je voelde goed aan waar ik rechtstreeks met Henk moest overleggen, hielp me op allerlei manieren, bijvoorbeeld met mijn toegang tot de Erasmus MC Medische Bibliotheek. Afspraken via jou klopten altijd. Ook al was ik een promovendus op afstand en ben ik fysiek in die jaren niet vaak bij jullie geweest, toch voelde het aan of het dichtbij was. Met telefoon, fax en vooral e-mail kwam alles prima over. Beste Hetty Mulder, je neemt het stokje van Betty over, ook jou dank ik voor de hulp die ik recent al van je mocht ontvangen.

Beste Paul Stumpel, ik dank je voor alle ontspannende rondjes golf en de mooie bewerking die je voor het omslagontwerp maakte van "het spastische jongetje" uit de beroemde publicatie van Little uit 1861.

Beste Renate Siebes, hartelijk dank voor de vlotte interactie en het gemak waarmee je de inhoud van het proefschrift hebt ge-layout. Het ziet er prima uit!

Ik dank de manuscriptcommissie of "kleine commissie", bestaand uit de professoren Arts (kinderneurologie) en Steyerberg (maatschappelijke gezondheidszorg), beide van het Erasmus MC en professor Becher (kinderrevalidatie, VUMC) en de promotoren hartelijk voor de aan het manuscript bestede aandacht. Deze commissie wordt op de

promotiedag tot "grote commissie" uitgebreid met de professoren Jongbloed (emeritus hoogleraar revalidatiegeneeskunde LUMC) en Grol (emeritus hoogleraar kwaliteit van zorg, Radboud Universiteit en Rijksuniversiteit Limburg) en de doctoren Maathuis (kinderrevalidatie UMC Groningen) en Roebroeck (revalidatie-onderzoek Erasmus MC). Ik dank hen voor hun bereidheid om te opponeren.

Hooggeleerde Jongbloed, gewaardeerde opleider uit het LUMC (voorheen Academisch Ziekenhuis Leiden), beste Hans, ik dank je voor je stijl van opleiden. Je liet je assistenten hun eigen denkwerk doen, stelde vragen, zag de hoofdlijn en gaf ruimte voor het ontwikkelen van de eigen stijl. Sterker nog: je moest er niet aan denken dat de assistenten een soort klonen van de opleider en dus van elkaar zouden worden.

Dank ben ik op deze plaats ook verschuldigd aan de partners van Surveillance of Cerebral Palsy in Europe, SCPE. Zij zullen uiteraard in het Engels nog een bedankje krijgen. Eind jaren '90 waren we als Nederlandse studie "Arnhem" partner in de eerste tranche van dit samenwerkingsverband van (toen) 14 population-based onderzoeken bij cerebrale parese. Het project werd gesubsidieerd door de Europese Gemeenschap. Het formuleren van een gezamenlijk begrippenkader rond de klinische classificatie van CP was een inspirerend gebeuren dat voor een groot deel in workshops bij ons op Groot Klimmendaal en in Berg en Dal plaatsvond. De SCPE-terminologie maakt nu deel uit van de internationale klinische praktijk en de eerder genoemde instructie-CD-rom is een uitvloeisel van deze inspanningen. Naast veel inhoudelijke inspiratie leverden SCPE-bijeenkomsten in Grenoble, Liverpool, Rome, Newcastle en Toulouse geanimeerde contacten op. We mochten in de keuken kijken van andere population-based studies of van soms lang bestaande CP-registers. Ik noem en dank hier Inge Krägeloh, Bengt en Gudrun Hagberg, Steven Jarvis, Allan Colver, Monica Topp, Peter Uldall, Christine Cans, Jerome Fauconnier, Jackie Parkes, Vicki McManus, Owen Hensey, Eva Beckung, Mary-Jane Platt, Ann Johnson, Maria-Giulia Torrioli, Catherine Arnaud en Richard Michaelis en anderen voor de inspirerende contacten binnen of naar aanleiding van SCPE. Om volledig te zijn: ook dank aan Fiona Stanley and Eve Blair uit Australië voor het vererende verzoek om Nederlandse prevalentiecijfers te mogen vermelden in hun grote werk voor MacKeith Press "Cerebral Palsies: Epidemiology and Causal Pathways" dat in 2000 uitkwam. Jammer genoeg was onze eigen prevalentiepublicatie toen nog niet geaccepteerd, wat ons verhinderde een "voorpublicatie" in een overzichtswerk te laten doen.

Het gaat te ver om alle instellingen, collega's, leidinggevenden en teamleden waar ik zoal als revalidatiearts mee werkte met naam te noemen. Niet te vergeten de kinderen en hun ouders die hun vertrouwen in mij stelden! Ik ben allen dankbaar, voor het plezier dat ik in het werk had en heb.

Ik eindig met dank aan de allerbelangrijkste personen in mijn leven.

Lieve José: heel veel dank voor de achtendertig jaar dat we het goed hebben samen. We mogen tevreden zijn met wat we hebben en met de ruimte die we elkaar geven en die we nemen. We steunen elkaar en spreken elkaar ook aan. Zo kun je nog eens ouder worden!

Lieve kinderen, Marije, Otto en Leonie, jullie zijn enorm belangrijk voor José en mij. Het voltooien van dit proefschrift ging (dat is door jullie bevestigd) niet ten koste van mijn aandacht voor jullie. Wij genieten van de momenten die we samen, met de aanhang, meemaken. Dank voor de vreugde die jullie ons brengen en dank Marije dat je mijn paranimf wilt zijn.

Dit proefschrift draag ik op aan de nagedachtenis van mijn ouders Otto Wichers (1903-1964) en Frederika Wichers-Ovinge (1909-1986). Otto Wichers, vele jaren lid van de Kamer van Toezicht voor het notariaat, gold als "het toppunt van onkreukbaarheid". Zij blijven voor mij het voorbeeld van authenticiteit, zorgvuldigheid en respect voor iedereen en zij gaven mij de liefde voor muziek.

Levensloop Marc Wichers

Marc Jan Wichers werd op 9 april 1952 geboren in Emmen, groeide op in Exloo (Drenthe) en Haren (Groningen) en haalde zijn H.B.S.-B-diploma aan het Zernike College in Groningen in 1969. Na een schooljaar als A.F.S.-uitwisselingsstudent in Phillips, Wisconsin, Verenigde Staten, studeerde hij geneeskunde in Groningen en werd arts in 1976. Hij werkte als dienstplichtig arts in het Militair Revalidatie Centrum in Doorn en werd in 1978 arts-assistent in opleiding chirurgie in het Bleuland Ziekenhuis in Gouda. Marc stelde daar vast dat chirurgie niet "zijn" vak was. In 1979 begon hij de opleiding tot revalidatiearts in het Academisch Ziekenhuis Leiden (opleider prof. J.C. Jongbloed).

Op 1 juli 1983 werd hij als revalidatiearts aangesteld in de Sint Maartenskliniek te Nijmegen. In diverse vaste detacheringen startte hij met een consulentschap of spreekuur de functie revalidatiegeneeskunde of bouwde deze uit. Detacheringen of consulentschappen betroffen het Maasziekenhuis Boxmeer, Canisius Wilhelmina Ziekenhuis, Radboud Ziekenhuis, Dekkerswald, Werkenrode, de regionale verpleeghuizen en instellingen voor mensen met een verstandelijke beperking. Naast de detacheringen werkte hij op poliklinische en klinische afdelingen van het revalidatiecentrum van de Sint Maartenskliniek en was hij de consulent voor de afdeling Reumatologie. Hij was een periode voorzitter van de Medische Staf van de Sint Maartenskliniek, participeerde in stafactiviteiten in het Canisius Wilhelmina Ziekenhuis en was betrokken bij de opleiding tot revalidatiearts die in die periode in Nijmegen werd opgezet.

Tevens was hij medisch adviseur van de zich toen vormende Landelijke Werkgroep Beenprothesedragers en van de Stichting SOHO (hulphonden voor personen met ernstige beperkingen) te Herpen (Noord-Brabant). In 1985 verscheen de door hem samengestelde bibliografie "Wetenschappelijke publicaties van revalidatieartsen en voordrachten gehouden voor de VRA" [uitgave VRA, ISBN 90-71285-01-4].

In 1993 stapte Marc over naar het revalidatiecentrum Groot Klimmendaal, richtte zich op poliklinische kinderrevalidatie en inhoudelijk management op unitniveau en was plaatsvervangend opleider kinder-revalidatiegeneeskunde.

Groot Klimmendaal gaf hem de gelegenheid om het voorwerk en het veldwerk te doen voor het onderzoek beschreven in dit proefschrift. Dit onderzoek was een van de 14 partners van het Europese samenwerkingsproject Surveillance of Cerebral Palsy in Europe SCPE, eerste tranche. Marc werkte mee aan meerdere SCPE-publicaties. Het werk binnen SCPE gaf aanleiding tot het maken van de Reference and Training Manual op CDROM. Deze CDROM werd in 2006 door Marc vertaald en is via de dr. W.M. Phelps-Stichting beschikbaar voor Nederlandse professionals.

In 2002 volgde een tweede overstap, ditmaal naar het revalidatiecentrum Leijpark te Tilburg, waar hij werkte als poliklinisch kinderrevalidatie-arts en inhoudelijk manager op sectorniveau.

In 2007 stelde Marc vast dat afwisseling hem goed beviel. Hij besloot voortaan aanstellingen voor niet langer dan een jaar af te spreken. Zo werkte hij periodes van zes maanden voor Revalidatie Friesland op de locatie Medisch Centrum Leeuwarden en in het Revant Revalidatiecentrum te Breda. Periodes van een jaar werkte hij voor Sophia Revalidatie op de locatie Groene Hart Ziekenhuis te Gouda, in het Jan van Breemen Instituut te Amsterdam en voor het Revalidatiecentrum De Hoogstraat te Utrecht op de locatie Mytylschool Ariane de Ranitz. Hij is nu, tot september 2012, verbonden als kinderrevalidatiearts aan het revalidatiecentrum De Vogellanden in Zwolle, deels op de locatie Mytylschool De Twijn.

Marc was van 1984 tot 1992 lid van de Wetenschappelijke Commissie van de Nederlandse vereniging van revalidatieartsen VRA. Hij coördineerde het inhoudelijk deel van het Lustrumsymposium `Revalidant en Revalidatie` van de VRA in 1990. Van 1992 tot 1999 maakte hij deel uit van het dagelijks bestuur als penningmeester. In 2000 ontving hij de 'zilveren balk' van de VRA. Marc is lid van de kindersectie van de VRA, verder van de BOSK, de International Society of Physical and Rehabilitation Medicine, de International Society for Prosthetics and Orthotics en de Nederlandse Vereniging voor Kinderneurologie.

Sinds 1975 is Marc getrouwd met José Bots, orthopedagoog (Rijksuniversiteit Groningen 1977). Zij is senior docent/onderzoeker bij het Fontys Opleidingscentrum Speciale Onderwijszorg. Marc en José hebben drie kinderen. Marije (1979) is wiskundige en werkt bij PostNL. Otto (1981) studeerde af in de Engelse taal en literatuur en is singer-songwriter (artiestennaam Lucky Fonz III). Leonie (1982) is arbeids- en organisatiepsycholoog en werkt bij Randstad.

Overige interesses van Marc zijn onder andere muziek (actief als pianist en passief middels concertbezoek), taal en golf. Vanaf 2005 was hij penningmeester van de Vereniging Vrienden van het Concertgebouw de Vereeniging te Nijmegen, sinds 2011 is hij voorzitter.