Introduction and outline of the thesis
Introduction

Cerebral palsy (CP) is the most common cause of a physical disability in children. In Europe the prevalence of CP is approximately 2.0 in every 1000 live-born children.\textsuperscript{1,2} Prevalence decreased significantly to the late sixties, increased significantly to the mid-eighties, then stabilized and decreased slightly in the nineties.\textsuperscript{3,4} Wichers et al. found a similar trend of prevalence of CP in the Netherlands: a rising prevalence from 0.77 in the seventies to 2.44 in the eighties.\textsuperscript{5}

Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture causing activity limitations, which are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication and behaviour, by epilepsy, and by secondary musculoskeletal problems.\textsuperscript{6} The severity of the motor impairment, as well as the cognitive, communicative and behavioural impairments are different for each patient,\textsuperscript{7,8} resulting in a large variability in functioning in all domains.

The International Classification of Functioning, Disability and Health (ICF) provides a classification scheme for understanding the functional outcomes of a disorder or disease.\textsuperscript{9,10} It classifies the functional outcomes at two levels: (1) body function and structure (impairments), and (2) everyday activities (limitations) and participation (restrictions). The ICF also recognises contextual factors, which may affect the outcomes in all categories: environmental and personal factors. In the study described in this thesis, we used the ICF as a conceptual framework.

During growth and development from childhood to adulthood, children with CP may develop a variety of secondary musculoskeletal problems, such as muscle shortening or joint contractures. During this development, adolescence is an important phase because in this period many changes take place such as growth spurt, puberty, psychosocial

![Figure 1.1 The International Classification of Functioning, Disability and Health.\textsuperscript{9}](image-url)
development and changing expectations by parents or school or by the children themselves. The musculoskeletal problems and the changes during adolescence may affect the course of motor functioning and may influence daily activities and social participation of adolescents with CP. With respect to motor function, much attention has been given to the ambulatory prognosis of young children with CP.\textsuperscript{11,12} However, it is unknown, whether a child will maintain his or her level of mobility during puberty, and into adolescence. Retrospective studies of young adults with CP reported deterioration of motor function and even loss of ambulation in a subgroup of adolescents and adults,\textsuperscript{13-16} but longitudinal studies describing the course of motor function in adolescents and (young) adults, are not yet available. While motor function or mobility is an important aspect of CP, it is also important to know how children and adolescents with CP function in daily activities and participate in social life, and which factors are related to the level and course of functioning. Studies focussing on activities and participation have reported more restrictions in children and adolescents in relation to the severity of CP.\textsuperscript{17-19} However, although the level of motor functioning and daily living skills were strongly related to severity of the CP, other studies reported that the level of social functioning and communication may also be influenced by personal and environmental factors.\textsuperscript{20-24} With respect to the course of social functioning, no longitudinal studies are known, but cross-sectional studies have reported higher social isolation in adolescents with CP in comparison with younger children with CP.\textsuperscript{25,26}

Describing the level and course of activities and participation in children with CP gives much information about the functioning of children with CP. However, because the objective level of functioning does not necessarily correspond to the subjective experience concerning their functioning,\textsuperscript{27,28} it is also important to investigate children's subjective perceptions of their functioning.\textsuperscript{27,28}

The WHO has defined quality of life (QOL) as a subjective perception of an individual about his own position in life. In the literature both QOL and health-related quality of life (HRQOL) are described. QOL refers to holistic well-being, while HRQOL focuses on personal valuations of daily experiences.\textsuperscript{29-33} Since the HRQOL can be defined as the individual perceptions of various domains, the ICF domains of functioning can serve as a basis.\textsuperscript{34} Because QOL and HRQOL concern the subjective perception and feelings of the children, the children themselves would in principle be the best informants about their QOL or HRQOL. However, because children with CP often have limitations in communication and cognitive abilities, parental caregivers or other proxy reporters frequently respond on QOL and HRQOL measures on behalf of these children. Recent literature reported a moderate level of concordance between parent- and child reports. This means that potentially, both perspectives may provide complementary information.\textsuperscript{35,36} Another aspect of children's subjective perception regarding their functioning is the way children think about themselves.\textsuperscript{37} Harter has described two types of constructs: global sense of self-worth (also called self-esteem) and perceived competence in distinct domains of functioning, such as motor, autonomous or social functioning.\textsuperscript{38} Global self-worth emerges during development, and is thought to be based partly on perceived competence in areas that are particularly salient to the individual child in the context of his or her situation and development stage.\textsuperscript{39}
The main aims of paediatric rehabilitation medicine are to improve participation and HRQOL of children and adolescents with disabilities. Paediatric rehabilitation medicine programmes may have a direct impact on the level of body function and structure, and may result in a higher level of activities and participation by improving the body functions. Care-givers need tools to detect children at risk for limitations or a decrease in activities and participation, to predict future development and they need tools for setting realistic goals. Also parent and children want to know which expectations they should have about future functioning. So, information concerning the level and course of functioning and subjective experiences of the children and the relationships with disease characteristics, personal factors and environmental factors is indispensable in order to improve paediatric rehabilitation of children with CP.

The aims of the PERRIN CP 9-16 year study were to describe the course of the activities and participation and to determine its relationship with disease characteristics, personal factors and environmental factors in children and adolescents 9 to 16 years. The second aim was to investigate their self-esteem and get insight into their health-related quality of life.

**PERRIN CP 9-16 year study**

The contents of this thesis are the results of the PERRIN CP 9-16 year study. This study has been performed as part of the PERRIN (PEdiatric Rehabilitation Research in the Netherlands) research program. At the time of the start of this research program, little was known about the course of activities and participation and about determinants of future daily activities and social participation of children with CP. To describe the development of functioning in these domains, there was a need for instruments that reflected the activities and participation of children. Although some measures already existed, these had not been validated for the Dutch situation at that moment. Furthermore, little was known about the form and content of rehabilitation programmes and about team collaboration within rehabilitation setting in the Netherlands. So, the main aims of the PERRIN program were to develop instruments to measure the activities and participation of children with CP, to describe rehabilitation programmes in Dutch paediatric rehabilitation and to investigate the course and determinants of activities and participation, self-esteem and health-related quality of life of children with CP.

To investigate the course and determinants of activities and participation, self-esteem and health-related quality of life, four cohorts of children with CP were followed during 2-4 years: one cohort of children aged 1 and 2 years, one cohort aged 5 and 7 years, one cohort aged 9, 11 and 13 years (presented in this thesis) and one cohort aged 16 to 20 years. During the course of the studies, researchers of all cohorts have intensively collaborated and looked for corresponding instruments in order to use them in the different age cohorts.

The PERRIN CP 9-16 study was started in 2001 as the PERRIN CP 9-15 study. Two

---

*Supported by a grant from the Netherlands Organisation for Health Research and Development (grant no. 1435.0011).*

*Additionally supported by the Johanna Children's Fund and the Bio Child Rehabilitation Fund.*
yearly follow-up assessments were planned. Rehabilitation centres, special schools for physically and mentally disabled children (in the Netherlands called Mytyl and Tyltyl schools) and outpatient clinics of departments of rehabilitation medicine in the northwest region of the Netherlands identified 244 children with CP, aged 9, 11 and 13 years of age. Of these 244, 110 children and their parents participated in the study. In 2005 the PERRIN CP 9-15 year study was extended with an additional follow-up assessment, so the follow-up period became three years. The study name was therefore changed to PERRIN CP 9-16.

As part of their participation in the study, the children and their parents visited the Department of Rehabilitation Medicine of the VU University Medical Center each year for a period of three years (four measurements). Two weeks before the visit, the parents of the children received a questionnaire concerning HRQOL of the children (TNO-AZL Children's Quality of Life Questionnaire – parent form). During the visit, a researcher asked standardized questions about diagnosis, epilepsy, performed a physical examination and classified the children according to the Gross Motor Function Classification System (GMFCS). In the second part of the visit a trained researcher (physician / physiotherapist) administered the Gross Motor Function Measure (GMFM) to describe the level of motor function. The children who were able to self-report completed a questionnaire concerning HRQOL (TACQOL – child form) and a questionnaire concerning self-esteem (Harter's Social Perception Profile for Children). At the same time a trained researcher of the Department of Clinical Child and Family Studies from the Faculty of Psychology and Education investigated the level of motor, daily and social functioning and communication in a semi-structured interview with the parents, by scoring the Paediatric Evaluation of Disability Inventory (PEDI) and Vineland Adaptive Behavior Scales (VABS). During this session, the parents also completed a questionnaire on behavioural problems of the children (Child Behavior Checklist; CBCL).

Outline of this thesis

Chapter 2, 3 and 4 present the results of the level and course of functioning at different domains. Chapter 2 is based on a cross-sectional study on 9 to 13 year-old children and describes the level of activities and participation in the domains of mobility, self-care, daily living skills, social functioning and communication. Multivariate analyses were performed to investigate the relationship with personal and disease characteristics. Chapter 3 and 4 are based on longitudinal analyses of the course of functioning. In Chapter 3 the course of motor functioning over two years was analysed in relation to motor impairments and age. Chapter 4 describes the course of social functioning and communication over three years in relation to disease characteristics, personal and environmental factors. The level of social function and communication was compared to the US norm data.

In Chapter 5 the relationship between self-worth, perceived competence and CP was investigated. Our study group was compared with a Dutch norm group. Multivariate analyses were performed to analyse the relationship with severity of the CP and behavioural problems.
The focus of Chapter 6 was the HRQOL of children with CP. Firstly, the HRQOL of our study group was compared to a Dutch norm group. Secondly, the differences in reported HRQOL between the parents and the children were investigated. Thirdly, the course over time of both child-reported and parent-reported HRQOL was analysed in relation to gross motor functioning and behavioural problems.

Chapter 7 is the general discussion in which the main findings of the study are discussed, and clinical implications and recommendations for future research are provided.

References


