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INTRODUCTION

Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that 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.[1]

The Gross Motor Function Classification System (GMFCS)[2] is considered to be one of the most important scales for the measurement of function in CP. It was designed primarily to help clinicians prognosticate about the motor function of individuals with CP. The GMFCS consists of a 5-level scale with descriptors divided into four age bands: 0 to < 2 years, 2 to < 4 years, 4 to < 6 years, and 6 to 12 years. Children at Level I are relatively capable and by the time they reach 6 to 12 years old they can walk without limitations and can run and jump to some extent, with difficulties arising only with balance, speed, and coordination. At the other end of the scale, children at Level V will never achieve self-mobility unless they can learn to use a powered wheelchair with extensive adaptations. The GMFCS is based on self-initiated movements only, concentrating mostly on truncal control in sitting and functional mobility, taking into consideration everyday performance, rather than best capacity.

The 'Surveillance of Cerebral Palsy' in Europe (SCPE)[3] has classified CP into three main subtypes. These are *spastic*, *ataxic* and *dyskinesic* CP. Agreement has been reached on the clinical findings associated with each classification sub-group as follows:

Spastic CP is characterised by at least two of: 1) Abnormal pattern of posture and/or movement, 2) Increased tone (not necessarily constantly), 3) Pathological reflexes (hyper-reflexia or pyramidal signs e.g. Babinski response). It may be unilateral or bilateral.

Ataxic CP is characterised by both of 1) Abnormal pattern of posture and/or movement, 2) Loss of orderly muscular coordination, so that movements are performed with abnormal force, rhythm and accuracy.

Dyskinesic CP is characterised by both of 1) Abnormal pattern of posture and/or movement, 2) Involuntary, uncontrolled, recurring, occasionally stereotyped movements of affected body parts. Dyskinesic CP may be either a) Dystonic CP, dominated by both hypokinesia and hypertonus, b) Choreo-athetotic CP, dominated by both hyperkinesia and hypotonus.

As described above, the primary deficit in CP is an injury to the brain, not to the musculoskeletal or cardio-respiratory systems. However, the limitations in movement imposed by the brain lesion can have a marked secondary effect on these systems,[4] which may in turn become more debilitating for an individual with

CP than the direct effects of the initial injury. The secondary changes contribute to a vicious cycle whereby the disability leads to deconditioning that, in turn, worsens the level of disability.[5] Muscles need to be loaded adequately and frequently to maintain strength.[6] Similarly, in people with CP as in all people, the heart and lungs need to be exercised at moderately intense levels on a regular basis to maintain endurance and fitness. People with CP are already at a disadvantage with respect to achieving adequate levels of physical functioning because muscles and the cardio-respiratory system are not fully developed before the brain injury occurs and, therefore, are likely to have a lower starting point as well as slowed progress in developing these structures.[7]

It is becoming increasingly apparent that many of the adverse secondary changes may be preventable or reversible, although the extent to which this is possible has not been sufficiently explored or aggressively challenged. Management of motor disorder in CP is shifting gradually from the long-standing approach of alleviating impairments once they have occurred to a more proactive approach of promoting the type and degree of activity that may retard the development of muscle weakness. Alleviating the energy cost of performing typical daily activities is now a more frequently recognized and stated goal of motor interventions.[8-10]

Muscle strength and endurance training can be undertaken to improve performance of everyday activities. However, fitness implies a more generalized and greater degree of conditioning.

While health prevention and promotion efforts for persons with disabilities lag behind those for the non-disabled population, the potential benefits are enormous in terms of participation and health-related quality of life across the lifespan.

As stated in the revised 'Guide to Physical Therapist Practice' of the American Physical Therapy Association,[11] the role of the physical therapist is 'to restore, maintain, and promote not only optimal physical function, but optimal wellness and fitness and optimal quality of life as it relates to movement', which marks a significant expansion from its traditional mission. Strength, endurance, and fitness goals should be included in physical therapy treatment plans for the patients.

Importance of improving strength, endurance and fitness in CP

Historically, programs to promote physical fitness, including strengthening and cardio respiratory fitness exercise, were discouraged for patients with CP due to the concern that spasticity and abnormal movement patterns would worsen.[12] Therefore, traditional treatment of CP has focused primarily on attempting to improve abnormal motor patterns and maintain muscle length for daily activity and positioning. The historically used approaches failed to address adequately the ensuing muscle weakness, atrophy, and negative effects of diminished amounts and intensity of activity on the cardio-respiratory system. While these secondary

consequences have been recognized in CP for many decades, in the past, only a handful of proponents such as Winthrop Phelps[13] advocated physically intensive intervention. Scientific evidence has not supported the historically concern about the negative influence of physical fitness[14-16] and current research indicates that resistive exercise and endurance training is an effective intervention to improve strength, aerobic capacity and function in children and adolescents with CP.[6,17]
Documentation exists showing that individuals with CP have substantial generalized weakness.[18,19] Durstine and colleagues[5] described the circular process whereby persons with a chronic illness or disability experience less physical activity, which then leads to a cycle of deconditioning and further physical deterioration and reduction in activity (Figure 1). Many persons with CP lead a sedentary lifestyle with a decreased amount, variety, and intensity of physical activity.[4] Therefore, it is not surprising that also low levels of cardio-respiratory fitness have been reported.[20-24]

Improving the ability to walk or performing other functional activities are often the primary therapeutic goals for children with CP.[25] Because of existing impairments, many children and adolescents with CP have at least difficulty with activities such as walking independently, walking stairs, running or navigating safely over uneven terrain.[26] Additionally, children with CP have distinctly subnormal aerobic and anaerobic capacity in comparison with typically developing peers.[20,27,28]

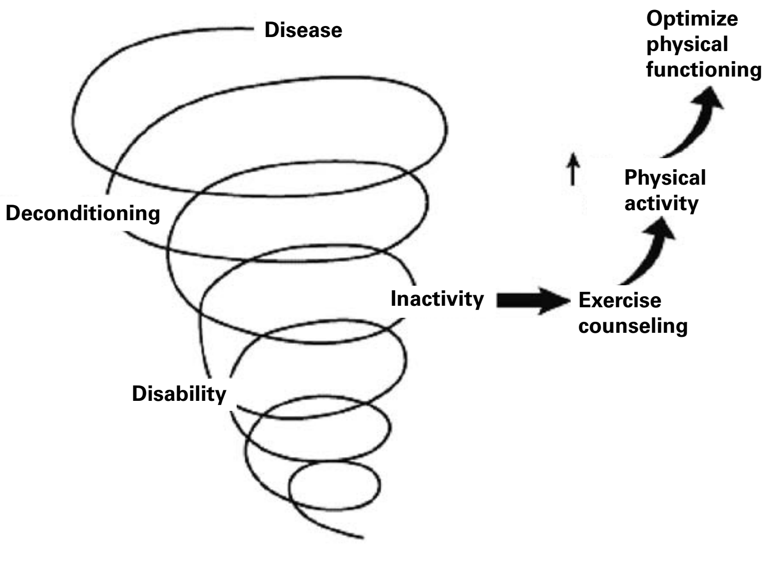

Figure 1. Cycle of deconditioning with physical inactivity. Disease can lead to inactivity and deconditioning. Deconditioning can lead to further inactivity and increase the potential for disability.

Also, muscle mass is low,[20] muscle strength is reduced[19,24,29] and energy cost of loco-motion is high.[30-32] Low levels on these fitness components may contribute to the difficulties in motor activities most children with CP encounter in daily life. More-over, evidence suggests that hypoactive children are more likely to become physically sedentary adults and that encouraging the development of physical activity habits in children will help us to establish activity patterns that continue into adulthood.[33]

The main question in this thesis is: "Do children with CP benefit from a fitness intervention program?" We focused on children that are classified at GMFCS level I and II. Due to the physical demands of the fitness tests and training exercises that were used throughout the study, recruitment solicited only children and adolescents who were classified at GMFCS level I (able to walk indoors and outdoors, and climb stairs without limitation) or level II (able to walk indoors and outdoors, and climb stairs holding onto a railing but experience limitations in walking on uneven surfaces and inclines, and walking in crowds or confined spaces).

The effects of this training program were considered by using the International Classification of Functioning, Disability and Health (ICF) framework for the description of health.[34] The ICF is applicable across cultures, age groups and sexes. In this framework, a person's disability can be considered in terms of impairments, activity limitations, and participation restrictions. The ICF classification served as a framework for the present study.

Principles of fitness training

Training to improve a child's performance obeys the three principles of training: specificity, overload and reversibility.[35,36]

Specificity

The application of the 'Specific Adaptation to Imposed Demands (SAID)' principle is essential in type of training.[37] Training programs are tailored to the exact demands of the activity and the individual needs of the child. This allows for specific gains to be made on the relative energy systems employed during training.

Specificity is an important principle in strength training, where the exercise must be specific to the type of strength required, and is therefore related to the particular demands of the event.

Overload

For training adaptations to occur the bodies systems must be overloaded beyond their normal levels. If these extra stresses are applied over a period of time the system will adapt and this becomes its new norm. During this adaptation-phase the bodies systems super or over compensate in order to cope with the next session. Training in this way along with sufficient recovery will allow for super compensation to occur thus resulting in an overall increase in fitness levels.

Reversibility or Detraining

During periods of exercise the human body makes adaptations to cope with the stresses placed on it. During periods of inactivity the human body will however reverse these adaptations in an attempt to return itself to a norm as this is the current level of stress placed upon it. Therefore gains that have been made will be lost.
Detraining is a de-conditioning process that affects performance due to the reduction in physiological capacity. During this period there is a loss of physiological adaptations associated with the training effect. Most training benefits are lost within a short period of stopping training. Most of the beneficial effects of training return to normal levels with 4-8 weeks dependent on the individual.

Measurements of fitness

Training effects are exercise mode specific.[38] Specificity of testing means that the modality of the testing tool needs to be similar to the type of activity the subjects train in. The outcome measures used in most studies in children with CP were not intervention-specific and often only focused on the International Classification of Function, Disability and Health (ICF) body function and activity level. However, to find results that are more exercise-related, intervention-specific tests should be used in future research. This may enhance the results of the studies and their interpretation. Since this study focuses on children that are able to walk independently (GMFCS I and II) we developed reliable and valid walking/running based exercise tests.

Age related physical activities

Recommended priorities for physical activities during childhood and adolescence relative to the development of skills and to behavioral, health, and fitness benefits are schematically illustrated in Figure 2. During the preschool and early school ages, general movement activities develop movement patterns and skills (dashed line in Figure 2). As these basic movements become established and skills improve, health, fitness, and behavioral components of physical activities increase in importance (solid line in Figure 2). Health related activities include those that emphasize cardiovascular and muscular endurance and muscular strength and those that involve weight bearing.

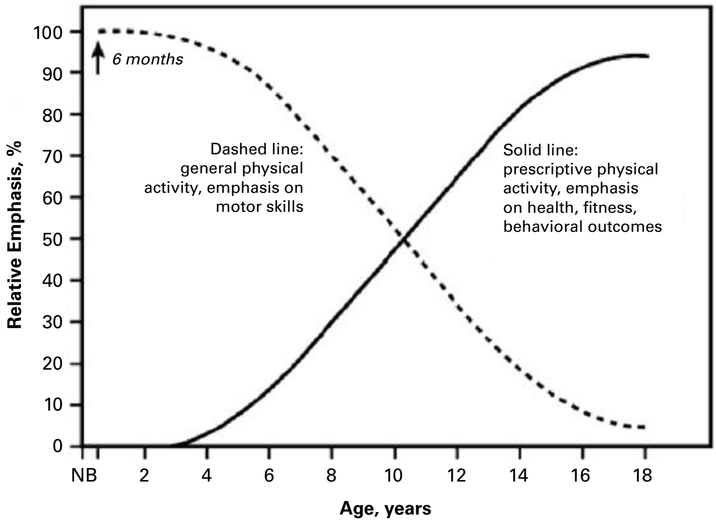

Figure 2. Changing emphasis of physical activity during childhood and adolescence.[39]

In this study we included children from 7 years of age and older, because we expected that children with CP of this age have established their motor patterns and skills, and are capable of increasing their fitness. Moreover, at this age children were expected to be able to understand and follow simple verbal commands.

Aim of this thesis

Based on the lack of exercise mode specific fitness-measures in children with CP, the first aims of this thesis are:

- Systematically review the literature regarding all types of exercise programs focusing on cardiovascular fitness (aerobic and anaerobic capacity) and/or lower extremity muscle strength in children with CP.
- To assess the psychometric qualities of measures for aerobic and anaerobic capacity that is specific for walking/running.
- To assess the psychometric qualities of a muscle strength measure that is exercise mode specific.

  Due to the scarce knowledge of the effects of a fitness intervention program in children and adolescents with CP, the second aim of this thesis is:
- To determine the effects of a functional fitness program in children and adolescents with CP not only on the domains of physical fitness, but also on gross motor function, self concept, participation and health-related quality of life.

Outline of this thesis

The first chapters address the assessment of fitness in children with CP. A review of the literature is presented in Chapter 2. In Chapter 3, a running based measure, the 10 m shuttle run test, is examined for its reliability and validity to measure aerobic capacity in children and adolescents with CP. In Chapter 4, two running based measures, the Muscle Power Sprint Test (MPST) and the 10x5 Meter Sprint Test, are being studied on its reliability and validity to measure respectively the mean and peak muscle power and agility. In Chapter 5, two ways to measure muscle strength are being studied. For Hand-Held Dynamometry the make-method and break-method are being compared regarding their reliability. Moreover, in this Chapter a new instrument (the 30-sec Repetition Maximum) to measure functional muscle strength is examined on its reliability.

The effects of a functional fitness program on aerobic and anaerobic capacity, agility, muscle strength, self-concept, gross motor function, participation and health-related quality of life are provided in Chapter 6. In Chapter 7, a summary and general discussion, focusing on the implications for research and clinical practice is presented.

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