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Knee contracture in children with cerebral palsy

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Knee contracture in children with cerebral palsy

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DOCTORAL DISSERTATION

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Docent Per Åstrand, The Karolinska Institute, Sweden

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Title and subtitle
Knee contracture in children with cerebral palsy

Abstract

Background: Lower limb contractures are a common problem among children with cerebral plasy (CP). Knee contractures affect both ambulant and non-ambulant children and may affect their ability to move, stand, walk and transfer, and increase the risk of asymmetric posture and further contracture development. To prevent severe contracture development it is crucial to identify children with reduced range of motion early and begin targeted treatment.

Purpose: To increase knowledge about the development of knee contractures; prevalence, risk factors, sequence of contracture development and the reliability of popliteal angle measurements.

Methods: Study I was a cross-sectional study of the total population of children with CP in Sweden. Data from the CPUP registry was used to identify factors associated with knee contracture. In study II, the interrater reliability of the unilateral- and bilateral popliteal angle tests were evaluated in children and youth with CP. Study III and IV were logitudinal cohort studies analysing the location and sequence of contracture development in the lower limb. Children at levels I-V of the Gross Motor Function Classification System (GMFCS) were included.

Results: Knee contractures occur in children at all GMFCS levels and the prevalence increases with lower motor function and higher age. Of all children with CP, 22% have a knee contracture. Knee contracture is strongly associated with short hamstring muscles. When measuring hamstring length in children with CP both unilateral and bilateral measurement of the popliteal angle are reliable.
While comparing hip, knee- and ankle contracture development, the ankle contracture occurs earliest in children with GMFCS level I or II and the knee contracture in children with GMFCS level III-V. Contracture developed in 34% of all legs and the median time to the first contracture was 10 years from first examination. Of legs with a primary contracture, 44% developed a second contracture. Children with a primary hip or ankle contracture were more likely to develop a second knee contracture. Children with a primary knee contracture developed either a hip or ankle contracture as a second contracture.

Conclusion: Knee contracture is a common problem for children with CP and occurs in children at all GMFCS levels. Knee contractures are associated with higher GMFCS level and are the first contractures to occur in children at GMFCS level III-V. A primary contracture affects the development of further contractures. Early interventions to prevent knee contractures should be considered.

Key words cerebral palsy, contracture, knee, ROM, musle length, reliability, children

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Play is the highest form of research

Albert Einstein

Table of Contents

Original papers

This thesis is based on the following original papers and are referred to in the text by their Roman numerals:

I. Cloodt E, Rosenblad A, Rodby-Bousquet E. Demographic and modifiable factors associated with knee contracture in children with cerebral palsy. Dev Med Child Neurol. 2018 Apr;60(4):391-396.

II. Cloodt E, Krasny J, Jozwiak M, Rodby-Bousquet E. Interrater reliability for unilateral and bilateral tests to measure the popliteal angle in children and youth with cerebral palsy. BMC Musculoskelet Disord. 2021 Mar 13;22(1):275.

III. Cloodt E, Wagner P, Lauge-Pedersen H, Rodby-Bousquet E. Knee and foot contracture occur earliest in children with cerebral palsy: a longitudinal analysis of 2,693 children. Acta Orthop. 2021 Apr;92(2):222-227.

IV. Cloodt E, Lindgren A, Lauge-Pedersen H, Rodby-Bousquet E. Sequence of flexion contracture development in the lower limb: a longitudinal analysis of 1,071 children with cerebral palsy. BMC Musculoskelet Disord. 2022 Jul 2;23(1):629.

Abbreviations

CI	Confidence Intervals
CP	Cerebral Palsy
CPUP	Cerebral Palsy Follow-Up Program and National Quality Registry
GMFCS	Gross Motor Function Classification System
GMFM	Gross Motor Function Measure
ICC	Intraclass Correlation Coefficient
KM	Kaplan Meier
OR	Odds Ratio
ROM	Range of Motion
SCPE	Surveillance of Cerebral Palsy in Europe
SD	Standard Deviation

Definitions

Ankle flexion contracture	Contracture of the ankle joint, placing the ankle in plantarflexion and preventing full dorsiflexion.
Cerebral palsy	A group of permanent disorders of the development of movement and posture, causing activity limitations which are attributed to non-progressive disturbances that occur in the developing fetal or infant brain. The motor disorders are often accompanied by disturbances of sensation, perception, cognition, communication, behaviour, epilepsy and secondary musculoskeletal problems [1,2].
Contracture	Reduced range of motion in a joint due to a permanent shortening of a muscle-tendon unit that occurs when soft tissue loses elasticity and cannot be stretched, either passively or by antagonistic muscles [3].
Hip flexion contracture	Contracture of the hip joint, placing the hip in flexion and preventing full extension.
Knee flexion contracture	Contracture of the knee joint, placing the knee in flexion and preventing full extension.
Spasticity	Disordered sensori-motor control, resulting from an upper motor neuron lesion, presenting as intermittent or sustained involuntary activation of muscles [4].

Thesis at a glance

Study	Questions	Methods	Results	Conclusions
I	What is the prevalence of knee contracture in children with CP and how are they associated with GMFCS level, age, sex, spasticity and muscle length?	Cross-sectional study of a total population of 3045 children with CP, born 2000-2014.	A knee contracture occurred in 22% of the children. The prevalence was higher in older children and in those with higher GMFCS level. Knee contractures were associated with reduced muscle length of the hamstrings.	Knee contracture is associated with higher GMFCS level, older age, and shorter muscle length; spasticity has a small effect. Maintaining muscle length, especially of the hamstrings, might be important for reducing the risk of knee contracture.
II	Are the unilateral and bilateral popliteal angle tests reliable measures when used in children and youths with CP?	Two independent physiotherapists examined 70 children and youths with CP, aged 5-22 years, at GMFCS level I-IV. Intraclass correlation coefficient (ICC) was calculated.	The interrater reliability was high for both measurements. The ICC values were 0.80 on the right and 0.86 on the left for the unilateral test and 0.82 on the right and 0.83 on the left for the bilateral test.	Both unilateral and bilateral measurement of the popliteal angle are reliable methods for estimating hamstrings length.
III	Which lower limb contracture occur earliest in children with CP?	Longitudinal cohort study of 2693 children with CP born 1990-2018 and registered in CPUP before 5 years of age.	A contracture developed in 34% of all legs and the prevalence increased with higher age and GMFCS level. The first contracture to occur in children with GMFCS level I or II was an ankle contracture, and a knee contracture was the first contracture to occur in GMFCS level III to V.	Early interventions to prevent knee and ankle contractures in children with CP should be considered.
IV	How is a primary hip, knee or ankle contracture associated with the time to and sequence of contracture development in adjacent joints?	Longitudinal cohort study of 1071 children with CP born 1990-2018, registered in CPUP before 5 years of age with a primary contracture of the hip, knee or ankle.	A second contracture developed in 44% of the legs. The frequency of multiple contractures increased with higher GMFCS level. Children with a primary hip or ankle contracture were more likely to develop a second knee contracture. Children with a primary knee contracture developed either a hip or ankle contracture as a second contracture.	Multiple contractures were associated with higher GMFCS level. Lower limb contractures appeared in specific patterns where the location of the primary contracture and GMFCS level were associated with contracture development in adjacent joints.

Populärvetenskaplig sammanfattning

Cerebral pares (CP) är ett samlingsnamn för olika typer av motorisk funktionsnedsättning. CP beror på en skada i hjärnan som inträffar antingen i fosterstadiet, vid födseln eller innan två års ålder. CP är den vanligaste orsaken till rörelsenedsättningen hos barn och ungdomar och varje år föds cirka 200 barn i Sverige som får diagnosen CP. Symptomen varierar från knappt märkbara till mycket begränsande för individen och innebär bland annat nedsatt rörelseförmåga och nedsatt förmåga att stabilisera kroppen.

Barn med CP har mindre och svagare muskler än andra barn. Muskeltillväxten är försämrad vilket gör att musklerna inte växer i samma takt som skelettet. För att kompensera för detta blir muskelsenorna istället längre och fibrerna inuti muskeln utdragna. Trots detta är det mycket vanligt att barn och vuxna med CP får en sådan muskelförkortning att de inte längre har full rörlighet i sina leder. Detta kallas kontraktur. En av de vanligaste kontrakturerna bland personer med CP är knäkontraktur som innebär att man inte kan sträcka ut knät till rakt läge. Detta påverkar både de som kan gå och de som inte är gångare, försämrar gångmönstret, gör det svårt att klara förflyttningar och gör det svårt att ligga och stå rakt.

Syftet med denna avhandling var att studera hur vanligt det är med knäkontrakturer hos barn med CP, vilka faktorer som kan kopplas till förekomsten, utvärdera två metoder att mäta muskellängd på lårets baksida (hamstringsmuskeln) och undersöka var och i vilken ordning kontrakturer i höft, knä och fotled uppstår.

Sedan 1994 finns ett uppföljningsprogram för barn med CP i Sverige (CPUP). Sedan 2005 är CPUP också ett nationellt kvalitetsregister och från 2009 inkluderas även vuxna med CP i programmet. Syftet med programmet är att följa individer systematiskt med mätningar av ledrörlighet, röntgen och andra undersökningar för att kunna ge förebyggande insatser i rätt tid om det upptäcks försämring. Personer med CP kan beskrivas utifrån sin grovmotoriska förmåga med hjälp av The Gross Motor Function Classification System (GMFCS) som är en femgradig skala baserad på förmågan att sitta, stå och förflytta sig. Personer i nivå I kan sitta och gå själv utan hjälpmedel medan personer i nivå V inte kan sitta utan stöd och har stora svårigheter att förflytta sig även med hjälpmedel.

I studie I undersöktes förekomsten av knäkontrakturer och eventuella samband med ålder, kön, GMFCS, muskellängd och spasticitet hos samtliga barn inkluderade i CPUP. I studie II testades två sätt att mäta längden på muskeln på lårets baksida (hamstringsvinkeln) för att se hur väl mätresultaten stämde överens mellan två oberoende bedömare. I studie III och IV följdes barn med CP över tid för att undersöka vilken led som först drabbades av en kontraktur och hur många barn som sen fick fler kontrakturer och i vilka leder.

Resultaten av studierna visade att knäkontrakturer förekom hos barn på samtliga GMFCS nivåer och att totalt 22% av alla barn med CP hade en knäkontraktur. Sannolikheten ökade med svårare grad av CP (högre GMFCS nivå) och med högre ålder. Barn med försämrad hamstringsvinkel hade särskilt stor risk att ha knäkontrakur och en hamstringsvinkel under 120 grader tio-dubblade sannolikheten för knäkontraktur medan spasticitet hade ett svagt samband med förekomst av knäkontraktur. Både unilateral och bilateral mätning av hamstringslängden är reliabla metoder, dvs tillförlitliga mätmetoder som visar god överensstämmelse mellan olika bedömare. Hos barn på GMFCS nivå I och II var det vanligast att den första kontrakturen uppstod i fotleden. Hos barn på GMFCS nivå III till V var det vanligast att första kontrakturen uppstod i knäleden. Den första kontrakturen uppstod efter en mediantid på 10 år från första bedömning. Totalt fick 34% av barnen en kontraktur i antingen höft, knä eller fotled. Av dessa var det 44% som sen fick ännu en kontraktur. Det var vanligast att få den andra kontrakturen i en intilliggande led till den först drabbade.

SLUTSATSER:

Knäkontrakur är vanligt hos barn med CP och förekommer hos barn på alla GMFCS nivåer. Förekomsten ökar med svårighetsgrad av CP och med ålder. En hamstringsvinkel under 120 grader tio-dubblar sannolikheten för knäkontraktur. Spasticitet i lår- eller vadmuskel har ett svagt samband med förekomsten av knäkontraktur.

Unilateral och bilateral mätning av hamstringslängd visar god reliabilitet.

Det är vanligast för barn på GMFCS nivå I och II att få en fotledskontraktur först, medan barn på GMFCS nivå III-V oftast får en knäkontraktur först.

Kontrakturer i nedre extremitet uppstår ofta tidigt. Tidigare insatser för att förebygga knä- och fotledskontrakturer bör övervägas. Insatserna bör anpassas efter barnets GMFCS nivå.

“Having cerebral palsy can be difficult sometimes. That is something you don’t always understand. I´m not as fast as the other children and everyone else can ride a bike. I notice this a lot. In school they have this rule; you must carry objects with two hands in the stairs. But I have to hold on to the rail with one hand when walking in stairs. Therefore, others must carry my belongings. I think about situations like this a lot. I think you should also think more about it. Children’s thoughts are important!”

Smilla, 10 years

Introduction

History of cerebral palsy

Cerebral palsy (CP) is the umbrella term for a group of brain disorders affecting motor function, movement and posture. CP is caused by an injury to the developing brain during pregnancy, at birth or before two years of age. CP is a permanent, but not degenerative, disorder [2].

CP was first described in 1862 by an English orthopaedic surgeon, William Little. He described children with spasticity and stiff extremities and understood that there was a connection between the neurological system and the muscle deformities. He described contractures and deformities as a result of the brain injury. He stated that CP was caused by difficult birth and classified the children based on their clinical symptoms into hemiplegic rigidity (one side affected), paraplegic rigidity (legs more affected than the arms) and generalized rigidity [5,6]. Little, however, never used the term “cerebral palsy” himself. The first recorded use of the term was by William Osler in 1888. Sigmund Freud (1856-1939) stated that CP could be caused by abnormal brain development even before birth. He also linked changes to the brain with types of paresis. In the early 1920s the American orthopaedic surgeon, Winthrop Phelps made an important contribution to the understanding of CP when advocating physical therapy, orthoses and nerve blocks as management of CP and being conservative to surgery. He grouped all movement disorders under the term dyskinesia and classified spasticity, athetosis, dyskinesia, ataxia and tremor as subtypes [7,8].

Definition

In 1959 Mac Keith et al. published a definition of CP as “a permanent but not unchanging disorder of movement and posture, appearing in the early years of life due to a non-progressive disorder of the brain, the result of interference during its development” [8]. In 1992 Mutch et al. defined CP as “an umbrella term covering a group of non-progressive but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stage of development” [7]. The most common definition of CP today is from Rosenbaum and colleagues [2] from 2006; “Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitations, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, epilepsy, and by secondary musculoskeletal problems".

The surveillance of Cerebral Palsy in Europe (SCPE), a collaboration of CP registers and surveys, have created a decision flow chart for clinicians with standardized inclusion and exclusion criteria of CP (Figure 1)[9,10].

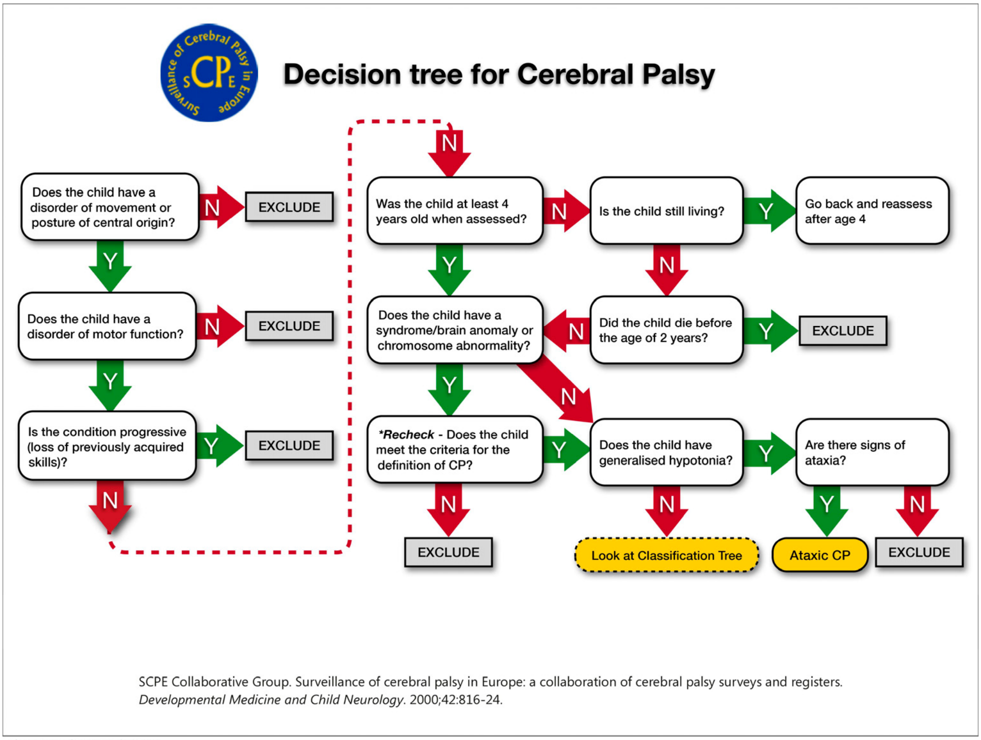

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Figure 1. The SCPE decision tree for inclusion or exclusion of cases of CP. The figure is reprinted with permission from Developmental Medicine and Child Neurology 2000;42:816-24 [10].

Prevalence

CP is the most common motor disability in children [10] with a prevalence of 1.7/1000 live births in Sweden[11] and 1.77/1000 live births in Europe [12]. Worldwide, over 17 million people live with CP and approximately 200 children in Sweden are diagnosed with CP every year. The prevalence is highest among children born before 28 weeks' gestation [13]. A decrease in the birth prevalence of CP has been reported from high income populations during the first decades of the 21st century [14,15,11]. The reasons for this are multifactorial, including a decline in maternal smoking, children born post-term and improved neonatal care [16]. CP is a lifelong condition and the estimated life expectancy has increased. For individuals with a mild form of CP the estimated life expectancy is similar to that of the general population [17].

Classifications of subtypes

The characteristics of CP depend on the location of the brain damage and the neurological symptoms. Spastic CP is the most common form.

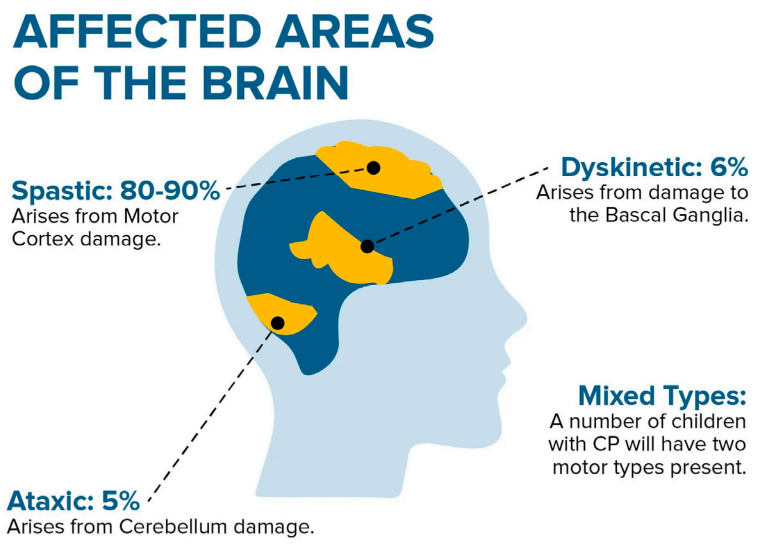


Figure 2. Location of brain damage and CP types. The figure is reprinted with permission from the United Cerebral Palsy Association of Hawaii.

Since the first description of CP subtypes were made by Little, several different classification systems of subtypes have been used. The Swedish classification by Hagberg was used for many years in Sweden [18]. Hagberg categorized CP into spastic hemiplegia, spastic diplegia, spastic tetraplegia, ataxic CP (divided into ataxic diplegia or congenital ataxia), dyskinetic CP (divided into dystonic CP or choreoathetotic CP) and mixed form. Since the beginning of the 21st century, registers and databases for children and adults with CP, have used the SCPE classification with subtypes divided into spastic unilateral, spastic bilateral, ataxic, dyskinetic and non-classifiable CP (Figure 3)[9,10]. The SCPE classification has been used in this thesis.

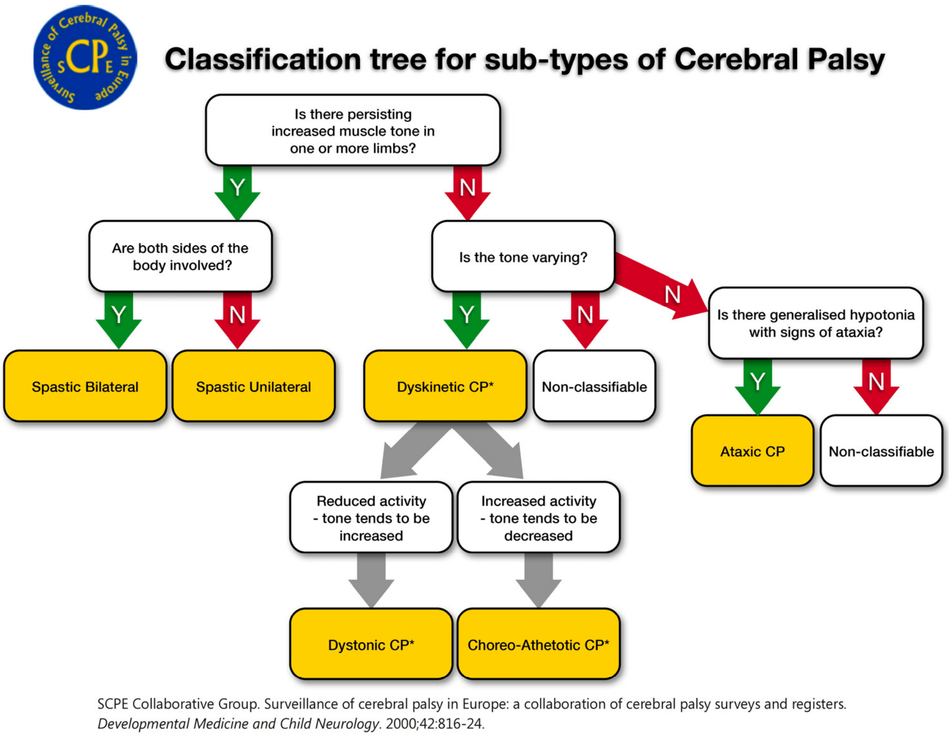


Figure 3. The SCPE hierachical classification tree of CP subtypes. The figure is reprinted with permission from Developmental Medicine and Child Neurology 2000;42:816-24 [10].

Classification of gross motor function

In addition to the classification of subtypes, children with CP are often described by their functional performance with a classification developed by Palisano et al. [19] known as the Gross Motor Function Classification System (GMFCS) (Figure 4). GMFCS is a 5-level classification system describing the child's self-initiated mobility such as sitting and transfer, and the use of assistive devices such as walkers and wheelchairs at different ages. Level I describes the highest level of function and level V the lowest. The GMFCS level tends to remain stable over time [20]. The GMFCS helps clinicians to objectively classify children with CP, predict future function, design treatment interventions and translate research into practice. The classification system has a high interrater reliability and validity, good content validity and high test-retest reliability[19,21].

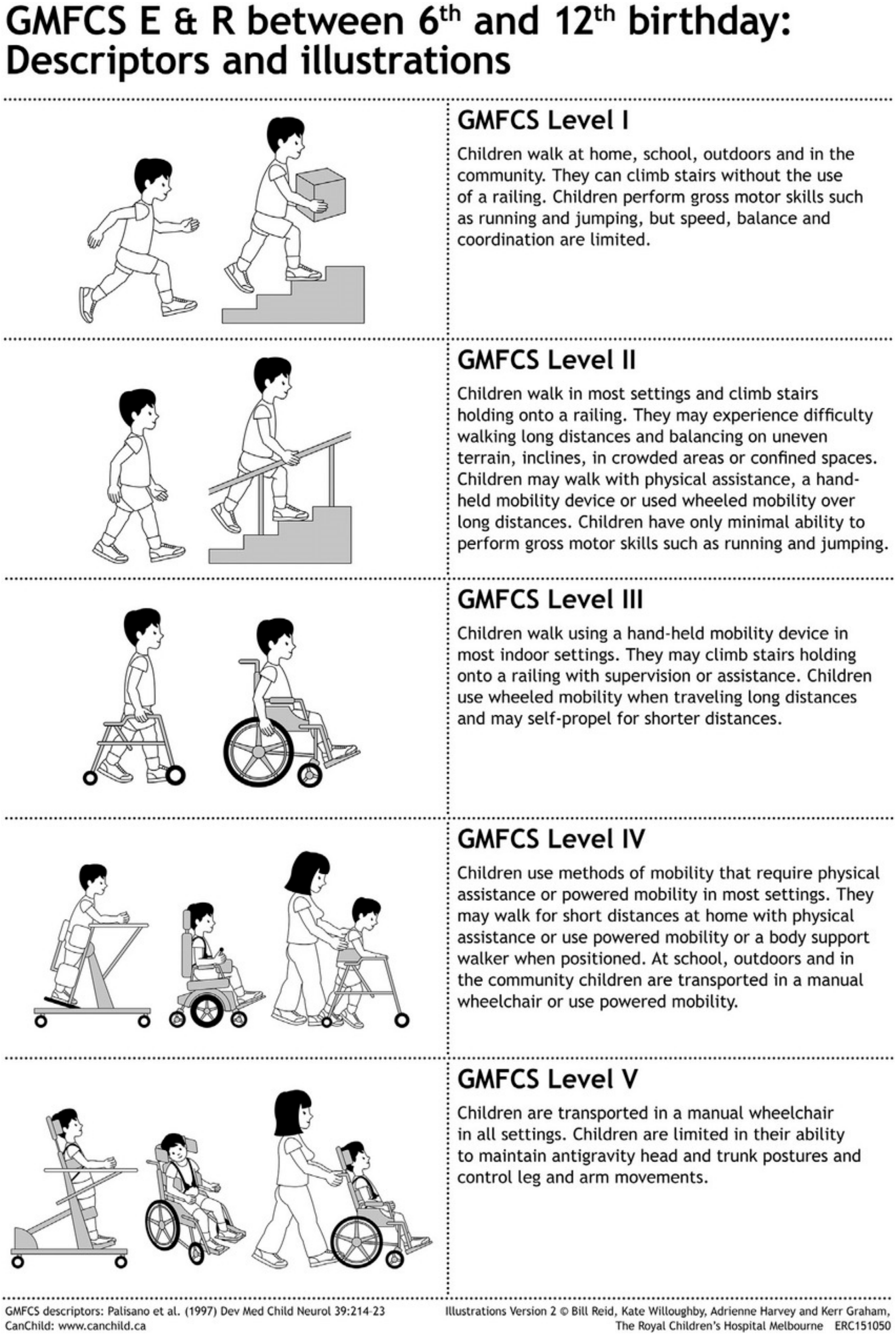


Figure 4. The Gross Motor Classification System (GMFCS), level I to V for ages 6-12 years. The figure is reprinted with permission from Professor H. Kerr Graham, Royal Children's Hospital in Melbourne.

The muscle in cerebral palsy

Muscles are essential for our life and for everything we do. The interest for, and the understanding of, the skeletal muscle in CP has increased during the last years. Until recently, the skeletal muscle was seen as an "end organ" affected secondary to the neurological impairment. Instead, a lot of focus and research has been on spasticity and spasticity reducing treatment to prevent secondary complications. The latest research has now challenged the role of spasticity in contracture development and showed that spasticity and contractures develop parallel with each other rather than as a result [22-24]. Treatment of spasticity has not managed to significantly prevent contractures and deformities [25,26].

Muscle pathology

The skeletal muscle in children with CP differs from skeletal muscles of children without CP [27,28]. Children with CP have reduced muscle size and studies have estimated a reduction of 20-40 % compared to children without CP [28-31]. In general, the muscles are shorter and smaller in cross-sectional area and thickness but have longer tendons [28]. The reduced muscle growth can be seen already in toddlers with CP and the level of reduction increases with lower levels of motor function (higher GMFCS levels) [32]. It is not known if the reduced muscle size is a result primarily of atrophy or an inability to grow [33]. The link between the neurological lesion and the cellular muscle pathology is not fully understood, and muscle size is not equally affected in all individuals with CP [29].

A skeletal muscle consists of bundles of muscle fibres. Each muscle fibre consists of myofibrils that are composed of thousands of sarcomeres in series. In children without CP the sarcomere overlaps to optimize muscle function. In children with CP, the sarcomeres are significantly less in number and longer in length [34], with a non-optimal overlapping which reduces the muscle's ability to generate force and speed (Figure 5) [22]. Even though the muscle is shorter than in children without CP, the sarcomeres are highly stretched, sometimes beyond their normal operating length [34]. Functionally, long sarcomeres generate very low active tension which results in a weak muscle [35].

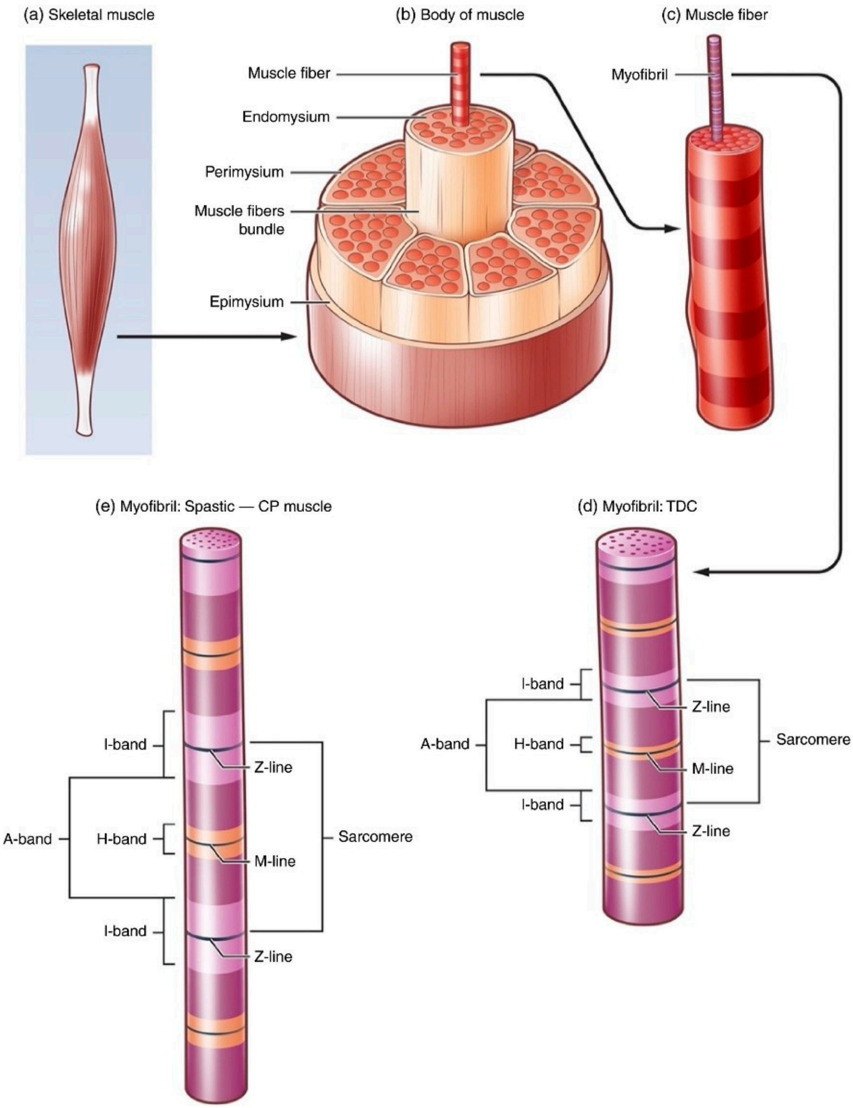


Figure 5. Skeletal muscle hierarchy. (a) In children with cerebral palsy (CP) the ratio of belly to tendons and the muscle cross-sectional area is significantly reduced compared to children without CP. (b) Muscle belly divided into bundles of muscle fibers surrounded by collagen-based epimysium, perimysium and endomysium. (c) Each muscle fiber is comprised by myofibrils, contractile organelles made of serial arranged sarcomeres. (d) In TDC, actin and myosin filaments overlap to optimize the sarcomere length. (e) In CP, sarcomeres are overly long with sub-optimal actin-myosin overlap and reduced force genreating potential[22]. The figure is reprinted with permisson from Professor H. Kerr Graham, Royal Children's Hospital in Melbourne**.**

The skeletal muscle in CP also contains less satellite cells compared to children without CP. The satellite cells are responsible for muscle growth and for repairing the muscle [36]. Biopsies from hamstrings muscles from individuals with CP show less than half as many satellite cells compared to children without CP. The decreased number of satellite cells might be a cause behind decreased longitudinal growth of muscles in CP that leads to fixed contractures and reduced ability to strengthen the muscles [37].

Knee contracture

Contractures are defined as reduced range of motion in a joint due to a permanent shortening of a muscle-tendon unit that occurs when soft tissue loses elasticity and cannot be stretched, either passively or by antagonistic muscles [3]. Contractures are a common problem for children with CP and one of the most discussed and challenging tasks in clinical settings [3,38].

A contracture in the knee joint prevents the knee from reaching full extension and forces the knee into a permanent flexed position. Therefore, knee contractures are often referred to as a "knee flexion contracture". Knee extension contractures, when the child cannot bend their knee to full flexion also occur but are much less common than the knee flexion contractures described in this thesis. From now on, a knee flexion contracture will be referred to as a "knee contracture".

Clinical implication

A contracture can involve ligaments, capsule, tendons and muscles and arises when the structures develop limited extensibility and increased stiffness [39]. The main symptom of a contracture is the limited ability to move a joint but contractures can also cause secondary symptoms[40].

The ability to move is an important activity in daily life. Decreased mobility and contractures may lead to limited levels of activity and difficulties affecting participation [41-44].

Ambulant children

For ambulant children a knee contracture primarily affects their gait pattern. Differences in gait patterns in persons with CP has been a cause for attention both in clinic and in research[40]. The most common gait abnormalities of the knee occur in the sagittal plane [45,46]. A knee contracture can lead to decreased walking speed, stride and step length, higher energy cost during walk and fatigue [47,48]. Stronger forces on the knee joint can also lead to pain [49]. The most common gait pattern associated with the knee is the crouch gait (also referred to as flexed knee gait). Crouch gait is defined as excessive flexion of the knee with increased hip flexion and/or ankle dorsiflexion throughout the stance phase of gait [45]. Crouch gait can be categorized in severity levels based on the degrees of knee flexion: mild 20-30°, moderate 30-40° and severe ≥40° [50]. The causes behind crouch gait are multifactorial including muscle weakness of ankle plantar flexors or knee extensors, spasticity or contracture of hip or knee flexors, lever arm dysfunction or a combination. Short hamstrings muscle has been seen as the primary cause of crouch gait and most treatments for crouch gait focus on surgical lengthening of hamstrings. However, other studies have shown that many persons with crouch gait have normal hamstrings length and that an increased shortening of the hamstring length seems to occur secondary to walking in crouch rather than contributing to the development of the gait pattern [51,52]. On the other hand, a knee contracture of -10 degrees of extension or worse might have a major effect on development of crouch gait [53]. Tight hip flexors [54] and weakness in gastrosoleus muscles [55] also seem to be important contributors to crouch gait.

Non-ambulant children

For non-ambulant children a knee contracture impacts both the ability to stand and the ability to transfer from sit to stand [44,56]. A knee contracture is also a predictor for scoliosis which mainly affects children at GMFCS level IV and V [57]. In supine lying, limited knee extension may cause the legs to tilt to one side resulting in a rotation of the hips and pelvis which can affect the spine. Ágústsson et al. [58] showed that adults with CP who were immobile and spend long periods in a lying position (>8 hours per day) had a higher likelihood for scoliosis and windswept hip deformities. Furthermore, knee and hip contractures are associated with postural asymmetries in both sitting and supine positions [59]. Children at higher GMFCS levels spend more time in sitting and lying. Both contractures and postural asymmetries are associated with pain [59] and pain is a common problem for children with CP, specially at GMFCS level V [60].

Management

Knee contractures are challenging to treat. The management normally focuses on increasing hamstrings length [61,62]. Non-operative treatment includes orthoses, serial casting, botulinum toxin injections and strength training [63-65]. However, the evidence for those treatments is limited. Using knee orthoses during night can be difficult for the child to tolerate and soft versions are often preferred. Even with the soft version, few families are able to maintain the treatment for more than 12 weeks in a row. Children with less severe knee contractures seem to tolerate the orthoses better [66]. Botulinum toxin injections to reduce spasticity of the hamstring muscles can provide temporary benefits for young children but are not as helpful for older children with fixed contractures [67]. Serial casting of the knee is also used as an initial treatment for knee contractures [68]. Although there are risks of skin break down and nerve stretch associated with the casting, it can in some cases successfully reduce resistant contractures and stretch the posterior capsule [66]. Serial casting is most likely to be a successful treatment in children under 12 years of age and for children with mild contractures [68]. The evidence of strength training to prevent or treat knee contractures and improve hamstrings length is limited [63]. Limiting the time in sitting and lying with flexed knees and increase and support standing for children at higher GMFCS levels are also part of the prevention and treatment of knee contractures [66].

Surgical treatments to improve knee extension and hamstrings length include hamstring lengthening, distal femoral extension osteotomy, distal tenotomy, hamstring transfer, rectus femoris transfer, patella tendon shortening, guided growth and knee capsulotomy [38]. Hamstring lengthening can improve knee extension and gait [38] but should be combined with strength training, orthoses and reduced time spent with flexed knees during the day [66]. The risk for recurrence is highest for non-ambulant children, children at older age at the time for surgery and for children without additional treatment or follow up [66,69].

CPUP: The follow-up program for cerebral palsy

The cerebral palsy follow-up program (CPUP) is a national surveillance program for children and adults with CP in Sweden. The program was initiated in 1994 as a collaboration between the orthopaedic departments and habilitation centres in southern Sweden. The aim of starting the program was to prevent the large number of hip displacements, contractures and deformities seen in children with CP. In 2005, the Swedish CPUP was certified as a national health care quality registry. Today, the program has a coverage rate of over 95% of all children with CP in Sweden, representing all 21 health care regions with a total population of 10.4 million inhabitants [70]. Since 2009, CPUP also includes adults with CP, and at present, over 2500 adults are followed in the program [71].

The priorities of CPUP are to prevent hip dislocations, severe contractures, scoliosis and other deformities with the goal of reducing pain and increasing quality of life for individuals with CP. The program also aims to describe the function and development of individuals with CP, evaluate treatment, increase knowledge about the diagnosis and improve collaboration between professionals and between professionals and families. The National Healthcare Quality Registry enables quality control and research [70,72].

The systematic follow-up through CPUP has reduced the incidence of hip dislocations from 8% to 0.5% in Sweden [73]. The prevalence of scoliosis, windswept deformities[74] and severe contractures has also decreased significantly since the start of the program and the need for surgery to treat severe contractures has been reduced [75].

Other countries have been inspired by CPUP and started similar follow-up programs such as Norway (2006), Denmark (2010), Scotland (2010), parts of Iceland and New South Wales in Australia (2012) and the Netherlands (2018).

Children with suspected CP are included in the program as early as possible. The diagnosis is verified by a neuropaediatrician from the age of 4 years. Children who do not fulfil the criteria for CP leave the program. Inclusion and exclusion criteria are in accordance with those identified by the SCPE (Figure 1) [9,10].

The clinical examination in CPUP

In CPUP, all children are followed with regular examinations by their local multi-professional habilitation team. The examination includes gross- and fine motor function, dominant neurological symptoms, measurements of ROM, spasticity assessment, spinal assessment, posture, mobility, physical activity, pain and also standardized radiographic examinations of the hip and spine. The received treatments are also reported such as surgeries, use of orthoses, botulinum toxin injections, serial casting, physiotherapy. The result of the examination is reported into the CPUP database [70]. The examiners follow standardized protocols (https://cpup.se/) and the frequency of examinations depends on the child's age and GMFCS level (Figure 6).

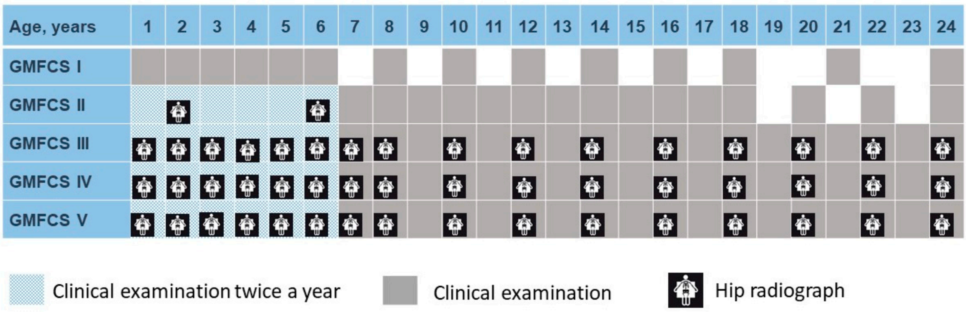


Figure 6. Guidelines for clinical examinations and hip radiograph within the Swedish Cerebral Palsy Follow-Up Program (CPUP).

Measuring range of motion (ROM)

The routine for measuring passive ROM in clinic is the goniometric measurement.

In CPUP, when measuring ROM of the hip extension, knee extension and ankle dorsiflexion the child is placed in a supine position on the examination table. When measuring hip extension, the tested leg is placed in extension outside the examination table. The contralateral leg is kept in hip and knee flexion (Figure 7).

Knee extension is measured with the hip and knee extended and ankle dorsiflexion is measured with the knee extended (Figure 8).

Figure 7. Hip extension measurement.

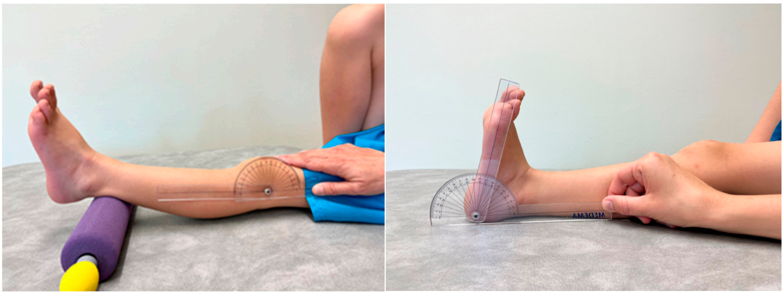


Figure 8. Knee extension measurement (left) and ankle dorsi flexion measurement (right).

The Modified Ashworth scale

To examine the spasticity level, CPUP uses the Modified Ashworth Scale [76]. The child is placed in a supine position. When testing a muscle that primarily flexes a joint; the examiner places the joint in maximum of flexion and then move to a position of maximal extension in one second. The examiner scores the muscle tone based on the following classification:

- **0** No increase in muscle tone.
- **1** Slight increase in muscle tone, manifested by a catch and release or by minimal resistance at the end of the ROM when the affected part(s) is moved in flexion or in extension.
- **1+** Slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the ROM.
- **2** More marked increase in muscle tone through most of the ROM, but affected parts(s) easily moved.
- **3** Considerable increase in muscle tone, passive movement is difficult.
- **4** Affected part(s) rigid in flexion or extension.

The popliteal angle measurement

The popliteal angle test aims to measure the passive length of the hamstring muscle. This method is frequently used in clinic. In order to measure the ROM of the knee joint, the examiner places the child in a supine position at the examination table and measures knee extension with the hip in 90˚flexion. There are different ways of performing the test, however, the unilateral and bilateral measurements are the most common [77]. For the unilateral test, the contralateral leg is placed and fixed in a fully extended position compared to the bilateral test where the contralateral leg is flexed at the hip to a position in which the anterior superior iliac spine and the posterior superior iliac spine are vertical (Figure 9).

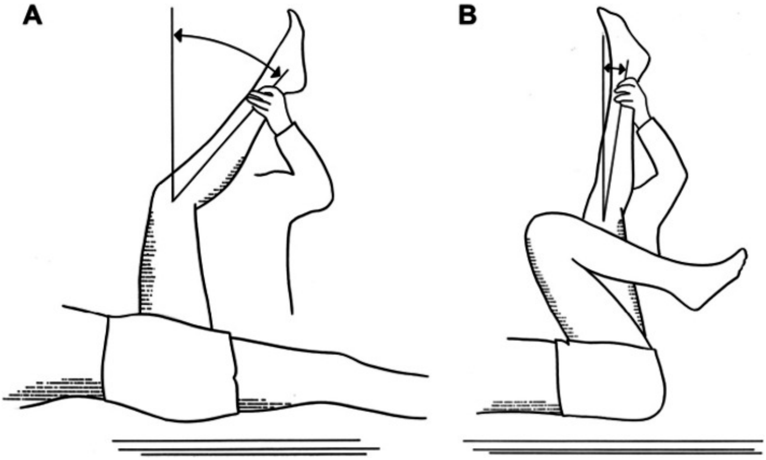


Figure 9. Tests to measure the popliteal angle. (a) The unilateral popliteal angle is measured with typical lordosis, contralateral hip extended, and the ipsilateral hip flexed to 90°. (b) Bilateral popliteal angle is measured with the contralateral hip flexed until the anterior superior iliac spine and the posterior superior iliac spine are vertical. Reprinted with permission from Identification and Treatment of Gait Problems in Cerebral Palsy, 2nd edition, by James R. Gage, Michael H. Schwartz, Steven E. Koop and Tom F. Novacheck, published by Mac Keith Press (www.mackeith.co.uk) in its Clinics in Developmental Medicine Series, 2009, 9781898683650.

The aims of this thesis

The overall aim of this thesis was to increase knowledge about the development of knee contractures; determine the prevalence, analyse risk factors, follow the time to and sequence of contracture development in the lower limb and evaluate the reliability of popliteal angle measurements in children with CP.

Study I To determine the prevalence of knee contracture and its association with GMFCS level, age, sex, spasticity and muscle length in children with CP.

Study II To evaluate the interrater reliability of unilateral and bilateral tests to measure the popliteal angle when used in children and youth with CP.

Study III To analyse whether contracture preventing hip extension, knee extension, or ankle dorsiflexion occurs first in children with CP at GMFCS level I to V.

Study IV To analyze if and how a hip, knee or ankle contracture affects the time to and sequence of contracture development in adjacent joints in children with CP at GMFCS levels I–V.

Materials and methods

Study design

Study I was a cross sectional study of children with CP followed by the CP follow-up program, analysing factors associated with knee contracture. Study II was a reliability study evaluating the interrater reliability of the unilateral and bilateral measurement of the popliteal angle. Study III and IV were longitudinal cohort studies analysing the occurrence and location of the first and following lower limb contractures in children with CP.

Participants and methods

Table 1. Participants in study I-IV.

Demographics	Study I (%)	Study II (%)	Study III (%)	Study IV (%)
Participants	3045	70	2693 (4751 legs)	1071
Age range	1-15	5-22	0-30	0-30
Male/female	1756/1289	38/32	1598/1095	636/435
GMFCS I	1330 (44)	17 (24)	1901 legs (40)	185 (17)
GMFCS II	508 (17)	31 (44)	765 legs (16)	206 (19)
GMFCS III	280 (9)	12 (17)	540 legs (11)	156 (15)
GMFCS IV	449 (15)	10 (14)	752 legs (16)	234 (22)
GMFCS V	478 (16)	0 (0)	793 legs (17)	290 (27)

Note: The sum of all percentages may not total 100% due to rounding

Study I

Data for all 3045 children at GMFCS level I-V with CP aged 1-15 years from the Swedish CP registry born between 2000 and 2014 was included. The latest physiotherapy examination for each child performed in 2014-2015 was extracted and used for analysis. Passive knee extension was analysed according to age, sex, GMFCS level, spasticity of the knee and plantar flexors and the length of the gastrocnemius and hamstring muscles. Passive ROM of the knee, ankle and popliteal angle was measured with goniometric measurement according to the standardized protocol in CPUP (https://cpup.se/in-english/manuals-and-evaluation-forms/). A knee contracture was classified as a mild contracture if the extension was -5˚ to -14˚ and as severe if the extension was -15˚ or worse. The hamstrings length was measured as the popliteal angle with the hip of the examined leg in 90˚ flexion and the contralateral leg fully extended. We classified a mildly reduced hamstrings length as a popliteal angle of 120˚ to 139˚ and a severely reduced hamstrings length as below 120˚. The gastrocnemius length was classified by measuring the ankle dorsiflexion with the hip and knee extended. A dorsiflexion of 9˚ to 0˚ were considered a mildly reduced muscle length and <0˚ as severely reduced. Muscle tone in the hamstring muscle and the gastrocnemius muscle was measured according to the Modified Ashworth Scale [76].

Study II

Measurements of the unilateral and bilateral test in 70 children and youth, age 5-22 years, with CP were analysed. The children were recruited at a rehabilitation centre in Poznan, Poland, between 10 June and 2 September 2019. Children with unilateral CP were excluded to ensure that no unaffected legs were analysed. All measurements were performed by two independent examiners on the same day for each child. Both examiners were recently graduated physiotherapists without previous experience of measuring children with CP. The test was performed according to figure 9.

Study III

The study included all available measurements from the start of the CP follow-up program in 1994 until the end of December 2018. Data for 2,693 children with a first examination in CPUP before 5 years of age were included in study III. The children were followed for an average of 5 years (range 0-24) during a study period of 24 years with 27,230 measurement occasions. ROM was measured as in study I. Repeated measures of passive hip extension, knee extension and ankle dorsiflexion were followed to analyse in which joint the first contracture occurred and when. A contracture was defined as a hip-, knee- or plantarflexion contracture of 10˚ or more. Legs that underwent soft tissue- or bony surgery and children with intrathecal baclofen pump (ITB) or who underwent selective dorsal rhizotomy (SDR) before the onset of the first contracture were excluded from further analysis at the date of surgery. For children with unilateral CP, only the affected leg was followed in the analysis. Children with surgery on the upper extremity or the spine, fractures, serial casting or botulinum toxin injections were included in the analysis. The frequency and location of additional contracture development was recorded. In total 1071 children born between 1990 and 2018 and registered in CPUP before the age of 5 years were included.

Study IV

Study IV included all available measurements of children with a lower limb contracture in the hip, knee or ankle to analyse the frequency of additional contracture development. Children with at least one examination after their first measured contracture from the start of the CPUP follow-up program in 1994 until the end of December 2018 were included. Data for 1,071 children included in CPUP before 5 years of age were followed for an average of 4.6 years (range 0-17 years). A contracture was defined as in study III and legs that underwent lower limb surgery were excluded at the date of surgery according to study III.

Statistics

IBM SPSS statistics version 23-26 (IBM Corp, Armonk, NY, USA) was used in all studies. R (R Foundation for Statistical Computing, Vienna, Austria) was used in study I, III and IV. STATA (version 14, Stata-Corp, College Station, TX, USA) was used in study III. P-values < 0.05 were considered significant for all analyses in this thesis. Categorical variables were described by frequency (n) and percentages (%) in all studies.

In *study I* age was categorised into five groups (1-3 years, 4-6 years, 7-9 years and 13-15 years). The Ashworth levels were divided into three groups (0, 1-1+ and 2-4). The ROM measurements were also categorised into normal ROM, mild contracture and severe contracture. Pearson x^2 test was used to analyse the categorical data and x^2 test for trend to analyse knee contracture in relation to GMFCS level and age. The Watson-Williams test for homogeneity was used to compare the mean direction of knee extension and levels of categorical variables. Multiple binary regression was used to analyse the outcome knee contracture (Yes/No) with explanatory variables; age, sex, GMFCS level, muscle length and spasticity. The results of the regression analysis were presented as odds ratios (ORs) with 95% confidence interval (CI). Nagelkerke R2 was used as a measure of goodness-of-fit.

In *study II* the interrater reliability was evaluated using the intra-class correlation coefficient (ICC) [78] and a CI of 95% with two-way random and absolute agreement for single measures. The ICC calculates the degree of disagreement and should exceed 0.75. The ICC was estimated for the right and left side separately.

In *study III* hip, knee and ankle contractures were dichotomized into two groups: contracture/no contracture. The follow up time for each leg was calculated with separate Kaplan-Meier (KM) curves drawn for each leg´s ROM to illustrate the proportion of contracture free legs at a given time. A 95% CI were generated for equally spaced time points every 2.5 year for each KM curve.

In *study IV* the time from the first to second contracture for each leg in each child was calculated using Cox proportional-hazards model, adjusting for GMFCS level. This was done both for all legs and separately according to the joint where the first contracture occurred. Legs with more than one contracture measured at baseline were only included in the descriptive analyses and when analysing the time between contractures. They were then excluded when analysing the sequences of contracture development. The x^2 test was used to analyse in which joint the second contracture occurred according to the GMFCS level; this analysis was performed separately for joints with a baseline contracture of the hip, knee, or ankle. The p-for-trend was calculated using logistic regression to analyse systematic, linear, associations of contracture development in these joints relative to the GMFCS levels.

Ethics

Ethical approval was granted by the Medical Research Ethics Committee at Lund University for study I, III and IV (383/2007, 443-99) and permission was obtained to extract data from the CPUP registry. Study II was approved by the Ethical Committee of Poznan University of Medical Sciences (nr 244/20) and written informed consent was obtained from a parent or guardian for participants under 18 years of age.

Results

Study I: Risk factors associated with knee contracture

A knee contracture was reported in 685 (22%) of the 3045 children. Of these, 12% had a mild contracture (-5˚ to -14˚) and 10% had a severe contracture (-15˚ or worse). The prevalence of knee contractures increased with higher GMFCS level. None of the children at GMFCS level I had a severe contracture compared to 35% of the children at level V. The prevalence of contracture also increased with older age and was slightly more common in males than in females (Table 2).

Table 2. Distribution of knee extension in relation to sex, age and GMFCS level.

		Knee extension		
Variable	**Level**	**Normal ROM ≥-4˚ *n* (%)**	**Mild contracture -5˚ to -14˚ *n* (%)**	**Severe contracture ≤ -15˚ *n* (%)**
Sex	Male	1300 (75)	220 (13)	199 (12)
	Female	993 (79)	149 (12)	117 (9)
Age (years)	1-3	461 (93)	30 (6)	5 (1)
	4-6	611 (85)	85 (12)	25 (3)
	7-9	576 (77)	90 (13)	66 (10)
	10-12	443 (67)	103 (16)	112 (17)
	13-15	262 (61)	61 (14)	108 (25)
GMFCS level	I	1214 (94)	73 (6)	0 (0)
	II	447 (89)	49 (10)	4 (1)
	III	199 (72)	43 (16)	33 (12)
	IV	236 (53)	96 (22)	112 (25)
	V	197 (42)	108 (23)	167 (35)

Note: GMFCS; Gross Motor Function Classification System, ROM; range of motion

The risk factors demonstrating strongest association with knee contracture were GMFCS level IV and V, severely reduced hamstrings length and an age over 10 years. Reduced hamstrings length with an angle below 120˚ increased the likelihood of having a knee contracture by almost ten times. Severely reduced muscle length in the gastrocnemius muscle also had a small association with knee contracture whereas spasticity had a very small effect (Table 3).

Table 3. Results of multiple binary regression for the association between contracture (yes/no) and sex, age, GMFCS level, hamstring- and gastrocnemius muscle length and spasticity of the knee and plantar flexors.

Variable	Level	OR	95% CI	*p*-value
Sex	Male	Ref.		
	Female	0.95	(0.71–1.27)	0.714
Age (y)	1–3	Ref.		
	4–6	1.88	(1.02–3.49)	0.044
	7–9	3.27	(1.80–5.94)	<0.001
	10–12	5.18	(2.84–9.45)	<0.001
	13–15	6.80	(3.62–12.77)	<0.001
GMFCS level	I	Ref.		
	II	0.95	(0.58–1.57)	0.849
	III	3.52	(2.14–5.79)	<0.001
	IV	7.91	(5.03–12.46)	<0.001
	V	13.17	(8.42–20.60)	<0.001
Muscle length				
Hamstring muscle	Normal length	Ref.		
	Mildly shortened	3.05	(2.24–4.17)	<0.001
	Severely shortened	9.86	(5.49–17.69)	<0.001
Gastrocnemius muscle	Normal length	Ref.		
	Mildly shortened	1.70	(1.24–2.32)	0.001
	Severely shortened	3.62	(2.31–5.66)	<0.001
Spasticity				
Knee flexors	0	Ref.		
	1–1+	1.49	(1.03–2.15)	0.035
	2–4	2.10	(1.27–3.47)	0.004
Plantar flexors	0	Ref.		
	1–1+	1.00	(0.70–1.42)	0.984
	2–4	0.80	(0.51–1.24)	0.317

Note: Results based on 2,168 of 3,045 individuals with complete data set. Nagelkerke R2=0.499. CI, confidence intervals, GMFCS; Gross Motor Function Classification System, OR; odds ratios, Ref; reference.

Study II: Interrater reliability of the popliteal angle tests

The mean popliteal angle was 31° measured with unilateral test and 15° in the bilateral test. The mean popliteal angle increased in both tests with higher age and there was a tendency for slightly higher values in males (Table 4).

Table 4. Popliteal angle with mean value and standard deviation (SD) for GMFCS, sex and age.

	Unilateral test Mean (SD)	Bilateral test Mean (SD)
GMFCS		
I	26.47 (10.17)	13.46 (9.9)
II	32.58 (10)	15.56 (10.02)
III	33.23 (7.84)	17.6 (8.13)
IV	32.38 (10.82)	16.25 (11.32)
Sex		
Female	29.14 (9.04)	12.81 (8.03)
Male	32.89 (10.54)	17.76 (10.61)
Age group		
0–6	28 (9.23)	11.63 (8.46)
7–12	30.64 (9.34)	15.03 (8.71)
13–18	32.98 (11.12)	17.31 (11.62)
19–22	47.5 (10.61)	33.75 (10.61)

Note: Average values for unilateral and bilateral popliteal angle tests of both examiners together presented for the different subgroups. GMFCS; Gross Motor Function Classification System

Both the unilateral and bilateral test to measure the popliteal angle in children and youth with CP had a high interrater reliability (Table 5).

Table 5. Interrater reliability for the unilateral and bilateral popliteal angle tests.

	Unilteral test		Bilateral test	
	ICC	95% CI	ICC	95% CI
Right				
Single measure	0.80	0.67-0.87	0.82	0.72-0.88
Left				
Single measure	0.86	0.78–0.91	0.83	0.74-0.89

Note: Intraclass Correlation Coefficients (ICC) with 95% Confidence Intervals (CI) for the right and the left leg

Study III: First lower limb contracture

A contracture in either the hip, knee or ankle joint occurred in 34% of all 4,751 legs during follow up. The median time for a first contracture was 10 years from the first examination. Within 10 years of follow up, 937 legs (20%) were operated on. The most common operation was soft tissue surgery of the hip (6.7 %) followed by soft tissue surgery of the foot and bony surgery of the hip. Contractures were more common in older age and at higher GMFCS levels. All lower limb contractures had the highest prevalence in children at level V. The most common contracture to occur first was an ankle contracture for children at GMFCS level I and II, and a knee contracture for children at GMFCS level III-V (Figure 10). After 10 years of follow-up, 10% of the children at GMFCS level I and 20% at level II had an ankle contracture compared to 40% of the children at level V. In addition, 70% of the children at level IV and 80% of the children at level V had a knee contracture after 10 years of follow up (Table 6).

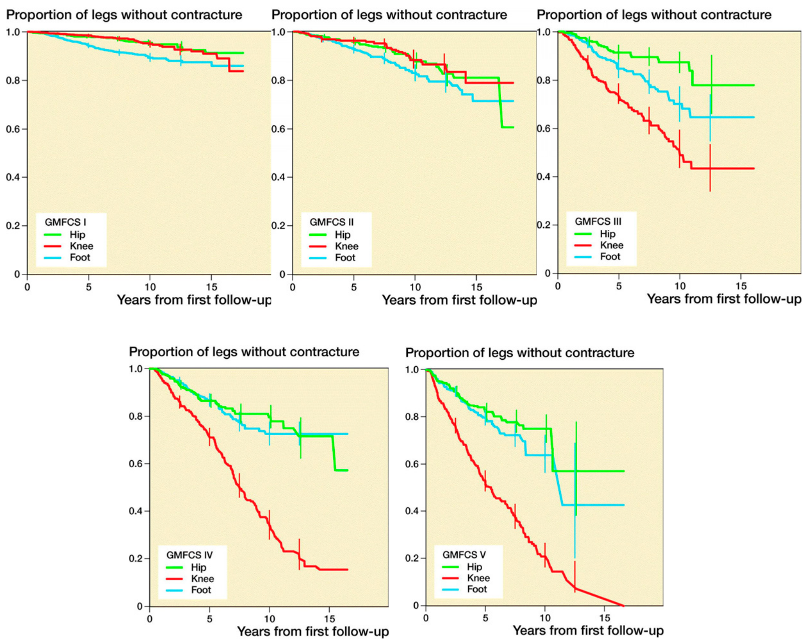


Figure 10. Proportions of contracture free legs stratified by GMFCS level and 95% pointwise confidence intervals for equally spaced time points every 2.5 years during the follow-up.
Note: GMFCS level; Gross Motor Function Classification System

Table 6. Proportion of legs free from hip, knee and ankle contracture, stratified by GMFCS level and 95% pointwise confidence intervals after 10 years of follow up.

GMFCS level	Hip	Knee	Ankle
I	1.0 (0.9–1.0)	1.0 (0.9–1.0)	0.9 (0.9–0.9)
II	0.9 (0.8–0.9)	0.9 (0.9–0.9)	0.8 (0.8–0.9)
III	0.9 (0.8–0.9)	0.5 (0.4–0.6)	0.7 (0.6–0.8)
IV	0.8 (0.8–0.8)	0.3 (0.3–0.4)	0.7 (0.7–0.8)
V	0.8 (0.7–0.8)	0.2 (0.2–0.3)	0.6 (0.6–0.7)

Note: GMFCS; Gross Motor Function Classification System

Study IV: Sequence of lower limb contracture development

Within the follow-up period, 720 legs (44%) developed additional contractures (Table 7). The second contracture occurred after an average of 5 years from the first contracture. The time from the first to second contracture varied depending on the location of the first contracture. Children with a primary hip or ankle contracture developed their second contracture earlier than those with a primary knee contracture.

Table 7. Location of the baseline contracture and prevalence of a second contracture during follow up and number of limb-years for children at GMFCS I-V.

GMFCS level	Location of the first contracture Legs n (%)				Second contracture Legs n (%)		Limb- years (mean)
	Hip	Knee	Ankle	> 1 joint	1 joint	> 1 joint	
I	41 (19)	83 (39)	78 (37)	10 (5)	53 (25)	2 (4)	899 (4.2)
II	43 (16)	98 (36)	112 (42)	16 (6)	101 (38)	2 (2)	989 (3.7)
III	40 (16)	124 (50)	66 (27)	19 (8)	115 (46)	3 (3)	747 (3.0)
IV	58 (14)	210 (52)	90 (22)	47 (11)	203 (50)	8 (4)	1,212 (3.0)
V	77 (15)	294 (59)	90 (18)	40 (8)	248 (50)	9 (4)	1,256 (2.5)
Total	259 (16)	809 (49)	436 (27)	132 (8)	720 (44)	24 (3)	5,103 (3.1)

Note: GMFCS Gross Motor Function Classification System

When analysing legs with only one baseline contracture and one follow-up contracture, hip contracture was the primary contracture in 259 legs. Of these, 47% developed a second contracture, most commonly in the knee (74%) and less frequently in the ankle (26%).

A knee contracture was the primary contracture in 809 legs. A second contracture occurred in 297 (37%) of these legs and the distribution between hip (47%) and ankle contracture (53%) as the second contracture was even. Children at GMFCS level I-IV were more likely to have an ankle contracture as their second contracture while children at level V were more likely to have a hip contracture.

Ankle contracture was the primary contracture in 436 legs, and a second contracture occurred in 151 (35%) of these legs. An ankle contracture was more likely to be followed by a knee contracture (79%) and less likely by a hip contracture (21%). Having an ankle contracture at baseline increased the likelihood of the second contracture to be a knee contracture in children at all GMFCS levels (Table 8).

Table 8. Results for chi-square and p- for trend analyses of the association between each baseline contracture and the location of a second contracture during follow up at each GMFCS level for legs that developed one single baseline contracture and one single follow up contracture.

First contracture	GMFCS level	Legs n	Second contracture Hip n (%)	Knee n (%)	Ankle n (%)	p value
Hip joint						0.002a
	I	8	–	5 (63)	3 (37)	< 0.001b
	II	21	–	10 (48)	11 (52)	
	III	19	–	12 (63)	7 (37)	
	IV	28	–	22 (79)	6 (21)	
	V	45	–	40 (89)	5 (11)	
	Total	121	–	89 (74)	32 (26)	
Knee joint						0.003a
	I	15	4 (27)	–	11 (73)	< 0.001b
	II	32	7 (22)	–	25 (78)	
	III	54	25 (46)	–	29 (54)	
	IV	80	36 (45)	–	44 (55)	
	V	111	65 (59)	–	46 (41)	
	Total	292	137 (47)	–	155 (53)	
Ankle joint						< 0.001a
	I	18	3 (17)	15 (83)	–	0.59b
	II	30	10 (33)	20 (67)	–	
	III	20	3 (15)	17 (85)	–	
	IV	40	7 (17)	33 (83)	–	
	V	43	9 (21)	34 (79)	–	
	Total	151	32 (21)	119 (79)	–	

Note: GMFCS; Gross Motor Function Classification System

Discussion

The overall aim of this thesis was to increase the knowledge about the development of knee contractures; prevalence, risk factors, sequence of contracture development and the reliability of popliteal angle measurements.

We found a prevalence of 22% for knee contractures in children with CP. Knee contractures were strongly associated with GMFCS level, age and short muscle length (Study I). When measuring hamstrings length, different types of measurements are used in clinic, with unilateral and bilateral popliteal angle test being the most common. We found that both tests had a high interrater reliability (Study II). When studying the contracture development in the hip, knee and ankle we found that the location of the first contracture depends on GMFCS level and that children at level I-II were most likely to develop an ankle contracture as their first contracture and children at level III-V a knee contracture. In total, 34% of all children developed at least one lower limb contracture during childhood (Study III). When following the development of further contractures, we found that 44% developed at least one additional contracture. The second contracture tended to occur in a specific pattern based on the location of the first contracture. The second contracture was most likely to occur in the closest adjacent joint (Study IV).

Prevalence of contractures

We found that 22% of all children with CP had a knee contracture. There is limited evidence about the prevalence of lower limb contractures from previous studies. A recent study from Denmark found a prevalence of 44% knee contractures in their population of children with CP followed within a similar program to CPUP [79]. They identified the biggest difference compared to our study in age group 0-3 years but when comparing the results, the prevalence of knee contractures was higher in all age groups and at all GMFCS levels in their study. A potential reason for this could be that Sweden has followed the CPUP program since 1994 compared to Denmark where the program was initiated 2010. Sweden has a yearly reporting rate of 90-95% [71] and Denmark 74% [80]. The health system in Denmark differs from the system in Sweden. In Sweden, the children are followed by multi-professional teams, working in close collaboration, while in Denmark, healthcare professionals are employed by different health care systems, mostly by the community-based healthcare providers. Clinicians in Denmark prescribes less orthoses and standing aids than Sweden and has less access to occupational therapists [81].

We found that knee contractures are present at all GMFCS levels but are very rare for children at GMFCS level I (6%) and level II (11%) compared to level V where more than half of the children are affected (58%). CP is a heterogenous group and being able to discriminate between groups within the CP diagnosis enables better predictions about development and function and directed treatment [19,82]. We found a clear difference between GMFCS levels in contracture prevalence and contracture development in study I, III and IV. The GMFCS level is relatively stable over time [20,83,84] and by following the child´s gross motor function curves (GMFM) and their GMFCS level we can predict future function [85,86]. Despite this, international studies report that less than half of the parents to children with CP know their child's GMFCS level [87]. It´s still a need for clinicians to include description of the GMFCS level and how this affect treatment plans and future ability in discussions with parents [88].

We also found that the prevalence of knee contractures increased with higher age. Even though CP is a non-progressive disorder it is well known that the secondary symptoms can change and worsen over time [29,89-92]. Study I confirmed that the oldest age group (13-15 years) had the strongest association with knee contracture (OR 6.80). However, a high GMFCS level had a stronger association with knee contractures than age (OR 7.91 for level IV and 13.17 for level V).

We found that 34% of all children with CP develop at least one lower limb contracture. The prevalence of multiple lower limb contractures developed with higher GMFCS levels. This correspond with previous findings [93].

After 10 years from first follow up, an ankle contracture is the most common flexion contracture for children at GMFCS level I-II with a prevalence of 10 to 20%. For children at GMFCS level III-V, knee contractures are the most common contracture after 10 years of follow up with a prevalence of 50 to 80%. Contractures increase the risk for pain [59] and our results correspond with the most common pain locations for children at level I and II, foot, and level III to V, knee and hip [94]. Pain decreases function and quality of life and is important to address and treat [95,96].

Clinical Measuring

In study II we found a high interrater reliability for the unilateral and bilateral popliteal angle tests. A previous study by Ten Berge et al. [97] showed a low interrater reliability for the unilateral test. They instructed the examiners to extend the knee three times before measuring the angle in the position where the "next endpoint of resistance was felt". It was not specified how much pressure the examiners applied to extend the knee. In our study, the examiners were instructed to extend the child´s leg with as much pressure as tolerated by the child and reach the end ROM. One challenge with comparing studies about hamstrings measurement is that not all studies specify whether the test is performed according to the unilateral or bilateral procedure [98,99]. Our study showed that the mean values are different between the two measurements. The positions of the pelvis yield different results measured in ROM [100] and the muscles activated during the test differ [77]. Therefore, it is important to specify how the test is performed in terms of the child's position and the pressure added by the examiner. Another challenge with measuring hamstrings length is that several studies have showed a limited correlation between the results of the test and the actual length of hamstrings [52]. The hamstrings muscle is a biarticular muscle, crossing two joints, which makes it difficult to measure the actual length in clinic. Instead, we measure the indirect length by the popliteal angle tests [101]. Previous studies have shown that the results of passive measurement of the popliteal angle tend to underestimate the maximal length of hamstring during gait [54,102]. Thompson et al. [52] found a correlation between the bilateral popliteal angle and maximal hamstrings length and no correlation between the unilateral test and hamstrings length. It is recommended that passive popliteal angle ROM is combined with gait analysis before treatment decisions are made and the outcome measure after treatment should be functional rather than a static test [52].

In study I, we found a limited association between knee contracture and spasticity in hamstrings or gastrocnemius measured with the Modified Ashworth Scale (MAS). Spasticity is a challenging term to define and assess in clinic. The term spasticity is used in different ways with different meanings. One aspect of it refers to the physical signs measured at clinical examinations and another aspect to the underlying pathological process in the child [103]. Studies are inconsequent in assessment methods for measuring spasticity and in a review by Scholthes et al. [104], 13 different assessment methods were used. The most commonly used assessments of spasticity are the Ashworth Scale and the Tardieu scale. The reliability of the Ashworth scale varies between studies and is questioned [105,106]. It can also be difficult to distinguish between a resistance of a muscle to stretch caused by alteration in reflex response or by muscle pathology and muscle stiffness in children with CP compared to children without CP [107].

In study I, only 2% of the children had a reported MAS grade 3 or 4 although 37% had a reduced hamstrings length and more than 10% of the children had a severe knee contracture. In a study by Fosdahl et al. [108] none of the children had a reported MAS grade higher than +1. This indicates that it is challenging to distinguish between levels of the scale and between spasticity, muscle stiffness and contracture [105,107,109]. Therefore, we regrouped MAS into 3 groups.

It is important to describe our findings from physical examinations, such as the perception of resistance to passive stretch rather than only referring to the concept as "spasticity" [103].

Sequence of contracture development

Our studies contribute to the existing body of knowledge since no previous studies have investigated the sequence of contracture development. Our studies showed that lower limb contractures occur early. After 5 years of follow up, over 50% of the children at GMFCS level V already have a contracture. When seeing contractures as a result of muscle stiffness, this corresponds to previous findings of early reduction of skeletal muscle volume and altered gene expression seen in young children with CP [110,111]. The main finding in study III was that the first contracture is likely to occur in different joints depending on the child's GMFCS level. Children at GMFCS level I and II most likely get an ankle contracture first and children at level III-V most likely a knee contracture. Only 13% of the children at GMFCS level I and 25% at level II get a lower limb contracture in the sagittal plane compared to 63% of the children at level V. Changes in the sagittal plane are the most common gait abnormality for ambulant children [45,46].

After 10 years of follow up, 80% of the children at GMFCS level V have developed a knee contracture. This shows that there is still more to learn and understand about contracture development to fully manage to prevent them [66]. Children with one contracture have an increased risk of additional contractures. We found that second contractures most often occur in the closest adjacent joint to the first contracture. For the knee joint, with two adjacent joints, the location of the second contracture was dependent on GMFCS level. Non-ambulant children were more likely to get a hip contracture after their initial knee contracture and ambulant children an ankle contracture [112]. Non-ambulant children usually spend more time sitting in a wheelchair and in lying. Adults with CP, in need of standing devices, spend less than one hour per day standing [113]. In supine position, a knee contracture might force the hip into flexion and gravity will tilt the legs to one side causing a windswept position [58]. Asymmetric postures are highly associated with the inability to change and maintain position [113], and increase the risk of having contractures [59]. When Petersson et al. [57] created a risk score for scoliosis in children with CP they identified knee contracture as an independent predictor for severe scoliosis.

For ambulant children, a knee or ankle contracture of -10° or worse changes kinematics and affects gait [114-116]. A knee contracture may force the child to walk in equinus due to lever arm dysfunction and an ankle contracture in children with unilateral CP might force the child to walk with a flexed knee due to leg length discrepancy [117]. The gastrocnemius muscle involves both the knee and ankle joint and affects knee flexion and plantar flexion which probably also explains the increased risk of an additional contracture in an adjacent joint.

Early detection and interventions

The results of our studies showed that lower limb contractures develop early and that the incidence increases with age. It is easier to get a better outcome from treatment if it´s initiated early [66,68]. Orthoses are commonly used to improve function or to prevent contracture development and deformities. They can be used to improve and maintain the biomechanical alignment of a joint or body segment. Ankle-Foot Orthoses (AFO) are the most common orthoses [118,119] and are used by 50% of all children with CP in Sweden, either to improve function (10%), ROM (11%) or both (30%) [120]. A study by Wingstrand et al. [120] showed that the use of AFO increased with higher GMFCS level and was most frequently used in young children aged 4-6 years. This corresponds to the results of study III showing that ankle contractures are most common at GMFCS level V and that contracture development starts early. Study IV showed that ankle contractures in children were most commonly followed by a knee contracture. Miller [66] recommends immobilizers for the knee when using AFOs at night. Stretching gastrocnemius at the ankle can pull the knee into flexion and potentially lead to a knee contracture. Our results show that this might be especially important for non-ambulatory children where knee contractures are common. It is important to be aware of the low evidence for orthoses to prevent and treat contractures [120-123]. The prescription should therefore be decided together with families and with the child's goals, comfort and function kept in focus. Interviews with families describe a frustration over both the provision and difficulties to wear orthoses but also a positiv perception that orthoses support mobility and participation [118,124].

Short time stretch does not increase ROM, reduce pain or improve function in people with CP [39,125]. Long-time stretching with orthoses [126], standing [127], active stretching (through exercise and activity) [128], stimulation of muscle growth through exercise [129] and maintaining biomechanical alignments through positioning [58] might have a positive effect on the child´s overall function and potentially reduce the risk of secondary contracture development. The evaluation of 10 years of multi-professional follow up of children with CP in Sweden showed a decreased prevalence of severe contractures [75].

Botulinum toxin injections are also frequently used to reduce spasticity, improve function and prevent secondary contracture development. Injections are most common in the gastrocnemius muscle and in particular for children at GMFCS level I [130]. Injections in gastrocnemius are most common in younger children, 4-6 years old [131]. This corresponds to our results showing that the first contracture to occur in children at GMFCS level I was an ankle contracture and that contracture development starts early. Injections in the hamstring muscles are most common for children at level IV-V which also correspond to our results showing that reduced hamstrings length are associated with higher GMFCS level. The injections of hamstrings are most common in older children, 10-12 years [131]. Our findings indicate that problems in hamstrings start early and, therefore, treatment should be initiated earlier if the aim is to contribute to a possible prevention of contracture development. Botulinum toxin treatment should be combined with orthoses, casting and therapy to affect ROM [123]. Botulinum toxin injections should be used thoughtfully. Many studies have reported changes in muscle tone after injection but not many report changes in function. Botulinum toxin injection can cause damage to the muscle and a decreased frequency of injections to once every 12 months is recommended [132].

In study I, we found a limited association between spasticity and knee contracture. As mentioned previously, spasticity as a phenomenon is difficult to define and address. The clinical model that muscles need to be stretched if they grow and that spasticity limits this ability and therefore must be treated is not proven [103]. When it comes to contracture development, reduction of spasticity in terms of botulinum toxin injections and selective dorsal rhizotomy has not been effective to reduce contractures long term which indicates a need for further knowledge about spasticity as a concept in children with CP [25,26,133]. However, a reduction of spasticity might still increase the child´s function, facilitate positioning or the use of orthoses, reduce pain, or improve gait and through that contribute to less deformities. To directly prevent contracture development, a deeper understanding of the musculoskeletal system and the interaction between components within the system is needed (Figure 11).

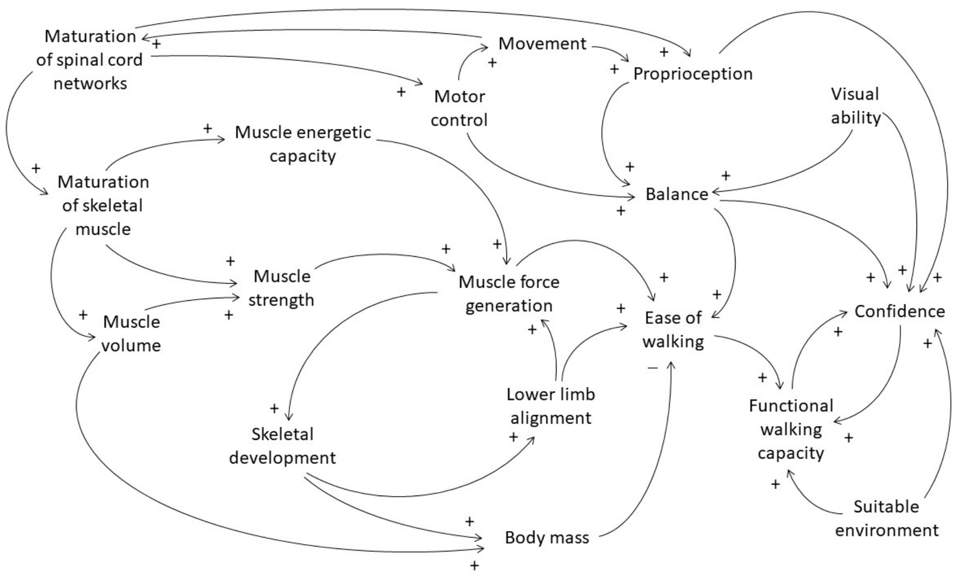


Figure 11. Reproduced with permission from 'The Musculoskeletal System in Children with Cerebral Palsy: A Philosophical Approach to Management by Martin Gough and Adam Shortland published by Mac Keith Press (www.mackeith.co.uk) in its Clinics in Developmental Medicine Series, 2022, 9781911612537.

Further, we determined a strong association between reduced hamstrings length and knee contracture. This finding corresponds to other studies describing interventions for hamstrings as the most common way to treat knee contractures [38,61,62]. Nordmark et al. [93] found that hamstrings starts to become shorter before the development of knee contracture in a total population of children with CP. The popliteal angle increases during childhood in children with CP at all GMFCS levels [93,108]. Short hamstrings muscle is challenging to treat and non-surgical interventions are most likely to succeed in younger children with a mild reduction of ROM or a mild contracture [66].

Study III showed that the age at first surgery varied between types of surgery and body segment. The median age was 4 years for soft tissue surgery of the hip, 6.5 years for soft tissue surgery of the foot/ankle, and 9 years for soft tissue surgery of the knee. The median age for bony surgery was higher for all joints. According to Hagglund et al. ankle surgery is the most common surgery for children at GMFCS level I-II and hip and spine surgery for level IV-V. Reduced hamstrings length can be more difficult to treat with a risk of overlengthening of the muscle [66] which can be a reason why knee surgery is performed at a higher age when other treatments might not have succeeded. Also, the risk for hip displacement is higher in younger children and this might be a reason for the lower median age of soft tissue surgeries of the hips.

Our findings that one contracture increases the risk for additional contractures and that a second contracture most often occurs in an adjacent joint indicates a need for postural management to facilitate good postural alignments and reduce the risk for further contractures. Children with asymmetric postures sustained over longer periods of time and those unable to change position have a higher risk for scoliosis, windswept hip deformity, hip contracture and knee contracture [59]. Asymmetric postures can be seen already during a child's first year of life with an association to a later development of deformities [134].

Systematic follow-up of children with CP in Sweden has reduced the number of severe contractures and the need for surgical treatment. Early detection and treatment for reduced ROM that interferes with function is an important reason to continue to follow children with CP in a standardised way [74,75,135].

Understanding the underlying mechanisms behind contracture development in children with CP is essential to develop treatment strategies[103]. In study IV, we found that lower limb contractures tend to occur in a specific pattern where the location of the first contracture affected the location of the second contracture. It is tempting to think that if we could manage to treat the first contracture we might automatically avoid the second one. Unfortunately, the musculoskeletal system is more complicated than that. Our results strengthen the existing body of knowledge about differences in contracture development between children at different GMFCS levels and the potential effect that one contracture has on adjacent joints due to biomechanical alignments [57-59]. We do, however, need to put this information in a larger causal loop to increase our understanding of the musculoskeletal system and to be aware of how our interventions can affect other parts of our patients' bodies or function. A contracture is both the result of, and the start of, an interaction between many aspects involving nutrition, movement, visual ability, body mass, muscle strength, balance, proprioception, confidence of the child etc. [103] (Figure 11).

Interventions focusing on body structure and function do not automatically lead to improvements in activity or participation [136]. Traditionally, disabilities have been seen as a "within-the-person" issue that treatment is expected to fix. The importance of environmental factors has often been neglected [137]. In 2001, the World Health Organization published The International Classification of functioning, disability and Health (ICF) as a framework for health professionals to think about health and how it might affect different aspects of a person's life. In 2011, Rosenbaum & Gorter developed an alternative framework based on ICF called "The F-words" (Figure 12)[138]. The aim of the F-words was to recognise that all areas are equally important and encourage clinicians to adopt this way of thinking when meeting children with disabilities. Our attempts to prevent, and treat contractures, must be added to the bigger framework representing the child's life. When evaluating treatment, activity and participation outcome measures should be added as a complement to ROM and spasticity to evaluate changes in quality of life [139-141]. Health care providers should always strive to promote child and family development and use our best available medical, technical and therapeutic resources to do so [137].

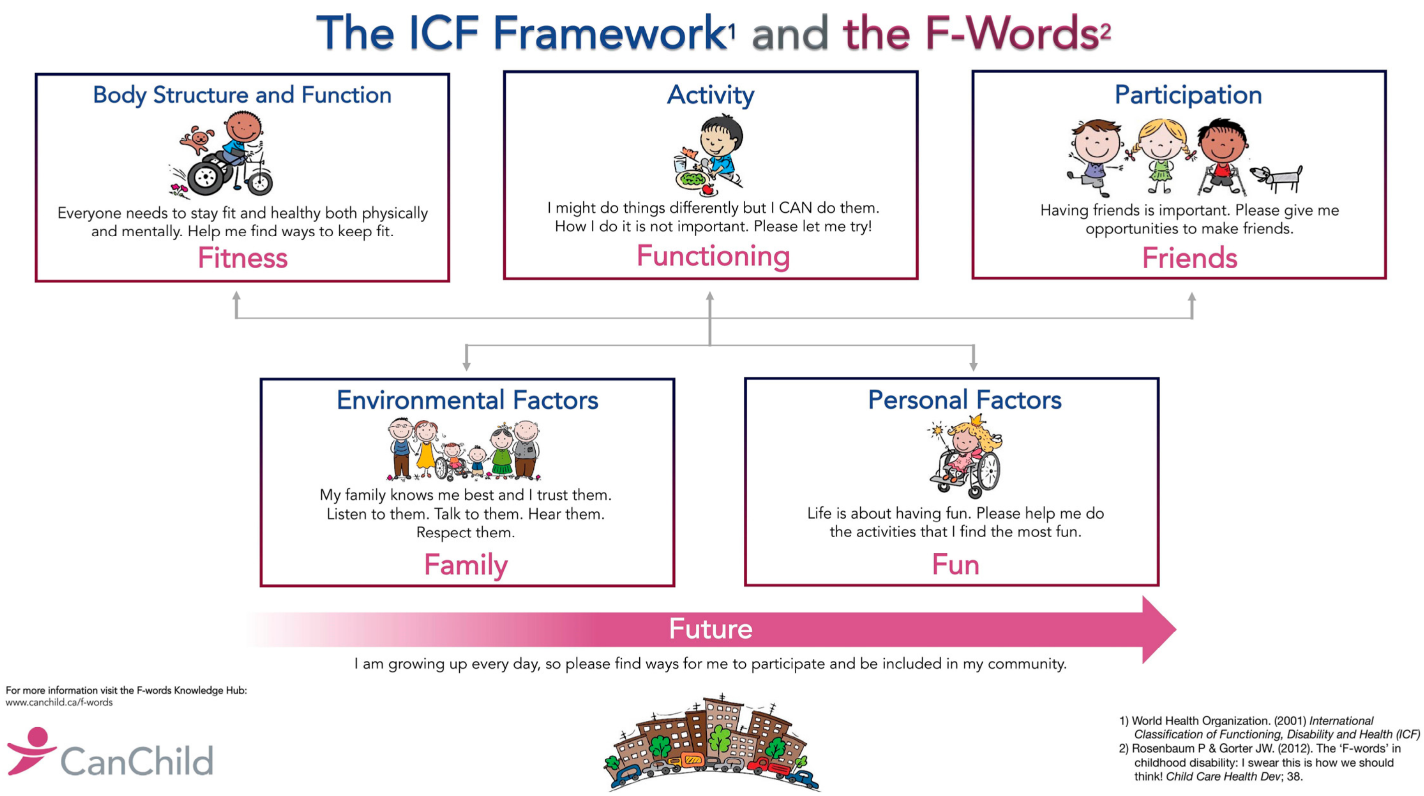


Figure 12. The F-words in childhood disability.

Limitations

There are several limitations to our studies. Three of our studies are based on data from a national CP registry. Register based data provides a large number of data but can be difficult to translate to clinical settings. Register studies can however be a good complement to randomized control trials which are challenging in our field due to small study populations and difficulties to double-blind[103]. Register data can be advantageous in the identification of associated factors, start- and endpoints and development over time. Furthermore, register data provides large, unselected datasets with repeated measurements collected in a consistent way over long periods of time.

In Sweden, all children with CP have access to free healthcare services including regular follow ups by multi-professional paediatric teams, assistive technology devices, orthoses and surgeries [142]. These factors should be taken into consideration when translating the results to other populations. Study III and IV cover a period of 24 years and interventions have changed over time from more hands-on therapy and "fixing the child" to new treatment options, technology and "fixing the environment" with focus on participation and activities in daily life.

A general limitation was the definition of a contracture. There is no standard definition of degrees in ROM for a contracture. In study I, we used two different cut-offs, mild and severe. In study II and III we wanted to use one cut-off and chose -10˚ for all joints since these values are often used in clinical practice and severely affect biomechanical alignment, gait, function and positioning [114-116,143]. We also ran the statistical analyses in study III and IV with 0˚ as a cut-off and got similar outcomes. The reliability of goniometric measurement varies with position and joint[144-146] but even though the reliability can be low, it is still the most commonly used method in clinical settings. We validated the data to correct measurement errors e.g. 180˚ of knee extension instead of 0˚ and reporting errors e.g. 300˚ of dorsiflexion instead of 30˚.

Another general limitation is that we did not include CP subtypes in our analyses due to the large number of missing reports. The CP subtype is usually verified between 4 to 7 years and we excluded children who were not reported into the register before the age of 5. There is also a lack of neuropaediatricians in Sweden and they are responsible for classifying the child's subtype.

Study I

We used the MAS to measure spasticity and the scale has a relatively low reliability. still, the Ashworth and Modified Ashworth scales are currently the only available tools in clinical practice. The study did not reflect the natural development of contracture since we included children in a systematic follow-up program who received treatment. It was a cross sectional study so we did not follow the contracture development over time and could not establish any causal relationships.

Study II

A potential that the examiner was influenced by the result of the first test when performing the second test. No children at GMFCS level V were included in the study. The popliteal angle is likely more difficult to measure in children at level V depending on contractures and severity of diagnosis. The lack of children at level V also affected the mean value of measured popliteal angles.

Study III and IV

We censored children who had lower limb surgery at the date of surgery. In Sweden, children at risk of hip dislocation have their first hip surgery early (median age 4 years) and often before a confirmed contracture. This may have affected the results especially for children at GMFCS level IV and V because these children most often receive hip surgery. We included children treated with botulinum toxin injections and serial casting and it can be argued that this interfered with the results. However, given the large number of treated children and the fact that the effect of casting and injections is not permanent, these children were included. The intervals between examinations varies between GMFCS levels which results in different number of examinations. This might affect the proportions of contractures but not the sequence for each GMFCS level.

Clinical implications

This thesis provides new knowledge that can help to improve clinical practice. We now know that knee contractures are a common problem for children with CP and affects children at all GMFCS levels.

Further, we should be extra observant on reduced dorsiflexion for children at GMFCS level I-II and reduced knee extension for children at GMFCS level III-V since these are the most common locations of the first lower limb contracture. We know that lower limb contractures occur early and therefore we should focus on early prevention. We know that it is common for children with one contracture to develop additional contractures. The location of the first contracture affects the location of the second contracture, and we should be aware of this when prescribing orthoses and positioning equipment to avoid further contracture development.

The majority of all children with higher GMFCS levels will develop a knee contracture during childhood. It is the first contracture to occur in children at GMFCS level III-V and increases the risk for other deformities such as windswept hips and scoliosis. Sweden has half as many knee contractures compared to Denmark which indicates that our multi-professional work has paid off and that through monitoring the child’s development, providing appropriate equipment, promoting standing, positioning and timely surgery we have managed to reduce the number of severe contractures.

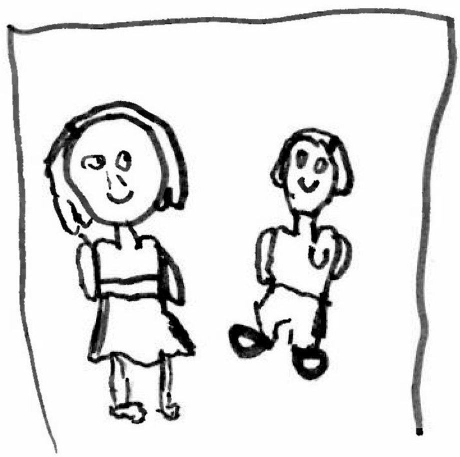
Knee contractures have a strong association with reduced muscle length and only a weak association with spasticity. We should therefore focus on helping the child maintain muscle length. However, we still do not fully understand the mechanisms behind muscle pathology and contracture development and which interventions that are the most successful and when. By stimulating healthy muscle growth, promote activity, increase standing and walking options, reduce time in sitting and optimize positioning we might contribute to better function and less secondary contracture development in children with CP.

Since there are different ways to measure popliteal angle in children with CP, and as the results depends on which test is being used, it is important to document how we performed the test. Both the unilateral and bilateral test are reliable. We should also be aware of the fact that true length of hamstrings and spasticity is difficult to measure in clinic and a combination of results from measurements, functional tests and analysis of gait are recommended.

I wish to end this thesis with a quote by Mulderij 2000 [147]:

"What I would like to emphasize above all is that we are not only treating a moving body, we are treating a child who is out to explore the world physically, to enjoy freedom of action, and to confer his or her own meanings on the world"

And as Smilla answered me when I asked her about CP, contractures and measures, in the foreword of this thesis, she doesn't even think about it. She thinks about difficulties in daily life and how to be able to participate with her friends. This provides us with important information that we have to bring our knowledge about knee contracture, measuring and sequence of contracture development into a bigger context and focus on the child's goals and their abilities when deciding on interventions and treatment.



"The small therapist"

"Oops I drew you too small." No, you did not. I´m the perfect size for my purpose. To ensure that your goals are in focus and not mine.

Conclusions

This thesis has described the prevalence of knee contractures and the development of lower limb contractures in children with CP.

Knee contractures are a common problem for children with CP at all GMFCS levels and are associated with higher GMFCS level, older age and reduced muscle length in hamstrings and gastrocnemius. Spasticity measured with MAS had a low association with the prevalence of knee contracture.

Both the unilateral and bilateral test to measure the popliteal angle showed high interrater reliability when used in children and youth with CP. Given the variability in measuring methods and results, it is important to describe what method is used and to include the results from different examinations, such as gait analysis and functional tests when deciding on treatment.

Lower limb contractures occur early and the risk increases with higher GMFCS level and older age. The first lower limb contracture to occur is an ankle contracture in children with GMFCS level I-II and a knee contracture in children at level III-V. Early interventions to prevent knee and ankle contractures should be considered.

Development of a second contracture is a common problem for children with CP. The time between the first and second contracture and the sequence of contractures are influenced by GMFCS level. A second contracture primarily occurs in the adjacent joint. This should be considered when treatment strategies are being formed.

In summary

Knee contractures affect children at all GMFCS levels and a majority of children at higher GMFCS levels will develop a knee contracture during childhood within the first ten years of follow up. Knee contractures are strongly associated with short hamstrings length. When measuring hamstrings length, both unilateral and bilateral popliteal angle measurements are reliable. Children at GMFCS level I-II most often develop an ankle contracture first and children at level III-V a knee contracture. The second contracture primarily develops in the closest adjacent joint. Treatment plans should be based on GMFCS level.

Future research

1. To investigate the effect of contractures on children’s quality of life.
2. To explore children’s experience of interventions to treat contractures.
3. To follow range of motion and contracture development during adulthood.
4. To analyse the effect of standing intervention to reduce contracture development.

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This was the first section I wrote for this thesis since gratitude has characterised this project from the beginning.

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Paper I

DEVELOPMENTAL MEDICINE & CHILD NEUROLOGY ORIGINAL ARTICLE

Demographic and modifiable factors associated with knee contracture in children with cerebral palsy

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ABBREVIATIONS

CPUP	Cerebral Palsy Surveillance Programme
ROM	Range of motion

AIM To identify the prevalence of knee contracture and its association with gross motor function, age, sex, spasticity, and muscle length in children with cerebral palsy (CP).

METHOD Cross-sectional data for passive knee extension were analysed in 3 045 children with CP (1 756 males, 1 289 females; mean age 8y 1mo [SD 3.84]). CP was classified using the Gross Motor Function Classification System (GMFCS) levels I (n=1 330), II (n=508), III (n=280), IV (n=449), and V (n=478). Pearson's χ^2 test and multiple binary logistic regression were applied to analyse the relationships between knee contracture and GMFCS level, sex, age, spasticity, hamstring length, and gastrocnemius length.

RESULTS Knee contracture greater than or equal to 5 degrees occurred in 685 children (22%). The prevalence of knee contracture was higher in older children and in those with higher GMFCS levels. Odds ratios (ORs) for knee contracture were significantly higher for children at GMFCS level V (OR=13.17), with short hamstring muscles (OR=9.86), and in the oldest age group, 13 years to 15 years (OR=6.80).

INTERPRETATION Knee contracture is associated with higher GMFCS level, older age, and shorter muscle length; spasticity has a small effect. Maintaining muscle length, especially of the hamstrings, is important for reducing the risk of knee contracture.

Despite preventive treatment, children with cerebral palsy (CP) often develop muscle contracture that limits their range of motion (ROM). Contracture often results from the chronic activation of a muscle, sometimes in combination with weakness of the antagonists, which occurs in people with CP.[1,2] Low joint mobility also affects adjacent joints and can cause postural asymmetry.[3] The mechanisms underlying contracture in CP are complex and are not fully understood.[4]

Knee flexion contracture frequently occurs in children with CP. A number of different factors can contribute to the development of knee contracture, including older age, spasticity, muscle imbalance, immobility, and prolonged sitting posture.[5,6] Limited knee extension affects both ambulant and non-ambulant children. For non-ambulant children, knee contracture may affect the ability to transfer and to assume an aligned standing or lying posture. For ambulant children, knee contracture can cause an asymmetric posture, impaired gait pattern, increased energy consumption, and difficulty wearing orthoses.[7]

Limited knee extension can lead to a gait pattern that imposes a greater pressure on the knee extensor apparatus and the patellofemoral joint, which can cause knee pain.[8] The proportion of children with pain increases with age and pain is most common in the lower extremities.[9] One in four adults with CP experiences knee pain,[10] which is associated with decreased joint mobility.

The Cerebral Palsy Surveillance Programme (CPUP) is a national quality registry in Sweden.[11] More than 95 per cent of all children with CP nationwide are included in this programme. In the CPUP, children are followed regularly by their local rehabilitation team and are examined every 6months, once a year, or every other year, depending on their age and level in the Gross Motor Function Classification System (GMFCS).[12]

To our knowledge, no studies have investigated multiple factors associated with knee contracture in a large population of children with CP. A deeper understanding of the factors that contribute to the development of knee contracture might help in the creation of treatment strategies.[1] The purpose of this study was to determine the prevalence of knee contracture and its association with GMFCS level, age, sex, spasticity, and muscle length in children with CP.

METHOD

Participants

This was a cross-sectional study based on data from the Swedish CP registry and the CPUP. The study participants were all 3 045 children with CP aged 1 year to 15 years, born between 2000 and 2014 who were reported to the registry in 2014 and 2015. The data for the most recent visit were used. The children represent all 21 counties in Sweden with a total population of 10 million inhabitants. The Research Ethics Committee at Lund University approved the study (LU-443-99), and permission to extract data was obtained from the registry holder.

Classifications and measurements

The diagnosis of CP was verified at the age of four years by a neuro-paediatrician, according to the inclusion and exclusion criteria of the Surveillance of Cerebral Palsy in Europe network.[13] Children younger than 4 years with presumed, although not yet confirmed, CP are included in the CPUP, while those who at a later stage do not fulfil the criteria for CP are excluded. In the present study, 521 children were younger than 4 years with presumed but not yet confirmed CP. Gross motor function was classified by the child's physiotherapist into levels I to V according to the expanded and revised version of the GMFCS.[12]

Passive knee extension was assessed by goniometric measurement in a standardized supine position according to the CPUP manual (http://www.cpup.se). In the CPUP, ROM is graded into three levels according to a traffic light system with separate cut-offs for GMFCS levels I to III compared with GMFCS levels IV to V. Green indicates a good passive ROM. Yellow indicates a reduced ROM and the need for increased observation and actions to improve ROM. Red indicates the development of a contracture that requires intervention. In this study, we used the same classification values for all GMFCS levels. We classified knee extension as a mild contracture if the extension was minus 5 degrees to minus 14 degrees and as a severe contracture if the extension was minus 15 degrees or less.

The hamstring length was classified according to the popliteal angle with the hip flexed to 90 degrees with the child in the supine position. We classified a popliteal angle of 120 to 139 degrees as indicating a mildly reduced hamstring length and below 120 degrees as a severely reduced hamstring length. The length of the gastrocnemius was classified according to the angle of the ankle joint in dorsiflexion with the hip and knee extended. Angles between 9 degrees and 0 degrees of dorsiflexion were considered to indicate mildly reduced muscle length and below 0 degrees indicated severely reduced muscle length.

Muscle tone of the knee and plantar flexors was measured according to the Modified Ashworth Scale.[14] In the statistical analyses, the Ashworth levels were divided into three groups: 0, normal muscle tone; 1 to 1+, slight increase in muscle tone; and 2 to 4, large increase in muscle tone. For all children in this study, information from only one leg, that with the lowest ROM, was used in the analyses. If the ROM was equal on both sides, we used the leg with the highest Ashworth level. For children with the same measurements in both legs, one leg was randomly assigned to be included in the analyses.

Passive knee extension was analysed according to GMFCS level, age, sex, spasticity of the knee and plantar flexors, and length of the hamstring and gastrocnemius muscles.

What this paper adds

- Knee contracture occurs in children with cerebral palsy at all Gross Motor Function Classification System (GMFCS) levels.
- Knee contracture in children is associated with short hamstring muscles, higher GMFCS level, and older age.
- Short hamstring muscles present a greater risk for knee contracture than spasticity.

Statistical analyses

In the statistical analyses, age was classified into five groups (1–3y, 4–6y, 7–9y, 10–12y, and 13–15y). The Ashworth levels were divided into three groups (0, 1 to 1+, and 2 to 4). Knee extension and length of the hamstrings (popliteal angle) and gastrocnemius (dorsiflexion) were also divided into three groups: normal, mild contracture, and severe contracture. Categorical variables are described by frequencies (n) and percentage (%).

Pearson's χ^2 test was used to analyse categorical data, while a χ^2 test for trend (linear-by-linear association test) was used to analyse trends in knee contracture according to GMFCS level and age. Since the outcome variable knee extension was originally measured as the angle of extension, it is essentially a circular data variable, that is, given as the angle of a circle.[15] To fully take this characteristic into account in the statistical analyses, the Watson–Williams test for homogeneity of means was used to identify differences in the mean direction of knee extension between the different levels of categorical variables. For these analyses, knee extension is described by mean direction with accompanying circular standard deviation.

Multiple binary logistic regression was used to analyse the simultaneous associations between the outcome knee contracture (yes/no) and the explanatory variables sex, age, GMFCS level, hamstring and gastrocnemius muscle lengths, and spasticity of the knee and plantar flexors. For the regression analysis, a child was classified as having a knee contracture if the knee extension was below 5 degrees or less (i.e. mild or severe contracture) and as not having a knee contracture if the knee extension was below 4 degrees or more. The results of the regression analyses are presented as odds ratios (ORs) with accompanying 95 per cent confidence intervals (CIs). Nagelkerke R^2 is used as a measure of goodness-of-fit. SPSS v23/24 (IBM Corp., Armonk, NY, USA) and R 3.3.2 (R Foundation for Statistical Computing, Vienna, Austria) with the package 'circular' version 0.4-7 were used for the statistical analyses, with p values less than 0.05 considered significant.

RESULTS

Data were obtained from a total of 3 045 children; 1 756 (58%) males and 1 289 (42%) females. Their mean age was 8 years 1 month (SD 3.84). The distributions according to age, sex, and GMFCS levels are presented in Table I.

Table I: Distribution of age, sex, and GMFCS level

Age (y)	*N* of children	Sex *n* (%)		GMFCS level *n* (%)				
		Male	Female	I	II	III	IV	V
1–3	521	288 (55)	233 (45)	212 (41)	90 (17)	61 (12)	79 (15)	79 (15)
4–6	737	440 (60)	297 (40)	358 (49)	98 (13)	76 (10)	100 (14)	105 (14)
7–9	683	401 (59)	282 (41)	282 (41)	124 (18)	60 (9)	106 (15)	111 (16)
10–12	665	390 (59)	275 (41)	301 (45)	114 (17)	46 (7)	91 (14)	113 (17)
13–15	439	237 (54)	202 (46)	177 (40)	82 (19)	37 (8)	73 (17)	70 (16)
Total	3 045	1 756 (58)	1 289 (42)	1 330 (44)	508 (17)	280 (9)	449 (15)	478 (16)

GMFCS, Gross Motor Function Classification System.

Knee contracture was reported in 685 (22%) of the 3 045 children; 369 (12%) had a mild contracture (−5° to −14°) and 316 (10%) had a severe contracture (−15° or less). Data for passive knee extension were missing for 67 children, 43 of whom were classified as being in GMFCS I. Slightly more males (*n*=419, 24%) than females (*n*=266, 21%) had a knee contracture. The percentages of children with a severe contracture were 12 per cent in GMFCS level III, 25 per cent in level IV, and 35 per cent in level V. A severe contracture was exhibited by 3 per cent of children in the 4- to 6-year-old group and 25 per cent of children in the 13- to 15-year-old group (Table II).

Significant differences were observed in knee extension related to sex (*p*=0.029), age (*p*<0.001), GMFCS level (*p*<0.001), length of the hamstring and gastrocnemius muscles (*p*<0.001), and spasticity of the knee and plantar flexors (*p*<0.001) (Table III). Knee extension was slightly lower in males than in females; the mean directions (circular SDs) were −2.4 degrees (0.2°) and −1.7 degrees (0.2°) respectively. Knee extension decreased with increasing age from 0.9 degrees (0.1°) for the 1-year to 3-year-old group and to −7.1 degrees (0.2°) for the 13-year to 15-year-old group. Knee extension also decreased with increasing GMFCS level from 1.5 degrees (0.1°) for children at GMFCS level I to −10.6 degrees (0.3°) for those at GMFCS level V. The largest difference in knee extension was related to the length of the hamstring muscle; the mean direction of knee extension (circular SD) was 1.0 degree (0.1°) for children with a normal muscle length compared with −19.5 degrees (0.3°) for those with a severe knee contracture (Table III).

In the multiple binary logistic regression model, age, GMFCS level, hamstring and gastrocnemius muscle lengths, and spasticity of the knee flexors were significantly

Table II: Distribution of knee extension in relation to sex, age, and GMFCS level

Variable	Level	Knee extension		
		Normal ROM ≥−4° *n* (%)	Mild contracture −5° to −14° *n* (%)	Severe contracture ≤−15° *n* (%)
Sex	Male	1 300 (75)	220 (13)	199 (12)
	Female	993 (79)	149 (12)	117 (9)
Age (y)	1–3	461 (93)	30 (6)	5 (1)
	4–6	611 (85)	85 (12)	25 (3)
	7–9	576 (77)	90 (13)	66 (10)
	10–12	443 (67)	103 (16)	112 (17)
	13–15	262 (61)	61 (14)	108 (25)
GMFCS level	I	1 214 (94)	73 (6)	0 (0)
	II	447 (89)	49 (10)	4 (1)
	III	199 (72)	43 (16)	33 (12)
	IV	236 (53)	96 (22)	112 (25)
	V	197 (42)	108 (23)	167 (35)

GMFCS, Gross Motor Function Classification System; ROM, range of motion.

Table III: Knee extension in relation to sex, age, GMFCS level, hamstring and gastrocnemius muscle lengths, and spasticity of the knee and plantar flexors

Variable	Level	*n* (%)	Knee extension (°), mean direction (CSD)	*p*-value[a]
Sex	Male	1 719 (58)	−2.43 (0.17)	0.029
	Female	1 259 (42)	−1.65 (0.16)	
Age (y)	1–3	496 (17)	0.90 (0.08)	<0.001
	4–6	721 (24)	0.28 (0.11)	
	7–9	672 (23)	−1.68 (0.14)	
	10–12	658 (22)	−4.26 (0.20)	
	13–15	431 (14)	−7.08 (0.24)	
GMFCS level	I	1 287 (43)	1.50 (0.07)	<0.001
	II	500 (17)	0.96 (0.08)	
	III	275 (9)	−2.44 (0.14)	
	IV	444 (15)	−7.13 (0.20)	
	V	472 (16)	−10.58 (0.25)	
Muscle length				
Hamstring muscle	Normal	1 825 (62)	0.97 (0.08)	<0.001
	Mildly shortened	924 (31)	−4.75 (0.17)	
	Severely shortened	189 (6)	−19.52 (0.30)	
Gastrocnemius muscle	Normal	1 744 (59)	−0.77 (0.15)	<0.001
	Mildly shortened	903 (31)	−2.48 (0.14)	
	Severely shortened	287 (10)	−5.61 (0.17)	
Spasticity				
Knee flexors	0	1 751 (69)	0.41 (0.11)	<0.001
	1–1+	548 (22)	−4.11 (0.18)	
	2–4	238 (9)	−10.70 (0.24)	
Plantar flexors	0	1 279 (53)	−0.32 (0.14)	<0.001
	1–1+	674 (28)	−2.04 (0.17)	
	2–4	480 (20)	−5.08 (0.19)	

[a] *p*-values for the difference in mean direction between group levels using the Watson–Williams test for homogeneity of means. CSD, circular standard deviation; GMFCS, Gross Motor Function Classification System.

Table IV: Results of multiple binary logistic regression for the association between knee contracture (yes/no) and sex, age, GMFCS level, hamstring and gastrocnemius muscle lengths, and spasticity of the knee and plantar flexors

Variable	Level	OR	95% CI	*p*-value
Sex	Male	Ref.		
	Female	0.95	(0.71–1.27)	0.714
Age (y)	1–3	Ref.		
	4–6	1.88	(1.02–3.49)	0.044
	7–9	3.27	(1.80–5.94)	<0.001
	10–12	5.18	(2.84–9.45)	<0.001
	13–15	6.80	(3.62–12.77)	<0.001
GMFCS level	I	Ref.		
	II	0.95	(0.58–1.57)	0.849
	III	3.52	(2.14–5.79)	<0.001
	IV	7.91	(5.03–12.46)	<0.001
	V	13.17	(8.42–20.60)	<0.001
Muscle length				
Hamstring muscle	Normal length	Ref.		
	Mildly shortened	3.05	(2.24–4.17)	<0.001
	Severely shortened	9.86	(5.49–17.69)	<0.001
Gastrocnemius muscle	Normal length	Ref.		
	Mildly shortened	1.70	(1.24–2.32)	0.001
	Severely shortened	3.62	(2.31–5.66)	<0.001
Spasticity				
Knee flexors	0	Ref.		
	1–1+	1.49	(1.03–2.15)	0.035
	2–4	2.10	(1.27–3.47)	0.004
Plantar flexors	0	Ref.		
	1–1+	1.00	(0.70–1.42)	0.984
	2–4	0.80	(0.51–1.24)	0.317

Results based on n=2 168 out of n=3 045 individuals with complete cases (complete data set and no missing variables included in the regression analysis), of which 407 (19%) had knee contracture. Nagelkerke R^2=0.499. CI, confidence intervals; GMFCS, Gross Motor Function Classification System; OR, odds ratios; Ref., reference.

associated with an increased risk of knee contracture (Table IV). The goodness-of-fit was quite high, with Nagelkerke R^2 equalling 0.499. However, neither sex nor spasticity of the plantar flexors was significantly associated with knee contracture. Mildly reduced (OR 3.05) and severely reduced (OR 9.86) hamstring length and reduced gastrocnemius length (OR 1.7–3.62) were associated with an increased risk of knee contracture. A higher degree of spasticity in the hamstrings (OR 1.49–2.10) was also associated with a slightly increased risk of knee contracture. Both reduced hamstring length and reduced gastrocnemius length were more strongly associated with knee contracture than increased spasticity in the knee or plantar flexors.

DISCUSSION

Despite the high prevalence of knee contracture in CP, few studies have investigated this issue in children with CP. We found that almost one out of four children of a total population of children with CP aged up to 15 years, which included all GMFCS levels, had a knee contracture of 5 degrees or more. Children at all GMFCS levels exhibited knee contracture, although this was more common in older children and in those with a lower level of motor function. A strong association was observed between reduced hamstring length and knee contracture. Reduced hamstring and gastrocnemius lengths conferred a higher risk for knee contracture than increased spasticity of the knee flexors, but no significant association was found between knee contracture and spasticity in the plantar flexors.

Our finding that 2 per cent of the 13-year to 15-year-olds had knee contractures compared with 10 per cent of the 4-year to 6-year-olds is consistent with previous studies that have reported an increase in the frequency of knee contractures with age.[5,6] It is also the most frequent contracture in adults with CP.[3] In our study, the frequency of knee contracture increased with age at all GMFCS levels, but was more frequent in children with a lower level of motor function. For example, none of the children at GMFCS level I had a severe contracture compared with 35 per cent of those at level V. Even in children at GMFCS level V who lack the ability to walk, knee contracture is highly associated with postural asymmetries, which may cause progressive deformity such as scoliosis, windswept hip deformity, or hip dislocation when the legs are tilted to the side in the supine position. Limited knee extension also obstructs the ability to find optimal lying and standing positions for these children.[3]

Knee contracture was strongly associated with reduced muscle length, particularly of the hamstrings, but also to the gastrocnemius muscles to some extent. In a study by Nordmark et al.[6] hamstring length measured as the popliteal angle started to reduce on average 4 years before the development of knee contractures in children with CP. Both the hamstring and gastrocnemius are two-joint muscles that pass the knee joint and thus affect its ROM. It is common for nonoperative interventions to maintain the length of the gastrocnemius.[16,17] Short hamstring muscles are a challenge and often require operative interventions such as hamstrings release[18] or extension osteotomy. This study shows that children with a hamstring length of less than 120 degrees have an almost 10-fold increase in the risk of knee contracture.

Molenaers et al.[19] described the importance of treating spasticity to prevent joint deformity. However, our data suggest that knee contracture is more strongly associated with reduced muscle length than with spasticity, which may also contribute to reduced muscle length. It is sometimes difficult to distinguish limb stiffness resulting from muscle shortening from that caused by spasticity. Because muscle stiffness increases with age, this difficulty in differentiating shortening from spasticity may occur more frequently in adolescents than in younger children.[20]

This study had several limitations. The Ashworth Scale is a widely used clinical measure of spasticity, but its validity and precision remain open to question.[14] In this study, only 1 of 2 844 children had a reported grade of 4 (rigid) on the Ashworth Scale for the knee flexors, and only 55 (2%) had a reported grade of 3, although more than 10 per cent of the children had a severe knee contracture. Several children with contracture and spasticity received treatment to reduce spasticity, such as the use of an intrathecal baclofen pump and botulinum neurotoxin injections, which may explain the low numbers reported as grade 3 or 4. In the present study, the low reliability of the Modified Ashworth Scale may have underestimated the effects of spasticity on knee contracture. The reliability is slightly higher for the original Ashworth Scale than for the Modified Ashworth Scale and, in this study, we grouped the 1 and 1+ levels according to the original scale.

Another limitation of this study was the cross-sectional design, which involved the collection of information from the last examination for each child and no longitudinal data to follow the development of contracture over time. Our study does not reflect the natural development of knee contracture because many of the children received treatment to prevent its development. Since the CPUP programme started, the number of children with severe contracture has been reduced and the children receive earlier treatment.[21] However, with no treatment, the relationship between reduced muscle length and development of knee contracture would probably have been even more pronounced. The cut-off value of greater than or equal to 5 degrees (mild contracture) affected the high frequency of knee contracture in the present study. A cut-off value of greater than or equal to 15 degrees (severe contracture) would have reduced the percentage by more than half, from 23 per cent to 11 per cent. The measurement of the popliteal angle was used as a surrogate value for hamstring length. Unless adjusted for, the popliteal angle measured in supine with the opposite leg straight may be affected by a limited hip flexion below 90 degrees or a hip flexion contracture pulling the pelvis forward. However only 218 of 3 045 children (7%) were reported to have a hip flexion contracture exceeding 5 degrees on either side.

One strength of this study is that it included a total population of children with CP at all GMFCS levels aged from 1 year to 15 years. Given the prevalence and distributions of sex and GMFCS levels, the study population is likely to be representative of children with CP in other countries with similar development.

The association between knee contracture and reduced hamstring and gastrocnemius muscle lengths is important for the treatment of children with CP. Chan and Miller[18] described the importance of early interventions and noted that children should not be allowed to develop severe contracture that causes them to lose function and ambulatory potential. Maintaining hamstring and gastrocnemius muscle lengths can reduce the risk of developing severe knee contracture and may prevent contracture of the ankle and hip.

In conclusion, knee contracture is frequent in children with CP at all GMFCS levels and is associated with higher GMFCS level, older age, and reduced length of the hamstring and gastrocnemius muscles.

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Paper II

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RESEARCH ARTICLE

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Interrater reliability for unilateral and bilateral tests to measure the popliteal angle in children and youth with cerebral palsy

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Abstract

Background: Short hamstring muscles can cause several problems for children with cerebral palsy. The results of the clinical measurement of hamstring length are often used in decision-making about treatment of children with cerebral palsy. There are different ways of performing this measurement. The aim of this study was to evaluate the interrater reliability of the unilateral and bilateral measurement of the popliteal angle in children and youth with cerebral palsy.

Methods: Two methods for estimating hamstring length using unilateral and bilateral measurements of the popliteal angle were applied in children with cerebral palsy. Both tests were applied bilaterally by two independent examiners on the same day for each child. The intraclass correlation coefficient (ICC) was calculated to evaluate the interrater reliability of both measurements. Seventy young people with cerebral palsy (32 females, 38 males, mean age 10 years 8 months, range 5–22 years) at Gross Motor Function Classification System levels I ($n = 17$), II ($n = 31$), III ($n = 12$) and IV ($n = 10$) were included.

Results: The interrater reliability was good for both measurements. The ICC values were 0.80 on the right and 0.86 on the left for the unilateral popliteal angle, and 0.82 on the right and 0.83 on the left for the bilateral popliteal angle.

Conclusions: Both unilateral and bilateral measurement of the popliteal angle is a reliable method for estimating hamstring length in children and youth with cerebral palsy.

Keywords: Cerebral palsy, Hamstring muscles, Physical examination, Range of motion, Reproducibility of results

Background

Reduced muscle length is a common problem among children with cerebral palsy because of the effects of this non-progressive brain disorder and secondary problems such as spasticity, immobility, pathological muscle growth and fewer satellite cells compared with children without cerebral palsy [1–3]. Short hamstring muscles can cause gait problems for ambulatory children and can lead to postural asymmetries in standing and lying for non-ambulant children. The hamstring muscles semitendinosus, semimembranosus and biceps femoris (long head) are two-joint muscles that cross both the hip and knee joint, and are active in hip extension and knee flexion movements. Short hamstrings can cause the "crouch gait" and can reduce knee extension during the stance phase of walking in children with cerebral palsy [4, 5].

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Two of five gait patterns described for unilateral cerebral palsy and three of four described for bilateral cerebral palsy involve limited knee extension and potential short hamstring muscles. The gait pattern can be described as equinus, jump knee and crouch gait [6]. Shortened hamstrings are strongly associated with a high risk for knee contracture, and muscle length has a stronger impact on knee contracture development than the level of spasticity [7]. Shortened muscle length of the hamstrings is a common reason for surgery in children with cerebral palsy [8, 9]. Hamstring lengthening is performed to improve gait and/or standing [4, 10].

The average routine for estimating hamstring length in the clinic is the goniometric measurement of the passive range of motion of the knee joint with the patient in the supine position and the hip in flexion [11]. There are different ways of performing this test, two of which are unilateral and bilateral measurements of the popliteal angle. The bilateral popliteal angle test is also called the 90–90 straight-leg test in the literature [12]. The results of the clinical measurement of hamstring length are often used in decision-making about treatment and when evaluating the treatment of children with cerebral palsy [9, 13, 14]. Given the lack of other convenient methods for estimating hamstring length in daily clinical practice and the need for this information for decision-making, it is important to know the reliability of the measurements in current use.

The aim of this study was to evaluate the interrater reliability of unilateral and bilateral tests to measure the popliteal angle when used in children and youth with cerebral palsy.

Methods

The popliteal angle was measured using the unilateral and bilateral tests in 70 children and youth with cerebral palsy. All children were recruited at a rehabilitation centre, between 10 June and 2 September 2019. The inclusion criterion was diagnosis of cerebral palsy. The exclusion criterion was unilateral cerebral palsy to ensure that no unaffected legs were included. All measurements were performed independently by two physiotherapists on the same day for each child. Both the left and right legs were measured, and the order of measurements was randomized for the two examiners. Before the start of the study, the examiners were instructed in performing the two tests using a goniometer and a standardized method. Both physiotherapists had recently graduated and had no previous experience measuring children with cerebral palsy.

The unilateral test was performed with the child in the supine position on the examination table. The examiner held the test leg in 90 degrees of hip flexion and the contralateral leg was fully extended and fixed. The examiner increased the knee extension until the leg was maximally passively extended. The movement was performed slowly (> 5 s for the entire range of motion) and with as much force as required to reach the passive end of motion. Reference marks for the goniometer ran along the femur to the greater trochanter and along the tibia to the lateral malleolus. The bilateral test was performed in a similar way as the unilateral test except that the child's contralateral leg was flexed at the hip to a position in which the anterior superior iliac spine and the posterior superior iliac spine were vertical (Fig. 1).

For both tests, the goniometric measurement of the popliteal angle was defined as the angle, in degrees, of the lower leg relative to full extension (where 0 degrees was equal to full extension). The angle was recorded in 5-degree intervals as 0, 5, 10 or 15 degrees.

Statistical analyses

The interrater reliability was evaluated using the intraclass correlation coefficient (ICC) and 95% confidence interval (CI) with two-way random and absolute agreement for single measures [15]. The ICC takes the degree of disagreement into account and should exceed 0.75 [16]. The ICC was estimated for the left and right sides separately and for the average value of the left and right side of each child. All analyses were performed by independent physiotherapists with no other knowledge of the participants using IBM SPSS Statistics (version 25).

Results

In total, 70 participants (38 males and 32 females) with cerebral palsy were included. Their mean age was 10.8 years (range 5–22 years). The participants were classified according to the Gross Motor Function Classification System (GMFCS) level as follows: I ($n = 17$); II ($n = 31$); III ($n = 12$) and IV ($n = 10$).

The mean popliteal angle was 31 degrees measured in the unilateral test and 15 degrees in the bilateral test, in which the pelvis is tilted slightly more posteriorly. The popliteal angle measured in both tests increased with higher GMFCS level (Table 1).

The mean popliteal angle for both tests also increased with older age and there was a tendency for slightly higher values in males than females (Table 2).

There was a high interrater reliability, with ICCs of ≥0.80 for both the unilateral and bilateral measurements of the popliteal angle (Table 3).

Discussion

This study showed that both the unilateral and bilateral tests to measure the popliteal angle have high interrater reliability for measuring hamstring length in children and youth with cerebral palsy. The participants in this study represented different ages and GMFCS levels. All

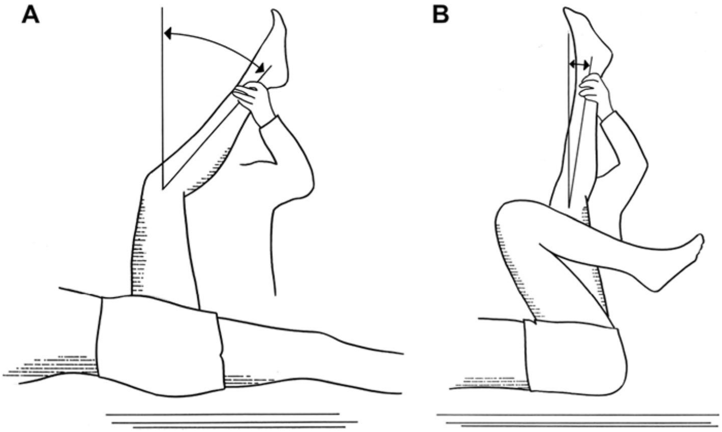

Fig. 1 Tests to measure the popliteal angle. Unilateral test (**a**) and bilateral test (**b**). Reproduced with permission from 'Orthopaedic Management in Cerebral Palsy' by Eugene E. Bleck published by Mac Keith Press (www.mackeith.co.uk) in its Clinics in Developmental Medicine Series, 1987, 0–632–01523-3

participants had bilateral cerebral palsy to ensure that only the affected legs were included. Previous studies have shown that measuring range of motion can be more difficult in children with cerebral palsy than in those without cerebral palsy [17, 18]. A previous study of 15 children with cerebral palsy by Ten Berge et al. [19] showed good intrarater reliability but low interrater reliability for the unilateral popliteal test.

Table 1 Mean value, standard deviation (SD) and range for the unilateral and bilateral popliteal angle tests

GMFCS level (n)	Type of test	Examiner	Mean	SD	Range
I (17)	Unilateral	1	27.35	10.1	10–50
		2	25.59	10.66	10–50
	Bilateral	1	13.53	10.5	0–35
		2	13.38	9.68	0–35
II (31)	Unilateral	1	33.31	10.69	15–65
		2	31.86	9.83	15–50
	Bilateral	1	16.21	9.9	0–35
		2	14.92	10.49	0–35
III (12)	Unilateral	1	33.96	7.65	20–50
		2	32.5	8.66	20–50
	Bilateral	1	17.71	8.36	0–35
		2	17.50	8.26	0–35
IV (10)	Unilateral	1	32.0	10.12	15–55
		2	32.75	11.99	20–60
	Bilateral	1	14.75	7.86	0–25
		2	17.75	15.96	0–60

Popliteal angles of the two examiners presented separately for each level of the Gross Motor Function Classification System (GMFCS)

The unilateral popliteal angle reported was lower in this study (mean 31 degrees) than in other studies. Choi et al. [20] reported a mean unilateral popliteal angle of 37 degrees in children with GMFCS I or II. Bell et al. [21] reported a mean angle of 28 or 59 degrees, in children with CP depending on walking function and White et al. [22] reported a mean of 57.6 degrees for 8 to 12-year-old children with CP but did not specify whether the test was bilateral or unilateral. None of the previous studies specified how much pressure the examiners

Table 2 Popliteal angle with mean value and standard deviation (SD) for age, sex and GMFCS

	Unilateral test Mean (SD)	Bilateral test Mean (SD)
GMFCS		
I	26.47 (10.17)	13.46 (9.9)
II	32.58 (10)	15.56 (10.02)
III	33.23 (7.84)	17.6 (8.13)
IV	32.38 (10.82)	16.25 (11.32)
Sex		
Female	29.14 (9.04)	12.81 (8.03)
Male	32.89 (10.54)	17.76 (10.61)
Age group		
0–6	28 (9.23)	11.63 (8.46)
7–12	30.64 (9.34)	15.03 (8.71)
13–18	32.98 (11.12)	17.31 (11.62)
19–22	47.5 (10.61)	33.75 (10.61)

Average values for the unilateral and bilateral popliteal angle tests of both examiners together presented for the different sub groups

Table 3 Interrater reliability for the unilateral and bilateral popliteal angle tests

	Unilateral test		Bilateral test	
	ICC	95% CI	ICC	95% CI
Right				
Single measure	0.80	0.67–0.87	0.82	0.72–0.88
Left				
Single measure	0.86	0.78–0.91	0.83	0.74–0.89

Intraclass Correlation Coefficients (ICC) with 95% Confidence Intervals (CI) for the right and the left leg

applied to extend the knee. The examiners in our study were instructed to extend the child's leg with as much pressure as tolerated by the child and to reach the end range of movement. The range of 5–60 degrees reported by Choi et al. [20] was similar to the 0–60 degrees observed in our study. This wide range highlights the variability of measurement of the popliteal angle in children with cerebral palsy, which would affect the mean value in a smaller study population. Also capsular contracture may affect the range of motion. Reduced popliteal angle is one criterion for surgery to lengthen the hamstrings, but this should not be the only criterion [19, 23, 24].

The unilateral and bilateral tests can yield different results, as measured in degrees, according to the position of the pelvis. Therefore, when reporting results, it is important to indicate the type of measurement used and to consider the pelvic position [12]. A more posteriorly tilted pelvis, as in the bilateral test, will automatically yield a smaller popliteal angle. The bilateral test allows for relaxation of the hip flexors of the contralateral leg [25] and may be easier to use when measuring children with hip flexion contractures. A study by Manikowska et al. [23] showed a greater muscle activation of the contralateral leg during the unilateral compared with the bilateral test. The authors suggested that this muscle activation may affect the result and, therefore, should be considered.

The speed at which the knee extends during measurement is also important. Choi et al. [20] used the sum of a fast speed and slow speed measure of the popliteal angle when considering both spasticity and mechanical resistance, which can be important when evaluating treatment [26]. The aim of our study was to compare the interrater reliability between two frequently used clinical test methods to measure the muscle length of the hamstrings.

The popliteal angle was measured with a goniometer by two independent physiotherapists on the same day for all participants to minimize the effects of true changes in muscle length over time. The ICC was used to evaluate interrater reliability, and the values for both tests exceeded the minimum of 0.75 recommended by Streiner et al. [16] for clinical tests.

One limitation of this study is the potential influence on the examiners by the first test when performing the second test. However, the two examiners performed their measurements independently of each other and with the same conditions. The examiners had no personal interest in the results of the study that may have affected their measurements.

Another limitation of our study is that no children at GMFCS level V were included. It is possible that children at GMFCS level V are more difficult to measure depending on the severity and number of contractures. The tests were performed passively in the supine position, and it is unlikely that this affected the measurements in this study. However, the lack of children at GMFCS level V likely affected the mean value obtained in the tests to measure popliteal angle.

Some authors have questioned the measurement of hamstring length and suggest that the popliteal angle is a substitute measure and not a true measure of hamstring length [24, 27]. Other ways of measuring hamstring length, for example software techniques, have been used to evaluate the results of hamstring lengthening in children with cerebral palsy [28, 29]. A study by Park et al. showed that hamstring length measured with musculoskeletal modelling software correlated with the results of popliteal angle measurement [27].

Conclusions

The popliteal angle is part of the regular examination of children with cerebral palsy. Given the variability in measuring methods and results, it is important to also include other examinations, such as gait analysis, when considering potential treatments [29].

This study showed high interrater reliability for both the unilateral and bilateral tests to measure popliteal angle. The ICCs were high and did not differ significantly between the two methods. Our finding suggests that both tests are reliable methods of measuring popliteal angle in children and youth with cerebral palsy.

Abbreviations

CP: Cerebral palsy; CI: Confidence interval; GMFCS: Gross Motor Function Classification System; ICC: Intraclass correlation coefficient

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Authors' contributions

EC designed the study, analysed the data and drafted the manuscript. JK designed the study, collected the data and improved and revised the manuscript. MJ designed the study, recruited the children and improved and revised the manuscript. ERB designed the study, analysed the data and improved and revised the manuscript. All authors approved the final draft.

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Availability of data and materials
The data that support the findings of this study are available from the corresponding author, upon reasonable request.

Declarations

Ethics approval and consent to participate
The study was approved by the Ethical committee of Poznan University of Medical Sciences (nr 244/20).
Written informed consent was obtained from a parent or guardian for participants under 18 years old.

Consent for publication
Consent to use data for research was provided by all families participating in this study.

Competing interests
The authors declare that they have no competing interests.

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Publisher's Note

Paper III

OPEN ACCESS

Knee and foot contracture occur earliest in children with cerebral palsy: a longitudinal analysis of 2,693 children

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Background and purpose — Joint contracture is a common problem among children with cerebral palsy (CP). To prevent severe contracture and its effects on adjacent joints, it is crucial to identify children with a reduced range of motion (ROM) early. We examined whether significant hip, knee, or foot contracture occurs earliest in children with CP.

Patients and methods — This was a longitudinal study involving 27,230 measurements obtained for 2,693 children (59% boys, 41% girls) with CP born 1990 to 2018 and registered before 5 years of age in the Swedish surveillance program for CP. The analysis was based on 4,751 legs followed up for an average of 5.0 years. Separate Kaplan–Meier (KM) curves were drawn for each ROM to illustrate the proportions of contracture-free legs at a given time during the follow-up. Using a clustered bootstrap method and considering the child as the unit of clustering, 95% pointwise confidence intervals were generated for equally spaced time points every 2.5 years for each KM curve.

Results — Contracture developed in 34% of all legs, and the median time to the first contracture was 10 years from the first examination. Contracture was most common in children with a higher Gross Motor Function Classification System (GMFCS) level. The first contracture was a flexion contracture preventing dorsiflexion in children with GMFCS level I or II and preventing knee extension in children with GMFCS level III to V.

Interpretation — Early interventions to prevent knee and foot contractures in children with CP should be considered.

Joint contracture is a common problem in children with cerebral palsy (CP) (Rosenbaum et al. 2007). Spasticity, muscle imbalance, inability to move, and muscle pathology constrain normal muscle growth and lead to dynamic contracture followed by static joint contracture over time because of tight muscles surrounding the joints (Barrett and Lichtwark 2010). Children with CP exhibit increased sarcomere length and reduced number of satellite cells, both of which affect the ability to maintain muscle length during development and bone growth (Barrett and Lichtwark 2010, Smith et al. 2013). The risk of contracture increases with age and higher level on the Gross Motor Function Classification System (GMFCS) (Nordmark et al. 2009). Some 60% of adults with CP experience contracture in the lower limbs (Agustsson et al. 2018).

Joint contracture prevents mechanical alignment of the joints, which affects standing and lying positions, and the quality and energy cost of gait (Raja et al. 2007). Contracture of the foot, knee, or hip joint may also affect adjacent joints and lead to severe postural asymmetries, windswept hips, and scoliosis (Agustsson et al. 2017, 2018, Pettersson et al. 2020). Contracture is often associated with pain, which occurs most frequently in the lower limbs of children with CP (Alriksson-Schmidt and Hägglund 2016, Blackman et al. 2018). To prevent severe joint contracture and reduce its effects on adjacent joints, it is crucial to identify children with reduced range of motion (ROM) to begin targeted treatment early (Chan and Miller 2014).

We analyzed whether contracture preventing hip extension, knee extension, or foot dorsiflexion occurs first in children with CP with GMFCS level I to V.

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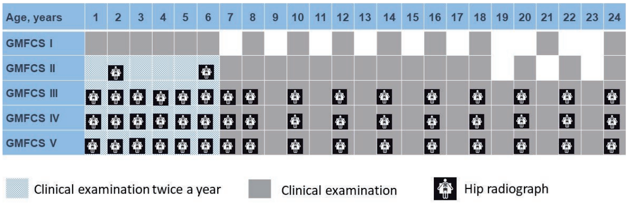


Figure 1. Guidelines for clinical examination and hip radiograph within the Swedish Cerebral Palsy Follow-up Program (CPUP).

Patients and methods

This was a prospective study based on register data from the Swedish Cerebral Palsy Follow-up Program (CPUP), which includes > 95% of all children with CP in Sweden. We analyzed all measurements reported from the start of the program in October 1994 until the end of June 2018 and included all children born 1990 to 2018 and registered in the CPUP before 5 years of age. Children registered at 5 years of age or later were excluded. Children included in the CPUP were examined every 6 months, once a year, or every other year, depending on their age and GMFCS level (Figure 1).

The systematic follow-up includes several variables such as reports of surgery, CP subtype, and clinical examinations of passive ROM and gross motor function. The full protocol is available at: https://cpup.se/in-english/manuals-and-evaluation-forms/.

Gross motor function was classified by the child's physiotherapist into level I to V according to the expanded and revised version of the GMFCS (Palisano et al. 2008). Passive hip extension, knee extension, and dorsiflexion were assessed by goniometric measurements in standardized positions. Hip extension was measured with the child in the prone position with legs over the end of the examining table and the pelvis straight. Knee extension and foot dorsiflexion were measured with the child in the supine position with the hip and knee extended.

Contracture was defined as hip and knee flexion contracture of 10° or more and plantarflexion contracture of at least 10° in extension of the hip, knee, or foot. Children with unilateral spastic CP were identified, and only the affected side was used in the analyses. For children with ataxic, dyskinetic, or spastic bilateral CP, both legs were included.

Each leg that underwent soft tissue surgery or bony surgery and both legs of children with an intrathecal baclofen pump (ITB) or who underwent a selective dorsal rhizotomy (SDR) operation before the onset of the first contracture were censored from the analyses at the date of surgery. If information on which leg was operated on was missing, both legs were censored at the date of surgery.

Reports of spasticity-reducing surgery, such as that to insert an ITB or SDR, and the date of surgery or pump insertion were extracted. Surgery of the lower limbs was recorded in the database and was grouped into either soft tissue surgery or bony surgery for the hip, knee, or foot along with the date of surgery for the left and right leg. Examples of the soft tissue surgeries performed were adductor tenotomy, hamstring or Achilles tendon lengthening, and tendon or muscles transfer. Examples of the bony surgeries reported were osteotomy, physiodesis, or arthrodesis. Surgery on the upper extremity or the spine, fracture, extraction of osteosynthesis material, or treatment with botulinum toxin injection were not censored and data for children who received these treatments were retained in the analyses.

Statistics

Hip extension, knee extension, and foot dorsiflexion were dichotomized into 2 groups: no contracture and contracture. Both legs for each child were then followed up from inclusion in the CPUP surveillance program until the last examination or the date of surgery. The follow-up time was calculated for each leg for each child, and separate Kaplan–Meier (KM) curves were drawn for each leg's ROM to illustrate the proportions of contracture-free legs at a given time during the follow-up. For children with unilateral spastic CP, only the affected side was included in the analyses. Using a clustered bootstrap method and considering the child as the unit of clustering, 95% pointwise confidence intervals (CI) were generated for equally spaced time points every 2.5 years for each KM curve. IBM SPSS Statistics (version 25.0; IBM Corp, Armonk, NY, USA), STATA (version 14, Stata-Corp, College Station, TX, USA), and R (R Foundation for Statistical Computing, Vienna, Austria) were used for the statistical analyses. Categorical variables are described by frequency (n) and percentage (%).

Ethics, funding, and potential conflicts of interest

The study was approved by the Medical Research Ethics Committee in Lund (383/2007, 443-99), and permission was obtained to extract data from the CPUP registry. The study received funding from Stiftelsen för bistånd åt rörelsehindrade i Skåne, Promobilia, Forte and Region Kronoberg. The funding sources had no decision-making role or influence on the study design, data collection, data analysis, data interpretation, or writing of the report. The authors declare that they have no conflicts of interest.

Results

2,693 children (1,598 boys, 1,095 girls) with an examination before the age of 5 years were reported between 1994 and

Table 1. Contracture events and number of person-years according to the Gross Motor Function Classification System I–V (GMFCS)

GMFCS level	n (%)	Contracture, legs (%)	Person-years (mean)
I	1,901 (40)	237 (13)	11,450 (6.0)
II	765 (16)	183 (24)	4,495 (5.9)
III	540 (11)	247 (46)	2,250 (4.2)
IV	752 (16)	436 (58)	3,166 (4.2)
V	793 (17)	499 (63)	2,406 (3.0)
Total	4,751	1,602 (34)	23,768 (5.0)

Table 2. Age at first hip, knee, and foot operation

Type of surgery	Age at first operation Median	1st quartile	3rd quartile
Hip, soft tissue	4.0	3.0	6.0
Hip, bony	6.5	4.0	10
Knee, soft tissu	9.0	7.0	12
Knee, bony	10	5.5	12
Foot, soft tissu	7.0	5.0	10
Foot, bony	11	8.5	14

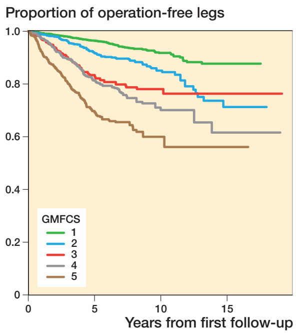
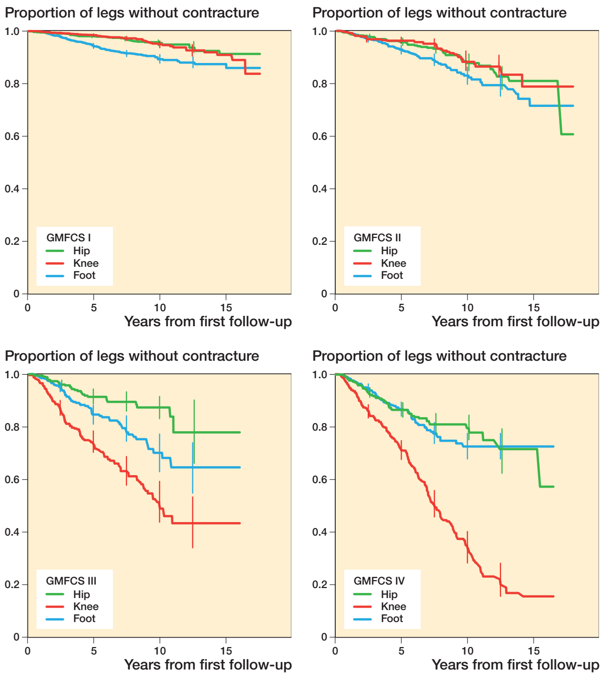


Figure 2. Proportions of operation-free legs from the time of the first measurement for each GMFCS level.



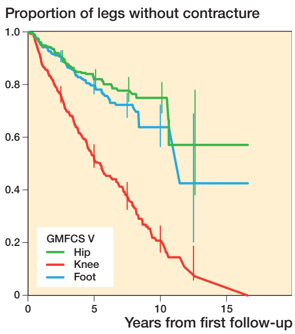


Figure 3. Proportions of legs free from hip, knee, and foot contracture, stratified by GMFCS level and 95% pointwise confidence intervals for equally spaced time points every 2.5 years during the follow-up.

2018. The analysis was based on 4,751 legs followed up for an average of 5.0 years (only the affected leg was included for children with unilateral CP). Contractures were in general most common at higher GMFCS levels (Table 1). There were 27,230 measurement occasions.

Within the 10 years of follow-up, 937 legs (20%) had been operated on. The most common operation was soft tissue surgery of the hip (316 legs, 6.7%), followed by soft tissue surgery of the foot (201 legs, 4.2%), and bony surgery (osteotomy) of the hip (151 legs 3.2%). The median ages were 4 years for soft tissue surgery of the hip, 7 years for soft tissue surgery of the foot, and 6.5 years for bony surgery of the hip (Table 2). The proportions of legs not operated on in children with different GMFCS levels are presented in Figure 2.

1,602 contractures were recorded. A contracture developed in 34% of all legs, and the median time to the first contracture was 10 years from the first examination within the follow-up program. Contracture occurred most frequently in children with a higher GMFCS level. The first contracture to occur in children with GMFCS level I or II was a foot or ankle con-

Table 3. Proportions of legs free from hip, knee, and foot contracture, stratified by GMFCS level, and 95% pointwise confidence intervals after 10 years of follow-up

GMFCS level	Hip	Knee	Ankle
I	1.0 (0.9–1.0)	1.0 (0.9–1.0)	0.9 (0.9–0.9)
II	0.9 (0.8–0.9)	0.9 (0.9–0.9)	0.8 (0.8–0.9)
III	0.9 (0.8–0.9)	0.5 (0.4–0.6)	0.7 (0.6–0.8)
IV	0.8 (0.8–0.8)	0.3 (0.3–0.4)	0.7 (0.7–0.8)
V	0.8 (0.7–0.8)	0.2 (0.2–0.3)	0.6 (0.6–0.7)

tracture, and a knee contracture was the first contracture to develop in children with GMFCS level III to V. Proportions of legs free from hip, knee, and foot contracture in each GMFCS level are presented in Table 3 and with separate KM curves in Figure 3.

Discussion

We identified that the first joint contracture to occur in the lower limb involved foot contracture for children with GMFCS level I or II and knee contracture for children with GMFCS level III to V. Children classified with GMFCS level I and II are ambulant whereas those classified with GMFCS level III to V in general rely on wheeled mobility and spend more time sitting with flexed knees. This could explain the sequence in which the contracture presents. This seems to follow the same pattern as previously reported for pain localization in children with CP, in which children with GMFCS level I or II report pain primarily in the feet and those with GMFCS III to V pain in the knees and hips (Alriksson-Schmidt and Hägglund 2016).

These results are consistent with those of a previous study (Nordmark et al. 2009) that reported decreasing ROM from 2 years of age in all lower limb joints. Together, these findings suggest that contracture should be treated early given that ROM seems to decrease over time. Cloodt et al. (2018) found that hamstring length, measured as the unilateral popliteal angle, and dorsiflexion of the foot are strongly associated with the development of knee contracture, whereas spasticity had a significantly smaller effect. Contracture in the lower limbs affects the mechanical position of both the affected and adjacent joints. Preventing normal movements and forces around the joint may increase the risk for additional contractures.

Our study excluded children whose first measurement was at 5 years of age or later. The reason for this was to increase the opportunity of identifying the first contracture and to reflect the natural development of contracture as much as possible because information on treatment and surgery before enrollment in the CPUP was not available in these children. Among the children included, their access to early service and follow-up during their first years of life should be considered. All children with CP in Sweden have access to free health care and interventions from multiprofessional habilitation centers (Alriksson-Schmidt et al. 2017).

Surgery or treatment with SDR and ITB can affect the development of contracture in a specific joint as well as in adjacent joints (McGinley et al. 2012).

There are several strengths and limitations of our study. Classification of subtype was missing in several cases and therefore not included. The contractures were recorded by goniometric measurement, which is a standard measurement in clinical settings but whose reliability varies according to the joint and position (Hancock et al. 2018, Kim et al. 2018). The results included in our study were based on repeated measurements taken by many different examiners, and this may have introduced information bias or measurement errors. In survival analysis, as we used, measurement error can introduce bias. However, this bias has been shown to be small when differences between groups are moderate in terms of hazard ratios (Oh et al. 2018). Furthermore, in some situations in which bias was found to be substantial, bias attenuates observed differences between groups. For our study, this would mean an underestimation of differences in risk of having a first contracture in a specific joint compared with another joint. We validated the data for incorrectly reported measurements to reduce the risk of incorrect outliers. Examiners in the CPUP are encouraged to practice and learn to perform the register's standardized measurements, which has been shown to be important for reliability (Fosang et al. 2003).

One limitation of our study was the cutoff values for defining contracture. We chose –10° as the cutoff for all joints to be consistent with the reference values used in the surveillance program. Hip or knee extension or foot dorsiflexion of –10° or less causes functional limitations and affects mechanical alignments. To evaluate the effects of this cutoff, we also ran the statistical analyses using less than 0° as the cutoff and found a similar outcome. For children with GMFCS level III to V, the statistical analyses indicated that the conclusion of this study was insensitive to the choice of cutoff values for contracture, that is, 0° or –10°. It is more difficult to draw a conclusion from the trend for GMFCS level I and II because the fewer contractures at these levels of motor function (Nordmark et al. 2009) make it harder to detect statistically significant differences.

Another limitation was the age of the children censored because of surgery. In the CPUP surveillance program, children at risk for hip dislocation have their first soft tissue surgery of the hip early (median age 4 years), which precedes the first foot and knee surgery by 3 and 5 years, respectively. In some cases, hip surgery is likely to occur before confirmed contracture of the hip because of the indication for surgery based solely on lateralization on radiographs (Hägglund et al. 2005). In our study, the operated leg was censored from analyses at the time of the first operation. This may have affected the results, especially for children with GMFCS level IV or V, because many of these children receive their hip operation early and therefore were not included in the analysis.

Botulinum toxin A injection is a common treatment for spasticity in children with CP in Sweden (Franzén et al. 2017) and it could be argued that this interfered with our results. However, given the large number of treated children and the fact that the effect of botulinum toxin is not permanent, these children were included.

The intervals between examinations in the CPUP are based on GMFCS level and age, from twice a year up to the age of 6 years and then every year (GMFCS II–V) or every second year (GMFCS I) (Figure 1). This indicates that some differences between GMFCS levels may stem from differences in detection, probably because of the different number of examinations. Although this may explain some of the differences in the proportion of contractures, it does not explain the fact that contractures occurred in a different sequence, with dorsiflexion first in children with GMFCS level I or II and knee extension first in children with GMFCS level III to V.

Our longitudinal study covers a time period of 24 years and during this period several interventions have changed. Physiotherapy interventions have changed from hands-on sessions to a greater focus on activity and participation. The use of assistive devices and orthoses has increased, and interventions are primarily integrated into the child's everyday life. There are regional differences in pediatric physiotherapy interventions in Sweden (Degerstedt et al. 2020). More intensive treatment has been associated with better outcome for the children (Storvold et al. 2020). Surgeries are more proactive rather than reactive today and the amount of surgeries has decreased (Hägglund et al. 2005).

Our study indicates that contractures in the lower limb occur early. It seems possible to avoid major surgeries by monitoring patients with CP and treating contractures early on with physiotherapy, orthoses, and botulinum toxin (Hägglund 2005, Novak et al. 2013). Knowledge regarding the occurrence of the first contracture is important to be able to provide early treatment.

An ankle–foot orthosis (AFO) is used widely to facilitate dorsiflexion and to improve walking ability, gait pattern, or foot alignment when standing. In Sweden, 50% of children with CP use an AFO, and the use of AFO starts early and increases with age up to 5 years (Wingstrand et al. 2014). A similar trend has been reported for the development of spasticity of the gastrosoleus muscle, which peaks at age 5–6 years in children with CP (Lindén et al. 2019). Knee orthoses can also be used to increase ROM, but this has not been evaluated widely (Laessker-Alkema and Eek 2016). Most children with CP in Sweden who require a standing device use individually molded hip–knee–ankle–foot orthoses for mechanical alignment of all the segments of the lower extremities.

Botulinum toxin is frequently used in children with CP in Sweden (Franzén et al. 2017). It is most frequently injected into the gastrocnemius muscle to improve gait and increase ROM. The treatment is usually followed by use of orthosis or in some cases serial casting. Botulinum toxin is more frequently used in younger children (Franzén et al. 2017) and its use corresponds to the development of spasticity that increases over the first 5 years of life and then decreases after 6 years of age (Lindén et al. 2019) and GMFCS level I or II. Our results are consistent with this observation that foot contracture occurred first in children with GMFCS levels I–II. Botulinum toxin injection into the hamstring muscles is most common in older children with GMFCS level IV or V (Franzén et al. 2017). Our study showed that knee contracture occurs first in children with GMFCS level IV or V but that contracture occurs at an early age.

It is reasonable to think that a contracture in a joint affects the adjacent joints, which are then at risk for developing contracture. With a knee joint contracture, the hip will not be extended and flexion contracture in connection with abduction or adduction is likely to occur. Knowledge concerning the sequence of the development of contracture in children at different GMFCS levels is crucial for treating the joints early on and preventing the sequence of contractures following the first one.

In conclusion, lower limb contracture occurs first in the foot of children with GMFCS level I and II and in the knee in children with GMFCS level III to V. Early interventions to prevent knee and foot contractures in children with CP should be considered.

Abbreviations

AFO: ankle–foot orthosis; CP: cerebral palsy; CPUP: Cerebral Palsy Follow-up Program (Sweden); GMFCS: Gross Motor Function Classification System; ITB: intrathecal baclofen pump; KM: Kaplan–Meier; ROM: range of motion; SDR: selective dorsal rhizotomy.

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Paper IV

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RESEARCH Open Access

Sequence of flexion contracture development in the lower limb: a longitudinal analysis of 1,071 children with cerebral palsy

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Abstract

Background: To prevent severe contractures and their impact on adjacent joints in children with cerebral palsy (CP), it is crucial to treat the reduced range of motion early and to understand the order by which contractures appear. The aim of this study was to determine how a hip–knee or ankle contracture are associated with the time to and sequence of contracture development in adjacent joints.

Methods: This was a longitudinal cohort study of 1,071 children (636 boys, 435 girls) with CP born 1990 to 2018 who were registered before 5 years of age in the Swedish surveillance program for CP and had a hip, knee or ankle flexion contracture of ≥ 10°. The results were based on 1,636 legs followed for an average of 4.6 years (range 0–17 years). The Cox proportional-hazards model adjusted for Gross Motor Function Classification System (GMFCS) levels I–V was used to compare the percentage of legs with and without more than one contracture.

Results: A second contracture developed in 44% of the legs. The frequency of multiple contractures increased with higher GMFCS level. Children with a primary hip or foot contracture were more likely to develop a second knee contracture. Children with a primary knee contracture developed either a hip or ankle contracture as a second contracture.

Conclusions: Multiple contractures were associated with higher GMFCS level. Lower limb contractures appeared in specific patterns where the location of the primary contracture and GMFCS level were associated with contracture development in adjacent joints.

Keywords: Cerebral palsy, Range of motion, Contracture, Joint, Hip, Knee, Foot

Background

Lower limb contractures are often present in children with cerebral palsy (CP). A contracture is defined as reduced joint range of motion (static contracture). The risk of contractures increases with age and severity of the disease [1, 2]. The causes responsible for the development of contractures are not fully understood but are usually referred to spasticity, muscle pathology, muscle weakness, biomechanical alignment, and positioning [3–5]. Contractures are associated with pain and affect the child's ability to walk, stand, or transfer [6]. Treatment is usually conservative when the contracture first appears and involves physical therapy, orthoses, casting, and treatment for spasticity. Preventive treatment might reduce the need for surgery and should be initiated as early as possible [7].

Children with CP are at risk of developing contractures in multiple joints [8]. It is likely that a single contracture increases the risk of further contractures because of the changes in biomechanical alignment and positioning.

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Previous studies have shown that hip and knee contractures increase the risk of scoliosis [9, 10]. Therefore, it is important to understand the pattern in which contractures occur to be able to customize effective treatment strategies and prevent severe contractures.

The first contracture to occur in the lower limb varies according to the Gross Motor Function Classification System (GMFCS) level [11]. Children at GMFCS level I or II are most likely to develop an ankle contracture first, whereas children at GMFCS levels III–V are most likely to develop a knee contracture first [12]. To our knowledge, no studies have investigated the association between the location of the first contracture with further contracture development in the lower limb in children with CP.

The aim of this study was to analyze how a hip, knee or ankle contracture affects the time to and sequence of contracture development in adjacent joints in children with CP at GMFCS levels I–V.

Methods

This was a longitudinal cohort study based on register data from the Swedish Cerebral Palsy Follow-Up Program (CPUP), which includes > 95% of all children with CP in Sweden. Children with at least one flexion contracture in the hip, knee, or ankle joint were included in this study. The study included all measurements reported in the registry since the start of the program in October 1994 until the end of June 2018. All children born in 1990–2018 who were registered in the CPUP before 5 years of age were included. Children registered at 5 years of age or later were excluded. According to CPUP, children are examined every 6 months, once a year, or every other year, depending on their GMFCS level and age [12].

The follow-up includes several variables, such as examination of gross motor function, passive range of motion (ROM), and surgeries reported by the child's multiprofessional habilitation team and orthopedic department. The full protocol is available at https://cpup.se/in-english/manuals-and-evaluation-forms/. The examiners have access to the child's previous measurements. The CPUP registry has a yearly reporting rate of 90–95%.

Gross motor function was classified as levels I–V according to the expanded and revised version of the GMFCS [13]. The GMFCS level from the child's most recent visit were used. Passive ROM was measured using a universal goniometer and standardized positions, and included hip extension (Thomas test), knee extension, and ankle dorsiflexion (knee extended). The range of motion was rounded to the nearest 0 or 5 degrees according to the manual. A contracture was defined as a flexion contracture of at least 10° of the hip and knee or at least 10° plantar flexion of the ankle. The primary outcome was the onset and location of a second contracture.

Table 1 Number of legs and involvement for children at GMFCS level I-V at baseline

GMFCS level	Children N (%)	Legs N (%)	Involvement	
			Unilateral CP Legs N (%)	All other subtypes Legs N (%)
I	185 (17)	212 (13)	115 (54)	97 (46)
II	206 (19)	269 (16)	68 (25)	201 (75)
III	156 (15)	249 (15)	14 (6)	235 (94)
IV	234 (22)	405 (25)	8 (2)	397 (98)
V	290 (27)	501 (31)	0 (0)	501 (100)
Total	1,071 (100)	1,636 (100)	205 (13)	1431 (87)

GMFCS Gross Motor Function Classification System

For children with unilateral CP, only the affected leg was included in the analyses. Children included at baseline had not received lower limb surgeries, selective dorsal rhizotomy operation or an intrathecal baclofen pump before their first hip, knee or ankle contracture occurred. Each leg that underwent soft tissue or bone surgery any time after enrollment, that potentially could influence the risk of further contracture development, was censored at the date of surgery. Examples of reported soft tissue surgeries were adductor tenotomy, hamstring or Achilles tendon lengthening, and tendon or muscle transfer. Examples of reported bone surgeries were osteotomy, physiodesis, or arthrodesis [12]. Both legs were censored from the analysis at the date of a selective dorsal rhizotomy or insert of an intrathecal baclofen pump, or if information about which leg was operated on was missing. Surgery to the upper extremity or the spine, treatment with botulinum toxin injection, oral baclofen medication, orthotics, or serial casting were not censored, and the data for children who received these treatments were retained in the analyses.

Statistical analysis

All legs with a contracture were followed up individually from the date of the first contracture until the development of the second contracture, the last examination, or date of surgery. The time from first to second contracture was analyzed, for each leg for each child, using the Cox proportional-hazards model, adjusting for GMFCS level. The model assumptions for proportional hazards was tested and fulfilled. This was done first for all legs and then separately according to the joint where the first contracture occurred. Legs with more than one contracture measured at baseline were included in the descriptive analyses (Tables 1 and 2) and when analyzing the time between contractures. They were however excluded when analyzing the sequences of contracture development (Table 3). The chi-squared test was used to analyze in which joint the second contracture occurred according to the GMFCS level; this analysis was performed separately for joints affected by a baseline contracture of the hip, knee, or ankle [14]. The p-for-trend was calculated using logistic regression to analyze systematic, linear, associations of contracture development in these joints

Table 2 Location of the baseline contracture, prevalence of a second contracture during follow-up and number of limb- years for children at GMFCS I-V

GMFCS level	Location of the first contracture				Second contracture		Limb-years (mean)
	Legs N (%)				Legs N (%)		
	Hip	Knee	Ankle	> 1 joint		> 1 joint	
I	41 (19)	83 (39)	78 (37)	10 (5)	53 (25)	2 (4)	899 (4.2)
II	43 (16)	98 (36)	112 (42)	16 (6)	101 (38)	2 (2)	989 (3.7)
III	40 (16)	124 (50)	66 (27)	19 (8)	115 (46)	3 (3)	747 (3.0)
IV	58 (14)	210 (52)	90 (22)	47 (11)	203 (50)	8 (4)	1,212 (3.0)
V	77 (15)	294 (59)	90 (18)	40 (8)	248 (50)	9 (4)	1,256 (2.5)
Total	259 (16)	809 (49)	436 (27)	132 (8)	720 (44)	24 (3)	5,103 (3.1)

GMFCS Gross Motor Function Classification System

Table 3 Results of the chi-squared and p-for-trend analyses of the associations between each baseline contracture and the location of a second contracture during follow-up at each level I to V of the Gross Motor Function Classification System (GMFCS) for legs that developed > 1 lower limb contracture. Only legs with one single baseline and one single follow up contracture were included

First contracture	GMFCS level	Legs N	Second contracture			*p* value
			Hip N (%)	Knee N (%)	Ankle N(%)	
Hip joint						0.002[a]
	I	8	–	5 (63)	3 (37)	< 0.001[b]
	II	21	–	10 (48)	11 (52)	
	III	19	–	12 (63)	7 (37)	
	IV	28	–	22 (79)	6 (21)	
	V	45	–	40 (89)	5 (11)	
	Total	121	–	89 (74)	32 (26)	
Knee joint						0.003[a]
	I	15	4 (27)	–	11 (73)	< 0.001[b]
	II	32	7 (22)	–	25 (78)	
	III	54	25 (46)	–	29 (54)	
	IV	80	36 (45)	–	44 (55)	
	V	111	65 (59)	–	46 (41)	
	Total	292	137 (47)	–	155 (53)	
Ankle joint						< 0.001[a]
	I	18	3 (17)	15 (83)	–	0.59[b]
	II	30	10 (33)	20 (67)	–	
	III	20	3 (15)	17 (85)	–	
	IV	40	7 (17)	33 (83)	–	
	V	43	9 (21)	34 (79)	–	
	Total	151	32 (21)	119 (79)	–	

[a] chi-squared test

[b] *p*-for-trend analysis

relative to the GMFCS level. IBM SPSS Statistics (version 26.0; IBM Corp, Armonk, NY, USA) and R 3.6.1 (R Foundation for Statistical Computing, Vienna, Austria) were used in the statistical analyses. Categorical variables were described in frequency (n) and percentage (%).

Results

In total 1,071 children with a lower limb contracture and at least one examination after their initial contracture were included at baseline. The analysis was based on 1,636 legs followed up for an average of 4.6 years (range 0–17 years) after the onset of the first contracture (Table 1). The distribution of the primary contracture varied between GMFCS levels, and multiple contractures were most common at higher GMFCS levels. Within the follow-up period, 916 of the legs (56%) did not develop further contractures, while 720 legs (44%) developed additional contractures (Table 2).

In total 132 legs had more than one contracture occurring at baseline. These were counted multiple times; both contractures occurring simultaneously were treated as the first contracture and the potentially following contracture(s) were treated as the second (or third) contracture (Fig. 1). Lower limb surgery was performed on 408 legs (25%) during the follow-up period and were censored from the analysis at the date of surgery. The number lost to follow-up was negligible.

The children's median age for a reported second contracture was 10.8 years. The risk of a second contracture increased with older age and higher GMFCS level (Fig. 2). The second contracture occurred after an average of 5 years from the first contracture (Fig. 3).

The time from the first to second contracture varied depending on the location of the first contracture. Legs with a primary hip or ankle contracture developed a second contracture earlier than those with a primary knee contracture (Fig. 4).

Legs with more than one baseline contracture were not included in the following analyses.

When analyzing legs with one single baseline contracture and one single follow-up contracture, hip contracture was the primary contracture in 259 legs (Table 3). A second contracture developed during follow-up in 121 (47%) of these legs, most commonly in the knee (74%) and less frequently in the ankle (26%). These results were dependent on the GMFCS level; children at GMFCS level II were more likely to develop an ankle contracture after the hip contracture, whereas those at GMFCS levels I or III–V were more likely to develop a knee contracture after the hip contracture.

A knee contracture was the primary contracture in 809 legs. A second contracture developed in 297 (37%) of these legs with an even distribution between hip (47%) and ankle (53%). A primary knee contracture was more likely to be followed by an ankle contracture for children at GMFCS level I-IV whereas a hip contracture was more likely to be the second contracture in children at GMFCS level V.

An ankle contracture was the first contracture to occur in 436 legs, and a second contracture developed in 151 (35%) of these legs, with more contractures affecting the knee joint (79%) and fewer affecting the hip joint (21%). Having an ankle contracture at baseline increased the risk of developing a knee contracture regardless of GMFCS level (Table 3).

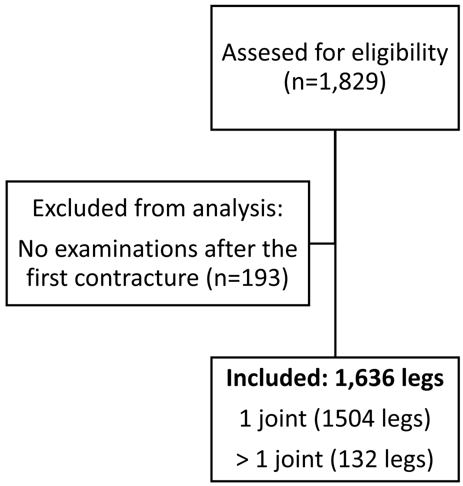

Fig. 1 Flowchart of the number of legs included in and excluded from the analyses

Discussion

We found that lower limb contractures appeared in specific patterns. A hip or ankle contracture primarily affected the knee joint, whereas a knee contracture affected both the hip and the ankle. Multiple contractures were most common at higher GMFCS levels. Of the children, 25% at GMFCS level I and 38% at level II developed more than one contracture during the follow-up compared with 50% of children at GMFCS IV and V. This corresponds with previous findings [2] that children at GMFCS IV and V are most likely to develop lower limb contractures.

The median age for developing a second contracture was 10 years. The second contracture was reported after a median of 5 years from the first contracture. The key problems in children with CP are muscle pathology,

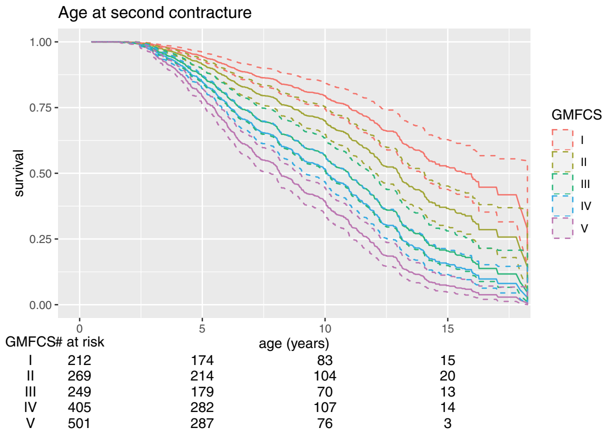

Fig. 2 Age at the development of a second contracture for children at GMFCS level I to V

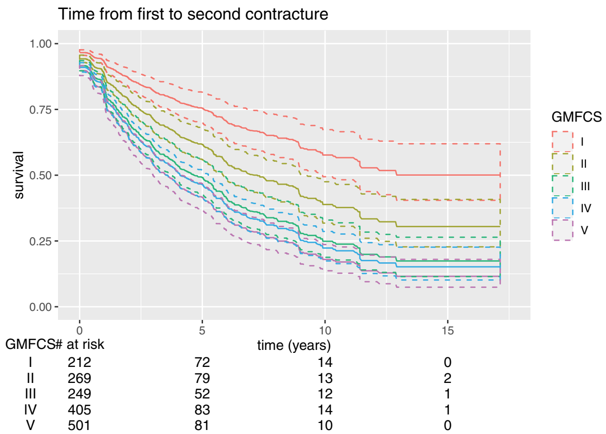

Fig. 3 Time in years from the first contracture to the development of a second contracture for children at GMFCS level I to V

loss of muscle control, abnormal muscle tone and muscle imbalance and these are associated with secondary problems, such as short muscles and contractures during childhood and growth. The secondary problems lead to tertiary abnormalities because the child must use compensatory strategies. It is important to understand the natural history and multifactorial pathology of contracture development [15, 16].

This study showed that the most common pattern in which the contractures appeared was a knee contracture at baseline followed by a hip or ankle contracture. The location of the second contracture varied with GMFCS level, and children at GMFCS level V were more likely to develop a hip contracture secondary to the knee contracture. Children with bilateral CP are, in general, more affected by deformities of the hip, whereas those with unilateral CP have more involvement of the ankle [17]. When Pettersson et al. [10] created a risk score for the development of scoliosis before 16 years of age in children with CP, limited knee extension was identified as an independent predictor. In supine lying, limited knee extension may force the hips into flexion and gravity is

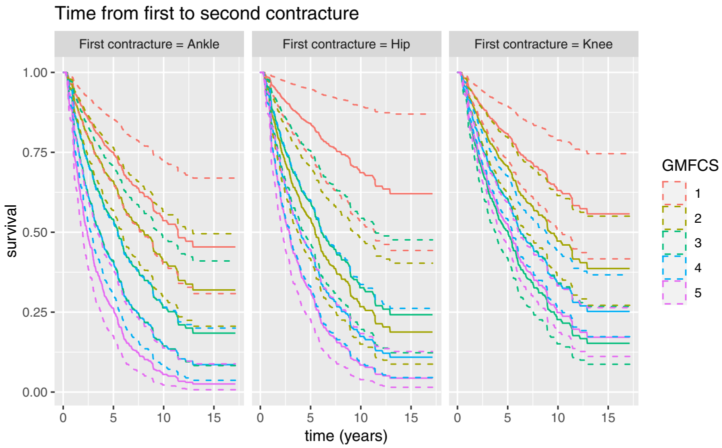

Fig. 4 Time to the development of a second contracture based on the location of the first contracture for children at GMFCS I to V

likely to tilt the legs to one side, causing the windswept position [18]. That may be one reason why a knee contracture increases the risk of a hip contracture. Children at GMFCS level IV or V are mostly nonambulatory and spend more time sitting with flexed knees and hips. This could also contribute to the sequence in which the contracture presents. Children at higher GMFCS levels have a high prevalence of postural asymmetries in sitting and lying, and they have difficulties to change position [19]. A knee contracture also increases the risk of an ankle contracture, possibly because of the gastrocnemius muscle, which is involved in both knee flexion and plantar flexion.

For ambulatory children with bilateral CP, a knee contracture may force them to walk in apparent equinus because of the lever arm dysfunction, which could lead to an ankle contracture. It is important to recognize apparent equinus when children walk on their toes because of the flexion contracture of the knee and not because of an ankle contracture [20]. For children with unilateral CP, a knee contracture can cause a limb-length discrepancy that makes them use toe walking as a compensatory strategy [21].

We found that children who developed multiple contractures and had an ankle contracture at baseline, were most likely to develop a knee contracture during the follow-up independent of their GMFCS level. In Sweden, children with limited dorsiflexion are usually treated with ankle–foot orthoses [22]. Most of these children use their orthoses during sleep. By preventing plantar flexion of the foot, a short gastrocnemius may force the knee joint into flexion and the child to sleep with flexed knees, which could cause a knee contracture over time [16]. This may lead to a vicious cycle with more time in sitting and less time in standing.

Limited dorsiflexion and equinus in children with unilateral CP may also compensate for the leg-length discrepancy by causing them to walk with a flexed knee on the longer leg [21]. In Sweden, botulinum toxin A injections are a common treatment for reduced ROM caused by increased muscle tone in the plantar flexors [23]. Together with isolated lengthening of the Achilles tendon, excessive injection of botulinum toxin A into the calf muscles can cause crouch gait, which is likely to persist and deteriorate over time with increased knee flexion [20].

There are several limitations to our study. Even though all subtypes were included, only spastic unilateral CP was identified to select the affected leg. The examinations started in 1994, prior to the introduction of the GMFCS. Therefore, we used the GMFCS level from the most recent report. However, the GMFCS levels show stability over time [24]. The contractures were recorded by goniometric measurement, which is a standard measurement in clinical settings but whose reliability varies according to the joint and position [25–27]. The results of our study were based on repeated measurements by many different examiners, and this may have introduced information bias or measurement errors that could influence the survival analysis. However, this bias has been shown to be small when differences between groups are small in terms of hazard ratios [14]. We validated the data for incongruent measurements to reduce the risk of extreme outliers caused by errors, e.g. clear reporting errors such as ROM of 360 degrees. The examiners involved in the CPUP are encouraged to practice the standardized measurements in the program, which has been shown to be important for reliability [28].

Another limitation was the cutoff value of a contracture because −10° in the hip, knee, and ankle joint can also affect the order in which a contracture appears. Full passive range of motion allows more extension of the hip and ankle joints compared with the knee joint. However, we could not find any literature supporting a cutoff exceeding 0° in any of the joints being considered a contracture. The statistical analyses were also run with a cutoff value of 0° and produced similar outcomes. There are few reference values for passive ROM in children with CP and no standard definition of what ROM is considered a contracture [29, 30]. Reference values for typically developing children are not representative when describing ROM in children with CP, who have reduced knee and ankle extension compared with hip extension [29]. We chose −10° as the cutoff for all joints since these values are often applied in clinical practice as severely affecting biomechanical alignment, gait, function and positioning. Hip extension less than 0° affects mobility and function in ambulatory children with CP, and a hip flexion contracture of -15° significantly decreases the physical functioning [31]. A knee flexion contracture of -10° increase the risk for crouch gait, and -10° of plantarflexion is the minimum required to significantly change kinematics and kinetics[30, 32, 33].

A further limitation of this study was the age of the children censored because of surgery. In the CPUP follow-up program, children at risk for hip displacement (migration percentage > 40) have their first soft tissue surgery of the hip early at a median age of 4 years. We wanted to follow the natural history of contracture development as much as possible, and it is likely that surgery of a joint affects the ROM both in the specific joint and the adjacent joints. Therefore, we censored legs at the date of surgery. This may have affected the results, especially for children at GMFCS level IV or V, who are more likely to have a surgery at a younger age. It is also possible that this decision led to underestimation of the influence of hip contracture on other joints. This may also partially explain why a hip flexion contracture was the least common contracture in our study. However, loss of hip extension seems less common than loss of knee extension and ankle dorsiflexion in children with CP at GMFCS I-II [29].

Children who received botulinum toxin treatment were not excluded from analyses and it could be argued that this interfered with our results since this treatment is more common in muscles affecting the knee and ankle than muscles affecting the hip. However, given the large number of children having this treatment[23] and the fact that the effect is not permanent, these children were included. Most children followed in CPUP receives ongoing interventions such as physiotherapy and treatment with orthoses. This should be taken under consideration when translating the results to other groups. This study covers a period of 24 years and interventions have changed during this time from more hands-on therapy to focus on participation and activities in daily life.

Contracture development starts early during the rapid growth in childhood and the risk of fixed contractures increases with age [2]. To prevent decompensation due to fixed deformities, clinicians should treat contractures as early as possible [34]. By monitoring children with CP, and treating contractures when they appear, with e.g. physiotherapy and orthoses, it may be possible to avoid the need for major surgery [7]. In the clinical setting, it is important to recognize how the sequence of contractures is likely to occur and to initiate prophylactic treatment of adjacent joints e.g. consider the knee joint when introducing Ankle–Foot Orthoses. For the orthopedic surgeon, knee contracture might indicate the need for closer monitoring of the hip to avoid dislocation [35].

Conclusions

Development of a second contracture is a common problem in children with CP and associated with higher GMFCS level. The time and sequence of secondary contracture development in the lower limbs are influenced by the location of the first contracture and GMFCS level. The second contracture primarily develops in the adjacent joint.

Lower limb contracture tends to appear in specific patterns, in which the location of the primary contracture affects the later development of secondary contracture in the adjacent joints. This should be considered when treatment strategies to prevent contractures are being formed.

Abbreviations

CP: Cerebral Palsy; CPUP: Cerebral Palsy Follow-up Program (Sweden); GMFCS: Gross Motor Function Classification System; ROM: Range of Motion.

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Authors' contributions

EC designed the study, collected and analyzed the data and drafted the manuscript. AL designed the study, analyzed the data and improved and revised the manuscript. HLP designed the study, improved and revised the manuscript. ERB designed the study, collected and analyzed the data, improved and revised the manuscript. All authors approved the final draft.

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Availability of data and materials

The dataset analyzed for this study are available in the CPUP registry. Permission to access data after ethical approval is granted by KVB Region Skåne. Requests to access the dataset should be directed to https://vardgivare.skane.se/kompetens-utveckling/forskning-inom-region-skane/utlamnande-av-patientdata-samradkvb/.

Declarations

Ethics approval and consent to participate

The study was approved by the Medical Research Ethics Committee in Lund, Sweden (383/2007, 443–99), and permission was obtained to extract data from the CPUP registry.

Consent for publication

Consent to use data for research was provided by all families participating in the registry.

Competing interests

The authors declare that they have no competing interests.

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Knee contracture in children with cerebral palsy

Children with cerebral palsy have an increased risk of developing knee contractures which affects their ability to stand, walk and participate in daily activities. The purposes of this thesis were to increase knowledge about the development of knee contractures; determine the prevalence, analyse associated factors, follow the time to and sequence of contracture development in the lower limb and evaluate the reliability of popliteal angle measurements in children with CP.

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