Pain and disabilities related to hip disorders in adults with severe cerebral palsy

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Adress of correspondence

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The work presented in this thesis was financially supported by the Johanna Kinderfonds, the Phelps Stichting voor spastici and the Anna Fonds.

The printing and post and packaging of this thesis were financially supported by Noppe Orthopedie and the Sophia Fonds.

Cover design	Eric Boldingh
Drawing cover	Anika de Ridder-van Leeuwen
Layout	Renate Siebes, Proefschrift.nu
Printed by	Ridderprint, Ridderkerk
ISBN	978-94-90791-13-1

VRIJE UNIVERSITEIT

Pain and disabilities related to hip disorders in adults with severe cerebral palsy

Klachten in relatie tot heupluxatie bij volwassenen met een ernstige cerebrale parese

ACADEMISCH PROEFSCHRIFT

ter verkrijging van de graad Doctor aan
de Vrije Universiteit Amsterdam,
op gezag van de rector magnificus
prof.dr. L.M. Bouter,
in het openbaar te verdedigen
ten overstaan van de promotiecommissie
van de Faculteit der Geneeskunde
op vrijdag 15 maart 2013 om 13.45 uur
in de aula van de universiteit,
De Boelelaan 1105

door

Eric Johan Karel Boldingh

geboren te Den Helder

promotor: prof.dr. G.J. Lankhorst

Kijk naar het licht en bewonder haar schoonheid.
Sluit de ogen en observeer.
Wat je eerst hebt gezien is er niet meer en wat je
erna zult zien is er nog niet.

Leonardo da Vinci

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Boldingh EJK[1], van der Heijden-Maessen HCM[1], Bos CFA[2], Bouwhuis CB[1]. Monitoring hip disorders in spastic cerebral palsy and its consequences for intervention; proposal to a surveillance and intervention algorithm. To be submitted.

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LIST OF ABBREVIATIONS

AI	acetabulum index
CCD angle	caput collum diaphyseal angle
CE angle	center edge angle
CMMS	columbia mental maturity scale
CO	Chiari osteotomy
CP	cerebral palsy
CPHCS	cerebral palsy hip classification system
CT	computed tomography
DVO	derotation varus osteotomy
FEHLA	femoral ephiphysis to Hilgenreiners line angle
GMFCS	gross motor function classification system
HO	heterotopic ossification
HSA	head shaft angle
FNA	femoral neck anteversion
FPS	faces pain scale
MP	migration percentage
NSA	neck shaft angle
PAICP	pain assessment instrument cerebral palsy
ROM	range of motion
VDRO	varus derotational osteotomy

INTRODUCTION

Chapter 1

Introduction and outline of this thesis

"Maybe it does bother them, but we don't know."

A clinical question turned into a scientific research project

In 1981, I was working as a rehabilitation resident at Leiden University Hospital. Sometimes we saw a young child with cerebral palsy (CP), and my teacher, Professor Hans Jongbloed, told me to assess the situation of the hips regularly by X-ray. "The hips show displacement and the patients, as adults, will be bothered by pain and other impairments. You should check the hips, so they can be operated upon when necessary."

Around 1987, I was working as a pediatric physiatrist at the rehabilitation center in Katwijk near Leiden. I told the parents of young children with CP that regular radiographs of the hips were necessary, and that if a disorder was found, surgery to correct it was needed.

Associated with the rehabilitation center was an institution for the residential care of multiply disabled adults, including many patients with CP, GMFCS levels IV and V. When on duty for the basic medical care, I met physiotherapist Peter Groot, who was caring for these people. I asked him whether these adult people had hip disorders, as I had learned in residency. "Sure", he sad, "we make X-rays once in a while". "And do they suffer from the disorder"? was my next question. "Maybe it does bother them, but we don't know" he answered.

Our conversation raised the basic question for this thesis.

Do people with severe cerebral palsy suffer from hip disorders, and if so, how can we help them ?

A quick literature search did not yield a consistent answer.

When we decided to start scientific research into this matter, starting in 1991, the question arose how to design the research project. Should we perform a prospective study with an intervention group and a control group? The problem in this field is the huge heterogeneity of symptoms in patients with CP, plus the diversity of surgical procedures that are applied in many different age groups.

After consulting Professor Lex Bouter, epidemiologist at VU University Amsterdam, we decided to perform a prevalence study, with independent X-ray variables like

migration percentage and deformity of the femoral head on the one hand, and dependent variables like pain, decubitus ulcers, fractures, sitting problems and nursing difficulties on the other. Statistical analysis could then identify correlations between the variables (Figure 1.1).

The next question was how to measure pain in this patient group, in which 85% of the patients have speech impairment and more than 60% have an intellectual disability.[1]

At an epidemiology course organized by Professor Bouter, I met Suzanne Arts. She had recently returned from Australia, where she had developed and tested a scale to measure pain in children, the faces pain scale (FPS), with the Australian group led by Bieri.[2] The scale had been validated for children aged four years or older.

The advantage of this scale is that it can be used by nonverbal people, who can for instance indicate the rank number of a drawing with their finger, or even by sticking out their tongue.

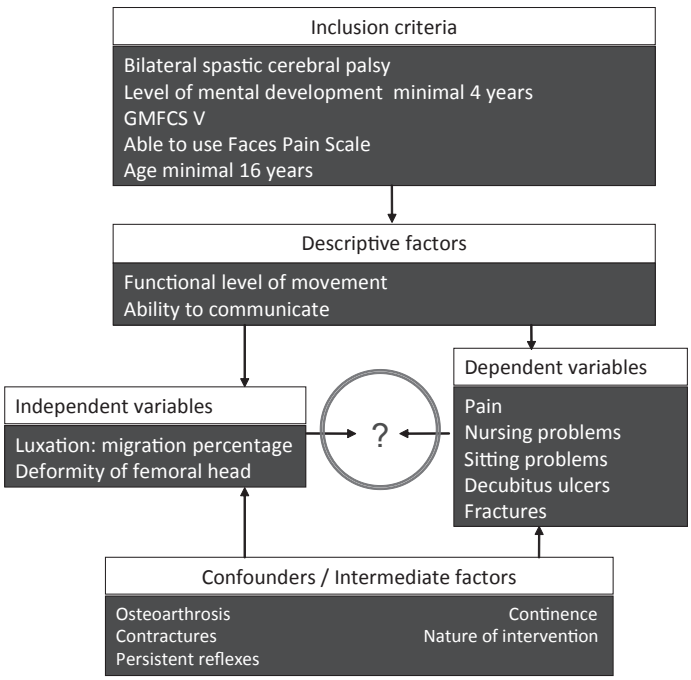

Figure 1.1 Research design.

The next problem was to assess a patient's developmental level, to ensure that they had a mental age of at least four years. In this preparatory study, we tried the Columbia Mental Maturity Scale[3] (CMMS). This scale, developed in the USA in the 1950s, offers drawings of both realistic and abstract design. Patients have to indicate which drawing is different from the other two or three. The task becomes increasingly difficult, and 25 correct answers indicate a developmental age of four years or more.

We decided to perform a validation study, in which the CMMS acted as a filter to assess whether a patient would be able to use the FPS as an instrument to indicate pain in the hip region.

We developed drawings of everyday situations, including events involving strain on the hips, such as being lifted from the bed (figure on front page) or putting on trousers. For the sake of comparison, drawings presented situations that are normally not painful, like listening to the radio, or situations that are very painful for most people, like being stung by a wasp or burning one's hand.

The next step was to compose a group of adult patients with CP, all non-walkers, as a research sample. We managed to find 160 patients, most of them living in a residential setting, others living independently or with their parents. Our research physiotherapist, Monique Jacobs-van der Bruggen, traveled throughout the country to find candidates and perform the measurements.

We asked the patients' own physiotherapist and their regular caregiver to predict whether the patient would be able to use the FPS and to predict the pain score on the items used. The combination of scales proved to be useful for our purpose. The results are presented in **chapter 2**.

The next question to answer was whether a hip disorder is related to pain. In addition to the CMMS and the FPS, we asked the patients for permission to make an X-ray of their pelvis, if their most recent X-ray was older than five years, so as to assess the present situation of the hip.

This enabled us to relate the FPS scores to the X-ray results at group level. A formula, proposed by Mr. Joop Kuik MSc, statistician at VU Medical Center Amsterdam, was used to establish the threshold for pain being significant for the patients, by comparing the scores in pain-positive, pain-negative and hip-straining situations. The results of this study are presented in **chapter 3**.

We also assessed the relationship between hip disorders and other important problems, viz. fractures, sitting problems, nursing difficulties and decubitus ulcers. Nursing difficulties were measured using part of the WEEFIM instrument.[4] The results of this part of the study are presented in **chapter 4**.

After this study, we concluded that a hip disorder in adult patients with severe CP (GMFCS V) results in a risk of approximately 30% of pain due to the disorder. Hence, we wondered what the best way would be to monitor an imminent or established hip disorder and how to treat it, once it had been identified. We performed a systematic review to assess the value of preventive and corrective interventions for hip disorders in CP. A lot of work for this review was done by Hélène van der Heijden-Maessen en Carola Bouwhuis, who read most of the articles. The result of this work is discussed in **chapter 5**.

Unfortunately, preventive or corrective interventions are not always successful; patients sometimes end up with incurable pain due to hip disorder. We therefore performed a systematic review about the value of palliative surgery in CP, the results of which are presented in **chapter 6**.

The only reliable way to identify a hip disorder in CP is to make an X-ray of the pelvis.[5] There has been a long debate on the question how often these radiographs must be made, and at what age the surveillance should start. Surveillance guidelines were published from the late 1970s onward.[6] A major breakthrough was achieved with the introduction of the GMFCS classification by Palisano et al. in 1997.[7]

The reliability of surveillance guidelines was dramatically improved by the publications by Soo (2006)[8] and Hägglund (2007).[9] They matched the incidence of hip disorders in CP to the GMFCS level, proving that the higher the GMFCS level, the greater the risk of hip displacement and the greater the need for careful surveillance. However, none of the guidelines published since 1979 contains recommendations for intervention.

From an early stage of my research, I made proposals to match the surveillance of hip disorders and the results of this surveillance with guidelines for intervention. As far as I am aware, this was – and remains – the first time that this combination was incorporated in a decision tree. My argumentation was that it is useless to offer recommendations for surveillance if you do not know what to do with the result of the monitoring, other than referring the patient to an orthopedic surgeon.

The first version of my decision tree was produced in 2003, after which it was frequently discussed at meetings of the Dutch Society of Pediatric Physiatrists, a division of the Netherlands Society of Physical and Rehabilitation Medicine, VRA.

The completion of the systematic reviews reported on in chapters 5 and 6 lent greater significance to the decision tree, as the review results confirmed the intervention recommendations in the tree. A review of the monitoring instruments, surveillance methods and intervention techniques on which the decision tree is based is presented in **chapter 7**.

Chapter 8 discusses the value of the results of our research and offers recommendations for further research and clinical practice.

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DEVELOPMENT AND VALIDATION OF A PAIN ASSESSMENT INSTRUMENT

Chapter 2

Assessing pain in patients with severe cerebral palsy: development, reliability, and validity of a pain assessment instrument for cerebral palsy

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Arch Phys Med Rehabil 2004;85(5):758-66.

ABSTRACT

Objective: To develop a Pain Assessment Instrument for CP patients (PAICP) and to study its test-retest reproducibility and construct validity.

Design: Cross sectional validation study.

Setting: Homes for severely handicapped.

Participants: 164 adult severe CP patients, caregivers and physiotherapists. Nine healthy children.

Interventions: The PAICP contains drawings of daily situations. Patients score experienced pain on a Faces Pain Scale. Reproducibility and construct validity was assessed in pilot study with CP patients and healthy children. Construct validity and agreement between the pain scores of the patients and proxies was assessed in 160 patients with severe CP.

Main outcome measures: Pain score on PAICP.

Results: Good construct validity and adequate test-retest reproducibility. A significant difference between the mean scores for "painful" and "not painful" situations. We found moderate agreement between the scores of the patients and proxies for daily activities, and only for those activities in which the proxies are personally involved.

Conclusion: The PAICP has adequate test-retest reproducibility and construct validity. It provides an indication of the pain experienced by patients in situations in which proxies are not personally involved and maybe more valid then proxy measures for other situations.

INTRODUCTION

It is very difficult to assess the severity of pain experienced by people with cerebral palsy (CP). The fact that many patients cannot express their pain verbally,[1] due to motor impairment, makes the use of nonverbal scales necessary. Some patients with CP also have visual impairments.[2,3] Another complicating factor in the assessment of pain is mental retardation, which frequently occurs in people with severe CP; approximately 40% of the CP population has an intelligence quotient below 70.[4] Although measuring pain in cognitively impaired patients is difficult,[5] recent studies have demonstrated that it is possible.[6,7] However, the common practice in the case of severely disabled persons is still to interview the caregivers to assess the amount of pain suffered by the patient.[8-11] Unfortunately, the pain scored by the patient often differs from the pain scored by the caregivers,[12-16] although one study[17] has reported a strong correlation.

To measure the relation between hip disorders and pain in patients with severe CP, an assessment instrument was developed and the present study was undertaken to investigate its test-retest reproducibility and construct validity.

METHODS

The assessment instrument

A literature search (PubMed and MEDLINE using key words cerebral palsy and pain from 1993 to 2002) was performed to identify scales for the self-rating of pain by people with severe CP. No available scales have been specifically developed for this group of patients. The scale that is needed should be appropriate for use with nonverbal patients with a low mental age, some of whom can only communicate by scanning symbols. Because mental retardation is not associated with a disturbance of face recognition[18] and CP is not associated with disturbances in body image,[19] researchers[20] suggest that a pain scale expressed in facial expressions can be used for people with low levels of mental development.

Recently, a high correlation between a visual analog scale and a Faces Pain Scale (FPS) was demonstrated.[21] It is known that children from the age of four can recognize and interpret symbols such as a drawn face.[22] Such a scale should range from a neutral to a painful face, instead of starting from a happy face, which suggests a different mood.[23]

The FPS developed by Bieri et al.[24] (Figure 2.1) might be suitable for this purpose. The scale, which consists of seven faces with expressions ranging from neutral to very painful, was designed and validated for use with children from the age of four years.[24] The intervals between the faces are almost equal.[25] The use of this scale with cognitively impaired people has recently been assessed, and its reliability and construct validity was acceptable in people with moderate cognitive impairment.[6] The original scale has recently been revised.[26]

Basing our design on the FPS, we developed the Pain Assessment Instrument for Cerebral Palsy (PAICP) to assess pain in patients with hip problems. The instrument consists of six drawings of daily situations that are usually not painful and six that usually are painful.

To be able to relate pain in the hip region to other types of pain, five drawings of situations that may be painful for those with hip problems were added (Figure A2.1, Appendix 2.2). There are four preliminary drawings with obvious answers introducing the method. The drawings are shown in random order (Table 2.1). The patient scores the amount of pain experienced in these situations according to the FPS.

Participants

We studied the instrument in two groups of subjects. First, we conducted a reliability study of the PAICP in a group of nine healthy children in their own homes and four

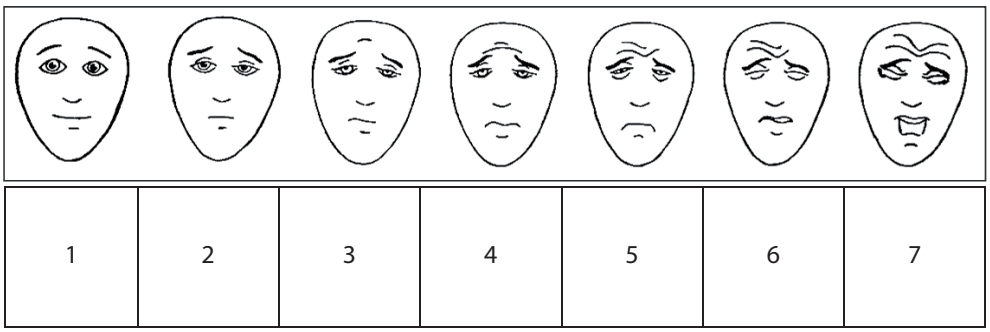

Figure 2.1 The score options assigned to items of the FPS. Numbers were not shown to patients. From Hicks CL, von Baeyer CL, Spafford P, van Korlaar I, and Goodenough B (2001). Adapted from Bieri D, Reeve RA, Champion GD, Addicoat L, and Ziegler JB. The Faces Pain Scale for self-assessment of the severity of pain experienced by children: development, initial validation, preliminary investigation for ratio scale properties. Pain 1990;41:139-50. Reprinted with permission.[24,26]

Table 2.1 The items of the Pain Assessment Instrument for CP (PAICP)

Item		Painful	Not painful	Possibly painful
Preliminary questions	Squeezing a hand in the door	X		
	Dirt in an eye	X		
	Removing plaster from the skin	X		
	Injection by a dentist	X		
Putting on trousers				X
Drinking hot tea		X		
Cleaning teeth			X	
Eating bread			X	
Burning hand		X		
Doctor using a stethoscope			X	
Combing hair			X	
Biting own tongue		X		
Being lifted from bed				X
Putting on sweater			X	
Listening to music			X	
Lying in bed				X
Physiotherapy for legs				X
Stubbing a toe		X		
Having a blood sample taken		X		
Sitting in a wheelchair				X
A wasp sting		X		

Painful: situations usually painful in daily life; Not painful: situations usually not painful; Possibly painful: hip straining situations

adults with severe CP in a home for severely handicapped persons. The adults were included if they had CP, were unable to walk independently, had a mental age of four or above, and were able to use an FPS. The ability to use an FPS (vision, ability to recognize and select) and the mental level was assessed beforehand using the Columbia Mental Maturity Scale[27] (CMMS), a nonverbal mental development test that has been validated for adults and young children with CP.[28,29] To use the FPS, a minimum score of 25 points on the CMMS is needed, which indicates a minimal mental age of four years.

The healthy children were three to seven years of age, and the adults with CP were 24 to 31 years of age.

Second, we conducted a construct validity study in 160 patients with severe CP. These subjects met the same criteria as those in the reliability study, and they all resided in The Netherlands.

The patients were recruited in nursing homes for severely handicapped persons and through rehabilitation centers in The Netherlands. Physicians and physiotherapists were asked to select patients according to the inclusion criteria and to predict whether they would meet the CMMS criteria for selection. Of the 160 patients, 87 (54%) were men and 73 (46%) were women. Their ages ranged between 16 and 84 years (mean age, 36y). Nineteen patients (12%) could not speak, and 11 used scanning symbols for communication.

Reliability study

The children and patients were asked to indicate the level of pain they usually had in situations shown in the drawings, and they rated the amount of pain they experienced in these situations using the FPS. They indicated the location of the pain experienced on a female or male drawing of the human body according to their gender (Figure 2.2). A standard list of instructions and questions was applied (Appendix 2.1). The researcher

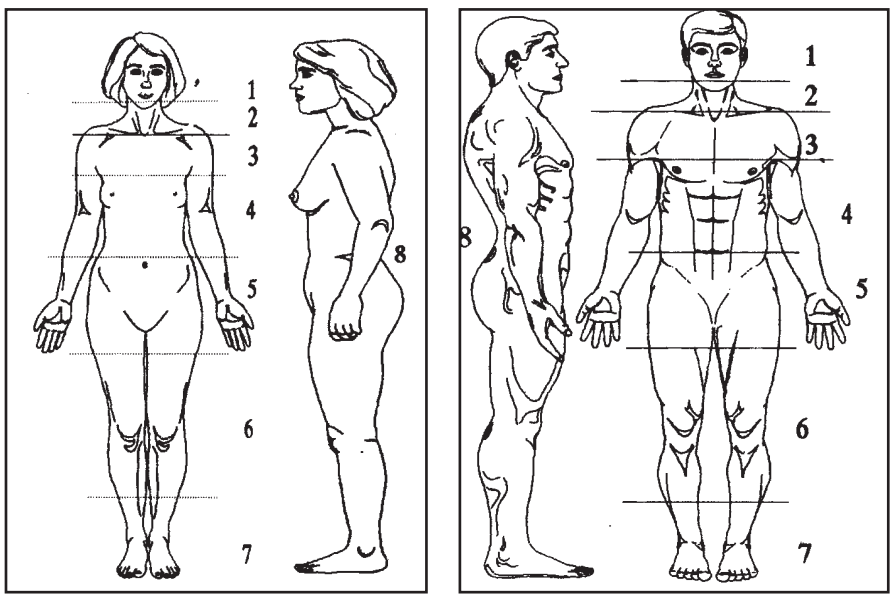

Figure 2.2 Drawings of human body.

recorded the number of the face, which ranged from one to seven. The test-retest reproducibility of the PAICP was assessed by applying the instrument twice with an interval of two weeks, and the test-retest analysis was performed with a modified κ.[30] The percentage of observed agreement between scores (OBS) was defined as the percentage of patients with the same score, ±1, on test and retest. Expected agreement (EXP) was defined as the expected percentage of patients with the same score, ±1, on test and retest just by chance, which is three out of seven. Modified κ was calculated as (OBS-EXP)/(1-EXP). The results were classified according to the Altman method.[31] Response to a question was considered reproducible if a modified κ of 0.4 or higher was reached.

Construct validity study

A total of 160 patients who met the inclusion criteria were asked to score the pain they experienced in the 21 situations. Subsequently, one main caregiver and one physiotherapist associated with the patients were asked to predict the scores of the patients in all situations on the PAICP, without being aware of the patients' scores. The construct validity was considered adequate if the drawings of situations that were usually painful produced a mean score of three or higher, and the situations that were usually nonpainful produced a mean score below three on the seven-point scale. Further, the mean score of all painful situations should exceed the mean score of all situations that were nonpainful. Nonparametric correlations between the PAICP scores of patients and caregivers were calculated using the Spearman ρ, because the answers were not normally distributed. Internal consistency of the PAICP was analyzed by calculating the Cronbach α.

RESULTS

The assessment instrument

The characteristics of the PAICP are shown in Figure 2.1 and Table 2.1.

Reliability study

Adequate test-retest reproducibility was found for all items, for both the healthy children and the patients. The modified κ was .48 or greater, with the exception of

Table 2.2 Test-retest reproducibility of the assessment instrument based on the Faces Pain Scale. Modified Kappa; reliability study; n=13

Category	Paired samples	OBS	Kappa*	Classification
Situations usually painful	Hot tea	70	0.48	Moderate
	Burn hand	75	0.56	Moderate
	Bite tongue	82	0.69	Good
	Injection dentist	100	1.00	Very good
	Blood sample	75	0.56	Moderate
	Wasp sting	91	0.84	Very good
Situations usually not painful	Clean teeth	100	1.0	Very good
	Eat bread	92	0.86	Very good
	Stethoscope	100	1.0	Very good
	Combing hair	100	1.0	Very good
	Putting on sweater	100	1.0	Very good
	Listening to music	92	0.86	Very good
Situations possibly painful	Putting on trousers	100	1.0	Very good
	Being lifted from bed	100	1.0	Very good
	Lying in bed	100	1.0	Very good
	Physiotherapy leg	92	0.86	Very good
	Sitting	100	1.0	Very good

OBS = observed percentage of patients with same score (plus or minus 1) in test and retest. EXP = expected percentage of patients with the same score (plus or minus 1) in test and retest just by chance, which is 3 out of 7 x 100% = 43%. *Modified Kappa= (OBS-EXP)/(1-EXP).

Table 2.3 Mean score items painful – not painful; main group; n=160

Category	Item	Mean score	Standard deviation
Situations usually painful	Hot tea	4.3	1.9
	Burn hand	4.7	1.9
	Bite tongue	4.0	1.9
	Injection dentist	4.1	2.0
	Blood sample	3.1	2.0
	Wasp sting	4.8	2.0
Situations usually not painful	Clean teeth	1.4	1.0
	Eat bread	1.1	0.5
	Stethoscope	1.2	0.6
	Combing hair	1.3	0.8
	Putting on sweater	1.4	1.1
	Listening to music	1.0	0.3
Situations possibly painful	Putting on trousers	1.6	1.4
	Being lifted from bed	1.8	1.6
	Lying in bed	1.6	1.4
	Physiotherapy leg	2.7	2.0
	Sitting	1.5	1.3

the question about "stubbing a toe" (κ=.37). In subsequent analyses, this question was replaced by the question "injection by a dentist" (Table 2.2).

Construct validity study

The mean scores met the predetermined criteria, with all mean scores of painful drawings being 3.1 or higher, and all mean scores of nonpainful drawings being 1.4 or lower (Table 2.3). The difference between the mean scores for painful and nonpainful drawings was statistically significant. A significant difference also existed between the drawings for situations that were possibly painful and situations that were usually not painful (Table 2.4). The Cronbach α analysis revealed good internal consistency (Table 2.5).

Table 2.4 Difference in mean scores between situations *usually* painful and not painful, on the Pain Assessment Instrument for CP (PAICP); main group, n=160

Group	Mean score (SD)		Difference of the mean	95% confidence interval*	
	Situations usually painful	Situations usually not painful		Lower	Upper
CP patients	4.11 (1.46)	1.25 (0.45)	2.86	2.63	3.08

Difference in mean scores between situations *possibly* painful and not painful, on the Pain Assessment Instrument for CP (PAICP); main group, n=160

Group	Mean score (SD)		Difference of the mean	95% confidence interval	
	Situations possibly painful	Situations usually not painful		Lower	Upper
CP patients	2.21 (1.29)	1.21 (0.45)	0.96	0.78	1.14

* Analysis paired samples t-test.

Table 2.5 Internal consistency of questions posed

Category	Cronbach's alpha	95% confidence interval
Situations usually painful	0.83	0.77–0.87
Situations usually not painful	0.65	0.55–0.73
Situations possibly painful	0.81	0.75–0.86

Caregivers were able to predict that the patients would meet the CMMS criteria for selection: 179 of the 219 caregivers (82%) made a correct prediction.

There was a statistically significant, but only modest, agreement between the patients' pain scores and the pain scores given by the caregivers and physiotherapists for the situations that could be painful for patients with hip problems. Two other situations showed significant correlations as well, but they were limited to situations in which caregivers applied direct care to the patients (i.e., cleaning teeth and combing hair; Table 2.6).

DISCUSSION

Measuring the pain experienced by people with severe CP has always been difficult. The PAICP showed adequate test-retest reproducibility and construct validity and

Table 2.6 Construct validity: correlations between scores of patients, physiotherapists and caregivers on the Pain Assessment Instrument for CP (PAICP) Scale range 1–7

Category	Question	Physiotherapist vs. patient	Care-giver vs. patient
		Correlation*, Significance**	Correlation*, Significance**
Situations usually painful	Hot tea	0.13	0.13
	Burn hand	0.02	0.14
	Bite tongue	-0.03	0.06
	Injection dentist	0.15	0.14
	Blood sample	-0.03	0.20
	Wasp sting	0.11	0.08
Situations usually not painful	Clean teeth	-0.03	0.35**
	Eat bread	0.05	-0.08
	Stethoscope	0.01	-0.10
	Combing hair	0.03	0.28**
	Putting on sweater	0.20	0.11
	Listening to music	-0.01	-0.01
Situations possibly painful	Putting on trousers	0.32**	0.26**
	Being lifted from bed	0.52**	0.23**
	Lying in bed	0.29**	0.37**
	Physiotherapy leg	0.34**	0.48**
	Sitting	0.36**	0.28**

* Spearman's ρ, ** significance at the 0.1 level (2-tailed).

appears to be suitable for use with patients with severe CP, several of whom were unable to speak and/or had the mental age of a toddler.

The usefulness of the FPS has recently been demonstrated for elderly people as well.[25] Chibnall and Tait[6] found the FPS is valid for assessing the most severe pain in the past and retrospective levels of pain, which was also the focus of the present study. Their study population consisted of people with moderate mental retardation; the participants in the present study had severe mental impairments. Preassessment with the CMMS worked well: every patient (n=160) who scored 25 or higher on the CMMS used the FPS easily.

The PAICP is an important improvement in scientific research among this group of patients. In the present study, we added drawings of situations affecting the hip, but the instrument can easily be amended to suit other specific situations that are the focus of research by changing the drawings accordingly.

One limitation of the instrument is that it can only be used in persons with a mental age of at least 4 years, which excludes patients with very severe CP. Another prerequisite is sufficient visual ability to see the drawings. It may therefore be necessary to increase the size of the pictures in some cases. People must also be able to indicate their choice. In the present study, some patients indicated their choice by just looking at a drawing or by sticking out their tongue a number of times corresponding to the number of the drawing.

Preassessment with the CMMS worked well for assessing patients' ability, not only with regard to their level of mental development, but also with regard to their ability to see the drawings and to indicate their choice.

Caregivers in the study were able to estimate well the level of intelligence of the patients they cared for and consequently their ability to use an FPS (82%), which makes the use of a pretest such as the CMMS unnecessary when caregivers are closely involved with their patients.

The instrument was developed in The Netherlands, based on a scale of drawings created in Australia. Almost all the drawings illustrate situations that have been experienced by most people with CP. Thus, the scale may have international applicability, apart from translation of the verbal instructions to the native language of the patient.

However, two of the drawings may have to be altered or replaced in some cases: the "burning hand" and, perhaps, the "wasp sting." The former may not have been

experienced by respondents who do not have radiators in their houses and the latter by respondents who have never been stung by a bee or a wasp. Future versions may need to use a hand in a fire or on a stove to illustrate burning a hand and perhaps to omit the wasp sting (or use it only for respondents who have experienced it).

The findings also indicate that caregivers and physiotherapists do not necessarily provide accurate estimates of the amount of pain experienced by patients with CP. The associations between self-reports and proxy reports were only modest at best in the current study. The estimate of proxies is better, but not optimal, in situations in which they perform an act for the patient that directly causes discomfort or pain, such as cleaning teeth or lifting the patient out of bed.

CONCLUSIONS

The amount of pain and discomfort rated by the patient using this instrument was quite different from the estimates made by the proxy. This finding emphasizes the usefulness of the instrument.

The PAICP enables the patient to rate the amount of pain he/she experiences in certain common situations that others would find clearly painful or clearly not painful. The instrument gives the caregiver a way to assess the pains impact and to select the treatment needed because of the pain. Without an instrument such as the PAICP pain experienced by a person with CP may otherwise not be signalled or treated.

Caregivers should not rely on their own estimate of the pain experienced by their patient but should make use of an instrument like the PAICP more often. The measure also gives people who are unable to explain their problems verbally or patients with a mental disability a new chance to express their needs.

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APPENDIX 2.1

Instructions and questions concerning the PAICP

I would like to know whether you experience pain from time to time.

I will show you a couple of drawings.

On the drawings you can see various situations; some of them are painful and some are not.

Look, this is an example (show drawing of squeezing hand).

Here you see seven faces; they are all different.

This one has a lot of pain (indicate face no. 7).

This one has no pain at all (indicate face no. 1).

These faces show that the pain increases in severity (indicate faces no. 2–6).

Now, which face do you think will match the situation in the drawing?

Here is another example (show drawing no. 2: dirt in eye).

Did you ever experience that?

Did that hurt?

Can you indicate how much it hurt, which face is the best match?

Can you indicate on this drawing where you felt the pain?

OK: next . . . et cetera.

APPENDIX 2.2

Figure A2.1 Drawings of daily situations

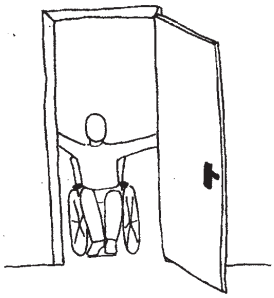

Figure A2.1.1
Squeezing hand in the door

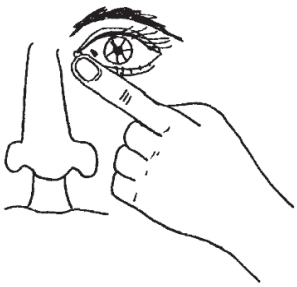

Figure A2.1.2
Dirt in an eye

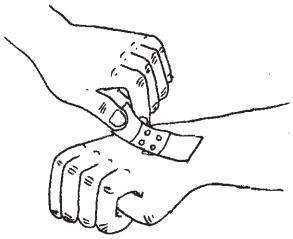

Figure A2.1.3
Removing plaster from skin

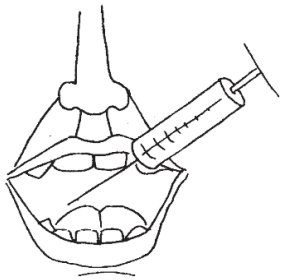

Figure A2.1.4
Injection by a dentist

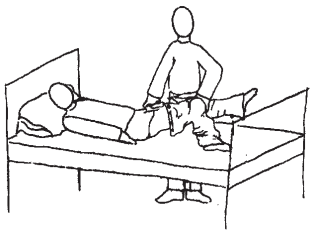

Figure A2.1.5
Putting on trousers

Figure A2.1.6
Drinking hot tea

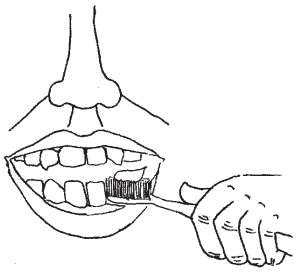

Figure A2.1.7
Cleaning teeth

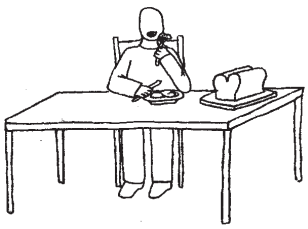

Figure A2.1.8
Eating bread

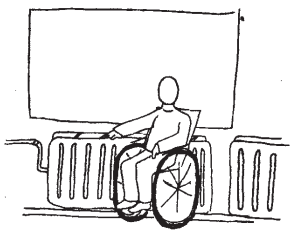

Figure A2.1.9
Burning hand

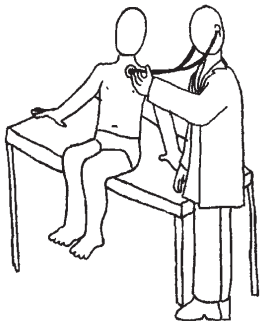

Figure A2.1.10
Doctor using stethoscope

Figure A2.1.11
Combing hair

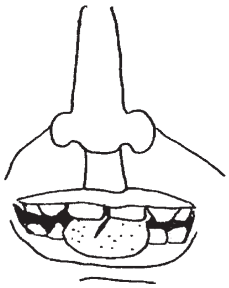

Figure A2.1.12
Biting own tongue

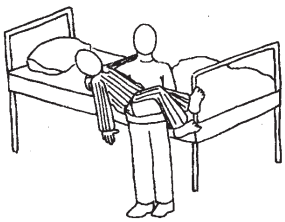

Figure A2.1.13a
Being lifted from bed 1

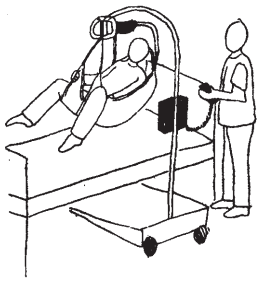

Figure A2.1.13b
Being lifted from bed 2

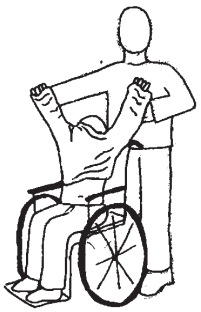

Figure A2.1.14
Putting on sweater

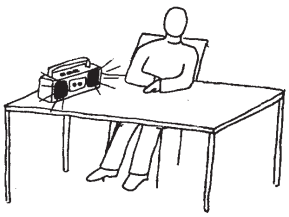

Figure A2.1.15
Listening to music

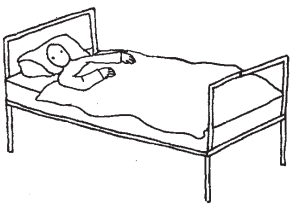

Figure A2.1.16
Lying in bed

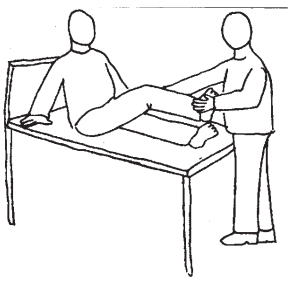

Figure A2.1.17
Physiotherapy for legs

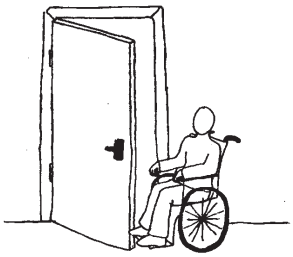

Figure A2.1.1 18
Stubbing a toe

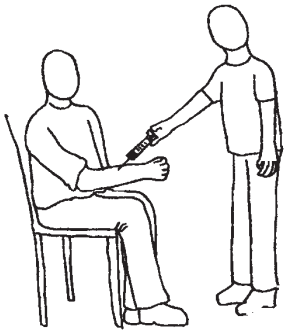

Figure A2.1.19
Having blood sample taken

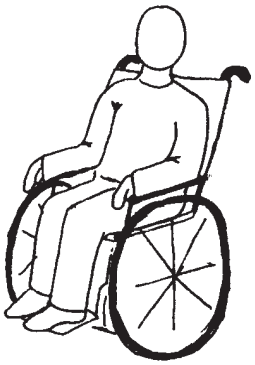

Figure A2.1.20
Sitting in a wheelchair

Figure A2.1.21
Wasp sting

RELATIONSHIP BETWEEN HIP DISORDERS, PAIN AND OTHER IMPAIRMENTS

Chapter 3

Determinants of hip pain in adult patients with severe cerebral palsy

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J Pediatr Orthop B 2005;14(2):120-5.

ABSTRACT

The purpose of our investigation was to study the relationship between radiographic results of the femoral head and pain in people with severe cerebral palsy. We conducted a cross-sectional study on hip radiographic results and pain in 160 patients with severe cerebral palsy.

Eighteen percent of our patients had hip pain in hip-loading situations. Migration and deformity were closely related.

There was a significant association with hip pain (odds ratio, 2.79; 95% confidence interval 1.01–7.70). There is a high prevalence of hip pain after unsuccessful femoral bone surgery. Migration and deformity of the femoral head are strongly interrelated, and are associated with pain.

INTRODUCTION

In severe cerebral palsy (CP), the femoral head tends to migrate from the acetabulum, and approximately 60% of the hips of non-ambulant CP patients will subluxate.[1] According to the method described by Reimers,[2] subluxation is defined as a migration of 30% or more.[3] One in six of these hips will progress to dislocation (migration of 100%).[3] Above a migration of 60%, dislocation will follow, unless prevented by surgery.[1,4,5] Finally, prevalence of dislocation in non ambulant patients is about 50–60%.[6,7] Migration can be visualized on a radiograph of the pelvis from the age of 18 months or earlier;[8] dislocation occurs at the age of 5–7 years.[9-12]

Subluxation and dislocation of the hip can be treated by conservative and operative procedures with the purpose of preventing severe problems in adulthood, such as pain, difficulties with care, seating problems, decubitus ulcers and fractures.[13] Pain in patients with CP is a frequent phenomenon, also in the hip region: in a sample of CP patients, 39% reported chronic pain in the hip region with a mean duration of 20 years.[14] However, there is still debate in the literature as to whether hip disorders are the cause of the pain.

This leads to differences in opinion concerning the necessity of (preventive) surgery.[15-17] Some authors state that pain is related to migration or dislocation in itself.[18] Others consider osteoarthritis to be an additional factor,[3] while pain is also explained by deformity of the femoral head.[19,20] Furthermore, although deformity of the femoral head is related to migration, it also occurs in non-migrated hips.[21]

Unfortunately, the methods used to assess pain in relation to migration are sometimes not clearly or accurately defined in the literature. Moreover, no author has yet used a method which enables CP patients to assess their own pain. The pain experienced by these patients, who are often mentally handicapped and are frequently verbally impaired, is difficult to assess.

In the literature, pain is often assessed by proxies, which makes reports of the relationship between migration and pain difficult to interpret.[1,10, 22,23]

The aim of this study was to investigate the relationship between radiographic results of the femoral head (migration, deformity, osteoarthritis), on one hand, and pain on the other hand, in a group of patients with severe CP. Use was made of an instrument that was specifically designed for the self-assessment of pain in this group of patients;

a CP Pain Assessment Instrument (PAICP), based on the Faces Pain Scale (FPS) developed by Bieri et al.[24] This instrument had been validated in an earlier study in healthy children and adults with CP.[25]

MATERIALS AND METHODS

Patients

A cross-sectional study was conducted to investigate the relationship between radiographic results (migration, deformity and osteoarthritis) of the femoral head and pain in a group of 160 patients with severe tetraspastic CP. The study was approved by the Medical Ethics Committee of the VU University Medical Center. The study population consisted of patients with CP, who were not able to walk independently with or without walking aids, with a minimum mental age of four years, and able to use an FPS.

The ability to use the PAICP (vision, ability to recognize and select) and the mental level of the patients was assessed in advance with the Columbia Mental Maturity Scale (CMMS),[26] a non-verbal mental development test which has been validated for use with adults and young children with CP.[27,28] To demonstrate ability to use the PAICP, a minimum score of 25 points is needed, which indicates a minimum mental age of 4 years.

Patients were recruited in homes for the severely handicapped and in rehabilitation centers in The Netherlands. Informed consent was obtained from the patients themselves or from their representatives.

Methods

The PAICP contains 21 drawings of daily occurring situations. It includes 6 situations that are usually not painful (P-). They result in a mean score of ≤1.4. Six drawings represent situations that are usually painful (P+), resulting in a mean score of ≥4.0. There are 4 drawings introducing the method. It also contains 5 hip loading situations (H) possibly painful for those with hip problems. The latter category consists of: putting on trousers, being lifted from bed, lying in bed, physiotherapy of the legs and sitting in a wheelchair.

The drawings were shown in random order (available on request from the first author) and the patient assessed the amount of pain experienced in the situations represented by the drawings with a score ranging from 1–7 (1 = no pain, to 7 = severe pain) and located the pain on a picture of the human body (male/female, according to the gender of the patient).

Because the perception of pain is a subjective matter, a method was sought with which to relate the potentially hip-related pain to situations that definitely cause pain and those that do not. Therefore, the scores for the drawings representing hip-loading situations were related to the scores for the other groups of questions, usually non-painful and painful.

Hip pain was mathematically related to the scores for the drawings of usually "painful" situations (P+) and the drawings of usually "non-painful" situations (P-). Patients were considered to suffer hip pain if the result of the formula $\{H-(P-)\}/\{(P+)-(P-)\} \geq 0.33$, and H>P-. H = median of answers concerning situations possibly painful for people with hip pain; P+ = median of usually painful situations; P- = median of usually not painful situations. P- has is minimal value of 1; P+ a maximum of 7. Therefore, P+ - P- has a maximal value of 6. The difference between H and P- must be at least 2 in order to consider the amount of pain as positive.

Scores of 0.33 and above as outcome of the formula represent patients that experience pain in daily returning situations as putting on trousers, being lifted from bed, lying in bed, receiving physiotherapy for legs and sitting. In this way, the patients were divided into two groups: those with hip pain and those with no hip pain. In the subsequent analyses, pain was considered as a dichotomous variable. Patients were interviewed and files were checked about surgery of the hip in the past and its indication. An anterior-posterior radiograph of the pelvis was then made, unless there was already a radiograph available that was not made more then 5 years before.

The migration percentage (MP)[2] was measured on an anterior-posterior photograph. This widely used method is known to be reliable in assessing displacement of the femoral head from the acetabulum.[8,29] The asymmetry of migration was assessed by calculating the difference between the migration percentages on each side. It was divided into four groups (Table 3.3). Femoral head deformity was scored on a 3-point scale, with the categories absent, moderate and severe.

Osteoarthritis was graded according to the Tönnis classification.[30] Grade 1 osteoarthritis is increased sclerosis of femoral head and acetabulum, a slight decrease in the height of the cartilage and slight osteophytes. Grade 2 refers to small cysts in the femoral head or acetabulum, a marked decrease in the height of cartilage and slight deviation from the round form of the femoral head. Grade 3 represents large cysts in femoral head and acetabulum, a severe decrease in cartilage up to complete absence of the joint cleft, severe deviation of the round form of the femoral head and avascular necrosis.

One author (MJvdB) conducted all the interviews, and another author (EB) scored all the radiographs, without any knowledge of patient characteristics. Logistic regression analysis using SPSS® version 10.0 (SPSS, Chicago, Illinois, USA) was performed with pain (yes / no) as dependent variable and migration and its difference, deformity and osteoarthritis of the femoral head as independent variables. Surgery was considered to be successful when the result was in concordance with the primary indication and no adverse side-effects such as pain or (sub) luxation occurred.

RESULTS

Patients

The age of the patients varied between 16 and 84 years, with a mean of 36 years. Of the 160 patients, 87 (54%) were male and 73 (46%) were female; 19 (12%) were not able to speak, and 11 (7%) of them used a scanning device for communication.

Prevalence of pain

Thirty patients (19%) had hip pain in hip-loading situations according to the study criteria. Pain occurred more frequently in women (12/63) than in men (6/77), but there was no differentiation in age.

Radiography results

The radiographs were subdivided, on the basis of the maximal migration percentage, into four groups, according to the standard method.[7] In 20 cases, the radiograph showed an

abnormal configuration of femur and pelvis, in which the migration percentage could not be calculated. On those radiographs, the femoral head was either absent, obviously totally disintegrated, or impossible to identify. Direct aberrant contact between (parts of) the femur and the pelvis was also classified as an abnormal configuration. It was decided to consider these patients as a separate fifth group (group V, Table 3.1).

The remaining 140 patients (groups I–IV) had a migration of less than 100% or a dislocation with a femoral head that was clearly recognizable (Group I: MP <30%; II: MP 30–59%; III: MP 60–89%; IV: MP >90%).

Relationship between radiographic results and pain

In cross-tab analysis of the five sub-groups, the relationship between pain and the situation of the femoral head was significant (Spearman's rho 0.367, p=0.000, Table 3.1). No relation was found between pain and gender.

Groups I–IV (migration less than 100% or dislocation)

In the 140 patients in groups I–IV, deformity proved to be a concomitant factor for pain (Table 3.2). Migration and deformity of the femoral head were significantly inter-related and combined for univariate logistic regression analysis (rs 0.46; p<0.01). Also, pain increased with the quantity of asymmetry and osteoarthritis, respectively (Table 3.3). Migration percentage and difference in migration put together were also closely related (rs 0.64; p<0.01). When arranged in one table, pain showed to be a factor dependent on multiple factors (Table 3.4).

Table 3.1 Determinants of pain: relationship between situation of the femoral head and pain

Migration of femoral head		N	With pain
Group			
I	<30%	89	8 (9%)
II	30–59%	35	5 (14%)
III	60–89%	9	2 (22%)
IV	>90%	7	3 (42%)
V	Abnormal configuration	20	12(60%)
Total		160	30(19%)

Spearman's Rho=0.37; p=0.000.

In univariate logistic regression analysis of groups I–IV, the migration and deformity factors were combined into two groups, because of their close inter-relationship: group one with migration less than 33% and no or moderate deformity, the second with migration >33% and/or severe deformity.

Table 3.2 Determinants of pain: relationship between deformity, migration and pain; group I–IV, n=140

Deformity	Migration	Pain		Total
		No	Yes	
None	<30%	29	3	32
	30–59%	7	1	8
	90–100%	1	0	1
	Total	37	4 (9.8%)	41
Moderate	<30%	43	3	46
	30–59%	8	1	9
	60–89%	3	0	3
	Total	54	4 (6.9%)	58
Severe	<30%	9	2	11
	30–59%	15	3	18
	60–89%	4	2	6
	90–100%	3	3	6
	Total	31	10 (24.4%)	41

Table 3.3 Determinants of pain: relationship between asymmetry, osteoarthritis and pain

Asymmetry of femoral head, groups I–IV	N	With pain
None (difference 0–10)	79	7 (9%)
Mild (difference 11–30)	35	4 (11%)
Moderate (difference 31–60)	17	3 (18%)
Severe (difference >60)	9	4 (44%)
Total	140	18
Osteoarthritis of femoral head, groups I–IV	**N**	**With pain**
None	57	6 (10%)
Mild	60	8 (13%)
Moderate	11	1 (9%)
Severe	12	3 (25%)
Total	140	18

The analysis showed a significant relationship ($p<0.05$) between the combined factor migration >33% or severe deformity and pain, with an Odd's ratio of 2.79. Also asymmetry of migration showed a significant relationship with pain (OR 8.23), indicating an increased relative risk.

Table 3.4 Relationship between osteoarthritis, migration, deformity and pain; group I–IV

Osteoarthritis	Migration	Deformity	Pain	
			No	Yes
None	<30%	None	19	1
		Moderate	17	1
		Severe	2	1
	30–59%	None	4	1
		Moderate	1	1
		Severe	1	1
	60–89%	Moderate	2	
		Severe	3	
	90–100%	None	1	
		Severe	1	
Mild	<30%	None	10	2
		Moderate	23	2
		Severe	4	1
	30–59%	None	2	
		Moderate	7	
		Severe	6	
	60–89%	Severe		1
	90–100%	Severe		2
Moderate	<30%	Moderate	2	
		Severe	2	
	30–59%	None	1	0
		Severe	3	1
	90–100%	Severe	2	
Severe	<30%	Moderate	1	
		Severe	1	
	30–59%	Severe	5	1
	60–89%	Moderate	1	0
		Severe	1	1
	90–100%	Severe		1
Total			122	18

Osteoarthritis was a factor that was not significantly related to pain (Table 3.5). If adjusted for the other factors, the odds ratios remained positive but not statistically significant, which illustrates their close inter-dependence.

Group V (abnormal configuration of the femoral head and pelvis)

This abnormal configuration was strongly associated with pain: 12 of the 20 patients (still) suffered from hip pain. In this sub-group only patients with ankylosis of femur and pelvis and a previous resection of the femoral head at the level of the minor trochanter currently had less than 100% pain (Table 3.6).

Results of surgery

Of the 160 patients, 132 had surgery for three major indications: pain, reduction of the femoral head and others/unknown. The results of this surgery are shown in Figure 3.1. Soft tissue surgery was most successful with reduction as indication (80%); result for indication pain was poor (54%). Pain was best cured by DVO (71%, Figure 3.1).

Table 3.5 Crude and adjusted[a] odds ratios (OR) and 95% confidence intervals (CI) for determinants of hip pain, groups I–IV, n=140

n=140	crude OR	adjusted OR[a]
Migration/deformity		
migration <33% and no/moderate deformity	1.00 (ref)	1.00 (ref)
migration ≥33% and/or severe deformity	2.79 [1.01–7.70]*	1.70 [0.39–7.34]
Osteoarthritis		
None	1.00 (ref)[b]	1.00 (ref)[b]
Mild	1.31 [0.42–4.04]	1.29 [0.40–4.15]
Moderate	0.85 [0.09–7.85]	0.54 [0.05–5.62]
Severe	2.83 [0.60–13.44]	1.60 [0.28–9.12]
Asymmetry of migration		
None	1.00 (ref)[c]	1.00 (ref)[b]
Mild	1.33 [0.36–4.86]	1.19 [0.30–4.81]
Moderate	2.20 [0.51–9.57]	1.45 [0.23–9.03]
Severe	8.23 [1.79–37.87]*	5.43 [0.82–35.8]

* $p<0.05$.
[a] adjusted for the other variables in the table.
[b] p for trend not significant.
[c] p for trend 0.011.

Table 3.6 Patients with abnormal configuration of femoral head and pelvis (group V; n=20)

Situation	Number	History of bone surgery	Pain as indication bone surgery	(Residual) hip pain
After proximal resection of the femoral head (subcapital)	5	5	4	4
After distal resection of the femoral head (level minor trochanter)	3	3	2	1
Ankylosis of femur and pelvis	4	4	2	2
Aberrant contact between (rest of) femur and pelvis	8	5	5	5
Total	20	17	13	12

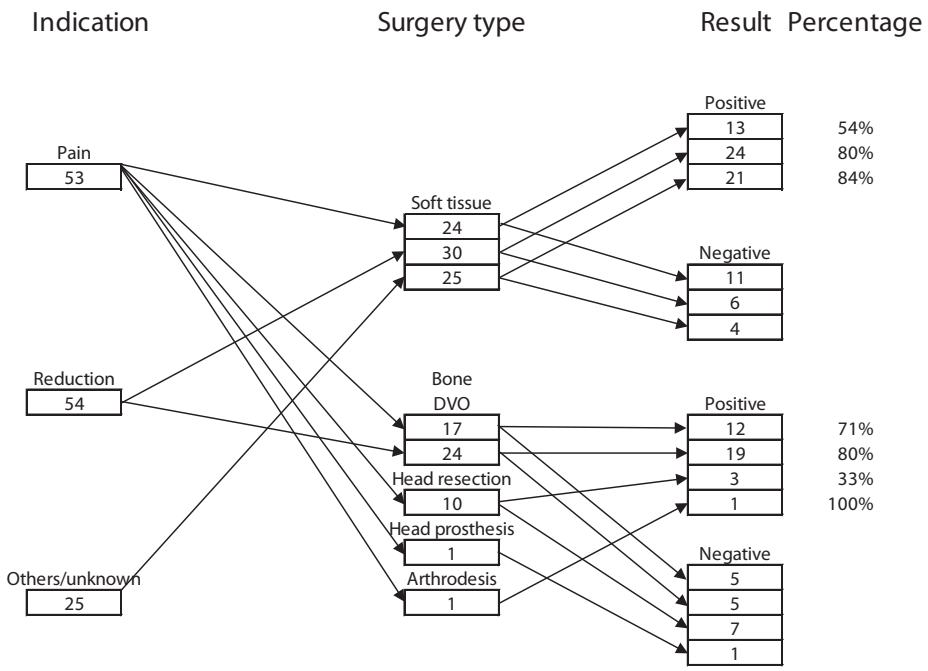

Figure 3.1 Indication, type of surgery and result. DVO = derotation varus osteotomy.

DISCUSSION

The overall prevalence of hip pain in patients with severe CP found in the present study (19%) is in agreement with the prevalence of pain in patients with CP found in other studies,[31,32] but less than reported by Hodgkinson et al., who found a percentage of 47%.[22]

However, their interviews were partly held with care-givers and with the patients themselves, as took place in the present study. Furthermore, they used a pain scale that had not been validated for this group of patients. The prevalence of osteoarthritis found in the present study was also in concordance with findings reported in the literature.[33]

A relationship between deformity and pain had already been found.[19] The results of the present study show that there is an association between migration of the femoral head, its deformity and pain. However, the design of the study did not make it possible to assess the temporal relationship between these factors and pain.

Migration and deformity are closely related, as was found in the analysis, and also in other studies,[21,23,33] but they do not seem to be completely inter-dependent. Although deformity generally increases with migration, it is not the result of migration alone. Eleven out of 89 patients with a migration percentage of less than 30% already had severe deformity of the femoral head (Table 3.2). At least one of the patients without osteoarthritis, migration and deformity suffered from pain; on the other hand, there was a patient with severe osteoarthritis and deformity and subluxation without pain (Table 3.4).

Also the relationship we found between asymmetry of the migration percentage and pain confirms that in people with CP the pain apparently is a multifactorial phenomenon that cannot be explained by migration, deformity or asymmetry alone.

Several patients in this study had undergone surgery to reduce the femoral head or to reduce pain. A number of these patients were in groups I–IV and their surgery had been successful (Figure 3.1). However, a number of operations had obviously not been successful: 20 of 160 patients had an abnormal configuration of femur and pelvis, mostly after bone surgery, which was associated with (residual) pain.

Only distal resection of the femoral head at the level of the minor trochanter was found to have no association with pain (Table 3.6). This in concordance with the findings of Cooperman et al.[19]

The results of the present study suggest that deformity, asymmetry and migration of the femoral head should ideally be prevented or treated, but the question that rises is: how and at what price?

Soft tissue or bone surgery to prevent or correct migration can be quite successful,[3,10] but surgery has a great impact on these already very disabled patients, and if soft tissue

surgery fails the decision to perform bone surgery should be even more carefully considered. Surgery in itself does not cure asymmetry,[34] and may even aggravate it when performed unilaterally.[35]

Furthermore, unsuccessful bone surgery creates a high risk for residual, severe pain. In the literature, the distal resection of the femoral head at the level of the minor trochanter is reported to be a successful salvage treatment in curing pain when other measures fail.[19,32] Our findings support that view.

Acknowledgements

The Johanna Children's Fund and the Dr. W.M. Phelps Foundation in The Netherlands funded this research. The data were analyzed with help of Mr. J. Kuik from the Department of Clinical Epidemiology and Biostatistics at the VU University Medical Center.

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Chapter 4

Radiographic hip disorders and associated complications in severe cerebral palsy

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J Pediatr Orthop B 2007;16(1):31-4.

ABSTRACT

We performed a cross-sectional study of 160 adult patients with severe cerebral palsy (CP) to study the relationship between radiographic hip disorders (migration and deformity of the femoral head), on the one hand, and complications such as handling problems, seating problems, decubitus ulcers, fractures and contractures, on the other hand.

Both migration and deformity were positively related to the need for a special seat in the wheelchair and adduction contractures of the hip.

We conclude that migration and deformity of the femoral head, if possible, should be prevented in patients with severe cerebral palsy.

INTRODUCTION

In approximately 60% of patients with severe cerebral palsy (CP) the femoral head subluxates from the acetabulum.[1] Subluxation is defined as at least 30% of migration, as described by Bagg[2] and Reimers,[3] dislocation as migration >100%. No clear answer can be found in the literature as to whether or not there is an association between radiologically assessed subluxation or dislocation of the hip and complications. Well-known problems experienced by patients with severe CP are pain, difficulties in care-giving, seating problems, decubitus ulcers, fractures and contractures.[4-6] The determinants of hip pain in patients with severe CP have been described, indicating that migration and deformity of the hip should be prevented to avoid pain.[7]

Problems with perineal hygiene are mentioned most frequently in the literature[8] and it is known that seating problems often occur in patients with severe CP.[9] Decubitus ulcers are reported in patients who are totally bed-bound.[8] Patients with severe CP are also prone to fractures.[10] Contractures of the lower extremities have been found in 64% of non-walkers.[5]

The few publications that report on the relationship between radiographic findings and contractures demonstrate that there is no association.[3,11]

The objective of our research was to study the relationship between subluxation and deformity of the hip, on the one hand, and handling problems, seating problems, decubitus ulcers, fractures and contractures on the other hand, in patients with severe CP.

METHODS

Patients

We conducted a cross-sectional study in 160 adult patients (age ≥16 years) with severe tetraspastic CP. They were all on level five on the GMFCS (gross motor function classification system) scale.[12] Fifty-eight percent of the patients lived in a nursing home. Patients with CP were included if they had a minimum mental age of four years according to the Columbia Mental Maturity Scale.[13] These patients were also recruited

for a study on the determinants of hip pain.[7] The Medical Ethics Committee of the VU University Medical Center approved the study protocol.

METHODS

Both the usual care-giver and the physiotherapist of each patient were asked to rate the difficulties in handling of the patient on a scale from 1 to 5 (Table 4.1). This produced a score ranging from 4 to 20. Handling problems were defined as a score of >12 out of 20 (at least four items rated as moderate). Patients without a regular caregiver or physiotherapist are indicated as 'missing'. Both these scales were developed for this study.

The chair was classified on a six-point scale indicating progressive adaptation (1 – standard, no addition; 2 – standard with additions; 3 – special molded seat back, regular seat; 4 – special molded seat, regular seat back; 5 – special molded seat and seat back; 6 – special molded seat and seat back, integrated). In the analysis, two categories were distinguished: no special seat (scores 1–3) and special molded seat (scores 4–6).

Subsequently, the patients were examined, for decubitus ulcers and contractures in the hip region and lower extremities, by our research physiotherapist (MJvdB). For fractures, the patients' files were examined and the patients' doctor was interviewed. The standard classification was used for decubitus ulcers,[14] and the sagittal, frontal, transverse, rotation system for the recording of angles[15] and a standard goniometer[16] to measure contractures. Patients were assessed while lying flat on a bed. Abduction of less than 30° of at least one of the hips was considered as hip adduction contracture. A hip and knee flexion contracture was defined as a limitation of 30° or more of extension of one of the hips or knees, respectively. Fractures were considered to be present if detectable on a radiograph of the lower extremity that was made within the previous 3 months.

An anterior–posterior radiograph of the pelvis was taken, unless there was one available that was made within the previous 5 years. We measured the migration percentage (MP^3) on the anterior–posterior radiograph. Femoral head deformity was scored on a three-point scale: categories absent (normal shape of the femoral head), moderate (femoral head not round but flattened in some parts) and severe (femoral head flattened, or with sharp edges as in 'Napoleons' cap' or otherwise deformed).

Table 4.1 Questions on handling problems

Question: Do you experience difficulties in handling due to limited motion or stiffness in:	
(Care-givers)	(Physiotherapists)
Washing perineum	Lifting from bed to couch
Drying after washing	Lying on the couch
Putting on trousers	Moving legs in hip joint
Lifting from bed to wheelchair	Transfer from prone to sitting

Score: 1 = None, 2 = Minimal, 3 = Moderate, 4 = Many, 5 = Serious, 9 = Missing data.

The radiographs of 20 patients showed an abnormal configuration of the femur and pelvis, in which the MP could not be calculated. The femoral head was either absent, obviously totally disintegrated or impossible to identify. Direct aberrant contact between (parts of) the femur and the pelvis was also classified as an abnormal configuration. Details on this group have been published separately.[7] We excluded these patients in the analysis regarding MP.

The remaining 140 patients had a femoral head that was clearly recognizable. The radiographs were subdivided, on the basis of the maximal MP, into four groups, according to the standard method: 0–29%, 30– 59%, 60–89%, 90–100%.[17]

One author (EB) scored all the radiographs, without any knowledge of patient characteristics. Differences in the mean MP were analyzed with t-tests for independent samples. Associations between hip deformity and complications were analyzed with χ^2 tests for trend. In addition, the associations between the outcome variables (complications) and radiographic findings (MP and deformity) were modeled with linear regression (for handling problems) or logistic regression (seating problems and contractures) adjusted for age and sex (SPSS® version 10.0; SPSS, Chicago, Illinois, USA).

RESULTS

Patient characteristics, prevalence of complications and results of radiography

A brief description of the patient characteristics and the prevalence of complications is shown in Table 4.2.

Table 4.2 Patient characteristics

Mean age (SD) (n=160)	36 (15.7)
% Male	54
Migration Percentage (MP) (n=160)	
Mean MP (SD)	44 (42.9)
% MP<30	55.6
% MP 30–59	21.9
% MP 60–89	5.6
% MP 90–100	4.4
% Abnormal hip	12.5
% Total	100
Hip deformity (n=140)	
% No	29.3
% Moderate	41.4
% Severe	29.3
% Total	100
Handling problems physiotherapist (n=125)	
Mean score (SD)	9.0 (3.6)
% with score >12	23
Handling problems care-giver (n=146)	
Mean score (SD)	10.0 (4.0)
% with score >12	14
Seating problems (n=160)	
% special moulded seat in chair	61
% Decubitus ulcers (n=160)	3
% Fractures (n=160)	0.6
Contractures (n=160)	
% Hip limited abduction	58
% Hip flexion contractures	14
% Knee contractures	32

SD = standard deviation; n = number of patients.

Correlation between radiograph results and complications

Results of radiography and handling problems

No significant difference was found in mean MP between patients with or without handling problems (Table 4.3). Handling problems were also not significantly associated with the level of hip deformity (Table 4.4). Regression analysis, with adjustment for age and sex, also showed no significant associations.

Table 4.3 Differences in mean migration percentage between patients with and without problems (n=140)

Item	Mean MP (SD) in patients without problems	Mean MP (SD) In patients with problems	Mean difference (95% CI)
Handling problems physiotherapist (score >12)	30.3 (20.6)	27.9 (20.9)	-2.4 (-11.6–6.8)
Handling problems care-giver (score >12)	32.3 (25.0)	28.9 (22.3)	-3.5 (-15.3–8.4)
Seating problems (yes / no)	25.9 (15.5)	35.2 (27.3)	9.3 (2.0–16.6)**
Hip limited abduction*	23.6 (17.2)	37.3 (26.3)	13,7 (6.4 –21.0)**
Hip flexion contractures*	31.6 (23.9)	28.1 (22.1)	-3.5 (-15.6–8.6)
Knee contractures*	29.9 (22.1)	34.4 (26.8)	4.5 (-4.1–13.1)

* left or right >30°; ** significant at p<0.05 level.

Table 4.4 Frequency of problems (%) in patients with different levels of hip deformity (n=140)

Item	No hip deformity	Moderate hip deformity	Severe hip deformity	Chi-squared trend
Handling problems physiotherapist	16.1%	21.3%	33.3%	p=0.10
Handling problems care-giver	20.0%	12.0%	16.7%	p=0.67
Seating problems (yes / no)	51.3%	51.8%	87.2%	p=0.001
Hip limited abduction*	39.0%	53.4%	75.6%	p=0.001
Hip flexion contractures*	9.8%	10.3%	17.1%	p=0.312
Knee contractures*	22.0%	31.1%	36.6%	p=0.150

* left or right >30°.

Results of radiography and seating

A clear relationship was found between migration and deformity of the femoral head, on the one hand, and the use of a special seat in the wheelchair, on the other hand (Tables 4.3 and 4.4). The relationship between deformity of the femoral head and the need for a special molded seat remained significant in logistic regression analysis, after correction for age and sex (p=0.01). The association between MP and the use of a special seat was borderline significant after adjustment for age and sex (p=0.08).

Results of radiography and decubitus ulcers and fractures

Owing to the low prevalence of these problems (Table 4.2), no associations could be investigated.

Results of radiography and contractures

The mean MP was significantly higher in patients with hip adduction contractures than in patients without contractures (Table 4.3). The prevalence of hip adduction contractures increased as the level of hip deformity increased (Table 4.4). In logistic regression analysis, after correction for age and gender, the associations remained significant.

DISCUSSION

Patients with severe cerebral palsy suffer from a wide range of complications, which is confirmed by the results of our study. We found some aspects of care to be in accordance with reports in the literature, such as the need for a special seat and the prevalence of contractures, but decubitus ulcers and fractures were rare in our population. The high level of care that is provided in The Netherlands for these patients could explain these findings.

Deformity of the hip was strongly associated with the need for a special molded seat and limited abduction of the hip. Especially, the relationship between femoral head deformity and the need for a special seat is remarkable: only 51% of the patients with no deformity of the femoral head had a special molded seat whereas 87% of the patients with a serious deformity needed one. The need of adapting a special seat means a considerable amount of time and money and decreases the possibility of interchanging chairs.

The relationship between migration and deformity of the femoral head and limited range of hip abduction is also prominent. Hip flexion contractures are not significantly related to hip deformity, but in patients with an abnormal configuration of the hip the prevalence of hip flexion contractures increases considerably. This illustrates the impact of hip abnormality on handling possibilities of these patients.

In conclusion, handling problems, seating problems and contractures are common in adults with severe CP. The need of providing a special molded chair and limited

abduction of the hip are strongly related to deformity of the femoral head. Abnormal configuration of the femoral head and pelvis decreases the possibility to extend the hips. It is therefore recommended that these orthopedic abnormalities are prevented or treated as they develop.

Acknowledgement

Data were analyzed with the help of Mr J. Kuik from the Department of Clinical Epidemiology and Biostatistics, VU University Medical Center, Amsterdam, The Netherlands.

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SYSTEMATIC REVIEW ON INTERVENTION METHODS

Chapter 5

Effectiveness of preventive and corrective surgical intervention on hip disorders in severe cerebral palsy: a systematic review

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Submitted for publication.

ABSTRACT

Purpose: This review presents an overview of studies on the effectiveness of preventive and corrective surgical interventions to treat hip luxation in patients with severe cerebral palsy.

Method: A systematic literature search was used to identify studies concerning surgical procedures for hip luxations and subluxations in severe Cerebral Palsy (GMFCS 4 and 5). A qualitative analysis and a best evidence synthesis were performed for soft tissue surgery and osteotomies.

Results: All 15 included studies were observational. Only one study involving soft tissue surgery was of sufficient quality. Nine of 10 studies involving osteotomies were of sufficient quality, including a total of 189 patients. The mean migration percentage (MP) at follow-up ranged from 6 to 29%. No relationship could be established between the effect of the surgical procedure and the patients' age or the duration of follow-up. The percentage of patients reporting pain decreased from 81% preoperatively to 5% at follow-up. Twenty-five percent had complications like osteoarthritis, ulcers or fractures.

Conclusions: There is insufficient evidence for the effectiveness of soft tissue surgery to stabilize the hip, due to insufficient quality of the retrospective observational studies. This review revealed indicative findings for an effect of bony surgery in stabilizing the hip. Timing of the procedure remains an issue. Multicenter trials could shed further light on this complicated subject.

INTRODUCTION

Hip problems involving displacement of the femoral head are common in cerebral palsy (CP). Their incidence is correlated with the severity of CP as measured by GMFCS: rates of almost 90% have been reported for patients with GMFCS IV and V.[1-3] Displacement frequently leads to complaints of pain, caregiving problems, seating problems, pressure sores, fractures and contractures.[4] As we showed in previous studies, severe and long-lasting subluxation or dislocation of the hip increases the risk of hip pain in adulthood from 10 to about 30%.[5] Pain is correlated with the migration percentage and deformity of the hip.[5] The above problems make it important to prevent or treat displacement of the hip in patients with severe CP.

Current treatments include soft tissue surgery to prevent migration and several types of bony surgery to correct the displacement. The results and effects of all these surgical procedures have been frequently described. Their outcomes are usually reported to be good, although unfavorable side effects and failures have been reported.[6]

Several years after soft tissue surgery, patients often require bony surgery such as femoral varus-derotational osteotomy (VDRO) with or without pelvic osteotomy, or even more palliative surgery. Unfortunately, when bony surgery fails, the prevalence of persistent hip pain is high.[5]

The indication for bony surgery is even more critical for non-walkers (GMFCS IV and V). The impact of bony surgery on these severely handicapped patients is high, and the complication rate is substantial. The question is therefore whether we should perform bony surgery in this group of patients, and whether there is sufficient evidence for the effectiveness of these procedures.

Until now, no systematic review on this topic has been published, except for the study by Stott and Piedrahata on the effect of soft tissue surgery.[7] This review concerns preventive soft tissue surgery, but does not differentiate between GMFCS categories. The authors concluded that radiographic hip subluxation improves in 33% of patients after surgery, but that the evidence is insufficient, due to the low quality of the 27 included studies. This is probably because the studies were retrospective and uncontrolled, and used different outcome measures.

We believe that constructing a flowchart for the purpose of monitoring and treating hip problems in severe CP requires more information about the effectiveness of

treatment. The best evidence synthesis enabled us to review the literature in a correct and comprehensive way.

The aim of the present systematic review is to determine the effectiveness (in terms of pain relief and decreased MP) of preventive (soft tissue) and corrective (bony) surgical interventions in hip subluxation in patients with severe cerebral palsy (GMFCS IV and V).

MATERIALS AND METHODS

Literature search

An extensive literature search was conducted by two independent researchers (CB and HvdH) using the following resources: MEDLINE (1966 until February 2009), CINAHL (1982 until February 2009), EMBASE (1982 until February 2009), Cochrane controlled trial register and PEDRO. This was supplemented by hand searching the reference lists of papers found in the above sources and publications by known researchers in this field.

The keywords used for the computerized search strategy in MEDLINE were 'cerebral palsy' [MeSH] or free text word, 'hip' [MeSH] or free text word, OR 'hip subluxation' [Mesh] or free text word OR 'hip dislocation' (free text word) AND 'surgical procedure, operative' [MeSH] or 'surgery' [Mesh] or free text word). The search was limited to the English, Dutch and German languages and full text articles.

The inclusion of articles, based on title and abstract, was decided by the same two independent reviewers (CB and HvdH). In case of uncertainty, they read the whole text, and any disagreements were resolved by discussion. If no consensus was reached, a third reviewer (EB) was consulted. Studies that met the following criteria were included:

- Diagnosis of severe cerebral palsy, i.e. GMFCS IV or V, spastic quadriplegia, total body involvement, independent sitters/dependent sitters, non-ambulatory, dependent non-walker, severe neurological involvement.
- Evidence of a hip subluxation or dislocation, objectively verified by X-ray (MP).

- Surgical intervention having been performed (soft tissue or bony surgery)
- Reason for the intervention described in terms of pain or subluxation/ dislocation by MP.
- Examination of the effect of surgical intervention in terms of the outcome measures 'diminished pain' or 'decreased MP', and a follow-up of at least 1 year.

Palliative interventions, like femoral head resection, were excluded. These interventions are performed when other options have failed, and as they require other inclusion criteria, the palliative methods have been described in a separate review article.[8]

Since we did not find any randomized controlled trials, we included all types of studies, except letters or case-reports.

Data analysis

The methodological quality of the selected studies was assessed by the same independent reviewers (CB and HvdH). Any disagreements were resolved by discussion, but if no consensus was not reached, a third reviewer (EB) made the final decision. The methodological quality was rated using the criteria formulated by Steultjens et al.[9] (Table 5.1). This is an adapted form of the list of criteria by Van Tulder et al.,[10] specifically designed for observational studies. The amended list includes seven criteria for internal validity, four descriptive criteria and two statistical criteria. Studies were considered to be of 'sufficient quality' if at least four criteria for internal validity, two descriptive criteria and one statistical criterion were met, as proposed by Steultjens et al.[9]

In view of the diversity among the studies in terms of outcome measures, interventions used and study designs (no RCTs), meta-analysis was not considered appropriate. Instead, we used a best-evidence synthesis according to Steultjens et al.[9] and Van Tulder et al.[10] (Appendix 5.1). Separate best-evidence syntheses were performed for soft tissue surgery and osteotomies. A descriptive summary of the results of the included studies is presented.

Since different outcome measures and definitions had been used in the studies, we recalculated the following outcome measurements using the data from the tables presented in the articles: mean pre- and post-operative percentage of subluxation/

Table 5.1 Criteria for methodological quality (other than controlled designs) (Steultjens et al.[9] and Van Tulder et al.[10])

Patient selection: a. eligibility criteria specified?
Interventions: b. intervention explicitly described? c. co-interventions described? d. compliance acceptable?
Outcome measurement: e. outcome assessor not involved in the treatment? f. outcome measures relevant? g. adverse effects described? h. withdrawal/dropout rate reported and acceptable? i. timing of follow-up measurements: short-term follow-up measurement performed? long-term follow-up measurement performed? j. timing of the outcome assessment comparable for all patients?
Statistics: k. sample size of the patient group described? l. did analysis include an intention-to-treat analysis? m. point estimates and measures of variability presented for the primary outcome measures?
Internal validity criteria: c, d, e, f, h, j, l. Descriptive criteria: a, b, g, i. Statistical criteria: k and m.
Studies were considered to be of 'sufficient quality' if at least 4 criteria for internal validity, 2 descriptive criteria and 1 statistical criterion were met.

dislocation and number of hips with a post-operative MP below 33%. This allowed us to make a better comparison between the outcomes of the different studies.

RESULTS

The literature search resulted in 224 abstracts (Figure 5.1). The first selection, based on title and abstract, yielded 44 articles. All of these studies were observational studies. Eleven studies were excluded because they incorporated all CP severity classes and it was not possible to separate them. Ten studies were excluded because they had used a

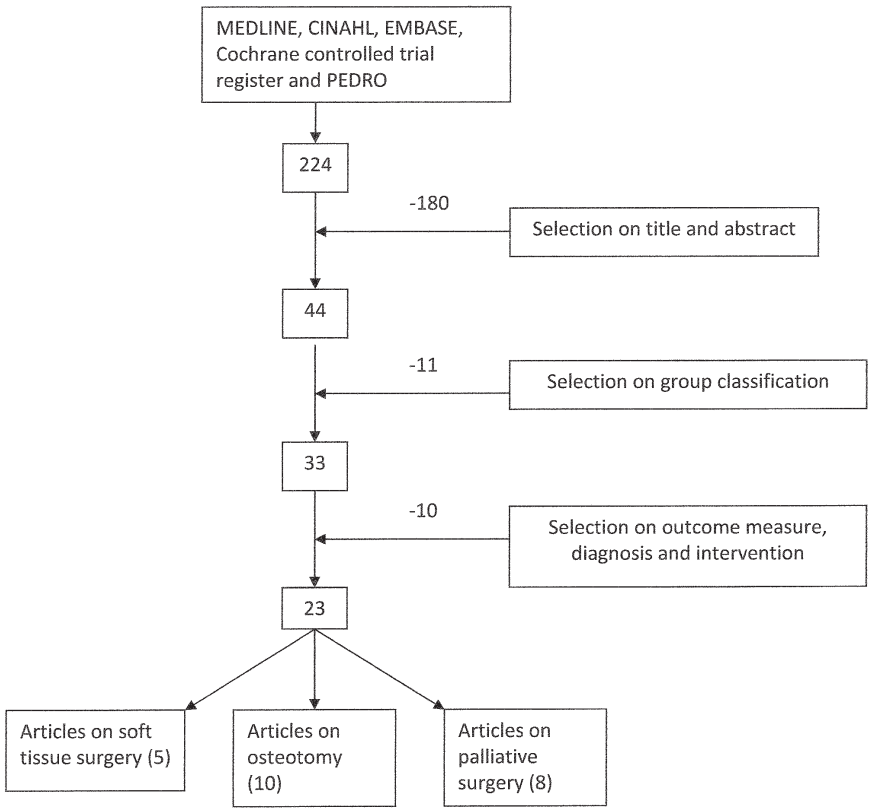

Figure 5.1 Literature search scheme systematic reviews.

different outcome measure than MP (neck shaft angle, center edge angle or acetabulum index), a different diagnosis or different interventions.

In the end, 23 articles met our inclusion criteria (5 regarding soft tissue surgery, 10 regarding osteotomies (VDRO/pelvic osteotomy) and 8 regarding palliative surgery). The palliative studies have been described elsewhere[8] and were excluded from the present study.

The quality assessment procedure was applied to five articles regarding soft tissue surgery (category 1) and 10 regarding osteotomies (category 2) (Table 5.2). Although the raters disagreed on 20% of the items, all disagreements were resolved after discussion, if necessary with the help of the third reviewer (EB).

Table 5.2 Methodological quality

Items First author	a	b	c	d	e	f	g	h	i	j	k	l	m	Sufficient quality	Score
Category 1: Soft tissue surgery															
Bowen 2006[15]	+	+	+	+	-	+	+	+	+	-	+	-	+	+	10
Cottalorda 1998[11]	+	+	+	+	-	+	+	-	+	-	+	-	+	-	
Kalen & Bleck 1985[12]	+	+	+	+	-	+	-	+	+	-	+	-	-	-	
Onimus 1991[13]	+	+	+	+	-	+	+	-	+	-	+	-	-	-	
Silver 1985[14]	+	+	+	+	-	+	+	-	+	-	+	-	+	-	
Category 2: Osteotomy															
Debnath 2006[6]	+	+	+	+	-	+	+	+	+	-	+	-	-	+	9
Herndon 1992[20]	+	+	+	+	-	+	+	+	+	-	+	-	+	+	10
Hoffer 1985[30]	+	+	+	+	-	+	+	-	+	+	+	-	-	+	9
Mubarak 1992[31]	+	+	+	+	-	+	+	+	+	+	+	-	-	+	10
Oh 2007[18]	+	+	+	+	-	+	+	-	+	+	+	-	+	+	10
Persiani 2008[17]	+	+	+	+	-	+	+	+	+	+	+	-	-	+	10
Robb 2006[16]	+	+	+	+	-	+/-	+	-	+	-	+	-	-	-	
Roposch 2005[26]	+	+	+	+	+	+	+	+	+	-	+	-	+	+	11
Sankar 2006[23]	+	+	+	+	-	+	+	+	+	+	+	-	-	+	10
Yun 2005[19]	+	+	+	+	-	+	+	+	+	-	+	-	+	+	10

See Table 5.1 for a description of a–m. + Sufficient methodological quality/item met; - Insufficient methodological quality/item not met.

Category 1 (soft tissue surgery)

Our initial selection included five articles involving soft tissue surgery. However, the quality of the studies by Cottalorda,[11] Kalen,[12] Onimus,[13] and Silver[14] was rated as insufficient in terms of the criteria formulated by Steultjens et al.[9] and Van Tulder et al.[10] This resulted in only one article (Bowen and Kehl 2006[15]) that met the quality criteria. The result of the quality assessment is represented in Table 5.2.

Characteristics and outcome measures (Table 5.3)

The study by Bowen and Kehl included a total of 70 hips in 38 patients. The mean age at surgery was 5.5 yrs, with a range of 2.4 to 9.7 years. The follow-up ranged from 1.7 to 19.2 years.

Bowen and Kehl found that 87% of the hips with a pre-operative MP<50% were stable at follow-up, versus 43% with a pre-operative MP >50% (p=0.031). The average patient age at surgery was 5.9 years for stable hips versus 3.8 years for unstable hips (p=0.001). Seventeen percent of all hips were unstable at follow-up and needed further surgery, consisting of femoral and/or pelvic osteotomy.

Best evidence synthesis

Because our quality assessment left only one article reporting a significant effect on hip migration in patients with CP of soft tissue surgery alone (Bowen[15]), we conclude that there is insufficient evidence for a favorable effect of soft tissue surgery of the hips in patients with severe CP.

Table 5.3 Characteristics and outcome of study involving soft tissue surgery (category 1)

First author	Hips (N)	Patients (N)	Mean age at surgery (range)	Duration of follow-up (range)	Co-interven-tions	MP preopera-tive	MP postoperative	Success Number of hips (%)
Bowen 2006[15]	70	38	5.5 (2.4–9.7)	7.4 years (1.7–19.2)	4 weeks A-frame	30% (15–79%)	37 of 42 hips with MP <30; 18 of 21 hips with MP 30–50 stable (stable = MP less than 15%)	58/70 (83%)

Category 2 (osteotomy)

Characteristics

Table 5.4 shows the main characteristics of the 9 included studies involving osteotomy. One study (Robb 2006[16]) was excluded due to insufficient quality. The remaining studies included 189 patients with 289 operated hips. The sample sizes ranged from 7 to 38 patients. Three studies involved only VDRO, two studies involved patients with VDRO or VDRO in combination with pelvic osteotomy (Persiani[17] and Oh[18]), and four studies involved combined osteotomy only. The various subcategories are shown in the tables.

The mean age of the patients at surgery was between 8 and 10 years, with a range of 2 to 22 years. The studies differed widely in terms of duration of follow-up.

Outcome measures

Migration percentage

Tables 5.5 and 5.6 show the outcome measures in category 2 (osteotomies).

The co-interventions were comparable (spica >4 weeks and physiotherapy). The MP decreased in all studies from means between 61 and 100% to a mean value at follow-up of between 6 and 29%. Various definitions have been used for a poor outcome or unstable hip at follow-up.

Forty-nine of the 289 hips (17%) were subluxated at follow-up and 9 (max 17) were dislocated. The percentage of patients who had an MP below 33% (meaning no subluxation) after surgery differed between the studies (53%–75% for VDRO alone and 85% or higher for the combined surgeries).

Best evidence synthesis

The review included 9 studies (with an uncontrolled design) of sufficient quality which used consistent outcome measures. The resulting best-evidence synthesis (Appendix 5.1) shows that there were indicative findings showing a favorable effect of VDRO/pelvic osteotomy in stabilizing the hip, after several years of follow-up. Significant improvement in MP after surgery was reported in three articles (Yun et al.,[19] Oh et al.,[18] and Debnath et al.[6]).

Table 5.4 Characteristics of studies involving osteotomies (category 2)

First author	Hips (N)	Patients (N)	Type of osteotomy	Indication for surgery	Mean age in yrs (range)	Duration of follow-up in yrs (range)
Herndon 1992[20]	48	32	VDRO	Treatment of dislocated/ subluxated hips	8.6 (4–21)	3.6 (1–8.9)
Hoffer 1985[30]	18	15	VDRO	Stabilizing hip. relieving/ preventing pain	13.3 (4–15)	11.5 (7–17)
Yun 2005[19]	69	38	VDRO	Treatment of subluxated/ dislocated hips	8.2 (2–16)	5.3 (2–13)
Persiani 2008[17]	13	13	VDRO with/without pelvic osteotomy (Chiari)	Treatment of subluxated/ dislocated hips	11 (6.2–22)	4.6 (2–9)
Oh 2007[18]	61	31	VDRO with/without pelvic osteotomy (Dega)	Preventing/relieving pain and improving ROM of hip	8.3 (2–16.2)	19.1 (10.8–21.4)[a]
Debnath 2006[6]	7	7	VDRO and pelvic osteotomy (Chiari)	Treatment of painful. dislocated/subluxated hips	14.1 (9.1–17.8)	13 (8–17.5)
Mubarak 1992[31]	18	11	VDRO and pelvic osteotomy (Salter)	Treatment of dislocated hips	8.4 (5.5–13)	6.8 (3.6–8.1)
Roposch 2005[26]	41	32	VDRO and pelvic osteotomy (modified Pemberton)	Relieving pain. improving ROM and improving sitting ability	9.5 (5.2–16.8)	5.3 (2–11.7)
Sankar 2006[23]	14	12	VDRO and pelvic osteotomy (modified Derqui)	Relieving pain. treatment of dislocation	10.6 (ND)	16.7 (12.4–19.5)

[a] Age instead of duration in years. ROM = range of motion; ND = not described.

Table 5.5 Outcome studies concerning osteotomies (category 2)

	Co-intervention	Definitions of outcome measures	Pre-operative assessments	Outcomes	Pain (N hips)	Additional outcomes
Herndon[20]	Spica cast 4–6 weeks Abduction orthosis 6 weeks full-time and 6–12 months bedtime	Good: CEA >15, coverage >80%, no operation needed, no pain Poor: recurrent dislocation, CEA <5, coverage <65%, pain	Only presented for the different outcome groups (good/fair/poor)	28 good, 15 fair, 5 poor 1 dislocated 4 subluxated	Pre ND FU: 5/48	Relation with age: mean age Poor 12 yrs, Fair 7 yrs, Good 9 yrs
Hoffer[30]	Spica cast 2–8 weeks	NS	3 dislocated 12 subluxated	1 dislocated 4 subluxated	Pre 5/18 FU: 2/18	2 patients better / one worse functional outcome ROM unchanged,
Yun[19]	ND	Subluxation MP 25–99% Dislocation MP 100% Failure: MP >40%/ resurgery	18 dislocated 14 subluxated mild 28 subluxated severe	18 failure (26%)* 10 MP >40%	ND	Mean abduction increased and mean flexion contracture decreased
Persiani[17]	Spica cast 6-8 weeks	Good: no pain, improved/stable functional outcome, MP <33%, Wiberg angle >20 Poor: pain, worse functional outcome, MP >50%, Wiberg <0	Mean MP 79%	VDRO 3 good, 3 fair, 2 poor CO 3 good, 1 fair, 1 poor	Pre: 9/13 FU: 2/13	1 better functional outcome
Oh[18]	ND	Success: stable hip (MP <25%) Failure: MP >60%, resurgery/ osteo-arthritis	17 MP 0-24% 8 MP 25-39% 13 MP 40-59% 8 MP 60-79% 15 MP 80-100% Spastic hip score	49/61 stable hip* 12 (20%) failure 4/12 VDRO 8/24 Combined	Pre: ND FU: 2/61	Dislocated hips: more unsatisfactory outcome in VDRO compared to VDRO plus pelvic osteotomy

	Co-intervention	Definitions of outcome measures	Pre-operative assessments	Outcomes	Pain (N hips)	Additional outcomes
Debnath[6]	Slings and Springs 3–4 weeks	Following Osterkamp et al. Good: pain-free/reduced hip Fair: subluxated hip/pain	2 dislocated 3 subluxated >50% 2 subluxated <50%	6 good, 1 fair, * non-dislocated	Pre: 7/7 Fu: 0	7 better personal hygiene
Mubarak[31]	Spica cast 6+4 weeks	Subluxation MP 30–99% Dislocation MP 100% Stable hip <30% MP	6 dislocated 12 subluxated	17/18 stable	Pre: ND Fu: 1/18	ROM mean abduction improved, 16 hips functional ROM
Roposch[26]	Abduction Petri cast 6–8 weeks	Subluxation MP 33–99% Dislocation MP 100%	18 dislocated 19 subluxated Mean MP 77%	Mean MP 13% 1 dislocated 5 subluxated	Pre: 41/41 FU: 1/41	39 hips functional ROM 1 dislocated hip: femoral head resection
Sankar[23]	Spica cast 4 weeks	Subluxation MP 33–99% Dislocation MP 100%	All dislocated	Mean MP 10% None sub/ dislocated	Pre: 14/14 FU: 0	No change in functional outcome 1 femoral head resection after 12 yrs increased abduction and extension ROM

ND = not described; CEA = centre edge angle; CO = Chiari osteotomy; MP = Reimers migration percentage; NS = not significant; Pre = pre-operative; FU = follow-up; ROM = range of motion. * statistically significant results.

Table 5.6 Adjusted values of migration percentage (MP) after osteotomies (category 2)

	N patients	N hips	Duration of follow-up (range)	Mean MP pre	Mean MP post	Pre subluxation (n)	Post subluxation (n)	Pre dislocation (n)	Post dislocation (n)	Post % MP <33%
Herndon[20,a,c]	32	48	3.6 (1–8.9)	68	19	ND	4	ND	1	ND
Hoffer[30,a]	15	18	11.5 (7–17)	61	29	12	4	3	1	72% (13)
Yun[19]	38	69	5.3 (2–13)	80	27	42	23[d]	18	10/69	74% (51)[b]
Persiani[17]	13	13	4.6 (2–9)	79	29.4	10	5	3	0	53% (7)
VDRO	8	8		75.6	32.8	6	3	2	0	50% (4)
CO	5	5		84.8	28.8	4	2	1	0	60% (3)
Oh[18]	31	61	19.1 (10.8–21.4)[e]	ND	ND	36[d]	2/36	15[d]	5/36	ND
Debnath[6]	07	07	13 (8–17.5)	81	17.8	5	1	2	0	85% (6)
Mubarak[31]	11	18	6.8 (3.6–8.1)	69	6.1	12	1	6	0	99.5% (17)
Roposch[28]	32	41	5.3 (2–11.7)	77	13	23	4	18	2	85% (35)
Sankar[23]	12	14	16.7 (12.4–19.5)	100	11	0	0	14	0	100% (12)

[a] Percentage coverage used. [b] <40% MP. [c] No definition of sublux/disloc. [d] Subluxation MP >40%. [e] Dislocation MP >80%. ND = not described.

The findings of these studies do not indicate a relationship between the effect of the surgical procedure and the patients' age and / or the duration of the follow-up, as the ranges reported in the different studies are too large.

Pain

Pain as an outcome measure was reported in eight studies. However, only five studies had recorded the number of patients with pre-operative pain. Pain was in most cases reported by parents and/or caregivers, although no exact definitions were provided. These five studies included a total of 93 patients, 76 of whom (81%) had pre-operative hip pain. At follow-up, 5 patients (5%) complained of pain. In the other three studies, 7% of the 107 patients had a painful hip at follow-up. However, no significance levels were recorded in any of these studies.

Best evidence synthesis

Although the above-mentioned data may suggest otherwise, there is insufficient evidence for a favorable effect of VDRO/pelvic osteotomy in reducing pain after several years of follow-up, according to the best-evidence synthesis. This is due to the abcense of significant levels.

Complications

Table 5.7 lists the complications of bony surgery reported in the category 2 studies.

Complications occurred in 25% (48 out of 191) of all patients. Herndon[20] and Debnath[6] reported 7 patients with heterotopic ossification. Osteoarthritis was only reported for the combined interventions (VDRO + pelvic osteotomy), with percentages of 8–28%. The mean percentage of fractures was 6%, although the study by Yun[19] found a percentage of 13%. Post-operative ulcers were reported in 5% of all patients.

DISCUSSION

This systematic review has examined the effectiveness of surgical interventions for hip displacement in children with severe cerebral palsy (CP), in the form of soft tissue and bony surgery to prevent further luxation or reduce the femur in the acetabulum.

Table 5.7 Complications after osteotomies (category 2)

Author		N patients	N hips	Number of complications	Percentage of complications (patients)	Percentage of complications (hips)
Herndon 1992[20]	VDRO	32	48	1 femur fracture 5 HO (by medial capsular release) without pain, 1 with pain	3% 16%	2% 10%
Hoffer 1985[30]	VDRO	15	18	1 pressure ulcer 1 avascular necrosis 2 limb length discrepancy	7% 7% 13%	5.5% 5.5% 11%
Yun 2005[19]	VDRO	38	69	5 fractures 6 ulcers	13% 15%	7% 9%
Persiani 2008[17]	VDRO with/without pelvic osteotomy (Chiari)	13	13	2 ulcer 1 superficial infection 2 fractures	15% 7,5% 15%	15% 7.5% 15%
Oh 2007[18]	VDRO with/without pelvic osteotomy (Dega)	31	61	5 osteoarthritis 3 windblown	16% 10%	8% 5%
Debnath 2006[6]	VDRO and pelvic osteotomy (Chiari)	07	07	2 late loss of joint space 2 progressive osteoarthritis 1 HO without pain	28.5% 28.5% 14%	28.5% 28.5% 14%
Mubarak 1992[31]	VDRO and pelvic osteotomy (Salter)	11	18	2 hamstrings release after 2 to 3 years 2 avascular necrosis 1 fracture	18% 18% 9%	11% 11% 5.5%
Roposch 2005[26]	VDRO and pelvic osteotomy (modified Pemberton)	32	41	None		
Sankar 2006[23]	VDRO and pelvic osteotomy (Modified Derqui)	12	14	3 fractures 1 osteoarthritis	25% 8%	21% 7%

HO = Heterotopic ossification; VDRO = varisating derotational osteotomy.

Soft tissue surgery

The studies we reviewed do not definitively prove the effectiveness of soft tissue surgery. In terms of the criteria by Steultjens et al.[9] and Van Tulder et al.,[10] only one retrospective study had sufficient methodological quality. This is due to insufficient internal and statistical validation in most studies. We therefore conclude that there is insufficient evidence for the effectiveness of soft tissue surgery in preventing (sub) luxation in children with severe CP.

Our findings are consistent with the results of the AACPDM systematic review performed by Stott and Piedrahata,[7] who studied the effect of soft tissue surgery in children including all GMFCS levels in CP. They reported an overall improvement in Reimers' migration percentage in 241 of 467 (52%) hips. However, their conclusions were weakened by the heterogeneity of the patient populations studied and the variety of the surgical procedures undertaken.

The question remains whether adductor tenotomy should be advocated in patients with spastic CP for whom an X-ray of the pelvis shows progressive subluxation of the hip. It is common knowledge that the success of soft tissue surgery depends on the patient's age. In his well-known paper, Scrutton (1989[21]) promoted performing soft tissue surgery before the age of five. Before this age, the acetabulum is supposed to possess the ability to remodel itself after the forces around femur and pelvis have been altered by means of adductor tenotomy.

Another important factor influencing the success rate of soft tissue surgery is the GMFCS level. Recent research in Australia has revealed that the success of adductor tenotomy is inversely related to the GMFCS classification.[22]

A third important factor is the preoperative level of migration. Two studies have advocated soft tissue surgery to prevent further luxation before MP has reached 50%.[12,15] Regrettably, all this clinical evidence is based on retrospective studies of insufficient methodological quality, and the single study that could be included in this review does not sufficiently support the clinical evidence.

Bony surgery

Our review shows that there are indications for a positive effect of VDRO/pelvic osteotomy in stabilizing the hip, which persists for several years of follow-up. The

studies involving bony surgery without pelvic osteotomy show a mean MP at follow-up of between 6 and 29%, a subluxation percentage of 15–47% and a dislocation percentage of 3–6%. The combined intervention (VDRO + pelvic osteotomy) shows better outcomes, with a lower number of re-subluxations and dislocations, compared to VDRO only.

Several studies have reported a relation between severe preoperative (sub)luxation (high MP) and a poorer outcome at follow-up. Our review also indicates that a high migration percentage may necessitate an additional pelvic osteotomy.[17,18] Yun[19] used an MP of 60% as a threshold to add pelvic osteotomy to VDRO. The likelihood that this additional procedure is needed increases with age. At higher age, longer duration of the subluxation increases the risk of dysplasia of the acetabulum, which makes additional pelvic surgery unavoidable.

On the other hand, the combination of VDRO and pelvic osteotomy may result in a higher rate of osteoarthritis.[6,18,23] Additionally, the complication rate of this procedure is quite high: up to 28.5% (Table 5.6). Treatment with only VDRO involves a moderate complication rate of 1–11%.

One must also keep in mind that adding pelvic osteotomy to the VDRO procedure implies the use of a postoperative spica cast, which means a prolonged period of immobilization. Using VDRO alone can considerably reduce the period in which the patient is bedridden.

Recently, a new approach to the peroperative assessment of the need for additional pelvic osteotomy has been published.[24] The authors advocate the use of peroperative fluoroscopy and arthrography for this purpose. Others have suggested using preoperative three-dimensional MRI for this purpose.[25]

These findings give rise to questions about the timing of bony surgery. Some have advocated performing surgery at a relatively young age (8–9 yrs), to make use of the ability of the bones to remodel themselves, and to prevent further migration and the concomitant acetabular dysplasia.[17] However, in view of the wide range of ages at the time of surgery and the large variety of follow-up durations within and between the studies in our systematic review, we were not able to relate the outcome measures to age. Discussing the timing of interventions also requires information about prior interventions. Unfortunately, the studies included did not provide sufficient information about preceding interventions.

Our study revealed no evidence about the usefulness of the different types of pelvic osteotomy. Several techniques were used, including personal modifications of more common techniques.[23,26] The number of individual techniques was too small to allow conclusions about the value of specific techniques.

General considerations

Although we were aware of the lack of randomized controlled trials, we expected that the method for quality assessment and best-evidence synthesis proposed by Steultjens et al.[9] and Van Tulder et al.[10] would enable us to make a statement on the usefulness of soft tissue and bony surgery in this group of patients with CP. Although we think we succeeded in this design, the conclusions are weakened by the variety of surgical procedures applied, the range of ages and the varying definitions used for the outcome measures.

There is a shortage of measures to assess the amount of pain expressed by the patients themselves. This is a limitation, especially for patients with intellectual disability, a common phenomenon in severe CP. Therefore, conclusions concerning the effect in terms of pain relief must be drawn with caution. The results suggest that bony surgery may reduce the pain that occurs in older children. In some cases, the pain related to subluxation may disappear when the subluxation progresses to total luxation. In that case, bony surgery can be avoided just by waiting for this process to take place.

We used MP as an outcome measure, as this allowed us to compare the outcomes of all studies. Throughout the literature, MP is a highly accepted measure to describe the severity of hip displacement. Nevertheless, the definitions used for subluxation differed between the studies. To improve the use of "(sub)luxation" as an outcome measure we tried to recalculate the number of subluxations using the definition proposed by Reimers.[27]

In the near future, the CP hip classification system proposed by Robin et al.[28] can prove to be useful for this category of patients. In addition to measures like MP and pain, other aspects such as quality of life can also be useful to measure. Recently, Narayanan et al. used their CPCHILD assessment instrument to prove that CP patients who are waiting for or are considered ineligible for hip surgery have a significant lower CPCHILD score than patients who have undergone hip surgery.[29]

Several studies were excluded from our review simply because they had included all GMFCS levels and we were not able to separate these levels. We focused on the group of patients with severe CP because the incidence of hip migration is higher in this group and because the timing of interventions, reasons for intervention and way of intervention differ from those in the group of patients who have the ability to walk.

CONCLUSION

Once a patient with severe cerebral palsy has been diagnosed with subluxation, a few options are available to prevent further progression and/or to reduce the femur. Since our literature search found only one article that met the criteria for sufficient methodological quality, we did not find enough evidence to unequivocally support soft tissue surgery as the method of choice in young children (below the age of five) with an MP below 50%. The clinical practice of performing soft tissue surgery at a young age because of the relatively low impact of the procedure on the patient and their parents is thus not supported by substantial scientific evidence.

If soft tissue surgery fails, varus derotational osteotomy can be a beneficial procedure in children with severe CP. For patients of a higher age and with severe acetabular dysplasia, pelvic osteotomy can be successfully added. One must keep in mind, however, that this procedure constitutes a major event for these vulnerable patients, and requires a long period of recovery. The complication rate is substantial, particularly when pelvic surgery is added to VDRO. This is the reason why some people reject bony surgery as a first choice treatment in patients with GMFCS IV and V, provided that there is no prolonged intractable pain. If bony surgery in these vulnerable patients is unavoidable, for instance because of prolonged intractable pain, the timing of surgery is crucial.

The debate about the type of intervention and the best timing is ongoing. Careful surveillance of the development of the acetabulum by means of frequent X-rays is needed to ensure that the acetabulum is capable of accommodating the femoral head, in case VDRO is necessary. The development of new assessment methods such as 3D CT can be helpful in this respect. Multicenter trials with larger groups of patients that can be followed for a longer period can provide further insights into this complicated matter.

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APPENDIX 5.1

Best evidence synthesis

Strong evidence	Provided by consistent, statistically significant findings on outcome measures in at least two high-quality RCTs
Moderate evidence	Provided by consistent, statistically significant findings on outcome measures in at least one high-quality RCT or high-quality CCT
Limited evidence	Provided by consistent, statistically significant findings on outcome measures in at least one high-quality RCT or Provided by consistent, statistically significant findings on outcome measures in at least one high-quality CCT
Indicative findings	Provided by consistent, statistically significant findings on outcome and/or process measures in at least one high-quality CCT or low-quality RCT (in the absence of high-quality RCTs) or Provided by consistent, statistically significant findings on outcome and/or process measures in at least two ODs with sufficient quality (in the absence of RCTs and CCTs)
No or insufficient evidence	If results of eligible studies do not meet the criteria for one of the above levels of evidence or In case of conflicting (statistically significant positive and statistically significant negative) results among the RCTs and CCTs or In case no eligible studies are available

RCT = randomized controlled trials; CCT = controlled clinical trials; OD = other design than controlled.

Chapter 6

Palliative hip surgery in severe cerebral palsy: a systematic review

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Submitted for publication.

ABSTRACT

We performed a systematic review of the results of palliative hip surgery in severe cerebral palsy. People with severe cerebral palsy frequently suffer from pain and other impairments due to dislocation or malformation of the hips. When preventive or reconstructive surgery fails, palliative intervention is undertaken. A number of salvage interventions have been described. We found articles on resection surgery of the femoral head, arthrodesis of the hip joint and total hip replacement. The published literature does not clearly favor one procedure over the others. The resection arthroplasty technique developed by Castle[1] is reported to yield the best results and fewer complications, and seems to eventually lead to a good outcome.

INTRODUCTION

Patients with severe cerebral palsy frequently develop subluxation or dislocation and associated deformity of the femoral head. The incidence increases with the GMFCS (Gross Motor Function Classification System) level:[2,3] patients with a GMFCS V level have an almost 90% risk of developing a migration percentage >33%.[2]

As we found in a previous study, luxation of the hip increases the risk of hip pain in adults from 10 to about 30%.[4] Luxation is also associated with seating problems due to hip adduction contractures, and with problems of perineal hygiene.[5] Hence, luxation of the hip in patients with severe cerebral palsy should be prevented or treated.

A large number of nonsurgical or surgical interventions to prevent or correct luxation of the hip have been applied, not all of them very successful, as the measures sometimes fail or cause unintended side effects. The result of unsuccessful bony surgery can be disastrous to the patient, as we found in our previous study: 12 of 17 patients with an abnormal configuration of the femoral head in relation to the pelvis suffered from pain in the hip after failed bony surgery.[4]

Several methods have been described to offer relief to patients in whom the hip disorder causes complications, with the aim of resolving pain, improving sitting comfort and reducing nursing problems. Most of these methods, however, like injection of botulinum toxin into the adductor muscles, have proved unsuccessful or relieve pain and discomfort only temporarily.[6] Concomitant loss of articular cartilage surface occurs frequently[7,8] and leaves no other option than palliative procedures, which include proximal femoral head resection, valgus osteotomy of the femur, total hip replacement and hip arthrodesis.

Several techniques for resection arthroplasty have been advocated in the literature. Our earlier study found that distal resection arthroplasty gave the best result in terms of pain relief.[4] Distal in this case means a resection of the femoral head below the trochanter minor. This technique, described by Castle and Schneider in 1978, involves an extraperiosteal dissection and division of the proximal femur at least 3 cm below the lesser trochanter. A capsular flap is constructed across the acetabulum and the quadriceps muscles are sutured over the end of the femur.[1]

The Castle technique was modified by McCarthy et al. to include interposition involving suturing the iliopsoas and gluteal muscles to the hip capsule ("closing" the acetabulum) and covering the femoral stump by suturing the lateral vastus muscle to the muscles and soft tissues on the medial side.[9]

Valgus osteotomy of the femur is a technique that was described several years ago by Schanz.[10] To some orthopedic surgeons, this is the preferred technique, for which they claim good results. The advantage of this technique is that it does not require a long postoperative treatment with traction or plaster of Paris. However, many surgeons refrain from the use of traction after resection as described by Castle.

Palliative surgery, which is obviously only performed when there are no other options, can be successful, but is very intrusive to these severely disabled patients, because of the need for bony surgery. There is yet no consensus about the indication for this type of intervention. Pain relief is frequently reported, but complications are also numerous, depending on the type of intervention and the type of postoperative treatment. Complications have been described, but as far as we know, no systematic review of the effectiveness of the different methods has been undertaken.

In order to develop a treatment algorithm for surveillance and treatment of patients with hip disorders related tot bilateral spastic cerebral palsy, we performed a systematic review of the effectiveness and complications of palliative surgery of the hips in patients with CP and GMFCS level IV or V.

METHOD

Literature search

Our research question was: *what is the effectiveness and what are the side effects of palliative surgery for hip problems in CP patients with GMFCS IV or V*. To answer this question, an extensive literature research was conducted by two independent researchers (CBB & HCvdH), consulting the following databases: Medline, Cochrane, Cinahl, Pedro and Embase. The keywords and MeSH terms were

- Cerebral palsy (MeSH and text word)
- Surgery/ surgical procedures (MeSH and text word)
- Hip (MeSH and text word)
- Hip (sub)luxation/dislocation (text word)
- Tendons/surgery (MeSH and text word)

Articles were included by two independent researchers (CBB & HCvdH) based on information from title and abstract. If this was not clear enough, they read the whole article. In case of disagreement, a third researcher (EJB) was asked to read the article and a final decision was made by consensus.

We distinguished between articles concerning salvage surgery and those reporting on prophylactic (soft tissue surgery) or reconstructive interventions (derotational varus osteotomy / pelvic osteotomy). The latter will be addressed in a separate study.

The following inclusion criteria had to be met:

- All types of studies concerning palliative intervention (resection arthroplasty, total hip replacement, arthrodesis and valgus osteotomy).
- Diagnosis CP; GMFCS IV or V; diagnosis of pelvis / femur relation by X-ray; indication for surgery stated.
- Articles had to explain the surgical procedure and report the effect in terms of indication; follow-up had to comprise one year or more.
- Articles had to be written in English, Dutch or German.

Quality assessment

The methodological quality of the selected articles was assessed by two independent researchers (EJB & HCvdH). Disagreements were resolved by discussion. If no consensus was reached, the third researcher was asked and a final decision was made. A list of criteria used by Steultjens et al.[11] was used to assess the methodological quality. This is a modified version of the list by Van Tulder et al., to assess uncontrolled studies (i.e. not RCT or CCT)[12] (Table 6.1). This list contains seven criteria for internal validity, four descriptive criteria and two statistical criteria. Studies were considered to be of sufficient methodological quality if at least four internal validity criteria, two descriptive criteria and one statistical criterion were met.

In view of the diversity of surgical methods included in the studies, and the type of studies (observational studies), meta-analysis or statistical analysis was not considered appropriate. We used a best evidence synthesis according to Steultjens et al.[11] (Appendix 6.1), and present a descriptive summary of the results.

Table 6.1 Criteria for methodological quality (other than controlled designs) (Steultjens et al.[11] and Van Tulder et al.[12])

Patient selection: a. eligibility criteria specified?
Interventions: b. intervention explicitly described? c. co-interventions described? d. compliance acceptable?
Outcome measurement: e. outcome assessor not involved in the treatment? f. outcome measures relevant? g. adverse effects described? h. withdrawal/dropout rate reported and acceptable? i. timing of follow-up measurements: short-term follow-up measurement performed? long-term follow-up measurement performed? j. timing of the outcome assessment comparable for all patients?
Statistics: k. sample size of the patient group described? l. did analysis include an intention-to-treat analysis? m. point estimates and measures of variability presented for the primary outcome measures?
Internal validity criteria: c, d, e, f, h, j, l. Descriptive criteria: a, b, g, i. Statistical criteria: k and m.
Studies were considered to be of 'sufficient quality' if at least 4 criteria for internal validity, 2 descriptive criteria and 1 statistical criterion were met.

RESULTS

The literature search identified 224 articles. The initial selection based on title and abstract yielded 44 articles (Figure 6.1). All concerned observational studies. Eleven studies were excluded because they had incorporated all severity classes of CP and it was not possible to separate them. Ten studies were excluded because they had used a different outcome measure than the migration percentage, a different diagnosis, or different interventions.

Finally, 23 articles met our inclusion criteria (5 regarding soft tissue surgery, 10 regarding osteotomies (DVO / pelvic osteotomy) and 8 regarding palliative surgery).

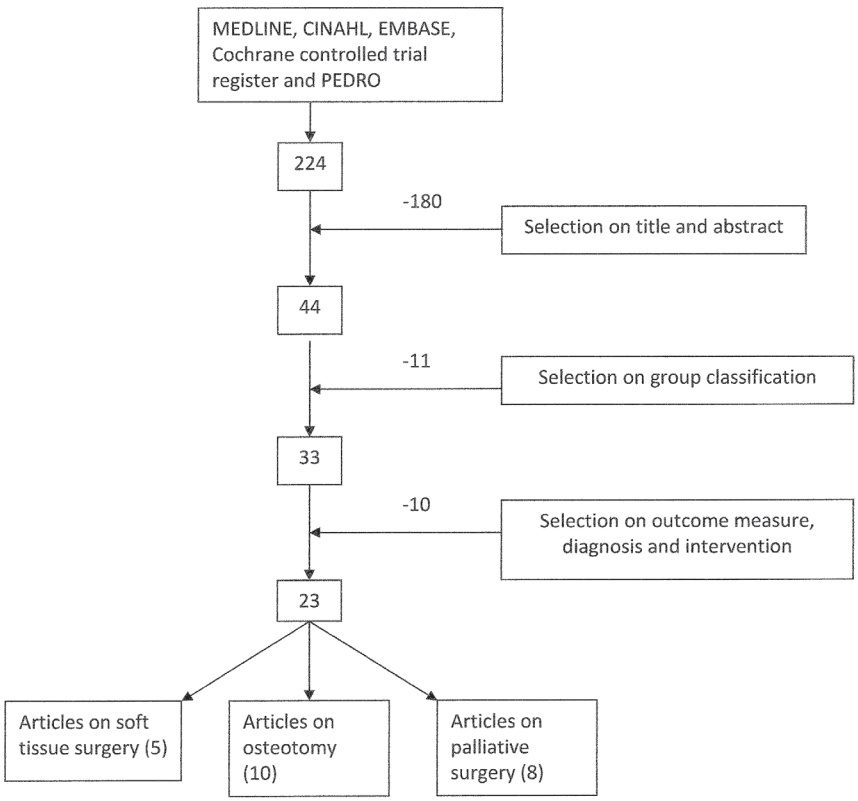

Figure 6.1 Literature search scheme systematic reviews.

The first 15 studies will be the subject of a separate review, while the eight studies on palliative surgery met our inclusion criteria and are described in the present review.

Methodological quality

All eight articles passed the methodological quality assessment (Table 6.2). We did not find any high-quality systematic reviews, or case-control studies. All articles were retrospective cohort studies. Patients were described as GMFCS category V,[23] or nonambulant.[14-17,24,25] One article described a mixed population, but the nonambulant patients could be derived from the tables.[18] The studies were divided into articles on femoral head resection as a palliative treatment (n=6), total hip replacement (n=1) and arthrodesis of the hip joint (n=1). The results of these studies are presented below as descriptive summaries within these different categories. Unfortunately, we did not find any article about Schanz valgus osteotomy that passed our quality criteria.

Table 6.2 Methodological quality

Article	Patient selection	Interventions	Outcome measurement							Statistics				Internal validity c, d, e, f, h, j, l	Descriptive criteria a, b, g, i	Statistical criteria k, m	Total score
	a	b	c	d	e	f	g	h	i	j	k	l	m				
Articles on resection of the femoral head																	
Abu-Rajab[23]	+	Castle	+	+	-	+	+	+	+	+	+	-	-	5	4	1	10
Baxter[24]	+	Castle	+	+	-	+	+	+	3 out of 4	-	+	-	-	4	4	1	9
Knaus[16]	+	McCarthy	+	+	ND	+	+	+	+	-	+	-	-	4	4	1	9
Lampropulos[25]	+	Castle	ND	+	-	+	+	+	+	+	+	-	-	4	4	1	9
Muthusamy[14]	+	McCarthy	+	+	-	+	+	+	+	+	+	-	-	5	4	1	10
Widmann[15]	+	McCarthy	+	+	+	+	+	+	+	+	+	-	-	6	4	1	11
Article on total hip prosthesis																	
Gabos[17]	+	+	+	+	-	+	+	+	5 out of 11	-	+	-	-	4	3	1	8
Article on arthrodesis																	
de Moraes Barros[18]	+	+	+	+	ND	+	+	+	+	+	+	-	-	5	4	1	10

See Table 6.1 for a description of a–m. + Sufficient methodological quality/item met; - Insufficient methodological quality/item not met; ND = not described.

Palliative resection of the femoral head

The articles reported on 82 patients, ranging in age from 3 to 45 years (Table 6.3). The Castle technique and the McCarthy technique were most frequently used, in 22 and 60 cases, respectively. There were only two studies in which the outcome assessor was not involved in the treatment. The follow-up period ranged from one to over 20 years. Unfortunately, the timing of the outcome assessment varied greatly across the studies, which makes the results difficult to compare. The impairments of the patients, i.e. pain, seating problems and nursing problems, were recorded but not clearly classified.

The heterotopic ossification (HO) grade was assessed in three studies using the McCarthy scale, while one study used the Brooker classification.[13]

Postoperative treatment consisted mostly of traction therapy, attached to the skeleton (26 cases) or the skin (22), or hinged fixation combined with plaster of Paris (34).

Pain

In all articles, pain was the primary indication for surgery. The level of pain was mostly assessed dichotomously, only pain or no pain being recorded. Only one author used a semi-quantitative scale.[14] One study used the amount of pain medication as a measure of pain.[15] Studies using the Castle technique reported 90–100% improvement of the pain (Tables 6.4A and 6.4B), while the improvement reported in the articles on the McCarthy technique ranged from 53 to 77%.

Best evidence synthesis

According to the best evidence synthesis (Appendix 6.1), the indicative findings of these studies suggest that pain relief can be achieved after a salvage resection of the femoral head. The articles of the highest quality, i.e. those by Abu Rajab et al.,[25] Widmann et al.,[15] and Muthusamy et al.,[14] report significant improvement in terms of pain experienced.

Seating / nursing problems

Pre-operatively, the different studies reported seating problems in 44–100% of the cases. Postoperatively, all patients were reported to no longer have seating disabilities.

Nursing problems were described in 38–100% of the cases before surgery. After surgery, these problems were reported to have completely resolved in 100% of the cases in

Table 6.3 Characteristics of studies using resection of the femoral head

Article	No. of patients	Age	Surgeon	Surgical technique	Pelvis X-ray pre-op	Treatment post-op	Follow-up	Outcome measure			Complications
								Pain	Seating problems	Nursing problems	HO
Abu-Rajab[23]	15	11–26	1	Castle & Schneider	Dislocation	3 wks skeletal traction	1–7 yrs	Recorded, but undefined	Recorded, but undefined	Recorded, but undefined	McCarthy classification
Baxter[24]	4	10–16	1	Castle & Schneider	Dislocation	3–6 wks skeletal traction	6 mo–5 yrs	Discomfort as inclusion criterion; pain as dimension, recorded, but undefined	Recorded, defined as detoriation of sitting ability vs. comfortable sitting	Recorded, defined as inability to maintain adequate perineal hygiene, vs. dramatic improvement	Not classified
Knaus[16]	20	3–27	ND	McCarthy	3 Subluxation (MP >33%); 24 dislocation (MP >90%)	2–5 wks skin traction (13); skeletal traction (1); plaster (5)	1–6.5 yrs	Recorded, defined as none, mild or severe	Recorded, but undefined. Follow-up included satisfaction question in 4 categories	Recorded, but undefined. Follow-up included satisfaction question in 4 categories	McCarthy classification

Article	No. of patients	Age	Surgeon	Surgical technique	Pelvis X-ray pre-op	Treatment post-op	Follow-up	Outcome measure			Complications
								Pain	Seating problems	Nursing problems	HO
Lampropulos[25]	3	13–16	ND	Castle & Schneider	Dislocation	Hinged external fixation at hip joint	15–24 mo	Recorded, but undefined	Recorded, but undefined	Recorded, but undefined	Not classified
Muthusamy[14]	27	2 groups: immature skeleton 6.8–15.8 yrs; mature skeleton 14.1–28.1 yrs	ND	McCarthy; intertrochanteric resection of femur	Serious subluxation (MP >50%) or dislocation	Plaster or hinged external fixation at hip joint	1–13.2 yrs	Self-developed questionnaire with three categories, including time needed for improvement and drug use	Self-developed questionnaire, several categories. Improved, unchanged or worse	Self-developed questionnaire, several categories, relating to range of motion Improved, unchanged or worse	McCarthy classification
Widmann[15]	13	10.7 / 45.5 yrs		McCarthy		6 skeleton traction, 9 skin traction, 5 radiotherapy	2.2 / 20.8 yrs	Recorded, drug dose defined	Recorded, span from bed defined	Self-developed questionnaire, several categories.	Brooker classification

ND = not described; HO = heterotopic ossification.

Table 6.4A Results reported in articles on resection of the femoral head

Article	Outcome measure						During surgery	
	Pain pre-op	Pain post-op	Seating problems pre-op	Seating problems post-op	Nursing problems pre-op	Nursing problems post-op	Arthrosis	Lateral defect of femoral head
Abu-Rajab[23] (Castle)	95%	90% improvement	57%	100% improvement	38%	100% improvement	93%	90%
Baxter[24] (Castle)				100% improvement		100% improvement	80%	
Knaus[16] (McCarthy)	17 serious; 2 mild, 1 none	8 out of 15 serious to none (53%); 7 serious to mild (47%)	8 out of 18 (44%)	100% improvement	16 out of 20 (80%)	15 out of 16 improved (94%)		
		Satisfaction at follow-up: 8 very satisfied, 6 satisfied, 3 dissatisfied, 2 unknown, 1 died						
Lampropulos[25] (Castle)	3 out of 3	3 out of 3 improved	3 out of 3	3 out of 3 improved	3 out of 3	3 out of 3 improved		
Muthusamy[14] (McCarthy)	27 out of 30 (90%)	4 out of 30 (13%)	100%	Significantly improved, by 4.4 hours	100%	12 out of 15 functional categories improved		
Widmann[15] (McCarthy)	Not quantified	Significant reduction of drug use, after mean of 5.6 months	Seating possible for mean of 3.2 hours	Seating possible for mean of 8.9 hours	54% nursing problems	100% improved		

Table 6.4B Results reported in articles on resection of the femoral head

Article	Complications							
	Migration of femur stump	HO	Other	Prolonged pain post-op	Death	Obturatorius neurinoma	Abduction contracture	Pressure ulcer
Abu-Rajab[23] (Castle)	[3 wks skeletal traction] Above acetabulum level: 0 At acetabulum level: 19 (90%) Below acetabulum level: 2 (10%)	12 out of 21 hips (57%). 11 out of 12 grade 1 (92%), 1 grade 2 (8%)	1 revision of stump 5%					
Baxter[24] (Castle)	[3–6 wks skeletal traction]	0%				1 = 25%	1 = 25%	1 = 25%
Knaus[16] (McCarthy)	[2–5 wks skin traction (13); skeletal traction (1); plaster (5)] Mean proximal migration 2.1 cm post-op, mean 4.8 cm at follow-up. With plaster more than with traction	21 out of 34 (62%). Grade 1 in 3 out of 21 (14%), grade 2 in 6 out of 21 (29%), grade 3 in 12 out of 21 (57%)	1 deep venous thrombosis	7 out of 20 (35%)	2 out of 20 (10%)			
Lampropulos[25] (Castle)	[Hinged external fixation at hip joint] 3 out of 4 hips below level of acetabulum post-op; 1 hip revision	3 out of 3 "light"						
Muthusamy[14] (McCarthy)	[Plaster or hinged external fixation at hip joint] 17% proximal migration, mostly with external fixation	41% in total: 11% mild, 22% moderate, 8% serious; more with external fixation, more with longer follow-up						
Widmann[15] (McCarthy)	[6 skeleton traction, 9 skin traction, 5 radiotherapy] 5 out of 18 had proximal migration = 28%; 9 out of 18 at level (50%); 4 out of 18 below level (22%)	2.7 on Brooker scale without radiotherapy; 0.8 with radiotherapy	2 out of 18 pneumonia 2 stump revisions					4 out of 18

HO = heterotopic ossification.

Table 6.5 Characteristics of studies using total hip replacement and arthrodesis

Article	Research type	GMFCS	No. of patients	Age	Surgical technique	Pelvis X-ray pre-op	Osteoarthritis	Treatment post-op	Follow-up	Outcome measure			Complications
Article on total hip replacement - description													
										Pain	Seating problems	Nursing problems	HO
Gabos[17]	Retrospective cohort study	V	11	11–20	Total hip or shoulder prosthesis in acetabulum	7 out of 11 with dislocation	100%	Abduction pillow of plaster with broomstick 4–6 wks	2 yrs–6 yrs 4 months	Pain upon hip motion: crying aloud or prolonged facial grimacing	Limited sitting tolerance, in minutes	Restricted perineal access for hygienic care	Booker classification
Article on hip arthrodesis – description													
Moraes Barros[18]	Retrospective cohort research	V	7	10–17	AO osteosynthesis	Several patients with history of failed soft tissue or bony surgery, such as McHale procedure or valgus osteotomy	2–4 months of plaster	2–7 yrs	Recorded, but undefined	All bedridden			

HO = heterotopic ossification.

Table 6.6 Results reported in articles on total hip replacement and arthrodesis

Article	Outcome measure				During surgery				Complications				
	Pain pre-op	Pain post-op	Seating problems pre-op	Seating problems post-op	Nursing problems pre-op	Nursing problems post-op	Osteo-arthritis	Lateral defect of femoral head	Abduction contracture	Other	Femoral fracture	HO	Dislocation
Article on total hip replacement – results													
Gabos[17]	100%	Complete relief in 10 out of 11 (91%)	100%	5 out of 11 unlimited sitting tolerance (45%)	91%	100% improved			1 out of 11 (9%)		4 out of 11 (36%)	Grade II in 4 out of 14 hips (29%) Grade III in 2 out of 14 hips (14%)	4 out of 14 hips (29%)
Article on hip arthrodesis – results													
Moraes Barros[18]	100%	100% resolved	All bedridden	5 out of 7 could sit comfortably			100%	100%		3 out of 7 non-union (42%)	Once		

HO = heterotopic ossification.

which the Castle technique was used, and in 92% of the cases in which the authors used the McCarthy technique.

Best evidence synthesis

Our best evidence synthesis (Table 6.2 and Appendix 6.1) reveals that there are indicative findings for a favorable effect in terms of seating and nursing problems after palliative resection of the femoral head. The articles of the highest quality, i.e. those by Abu Rajab et al.,[25] Widmann et al.,[15] and Muthusamy et al.,[14] reported significant improvement in seating and nursing problems.

Complications

Postoperative proximal migration of the femur occurred in only one of the 22 cases (5%) in which the Castle technique was used, compared to 17–28% of the cases in which the McCarthy technique was used.

Heterotopic ossification occurred in 0–57% of the cases in which the Castle technique was used, but was in most cases mild. Studies using the McCarthy technique reported a higher frequency of HO, ranging from 30 to 62%.

Other complications of surgery were reported in 22 of the 82 cases. The type of complications was diverse, the most frequently reported problems being postoperative pain (7 out of 20 cases in the study by Knaus,[16] using the McCarthy technique) and pressure ulcers (4 out of 18 cases in the study by Widmann,[15] using the McCarthy technique). Age did not influence the incidence of complications.[14]

Total hip arthroplasty

The article by Gabos[17] concerns 11 patients, ranging in age from 11 to 20 years (Table 6.5). Total hip or shoulder endoprosthesis was used, depending on the size and shape of the acetabulum. The follow-up period ranged from 2 to 6.5 years. Pain was assessed using the criterion of patients crying out or grimacing. Sitting tolerance was measured in minutes. The Brooker classification for HO was used.

Postoperative treatment consisted of an abduction pillow of plaster of Paris, applied for 4–6 weeks.

Pain

Pain was the primary indication for surgery. Pain relief after surgery was reported in 10 out of 11 patients (91%, Table 6.6).

Seating / nursing problems

Pre-operatively, all patients had seating problems. Postoperatively, 5 out of 11 patients (45%) were reported to have unlimited sitting tolerance.

Nursing problems improved from a prevalence of 91% before surgery to complete resolution in all cases.

Best evidence synthesis

The articles provided no or insufficient evidence for the effect of hip arthroplasty in terms of decreasing pain or seating/nursing problems, as these problems were insufficiently defined and there was only one article that met our quality criteria.

Complications

Dislocation of the prosthesis occurred postoperatively in 4 out of 14 hips (29%).

HO occurred in 5 out of 11 cases (45%), 3 with grade 2 and 2 with grade 3. There was a relatively high incidence of femoral fractures: 4 out of 11 (36%).

Arthrodesis of the hip joint

The article by De Moraes Barros[18] described 7 patients with GMFCS level V, ranging in age from 10 to 17 years (Table 6.5). The majority of the cases concerned patients with a history of failure of soft tissue or bony surgery, such as a McHale procedure.

The follow-up period ranged from 2 to 7 years. Pain was recorded, but not quantified in any way. Sitting problems were not reported because all patients were bedridden. HO and nursing problems were not reported.

Postoperative treatment consisted of plaster of Paris, applied for 2–4 months.

Pain

Pain was the primary indication for surgery in all patients. Pain relief after surgery was reported for all patients (Table 6.6).

Seating problems

Pre-operatively, all patients were bedridden. Postoperatively, 5 out of 7 patients were reported to be able to sit in a chair.

Best evidence synthesis

There was no or insufficient evidence for the effectiveness of arthrodesis in terms of reduced pain or seating/nursing problems, because these problems were insufficiently defined and there was only one article that met our quality criteria.

Complications

Non-union of the arthrodesis was reported in 3 of 7 patients (42%). One patient suffered a femoral fracture.

DISCUSSION

Pain associated with hip disorders can be a huge problem for patients with severe cerebral palsy (CP). In our previous study of determinants of pain in CP, 20 out of 160 patients suffered from pain due to an abnormal configuration of the femur in relation to the pelvis, frequently after failed bony surgery.[4] When persistent pain is not treatable with conservative methods, surgery can be helpful.

Our systematic review was performed to describe the effects and complications of salvage surgery (resection arthroplasty, total hip replacement, arthrodesis and valgus osteotomy) in adults and children with hip dislocation associated with severe CP. Studies assessing surgical procedures have mostly been retrospective cohort studies rather than controlled trials, and were in that sense of low quality. Our systematic review used the method developed by Van Tulder et al.[11] and Steultjens et al.[12] to assess the methodological quality of the non-controlled studies we included. Eight studies met the criteria, enabling us to draw some cautious conclusions. Based on the best-evidence synthesis, there were only indicative findings for the efficacy of the palliative resection procedures. The tables present a descriptive summary.

The majority of the articles that satisfied our selection criteria suggest that femoral resection is the most preferable method to treat these patients, and that the Castle procedure is to be preferred to the McCarthy procedure. This conclusion should be drawn

with caution, as statistical analysis was not possible. The Castle procedure has a higher success rate in terms of pain (90–100% versus 53–77%), a higher rate of improvement in terms of seating problems (100% versus 92%), and a very low rate of proximal migration of the femur (5% versus 17–28%). In addition, the rate of postoperative HO is lower in patients treated with the Castle technique (with reported values of 0–57% and low grades, versus 30–62%). The careful resection of the periosteum described by McCarthy to prevent HO may have a counterproductive effect.

Valgus osteotomy of the femur, as described by Schanz,[10] is advocated by several orthopaedic surgeons. This technique was originally described for patients with congenital dislocation of the hip. We found only a few articles on the use of this technique in the literature, and none of them met our quality selection criteria. Some surgeons prefer a procedure as described by Girdlestone[19] to cure otherwise incurable pain in patients with cerebral palsy. We did not find any articles about the value of this technique that met our quality criteria. We point out that in our own research all of the patients who had undergone a proximal resection of the femoral head because of pain all still suffered from pain.[4] Neither did we find any articles about the McHale modification of the Castle technique,[20] that could meet our quality criteria.

The only study included in our review that used hip replacement reported a relatively low success rate in terms of sitting tolerance (45%) and a rather high complication rate. Dislocation of the prosthesis occurred in 29% of the cases, and HO was present in 45%. In addition, femoral fractures were relatively frequent (36%).

Arthrodesis of the hip joint can result in a pain-free hip, but the rate of complications, such as non-union, is high (42%). In addition, the postoperative immobilization period (2–4 months) can be a serious burden to this vulnerable group of patients. Frequently, the condition of the contralateral hip and the spine prevents successful arthrodesis.[21]

Although less favorable outcomes have been reported for hip replacement and hip arthrodesis, our conclusion must be drawn with some caution, as both techniques were only reported in one article each that met our inclusion and methodological quality criteria. Therefore all we can say is that there is so far insufficient evidence for a positive effect on pain and seating problems.

Regrettably, none of the intervention methods (resection surgery, hip replacement or arthrodesis) provides the ultimate solution to treat the severely deformed painful hip in CP. The published literature does not clearly favor one procedure over the others.

However, the technique for which the largest number of results has been published is subtrochanteric resection, which seems to eventually lead to a good outcome. Bony intervention, however, implies no instant cure: we note that it can take up to three to six months postoperatively before maximum pain relief is obtained.[14,15]

Heterotopic ossification (HO) is a complication that is frequently occurring in patients after hip surgery. In their review, Balboni et al.[22] mentioned two ways to prevent HO after surgery. Of these two, radiation therapy is considered to be superior to nonsteroidal anti-inflammatory drug treatment. Only one of the studies included in our review used prophylactic treatment,[15] (radiation therapy) with good result. Other authors just measured the degree of postoperative HO without using prophylaxis. Muthusamy et al.[14] found less HO in patients who were treated with a spica cast, compared to treatment with an external fixator. The question remains whether HO actually troubles the patient after surgery in the long term.

Dislocation after total hip replacement occurred frequently in the study described by Gabos.[17] The dislocation rate of 29% is quite high. No explanation for this phenomenon can be given within the context of this review.

Unfortunately, the results of our review are weakened by the variation in surgical procedures, the age of the patients at the time of surgery, and the definitions used for the outcome measures. The studies mostly failed to measure the level of pain by instruments that assess the pain expressed by the patients themselves. This is especially necessary for patients with mental retardation, which is common in severe CP. Therefore, conclusions about the effect in terms of pain relief must be drawn with caution. Post surgical management is also very diverse in the various articles, making the result of the interventions even more difficult to compare.

In our algorithm for the surveillance and treatment of hip disorders in patients with bilateral spastic cerebral palsy (see Appendix 6.2), we used the GMFCS classification to indicate the most favorable palliative intervention. Total hip replacement is to be preferred for patients with GMFCS I-III. In patients with GMFCS IV and V, a Castle procedure or Schanz osteotomy is the method of choice.

CONCLUSION

In conclusion, our review suggests that there are research findings indicating a favorable effect on pain and sitting/nursing problems of palliative surgery in patients with severe cerebral palsy and intractable pain and hip problems. The published literature does not clearly favor one procedure over the others. The resection arthoplasty technique developed by Castle is reported to yield the best results and fewer complications, and seems to eventually lead to a good outcome.

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APPENDIX 6.1

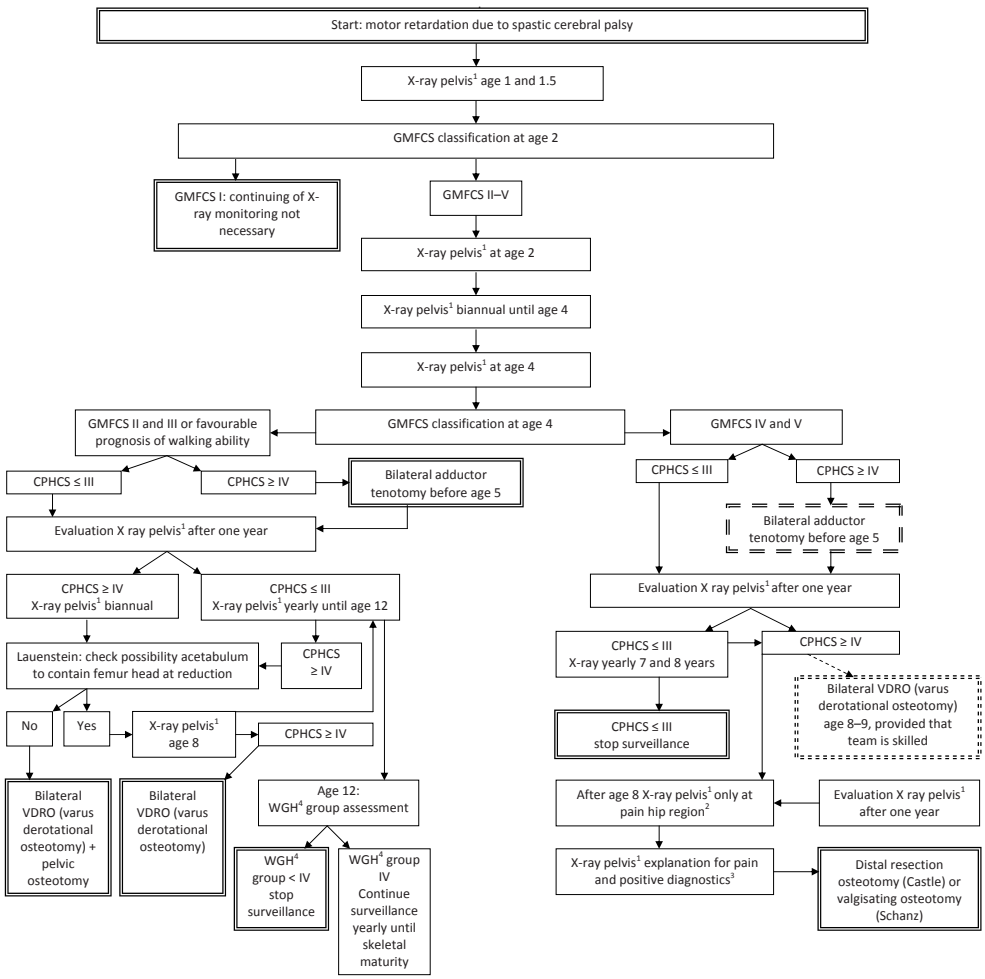
Best evidence synthesis

Strong evidence	Provided by consistent, statistically significant findings on outcome measures in at least two high-quality RCTs
Moderate evidence	Provided by consistent, statistically significant findings on outcome measures in at least one high-quality RCT or high-quality CCT
Limited evidence	Provided by consistent, statistically significant findings on outcome measures in at least one high-quality RCT or Provided by consistent, statistically significant findings on outcome measures in at least one high-quality CCT
Indicative findings	Provided by consistent, statistically significant findings on outcome and/or process measures in at least one high-quality CCT or low-quality RCT (in the absence of high-quality RCTs) or Provided by consistent, statistically significant findings on outcome and/or process measures in at least two ODs with sufficient quality (in the absence of RCTs and CCTs)
No or insufficient evidence	If results of eligible studies do not meet the criteria for one of the above levels of evidence or In case of conflicting (statistically significant positive and statistically significant negative) results among the RCTs and CCTs or In case no eligible studies are available

RCT = randomized controlled trials; CCT = controlled clinical trials; OD = other design than controlled.

APPENDIX 6.2

A decision tree for monitoring hip disorders in spastic cerebral palsy and its consequences for intervention



[1] X-ray pelvis: anterior-posterior. Evaluation Shenton´s line and Migration Percentage (MP). Lauenstein X-ray only when planning adductor tenotomy and/or VDRO. CPHCS: CP Hip Classification System (Robin J, Kerr Graham H, et al. A classification system for hip disease in cerebral palsy. Dev Med Child Neurol 2009;51:183-92).
[2] Pain hip region: to be diagnosed on X ray pelvis and with PAICP (Pain Assessment Instrument for CP).
[3] Diagnostics: Pain really in hip region? Muscles? Temporarily stop physiotherapy; analgetics; Botulinum Toxin; wait and see (1/2 year); spasticity treatment.
[4] WGH: Winters, Gage and Hicks hemiplegia classification. == cadre: intervention doubtful, see this thesis.

SYNTHESIS: SURVEILLANCE AND INTERVENTION RECOMMENDATIONS

Chapter 7

Monitoring hip disorders in spastic cerebral palsy and its consequences for intervention; proposal to a surveillance and intervention algorithm

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To be submitted.

ABSTRACT

Hip disorder is one of the most frequent complications in patients with cerebral palsy. The risk of this disorder is associated with the severity of the patient's neurological condition and therefore with their functional level. Since it is generally assumed that hip disorder may lead to certain health risks, early monitoring of the hips by radiological examination of the pelvis is important to enable well-timed intervention.

There is no consensus in the literature about the age at which and the frequency with which such examinations should take place. This paper presents a surveillance protocol based on a review of the literature and on our own research and clinical experience. The protocol links the results of the examination to the required intervention. The interventions are based on a systematic review of the literature.

INTRODUCTION

Disorder of the hip is one of the most frequent complications in patients with cerebral palsy. The incidence of the disorder is associated with neurological severity and therefore with the functional level. It is especially the patients with GMFCS levels IV and V who are at risk for hip disorder.[1-3] Since it is generally assumed that hip disorder may lead to certain health risks, early intervention is important, so early and regular radiological examination of the pelvis is required.

There is no consensus in the literature about the age, frequency and nature of the examination,[4-6] nor about the proper intervention to be applied when specific radiological results are obtained. This paper presents a protocol based on a review of the literature as well as our own research and clinical experience.

SUBLUXATION OR DISLOCATION OF THE HIP IN CEREBRAL PALSY

Unfortunately, the definitions of subluxation and dislocation vary widely in the literature. The common measure of subluxation is the migration percentage defined by Reimers (MP), which is measured on a standard anteroposterior radiograph of the pelvis[7] (Figure 7.1). The term subluxation is used when the femoral head is partly out of the acetabulum but is still in contact with it.[8] Authors use different migration percentages, ranging from 30%[9] to 33%,[7] as a cut-off value for subluxation.

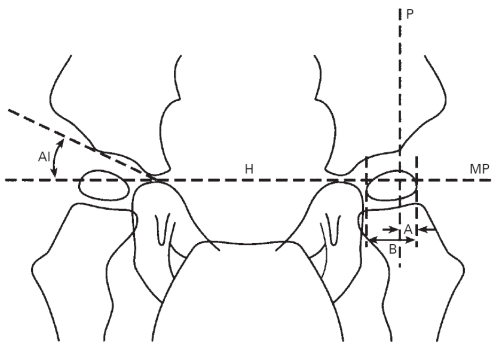

Figure 7.1 Measurement of Migration Percentage (a/b x 100%). H = Hilgenreiner's line; P = Perkins' line; AI = Acetabulum index. (Wynter M, Gibson N, Kentish M, et al. The consensus statement on hip surveillance for children with cerebral palsy: Australian standards of care. J Pediatr Rehabil Med 2011;4:183-95.)

In dislocation there is loss of contact between the femoral head and the acetabulum.[10] However, some authors use a specific migration percentage (70–80%) to discriminate between subluxation and dislocation.[11,12]

RISK FACTORS

Individuals with severe spastic cerebral palsy who cannot walk because of total body involvement (GMFCS levels IV and V[2,3]) are most vulnerable to developing displacement of the hip. Other factors associated with displacement include the persistence of neonatal reflexes,[13] causing asymmetry and windblown hip syndrome.[14] Some authors have reported that dystonic individuals are at greater risk for displacement.[15] Theoretically, the absence of axial weight bearing may be another risk factor, but no evidence for this view has been found.[16] Although pelvic obliquity and scoliosis play an important role in the development of hip displacement, it is not clear whether they are a cause or a result of displacement, or merely associated with it.[10,17]

RADIOGRAPHIC INVESTIGATION

The only reliable way to establish the diagnosis of hip disorder is through radiographic investigation of the pelvis.[4] Physical examination (degree of hip abduction) cannot totally replace the radiographic investigation, as it only provides some indication.[15] No straightforward relation has been found between physical examination and radiographic findings.[3] In a radiological study of 123 CP hips, Reimers[7] found only a marginal association between clinical hip abduction and the corresponding migration percentage. A hip could very well be subluxated even with 60° abduction, though when the abduction was less than 30°, the hip "must have migrated beyond the acetabular rim".

In an earlier study, we found the mean MP to be significantly higher in patients with hip adduction contractures as compared to those without contractures. The prevalence of hip adduction contractures increased as the level of hip deformity increased.[18]

Monitoring the development of hip displacement requires repeated radiological investigation of the pelvis.

The **migration percentage** (MP, Figure 7.1) was defined by Reimers[7] as the percentage of the femoral head projecting outside Perkins' line. This measure can be used in children and adults aged 12 months or over. The measure indicates the position of the ossific nucleus of the capital femoral epiphysis, in contrast to the CE angle of Wiberg, which is based on the position of the center of the cartilaginous femoral head. The intra- and inter-observer reliability of the MP method is good, especially when the same measurer performs the measurement over time.[19-21]

The MP varies with abduction and adduction of the femur but is not significantly affected by the degree of internal or external rotation of the femur.[22] Most authors use a view in anteroposterior direction with the hips in an anatomically neutral position and the knees hanging down.[23] This carries the risk of a radiograph with the pelvis tilted forward, thus misrepresenting the configuration of the relation between femoral head and acetabulum. Correct positioning of the patient (Figure 7.2) is therefore important in measuring the MP in each radiograph.[24] A standard measuring error of 10% has been found.[7]

Many authors have claimed that a radiograph according to **Lauenstein**, i.e. an anteroposterior radiograph of the pelvis with maximal abduction of both hips, should be part of the routine investigation of the hip. In this maneuver, a subluxating hip can

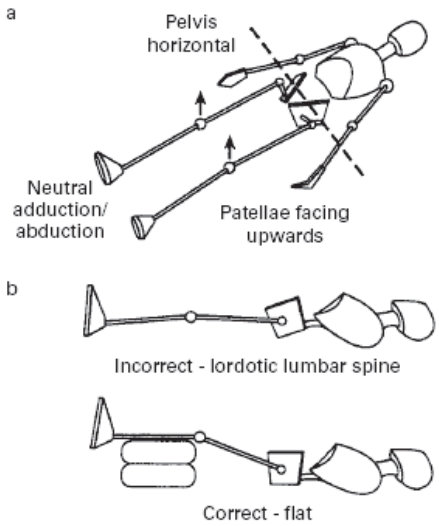

Figure 7.2 Correct positioning of patient in pelvic X-ray. (Wynter M, Gibson N, Kentish M, et al. The consensus statement on hip surveillance for children with cerebral palsy: Australian standards of care. J Pediatr Rehabil Med 2011;4:183-95.)

be redressed in the acetabulum while fully abducted. This information is important only when surgery of the acetabulum is considered. The radiograph has no predictive value for the rate of success of adductor tenotomy, and should therefore not be part of the routine radiological investigation. A Lauenstein radiograph can be useful in predicting the ability of the femoral head to be reduced in the acetabulum after a proximal femoral varus and derotation osteotomy (VDRO).

Another frequently used measure of displacement of the hip, mainly used in congenital hip dysplasia, is the **Centre Edge (CE) angle** or angle of Wiberg.[25] The CE angle cannot be used in children under the age of 5, however, because of the difficulty of determining the center of the femoral head in childhood.[23,26] In view of these restrictions, the CE angle should not be used for children with CP.

Shenton's line is an arc drawn on the radiograph from the trochanter minor along the medial inferior margin of the femoral head to the superior medial border of the obturator foramen. It indicates the position of the femoral head in relation to the acetabulum. A broken line expresses a proximal displacement of the femoral head (occurring at a displacement of at least 8 mm).[7,27] This situation indicates subluxation.[28]According to Vidal, a broken line does not have a predictive value until the MP is 20%.[27]

The **acetabular index** or acetabular angle is a measure of acetabular dysplasia[29] (Figure 7.1), i.e. the angle between the horizontal line of Hilgenreiner and a line between the medial and lateral borders of the acetabulum. Though the intra-rater reliability is good,[20] this measure can only be used in children under 12 years of age, in whom the ossification of the epiphysis of the pelvis has not yet been completed.[23] The acetabular angle increases only after the age of 30 months and at a migration percentage of at least 20%.[27] By comparison, the migration percentage can be measured from the age of 12 months, and it increases before the acetabular angle does.[27,30] The migration percentage should therefore be preferred to the acetabular angle.

Femoral anteversion is represented by the intersection of the coronal plane passing through the femoral condyles and the oblique plane passing through the femoral neck. Ruwe described a clinical measurement that proved to be more accurate than computed tomographic scanning and Magilligan radiological determination. The method has a low inter-observer variability and can be quickly performed.[31] However, the increase in femoral anteversion is mainly important for the estimation the effect of a VDRO

and not for the – surgical – management of subluxation or dislocation. The femoral neck anteversion does not diminish after birth as in normal developing children and therefore is increased in patients of all GMFCS levels.[32]

The **neck-shaft angle (NSA)** (Figure 7.3) is the smallest angle between the femoral neck and shaft, which can be demonstrated radiologically by rotation of the femur.[33] Other names for this measure are inclination angle and CCD (caput-collum-diaphyseal) angle. It is clear that rotation influences the value of the angle, making standardized measurement difficult.[34] The angle further depends on anteversion of the femoral shaft, which cannot be measured on a plain radiograph of the pelvis. Many authors agree that there is considerable individual variation[23] and a large standard deviation in patients with cerebral palsy.[35] The NSA correlates with the GMFCS level and the shape of the proximal femoral epiphysis and is to be preferred to the Head Shaft Angle (HSA).[36] In cerebral palsy, the neck shaft angle increases with GMFCS level (Figure 7.3).[32] Authors explain this phenomenon by lack of weight bearing in patients with high GMFCS levels.

In Australia, a combination of MP, AI, NSA and a new measure, **FEHLA** (Femoral Ephiphysis to Hilgenreiners Line Angle) was proved to be the best predictor of later hip displacement in preschool age children with CP.[37] An angle of less than 11° is found to be predictive of displacement in the future.

Recently, a new classification system for cerebral palsy hip status has been developed by the Australian group led by Robin. The Cerebral Palsy Hip Classification System

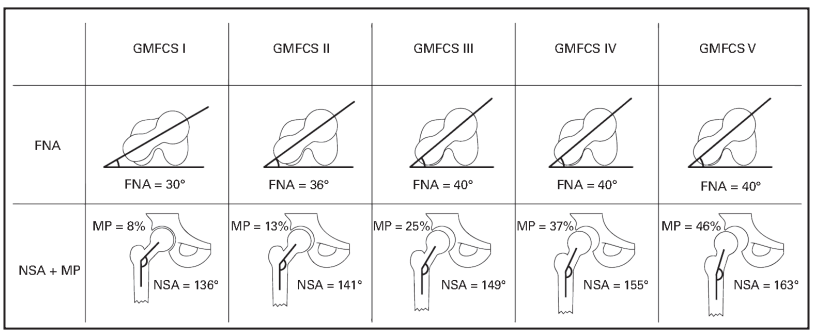

Figure 7.3 Relationship between GMFCS level and Neck Shaft Angle (Robin J, Dobson F, Baker R, Selber P, Kerr Graham H. Proximal femoral geometry in cerebral palsy: a population-based cross sectional study. J Bone Joint Surg [Br] 2008;90(B):1372-9.). FNA = femoral neck anteversion; NSA = neck shaft angle; MP = migration percentage.

(CPHCS) combines elements of Shenton's arc, the shape of the femoral head, the configuration of the acetabulum and pelvic obliquity. This confirms the intimate relationship between these elements. The classification contains quantitative (MP)

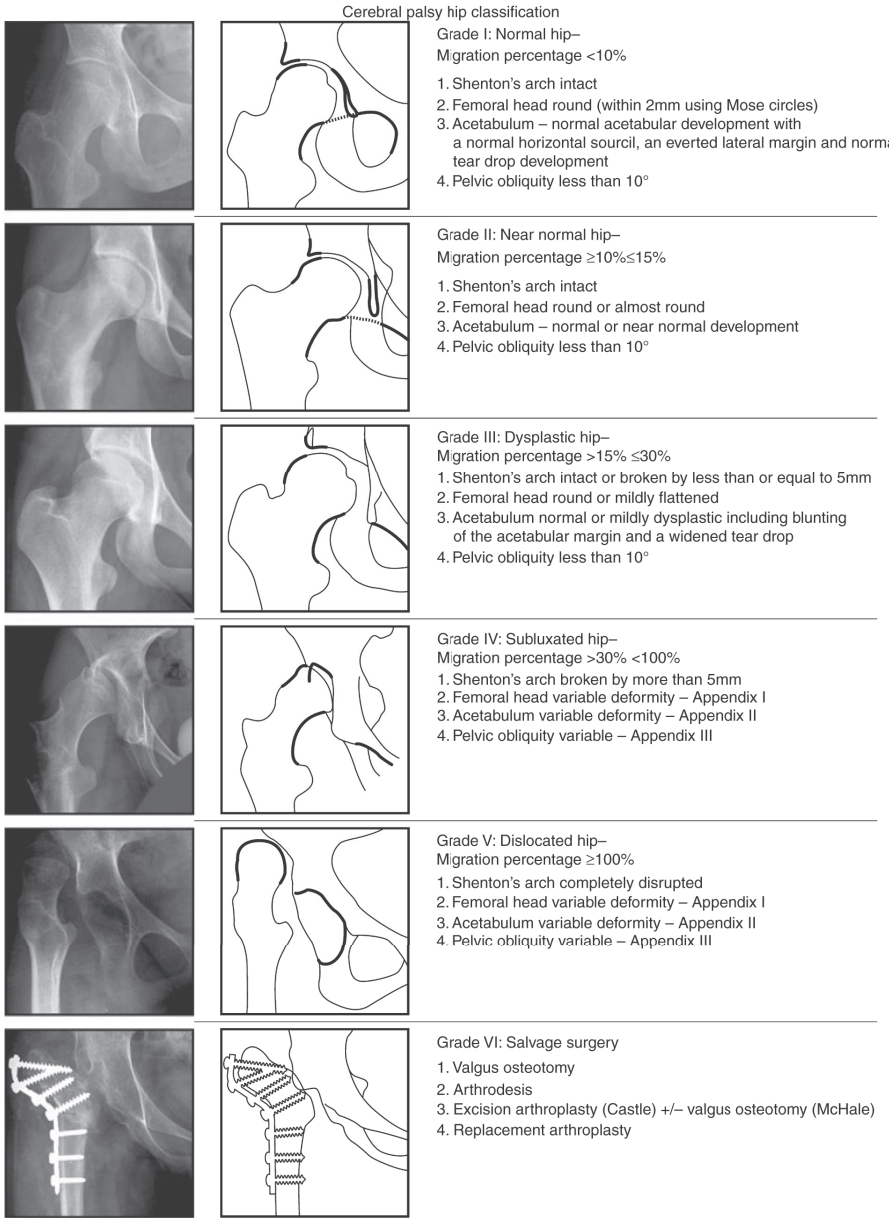

Figure 7.4 The CPHCS classification system (Robin J, Kerr Graham H, Baker R, Selber P, Simpson P, Symons S, Thomason P. A classification system for hip disease in cerebral palsy. Dev Med Child Neurol 2009;51:183-92).

and qualitative measures. There are five levels; the sixth level describes the situation after salvage surgery[38] (Figure 7.4). The advantage of this instrument is that it is simple and easy to use. Many researchers are using it, and the instrument is rapidly reaching the status of the new state of the art in the literature. In a recent study, the inter- and intra-observer reliability was found to be excellent, ranging from 0.88 to 0.94.[39] The reliability of the CPHCS system was also confirmed by three-dimensional computed tomography.[40]

PATHOPHYSIOLOGY OF HIP DISPLACEMENT

The natural history of disorder of the hip in spastic quadriplegic individuals is not yet fully clear. The skeleton of children with cerebral palsy is known to be normal at birth.[7,30] In normal children, subluxation of the hip does not occur, as migration is less than 1% per year.[7] In severe cerebral palsy, there is a dynamic imbalance of the muscles of the hip, which causes shortening of muscles and deformation of the originally normal cartilage and bone. The physiological degree of anteversion does not decrease as it does in normal children, but persists due to muscle imbalance and delayed weight bearing.[30] The spastic adductors and flexors of the hip, including the iliopsoas muscle, force the femoral head in a superior lateral direction. Dysplasia of the acetabulum and deformity of the femoral head occur simultaneously.[28] Acetabular dysplasia is seen only after subluxation or dislocation of the hip has occurred.[30] This resembles the acetabular dysplasia seen in the congenital dysplastic hip,[34] but the acetabulum in CP is shallower and shows more posterior deficiency.[41] Finally, degenerative arthritis can occur in association with subluxation or dislocation.[12] Histologically, inflammatory factors cause sensitization of the nociceptors in the cartilage of femoral head and pelvis,[42] which explains the patients' pain experience.

NATURAL HISTORY OF MIGRATION OF THE HIP

Migration of the hip in CP can occur very early in life, within the first two years.[43] The migration percentage increases from the age of twelve months, but the acetabular angle does not increase until after the age of 30 months.[27] The migration rate is related to the neurological status and the ambulation level.[2,7] Quadriplegic individuals who

are unable to walk can have a migration of up to 9.5% a year,[7,44] whereas the hips of independent walkers can migrate by 4% a year.[27] It takes an average of five years for a hip to dislocate completely.[45] At migration percentages of 50% or more, all hips will eventually dislocate if no intervention is performed.[9,46,47]

HIP DISORDER AND COMPLICATIONS

It is common opinion that untreated hip disorder in children with spastic cerebral palsy can lead to pain and problems with nursing care, impaired sitting, decubitus ulcers and fractures. In an earlier study, we confirmed the relationship between subluxation and deformity of the femoral head and pain in adults with severe cerebral palsy.[48] The risk of pain associated with hip disorder was about 30% in our research sample.

Pain associated with subluxation is just one of the inconveniences people with severe CP have to bear.[49-51] For instance, physiotherapy of the legs proved to be quite painful in our study group, confirming the findings of a study by Hadden.[52,53] In the same sample, both migration and deformity were positively related to the need for a special seat in the wheelchair, and adduction contractures of the hip.[18]

We conclude that migration and concomitant deformity of the femoral head in patients with severe cerebral palsy should, if possible, be prevented. However, treating an imminent or present subluxation in patients who are unable to walk (GMFCS IV and V) by means of bony intervention, especially when there are no symptoms of pain or other impairments, is still very controversial. Careful consideration is essential to assure that the burden of the intervention does not outweigh its benefits.

INTERVENTION

Many interventions have been recommended to prevent subluxation in patients with cerebral palsy. Conservative options include physiotherapy, special mattresses and standing devices. Although these have been claimed to prevent dislocation,[47,54] this has never been proved beyond doubt.[16,55,56]

Soft tissue surgery

Bilateral tenotomy of the adductors[57] is a very widely used preventive intervention to stop imminent subluxation as seen on a pelvic X-ray. The aim of adductor tenotomy in CP is to remodel the femoral head / acetabulum configuration. Remodeling of the acetabulum is possible when reduction of the hip is performed at an early age, i.e. before the age of four years.[58-60] Hence, it is common clinical practice to perform adductor tenotomy before the fourth[7] and not later than the fifth year of age.[61] A migration percentage of 30–40% or more is used as a threshold indication for adductor tenotomy.[62] Unfortunately, the effect of this intervention has never been proved.

Systematic review

In a systematic review of the value of preventive adductor tenotomy, we found only one article (Bowen 2006) that met our quality criteria.[63] Bowen found that 87% of the hips with a pre-operative MP<50% were stable at follow-up, versus 43% with a pre-operative MP >50% (p=0.031). The average patient age at surgery was 5.9 years for stable hips versus 3.8 years for unstable hips (p=0.001). Seventeen percent of all hips were unstable at follow-up and needed subsequent surgery consisting of femoral and/or pelvic osteotomy.[64]

Recently, a survey among 330 patients revealed that the success rate of adductor tenotomy is inversely correlated with the GMFCS level: the higher the level, the lower the success.[65] The success rate in patients with GMFCS level II was 94%, that in GMFCS V only 14%. Shore et al.[65] discuss the role of the adductor muscles in the etiology of hip disorders. The poor result of adductor tenotomy in patients with a high GMFCS level suggests that a different factor, for instance the lack of weight bearing plays a more important role then we thought until now. In addition, the m. gluteus medius in these patients may be too weak to alter the configuration of femoral head and acetabulum after reducing the force of the adductor muscles.

Clinical experience

Clinical experience, not confirmed by our systematic research, is that adductor tenotomy, if performed, is best done symmetrically. Unilateral adductor tenotomy, especially if performed before the age of nine years, carries a high risk of causing subluxation in the contralateral hip.[57,66,67] Postoperative bracing with the hips in

abduction is recommended.[68] Full neurectomy of the obturator nerve is discouraged, as it leaves the patient with an abduction contracture in many cases. Overcorrection is avoided by preserving the adductor brevis and by performing only a partial neurectomy of the medial branches of the anterior obturator nerve.[69] An adductor tenotomy cannot establish a reduction of a dislocated hip in older children.[57,61] Additionally, when the MP is greater than 50% soft tissue procedures alone will not prevent dislocation of the spastic hip.[7,13,61]

Bony surgery

When adductor tenotomy fails, the next possible step to reduce the femoral head in the acetabulum is a femoral varus and derotational osteotomy (VDRO). This operation must sometimes be combined with pelvic osteotomy to create a proper acetabulum and establish good containment of the femoral head in the acetabulum.

Systematic review

In a systematic review we found that VDRO is frequently a successful intervention to reduce the luxating femoral head in the acetabulum.[63] The success rate, expressed in reduction of the MP, is 53%–75% for VDRO alone and 85% or higher for interventions in which pelvic osteotomy is added to the VDRO. Unfortunately, the complication rate for interventions in which pelvic osteotomy is added is quite high: up to 28%, with osteoarthritis being the main complication. The complication rate of VDRO alone is up to 15%, mainly consisting of heterotopic ossification, fractures and ulcers.[63] However, our best-evidence synthesis could not establish sufficient evidence for a positive effect of VDRO/pelvic osteotomy to reduce pain after several years of follow-up.[63]

Should pelvic osteotomy be added to VDRO?

The question whether pelvic osteotomy should be added to VDRO is a controversial one. Many orthopedic surgeons make their decision peroperatively, based on judging on intuition and experience whether pelvic osteotomy is needed to provide good containment of the femoral head. Song and Carroll recommended that patients with MP >70% should undergo concomitant acetabular procedures.[70] Recently a study appeared in which a more objective means was described to assess the shape of the acetabulum preoperatively by three-dimensional CT scan.[71]

Other authors recommend a sequence of first performing VDRO and soft tissue release, assessing the reduction using fluoroscopy and arthrography during the operation and proceeding with open reduction and/or pelvic osteotomy if reduction and/or femoral head coverage are inadequate.[72]

Another question that remains to be answered is whether the cartilage of the femoral head and acetabulum is still healthy enough to obtain a pain-free joint after the femoral has been reduced in the acetabulum. Shore[73] advocated delayed gadolinium-enhanced magnetic resonance imaging of cartilage (dGEMRIC). This technique has been shown to identify early signs of hip osteoarthritis. The results correlate well with clinical symptoms of hip pain.[73]

Timing of bony surgery

The age at which VDRO should be performed is also controversial. Some surgeons advocate early bony surgery in children whose hip is dislocating, as an addition to adductor tenotomy, just to ensure good containment of the femoral head in the acetabulum.[74] Khalife et al. recommended operating on subluxated hips at an early stage, before Reimer's MP reaches 70% and preferably before the age of eight years, in order to decrease the risk of avascular necrosis.[75] Others point out that the younger the child, the greater the risk of incorrectly remodeling the femoral head / acetabulum configuration, due to a recurrent disequilibrium of the muscles around the hip.

On the other hand, the risk that pelvic osteotomy will have to be added to VDRO increases with age: the older the child, the greater the risk that the acetabulum is so deformed that it can no longer contain the femoral head adequately. One study showed that the increase in neck shaft angle (NSA) will be sustained if derotation osteotomy is performed after the age of eight.[76]

At the Wilmington conference on hip disorders in CP (2010), consensus was reached on a preferred age of between eight and nine years for VDRO in patients in whom adductor tenotomy has failed to reduce the femur. Careful monitoring of the development of femur and acetabulum in the years after adductor tenotomy is necessary to determine the right age for surgery (whether or not combined). The additional measurements described above supplied by regular Lauenstein X-ray, will be useful to assess the situation.

Surgery: unilateral or bilateral?

Another controversy exists about the question whether VDRO should always be performed bilaterally or only on the affected side. The probability of instability of the untreated contralateral hip has been reported to be about 44%.[77]

One author has advocated that osteotomy, if performed unilaterally, should be done after the ninth year, when the skeleton has sufficiently matured to reduce the risk of subluxation of the contralateral hip.[57] A recent study reported that VDRO should always be performed bilaterally when the unstable hip rate of the contralateral stable hip during observation is 27% or over.[78]

ALWAYS PREVENT OR CURE HIP DISORDERS?

The indication for intervention depends on the patient's GMFCS classification. Though not always successful, soft tissue surgery is still common practice for all GMFCS levels, once imminent subluxation is diagnosed by X-ray. The preferred age is around four years. Since adductor tenotomy is a relatively minor surgical intervention, most authors recommend this operation to prevent the hip from dislocating, even in children with a poor locomotor prognosis.[13] However, the relatively poor results of adductor tenotomy in this category should be thoroughly discussed with the parents.

If soft tissue surgery fails in walking children (GMFCS I–III) who are at risk for dislocation, osteotomies should always be performed, preferably at age eight, to preserve the walking potential.

However, radical femoral or acetabular bony interventions remain controversial in those children who are severely spastic, neurologically immature and have a poor locomotor prognosis (GMFCS IV–V). Some authors always recommend VDRO, others are more conservative as regards non-walkers.[79,80] One must keep in mind that bony surgery has a large impact on the vulnerable patients with GMFCS levels IV and V, who are frequently bedridden and not very well able to express themselves due to communication problems (85%) and / or intellectual disability (60%).[111] The simple fact that an X-ray shows progressive subluxation or dislocation of the hip after a failed adductor tenotomy does not justify major surgery like VDRO, whether or not combined with pelvic osteotomy. As mentioned above, the complication rate of VDRO is quite high, and increases further when pelvic osteotomy is added to the procedure.[63]

We therefore recommend a wait-and-see policy, provided that the patient has no pain or serious problems regarding aspects like nursing or sitting.

SALVAGE SURGERY

The opponents of bony interventions in non-walkers point out that proximal femoral resection in a painful dislocated hip in adults is very often successful.[81,82] They advocate the technique used by Castle and Schneider,[83] which has later been modified by others.[84] The Castle technique was modified by McCarthy et al. to include interposition involving suturing the iliopsoas and gluteal muscles to the hip capsule ("closing" the acetabulum) and covering the femoral stump by suturing the lateral vastus muscle to the muscles and soft tissues on the medial side.[85]

Systematic review

In our recent systematic review,[86] we concluded that, regrettably, none of the intervention methods (resection surgery, hip replacement or arthrodesis) provides the ultimate solution to treat the severely deformed painful hip in CP. The published literature does not clearly favor one procedure over the others. However, the technique for which the largest number of results has been published is subtrochanteric resection, which seems to eventually lead to a good outcome.

Bony intervention, however, implies no instant cure: we note that it can take up to three to six months postoperatively before maximum pain relief is obtained.

If resection surgery is chosen, the Castle procedure is to be preferred to the McCarthy procedure in these cases. The Castle procedure has a higher success rate in terms of pain (90–100% versus 53–77%), a higher rate of improvement in terms of sitting problems (100% versus 92%), and a very low rate of proximal migration of the femur (5% versus 17–28%). In addition, the rate of postoperative heterotopic ossification (HO) is lower in patients treated with the Castle technique (with reported values of 0–57% and low grades, versus 30–62%). The careful resection of the periosteum described by McCarthy to prevent HO may have a counterproductive effect. Heterotopic ossification is a frequent complication after bony surgery of the hip. It can be reduced by applying postoperative radiation therapy.

Clinical experience

Valgus osteotomy of the femur, as described by Schanz,[87] is advocated by several orthopedic surgeons. This technique was originally described for patients with congenital dislocation of the hip. We found only a few articles on the use of this technique in the literature, and none of them met our quality selection criteria. Schejbalova et al.[88] reported 98% pain relief in their patient group, but Hogan et al.[89] reported a 63% complication rate. One of the main problems is persistent pain and postoperative problems with pressure ulcers from the retained femoral head.

McHale described a variant in which femoral head resection is combined with valgus osteotomy.[90] There is disagreement in the literature about the complication rate in this procedure. Leet et al. reported a lower rate of complications than with the Castle procedure.[91] By contrast, Van Riet et al. reported a high complication rate, including heterotopic ossification, residual pain caused by hardware, fracture and pulmonary complications, for which a second procedure was necessary in several cases.[92] Some surgeons prefer a procedure as described by Girdlestone[93] to cure otherwise incurable pain in patients with cerebral palsy. We did not find any articles about the value of this technique that met our quality criteria. In our own research, all of the patients who had undergone proximal resection of the femoral head because of pain still suffered from pain.[48]

BOTULINUM TOXIN

The role of botulinum toxin in preventing subluxation of the hip in cerebral palsy is – again – controversial. Some authors recommend the use of botulinum toxin to prevent subluxation, especially for children below the age of two and with an initial MP of >30%,[94] but a recently published randomized controlled study found no significant effect of botulinum toxin combined with a hip abduction orthosis on migration of the hip, compared with no such treatment.[95] The age of the patient receiving the botulinum toxin seems to be crucial in this matter and is a possible explanation of the failure, as the population in the recent study had an average age of 38 months.

A systematic review from the Cochrane Collaboration did not reveal strong controlled evidence to support the use of botulinum toxin.[96] An explanation of the poor results of this treatment in the long term could be the delay of surgery, thus missing the window of opportunity in which the acetabulum is able to restructure itself after adductor

tenotomy or bony surgery.[47] However, some studies have found decreased pain in the hip region after injection with botulinum toxin.[97] In addition, botulinum toxin injection can be useful as a preoperative measure in total hip surgery.[98] However, botulinum toxin administration often leads to temporary general side-effects, in 23.2% of the cases, especially in children with GMFCS levels IV and V.[99]

SURVEILLANCE OF HIP DISORDERS IN CP

The only secure way to assess whether a hip in a patient with CP is at risk is to make a radiograph of the hips. Other methods are not reliable enough,[4] so X-rays of the hip must be taken frequently. There is, however, no consensus on how frequently a radiographic examination of the pelvis should be performed.[4] Many different recommendations have been made since 1979,[100] and only a few of these are evidence-based.

Scrutton and Baird[4] recommended that all children with bilateral CP who cannot walk more than 30 steps at the age of 30 months should have an X-ray of the pelvis. Depending on the results, they recommended further X-rays every six months for those with severe involvement. They suggested orthopedic referral if the migration percentage is 30% or more and orthopedic intervention before the age of four.

Miller[46] reported on a retrospective study among 45 children with cerebral palsy aged 2–18 years (64 hips) with a migration percentage of 30–75% in at least one hip. In this study, no stratification was made for the degree of spasticity and the ambulation level. The group aged 8–18 years with an MP of 30-60% never showed a rate of progression greater than 20% a year. Since the MP has a standard measuring error of 10%, Miller concluded that a radiograph once a year would be enough in this group. The group of children older than 18 years with the same migration percentage was found to have a progression rate of at most 5% a year, so a radiograph once every two years would be enough. The progression rate in the group of children under eight years of age was comparable to that in the 8–18 years group, probably because only those who did not have dislocated hips were selected. Children who were more severely affected were not included in the study, and no stratification was made for the degree of spasticity or the ambulation level.

Other authors have recommend radiographs at least every year to "avoid being surprised".[15,41,55] Dobson recommended radiological examination of the hips at 18 months of age in all children with bilateral cerebral palsy and at 6–12-monthly intervals

thereafter.[5] Cooke advocated radiographic screening by measuring the so-called acetabular index at the ages of two and four years.[15]

Gordon and Simkiss[6] performed a systematic review of the evidence for hip surveillance in children with CP. They share the advice by Scrutton et al.[101] that radiographic monitoring for all children with bilateral CP should be performed from the age of 30 months. The migration percentage is a more valuable instrument than the acetabular index (angle) and remains useful after the age of eight years. Children with a migration percentage greater than 33% are likely to need further treatment. Progression of the migration percentage by more than 7% a year requires careful monitoring, and orthopedic referral should be considered.[6]

Hägglund et al.[102] described the results of an intervention program for children with CP in Sweden. They reported that none of the hips with a migration percentage of more than 42 returned to normal without operative treatment. They recommended radiological follow-up at 6-month intervals before a decision about surgery is made for hips with an MP between 33 and 40%.

GMFCS

A major breakthrough was achieved by the use of the GMFCS system[103] as the basis for monitoring the development of the hips in CP. After Soo[2] had pointed out that the risk of subluxation increases linearly with the category in the GMFCS classification, this was confirmed by Hägglund.[3] He therefore advocated a surveillance program based on the GMFCS classification. Only the patients in categories III-V should have an X-ray of the pelvis as a monitoring measure, starting at the age of one year, or as soon as the diagnosis has been confirmed. These children should be checked once a year until the age of eight years. For children with GMFCS II, Hägglund estimated the risk to be low enough to recommend radiographic examination at two and six years of age only. Further radiographic examination is needed if the MP exceeds 33%, or if clinical investigation reveals a decreasing hip ROM.

We conclude that X-ray examination including measurement of the MP according to Reimers is sufficient for monitoring. These recommendations have now been incorporated in the CPUP[1] recommended routines in Western Sweden.[3] In a recent

[1] CPUP - Swedish National Health Care Quality Programme for prevention of hip dislocation and severe contractures in Cerebral Palsy

article, Terjesen supports the advice of Hägglund to start making a radiograph in patients with GMFCS level IV and V at the age between one and two.[44]

Australian standard

In Australia, the Australian Standards of Care for Hip Surveillance in Children with Cerebral Palsy were established in 2007 and 2008.[104] A working party of physicians and therapists with national representation produced a draft document, which was discussed at the national CP conference. Finally, specific orthopedic surgeons were asked to complete a detailed survey. The consensus created a guideline for referral to an orthopedic surgeon in specific circumstances. The consensus does not provide a guideline for intervention other than referral to a pediatric orthopedic surgeon. The guideline is based on repeated assessment of the GMFCS level and measuring the migration percentage on a pelvic X-ray. The algorithm also includes clinical assessment of the patient's situation. Patients with GMFCS level I should have repeated X-rays of the pelvis until the age of five.[73] In patients with GMFCS levels I–III, extra attention should be given to walkers with hemiplegia group IV, according to Winters, Gage and Hicks.[105] Their surveillance should be continued until skeletal maturity. Patients with GMFCS levels IV and V should be monitored until the age of seven, and thereafter if there is evidence of pelvic obliquity or scoliosis. For all GMFCS levels, surveillance should start at age 12–24 months. Referral is recommended if the MP exceeds 30%, or if the MP is unstable (changing by 10% a year), or if there is pain in the hip region or if other orthopedic conditions have been identified.[73]

Unfortunately, the Australian consensus is not yet based on the Cerebral Palsy Hip Classification System (CPHCS), but on repeated assessment of GMFCS, MP and clinical findings. As mentioned above, the CPHCS system is a convenient and reliable instrument to describe the state of the hip, as seen on an X-ray.

Hazards of frequent X-ray monitoring

The hazards of taking repeated X-rays of the pelvis are very low. In recent decades, the dose needed for a pelvic radiograph has decreased from about 10 mGy in the 1950s to a few tenths of a milligray today.[106] Gonad protection during the examination is no longer necessary.[106] The impact of an X-ray of the pelvis is between 0.008 and 0.098 mSv. By comparison, a long-distance airplane flight taking 10 hours results in a dose of 0.05 mSv. The risk of developing a fatal malignancy due to radiation exposure for one year old patients is 1.8 per million.[107]

PROPOSAL FOR A SURVEILLANCE AND INTERVENTION ALGORITHM (see Appendix 7.1)

Starting points

In our view, the Australian guidelines present only the first part of the story, while some of its components are debatable.

Surveillance of luxation of the hip in patients with cerebral palsy only makes sense when measures are undertaken to prevent or treat an imminent or established hip disorder. We therefore present a surveillance algorithm that includes guidelines for intervention. Our algorithm was discussed at meetings of the Dutch Association of Pediatric Physiatrists, and consensus was reached. Our flowchart is also based on repeated radiographs of the pelvis, but uses the CPHCS level as a threshold for intervention.

The Dutch consensus statement includes three major interventions once migration above CPHCS level III has been diagnosed on X-ray. These are preventive symmetrical adductor tenotomy around the age of four, corrective derotational varisating osteotomy around the age of eight and palliative intervention in adulthood in case of pain that cannot be relieved by other means (Figure 7.5).

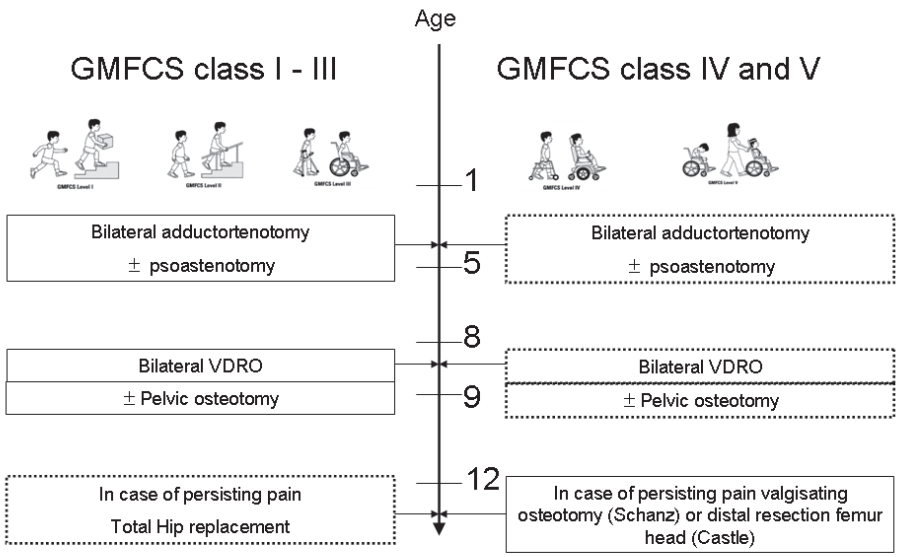

Figure 7.5 Interventions related to GMFCS level and age.

Details of the Dutch consensus and comparison with the Australian guidelines

GMFCS levels I–III

In patients with these levels, a major effort must be made to maintain a proper relationship between femoral head and acetabulum. If the configuration is disturbed, non-disturbed walking is seriously impaired. If migration progresses rapidly before the age of 2 years, treatment of the adductors with botulinum toxin can be helpful according to Pidcock et al.[94] However, their study is based on only a few patients. Additionally, in the age group in patients with an initial radiograph below 24 months, 4 patients showed an improvement of MP after botulinum toxin injection, whereas 7 patients showed a worsening.

If the CPHCS level exceeds III, a symmetrical adductor tenotomy should be performed around the age of four. Adductor tenotomy in this group of patients is a rather successful intervention, as recent research has shown. The success rate is 94% for patients with GMFCS level II, and 49% for level III.[65]

Monitoring of the pelvis should start at the age of one and be repeated every six months. This allows the rate of progression of the migration to be monitored and offers the opportunity to prepare the parents for a possible intervention. In the case of rapid progression, adductor tenotomy can be performed at an earlier age than four years.

The incidence of migration at a very young age is not extremely low. Hägglund[3] found that 4% of the patients below the age of two had an MP above 33.

If adductor tenotomy is not successful, derotational varisating osteotomy is recommended around the age of eight years. Careful monitoring of the femoral head / pelvic configuration is needed to time the moment of surgery. A Lauenstein radiograph should be added to the regular anteroposterior X-ray to assess whether the acetabulum can contain the femoral head after bony surgery.

If the intervention is performed at too early an age, there is a higher risk of recurrent incongruity between femur and acetabulum, because of the remaining capacity of the bones to deform under the influence of the spastic muscles. If VDRO is performed too late, the necessity of adding pelvic osteotomy increases. As mentioned earlier, Khalife et al. recommended operating on subluxated hips at an early stage, before Reimer's MP reaches 70%, in order to decrease the risk of avascular necrosis.[75]

The Australian consensus statement advocates to continue making radiographs in patients with GMFCS level I until the age of five. The Dutch consensus does not support this advice, as hip displacement in patients with GMFCS I is very rare. In addition, a review of five years of surveillance in Australia by Kentish (2011) found an MP exceeding 30 in only 3% of patients with GMFCS I (n=10). In nine of these ten patients, the MP spontaneously reduced to below 30.[108] The child starting to walk and thus introducing weight bearing seems to be a favoring factor in reducing the migration to normal.[109] According to the Dutch consensus, X-ray monitoring can be discontinued when the first radiograph of a child with GMFCS I shows an intact hip.

According to the Dutch consensus, the situation of the hip in patients with GMFCS II and III should be monitored carefully between the ages of four and eight (after a possible adductor tenotomy), to assess the emergence or increase of migration and evaluate the result of the tenotomy performed, and to decide about VDRO, whether or not combined with pelvic osteotomy. The Australian consensus statement only indicates a review X-ray at the age of eight, provided the MP is stable.

In accordance with the Australian consensus statement, walkers in Winters, Gage and Hicks hemiplegia group IV should remain under surveillance until skeletal maturity.

GMFCS levels IV and V

Before the age of four, the Dutch guideline for these levels is the same as for GMFCS levels II–III. Adductor tenotomy is recommended in patients with CPHCS above III, even though the success rate is only 27% in patients with GMFCS level IV and 14% for GMFCS level V.[65] In our clinical practice, we discuss the chances of success with the parents and point out that the intervention poses an acceptable burden for most parents and children, being a soft tissue intervention only.

Some parents take the risk of not having their child operated, given the low success rate of soft tissue surgery and information about chances of pain incidence and possibilities of intervention. The risk of pain related to hip disorder in adults is about 30%.[48] A salvage procedure in adolescence or adulthood, in case of intractable pain, relieves the pain in about 90% of cases.[86] This is why the decision tree (and Figure 7.5) shows adductor tenotomy in patients with GMFCS IV and V in a dotted box.

If soft tissue intervention fails and/or migration progresses, we discuss with the parents the possibility of bony surgery (in this case VDRO with or without pelvic osteotomy). It is our experience that bony surgery in this vulnerable group of patients has a large impact. There are indications that bony hip surgery is a procedure more painful than spine fusion.[110]

In addition, bony surgery in this category of patients can only be successful if performed by an experienced surgeon, supported by an experienced rehabilitation team in a sophisticated setting. If bony surgery fails, the risk of persistent pain is high, as we found in a prevalence study in 160 patients with spastic cerebral palsy level GMFCS V. 12 out of 17 patients suffered from pain due to failed bony surgery.[48] Postoperative care is crucial, including expertise in pain management, communication techniques for non-verbal patients etc.

After the age of eight, when corrective osteotomy would include additional pelvic osteotomy, monitoring by X-ray should be discontinued. The chance that a persistent hip luxation or dislocation will cause no pain or other impairments is about 70%, as we found in a study among 160 patients with GMFCS V.[18,48] If the hip disorder causes complaints after the age of ten, an X-ray of the pelvis can be made to assess the relationship with the pain or impairments. If there is such a relationship, a palliative intervention can be performed, such as Schanz osteotomy or distal resection according to Castle.

We do not agree with the Australian advice to continue monitoring patients with GMFCS IV and V after the age of eight until skeletal maturity. We can envisage no therapeutic consequences of the radiographic findings. When complaints occur, a diagnostic X-ray will be helpful to make decisions about the best way to resolve the problems. In addition, a study by Kentish of 212 cases in an Australian cohort found that only six patients (= 2.8%) with levels GMFCS IV and V developed hip displacement after the age of 12.[108]

We do not support the Australian advice to continue monitoring if pelvic obliquity or scoliosis is present. Once again, the therapeutic consequences are not clear, and no relationship between scoliosis and hip displacement has been proved.[10] Porter et al.[17] found an association between scoliosis and windswept deformity of the hips, but their study was a prevalence study and does not provide evidence for the temporal sequence of events.

CONCLUSION

In conclusion, we recommend a pelvic X-ray for every child with an impairment who does not walk or has a poor walking prognosis at the age of one. We distinguish three phases of monitoring and treatment. The first concerns patients aged between one and four years, in whom adductor tenotomy is the treatment of choice when a hip displacement is diagnosed. The second concerns patients with a hip disorder aged between eight and nine years, which can be treated with VDRO, whether or not combined with pelvic osteotomy. The third concerns the period after the age of nine years, in which palliative surgery can be used to treat patients with pain or impairments that cannot be cured by other means.

We use the CPHCS system to decide whether an intervention is needed; we think that adductor tenotomy is useful at every GMFCS level, provided the reason for and success rate of the intervention are explained to the parents. Bony intervention should be reserved for patients with GMFCS level I–III, and only used for patients with higher levels of GMFCS in exceptional cases.

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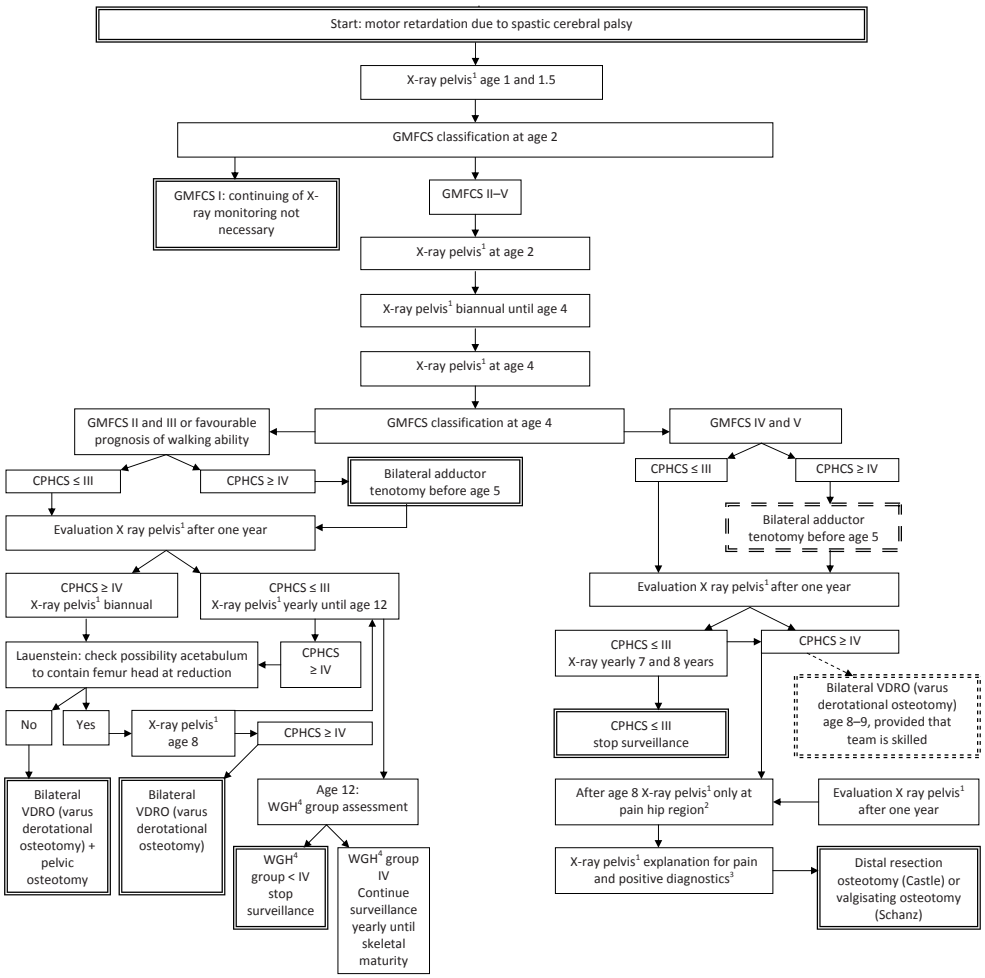
APPENDIX 7.1

A decision tree for monitoring hip disorders in spastic cerebral palsy and its consequences for intervention



[1] X-ray pelvis: anterior-posterior. Evaluation Shenton´s line and Migration Percentage (MP). Lauenstein X-ray only when planning adductor tenotomy and/or VDRO. CPHCS: CP Hip Classification System (Robin J, Kerr Graham H, et al. A classification system for hip disease in cerebral palsy. Dev Med Child Neurol 2009;51:183-92).
[2] Pain hip region: to be diagnosed on X ray pelvis and with PAICP (Pain Assessment Instrument for CP).
[3] Diagnostics: Pain really in hip region? Muscles? Temporarily stop physiotherapy; analgetics; Botulinum Toxin; wait and see (1/2 year); spasticity treatment.
[4] WGH: Winters, Gage and Hicks hemiplegia classification. == cadre: intervention doubtful, see this thesis.

Chapter 8

General discussion

"Maybe it does bother them, but we don't know."

Do we know now?

And how can we apply the things we have learned to a particular case?

This is the story of William S. He was born in June 2009, after 26 weeks of pregnancy. Soon after his birth, many problems emerged. He was diagnosed as suffering from cerebral palsy, GMFCS classification V. There was periventricular leucomalacia, and he had cortically impaired vision. There was central hypotonia of the trunk and extension adduction spasm of the legs. The hips could be passively abducted by about 30 degrees, with a catch on abduction. There were no problems with nursing, changing diapers etc, nor symptoms of pain. He was unable to speak.

We made an X-ray of his pelvis biannually, starting from his first birthday. The pelvic X-ray made in October 2011 showed a slightly disrupted Shenton's line on the right side, while on the left side, we saw an intact Shenton's arc. There was no migration on the left side, but on the right side, he had a migration percentage (MP) of 30%. The neck shaft angle was 170 degrees on both sides.

We repeated the X-ray in May 2012 (Figure 8.1), and found that Shenton's line had disrupted further, while migration had increased to 60%. The situation was alarming. His mother S. visited my outpatient clinic recently to discuss the X-ray and to hear my advice about the next step in his treatment.

What should my advice be? Surgery? At what age? What kind of surgery? Adductor tenotomy? Bony surgery already?

What have we learned from the research described in this thesis, and what are the limitations of our research findings? That is what this chapter discusses.

In **chapter 2**, we have described the development of an instrument designed to enable people with CP, especially those with communication problems, to indicate and rate the level of pain they experience in daily life by *themselves*. As far as we know, this is the first time that this possibility has been provided for these severely handicapped people. The pain assessment instrument for patients with cerebral palsy (PAICP) turned out to be suitable for this purpose, allowing many patients to express the severity of their burden for the first time in their lives.

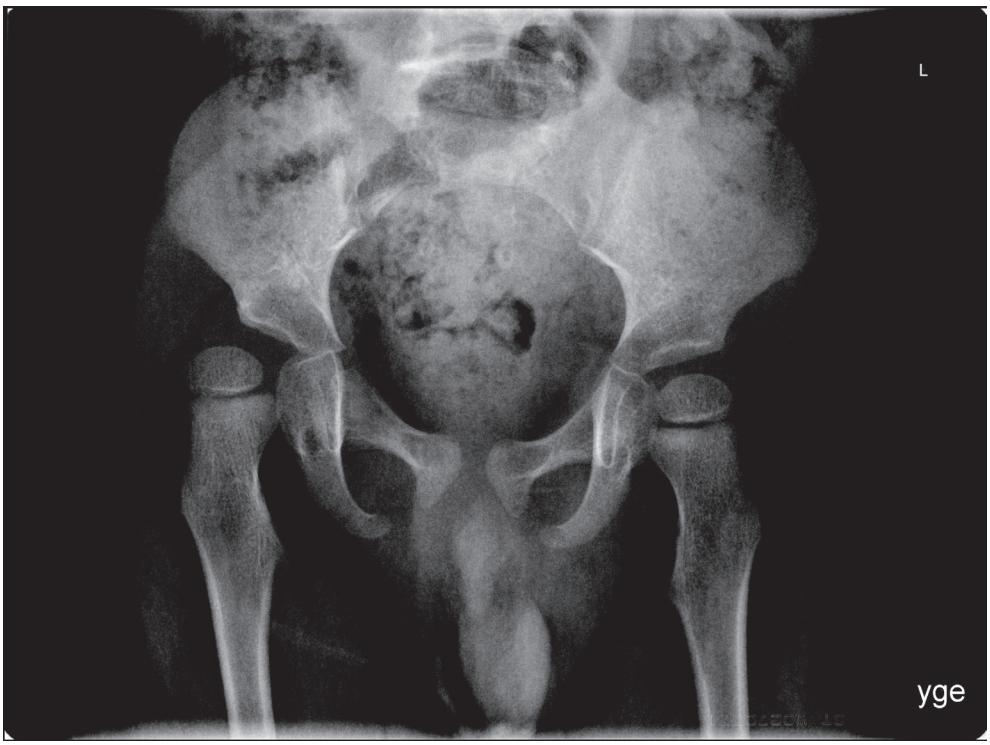


Figure 8.1 X-ray of the pelvis in May 2012, age 2 years 11 months.

We learned that caregivers and physiotherapists were well able to predict whether their pupil had reached a mental developmental age of four years, which is necessary to use the PAICP. The Columbia Mental Maturity Scale (CMMS) proved to be a useful instrument to assess the mental level of the patients. Although the caregivers and physiotherapists knew their pupils well in this respect, their ability to predict the level of pain the patients experience in daily life was rather disappointing. Caregivers and physiotherapists could only sufficiently predict the score indicating the level of pain experienced by their pupils, insofar as it concerned activities that they were actually involved in.

Fortunately, the prediction of the level of pain specifically associated with the patients' hip disorders was quite satisfactory, making the PAICP instrument very useful to assess pain relating to these disorders in people with severe CP.

The study also taught us that an instrument like PAICP should be applied to assess pain levels in activities that caregivers or therapists are not closely involved in.

We also found that undergoing physiotherapy of the legs, a common therapy in this group of patients, is a painful experience for them. *Further research* is needed to evaluate the benefit of the physiotherapy these patients receive, in order to prevent or treat the joint contractures they all develop.

In **chapter 3**, we described the results of a study using the PAICP to assess 160 patients with CP level V (in the GMFCS classification). Patients were initially recruited at large homes for the handicapped. When the study started, four large homes were available for participation, with a total of 697 residents. We informed the doctors working at the homes about the inclusion criteria of the study.

The criteria were: a diagnosis of spastic bilateral cerebral palsy, classified according to Hagberg (1975);[1] inability to walk independently (= without the help of a person), with or without aids (which nowadays would be called GMFCS level V); age at the time of research ≥16 years; mental developmental age ≥4 years; sufficient visual capacity to distinguish drawn figures; ability to make and indicate a choice from a set of drawings, if necessary in a nonverbal manner, and if so, the ability to use the faces pain scale as an instrument to indicate the level of pain they experience in daily life situations; availability of a radiograph of the pelvis not older than five years; and a minimum age of the patient of 16 years at the time of the examination.

Table 8.1 Number of participants and drop-outs

Location	Invited	No response	No permission	Not meeting selection criteria	Preselection participants	Failed CMMS preselection	No radiograph	Final participants
Large residential homes	158	0	62	9	87	17	5	65
Rehabilitation centers, outpatient departments	209	65	48	14	82	15	6	61
Smaller residential homes	97	16	33	9	39	7	5	27
Others, including Fokus foundation				1	11	1	3	7
Total	464	81	143	33	219	40	19	160

After having assessed 65 patients in these four large homes, we approached the smaller homes for the handicapped. Homes were selected throughout the Netherlands based on the number of residents. In addition, we asked physiatrists at the Dutch rehabilitation centers to approach patients who attended their outpatient departments and ask them to participate in the research. We also asked a Dutch society providing independent housing for people with physical impairments ("Fokus") to give us the addresses of their homes, so we could contact their inhabitants. This enabled us to assess 160 patients for our study (Table 8.1).

We think that this method to select patients for our research has minimized selection bias. We think we were able to assess a large percentage of the group of people that could meet our selection criteria, though the percentage was not exactly known at the time of our research. The prediction by the doctors and caregivers of the mental level of the patients was correct in 80% of the cases: only 40 of the 219 people failed to pass the preselection procedure, which was based on the CMMS instrument (Table 8.1).

The main result of this study was an answer to the question: *Hip disorder, does it bother them?* The answer is: *yes, it does*. If an adult with GMFCS level V has a hip disorder, this increases the risk of pain from a basic level of about 10% – not related to hip disorder – to a risk associated with hip disorder of about 30%. The pain level is related to the level of migration of the femoral head out of the acetabulum, which in turn is related to deformity of the femoral head and to asymmetry of migration.

The relatively large percentage of people experiencing pain after failed bony surgery (12 out of 17) is a very troublesome finding. This concerns patients who were operated upon in the past to cure pain, or sometimes just because of an X-ray showing migration, with very poor results. Instead of relief, they experience pain every day in situations like putting on trousers or being lifted from the bed.

This implies that doctors should be very reticent about applying bony surgery to these vulnerable patients, especially for indications other than curing persistent pain. It also means that these patients should only be treated in specialized centers, by orthopedic surgeons, physiatrists, nurses and care teams experienced in this treatment.[2]

We used a formula that enabled us to relate the amount of pain experienced in the hip region to various painful and non-painful situations in daily life. This increased the value of the measurement, as pain is a subjective experience for an individual person, only partly objectified by an instrument. The formula made the pain experience

dichotomous: the patient either experiences pain or not. This made it possible to use the statistics we applied, and to assess relationships with hip disorders seen on X-ray.

The use of X-rays made a maximum of five years ago was a compromise. On the one hand, it enabled us to limit the costs and inconvenience for patients of having an X-ray taken. On the other hand, X-rays made specifically for our study provided the doctor treating the patient with useful information about the situation of their patient's hip, so the results of our research could be used to benefit that individual patient.

A limitation of our study is that, theoretically, the status of a patient's hip could have changed during the period of five years between the moment an X-ray was made and the time of our assessment. As we assessed adults, we think the risk of this happening was not very high. As far as we know, there are no reports of changes in the configuration of the hip in CP patients after reaching adulthood. This could, however, be a possible topic for *further research*.

Another limitation was that we used a scale for the measurement of femoral head deformity that was not validated for this research, as we found no validated scale for the assessment of femoral head deformity in CP at the time of our research. However, the scale we used was highly suitable for the purpose, as the relationship we found between migration and deformity was in agreement with earlier reports we found in the literature.

We found that many participants experienced pain after failed bony surgery, and that a distal resection of the femoral head appeared to be a convenient procedure as a salvage measure. However, our conclusions in this chapter should be interpreted with caution. In most cases, we obtained the history of the patients by questioning their doctors, caregivers or physiotherapists. These people were frequently not aware of the exact indication for the hip surgery previously applied to their patient, and did not know whether the indication was pain, discomfort or just an abnormal X-ray. Surgery had frequently been performed long ago, and many files were missing or incomplete. In that case, we had to rely on reports by the patients or their relatives about the indication for past surgery. Files of patients with a long history of cure and care should be updated carefully, so as to improve the quality of treatment and facilitate scientific research.

Another limitation with regard to the interpretation of the results is that we were only able to assess patients with a mental developmental age of four or higher. Although measures to assess pain in people with a lower mental developmental age have been

developed, these instruments are based on observations by others, not ratings by patients themselves as in our method.

We assume that the pain due to hip disorder experienced by CP patients with a lower level of mental development does not differ from that of patients with a higher level. However, their threshold for pain may be higher,[3] so symptoms of pain may be easily missed. Extra attention must be paid to the assessment of the physical situation of these patients, once the suspicion has arisen that a patient with a low level of mental development has a hip disorder. A diagnostic local anesthesia of the hip joint can help make the right decision in these cases.

We could not explain the hip pain in about 10% of the patients, whose hip X-ray revealed no cause of the pain. Studies have shown that many CP patients experience pain, which can often not be explained by X-ray or other diagnostic methods.[4]

In the literature, pain in CP patients with hip disorders is often reported to be related to osteoarthritis of the hip joint, and clinical experience shows that surgeons performing salvage surgery by distal resection of the femoral head frequently observe extensive destruction of the cartilage of the femoral head after removal. Our research could not confirm a relationship between pain and this single factor; it was the combination of asymmetry of migration, hip deformity and osteoarthritis that was associated with pain.

Our finding that asymmetry of migration was an important factor causing pain rather surprised us, and we did not find an explanation for this specific phenomenon. It underlines the view that the pain experienced by people with CP appears to be a multifactorial phenomenon, which cannot be explained by migration, deformity or asymmetry alone. It also indicates that one should try to obtain or preserve symmetry between the two hips in this respect, for instance by applying soft tissue and bony surgery bilaterally.[5,6]

Future research with new techniques, such as the new 3D CT scan, but also histological investigation, as studied by Maslon et al.,[7] could provide a better explanation for the factors contributing to pain in the hip joints of CP patients.

Chapter 4 describes the results of a study into the relationship between X-ray findings on the one hand and impairments, other than pain, on the other. Generally speaking, 14% of the patients were difficult to care for by their caregivers and 23% were difficult to handle by the physiotherapists, according to the scale we developed. This is not

surprising, as people with GMFCS level V are frequently difficult to handle due to their spasticity, joint contractures, persistent reflexes etc. More than 60% of the patients needed a special molded seat in the chair, and this number increased with the level of hip deformity. Hip deformity also was related to hip adduction contractures.

A limitation of this study was that we used a special self-developed scale to assess the handling difficulties, as existing scales were not suitable for our purpose.

Further research could be aimed at facilitating the care for these patients, for instance by means of antispastic drugs or botulinum toxin to facilitate seating, etc.

After the studies reported on in chapters 3 and 4, we concluded that the problems of seating and the contractures of the adductor muscles form an extra argument, in addition to pain, to try to prevent or treat hip disorders in patients with severe cerebral palsy. The question that remained to be answered was what intervention would be effective to prevent or treat this problem in these vulnerable patients. The answer was sought in two systematic reviews, described in chapters 5 and 6.

Systematic review

The systematic review technique is preferred in cases in which a meta-analysis of articles is not appropriate. In our case, major differences between the published studies in terms of outcome measures, interventions used and study designs (no RCTs) precluded the use of meta-analysis.

After the best evidence synthesis technique had been introduced by Slavin in 1995,[8] it was further improved by Van Tulder et al.,[9] who added the use of qualitative criteria to the system, thus weighting the quality of articles and facilitating the choice of articles to be included in the review.

In **chapter 5**, we describe a systematic review regarding the value of soft tissue surgery and bony surgery in preventing or correcting hip displacement in cerebral palsy. Unfortunately, only one article on soft tissue surgery (Bowen and Kehl[10]) met our selection criteria. There was therefore little evidence for the predominant view in clinical practice and the experience of many doctors involved in this treatment, namely that adductor tenotomy is a useful intervention for patients with spastic CP.

The recent article by Shore et al.[11] shows that the success rate of adductor tenotomy is relatively low in patients with high levels in the GMFCS classification. This makes

the current clinical practice of offering adductor tenotomy to parents with a young child with GMFCS level V disputable.

In any case, the findings of our studies described in chapters 3 and 4, combined with the review in chapter 5, the new data from Shore's article[11] and the information from chapter 6 make it possible to inform the parents much better than before. The new data enable us to discuss with the parents the chances of success of intervention or non-intervention, expressed in percentages, providing them with arguments to make their own choice for their child.

In our review, the answer to the question whether soft tissue surgery or bony surgery can resolve *pain* remains unclear. Clinical experience indicates that pain seldom occurs in young children. Mothers sometimes report pain in their children, which they observe while changing a diaper or upon other involuntary movements of the hip. If an X-ray confirms a hip disorder, adductor tenotomy is the usual treatment for children younger than five years. Clinical experience also indicates that if a patient aged between five and twelve years shows symptoms of pain, and a hip disorder is confirmed by X-ray, bony surgery is useful as a treatment.

The article by Bowen and Kehl does not provide information on pain as an indication for surgery in the patients they describe. It seems their patients were operated as a result of X-ray assessment, not because of complaints like pain or nursing problems. This lack of information makes the results of the article less valuable. In addition, they used a very low criterion of 15% as a threshold for subluxation in their patients.

The articles on bony surgery are not clear about the definition they used for pain experienced by the patients. None of the articles offers a clear-cut definition of pain, and none of the studies involved the use of a pain scale, neither for caregivers, nor for the patients themselves.

In clinical practice, careful assessment of patients is necessary to ensure that the pain they perceive can be related to their hip disorder and not to events like accidental overstretching of the hamstrings by the physiotherapist. Sometimes a temporary interruption of stretching activities by the physiotherapist is enough to make the pain subside. The use of the PAICP is expected to improve the quality of the diagnosis and the evaluation of the outcome in case of intervention.

The effect of bony surgery as a treatment for *pain* is spectacular, but the lack of a clear definition means we cannot accept the pain relief described in the articles as evidence. A striking aspect is that pain as an indication for bony surgery is more frequently mentioned in the articles describing surgery combining varus and derotational osteotomy (VDRO) and pelvic osteotomy. Nevertheless, bony surgery is being used in routine clinical practice to treat patients with pain. As described in chapter 3, pain relief was mentioned in 71% of the cases of bony surgery.

As shown in Table 5.5 of our review, adding pelvic osteotomy to VDRO has led to an increased success rate: the success rate of VDRO alone is about 60%, while reported success rates for the combination with pelvic osteotomy exceed 85%. However, the differences are too small to allow definite conclusions to be drawn about the benefit of combined surgery compared to VDRO alone.

Osteoarthritis as a complication has been described only for the combined intervention (VDRO + pelvic osteotomy), with rates of 8–28%. This suggests that the risk of osteoarthritis is increased by the addition of pelvic osteotomy to VDRO. As we mentioned in our review, surgeons must decide whether pelvic osteotomy should be added to VDRO in individual cases. The use of modern diagnostic techniques like a preoperative three-dimensional CT scan can help resolve the issue.[12] In addition, we must keep in mind that bony hip surgery is itself a painful procedure, which was recently shown to be more painful than spinal fusion.[13]

Further research will be facilitated by unambiguous use of definitions, cut-off points, classification systems and assessment instruments and a sufficient duration of follow–up. The definitions of subluxation, luxation and dislocation used in the literature are unfortunately still highly diverse. For instance, no clear cut-off points have been established for the definition of subluxation versus a normal situation on the one hand and of subluxation versus luxation on the other. When we recalculated the mean pre- and post-operative percentages of subluxation/dislocation from a variety of articles, we chose a 33% cut-off value as proposed by Hägglund as a threshold for reaction or intensified observation.[14]

The article by Shore et al.[11] proves the value of the GMFCS classification in the matter of hip disorders in CP. The GMFCS classification should be used as an important method in all future research projects concerning interventions in CP patients. The use of modern instruments like CPHCS[15] and CPCHILD[16,17] and the PAICP will improve the value of

research significantly. The value of the new FEHLA[18] instrument in predicting the future development of hip displacement remains to be proved in routine clinical practice.

Shore et al.[11] also underline the importance of a follow-up period lasting at least six years, in order to sufficiently evaluate the effect of soft tissue surgery. They agree with Turker and Lee, who also stated this in their article.[19]

On the whole, research in the complicated field of hip disorders in cerebral palsy would gain in quality if study designs involved experimental groups as well as controls, used sophisticated assessment instruments and had a sufficiently long period of follow-up.

Chapter 6 reports on a review of the possibilities to cure the pain and other impairments in adult patients with otherwise incurable complaints associated with hip disorders. The review shows that there are successful methods of palliative surgery available to reach this goal. Regrettably, none of the intervention methods (resection surgery, hip replacement or arthrodesis) provides the ultimate solution to treat the severely deformed painful hip in CP. The published literature does not clearly favor one procedure over the others. However, the technique for which the largest number of results has been published is subtrochanteric resection, which seems to eventually lead to a good outcome. The succes rate of *distal resection* of the femoral head in terms of curing pain is 90–100%, as reported in the articles meeting our quality standards. Distal resection means resection below the level of the trochanter minor of the femur. Bony intervention, however, implies no instant cure: we note that it can take up to three to six months postoperatively before maximum pain relief is obtained.

A more proximal approach like the resection described by Girdlestone[20] results in a very high risk of residual pain, as we described in chapter 3.

In our opinion, when opting for femoral head resection surgery, the Castle[21] is to be preferred over the McCarthy procedure,[22] achieving better results and fewer complications. The explanation for this difference is not quite clear; perhaps the careful resection of the periosteum described by McCarthy to prevent heterotopic ossification (HO) has a counterproductive effect. In addition, it is unclear why the McCarthy technique yields a much lower percentage of success in terms of curing pain than the Castle procedure (90–100% versus 53–77%).

A disadvantage of the femoral resection technique is that traction on the operated leg is needed for several weeks after the operation. On the other hand, many surgeons refrain from traction after a Castle procedure.

Many surgeons prefer the Schanz technique,[23] which does not require traction. We did not find any articles on this technique that met our quality criteria, and the literature is also contradictory. One author[24] reported 98% pain relief in their patient group, but others reported a 63% complication rate.[25] One of the main problems is persistent pain and postoperative problems with pressure ulcers from the retained femoral head.

Total hip replacement can be useful for patients with GMFCS levels I–III, but this technique also has a high complication rate. Femoral fractures have been reported in 36% of the cases, and heterotopic ossification in 29%.[26] Recently, the preoperative use of botulinum toxin was advocated to reduce postoperative complications.[27]

Arthrodesis of the hip joint can relieve pain in these patients, but this technique also has a very high complication rate. The only article that met our quality criteria reported a non-union rate of 42%.[28]

Reports on palliative surgery also suffer from poor definitions of pain, nursing problems and seating problems, hampering the interpretation of results. The best-evidence synthesis method proved to be useful in this respect. Authors have used different methods of reporting heterotopic ossification. Some use no classification, while others use the McCarthy classification[16] or the Brooker classification.[29] Regardless of which scale is used, it is clear that heterotopic ossification is a major problem complicating surgery in adults with CP. *Further research* should be aimed at preventing this phenomenon in the future.

Decision tree for monitoring and treatment

In recent years, **botulinum toxin** has been promoted as a useful addition to the techniques available to cure problems associated with hip disorders in CP.

In **chapter 7**, we describe the limited role of botulinum toxin in preventing or curing hip disorders. The chapter also gives an overview of the research we did in our group of 160 patients, combined with a systematic review of the effects of surgery in preventing or curing hip disorders associated with cerebral palsy. This has resulted in a **decision tree** for monitoring and treatment, based on GMFCS level, age and results of the intervention. As far as we know, this is the first such decision tree to be presented.

We started our research with the question whether CP patients are bothered by hip disorder. The answer is yes. Around thirty percent of the patients with abnormalities

on pelvic X-ray suffer from pain due to the disorder. Patients with hip disorder due to CP need a specially moulded seat and they are affected by hip contractures.

Since we performed a cross-sectional study, we could only use the patients' current situation, in many cases without details of their history. 132 out of 160 patients had undergone hip surgery, indicating the seriousness of their situation. The surgery had been without result in quite a number of the patients, as their hip was still subluxated and deformed, and a relatively high proportion suffered from pain, obviously despite surgery or even as a complication of it.

Subsequently, we studied publications on the value of interventions for hip disorders in CP patients. Unfortunately, our systematic reviews yielded no clear results for the majority of intervention methods. As regards soft tissue surgery, total hip replacement and arthrodesis of the hip, only one article satisfied our quality criteria. As regards bony surgery for correction and palliative intervention, only a best evidence synthesis provided a clue as to the value of the intervention.

Articles suffered from small sample sizes, unclear definitions of pain measurement and short follow-up. The natural history of hip disorders in childhood still is unclear, and the effects of interventions like postural correction and botulinum toxin treatment are still ambiguous.

Much research remains to be done to improve the prevention and intervention methods in the field of hip disorders in CP.

Future research

Future research should focus on epidemiological studies of the incidence and prevalence of hip disorders and the value of interventions, similar to the studies performed in Australia and Sweden. The registration system for patients with cerebral palsy, the development and application of the GMFCS assessment system and the registration of patients' hip status in these countries has meant a major breakthrough in the research into the epidemiology of hip disorders in CP and their treatment. Research into CP in the Netherlands can never acquire sufficient quality without a proper registration system, as was discussed by my colleague Marc Wichers in his thesis.[30]

The most controversial issue in our research was the value of soft tissue surgery in young patients with cerebral palsy and GMFCS IV or V. It is doubtful whether we should advise parents to have this surgery performed on their child, in view of the relatively poor effect on the hip disorders in these patients, as described by Shore et al.[11]

The value of this intervention could be evaluated by a controlled study, in which patients could be randomly assigned to two groups that are comparable in terms of patient characteristics. One group could have an adductor tenotomy performed, while the other could be treated with conservative therapy only. As Shore et al. pointed out, a follow up period of six years would be the minimum.[11]

As soon as an appropriate registration system has become available, research could also focus on the debate about the value of concentrating surgery of the lower limbs among children – and adults – with CP in specialized centers. Determining the value of such centers by assessing the results of interventions, and above all, the level of satisfaction of patients and parents, would be an important tool to improve the care for this vulnerable patient group. A controlled study could be performed at these centers to assess the value of bony surgery in patients with CP and GMFCS level IV or V, which is even more controversial than soft tissue surgery in these patients.

As mentioned above, further research will be facilitated by unambiguous use of definitions, cut-off points, classification systems, assessment instruments and sufficiently long follow-up after an intervention.

Further research is also needed to evaluate the benefit of the leg-stretching physiotherapy that CP patients receive to prevent or treat the joint contractures they all develop. In our research, we found that this intervention, a common therapy in this group of patients, is a very painful experience for them.

What does all this mean for the advice we should give to the parents of our patient William S.?

According to the advice I received from my teacher during my residency, the answer to the above question would be simple: the patient should be operated soon, because the hip disorder would bother him in adulthood or even earlier. At that time, the late 1970s, there was a considerable chance that a patient would meet an orthopedic

surgeon who would perform an adductor tenotomy only on the affected side, or would add a varisating derotational osteotomy to the procedure, 'just to make sure that there would be an effect'.

On the other hand, as I reported in the introduction to this thesis, a physiotherapist at a residential care centre in Katwijk told me that he was "not sure whether the patients are bothered by their hip disorder". In view of these contradictory opinions, I would not have been sure at the time what advice to give to the mother of William, that is, whether to opt for surgery or no surgery.

That was the reason why I started this research project. Now, after 21 years of studying the subject of complaints associated with hip disorders in patients with CP, I am still not completely sure, but I do know more about relevant chances of success of the various treatment options in this field.

Until the appearance of the study by Shore et al.[11] about the success rate of adductor tenotomy in relation to GMFCS level, everything seemed clear enough. The advice would be to have an adductor tenotomy performed, with a chance of success of around 60%, as explained below. The advice for surgery would be supported by the report by Zarrinkalam et al.,[17] stating that the overall function of a child with severe cerebral palsy would improve after surgery, as measured by the CPCHILD score.

In our research, described in chapter 3, we discovered that the risk of experiencing pain relating to hip disorder in adolescents and adults with severe CP is about 30%. Therefore, I could tell the mother of William that my teacher was quite right: an adult with severe CP with a hip disorder has a 30% risk of experiencing pain. However, one could also take the optimistic view and say that the chance of *not* experiencing pain would be about 70%. In addition, based on the research described in chapter 4, I could warn the mother that William would be at risk of needing a special molded chair and to develop an adduction contracture of the hips, although this would probably not hamper nursing activities.

Based on the research reported in chapter 5, we could assure the mother that an adductor tenotomy performed before the age of five, preferably around the age of four, would offer a high chance of success in terms of reducing the migration percentage (MP). In the only article with sufficient quality, Bowen & Kehl described that 87% of the hips with a pre-operative MP <50% were stable at follow-up.[10]

In cases like that of William S., we would advise the parents to have the surgeon perform a bilateral adductor tenotomy, with a 60–80% chance of success and that the risk of experiencing pain in adulthood after a successful tenotomy would be only 10% (chapter 3).

If, by chance, William should fail to benefit from the adductor tenotomy, the odds of a successful varisating derotational osteotomy, possibly combined with a pelvic osteotomy, would be 53%–75% for VDRO alone and 85% or higher for the combined surgeries.

However, careful consideration would be appropriate when choosing the setting for the bony surgery. Only an experienced orthopedic surgeon should perform the operation, which would best take place in a specialized center that cares for vulnerable, non-verbal patients like William S. Choosing a non-experienced orthopedic surgeon would increase the risk of post-operative failure, as described in chapter 3. In our research, we came across a number of operations that had obviously not been successful: 20 of our 160 patients had an abnormal configuration of the femur and pelvis, mostly after bony surgery, which was also associated with (residual) pain.

To reassure the mother, I would tell her that if William should experience pain in his hip because of residual hip disorder in adulthood, a salvage procedure such as distal resection of the femoral head or a valgisating rotational osteotomy would yield a 90–100% chance of curing the pain, as described in chapter 6.

This was the situation until March 2012, when the article by Shore et al. appeared about the success rate of adductor tenotomy in relation to the GMFCS level. They reported that the success rate of adductor tenotomy in patients with GMFCS level V, in other words patients like William S., was only 14%.[11]

So what would my advice to the mother of William S. be now?

Probably no surgery in the short term.

The risk of persisting pain in adulthood without surgery in childhood and after a possible salvage procedure in adulthood would only be about 3%. This is calculated as follows. There is a 30% risk of pain associated with hip disorder in adulthood (chapter

3), but a chance of 90–100% that this pain can be cured by a salvage procedure (chapter 6); 30% of 90% is about 3%.

This is why, in our decision tree (chapter 7), we changed the box around the words 'adductor tenotomy' to a dotted box for patients with GMFCS level V.

We think we should inform parents about chances, that is, the chances of the success of soft tissue surgery, bony surgery and salvage surgery, as applied to their child's particular situation.

This is the main thing we have learned.

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Summary

Chapter 1 of this thesis presents an introduction and outline of the thesis, and explains the rationale for the research. While working as a physiatrist for children, I was confronted with the question: are patients with cerebral palsy (CP) who develop hip disorders bothered by this disorder, in their youth, in adulthood and / or later? And if so, how can we help these people by preventing or curing this problem?

The chapter reports about the development of the research design and a suitable instrument to measure pain in the patients of the study sample. After the study among 160 patients with CP level V (in the GMFCS classification) had been completed, a systematic review of the literature about preventive, corrective and palliative surgery was set up.

The results of the patient study and the systematic review enabled us to develop a decision tree for the treatment of hip disorders in spastic cerebral palsy. The decision tree was presented and discussed at the regular meetings of Dutch pediatric physiatrists, and is now considered to be the Dutch consensus.

Chapter 2 describes the development of a pain assessment instrument (PAICP) for patients with cerebral palsy, which was then tested in a cross-sectional validation study. Measurements were made at homes for the severely handicapped and in the home environment of the patients. The participants were 164 adult patients with severe CP, their caregivers and physiotherapists. The study was preceded by a pilot study with nine healthy children.

The PAICP instrument uses drawings of everyday situations. Patients score the pain they experience on a faces pain scale. Reproducibility and construct validity were assessed in a pilot study with CP patients and healthy children. The construct validity and agreement between the pain scores of the patients and proxies was assessed in 160 patients with severe CP. The main outcome measure was the pain score on PAICP.

The results showed that the instrument had good construct validity and sufficient test-retest reproducibility. A significant difference was found between the mean scores for 'painful' and 'non-painful' situations. We found moderate agreement between the scores of the patients and the proxies regarding daily activities, and only for those activities in which the proxies were personally involved. There was significant agreement in the scores of the patients and their proxies in hip straining situations.

We concluded that the PAICP has satisfactory test-retest reproducibility and construct validity. It provides an indication of the pain experienced by patients in situations in

which proxies are not personally involved and may also be more valid than proxy measures for other situations.

A striking finding was that stretching of the legs by physiotherapists, a common procedure in this group of patients, is actually a painful experience for them. Further research is needed to evaluate the benefit of the physiotherapy these patients receive to prevent or treat the joint contractures they all develop.

In **chapter 3** we report on a study of the relationship between migration and deformity of the femoral head and osteoarthritis on the one hand, and pain on the other hand, in people with severe CP. Measurement instruments that enable the patients themselves to indicate the severity of their pain had not previously been used to study this relationship.

A cross-sectional study was carried out to investigate the relationship between hip X-ray findings (migration, deformity and osteoarthritis) and pain in 160 adults with severe CP. The patients were unable to walk independently (GMFCS level V), and had a minimum mental age of four years. They scored their pain on the PAICP instrument. Their scores ranged from 1–7. Pain was dichotomized, and logistic regression analysis was performed to evaluate the associations between X-ray findings and hip pain.

Thirty patients out of a total of 160 (19%) had hip pain in hip-straining situations. If an adult with GMFCS level V has a hip disorder, this increases the risk of pain from a basic level of about 10% – not related to hip disorder – to a risk associated with hip disorder of about 30%. Severe deformity of a femoral head was present in 41 (30%) patients. Osteoarthritis was found in 23 (16%) patients. Twenty patients who had an abnormal configuration of the femur and pelvis, mainly after (unsuccessful) bone surgery (n=17, 85%), were analyzed as a separate group. In this group, 12 (60%) patients suffered from pain in hip straining situations.

Among the remaining 140 patients, migration and deformity of the femoral head were found to be closely related. Univariate logistic regression analysis of these combined factors showed a significant association with hip pain (odds ratio 2.79 (1.01–7.70)). Pain was also related to severe asymmetry, expressed as a difference in migration percentage in the left versus right hip. Osteoarthritis was not significantly associated with pain.

We conclude that there is a high prevalence of hip pain after unsuccessful bony hip surgery in patients with severe CP. Migration and deformity of the femoral head

are closely inter-related, and are associated with pain. Pain is also associated with asymmetry of migration of the femoral head.

Treatment should aim to prevent migration and deformity of the femoral head and to maintain as much symmetry as possible. If bony surgery is considered, the risk of residual pain due to failed surgery and/or complications is an important aspect that should be taken into consideration.

Further research could investigate changes in the configuration of the hip in CP patients after they reach adulthood.

It is not clear why 10% of patients suffer from pain in the hip region without abnormalities on the pelvic X-ray that could explain this pain. Future research with new techniques, such as the new 3D CT scan technique, but also histological investigation, might offer new insights into the factors contributing to pain in the hip joints of CP patients. Further research would be facilitated by unambiguous use of definitions, cut-off points, classification systems and assessment instruments and a sufficient duration of the follow-up.

Chapter 4 presents a study of the relationship between hip disorders (migration and deformity of the femoral head) on the one hand, and handling problems, seating problems and impairments (decubitus ulcers, fractures and contractures) on the other. This was a cross-sectional study among 160 adult patients with severe CP, the same sample used in the study described in chapter 3.

Although no significant associations were found between hip disorders and handling problems, both migration and deformity of the femoral head were positively related to the need for a special molded seat in the wheelchair, and were negatively associated with the ability to abduct the hip. Patients with seating problems had a higher mean migration percentage then patients without seating problems; the difference being significant. Also, limited passive hip abduction was significantly related to a higher mean MP in the group. Seating problems were found in 51.3% of the patients with no hip deformity, 51.8% of the patients with moderate deformity and 87.2% of the patients with severe deformity. Limited abduction was found in 39.0, 53.4 and 75.6% of patients with no, moderate and severe hip deformity, respectively.

We conclude that migration and deformity of the femoral head should be prevented in patients with severe CP. Even in the hands of experienced caregivers and therapists, people with CP GMFCS level V are difficult to handle due to their spasticity, joint

contractures, persistent reflexes etc. Further research could be aimed at facilitating the care for these patients, for instance by using antispastic drugs, administering botulinum toxin to facilitate seating, etc.

Chapter 5 presents the results of a systematic review of the effectiveness of preventive and corrective surgical interventions to treat hip luxation in patients with severe cerebral palsy. Data sources were obtained by a systematic literature search in MEDLINE, CINAHL, EMBASE, Cochrane controlled trial register and PEDRO.

We applied the following inclusion criteria: severe cerebral palsy (GMFCS levels IV and V), evidence of hip subluxation or dislocation (assessed as migration percentage (MP)), use of surgical procedure, and using decreased MP and pain relief as outcome measures. A qualitative analysis according to Steultjens et al.[1] and Van Tulder et al.[2] was performed for soft tissue surgery and osteotomies. We used the best-evidence synthesis technique to summarize the results of the included studies. All studies were observational.

Only one of the five studies involving soft tissue surgery was of sufficient quality. Nine of the ten studies involving osteotomies were of sufficient quality, including a total of 189 patients. As a result of osteotomy, the mean MP at follow-up ranged from 6 to 29%, with better results for the combination of varus derotational osteotomy (VDRO) and pelvic osteotomy than for VDRO only. No relationship could be established between the effect of the surgical procedure and the patients' age at the time of the surgery or the duration of follow-up. The percentage of patients reporting pain decreased from 81% preoperatively to 5% at follow-up, though the evidence level was insufficient. Twenty-five percent had complications like osteoarthritis, ulcers or fractures.

We conclude that there is insufficient evidence for the effectiveness of soft tissue surgery in stabilizing the hip, due to insufficient quality of the retrospective observational studies.

There are indicative findings for an effect of bony surgery in stabilizing the hip. Timing of the procedures remains an undecided issue. Multicenter trials with larger groups of patients that can be followed for a longer period could shed further light on this complex subject. We had to recalculate the mean pre- and post-operative percentages of subluxation/dislocation from a variety of articles due to different definitions of outcome measures in the articles. Further research would be facilitated by unambiguous use of definitions describing hip disorders and the use of cut-off points in, for instance, the definition of subluxation versus normal on the one hand and subluxation versus luxation on the other.

Chapter 6 presents the results of a systematic review of the effectiveness of palliative hip surgery in severe CP. Palliative intervention is undertaken when preventive or corrective surgery fails or has not been performed. A number of salvage interventions have been described.

Just as in chapter 5, we applied the following inclusion criteria: severe cerebral palsy (GMFCS IV and V), evidence of hip subluxation or dislocation (assessed as migration percentage (MP)), use of surgical procedure, and using decreased MP and pain relief as outcome measures. We performed a qualitative analysis according to Steultjens et al.[1] and Van Tulder et al.,[2] using the best-evidence synthesis technique to summarize the results of the included studies. All studies were observational.

We found articles on resection surgery of the femoral head, arthrodesis of the hip joint and total hip replacement. According to the best evidence synthesis, the indicative findings of these studies suggest that relief of pain and seating and nursing problems can be achieved after a salvage resection of the femoral head. Regrettably, none of the intervention methods (resection surgery, hip replacement or arthrodesis) provides the ultimate solution to treat the severely deformed painful hip in CP. The published literature does not clearly favor one procedure over the others. However, the technique for which the largest number of results has been published is subtrochanteric resection, which seems to eventually lead to a good outcome. Our review concludes that, if resection surgery is to be performed, distal resection and interposition surgery according to Castle and Schneider[3] should be the method of choice. The percentage of pain relief after this intervention is 90–100%. Bony intervention, however, implies no instant cure: we note that it can take up to three to six months postoperatively before maximum pain relief is obtained.

As for the technique of valgisating derotation osteotomy as described by Schanz, which is practiced by a number of orthopedic surgeons, we found no articles that could pass our quality criteria. One author reported 98% pain relief, while another described a complication percentage of 63% for this technique.

We only found one article that could pass our quality criteria that reported on other techniques like total hip replacement respectively arthrodesis of the hip joint. These techniques can produce pain relief, but also cause a relatively large number of complications. In total hip replacement, dislocation of the prosthesis occurs in 29% of the cases, heterotopic ossification (HO) is present in 45%. In addition, femoral fractures

are relatively frequent (36%). Arthrodesis of the hip joint can result in a pain-free hip, but the rate of complications, such as non-union, is high (42%).

Bony surgery can be used as a palliative intervention to relieve pain and discomfort in patients with cerebral palsy and hip disorder. Unfortunately, however, heterotopic ossification is a major problem complicating surgery in these patients. Research should be aimed at preventing this phenomenon in the future.

Chapter 7 synthesizes the results of the research described in chapters 2–4 and the reviews described in chapters 5 and 6. It also describes measurements to assess subluxation and the pathophysiology and natural history of the phenomenon. Methods of intervention are explained and their value evaluated. The question whether surgery should be performed symmetrically is discussed, as is the correct timing of surgery. The chapter also evaluates the advantages and disadvantages of surgery to prevent or treat hip disorders, and comments on the role of botulinum toxin. In addition, it discusses the different options for surveillance of hip disorders and the corresponding measures for prevention or treatment. The small hazards of frequent X-ray assessment are also discussed.

All this culminates in a proposal for a **decision tree** for the surveillance and intervention of hip disorders in spastic CP. The algorithm is based on our research and on the literature assessed in our reviews. It is now regarded as the Dutch consensus and was discussed at the general meetings of Dutch pediatric physiatrists. We compare the algorithm with the Australian consensus on surveillance, as published in 2008.

The decision tree uses the GMFCS level as a basis for intervention and is divided into three periods based on the age of the patient. The first period concerns the ages between one and four years, in which a bilateral adductor tenotomy is the preferred treatment option for most patients when an imminent hip displacement is diagnosed. The second period concerns the ages between eight and nine years, in which patients with hip disorder can be treated with VDRO, whether or not combined with pelvic osteotomy. The third period is the period after the age of nine years, in which palliative surgery can be used to treat patients with otherwise incurable pain or impairments.

We use the cerebral palsy hip classification system (CPHCS) to decide whether an intervention is needed; we think that adductor tenotomy can be useful at every GMFCS level, provided the parents are fully informed about the reason for the operation and its success rate. Bony interventions should be reserved for patients with GMFCS levels I–III, and only exceptionally used in patients with higher GMFCS levels.

Chapter 8 presents a general discussion of the research undertaken for this thesis, using the problems of a specific patient as the point of departure. The chapter addresses the main issues of the research studies and reviews we undertook, and applies them to the patient's problems. In this chapter we also put the results of our research into perspective: our patient study was only a cross-sectional and not a prospective study. Only a few articles in the reviews satisfied our quality criteria, so we had to use the best evidence method to draw conclusions.

The main conclusion is that hip disorders in spastic CP cause pain and impairments, and should if possible be prevented or treated. But not at any price: careful assessment, evaluation and discussion are necessary for each individual patient and evaluation and treatment should be performed by an experienced and patient-oriented team of professionals with affinity for the specific population.

The general discussion also includes some recommendations for future research. A suggestion is made for further research into the value of soft tissue surgery for young CP children with a high GMFCS classification. A plea is made for a properly designed system of epidemiological research into the incidence and prevalence of CP in the Netherlands and the effects of interventions, comparable to the research performed in Australia and Sweden. Such a system would make it possible, for instance, to determine the value of concentrating surgical interventions for CP children and adults in specialized centers.

In addition, further research should address the value of the common practice of passively stretching the hips and knees of CP patients, as our study shows that this is a very painful procedure for the patients.

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Samenvatting
(Summary in Dutch)

Hoofdstuk 1 bevat een inleiding en overzicht van dit proefschrift. We beschrijven de aanleiding voor het onderzoek. Als kinderrevalidatiearts werd de auteur geconfronteerd met de vraag: patiënten met een cerebrale parese ontwikkelen heupaandoeningen. Is dat een aandoening die klachten veroorzaakt, in hun jeugd, bij het bereiken van de volwassen leeftijd en/of later? En zo ja, hoe kunnen we deze mensen helpen door het voorkómen of verhelpen van deze klachten?

We beschrijven de totstandkoming van de onderzoeksopzet en de keuze van het juiste instrument om pijn te meten bij de patiënten van de onderzoeksgroep.

Na het onderzoek bij 160 patiënten met cerebrale parese (CP) en GMFCS-niveau V, werd een systematische review van de literatuur over preventieve, correctieve en palliatieve chirurgie uitgevoerd.

Na het patiëntenonderzoek en de systematische review was het mogelijk om een beslisboom voor de behandeling van heupafwijkingen bij patiënten met spastische cerebrale parese te ontwikkelen. De beslisboom is gepresenteerd en besproken in de reguliere vergaderingen van de Nederlandse kinderrevalidatieartsen en wordt beschouwd als de 'Dutch consensus'.

Hoofdstuk 2 beschrijft de ontwikkeling van een pijnmeetinstrument (PAICP) voor patiënten met CP. Het onderzoeksdesign was een puntprevalentieonderzoek. Patiënten werden gerekruteerd in instellingen voor (meervoudig) gehandicapten, maar ook thuiswonende CP-patiënten namen deel. De deelnemers waren 164 volwassen ernstig aangedane CP-patiënten, hun verzorgers en hun fysiotherapeuten. Een pilotstudy werd gedaan bij negen gezonde kinderen.

De PAICP bevat tekeningen van dagelijkse situaties. Patiënten scoren de ervaren pijn op een pijnschaal met gezichten (Faces Pain Scale). De reproduceerbaarheid en constructvaliditeit werden onderzocht in een pilotstudy bij CP-patiënten en gezonde kinderen. De constructvaliditeit en de overeenstemming tussen de pijnscores van de patiënten en hun verzorgers en therapeuten werd beoordeeld bij 160 patiënten met ernstige CP. De belangrijkste uitkomstmaat was de pijnscore op de PAICP.

Er bleek een goede constructvaliditeit en adequate test-hertest reproduceerbaarheid te zijn. Er werd een significant verschil gevonden tussen de gemiddelde scores voor 'pijnlijke' en 'niet pijnlijke' situaties. We vonden verder een matige overeenkomst tussen de scores van de patiënten en verzorgers/therapeuten voor de dagelijkse activiteiten, en alleen voor die activiteiten waarbij de naasten persoonlijk betrokken zijn. Er was

een significante overeenkomst tussen de scores van de patiënten en hun naasten betreffende heupbelastende dagelijkse situaties.

De conclusie wordt getrokken dat de PAICP een adequate test-hertest reproduceerbaarheid heeft en een goede constructvaliditeit. Het instrument geeft een indicatie van de pijn die patiënten ervaren in situaties waarin naasten niet persoonlijk betrokken zijn en is naar onze mening beter bruikbaar dan inschattingen van naasten in andere situaties waar naasten niet rechtstreeks betrokken zijn.

Opvallend is, dat het ondergaan van fysiotherapie van de benen, het zogenaamde doorbewegen, een gebruikelijke behandeling bij deze groep patiënten, een pijnlijke ervaring voor hen is. Verder onderzoek is nodig om het nut van de behandeling te evalueren die deze patiënten krijgen om de contracturen die ze allen ontwikkelen te voorkomen of te behandelen.

In **hoofdstuk 3** presenteren we de resultaten van een onderzoek naar de relatie tussen migratie, deformatie van de heupkop en arthrose enerzijds, en pijn anderzijds, bij mensen met ernstige CP. Meetinstrumenten waarmee de patiënt zelf de ernst van de pijn kan aangeven waren tot nu toe niet gebruikt om deze relatie te onderzoeken.

Een cross-sectionele studie werd uitgevoerd om de relatie tussen waarden op een röntgenfoto van de heup (migratie, deformatie en artrose) en pijn bij 160 patiënten met ernstige CP te onderzoeken. De patiënten waren niet in staat om zelfstandig te lopen (GMFCS-niveau V) en hadden een minimale mentale ontwikkelingsleeftijd van 4 jaar. Ze gaven hun pijn aan op de hiervoor ontworpen pijnschaal (PAICP) door middel van scores variërend van 1–7. Pijn werd gedichotomiseerd, en een logistische regressieanalyse werd uitgevoerd om de relatie tussen de röntgenbevindingen en pijn in de heup te beoordelen.

Dertig patiënten van de 160 (19%) hadden pijn in de heup in heupbelastende situaties. Als een volwassene met CP, GMFCS niveau V, een heupafwijking heeft, verhoogt dit de kans op pijn van een basaal niveau van 10% – niet gerelateerd aan de heupafwijking – naar een kans op pijn gerelateerd aan de heupafwijking van rond de 30%. Ernstige deformatie van een heupkop was aanwezig bij 41 (30%) patiënten. Arthrose werd gevonden bij 23 (16%) patiënten. Twintig patiënten met een afwijkende configuratie van femur en het bekken, voornamelijk na (mislukte) botchirurgie (n=17, 85%), werden geanalyseerd als een aparte groep. In deze groep hadden 12 (60%) patiënten last van pijn in de heupbelastende situaties.

Van de overige 140 patiënten bleken migratie en deformatie van de heupkop nauw gerelateerd. Deze gecombineerde factoren vertoonden bij univariate logistische regressieanalyse een significante associatie met pijn in de heup (odds ratio 2,79 [1,01 tot 7,70]). Pijn was ook gerelateerd aan ernstige asymmetrie, uitgedrukt als het verschil in migratiepercentage van de linker- ten opzichte van de rechterheup. Artrose bleek niet significant geassocieerd met pijn.

We concluderen dat er een hoge prevalentie is van pijn in de heup na een mislukte botoperatie van de heup. Migratie en deformatie van de heupkop zijn sterk aan elkaar gerelateerd, en zijn geassocieerd met pijn. Pijn is ook gerelateerd aan asymmetrie van migratie van de heupkop. De behandeling moet daarom gericht zijn op het voorkómen van migratie en deformatie van de heupkop en het behoud van zoveel mogelijk symmetrie. Als botchirurgie wordt overwogen, moet het risico op persisterende pijn door mislukte chirurgie en/of complicaties serieus in de overwegingen worden betrokken.

Suggestie voor verder onderzoek is de mogelijke verandering van de configuratie van de heup bij CP-patiënten na het bereiken van de volwassen leeftijd.

We memoreren dat het niet duidelijk is waarom 10% van de patiënten last van pijn in de heupregio aangeeft zonder dat hier een verklaring voor is te zien op de röntgenfoto van het bekken. Toekomstig onderzoek met nieuwe technieken, zoals de nieuwe 3D-CT-scantechniek, maar ook histologisch onderzoek, kan wellicht een verklaring opleveren voor de factoren die bijdragen aan pijn in de heup van CP-patiënten.

Voortgezet onderzoek zal aan waarde winnen door het gebruik van eenduidige definities, drempelwaarden, classificatie en meetinstrumenten, alsmede het hanteren van een ruime follow-up-duur.

Hoofdstuk 4 geeft de resultaten van een onderzoek weer naar de relatie tussen heupafwijkingen (migratie en deformatie van de heupkop) aan de ene kant, en hanteringsproblemen, zitproblemen en beperkingen (decubitus, fracturen en contracturen) aan de andere kant. Het onderzoeksdesign is een cross-sectionele studie van 160 volwassen patiënten met ernstige CP, dezelfde populatie als beschreven in hoofdstuk 3.

Er werden geen significante associaties gevonden tussen heupaandoeningen en hanteringsproblemen. Zowel migratie als deformatie waren positief gecorreleerd met de noodzaak van het gebruik van een speciale zitting in de rolstoel, en waren negatief gerelateerd aan de mogelijkheid tot passieve abductie van de heup. Patiënten met

zitproblemen hadden gemiddeld een hoger migratiepercentage (MP) dan patiënten zonder zitproblemen; het verschil was significant. Ook een beperkte passieve abductie van de heup was significant gerelateerd aan een hoger gemiddeld MP.

Zitproblemen werden gevonden in 51,3% van de gevallen (bij patiënten zonder heupkopdeformatie), 51,8% (bij matige deformatie), 87,2% (bij ernstige deformatie van de heupkop) en beperkte abductie bij 39,0%, 53,4% en 75,6% van de patiënten met respectievelijk geen, matige en ernstige deformatie van de heupkop.

We concluderen opnieuw dat migratie en deformatie van de heupkop zoveel mogelijk moeten worden voorkomen bij patiënten met ernstige CP. Zelfs in de handen van ervaren verzorgers en therapeuten zijn mensen met CP en GMFCS-niveau V moeilijk te hanteren als gevolg van hun spasticiteit, contracturen, persisterende reflexen, etcetera. Verder onderzoek zou kunnen worden besteed aan de facilitering van de zorg voor deze patiënten, bijvoorbeeld door het gebruik van antispastische medicatie, de toepassing van botuline toxine als middel om het zitten te vergemakkelijken, etcetera.

Hoofdstuk 5 presenteert de resultaten van een systematic review naar de effectiviteit van preventieve en correctieve chirurgische interventie om heupluxatie te behandelen bij patiënten met ernstige CP. Gegevensbronnen waren een systematisch literatuuronderzoek in MEDLINE, CINAHL, EMBASE, Cochrane gecontroleerd trial register en PEDRO.

We gebruikten de volgende inclusiecriteria: ernstige CP (GMFCS IV en V), aangetoonde subluxatie of dislocatie van de heup (gemeten door middel van het MP, de aard van de chirurgische ingreep, en een verminderd MP en afname van pijn als uitkomstmaten.

Een kwalitatieve analyse volgens Steultjens et al.[1] en Van Tulder et al.[2] werd uitgevoerd voor de weke-delen-chirurgie en osteotomieën. We gebruikten de techniek van de best evidence synthese om de resultaten van de geïncludeerde artikelen weer te geven. Alle onderzoeken waren observationeel.

Slechts één van de vijf artikelen over weke-delen-chirurgie was van voldoende kwaliteit. Negen van de tien artikelen betreffende osteotomieën waren van voldoende kwaliteit, met een totaal van 189 patiënten.

Het gemiddelde MP bij follow-up na een botoperatie varieerde van 6–29%, met betere resultaten voor de combinatie van variserende deroterende osteotomie (VDRO) en bekkenosteotomie dan voor VDRO alleen.

Er kon geen relatie worden aangetoond tussen het effect van de chirurgische ingreep en de patiëntenleeftijd bij operatie of de duur van de follow-up.

Het percentage patiënten met pijn nam af van 81% preoperatief tot 5% bij follow-up, maar met onvoldoende bewijskracht. Vijfentwintig procent had complicaties zoals artrose, ulcera of fracturen.

We concluderen dat er onvoldoende aantoonbaar bewijs is voor de effectiviteit van weke-delen-chirurgie om de heup te stabiliseren, als gevolg van onvoldoende kwaliteit van de retrospectieve observationele artikelen. Er zijn wel aanwijzingen voor een effect van botchirurgie op het stabiliseren van de heup. De juiste timing van de interventies blijft een probleem. Multicenteronderzoek met grotere groepen patiënten die gevolgd kunnen worden voor een langere periode zou meer licht kunnen werpen op dit ingewikkelde onderwerp.

In het kader van de review moesten we het gemiddelde pre- en postoperatieve percentage van de subluxatie/dislocatie zoals weergegeven in diverse artikelen opnieuw berekenen. Dit vanwege de niet-eenduidige definiëring. Verder onderzoek zal aan waarde winnen door ondubbelzinnig gebruik van definities bij het beschrijven van heupaandoeningen en het gebruik van afkappunten in bijvoorbeeld de definitie van subluxatie versus normale configuratie enerzijds en subluxatie versus luxatie anderzijds.

Hoofdstuk 6 bevat de resultaten van een systematische review over de resultaten van palliatieve heupoperaties bij ernstige CP. Als preventieve of correctieve operaties zijn mislukt of niet uitgevoerd en er zijn persisterende klachten, is palliatieve interventie aangewezen. Een aantal palliatieve maatregelen wordt beschreven.

Zoals beschreven in hoofdstuk 5, gebruikten we de volgende criteria bij de review: ernstige CP (GMFCS IV en V), aangetoonde subluxatie of dislocatie van de heup (gemeten door middel van het migratiepercentage [MP]), de aard van de chirurgische ingreep, en een verminderde MP en afname van pijn als uitkomstmaten. Er werd een kwalitatieve analyse volgens Steultjens et al.[1] en Van Tulder et al.[2] uitgevoerd. We gebruikten de techniek van de best evidence synthese om de resultaten van de geïncludeerde artikelen weer te geven. Alle onderzoeken waren observationeel.

We vonden artikelen over resectiechirurgie van de heupkop, arthrodese van de heup en totale heupprothese. De door ons gebruikte techniek van de best evidence synthese

gaf als resultaat, dat er indicaties zijn dat pijn, zit- en verzorgingsproblemen kunnen worden verholpen door een palliatieve resectie van de femurkop. Helaas is er geen palliatieve ingreep voorhanden, die ondubbelzinnig de garantie oplevert, dat patiënten met niet op een andere manier te behandelen pijn hun pijn zullen kwijtraken. De techniek van de subtrochantere kopresectie is echter de techniek waarover de meeste resultaten zijn gepubliceerd en waarvan de beste effecten zijn beschreven. Ons oordeel is dat, indien de femurkopresectie wordt gekozen, de distale resectie- en interpositie-operatie volgens Castle en Schneider[3] de methode bij uitstek in deze gevallen is. Het percentage pijnverlichting na deze interventie is 90–100%. Het effect van dit soort botchirurgie kan echter nog wel eens op zich laten wachten; het kan tot een half jaar duren voordat pijnreductie is bereikt.

Over de techniek van valgiserende derotatie-osteotomie zoals beschreven door Schanz, die wordt uitgevoerd door een aantal orthopedisch chirurgen, vonden we geen literatuur die voldoet aan onze kwaliteitscriteria. Eén auteur meldt 98% verlichting van de pijn maar een ander beschrijft een complicatiepercentage van 63% bij deze techniek.

Over totale heupvervanging respectievelijk arthrodese van de heup vonden we slechts één artikel dat paste binnen onze kwaliteitscriteria. Deze technieken kunnen pijnreductie opleveren, maar kunnen ook leiden tot een relatief groot aantal complicaties. Na plaatsing van een totale heupprothese, komt dislocatie van de prothese voor bij 29% van de gevallen, heterotope ossificatie (HO) is aanwezig in 45%. Bovendien zijn femurfracturen relatief frequent (36%). Verder kan arthrodese van het heupgewricht resulteren in een pijnvrije heup, maar ook hier is de kans op complicaties, zoals non-union hoog (42%).

Botchirurgie kan verlichting van pijn en ongemak als palliatieve ingreep bij patiënten met CP en heupafwijkingen opleveren. Helaas is heterotope botvorming een groot probleem als complicatie bij deze patiënten. Onderzoek gericht op het voorkomen van dit fenomeen in de toekomst zou nut hebben.

Hoofdstuk 7 beschrijft de synthese van de resultaten van het onderzoek zoals beschreven in de hoofdstukken 2–4 en de reviews, zoals beschreven in hoofdstuk 5 en 6. Methoden om subluxatie te meten en de pathofysiologie en de natuurlijke historie van de heupafwijkingen worden beschreven. Methoden van interventie worden besproken en op waarde geschat. De vraag of een operatie symmetrisch moet worden uitgevoerd en de juiste timing van operaties worden besproken. Voor- en nadelen van een operatie om

heupafwijking te voorkomen of te behandelen worden besproken en de rol van botulinum toxine wordt becommentarieerd. We bespreken de verschillende manieren van het monitoren van heupafwijkingen en de daaruit voortvloeiende maatregelen ter preventie of behandeling. Het geringe risico van frequent röntgenonderzoek wordt besproken.

Dit alles cumuleert in een voorstel voor een **beslisboom** voor de monitoring en interventie van heupafwijkingen bij spastische CP. Het algoritme is gebaseerd op ons onderzoek en op de literatuur in onze reviews. We noemen het de Nederlandse consensus welke is besproken in de algemene vergaderingen van de Nederlandse Kinderrevalidatieartsen, onderdeel van de Vereniging van Revalidatieartsen (VRA). We vergelijken het algoritme met de Australische consensus over surveillance, zoals gepubliceerd in 2008.

De beslisboom heeft het GMFCS-niveau als basis voor de interventie en is verdeeld in drie fasen op basis van leeftijd van de patiënt. Ten eerste: leeftijd tussen 1 en 4 jaar waarin een dubbelzijdige adductorentenotomie de voorkeursbehandeling is als een (dreigende) subluxatie van de heup wordt gediagnosticeerd. Ten tweede, de leeftijd tussen 8 en 9 jaar, waarin patiënten met heupafwijking kunnen worden behandeld met een VDRO, eventueel gecombineerd met een bekkenosteotomie. Ten derde: de periode na de leeftijd van 9 jaar, waarin een palliatieve operatie een patiënt met niet op een andere manier behandelbare pijn of beperkingen soelaas kan bieden.

We gebruiken het cerebral palsy hip classification system (CPHCS)-systeem om te bepalen of een interventie nodig is. We denken dat een adductorentenotomie zinvol kan zijn bij elk GMFCS-niveau, op voorwaarde dat de ouders wordt uitgelegd wat de reden is van een ingreep en de kans op succes wordt besproken. Botchirurgie moet worden gereserveerd voor patiënten met een GMFCS niveau I–III, en slechts bij uitzondering worden toegepast bij patiënten met een hoger GMFCS-niveau.

Hoofdstuk 8 bevat de algemene discussie. Een patiëntencasus wordt gepresenteerd en de belangrijkste punten van het onderzoek en de reviews herhaald en toegepast op het patiëntprobleem. We relativeren het resultaat van ons onderzoek: er werd een prevalentie-onderzoek uitgevoerd en geen prospectief onderzoek. Slechts een klein aantal artikelen in de reviews voldeed aan onze kwaliteitscriteria, waardoor we genoodzaakt waren de 'best evidence' methode toe te passen om conclusies te kunnen trekken.

Belangrijkste conclusie: heupafwijkingen bij spastische CP veroorzaken pijn, leveren beperkingen op en moeten zo mogelijk worden voorkomen of behandeld. Echter niet

tot elke prijs: zorgvuldige beoordeling, evaluatie en discussie zijn nodig bij iedere individuele patiënt en deze dienen te worden uitgevoerd door een ervaren team van professionals, met affiniteit voor de doelgroep.

De algemene discussie bevat ook enkele aanbevelingen voor toekomstig onderzoek. Er wordt een voorstel gedaan voor verder onderzoek naar de waarde en effectiviteit van weke delen chirurgie bij jonge CP-kinderen met een hoge GMFCS-classificatie. Een pleidooi wordt gehouden voor het goed inrichten van epidemiologisch onderzoek naar incidentie en prevalentie van CP in Nederland en de effecten van interventie, naar voorbeeld van Australië en Zweden. Een dergelijk systeem zou het mogelijk maken de waarde te bepalen van de concentratie van chirurgische interventie bij CP-kinderen en -volwassenen in enkele gespecialiseerde centra.

Daarnaast wordt een pleidooi gehouden voor verder onderzoek naar de waarde van het passief doorbewegen van de heupen en knieën bij CP-patiënten, aangezien uit ons onderzoek blijkt dat dit een zeer pijnlijke handeling is voor de patiënten.

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Dankwoord (Acknowledgements)

In de ruim 21 jaar die dit onderzoek geduurd heeft hebben meerdere mensen me geholpen om het onderzoek met goed resultaat te kunnen afronden. Ik zal de geschiedenis chronologisch doornemen en kom daarbij vanzelf op de namen van hen, die hebben geholpen om het onderzoek tot een succes te maken. Ik dank hen allen voor de steun die ze me gaven.

Zoals in hoofdstuk 1 aangegeven was het mijn leermeester en opleider, Hans Jongbloed, die me in aanraking bracht met het probleem van de heupluxatie bij spastische kinderen. Het was gedurende mijn opleidingstijd in het Academisch Ziekenhuis Leiden. In mijn opleidingstijd in Leiden was voldoende tijd voor wetenschappelijk werk, zoals past bij een academische setting. Ik had de gelegenheid om onderzoek te doen in het bewegingslaboratorium in Leiden op het terrein van de oude Anna Kliniek, samen met Daan Wijkmans en Teun van Pijkeren. Dit leidde tot enkele publicaties van mijn hand, onder andere over gangbeeld, innovatieve orthesen en schoenen en over prothesiologie. De details zijn weergegeven in de lijst met publicaties.

Toen ik in 1982 specialist werd in het Rijnlands Revalidatie Centrum in Katwijk aan Zee kreeg ik van de directie gelegenheid om wetenschappelijk onderzoek te doen, gedurende een halve dag per week. Aanvankelijk kwam daar niet veel van, maar in 1991 vond ik dat het er maar eens van moest komen. Een onderwerp had zich aangediend via de fysiotherapeut van het voorzieningencentrum, Peter Groot, zoals beschreven in hoofdstuk 1. Hélène van der Heijden, toen assistent revalidatie, hielp me met de opzet en biedt tot heden grote steun bij de voortgang van het onderzoek en de daarbij behorende publicaties. Fijn dat ze paranimf wil zijn bij de promotie.

Begin 1992 sprak ik met professor Guus Lankhorst over de onderzoeksplannen en hij constateerde met mij dat het niet eenvoudig was om tot een goede onderzoeksopzet te komen. Hij raadde me aan de cursus epidemiologie te volgen bij professor Lex Bouter, toen hoogleraar epidemiologie aan het EMGO, onderdeel van VUMC. Samen met Hélène volgde ik in juni 1993 de cursus epidemiologisch onderzoek van Lex Bouter in Rolduc. Een betere cursus heb ik daarna niet meer gevolgd! Professor Bouter nam de tijd om met mij tijdens de cursus de conceptopzet te bespreken en we kwamen tot de conclusie dat een puntprevalentieonderzoek geschikt was, met als onafhankelijke variabelen de bevindingen op de röntgenfoto en als afhankelijke variabelen de metingen aan de patiënten.

De cursus in Rolduc leverde nog meer op: medecursist Suzanne Arts deed me het idee aan de hand om een pijnschaal met gezichten, ontworpen voor kinderen voor het onderzoek te gebruiken. We bespraken enkele malen de onderzoeksopzet met Hélène van der Heijden en collega Peter de Koning in het huis van Peter aan de Lange Mare in Leiden.

Intussen was ik in 1993 van werkplek veranderd naar Heliomare en medisch directeur Arie Prevo bood me de faciliteit om het onderzoek uit te voeren en af te ronden voor een halve dag per week gedurende drie jaar. Die drie jaar liep helaas een beetje uit…

Toen ik in Heliomare in 1997 lid van de Raad van Bestuur werd luisterde de Raad van Toezicht met welwillende belangstelling naar mijn plannen en verlengde de onderzoeksfaciliteit. Het komt niet iedere dag voor dat een revalidatiearts van je centrum promoveert.

Manager Frans Broekhuizen van Revalidatiecentrum Katwijk bood me ook na mijn vertrek als revalidatiearts uit het centrum de faciliteit om eenmaal per week gebruik te maken van een kamer in het Zeehospitium om ongestoord en zonder internet aan het onderzoek te werken, een faciliteit die het centrum me tot de huidige dag biedt. Ik geniet er nu de gastvrijheid van AVG-arts Olga Hutten.

Ik verdiepte me in de literatuur over de gezichtsherkenning en pijnregistratie bij kinderen en meervoudig gehandicapten, daarbij geholpen door logopediste Marian Fransen. We bedachten dat er tekeningen moesten komen van dagelijks voorkomende situaties, zowel pijnlijk als niet pijnlijk, en daarnaast heupbelastende situaties ter vergelijking. Anika de Ridder-van Leeuwen maakte de tekeningen, die we op willekeurige volgorde legden door ze op de grond te gooien en vervolgens weer op te rapen…

We voerden een pilotonderzoek uit bij de gezonde kinderen van Marleen Eberwijn, Cate Bos, Ans Teeuwisse en Marjorie Olde Monnikhof en deden een proefonderzoek bij enkele bewoners van woonvorm Voorzieningencentrum van het Rijnlands Zeehospitium in Katwijk. Het resultaat was veelbelovend en een definitieve onderzoeksopzet kon worden voorgelegd aan de begeleidende hoogleraren en vervolgens aan de potentiële financiers.

Op 18 januari 1995 had ik overleg met Guus Lankhorst en Lex Bouter en werd de opzet van het onderzoek door hen goedgekeurd. Een subsidieaanvraag werd in het begin van 1996 ingestuurd en ik kreeg subsidie van het Johanna Kinderfonds (toen Fonds

Johanna Stichting geheten), de Phelps Stichting en het Anna Fonds. In december 1996 had ik een overleg met Michael Rutgers, directeur van het Fonds Johanna Stichting, die zich positief uitliet over de onderzoeksopzet. Inderdaad verleende het fonds begin 1997 de gevraagde subsidie.

In de zomer van 1997 kon na een open sollicitatieronde Monique Jacobs-van der Bruggen worden aangesteld als onderzoeksmedewerker / research fysiotherapeut in dienst van Heliomare. Zij deed vanaf september 1997 de metingen bij de proefpersonen in het gehele land en deed vervolgens mee aan de analyses en de publicatie van de resultaten. De automatiseringsafdeling van Heliomare zorgde voor een adequate laptop.

Ik ben veel dank verschuldigd aan alle bijna 200 proefpersonen en potentiële proefpersonen, met hun artsen, paramedici en verzorgers, die geheel belangeloos meewerkten aan het onderzoek. Hun medewerking heeft geleid tot meer inzicht in hun complexe problematiek, die helaas onderbelicht is in de Nederlandse gezondheidszorg. Hopelijk draagt dit proefschrift bij tot meer aandacht voor de problemen waarmee deze mensen te kampen hebben. Ik heb voor hen een nieuwsbrief gemaakt met een samenvatting van de resultaten.

De resultaten van het onderzoek werden in het voorjaar van 2000 mede geanalyseerd door Joop Kuik, statisticus in het VUmc. Hij bedacht de formule om onderscheid te maken tussen *wel* pijn in de heup en *geen* pijn in de heup, door dit te relateren aan in het algemeen pijnlijke en niet pijnlijke situaties.

Na de publicatie van de eerste drie artikelen deed zich in de tweede helft van 2006 de vraag voor hoe het onderzoek kon worden afgerond. Het uitvoeren van een systematic review lag voor de hand, maar de manier waarop dat moest gebeuren en door wie was een discussiepunt. In juni 2007 bood Carola Bouwhuis, toen net gestart als AIOS revalidatiegeneeskunde, haar hulp aan. Ze is gepromoveerd op een kindergeneeskundig onderwerp en was bereid om, mede in het kader van haar opleiding, mee te denken en mee te werken aan de benodigde systematic reviews. Samen met Hélène van der Heijden las ze veel artikelen en schreef ze de opzet voor hoofdstuk 5, waarvan ze eerste auteur werd. Heleen Beckerman, staflid van het VUmc, dacht op verzoek van Guus Lankhorst mee over de opzet van de systematic reviews.

Vanaf september 1999 verplaatste ik mijn werkzaamheden naar Sophia Revalidatie in Den Haag, waar ik parttime lid werd van de Raad van Bestuur en revalidatiearts in het Haags Tyltylcentrum de Witte Vogel.

Ook in Sophia Revalidatie verleende de Raad van Commissarissen me faciliteiten om het onderzoek voort te zetten en af te ronden. Met de gezamenlijke gedachte dat het onderzoek rond 2002 af te ronden zou moeten zijn, werd me een dag per week de gelegenheid gegeven onderzoek te doen. In de loop der tijd bleken de werkzaamheden als RvB-lid en revalidatiearts echter zo tijdrovend, dat ik na overleg met - en enigszins op aandringen van – collega RvB-lid Hans Borgsteede de tijdsbesteding aan het onderzoek terugdraaide naar eenmaal per twee weken een dag, wat in de praktijk meestal neerkwam op eenmaal per twee weken een halve dag.

De Raad van Commissarissen informeerde regelmatig en langzamerhand enigszins besmuikt naar de voortgang van het onderzoek, met name via raadslid Harm Garvelink, maar verbond geen negatieve consequentie aan mijn geruststellende woorden.

Regelmatig werd de voortgang van het onderzoek besproken in de sectie kinderrevalidatie van de VRA, voorheen kinderplatform genoemd. De collega's dachten intensief mee en gaven er blijk van blij te zijn met richtlijnen voor de behandeling van heupluxatie bij CP in de vorm van een stroomschema, ook al werd de inhoud daarvan regelmatig bijgesteld. Vooral professor Jules Becher viel – als hij er was – op door zijn betrokken en deskundige commentaar. De voordrachten leverden maar liefst drie kinderplatformmokken op…

Pas na het beëindigen van het lidmaatschap van de RvB in september 2011 kwam er schot in de zaak, omdat ik naast het werk als revalidatiearts voor 0.5 FTE meer vrije tijd kreeg om het onderzoek af te ronden.

Gedurende de loop van het onderzoek en de publicaties kreeg ik veel steun van orthopedisch chirurg Kees Bos. Hij was toen ik hem leerde kennen rond 1991 staflid kinderorthopedie in het LUMC en consulent orthopedie in het Rijnlands revalidatiecentrum, als opvolger van Ab Verbout. Later werkte ik met hem samen in Den Haag, waar hij werkzaam was in het Juliana Kinderziekenhuis tot zijn pensionering in december 2009. Hij werd mede-auteur van alle publicaties en dacht steeds actief mee. Zijn opvolger Hubert Oostenbroek zette die goede traditie in de dagelijkse praktijk van de patiëntenzorg voort, samen met zijn collega Femke van Erp Taalman Kip.

De correctie van de tekst op vloeiend Engels werd uitgevoerd door de heer Jan Klerkx, die niet alleen lette op de taalproblemen, maar ook wees op tegenstrijdigheden, onlogische formuleringen, etcetera.

De ruwe tekst vanuit Word werd omgevormd tot een mooi boekje door Renate Siebes, die uitzonderlijke kwaliteit van werk koppelde aan een buitengewoon tempo van werken. Het omslagontwerp werd gemaakt door Anika de Ridder-van Leeuwen, de beslisboom werd drukklaar gemaakt met hulp van Anne Kouwenhoven.

Toen in maart 2012 eindelijk zicht kwam op voltooiing van dit proefschrift werd de leescommissie aangezocht, bestaande uit prof. dr. O.F. Brouwer, dr. C.G.B. Maathuis, dr. H.J. Pruijs, dr. J.A. van der Sluijs en prof. dr. H.C.W. de Vet. Ook prof. dr. J.G. Becher maakt deel uit van de promotiecommissie. Met bijna ieder van hen had ik in de loop der jaren een inspirerend contact over de intrigerende wereld van de heupluxatie bij CP-patiënten.

Promovendi hebben nogal eens de gewoonte hun partner te bedanken voor de gederfde tijd die ze samen hadden kunnen doorbrengen als de promovendus niet zoveel tijd aan het onderzoek zou hebben besteed. In mijn geval gaat die vlieger niet op. Ook mijn dochters Maartje en Anne hebben er niet onder geleden.

Tot september 2011 werkte ik slechts gemiddeld eenmaal per twee weken aan het onderzoek. Lex Bouter zei niet voor niets in 1995 dat hij niet verwachtte dat ik met deze tijdsbesteding ooit zou kunnen promoveren. Pas na september 2011 ging een deel van mijn vrije tijd op aan het onderzoek.

Het was een leuk onderzoek, en het sloot aan op mijn praktijk. Ik zeg altijd: promotieonderzoek moet een vraagstelling hebben waar ze je 's nachts voor kunnen wakker maken. In mijn geval was dat zo. Het is een onderwerp dat je niet loslaat.

Iedereen bedankt voor de fijne samenwerking !

Curriculum vitae

Eric Boldingh was born in Den Helder on 8 September 1951. He attended primary school in Sassenheim and Heerenveen, and secondary school in Heerenveen and Oosterwolde (Friesland). Between 1968 and 1975, he studied medicine at the Vrije Universiteit in Amsterdam.

In 1975 and 1976, he trained to become a general practitioner in Bennebroek and Leiderdorp. Between 1976 and 1978, he performed his military service in the Dutch Royal Navy, at the outpatient rehabilitation clinic in Driehuis Westerveld.

Between 1978 and 1982, he was a resident of rehabilitation medicine at the rehabilitation center in Katwijk (Zuid-Holland) and at the university hospital in Leiden.

From 1982 until 1987, he worked as a physiatrist treating adults at the rehabilitation center in Katwijk, and from 1987 until 1993 he was a physiatrist for pediatric rehabilitation at the same center. He started the research for this thesis in 1991.

In 1993, he transferred to Rehabilitation Center Heliomare in Wijk aan Zee, to work as a physiatrist for pediatric rehabilitation there. From 1997, he also was CEO for patient care at Heliomare.

In 1999 he moved to Sophia Rehabilitation Center in The Hague to become CEO for patient care there (until September 2011), which he combined with working as a physiatrist for pediatric rehabilitation and patients with multiple handicaps, which he has continued until the present. About 30% of his patients are CP patients, most of them with GMFCS levels IV-V.

Eric has been married to Riekje de Kloet since 1976, and has two daughters, Maartje (1977) and Anne (1979), and four grandchildren, Fenna (2005), Jens (2008), Thijmen (2010) and Laurens (2012). He has lived in Sassenheim since 1978.

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