Prevention of severe contractures might replace multilevel surgery in cerebral palsy: results of a population-based health care programme and new techniques to reduce spasticity

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During the 1990s three new techniques to reduce spasticity and dystonia in children with cerebral palsy (CP) were introduced in southern Sweden: selective dorsal rhizotomy, continuous intrathecal baclofen infusion and botulinum toxin treatment. In 1994 a CP register and a health care programme, aimed to prevent hip dislocation and severe contractures, were initiated in the area. The total population of children with CP born 1990–1991, 1992–1993 and 1994–1995 was evaluated and compared at 8 years of age. In non-ambulant children the passive range of motion in hip, knee and ankle improved significantly from the first to the later age groups. Ambulant children had similar range of motion in the three age groups, with almost no severe contractures. The proportion of children treated with orthopaedic surgery for contracture or skeletal torsion deformity decreased from 40 to 15% ($P=0.0019$). One-fifth of the children with spastic diplegia had been treated with selective dorsal rhizotomy. One-third of the children born 1994–1995 had been treated with botulinum toxin before 8 years of age. With early treatment of spasticity, early non-operative treatment of contracture and prevention of hip dislocation, the need for orthopaedic surgery for contracture or torsion deformity is reduced, and the need for multilevel procedures seems to be eliminated. *J Pediatr Orthop B* 14:269–273 © 2005 Lippincott Williams & Wilkins.

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Introduction

Children with spastic or dystonic types of cerebral palsy (CP) have a risk of developing contractures and secondary skeletal torsion deformities [1–3]. If prevention and non-operative treatment fail, orthopaedic surgery is often indicated.

A child operated on with muscle or tendon lengthening at a young age has an increased risk for relapse and need for repeated operations [1,4]. Children operated on when older have an increased risk for developing more severe contractures and more skeletal deformities during the 'waiting time' for surgery [5].

In the 1990s three new methods to reduce spasticity were introduced: selective dorsal rhizotomy (SDR), continuous intrathecal baclofen infusion (ITB) and botulinum toxin treatment. These techniques made it easier to prevent development of severe contracture, by reducing the muscle tone.

We have analysed the first 10 years result of a health care programme for children with CP, aimed at reducing severe contractures by early treatment of spasticity and early, preferably non-operative, treatment of contractures.

Materials and methods

In 1994, a CP register and a health care programme for children with CP in southern Sweden was initiated, comprising all children with CP born in the area since 1990. The goals are to prevent hip dislocation and severe contractures. The study area (the counties of Skåne and Blekinge) has a population of 1.3 million inhabitants. The total population born in 1990 and later was systematically searched in 1998 and 2002 to find all children with probable CP. In children 4 years of age or older the CP-diagnosis and subtypes were established. The prevalence of CP in children 4–7 years of age living in the area on 1 January 1998 was 2.4 per 1000 children [6] and their gross motor function and disabilities were studied [7].

CP was defined as 'an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of development' [8]. Only children who lived to 2 years of age and had their brain

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lesion before that age were included. The CP-subtype was determined after the fourth birthday according to Hagberg *et al.* [9]. The gross motor function was classified into five levels according to the gross motor function classification system (GMFCS) [10], which is an age-related five-level system, in which level I is the most and level V the least independent.

The health care programme includes a continuing standardized follow-up of each child with a recording form. The child's local physiotherapist and occupational therapist fill in the recording form twice a year until 6 years of age, then once a year. The recording form includes the CP-subtype, GMFCS, measurements of passive range of motion (ROM) with a goniometer in stated and standardized positions and joint angles. In addition gross motor function, clinical findings, use of orthoses and treatment are recorded. The results are computerized, and the local team receives a report showing the child's development over time. The programme also includes a standardized radiographic follow-up of the children's hips [11]. The hip programme includes the children born 1992 or later.

In 1992 the first child was operated with ITB, in 1993 the first child was operated with SDR, and in 1998 treatment with botulinum toxin began.

In the present study the children born 1990–1995 have been analysed at 8 years of age. In this period, 221 children born in the area developed CP. Only those children born in the area and those who had moved into the area before 2 years of age ($n = 16$), and were still living in the area at 8 years of age and participating in the follow-up programme were included in the study ($n = 209$) (Table 1). The material was divided according to year of birth (1990–1991, 1992–1993 and 1994–1995). Children born 1990–1991 were not included in the hip programme. Children born 1992–1993 were 2–3 years of age when the health care programme started, and 5–6 years old when treatment with botulinum toxin began. Children born 1994–1995 were included in the health care programme as soon as possible CP was noted, and they were 3–4 years of age when treatment with botulinum toxin began.

The number of children related to CP-subtype and the gross motor function were similar in the three groups (Table 2). In the age group born 1992–1993, there was a higher proportion of children with dyskinetic CP.

We analysed the passive range of motion in hip, knee and foot for these children at 8 years of age, and the treatment for spasticity and orthopaedic operations performed up to 8 years of age. Only orthopaedic operations on the lower extremities were included. In addition, the operations performed between 8 and 12 years of age were recorded in the children born 1990–1991.

Fishers exact test was used to compare the number of operations in the three age groups. The Mann–Whitney test was used to compare the ROM between the three age groups.

Results

In children with independent or dependent walking ability (GMFCS 1–3), the passive ROM was almost equal at 8 years of age in the three age groups, both regarding the median and the – 90 percentile values (Table 3). Of the 157 children, 154 could extend their hips to at least -10°, 156 had knee extension to at least -10°, and 153 could extend their feet dorsally to at least a neutral position.

In children without walking capacity (GMFCS 4–5), the ROM was lower in children born 1990–1991 and improved in the later age groups (Table 4). Improvement was seen both regarding median and the – 90 percentile values. For children born 1994–1995 the ROM was almost similar to children with walking capacity regarding hip and foot motion, while the – 90 percentile values for knee extension and popliteal angle was better in the children with walking capacity.

The 23 children not participating in the follow-up programme did not differ from the participants according to gender and CP-subtype.

There has been a marked change in treatment during the study period (Table 5). Treatment of spasticity with SDR and botulinum toxin has increased. Orthopaedic surgery for contracture, rotational deformity, foot deformity and salvage operations for dislocated hips have decreased from 40 to 15% ($P = 0.0019$) (Fig. 1). Of the 41 children operated for contracture, 32 had a single

Table 1 The number of children with cerebral palsy in the total population, and in the study groups

	Born 1990–1991	Born 1992–1993	Born 1994–1995	Total
Born in the area 1990–1995	74	77	70	221
Moved into the area before 2 years of age	6	7	3	16
Moved out from the area before 8 years of age	1	1	1	3
Died before 8 years of age	1	0	1	2
Not participating in health care programme	9	8	6	23
Number of children in present study	69	75	65	209

Table 2 Sub-diagnosis and gross motor function classification system (GMFCS)-score in 209 children with cerebral palsy, born 1990–1995

Subdiagnosis	Born 1990–1991 ($n=69$)	Born 1992–1993 ($n=75$)	Born 1994–1995 ($n=65$)	Total ($n=209$)
Spastic				
Hemiplegic	21	25	23	69
Tetraplegic	7	10	3	20
Diplegia	32	28	31	91
Ataxic	4	2	5	11
Dyskinetic	5	10	3	18
GMFCS				
Level I	29	32	35	96
Level II	16	10	9	35
Level III	11	10	5	26
Level IV	6	9	12	27
Level V	7	14	4	25

Table 3 Range of motion (−90 percentile, median) at 8 years of age in children with cerbral palsy with walking capacity [gross motor function classification system (GMFCS) 1–3] in southern Sweden: comparison between children born 1990–1991, 1992–1993 and 1994–1995

	Born 1990–1991 ($n=56$)		Born 1992–1993 ($n=52$)			Born 1994–1995 ($n=49$)		
	−90%	Median	−90%	Median	*P*	−90%	Median	*P*
Hip extension	0	0	0	0	0.32	0	0	0.013
Hip abduction	30	40	29	40	0.48	30	40	0.92
Hip external rotation	30	40	25	35	0.32	30	40	0.013
Hip internal rotation	40	55	35	50	0.58	40	55	0.59
Popliteal angle	132	150	130	150	0.59	130	145	0.46
Knee extension	−3	0	0	0	0.37	0	0	0.014
Foot dorsiflexion extended knee	0	10	0	10	0.63	5	10	0.64
Foot dorsiflexion flexed knee	1	20	10	20	0.81	10	20	0.62

P value for comparison with children born 1990–1991. For each child the lowest value of the left and right leg was used.

Table 4 Range of motion (−90 percentile, median) at 8 years of age in children with cerebral palsy without walking capacity [gross motor function classification system (GMFCS) 4–5] in southern Sweden: comparison between children born 1990–1991, 1992–1993 and 1994–1995

	Born 1990–1991 ($n=13$)		Born 1992–1993 ($n=23$)			Born 1994–1995 ($n=16$)		
	−90%	Median	−90%	Median	*P*	−90%	Median	*P*
Hip extension	−19	−10	−10	0	0.006	0	0	0.001
Hip abduction	8	20	16	33	0.01	23	30	0.009
Hip external rotation	20	35	20	45	0.06	28	48	0.001
Hip internal rotation	25	55	30	40	0.92	26	60	0.51
Popliteal angle	115	130	120	135	0.25	125	143	0.028
Knee extension	−38	−15	−25	−5	0.34	−22	0	0.11
Foot dorsiflexion extended knee	−9	0	−5	10	0.23	0	10	0.20
Foot dorsiflexion flexed knee	0	15	10	20	0.13	13	23	0.083

P value for comparison with children born 1990–1991. For each child the lowest value of the left and right leg was used.

Achilles–gastrocnemius lengthening (in some cases combined with lengthening of the tibialis posterior tendon), five had a single lengthening of the medial hamstring muscles, and four children were operated with combined Achilles–gastrocnemius and hamstring lengthening. As a consequence of the hip prevention programme, the number of operations to prevent hip dislocation has increased from 9 to 17% ($P = 0.20$).

In the 69 children born 1990–1991, a further 19 orthopaedic operations were performed between 8 and 12 years of age; four operations to prevent hip dislocation, two salvage operations for painful hip dislocation, one osteotomy for torsion deformity, five operations for foot deformity, and seven operations for contracture. The operations for contracture were a single Achilles–gastrocnemius lengthening in all cases.

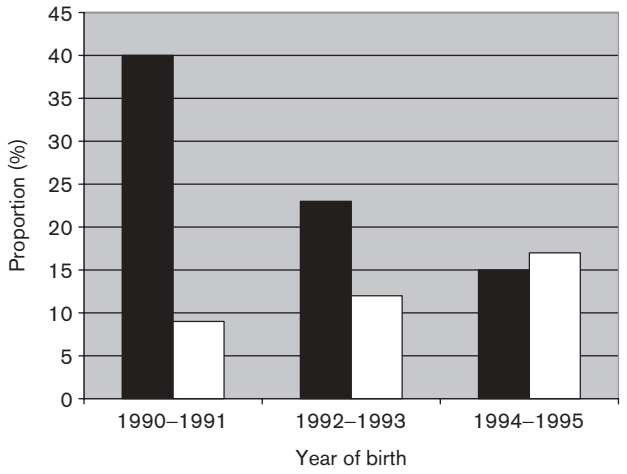
Discussion

This report is based, to our knowledge, on the first prospective follow-up study of a health care programme of a total population of children with CP.

Table 5 Number of children treated with selective dorsal rhizotomy, intrathecal baclofen infusion, botulinum toxin and orthopaedic surgery at 8 years of age: comparison between children born 1990–1991, 1992–1993 and 1994–1995

	Born 1990–1991 ($n=69$)	Born 1992–1993 ($n=75$)	Born 1994–1995 ($n=65$)
Selective dorsal rhizotomy	3	7	8
Intrathecal baclofen infusion	1	2	1
Botulinumtoxin	1	6	24
Orthopaedic surgery			
Salvage for dislocated hip	2	0	0
Osteotomy–torsion deformity	3	0	0
Foot deformity	5	3	1
Contracture	18	14	9
Hip surgery to prevent dislocation	6	9	11

Fig. 1



Proportion of children with cerebral palsy treated with orthopaedic surgery before 8 years of age. Comparison between children born 1990–1991, 1992–1993 and 1994–1995. ■, Operations for deformity, contracture or salvage operation for dislocated hip; □, Hip surgery to prevent dislocation.

Growth in length of a muscle-tendon is stimulated by the growth in length of the skeleton and the muscle excursion [12]. Spasticity results in reduced muscle excursion leading to failure of muscle growth, with contracture seen as restricted joint range [1]. Reduced muscle excursion due to muscle weakness, inability to stand or walk also contributes to contracture development. If untreated, a contracture often results in restricted muscle excursion in adjacent joints, for example a knee flexion deformity results in flexion deformity of the hip and equinus deformity of the ankle joint. Spasticity and contracture also influence the skeletal growth, with development of torsion deformities giving the child a more and more complex musculoskeletal problem to treat.

Non-operative methods to prevent or treat contractures includes physical therapy with range of motion and stretching exercises, use of orthoses and standing device, serial casting (sometimes in combination with botulinum toxin). Non-operative treatment of contracture should be initiated as early as possible [1].

Operative treatment includes muscle–tendon lengthenings or skeletal procedures. A short muscle has a short active and passive excursion. Lengthening results in transition, but often not increase, of the active excursion, the increased ROM is only passive [1]. A lengthening also often results in weakening of the muscle [1,5,13].

Operation with muscle–tendon lengthening at an early age is known to increase the risk for recurrence and need for repeat surgery [4,5]. This is probably due to either that not all contractures being treated at first operation or high muscle tone making postoperative prevention of recurrence impossible. Several authors advocate delay of operation to decrease the need for repeat surgery, and to preoperatively be able to analyse the child's 'adult' gait pattern [1,2]. Consequently muscle–tendon surgery is often regarded ideal at age 6–8 years of age [1,2,5]. The drawback with delayed treatment of contracture is the risk of deformity spread to adjacent joints and to the skeleton during the 'waiting-time' for surgery. Sometimes an instrumented gait analysis is needed to rule out all problems, and the result is a multilevel procedure [14,15]. It is also more difficult to treat the child with established contractures during the years of waiting for surgery.

The new techniques to reduce spasticity, SDR, ITB, and botulinum toxin, give a new possibility to prevent and treat contractures. In the 1994–1995 group, one-third of the children have had treatment with botulinum toxin before 8 years of age. Among children with spastic diplegia, one-fifth have been treated with SDR. These children with severe spasticity are probably those who previously would have developed severe contracture with need for orthopaedic multilevel surgery.

Decision on operative treatment is not based on impairment of ROM, but on functional limitations in activities of daily living such as sitting, standing and walking. However, if a child does not have a contracture, and if severe spasticity is treated by methods reducing

the muscle tone, there is often no need for orthopaedic surgery to improve function. The development of torsion deformity is also decreased. Orthopaedic surgery is still important for prevention of hip dislocation, and to correct and stabilize scoliosis and foot deformity.

In the present material, no child has been treated with multilevel surgery, and at 8 years of age there is no child with problems indicating need for multilevel surgery. A small percentage had minor contractures at 8 years of age. These are most often single joint contractures that are detected early by the recording form, and treated non-operatively, often by intensified physiotherapy, serial casting or orthoses. The children born 1990–1991 are at present 12–13 years of age, and only seven further orthopaedic operations for contracture, all Achilles–gastrocnemius lengthening, have been performed from age 8–12 years among these children.

For early detection and treatment in a population a CP register and a standardized follow-up programme is needed. The main challenge to run this population-based follow-up programme is to early identify all children with CP in the population [6]. The follow-up programme has been developed in collaboration with all local team members (paediatricians, physiotherapists, occupational therapists, orthotists, orthopaedic surgeons) in the area, and all interventions have the support of both the local team and the paediatric orthopaedic surgeon.

In conclusion, with the new techniques to reduce spasticity and with a population-based screening programme it seems possible to prevent development of severe contractures in children with CP, reducing or eliminating the need for multilevel procedures. Our strategy for treatment involves early treatment of spasticity with the new techniques, SDR, ITB and botulinum toxin; early detection and treatment of contractures; early detection and treatment of lateral migration of hips.

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