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ORIGINAL ARTICLE

Scoliosis in children with severe cerebral palsy: a population-based study of 206 children at GMFCS levels III–V

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Abstract

Purpose To evaluate the prevalence of scoliosis and the rate of scoliosis progression in children with severe cerebral palsy (CP) at GMFCS levels III–V.

Methods Two hundred and six children (86 girls, 120 boys), born 2002–2008, were recruited from The Norwegian Quality and Surveillance Registry for Cerebral Palsy (NorCP). Inclusion criteria were bilateral CP and GMFCS levels III–V. Scoliosis was evaluated annually by examination of the spine by a physiotherapist. Radiographic examination was performed in children with moderate or severe scoliosis at clinical evaluation. The Cobb angle was used as a measure of curve magnitude.

Results Scoliosis, defined as Cobb angle ≥ 10°, occurred in 121 children (59%). Severe scoliosis (Cobb angle ≥ 40°) developed in 80 of the 206 patients (39%) at a mean age of 10.9 years (range 5–16) and was more prevalent in children at GMFCS level V (62%) than at levels IV (19%) and III (6%). Initial Cobb angle, Cobb angle ≥ 30° at age 10 years, and GMFCS level V were independent risk factors for severe scoliosis. In children at GMFCS level V, the rate of scoliosis progression decreased with age from a mean of 9.7° per year at age 3–5 years to 2–3° per year in children ≥ 11 years.

Conclusions The prevalence of scoliosis among children with CP increased with decreasing motor functional level. The most important risk factors for progression of scoliosis were high initial Cobb angle, Cobb angle ≥ 30° at age 10 years, and GMFCS level V.

Keywords Scoliosis · Cerebral palsy · Natural history · Gross motor function classification system · Cobb angle

Introduction

Children with cerebral palsy (CP) are at increased risk of scoliosis, especially those with the greatest degrees of disability. The estimated prevalence of scoliosis ranges from 25 to 64% in institutionalized patients [1–3]. According to a systematic review [4], this wide variability probably reflects differences in the definitions of scoliosis, functional levels of the patients, types of CP, and age groups. To provide more precise and reliable data on the prevalence of scoliosis, population-based studies that include defined subgroups are required, but so far, only a few such studies have been published [5, 6].

Severe scoliosis in patients with CP often leads to additional motor dysfunction, compromised pulmonary function, decubitus ulcers, and increased nursing demands [1, 7]. Early detection of scoliosis and systematic follow-up are needed to avoid or reduce the risk of these problems. Screening for scoliosis has been incorporated in the population-based Swedish and Australian CP registers [5, 6] and has also been adopted in the population-based surveillance programme that was used in the present study.

A systematic review of progression of scoliosis in severe CP, published before population-based studies were available [4], concluded that the studies had a low level of evidence and a heterogeneous methodological quality, indicating that it was impossible to draw firm conclusions about risk factors for curve progression. Recent population-based studies have shown that degree of motor function, as classified by the GMFCS (Gross Motor Function Classification System) is a strong predictor for the development of scoliosis

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[5]. There is, however, a lack of population-based data on other parameters of importance for optimal spine surveillance. The rate of progression of the Cobb angle according to the level of motor function and increasing age has not been clarified, and the association between clinical and radiographic spine screening needs further investigation [8].

In order to improve existing screening programmes for surveillance of scoliosis in CP, the aims of the present population-based study were to:

1. Evaluate the prevalence of scoliosis according to patient age and GMFCS level in children with severe CP.

2. Assess the rate of progression of scoliosis and the risk factors for deterioration.

Materials and methods

The patients included in this retrospective population-based study were children enrolled in The Norwegian Quality and Surveillance Registry for Cerebral Palsy (NorCP). The inclusion criteria were children born in the 7-year period 2002–2008 and living in one of the 10 south-eastern counties and GMFCS levels III–V [9].

Clinical examination

Clinical examination of the spine was performed once a year by a physiotherapist at the Child Rehabilitation Services, one in each county. The examination was performed with the patient in the standing position, (GMFCS level III) or sitting position (GMFCS level IV or V). The forward bending test was included in the clinical examination. The degree of scoliosis was classified subjectively as mild, moderate, severe, or uncertain.

Radiographic examination

According to the NorCP protocol, radiographs of the spine should be taken in all patients with moderate or severe degrees of scoliosis at clinical examination. Radiographs were taken in the standing position for children who had unsupported standing function (GMFCS level III or IV), and in the sitting position (with support by a caregiver if needed) for those without standing function (GMFCS level IV or V). Scoliosis was characterized by location in the spine, shape of the curve, apex, and Cobb angle, which indicates the degree of scoliosis. These assessments were performed by a radiologist and were quality-assured by a second observer, an orthopaedic spine surgeon with experience in neuromuscular scoliosis (SV and TK).

For each patient, all available radiographs of the spine from the beginning of 2004 to 2021 were assessed. The radiographs were taken at the local hospital or the university hospital and stored in our Picture Archiving and Communication System (PACS; Sectra, Linköping, Sweden). Although mild spinal deformities are of less clinical importance, Cobb angles $\geq 10°$ were registered. Scoliosis was termed mild for a Cobb angle of 10–19°, moderate for an angle of 20–39°, and severe for an angle of $\geq 40°$.

To test the quality of the clinical screening, we analysed the association between the clinical and radiographic results. The clinical gradings within 1 year before or after the first radiograph showing a scoliosis $\geq 10°$ were compared with the radiographic gradings based on the Cobb angle.

For assessment of risk factors for deterioration of the scoliosis, these variables were analysed: gender, GMFCS level, CP type, intrathecal baclofen treatment, epilepsy, initial Cobb angle, and Cobb angle at age 10 years.

Statistical analysis

Statistical analyses were performed using IBM SPSS Statistics (version 28; IBM Corp., Armonk, New York, USA). Categorical variables were analysed using the Chi-square test and continuous variables using the t-test for independent samples and analysis of variance. Univariable logistic regression was used to identify the risk factors for scoliosis involving a Cobb angle $\geq 40°$. Variables with p values < 0.05 were evaluated using multivariable regression analysis. Kaplan–Meier analysis was used to evaluate the percentage of patients with a Cobb angle $\geq 40°$ according to age and functional level.

The progression of scoliosis in patients at GMFCS level V or IV was analysed using a nonlinear model comprising a fractional polynomial linear regression, with maximum one degree of the fractional polynomial, and robust standard error for repeated measurements in the same subject. The plots are presented with the linear prediction (estimated mean from the regression model) and 95% confidence intervals. We estimated the mean Cobb angle and Cobb angle increase per 2-year age interval from the fitted nonlinear models.

Ethics

This study was approved by the Regional Committee for Research Ethics (REC no. 107726) and the hospital's Privacy and Data Protection Officer. Informed consent was obtained from the caregivers of all participants. There were no conflicts of interest.

Results

A total of 206 children (86 girls and 120 boys) were included in the study. The distribution according to GMFCS was level III in 50 children, level IV in 47 children, and level V in 109

children. The movement disorder was bilateral spastic CP in 153 children, dyskinesia in 45 children, and ataxia in 8 children.

Scoliosis was defined as a Cobb angle ≥ 10° and occurred in 121 children (59%). The GMFCS distribution showed that scoliosis was more frequent in children at GMFCS level V than in children with better motor function (Table 1). The mean Cobb angle at the initial radiographic examination was 31.4° (range 10–87) at a mean age of 9.6 years (range 2–18).

Table 1 Scoliosis prevalence in 206 patients with CP at GMFCS level III–V

	N	Cobb angle ≥ 10°		Cobb angle ≥ 40°	
		N	%	*N*	%
GMFCS III	50	10	20	3	6
GMFCS IV	47	22	47	9	19
GMFCS V	109	89	82	68	62
All patients	206	121	59	80	39

CP cerebral palsy, *GMFCS* gross motor function classification system, *N* number of patients

The mean initial Cobb angle was 26° at GMFCS level III, 27° at level IV, and 33° at level V; the differences were not significant ($p = 0.505$). The scoliosis was convex to the left side in 70 patients and convex to the right in 51 patients. The apex of the scoliosis was in the middle thoracic spine (T5–T8) in 17 children, in the lower thoracic spine (T9–T12) in 46, and in the lumbar spine (L1–L4) in 58 children.

Eighty of the 206 patients (39%) developed a severe scoliosis with a Cobb angle ≥ 40°, 33 girls and 47 boys, at a mean age of 10.9 years (range 5–16). Severe scoliosis was much more prevalent in nonambulatory children at GMFCS level V (62%) than in those at level IV (19%) and level III (6%) (Table 1). The potential risk factors for deterioration to a Cobb angle ≥ 40° are shown in Table 2. GMFCS level, initial Cobb angle, Cobb angle ≥ 30° at age 10 years, epilepsy, and intrathecal baclofen treatment were significant risk factors in univariable analysis. In multivariable logistic regression, the initial Cobb angle, Cobb angle ≥ 30° at age 10 years, and GMFCS level V remained as independent (true) risk factors.

The analysis of scoliosis progression was based on a total of 372 radiographs in 119 children at the age of 3–17 years, with a mean of 3.1 radiographs (range 1–9) per patient. The Cobb angle increased during the follow-up

Table 2 Potential risk factors for scoliosis with a final Cobb angle ≥ 40° in 206 children at GMFCS levels III–V

Parameters	*N*	Scoliosis		*p* value	
		Cobb angle < 40°	Cobb angle ≥ 40°	Univariable	Multivariable
Sex (*N*)					
Boys	120	73	47	0.908	
Girls	86	53	33		
GMFCS (*N*)					
Level III	50	47	3	< 0.001	0.004
Level IV	47	38	9		
Level V	109	41	68		
CP type* (*N*)					
Spastic	153	89	64	0.308	
Dyskinetic	45	31	14		
Intrathecal baclofen (*N*)					
No baclofen	170	116	54	< 0.001	0.162
Baclofen	36	10	26		
Epilepsy** (*N*)					
No epilepsy	73	64	9	< 0.001	0.898
Epilepsy	104	46	58		
Cobb at 10 years (*N*)					
Cobb < 30°	78	39	39	< 0.001	< 0.001
Cobb ≥ 30°	43	2	41		
Initial Cobb°, mean (SD)					
121	121	19.2 (6.3)	37.6 (18.6)	< 0.001	< 0.001

GMFCS gross motor function classification system, *N* number of patients, *CP* cerebral palsy

*eight patients with ataxia were not included because of the low number of cases

**data on epilepsy were not available or were uncertain in 29 patients

in most of the patients (Fig. 1), but the pattern of increase differed (Fig. 2). The youngest children at GMFCS level V had the highest rate of scoliosis progression (Table 3). Their mean progression was 9.7° per year at patient aged 3–5 years and 4–6° at age 5–10 years. From the age of 11 years to skeletal maturity, the progression decreased to a mean of 2–3° per year. In children at GMFCS level IV, the number of radiographs in patients aged < 11 years was too small for meaningful analysis. From the age of 11 years, the Cobb angle increased 2–3° per year up to skeletal maturity (Table 3).

Kaplan–Meier survival analysis in which the end point was the percentage of patients with a Cobb angle < 40° was performed for children with GMFCS levels IV and V. Patients at level IV had markedly better prognosis than those at level V (Fig. 3). At age 10 years, 91% of children at GMFCS level IV and 61% at level V had a Cobb angle < 40°. Most of the children were diagnosed with severe scoliosis after age 10 years. At the age of 15 years, the percentages of patients with a Cobb angle < 40° had decreased to 64% at GMFCS level IV and 25% at level V.

Thirty-one patients (28 at GMFCS level V and three at level IV) underwent surgical treatment of their scoliosis during the follow-up period. The mean age at the time of surgery was 13.4 years (range 6–17). The mean preoperative Cobb angle was 90.5° (range 49–140). In patients who had not undergone spine surgery, the mean Cobb angle at the last radiograph was 63.1° (range 11–137) at a mean age of 13.4 years (range 4–18).

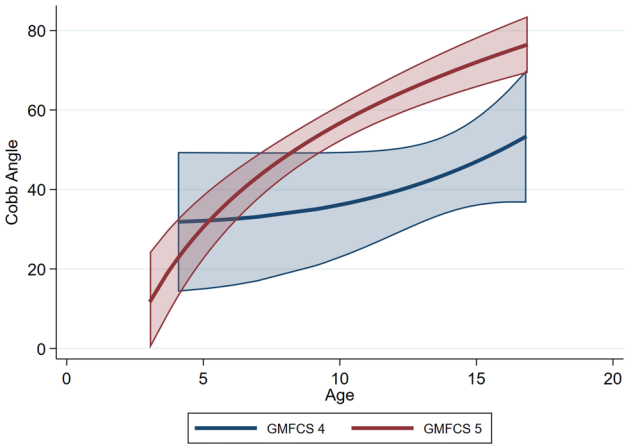

Fig. 2 Scoliosis progression at different Gross Motor Function Classification System (GMFCS) levels presented for patients aged 3–17 years (GMFCS level IV: blue curve; level V: red curve)

The associations between the clinical and radiographic gradings are shown in Table 4. Since clinical evaluation within one year before or after the first radiograph showing a Cobb angle of ≥ 10°, was not available in 13 patients, the analysis was performed in the remaining 108 patients. The Chi-square test showed a significant association between the two gradings

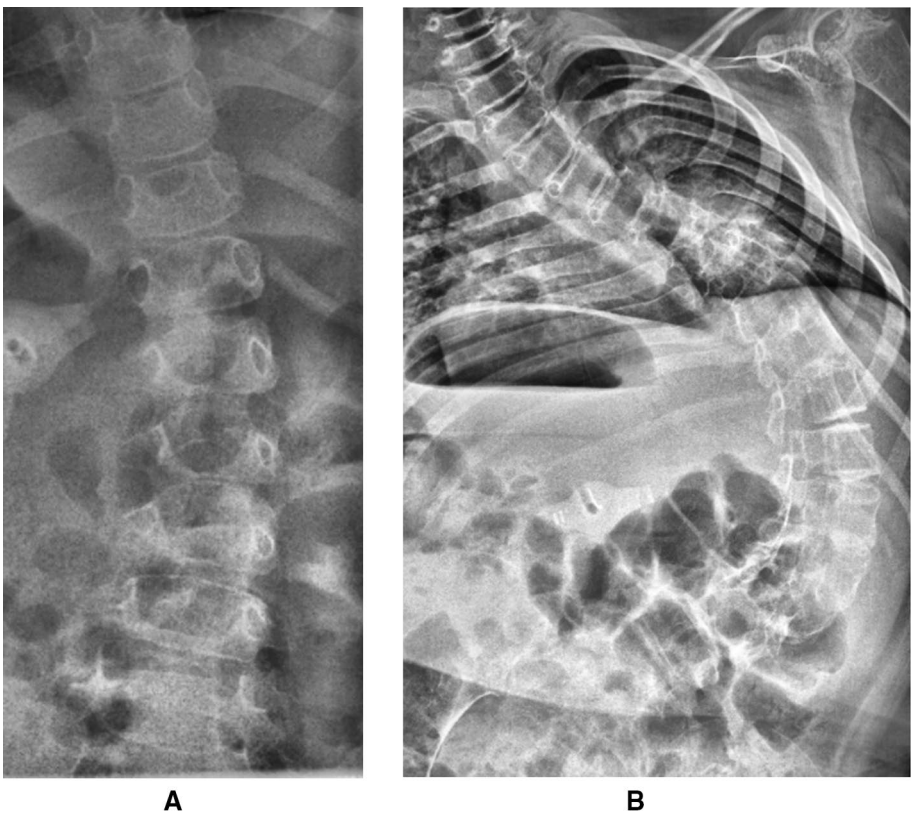

Fig. 1 **A** Posteroanterior radiograph of a 4-year-old girl, with moderate scoliosis that was convex to the right side and a Cobb angle of 29°. **B** Posteroanterior radiograph of the same girl at 11 years of age. The scoliosis has increased markedly, and the Cobb angle is 118°

Table 3 Progression of scoliosis in patients at GMFCS level V and IV, given as Cobb angle increase per year according to patient age

Parameters	Age group (years)	N1	N2	Mean Cobb angle (°)	Annual increase in Cobb (°)	95% CI	*p* value
GMFCS V	3–4.9	12	15	30.5	9.7	7.1–12.2	< 0.001
	5–6.9	22	29	43.2	6.4	4.7–8.1	< 0.001
	7–8.9	32	39	52.7	4.8	3.5–6.0	< 0.001
	9–10.9	41	58	60.3	3.8	2.8–4.8	< 0.001
	11–12.9	44	62	66.6	3.2	2.3–4.0	< 0.001
	13–14.9	40	65	72.0	2.7	2.0–3.4	< 0.001
	15–16.9	23	34	76.7	2.4	1.7–3.0	< 0.001
GMFCS IV*	11–12.9	10	17	41.6	2.0	–0.8–4.8	0.149
	13–14.9	12	19	47.0	2.7	–1.1–6.5	0.149
	15–16.9	7	10	54.1	3.5	–1.4–8.4	0.149

GMFCS gross motor function classification system, *N1* number of patients, *N2* number of radiographs, *CI* 95% confidence interval

*the analysis was not performed in patients < 11 years because the number of patients in each age span was too small (< 5)

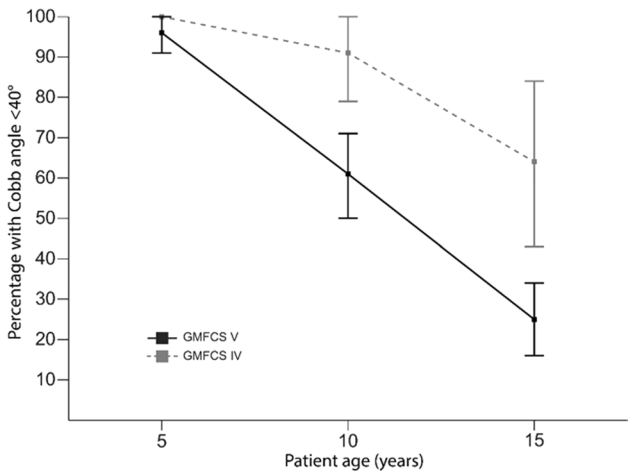

Fig. 3 Kaplan–Meier survival analysis of GMFCS levels IV and V, where the end point is the percentage of patients not having a Cobb angle ≥ 40° (broken line: GMFCS IV; unbroken line: GMFCS level V

($p < 0.001$). Clinical scoliosis was noted in 91 patients and was graded "uncertain" in 23 patients. In the remaining 68 patients, there was full agreement (mild, moderate, or severe) between the clinical and radiographic gradings in 51 cases (75%). Clinical examination noted scoliosis in 26 of 36 patients with mild radiographic scoliosis (Cobb angle 10–19°), in 30 of 37 patients with moderate scoliosis, and in all the patients with severe scoliosis (Cobb angle ≥ 40°). Thus, the reliability of the clinical screening increased with the magnitude of the scoliosis.

Table 4 Association between initial radiographic examination in 108 patients with Cobb angle ≥ 10° and clinical examination within 12 months before or after the radiographic examination

Clinical scoliosis		Cobb angle		
		10–19° mild	20–39° moderate	≥ 40° severe
No scoliosis	17	10	7	0
Mild	27	19	6	2
Moderate	18	0	12	6
Severe	23	1	2	20
Uncertain grade	23	6	10	7
Total	108	36	37	35

No clinical examination was available in the case reports for 13 of the 121 patients

Discussion

The most important findings were that the prevalence of scoliosis increased with decreasing functional capacity, that high initial Cobb angle, Cobb angle ≥ 30° at age 10 years, and GMFCS level V were independent risk factors of deterioration, and that the annual increase in Cobb angle was more than twice as large in children at GMFCS level V aged < 10 years than in older children.

Population-based studies are needed to provide reliable data on scoliosis prevalence and development in subgroups. In the present study, the prevalence of scoliosis with a Cobb angle ≥ 10° was 59% in children at GMFCS levels III–V, which is consistent with the prevalence of 65% in a population-based study from Australia [6]. More clinically interesting is the prevalence of severe scoliosis, defined as a Cobb angle ≥ 40°. Given that the prevalence increases with age and GMFCS levels, these factors should

be considered. The rates of severe scoliosis at age 10 years in our study were 9% in children at GMFCS level IV and 39% in those at level V in our study, which are somewhat higher than the 5% and 20%, respectively, in a population-based Swedish study [5]. At skeletal maturity, the percentages of children in our study with Cobb angle ≥ 40° were 19% at GMFCS level IV and 62% at level V, which are slightly lower than the 35% and 75%, respectively, at patient aged 20 years in the Swedish study. However, the latter patients were a few years older, and the degree of scoliosis continues to increase after skeletal maturity. Our rates also correspond with those in an Australian study that reported rates of severe scoliosis of 18% in adults at GMFCS level IV and 48% at level V [6].

Previous studies that were not population-based have shown wide differences in the progression of the Cobb angle in nonambulatory children, from 3.5° to 12.2° per year [2, 10, 11]. Since the rate of progression is important for a better understanding of scoliosis development and for improvement of surveillance, analysis according to age groups and GMFCS levels is required. The present study had an age range of 3–17 years and a mean of three radiographs per patient, which allowed us to assess the annual progression during childhood and adolescence. The annual progression in Cobb angle was greater in children at GMFCS level V than in those at levels IV and III. The progression at GMFCS level V was greatest in younger children, with a mean yearly increase in Cobb angle of approximately 6° in the age span 5–7 years, decreasing to 2–3° per year in children older than 11 years. We have found no previous population-based analysis of curve progression according to age. In a study of nonambulatory children that was not population-based, Gu et al. [2] found a mean annual progression in Cobb angle of 4.6° in the age span 7–11 years, which is similar to the rates of 4–5° in our study. However, whereas Gu et al. [2] reported that the Cobb angle increased more rapidly later in childhood than earlier in childhood, we found the opposite trend with larger annual progression in children < 10 years of age.

GMFCS level V was a risk factor for curve progression, which confirmed the findings of previous studies [5, 6, 12]. The initial Cobb angle, which seems not to have been evaluated in previous studies, was also an independent risk factor of progression. The risk factors should be known before the adolescent growth spurt. Therefore, Yoshida et al. [13] examined the Cobb angle at patient aged 10 years and found that an angle ≥ 30° was a significant predictor of increased curve progression. This was confirmed in the present study. In a recent population-based Swedish study [12], the independent predictors of Cobb angle ≥ 40° were female sex, GMFCS levels IV and V, epilepsy, and limited knee extension. We did not examine the latter variable and did not find that female sex and epilepsy were independent predictors. Willoughby et al. [6] reported that a Cobb angle ≥ 40° was more likely to occur in patients with dystonia than in those with spastic CP, but this was not confirmed in our study. Intrathecal baclofen treatment was a risk factor in the univariable but not in the multivariable analysis, probably because most of the patients who had received baclofen were at GMFCS level V, which was an independent risk factor.

In accordance with a previous study based on only 28 patients [8], we found a significant association between clinical and radiographic gradings. A positive aspect was that the reliability of the clinical examination increased with increasing degree of scoliosis, and that no cases of severe radiographic scoliosis had been missed in the clinical examinations.

Our study has some limitations. First, the number of patients was small in some of the subgroups, which may have affected the reliability of the statistical analyses. Second, the results of the clinical evaluations within one year before and after the radiographic examination were not available in several patients, indicating that the screening routines had not always been followed. Third, the clinical classification by the physiotherapists was based on categories that were not clearly separated, which is a common limitation in subjective gradings. The main strength of this study is that the patients were recruited from the population-based NorCP register and that the follow-up was good. The register is based on data from > 93% of patients with CP in Norway [14].

What are the clinical implications of the present study? The NorCP surveillance programme has so far no specific recommendations for the frequency of radiographic follow-up. The observation that one third of children with mild clinical scoliosis had moderate or severe radiographic scoliosis, indicating that a spine radiograph should be taken even in such mild cases. Given the pronounced trend to deterioration in nonambulatory children, a plan for radiographic follow-up in this group should be specified and should include annual radiographs for at least a few years after detection of scoliosis in order to elucidate the annual Cobb angle progression. This routine may provide important information to patients and caregivers and improve the timing of non-surgical and surgical treatment.

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Declarations

Conflict of interest None.

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