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Original Article

Natural history of scoliosis in cerebral palsy and risk factors for progression of scoliosis

Kiyoshi Yoshida [a, *], Ichiro Kajiura [b], Tsunehiko Suzuki [b], Hidehiko Kawabata [b]

[a] Department of Orthopaedic Surgery, Osaka University Graduate School of Medicine, Japan
[b] Department of Orthopaedic Surgery, Osaka Rehabilitation Hospital for Children, Japan

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ABSTRACT

Background: Scoliosis in cerebral palsy (CP) often occurs and causes a disturbance in daily life. The purpose of this study was to investigate the natural history of scoliosis in cerebral palsy and determine risk factors for the progression of scoliosis using multivariate analyses.
Methods: We revised 113 patients with CP (47 males and 66 females) who had scoliosis with a curve of at least 10° were reviewed and retrospectively investigated these cases of scoliosis and analyzed the risk factors for the progression of this condition.
Results: The mean follow-up period was 16.5 years and the mean age at onset of scoliosis was 6.6 years (range: 1–16 years). In 59 patients (52%), the age at onset of scoliosis was under 6 years. On the final radiographs, the mean Cobb angle was 55.1° (range: 10° to 169°). After the age of 20 years, 13 of 40 patients (32.5%) had a progression of over 10° in scoliosis. Multivariate analyses showed the risk factors for the progression of scoliosis to be hip displacement ($p = 0.0038$), the onset of scoliosis before the age of 6 years ($p = 0.0024$), and 30° of the Cobb angle before the age of 10 years ($p < 0.001$). A subtype of CP (spastic quadriplegia) was identified as a potential risk factor.
Conclusions: After the age of 20 years, 32.5% patients had a progression of over 10° in scoliosis. Risk factors for the progression of scoliosis in CP included hip displacement, early-onset scoliosis, and Cobb angle of 30° before the age of 10 years.
Level of evidence: Prognostic level IV - case series.

1. Introduction

Scoliosis in cerebral palsy (CP) frequently occurs; its incidence has been reported from 25% to 69% [1–4]. Severe scoliosis often causes a disturbance in daily life and places a high demand on nursing care. Treatment for scoliosis in CP is primarily includes brace treatment or surgical operation. Brace treatment in CP is often difficult to continue. Recently, surgical treatment has been recommended and performed for various indications. However, there is no consensus on the treatment of scoliosis in CP and indications of surgical treatment [4,5]. This may be because there are few studies on the natural history of scoliosis in CP and on risk factors for the progression of scoliosis [6–8]. Many factors may influence the progression of scoliosis, and the condition can continue to progress even after growth maturity. Previously We reported the natural history of scoliosis with 89 patients [9]. We divided into 2 groups and univariate analysis of several factors was performed. We concluded the risk factors for the progression of scoliosis were Gross Motor Function Classification System (GMFCS) [10] level V, Spastic Quadriplegia, hip displacement, the early-onset scoliosis and Cobb angle of 30° before the age of 10 years. Because our previous study used univariate analysis, we needed to increase the number of cases and use multivariate analysis.

In this study, we investigated the natural history of scoliosis in CP in depth and analyzed risk factors for the progression of scoliosis using multivariate analysis.

2. Materials and methods

A retrospective medical review of 113 patients with CP who had scoliosis with a curve of at least 10° as analyzed by the Cobb method on total spine radiographs at Osaka Rehabilitation Hospital

* Corresponding author. Department of Orthopaedic Surgery, Osaka University Graduate School of Medicine, 2-2 Yamadaoka, Suita, Osaka, 565-0871, Japan. Fax: +81 6 6879 3559.
E-mail address: yoshida-kiyoshi@umin.ac.jp (K. Yoshida).

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for Children was performed. None of the patients received continuous treatment for scoliosis, such as brace treatment or surgical operation. The study was approved by the institutional review board and informed consent was obtained from all individual participants included in this study.

All patients were followed up by orthopedists and had several radiographs of the total spine in the supine position to monitor the progression of the curvature. The severity of scoliosis was measured by the Cobb method, and the pattern of the spinal curve, location of the curve, and number of vertebrae involved were examined. In cases with a double curve, the major curve was measured. The progression of the curvature after growth maturity was also measured. In this study, 40 patients had radiographs of the total spine after the age of 20 years. In this study, the progression of scoliosis was defined as more than 10° progression after the age of 20 years.

Risk factors for the progression of scoliosis regarding CP subtype, physical mobility, hip dislocation, scoliosis onset, and the Cobb angle before growth maturity in patients who had a follow-up after the age of 18 years were investigated. Regarding the subtype of CP, patients were divided into spastic, dyskinetic, and ataxic subtypes. Spastic CP included spastic quadriplegia, diplegia, and hemiplegia. These were classified into two groups: spastic quadriplegia and others. Physical mobility was classified according to GMFCS. Patients were classified into two groups based on the following: 1) those classified as GMFCS level V (bedridden) and 2) others. In addition to spinal radiographs, hip dislocation via anteroposterior radiographs of the hip was examined. In this study, a normal hip was defined as when the migration percentage of the hip was under 50% and a displacement hip was defined when the migration percentage was over 50%. Patients were classified into three groups as follows: 1) normal bilateral hip, 2) unilateral hip displacement, and 3) bilateral hip displacement. For age at onset of scoliosis, patients were divided into two groups: 1) before the age of 6 years and 2) after the age of 6 years. In analyzing risk factors for the progression of scoliosis regarding age and the Cobb angle, patients were divided into two groups: 1) under 30° and 2) over 30° before the age of 10 years.

2.1. Statistical analysis

Statistical analysis was performed using JMP Statistical Software, Version 11 (SAS Institute Inc., Cary, NC). Each parameter was analyzed using the Mann–Whitney test. Cobb angle between normal hip and unilateral and bilateral hip displacement was tested by one-way ANOVA. Differences were considered statistically significant at $p < 0.05$. Factors significant on univariate analysis were evaluated by multiple regression analysis. The threshold for including variables in the multiple linear regression analysis was $p = 0.05$.

3. Results

3.1. Demographic data

Forty-seven of the 113 (42%) patients were males, and 66 (58%) were females. The mean age at the first radiograph was 4.1 years (range: 0–17 years) and the mean follow-up period was 16.5 years (range: 6–32 years). There were 73 patients who had a follow-up over the age of 18 years. Forty patients who had a follow-up over the age of 20 years and mean follow-up after 20 years was 6.0 years (range 1–14 years). Eleven patients (10%) were able to walk (GMFCS level I ~ III). 19 patients (17%) were classified as level IV, and 83 patients (73%) were level V in GMFCS. A total of 72 patients (64%) had spastic quadriplegias (16 dyskinetic, 12 spastic diplegia, 6 ataxia, and 7 others).

3.2. Scoliosis characteristics

Eighty-four patients (74%) had single curves, and 29 patients (26%) had double curves. In the major curves, 46 patients (41%) had thoracic curves, 41 patients (36%) had thoracolumbar curves, and 26 patients (23%) had lumbar curves. The number of vertebrae involved in the major curves was an average of 7.1 (4–14).

The mean age at onset of scoliosis was 6.6 years (range: 1–16 years). In 59 patients (52%), the age at onset of scoliosis was under 6 years. On the final radiographs, the mean Cobb angle was 55.1° (range: 10° to 169°) (Fig. 1).

3.3. Progression of the curvature after growth maturity

After the age of 20 years, 13 of 40 patients (32.5%) had a progression of over 10° in scoliosis. The mean Cobb angle at the growth maturity was 60.3° (range 22–169°) with the patients who had no progression of over 10° and 84.9° (range 12–126°) with the patients who had a progression over 10°. There was no significant difference in the progression after growth maturity for the degree of curvature at growth maturity, the subtype of cerebral palsy, and the GMFCS level.

3.4. Risk factors for the progression of scoliosis

To determine the important prognostic factors for the progression of scoliosis, ten factors were evaluated by uni- and multivariate analyses. Table 1 shows the distribution of sex, the pattern of the curve, the location of the major curve, the subtype of CP, the GMFCS level, hip displacement (unilateral or bilateral hip displacement), the onset of scoliosis, and Cobb angle at the age of 10 years. In patients with spastic quadriplegia, GMFCS level V, hip displacement (both unilateral and bilateral hip displacement), the onset of scoliosis before the age of 6 years and Cobb angle of 30° before the age of 10 years, a significant progression was revealed by univariate analysis. In the multiple linear regression analysis, the important prognostic factors for progression were hip displacement ($p = 0.0018$), the onset of scoliosis before the age of 6 years ($p = 0.0001$) and Cobb angle of 30° before the age of 10 years ($p < 0.001$) (Table 2). The subtype of CP ($p = 0.071$) were identified as potential risk factors for progression.

4. Discussion

Several studies have reported the frequency of scoliosis in CP. Balmer et al. [1] reported that 21 of 100 (21%) children in an outpatient clinic had scoliosis. Madigan et al. [3] reported 64%, and Koop et al. [11] reported 77%. However, there are few reports of the natural history of scoliosis in CP. Saito et al. [4] reported a detailed investigation of the natural history of scoliosis in CP from childhood to adult life. They reported that 54 of 79 (68%) institutionalized patients with spastic CP had scoliosis. They analyzed 37 patients via radiographic examination and the details of the patients (e.g., mental retardation, nutritional status, joint contractures, and frequency of pneumonia occurrence). They also reported that 11 of 13 (85%) patients had a spinal curve of more than 40° at the age of 15 years, and their scoliosis progressed to more than 60°. They also reported that scoliosis of more than 60° developed in patients with total body involvement (67%), those who were bedridden (100%), and those with thoracolumbar curves (57%). In their conclusions, risk factors for the progression of scoliosis in spastic CP included a spinal curve of 40° before the age of 15 years, total body involvement, bedridden, and thoracolumbar curve. Gu et al. [5] investigated the natural history of scoliosis in 110 patients with nonambulatory spastic tetraplegic CP. They reported that the risk

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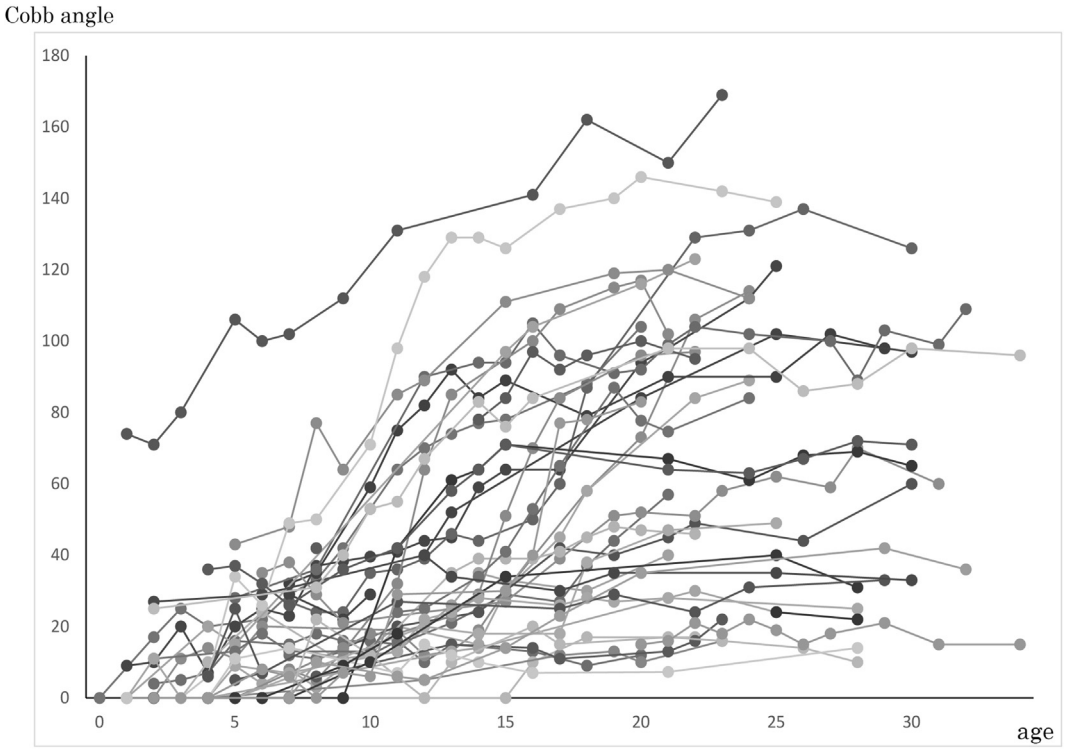

Fig. 1. Curve progression in 113 patients.

Table 1
The risk factor for the progression of scoliosis.

		Cobb angle	p value
Sex	male	68.6 ± 46.1	0.34
	female	58.9 ± 39.8	
Subtype of CP	spastic quadriplegia	64.3 ± 41.6	<0.01
	others	37.0 ± 28.3	
GMFCS	level V	70.7 ± 41.8	<0.01
	others	34.4 ± 31.8	
Location of curve	thoracic	65.1 ± 45.8	0.76
	thoracolumbar	56.1 ± 43.9	
	lumbar	64.1 ± 34.3	
Curve pattern	single curve	59.9 ± 43.1	0.37
	double curve	69.8 ± 40.7	
Onset of scoliosis	under 6 years	79.8 ± 41.7	<0.01
	over 6 years	44.0 ± 33.9	
Cobb angle at 10 years	under 30°	46.2 ± 30.9	<0.01
	over 30°	115.1 ± 25.1	
Hip displacement	normal	44.9 ± 30.8	
	unilateral displacement	79.8 ± 39.7	0.02
	bilateral displacement	102.1 ± 46.7	<0.01

Table 2
The risk factor of the progression of scoliosis: multivariate analysis.

	Coefficients	Standard error	95% CI	t value	p value
Sex	3.57	3.09	(−2.6–9.7)	1.15	0.253
Subtype of CP	−6.74	3.67	(−14.1 to −0.6)	−1.83	0.071
GMFCS	−4.05	4.11	(−12.3–4.2)	−0.99	0.328
Onset of scoliosis	−2.98	0.86	(−4.7 to −1.3)	−3.45	0.0001
Cobb angle at 10 years	−21.56	4.11	(−29.7 to −13.4)	−5.26	<0.0001
Hip displacement	−10.88	3.33	(−17.5 to −4.2)	−3.26	0.0018

factors for the progression of scoliosis were age, weight, height, and, most significantly, Cobb angles greater than 40° by the age of 12 years. The number of patients in their study was large, but their follow-up period was short, and half of the patients had had fewer than four spine radiographs. Bunke et al. [7] investigated 116 children with scoliosis and reported that the risk factors for developing scoliosis were GMFCS level and age. In their results, children with a GMFCS level IV and V had a 50% risk and most children had scoliosis before they were 8 years old. Kalen et al. [12] investigated 14 adult patients with scoliosis (Cobb angle > 45°) and 42 patients with mild scoliosis or no curves. The former group had more orthopedic deformities involving the pelvic obliquity, hip dislocation, and physical capability.

In the relationship between hip deformities and scoliosis, the conclusion is controversial. Senaran et al. [13] investigated 23 patients with unilateral hip dislocation. They compared the mean progression rate of scoliosis between the unilateral hip dislocation and the normal hip group, and the mean progression rate was not significantly different. Pelvic obliquity was significantly increased in the unilateral hip dislocation group, but the progression rate of scoliosis was not significantly different between the two groups. They concluded that a unilateral hip dislocation was not a risk factor for the progression of scoliosis. Porter et al. [14] investigated the relationship between the direction of scoliosis and hip deformities. They concluded that the direction of scoliosis and hip deformities had a significant relationship. In this study, both unilateral and bilateral hip displacement had a significant effect on the progression of scoliosis. Our results were not consistent with some previous studies, and this was because of the difference in the number of patients and the definition of hip dislocation and displacement. Many reports have used the migration percentage as the definition of hip dislocation and displacement [15–17], but there is no consensus for hip dislocation and subluxation. We defined hip displacement as greater than 50% and hip displacement

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included hip subluxation and dislocation. This was because both hip dislocation and subluxation caused the pelvic obliquity. In this study, we excluded the factor of pelvic obliquity, because pelvic obliquity can be greatly influenced by the position and muscle tonus at the time of radiographs and the joint contracture of the hip. In many patients, pelvic obliquity varied much during follow-up and we could not perform the statistical analysis in pelvic obliquity.

We analyzed several patients and many factors regarding the progression of scoliosis. Our results suggest that the risk factors for the progression of scoliosis in CP were hip displacement, early-onset scoliosis, and Cobb angle of 30° before the age of 10 years, using multivariate analysis. Scoliosis was reported to progress rapidly during a growth spurt. Since we believed that the risk factors for progression should be known before a growth spurt, we investigated the Cobb angle at the age of 10 years. Thirty degrees was the lowest number at which statistically significant differences could be found.

Early-onset scoliosis has been reported to progress in children and cause remarkable restriction of daily life [18,19]. In the age at onset of scoliosis, we divided the patients into two groups, onset at younger than 6 years and older than 6 years. Early onset scoliosis had a significant effect on the progression of scoliosis in our results.

There are very few reports of the curve progression after growth maturity. Thometz et al. [20] analyzed the natural history of scoliosis after growth maturity. They reviewed 51 adult patients with CP. The patients, whose curve was more than 50° at growth maturity, had a higher progression rate than patients whose curve was less than 50°. In our results, 32.5% of patients exhibited progression of scoliosis after the age of 20 years. However, there was no significant difference in the degree of curvature at growth maturity, the subtype of CP, and physical capability. Our data did not have a homogeneous distribution in the patient group (the majority of patients had a spastic CP and GMFCS level V). We need to investigate a larger number of patients with CP to determine the progression of scoliosis after growth maturity.

Conflicts of interest

None of the authors have any conflicts of interest of disclosures in relation to this work.

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