Original article

Progressive age and other factors affecting scoliosis severity in cerebral palsy patients

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Abstract

Objective: This study aimed to investigate the age at which scoliosis progresses to a severe condition and identify the factors related to severe scoliosis in patients with cerebral palsy.

Patients and Methods: This retrospective study included 51 patients aged ≥15 years. The Cobb angle was measured over time using radiographs. Patients were divided into the following groups according to their final Cobb angle: <60°, 60°–100°, and ≥100°. The age at which the Cobb angle was ≥20° in the patients was compared among the groups. Moreover, the age at which a significant difference in the Cobb angle occurred in the groups was considered the age at which the scoliosis worsened. Association of the final Cobb angle with factors such as the location of curve, Gross Motor Functional Classification System (GMFCS), capability of turning over, orthosis use, hip dislocation, tracheotomy, and gastric fistula was examined.

Results: The mean age at which the Cobb angle was ≥20° was significantly lower in the ≥100° group. From 9 years of age, a significant difference was noted in the Cobb angle between the <60° group and ≥100° group. Between 13–19 years, a significant difference in the Cobb angle was observed among the three groups. Furthermore, GMFCS, capability of turning over, hip dislocation, and gastric fistula were the factors showing a significant difference among the three groups.

Conclusion: Scoliosis progressed to the severe form (Cobb angle ≥100°) at 9 years of age. Moreover, scoliosis is aggravated during the growth period. Severe cerebral palsy with low motor function levels and problems with internal functions was considered the cause of scoliosis deterioration.

Key words: cerebral palsy, scoliosis, Cobb angle

Introduction

Scoliosis is the most common complication occurring in patients with cerebral palsy (CP), with an incidence of 21–68%[1–3]. Asymmetric spasticity and the effect of gravity are considered as factors related to the development of scoliosis in patients with CP[2, 4]. Surgical interventions may be considered for progressive scoliosis in patients with CP, in which curve progression could cause loss of ambulation, decreased sitting tolerance, or pain[5]. Furthermore, severe scoliosis can cause ventilation dysfunction[6] and gastrointestinal disorders. Therefore, severe scoliosis could worsen the general condition of patients with CP.

In order to plan the prevention and treatment strategy for scoliosis in patients with CP effectively, it is necessary to know the natural course of scoliosis. Therefore, this study aimed to determine the age at which the condition progresses to severe scoliosis and identify factors related to severe scoliosis in patients with CP.

Patients and Methods

The ethics committee of Ibaraki Prefectural University of Health Sciences reviewed and approved this retrospective study (approval number: e88). We adopted an opt-out style to maintain the rights of the patients to end their participation in the study whenever they desired.

This retrospective study included patients with CP, aged ≥15 years, who exhibited a Cobb angle of ≥20° as of 2018.

All participants included were patients at our hospital. Patients who had undergone scoliosis surgery were excluded, but those who received orthotic treatment were included. Full spinal radiographs of all patients were reviewed.

The severity of the scoliosis was measured using the Cobb method based on supine whole-spine radiographs. Moreover, we examined the type of curve (single, double), location of the apex vertebra (thoracic T1–T11, thoracolumbar T12–L1, and lumbar L2–L4), and convex side (left and right). In cases with a double curve, the major curve was measured.

The patients were divided into the following groups according to the Cobb angle during the final follow-up: <60° group, 60°–100° group, and ≥100° group. The age at which the patients in the three groups were diagnosed with a Cobb angle of ≥20° was compared. Furthermore, the age at which a significant difference in the Cobb angle occurred in the three groups was considered the age at which the scoliosis worsened.

Factors related to the severity of scoliosis were also examined. The association between the Cobb angle at the final follow-up and each factor was investigated. The factors expected to be associated with scoliosis were location of the curve (thoracic, thoracic lumbar, lumbar), Gross Motor Functional Classification System (GMFCS), capability of turning over, use of orthosis, presence/absence of hip dislocation, presence/absence of tracheotomy, and presence/absence of gastric fistula. The GMFCS comprises of levels I–V. Capability of turning over was defined as the ability to turn from the supine position to the lateral position, from the lateral position to the prone position, from the prone position to the lateral position, etc. Hip dislocation was defined as a migration percentage of ≥100% as measured on the radiographs.

Statistical analysis

For the factors related to the severity of scoliosis, the comparison of data between two groups was analyzed using the Mann-Whitney test, whereas comparison of data among the three groups was performed using the Kruskal-Wallis test. When the results were significantly different, they were examined using Bonferroni correction to determine the dif-

ference among the groups. All statistical analyses were performed using the Statistical package for the Social Sciences software version 25 (SPSS Inc., Chicago, Illinois, USA). A significance level of 5% was chosen for all tests ($P<0.05$).

Results

A total of 51 patients (33 male and 18 female) were included in the analysis. Regarding the type of CP, 42 patients were diagnosed with spastic CP, 7 with dyskinetic CP, and 2 with hypotonic CP. In terms of the classification of the paralysis, 45 patients had quadriplegia, 5 had diplegia, and 1 had hemiplegia. One patient had GMFCS level I, 2 had level II, none had level III, 6 had level IV, and 42 had level V. The mean age of the first radiographic examination of the patients was 9.4 years (2–23 years). The average age of the final investigation was 20.2 years (15–29 years). The average duration between the first radiograph and the final investigation was 10.8 years (3–19 years) (Table 1).

Figure 1 shows the development of the Cobb angle in all cases. The average final Cobb angle was 73.4° (20°–163°). Based on the Cobb angle at the final investigation, 18, 20, and 13 patients were categorized into the <60°, 60°–100°, and ≥100° groups, respectively.

In 44 patients, the age at which the Cobb angle was ≥20° was confirmed (Figure 2). The average age in the <60°, 60°–100°, and ≥100° groups was 13.6 years, 11.3 years, and 7.6 years, respectively. The age at which the Cobb angle was ≥20° in the ≥100° group was significantly lower than that of the <60° and 60°–100° groups ($P=0.044$ and $P=0.044$, respectively).

Figure 3 shows the different mean values of the Cobb angle by age in the three groups. A significant difference was observed in the Cobb angle from the age of 9 years between the <60° and ≥100° groups. From 13–19 years, a significant difference was observed among the three groups.

Table 2 shows the factors associated with the average of the final Cobb angles, and GMFCS, capability of turning over, hip dislocation, and gastric fistula showed significant differences with respect to the final Cobb angles in the three groups.

Regarding the location and type of curve, thoracic sco-

Table 1 Patient characteristics

Male : Female (n)	33 : 18
Type of cerebral palsy (n)	Spastic 42, Dyskinetic 7, Hypotonic 2
Classification of palalysis	Quadriplegia 45, Diplegia 5, Hemilplegia 1
GMFCS stage (n)	I 1, II 2, III 0, IV 6, V 42
Mean age at the first X-ray	9.4 (2–23) years of age
Mean age at last investigation	20.2 (15–29) years of age
Mean follow-up period	10.8 (3–19) years

*$P<0.05$.
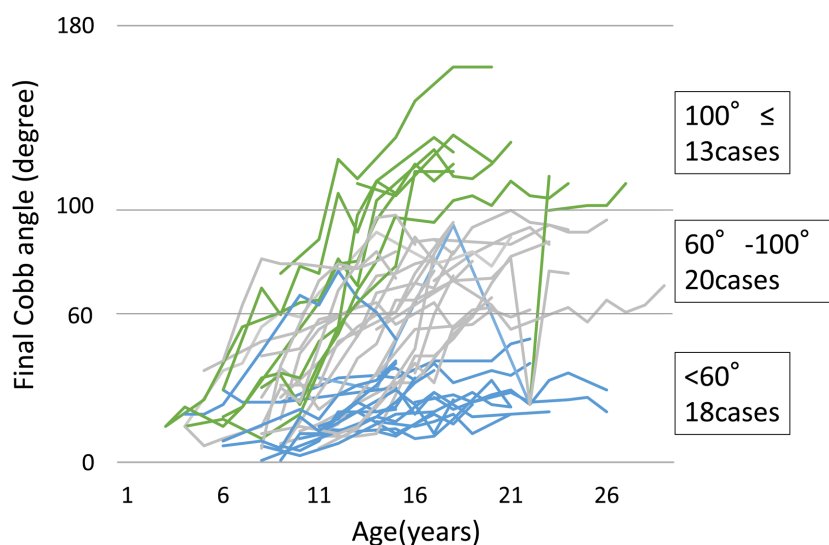
Figure 1 Development of the Cobb angle in all cases.
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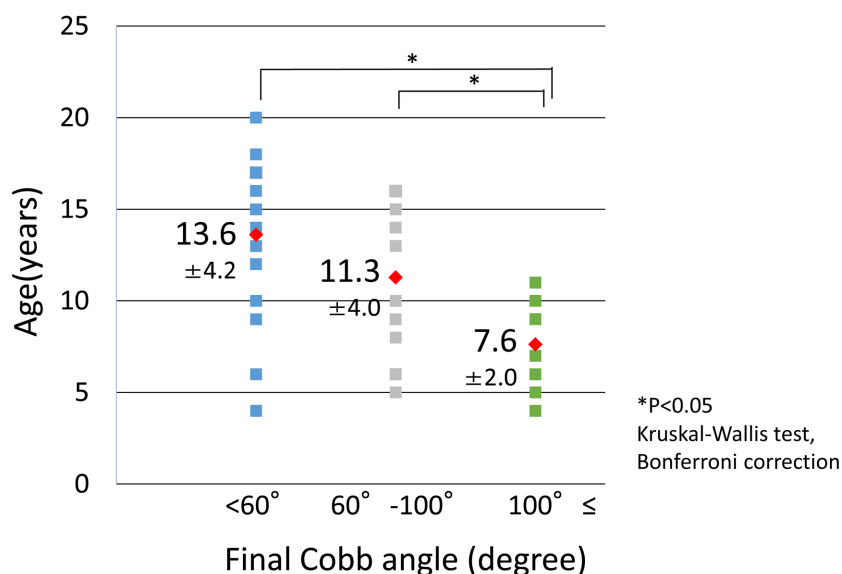

Figure 2 Detection age of Cobb angle ≥20°.
The average ages in the <60°, 60°–100°, and ≥100° groups were 13.6 years, 11.3 years, and 7.6 years, respectively. The age at which the Cobb angle reached ≥20° in the ≥100° group was significantly lower than that of the <60° and 60°–100° groups (P=0.044 and P=0.044, respectively).

liosis was observed in 16 cases (right of the convex side in 13 cases, left of the convex side in 3 cases), thoracic lumbar scoliosis in 20 cases (9 in the right, 11 in the left), and lumbar scoliosis in 15 cases (2 in the right, 13 in the left). Thirty-three patients had a single curve, whereas 18 patients had a double curve.

A total of 42 cases were classified as GMFCS level V, whereas 9 cases were at levels I–IV. Twenty patients (9 patients with GMFCS levels I–IV, 11 with level V) were capable of turning over, whereas 31 could not. Fourteen patients wore braces, whereas 37 patients did not. The types of orthoses used were rigid under-arm brace in 2 patients and dynamic spinal brace (DSB) in 12 patients. Two patients changed from a rigid brace to a DSB. The average Cobb
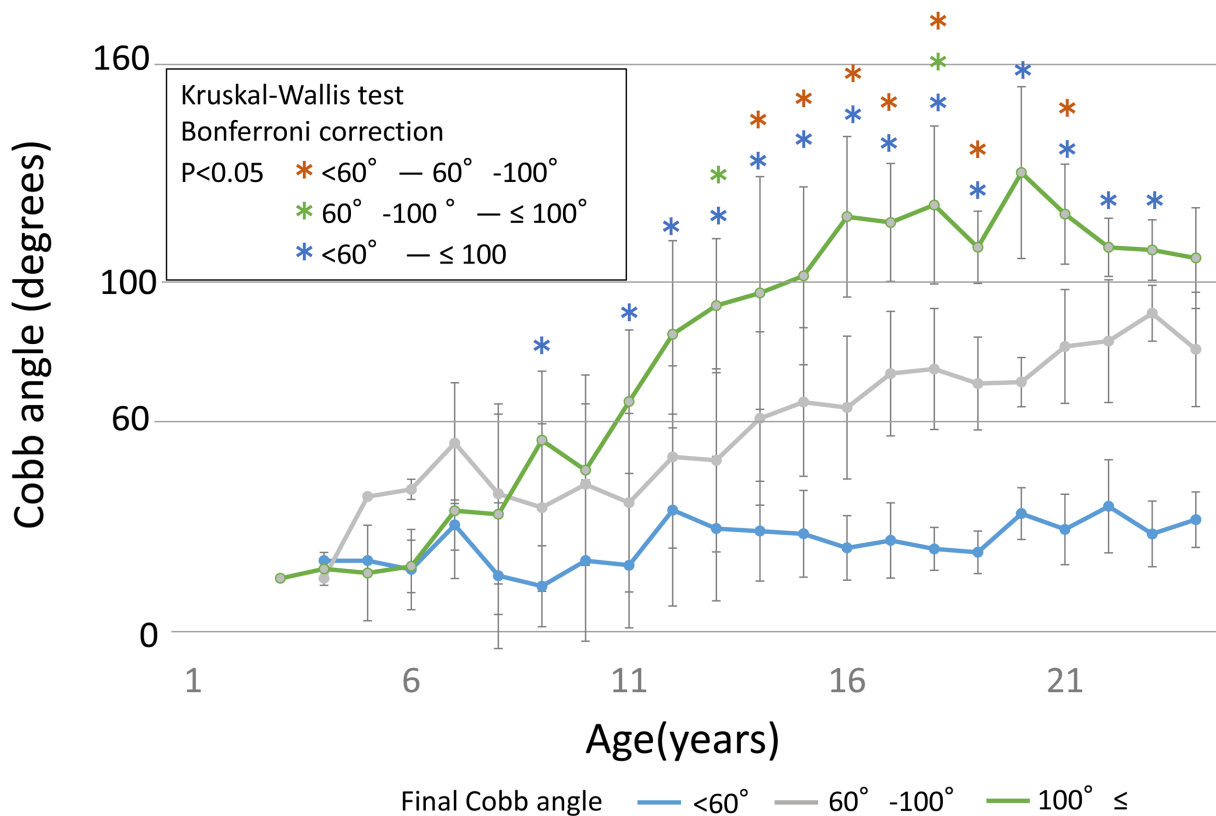
Figure 3 Mean values of the Cobb angle at different ages in the three groups.
A significant difference was observed in the Cobb angle at the age of 9 years between the <60° and ≥100° groups. From 13–19 years, a significant difference was observed among the three groups.

angle and age at the time of starting the brace were 54.8° (18°–106°) and 14.8 years (8.7–20.9 years), respectively. The average duration of wearing the brace was 3.3 years (1.4–6.6 years). Nineteen patients had hip dislocation with migration percentage of ≥100%, while 32 patients had migration percentage of <100%. Ten patients underwent tracheostomy, and 25 had gastric fistula.

Discussion

The patients with CP with the most severe scoliosis (Cobb angle ≥100°) developed the condition at a young age (average age 7.6 years), and the scoliosis progressed from 9 years of age. After the age of 13 years, the difference in Cobb angle among the <60°, 60°–100°, and ≥100° groups became significant, indicating that the scoliosis in these patients tended to worsen during the growth phase. This is comparable with the findings of a previous study that reported that scoliosis in CP is similar to adolescent idiopathic scoliosis[3]. During the growth phase, spasticity is enhanced by the relative shortening of the muscle length due to the rapid growth of bone, which is considered as the cause of the worsening of the scoliosis of CP during the growth spurt.

In addition, in severe cases of CP, paralysis, spasticity, and imbalance of the left and right sides are intrinsically strong; thus, it is believed that scoliosis develops and progresses early. Therefore, since the early onset cases can rapidly progress to severe cases, early interventions are necessary.

In this study, the factors associated with scoliosis severity were GMFCS level V, capability of turning over, hip dislocation, and gastric fistula. There are several reports on the factors related to severe scoliosis. Previous studies reported that the risk factors of severe scoliosis were GMFCS[7, 8], being bedridden[3, 4], spastic quadriplegia[4, 7, 9], hip displacement[7], thoracolumbar curve[3, 4], and early onset[4, 7]. The risk factors for scoliosis analyzed in this study (GMFCS level V, capability of turning over, hip dislocation, gastric fistula) were associated with severe CP. The scoliosis in patients with severe motor dysfunction tends to be severe.

In our study, the final Cobb angle was significantly larger in patients with unilateral and bilateral hip dislocations. There are several reports on the relationship between hip dislocation and scoliosis, but it has not yet been clarified[7, 10, 11]. Hip dislocation occurs due to the imbalance in spasticity and muscular strength of the iliopsoas, adductors, and hamstrings. Hip dislocation can lead to worse pelvic obliquity[10]

Table 2 Factors related to severity of scoliosis

		Cases	Average (degree)	*P*-value
Location of the curve	Thoracic	16	73.6 ± 41.2	0.704
	Thoaciclumber	20	78.3 ± 34.7	
	Lumber	15	66.5 ± 37.0	
GMFCS	I–IV	9	24.8 ± 6.8	0.000*
	V	42	83.8 ± 32.0	
Turniug over	Possible	20	40.9 ± 24.8	0.000*
	Impossible	31	94.3 ± 27.3	
Orthosis	Wearing	14	72.6 ± 21.9	0.874
	Not wearing	37	73.6 ± 41.6	
Hip dislocation	Presence	19	95.9 ± 29.8	0.002*
	Absence	32	61.1 ± 35.1	
Tracheotomy	Presence	10	91.3 ± 40.7	0.169
	Absence	41	69.0 ± 35.3	
Gastric fistula	Presence	25	90.1 ± 33.3	0.001*
	Absence	26	57.2 ± 33.7	

*$P<0.05$.

and even worse scoliosis. It is also necessary to investigate whether progression of scoliosis can be prevented by hip joint treatment.

Studies investigating the effect of using orthoses to prevent scoliosis progression are limited. Olafsson *et al*.[12] reported that successful treatment was observed in ambulating patients with muscle hypotonia and short thoracolumbar/lumbar curves measuring <40°, as well as in non-ambulating patients with spastic short lumbar curves. Moreover, it is difficult for caregivers to assist patients with CP to wear the orthosis made of hard materials, and patients may develop redness or decubitus ulcers in the bony protrusions. Therefore, compliance with wearing orthoses is poor. Kajiura *et al*.[13] developed the DSB, a novel device with the basic concept of automatic correction by maximizing posture control. Nakamura *et al*.[14] reported that DSB treatment may be effective when worn during the early stage of scoliosis. In our report, we did not find a significant difference in the final Cobb angle between patients who wore a brace and those who did not. It is important to consider that the age at which the patients started wearing the brace was 14.8 years, and the initial Cobb angle was 54.8°; thus, the time when the brace was started tended to be in the later stage of the scoliosis.

For the scoliosis of patients with CP, surgery is the definitive treatment for correcting the deformed spine and preventing its progression[15]. However, patients with CP often have complications such as respiratory disorders, gastrointestinal disorders, osteoporosis, and epilepsy; thus, spine surgery may be highly risky. In recent years, botulinum toxin injection[16] and intrathecal baclofen[17] have been used as treatment methods for controlling spasticity. Our study found that patients with early onset of scoliosis developed more severe scoliosis than those with 100° Cobb angle. For these cases, it is necessary to control spasticity during the early stage through methods such as internal medicine, botulinum toxin injection, and intrathecal baclofen. In addition, surgery for the scoliosis should be considered in certain cases.

The limitation of this study is that the sample size was small and the follow-up period was not sufficiently long, because this was a retrospective observational study. The scoliosis in patients with CP was reported to progress even after bone maturation[4, 18]. Thus, future studies involving a large number of patients and with a long-term follow-up duration, especially the period after bone maturation, are warranted.

Conclusion

Patients with CP with the most severe case of scoliosis, manifested as Cobb angle of ≥100°, showed scoliosis progression starting from 9 years of age. Therefore, early interventions to prevent scoliosis progression are necessary. Moreover, severe scoliosis is aggravated during the growth period, and GMFCS, capability of turning over, hip dislocation, and gastric fistula are the factors associated with scoliosis severity. Severe CP with low motor function levels and problems with internal functions was considered as the cause of deterioration of the scoliosis.

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