Introduction

Most children with severe cerebral palsy (CP) and chromosomal disorder have mental retardation and scoliosis, and their spinal deformity is usually progressive. The incidence of scoliosis in patients with CP has been reported to be >70% [1,2]. Despite brace treatment, the progression of scoliosis deformity does not cease even after reaching skeletal maturity; hence, quality of life and functional abilities of these patients significantly deteriorate [3,4]. Thus, scoliosis surgery is advised in children with severe and progressive spinal deformities to prevent functional deterioration of their cardio-pulmonary status and balance during sitting posture.

Recently, many authors have reported good treatment outcomes for spinal deformity correction using modern surgical techniques and instrumentation in patients with neuromuscular disorders [5,6]. Besides, patient/caregiver questionnaires regarding their satisfaction with surgery outcome has demonstrated high satisfaction scores [7]. However, the incidence of postoperative surgical complications is high [8,9]. Thus, surgery for spinal deformity in handicapped children (HC) remains controversial.

Abstract:

Introduction: This study aimed to assess treatment outcomes and caregivers’ satisfaction regarding scoliosis surgery for handicapped children.

Methods: Handicapped children are, by definition, noncommunicative and/or nonambulatory. We recruited 26 handicapped children who were followed-up for >1 year after a scoliosis surgery. We recruited 40 patients with adolescent idiopathic scoliosis (AIS) who underwent a surgery during the same period as controls. We used a posterior approach in all the children. We determined preoperative body mass index (BMI), main Cobb angle, Cincinnati correction index (CCI), and fusion level; intraoperative time and blood loss per level; and postoperative complications. We also assessed caregivers’ satisfaction with surgical treatments for these patients using the modified Bridwell’s questionnaire.

Results: We have described the results as handicapped children/AIS. Median preoperative BMI was 16.1/18.6 kg/m². Preoperative and final Cobb angles were 94.2°/59.7° and 39.7°/17.0°, respectively and CCI was 2.0/1.7. The number of fusion levels was 14.6/9.0. The operative time and blood loss per level were 40.1/44.1 minutes and 264/138 ml, respectively. Postoperative complications in handicapped children were adynamic ileus in 8 cases, dysphagia in 5, pneumonia in 3, urinary tract infection in 2, and superior mesenteric artery syndrome (SMA), surgical site deep infection, infectious enteritis, agitation, and liver dysfunction in 1 each. However, in the AIS group, there was only 1 case of SMA. Median caregivers’ satisfaction score on the 0-10 visual analog scale was 9. Caregivers for 19 of the 26 handicapped cases (73%) recommended surgical treatment to caregivers of other children with the same disease.

Conclusions: Surgical treatment for neuromuscular and syndromic scoliosis was associated with a high rate of postoperative complications. However, the caregivers’ satisfaction score after surgery was high.

Keywords:
Neuromuscular scoliosis, adolescent idiopathic scoliosis, caregivers’ satisfaction, questionnaire assessment, complication rate
We have been surgically treating patients with neuromuscular and syndromic scoliosis since 2012. This study aimed to assess treatment outcomes and caregivers’ satisfaction regarding scoliosis surgery for HC.

**Materials and Methods**

We recruited 26 HC, by definition, those with communication and/or ambulatory disabilities, who underwent scoliosis surgery from April 2012 to November 2015. Follow-up period was >1 year after surgery. The diagnoses included CP in 14 cases, chromosomal disorder in 5 (5p- in 3, 4p- in 1, and 1p36 del in 1), muscular disorder in 2 (Duchenne muscular dystrophy and unclassified congenital myopathy), and other syndromes in 5 (Coffin-Lowry syndrome, Rett syndrome, Smith-Magenis syndrome, and unknown in 2). Preoperative comorbidities in HC included epilepsy in 15 cases; otitis media in 3; asthma, mild atrial septal defect, and use of noninvasive positive-pressure ventilation in 2 cases each; and hydrocephalus, enlarged adenoids, and alpha thalassemia in 1 case each. No child had cardiovascular disturbance, urinary system disorder, or metabolic disease. Seventeen cases belonged to the noncommunicator and 21 to the nonambulatory categories. The median age at surgery was 14.9 years (11.6-19.4), and the median follow-up period was >1 year after surgery. The diagnoses included CP in 14 cases, chromosomal disorder in 5 (5p- in 3, 4p- in 1, and 1p36 del in 1), muscular disorder in 2 (Duchenne muscular dystrophy and unclassified congenital myopathy), and other syndromes in 5 (Coffin-Lowry syndrome, Rett syndrome, Smith-Magenis syndrome, and unknown in 2). Preoperative comorbidities in HC included epilepsy in 15 cases; otitis media in 3; asthma, mild atrial septal defect, and use of noninvasive positive-pressure ventilation in 2 cases each; and hydrocephalus, enlarged adenoids, and alpha thalassemia in 1 case each. No child had cardiovascular disturbance, urinary system disorder, or metabolic disease. Seventeen cases belonged to the noncommunicator and 21 to the nonambulatory categories. The median age at surgery was 14.9 years (11.6-19.4), and the median follow-up period was 1.7 years (1.0-4.9).

We additionally recruited 40 children with adolescent idiopathic scoliosis (AIS) in the control group who underwent scoliosis surgery during the same period as that of the HC group. There was Lenke curve type 1 deformity in 24 cases, type 2 in 9, type 3 in 2, type 5 in 3, and type 6 in 2. The median age at surgery was 14.3 years (11.4-17.2), and the median follow-up period was 1.9 years (1.0-4.8). Posterior approach was used in all the children, and all surgeries were performed by a single surgeon (NN).

We determined preoperative body mass index (BMI), main Cobb angle, flexibility\(^{10}\), Cincinnati correction index (CCI)\(^{10}\), and fusion level; intraoperative time and blood loss per level; postoperative non per oral and bed rest periods, hospitalization duration, complications, and number of other departments consulted. Additionally, in the HC group, we assessed caregivers’ satisfaction with surgical treatment for these patients using a specially designed questionnaire for caregivers of children with neuromuscular spinal deformities. The questionnaire was a modified evaluation method (ordinal variable) of Bridwell’s questionnaire\(^{10}\), with a 0-10 visual analog scale and several added questions.

Comparisons between the HC and AIS groups were performed using the Mann-Whitney U test, and those between the preoperative and postoperative questionnaire results were performed using the Wilcoxon signed-rank test. P-values of <0.05 were considered significant. SPSS version 24 (IBM Corporation, Armonk, NY) was used for statistical analyses.

**Results**

Table 1 shows the pre-, intra-, and postoperative data of both the groups. Compared to that of the AIS group, HC group had lower BMI (p<0.05), larger preoperative Cobb angle (p<0.05), lower flexibility of the spinal deformity (p<0.05), and larger postoperative remnant Cobb angle (p<0.05). However, CCI for the evaluation of the correction rate, considering preoperative flexibility, did not show statistically significant difference between the groups. In terms of surgery, the HC group showed longer fixation levels (p<0.05) and higher blood loss (p<0.05) than those of the AIS group. Post-surgery, the HC group had more number of nil per oral days (p<0.05) and a longer hospitalization period (p<0.05) than those of the AIS group. Meanwhile, there was no statistical difference in the time taken for initiation of wheelchair use between both the groups.

Postoperative complications are described in Table 2. Al-

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**Table 1. Pre, Intra, and Postoperative Data of Patients: Demographics and Radiographics.**

|                      | HC                 | AIS                 | p-value |
|----------------------|--------------------|--------------------|---------|
| **BMI**              | 16.1 (10.8-21.8)   | 18.6 (12.4-26.9)   | <0.05   |
| Preoperative Cobb angle (°) | 94.2 (65-129)    | 59.7 (45-87)       | <0.05   |
| Preoperative flexibility (%) | 35.2 (9.9-61.2)  | 45.7 (16.7-71.1)   | <0.05   |
| Postoperative 1 month |                    |                    |         |
| Cobb Angle (°)       |                    |                    |         |
| Final visit          | 39.7 (17-106)      | 17.0 (4-30)        | <0.05   |
| CCI                  | 2.0 (1.1-4.4)      | 1.7 (0.9-4.4)      | 0.3     |
| Fused levels (FL)    | 14.6 (11-17)       | 9.0 (5-13)         | <0.05   |
| Surgical time/FL (minutes) | 40.1 (32.3-58.4) | 44.1 (35.6-76.8)  | <0.05   |
| Estimated blood loss/FL (ml) | 264 (105-762.2) | 138 (42.3-396.8)  | <0.05   |
| NPO days             | 6.9 (3-28)         | 3.1 (2-15)         | <0.05   |
| Days of bed rest     | 7.6 (4-90)         | 5.3 (3-9)          | 0.1     |
| Hospitalization period (weeks) | 5.6 (2-22.9) | 3.4 (2.1-6)      | <0.05   |

Data are expressed as median (range; min-max).

HC, Handicapped children; AIS, Adult idiopathic scoliosis; BMI, Body mass index; CCI, Cincinnati correction index; NPO, Nil per oral; FL, Fused levels
though the AIS group had only 1 case with superior mesenteric artery syndrome (SMA), the HC group had many complications, including adynamic ileus, dysphagia, pneumonia, and other complications. The complications in ambulatory cases included adynamic ileus and severe agitation in 1 case each. Other complications were associated with nonambulatory deformities. Regarding postoperative management, the HC group required the cooperation from physicians of other specialties as well (Table 3).

Table 4 represents detailed results of the caregivers’ questionnaires. For 12 parameters, there was a statistically significant improvement in the caregivers’ scores between the postoperative and final visits: sitting balance (p<0.05), defecation (p<0.05), transfer (p<0.05), patient’s quality of life (p<0.05), and respiratory function (p<0.05). Median score for caregivers’ overall satisfaction was 10.0 (0.0-10.0). The final question “Would you recommend this surgery for other children with the same disease?” was affirmatively answered by 19 of the 26 caregivers (73%). Six caregivers were of the following opinion: “It is a major surgery; the patient and caregiver should themselves decide.” The caregiver of only 1 case, who suffered severe deep infection and required the removal of the implant, said he/she would not recommend the surgery.

Discussion

HC are often afflicted with severe spinal deformities, which have poor outcomes with brace treatments. In these cases, good treatment outcomes have been reported with scoliosis surgery using modern instruments. The surgery in these patients is aimed at correcting the spinal deformity, avoiding respiratory dysfunction, and maintaining stability while sitting. Additionally, improvements in bowel function and patient’s quality of life have also been reported, and the satisfaction scores of caregivers are high.

In our study, the preoperative profiles of the HC group were statistically different from those of the AIS group. HC were malnourished (lower BMI), had more severe deformities, and showed lower flexibility in their spine. Several cases had a CCI of >3 (Fig. 1); hence, the evaluation of flexibility of the deformed spine in HC was more difficult than in those with AIS. In the HC group, the surgeries were of long durations and with high amount of bleeding. This implies that the invasion of surgery for HC is excessive; resuming oral intake is later in HC than in those with AIS. On the other hand, there was no significant difference in the time taken to initiate ambulation on a wheelchair between the two groups as we encouraged the patients to sit up early after surgery to prevent respiratory complications. However, the HC group still developed many complications, which required seeking treatment help from physicians of other specialties. The duration of hospitalization in the HC group was longer than in the AIS group due to the complications. We believe that such surgeries need a multidisciplinary team.

The caregivers’ questionnaires did not see a reduction in score from the preoperative to postoperative period for any of the 12 parameters. There were significant differences in domains, including sitting balance, defecation, transfer, patient’s quality of life, and respiration. Despite post-surgical

### Table 2. Postoperative Complications.

| Condition                                      | HC | AIS |
|-----------------------------------------------|----|-----|
| Adynamic ileus                                 | 8  |     |
| Dysphagia                                      | 5  |     |
| Pneumonia                                      | 3  |     |
| Urinary tract infection                        | 2  |     |
| SMA                                           | 1  | 1   |
| Infectious enteritis                           | 1  |     |
| Agitation (as a psychiatric disorder)          | 1  |     |
| Surgical site deep infection (needed removal)  | 1  |     |
| Liver dysfunction                              | 1  |     |

Table 3. Physicians from Other Specialties Consulted during Postoperative Management.

| Specialty                          | HC | AIS |
|------------------------------------|----|-----|
| Pediatric neurology                | 10 |     |
| General pediatrics                 | 7  |     |
| Pediatric surgery                  | 3  | 1   |
| Pediatric emergency care           | 3  |     |
| Pediatric urology                  | 2  |     |
| Rehabilitation (swallowing)        | 1  |     |
| Infection control                  | 1  |     |

### Table 4. Detailed Results of Caregivers’ Questionnaires.

| Domain                          | VAS Preoperative | VAS Postoperative | p-value |
|---------------------------------|------------------|-------------------|---------|
| Sitting balance                 | 2.3 (0.0-10.0)   | 8.9 (0.0-10.0)    | <0.05   |
| Digestion                       | 2.0 (0.0-5.0)    | 3.0 (1.0-5.0)     | 0.36    |
| Defecation                      | 6.1 (0.0-10.0)   | 7.5 (5.0-9.5)     | <0.05   |
| Sleep                           | 7.1 (0.5-10.0)   | 7.3 (0.0-10.0)    | 0.74    |
| Transfer                        | 5.0 (0.0-10.0)   | 7.0 (5.0-9.5)     | <0.05   |
| Perineal care                   | 5.0 (0.0-10.0)   | 5.0 (0.0-10.0)    | 0.27    |
| Changing clothes                | 3.7 (0.0-10.0)   | 5.0 (0.0-10.0)    | 0.08    |
| Patient’s quality of life       | 5.0 (2.1-10.0)   | 7.0 (4.8-10.0)    | <0.05   |
| Caregiver’s quality of life     | 5.0 (1.4-10.0)   | 5.0 (0.0-9.5)     | 0.07    |
| Respiration                     | 6.7 (0.0-10.0)   | 7.8 (0.0-10.0)    | <0.05   |
| Analgesic medication            | 1.0 (1.0-1.0)    | 1.0 (0.0-4.0)     | 0.1     |
| Sociality                       | 10.0 (4.6-10.0)  | 10.0 (4.6-10.0)   | 0.59    |

Data are expressed as median (min-max). VAS, Visual Analog score.
scores for several cases in the handicapped children group were >3. Cincinnati correction index is calculated as postoperative correction/preoperative flexibility. The formulae are shown below: Preoperative Flexibility (\%) = (Preoperative Cobb angle–Supine bending Cobb angle) / Preoperative Erect Cobb Angle × 100. Postoperative Correction (\%) = (Preoperative Cobb angle–Postoperative Cobb angle) / Preoperative Erect Cobb Angle × 100.

Figure 1. The distribution of Cincinnati correction index.
AIS, adolescent idiopathic scoliosis

Figure 2. Female aged 14 years with cerebral palsy and nonambulatory deformities.
It was difficult for her to sit by herself, and she presented symptoms of gastroesophageal reflux disease. Preoperative Cobb angle was 125°.

complications, the overall satisfaction score for this surgical treatment was very high among the caregivers of HC (Fig. 2a, 2b, 3a, 3b). When asked if they would recommend this surgery for children with similar deformities, 73% of the caregivers gave positive replies; the caregiver of only 1 case with Duchenne muscular dystrophy who postoperatively developed deep infection of the surgical site gave negative reply.

The efficacy of this surgery for HC remains unclear among pediatricians, pediatric orthopedic surgeons, and physiotherapists who usually encounter and manage these cases. When we initiated this surgery for HC in 2012, the associated medical staff had their reservations about the outcomes, as previously reported by Lubicky15). However, as the outcomes became apparent, the number of parents opting for this treatment for their children with similar deformities increased.

Surgical treatment for neuromuscular and syndromic
scoliosis was associated with a high rate of postoperative complications. However, it had a high rate of caregiver satisfaction. The medical team caring for HC should be aware of the risks and benefits associated with scoliosis surgery.

Conflicts of Interest: The authors declare that there are no conflicts of interest.

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