

Case report

Anorexia and supracondylar fracture of the femur following surgery for bilateral lower limb joint contracture in a case of severe cerebral palsy with mental retardation: a case report

Ryoko Takeuchi¹, Hiroataka Mutsuzaki¹, Yuki Mataki¹, and Hiroshi Kamada²

¹Department of Orthopedic Surgery, Ibaraki Prefectural University of Health Sciences, Japan

²Department of Orthopedic Surgery, University of Tsukuba, Japan

Abstract

Objective: Postoperative management of children having cerebral palsy (CP) with mental retardation is difficult. This report presents a case of severe CP with mental retardation, which resulted in anorexia and condylar fracture of the femur following a surgery for bilateral lower limb joint contracture.

Case report: A 14-year-old male was diagnosed with CP having severity of Gross Motor Functional Classification System Level V, and severe mental retardation. Due to bilateral spasticity in hip adduction, knee flexion, and valgus of the feet, tenotomies of the adductors of both hips, bilateral lengthening of the hamstrings and bilateral tenotomy of the peroneal tendons were performed. At 4 weeks postoperatively, knee casts were used; thereafter, knee extension braces were attached for 12 weeks. After surgery, he showed severe anorexia for 8 weeks and weight loss of 8.8 kg (from 35.2 kg to 26.4 kg) during the 4 months of hospitalization. Six months postoperatively, he was diagnosed with supracondylar fracture of the right femur.

Discussion: Severe CP with mental retardation requires attention to the physical symptoms caused by stress related to the hospitalization, surgery, fixation by casts, and braces. Moreover, care must be taken not to increase the risk of fracture by fixation or immobility of the postoperative cast, because it can lead to the progression of low bone mineral density.

Key words: cerebral palsy, mental retardation, anorexia, fracture, contracture

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Introduction

Surgery for joint contracture, hip dislocation, and foot deformity due to spasticity is common in children with cerebral palsy (CP). However, CP is often complicated with mental retardation. In addition, stress related to hospitalization, surgery, fixation by casts and braces might cause physical symptoms in children having CP with mental retardation; hence, postoperative management is difficult. Children with severe CP are at a risk for low bone mineral density (BMD) and low-impact fracture¹. Risk factors for low BMD and fracture are severe CP, non-ambulation, anticonvulsant medication, joint contractures, immobilization after surgery, and poor nutrition^{2–6}. In this report, the patient had severe CP with mental retardation and was predisposed to the risk of low BMD due to antiepileptic medication and non-ambulation. In addition, bone atrophy was aggravated by anorexia after surgical stress and immobilization, which resulted in supracondylar fracture of the femur.

Case report

This case involved a 14-year-old male patient with CP. He was the first born in a twin pregnancy at 32 weeks gestation and weighed 1,968 grams. He was diagnosed with perivascular leukomalacia 1 month after birth. At the age of 7 months, he first visited the pediatrics department of our hospital and rehabilitation was initiated. At the age of 10 years, the spasticity in hip joint adduction and knee flexion

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Correspondence: Ryoko Takeuchi, MD, PhD, Department of Orthopedic Surgery, Ibaraki Prefectural University of Health Sciences, 4669-2 Ami, Ami-machi, Inashiki-gun, Ibaraki 300-0394, Japan
E-mail: a_boor88@yahoo.co.jp

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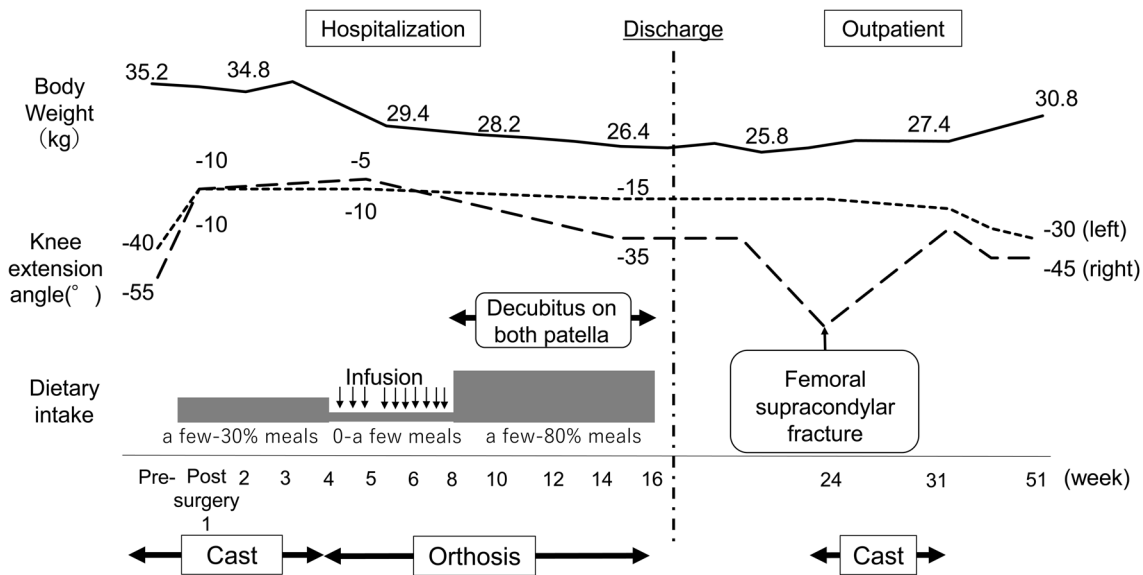


Figure 1 Progression of the patient's condition.

contracture progressed, and botulinum toxin therapy was started. However, the spasticity and contracture gradually worsened, with bilateral deformities of the feet and attempts at standing became difficult. At the age of 14 years, changing diapers and clothes became difficult. Hence, he was admitted to our orthopedic department for surgery.

Physical examination findings were as follows: height, 154 cm; weight, 32.5 kg; body mass index, 14.8 kg/m²; and Gross Motor Function Classification System (GMFCS), level V. He had impaired head control and turning over was possible only up to the prone position, but the flexed lower limbs tangled. Communication was impossible because of severe mental retardation and vocal discomfort. Due to complicated epilepsy, phenobarbital and carbamazepine were administered since the age of 5 months.

On examination, the knee extension angles were -55° (right) and -40° (left), popliteal angles were 90° (right) and 70° (left), knee flexion angles were 145° (right) and 135° (left), and hamstring muscles were shortened. Bilateral flexion contracture of the knees was detected. The hip abduction angles were 30° (right) and 10° (left), and abduction was severely restricted at the left hip joint. Both feet had severe valgus deformity. Changing the diapers and clothes was difficult due to the knee flexion contracture, hip joint adduction spasticity, and deformed valgus feet.

An X-ray image showed left convex lumbar scoliosis with a Cobb angle of 34° . The hip joints were not dislocated, and the migration percentages were 25% (right) and 25% (left).

Under general anesthesia, surgery was performed for Z-

lengthening of the semitendinosus, fractional lengthening of the semimembranosus, dissection of the gracilis muscle, and fractional lengthening of the lateral biceps femoris muscle at the popliteal region on both sides. In addition, bilateral tenotomy of the adductor longus at the groin and tenotomy of the peroneal tendon at the distal fibula were performed. The knee extension angle immediately after surgery improved to -10° on both sides. At 4 weeks postoperatively, a knee cast was fixed; thereafter, knee extension braces were attached for 12 weeks. The knee orthosis was made by molding, and the extension angle of the orthosis was adjusted according to the affected joint.

After surgery, hospitalization continued, and rehabilitation was initiated. Figure 1 shows the course of the patient's condition including dietary intake, weight, and knee extension angles during the 16-week hospitalization period and 35 weeks after discharge.

His dietary intake was poor, and it was only from several mouthfuls to 30% of his regular diet, for one month from the day after surgery. However, when the cast was changed to an orthosis brace, the food intake further decreased. He was listless and drowsy. Rehabilitation only included range of motion (ROM) exercises, and attempts at standing were poor. The urine volume decreased, and intravenous infusion was initiated. His nutritional status was evaluated by a nutrition support team (NST). After assessment of his eating ability, his diet was changed by an occupational therapist, and home-prepared meals and take-away food were allowed. Eight weeks postoperatively, the dietary intake gradually increased, and he consumed 80% of his meals. Albumin,

Table 1 Nutrition blood test

Post surgery (weeks)	Pre	Post 1Day	4	5	6	8	18	23	24	25	44
Alb (g/dl)	4.2	4.2	4.3	4.4	4.0	4.3	4.2	3.7	3.6	3.5	4.1
T-Cho (mg/dl)	–	–	–	184	177	–	178	155	159	159	136
Lym ($\times 10^3/\mu\text{l}$)	2.53	1.23	1.84	1.48	1.77	–	2.14	1.07	1.91	2.34	2.63

Alb: albumin; T-Cho: total cholesterol; Lym: lymphocyte counts.



Figure 2 X-ray images with antero-posterior and lateral views showing the right femoral supracondylar fracture. (a) At the time of fracture (2 months after discharge (6 months after surgery)). (b) At the time of fracture fusion.

total cholesterol, and lymphocyte counts are shown in Table 1. No major abnormalities were found in these values during hospitalization. However, at this time, his body weight decreased from 35.2 kg to 28.2 kg, and decubitus ulcers developed on both patellas, caused by the knee extension orthosis. As a result, the knee orthosis was not sufficiently attached, and the limitation of the right knee extension recurred. Four months postoperatively, the decubitus healed, and he was discharged from the hospital. At discharge, the knee extension angles were -35° (right) and -15° (left), and his body weight was 26.4 kg. Changing diapers and pants became easier, and the tangling of the lower limbs when turning over in bed disappeared.

At 2 months after discharge (6 months postoperatively), the patient presented at the pediatric department for fever and was admitted due to swelling of the right knee and diagnosis of a right femoral supracondylar fracture as shown on the X-ray (Figure 2a). With manual reduction under general anesthesia, fixation of the cast above the knee was performed. Seven weeks after reduction, the fracture was fused, and the femoral bone length was shortened by 14 mm (Figure 2b). After the cast was removed, the bone density of the femoral neck was measured with dual energy X-ray absorptiometry. Bone density values were 0.286 g/cm^2 on the right and 0.397 g/cm^2 on the left. The percentage of Z score

(the % value compared with the average value of the same age) was 36% on the right and 50% on the left.

His body weight gradually increased, and the weight recovered to 30.8 kg at 7 months after discharge (11 months postoperatively). The knee flexion contracture continued to worsen; however, there was no difficulty in changing his diapers and pants.

Written informed consent was obtained from the parent of the patient for publication of this case report and accompanying images.

Discussion

This report presents a case of anorexia and supracondylar fracture of the femur, following surgery for bilateral lower limb joint contracture in a patient with severe CP and mental retardation. Surgery and cast fixation can often cause physical symptoms such as food refusal, insomnia, and urinary incontinence, which are commonly seen in children of CP with mental retardation. Our patient had severe mental retardation and feeding was difficult. He was exposed to various stressors such as unfamiliar surroundings during hospitalization, pain due to surgery, fixation by cast and brace. As a result, he suffered stubborn anorexia. In such patients, it is important to obtain information from

the family, pediatrician, and nurse prior to surgery, about predictable patient responses. In this case, we focused on his strong refusal to cooperate with the NST, occupational therapist, and ward nurse. Thus, a multi-functional approach was necessary.

Children with severe CP are at a risk for low BMD and low-impact fracture⁹. The incidence of fractures with CP is reported at 15–37%^{2, 4, 7, 8}. Such fractures occur in the lower limb, especially the femurs^{9, 10}, and are associated with low BMD³. Henderson *et al.*⁷ reported that osteopenia (BMD z score <−2.0) was found in the femurs of 77% of 117 children with CP and 97% of all study participants who were unable to stand. In addition, King *et al.*⁸ reported that patients with non-ambulatory spastic quadriplegia with a history of fracture had significantly lower lumbar spine-BMD z score compared to those without a history of fracture. Risk factors for low BMD and fracture are severe CP as evaluated by gross motor function, non-ambulation, anticonvulsant medication, joint contractures, and immobilization after surgery, poor nutrition, greater body fat, and feeding gastrostomy^{2–6}. In this case, the patient was non-ambulatory with GMFCS level V and was on oral anticonvulsants, such as phenobarbital and carbamazepine, since the age of 5 months. In addition to the post-surgical cast fixation, the poor dietary intake and exacerbation of the bone atrophy led to a femoral supracondylar fracture. The patient's bone density was found to be remarkably decreased to 36% (right) and 50% (left), although it was an examination after casting for fracture.

To prevent fractures in children with CP, treatment to increase peak bone mass is necessary since early childhood. Adequate dietary intake of calories, protein, calcium, phosphorus, antiepileptic drugs, vitamin D, calcitonin preparation, vitamin K, and bisphosphonates should be administered. Recently, the effect of bisphosphonates on osteoporosis in CP children has been reported^{11, 12}. Additionally, there are several reports on the effect of weight bearing exercises in the improvement of BMD in children with CP. Moreover, weight bearing was reported to increase the BMD of the femur and vertebra of non-ambulatory CP patients^{4, 13, 14}. Other weight-bearing exercises have been reported to decrease the spasticity of the lower extremity with prolonged stretching¹⁵. Hence, weight-bearing exercises are important in children of CP with GMFCS levels IV and V in whom walking is impossible. Moreover, for continuous weight-bearing exercises, any contractures of the major joints should be avoided, and early intervention is necessary. In this case, the patient developed severe anorexia after surgery. In addition to the original osteoporosis, the poor nutritional status and immobility due to braces and casts accelerated the bone atrophy of the patient, resulting in su-

pracondylar fracture of the femur. Hence, it is necessary to explain to the patients' family and caregivers that there is a likelihood of fracture and careful attention is required.

Severe CP with mental retardation requires attention to the physical symptoms caused by stressors such as hospitalization, surgery, and fixation by casts and braces. Moreover, care must be taken not to increase the risk of fracture by fixation of the postoperative cast, which causes progression of low BMD due to immobility.

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