

Fractures in Duchenne Muscular Dystrophy —chiefly about their causes—

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ABSTRACT

Among 148 Duchenne muscular dystrophy children, nineteen cases (twenty-six fractures) were associated with bone fractures involving twelve femurs, nine humeri, four tibiae and one metatarsal bone. Seven out of twenty-six cases had experienced fractures twice. The causes of fractures were falls in fifteen cases, collision against surrounding objects in five, body position change in four and unknown in two. Femoral fractures were dominant during the wheelchair-bound phase, while, humeral fractures were dominant during the ambulatory phase. As these children lack sitting and standing balance as well as normal muscular power, we have to be careful of falls to prevent bone fractures when they are in a sitting or standing posture. Most of these fractures seem to be prevented if careful attention was paid during rehabilitation exercise, transfer and body position change etc.

Children with Duchenne muscular dystrophy (DMD) easily fall down due to poor balance and progressive muscular weakness, and as a result have bone fractures. However, few reports have been made about precise causes of bone fracture.

Therefore, we reviewed the DMD children with bone fractures, who had been hospitalized in National Sanatorium Hara Hospital during the past twenty-one years, in order to clarify the causes in detail and to prevent bone fractures.

SUBJECTS

For the period of twenty-one years, we have collected twenty-six fractures (nineteen cases) in DMD patients. The ages of fractures were 9-21 years old (mean 13.8). At the time of fractures three cases were of class 3, three of class 4, four of class 5, eight of class 6, five of class 7 and three of class 8, respectively (mean class 5.7). The class of physical activity capacity was presented by Ueda's classification (Table 1).

Table 1. Class of Physical Activity Capacity

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- | |
|---|
| 1. walks and climbs stairs without assistance |
| 2. walks and climbs stairs with assistance |
| 3. walks and rises from chair but cannot climb stairs |
| 4. walks but cannot rise from chair |
| 5. cannot walk but can crawl on all fours |
| 6. cannot crawl on all fours but can creep with other means |
| 7. cannot creep at all but keeps sitting posture unassisted |
| 8. totally dependent in bed |
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RESULTS

I. Site of Fractures

Twelve fractures occurred in femur, nine in humerus, four in tibia and one in metatarsal bone. Seven femurs were fractured on the right side and five on the left, respectively. All femoral fractures were seen at distal metaphyse. Two humeral fractures were seen on the right side and seven on the left side. The proximal metaphyse of humerus was fractured in three, diaphyse in four and the distal metaphyse in two (Fig. 1). The high incidence of left side fractures

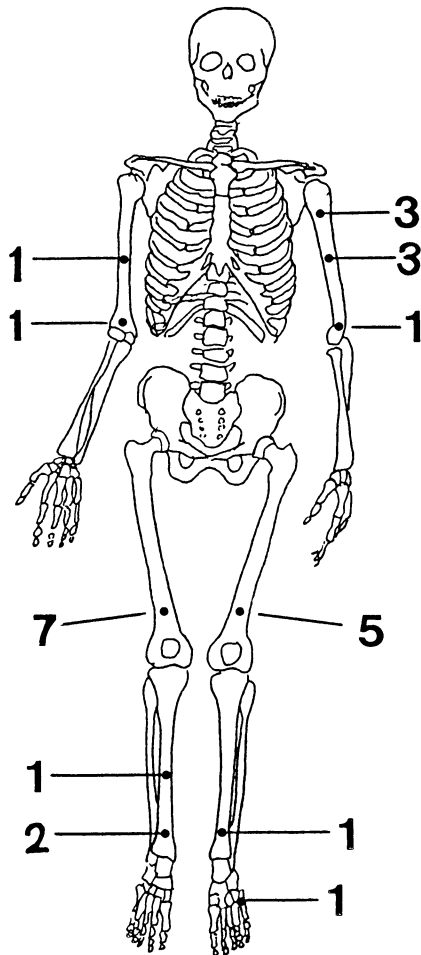


Fig. 1. Sites of fractures
Twelve fractures were seen in femur, nine in humerus, four in tibia and one in metatarsal bone.

might be related to the spinal deformity of right convex curve. The mean age at the time of fractures are 14.0 and 12.4 years old in the femur and humerus, respectively. Two cases were of class 5 at the time of femoral fractures, six of class 6, three of class 7 and one of class 8, and as for humeral fractures two was of class 2, three of class 4, two of class 5, one of class 7 and one of class 8, respectively. Five out of nine humeral fractures were caused during the ambulatory phase, while on the other hand, all of twelve femoral fractures during the wheelchair-bound phase.

II. Causes of Fractures

Fifteen fractures were caused by falls, five by colliding against surrounding objects, four by changing body position and two by unknown etiology. Among fifteen fractures due to falls, humerus was fractured in seven, femur in six, tibia in one and metatarsal bone in one, respectively. Two femurs, two tibiae and one humerus were fractured due to colliding against surrounding objects. Among four fractures due to body position change femur was fractured in three, tibia in one and no fracture was seen in the upper extremity (Table 2). Among twenty-six fractures, twenty-one were caused in the hospital and five in the patients' home during staying overnight.

A. Fractures in the hospital (21 fractures)

At the time of fractures five cases were on rehabilitation program, five in being transferred, four in riding on a wheelchair, three in changing the body posture, two in walking and the remaining two cases had fractures without be-

Table 2. Relationship between bones and causes of fractures

causes	Humerus			femur	tibia	metatarsal bone
	proximal metaphyse	diaphyse	distal metaphyse			
Falls	3	3	1	6	1	1
Collision against objects	0	1	0	2	2	0
Body Position Change	0	0	0	3	1	0
Unknown	0	0	1	1	0	0

ing realized. Among five fractures during rehabilitation exercise, two were due to falls from sitting posture on the mat, two due to falls in walking and one due to falls when standing up. All these fractures were caused due to loss of balance, which suggests that supervised rehabilitation exercise is mandatory for DMD children. All DMD patients who suffered from fractures during transfer were wheelchair-bound and two out of five fractures were caused during transfer from bed to wheelchair, one from toilet to wheelchair, one from bus to wheelchair and the remaining one from wheelchair to bath. As for the four children suffering from fractures during riding on a wheelchair, one had a fracture trying to brake, one trying to catch some object bending forward and the remaining two due to falling down out of a wheelchair.

B. Fractures in the domestic situation (5 fractures)

Four out of five fractures were caused by falls and at the time of fracture two of the four cases were sitting on the mat, one sitting on a toilet seat and the remaining one was held in his mother's arms. Only one patient had his right tibia malleolus fractured due to sprain of ankle joint, when he was made to sit up from supine posture by his mother.

DISCUSSION

The incidence of fractures was rated about 12.8% among 148 DMD children, who had received in-door services at our hospital for the past twenty-one years. Inoue²⁾ also reported fractures of 12.7% among 141 DMD children, who were admitted into his hospital.

As these data indicate, fractures are likely to occur in DMD children because they lack, (1) normal standing and sitting balance, (2) a normal bulk of muscle and fat with which to protect themselves during falling down and colliding against objects. These factors prohibit patients from reacting to situation that may lead them to unbalance or that may occur as a result of collision. Moreover, roentgen features of DMD show the signs^{4,5)} of, firstly, cocentric bone atrophy, secondly, soft tissue wasting, thirdly, periarticular osteoporosis, fourthly overgrowth of the epiphysse (Fig. 2). These factors might be altogether related to the high incidence of long bone fractures. Therefore, we have to seriously

consider the risks inherent in taking care of these disabled.

I. Fractures due to falls

Four out of fifteen fractures due to falls were concerned with a wheelchair. That is, the first case had the proximal metaphyse of left humerus fractured because a wheelchair was not braked on the descent. The second and the third case fell down from wheelchairs and had femoral fractures, when the former was trying to brake and the latter trying to catch an object in front of him. The last case of class 3 had fractures of left humeral proximal metaphyse, because a wheelchair fell down on the road being caught in the gutter, when fleeing from our hospital to go his home using an other's wheelchair on the midnight of his entrance into our hospital. Siegel⁶⁾ reported that the most frequent cause of fractures is falling down from a wheelchair and also recently, Hays³⁾ reported a case who was discovered belted in his motorized chair immobilized in a muddy pathway with a caster fork broken. This kind of accident might occur in America, because these care is taken at home against the possible dangers above mentioned and they ride on wheelchairs outdoors as well as indoors and they usually have close contact with neighboring community. Wheelchairs serve as their legs, whereas, in our country wheelchairs are mainly used in the hospital because most of the children are cared in the hospital. Even children who stay at home can not use wheelchair effectively because Japanese houses are not built to suit a wheelchair life. For example, corridors are extremely narrow and there are so many steps between rooms which prove serious barriers for a wheelchair. At our hospital, children sit in wheelchairs during the whole daytime except when they are on rehabilitation program on the mat and sit on a toilet seat, though, they seldom go outdoors using wheelchair. Therefore, outdoor accidents of wheelchairs are extremely rare. In the present study, only one patient was fractured outdoors. Nine of twenty-six fractures (35%) were related to wheelchair, that is, five of nine fractures were caused when transferring patients and four by falls from wheelchair as mentioned before.

As for the remaining eleven fractures due to falls, six cases were sitting at the time of fractures, that is, three were on an exercise mat,

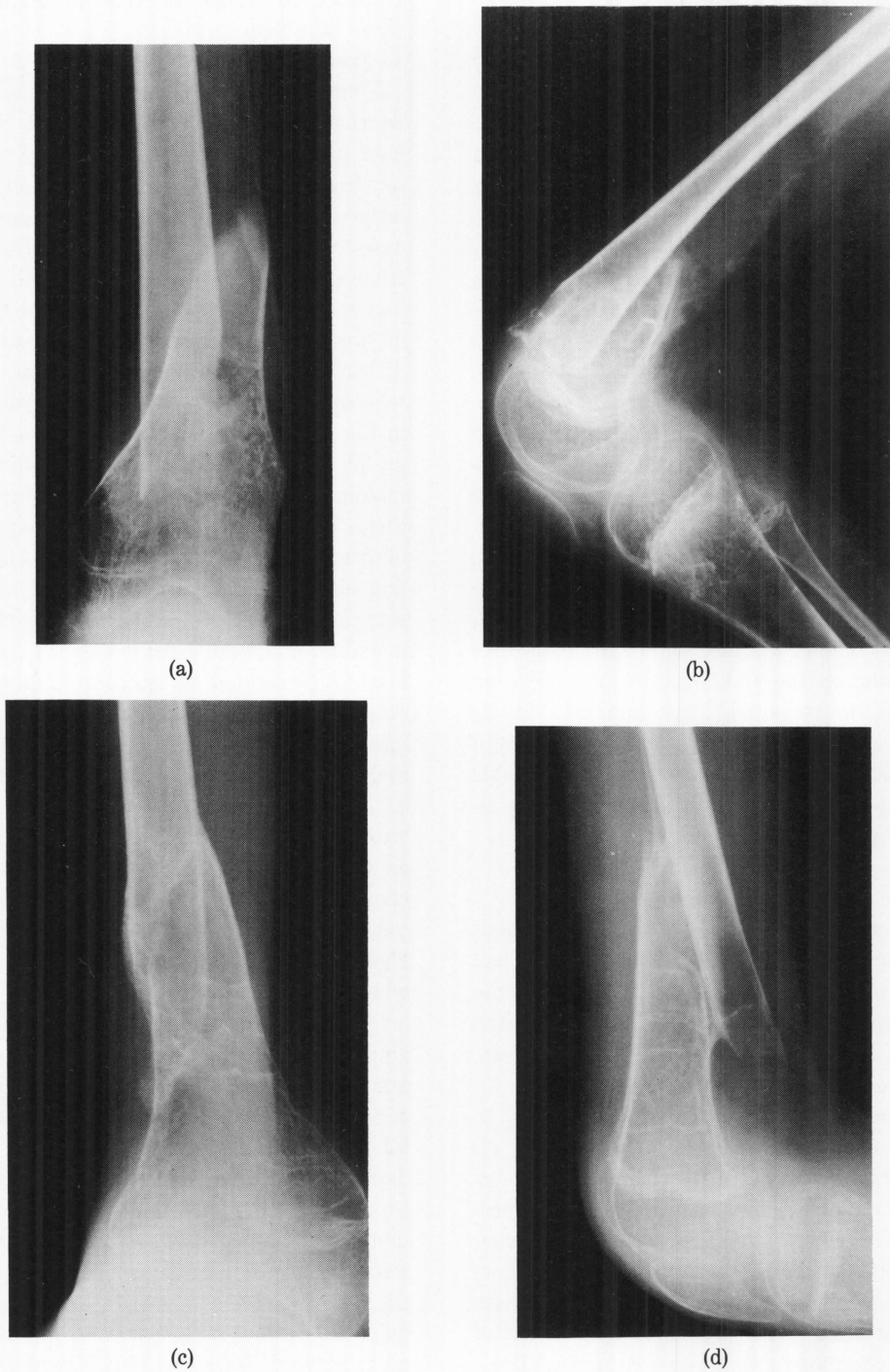


Fig. 2. Case. K.K. 17 year old boy of class 8

(a), (b). Left femur was fractured at distal metaphyse due to falls from supported sitting posture during rehabilitation exercise. Roentgen shows the typical findings, that is, narrow diaphyse, periarticular osteoporosis and overgrowth of epiphyses.

(c), (d). 5 months after fracture. Long leg cast was applied for three months. Fracture is healed with deformity and marked osteoporosis.

two on the mat at parents' home and one on a toilet seat.

The incidence of falls as a cause of fracture is much higher in the light class than in the severe class, because the less disabled are more active in their daily life than the severely disabled. For example, six out of eight fractures of class 6 were caused by falls due to loss of balance, whereas, none out of five fractures of class 7 was caused by falls. Sitting balance of these children of class 6 is still maintained, though, these accidents of fractures suggest that once the center of gravity breaks, they are likely to lose their balance and finally fall down. Therefore, safety belt should be fastened even for the patients of class 6, and their daily activities should be more frequently supervised.

II. Fractures by body position changes

Patients of an advanced class are unable to change their body postures by themselves during sleeping, which is performed spontaneously in the normal humans. Therefore, most of patients complain of pain or discomfort around the trunk or extremities at midnight and frequently call nurses to ask for body position change. This is not so much trouble in the hospital care, because nurses usually change their postures every two or three hours to make children sleep well. However, when children are cared at their homes, complaints of this kind prevent their family from sleeping well. This is one of the possible reasons why most of DMD children are cared at the hospital. At our hospital one team with two nurses performs position change during midnight at the utmost care not to give any trauma to these children. The knack of body position change is to roll a child as a log. Even with the utmost caution, two out of four fractures were caused at midnight during body position change and the remaining two were caused during body position change from supine to sitting posture at patients' home and from supine to lateral for cleaning the body.

III. Fractures during transfer

During transfer of the patients, fractures are likely to be caused due to colliding against surrounding objects. DMD children cannot only

avoid obstacles by themselves, but also due to less soft tissue and less bulk of muscle they can not absorb impact effectively when part of body collides against hard objects. All fractures caused during transfer were related to a wheelchair, that is, these accidents occurred when children were transferred from wheelchair to bed, toilet or mat. The objects against which patients collided were bed railing, floor, leg cast and foot rest of a wheelchair and a door of vehicle.

Fractures of DMD are easily overlooked because of minimal displacement of fractures and less pain due to little muscle spasm. Therefore, sometimes a precipitating cause of fracture was unable to be found. In our study two out of twenty-six fractures were caused without being noticed. Once patients have fractures, they easily develop marked muscle weakness and joint contractures, so that prevention of fractures is extremely important.

These affirm the need for careful nursing and supervision as well as the technique of transfer and body position change to prevent possible fractures.

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