THE MANAGEMENT OF EQUINUS DEFORMITY IN DUCHENNE MUSCULAR DYSTROPHY

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Equinus deformity of the ankle is one of the serious orthopaedic problems associated with Duchenne muscular dystrophy. Sixty-nine patients (age range 4 to 17 years) were treated, 43 conservatively and 26 operatively. They were followed up at six-monthly intervals for a minimum of two years and a maximum of six years. The patients were divided into three groups: independently mobile, mobile in calipers, and wheelchair-bound. It was found that conservative treatment could at best only minimise progression of the deformity. The indications for surgery, the operative procedure and the postoperative management are described; all varied according to the stage of the disease. The postoperative follow-up suggests that, though the deformity recurs, the patients have several years of benefit from the procedure.

Duchenne muscular dystrophy remains an incurable condition in which treatment can only be supportive. It is important therefore that any treatment offered is carefully considered with adequate knowledge of its implications. There are many interrelated problems that make a rigid regimen of treatment impossible. No consensus of opinion exists on the management of equinus deformity, one of the more serious problems, which begins at about five years of age and reaches its final stages in the late teens (Gardner-Medwin 1979).

In this paper we have examined the results of our treatment of equinus deformity.

PATIENTS AND METHODS

Of 105 boys with Duchenne muscular dystrophy currently attending our muscle clinic, 69 have been followed-up for a minimum of two years and a maximum of six years; the ages when they were first seen ranged from 4 to 17 years. This study has been confined to these 69 boys. The diagnosis was confirmed by enzyme tests, muscle biopsy, family history and the clinical course. All patients had an initial assessment in which general mobility, the power of individual muscle groups, joint contracture, lung function, and the presence of scoliosis were charted in detail. These assessments were repeated every six

months to record the progression of the disease. Once equinus was detected the angle between the sole of the foot and the horizontal was measured with a goniometer.

Conservative treatment

This varied according to the stage to which the condition had progressed. The patients were therefore considered in three groups.

Group 1 (independently mobile). Tightness of dorsiflexion of the ankle could be detected clinically as early as three years of age, although the walking pattern was quite normal at this stage. Regular passive stretching was advised. The contracture of the calf muscles increased progressively until by the age of six or seven years the foot could no longer be brought into the neutral position; that is, there was fixed equinus of the ankle. Stretching exercises supervised by a physiotherapist were continued, and individually moulded polyethylene night splints were provided, special care being taken to ensure that these were comfortable.

Group 2 (mobile in calipers). When the boys became so weak that they were about to become wheelchair-bound, cosmetic long-leg calipers were provided, in an attempt to prolong walking and standing (Spencer and Vignos 1962). The physiotherapy which they needed required cooperation from the patient and his family.

The footpiece of the caliper was moulded to fit the ankle and foot comfortably. Prolonged weight-bearing in the caliper had a stretching action on the calf muscles. The use of night splints was continued and if the boys were sitting for long periods, day splints also were sometimes provided.

Group 3 (wheelchair-bound). Stretching of the calf muscles was continued as part of a regular programme of physiotherapy. The boys spent most of the day sitting in a wheelchair. Support was provided for the feet to prevent them from dangling downwards; either the footplates of

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the wheelchair were adjusted or day splints were provided. When the foot deformity became severe, great resistance to wearing splints was encountered. It was therefore essential to check carefully that the splints were comfortable; if uncomfortable they would not be worn.

Operative treatment

The indications for operation were different in the three groups.

Group 1 (four patients). As weakness progressed the boys remained mobile by adopting a typical gait pattern: the hips and knees were held slightly flexed and a pronounced lumbar lordosis kept the centre of gravity of the body directly above these joints. Dynamic equinus completed the typical posture so that the children walked on tiptoe. Ultimately equinus contracture developed and when this fixed deformity was sufficient to interfere with balance, the calcaneal tendon was elongated surgically. It was most important that the tendon was not overlengthened as this could lead to loss of control of the ankle and to deterioration in the ability to walk. Allowing the foot to be brought to the neutral position was all that was required.

Group 2 (six patients). Of the three groups this was the most precarious, as incorrectly timed or mismanaged operative treatment could result in a loss of the ability to walk. However, a small number of boys who had adjusted well to calipers and who had sufficient power to use them, developed excessive equinus which interfered with their ability to stand or walk, and they need operation; their postoperative management was crucial and will be described later.

Group 3 (16 patients). This comprised the largest group. Those patients with marked foot deformity that had occurred within a few years of their becoming wheelchair-bound, were corrected surgically, even if the deformity was painless. Foot deformity which occurred in later stages of the disease (the late teens) was overshadowed by other problems; at that stage the indications for operative treatment were pain in the foot, pressure sores, or inability to wear normal shoes.

As the equinus contracture progressed it was associated with increasing varus deformity of the ankle and foot. Ultimately this could lead to subluxation of the midtarsal joints which could be painful. One boy had a stress fracture through the neck of the talus in each foot. Often the head of the talus became prominent beneath the skin on the lateral side of the foot. Despite efforts to fit suitable footwear, pressure sores were prone to develop.

Even in the boys who were mobile and active in wheelchairs severe equinus deformity was often disfiguring. In a few cases where such boys were unable to wear normal footwear and would not go out of doors because they were self-conscious in felt boots, we corrected the ankle equinus surgically.

Pre-operative investigations. Problems relating to anaesthesia in children with progressive muscular dystrophy

are well described and may include sudden death (Boba 1970; Ellis 1980). Careful pre-operative assessment of all patients was carried out, including lung function tests, electrocardiograms and serum electrolyte determinations. Suxamethonium, a neural blocking agent sometimes used in general anaesthesia, was strictly avoided as reports suggest it may be responsible for hyperkalaemic cardiac arrest and sudden death (Genever 1971).

As muscle strength decreased and particularly as scoliosis developed, the respiratory reserve declined (Burke *et al.* 1971). In our series the boys in Group 1 and Group 2 had an average lung function of over 75% of the predicted value, which was sufficient for general anaesthesia. Those in Group 3 had an average lung function of 60% of the predicted value.

In the later stages of the disease cardiomyopathy may occur resulting in a lowering of the cardiac reserve. In two patients (aged 9 and 14 years), the pre-operative electrocardiogram showed ST elevation.

Operative technique. This was similar in boys of Groups 1 and 2. Through a posteromedial incision the calcaneal tendon was identified and lengthened in Z fashion. The choice of suture material did not appear to be important; chromic catgut, silk, or nylon were used without any problem. All wounds were closed in layers and dressed. In boys of Group 3 simple elongation of the calcaneal tendon was often insufficient to allow passive dorsiflexion of the ankle to the neutral position. In these circumstances the tendons of flexor hallucis and flexor digitorum longus were divided and that of tibialis posterior divided or elongated. Posterior capsulotomy of the ankle and subtalar joints was carried out as necessary.

Of the 26 boys treated surgically, 25 had bilateral operations and one unilateral (the parents of this boy declined treatment to the second foot). In this boy, who was from Group 3, the foot deformity was gross and could not be fully corrected by lengthening the calcaneal tendon alone; the flexor hallucis, flexor digitorum longus and tibialis posterior were divided, but skin closure was not possible and the wound was allowed to heal by second intention. We have not been in favour of percutaneous tenotomy because of the risk of overlengthening the tendon in Group 1 patients and because it may be insufficient in Group 3 patients.

Postoperative management. Group 1 patients had a short-leg plaster applied with the foot held in the neutral position. Quick-setting plaster was used so that, with the help of the physiotherapist, walking was possible within 12 hours of operation. Prolonged bedrest was avoided as this causes marked deterioration of muscles. The boys in Group 2 were placed in long-leg plasters and, with the help of the physiotherapist, they too walked within 12 hours of operation. The risk of not walking again because of progressive weakness of muscle after treatment was significant, but could be overcome by a programme of vigorous physiotherapy. Group 3 patients were placed in a short-leg plaster with the foot in the neutral position.

Fig. 1

Natural progression of the equinus deformity in the 69 patients assessed at six-monthly intervals (185 observations in all). In each of the three groups the average fixed equinus (*) is shown with one standard deviation above the average. Observations made after operative treatment are not included.

Fig. 2

Figures 2 to 4—The effect of operative treatment on the three groups of boys. The bold curve derived from Figure 1 shows the average equinus occurring in the series when conservative methods alone are used. The dotted line shows the average equinus immediately before (first closed circle) and for the follow-up period after operation. Figure 2—Effect of operative treatment on the four Group 1 boys.

Fig. 3
Effect of operative treatment on the six
Group 2 boys.

Fig. 4
Effect of operative treatment on the 16
Group 3 boys.

All plasters were changed at two weeks when the sutures were removed and casts taken for splints. At six weeks the casts were removed, and boys in Groups 1 and 3 were provided with night splints only; boys in Group 2 were provided with ischial-bearing cosmetic calipers and night splints.

RESULTS

Conservative treatment. Figure 1 shows the natural progression of the equinus deformity in the 69 patients. It includes 185 observations made at six-monthly assessments; those made after operation were excluded. The results for each age group are shown as an average, with

the vertical line indicating one standard deviation above the average. We did not attempt to quantify the varus component but, in general, this deteriorated in proportion to the fixed equinus.

The average age at which the patients lost their mobility was 8.8 years (23 observations) and the average time at which they became wheelchair-bound, 10.5 years (19 observations). The period between these two values, 20 months, represents the average period of time that the calipers prolonged walking. It can be seen that at 10.5 years, the average age when the patients became wheelchair-bound, there was a marked increase in the equinus deformity.

Operative treatment. Figures 2, 3 and 4 show the follow-up results in the three groups. The smoothed curve is derived from Figure 1 and shows the average equinus. The follow-up begins (in each group) at the average age at which operation was performed. It begins with the average pre-operative equinus deformity and shows the subsequent six-monthly assessment values. All points on these graphs are a mean of at least four patients.

Complications. We have detailed records on the use of general anaesthesia in 30 patients. (Casting for splints, when the sutures were removed at two weeks after operation, was usually done without anaesthesia.) There were no cases of cardiac or respiratory arrest. Two boys had transient sinus tachycardia in the postoperative period but this subsided within 12 hours (Wislicki 1962).

The only other problem encountered was one case of delayed wound healing in the boy with a unilateral operation. Ultimately his wound healed without loss of correction.

DISCUSSION

In compiling the statistics for Figure 1, we were not able to differentiate between the various forms of conservative treatment. The provision of physiotherapy for these children, who come from a large catchment area, varied considerably. There was also variation in the degree of compliance by patients and parents, which meant that adherence to treatment regimens was unpredictable. For example, the wearing of splints tended to be intermittent, the most important factor being comfort. Clearly conservative treatment can at best only minimise the

progression of the equinus deformity, it cannot prevent it. Progressive foot deformity is an inevitable part of Duchenne muscular dystrophy.

The protective effect that prolonging the ability to walk has on retarding the development of contractures has been described by Siegel (1978). Our follow-up supports this finding, as there was a marked increase in the degree of fixed equinus when the children became wheelchair-bound. One of the benefits of calipers was in delaying the time when the boys became permanently confined to their wheelchairs.

The place of operative treatment is controversial; the mere presence of fixed equinus is not in itself an indication. From Figures 2, 3 and 4, it can be seen that initially those patients requiring operative treatment had excessive equinus for their age, as judged by the smooth curve representing equinus in the series as a whole. Operation was always able to eliminate the equinus, but eventually it always recurred.

As postoperative compliance with night splints varies, it is not possible to state whether their regular use reduces the rate or severity of recurrence. The pattern of recurrence paralleled the curve for the whole series. The effect of operation is not, therefore, permanently curative, but it transfers the patient from having equinus which is excessive for one particular stage of the disease, to having minimal equinus for that stage. This may appear at first sight to be only a slight gain, but in the context of Duchenne dystrophy, with the innumerable problems that develop, it is well worth while.

It has been suggested (Miller *et al.* 1982) that posterior tibial tendon transfer might prevent recurrence of the deformity. This procedure may be worth considering in Group 3, but in Groups 1 and 2 it may lead to loss of standing and walking balance because of the resulting dorsiflexion.

Providing the indications for operation are present, that pre-operative assessment is meticulous, postoperative management is vigilant and that the implications of operation are understood, then our results suggest that operative treatment has a part to play in the management of equinus in patients with Duchenne muscular dystrophy.

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