Scoliosis in Duchenne muscular dystrophy: aspects of orthotic treatment

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Abstract

The x-linked Duchenne muscular dystrophy (DMD) is the most frequent generalized muscle disorder arising from a lack of the sarcolemmic protein "dystrophin". Patients with DMD develop in the majority a progressive scoliosis when they cease walking and/or standing at the age of 10 years and become confined to a wheelchair. Increasing muscle weakness leads to a progression of the curvature, the pelvic tilt and problems in sitting. Together with the simultaneous progressive weakness of the respiratory muscles a restrictive pulmonary insufficiency will occur. Surgical stabilization of the spine (> 20° Cobb, forced vital capacity > 35%) by an adequate multisegmental instrumentation enabling early mobilization is now the treatment of choice.

However, orthotic treatment may offer an acceptable compromise in exceptional cases, if the patient rejects surgical intervention or is in the late (inoperable) stages of the disease. Such a treatment is superior to a primary sitting support provision with insufficient possibilities of correction.

The authors' experiences with 48 scoliosis orthoses made for 28 patients with DMD are reported. A "double plaster" cast has emerged as the best method to optimize adaption, especially in severe curvatures and the time taken for manufacturing the orthosis. A great deal of experience, patience and the consideration of the patients' individual demands are inevitable for a successful orthotic treatment.

Introduction

Besides contractures and deformities of the lower limbs, scoliosis is the most important orthopaedic problem in treating patients with progressive generalized muscle disorders (Forst, 1988). The particular problem of scoliosis in muscle disorders results from the fact that lung function is restricted both by spinal curvature and muscular weakness, which also affects the respiratory muscles. In contrast to idiopathic scoliosis there is a significant decrease of vital capacity even in scoliosis with only small curvature.

Rideau (1987) could prove for example that 13-year-old patients suffering from DMD with a curvature of 23° Cobb have a forced vital capacity (FVC) of only 40%. In addition many authors (Rideau et al, 1981; Jenkins, 1982; Kurz et al, 1983; Rideau, 1987) showed that after an increase up to the age of 10-12 (beginning of the wheelchair stage) vital capacity in DMD develops a plateau stage. Direct conclusions concerning the type of progression (rapid/slow) of the disease can be drawn from the time of occurrence and the level of this plateau stage (Rideau, 1987). A further progressive decrease of vital capacity of an average 200ml per year follows by which the average life expectancy can be estimated (Rideau, 1987). The knowledge of this typical decrease of absolute and FVC is decisive for the correct indication of surgical stabilisation of the spine in advanced stages of DMD. The time when patients with DMD develop a scoliosis coincides with the time they lose their ability to walk and stand. Patients with DMD cease walking ability at an age of 9-12 years. Most scolioses in DMD will not occur before the beginning of the wheelchair stage. The

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treatment of choice for scoliosis in DMD is nowadays the early surgical stabilization of the spine in the very beginning of the wheelchair stage, because the surgical risks arising from significant respiratory insufficiency and cardiomyopathy in later stages can be reduced. Optimal conditions are a proved progressive curvatures of more than 20° Cobb and a preserved FVC of at least 35%. The instrumentation (e.g. Luque, CD, Isola, etc.) extends from D2 down to L5 or SI depending on the pelvic tilt and should enable an early mobilisation of the patient.

This study reports a wide experiences with patients with DMD who refused spinal stabilisation or could not be treated surgically because of their general condition in the late stages of the disease. The indications and limits of orthotic treatment as well as the particular technical features in the adaption and manufacturing of the authors’ scoliosis orthoses are described.

Methods

Patients

A total of 224 patients with DMD (proved by lack of dystrophin) with an average age of 8.2 years at the first consultation in the authors’ department were examined and treated comprehensively by various conservative and surgical orthopaedic procedures. Some 132 patients were still able to stand and walk independently, 92 were in the wheelchair stage. After their first examination patients were first of all regularly examined by a standardized method every six months or in case of rapid evolution every three months. If the first clinical signs of a scoliosis occur an anteroposterior and a lateral x-ray has to be taken of the total spine in sitting position. Close clinical controls have to follow three or four monthly in order to record a progression of the curvature.

At Cobb angles of more than 20° with a proved progression a "prophylactic" stabilization of the spine is recommended using the Isola system between D3 and L5 or SI respectively depending on the extent of pelvic tilt. Patients who were inoperable or rejected an operation were treated with an orthoses (see below) and regularly examined every six months. The follow-up examinations included regular total-spine x-rays and pulmonary function tests.

Scoliosis orthosis

A thoraco-lumbo-sacral orthosis (TLSO) was made of low-pressure polyethylene and closed ventrally. Correcting pressure points have to use large pressure pads at the relevant points because of partly very thin subcutaneous adipose tissue. The extent of a possible correction is particularly determined by the rigidity of curvature, which is not great in early stages of DMD.

The plaster cast for the scoliosis orthosis is always made while sitting on a stool. After a good modelling of the pelvic contours it is important to seek the best correction of the scoliosis when fixing the plaster bandages in the trunk area. According to the rigidity of curvature manual pressure is exerted on the rib hump and is counteracted ventrally in accordance with derotation. The trunk is extended as well as possible and thus a "pre-correction" can be achieved.

There is no strict pattern for modelling the plaster positive but this has to take into account the individual patient (curvature, rigidity, etc.) An abdominal pad is not used so that ventilation is not restricted. In the abdominal area of the plaster model plaster is only slightly removed.

Fig. 1. Plaster positive modelled and corrected
In severe lumbar scoliosis a cuneiform mass of plaster is removed so that there is enough space between the lower ribs and the iliac crest to avoid impingement. At the same time plaster is removed at the armpit and at the greater trochanter of the opposite convex side. Depending on the extent of thoracic curvature a rigid lock is not used above the sternum.

In the rare case where patients are still able to stand and walk a hyperlordosis of the lumbar spine in the orthosis has to be tolerated to balance the forward movement of the trunk, which is caused by a hip contracture so that plumb line and balance are not impaired.

It is most important that after having finished the "primary" plaster positive (Fig. 1), a pre-plaster orthosis (negative) is made (Fig. 2), cut to size and tried on the patient. A new plaster positive is produced incorporating any modifications required in the pre-plaster orthosis. Over this positive the polyethylene orthosis is made according to known techniques. The advantage of this method of "double-plaster cast" is the fact that the pre-orthosis, made of plaster, can be easily and quickly changed or even wholly renewed if this is necessary. So the treatment as an in-patient to try and produce the final orthosis can be considerably reduced, to about one week on average, even in complicated cases with severe scoliosis (Fig. 3).

The orthosis can be reshaped at those parts where it causes bruises and the places for necessary correction pressure pads marked. In a further step the orthosis is finished by fixing an elastic lock above the sternum, a leather abdominal tab, 3 straps as abdominal lock and pressure pads of soft foam rubber. For a better air circulation the polyethylene orthosis is perforated at parts that do not fit closely to the skin.

Patients suffering from DMD cannot evade correction pressure by their own muscular strength, but will sink in the orthosis after a while. For this reason the skin has to be checked every 30-60 minutes after the polyethylene orthosis has been manufactured. This is the only way to detect and thus avoid developing bruises. This part of the work will take a lot of time, and a great deal of practical experience is needed until the orthosis is finally finished.
especially in patients at a late stage of their disease with severe curvatures.

**Results**

The age at first appearance of the scoliosis is listed in Table 1. Only those patients were included who were checked regularly before developing a scoliosis. The Cobb angles at the first detection respectively vary from 12° up to 117°. There are older patients with severe scoliosis at the first consultation. Some 81% of the patients were in the wheelchair stage when they developed scoliosis. The distribution of scoliosis to the respective parts of the spine is listed in Table 1. Regarding the main curvature 58% of the patients with muscular dystrophy showed a c-shaped thoracolumbar scoliosis with its convex side to the right. In addition to the scoliosis almost 100% had a more or less distinct pelvic tilt (0°-15°). The majority of these patients complained about pain when sitting in a wheelchair because of the one-sided load on one buttock. Some of them showed pressure sore formation and relapsing sciatic irritation. Many of the patients in advanced clinical stages had a severe restrictive pulmonary insufficiency and were in such a bad general condition of health, that the indicated

<table>
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<th>No.</th>
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<th>age at detection of scoliosis</th>
<th>extension of scoliosis</th>
<th>Cobb angle</th>
<th>side</th>
<th>shape</th>
<th>reason for corset treatment</th>
<th>number of corsets</th>
<th>previous early lower limb surgery</th>
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*missed follow-up for 2 years
surgical treatment of the scoliosis was impossible, mainly because of pulmonary problems. A large majority wished to improve their sitting position by the orthotic treatment and to achieve a greater length of the trunk. Some 28 of 44 patients with scoliosis in DMD agreed to wear an orthosis either because they themselves or their parents rejected surgical intervention or because their vital capacity did not allow an operation. Sixteen patients agreed to a surgical stabilization of the spine.

Because of growth and/or changed possibilities of correction scoliosis orthoses have to be altered during treatment (Table 1). Almost all the patients accepted their orthosis after an appropriate fitting. There is no sense in classifying the Cobb angles before and after orthotic treatment because each patient wears individually manufactured orthoses and the grade of scoliosis and the age of the patients is too different.

In a great number of patients it was possible to obtain data of lung function before and after orthosis adaption. Because of the heterogeneous population (age, general condition of health, symptoms) the results of these data can only be summarized in general terms: in all cases there was no measurable reduction of vital capacity or increase of residual volume after orthosis adaption. However, only a very slight improvement of vital capacity (maximal 100ml) and just an insignificant decrease of residual volume (maximal 10%) could be achieved.

Discussion

Duchenne muscular dystrophy has a characteristic and uniform clinical course. In the first stage (up to the age of 9-10) patients are able to walk and lead a relatively normal life. At an average age of 9.35 years (Forst and Forst, 1995) they cease walking and later lose standing ability due to increasing muscle weakness and contractures of the lower limbs. The patients will be confined to a wheelchair. Almost 90% of the patients in the wheelchair stage develop a scoliosis (Wilkins et al., 1976; Cambridge et al., 1987; Galasko et al., 1992) and 100% a marked respiratory insufficiency when they pass to the terminal stage. Even if the DMD cannot be cured it is possible to influence its clinical course by orthopaedic treatment. Operations on the lower limbs are performed when contractures begin to appear and a prolongation of walking and standing ability has been achieved later on with the aid of calipers (Forst and Forst, 1995). By keeping the orthotic assisted standing ability the formation of scoliosis, which is known to develop during the early wheelchair stage, can be delayed.

The orthopaedic treatment of DMD is a prophylaxis orientated comprehensive conception. From a therapist's point of view it is not the muscular weakness but the scoliosis which has to be seen as the most important problem. In contrast to idiopathic scoliosis, scoliosis in DMD is progressive even after the end of growth. As a consequence of the scoliosis and its resulting pelvic-tilt, patients complain about an impaired sitting position with loss of trunk stability and head control. Furthermore there is a deterioration of their angle of view and their cosmetic appearance. In particular they complain about the progressive impairment of pulmonary function.

A scoliosis treatment, especially the surgical treatment of scoliosis, aims at an increase of the quality of life by improving the sitting position and a changed respiratory condition improving nursing possibilities. In principle there are the following advantages of an early surgical treatment of scoliosis in DMD: there is less backache, less pain while sitting and no impingement of the ribs on the iliac crests. Both the cosmetic appearance and the sitting position are improved and, in addition, the handling of the wheelchair is facilitated. The prophylactic scoliosis operation, i.e. an early surgical treatment, offers an easier possibility of correction, an easier postoperative adaption, less respiratory complications as the patient has a better general condition of health at the time of operation and less blood loss. Many authors recommend the early surgical stabilisation of the spine (Jenkins et al., 1982; Kurz et al., 1983; Rideau et al., 1987; Gibson et al., 1987; Garfin et al., 1988; Miller et al., 1991; La Prade et al., 1991; Shapiro et al., 1992; Galasko et al., 1992; Hopf et al., 1994; Heller and Forst, 1996). A spinal stabilization in DMD is indicated at a Cobb angle of more than 20° and proved progression of the curve. An operation should be performed at a time when FVC is above 35% and with no cardiomyopathy to achieve a better postoperative performance.

Indications and limits of orthotic treatment vary. Vital capacity cannot be influenced by this
measure. A temporal improvement of pelvic-tilt, Cobb angle, comfort in sitting and of cosmetic appearance is however possible. A scoliosis orthotic treatment requires the provision of detailed information to the patients and their parents about a possible surgical treatment, which is definitely to be preferred. Both risks and limitations of orthotic treatment have to be mentioned. An orthosis should be adapted in case of a progressive scoliosis with a Cobb angle of more than 20° if both patients and parents reject a surgical intervention or if the general condition of health does not allow a surgical stabilisation of the spine.

The most important problem of orthotic treatment in DMD is the frequent formation of bruises for lack of soft tissue bolstering. Very often the orthosis has to be altered because of growth, loss or increase in weight and functional adaption. At the beginning of the treatment patients most often complain about pain at the buttocks and the iliac crest when sitting, problems in trunk control by a new regulation of the balance, problems in head control and their cosmetic appearance.

Many authors absolutely refuse orthotic treatment in muscle diseases, as they do not think there is an efficient improvement such as a positive influence on curvature or a decrease of progression. Up to now there have been only few reports on orthotic treatment of patients with muscle diseases in the international literature. These concentrate on DMD and spinal muscular atrophies (Morris et al., 1961; Gibson and Wilkins, 1975; von Kriichten, 1980).

Gibson and Wilkins (1975) emphasize that in muscle diseases an orthosis can support a spine that is still rather straight much better than one that is already deformed. For this reason they recommend an early start of orthotic treatment at the beginning of the wheelchair stage before a formation of scoliosis. Gibson and Wilkins (1975) found out that a great number of patients with DMD show primary kyphosis and that only a few of them develop lumbar scoliosis. For them the "ideal" orthosis has to work most efficiently in the sitting position. In addition they demand the horizontal iliac crest and hyperextension of the spine. They use an orthosis which is closed dorsally, exactly modelled to the iliac crest and which stabilizes the pelvis as it goes down to the greater trochanter. The trunk is mainly supported at the sternum and the rib-hump. An extension of the spine is achieved by pressure pads in the area of thoracolumbar transition together with the front elements of the orthosis (3-point fixation of the spine). Spine and pelvis are supposed to build a functional unit so that changing positions of the spine cause corresponding changes of the pelvis. For the manufacturing of the orthosis a plaster cast is made in the sitting position under slight distraction with a cervical device. The iliac crest is strongly moulded with a long compress because it is the pelvic contour that mainly supports the chest in the finished orthosis. To avoid an impeding of diaphragm movements there has to be enough space in the abdominal area. The whole front part of the orthosis is left intact to exert continuous pressure on the abdominal area (liquid cylinder effect), which then effects the straightening and partly takes the load off the spine (Morris et al., 1961).

Von Kriichten (1980) emphasized that the "ideal" orthosis for treating scoliosis in DMD has to avoid the development of the scoliosis. Furthermore it has to facilitate a comfortable sitting position, unrestricted respiratory function and aesthetic acceptability. The orthosis should not restrict daily life activities and must be easy to handle (dressing and undressing). The so-called "T-Orthosis", which was developed in Copenhagen, goes back to the Boston and Toronto Brace for idiopathic scoliosis (von Kriichten, 1980). It is mainly made of polypropylene and consists of a pelvic part which is inclined in the front plane and which is supposed to exert pressure on the lordotic lumbar spine. The proximal trunk parts are supported by modelling a double "T". The orthosis is closed dorsally by three straps. In this orthosis the deterioration of vital capacity is said to be far less than the reduction of respiratory function in untreated scoliosis.

Young et al. (1984) report about 9 patients with DMD, who did not show a scoliosis but had to use a wheelchair. They were treated with a modified metal Calot-supporting orthosis, moulded of medium dense polyethylene and an individually made "leather jacket". The orthosis fixed the lumbar area in the lordotic position that the patients were able to achieve in a short, active attempt to raise the pelvis without the orthosis when sitting. Both patients and
physicians in charge preferred the orthosis to the "leather jacket". The modified Calot-orthosis restricted lung function far less than the specially modelled "leather jacket".

Rideau et al. (1984) treated 10 patients with DMD by spinal orthosis or special seating in the wheelchair. In spite of these measures all their patients were unable to sit and showed severe scoliosis in the final stage of their disease. The authors concluded that in the progressive clinical situation the development of severe spinal deformation could not be avoided by a sole conservative treatment. For this reason they now recommend the prophylactic stabilisation of the spine in DMD right at the beginning of the wheelchair stage in order to avoid the appearance of advanced scoliosis.

The treatment of muscular diseases by sitting supports is also only rarely reported in the literature. In principle one can say that sitting supports should not be used in the primary treatment of scoliosis (Albrecht, 1983; Forst, 1988). Special seating can never work as a correction because of the three-dimensionality of the scoliosis. A treatment with a suitable seat may however be indicated as a palliative measure in untreated scoliosis with distinct spine and thorax deformation in very late stages (Robin, 1989).

Reports about a low acceptance of orthoses in muscle diseases (Bossingham et al, 1977) have to be judged in connection with the construction features. A great number of orthoses aim to cause an extreme hyperlordosis in the lumbar region, which is difficult for the patient to tolerate in the authors' experience. For this reason the aim is a medium pelvic inclination which basically corresponds to the pelvis position in sitting.

After initial difficulties in fitting scoliosis orthoses to patients with muscle diseases the authors found that bruises at the iliac crest and of the skin could be almost completely avoided by using foam rubber as pressure pads and by supporting the pelvis at the trochanters. Patients, who even in the orthosis showed a trunk deviation because of scoliosis curvature, had to be additionally supported by lateral pressure pads in an appropriately adapted wheelchair (fixed seat, fixed back with head support). In these clinical pictures the efficiency of the orthotic treatment largely depends on the empathy of the treating team (physician, technician, physiotherapist) and on the patient's (parents') cooperation. Regular follow-up examinations every 3 to 6 months focus on an early detection and correction of changes due to growth or the underlying disease.

In most of the cases the patients felt restricted because of problems when sitting, but also because of cosmetic reasons (trunk length). Even in severe scoliosis they never complained about respiratory problems. For this reason a treatment has to be offered, that is "between" an operation and the "palliative" measure of sitting supports. Yet, patients or their parents still have to be informed that an orthosis is only a "temporary compromise", which is never as effective as an early surgical stabilisation of the spine.

**Conclusion**

Without doubt the early surgical treatment of scoliosis in patients with generalized muscle disorders is the most effective and thus most appropriate method, even if it has not been settled, which instrumentation gets the best results in this clinical picture with regard to stability, derotation and possible complications.

A primary treatment of children (even at the beginning of scoliosis) with sitting supports has to be rejected even in "anatomic adaption" as there is no effective approach because of the existing elements of deformation of the scoliosis (lateral deviation and rotation).

The decision in favour of a treatment with a scoliosis orthosis in muscle diseases must only be made after careful consideration. From the authors' point of view the scoliosis orthosis is only a temporary compromise with limited (but acceptable) efficiency, which should be reserved for patients who reject an operation or patients who are inoperable because of their general condition of health. If the patient has decided on an orthotic treatment the physician should always (depending on the patient's age and situation) point to the necessity or efficiency as well as the status of a surgical stabilisation of the spine. In this way it can be avoided that the surgical intervention is considered as "a last resort" after a (predictably) unsuccessful orthotic treatment with a then much worse starting position (pelvic-tilt, marked curvature, thoracic deformity).

The use of a scoliosis orthosis in muscle diseases requires wide experience and patience.
on the part of both orthopaedic technician and physician to ensure the acceptance of the orthosis by the best possible wearing-comfort.

REFERENCES


