Lower extremity surgery in muscular dystrophy

J. Griffet, L. Decrocq, H. Rauscent, C. Richelme, M. Fournier

Pediatric Surgery Department, Parental couple—Child Hospital, BP 217, 38043 Grenoble cedex 09, France
Children Hospitals, Lenval Teaching Medical Center, avenue de la Californie, 06200 Nice, France
Pediatric Medicine and Rehabilitation Department, Pontchaillou Teaching Medical Center, 35033 Rennes cedex, France
Neuro-Sciences Unit, Physical Medicine Department, Archet Hospital 1, BP 3079, 06202 Nice cedex 3, France
Joseph-Fourier University, Medicine Faculty, domaine de la Merci, 38700 La Tronche, France

Accepted: 12 April 2011

KEYWORDS
Myopathy;
Tenotomy;
Tendon transfer;
Children;
Duchenne muscular dystrophy

Summary
Introduction: The natural progression of neuromuscular diseases results in inevitable musculo-tendinous contractures, most often in spite of early treatment. Surgery corrects this. The aim of this study was to evaluate the results of tendon surgery in these cases.

Materials and methods: Twenty children with muscular dystrophy underwent hip surgery (tensor fascia lata tenectomy, rectus femoris and sartorius tenotomy), knee surgery (gracilis, semitendinosus and semimembranosus tenotomies) and/or ankle surgery (lengthening or tenotomy of the Achilles tendon with or without posterior tibial tendon transfer). Articular range of motion was evaluated preoperatively, 6 months after surgery, at 1 year and at the final follow-up (7.4 years).

Results: Three children underwent surgery before they had lost their walking capacity, eight soon afterwards, and nine long afterwards. Surgery was bilateral in all cases: 38 hips, 12 knees and 36 ankles with 22 posterior tibial tendon transfers. Hip extension, which was limited by a 30° flexion contracture improved to 10° and stabilized at 14° at the final follow-up. Adduction which was −19° before surgery increased to 35° and had stabilized at 32° at the final follow-up. Knee flexion (38°) was only slightly improved (24°) and had regressed at the final follow-up. Equinus deformity (42°) was corrected to 9° of dorsal flexion with a slight loss (2°) at the final follow-up. Varus was improved from 11° to 6°, but this had regressed at 1 year (10°) and at the final follow-up (11°). Achilles tendon tenotomies resulted in 28° of dorsal flexion at 6 months compared to 20° with lengthening. Correction of varus was better with posterior tibial tendon transfer at 6 months, 1 year and at the final follow-up (11° versus 5°). In the three patients who underwent surgery before losing their walking ability, one continued walking for 1 year and two for 6 months. None of the eight patients who underwent surgery soon after losing ambulation were able to regain walking. All patients were able to continue device assisted upright positioning for a mean 3 years.
Introduction

Neuromuscular diseases which begin in childhood, such as Duchenne muscular dystrophy are progressive myopathies affecting the lower limbs and/or spine. Progression varies depending on the type of disease, but generally results in joint stiffening making it impossible to walk. The orthopedic deformities of the lower limbs which are a result of musculotendinous contractures due to fibroadipose degeneration of the muscle fibers, also make it particularly difficult to install the child in his/her wheelchair, to use orthotic devices (upright positioning or walking devices, braces) or even to wear shoes.

Even very early management with rehabilitation and orthotic devices are usually not enough to prevent the development and progression of symptoms [1]. Surgery can correct these deformities and allow the child to be placed comfortably in wheelchairs, braces or upright positioning devices. Recovery of assisted upright positioning is one of the aims of this technique, but the main aim is to release contractures to improve the child’s and his/her family’s quality of life.

Following the studies by Spencer [2] and Vignos [3] in North America or Roy [4], Dubousset [5] or B. Glorion [6] in France, numerous reports have confirmed that surgery can correct these deformities, facilitate upright positioning or preserve the ability to walk. These studies are based on those by Demos [7] who showed that preventing these position deformities was essential to prolong ambulation.

The aim of this study was to evaluate the results of surgery in the soft tissues and the orthopedic benefits to the child.

Materials and methods

This monocentric retrospective study included all children with Duchenne muscular dystrophy or a similar disease who underwent lower limb surgery at the University Hospital of Nice, France. Twenty-one files were collected and 20 were included (one patient was lost to follow-up) from February 1990 to April 2009. Twelve patients who consulted and were being followed-up during this same period did not undergo lower limb surgery. These children were regularly followed up at a multidisciplinary consultation for neuromuscular diseases and underwent a standard clinical examination every 6 months. The decision to operate was a consensus opinion of this group. A single surgeon performed all operations.

In patients with Duchenne Muscular Dystrophy, the surgical techniques proposed by Bonnet and et al. [8] were used. Associated interventions depended on articular involvement; in hips: extensive tenotomy of the m. tensor fascia lata and tenotomy of the m. rectus femoris and m. sartorius; in the knee, tenotomy of the m. gracilis, m. semitendinosus and m. semimembranosus; in the ankles, lengthening or tenotomy of the Achilles tendon with or without posterior tibial tendon transfer. Immediately after surgery, patients were placed in hip-foot plaster braces or thigh-foot braces which were changed to a circular plaster cast between D8 and D10. The patient was immobilized for 21 days then physical rehabilitation was begun with night postural braces and upright positioning devices if this was possible.

The choice of the intervention was based on the musculotendinous contractures identified during the preoperative clinical examination. The degree of hip flexion—extension, adduction—abduction and rotation, knee flexion—extension and ankle dorsal-plantar flexion was evaluated. Patients underwent postoperative clinical follow-up at 6 months, 1 year and the final follow-up (mean 7.4 years; range 1.4—14.7 years).

Statistical analysis

Results were expressed as means and ranges with a standard deviation (SD). To analyze the immediate and long term results of surgery in each patient, the preoperative range of motion of hips, knees and ankles were compared to those obtained at the 6 month, 1 year and final follow-up with the Wilcoxon test for paired series. The influence of lengthening or tenotomy of the Achilles tendon on equinus deformity or varus and of posterior tibial tendon transfer or its absence on varus and varus-valgus range of motion were studied with the Mann-Whitney test for non-paired groups. Statistical analyses were performed with Statview® 5.0 software (SAS Institute, Cary, NC, USA). P < 0.05 was considered significant.

Results

Most of the 20 files analyzed involved Duchenne muscular dystrophies (17 cases), there were two cases of congenital muscular dystrophy and one case of progressive muscular dystrophy.
dystrophy (PMD). All diagnoses were confirmed by muscular biopsy and genetic analysis. There were 19 boys and one girl with PMD. The mean age at surgery was 12.4 years old (6.7–26.2 years-old) (Table 1). The age when ambulation became impossible was noted.

Three children underwent surgery before they could no longer walk, eight soon afterwards (less than 1.5 years) and nine long afterwards (Table 1). Sixteen children underwent secondary spine surgery.

Six patients underwent surgery for all three joints, two for the hips, 11 for the hips and ankles and one for the ankles. Surgery was bilateral in all cases. Thirty-eight hips underwent surgery, 12 knees and 36 ankles. During ankle surgery, the Achilles tendon was lengthened 18 times and cut in one case. The different surgical procedures are described in Table 1.

In the 38 operated hips, preoperative range of motion was reduced in 19 children and flessum-abductum was present in 13 children. Improvement of preoperative flessum (38°) was moderate (24°) and regressed at the final follow-up (35°) (Table 2).

Table 1  All cases operated with surgical techniques used.

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Diagnosis</th>
<th>Age walking was lost</th>
<th>Age at surgery</th>
<th>Hips soft tissues</th>
<th>Knees</th>
<th>Achilles lengthen</th>
<th>Achilles tenotomy</th>
<th>Transfer TP</th>
<th>Tenotomy TP</th>
<th>Transfer TA</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 G</td>
<td>G</td>
<td>PMD</td>
<td>8</td>
<td>9.5</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 G</td>
<td>G</td>
<td>CMD</td>
<td>9</td>
<td>6.7</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3 B</td>
<td>B</td>
<td>CMD</td>
<td>12</td>
<td>13.7</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4 B</td>
<td>B</td>
<td>DMD</td>
<td>10</td>
<td>14.4</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5 B</td>
<td>B</td>
<td>DMD</td>
<td>9.5</td>
<td>14.3</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6 B</td>
<td>B</td>
<td>DMD</td>
<td>9.5</td>
<td>9.1</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7 B</td>
<td>B</td>
<td>DMD</td>
<td>9</td>
<td>9.9</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8 B</td>
<td>B</td>
<td>DMD</td>
<td>10.5</td>
<td>10.0</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9 B</td>
<td>B</td>
<td>DMD</td>
<td>11.5</td>
<td>12.1</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10 B</td>
<td>B</td>
<td>DMD</td>
<td>12</td>
<td>15.0</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>11 B</td>
<td>B</td>
<td>DMD</td>
<td>11</td>
<td>14.7</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>12 B</td>
<td>B</td>
<td>DMD</td>
<td>11.5</td>
<td>12.3</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>13 B</td>
<td>B</td>
<td>DMD</td>
<td>12.5</td>
<td>15.1</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>14 B</td>
<td>B</td>
<td>DMD</td>
<td>9.5</td>
<td>10.3</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>15 B</td>
<td>B</td>
<td>DMD</td>
<td>12</td>
<td>26.2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>16 B</td>
<td>B</td>
<td>DMD</td>
<td>11</td>
<td>12.2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>17 B</td>
<td>B</td>
<td>DMD</td>
<td>8.5</td>
<td>8.6</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>18 B</td>
<td>B</td>
<td>DMD</td>
<td>11</td>
<td>11.5</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>19 B</td>
<td>B</td>
<td>DMD</td>
<td>9</td>
<td>8.9</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>20 B</td>
<td>B</td>
<td>DMD</td>
<td>9.5</td>
<td>13.9</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

TP: M. tibalis posterior, TA: M. tibalis anterior; PMD: Progressive muscular dystrophy; CMD: Congenital muscular dystrophy; DMD: Duchenne Muscular Dystrophy.

At the final follow-up, there was no longer any difference between the two approaches. A comparison of the group with posterior tibial transfer to that without transfer shows that correction for varus was better at 6 months, 16° at 1 year and 16° at the final follow-up. A comparison of Achilles tendon lengthening and tenotomy shows that tendonotomy resulted in an improvement of 28° in dorsal flexion at 6 months versus 20° for lengthening, even if this difference was not statistically significant (P = 0.190). At the final follow-up, there was no longer any difference between the two approaches. A comparison of Achilles tendon lengthening and tenotomy shows that tendonotomy resulted in an improvement of 28° in dorsal flexion at 6 months versus 20° for lengthening, even if this difference was not statistically significant (P = 0.190).

Two slight complications occurred, one hypoesthesia of the arch of the foot which resolved and one superficial infection of the incision site in the ankle. One child died immediately after secondary knee surgery.

All children had postoperative posture braces, either hip-foot in case of knee flessum, or thigh-foot if there was no knee flessum. Six months later, only six children were still wearing braces. All were able to wear normal shoes, and could be comfortably installed in their wheelchair.

Fourteen children had electric wheelchairs with upright positioning assistance, six had simple electric wheelchairs,
but three of these had upright positioning assistance with a separate device. In the three children who underwent surgery before being unable to walk, one continued to walk for 1 year and two for 6 months with hip-foot braces with ischiatic support. None of the eight patients who underwent surgery soon after being unable to walk were able to walk again. They all continued device assisted upright positioning a mean 3 years after surgery.

Discussion

Musculotendinous contractures are inevitable in most children with myopathy. Early rehabilitation therapy and orthotic assistance are known to be indispensable in these patients [9]. Seeger et al. [10] showed that the equinus deformity progressed faster if physical therapy was discontinued and braces were not worn regularly. For Bakker et al. [1], the benefits of rehabilitation and braces disappear rapidly over time. Corticosteroid treatment has changed the course of this disease in the past few years (less significant rapid progression) and the indications for surgery [11,12]. Most of our cases are older and patients did not receive this treatment. This justified the indications for surgical treatment. However, the role surgery is not always clear in teams managing these children. Should surgery be performed? At what age? For what purpose? The aim of this study was to try to respond to these questions.

Lower limb surgery in patients with neuromuscular diseases is useful because it results in improvement by releasing contractures [5,6,8,13–18]. Dubousset and Que-neau [5] insists that surgery should be indicated in cases of asymmetry which worsens the progression of scoliosis. In our studies in Duchenne muscular dystrophy, surgery was especially beneficial in the hip with 20° improvement in extension and 55° in adduction. These results were stable over time with a long-term follow up of more than 7 years. However, the surgical techniques used were important. We used the surgical technique described by Glorion et al. [6,8] who recommended wide tenectomy of the m. tensor fascia lata extending to the lower third of the thigh. This procedure was probably the reason for our stable long-term results. Based on Dubousset’s recommendations [5], surgery was rarely performed in the knee. Our results confirm that improvement was not significant (from 38° to 24°) and correction was lost at the final follow-up. Surgery was limited to significant flessum (more than 30°), which makes it difficult for the child to sit or lie down. Indeed this degree of flessum often resists physical rehabilitation, and makes orthotic devices very uncomfortable. For the ankles, Achilles tenotomy resulted in better dorsal flexion (28° versus 20°) while posterior tibial tendon transfer seems to result in better varus correction (5° versus 11°). For Miller et al. [18], the best indication for posterior tibial tendon transfer is Duchenne muscular dystrophy for which they obtained 26/28 excellent results. Bach and McKeon [19] feels that walking is prolonged when this technique is used.

On the other hand, the results of surgery before or soon after ambulation has been lost, which is recommended by several authors [6,13–17] were not as beneficial as expected, with the ability to walk maintained for approximately 1 year, but with none of the patients able to begin walking again. However, there were very few cases in our study. In a large study of 428 patients, Forst and Forst [15] report that walking was prolonged by a mean 1.25 years in patients operated at the age of 6.56 years. However, for this author, the goal of early surgery should not be continued walking, but to maintain an upright posture by limiting deformities by releasing contractures [16]. For Forst, more stable long-term results are obtained with early surgery. Smith et al. [17] performed subcutaneous tenotomies before the ability to walk had been lost, and in his study continued walking was possible for 2 years after surgery with orthotic assistance. Goertzen et al. [14] showed that early surgery improved the diagnosis of scoliosis.

All the children who underwent surgery were able to wear normal shoes and did not require orthopedic shoes. Installation in a wheelchair was facilitated and assisted upright

### Table 2 Results of flessum-abductum in hips, flessum in the knee and varus-equinus in the foot.

<table>
<thead>
<tr>
<th></th>
<th>Preoperative (mean, SD)</th>
<th>Postoperative: 6 months (SD, mean)</th>
<th>Postoperative: Final follow-up (7.4 years) (mean, SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hips (38)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Flessum</td>
<td>30° (18°)</td>
<td>10° (20°)</td>
<td>14° (14°)</td>
</tr>
<tr>
<td>Adduction</td>
<td>-19° (-25°)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Knees (12)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Flessum</td>
<td>38° (17°)</td>
<td>24° (19°)</td>
<td>35° (32°)</td>
</tr>
<tr>
<td><strong>Ankles (36)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Equinus</td>
<td>42° (8°)</td>
<td>9° (12°)</td>
<td>11° (17°)</td>
</tr>
<tr>
<td>Varus</td>
<td>11° (7°)</td>
<td>6° (5°)</td>
<td>11° (11°)</td>
</tr>
</tbody>
</table>

SD: standard deviation.
positioning was preserved. For Bonnet et al. [8] surgical treatment, especially early, is justified because it improves the child’s and family’s quality of life. Unfortunately, our long-term retrospective study did not take this parameter into account, because there was no quality of life scale. Because many of these children have since died, this parameter could not be retrospectively evaluated, but it should be taken into account in future studies. Leitch et al. [20] did not find any difference in the ability to wear shoes, or in pain between operated vs non-operated children. On the other hand, the degree of equinus deformity was much greater in those without surgery.

We can therefore conclude that lower limb surgery for the hip and ankles makes it possible to release the contractures in these joints. In the knee, it seems reasonable to limit surgery to cases of flexion greater than 30°. Extensive tenotomy of the m. tensor fascia lata and posterior tibial tendon transfer improve results. Achilles tendon tenotomy results in greater immediate improvement of dorsal flexion.

This surgery is indicated as soon as contractures are significant and interfere with walking. In our opinion, the best period for surgery is when walking is about to be lost. However, the goal should not be to continue walking, even with orthotic support, but to release contractures so the upright position can be maintained as long as possible. Goertzen et al.’s [14] results suggest that this type of surgery can be beneficial in slowing the progression of scoliosis in cases of asymmetric contractures by decreasing the oblique pelvis and lumbar lordosis.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

Acknowledgements

We would like to thank Dr. Amandine Rubio for her help with the statistical analysis.

References


