Clin Orthop Relat Res
DOI 10.1007/s11999-010-1596-x

SYMPOSIUM: MYELOMENINGOCELE

Does Threshold Nighttime Electrical Stimulation Benefit Children With Spina Bifida?

A Pilot Study

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Abstract

Background Electrical stimulation is an established treatment for muscle weakness. However, traditional tectonic stimulation is poorly tolerated in children as a result of discomfort. Threshold electrical stimulation performed at night reportedly increases muscle strength in a variety of neuromuscular conditions and has been well tolerated in children.

Questions/purposes We asked whether threshold electrical stimulation treatment at night would increase strength and function in children with myelomeningocele.

Methods In a pilot study we prospectively followed 15 treated children who served as his or her own control. The patients were provided with a stimulator and instructed to use it on areas of muscular weakness during sleep, six nights per week. Followup phone calls at 2 weeks and then monthly were performed by a physical therapist to address patient concerns. Assessments of muscle strength, monofilament sensation, and physical function using the Functional Independence Measure for Children were to be performed at 3-month intervals up to 12 months and compared with the pretreatment assessment.

Results Only seven of the 15 subjects completed 9 months of treatment and none finished the 1-year study. The most frequent complaint was the treatment schedule was too intensive for the benefits received. We found small gains in muscle strength, gait, and bowel continence but no changes in physical function.

Conclusions Although threshold electrical stimulation made small improvements in muscle strength, the currently recommended treatment schedules are not practical for patients with myelomeningocele.

Level of Evidence Level IV, therapeutic study. See Guidelines for Authors for a complete description of levels of evidence.

This study was funded by Kosair Charities, Inc, Louisville, KY. The institutions of the authors have also received research funding from Shriners Hospitals Research grants, DePuy, Smith and Nephew, and Zimmer for projects unrelated to this study.
Each author certifies that his or her institution approved the human protocol for this investigation, that all investigations were conducted in conformity with ethical principles of research, and that informed consent for participation in the study was obtained.
This work was performed at the Shriners Hospital for Children, Lexington Hospital, Lexington, KY, USA.

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Introduction

Most children with myelomeningocele are left with some degree of lower extremity paralysis and neurogenic bowel and bladder dysfunction even with neurosurgical closure of the exposed spinal roots. Leg paralysis is managed by strengthening available weak muscles and the use of assistive devices to facilitate mobility. Bowel and bladder dysfunction is managed by bowel and catheterization programs as well as surgical diversion.

Neuromuscular electrical stimulation has been used to treat disuse atrophy of muscles in a variety of clinical conditions, including paralysis [4, 12]. In most treatments, the muscle stimulation is of such high intensity that actual contractions are induced. Because of discomfort, children poorly tolerate tectonic electrical stimulation. However,

Published online: 29 September 2010

muscle stimulation over long periods of sleep with low-intensity or threshold electrical stimulation (TES) has been tolerated in children with better success [8–10].

Pape et al. [8] found that TES used in children with spastic cerebral palsy improved their total gross motor, balance, locomotor, and receipt/propulsion skills as measured by the Peabody Developmental Motor Scales. Steinbok et al. [10] looked at the effect of randomly adding TES to their traditionally intensive postoperative physical therapy program after selective rhizotomy for spasticity reduction in patients with cerebral palsy. At 1 year post-rhizotomy, those children who used TES had a statistically better outcome based on the Gross Motor Functional Measure, a validated outcome instrument for children with cerebral palsy. Subsequent randomized studies in spastic cerebral palsy have demonstrated some effects of TES with subjective reports or measures but were unable to show an effect using objective measures [2, 9]. A recent review of electrical stimulation in cerebral palsy concluded the existing studies in the literature unfortunately had "insufficient statistical power to provide conclusive evidence for or against these modalities" [6].

For children with myelomeningocele, the problem is mostly flaccid paralysis, although they may have some spasticity. A pilot study with seven patients showed an improvement in gait for two patients and increased leg movement and strength in others (KE Pape-personal communication).

We therefore evaluated (1) the use of night TES on trunk and lower extremity muscle strength, joint motion, sensation, and walking in children with myelodysplasia; and (2) changes in physical functioning using the standardized Functional Independence Measure for Children (WeeFIM) [13].

Patients and Methods

We prospectively enrolled a convenience sample of 15 children with myelomeningocele aged 4 to 12 years (mean = 7 years old) between 1999 and 2000. The study was not blinded or randomized. Each subject served as his or her own control by comparing pretreatment with post-treatment assessments. There were eight boys and seven girls. Their paralysis had resulted in quadriceps muscle strength of 2/5 or less (thoracic/high lumbar n = 3) or 3/5 or more (low lumbar/lumbosacral n = 12). To be accepted into the study, all patients had to be involved in a stable physical therapy program, which remained unchanged during the treatment period. They also had to have a history of clinical compliance at our institution, no plans for surgery, and were able to make a 1-year commitment for treatment. Of the 15 enrolled children, only seven completed at least 9 months, and no patient completed the full year. The most common reason, cited by the families of 11 patients, was that the treatment schedule was too intensive. Three patients had unexpected fractures or surgery resulting in discontinuation of the treatment during this study. One patient each did not complete the study because they saw no improvement, failed to report problems with a malfunctioning unit, or were lost to followup. For the seven patients completing at least 9 months of the study, a separate analysis was performed. There were five boys and two girls with a mean age of 7 years (5–11 years). Among the seven patients completing at least 9 months of the study, two patients had thoracic/high lumbar paralysis and five patients a low lumber/lumbosacral paralysis. This study was conducted under the supervision of the Institutional Review Board at the University of Kentucky.

The stimulator we used, NeuroTech NT2000-TES (Biomedical Research Ltd, Donegal, UK), is clinically available and has a number of safety features making it practical for use in children. It has an automatic shutoff to prevent burns and shocks, a microchip to allow for program storage, and an output limiter to prevent overstimulation. The lead wire design prevents insertion into a wall socket as well as a concealed control panel to prevent unauthorized manipulation of the settings. The wave form parameters are as follows: 35-Hz signal with a contraction time of 12 seconds and relaxation time of 12 seconds. The pulse width is 280 msec with a 2-second ramp-up time and a 1-second ramp-down time.

Our program used electrode placements with one pair on the lumbar trunk and another pair on the gluteal muscles for patients with thoracic/high lumbar paralysis. For patients with lumbar/lumbosacral paralysis, one pair was placed over the gluteal and another pair over the quadriceps muscles. The pair of surface electrodes was placed over each muscle belly with approximately 1 inch between them. They were alternated each night from right to left sides of the body. Our physical therapist (SR), trained in the use of electrical stimulation, set the amplitude when a comfortable tingling sensation was felt in each muscle. This makes blinded studies with this technique difficult. Most patients' simulators were set at less than 7 mA. Families were instructed by our therapist to use the stimulator six nights per week. Our therapist made followup phone calls to the families at 2 weeks and monthly intervals during the treatment. She performed followup assessments at the hospital initially and at 3-month intervals up to 1 year. Patients kept treatment logs, but the stimulators did not measure compliance.

The assessments used were physical examinations, including uninstrumented manual muscle testing of the hip flexors, extensors, abductors and adductors, and knee flexors and extensors [5]; passive ROM of the hip, knees, and ankles [7]; and monofilament sensory testing of the

legs [11] by the same investigator (SR). The patients' walking was assessed by observational review of walking videos for velocity and walking mechanics and quantified further by the Progressive Ambulation Scale [8], which is a 10-point scale ranging from independent sitting (1 point) to independent walking (10 points). Overall physical functioning was measured by the standardized WeeFIM [13]. This outcome instrument was administered by a credentialed clinician (SR). The patient was instructed to perform certain tasks and their ability to perform the task was rated on a scale of 1 to 7 from total assistance to complete independence without devices timely and safely. The self-care score involves tasks in eating, grooming, bathing, dressing upper body, dressing lower body, toileting, bladder, and bowel for a total possible score of 56. Because all the subjects in this study walked, the locomotion score assessed walking 150 feet and going up and down one flight of stairs for a total possible score of 14. The mobility score includes the locomotion score plus chair, toilet, and tub/shower mobility for a total possible score of 35.

Outcome measures were compared with each subject's pretreatment state using an analysis of variance using StatView 2.0 software (StatView, Cary, NC). To determine sample size for a definitive study, we performed a post hoc power analysis using Lenth RV (2006-9) Java Applets for Power and Sample Size (www.stat.uiowa.edu/~rlenth/Power; accessed December 31, 2009).

Results

For the seven patients completing 9 months of treatment, all had improved strength in one or more lower extremity muscle groups. Hip flexion improved one grade for two patients, one to two grades for one patient, and declined one to two grades for one. Hip extension strength remained unchanged for all. Hip abduction increased one to two grades in two patients and declined one to two grades in one. Hip adduction improved two grades in two patients, one grade in two, and one to two grades in one and declined one to two grades for one. Knee extension improved one grade for three patients and one to two grades for two. Knee flexion increased one to one to two grades in one patient and one to two grades in two but declined one to two grades in one.

Passive joint motion of the hips, knees, and ankles was maintained during 9 months of treatment to within 10° of their pretreatment examination in all patients with the exception of one patient whose knee flexion declined from 150° to 120°. This decline did not interfere with function.

We observed no changes in threshold sensation with monofilament testing.

Only one of the seven patients changed on the Progressive Ambulation Scale and that patient improved by one level. Among the seven patients completing at least 9 months of the study two used long leg braces, and one of these who also used a walker improved his gait velocity and switched from a swing-through to a reciprocal walking pattern. Two patients used short leg braces and a walker; one progressed to crutches and the other improved their hip control and gait velocity. Two of the seven patients used short leg braces and crutches before treatment, one of whom had improvement in trunk sway during walking; the other patient, despite improvement in muscle strength, had no improvement in gait but he also had a component of arthrogryposis, which results in muscle fibrosis. The final patient used short leg braces and no assistive devices for walking before treatment. He had better control of foot progression with less trunk sway during walking.

Functional assessment for the 10 patients who finished 3 months of treatment indicated no change in the self-care, mobility, or locomotion subscales of the WeeFIM ($p = 0.98$, 0.86, and 1.0, respectively). We noted variable changes in WeeFIM scores for the seven patients followed for 9 months of treatment but with similar (self-care $p = 0.68$, mobility $p = 0.99$, and locomotion $p = 0.91$) pre- and posttreatment means. In the self-care domain, the group mean initially was 29.2 compared with 30.7 after 9 months of treatment. One patient decreased one level in functional ability. However, one patient each increased by one, two, or three levels. Three patients remained unchanged. In the mobility domain, the group mean remained unchanged with treatment at 16.3. Two patients decreased two levels, whereas one patient increased five levels. Four patients remained unchanged. The group mean for locomotion started at 10.7 and actually declined to 10.6 after 9 months of treatment. Based on the small beneficial effect on the WeeFIM self-care subscore and the variability in our study, a post hoc power analysis indicates that to find a 20% effect, a study such as ours would have to include 393 subjects.

All of the subjects were on bladder and bowel programs for incontinence. Subjective reports from most families indicated improved bowel continence but we did not objectively determine these improvements.

No adverse effects were noted with the electrodes or treatment. Malfunction in one of the units required that it be replaced.

Discussion

Physical function for children with myelomeningocele is affected by their muscle weakness. Tectonic muscle stimulation is a common treatment for muscle weakness in

adults but poorly tolerated in children. However, threshold nighttime electrical stimulation is well tolerated by children and reportedly improves muscle strength and function in cerebral palsy and myelomeningocele [8, 10, KE Pape-personal communication]. Our pilot study examined the use of TES in 15 children with myelomeningocele and evaluated its effects on muscle strength, joint motion, sensation, walking, and physical function as assessed by the WeeFIM.

Our study has certain limitations. First, as a pilot study, the limited numbers of subjects has the potential for Type II errors. A post hoc analysis on our most positive finding at 9 months on physical function, the WeeFIM self-care subscore, indicates that to demonstrate a clinically meaningful change of 20% would require a study with approximately 400 subjects. Second, children with myelomeningocele change over time. The absence of a control group makes it difficult to compare the changes in our patients with the natural history for the population in general. We chose subjects who had a demonstrated compliance in a stable physical therapy program because we knew the research study would require patient and family participation. Because most of our patients were receiving some physical therapy, we selected those in which the program and the patient participation were stable to minimize the effects on outcome variables of stopping or starting physical therapy during the study period. Third, the lack of blinding and randomization to treatment may introduce bias with outcome measures by the patients, therapists, and investigators. Because TES requires a tingling sensation to be produced, it makes blinding of the patient to the treatment difficult.

During TES treatment, all patients experienced improvement in strength in at least one muscle group. This compares with the previous study of Pape et al. (personal communication) in which they demonstrated qualitative changes of increased leg movement and strength in seven patients with spina bifida.

We found no meaningful change in joint motion. Similarly, no change in motion was observed by Dali et al. [2] in TES treatment in children with cerebral palsy. We also found no change in lower extremity sensation. Other studies have not reported on sensation, although patients in the study by Balcom et al. [1] reported increased sensation of pelvic fullness aiding in their urinary continence. Walking videos demonstrated improvements in walking velocity or gait mechanics in four of seven patients. However, walking function, measured by the Progressive Ambulation Scale, improved in only one patient. Dali et al. [2] also noted visual and subjective assessments that showed improvement with TES could not be borne out by objective measures.

Similarly, objective measures of physical functioning were statistically unchanged as measured by the WeeFIM. Only two studies have reported statistical improvements in functional scores using TES and both were in patients with cerebral palsy. Pape et al. [8] found improvements in the Peabody Developmental Motor Scales and Steinbok et al. [10] found improvement in the Gross Motor Function Measure over controls after selective posterior rhizotomy.

Most families in our study reported improved bowel continence. Improvements in bowel dysfunction have also been noted in children with myelomeningocele who were treated with intravesical electrical stimulation for their urinary dysfunction [3]. After a pilot study of TES to the lower extremities resulting patients reporting improvement in both bowel and bladder sensation, Balcom et al. [1] investigated the effect of TES on bladder function in 11 children with myelomeningocele. After 9 months of treatment to the abdominal, trunk, and gluteal areas, patients subjectively reported improved sensation of pelvic fullness. However, the first sensation of bladder fullness was not improved on cystometrogram. Bladder capacity, however, was improved, when adjusted for age, from 59% to 77% of normal. The mechanism of this is unknown but believed to be a result of increased protein synthesis, enhanced blood flow, or muscle innervation [1].

Our patients experienced no adverse effects with the threshold nighttime stimulation. Other authors report low rates of complications; most commonly were skin irritation from the electrodes or adhesives [2, 8].

We believe our most important finding was the high dropout rate from treatment in these children. Only seven of 15 patients completed 9 months of treatment and none completed a full year. The most commonly reported reason was the intensity of the treatment regimen. The intensity of our treatment program was similar to other TES studies that had reported 63% to 100% compliance for 6 to 9 months of treatment [1, 2, 8–10]. Dali et al. [2] found 13 of 82 patients left their study because they lost interest or found the treatment too inconvenient. Complying with an intensive treatment program may be a particular problem for the myelomeningocele population because the caregiver burden for them is already high. Any modality that does not produce immediate and dramatic results may be too much for families to continue. Although we no longer use this treatment at our center, further studies looking at the effectiveness of less frequent treatment schedules may be warranted. If efficacious, they would likely result in improved patient compliance and therefore outcome.

Based on reports of success with night TES, there was initial enthusiasm for this treatment by both patients and staff. After 9 months of treatment, we did demonstrate small gains in muscle strength, gait, and bowel continence. Despite these small improvements in muscle strength, TES produced no substantial change in physical function as measured by the WeeFIM. We did demonstrate a higher

rate of dropout rate of families than reported by others; the most frequent reason was the intensity of the treatment regimen.

Acknowledgments We thank David Pienkowski, PhD, University of Kentucky, Department of Orthopaedic Surgery, for his assistance in the data analysis and Christin Minter, MA, for her assistance in maintaining the grant accounting and obtaining permission of the Institutional Review Board.

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