

Pain in Neuromuscular Disease

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Although pain is a symptom commonly associated with several chronic debilitating neuropathic conditions, including diabetes and Guillain-Barré Syndrome, pain has not been described in the literature as a significant clinical problem in the major neuromuscular diseases (NMD). However, in our clinical experience many NMD patients complain of pain. This pain can have a negative impact on the functional abilities of those who experience it. There is a need for research to examine the frequency and extent of NMD-related pain and to examine the impact of this pain on psychological and physical functioning in persons with NMD.

Our goals in this project were to investigate the frequency, severity, and nature of pain in persons with neuromuscular disease; to investigate the relationship between pain, physical impairment, psychological well being and ability to perform activities of daily living; and to explore the effectiveness of pain therapies in the treatment of NMD-related pain. Our findings from this study have been published in four articles (Carter et al., 1998; Abresch et al., 2002; Jensen et al., in press a, in press b).

In this project we documented that, with the exception of those participants with spinal muscular atrophy, persons with slowly progressive NMD experience significant pain that was comparable to the pain reported by subjects with osteoarthritis and chronic lower back pain. Those who reported significant pain also reported lower levels of general health, vitality, social function, and physical activity. To a lesser degree pain was related to increased fatigue, reduced ability to cope adequately with stress, and sleep disturbances.

We also investigated a variety of therapies designed to reduce or to better manage pain. Among these were physical therapy, nerve blocks, biofeedback/relaxation training, acupuncture, magnets, massage, chiropractic visits, hypnosis, counseling/psychotherapy, mexiletine, neurontin, tricyclic antidepressants, narcotics/opioids, acetaminophen, aspirin/ibuprophen, muscle relaxants including benzodiazapines, or carbamazepine.

We found no single intervention that provided a great deal of pain relief for all patients, but many of the treatments appeared to provide at least some relief for some patients. The most common treatments utilized were over-the-counter analgesics: ibuprophen, aspirin, and acetaminophen. These provided only moderate pain relief. Physical therapy, narcotic analgesics, and massage were the next most commonly used treatments. Only the latter two were associated with moderate relief and more than half of the patients discontinued their use. The least used treatment was chiropractic visits. These seemed to provide the

greatest relief. We concluded that none of these treatments provided effective long-term relief for all of the patients. This means that any patient with NMD pain who is interested in pain relief deserves a trial of each pain treatment to determine which treatment or treatments are most effective for him/her. The findings from this study suggest that, in their order of effectiveness, the following treatments provide pain relief: chiropractic manipulation (tried by only a few patients), narcotic analgesics, muscle relaxants, massage, acupuncture, ibuprophen/aspirin, hypnotic analgesia, neruontin, physical therapy, counseling, tricyclic antidepressants, biofeedback/relaxation training, acetaminophen, carbamazapine, and magnets.



Because fatigue is known to significantly impact quality of life, we also conducted an open label trial of a novel pharmaceutical agent, modafinil (Provigil) to determine whether it would be effective for treating fatigue in amyotrophic lateral sclerosis (ALS). Fifteen patients with ALS were treated for two weeks with either 200 or 400 mg of modafinil. Reported side effects of the medication were mild, and included diarrhea, headache, nervousness, and insomnia. Side effects did not result in any study dropouts. Following treatment, mean scores on the Fatigue Severity Scale (FSS) and on the Epworth Sleepiness Scale (ESS), significantly Mean scores on the self-report version of the Functional Independence Measure (FIM-SR) increased significantly. This pilot study suggests that modafinil may reduce symptoms of fatigue in ALS. Further investigation of modafinil in this setting is warranted.

We anticipate that the independence and quality of life of persons with slowly progressive NMD will be improved by a better understanding of the relationship between pain and the performance of activities of daily living. As our results are disseminated, physicians will become more aware of the problem of pain in NMD and will address it in their practices to provide their patients with the opportunity for a reduction in their pain and, as a result, an improved quality of life.