The Effect of Home Exercise and Electrical Stimulation on a Patient with Benign Hypermobility Syndrome

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Abstract:

Background and purpose: Benign hypermobility syndrome may be an unrecognized underlying cause of arthralgias, myalgias, clumsiness, and dislocations. The marked laxity of structures supporting the joint allows excessive joint motion, causing pain without evidence of inflammation. This study examined the influence of a home exercise program including the application of an electrical muscle stimulator (EMS) unit on the symptoms of pain and decreased activity level in a patient with benign hypermobility syndrome.

Case description: The patient was a healthy 22 year old male who was limited in his ability to sleep, to participate in sports, and to perform the requirements of his job secondary to muscular pain associated with benign hypermobility syndrome. He performed a daily home exercise program, including application of an electrical muscle stimulator, for a period of six months.

Outcomes: After six months, the patient was able to sleep and to perform his job without pain. Participation in sports remained limited.

Discussion: The intervention utilized helped this patient control his symptoms of pain that interfered with sleep and work.

Key words: benign hypermobility syndrome, exercise, EMS, chronic pain

Introduction:

Benign hypermobility syndrome (BHS) may be an underlying cause for chronic pain in both children and adults.[4,5] It may present as migratory pain without evidence of swelling or inflammation. It is important for physical therapists to recognize this syndrome so that proper intervention can occur.

Benign hypermobility syndrome (BHS) tends to be a familial trait characterized by excessive motion in the spine, temperomandibular joint, and in the peripheral joints.[1, 2, 3, 4, 5] This occurs because of deficiencies in the formation of type I collagen, which is a major component of the joint capsule and ligaments. The usual mechanism for maintaining joint stability is thereby impaired.[6] It is distinguished from other syndromes involving hypermobility, such as Ehlers-Danlos syndrome and Marfan syndrome by lack of involvement of the skin extensibility and by genetic testing. People with BHS often excel in activities that require flexibility, such as gymnastics, ballet, or wrestling.[7] They also do well in musical performance, especially the stringed instruments and piano. They develop pain when the poor joint stability leads to frequent, though mild, joint subluxations, or when they need to maintain a prolonged static posture.[5, 8] Most patients report that sleep is interrupted frequently by pain. BHS is relatively easy to detect, using Beighton's joint assessment scoring system.[9] Most patients try to control the pain through non-steroidal medications, through the use ofmodalities such as heat and cold, and/or by modification of activity. No one treatment has been shown to be effective. This article describes one patient who presented to physical therapy with BHS. He adhered to a graded home exercise program that was augmented by electrical muscle stimulation, for

a period of six months. His response to the intervention was slow, but after six months, most of his symptoms were resolved.

Literature Review

To determine a diagnosis of benign hypermobility syndrome (BHS), the examiner uses a Beighton scoring system to assess joint hypermobility. A score of four or greater is required for diagnosis. A patient receives one point for each of the following findings: knee or elbow hyperextension beyond 10°, the ability to touch the palms of the hands to the floor with knees straight, passive extension of the 5th MCP joint beyond 90°, and passive apposition of the thumb to the forearm. Since points can be given one for each elbow, knee, thumb, or 5th finger, there is a possible total score of nine points.[9] Exam reveals an absence of inflammatory signs, including tenderness, swelling, redness, or warmth. Laboratory tests for complete blood count, erythrocyte sedimentation rate, rheumatoid factor, antinuclear antibody titer, and levels of serum immunoglobulin and complement are all negative.[3] The examiner will also elicit a history of multiple joint subluxations, chronic joint pain, and often a family history of similar symptoms. [3,4] Children with BHS often experience gross motor delays due to poor joint stability. They report being "late walkers".[3] They may complain of being clumsy. Pain often develops during adolescence, and continues into adulthood. In Sachetti's study, adults with BHS typically report eight painful joints or periarticular regions, with an average intensity of 5/10 on the McGill scale. Headaches affect half of the patients.[5] Psychological well being is affected. Lumley reported that these patients experience higher levels of depression and anger, directed not only at the pain, but also at the perception that there is no systematic approach to treatment. [10]

It seems reasonable to wonder whether the increased articular gliding that occurs as a result of poor restraint mechanisms and by poor proprioception would lead to an increase in degenerative changes. Lewkonia reports that there is an increase in osteoarthritis in particularly mobile joints, such as in the mid-cervical spine, and in the patello-femoral joint. [11]

Most research concerning joint laxity centers on Ehlers-Danlos syndrome (EDS). There are nine types of EDS; type III is clinically indistinguishable from BHS.[4] In EDS type III, it appears that the laxity may be due to a collagen processing defect or an abnormality in some other non-collagen connective tissue protein system. This collagen is a primary component of the joint capsule and ligaments, which are the joints' chief restraints.[6] In addition to the laxity allowed by poor collagen, the proprioceptors that are responsible for the development of appropriate muscle tension, are compromised. In normals, there is increased kinsaesthetic activity at the end range of motion. This activity was shown to be deficient in a study of ten sedentary subjects with HMS studied by Hall et al. Hall further studied twelve ballet dancers with HMS, and determined that their threshold for proprioception was actually better than the control group. Hall concluded that the training undergone by the ballet dancers actually improved their proprioception and their joint stability.[8]

Two other studies imply that activity can influence the painful symptoms of BHS. Larsson et al studied 660 musicians at the Eastman School of Music in Rochester. When compared to musicians with normal joint mobility, the musicians with hypermobility had a greater incidence of pain in joints that must maintain prolonged static postures while practicing or playing. For example, violinists with hypermobility had more painful

symptoms in their spine and knees. However, in joints involved in the music activity, the musicians with hypermobility were more comfortable than those musicians with normal joint mobility. For example, those with BHS had fewer symptoms of elbow, wrist, or finger pain. They made an interesting conclusion that musicians with normal mobility in the spine, but hypermobility of the elbow, wrist, and fingers are most likely to enjoy a long and comfortable career. [12,13]

Decoster et al studied 310 lacrosse players to determine whether BHS predisposed these athletes to a higher injury rate. They found that 24% of the players had BHS according to the Beighton scoring scale.[9] Over the course of the season, the overall injury rate was not significant with reference to those athletes with normal mobility. The difference was in the type of injury incurred. Hypermobile athletes suffered more ankle injuries, while their counterparts experience more muscle strains.[14]

Russek reported a physical therapy intervention that significantly reduced multiple joint pain due to BHS in her patient who was also a physical therapist. They employed joint protection strategies that included wearing supportive splints as needed, and she avoided bringing joints to the end of their range of motion. Some behaviors were modified to avoid stressing joints at end range. Since this patient was already involved in regular exercise, new exercises were not introduced. The patient felt that her pain decreased by 50%, and credited the improvement to her new understanding of the underlying problem.[15]

Case Study

The subject was a 22 year old white male in excellent general health and with a long history of joint problems. In high school, he subluxed both patellae and both

shoulders several times. At his initial visit to physical therapy, he complained of diffuse spinal pain. His chief complaint was poor tolerance of static postures, especially driving and sleeping. He slept only 2 to 3 hours at a time before being wakened by painful muscle guarding in the lumbar and thoracic regions, and in the hamstrings. His job as a wood stove sales representative required about 500 miles of driving per week, and painful muscle guarding accompanied this activity. He had tried unsuccessfully to manage his pain by maintaining good posture and by taking muscle relaxants and pain medications. He has found that he can avoid traumatic dislocations by avoiding physically stressful positions or activities. At that time, he avoided all athletic activities. He reported that he was clumsy, frequently bumping into things, resulting in contusions.

Exam:

Exam revealed hypermobility in knee hyperextension (24° recurvatum) and the ability to oppose both thumbs to the forearm. He reported that he can usually place both palms on the floor with knees straight, but was restricted now due to back pain. According to the Beighton scale, he scored 5 of 9 possible points, confirming a diagnosis of BHS. There was also intervertebral hypermobility (Grade 4, Paris scale)[16], but he denied radiculopathy. Manual muscle test revealed 3+/5 to 4/5 strength in the proximal musculature, and there was atrophy of both vastus medialis oblique muscles. Unilateral stance with vision occluded was limited to 3 seconds bilaterally. He required Actrin, lOO mg to help with sleep, about once a week.

Assessment:

Ligamentous laxity requires that the muscles maintain tension to provide the joint with

stability. 3+/5 to 4/5 strength is inadequate to provide this support comfortably. In addition, the deficit in ligamentous function contributes to poor proprioception and increased articular gliding, causing generalized clumsiness and poor balance. Problem list, therefore, included:

- Poor tolerance of static postures, limiting sleep to 2 or 3 hours per night and impairing his ability to drive more than one hour.
- Poor balance
- Clumsiness
- Limited sports participation in order to prevent subluxations
- Pain graded 5/10 on a McGill scale in the thoracic and lumbo-sacral regions, knees and shoulders

Goals:

1. Patient will sleep 6 to 8 hours without interruption by painful muscle guarding
2. Patient will drive 4 hours per day without painful muscle guarding.
3. Patient will no longer bump into things, eliminating contusions.
4. Patient will be able to participate in controlled athletic activities such as running, biking, and swimming.
5. Patient will increase distance of golf drive from 250 yards to 300 yards.

Treatment Plan:

The literature review suggested that stability could be improved by training the muscles and proprioceptors around the joint. Musicians, lacrosse players, and ballet dancers with BHS all were less symptomatic in joints that were exercised. The focus of treatment, then, was to develop stability around the joints through exercises designed to improve muscle strength and proprioception. Progressive resistive exercises (PREs) for the postural muscles were emphasized. Also, he applied an electrical muscle stimulator (EMS) unit to the paraspinal muscles. He attended physical therapy once a week for 5 weeks, until a home exercise program was well established. The progression of exercises is described in Table 3.

The first session was primarily devoted to evaluation and establishing goals. He

was issued an EMS unit for the purpose of re-educating the thoraco-lumbar paraspinal muscles. He was taught to apply it ten (10) seconds on, thirty (30) seconds off for 30 continuous minutes per day. He turned up the intensity until there was a visible contraction.

On the second visit, exercises were introduced to strengthen the slow twitch muscle fibers in the trunk, quadriceps, rhomboids, and anterior deltoid (See Table 3).

At the third visit, he reported good adherence to the home exercise program, but he had to perform them in the evening due to pain. On most nights, pain wakened him at 2:30 a.m., and he was unable to get back to sleep. He was most comfortable in his Lazy Boy chair. Exam revealed a long right thoracic C-curve, which I corrected in the clinic using a muscle energy technique. Exercises were taught to strengthen the gluteal muscles (bridging), trunk rotation (simulated baseball swing, using black theraband for resistance), and the quadratus lumborum (hip hiking).

At the fourth visit, the subject reported that he was interspersing the exercises throughout the day to make them more tolerable. He slept at least 4 hours a night this week. He swam and went white water rafting as a team building experience at work, and felt better after the activity. He slept six (6) hours both of those nights. New exercises were introduced: unilateral bridging with the contralateral leg maintaining a 30° straight leg raise (SLR) and we increased rhomboid resistance from green to blue theraband.

At the fifth visit, the subject reported having good days and bad days. He needed reassurance, and therefore I recommended that he recheck with his physician. I reassured him that it takes time to build enough strength to provide adequate stability, and encouraged him to continue with the exercises. We replaced the “dead bug’ exercise

with abdominal crunches, and increased the wearing time of the EMS unit on the thoracolumbar paraspinal muscles to 45 continuous minutes per day. He turned it up until there was a visible contraction, ten (10) seconds on and thirty (30) seconds off.

During the sixth week, telephone consult replaced an office visit. He reported a significant improvement in sleep this week, to 5 hours most nights, and 6 hours on two nights. He asked to increase theraband from blue to gray for rhomboids, and I agreed. He continued with the established exercise plan. Because of his improvement, he decided to not consult with his physician at this time.

The subject continued with the home exercise program. Four months after his first visit, he reported that he no longer needed medication to sleep. He found that applying EMS just before bedtime was most beneficial. He slept four (4) hours most nights, and napped for two (2) hours each day. His golf drive increased from 250 to 300 yards. He continued the home exercise program.

Six months from the first visit, the subject reported a tremendous improvement in symptoms. He was then sleeping six (6) to eight (8) hours per night. He was able to drive four (4) hours per day for his job with no difficulty. His job has been demanding, and his time for leisure activities has been limited.

Discussion:

During adolescence, this subject developed difficulty maintaining static postures. Also, his ability to participate in athletic activities was impaired by frequent shoulder and knee subluxations. He felt clumsy during everyday activities. He had seen several orthopedic surgeons over the years for immediate care of his patellar and shoulder subluxations, but the diagnosis of benign hypermobility syndrome had not been

recognized. Now, at age 22, he was having difficulty performing the driving responsibilities associated with his job and the recreational activities that he would like to pursue. Further, his poor sleep patterns influenced his ability to maintain alertness during the day. He presented to physical therapy for pain management strategies and to increase his ability to sleep and to participate in recreational activities. The home exercise program that was prescribed emphasized strengthening the slow twitch muscle fibers of the proximal muscles. This type of exercise was selected because they are considered to be the muscles used for stability. Also, the exercises had to be suitable for performance in a hotel room to accommodate the travel required by subject's work. After many years of being stretched beyond the normal physiological length, the activity of the golgi tendon organs were considered to be impaired. So, to complement the stability exercises, the EMS was applied to help re-educate the paraspinal muscles. Although the subject was very faithful to the home exercise program, and with using the EMS, his response to the intervention was very slow. It took six months for the goals to be achieved.

The results are encouraging. This case study demonstrated that diligent adherence to a home exercise program, with emphasis on slow twitch muscle fibers, combined with use of an EMS on proximal musculature significantly reduced the pain associated with BHS. Other improvements include uninterrupted sleep and the ability to perform the driving duties associated with his work. The improvement was very gradual, and good resolution of symptoms was not achieved until the subject exercised and applied the EMS for six (6) months. He complained that a good deal of "clicking" occurred in the low back when he did the abdominal bracing exercise ("dead bug"). He was sleeping poorly, unable to find a comfortable position. I felt that further research could address whether

including other types of exercise would have helped him reach his goals more quickly. Training the fast twitch muscle fibers through plyometrics, propriociptive activities, and high repetition, low weight PRE's may have been helpful.

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