

## Role of Osteopathic Manipulative Treatment in the Management of Stiff Person Syndrome

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**Stiff person syndrome (SPS) is a rare and disabling central nervous system disorder first described in 1956 and characterized by fluctuating rigidity and stiffness, gait impairment, and painful spasms of the axial and limb musculature. Although an underlying mechanism of impaired synaptic  $\gamma$ -aminobutyric acid–ergic inhibition has been proposed, the exact mechanism remains unclear. The glutamic acid decarboxylase antibody, a marker for SPS, is a strong indication of disease and has been reported in approximately 70% of patients. The current treatment of choice is benzodiazepines and baclofen, both of which reduce motor unit potential firing and, therefore, decrease stiffness and spasms. However, patients continue to have substantial disability with pharmacologic therapy alone. This case report demonstrates the potential of osteopathic manipulative treatment as an adjunct to medication in the management of SPS. By decreasing somatic dysfunction and reducing the frequency of exacerbations, osteopathic manipulative treatment may alleviate the symptoms and overall morbidity associated with this disease.**

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**S**tiff person syndrome (SPS) was first described by Frederick Moersch, MD, and Henry Woltman, MD, PhD, in 1956 as progressive muscle rigidity and stiffness, primarily of the axial skeleton, that often manifests in patients aged approximately 35 to 39 years.<sup>1</sup> The disease course is characterized by intermittent and increasingly severe muscle spasms of the back and proximal extremities, resulting in postural deformities.<sup>1-4</sup> This rare condition has a prevalence of roughly 1 to 9 per million persons.<sup>5</sup> Although the ultimate cause of SPS has yet to be identified, several features suggest an autoimmune disease affecting the central nervous system (CNS) and spinal cord.<sup>1,6,7</sup> Occurring twice as frequently in women as men, SPS is often associated with other autoimmune conditions, including thyroiditis, vitiligo, and pernicious anemia.<sup>8</sup> Reports have cited the presence of glutamic acid decarboxylase (GAD) antibodies in up to 70% of patients with SPS.<sup>6,7,9</sup> A continuous motor unit potential firing pattern has been observed, even in patients at rest, along with a reduced level of  $\gamma$ -aminobutyric acid (GABA)–ergic tone, which results in the characteristic muscle stiffness. A history of traumatic injury has also been implicated as a mechanism behind SPS, although convincing data have yet to be reported.<sup>10,11</sup>

Various treatment regimens have been implemented for pain management, such as corticosteroids, intravenous immunoglobulin, and plasmapheresis. Benzodiazepines and

baclofen, in particular, have shown to be most beneficial in providing long-term relief by increasing GABA activity and reducing motor unit potential firing.<sup>2,10,12-14</sup> Although benzodiazepines and baclofen have shown symptom improvement, no controlled clinical trials have been conducted to establish definitive, evidence-based criteria for their use in patients with SPS.<sup>14</sup>

In the following case, we report the use of osteopathic manipulative treatment (OMT) in conjunction with pharmacologic therapy in a symptomatic patient with SPS.

## Report of Case

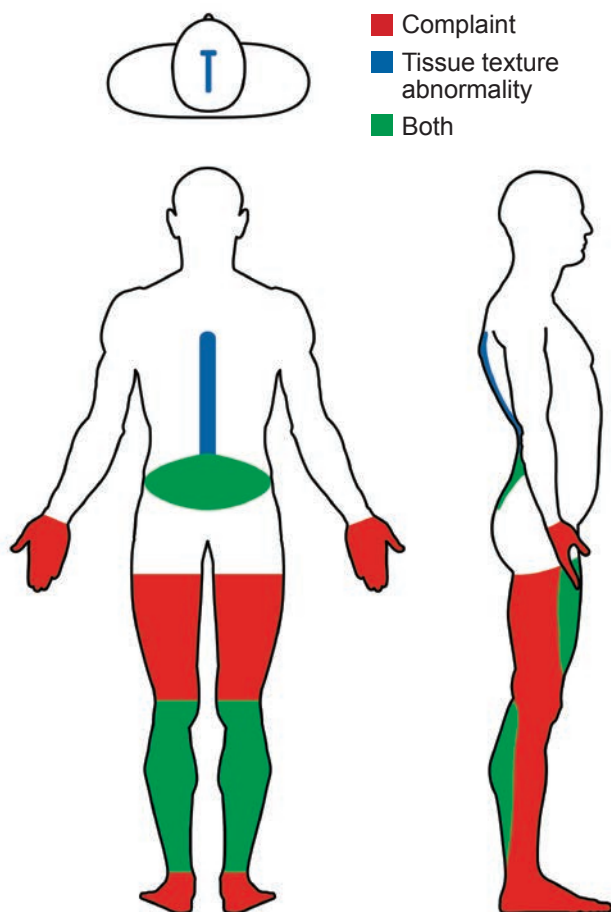
A 52-year-old man presented to the clinic with a 10-year history of diagnosed SPS. He reported current daily symptoms of pain localized particularly to his lower back, hands, and lower extremities. Aggravating factors included sitting for extended periods, excessive movement, and cold temperatures. His symptoms had been partially alleviated by benzodiazepines and opioids.

His SPS symptoms began after he sustained injuries in a motor vehicle accident 23 years earlier, in which he had been expelled from the vehicle through the windshield and had amnesia for 3 days. A second motor vehicle accident 11 years later resulted in worsening of his symptoms. Two years after the second accident, he was evaluated by a neurologist. Electromyogram and serum antibody study results led to the diagnosis of SPS. Other comorbidities included a chronic primary hypercoagulable state and erectile dysfunction, both of which were controlled with medication and stable at the time of the current visit. His family history was significant for an unspecified autoimmune disorder in his mother and sister, records for whom were unobtainable.

At the time of presentation, the patient expressed interest in establishing care for medication management alone. He was reluctant to try any additional therapies, having had poor responses to “overzealous” physical therapy. His initial appointments were scheduled 1 month apart per clinic policy regarding new narcotic prescriptions.

At the third follow-up visit, the patient expressed a willingness to undergo OMT as an adjunct to medication for his pain management, although he was skeptical about its effectiveness. A physical examination was conducted in standing, seated, and prone positions, with the following palpatory findings noted: restricted motion and tissue texture changes of the sagittal suture; chronic tissue texture abnormalities on the left at spinal levels T6-T11; decreased right rotation within the gross range of motion in the thoracic spine; bilateral thoracic and lumbar paraspinal muscle tightness and spasm; chronic tissue texture abnormalities bilaterally at spinal levels L1-L5; decreased left rotation within the gross range of motion in the lumbar spine; and excessive muscle tension of the right and left soleus and gastrocnemius, hip abductors, and vastus muscles. A summary of these findings is illustrated in the *Figure*. Of note, the patient was unable to comfortably lie in the supine position because of low back pain exacerbation. Flexion at the hips and knees did not produce relief.

On the basis of the patient’s medical history and physical examination results, an OMT regimen was applied to manage the palpatory findings. Owing to the cerebral trauma history, treatment was initiated at the head. Ligamentous articular strain, a technique in which the goal of treatment is to balance the tension in opposing ligamentous structures where abnormal tension is present, and neurofascial release, a light-pressure palpation procedure using several inherent body forces to normalize structural and functional relationships in the body, were used to manage the sagittal suture restrictions and overlying fascial restrictions.<sup>15,16</sup> Release of these fascial restrictions was noted to have a positive relaxing effect on the resting tone of the lower extremities. Further osteopathic cranial manipulative medicine (OCMM) techniques, including V-spread, balanced membranous tension, venous sinus drainage, and disengagement of the sagittal suture, were used to manage the remaining cranial dysfunction, which was



**Figure.** Areas of the body of a 52-year-old man with stiff person syndrome shaded to reflect pain reported by the patient and tissue texture abnormalities documented by his osteopathic physician.

again noted to have a positive relaxing effect on the thoracic, lumbar, and lower extremity dysfunctions.<sup>15</sup> The patient was observed to have a more erect gait at the conclusion of treatment.

The patient agreed to continue with OMT, but he was unwilling to schedule any appointments exclusively for treatments because of his skepticism about OMT. Consequently, the next OMT session did not occur until his 4-month follow-up. At that appointment, the patient reported marked improvement for approximately 3 days after the previous OMT session before symptoms returned to baseline. Physical examination revealed an externally rotated right parietal bone with right coronal suture restriction and restricted motion of the sagittal suture; bilateral piriformis spasms; restriction of the upper and middle poles of the sacroiliac joint on the right; neutral T9-T11 spinal levels, sidebent right and rotated left; neutral L2-L5 spinal levels, sidebent left and rotated right, with bilateral chronic tissue texture abnormalities, including bilateral paraspinal and quadratus lumborum muscle tension; and excessive muscle tension of the bilateral hamstring, soleus, gastrocnemius, and vastus muscles.

In addition to performing OMT on the patient's head in a similar fashion to the previous visit, neuro-fascial release was initiated at several sites of muscle tension. Techniques performed on the head and lower body had a similar effect in decreasing resting muscle tone. Expanding OMT to the sacrum and pelvis further produced a decrease in muscle tension, such that the patient was more comfortable on the treatment table; however, the patient remained unable to tolerate lying in a supine position.

At his third clinic appointment 2 months later, the patient and his wife reported that his symptoms had remained improved for several weeks after receiving OMT at the previous clinic appointment. Additionally, the patient noted that he was able to reduce his usual narcotic dosage by half. Because of his improved symptoms and reduced need for narcotics, the patient requested an in-

crease in OMT frequency. Three subsequent treatments were scheduled at 2-week intervals.

The findings at the beginning of each of these visits continued to be predominantly cranial suture restrictions with gradual visit-to-visit improvement; other findings were similar to the above-mentioned patterns of muscular tension throughout the body, likewise with visit-to-visit improvement. The patient not only tolerated the supine position by the end of the first of these visits, but in the second and third visits he was also able to stay in this position comfortably for the duration of his hour-long appointments. At the most recent visit, the patient reported experiencing a flare in his symptoms due to cold weather, but he was able to lie supine despite his subjective increase in pain. The patient has since been lost to follow-up.

## Discussion

The proposed mechanism behind SPS relates to involuntary firing of motor neurons thought to be a result of an autoimmune response to GAD, which in turn results in a lack of the inhibitory neurotransmitter GABA. The lack of inhibition by GABA creates an excitatory state that seems to be relieved by benzodiazepines. Further, 1 of the 2 isoforms of GAD is only detectable in the CNS.<sup>13</sup> For patients with SPS in whom benzodiazepines are inadequate, other avenues must be explored to better achieve symptomatic relief from this debilitating musculoskeletal disease. Although benzodiazepines and baclofen reduce motor unit potential firing to decrease stiffness and spasms in these patients, the efficacy of OMT as an adjunctive treatment option in this patient population has not been studied, to our knowledge.

In the current case, the OCMM techniques seemed to alleviate the somatic dysfunctions, which approximates normal inhibition at the neural level. Objective improvement was documented by the relaxation of muscle tone in widespread body locations, the new

ability to lie supine comfortably, and the decrease in total narcotic dose. Although OMT may have improved the symptoms associated with his traumatic brain injuries, the effect on the resting muscle tone and the overall decrease in muscle tension observed after treatment further indicates that the underlying pathophysiology of SPS was affected. Continued OMT would be needed to assess the ultimate response in symptoms and long-term quality of life.

The natural follow-up with this patient would have been to compare his GAD antibody levels before and after OMT. Before-and-after electromyography assessment could also have determined the effectiveness of OCMM. Electromyogram recordings of muscle spasms in patients with SPS resemble normal muscle contractions, except that the motor unit potentials are active at rest and not inhibited by the contraction of antagonist muscle groups.<sup>13</sup>

The current case demonstrates the potential benefit of adjunctive OMT in SPS management. Although the patient showed improvement in the palpatory findings in all regions identified, he continued to have painful exacerbations triggered by cold intolerance.

## Conclusion

The current case report demonstrates the improvement in palpatory findings and associated somatic dysfunctions achieved through the use of OMT and OCMM in a patient with SPS. This observed improvement in resting muscle tone and tension supports our belief that OMT affects the underlying pathophysiology of SPS. Further studies, however, are needed to confirm this hypothesis in a larger patient population and potentially in patients with other autoimmune musculoskeletal diseases. We propose OMT as an adjunctive therapy to provide symptom improvement in persons with SPS and other autoimmune movement disorders who do not obtain adequate relief with medication management alone.

## References

1. Moersch FP, Woltman HW. Progressive fluctuating muscular rigidity and spasm ("stiff-man" syndrome); report of a case and some observations in 13 other cases. *Proc Staff Meet Mayo Clin.* 1956;31(15):421-427.
2. Hadavi S, Noyce AJ, Leslie RD, Giovannoni G. Stiff person syndrome. *Pract Neurol.* 2011;11(5):272-282. doi:10.1136/practneurol-2011-000071.
3. Helfgott SM. Stiff-man syndrome: from the bedside to the bench. *Arthritis Rheum.* 1999;42(7):1312-1320.
4. Kuhn WF, Light PJ, Kuhn SC. Stiff-man syndrome: case report. *Acad Emerg Med.* 1995;2(8):735-738.
5. Holmøy T, Geis C. The immunological basis for treatment of stiff person syndrome. *J Neuroimmunol.* 2011;231(1-2):55-60. doi:10.1016/j.jneuroim.2010.09.014.
6. Levy LM, Dalakas MC, Floeter MK. The stiff-person syndrome: an autoimmune disorder affecting neurotransmission of  $\gamma$ -aminobutyric acid [review]. *Ann Intern Med.* 1999;131(7):522-530.
7. Solimena M, Folli F, Denis-Donini S, et al. Autoantibodies to glutamic acid decarboxylase in a patient with stiff-man syndrome, epilepsy, and type I diabetes mellitus. *N Engl J Med.* 1988;318(16):1012-1020.
8. NINDS stiff-person syndrome information page. National Institute of Neurological Disorders and Stroke website. <http://www.ninds.nih.gov/disorders/stiffperson/stiffperson.htm>. Accessed April 21, 2015.
9. Raju R, Rakocevic G, Chen Z, et al. Autoimmunity to GABA<sub>A</sub>-receptor-associated protein in stiff-person syndrome. *Brain.* 2006;129(pt 12):3270-3276.
10. Brashear HR, Phillips LH. Autoantibodies to GABAergic neurons and response to plasmapheresis in stiff-man syndrome. *Neurology.* 1991;41(10):1588-1588.
11. Hall CD, Haworth CC. "Stiff man syndrome" and trauma. *Br Med J.* 1971;3(5773):531.
12. Murinson BB. Stiff-person syndrome [review]. *Neurologist.* 2004;10(3):131-137.
13. Vasconcelos OM, Dalakas MC. Stiff-person syndrome. *Curr Treat Options Neurol.* 2003;5(1):79-90.
14. Ciccoto G, Blaya M, Kelley RE. Stiff person syndrome. *Neurol Clin.* 2013;31(1):319-328. doi:10.1016/j.ncl.2012.09.005.
15. Educational Council on Osteopathic Principles. *Glossary of Osteopathic Terminology.* Chevy Chase, MD: American Association of Colleges of Osteopathic Medicine; 2011.
16. Davidson SM. Neurofascial release. Healthabounds2 website. <http://healthabounds2.com/scholarly-paper-neurofascial-release/>. Accessed April 21, 2015.

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