




Volume 1, Issue 4B, September 2006

At the National Rehabilitation Information Center, Information Specialists field requests on a wide range of disability rehabilitation issues. While a dissemination center for NIDRR funded research, NARIC serves persons with disability and their families by providing information and referral on a variety of rehabilitation and disability topics. This two part edition of *reSearch* focuses on two specific topics affecting our consumers: Locked-In and Stiff-Person Syndromes.

According to the National Institute of Neurological Disorders and Stroke Information Page,









Stiff-Person Syndrome (SPS) is a rare neurological disorder with features of an autoimmune disease. SPS is characterized by fluctuating muscle rigidity in the trunk and limbs and a heightened sensitivity to stimuli such as noise, touch, and emotional distress, which can set off muscle spasms. Abnormal postures, often hunched over and stiffened, are characteristic of the disorder. (<http://www.ninds.nih.gov/disorders/stiffperson/stiffperson.htm>)

Our search resulted in a range documents from the most current through the past five years of SPS research. Searches in NARIC REHABDATA and National Library of Medicine database, PubMed resulted in over 40 search terms. A sample of these terms is listed below:

-  Antibodies
-  Anticonvulsants
-  Anti-Inflammatory Agents
-  Atrophy
-  Autoimmune Disorders
-  Cerebellar Diseases
-  Chronic Disease
-  Cognition Disorders
-  Depressive Disorders
-  Drug Therapy (combination)
-  Electric Stimulation Methods
-  Muscle Relaxants (therapeutic use)
-  Muscle Rigidity/Spasticity/Spasm
-  Nerve Tissue Proteins
-  Neural Inhibition

reSearch: *Stiff-Person Syndrome*

A collection of research reviews on rehabilitation topics from NARIC and other information resources.

-  Neuromuscular Agents (therapeutic use)
-  Neuromuscular Disorders
-  Physical Therapy
-  Pregnancy
-  Quality of Life
-  Research Methodology
-  Rehabilitation
-  Stiff-Person Syndrome

There is a limited amount of information available on SPS. Our search of REHABDATA resulted in one document ranging from 2001. The PubMed database search resulted in 23 documents ranging from 2006-2001. The complete citations are included at the end of this research brief.

In addition to document searches, NARIC searched its Program database of National Institute on Disability and Rehabilitation Research (NIDRR) projects to locate grantees/projects related Stiff-Person Syndrome. While there are no specific grantees/projects on SPS there are several NIDRR and non-NIDRR funded projects and organizations that address the symptoms and disabilities presented by this syndrome which may be useful to our patrons:

Practical Clinical Trial of Cognitive Rehabilitation in Neurologic Illness
Project Number: H133G050063
(Active)

Additional Organizations & Institutes:

National Organization for Rare Disorders (NORD)
Toll Free: 800/999-6673
Email: orphan@rarediseases.org
www.rarediseases.org

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)
Toll Free: 877/226-4267
Email: NIAMinfo@mail.nih.gov
www.niams.nih.gov

Support Group on Stiff-Person Syndrome:
www.stiffman.org

Document from NARIC's REHABDATA search listed are listed below:

2001

Dalakas, M.C., Fujii, M., Li, M., Lutfi, B., Kyhos, J., & McElroy, B. (2001). **High-dose intravenous immune globulin for stiff-person syndrome.** *New England Journal of Medicine*, 345(26), 1870-1876.

NARIC Accession Number: J43854

ABSTRACT: Study evaluated the efficacy of intravenous immune globulin for people with stiff-person syndrome. Sixteen participants were assigned randomly to receive either intravenous immune globulin or placebo for three months, followed by a one-month washout period, then three months of therapy with the alternative agent. Efficacy was judged by improvement in scores on the distribution-of-stiffness index and heightened-sensitivity scale from baseline to the second and third month of each treatment phase. Among patients who received immune globulin first, stiffness scores decreased substantially and heightened-sensitivity scores decreased significantly during immune globulin therapy but returned to baseline during placebo administration. In the group that received the placebo first, the scores remained constant during placebo administration but dropped significantly during immune globulin administration. Eleven patients who received immune globulin were able to walk more easily, their frequency of falls decreased, and they were able to perform work-related or household tasks. The beneficial effects of immune globulin lasted from six weeks to one year.

Documents from the National Library of Medicine PubMed search at <http://www.pubmed.com/> are listed below:

2006

Elkassabany, N., Tetzlaff, J.E., & Argalious, M. (2006). **Anesthetic management of a patient with stiff person syndrome.** *Journal of Clinical Anesthesia*, 18(3), 218-20.

PMID: 16731326

ABSTRACT: We describe the successful anesthetic management of a patient with SPS undergoing a right inguinal hernia repair, using a somatic paravertebral block supplemented with conscious sedation. We also

present the implications of general anesthesia in patients with SPS. The use of regional anesthetic techniques in patients with SPS has the advantage of avoiding exposure to muscle relaxants. The use of general anesthesia in patients with SPS carries the risk of postoperative hypotonia due to enhancement of gamma-aminobutyric acid action on synaptic transmission by drugs that have a gamma-aminobutyric acid agonistic action.

Potter, K. (2006). **Physical therapy during in-patient rehabilitation for a patient with Stiff-Person Syndrome.** *Journal of Neurologic Physical Therapy*, 30(1), 28-38.

PMID: 16630369

ABSTRACT: BACKGROUND AND PURPOSE: Patients with SPS typically show stiffness and spasms, primarily of the trunk and proximal lower extremities. The purpose of this case report is to provide an overview of SPS and a description of the specific physical therapy management strategies used during a brief inpatient rehabilitation stay for a patient with SPS, illustrating the use of the patient/client management model in the Guide to Physical Therapist Practice. **CASE DESCRIPTION:** The patient was a 33-year-old with a three-year history of SPS. He spent 10 days in an inpatient rehabilitation hospital where he received physical therapy daily. The initial examination revealed impairments of pain, range of motion, reflex integrity, and motor function, along with abnormalities of posture, balance, and function. The procedural interventions included therapeutic exercise and functional retraining. Stretching exercises were categorized according to their priority and level of difficulty to accommodate for the patient's varying symptoms, and relaxation exercises aimed to reduce the severity of the patient's spasms. The functional retraining program included transfer and progressive gait training. **OUTCOMES:** The patient showed improvements in ankle range of motion, posture, and gait (distance, speed, and independence), despite continued problems with stiffness, spasms, and pain. **DISCUSSION:** Physical therapists working with patients with SPS have challenges related to the paucity of information in the literature. The chronic, progressive, and variable nature of SPS indicates the need for life-long management, with the inclusion of an exercise program that can be adjusted accordingly, given the frequently changing symptoms experienced by the patient. As is shown with this case, it appears

that physical therapy can improve function and some of the impairments associated with SPS.

2005

Ameli, R., Snow, J., Rakocevic, G., & Dalakas, M.C. (2005). **A neuropsychological assessment of phobias in patients with stiff person syndrome.** *Neurology*, 64(11), 1961-3.

PMID: 15955955

ABSTRACT: A neuropsychological assessment was performed in ten patients with SPS to determine whether their anxiety and phobic symptoms precede stiffness and spasms or represent a reaction to disability. No neurocognitive dysfunction was noted. Patients perceived fears and anxiety as realistic and caused by SPS rather than due to an inherent phobic neurosis.

Baker, M.R., Das, M., Isaacs, J., Fawcett, P.R., & Bates, D. (2005). **Treatment of stiff person syndrome with rituximab.** *Journal of Neurology, Neurosurgery, and Psychiatry*, 76(7), 999-1001.

PMID: 15965211

ABSTRACT: This case report is about the novel use of the anti-CD20 antibody, rituximab, in the treatment of a 41-year-old woman with stiff person syndrome. She was admitted to hospital as an emergency with prolonged and painful extensor spasms affecting the neck and back, arms, and legs. The disease had progressed despite a favourable initial response to conventional treatment with intravenous immunoglobulin and cytotoxics. Treatment with rituximab induced a lasting clinical remission.

Burns, T.M. (2005). **A step forward for stiff-person syndrome.** *Lancet*, 365(9468), 1365-7.

PMID: 15836869

ABSTRACT: No abstract is available.

De la Torre-Laviana, F.J., Redondo-Verge, L., Izquierdo, G., Mir-Mercader, J., Cid-Boza, M.C., Dinca-Avarvarei, L., & Marcos-Gonzalez, A. (2005). **Stiff man syndrome: a specific immunological condition affecting the central nervous system.** *Revista de neurologia*, 40(3), 190-2. (Article is in Spanish).

PMID: 15750907

ABSTRACT: No abstract is available.

Economides, J.R. & Horton, J.C. (2005). **Eye movement abnormalities in stiff person syndrome.** *Neurology*, 65(9), 1462-4.

PMID: 16275836

ABSTRACT: The authors describe a 38-year-old woman with SPS and gaze-holding nystagmus, limited abduction, vertical and horizontal ocular misalignment, deficient smooth pursuit, and impaired saccade initiation. There was no evidence of ocular myasthenia, indicating that abnormalities of ocular motor function can occur as a primary manifestation of SPS, perhaps from depletion of GABA.

2004

Khasani, S., Becker, K., & Meinck, H.M. (2004). **Hyperekplexia and stiff-man syndrome: abnormal brainstem reflexes suggest a physiological relationship.** *Journal of Neurology, Neurosurgery, and Psychiatry*, 75(9), 1265-9.

PMID: 15314112

ABSTRACT: BACKGROUND AND

OBJECTIVES: Hyperekplexia and the stiff-man syndrome (SMS) are both conditions with exaggerated startle suggesting abnormal brainstem function. Investigation of brainstem reflexes may provide insight into disturbed reflex excitation and inhibition underlying these movement disorders. PATIENTS AND METHODS: Using four-channel EMG, we examined four trigeminal brainstem reflexes (monosynaptic masseter, masseter inhibitory, glabella, and orbicularis oculi blink reflexes) and their spread into pericranial muscles in five patients with familial hyperekplexia (FH), two with acquired hyperekplexia (AH), 10 with SMS, and 15 healthy control subjects. RESULTS: Both FH/AH and SMS patients had abnormal propagation of brainstem reflexes into pericranial muscles. All patients with hyperekplexia showed an abnormal short-latency (15-20 ms) reflex in the trapezius muscle with a characteristic clinical appearance ("head retraction jerk") evoked by tactile or electrical stimulation of the trigeminal nerve, but normal monosynaptic masseter reflexes. Inhibitory brainstem reflexes were attenuated in some FH/AH patients. Four of 10 patients with SMS had similar short-latency reflexes in the neck muscles and frequently showed widespread enhancement of other excitatory reflexes, reflex spasms, and attenuation of inhibitory brainstem reflexes. CONCLUSION: Reflex excitation is exaggerated and inhibition is

attenuated in both stiff-man syndrome and familial or acquired hyperekplexia, indicating a physiological relationship. Reflex transmission in the brainstem appears biased towards excitation which may imply dysfunction of inhibitory glycinergic or GABAergic interneurons, or both.

Goeb, V., Dubreuil, F., Cabre, P., Jean-Baptiste, G., & Arfi, S. (2004). **Lupus revealing itself after a stiff-person syndrome.** *Lupus*, 13(3), 215.

PMID: 15119554

ABSTRACT: No abstract is available.

Hassin-Baer, S., Kirson, E.D., Shulman, L., Buchman, A.S., Bin, H., Hindiyeh, M., Markevich, L., & Mendelson, E. (2004). **Stiff-person syndrome following West Nile fever.** *Archives of Neurology*, 61(6), 938-41.

PMID: 15210535

ABSTRACT: BACKGROUND: SPS is a rare autoimmune disorder associated with antibodies against glutamic acid decarboxylase (GAD), the key enzyme in gamma-aminobutyric acid synthesis. In most cases, a trigger cannot be identified. OBJECTIVE: To describe a 41-year-old man who developed stiff-person syndrome and antibodies to GAD following acute West Nile virus infection. DESIGN: A case report and a search in GenBank for common epitopes. RESULT: The search revealed a stretch of 12 amino acids in the NS1 protein of West Nile virus with a high degree of homology to the GAD65 region (an isoform of GAD) containing the PEVKEK motif. CONCLUSION: Cross-reactivity between antibodies directed against West Nile virus and GAD may have contributed to the development of stiff-person syndrome in this patient.

Murinson, B.B. (2004). **Stiff-person syndrome.** *The Neurologist*, 10(3), 131-7.

PMID: 15140273

ABSTRACT: BACKGROUND: SPS is a rare neurologic disorder with autoimmune features. It is characterized by progressive, severe muscle rigidity or stiffness most prominently affecting the spine and lower extremities. REVIEW SUMMARY: Superimposed muscle spasms result in simultaneous contraction of agonist and antagonist muscles which are detectable by electromyography (EMG) and relieved by administration of benzodiazepines. The exacerbation of SPS by emotional stressors often results in the

referral of these patients for psychiatric assessment although this was more common before the discovery of an association with antibodies to glutamic acid decarboxylase (GAD antibodies). Formerly known as stiff-man syndrome, the female to male ratio is two to one and the principle paraneoplastic variant is associated with breast cancer. Although rare, this is a disease of middle age that severely curtails the functional capacity of those it strikes. It is frequently associated with diabetes and other autoimmune diseases. IVIg is recently demonstrated to be effective in the treatment of SPS; diazepam remains useful in managing the symptoms. CONCLUSIONS: This article summarizes the history of SPS, describes important clinical features, discusses management, touches upon areas of uncertainty, and postulates some avenues for research.

Weatherby, S.J., Woolner, P., & Clarkem, C.E. (2004). **Pregnancy in stiff-limb syndrome.** *Movement Disorders*, 19(7), 852-4.

PMID: 15254953

ABSTRACT: To our knowledge, pregnancy in a patient with stiff-limb-syndrome (SLS) has not been reported. We present the case of a woman with SLS who improved during pregnancy, delivered a normal healthy baby by forceps-assisted vaginal delivery, and suffered a mild postpartum "relapse."

2003

Bartsch, T., Herzog, J., Baron, R., & Deuschl, G. (2003). **The stiff limb syndrome—a new case and a literature review.** *Journal of Neurology*, 250(4), 488-90.

PMID: 12760383

ABSTRACT: No abstract is available.

Lohmann, T., Londei, M., Hawa, M., & Leslie, R.D. (2003). **Humoral and cellular autoimmune responses in stiff person syndrome.** *Annals of the New York Academy of Sciences*, 998, 215-22.

PMID: 14592879

ABSTRACT: SPS is a chronic autoimmune disease associated with humoral and cellular immune responses to glutamic acid decarboxylase (GAD) 65. Another chronic autoimmune disease, type 1 diabetes (T1D), is also associated with autoimmune responses to this antigen, but T1D patients develop SPS only extremely rarely and only a third of SPS patients develop T1D

(mostly mild manifestations in adulthood). In a previous study, we described important differences between T1D and SPS in the autoimmune response to GAD 65: (1) T cells of SPS patients recognize epitopes in the middle of GAD 65 (amino residues 81-171 and 313-403), whereas patients with T1D preferentially recognize another middle (161-243) and a C-terminal region (473-555); and (2) GAD antibodies (Abs) were nearly exclusively of the Th1-associated IgG1 type in T1D, whereas SPS patients had both Th1- and Th2-associated IgG4 and IgE GAD Abs. These differences were not simply related to different HLA alleles. Fine epitope mapping revealed further distinct T cell epitopes in both diseases despite similar HLA background. Therefore, a single autoantigen can elicit different immune responses causing distinct chronic autoimmune diseases possibly related to a Th1 or Th2 bias of the disease.

Piovano, C., Piattelli, M., Spina, T., Iervese, G., & Bosco, G. (2003). **The stiff-person syndrome. Case report.** *Minerva anesthesiologica*, 68(11), 861-5. PMID: 12538969

ABSTRACT: The stiff-person syndrome is a rare and disabling disorder, characterized by muscle rigidity with superimposed painful spasms involving axial and limb musculature. The clinical symptoms are continuous contraction of agonist and antagonist muscles caused by involuntary motor-unit firing at rest and the spasms that are precipitated by tactile stimuli, passive stretch, volitional movement of affected or unaffected muscles, startling noises and emotional stimuli. Both the rigidity and the spasms are relieved by sleep, general anaesthesia, myoneural blockade and peripheral nerve blockade. The cause of the stiff-person syndrome is unknown but an autoimmune pathogenesis is suspected because (1) the presence in the cerebrospinal fluid (CSF) of antibodies against glutamic acid decarboxylase (GAD), the rate-limiting enzyme for the synthesis of the inhibitory neurotransmitter gamma-aminobutyric acid (GABA), (2) the association of the disease with other autoimmune disorders, (3) the presence of various autoantibodies and (4) a strong immunogenetic association. The stiff-person syndrome is clinically elusive but potentially treatable and should be considered in patients with unexplained stiffness and spasms. Drugs that enhance GABA neurotransmission, such as diazepam, vigabatrin and baclofen, provide modest relief of clinical symptoms. Immunomodulatory agents such as steroids, plasmapheresis and intravenous

immunoglobulin, seem to offer substantial improvement.

Szczepanska-Szerej, A., Kulka, M., Wojczal, J., & Stelmasiak, Z. **Botulinum toxin A in the treatment of stiff man syndrome.** *Neurologia i neurochirurgia polska*, 37 Suppl 5, 175-81. (Article is in Polish). PMID: 15098344

ABSTRACT: SPS is a very rare disorder characterised by progressive fluctuating muscle rigidity and episodic spasm. So far, only two reports have demonstrated a significant clinical improvement in the patients with SPS when muscles were injected with Botulinum Toxin A (BTA). We investigated the effectiveness of intramuscular injections of BTA in a patient with clinical, biochemical and electrophysiological evidence of SPS. A 41-year-old woman with coexisting epilepsy and insulin-dependent diabetes mellitus was hospitalised in our Department because of stiffness and paroxysmal spasm of trunk and proximal limbs muscles. Because of not sufficient results of the pharmacological treatment the injections of BTA into involved muscles were done. Clinical observations included measure of pain, frequency of spasm, well-being and selection's activities were performed at baseline and in one, two, seven, 11, 16, 20, weeks. Significant improvement started one week after injections and lasting about 4 months was observed. Using BTA injections into involved muscles for the treatment of SPS can be followed by marked functional improvement and reducing the need for systemic drugs.

Vasconcelos, O.M., & Dalakas, M.C. (2003). **Stiff-person Syndrome.** *Current Treatment Options in Neurology*, 5(1), 79-90. PMID: 12521565

ABSTRACT: SPS is a progressive neurologic disorder characterized by (1) stiffness that is prominent in axial muscles, with co-contraction of agonist and antagonist muscles; (2) sudden episodic spasms; and (3) absence of another disease that causes similar symptoms. The diagnosis of SPS is based on clinical grounds and requires a high degree of suspicion. The diagnosis is, however, aided by electromyography, which demonstrates motor unit firing at rest simultaneously from the agonist and antagonist muscles, and by high serum titers of antibodies against glutamic acid decarboxylase (GAD), the rate-limiting enzyme for the

synthesis of gamma-aminobutyric acid (GABA), which is the brain's main inhibitory neurotransmitter. The reduced GABA level in the brain and cerebrospinal fluid explains the patients' stiffness and justifies the clinical improvement observed by drugs enhancing GABAergic transmission. The association of SPS with other autoimmune disorders or autoantibodies, the intrathecal GAD-specific immunoglobulin G antibody synthesis, and the suppression of GABA by the patient's antibodies supports the autoimmune nature of SPS and justifies the use of immunotherapies. At present, GABA-enhancing agents, such as benzodiazepines, valproate, vigabatrin, tiagabine, gabapentin, and baclofen, provide symptomatic relief. Plasmapheresis, steroids, and periodic intravenous immunoglobulin infusions provide additional and lasting benefit. In this article, the treatment options for patients with SPS are discussed based on the authors' experience and that of others. The beneficial effects from the first controlled study conducted in SPS using intravenous immunoglobulin are presented.

Yeow, T.P. & Tomkin, G.H. (2003). **A delayed diagnosis of stiff person syndrome.** *Hospital Medicine*, 65(3), 178-9.

PMID: 15052911

ABSTRACT: No abstract available.

2002

Gerschlager, W., & Brown, P. (2002). **Effect of treatment with intravenous immunoglobulin on quality of life in patients with stiff-person syndrome.** *Movement Disorders*, 17(3), 590-3.

PMID: 12112212

ABSTRACT: The therapeutic effects of intravenous immunoglobulin (IVIG) on the SPS have been described exclusively in case reports or open-label studies in terms of clinical outcomes. We investigate whether IVIG improves quality of life (QoL) in the SPS. Six patients with the classic form of SPS completed a generic QoL instrument, the SF-36, and a Visual Analogue Scale (VAS) before treatment as well as two weeks after completion of a course of IVIG. There was significant improvement in the SF-36 subscores for pain, social functioning, general mental health, and energy-vitality with treatment. The VAS also improved

significantly. We conclude that treatment with IVIG improves QoL in the SPS. Copyright 2002 Movement Disorder Society.

Gerschlager, W., Schrag, A., & Brown, P. (2002). **Quality of life in stiff-person syndrome.** *Movement Disorders*, 17(5), 1064-7.

PMID: 12360560

ABSTRACT: SPS is a rare, chronic disorder characterized by painful spasm and stiffness. We investigated the quality of life (QoL) in SPS patients, and identified factors associated with impairment in patients' QoL. Twenty-four SPS patients (10 men, 14 women; mean age +/- S.D., 52.6 +/- 9.5 years) completed the medical outcomes study Short Form health survey (SF-36), the Beck Depression Inventory (BDI), and a questionnaire asking for sociodemographic and clinical details. Extent of the disease was assessed using a distribution of stiffness score. SPS patients showed markedly reduced mean scores for all dimensions of the SF-36 when compared to norms from the general population of the United Kingdom. QoL scores showed a strong correlation with the extent of the disease. Depression was a common finding; 14 of 24 patients had depressive symptoms as evidenced by the BDI. There was a significant and strong correlation between the BDI score and several SF-36 subscores. This is the first study to address QoL in patients with SPS. We have shown that SPS has a significant impact on patients' reported QoL. The association between depression and QoL highlights the importance of recognizing and treating depression in SPS. Copyright 2002 Movement Disorder Society

Meinck, H.M. & Thompson, P.D. (2002). **Stiff man syndrome and related conditions.** *Movement Disorders*, 17(5), 853-66.

PMID: 12360534

ABSTRACT: The stiff man syndrome (SMS) and its variants, focal SMS, stiff limb (or leg) syndrome (SLS), jerking SMS, and progressive encephalomyelitis with rigidity and myoclonus (PERM), appear to occur more frequently than hitherto thought. A characteristic ensemble of symptoms and signs allows a tentative clinical diagnosis. Supportive ancillary findings include (1) the demonstration of continuous muscle activity in trunk and proximal limb muscles despite attempted relaxation, (2) enhanced exteroceptive reflexes, and (3) antibodies to glutamic acid decarboxylase (GAD) in

both serum and spinal fluid. Antibodies to GAD are not diagnostic or specific for SMS and the role of these autoantibodies in the pathogenesis of SMS/SLS/PERM is the subject of debate and difficult to reconcile on the basis of our present knowledge. Nevertheless, evidence is emerging to suggest that SMS/SLS/PERM are manifestations of an immune-mediated chronic encephalomyelitis and immunomodulation is an effective therapeutic approach. Copyright 2002 Movement Disorder Society

Molloy, F.M., Dalakas, M.C., & Floeter, M.K. (2002). **Increased brainstem excitability in stiff-person syndrome.** *Neurology*, 59(3), 449-51.

PMID: 12177385

ABSTRACT: The recovery cycle of the R2 component of the blink reflex was studied in five patients with SPS and in seven healthy control subjects. R2 recovery was enhanced in patients with SPS. This result is suggestive of hyperexcitability of brainstem interneuronal circuits in SPS. Hyperexcitability may result from abnormal input from suprasegmental structures or loss of inhibition by interneurons and is compatible with the proposal that there is a widespread dysfunction of central inhibitory mechanisms in SPS.

2001

Thompson, P.D. (2001). **The stiff-man syndrome and related disorders.** *Parkinsonism & Related Disorders*, 8(2), 147-53.

PMID: 11489680

ABSTRACT: The stiff-man syndrome (SMS) is characterised by rigidity and spasm of predominantly axial and proximal limb muscles. The cause of the condition is unknown but the finding of antibodies to glutamic acid decarboxylase (GAD) in approximately 60 percent of patients has suggested an autoimmune basis. Pathological findings are limited to a small number of cases, which are reviewed in this paper. In some, evidence of an inflammatory aetiology has been found, and there appears to be overlap with progressive encephalomyelitis with rigidity (PER), which may present with a similar clinical picture. The spontaneous muscle activity in SMS and PER is of central origin, related to release of polysynaptic spinal and brainstem reflexes. The SMS is readily distinguished from the continuous muscle activity, spasm and cramps of Isaac's syndrome and neuromyotonia, which originate in the

peripheral nervous system. Fasciculations, myokymia, myotonia and complex repetitive discharges are characteristic of these peripheral neuromuscular disorders.



Clinical Trials

Stiff-Person Syndrome

Scholarly articles are only one aspect of research. Clinical trials in which medications, medical treatments and other interventions are tested on human subjects is a vital part of understanding, treating, and possibly curing a particular disease, syndrome, or disability.

We searched two online databases: RehabTrials.org and ClinicalTrials.gov for current studies related to Stiff-Person Syndrome. ClinicalTrials.gov resulted in two current studies recruiting patients.

The first study “Rituximab to Treat Stiff Person Syndrome” is sponsored by the National Institute of Neurological Disorders and Stroke (NINDS).

ClinicalTrials.gov Identifier: NCT000914897

ABSTRACT: The purpose of this study is to test whether rituximab (Rituxan™) can relieve symptoms of SPS. People with SPS may have certain proteins in their blood called anti-GAD antibodies that may cause some of the symptoms of the diseases. Rituximab, a drug approved to treat lymphomas, targets certain white blood cells that produce antibodies. This study will determine if rituximab can be also effective in patients with SPS who have high anti-GAD antibodies.

The second study “Cause, Development, and Progression of Stiff-Person Syndrome” is sponsored by the National Institute of Neurological Disorders and Stroke (NINDS).

ClinicalTrials.gov Identifier: NCT00030940




ABSTRACT: The purpose of this study is to explore the role of various immune factors involved in producing the disease symptoms in SPS and follow disease progression in patients. SPS is a progressive disease in which unexpected noises, touches or stressful events set off muscle spasms and stiffness. This syndrome is thought to be an autoimmune disease in which the body produces antibodies that attack certain healthy tissues. The results of this study will provide a greater understanding of SPS and provide researchers with data in which to design new therapies.

For further information on these clinical trials please visit www.clinicaltrials.gov and search by the ClinicalTrials.gov Identifier. For general information about rehabilitation and/or clinical trials please visit www.rehabtrials.org and www.clinicaltrials.gov.

About reSearch:

reSearch is a new information product from the National Rehabilitation Information Center (NARIC). Each issue is based on real-world queries received by our information specialists from researchers, educators, and rehabilitation professionals around the world.

We search several sources both in-house and online, to fill these requests including:

-  REHABDATA and the NIDRR Program database
-  Education Resources Information Center
-  National Clearinghouse of Rehabilitation Training Materials
-  Campbell and Cochrane Collaborations
-  PubMed and other National Library of Medicine databases
-  Agency for Health Care Policy and Research databases
-  Center for International Rehabilitation Research Information and Exchange

and other reputable, scholarly information resources.

We hope you find these *reSearch* briefs informative in your own research.

- NARIC Information and Media Team