



The Carousel Network

**Chronic Neuroimmune Disease
Information and Support for Sonoma County**
122 Calistoga Road, #216
Santa Rosa, CA 95409
www.cndsinfo.net

Sjögren's Syndrome

Sjögren's (pronounced "SHOW-grins") syndrome is an autoimmune disease in which the body's immune system mistakenly attacks its own moisture producing glands. Sjögren's is one of the most prevalent autoimmune disorders, striking as many as 2-4 million Americans. Nine out of ten patients are women. The average age of onset is late 40s although Sjögren's occurs in all age groups in both women and men.

The Carousel Network (TCN) offers information on the various diseases and disorders associated with chronic neuroimmune diseases, such as chronic fatigue syndrome, fibromyalgia, multiple chemical sensitivity, autoimmune thyroid disease, etc. The information is intended to help patients and caregivers make informed decisions about the patient's health, diagnostic testing, and treatment in conjunction with their health care practitioners. TCN does not diagnose patients nor recommend specific medical or palliative treatments.

The Carousel Network is a 501(c)3 nonprofit supported by memberships and donations.

Membership is \$20/year; make checks payable to The Carousel Network, POB 366, Fulton CA 95439-0366.

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Sjögren's Syndroms

Sjorgren's Syndrome Foundation, Inc.

Introduction

Sjögren's syndrome is an autoimmune disease in which the body's immune system mistakenly attacks its own moisture producing glands. Sjögren's is one of the most prevalent autoimmune disorders, striking as many as 2-4 million Americans. Nine out of ten patients are women. The average age of onset is late 40s although Sjögren's occurs in all age groups in both women and men.

The hallmark symptoms are dry eyes and dry mouth. Sjögren's can also cause dryness of the skin, nose, and vagina. Sjögren's also may affect other organs, such as the kidney, GI tract, blood vessels, lung, liver, pancreas, and the central nervous system. Many patients experience debilitating fatigue and joint pain.

Symptoms can plateau, worsen, or go into remission. While some people experience mild symptoms, others suffer debilitating symptoms that greatly impair their quality of life.

When Sjögren's syndrome occurs alone and no other connective tissue disease is present, it is called "Primary Sjögren's." When Sjögren's syndrome is accompanied by a connective tissue disease, such as rheumatoid arthritis, lupus, or scleroderma, it is called "Secondary Sjögren's." The term "secondary" in no way implies that Sjögren's syndrome is less important than the other co-morbid illness. Approximately half of people with Sjögren's have Primary, and the other half have Secondary Sjögren's.

Diagnosis

Early diagnosis and treatment are important for preventing complications. Nevertheless, it sometimes takes a Sjögren's patient 2 years to get this diagnosis. Sjögren's symptoms may mimic other diseases, such as lupus, multiple sclerosis, or rheumatoid arthritis. Furthermore, dryness can occur for other reasons, such as a side effect of medication like anti-depressants or high blood pressure medication.

Rheumatology is the medical specialty that has primary responsibility for diagnosing and managing Sjögren's syndrome. Ophthalmologists and dentists are also specialists who diagnose symptoms associated with Sjögren's.

Once Sjögren's syndrome is suspected, the rheumatologist will take a medical history and do a series of blood tests to confirm the diagnosis. S/he will also refer the patient to an ophthalmologist for further tests, and to an oral pathologist for additional procedures.

The following list includes some of the blood tests that the rheumatologist will order:

ANA (Anti-Nuclear Antibody)

ANAs are a group of antibodies that react against normal components of a cell nucleus. They are present in a variety of autoimmune diseases, so the test is not disease specific. About 70% of Sjögren's patients have a positive ANA test result.

SSA and SSB

The antibodies SSA (or RO) and SSB (or LA) are often found in Sjögren's syndrome; 70% of patients are positive for SSA and 40% are positive for SSB.

RF (Rheumatoid Factor)

This antibody test is indicative of a rheumatic disease, but, like the ANA test, is not specific to Sjögren's syndrome. In Sjögren's patients, 60-70% have a positive RF.

ESR (Erythrocyte Sedimentation Rate)

This test measures inflammation. An elevated ESR can indicate an inflammatory disorder, including autoimmune and connective tissue diseases, like Sjögren's syndrome.

IGs (Immunoglobulins)

Immunoglobulins are normal blood proteins. They are usually elevated in Sjögren's.

The following are tests that the ophthalmologists will perform to test for dry eye:

Schirmer Test

Small pieces of filter paper are placed between the lower eyelid and eyeball. The amount of wetting in 5 minutes gives a rough estimate of tear production.

Rose Bengal and Lissamine Green

These dyes are used to observe abnormal cells on the surface of the eye.

Slit-lamp Exam

This test provides an indication of the volume of tears by magnifying the eye and viewing it in its resting state. The amount of tears is then examined.

The dentist or oral pathologist will perform the following tests:

Parotid Gland Flow

This test is a quantitative measure of the amount of saliva produced over a certain period of time.

Salivary Scintigraphy

This test measures salivary gland function by injecting radioactive material into the salivary glands.

Sialography

This is an x-ray of the salivary-duct system taken after a radiologically sensitive dye is injected.

Lip Biopsy

This test is used to confirm lymphocytic infiltration of the minor salivary glands. An incision of approximately two centimeters is made on the inside surface of the lower lip. Minor salivary glands are removed and examined under the microscope.

Treatment

While, there is no known cure for Sjögren's syndrome, many problems can be treated symptomatically with over-the-counter and prescription medications. Other helpful tips for coping with Sjögren's symptoms are also available from the Sjögren's Syndrome Foundation.

Over-the-counter moisture replacement therapies are available to ease the symptoms of dryness. These include preservative-free artificial tears, artificial salivas, unscented skin lotions, saline nasal sprays, and vaginal lubricants. The Sjögren's Syndrome Foundation maintains an updated list of these products. The Foundation also offers tips for daily living in *The New Sjögren's Syndrome Handbook* and in its *Moisture Seekers* newsletter.

Two prescription medications, Salagen (pilocarpine hydrochloride) and Evoxac (cevimeline), are available to treat the dry mouth associated with Sjögren's. Depending on the nature and severity of symptoms, other medications include non-steroidal anti-inflammatory drugs (NSAIDs), steroids, and immunosuppressive drugs.

There are also non-medication strategies for dealing with the various symptoms of Sjögren's syndrome. *The New Sjögren's Syndrome Handbook* contains a chapter covering many helpful ideas. *The Moisture Seekers Newsletter* is also a great source of helpful hints.

THE MANAGEMENT OF DRY EYE IN SJÖGREN'S SYNDROME

Michael A. Lemp, MD

Sjögren's Syndrome Foundation Publication

The term "dry eye" covers a variety of conditions affecting the surface of the eye. These conditions are characterized by either a quantitative or qualitative deficiency in tear production, with a

drying effect on the ocular surface. Sometimes dry eyes are associated with dryness in other parts of the body and with systemic inflammation such as in Sjögren's syndrome. In other cases, however, the dryness is limited to the eyes alone.

The reasons for decreased tearing and its effect on the ocular surface are complex but are beginning to yield to systematic scientific inquiry. Recent evidence has demonstrated a strong relationship between hormonal changes in the body and the body's immune system in initiating and sustaining changes in the lacrimal glands and other structures such as the oil glands of the eyelids.

Artificial Tears and Ointments

The use of artificial tear solutions has been a mainstay in the treatment of dry eyes for many years. Artificial tears are formulated to mimic the composition of natural tears. Natural tears, however, contain such an array of substances in small amount that it is impractical to formulate artificial tears exactly the way nature produces tears. To be useful, however, artificial tears should closely mimic the composition of natural tears in their electrolyte compositions, their tonicity and their pH. These terms refer to how thick the tears are, how much "salt" they contain, and their acid balance. Most artificial tears meet these criteria; otherwise they would sting and be uncomfortable upon insertion. The main limitation of artificial tears is their relatively short duration of action. Many manufacturers add thickening agents such as polymers in an attempt to prolong the effectiveness of the eyedrops.

All eyedrops which are sold in bottles for multiple use must contain preservatives to protect against contamination of bacteria. Unfortunately, the generally used preservatives are also irritating to the eye and, when used frequently, can cause ocular irritation beyond that experienced from the dry eye itself. Recently manufacturers have begun producing unit dose dispensers (use once and throw away) which can be sterilized at the factory to protect against contamination. The packaging is expensive but preservative-free artificial tears are much gentler on the ocular surface and are, in general, recommended for anyone using an artificial tear more than three or four times per day. A new product, from CibaVision, contains a preservative in a multi-use bottle which is deactivated by ingredients in the natural tears and is reportedly less irritating.

Ointments and Lacriserts

No currently available artificial tear lasts throughout the night, whereas ointments do. Ointments increase the lubricity between the lid and the surface of the eye which can last during the hours of sleep. Some people however, find ointments irritating.

Lacriserts® (Rx) are small pellets which dissolve when placed under the lower lid (in very dry eyes, they must be "started" with a drop of artificial tears) where they release tears over many hours. They can be useful for certain patients with moderate to severe dry eyes. The drawbacks, however, include formation of a thick tear layer over the lower lid which can interfere with reading. Also, they sometimes fall out of the eyes.

Bandage Contact Lenses and Prescription Medications

In general, contact lenses are contra-indicated for patients with dry eyes as they increase the risk of infection. In some instances, however, particularly where there is a surface problem that needs temporary aid in healing, a special bandage contact lens can be useful in lessening the friction between the upper lid and the surface of the eye allowing the sore area to heal.

In particularly severe forms of dry eyes in which there is a large immunological component, some prescription medicines that decrease inflammation and immunologic activity are indicated. These are strong medications which can have side effects and should only be used under strict supervision of an ophthalmologist thoroughly familiar with their use.

Punctal Occlusion

Blocking of the drainage of tears in order to preserve moisture on the surface of the eye can be highly effective in certain patients with moderate to severe dry eyes. In this procedure either one or two of the openings in the upper and lower eyelids can be surgically blocked. This is usually done by the application of heat or laser. These closures are usually permanent, although they can reopen. Studies have shown that the reopening rate is much higher with laser treatment than with direct heat application.

The use of temporary blocking agents such as silicone plugs or collagen implants can also be helpful. I find the use of silicone plugs, particularly in the inferior puncta, to be an excellent way to assess the effects of tear blockage.

Patient Management

In my experience, people with dry eyes can be effective managers of their condition. The ocular surface is exquisitely sensitive to changes and the patient is frequently the first to recognize that. I believe such self-management, however, should be guided by professional expertise and the development of a treatment plan which includes periodic monitoring. Working with your doctor with periodic visits will maximize your chances for effective management of dry eyes.

LIVER ABNORMALITIES ASSOCIATED WITH SJÖGREN'S SYNDROME

Karen L. Lindsay, MD

Sjögren's Syndrome Foundation Publication

Introduction

Sjögren's syndrome is a chronic, autoimmune disease that can affect a number of organ systems, including the lungs, kidneys and liver. When it comes to involvement of the liver, there are three different, separate and distinct liver diseases associated with Sjögren's syndrome: primary biliary cirrhosis (PBC), chronic active hepatitis of the autoimmune type and hepatitis C virus infection.

Both PBC and chronic active hepatitis of the autoimmune type are primary idiopathic immunologic autoimmune disorders; that is, the cause of either is not known. These diseases are manifestations of the body making antibodies against itself, against liver cells. Hepatitis C virus infection, on the other hand, is a distinct entity caused by the hepatitis C virus, a recently described virus.

Incidence of Liver Abnormalities

It appears that the frequency of liver involvement in SS patients may be greater than once thought. A large study done in Europe in patients classified as having primary SS, defined, by these investigators, as symptoms limited to sicca syndrome (dry eyes and dry mouth), found that 25-30% of patients had abnormal liver blood tests. When these abnormalities were evaluated further, 10% of patients thought to have only primary SS were found to have primary biliary cirrhosis (PBC). Another 4% had chronic active hepatitis of the autoimmune type. The important aspect of this study is that even when patients are thought clinically just to have primary SS, they can, in fact, have either of these two autoimmune liver diseases.

Sjögren's Syndrome Associated Liver Diseases

Primary Biliary Cirrhosis (PBC)

This disease is characterized by immune-mediated destruction of small bile ducts inside the liver. We do know a great deal about the features of this disease, and we also have a treatment. One of the remarkable characteristics of the disorder, like many autoimmune diseases, is that it affects primarily females; the vast majority of patients, at least 85%, are women.

It is important to recognize that the disease starts at one point in time, perhaps when patients are in their 20s, 30s, 40s or 50s, and slowly develops over decades. The sequence of events and the order in which patients develop the various manifestations of the disease may vary somewhat, but it is clearly a chronic disease.

The predominant symptoms of primary biliary cirrhosis are itching and fatigue. The itching characteristically occurs all over the body, to varying degrees of intensity, with increased skin pigmentation in the areas of itching, frequently noticeable on the back. There may also be visible cholesterol deposits in the skin.

The simplest way to evaluate whether or not someone with itching has PBC is to perform a routine liver panel blood test. If the itching is caused by PBC, then the tests will be abnormal. That's how we differentiate the itching of PBC from all the other many causes of itching.

Patients with PBC have abnormal liver blood tests with a distinct pattern, as well as abnormal liver biopsies characteristic of this disease. The liver biopsy tissue should be examined by a pathologist

skilled in interpreting liver diseases. At least 20-30% of the patients I see with PBC have had previous liver biopsies that have not been read correctly by the pathologist, simply because the pathologist was not thinking of PBC.

The treatment available for PBC, ursodeoxycholic acid (Actigall), is moderately effective, although its effect on long-term survival, at this point, is not known. Actigall is effective, however, in relieving the symptoms and the biochemical abnormalities of PBC. Use of Actigall began experimentally in this country in 1983, although, prior to that time it had been used in Europe and Japan for other conditions. To my knowledge, there have been no reports of any serious side effects with Actigall. Should the patient's disease advance to the point of requiring liver transplantation, PBC patients, fortunately, do very well.

Regarding the frequency of Sjögren's syndrome in patients with PBC, about one third of patients have symptoms of dry eyes and dry mouth, and about 40% have antibodies characteristically found in Sjögren's syndrome. In a few studies, when more extensive evaluation was done with biopsies of salivary glands, abnormalities were found in 95% of the patients. So, although it may not be clinically obvious, microscopic evidence of Sjögren's syndrome may be very common in patients with PBC.

Chronic Active Hepatitis of the Autoimmune Type

This liver disease also affects primarily females, in that 85% of the patients are women. It is characterized by abnormal liver blood tests and characteristic features on liver biopsy. Most patients have nuclear antibody and smooth muscle antibody. At least 50% have extrahepatic abnormalities of the skin, arthritis, or Sjögren's syndrome. Immunosuppressive medications are usually very effective in controlling this liver disease.

Hepatitis C Virus Infection

The third distinct liver disease associated with Sjögren's syndrome is the newly recognized hepatitis virus infection, hepatitis C. For many years, physicians recognized hepatitis occurring after blood transfusion and in IV drug abusers. It was thought to be due to a chronic viral infection. Through sophisticated molecular biology techniques, the virus was identified only five years ago and named hepatitis C virus.

This infection, in humans, almost always becomes chronic, that means the individual cannot get rid of it. Only about 60% of patients with chronic hepatitis C virus infection have abnormal liver blood tests. The others have the infection without abnormalities in liver blood tests, but on liver biopsy, abnormalities of the liver can be found. This chronic infection goes on to progressive liver disease and can result in liver failure. A treatment, alpha interferon, is successful in at least 10—20% of patients.

Since identification of this virus is so recent, a great deal still needs to be understood about its relationship to Sjögren's syndrome. Hepatitis C is diagnosed by the presence of the antibody to the hepatitis C virus in the blood. In one study of 49 patients with hepatitis C virus infection, salivary gland biopsies were taken to determine if there was evidence of Sjögren's syndrome in the salivary glands. About half of the patients had what was described as lymphocytic capillaritis, an abnormality that is somewhat different from the features seen in classical Sjögren's syndrome. Only a small number of the original group actually had sicca syndrome or the characteristic antibody.

Work on hepatitis C is just emerging, but I suspect that as time passes and we have the opportunity to look at patients with hepatitis C virus infection more carefully, we will identify more Sjögren's syndrome.

Diagnosis

If you are a patient with Sjögren's syndrome and concerned about liver involvement, ask your doctor if specific liver blood testing has been done. A Sjögren's syndrome patient with any liver blood test abnormality should be evaluated further.

Not all doctors are familiar with these specific liver diseases. If your doctor leaves you with questions about whether or not you have a liver problem, and you have abnormal liver blood tests, then it's best to see a gastroenterologist who has been specifically trained in liver disease and can answer those questions. An abnormal liver test, by definition, means there is something wrong with the liver, but sometimes, only a specialist can determine what the problem is, whether it is serious, whether it is related to Sjögren's syndrome and what else needs to be done. In some cases, the abnormality may be due to medications.

THE ROLE OF ANTINUCLEAR ANTIBODIES IN SJÖGREN'S SYNDROME DIAGNOSIS

Steven E. Carsons, MD

Sjögren's Syndrome Foundation Publication

The presence of antibodies directed against the nucleus of the cell is a hallmark of auto immune disease. Since the 1960s, the ANA (antinuclear antibodies) has become the most important screening test for autoimmune diseases. Many patients and physicians associate a positive ANA with the diagnosis of systemic lupus erythematosus (SLE); however, ANA is not specific and can be present in a wide variety of autoimmune disorders including Sjögren's syndrome (Table 1).

What does a positive ANA mean?

Several characteristics of ANAs are helpful in deciding their disease association and specificity. These include the titer and pattern. The titer is the degree of positivity of the test and is expressed as a ratio of the dilution of the patient's serum (watery portion of the blood after coagulation) required to produce a positive test, i.e., 1:80; 1:320. The higher the denominator (the second number) the more strongly positive the ANA. Most laboratories set a positive cutoff at 1:40 or 1:80. This is somewhat arbitrary; however, it is important to note that values below these levels may not be clinically significant.

ANA Patterns

When viewed under a fluorescent microscope, ANAs form visible patterns in the cell nucleus. Four major patterns are recognized: rim, homogeneous, nucleolar and speckled. The presence of a speckled pattern is commonly seen in Sjögren's syndrome and overlap connective tissue disease syndromes, whereas a rim or homogeneous pattern is more typical of lupus.

Although ANAs are associated with autoimmune disorders, their presence is never diagnostic of any particular rheumatic disease including Sjögren's syndrome. Rheumatic diseases are diagnosed on the basis of classical clinical finding; laboratory data is used to corroborate that diagnosis.

Test Specificity—SSA and SSB

Specificity has been increased in some newer tests that break down a positive ANA into several of its components (Table 2). These tests can be used in the follow up evaluation of a patient with a positive ANA. Some of these tests may help a clinician learn more about a patient's condition than the basic ANA for a particular rheumatic disease. The so-called Sjögren's Antibodies, SSA and SSB, are present in 70% and 40% respectively of patients with Sjögren's syndrome. These are also referred to as anti-Ro and anti-La. However, these antibodies are not entirely specific for Sjögren's syndrome; 30% of patients with lupus (SLE) are SSA positive.

The Role of "Diagnostic Panels"

All of the tests described above can be individually ordered by a physician and have been available in most cases for the past ten years. More recently, clinical diagnostic laboratories have offered "diagnostic panels" to physicians. These panels simply offer a group of the autoantibody tests at a potential cost savings over the price of individual tests ordered separately. Panels may be more costly, however, if the physician orders an entire panel instead of choosing the one or two most relevant tests for that patient's clinical condition. Such panels include: ANA Rheuma Screen™, ANalyzer™, Rheumatoid profile, and Autoimmune profile. Although these panels may have slight differences in their sensitivities, it is important to note that they do not represent significant diagnostic advances for Sjögren's syndrome patients.

The ability to analyze antinuclear antibodies has provided the physician with an important tool in the diagnosis and management of auto immune disorders. It is important to remember that no single blood test is diagnostic for Sjögren's syndrome or any rheumatic disease in the absence of a complete clinical evaluation. Commercially available ANA panels add convenience, and in some instances may be cost effective, but do not represent significant diagnostic advances in the management of Sjögren's syndrome.

THE PERIPHERAL NERVOUS SYSTEM IN SJÖGREN'S SYNDROME

Elaine L. Alexander, MD, PhD

Sjögren's Syndrome Foundation Publication

The nervous system is the “essence of human beings or being human.” Basically, the nervous system can be divided into the central (CNS) and peripheral (PNS) nervous systems. The CNS is composed of the brain and spinal cord. Although some patients with Sjögren's syndrome can develop CNS or PNS disease (or both), this article will focus on defining and describing PNS disease in Sjögren's syndrome.

Description of the Peripheral and Autonomic Systems

The PNS can be viewed as the “executor” of impulses or signals generated by the brain and spinal cord. The spinal cord connects the brain to the peripheral nerves. Anatomically, the PNS is composed of the cranial nerves, spinal nerves with their roots, rami and ganglia (relay stations that send impulses to and from the CNS), the peripheral nerves, and the peripheral components of the autonomic nervous system.

There are three basic major types of peripheral nerves: sensory, motor and autonomic. These nerves stimulate sensory organs, the musculoskeletal system and internal organs. Sensory nerves serve as “sensors” of the environment, picking up signals or input from such organs as the eyes, ears, organs of taste and smell, mucous membranes covering or lining many tissues, and skin. These sensory messages are relayed to the brain for interpretation, integration, planning, and execution of subsequent actions.

The motor system responds to these sensory stimuli and initiates voluntary/involuntary activity. Motor actions are mediated by motor nerves. Motor actions include such activities as blinking, swallowing, speaking, writing, standing, walking, running, etc.

The autonomic system is another very important “noncentral” part of the nervous system and can be classified into the sympathetic, parasympathetic and gastrointestinal systems. The autonomic nervous system is the key to regulation of internal organ function. It controls vital functions of the lungs, cardiovascular system, gastrointestinal organs, and bladder and sexual function. Malfunction of any one or more parts of this system potentially can result in significant clinical symptoms of dysfunction.

The types of abnormalities that can affect the peripheral or autonomic nervous systems in Sjögren's syndrome patients are described below.

Clinical Spectrum of PNS Disease in Sjögren's Syndrome

The sensory, motor and autonomic components of the peripheral nervous system alone or in combination may be affected in Sjögren's. In many respects, the spectrum of PNS disease in Sjögren's syndrome resembles the many types of neuropathy seen in diabetes.

Sensory Neuropathy

Among the most common PNS manifestations of primary Sjögren's syndrome is a disease of the nerves (neuropathy) involving sensory nerves supplying the feet and hands. The neuropathy usually involves the lower extremities more than the upper extremities.

Cranial Nerves

There are twelve cranial nerves defined as sensory or motor (or both) nerves that stimulate structures within the cranium (above the neck). These nerves carry out important functions such as vision, taste and smell, coordination of movement of eye muscles, hearing and speech, chewing, swallowing, etc. Sjögren's syndrome patients may develop abnormalities of one or more of these cranial nerves.

Entrapment Neuropathy

Another common type of peripheral nervous system involvement in Sjögren's syndrome includes the “entrapment syndromes”—carpal, ulnar and tarsal syndromes. In these syndromes, the neurologic symptoms correspond to the abnormalities caused by compression of the nerves in the arm, wrist and ankle. Entrapment of the network of nerves stimulating the upper extremities and lower extremities has also been observed in Sjögren's syndrome.

Diagnosis

The main approach to the objective confirmation of suspected clinical PNS disease in Sjögren's syndrome is electro-physiologic. Electrophysiologic changes occur only after significant damage has been done to the PNS. Electrophysiologic studies include nerve conduction studies, electromyography, and quantitative sensory testing. Spinal cord (MR1) studies may be useful. Finally, nerve biopsies are used in certain cases to establish diagnosis and exclude other causes. Cerebrospinal fluid analysis should be performed in all cases. Central nervous system disease should be sought and evaluated in Sjögren's syndrome patients with PNS disease, because CNS and PNS disease co-exist in some patients.

Therapy

The current treatment of peripheral neuropathies can be divided into two fundamental approaches: symptomatic management and immunosuppressive therapy. Symptomatic treatment including salicylates (aspirin), acetaminophen, nonsteroidal antiinflammatories, anti-depressants, and anti-epileptics. Supervised exercise, physical therapy, and rehabilitation are essential and important adjuncts to therapy.

The main reason(s) the peripheral and autonomic nervous systems may be impaired in Sjögren's syndrome relate to an inflammatory or autoimmune attack on the nerves or ganglia or both. Thus, immunosuppressive therapy may be indicated in progressive cases of peripheral neuropathy resistant to symptomatic measures. Immunosuppressive therapy includes corticosteroids (prednisone), hydroxychloroquine (plaquenil), azathioprine, methotrexate, and cyclophosphamide. Plasmapheresis and intravenous gammaglobulin has been used in some cases.

New Directions for More Effective Therapy

None of the current therapies for the treatment of peripheral neuropathy specifically addresses either the initiation of the vascular inflammatory process or the reversal of existing pathology. In addition, all of the immunosuppressive agents have significant side effects. Axonal degeneration (destruction of the fibers that go from the neuron cell body to the target organ—muscle, sweat gland, etc.) and, to a lesser extent, demyelination (damage or destruction of the nerve-covering that conducts impulses), occur in peripheral neuropathy. In addition ganglia neurons are attacked by lymphocytes and die.

Recombinant human neurotrophic or growth factors (factors that protect neurons, ganglia cells and nerves from death and damage, as well as, facilitate repair of damaged nerves and muscles) and small molecules that either upregulate the expression of neurotrophic factors or molecules that mimic them, hold tremendous future promise for more effective and targeted therapy in Sjögren's syndrome associated peripheral and central nervous system disease.

Neurotrophic factors that might be beneficial in the treatment of neuropathy in Sjögren's syndrome include insulin-like growth factor (IGF-I), nerve growth factor (NGF), neurotrophin 3 (NT3) and new molecules currently under development. IGF-I has the potential for preventing or reducing the vascular-inflammatory response, protecting ganglia neurons, and enhancing nerve regeneration, collateral sprouting and remyelination of damaged axons. Such agents currently are being used in clinical trials of other neuropathies associated with diabetes and chemotherapy.

Clinicopathological Findings Consistent with Primary Sjögren's Syndrome in a Subset of Patients Diagnosed with Chronic Fatigue Syndrome: Preliminary Observations

David A. Sirois And Benjamin Natelson

J Rheumatol 2001;28:126-31

www.jrheum.com/abstracts/abstracts01/126.html

OBJECTIVE. Some patients diagnosed with chronic fatigue syndrome (CFS) have symptoms commonly observed in Sjögren's syndrome (SS), particularly xerophthalmia and xerostomia, leading to speculation that some patients with CFS might have primary SS or that the 2 disorders share common pathophysiological features. We investigated the prevalence of symptoms of mucosal dryness, salivary gland pathology, lacrimal hyposecretion, and autoantibodies (antinuclear antibody, SSA/SSB) among patients diagnosed with CFS.

METHODS. Twenty-five subjects with CFS and 18 healthy control subjects were interviewed and examined, had a Schirmer test and fluorescein tear dilution, and underwent minor salivary gland (MSG) biopsy. Antibody to nuclear antigen as well as anti-La (SSA) and anti-Ro (SSB) antibody were available for subjects with CFS. Pathologists unaware of the subject group assignment examined labial salivary gland biopsy specimens and calculated a standard MSG score for each specimen.

RESULTS. Mucosal dryness was reported by 13/25 (52%) subjects with CFS, of which 8 (32%) also had MSG score, low Schirmer test value, and symptoms consistent with primary SS ($p = 0.05$). No control subject met diagnostic criteria for primary SS. MSG focus scores ≥ 1 were common among both groups (CFS 14/25; controls 15/18). MSG results without pathological alteration were rare, seen in only one control and no CFS patients. Low Schirmer values were found in 10/25 (40%) CFS patients and 1/18 (6%) control ($p = 0.01$).

CONCLUSION. A subset of patients with CFS may have primary SS.

FOR ADDITIONAL INFORMATION AND RESOURCES:

Organizations:

National Sjogren's Syndrome Association

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Beachwood, OH 44122
(216) 292-3866; Fax: (216) 292-4955 (800) 395-6772
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Sjögren's Syndrome Foundation, Inc. (SSF)

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www.sjorgrens.org

SSF members receive the *MoistureSeekers* newsletter (9 issues/year) and other information. Individual memberships: \$25/year Health care professional: \$40/year. The following book, newsletter and videos are available through the SSF:

The New Sjögren's Syndrome Handbook. By Carsons, Harris. Hardcover, 256 pages. Published by Oxford University Press, 1998. SSF Members: \$20; Non-members: \$25

The MoistureSeekers Newsletter. A publication of the Sjögren's Syndrome Foundation. A year of *The Moisture Seekers Newsletter*. Past issues have been reformatted and contain all medical and patient information as originally published. Each volume is bound in a notebook. Volume 14 - 1996, Volume 15 - 1997, Volume 16 - 1998 and Volume 17 - 1999 are available; \$19.95 per annual issue.

International Conference on Sjögren's Syndrome Video

Two tape set (4 1/2 hours). A panel of international experts discuss the various aspects of Sjögren's syndrome; lectures and question and answer sessions. \$42.50

Xerostomia (Dry Mouth) Video

30-minute video tape, *The Health Care Professionals' Guide to Xerostomia*, explains the many causes of dry mouth and current treatments. For both health care providers and patients. \$29

The SSF website has a library of .pdf formatted articles that may be read and printed out.

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Treatment Centers

Sjögren's Syndrome Clinic

University of California
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Federal Agencies Funding/Studying Sjögren's Syndrome Research

NIAMSD/National Institute of Arthritis and Musculoskeletal and Skin Diseases
Information Clearinghouse
National Institutes of Health
1 AMS Circle
Bethesda, Maryland 20892-3675
877-226-4267 301-496-8188
www.nih.gov/niams/

NINDS/National Institute of Neurological Disorders and Stroke
National Institutes of Health
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