

lupus  **FACT SHEET**

Lupus Canada

The Nervous System and Lupus

Neurological manifestations of systemic lupus erythematosus (SLE or lupus) are common and vary from mild to severe. People with lupus can experience bouts of memory loss, headaches, strokes and cognitive dysfunction, which generally means difficulty concentrating or reasoning. These symptoms arise when lupus affects the body's nervous system.

There are no definitive answers as to how or why lupus affects the nervous system. It is known that the nervous system requires an uninterrupted flow of blood to supply its tissues with oxygen and nutrients necessary for normal functioning. There are theories regarding how lupus may cause the many symptoms of nervous system involvement:

Symptoms include difficulty finding words and retrieving information.

- Nerve tissue may be damaged when antibodies attack nerve cells or blood vessels.
- Nutrients and oxygen are delivered through blood vessels that feed the brain, spinal cord and nerves. If blood flow is slowed or interrupted, the cells of the nervous system are injured and unable to function normally. Symptoms develop as a result.
- The symptoms vary depending on the location and extent of the tissue injury.

Your doctor may order one or more laboratory tests to determine whether or not your lupus is causing neurological involvement and to what extent. These tests may include magnetic resonance imaging (MRI), electroencephalogram (EEG), electromyogram (EMG), computed tomography (CT) scan, lumbar puncture (spinal tap) or blood tests. X-rays may also be used on occasion to diagnose neurological damage.

- CT and MRI obtain images of the brain that reveal strokes, tumours, bleeding and abscesses.
- EEG detects abnormal electrical activity of the brain, which may be associated with seizures (epilepsy).
- EMG and nerve conduction studies detect abnormal electrical activity of muscles and nerves.
- Spinal fluid analysis reveals distinct changes in the spinal fluid that point to various disorders (infection, inflammation, etc).
- Blood tests may show bleeding problems, or antibodies that can lead to abnormal blood clotting (e.g., antiphospholipid antibodies, lupus anticoagulant).



UNDERSTANDING YOUR NERVOUS SYSTEM

The nervous system is made up of the brain, the spinal cord and the nerves throughout the body. It has three distinct parts:

- The **central nervous system** (CNS) consists of the brain and spinal cord.
- The **peripheral nervous system** is comprised of nerve fibres that supply the skin and muscles with the power needed for sensation and movement.
- The **autonomic nervous system** is the nerve supply to glands and other internal organs.

While we don't completely understand the mechanisms by which lupus affects the central nervous system, we know that there are several neurological syndromes that affect many people. Although cognitive impairment is believed to be very common in lupus, few patients undergo specific testing to document it. Symptoms include difficulty finding words and retrieving information. People with the condition often refer to it as "lupus fog."

A five-year study on cognitive function, called Brain CONNECTIONS, evaluated lupus patients every four months with a wide variety of tests. In one test, participants were given a list of words and asked to recall them after about 10 minutes. Although most participants had problems recalling the words, they were able to pick out the original 10 words from a list of 20 words. This indicates interference with access to the information that exists in the brain.

Researchers with the Brain CONNECTIONS study also measured cognitive dysfunction and biological changes in the participants and found a high incidence of anatomic brain abnormalities. People whose MRIs showed brain abnormalities were more likely to have problems with focussed attention and reaction time, and also had higher lupus disease activity. This suggests that both functional and structural damage to the brain is evident early in the disease process, which underscores the need for research into the underlying mechanism of the brain's involvement in lupus.

Important neurological syndromes seen in lupus

Central nervous system vasculitis

The most serious neurological syndrome associated with SLE is central nervous system vasculitis, an inflammation of the brain's blood vessels. Most episodes (more than 80 percent) take place early, within five years of lupus diagnosis.

Antibodies attack the blood vessels, causing inflammation,

which can seriously affect the flow of blood.

A relatively small number (some estimate 10 percent) of people with lupus develop central nervous system vasculitis, experiencing such symptoms as high fevers and seizures, and possibly psychotic or bizarre behaviour. Central nervous system vasculitis usually requires hospitalization and is treated with high doses of corticosteroids and other drugs.

Cerebrovascular accidents (strokes)

A fairly large number (some estimate one-third of all people with lupus) have anticardiolipin antibodies or a lupus anticoagulant.



PROBLEMS WITH MEMORY?

- Get a physical exam to make sure your memory loss isn't due to another medical condition.
- Pay attention when you receive new information. Repeat it or write it down. Verify any details you aren't sure about.
- Don't clutter your life with things that aren't important.
- Focus on one task at a time.
- Take good care of yourself: exercise, eat well and get adequate sleep.
- Learn memory techniques, such as associating a person's name with an image or repeating the name several times in conversation.

These antibodies increase blood clot development (despite the fact that the latter has the term “anticoagulant” in its name – this is a title that has stuck over the decades). An estimated one-third of these individuals may develop blood clots in various parts of the body. When a blood clot occurs in the central nervous system, it can cause a cerebrovascular accident, or stroke. Symptoms of stroke include sudden weakness or numbness in the face, arm or leg, or difficulty speaking.

The risk of stroke in lupus may be increased even without anticardiolipin antibodies or a lupus anticoagulant. Depending on the person’s risk factors, the risk of stroke may be reduced with medications, such as blood thinners and aspirin. Blood pressure control and cholesterol treatment are important. Smoking should be eliminated.

Lupus headache

Compared with the general population, people with lupus are twice as likely to have migraine-like headaches. People with lupus who also have Raynaud’s phenomenon are even more likely to have severe headaches. Headaches are



MAYBE IT'S YOUR MEDS

Medications used to treat lupus can cause side effects that are similar to the symptoms of central nervous system lupus. Non-steroidal anti-inflammatory drugs (NSAIDs) occasionally cause headache, dizziness and, rarely, meningitis-like symptoms. Anti-hypertensive medications may be associated with loss of libido or depression. Corticosteroids are associated with agitation, confusion, mood swings and, in high doses, psychosis. Withdrawal from steroids can lead to fatigue, aching and weakness.

When you are prescribed a new medication, be sure to discuss possible side effects with your doctor or pharmacist. If you experience any of these symptoms, talk to your doctor immediately.

managed using painkillers, anti-inflammatory drugs, specific migraine therapies and sometimes tricyclic antidepressants.

Mood Disorders

The signs and symptoms of neuropsychiatric lupus may be very subtle, such as mild headaches, altered mental activity or depression. In the most severe form, seizures or partial paralysis can occur. The American College of Rheumatology lists 19 neuropsychiatric syndromes associated with lupus. In one study, neuropsychiatric lupus syndromes were present in

80 percent of people with lupus; these included anxiety disorder (24%), major depressive-like episode (28%), mood disorder with depressive features (19%), mood disorder with manic features (3%), mood disorder with mixed features (1%) and psychosis (5%).

Depression is an important symptom of lupus. Depression in lupus is often wrongly attributed to having a chronic illness and all that goes with it. In fact, lupus itself causes depression, and the management of lupus — treating the underlying disease as well as possibly adding antidepressant therapy — often lifts the depression. Newer, milder antidepressants that have fewer severe side effects are now available.

During severe lupus flares, people with the disease can experience a variety of psychiatric disorders varying from mild personality disorders to severe psychotic behaviour. Some people with lupus are wrongly diagnosed as having schizophrenia at the onset of their illness. Treatment of the lupus usually results in total improvement in the psychiatric symptoms.



FOR MORE INFORMATION

- Lupus Canada: www.lupuscanada.org. This site includes electronic versions of the *Living Well with Lupus* fact sheets.
- *Lupus: The Disease with a Thousand Faces*, edited by Dr. Sasha Bernatsky and Dr. Jean-Luc Senécal, Key Porter Books (2004) ISBN 1-55263-603-8. Contact Lupus Canada to order this book.
- Lupus Foundation of America: www.lupus.org
- National Institute of Neurological Disorders and Strokes (NINDS) Information Clearing House: www.ninds.nih.gov
- LUPUS An International Journal, Vol. 12, No. 12 2003 Special issue on Central Nervous System Lupus, published by Arnold Journals, United Kingdom: arnoldjournals@hodder.co.uk

Disclaimer

Systemic lupus erythematosus is an autoimmune disease that affects thousands of Canadians, mostly women in their childbearing years. Symptoms vary greatly from person to person and treatment is highly individualized. Patients are urged to contact their physician or healthcare professional with any questions or concerns they might have.

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