Lupus: Deciphering the Clues

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Lupus Foundation of America
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The Lupus Foundation of America, Inc. (LFA) is the foremost national nonprofit health organization dedicated to finding the causes of and cure for lupus and providing support, services and hope to all people affected by lupus.

The LFA energetically pursues its mission through five program objectives:
- Provides direct financial support to researchers through LFA's own national research program, Bringing Down the Barriers
- Advocates for increased public and private sector support for research on lupus
- Translates research findings into medically sound information and programs for people with lupus, their families, physicians, and other health professionals
- Heightens public awareness of the impact of lupus
- Supports individuals with lupus, their families and caregivers

Acknowledgements

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Purpose and Objectives
The purpose of this continuing education course is to review the pathophysiology, diagnostic criteria, physical/socioeconomic impact, and standards of care in patients with lupus erythematosus (lupus).

The target audience is professional nurses, pharmacists and pharmacy technicians.

After successful completion of this course, the healthcare professional will be able to:
1. Define and classify the various forms of lupus.
2. Describe the epidemiology and pathogenesis of lupus.
3. Identify the American College of Rheumatology symptoms and laboratory criteria for the diagnosis of lupus.
4. Identify the potential impact of lupus to the various parts of the body.
5. Describe current drug therapy and disease management of the lupus patient.

Introduction
Lupus erythematosus (lupus) has been identified as a distinctive disease since the mid-1800s. In 1846, the Austrian dermatologist Ferdinand von Hebra described a distinctive rash – mainly on the face, on the cheeks and nose in a distribution not dissimilar to a butterfly.

The first published illustrations of lupus erythematosus were included in von Hebra’s text, Atlas of Skin Diseases, published in 1856. The existence of a disseminated or systemic form of lupus was firmly established in 1904 when Moriz Kaposi first described the systemic nature of the disorder: Kaposi proposed that there were two types of lupus erythematous; the discoid form and the disseminated form. The knowledge of pathogenesis and treatment of lupus has experienced an explosion in this century, especially over the past fifty years (Hochberg, 2003).

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Although the healthcare profession’s knowledge about lupus has increased dramatically, there is still much to be learned and research is ongoing to better diagnose and treat this disease process.

The term lupus (Latin for wolf) is attributed to the thirteenth century physician Rogerius who used it to describe erosive facial lesions that were reminiscent of a wolf’s bite.

Why Understanding Lupus is Important
Because of the heterogenic nature of lupus, as well as its ability to mimic other autoimmune diseases in physical presentation and laboratory findings, the diagnosis of lupus can be a prolonged and frustrating experience for both the patient and healthcare provider. To care effectively for a lupus patient, the healthcare professional needs an up-to-date understanding of the disease, its many manifestations, and its changing and often unpredictable course (NIAMS, 2006).

Lupus is also a disease of flares and remissions. Lupus can range from mild to life-threatening and should always be treated by a physician. With good medical care, most people with lupus can lead a full life.

Lupus Explained
Lupus is a multisystem disease in which autoantibody production can lead to inflammation and tissue damage in any part of the body. The pathogenesis of lupus is thought to be a combination of predisposing genetic factors and environmental factors such as medication, or infectious agents that trigger an abnormal immune response. This occurs when:

- Suppressor T cells fail to suppress, in other words there are defects in cell signaling
- There are defects in immune tolerance
- Apoptotic cells (cells going through the normal dying process) promote the creation of autoantibodies
- And/or there is loss of T regulatory cells that control auto-reactivity. This leads to the formation of autoantibodies and immune complexes, which promote inflammation and tissue damage (Wallace, 2008)

More Info

- Lupus is not contagious, even through sexual contact.
- Lupus is not like or related to cancer. Cancer is a condition of malignant, abnormal tissues that grow rapidly and spread into surrounding tissues. Lupus is an autoimmune disease, as described above.
- Lupus is not like or related to HIV (Human Immune Deficiency Virus) or AIDS (Acquired Immune Deficiency Syndrome). In HIV or AIDS the immune system is underactive; in lupus, the immune system is overactive.

Forms of Lupus
There are generally four recognized forms or types of lupus:

- **Systemic Lupus Erythematosus** (accounts for approximately 70% of all cases of lupus)
- **Cutaneous Lupus Erythematosus** (affecting only the skin - accounts for approximately 10% of all lupus cases)
- **Drug-induced Erythematosus** (accounts for approximately 10% of all cases of lupus)
• **Neonatal Lupus** (about 10% of women with anti-Ro antibodies and about 3% of all women with lupus – will give birth to a child with a syndrome known as neonatal lupus)

**Prevalence**
Lupus impacts people from all countries, although lupus disproportionately affects individuals of certain races and ethnicities. Lupus strikes mostly women of childbearing age; eight of ten new cases of the disease develop among women ages 15 to 44.

Individuals of African-Americans, Asian, and Hispanic origin have a higher prevalence of lupus than Caucasians.

Based on limited available epidemiological data on lupus, researchers believe at least five million people worldwide have lupus, with more than 100,000 new cases diagnosed every year worldwide and 16,000 new cases in the United States, although these estimates likely are low.

**More info**
- More than 1.5 to 2 million Americans have some form of lupus.
- 90% of individuals with lupus are women.
- 80% of women with the disease are between the ages of 15 and 45.
- Women of color are at highest risk.
- For every Caucasian woman with lupus, there are 2 to 3 women of color who have it.

**Test Yourself**
**Lupus:**
A. Is an infectious disease.
B. Is an inflammatory disease.
C. Generally only affects one organ system.
D. Progresses at the same rate in all patients.

The correct answer is: B. Lupus is a multisystem disease in which autoantibody production can lead to inflammation and tissue damage.

**The most common form of lupus is:**
A. Neonatal Lupus.
B. Drug-Induced Lupus Erythematosus.
C. Systemic Lupus Erythematosus.
D. Cutaneous Lupus Erythematosus.

The correct answer is: C. Systemic Lupus Erythematosus accounts for approximately 70 percent of all cases of lupus.

**The Role of Genetics, the Environment and Hormones in Lupus**
Most people with lupus are unable to correlate a specific factor (e.g. family history, environmental exposure), illness or event (e.g. pregnancy) with their subsequent diagnosis of lupus. Generally, a
A combination of genetic predisposition, environmental exposure and hormones has been shown to be likely triggers for the development of lupus.

Genetics, environmental exposure and hormones are likely triggers for the development of lupus.

More on Genetics
No gene or group of genes has been proven to cause lupus. Lupus does appear to be more common in certain families, and when one of two identical twins has lupus, there is an increased chance that the other twin will also develop the disease. These findings, as well as others, strongly suggest that genes are involved in the development of lupus.

Although lupus can develop in people with no family history of lupus, there are likely to be other autoimmune diseases in some family members. Certain ethnic groups (African-Americans, Asians, Hispanic/Latinos, Native Americans, Native Hawaiians, or Pacific Islanders) have a greater risk of developing lupus, which may be related to genes they have in common.

Environment
While a person’s genes may increase the chance that he or she will develop lupus, it takes some kind of environmental trigger to set off the illness or to bring on a flare. Examples include:

- Exposure to ultraviolet rays from the sun or fluorescent light bulbs
- Sulfur drugs, which make a person more sensitive to the sun, such as: trimethoprim-sulfamethoxazole (Bactrim®) and (Septra®); sulfisoxazole (Gantrisin®); tolbutamide (Orinase®); sulfasalazine (Azulfidine®); other sulfa-containing medications such as hydrochlorothiazide
- Sun-sensitizing tetracycline drugs
- Penicillin or other antibiotic drugs such as: amoxicillin; ampicillin; cloxacillin
- Viral or bacterial infections
- Anything that causes stress to the body, such as physical exhaustion, surgery, physical injury, pregnancy, or giving birth
- Emotional stress, such as a divorce, illness, death in the family, or other life complications

Hormones
Hormones are the body’s messengers and they regulate many of the body’s functions. Because of the close link between the immune and endocrine systems, hormonal changes may play a part in lupus flares.

“it has been established that there are close links between the endocrine system and the immune system. For example, female sex hormones have the ability to stimulate the helper T lymphocytes, so-called because they help B lymphocytes to produce antibodies. In contrast, male sex hormones encourage suppressor T lymphocytes, which tend to block the production of antibodies.” (Isenberg & Manzi, 2008)

For instance, some women have increased lupus symptoms before menstrual periods and/or during pregnancy, when estrogen production is high. This may indicate that estrogen somehow regulates the severity of lupus. However, it does not mean that estrogen, or any other hormone, causes lupus.
Test Yourself
Which of the following are possible triggers for lupus?

A. Genetics  
B. Hormones  
C. The Environment  
D. All of the above  

The correct answer is: D. All three responses- genetics, the environment and hormones are believed to be triggers for Lupus.

Symptomatology
- Because lupus can affect so many different organs, a wide range of symptoms can occur.
- Symptoms may wax and wane.
- Different symptoms may appear at different times during the course of the disease. Individual symptoms often appear independently of the others.
- Severe symptoms may occur with relatively normal labs values, or abnormal lab values may exist with no symptoms or very mild symptoms.

More Information: Lupus Symptoms in Different Body Parts

Brain and Nervous System
People with lupus can experience a number of complications when their nervous system is affected. The symptoms may develop suddenly or may come and go over time, but they will vary depending upon the location and extent of the tissue injury. Symptoms include: headaches, confusion, fatigue, depression, strokes, seizures, and vision problems.

Eyes
Changes in vision can be a result of lupus or of the corticosteroids and antimalarials used to treat lupus. Problems can include inflammation of the eye, glaucoma, cataracts, general changes in vision, and blocked tear ducts. Rarely, the retina will be involved.

Nasal and Oral
The linings of the mouth and nose are among the most common targets for lupus involvement. Approximately 95 percent of people with lupus will experience sores or lesions inside their mouth or nose. The gums and teeth also can be affected by lack of saliva due to another common condition called Sjögren's syndrome.

Heart and Lungs
Heart disease is a major complication of lupus and is now a leading cause of death among people with lupus. The most common way that lupus affects the heart is through inflammation of the pericardium, the sac that surrounds the heart. Inflammation caused by lupus may also affect the lungs in many ways, and can involve themselves, or the blood vessels within the lungs.

Skin
Approximately two-thirds of people with lupus will develop some type of skin disease. Skin disease in lupus can cause rashes or sores (lesions), most of which will appear on sun-exposed areas, such as the face, ears, neck, arms, and legs.
Gastrointestinal System
People with lupus may be affected by problems in any area of the GI system, including the esophagus, stomach, and intestines. Lupus can also affect surrounding organs such as the liver, the pancreas, the bile ducts, and the gallbladder. Not all of these problems will be directly related to lupus disease activity; some may be traced to side effects of medication or other diseases that may be present.

Reproductive System
While women with lupus can and usually do have successful pregnancies and healthy babies, they are at higher risk for a miscarriage or premature birth. All lupus pregnancies are considered high risk and should be planned and monitored closely by a team of doctors with experience in lupus and pregnancy.

Blood Disorders
Blood disorders are common in lupus. Hematologists, physician-specialists in blood disorders, are often asked to be involved in the evaluation and treatment of people with lupus. The most common concerns are anemia (low amount of hemoglobin or low red blood cell count), thrombosis (blood clotting), and low platelet count.

Musculoskeletal System
More than half of the people who develop lupus mention pain in their joints as their first symptom. It is also not uncommon for people with lupus to experience muscle aches and pain or to have inflammation in certain muscle groups, which causes weakness and loss of strength.

Note! Severe systemic lupus may cause a temporary pattern of hair loss that is then replaced by new hair growth and can occur for other reasons besides scarring on the scalp.

Common Symptoms
The most common symptoms of lupus, for both females and males, are:
- Extreme fatigue
- New or different headaches
- Painful or swollen joints
- Fever
- Anemia
- Edema in feet, legs, hands, and/or around eyes
- Pain in chest on deep breathing (pleurisy)
- Malar (butterfly-shaped) rash across cheeks and nose
- Photosensitivity
- Hair loss
- Abnormal blood clotting
- Raynaud’s syndrome
- Mouth or nose ulcers

Lupus Symptoms Imitate Other Diseases
Lupus is sometimes called "the great imitator" because its symptoms are often like the symptoms of other diseases including:
- Rheumatoid arthritis
- Blood disorders
- Fibromyalgia
- Diabetes
- Thyroid disease
- Lyme’s disease
- Several cardiac, pulmonary and joint diseases

**Diagnosing Lupus**

Because many symptoms of lupus mimic those of other illnesses, lupus can be a difficult disease to diagnose. The diagnosis of lupus is based on a combination of physical symptoms, and laboratory/biopsy results. For most people lupus is not a quickly identifiable diagnosis. It is a diagnosis that evolves over time.

Lupus is also characterized by abnormalities in many laboratory test results. These abnormalities are patient specific and they can vary significantly during the course of a person’s disease. The serial evaluation of an individual’s tests along with the physician’s observations and the patient’s history determine the diagnosis of lupus, its course, and the treatment regimen.

All laboratory values must be interpreted in light of the patient’s present status, other correlating laboratory test results, and coexisting illnesses.

**Lupus Management**

The goal of lupus treatment is to moderate or weaken the immune system thereby decreasing or interrupting the production of autoantibodies. The treatment includes pharmacologic interventions and lifestyle changes.

Each person with lupus is unique, so each treatment plan is unique. When considering the best pharmacologic treatment, the prescriber must consider active symptoms as well as potential manifestations of lupus (e.g. non-organ-threatening disease, organ-threatening disease, organ-specific disease).

**Lupus Management**

Some people have very active immune systems. Others have immune systems that are not as active. But the immune system is not the only factor. Each patient reacts to each drug differently. The exact same medication given to five people could result in five different responses.

"There are probably many factors that determine how well a medication will work, including age, hormonal status, systems and symptoms involved, and even the diet of the patient. Severity of the disease is also important. For example, someone with a life-threatening case of lupus may require a formula of several immunosuppressives, rather than just one drug. All of this – and – more must be considered when prescribing medication (Lahita & Phillips, 2004)."
Test Yourself
Which of the following are common symptoms for lupus?
- Anemia
- Fatigue
- Painful joints
- Photosensitivity
- Excessive hair growth
- Edema of the legs, hands or feet
- Loss of pigmentation on the cheeks

The correct answer is: Anemia, fatigue, painful joints, photosensitivity and edema are only a few of the possible symptoms of Lupus. However excessive hair growth and loss of pigmentation on the cheeks are NOT symptoms of lupus.

Types of Lupus
As previously mentioned, there are four distinct forms of lupus. We will discuss each one individually including manifestations, diagnosis, and treatment.

Systemic Lupus Erythematosus (SLE)
Systemic lupus erythematosus (SLE) is the most common form of lupus, and the most complicated to diagnose and treat. SLE is what most people refer to as “lupus”; however in this course we will use the term SLE. SLE can be mild or severe and can manifest itself a variety of ways (NIAMS, 2006):

General manifestations:
- Fatigue
- Fever
- Joint pain and inflammation

Systemic Lupus Erythematosus (SLE)
Specific Manifestations
- Dermatologic: butterfly rash, photosensitivity, discoid lupus erythematosus, subcutaneous lupus erythematosus, mucosal ulcers, alopecia, bruising
- Musculoskeletal: arthralgias, arthritis, other joint complications
- Hematologic: anemia, decreased white blood cell (WBC) count, thrombocytopenia, lupus anticoagulants, false-positive venereal disease research laboratory test (VDRL), elevated erythrocyte sedimentation rate (ESR), lymphopenia
- Cardiopulmonary: pericarditis, myocarditis, myocardial infarction, vasculitis, pleurisy, valvular heart disease
- Renal: asymptomatic microscopic renal involvement, renal failure, fluid and electrolyte imbalance, edema
- Nervous System: cranial neuropathies, cognitive impairment, mental changes, seizures, stroke, peripheral neuropathy, meningitis, coma, psychosis
- Gastrointestinal (GI): anorexia, ascites, pancreatitis, mesenteric or intestinal vasculitis
- Ophthalmologic: cytoid bodies, dry eyes

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Systemic Lupus Erythematosus (SLE)

Other Key Issues

Pregnancy: Lupus flare, miscarriage or stillbirth, pregnancy-induced hypertension, neonatal lupus

Infection: Increased risk of respiratory tract, urinary tract, and skin infections; opportunistic infections

Nutrition: Weight changes, poor diet, appetite loss

Systemic Lupus Erythematosus (SLE)

Diagnosis
The diagnosis of SLE is made by a careful review of:

- Current symptoms
- Laboratory test results
- Medical history
- Medical history of close family members

The American College of Rheumatology (ACR) established criteria for SLE in 1982, which were most recently revised in 1997. These criteria establish guidelines for diagnosis and consist of 4 cutaneous, 4 systemic and 3 laboratory criteria. Many patients have three of the listed criteria plus another feature not in the current criteria.

Systemic Lupus Erythematosus (SLE)

Diagnosis
At the present time a “Revision and validation of the American College of Rheumatology (ACR) diagnostic criteria for SLE” study is underway to determine whether updating the criteria used to diagnose SLE will increase the accuracy of distinguishing SLE from other overlapping diseases. The goal is that the new criteria will identify more patients who have lupus (true positives) and fewer without lupus (true negatives).

Systemic Lupus Erythematosus (SLE)

Laboratory Findings
Laboratory findings are an important part of diagnosing SLE. The following lab values are often seen in patients with SLE.

- **Positive ANA** – The Anti-nuclear antibody test (ANA) determines if autoantibodies to cell nuclei are present in the blood.

- **Presence of anti-dsDNA** (Antibodies to double-stranded DNA). Anti-dsDNA antibodies are found in half of the people with SLE, but the disease can still be present even if these antibodies are not detected. The antibodies form in response to DNA exposed during apoptosis and can bind to proteins and deposit in tissues (like the kidney) and damage the organ.

- **Presence of anti-Sm antibodies** – Sm proteins are found in the cell nucleus, Anti-SM antibodies are found in 30-40% of people with SLE. However, the presence of this antibody is very specific to SLE and almost always indicates that the individual has the disease.

A closer look at the Anti-nuclear Antibody (ANA)

Antinuclear antibodies (ANA) are antibodies that connect, or bind, to the nucleus -- the "command center" -- of the cell. This process damages, and can destroy, the cells.
The ANA blood test is a sensitive test for lupus, since these antibodies are found in 97% of people with the disease. When three or more typical features of lupus are present -- such as involvement of the skin, joints, kidneys, lungs, heart, blood, or nervous system -- a positive ANA test will confirm a diagnosis of lupus. However, a positive ANA test result does not always mean an individual has lupus. The ANA can be positive in people with other illnesses, or positive in people with no illness. The ANA can also change from positive to negative, or negative to positive, in the same person. Still, lupus is usually the diagnosis when the ANA test is positive.

The ANA may show a negative reading in some patients who have received immunosuppressant therapy only to later return back to a positive reading.

Systemic Lupus Erythematosus (SLE)

**Laboratory Findings**

**Low C3 and C4, Serum (blood) complement test** - Complement is the name of a group of proteins that protect the body against infections. They work by strengthening the body’s immune reactions. Complement proteins are used up by the inflammation caused by SLE, so persons with inflammation due to active SLE often have low complement levels. There are nine protein groups of complement, so complement is identified by the letter C and the numbers 1 through 9. Low levels of C3 or C4 may indicate active lupus.

Systemic Lupus Erythematosus (SLE)

**Treatment**

The goal of treatment for those with SLE is to moderate the immune system. Medications are important for managing many SLE patients. An array of drug therapies is now available, and this has increased the potential for effective treatment and improved patient outcomes.

Treatment varies depending on the organ systems involved in the individual's disease.

Systemic Lupus Erythematosus (SLE)

**Approved Therapies**

At the present time there are four FDA approved therapies for treating the symptoms of SLE. The four FDA approved therapies include: Corticosteroids (prednisone, prednisolone, methylprednisolone, and hydrocortisone), Antimalarials (hydroxychloroquine (Plaquenil®) and chloroquine), the monoclonal antibody belimumab (Benlysta®), and Aspirin. Healthcare providers often rely on off-label use of medications approved for other indications (some of them being highly toxic and presenting a host of adverse side effects). Since there are no formal or standardized disease management guidelines or treatment protocols for lupus, and because each patient presents differently, physicians are essentially using case-by-case investigational methods to determine the most appropriate treatments for their patients (LFA, MASC- Standards of Care Subcommittee, 2010).

**Aspirin**

Aspirin has pain-reducing, anti-inflammatory, and anticoagulant properties that can control some of the symptoms of lupus.

**Antimalarial Agents: hydroxychloroquine (Plaquenil®), chloroquine (Aralen®), and quinacrine (Atabrine®)**

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Antimalarials have a weak anticoagulant effect and are thought to have a beneficial effect stabilizing cell membranes and warding off complications of long-term inflammation in blood vessels. They may inhibit proteins called Toll-Like Receptors, which play a role in the initial body defense against microbes. Toll-Like Receptors may contribute to autoimmunity by triggering the production of interferon alpha in response to circulating genetic material and/or autoantibodies (Dr. Joan Merrill, LFA – MSAC, 2010).

Whatever the net effects of antimalarials, their impact on immune suppression is relatively mild. Thus they are relatively safe, but not usually dramatic in dampening significant lupus flares once they start. They are often useful for rash and arthritis symptoms, but often are given along with steroids or stronger immune modulators when flares are moderate to severe. Withdrawal of hydroxychloroquine, the most commonly used antimalarial for lupus, has been shown to be associated with increased flares, suggesting a protective effect of these agents in preventing some flares of lupus (Dr. Joan Merrill, LFA – MSAC, 2010).

Hydroxychloroquine (HCQ, Plaquenil®) is the agent most frequently prescribed, followed by chloroquine and quinacrine. The antimalarials generally do not work as quickly as corticosteroids and may take several months to reach full effectiveness.

Hydroxychloroquine (HCQ, Plaquenil®) is usually dosed at 200mg/day or 200 mg twice a day. Dosing often depends on the individual and many side effects can be avoided if doses remain at, or below, 6.5 mg/kg body weight (Isenberg & Manzi, 2008).

A major potential side effect of antimalarial use is the possible damage to the retina at the back of the eye. It is important to note that retinal damage due to the use of antimalarials is dose-related, and the low doses currently used in the treatment of lupus are rarely associated with this condition.

It is necessary for the patient to see an eye doctor or ophthalmologist prior to beginning treatment with antimalarials in order to have a baseline examination. A follow-up examination every three to six months thereafter is also necessary. Eye exams of the macula to rule out fine pigmentary disturbances should be performed at least every year.

Chloroquine is dosed at 250 mg/day (3.5 mg/kg/day) with effects seen within 3 – 4 weeks sooner than that of hydroxychloroquine (HCQ). (Isenberg & Manzi, 2008) (chloroquine 250 mg = hydroxychloroquine 200 mg)

Quinacrine, which is only available from compounding pharmacies, has a rapid onset of action similar to chloroquine and is usually dosed at 100 – 200 mg/day (2.5 mg/kg/day) (Isenberg & Manzi, 2008).

**Corticosteroids including prednisone, prednisolone, methyl-prednisolone, hydrocortisone**

Prednisone is a synthetic corticosteroid preparation most often used in the treatment of SLE in the United States. It may be given as infrequently as once every other day to try to decrease risk of some side effects or as frequently as several times daily, in order to prevent some immediate side effects such as sleeplessness or nervousness. Generally, 10 mg per day or less is considered to be a low dose; more than 40 mg per day would usually be considered a high dose, but some individuals are more sensitive than others to lower doses, and a person’s weight can influence the dosing requirements (Dr. Joan Merrill, LFA – MSAC, 2010).
Prednisolone is also frequently used for SLE treatment around the world. A dose of 4 mg prednisolone has similar effects to 5 mg prednisone. When prednisone enters the body it is broken down in the liver to prednisolone. For this reason prednisolone is a better treatment for patients with liver inflammation since the liver may be involved in patients with lupus (Dr. Joan Merrill, LFA – MSAC, 2010).

Although specific doses vary a great deal from patient to patient a rough estimate suggest that patients with arthritis, pleuritis or pericarditis might often be started on doses of 20 - 30 mg/day of prednisone or prednisolone. In contrast, patients with severe kidney involvement, very low platelet counts and/or severe anemia might receive 60 mg or more. Many patients require a “steroid maintenance dose of 5 - 10 mg for many months after the acute problem has been brought under control (Isenberg & Manzi, 2008).

Steroids may also be given by intra-muscular (IM) injection or as a more shallow injection into the skin for discoid rashes, or they may be injected directly into an inflamed joint. Occasionally, very large doses of steroids may be given to critically ill patients for a short period of time in order to achieve high anti-inflammatory levels. This treatment, referred to as pulse steroids, may involve giving 500-1000 mg of methyl-prednisolone intravenously, either once or every other day for three days.

**BENLYSTA®**

BENLYSTA® is a human monoclonal antibody that specifically recognizes and blocks the biological activity of B-lymphocyte stimulator, or BlyS® (pronounced bliss), a naturally occurring protein which was discovered by scientists at Human Genome Sciences (HGS). Elevated levels of BlyS prolong the survival of B cells which can contribute to the production of autoantibodies – antibodies that target the body’s own tissues. Studies have shown that BENLYSTA® can reduce autoantibody levels and help control autoimmune disease activity

BENLYSTA® is indicated for the treatment of adult patients with active, autoantibody-positive, SLE who are receiving standard therapy.

Recommended dosage regimen is 10 mg/kg at 2-week intervals for the first 3 doses and at 4-week intervals thereafter. Reconstitute, dilute and administer as an intravenous infusion only, over a period of 1 hour (2011 Human Genome Sciences, Inc., and The GlaxoSmithKline Group of Companies).

BENLYSTA® was generally well-tolerated. The most commonly reported adverse reactions included nausea, diarrhea, fever, inflammation of the nose and throat, bronchitis, insomnia, pain in extremity, depression, and migraine.

**Systemic Lupus Erythematosus (SLE)**

*Other Therapies*

Other therapies used in the treatment of SLE include:

*Immunosuppressive medications*

*Immunosuppressive medications are used to control inflammation and the overactive immune system, especially when steroids have been unable to bring lupus symptoms under control, or when a person cannot tolerate high doses of steroids.*

- Cyclophosphamide (Cytoxan®) is used in the treatment of severe SLE when nephritis or vasculitis has become an issue or in the presence of serious CNS involvement.
- Cyclophosphamide is given in high doses via intravenous infusion. Treatment regime varies for patients but is generally given monthly for 6 months. Patients are evaluated for a response to determine specific treatment management which may include continuation of Cytoxan or a change of medication to a maintenance drug, such as CellCept®.

- Azathioprine (Imuran®) 2- 2.5 mg/kg/per day is effective in treating moderate to severe disease activity, including kidney disease and other organ-threatening manifestations (Isenberg & Manzi, 2008).

- Methotrexate is effective in treating many manifestations of SLE. It is particularly helpful in patients with SLE arthritis, skin rashes, and may allow reduction in corticosteroid dosing.

- There is a greater risk of toxicity in lupus patients using methotrexate then seen in RA patients. Toxicity danger increases with renal insufficiency and use of methotrexate for patients with serum creatinine above 2 mg/dL is dangerous. Liver dysfunction and pneumonitis can occasionally occur with the use of methotrexate. Regularly monitor blood counts and liver enzymes; modify dosage if side effects are noted.

**Anticoagulants:**

**Anticoagulants: low-dose aspirin, heparin (Calciparine®, Liquaemin®) and warfarin (Coumadin®)**

- Lupus can cause life-threatening blood clots. These drugs are used as a prophylactic treatment to avoid potential clotting issues.

- Dosage of anticoagulants is based off of frequent monitoring of prothrombin time (Protime, PT) and varies from patient to patient.

- Caution patients about the use of aspirin products when using warfarin.

**NSAIDs**

- Non-Steroidal Anti-Inflammatory Drugs suppress inflammation and are especially useful for joint pain and stiffness. Examples of NSAIDs are ibuprofen (Advil®, Motrin®), naproxen (Naprosyn®), indomethacin (Indocin®), nabumetone (Relafen®), and celecoxib (Celebrex®).

**Systemic Lupus Erythematosus (SLE)**

**Investigational Treatments**

Many medications and drug products, both old and new, are now being studied in people with SLE. Some of these investigational treatments include:

**Biologics** are genetically engineered compounds that interfere with the action of cells involved in the immune response. Biologics that may be used in SLE may include abatacept (Orencia®) and atacicept (TACI-Ig).

**Immunosuppressives (Immune Modulators)** - These include Leflunomide (Arava®) and Thalidomide (Thalomid®).

**Monoclonal antibody therapy** targets both B and T lymphocytes, the white blood cells responsible for autoantibody production in lupus. Examples of monoclonal antibody therapy include Epratuzumab, Rituximab (Rituxan®).

**Organ Transplant Anti-Rejection Medications** – These drugs were developed to stop the body from rejecting a transplanted organ and target the white blood cells that initiate and increase inflammation and tissue damage in the joints, skin, and kidneys of people with lupus.
• Mycophenolate mofetil (CellCept®) is generally well tolerated at a dosing of 500 to 1500 mg twice daily (Isenberg & Manzi, 2008).

**Stem Cell Transplantation** - Both hematopoietic and autologous stem cell transplantation have been used in Europe and in the U.S. to treat autoimmune conditions that have not responded to any other treatments. Both procedures are currently in clinical trials for severe lupus. Stem cell transplantation is a high-risk procedure. Although it can be effective, and symptoms of lupus may become more easily controlled, some will experience a recurrence of their lupus over time.

**Test Yourself**

**One of the key blood test results that help diagnose SLE is:**

A. Positive ANA.
B. Negative serum acetone.
C. A low white blood cell count.
D. Elevated BUN and Creatinine.

**The correct answer is:** A. A positive ANA, although not unique to SLE, is one of the hallmarks used in helping differentiate SLE from other disorders.

**Which of the following agents are commonly used to treat SLE?**

A. Antiretrovirals, Acetaminophen and Warfarin
B. Corticosteroids, Anti-malarial agents, and Aspirin
C. Digoxin, Topical Steroids and Calcium Channel Blockers

The correct answer is: B. Corticosteroids, anti-malarial agents and aspirin are commonly used to treat SLE.

**Cutaneous (skin) Lupus Erythematosus**

A second form of lupus is called Cutaneous Lupus.

• As a standalone disease cutaneous lupus is limited to the skin but can also be a symptom component of systemic lupus.

• Cutaneous lupus can occur in one of three forms: chronic cutaneous (discoid) lupus erythematosus, subacute cutaneous lupus erythematosus, and acute cutaneous lupus erythematosus (malar rash).

• Approximately 10% of people who have discoid lupus will develop systemic lupus. However, it is likely that these people already have systemic lupus, with the skin rash as their main symptom.

**Cutaneous (skin) Lupus Erythematosus**

**Manifestations**

• Symptoms include rashes/lesions, hair loss, vasculitis, ulcers, photosensitivity.
  
  • Most common rash (discoid) is raised, scaly and red but not itchy.
  
  • Another common rash is a malar rash, known as the “butterfly rash.”

**Cutaneous (skin) Lupus Erythematosus**

**Diagnosis**

• Because of the great deal of variability in the way that the skin rashes may appear, cutaneous lupus can be quite difficult to diagnose. However, a **skin biopsy** may be performed to ensure correct diagnosis through microscopic visualization.
Physicians look for characteristic and unique signs, e.g., malar rash, oral ulcers, or alopecia.

**Cutaneous (skin) Lupus Erythematosus**

**Laboratory Findings**
- Lab studies used for the diagnosis of SLE may also be utilized to look for autoantibodies and to help determine if the cutaneous lupus is a standalone diagnosis or a component of SLE.
  - A positive ANA can be seen in about 5% of those with discoid lupus particularly if the individual has concurrent symptoms of joint pain.
  - About two thirds of those with subacute cutaneous lupus may have a positive ANA.
  - Acute cutaneous lupus is closely associated with SLE and virtually all of these individuals will have a positive ANA as well as possible evidence of other autoantibodies.

**Cutaneous (skin) Lupus Erythematosus**

**Treatment**
- **UV protection**
  - Minimize or avoid sun exposure.
  - SPF, 30 or greater, sunscreens with both high UVB (sunburn range) and UVA (non-sunburn range) block (e.g. mexoryl, avobenzone or physical blockers like zinc or titanium dioxide).
  - Sun protective clothing.

- **Topical therapy**
  - Topical corticosteroids are used for their anti-inflammatory properties to treat cutaneous lesions that are not widespread throughout the body surface.
  - Corticosteroid injections are used if the lesion does not respond to cream or ointment and are injected directly into the lesion.
  - Topical immunomodulators (Pimecrolimus (Elidel®) or tacrolimus (Protopic®) is a new class of drugs that has been shown to suppress the immune system activity in the skin and can be used to treat serious skin conditions without the side effects found in corticosteroids. Additional therapy may be required in conjunction with these treatment options to control the skin lesions. This can include topical steroids if the skin lesions are not too severe.

- **Oral anti-inflammatory medications** provide systemic treatment for those with moderate to severe cutaneous lupus that has shown a poor response to topical therapy or is so widespread that topical therapy may not be effective.
  - Antimalarials
  - Dapsone, retinoids, thalidomide
  - Methotrexate, azathioprine (Imuran®), mycophenolate mofetil (CellCept®), cyclosporine
  - Thalidomide (Thalomid®) has been shown to greatly improve cutaneous lupus that has not responded to other treatments.
Test Yourself
When teaching your patient with Cutaneous Lupus, you will encourage her to avoid the sun and to use sunscreen protection.

A. True
B. False

The correct answer is: A. Minimizing and/or avoiding sun exposure is one of the key methods to manage Cutaneous Lupus.

Drug-Induced Lupus Erythematosus

Drug-induced lupus is a condition caused by a reaction to certain medications. There are at least 46 different drugs in use that can cause drug-induced Lupus Erythematosus. Men and women at any age can develop the condition. The signs and symptoms of drug-induced lupus are similar to systemic lupus but tend to be milder and disappear when the drug is stopped (Isenberg & Manzi, 2008). Complete disappearance of symptoms can sometimes take months, and the disappearance of abnormal autoantibodies may take a few years.

Drug-Induced Lupus Erythematosus

Diagnosis

As with systemic lupus, most people with drug-induced lupus develop antinuclear antibodies. The ANA’s in drug-induced lupus are primarily autoantibodies that are able to react with a histone-DNA complex (the major component of the nucleus of all cells). A special laboratory test to detect certain antibodies to the histone-DNA complex is a sensitive marker for lupus-like disease brought on by many drugs. Drug-induced lupus typically comes after many months or years of continuous therapy with the causative drug.

Drug-Induced Lupus Erythematosus

Diagnosis

Medications most commonly associated with drug-induced lupus include:

- Anti-convulsant medications: phenytoin (Dilantin®)
- Anti-hypertensive medications: hydralazine (Apresoline®)
- Anti-arrhythmic medications: procainamide (Pronestyl®), quinidine (Quinaglute®)
- Drugs to treat acne: minocycline (Minocin®), isotretinoin (Accutane®)

The risk for developing lupus-like disease from any of the other 43 drugs is low or very low and with some of the drugs, only one or two cases have been reported.

Drug-Induced Lupus Erythematosus

Treatment

- Stop the offending drug.
- Support the patient until the symptoms wane.
- Some patients with moderate to severe symptoms or possible organ involvement (very rare) may require treatment similar to those with SLE until symptoms improve.

Test Yourself

Symptoms of Lupus generally disappear after the offending drug is discontinued.
Neonatal Lupus

Neonatal lupus is a condition that can occur when anti-SSA/Ro and/or anti-SSB/La antibodies (antibodies common in patients with lupus) cross the placenta in pregnancy from the mother to her developing baby. Babies born to women who are positive for anti-SSA/Ro and/or anti-SSB/La antibodies (even women who do not have lupus) are at greater risk for neonatal lupus, although this condition remains rare.

It is important to note that the baby does not have lupus and often neither does the mother. The mother may be totally healthy. The symptoms seen in the baby may be the first sign of the possible development of lupus in his or her future.

A number of symptoms are seen in infants who are born with neonatal lupus that may include a skin rash, liver problems, or low blood cell counts. These symptoms go away over time as the infant’s own immune system develops, and the mother’s antibodies are cleared from the baby’s system.

Neonatal Lupus

Complications

The most worrisome complication involves the heart. Babies can be born with a potentially life-threatening heart condition known as congenital heart block (CHB). In some cases the heart function itself may also be abnormal. It is possible to diagnose this condition in the baby while it is still in the womb (usually between 18 to 24 weeks) by seeing an abnormally slow heart beat during a fetal echocardiogram. If the heart block is not complete, it may resolve while the fetus is in the womb. If the block is complete, the condition is permanent and a baby may require a pacemaker at the time of birth or later in life.

Neonatal Lupus

Genetics

Simply because a pregnant woman has anti-SSA/Ro and/or anti-SSB/La antibodies does not automatically mean that her baby will develop CHB. In fact, the first offspring of only 2% of pregnant women who test positive for anti-SSA/Ro develop CHB; furthermore, in those women whose first babies did have CHB, there is only an 18% recurrence in future pregnancies. Also, when an anti-SSA/Ro mother has identical twins, 65% of the time one will develop CHB and the other will not or one may have complete block and the other incomplete. Genetics may be important as well as the in utero environment. All of this evidence suggests that there are other additional factors besides anti-SSA/Ro and/or anti-SSB/La that determine whether a baby will develop CHB (Sammaritano, 2011).

Neonatal Lupus

Diagnosis

At birth or within 3 months or so of birth, the infant may have a skin rash, liver problems, or low blood cell counts, but these symptoms disappear completely after ABOUT 6 months with no lasting effects.

As previously mentioned, some infants with neonatal lupus are born with congenital heart block. This block can be 2nd degree or 3rd degree heart block.
Treatment
Treatment of neonatal lupus depends on how the symptoms present. The most concerning issue is congenital heart block. The treatment depends on the severity of the block and may require placement of a pacemaker.

Currently, the best approach is frequent echocardiograms between 18 and 24 weeks gestation. Yale School of Medicine researchers have found in a new study published in the American Journal of Reproductive Immunology (January 21, 201) that an anti-malaria drug combination might be useful in helping to prevent pregnancy complications in women with lupus. Vikki M. Abrahams, associate professor in the Department of Obstetrics, Gynecology & Reproductive Sciences at Yale, and first author and Yale medical student Caroline Albert, deduced that a combination therapy that included hydroxychloroquine (taken by the mother) may be preventative and beneficial to pregnant patients. The rationale is that this drug may prevent inflammatory cells from causing scarring of the heart cells in the fetus (Albert et al., 2013).

Test Yourself
The most dangerous complication of neonatal lupus is:
A. Heart block.
B. Brain aneurysm.
C. Renal disorders.
D. Liver dysfunction.
The correct answer is: A. Heart block is a life threatening condition that may occur in an infant with Neonatal Lupus.

Lifestyle Considerations for Patients with Lupus
Lupus is a chronic disease that requires long term management. The management of the disease impacts the lifestyle of the individual with lupus.

Medication Adherence

No matter how a person’s lupus may manifest itself or the effectiveness of therapies used to control the symptoms, it is essential that healthcare providers emphasize the importance of medication adherence.

A recent report by the World Health Organization (WHO) revealed that 50% of patients with chronic disease do not take their medication as prescribed. Poor medication adherence leads to increasingly poor health outcomes for patients and has a significant negative economic impact on healthcare resources (Partners HealthCare: The Center for Connected Health, 2010).

Further studies conducted in London in recent years indicated that 50% of patients with lupus who experienced kidney failure were non-adherent to their recommended medicines (Lupus Now Magazine 2009 - Report on 2008 ACR/ARHP Scientific Meeting).

Non-adherence to medications among teens and young adults is thought to be a likely cause of most flares, and may be traced to the still developing human brain in this age group (Lupus Now Magazine 2009 - Report on 2008 ACR/ARHP Scientific Meeting). Education and support are essential so that the young person knows and understands the importance of adhering to the regimen.
Objective:
Consistent adherence with medication treatment plan.

Interventions
1. Create rapport with your patients.
2. Fully explain all medications to the patient to include possible side effects and potential problems that may result from non-adherence.
3. Provide the patient with strategies on how to integrate the medication treatment plan into his/her activities of daily living.
   a. Prepare medications in advance by using a pill box to help organize one or even two weeks' worth of medications.
   b. Use alarm clock or cell phone alarm to alert patient when it is time to take medication.
   c. Encourage family and friends to form a support system with the patient (particularly for teens and young adults), who will help reinforce medication adherence.
4. Verify adherence with medication treatment plan during follow-up appointments.
5. Remain vigilant for signs of non-adherence, as the patient may be embarrassed and not admit to being non-adherent.

Consider intravenous medications when concerns about non-adherence affect disease management.

Photosensitivity
The American College of Rheumatology loosely defines photosensitivity as "a skin rash as a result of unusual reaction to sunlight." By using this definition, photosensitivity has been identified in one-half to three-fourths of people with systemic lupus. Fluorescent and halogen lights are also a source of UV emissions significant enough to cause photosensitive reactions in patients with lupus.

In people with cutaneous lupus, photosensitivity affects 50% of those with discoid lupus and 70-90% of those with subacute cutaneous lupus.

Flares of systemic lupus disease, including joint pains and fatigue, can also be triggered by exposure to UV light.

Objective:
Minimize the effects of UV exposure.

Interventions
1. Teach the patient to minimize direct exposure to UV from sun and/or fluorescent and halogen light bulbs. (Glass does not provide complete protection from UV rays) (NIAMS, 2006).
2. Instruct the patient to use sunscreen of SPF 30 or greater and encourage the use of broad-brimmed hats and tightly-woven, loose-fitting clothing with long sleeves and long pants.
3. Teach patients about the benefits of shields that cover fluorescent bulbs to minimize indoor exposure. Recommend shields with nanometer readings of 380 to 400.
4. Assess the need for window films on the patient’s car.
5. Instruct the patient that certain drugs can increase sensitivity to the sun, including antihistamines, diuretics, non-steroidal anti-inflammatory drugs (NSAIDs) and antibiotics, including tetracycline or "sulfa" antibiotics. Extra precautions should be made if taking these medications.
6. Special emphasis on the effects of photosensitivity is necessary if the patient is taking any of these medications.
7. Special emphasis on the effects of photosensitivity should be given to people of color (e.g. African American, Hispanic, Asian, and Native American).

**Extreme Fatigue**

Extreme fatigue is one of the hallmarks of lupus.

- 80% of people with lupus experience significant fatigue
- It is unclear why fatigue occurs in so many people with lupus, but many factors appear to play a role
  - Disease activity
  - Pain
  - Medications
  - Poor physical conditioning
  - Mental health/Depression
  - Lack of social support (NIAMS, 2006)

**Objective:**

Minimize fatigue.

**Interventions**

1. Assess the patient’s general fatigue level.
2. Assess for the presence of depression, anxiety, and other stressors.
3. Conduct an assessment to determine the patient’s daily activities that contribute to fatigue.
4. Help the patient to develop an energy-conserving plan for completing daily and other activities and work. Many people with lupus need to take a daily nap. Encourage a nap earlier in the day to prevent any issues with insomnia.
5. Encourage the patient to get 8 to 10 hours of sleep at night.
6. Encourage exercise as tolerated.

**Diet and Nutrition**

There are no specific dietary restrictions for the person with lupus. However, encourage patients to maintain a nutritional, heart healthy and well balanced diet. If patients have cardiopulmonary or renal symptoms, or excessive weight gain due to steroids, a salt free diet, low protein diet, weight reducing
diet, or a combination of the three may be recommended. Referral to a dietician may be beneficial for some patients (NIAMS, 2006).

**Objective:**
Maintain weight at optimal range; minimize GI complications due to medications.

**Interventions**
1. Assess the patient’s prescription and nonprescription drug regimen and dosages for medications that may impact patient’s nutritional status.
2. Assess the patient’s usual daily dietary intake by recommending that the patient keep a food diary.
3. Educate patient about which medications to take with food.
4. Develop a dietary plan with the patient that encourages healthful eating. If the patient has nutrition-related lupus complications, refer to a dietician for counseling.
5. Encourage exercise as tolerated.
6. Record the patient’s weight at each visit.
7. Instruct the patient to weigh self at home once a week, record it, and bring to all patient visits.

**Smoking**
Cigarette smoking has been linked to the development of many autoimmune diseases, including lupus. Evidence shows that smoking complicates and accelerates the negative effects of lupus. Smoking can impair the body’s entire circulatory system, including heart and blood vessels, and can increase the risk for atherosclerosis, a condition seen more frequently in patients with lupus. If one smokes and has lupus he or she greatly increases the risk of damaging one’s heart.

Flares of cutaneous lupus have also been linked to hydrazine, an ingredient found in tobacco.

**Objective:**
Cessation of tobacco use.

**Interventions**
1. Assess smoking habits of the patient.
2. Provide support, guidance and instruction on the safest techniques of smoking cessation.
3. Provide contacts for local smoking cessation classes.

**Infection**
Patients with lupus are more likely to contract infections than completely healthy people, even when they are not taking corticosteroids. The most common infections are in the:

- Respiratory tract (colds, sore throats, sinusitis, bronchitis, and pneumonia)
- Urinary tract (bladder or kidney infection)
Skin (boils, cellulitis, and infected cuts)

If a person with lupus is taking higher dose corticosteroids every day the risk of infection is significantly higher. Lower doses, or doses taken every other day, are associated with significantly less risk of infection (LFA – Infections and Immunizations, 2010) (NIAMS, 2006).

There have been a few reports of some patients experiencing a flare in their disease following an immunization, but this association has not been proven. It is true that immunizations may be less effective in patients with lupus, particularly those on high doses of corticosteroids or other immunosuppressive agents. Immunizations with killed vaccine (e.g. influenza, tetanus, pneumococcus) are considered safe. The safety of live vaccines (e.g. polio, mumps, measles, rubella, and yellow fever) in lupus patients on high doses of corticosteroids (more than 10 mg prednisolone per day) and other immunosuppressive drugs has not been confirmed and should be avoided (Isenberg & Manzi, 2008).

Objective:
Minimize incidence of infection.
Educate the patient about immunizations.

Interventions

1. Assess the patient’s current medications, particularly those that promote susceptibility to infection such as corticosteroids and immunosuppressives.
2. Teach the patient to use good hand-washing and personal-hygiene techniques.
3. Teach the patient the signs and symptoms of infection and reinforce the importance of reporting them to the physician.
4. Encourage the patient to eat a balanced diet with adequate calories to help preserve the immune system.
5. Teach the patient to minimize exposure to crowds and people with infections or contagious illnesses.

1. Check the patient’s current immunization status.
2. Teach the patient that infections can be minimized with immunizations.
3. Encourage the patient to consult with their provider about vaccines and/or allergy shots that are being considered.

Stress

The immune system is one way the body interacts with the outside world. In people with lupus, extreme stress can often contribute to flares and increased symptoms.

Emotional stress (death of a loved one, divorce, or loss of a job) activates the immune system through the brain, where emotions are translated into nerve signals that affect hormone and immune cell function. Also, there are immune signal receptors in the brain which respond to white blood cells and alter mood and brain functions, including sleep and appetite.
Physical stress (sleep deprivation, a poor diet, lack of exercise, injury, or major surgery) also affects immune function and can lead to a flare in lupus symptoms.

**Objective:**
Identify stressors and develop coping techniques.

**Interventions**
1. Assess for presence and degree of anxiety or life stressors.
2. Encourage patient to find positive coping mechanisms for stressors.
3. Encourage the patient to accept help from others, such as counseling or a support group.
4. Discuss complementary coping activities such as biofeedback, yoga and/or acupuncture.
5. Encourage development of a consistent exercise regime.

**Depression**

Depression often goes unrecognized in those who have other medical illnesses because it presents symptoms similar to those of the underlying medical condition. Some psychiatric and medical studies state that 15% of those with a chronic illness suffer from clinical depression; others place this figure as high as 60% (LFA, 2010). Although clinical depression is certainly more common in people with chronic medical illness, (e.g., lupus) than in the general population, not every patient with a chronic illness suffers from clinical depression (LFA, 2010).

For patients with lupus, symptoms of depression that may be attributed to their lupus condition include:
- Inactivity
- Loss of energy and interest
- Insomnia
- Pain intensification
- Diminished sexual interest and/or performance

There is no single cause of depression in patients with lupus. Various factors contribute to depression in chronic illness such as lupus.

- The most common cause is the emotional drain from the continuous series of stresses and strains associated with coping with the chronic illness and medical condition.
- Other causes may be the many sacrifices and losses required by the continuous life adjustments that a patient with a chronic illness must make.
- Various medications used to treat lupus, such as corticosteroids (e.g., prednisone), may bring about depression.
- Lupus involvement of certain organs (e.g., the brain, heart, or kidneys) can lead to clinical depression (NIAMS, 2006).
- A lupus flare, both because you feel ill, and because it may seem as though you are never going...
Objective:
Recognize the signs and symptoms of depression and initiate a plan of care.

Interventions
1. Assess the patient for the signs and symptoms of depression.
2. Assess the patient’s interpersonal and social support systems.
3. Encourage the patient to express feelings.
4. Initiate a referral to a mental health counselor or psychiatrist.

Neurocognitive Dysfunction

Neurocognitive dysfunction can occur in patients with lupus. Symptoms include memory impairment, slow processing speed, and trouble finding words. It is estimated that 25 to 80% of people with systemic lupus experience neurocognitive dysfunction. While most patients have mild to moderate problems, the symptoms commonly fluctuate over time and occasionally can become severe (Utset, Fink and Doninger, 2005).

Objective:
Recognize the signs and symptoms of neurocognitive impairment and initiate a plan of care.

Interventions
1. Assess the patient for any changes in cognition, memory, and/or mental status.
2. Provide patient tips that may help ease the frustrations of cognitive impairment caused by lupus.
   - Focus attention when receiving new information. Repeat information back to the provider, ask for clarification, ask for memory tools, and/or write the information down. Verify any details.
   - Focus on one task at a time.
   - Take good care of your body; exercise, eat well, and get enough sleep.
   - Learn and use memory techniques, such as associating a person’s name with an image, or repeating the name several times in conversation.
   - Try to stay organized. One helpful hint is to use a year-long calendar notebook so that all appointments, plans, contact information, and reminders can be kept in one place.
3. Initiate a referral for neuropsychiatric evaluation if ordered by physician.

Conclusion
Lupus is a multifaceted autoimmune disease that can range from mild to life-threatening and affect any system of the body. When the disease is detected early it can often be treated successfully, and significant long-term complications may be prevented.
Health professionals play an integral role in the quality of life and positive patient outcomes for those living with lupus when they are knowledgeable about the complexities of the signs, symptoms and treatment options of lupus. In addition, educating those living with lupus is critical to helping them understand this often unpredictable disease. Education can also assist with increased treatment adherence and lifestyle changes that are essential for patients to live their best life with lupus.

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