



Health Providers'

**LUPUS
TOOLKIT**

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INTRODUCTION

The term “lupus” was first used in 1200 A.D. to describe ulcerations on the face. Lupus is a Latin word that means wolf. There are two theories as to why the disease was first described in terms of the “wolf”. The common theory is that the skin rash seemed to eat away at the skin and destroy it, resembling skin which had been bitten by a wolf¹. The rash associated with lupus, classically referred to as the “malar butterfly rash,” commonly occurs on the bridge of the nose extending above the eyes and across the top of the cheeks in a butterfly shape that is distinctive but not necessary for the diagnosis of lupus.

Lupus has been called “the great mimicker” since patients can present with an array of different symptoms that are non-specific and may be vague, making the disease difficult to diagnose. These symptoms may be spaced out over time. Although 90 percent of people with lupus will have a positive ANA (anti-nuclear antibody) test, there is no single laboratory test that can be used to definitively diagnose it. For this reason, health providers need to be knowledgeable about the signs and possible symptoms of lupus. Many individuals have symptoms for years before they are accurately diagnosed. The astute health provider who connects the dots can allay patient fears by providing a framework for understanding their disease and will help them find a path for treatment and management.

There is ample clinical information available for providers on lupus. The information contained in this toolkit is not meant to substitute for the clinical information that is obtainable with a simple modicum of effort. Rather, it is meant to be a quick reference and a supplement enabling health providers to take a more holistic approach with their patients, those already diagnosed and undiagnosed with lupus. That is to say, to take the patient’s point of view more into account and appreciate the broad spectrum of the effect of lupus on their daily lives. The intent of the toolkit is to allow the health provider to be both “healer” and “helper” to patients living with lupus.

Like art, medicine is an exacting mistress, and in the pursuit of one of the scientific branches, sometimes, too, in practice, not a portion of a man’s spirit may be left free for other distractions, but this does not often happen. On account of the intimate personal nature of his work, the medical man, perhaps more than any other man, needs that higher education of which Plato speaks, - “that education in virtue from youth upwards, which enables a man eagerly to pursue the ideal perfection.” It is not for all, nor can all attain to it, but there is comfort and help in the pursuit, even though the end is never reached.

-- Sir William Osler, *The Master-Word In Medicine*²

SOME FACTS ABOUT LUPUS³

- The Lupus Foundation estimates approximately 1.4 million Americans have some form of lupus.
- More than 16,000 Americans develop lupus each year.
- Although lupus affects men and women, 90 percent of individuals diagnosed are women.
- Systemic Lupus Erythematosus (SLE) can occur at any age, but it mostly affects women in their childbearing years.
- 80 percent of those diagnosed with lupus develop it between the ages of 15 and 45, however as many as 40 percent of cases may present after age 60.
- SLE affects 1 in 3,000 persons in the United States, striking women five to nine times more often than men.
- Lupus is two to three times more prevalent among people of color (African Americans, Hispanics, Asians, and Native Americans).
- Two out of three lupus patients report a complete or partial loss of their income because they are unable to work due to their condition.
- A survey of Lupus Foundation of America members suggests that more than half of those afflicted with lupus suffered at least four years, and saw three or more doctors before obtaining a correct diagnosis.
- Of all SLE deaths during 1979-1998, 36.4% occurred among persons aged 15-44 years old. For each year crude death rates increased with age, were greater than 5 times higher among women than men, and were greater than 3 times higher among blacks than whites. Death rates were highest among black women and increased 69.7% among those 45-64 years of age, with little difference in rates among other age groups⁴.

CLASSIFICATIONS OF LUPUS

Lupus is an autoimmune disease. It is a chronic, inflammatory, multi-system disease of unknown etiology and can be potentially fatal. There are three types of lupus:

- 1) Discoid (Cutaneous)
- 2) Systemic
- 3) Drug induced

Discord Lupus

Discoid (cutaneous) lupus is characterized by its limitation to the skin. A rash may appear on sun exposed areas, generally the face, ears, neck or scalp, back and arms. Generally it does not involve the internal organs, however, approximately 10 percent of cases may evolve into the systemic form. It is possible that those with this form may have systemic lupus with the discoid rash as their main symptom.

The discoid lesion is characteristically a circular, raised, reddish rash. The center may be scaly and lighter in color than a darker outer ring. The lesions are rarely itchy or painful and are patchy. As they heal the sharply defined skin plaques may scar and become hyperpigmented or hypopigmented. They may cause bald areas on the scalp associated with alopecia. Discoid lesions demonstrate histologic changes in the skin that are not seen in normal skin.

Systemic Lupus Erythematosus (SLE)

Systemic lupus (SLE) is more severe than discoid lupus. It can affect almost any organ system. No two individuals with SLE have exactly the same symptoms. SLE significantly affects the lungs, kidneys, heart, and brain as well as other systems. Auto-antibodies caused by lupus produce immune complexes leading to inflammation and injury to the tissues. This can be partially manifested as arthritis, nephritis, and serositis resulting in illness such as pericarditis and or pleuritis. Individuals with SLE may also have discoid lupus lesions of the skin as noted above.

Drug Induced Lupus

Certain prescribed medication can induce the symptoms of lupus. The characteristic syndrome includes pleuro-pericardial inflammation, fever, rash and arthritis. Lupus-like symptoms tend to occur after taking the medication for a period of time, usually three to six months. This is the result of a hypersensitivity reaction. Symptoms usually resolve within a few days to weeks after stopping the medication. Drugs associated with lupus are listed on the *right*.

Drugs associated with lupus⁵ include but are not limited to the following:

Atenolol (Tenormin)
Captopril (Capoten)
Carbamazepine
Chlorpromazine HCL
(Thorazine)
Clonidine HCL (Catapres)
Danazol (Danocrine)
Diclofenac (Cataflam, Voltaren)
Disopyramide (Norpace)
Ethosuximide (Zarontin)
Gold Compounds
Griseofulvin
Hydralazine (Apresoline)
Ibuprofen
Interferon alfa
Isoniazid
Labetolol HCL (Normadyne, Trandate)
Leuprolide acetate (Lupron)
Levodopa
Lithium carbonate
Lovastatin (Mevacor)
Mephenytoin (Mesantoin)
Methyldopa (Aldomet)
Methysergide maleate (Sansert)
Minoxidil (Loniten, Rogaine)
Nalidixic acid
Nitrofurantoin (Furadantin, Macrobid, Macrochantin)
Oral contraceptives
Penicillamine
Penicillin
Phenelzine sulfate (Nardil)
Phenytoin sodium (Dilantin)
Prazosin (Minipress)
Procainamide HCL (Pronestyl,)
Promethazine HCL (Phenergan)
Propylthiouracil
Psoralen
Quinidine
Spironolactone (Aldactone)
Streptomycin sulfate
Sulindac (Clinoril)
Sulfaxalazine (Azulfidine)
Tetracycline
Thioridazine HCL (Mellaril)
Timolol maleate (Betimol, Timoptic)
Tolazamide (Tolinase)
Tolmetin sodium (Tolectin)
Trimethadione (Tridione)

DIAGNOSTIC CRITERIA FOR SLE

There are eleven criteria for diagnosis of systemic lupus. These criteria were developed by the American College of Rheumatology (ACR) and then revised in 1982. The criteria are listed in the table below⁶. A person is considered to have systemic lupus erythematosus if four or more of any of the eleven criteria are present, serially or simultaneously, during any interval of observation. Note that the diagnosis of SLE is made by an individual's medical history, a constellation of non-specific signs and symptoms, and a small set of laboratory tests related to immune function.

Criterion	Definition
1. Malar rash	Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds
2. Discoid rash	Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions
3. Photosensitivity	Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation
4. Oral ulcers	Oral or nasopharyngeal ulceration, usually painless, observed by physician
5. Arthritis	Nonerosive arthritis involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion
6. Serositis	a) Pleuritis--convincing history of pleuritic pain or rubbing heard by a physician or evidence of pleural effusion
	<i>OR</i>
	b) Pericarditis--documented by ECG or rub or evidence of pericardial effusion
7. Renal disorder	a) Persistent proteinuria greater than 0.5 grams per day or greater than 3+ if quantitation not performed
	<i>OR</i>
	b) Cellular casts--may be red cell, hemoglobin, granular, tubular, or mixed
8. Neurologic disorder	a) Seizures--in the absence of offending drugs or known metabolic derangements; e.g., uremia, ketoacidosis, or electrolyte imbalance
	<i>OR</i>
	b) Psychosis--in the absence of offending drugs or known metabolic derangements, e.g., uremia, ketoacidosis, or electrolyte imbalance
9. Hematologic disorder	a) Hemolytic anemia--with reticulocytosis
	<i>OR</i>
	b) Leukopenia--less than 4,000/mm ³ total on 2 or more occasions
	<i>OR</i>
	c) Lymphopenia--less than 1,500/mm ³ on 2 or more occasions
	<i>OR</i>
	d) Thrombocytopenia--less than 100,000/mm ³ in the absence of offending drugs
10. Immunologic disorder ⁷	a) Anti-DNA: antibody to native DNA in abnormal titer
	<i>OR</i>
	b) Anti-Sm: presence of antibody to Sm nuclear antigen
	<i>OR</i>
	c) Positive finding of antiphospholipid based on 1) abnormal serum level of IgG or IgM anticardiolipin antibodies, 2) a positive test result for lupus anticoagulant using a standard method, or 3) a false-positive serologic test for syphilis known to be positive for at least 6 months and confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody adsorption test.
11. Antinuclear antibody	An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with "drug-induced lupus" syndrome.

SIGNS, SYMPTOMS & PROGNOSIS

Lupus can be manifested by a wide array of signs and symptoms that may be vague and can evolve over time. In addition, individuals with lupus report other immune diseases. One of three patients responding to a Lupus Foundation survey reported they had another autoimmune disease in addition to lupus, further complicating the ability to make the diagnosis of lupus. There is no one definitive finding, sign or symptom that makes the diagnosis.

The clinical presentation varies between different patients and in a single patient the disease activity may vary over time. A summary of the most common symptoms and clinical features of lupus and the percentage of people with lupus who have these symptoms is indicated as follows⁸:

<u>Arthralgia</u>	<u>95%</u>
<u>Fever more than 100 degrees F (38 degrees C)</u>	<u>90%</u>
<u>Swollen joints (arthritis)</u>	<u>90%</u>
<u>Prolonged or extreme fatigue</u>	<u>80%</u>
<u>Skin rashes</u>	<u>74%</u>
<u>Anemia</u>	<u>71%</u>
<u>Kidney involvement</u>	<u>50%</u>
<u>Pleurisy</u>	<u>45%</u>
<u>Malar rash</u>	<u>42%</u>
<u>Photosensitivity</u>	<u>30%</u>
<u>Hair loss</u>	<u>27%</u>
<u>Abnormal blood clotting problems</u>	<u>20%</u>
<u>Raynaud's phenomenon</u>	<u>17%</u>
<u>Seizures</u>	<u>15%</u>
<u>Mouth or nose ulcers</u>	<u>12%</u>

Additionally, there is a risk of first and second trimester fetal loss and premature birth and patients are often allergic to a variety of antibiotics, especially sulfonamides.

Health providers must also be aware of the neuro-psychiatric features of this disease. These may present as neurosis, psychosis, adjustment reactions, and organic brain dysfunction such as learning, memory, and other cognitive dysfunction. Other manifestations of lupus by organ system include:

Gastrointestinal:

Abdominal pain
Nausea
Vomiting

Neuropsychiatric:

Cranial neuropathies
Organic brain syndrome
Peripheral neuropathies
Psychosis
Transverse myelitis

Hematologic:

Leukopenia,
Thrombocytopenia
Lymph node enlargement

Pulmonary:

Pulmonary hypertension

Renal:

Casts Nephrotic syndrome
Hematuria Proteinuria

Prognosis

Life-threatening complications are most common in the first five years of the manifestations of lupus. The prognosis varies widely, depending on the organs involved and the intensity of the inflammatory reaction. Years ago, most individuals with systemic lupus survived less than five years. More recently with the ability for earlier diagnosis and changes in treatment modalities the long-term prognosis is good. The ten year survival rate is now greater than 95 percent.

Morbidity is mostly caused by renal and neurologic manifestations of the disease. These are associated with a worse prognosis compared to some other manifestations of lupus. The main cause of death is infection due to the immunosuppressive side effects of medications used to treat this chronic disease. Overall, prognosis is usually worse in men and children than in women. Symptoms that begin after the age of sixty tend to be more benign and flares are rare after menopause.

LABORATORY TESTING FOR LUPUS

The lupus erythematosus cell test (LE cell prep) was the first laboratory test created to test for lupus. It is somewhat sensitive, but not specific for lupus despite the name of the test. Persons with other rheumatic conditions, such as scleroderma, or other medical conditions such as hepatitis, and those taking certain medications may also have a positive test.

The Antinuclear Antibody (ANA) test is the most commonly performed test when a diagnosis of lupus is suspected. The clinician must be aware, however that the ANA test is not specific for the diagnosis of lupus. The ANA is over 90% percent positive in patients with active lupus, including drug induced lupus. However, it is also positive in patients with various connective tissue diseases and rheumatoid arthritis, and in 5-10% of patients without any systemic rheumatic disease. A negative ANA test practically excludes a diagnosis of lupus. A positive ANA, in combination with clinical symptoms and other laboratory findings associated with SLE noted above, indicates a high probability of lupus. ANA testing can help confirm the diagnosis of SLE in this context. The false positive rate is 3% overall but increases with age.

Anti-Smith (Anti-Sm) antibody is highly specific for systemic lupus but is only found in 30% of individuals with lupus. Antibodies to double stranded DNA (dsDNA), also highly specific for lupus, is found in 60 to 80 percent of patients with active SLE.

Of note, a false positive VDRL or RPR, tests for syphilis, should prompt clinicians to consider lupus as a possible cause. Patients with a false positive VDRL/RPR should be queried about symptoms and have their medical history reviewed to determine if they may have signs or symptoms of lupus.

It is prudent to consider patient referral to a rheumatologist for further patient evaluation and interpretation of test results for any individual patient. A rheumatologist can assist with early recognition of the patient with suspected lupus.

LUPUS TREATMENT APPROACHES

MEDICATIONS

There is currently no known cure for lupus. However, early recognition and the appropriate medical management of lupus can significantly help to ameliorate the effects of the disease and the patients ability to cope with its chronic nature, including its tendency for recurrent exacerbations (lupus flares) and remissions. In most cases, lupus can be managed in an outpatient setting.

Treatment of lupus is mainly focused on medication. Therapy is determined by the severity of the disease. Current treatment relies on the use of the following classes of medication, which can be used alone or in combination with other agents.

- Salicylates
- Non-steroidal anti-inflammatory drugs (NSAIDs)
- Antimalarials (Hydroxychloroquine)
- Corticosteroids (Topical, intralesional injections, oral, and intravenous)
- Cytotoxic agents (Azathioprine, Cyclophosphamide, Methotrexate)

Patients should be monitored for the effectiveness of these medications and health providers should be well versed with their side effects. Medication side effects are sometimes indistinguishable from symptoms of the disease.

In addition to medical treatment, patients need to be encouraged to fully engage in lifestyle modification that can help with their symptoms and to participate in the self-management of their disease. (See below).

LIFESTYLE MODIFICATION AND PATIENT SELF-MANAGEMENT

Sunlight Exposure

Exposure to the sun must be avoided or minimized as much as possible. Ultraviolet rays (UVA, especially UVB, and UVC) of the sun can lead to lupus flares, causing skin rashes and can worsen systemic manifestations of lupus. In fact, photosensitivity can be a symptom of lupus.

You should teach patients to minimize direct exposure to the sun and to florescent and halogen light bulbs. Florescent light bulbs mainly emit UVC light. Exposure to photocopiers, which emit UVA light, should be avoided as well. An acrylic shield can be fitted over florescent lights to eliminate UVB emission. Individuals should contact their employers to try to arrange this for their work areas.

Sunscreen with a SPF of fifteen or higher should be used and protective clothing should be worn to minimize exposure to the body. The neck, the temples and ears are often missed by patients when using sunscreens. These areas also tend to be the areas where photosensitive lupus patients frequently experience problems. Use of sunscreens help to prevent rashes. Certain topical medications can increase the risk of photosensitivity as well as some hair dyes and over the counter skin creams.

Advise patients to wear broad rimmed hats, long sleeves and pants to help minimize exposure. Thickness of the clothing is more important than color. Providers and patients should be aware that cloudy days are not protective. Care should also be taken with exposure to light through the glass of car windows.

Advise patients to take a multi-vitamin containing vitamin D everyday, since they will be limiting their skin exposure to daylight. Outdoor activities should be minimized between the peak hours of 10:00 am and 4:00 pm⁹.

Rest and the Assessment of Fatigue

Fatigue is a particularly common disease manifestation and one area in which patients can take some control. To minimize fatigue, advise lupus patients to:

- Be mindful of their energy levels during the day and note times of the day when energy levels tend to be higher. Slow down or stop before they get too tired.
- Plan activities during the times of the day when they tend to have higher energy levels
- Not to do everything at once. If possible, they should plan ahead and spread activities over the day and week and they should set priorities for how they will spend their time and energy.
- Get plenty of rest and adequate sleep. When possible, they should plan to take a nap during the day.
- Try to avoid stress, take time to relax and learn about relaxation techniques.
- Ask and let their friends and family help them with whatever they may have to do.

Assessment of fatigue starts with obtaining a careful history to characterize the individual's fatigue. Health providers should try to identify all the factors that may be contributing to its development. An evaluation of fatigue should include any identifiable pattern of fatigue, including onset, duration, and intensity as well as aggravating and alleviating factors. The assessment should also include the following:

- Type and degree of disease and of treatment-related symptoms and/or side effects.
- Current medications.
- Sleep and/or rest patterns
- Dietary intake and any appetite or weight changes.
- Effects of fatigue on activities of daily living and lifestyle.
- Psychiatric evaluation, including evaluation for depression.
- Job performance, financial resources, or other stressors
- Other contributing factors e.g. anemia or shortness of breath

Keep in mind that fatigue may also be a sign of depression. Depression should be regularly assessed and treated appropriately.

There are a number of instruments to assess the severity of fatigue. One such instrument is the Fatigue Severity Scale (FSS) included as one of the tools in this kit. The FSS scores a series of subjective questions to help differentiate physical fatigue from fatigue associated with depression. It can be used for individuals with lupus and other chronic diseases that have fatigue as a major component.

Also included is an Energy Calendar which you can provide to your patients to help them begin tracking their fatigue levels over the course of the day and to determine peak times

when energy levels are high.

Diet & Nutrition

There are no specific dietary guidelines for lupus. However, the general guidelines for good nutrition hold for lupus as well as other medical conditions. A diet comprised of plenty of fruits, vegetables, whole grains and low fat proteins which minimize animal fat intake is generally advisable. Lupus patients may receive steroids for treatment that can put them at risk for osteoporosis. Therefore advising patients to make sure to have a good intake of dietary calcium and possible oral supplementation over the counter or by prescription is wise. Vitamins that include supplements of vitamin D can be useful as well, since patients may not get adequate light exposure to generate vitamin D through the skin due to minimized exposure to the sun to prevent photosensitivity and lupus flares.

Antioxidants are thought to be helpful to reduce the risks of inflammation that can lead to arthritis symptoms and may be beneficial. Because lupus patients, particularly those with SLE, may be at risk for developing kidney disease and subsequent hypertension, it is important that low salt intake be a focus for those with this complication.

PREGNANCY AND CONTRACEPTION

Since the overwhelming majority of people with lupus are women and most are diagnosed during their childbearing years, issues related to lupus and pregnancy will arise. Lupus does not decrease the likelihood that a woman will become pregnant but it can increase the risks of pregnancy and lead to complications.

The pregnancy of any woman with lupus is automatically considered an obstetrically high-risk pregnancy, though many pregnancies are completely normal and result in normal babies. However, in the event a woman with lupus becomes pregnant it will be important that she have an obstetrician that is experienced with managing high-risk pregnancies and that she is monitored closely.

About one-third of women with lupus have improvement of their lupus symptoms during pregnancy. One-third experience worsening of their lupus, with an increase in flares, and another one-third experience no change in their lupus during pregnancy. There is no way to predict who will have problems, but those that have been in remission for more than six months before becoming pregnant seem to have a lower risk of flares and their babies do better.

- More than 50 percent of all systemic lupus pregnancies are completely normal.
- About 25 percent of pregnant women with lupus deliver a baby prematurely.
- Fetal loss, due to spontaneous abortion or miscarriage and stillbirth is increased in women with lupus.
- About 25 percent of cases are complicated by problems with hypertension and/or pre-eclampsia during pregnancy
- There may be an increased risk of the baby developing neonatal lupus syndrome (see below)
- Many of the medications used to treat lupus are safe but some can be hazardous to the baby

- About 33 percent of women with lupus have lupus anticoagulant, anti-phospholipid or anti-cardiolipin antibodies that can cause blood clots in the mother and to the placenta causing slowed fetal growth or miscarriage

Mothers with lupus may pass on anti-Ro and anti-La autoantibodies. One-fourth to one-third of those with lupus has these autoantibodies. When these are present in the mother, newborns can develop the neonatal lupus syndrome.

Neonatal lupus syndrome consists of skin rashes with sensitivity to light, thrombocytopenia, elevated liver transaminases and hepatosplenomegaly, with or without congenital heart block. The autoantibodies accumulate in the baby's heart where they can cause thickening and scarring of the heart muscles. On rare occasions this can lead to congestive heart failure before birth and may require pacemaker placement. The skin rashes are relatively minor in the constellation of neonatal lupus and will usually disappear without treatment by the time the baby is one year old.

Risks and benefits of pregnancy should be discussed. For all the reasons above it is recommended that women with lupus receive family planning counseling. The timing of pregnancy, if desired, should be discussed. Health providers or other appropriate health professionals should also discuss and provide the various kinds of contraception available as needed. When pregnancy is desired, it is best for the pregnancy to be planned and for the appropriate support to be provided. In the event of an unplanned pregnancy the women will need to weigh the risk and benefits of continuing the pregnancy.

Primary care providers need to be knowledgeable about the risks of pregnancy for women with lupus so that they can be supportive and provide information to their patients and or the appropriate referrals.

PSYCHOSOCIAL ASPECTS

Whenever anyone has an illness there is always a psychosocial aspect to the illness. This is even more important when one has a chronic disease, such as lupus. Due to the very nature of chronic illness, people often have to deal with the psychosocial effects of their disease as it relates to family, relationships, work, and their own sense of self. The effects of lupus on one's quality of life, lifestyle and appearance due to the occurrence of rashes, scarring, and hair loss certainly affect one's self image. This and dealing with how others, who may not fully understand lupus, perceive individuals with lupus all contribute to the psychosocial response of those living with it. It is important that the clinician pay close attention to this aspect of lupus so that they can assist those that have lupus to cope with the non-medical effects of the disease.

Many people with lupus wind up losing their jobs, their spouses or partners, and friends because of their condition. Emotions of anger, frustration, fear, feelings of isolation and others are bound to manifest themselves in some way. The need to be vigilant in watching for depression cannot be overstated. Providing avenues for mental, physical, and spiritual support are what individuals living with lupus want.

Exercise, physical/occupational therapy, peer support groups, mental/behavioral health, and discussions related to the availability of spiritual support may all come into play in helping

individuals with lupus feel better and to take a positive approach to their chronic illness.

When a group of women who have lupus were asked what people with lupus can expect during a focus group for preparation of a lupus patient information kit the following responses were given:

Being misunderstood by family and everyone	Major dental problems
Bone lockage	Memory loss
Chest pain	Menopause flare-ups
Confusion	Mental fog
Dark circles under their eyes	Mood swings
Depression	Mouth blisters and sores
Dry mouth	Muscle tightening
Employers to misunderstand	Nausea
Feelings of isolation	Nervous system disorder
Hair loss	Nose bleeds
Inability to balance a check book	Pain
Insomnia	Pale skin
Joint contractions	Shortness of breath
Joint swelling	Sores in the nose
Loss of appetite	Strange rashes
Loss of energy	Vision problems
	Weight gain

Certainly, these individuals and others diagnosed and undiagnosed with lupus have a lot to cope with. Counseling should be considered and provided early on. These women also expressed a desire for their providers to be sensitive to all their needs. A health provider that focuses on how the patient is doing in areas other than the clinical condition and that listens can go a long way.

Establishing a rapport and regular communication between the primary care provider and the treating rheumatologist can be helpful in the approach to managing individuals with lupus. Providers should work with their patients on self-management techniques, as well, and try to engage them in taking control of their illness and partnering to assist with its management. A family meeting, if agreed to by the patient, to help the family understand lupus better and how they might be helpful may be useful. This can be arranged with a use of an informed and supportive therapist or social worker.

TALKING WITH YOUR PATIENTS ABOUT LUPUS & WHAT LUPUS PATIENTS WANT YOU TO KNOW

During February and March of 2003, two focus groups were held with people living with lupus at different stages and with different lengths of time since their initial formal diagnosis. They were asked to provide information that they thought would be important for providers to know from their perspective. Their responses were based upon their experiences and own perceived needs. The focus group questions and the responses from individuals living with lupus are summarized in bullet format below.

What Do You Wish You Could See?

- More support groups
- More visibility regarding lupus e.g. billboards
- Physicians who understand and are sympathetic
- More information, so patients know what lupus is, that it is not contagious, etc. and a clearing up of misinformation:
 - o You can be ill and look great
 - o Lupus is not contagious
 - o Information about some other illnesses related to lupus (e.g. heart, liver, pancreatic, Raynaud's disease, hair, skin, etc.)
 - o Women can have other autoimmune diseases in addition to lupus
 - o At what point should doctors ask if their patient has lupus?
 - o Is there a standard test for lupus, why can't there be a standard test for people, especially women of color, so can diagnose lupus quicker?
- Information for family members, especially for children and partners (support groups and counseling for each group – children, spouses, patients)
- More information about what lupus is
- Conveniently located support groups
- Answering questions e.g. does it run in families?
- Getting early diagnosis and treatment

What were some of the challenges you faced before getting diagnosed with lupus?

(Items listed are what physicians actually told individuals was causing their symptoms and/ or the diagnosis given for presenting symptoms of what was actually lupus.)

- Doctor thinking I was crazy, that I needed psychiatrist
- Thought it was aging, carpal tunnel and dry eyes
- I was told it was a virus when I had a loss of feeling in my leg
- Sunburn lesions, AIDS-related skin issues, people thought I had AIDS
- My doctor thought I was a hypochondriac
- Eye problems
- Red spots on legs
- Ulcers
- Sores
- Seizures and epilepsy
- Kidney failure
- Infertility
- Hair loss
- A cold, nose bleeds,
- Arthritis
- Lumps in my neck, lymph nodes

- Uterine issues
- A collagen vascular disease
- Mumps
- Thyroid problems
- Anemia
- Psychiatric, psychosomatic
- Depression, migraines
- Menopausal systems
- Low grade fevers

What led to your diagnosis?

- Butterfly rash
- Dry eyes
- Death of a sibling from lupus
- Oral health – dentists can play a role in diagnosing lupus

What would have made your diagnosis easier?

- Doctors more knowledgeable regarding lupus
- Doctors more sensitive to women’s concerns – cultural competency training, awareness that a small number of men have it
- Include lupus diagnostics in check-ups, standardization
- Not making culturally-based assumptions regarding lifestyle (i.e. young black woman has syphilis when she really has lupus)
- Making emergency care easier. Triage nurses need to understand the seriousness of lupus. A connection to doctors – choosing where you are sent – access to medical records in an emergency
- Medical alert bracelets

What makes for a good relationship with doctors/providers?

- Knowledge and validation
- A good primary doctor, that spends one-on-one time and has a good rapport
- Primary doctor with a relevant specialty
- Communication across doctors
- Everything is documented, nothing slips through the cracks, doing diligence
- Primary doctor needs to coordinate communication with other doctors and care and understand that’s their role
- Sensitivity to patients’ personal being and not making assumptions regarding lifestyle, sexuality, etc.
- Understanding regarding bruising
- Understanding regarding medication induced illnesses
- More specific training regarding lupus (doctors, staff, triage nurses)
- More understanding regarding drugs, interactions, treatment modalities
- Understanding of the need for emotional support
- Understanding how to help people who are in denial

Is there anything else you would like to see related to support services?

- More information regarding symptoms
- More healthcare provider training to recognize symptoms
- Training for dentists
- Access to “The Ride” program so we can make appointments

- Convenient groups, possibly in community health centers
- More cognitive, behavioral therapist availability
- Check more for heart disease and make people aware of the link to lupus
- Lupus “look good, feel better” programs
- Wellness programs for people with chronic pain
- Work on handicap accessible alterations, materials, aids, etc.
- Legal services, e.g. to help obtain SSI
- Financial planners to inform how to manage money when you have a major illness
- More information about access to home health aids
- Social worker increased understanding of lupus and people of color
- Relationship based counseling (marriage, family, etc)

HELPING YOUR PATIENTS WITH SELF-MANAGEMENT

The approach to managing chronic disease is changing. The medical environment is moving from the paternalistic approach of the health provider as the all knowing expert, to the health provider engaging and collaborating with patients as decision-making partners in their care. The goal of engagement is to help the individual understand their health condition and behaviors and to assist them in developing strategies to live with their health condition as fully and as productively as possible.

Self-management patient education teaches problem-solving skills and engenders self-efficacy. This extends from the Chronic Care Model developed by Ed Wagner, MD, MPH, Director of the MacColl Institute for Healthcare Innovation and colleagues of Group Health Cooperative of Puget Sound. The Chronic Care Model outlines six areas of change, including Patient Self-Management, to improve chronic care management¹⁰.

Patients with chronic conditions already self-manage their illness every day by making decisions about whether they will be proactive, reactive or take no action at all in the approach and response to their illness. Health providers can help individuals living with lupus to shape a positive response to their disease by encouraging them to participate in self-management of their disease and thereby affirm the importance of the patients having a role in their overall care.

The patient self-management concept means that providers:

- Work together with patients to define problems, set priorities, establish goals, create treatment plans, and solve problems.
- Use self-management tools that are based on evidence of effectiveness
- Set and document self-management goals collaboratively with patients
- Receive training on how to help patients with self-management goals
- Consider use of patient group visits to support self-management
- Tap community resources to assist achievement of self-management goals.

There are four steps in providing self-management support:

- (1) collaborative goal setting between patient and health provider,
- (2) identification of barriers and challenges to achieving goals,
- (3) personalized problem-solving and
- (4) follow-up support.

Goal setting can be elaborated by exploring or asking about any problems, issues, or concerns your patient may have because they have lupus. Then clarify feelings that may be expressed or meanings associated with the issues identified to ensure you and your patient understands what has been said. Once problems have been clearly identified have the patient think about and list possible solutions. Guide them in prioritizing the problems. Work with your patient to then develop goals and set out a plan to achieve the goals set and to address any anticipated barriers or challenges and then have them commit to an action plan.

Ask questions such as, What are you going to do? How? When? How will you know you have succeeded? How confident are you that you can achieve the goal(s) that have been set? Once these have been answered review the plan and have the patient write them down, have your patient contract with you to review the plan again and the outcomes at the next visit or on a

time line that both of you agree with. Then set a follow-up plan for the other problems, barriers, or challenges that were initially identified or that may come up later

Remember the critical aspects of improving chronic care through self-management are 1) emphasis on the patient role, 2) a standardized assessment, 3) effective interventions, and 4) care-planning and problem solving¹¹. This involves a standardized approach to each individual in assessing their knowledge of their condition, skill, and level of confidence to manage symptoms. A Self-Management Support Tool is included as part of the toolkit to assist you and your patients in this process.

Health providers can be both healer and helper. It is important for providers to assist lupus patients and others living with a chronic illness to live as fully and productively as possible. It is hoped that this resource toolkit will help you to facilitate this process with your patients.

HEALTH PROVIDER TOOLS

TAKE THE LUPUS TEST

- Have you ever had achy, painful and/or swollen joints for more than three months?
- Do your fingers and/or toes become pale, numb or uncomfortable in the cold?
- Have you ever had any sores in your mouth for more than two weeks?
- Have you ever been told that you have a low blood count(s) – anemia, low white cello count or a low platelet count?
- Have you ever had a prominent redness or color change on your face in the shape of a butterfly across the bridge of your nose and cheeks?
- Have you ever had an unexplained fever over 100 degrees for more than a few days?
- Have you ever had sensitivity to the sun where your skin “breaks out” after being in the sun, but it’s not a sunburn?
- Have you ever had chest pain with breathing for more than a few days (called pleurisy)?
- Have you ever been told you have protein in your urine?
- Have you ever experienced persistent extreme fatigue/exhaustion and weakness for days or even weeks at a time even after 6–8 hours of restful sleep?
- Have you ever had a seizure or a convulsion?

If you answer “yes” to at least three of these questions, there is a possibility you may have a condition called Lupus. It is suggested you:

- Discuss these symptoms with your doctor.
- Take a list of the medications you are using or bring your medications with you to your visit.
- Get tested for lupus

CHECKLIST FOR LUPUS SIGNS AND SYMPTOMS FOR HEALTH PROVIDERS

American Rheumatism Association (ARA) 11 Criteria

Symptom or Criteria	Description
Malar Rash	Red rash over cheeks and the bridge of the nose
Discoid Rash	Red, scaly rash on the face, scalp, ears, arms, or chest
Photosensitivity	Unusual reaction to the sun
Oral ulcers	Small sores on the moist lining of the nose or mouth
Arthritis	Pain in the joints of the hands, arms, shoulders, feet, legs, hips or jaws which may move from joint to joint and be accompanied by heat, redness and swelling
Serositis	Inflammation of the lining of the heart. Documented by ECG or a pericardial rub heard by auscultation of the heart
Renal/Kidney	Excessive protein and/or cellular casts in the urine
Neurologic	Seizures and/or psychosis
Blood	Decrease in the number of red and white blood cells or platelets
Immunologic	Positive Anti-DNA test
Antinuclear Antibody	Positive Antinuclear Antibody (ANA) test
Other:	False-positive syphilis test (not an ARA 11 criteria)

FATIGUE SEVERITY SCALE (FSS)

There are a number of instruments to assess the severity of fatigue. One such instrument is the Fatigue Severity Scale (FSS). The FSS is designed to differentiate fatigue from clinical depression. It consists of a number of statements that individuals subjectively rate on a scale of 1 to 7 depending upon how much they feel the statement applies to them over a period of time. The subject is asked to read each statement and circle a number from 1 to 7, depending on how appropriate they feel the statement applies to them over the preceding week.

A low score for any one particular statement indicates that the patient does not think the statement applies to them and a high score indicates a level of agreement with the statement.

During the past week, I have found that:	SCORE (<i>Circle a number</i>)						
1. My motivation is lower when I am fatigued	1	2	3	4	5	6	7
2. Exercise brings on my fatigue	1	2	3	4	5	6	7
3. I am easily fatigued	1	2	3	4	5	6	7
4. Fatigue interferes with my physical functioning	1	2	3	4	5	6	7
5. Fatigue causes frequent problems for me	1	2	3	4	5	6	7
6. My fatigue prevents sustained physical functioning	1	2	3	4	5	6	7
7. Fatigue interferes with carrying out certain duties and responsibilities	1	2	3	4	5	6	7
8. Fatigue is among my three most disabling symptoms	1	2	3	4	5	6	7
9. Fatigue interferes with my work, family, or social life	1	2	3	4	5	6	7

The scoring is done by calculating the average response to all the questions (adding up all the answers and dividing by 9).

People with depression alone score about 4.5

People with fatigue related SLE average about 6.5*

*Also relates to fatigue of Multiple Sclerosis (MS), or Chronic Fatigue Immune Dysfunction Syndrome (CFIDS)

Daily/Weekly Energy Calendars

How energetic do you feel today?

This calendar will help you keep track of your energy level over the next week.

1. Fill in the circle that reflects how you feel. Do this for a couple of weeks to track your energy patterns.
2. At the end of the week, draw a line connecting the circles to see how much you're improving over time.
3. Take this calendar to your next doctor's visit and discuss how you've been feeling.

Week: _____

DAY	Monday			Tuesday			Wednesday			Thursday			Friday			Saturday			Sunday			
	AM	N	PM																			
Normal	<input type="radio"/>																					
Tired	<input type="radio"/>																					
Very Tired	<input type="radio"/>																					

AM = Morning

N = Afternoon

PM = Evening

You can make copies of these calendars to use as often as you like.

SELF-MANAGEMENT SUPPORT TOOL

Date: _____

Patient Name: _____ DOB: _____

Medical Record Number: _____

Health Provider Name: _____

1. The healthy change I want to make is:(be specific: What, When, How Where, How Often)

2. My goal for the next month is:

3. The steps I will take to achieve my goal are:

4. The things that could make it difficult to achieve my goal include:

5. My plan for overcoming these difficulties includes:

6. Support/resources I will need to achieve my goal include:

7. My confidence that I can achieve my goal:
(On a scale of 1 –10, with 1 being not confident at all, 10 being extremely confident) _____

Review date: _____ with _____

Patient Signature: _____

APPENDIX

CHRONIC CARE MODEL COMPONENTS

Self-Management

- Providers and patients work together to define problems, set priorities, establish goals, create treatment plans, and solve problems
- Use self-management tools that are based on evidence of effectiveness
- Set and document self-management goals collaboratively with patients
- Train providers on how to help patients with self-management goals
- Use group visits to support self-management
- Tap community resources to achieve self-management goals

Decision Support

- Embed evidence-based guidelines in the care delivery system
- Establish linkages with specialists to assure that primary care providers have access to expert support
- Provide skill-oriented interactive training programs for all staff in support of the chronic illness improvement
- Educate patients about guidelines

Clinical Information System

- Establish a registry of patients
- Use registry to provide feedback to generate reminders and care-planning tools for individual patients
- Use registry to provide feedback to care team and leaders to guide course of treatment, anticipate problems and track progress

Delivery System Design

- Use registry to review care and plan visits
- Assign roles, duties, and tasks for planned visits to a multidisciplinary care team.
- Use planned visits in individual and group settings, plan in advance based on patient's needs and self-management goals
- Make follow-up a part of standard procedure

Organization of Health Care

- Align the effort to improve care with a quality management system
- Translate the effort to improve care into clear goals reflected by the health center's policies, procedures, business plan and financial planning
- Leaders give team members and personnel the resources and support they need to pursue improvement

Community

- The health center forms alliances and partnerships with state programs, local agencies, etc. to develop support programs and policies
- Link to community resources
- Raise community awareness through networking, outreach and education
- Provide list of community resources to patients, families and staff

PROVIDER & PATIENT RESOURCES

RESOURCES FOR HEALTH PROVIDERS

Lupus: A Patient Care Guide for Nurses and Other Health Professionals

A guidebook with detailed review and information on lupus, includes patient information sheets covering a range of topics.

Web: www.niams.nih.gov/hi/topics/lupus/lupusguide/outline.htm

For copies, contact:

NIAMS Information Clearinghouse

1 AMS Circle

Bethesda, MD 20892-3675

Toll-free: 877-22 NIAMS (226-4267)

TTY: 301-565-2966

Fax: 301-718-6366

*Free copies (limited number while available) can be obtained from the

Massachusetts League of Community Health Centers
Department of Clinical Health Affairs

100 Boylston Street, Suite 700

Boston, MA 02116

Phone: 617-426-2225

Fax: 617-426-9864

Web: www.massleague.org

RESOURCES FOR PATIENTS

EDUCATIONAL MATERIALS

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)

Patient education booklets NIAMS Information Clearing House:

The Many Shades of Lupus: Information for Multicultural Communities

NIH Publication No. 01-4958

Web: www.niams.nih.gov/hi/topics/lupus/shades/index.htm

Handout on Health: Systemic Lupus Erythematosus

NIH Publication No. 97-4178

Web: www.niams.nih.gov/hi/topics/lupus/slehandout/index.htm

For copies, contact:

NIAMS Information Clearinghouse

1 AMS Circle

Bethesda, MD 20892-3675

Toll-free: 877-22 NIAMS (226-4267)

TTY: 301-565-2966

Fax: 301-718-6366

SPANISH LANGUAGE MATERIALS (EN ESPAÑOL)

¿Tengo Lupus? / Do I Have Lupus?

(*A free, bilingual booklet in Spanish and English)
Published by the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS). For copies of the booklet contact:

NIAMS Information Clearinghouse

1 AMS Circle

Bethesda, MD 20892-3675

Toll-free: 877-22 NIAMS (226-4267)

TTY: 301-565-2966

Fax: 301-718-6366

Web: www.niams.nih.gov/hi/topics/lupus/tengo/index.htm

Asociación Lupus Argentina

An Internet website with many informative and educational articles. Website is in Spanish. Titles include - What is Lupus? (Qué es el Lupus), Lupus and the Sun (El Lupus Y el Sol), Living with Lupus (Conviviendo con Lupus).

Web: www.drwebsa.com.ar/alua

SUPPORT GROUPS

Women of Courage (WOC) Support Group

A grassroots volunteer effort and support group for women of color living with lupus. Contact: Beverly Gibson, Interim President.

111 Locust Street, No. 3C3
Woburn, MA 01801
Phone: 617-417-0836

Lupus Foundation of America, Massachusetts Chapter

Monthly support groups and programs. Check calendar at www.lupusmass.org for current locations and programs or call the toll free number.

40 Speen Street, Suite 205
Framingham, MA 01701
Toll Free: 877-NOLUPUS (877-665-8787)
Web: www.lupusmass.org

Minorities with Disabilities Advocacy Center/ Vivienne S. Thomson Independent Living Center

Now located at the ABCD Independent Living Center.

Peer support and peer counseling, skills training for independent living, social services system advocacy and self and co-advocacy skills training. Also provides information and referrals to rehabilitation agencies, other disability resource agencies and other services for people with disabilities.

Phone: 617-288-9431

Lupus Foundation of Greater Washington

Support information, education, chat and discussion groups for people with lupus.

Web: www.lupusgw.org

Arthritis & Lupus Support Group, New England Baptist Hospital

Contact: Janice Sullivan, 617-754-5610.
125 Parker Hill Avenue
Boston, MA 02120
Phone: 617-754-5410

ORGANIZATIONS/ AGENCIES

Massachusetts Department of Public Health
Bureau of Environmental Health Assessment
Phone: 617-624-5757

Lupus Foundation of America, Inc.

2000 L Street, N.W., Suite 710
Washington, DC 20036
Phone: 202-349-1155
Toll-free: 800-558-0121 (information request line)
Fax: 202-349-1156
E-mail: lupusinfo@lupus.org
Web: www.lupus.org

Lupus Foundation of America, Massachusetts Chapter

40 Speen Street, Suite 205
Framingham, MA 01701
Toll-free: 877-NO LUPUS (665-8787)
Web: www.lupusmass.org

Lupus Foundation of Greater Washington

Phone: 703-644-0058
Email: info@lupusgw.org

END NOTES

- ¹ Lupus Trust of New Zealand, A History of Systemic Lupus Erythematosus
- ² Shigeaki Hinohara, M and Hisae Niki, M.A. OSLER'S "A WAY OF LIFE" & OTHER ADDRESSES, WITH COMMENTARY & ANNOTATIONS, p.255
- ³ Lupus Foundation of America, Inc.
- ⁴ MMWR Report May 23, 2002, 51(17), 371-4
- ⁵ Greenberg, Brent MD, Michlaska, Margaret MD, Systemic Lupus Erythematosus: Measures to keep this unpredictable disease under control. Postgraduate Medicine 1999; 106 (6): 213-23
- ⁶ Arthritis and Rheumatism, Vol. 25, No. 11 (November 1982), Pg. 1271-7. Criteria items 10 (a) is deleted and 10(d) revised in 1997. Table corrected from the original to indicate 1997 revisions. See next footnote for revision reference.
- ⁷ Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus [letter]. Arthritis Rheum 1997; 40:1725
- ⁸ Introduction to Lupus, LUPUS Foundation of America, Inc. pamphlet Revised 2/00
- ⁹ Dr. Thomas Millard, Research Registrar in Dermatology, Department of Photobiology St. John's Institute of Dermatology, St. Thomas' Hospital, London. (Internet source)
- ¹⁰ Other areas of the chronic care model are outlined in the Appendix.
- ¹¹ Improving Chronic Illness Care, Supporting Patient Self-Management: Evidence and Techniques, PowerPoint

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Sadly, Bobbie Drake-Saucer, President of the Women of Courage, whose energy in the fight for better understanding and support for persons living with lupus was never ending did not live to see the completion of this project. She died suddenly before this project was completed. She will be missed, but her spirit and passion live on.

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Gisele Thornhill, MD, MPH

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