



# **Clinical Practice Guidelines**

# **SYSTEMIC LUPUS ERYTHEMATOSUS**

**Program Update: 11/30/2014**

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# **Accordant Clinical Practice Guidelines: Systemic Lupus Erythematosus**

## **INTRODUCTION**

The AccordantCare™ program works with health plans to assess, monitor, and support those with certain complex, chronic conditions. The focus of the program is to improve health outcomes and prevent or limit disease-related complications. AccordantCare offers unique services at no additional charge to the patient, putting them in a strong position to adhere to their treatment plan.

There are several ways AccordantCare augments physicians' efforts. Through regular telephone contact, AccordantCare nurses:

- Keep patients informed about the disease process
- Coach patients in self-motivation and self-care skills
- Encourage patients to alert their physician when new symptoms arise
- Direct patients to resources that help pay for medication, transportation, home modifications, etc.
- Ensure preventive and screening measures are accomplished
- Provide emotional support to patients and caregivers
- Screen for depression
- Find local support groups

We invite physicians to make use of the services offered by AccordantCare and to suggest ways we can further patients' treatment goals. To offer feedback, get more information, ask questions or voice concerns call toll-free 1-800-948-2497 to speak with a program representative from 8 a.m. to 9 p.m., Monday through Thursday, and from 8 a.m. to 5 p.m. on Friday, Eastern Time. Messages left after hours will be returned the next business day.

### **Intent of Guidelines**

The purpose of this Clinical Practice Guideline is to describe current patterns of practice where there is no fully established national guideline for diagnosis and management. It is not meant to dictate care of patients. Decisions about care are made by the physician and the patient based on the individual needs of that patient.

A patient's health plan may or may not pay for the all medicines, tests, equipment, or services mentioned in this document. Benefits should be checked with the individual's health plan to assure payment.

## **DISEASE OVERVIEW**

Systemic lupus erythematosus (SLE) is a complex, chronic, relapsing, inflammatory autoimmune disease that often manifests in the skin, joints, blood, and kidneys.<sup>1</sup> Immunologic abnormalities result in a buildup of immune complexes in tissues leading to inflammation, tissue injury, and pain.

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About 20% of SLE patients also have discoid skin lesions.<sup>2</sup> Discoid lupus involves the skin and usually appears as a rash on the face, neck, and scalp. The lesions are disc-shaped and patchy, and they may scar. Discoid lupus may also occur in the absence of systemic disease, but Accordant does not enroll these patients.

Use of the following drugs is known to potentially cause drug-induced lupus:

- Chlorpromazine
- Hydralazine
- Isoniazid
- Methyldopa
- Procainamide
- Tumor-necrosis factor (TNF) antagonists<sup>3</sup>

The symptoms of drug-induced lupus are similar to those of SLE and usually diminish when the offending agent is discontinued.<sup>2</sup>

### Prevalence and Survival

Estimates vary widely, but about 300,000 to 1.5 million people suffer from SLE in the United States.<sup>4</sup> Nine out of ten lupus patients are women.<sup>5</sup> Compared with Caucasians, lupus is two to three times more common among African-Americans, Hispanics, Asians, and Native Americans.<sup>5</sup>

Five-year survival rates have increased from 50% of patients to 88% to 96%. Ten-year survival rates range from 95% for younger patients to 71% for older patients,<sup>6</sup>

### Economic Costs

A substantial economic burden is associated with SLE in terms of the utilization of healthcare resources, as well as with the losses of productivity due to patients' diminished capacity to work. A 2008 study found that the average annual direct healthcare cost of patients with lupus was \$12,643.<sup>7</sup> The mean annual productivity costs (lost hours of productive work) for participants of employment age (between the ages of 18 and 65) was \$8,659. Thus, the mean annual total costs associated with lupus (combining direct costs and productivity costs for subjects of employment age) is estimated to be \$20,000.<sup>5</sup>

## DIAGNOSIS OF DISEASE

For SLE patients to be efficiently treated and to experience early remission—which is more likely to last over time and thus allow for safe treatment tapering—early diagnosis is essential. The proper use of classification criteria, the search for useful lupus biomarkers, and the increased education of general practitioners in identifying lupus signs among their patients—all can help achieve the goal of earlier diagnosis and treatment.<sup>8</sup>

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No single test or clinical finding can positively diagnose SLE. Early screening may include a complete blood count (CBC), liver and kidney screening panels, lab tests for antinuclear antibodies, a test for syphilis (which may be falsely positive in the presence of antiphospholipid antibodies), blood chemistries, urinalysis, and erythrocyte sedimentation rate (ESR).

The table below contains a set of classification criteria published by the American College of Rheumatology (ACR). Though not specifically designed for diagnosis, it is often used as such. According to the ACR group, if "...4 of the 11 criteria are documented, the diagnosis of SLE can be made with about 95% specificity and 85% sensitivity."<sup>9</sup>

**1997 Update of the 1982 American College of Rheumatology Classification Criteria for SLE**

Item	Definition
Malar rash	Fixed erythema, flat or raised, over the malar eminences, sparing the nasolabial folds
Discoid rash	Erythematous raised patches with adherent keratotic scaling and follicular plugging: atrophic scarring may occur in older lesion
Photosensitivity	Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation
Oral ulcers	Oral or nasopharyngeal ulceration, usually painless, observed by a physician
Nonerosive arthritis	Involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion
Pleuritis or pericarditis	<ul style="list-style-type: none"> <li>a. Pleuritis—convincing history of pleuritic pain or rub heard by a physician or evidence of pleural effusion</li> <li>OR</li> <li>b. Pericarditis—documented by electrocardiogram or rub or evidence of pericardial effusion</li> </ul>
Renal disorder	<ul style="list-style-type: none"> <li>a. Persistent proteinuria &gt;0.5 g per day or &gt;3+ if quantitation not performed</li> <li>OR</li> <li>b. Cellular casts—may be red cell, hemoglobin, granular, tubular, or mixed</li> </ul>

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Item	Definition
Neurologic disorder	a. Seizures—in the absence of offending drugs or known metabolic derangement, e.g., uremia, ketoacidosis, or electrolyte imbalance  OR  b. Psychosis—in the absence of offending drugs or known metabolic derangement, e.g., uremia, ketoacidosis, or electrolyte imbalance
Hematologic disorder	a. Hemolytic anemia with reticulocytosis OR b. Leukopenia— $<4,000/\text{mm}^3$ on $\geq 2$ occasions OR c. Lymphopenia— $<1,500/\text{mm}^3$ on $\geq 2$ occasions OR d. Thrombocytopenia— $<100,000/\text{mm}^3$ in the absence of offending drugs
Immunologic disorder	a. Anti-dsDNA: antibody to native DNA in abnormal titer OR b. Anti-Sm: presence of antibody to Sm nuclear antigen OR c. Positive finding of antiphospholipid antibodies based on: 1) an 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 test result for at least 6 months and confirmed by <i>Treponema pallidum</i> immobilization or fluorescent treponemal antibody absorption test
Positive antinuclear antibody	An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time in the absence of drugs

Adapted from Guidelines for referral and management of systemic lupus erythematosus in adults. American College of Rheumatology Ad Hoc Committee on Systemic Lupus Erythematosus Guidelines. *Arthritis & Rheumatism*. 1999;42(9):1785-1796.

**APPROACH TO MANAGEMENT OF SLE**

**Complications of SLE**

Nephritis remains one of the most devastating complications of lupus. The 2012 *ACR Guidelines for Screening, Treatment, and Management of Lupus Nephritis* recommend<sup>10</sup> that all patients with clinical evidence of previously untreated active lupus

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nephritis (LN) undergo renal biopsy (unless strongly contraindicated) so that glomerular disease can be classified by ISN/RPS criteria and evaluated. End-stage renal disease may require dialysis or kidney transplant.<sup>1</sup>

It is important to practice aggressive screening and management of the traditional CVD risk factors (including smoking, high blood pressure, etc.).<sup>11</sup>

Infections in people with SLE often result in hospitalizations.<sup>12</sup> Bacterial, viral, and fungal infections remain an important cause of mortality and morbidity in patients with SLE, with the respiratory and urinary tracts as well as the skin and soft tissues being the most common sites of infection.<sup>13</sup>

### Treating SLE

In 2013 an international task force developed treat-to-target recommendations for managing patients with SLE. This task force defined four overarching principles for care<sup>14</sup>:

1. Management of SLE should be based on shared decisions between informed patients and their physician(s).
2. The goals of treatment should be to
  - ensure long-term survival;
  - prevent organ damage; and
  - optimize health-related quality-of-life by controlling disease activity and minimizing comorbidities and drug toxicity.
3. Management may have to be multidisciplinary.
4. Patients need regular long-term monitoring and review and/or adjustment of therapy.

### General Treatment Recommendations

The treat-to-target task force agreed on 11 recommendations for treating patients with SLE<sup>14</sup> (see Table 1).

**Table 1. Treat-to-Target Recommendations for SLE<sup>14</sup>**

1. The goal of treatment should be the remission of systemic symptoms and organ manifestations. If remission cannot be reached, the goal should be the lowest possible disease activity, measured by a validated lupus activity index and/or by organ-specific markers.
2. Prevention of flares (especially severe flares) is a realistic therapeutic goal.
3. Treatment in clinically asymptomatic patients should not be escalated based solely on stable or persistent serological activity.
4. Since damage predicts subsequent damage and death, prevention of damage

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accrual should be a major therapeutic goal.
5. Factors negatively influencing health-related quality of life, such as fatigue, pain, and depression, should be targeted.
6. Early recognition and treatment of renal involvement in lupus patients is strongly recommended.
7. For LN, following induction therapy, at least three years of immunosuppressive maintenance treatment is recommended to optimize outcomes.
8. Lupus maintenance treatment should aim for the lowest glucocorticoid dosage needed to control disease. If possible, glucocorticoids should be withdrawn completely after appropriate tapering.
9. Prevention and treatment of morbidity associated with antiphospholipid syndrome (APS) should be a therapeutic goal in SLE. Treatment recommendations are the same as those for primary APS.
10. Regardless of the use of other treatments, serious consideration should be given to the use of antimalarials.
11. Relevant adjunctive therapies to any immunomodulation (antihypertensives, lipid-lowering agents, antihyperglycemics, antiplatelet/anticoagulants, immunizations, and bone-protecting agents) should be considered to control comorbidity in SLE patients (.

### **Medicines for Treating SLE**

Mild SLE is generally treated with some combination of oral and/or topical corticosteroids, NSAIDs, antimalarials, and sunscreen.<sup>1</sup> Nonacetylated salicylates are a good choice for mild symptoms of arthritis, arthralgia, and myalgia because they inhibit prostaglandin synthesis less, cause fewer GI bleeds, and do not impair platelet function.<sup>15</sup> In the absence of contraindications, hydroxychloroquine should be used in every patient.<sup>11</sup>

Severe SLE, on the other hand, requires more aggressive treatments which may include high-dose corticosteroids, immunosuppressive or cytotoxic agents (alone or in combination with corticosteroids), intravenous immunoglobulin (IVIG), and plasmapheresis.<sup>1</sup>

Photosensitive patients need a sunscreen with a sun protection factor (SPF) of 15 or higher that protects against both ultraviolet (UV)-A and UV-B rays.<sup>16</sup> As photosensitivity may develop after the diagnosis, all lupus patients should be encouraged to avoid excessive sun exposure.<sup>17</sup>



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NSAIDs and salicylates

Corticosteroids

- prednisone (e.g., Deltasone<sup>®</sup>)
- methylprednisolone (e.g., Medrol<sup>®</sup>)

Antimalarials

- hydroxychloroquine (Plaquenil<sup>®</sup>)

Immunomodulating and cytotoxic drugs

- mycophenolate mofetil (CellCept<sup>®</sup>)
- cyclophosphamide (Cytoxan<sup>®</sup>)
- azathioprine (e.g., Imuran<sup>®</sup>)
- methotrexate (e.g., Rheumatrex<sup>®</sup>)
- belimumab (Benlysta<sup>®</sup>)
- chlorambucil (Leukeran<sup>®</sup>)
- cyclosporine (e.g., Gengraf<sup>®</sup>)

IVIg therapy has been used to treat refractory cases of SLE, particularly those unresponsive to corticosteroids.<sup>18</sup>

Plasmapheresis is a temporary remedy that is not often employed in the United States.<sup>15</sup>

### PREVENTION AND MANAGEMENT OF COMPLICATIONS

Accordant helps patients prevent and manage complications by teaching early warning signs, encouraging adherence to treatment plans, offering supportive care, and recommending physician contact where needed. The goals and cooperative interventions below do not represent a comprehensive list of complications but reflect some of the more common clinical situations specific to SLE. General health topics (e.g., age-appropriate cancer screening) are beyond the scope of this document.

**Goal:** Improve self-management skills

**Cooperative interventions** include teaching patients the importance of:

- personal motivation building;
- prevention-focused, behavioral self-management skills development;
- confidence and communication;
- adhering to treatment plan; and
- knowledge development.

**Goal:** Prevent thromboembolic events or fetal loss from antiphospholipid syndrome (APS).

**Cooperative interventions** include teaching patients to:

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- recognize the risk of recurrent miscarriage<sup>16</sup>, premature birth, and high blood pressure in women of childbearing age<sup>19</sup>;
- recognize the symptoms of stroke<sup>20</sup>, deep vein thrombosis, and pulmonary embolism and seek immediate medical attention in case of such an event<sup>21</sup>;
- understand the importance of compliance with anticoagulation therapy for stroke prevention<sup>22</sup>;
- control hypertension<sup>23</sup>;
- control hyperlipidemia<sup>23</sup>;
- avoid oral contraceptives<sup>21</sup>;
- avoid estrogen replacement therapy<sup>21</sup>;
- avoid smoking<sup>21</sup>; and
- avoid prolonged immobilization<sup>21</sup>.

**Goal:** Minimize pain and disability associated with arthritis/arthralgia

**Cooperative interventions** include teaching patients to<sup>16</sup>:

- understand that not all joint pains are due to a lupus flare and to look specifically for inflammation, swelling, and/or warmth in a painful joint as a possible sign for a lupus flare;
- apply heat or cold as appropriate;
- use warm showers or baths to lessen stiffness and pain;
- avoid strenuous activity and not place weight on an inflamed joint;
- talk to a doctor about a referral for physical therapy for ROM exercises;
- discuss occupational therapy with a doctor if ADL are impaired;
- understand the importance of a regular exercise plan that can be carried out during periods of remission; and
- use crutches, a walker, or a cane as needed.

**Goal:** Prevent cardiovascular disease by emphasizing treatment adherence and control of modifiable risk factors

**Cooperative interventions** include teaching patients to:

- be aware of their increased risk of atherosclerotic disease<sup>24</sup>;
- recognize the signs and symptoms of a heart attack or stroke and agree to seek immediate medical attention<sup>20</sup>;
- understand the importance of controlling hyperlipidemia and hypertension; and
- understand the importance of stopping smoking.

**Goal:** Minimize the impact of central nervous system lupus by helping patients understand the need to adhere to their treatment plan and gain control over modifiable risk factors for stroke.

**Cooperative interventions** include teaching patients to:

- understand that CNS lupus can manifest as vasculitis, cognitive dysfunction, lupus headache, stroke, peripheral nervous system lupus and the symptoms associated with each<sup>25</sup>;
- understand that treatment options depend on the source of the symptoms;
- control hyperlipidemia and hypertension;

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- understand the importance of reporting signs and symptoms of cardiac problems to a doctor;
- include significant others in patient care as appropriate to<sup>16</sup>:
  - discuss ways of strengthening support networks
  - help family identify potential coping skills, environmental supports, and community services for dealing with chronically ill people
  - encourage patient and family members to consider professional counseling
  - assist patient and family in identifying and removing potentially dangerous items in the environment
  - involve family members in planning of patient's care and safety measures
  - assess patient's ability to safely administer own medications

**Goal:** Minimize the severity of dermatological manifestations

**Cooperative interventions** include teaching patients to<sup>16</sup>:

- minimize direct exposure to ultraviolet (UV) rays from sun and from fluorescent and halogen light bulbs;
- use a sunscreen with an SPF of 15 or greater and wear protective clothing, such as hats and long-sleeve shirts;
- understand that treatment depends on the type and severity of lesions;
- seek a referral to a dermatologist for severe and/or disfiguring lesions;
- take medications as ordered that may help to alleviate discomfort and itching;
- avoid topical applications, such as hair dyes and skin creams and the use of certain drugs that increase sun sensitivity;
- use a hypoallergenic concealing makeup; and
- try a soft-food diet, lip balms, and warm saline rinses for mouth lesions.

**Goal:** Decrease disability related to fatigue and distinguish between fatigue and infection or anemia.

**Cooperative interventions** include teaching patients to<sup>16</sup>:

- understand the medical causes of fatigue;
- realize that fatigue is the result of illness and treatment;
- recognize the warning signs of fatigue;
- know the difference between fatigue and depression;
- recognize symptoms of infection;
- identify activities that cause fatigue; and
- identify activity changes that may decrease fatigue:
  - get eight hours or more of sleep each night
  - plan for rest periods throughout day
  - work exercise into daily activities.
  - divide large tasks into several small steps
  - schedule important activities at times of less fatigue.

**Goal:** Decrease complications related to fever and acute inflammatory episodes

**Cooperative interventions** include teaching patients to:

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- understand that high fevers can occur during a lupus flare, but they may also signal an infection or a drug reaction<sup>16</sup>;
- take temperature at least once a day for a week to determine the normal, baseline temperature<sup>16</sup>;
- agree to call their doctor if a fever is elevated above an agreed-upon baseline<sup>16</sup>;
- monitor temperature during a lupus flare<sup>16</sup>;
- look for signs and symptoms of infection, particularly urinary and respiratory infections, and report to their doctor<sup>16</sup>; and
- increase fluid intake to prevent dehydration.<sup>26</sup>

**Goal:** Minimize flares and accompanying complications through medication adherence and other self-management actions

**Cooperative interventions** include teaching patients to prevent flares by:<sup>16</sup>

- complying with medical therapy;
- limiting exposure to sun and other sources of UV rays;
- checking with a doctor before receiving immunization;
- getting adequate rest;
- exercising regularly;
- maintaining a balanced diet;
- checking with a doctor or nurse before taking OTC medications;
- testing OTC preparations used on skin or scalp;
- checking with a doctor before discontinuing medication(s);
- planning pregnancy; and
- delaying elective surgery until lupus is controlled.

**Goal:** Prevent complications from hematological manifestations (e.g., anemia, thrombocytopenia)

**Cooperative interventions** include teaching patients to<sup>16</sup>:

- recognize the risk for bleeding (e.g., low platelet count, anemia, thrombocytopenia) and to report episodes to a doctor;
- take iron preparation medications if prescribed;
- recognize the symptoms of infection and report to a doctor;
- reduce the risk of infection by using good hand-washing and personal-hygiene techniques and minimizing exposure to people with infections or contagious illnesses;
- discuss risks and benefits of immunizations with a doctor and avoid preparations made with live viruses;
- develop a plan to conserve energy; and
- follow the basics of good nutrition.

**Goal:** Minimize the incidence and complications of infection and help patients distinguish between an infection and a flare

**Cooperative interventions** include teaching patients to<sup>27</sup>:

- understand they are susceptible to infection because the disease process weakens their immune system;

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- know that their risk of infection increases if taking corticosteroids or immunosuppressants;
- learn the signs and symptoms of infection and report them to a doctor;
- understand that the flu shot is effective and not associated with flares in lupus patients taking immunosuppressants<sup>28</sup>, but that the nasal spray (live) vaccine must be avoided;
- discuss the human papillomavirus (HPV) vaccine with their doctor if 26 years of age or younger<sup>29,30</sup>;
- discuss the pneumonia vaccines with their doctor. This vaccination is indicated in all potentially immunosuppressed patients, including those with SLE<sup>31</sup>;
- discuss the Tdap (tetanus, diphtheria, and pertussis) vaccine: the Tdap vaccine is usually given at age 11 or 12 or as soon as possible thereafter;<sup>32</sup>
- discuss the zoster vaccine with their doctor if 50 years of age or older<sup>33</sup>;
- use good hand-washing and personal-hygiene techniques;
- minimize exposure to crowds and people with infections or contagious illnesses;
- talk to their doctor about taking antibiotics before dental procedures or surgery; and
- consult their doctors before considering allergy shots.

**Goals:** Detect renal disease early, preserve renal function, prevent disease flares, minimize proteinuria<sup>34</sup>, avoid harm from treatment, and improve quality of life/survival.<sup>35</sup>

**Cooperative interventions** include teaching patients to:

- understand that MMF is as effective as an induction agent for LN as cyclophosphamide and has fewer adverse events<sup>36</sup>;
- adhere to medicine schedules for LN and know that nonadherence is often a cause of treatment failure<sup>36</sup>;
- monitor blood pressure at home<sup>10</sup>;
- understand the importance of complying with medications to control high blood pressure;
- watch for signs and symptoms of renal complications (e.g., swelling, “foamy” urine) and report them promptly to a doctor;
- follow up with a nephrologist if necessary;
- report dyspnea;
- weigh daily to monitor fluid retention;
- consult a dietitian about dietary changes that accommodate alterations in renal status;
- understand that pregnancy in the context of active lupus nephritis is dangerous and that reliable birth control should be used;
- understand the risks associated with treatment of lupus nephritis (e.g., birth defects with mycophenolate mofetil, infection with immunosuppressants);
- watch for the signs and symptoms of urinary tract infection and report them to a doctor;
- learn that corticosteroid therapy may mask the usual symptoms of infection;
- take antibiotics for urinary tract infection as prescribed;

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- understand that children with lupus are more likely to have renal involvement than adults and that nephritis is often a presenting feature and<sup>35</sup>;
- know that the diagnosis, management, and monitoring of pediatric lupus nephritis are largely based on extrapolation from evidence obtained in adults.<sup>35,37</sup>

**Goal:** Maintain healthy vision

**Cooperative interventions** include teaching patients to:

- recognize the symptoms of eye problems (e.g., blurry or cloudy vision, sensitivity to light) and to report any changes in vision to their doctor immediately;
- understand the risk of retinal injury associated with hydroxychloroquine; and
- obtain a baseline ophthalmologic exam preceding hydroxychloroquine therapy and monitoring exams as directed by their physician.

**Goal:** Avoid hospitalization, surgery, and disability associated with osteoporotic fractures

**Cooperative interventions** include teaching patients to<sup>38</sup>:

- know the importance of risk assessment;
- know the importance of BMD testing<sup>39</sup>;
- recognize the importance of calcium and vitamin D supplements;
- discuss treatment options to prevent and/or repair bone loss with a physician, especially if taking corticosteroids;
- learn fall prevention strategies;
- improve home safety;
- participate in exercise that improves balance, agility, strength, and posture. Encourage members to get physician approval of new exercise programs;
- understand the importance of smoking cessation; and
- learn the importance of reducing alcohol consumption.

**Goal:** Minimize risk of pregnancy-induced complications and fetal loss

**Cooperative interventions** include teaching patients to<sup>16</sup>:

- understand the importance of pre-pregnancy counseling, especially in the context of lupus nephritis, antiphospholipid antibodies, or pulmonary hypertension<sup>40</sup>;
- understand that pregnancy in the context of pulmonary hypertension<sup>41</sup> or active lupus nephritis<sup>19</sup> endangers the life of the mother and should be avoided;
- talk to their doctor about the risks associated with oral estrogen contraceptives (the pill) for women of child bearing age;
- try to conceive during a time when the disease is under control;
- find an obstetrician experienced in high-risk pregnancies;
- plan delivery at a hospital that can handle high-risk patients and premature newborns;
- keep all doctors' appointments;
- monitor blood pressure;
- recognize the symptoms of miscarriage, pregnancy-induced hypertension, pre-eclampsia, and eclampsia;

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- understand that pregnancy in the context of active lupus nephritis and/or antiphospholipid antibodies increases the risk of hypertension and premature birth<sup>19</sup>;
- rest, exercise, and eat a well-balanced diet;
- abstain from cigarettes and alcohol; and
- take medications as prescribed.

**Goal:** Identify signs and symptoms of depression and promote appropriate treatment

**Cooperative interventions** include teaching patients to<sup>16</sup>:

- understand the need for systematic neuropsychiatric assessment to provide prompt diagnosis and treatment<sup>42</sup>;
- understand the symptoms of depression (e.g., loss of interest in favorite activities, insomnia);
- express feelings and needs;
- accept that feelings of denial and anger are normal;
- explore sources of potential support and community resources;
- consider possible ways of concealing skin lesions and hair loss;
- discuss interpersonal and social conflicts that arise;
- accept help from others, such as counseling or a support group;
- learn the major signs and symptoms of depression;
- discuss a possible referral to a mental health counselor or psychiatrist; and
- utilize support groups, educational and self-management programs offered by the Lupus Foundation of America and the Arthritis Foundation.

**Goal:** Minimize the impact of pulmonary hypertension

**Cooperative interventions** include teaching patients to:

- understand the symptoms of pulmonary hypertension but also that it may be asymptomatic<sup>43,44</sup>;
- understand the treatment options and to comply with their treatment regimen;
- understand the side effects and precautions associated with medications<sup>45</sup>;
- understand that pregnancy in the context of pulmonary hypertension is life-threatening and should be avoided using effective methods of birth control. Talk to their doctor about the risks associated with oral estrogen contraceptives (the pill); and
- understand the importance of flu and pneumonia vaccines.

**Goal:** Minimize attacks of Raynaud's phenomenon and prevent infection and/or deformities that may result over time.

Teach patients to<sup>16</sup>

- understand that there is a risk of ulceration or deformity of digits if no measures are taken to prevent attacks or attacks are not treated immediately;
- recognize an attack and take immediate action (e.g., go indoors, run warm [not hot] water over hands and feet);
- report attacks to their doctor;
- keep hands and feet warm;

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- keep other body parts warm (e.g., wear hats and layered, loose clothing);
- avoid too much air conditioning;
- avoid smoking;
- use insulated drinking glasses; and
- wear gloves when handling frozen or refrigerated foods, or whenever outside in cold weather.

### **PATIENT FOLLOW-UP**

Frequency of appointments is based on individual patient needs and will vary according to disease duration, level of disease activity, and medication issues. Many patients require regularly scheduled appointments that includes evaluation of<sup>46</sup>:

- Skin rash
- Joints
- Drug side effects (e.g., osteoporosis, liver and kidney toxicities, retinopathy)
- Thrombosis or miscarriage in the context of antiphospholipid antibodies
- Cardiovascular risk factors (monitor for development of vascular disease)

### **PATIENT EDUCATION**

The Accordant Health Communities website at <https://www.accordant.com> offers resources for patients with lupus.

Other approved and informative websites for patient education include the following:

Lupus Foundation of America  
<http://www.lupus.org>

American College of Rheumatology at:  
<http://www.rheumatology.org/practice/clinical/patients/>



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