RHEUMATOLOGY NURSE
PRACTICE

Inside this Issue

ISSUE 9 | VOLUME 3

- How is a lupus flare defined?
- How common are disease flares among patients diagnosed with systemic lupus erythematosus (SLE)?
- What triggers have been identified that may contribute to SLE flares?
- What evidence supports the use of various treatment options to help manage SLE flares?

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PRESENTATION, ASSESSMENT, & TREATMENT OF

LUPUS FLARES

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TARGET AUDIENCE

This activity has been designed to meet the educational needs of nurses and nurse practitioners. Other healthcare providers may also participate.

ACTIVITY DESCRIPTION

In this issue of *Rheumatology Nurse Practice*, we will examine how SLE patients experience a flare, the varying symptoms of flares, and the options for their treatment. We will also take a look at the questions that nurses and nurse practitioners can ask patients with SLE at every visit—both related and unrelated to disease flares—to help monitor and manage potentially disruptive changes in disease activity.

LEARNING OBJECTIVES

After participating in the activity, learners should be better able to:

- Define a lupus flare based upon expert recommendations
- Differentiate an SLE flare from ongoing disease activity
- Offer guidance to patients with SLE on risk factors of disease flares that they may want to avoid
- Develop a checklist of questions to ask patients with SLE at every routine visit

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Systemic lupus erythematosus (SLE) is an immune-mediated inflammatory disease that affects multiple systems in the body and has a highly variable course for many patients. After an acute or subacute presentation of the disease, some patients may experience a long period of quiescence with few symptoms, while others may have chronic or persistent disease activity.[1]

Often, SLE disease activity is characterized by an unpredictable pattern of remission and flares.[2,3] The rate and severity of flares during a year-long course of treatment are recognized as important predictors of long-term disease outcome.[4]

Flares and the Patient Experience

Lupus flares are common, with mild, moderate, or severe flares occurring in approximately 25% to 35% of patients with SLE treated with antimalarial and/or immunosuppressive therapy.[5] During a flare, patients often report feeling malaise and profound fatigue; many also report increasing alopecia, fever, and joint pain. These illness-related symptoms interrupt a patient's lifestyle, impact their quality of life, and often necessitate changes in therapy.[6]

Self-reported fatigue is a prominent symptom in patients with SLE, present in up to 90% of all individuals. While its severity can fluctuate from day to

Drug Names Included Within This Issue

GENERIC	BRAND
Hydroxychloroquine (HCQ)	Plaquenil
Mycophenolate mofetil (MMF)	CellCept
Belimumab	Benlysta
Azathioprine	Imuran
Cyclophosphamide	Cytoxan, Neosar
Furosemide	Lasix
Labetalol hydrochloride	Normodyne, Trandate
Lisinopril	Prinivil, Zestril
Epoetin alfa	Epogen, Procrit

day, it is typically worse during a flare.[7] The range of a patient's fatigue may be difficult for providers to quantify as it often occurs on a broad continuum, with both emotional and cognitive impacts that can have profound effects on a patient's activities of daily living, social and family activities, and employment.[7-9]

One study retrospectively examined 101 female patients with SLE to determine their perception of recent flare symptoms and triggers. The most common reported symptoms during disease flares were joint and muscle pain (73.3%), fatigue (65.4%), butterfly or malar rash (29.7%), and headaches (23.8%). Although stress and anxiety are typically categorized as flare triggers, in this study, patients often self-reported them as symptoms of flares, along with depression and mood disturbance.[2]

Other patient-reported flare symptoms in this study were high or frequent fevers (14.9%), joint swelling (14.9%), abdomen discomfort/gastrointestinal problems (13.9%), brain fog/cognitive clouding (11.9%), and ultraviolet (UV) sensitivity (10.9%). Symptoms reported by <10% of patients included shortness of breath, increase in Raynaud's episodes, infection, chest pain, skin dryness, skin changes, dizzy spells, ulcers in the mouth or nose, vision changes, sleep disturbances, and hair loss.[2]

Another study that looked at the experience of 120 SLE patients (90% female) with a median follow-up of almost 6 years found that 71% had one flare over that time period, 12 had two flares, and 5 had more than two flares. The most frequently recorded general patient symptoms that resulted in therapy changes (Figure 1) were musculoskeletal (67%), general (35%), mucocutaneous (32%), and hematologic (28%).[1]

Defining an SLE Flare

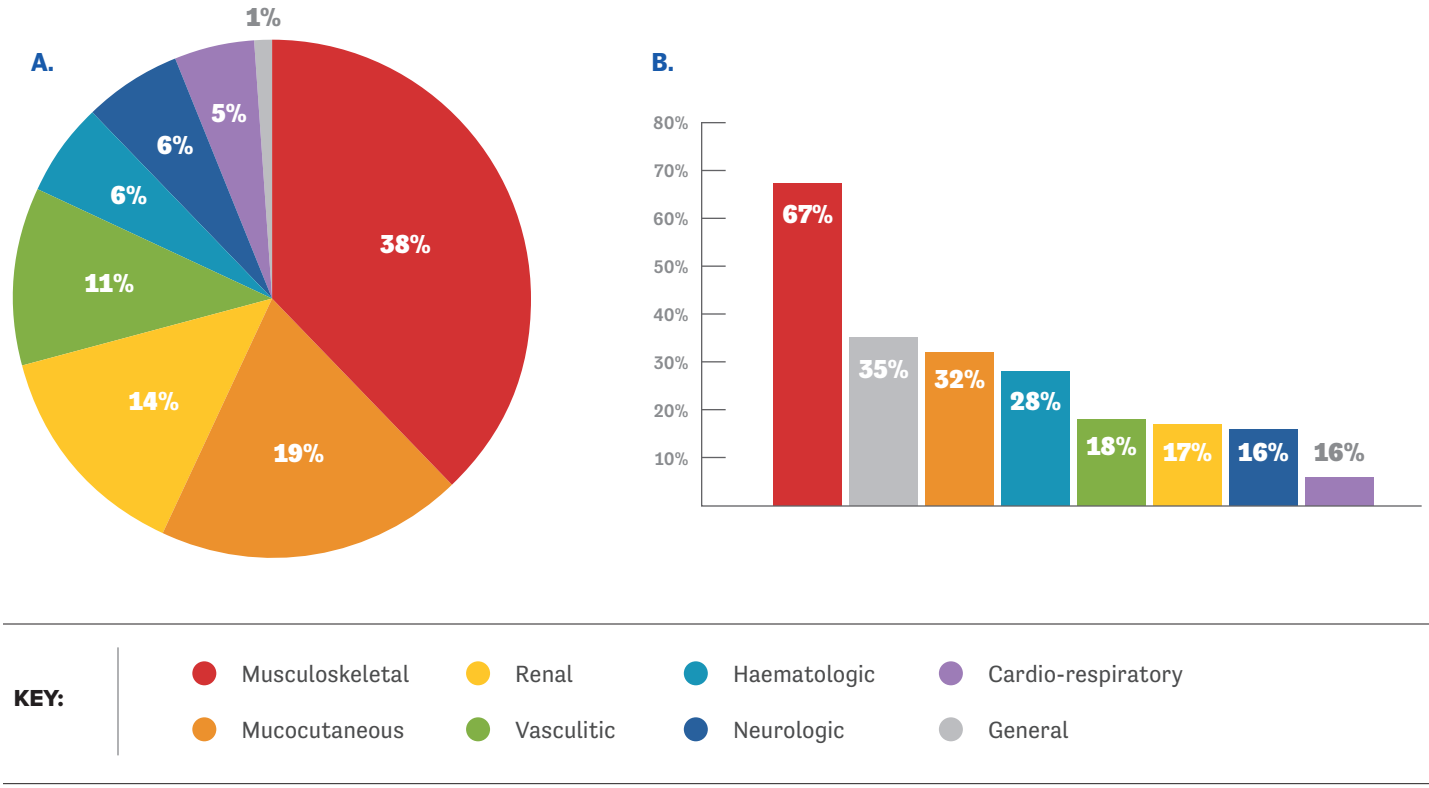
In 2011, the Lupus Foundation of America convened an international working group to provide a formal definition of an SLE flare (Table 1). Using a series of web surveys and face-to-face meetings, this group eventually defined an SLE flare as "A measurable increase in disease activity in one or more organ systems involving new or worse clinical signs and symptoms and/or laboratory measures. It must be considered clinically significant by the assessor, and usually there would be at least consideration of a change or an increase in treatment." The rate and severity of these flares during a year-long course of treatment are considered important predictors of disease outcome.[4]

Additional findings from the working group included the following:[4]

- A flare begins at the first sign or laboratory measurements of an increase in disease activity and ends when a patient's disease activity has returned to its pre-flare level, regardless of whether the patient's disease is fully quiescent.
- Flares can affect multiple organs in the body and can vary widely in severity and duration.
- Flares can occur at any level of disease activity and can be mild, moderate, or severe.
- A mild flare may affect a patient's quality of life but does not necessarily lead to changes in medication, although changes or increases in treatment should be considered. A moderate flare needs treatment and management. A severe flare implies a threat to an organ or life and may be different for different organ systems.
- A flare is different than ongoing clinical disease in that a flare is a change—a temporary increase of disease activity or a worsening in symptoms that were not present previously.
- Ongoing clinical disease is a continuum of worsening and is not considered a flare.

Pediatric rheumatologists have also moved toward developing criteria for global flares in juvenile SLE (jSLE). These criteria are similar to the international definition for adult SLE flares. The pediatric group defines a jSLE flare as "A measurable worsening of jSLE disease activity in at least one organ system, involving new or worse

Figure 1 *Clinical Manifestations of SLE Flares*[1]



A. *Frequency of clinical manifestations determining change in treatment at the time of flare;*

B. *Clinical manifestations recorded at the time of flare, irrespectively of whether they were the main reason for therapy changes*

signs of disease that may be accompanied by new or worse SLE symptoms. Depending on the severity of the flare, more intensive therapy may be required."[10]

Flare Triggers

A disease flare can sometimes occur with no apparent cause, perhaps due to progressive inherent buildup of autoimmunity.[11] However, several environmental triggers of SLE flares have been identified, including exposure to cigarette smoke and various chemicals such as mercury and silica dust.[12]

UV light exposure is a known trigger of disease flares. The ability of UV light to induce a flare is apparently dose dependent, with intermediate- and high-dose UVB exposure promoting proinflammatory apoptosis and necrosis, which is accompanied by the release of autoantigens and proinflammatory cytokines that may then trigger an inflammatory response.[11,12]

Patients with SLE are often advised to avoid UV light, although this can subsequently result in a vitamin D deficiency, which has recently been linked with higher lupus disease activity.[12] A 2012 study of 106 recurrently active patients with major renal SLE manifestations found a significant decrease in 25(OH)D serum levels among non-African American patients during flares occurring during low-daylight months (October-March). The decrease was approximately 3-fold larger than the expected reduction in 25(OH)D levels during low-daylight months. Consequently, the larger than normal decline in vitamin D was considered a mechanism of SLE flare.[13]

In that study, flares that occurred during high-daylight months were associated with a small but significant increase in the 25(OH)D level in the non-African American patients. Researchers surmised that even though normal 25(OH)D levels may be protective against flares, this mechanism of protection may be overridden during high-daylight months by other mechanisms, such as UV exposure. The flare rate among African American patients in the study did not differ between low- and high-daylight months.[13]

Another single-center study that followed 202 patients with SLE from baseline to 24-month follow-up observed flares in 16.8% of the patients. For this population of patients, the predictors of a flare included SLE diagnosis at age ≤25 years, severe disease as defined by use of immunosuppressors at baseline, or previous lupus nephritis. The risk of flare was increased more than 2-fold for patients diagnosed at age ≤25 or younger,

Table 1 *International Consensus Definitions of Lupus Flares*[4]

ORIGINAL QUESTION	FINAL DEFINITION
What is a flare?	A flare is a measurable increase in disease activity in one or more organ systems involving new or worse clinical findings and/or laboratory measurements. It is a temporary event and must be considered clinically significant by the assessor and usually there would be at least consideration of a change or an increase in treatment.
Is there a difference between disease activity and flare? If so, what is it?	Yes. Disease activity encompasses all the signs and symptoms or laboratory abnormalities related to lupus pathophysiology. It is determined at one point in time and is unrelated to the prior amount of disease activity. A flare is an increase in disease activity as compared to a previous assessment and usually there would be at least consideration of a change or increase in treatment modality.
When does flare begin?	The flare begins at the first sign or laboratory measurements of an increase in disease activity, whether or not the flare has reached its maximal disease activity at that point.
When does flare end?	A flare ends when disease activity returns to the pre-flare level, not necessarily fully quiescent disease, or the disease activity has been stable for a given period of time.
Is there a minimal threshold of disease activity to define mild, moderate, or severe flare?	A flare can occur at any level of disease activity. These definitions will require a combination of ''change'' and ''total activity.'' Severe flare implies a threat to an organ or to life. However, this is not easy to define, and may be different for different organ systems. Several instruments (BILAG, SLEDAI, SLAMR, CLASI) could be used to define the thresholds, and studies are needed to quantify these definitions.
Is the degree of change important in defining mild, moderate, or severe flare?	The degree of change may associate with the degree of flare over a broad range of disease activity levels. A change in the intention to treat, even without major change in disease activity level, should be considered to define the severity of flare.
Are there examples of thresholds in which a small change could signify a severe flare?	Yes, if it leads to institution of aggressive immune suppression.
Do both degree of change and threshold of flare need to be defined?	Yes, but there may be some examples where one or the other would suffice to designate a severe flare. The approach should be pre-specified in clinical trials.
Is it important to distinguish between mild flare and moderate flare?	To distinguish between severe and non-severe flares is clinically more important than to distinguish between mild and moderate. Mild flares usually do not lead to medication changes yet still might be important to quality of life and they might also be important in clinical trials. Moderate flares need to be recognized opportunely in order to be properly treated and controlled. These terms need more precise definition even though it also depends on the organ involved.
Is there a way to define a mild flare that would not be confused with symptoms not attributable to lupus?	Probably—this is a major problem. It depends on careful differential diagnosis by the clinician and clinical skills. Specific biomarkers help to indicate the presence or absence of lupus disease activity.
How do you differentiate a flare from ongoing clinical disease?	A flare is a change, an increase of disease activity or a worsening in symptoms that were not present previously that is temporary, while ongoing clinical disease is a continuum worsening and would not be considered a flare.
How do you differentiate flare from progressive organ damage?	A flare represents a potentially reversible increase in disease activity resulting from immunologic or inflammatory activity. Progressive organ damage occurs secondary to treatment toxicity or as a consequence of scarring from inflammatory insults. Clinical skill is critical to distinguish between the two entities and currently available assessment may be useful. The identification of future biomarkers should facilitate our ability to differentiate a flare from progressive organ damage.

3-fold for those treated with immunosuppressor therapy, and 4-fold for patients with previous lupus nephritis.[14]

Hormones and pregnancy have also been cited as potential triggers of disease flares, with estrogens possibly enhancing immune system activation, although there is disagreement about their role in triggering disease activity. Most flares in pregnant women occur in the first trimester, and up to 30% of SLE patients experience postpartum flares between 2 and 8 weeks following delivery.[6]

Some research has found that the risk of SLE flare increases dramatically in women who have had active lupus in the 6 months prior to pregnancy and discontinued medications such as hydroxychloroquine and azathioprine.[15] An analysis of The Hopkins Lupus Pregnancy Cohort from 1987 to 2002 found that the risk for significant SLE activity during pregnancy was 7.25-fold higher if a patient had recently active lupus prior to conception; other studies have identified a lesser increase in risk of flare during pregnancy among women with active SLE at conception. Other identified risk factors of flare during pregnancy are discontinuation of antimalarial therapy and history of highly active lupus in the years prior to pregnancy.[16]

A final meta-analysis that included 37 studies of pregnant women with SLE found an overall flare rate of 25.6%, mostly in patients with previous lupus nephritis. The largest study included within this meta-analysis found severe flare rates of 2.5% by week 23 of pregnancy and 3.0% by week 35.[11]

It is unclear whether hormones, hormone replacement therapy, or oral contraceptives trigger SLE flares. An early trial of high-dose estrogen-containing oral contraceptives in patients with active renal involvement found an increased risk of flare. Other studies, however, have found no impact of low-dose estrogen-containing oral contraceptives in risk or rate of flares.[11] Some studies have found a decrease in the number or severity of disease flares in women with SLE after menopause.[17]

Colds and viral infections have also been implicated as possible flare triggers. Epstein-Barr virus may precede or be associated with disease flares,[11] while other active cytomegalovirus infections have been detected at the time of flares.[18] The immunologic response to Epstein-Barr virus seems to depend on a person's genetic background, with the immune response to the infection playing a significant role in development of early autoantibodies.[19]

Other active viral infections detected during flares have included parvovirus B19, herpes simplex, varicella zoster virus, hepatitis A, as well as other less frequently reported viruses. Invasive fungal infections have also been associated with high disease activity.[18]

Higher levels of C-reactive protein (CRP) at baseline are also associated with a greater prevalence of self-reported flares. The long-term follow-up of patients in the Carolina Lupus Study found that the prevalence of active disease/flare was 31% in patients with CRP <3 µg/mL, compared with a prevalence rate of 47% in patients with CRP >10 µg/mL.[20-22]

Assessment of SLE Flares

A thorough history and physical examination that includes all major organ systems should be conducted at every visit for a patient with SLE. Any new symptoms or changes in symptoms should prompt further examination.[23]

Several global scoring systems have been used over the past several decades to assess lupus disease activity. These include the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI), the Systemic Lupus Activity Measure (SLAM) index, and the European Community Lupus Activity Measure; they each provide an overall measure of disease activity. More specific instruments that assess disease activity in single organs include the British Isles Lupus Assessment Group (BILAG) Index. Revisions have been made to SLEDAI and BILAG: the SLEDAI-2K and the BILAG 2004 (Table 2).[24-27]

SLEDAI-2K assesses disease activity and includes items such as the presence of ongoing rash, alopecia, or mucosal ulcers. SLAM measures global disease activity within the previous month. BILAG 2004 captures changes in disease manifestations over time, records disease activity in each organ system rather than giving a global score, and is based on intention to treat.[25,28] The BILAG can provide a more comprehensive overview of activity in eight organs/systems at a single point in time.[23]

Even though each of the SLE activity measures have both benefits and shortcomings, researchers advise that, given that patients with SLE have complex and different manifestations of the disease, these forms are most helpful when applied consistently and uniformly through proper and simple training.[25]

Although global scoring systems are used in large-scale international trials of new biologics, there are few with the ability to identify clinical flares. Therefore, several studies have examined the diagnostic value of flare-specific scoring systems that help determine the time to flare, numbers of flares, and severity of flares.

The Safety of Estrogens in Lupus Erythematosus National Assessment (SELENA) flare index (SFI) is a composite of SELENA-SLEDAI that helps identify and delineate mild, moderate, and severe flares, as well as physician-rated disease activity. The revised SFI (rSFI) suggests specific clinical manifestations for each organ system and categorizes flares into mild, moderate, and severe on the basis of the treatment decision.[29]

Table 2 *Strengths, Weaknesses of SLE Disease Activity Measurements*[25]

Measurement	Strengths	Weaknesses	Time Required to Complete
SELENA-SLEDAI Flare Index (SFI), revised SFI (rSFI)	rSFI suggests specific clinical manifestations for each organ system; categorizes flares into mild, moderate, severe	Training essential for optimal performance	20 minutes
The BILAG-Based Composite Lupus Assessment (BICLA)	Strict criterion needed to define flare	Significant time needed to complete form; training essential for optimal performance	50 minutes
SLAM and SLAM-R	Includes disease activity and severity calculations	Scoring relies heavily on patient self-reports	15 minutes
SLEDAI, SELENA-SLEDAI, SLEDAI-2K	Practical, used commonly for clinical, research purposes	SLEDAI versions do not capture improving or worsening disease, and do not include severity within an organ system	10 minutes
BILAG and BILAG-2004	Disease activity and severity calculations are included	Significant time needed to complete form; formal training essential for optimal performance	50 minutes
SLE Responder Index (SRI), SRI-50	SRI-50 superior to SLEDAI-2K for identification of patients with >50% improvement	SRI may miss the signal toward improvement	15 minutes

What to Ask Your Lupus Patients

Caring for patients with SLE not only involves diagnostic testing but also an understanding of how the patient perceives and experiences his/her illness. The healthcare team can play a vital role in educating patients about their disease symptoms and diagnostic test results, the importance of adhering to prescribed therapy, and environmental and other types of triggers that can result in heightened disease activity and flares.[8,23,32,38]

Talking with patients at every visit by asking questions and discussing the rationale behind their answers is important to help support patients and allow them to better understand the care they are receiving and steps they can take in their everyday life to cope with SLE and potentially prevent disease progression.[38] Encouraging patients to ask questions about their disease, their quality of life, and the care they are receiving can lead to improved shared decision-making and better adherence with therapy.[39]

The BILAG-2004 flare index was developed to help identify flares and is based on eight organ systems, each assessed on a scale from A to E. Grade A indicates very active disease; Grade B, moderate disease; C, mild stable disease; D, no disease activity but suggestive of a system that was previously affected; and E, no current or previous disease activity. A severe flare is defined as the presence of at least one A score, a moderate flare as the presence of at least two B scores, and a mild flare as the presence of one B or at least three C scores on the index.[25,30]

A comparison of the BILAG-2004 flare index with the SFI and the physician's global assessment found the highest inter-rater reliability with the BILAG-2004. Good agreement was found between the indices for distinguishing flares from non-flares, but there was less consistency in the ability to identify mild and moderate flares.[24]

Due in part to the variety of tools in use, an international consortium recruited more than 40 rheumatologists to address the dilemma of how to accurately distinguish flares from ongoing, persistent disease.[31]

This group of rheumatologists used nearly 1,000 paper-based individual case histories to determine the capacity of three flare activity instruments: BILAG 2004, SFI, and the rSFI. Investigators found a relatively high level of agreement among the flare instruments, indicating they are reliable to use in clinical practice to distinguish mild from moderate and severe lupus. However, the problem of capturing flare severity accurately, in particular distinguishing severe from moderate cases, continues and more research is needed, underlining the need for clinical examination and judgment.[25,31]

Assessment of disease activity as manifested by flares is crucial to a patient's well-being. This assessment can involve use of an activity index such as the BILAG 2004 or SFI, as well as assessment of the patient's quality of life since the previous visit. Other scoring systems to assess disease activity include the Health Assessment Questionnaire (HAQ), a quality-of-life index commonly used in other inflammatory diseases such as rheumatoid arthritis. Another common form in other conditions, the Short Form-36 (SF-36) index, can assess health status over the previous month.

It is also important that an accurate drug history is taken at each patient visit. Problems in SLE often arise due to patient noncompliance with drug treatment. Patients receiving disease-modifying antirheumatic drugs (DMARDs) should be monitored with regular full blood counts and liver function tests.[23]

Diagnostic Testing

Routine diagnostic testing and assessment can also be used to monitor disease activity and damage in patients with SLE. While these tests do not need to be performed at every patient visit, it is appropriate to order them at the time of a possible or perceived flare. The lab tests may include the following:[1,23,32]

Here are some questions you may want to consider asking patients with SLE at each visit:

- Can you describe your level of fatigue and joint pain since your last office visit? Is it worse, about the same, or better?
- Do you feel as though you need to nap more often than you did a month ago? How many hours of sleep do you usually get each night? Do you think that's enough for you?
- Have you missed any of your medications this past week? How often do you think you forget to take your medications in one week?
- Are you taking any supplements that your doctor did not prescribe?
- Have you added any new medications for other medical conditions since your last visit?
- Do you have any concerns or questions about the medications you are taking?
- Have you recently gained or lost weight?
- Are you being careful about spending time in the sun? Do you use sunscreen and wear protective clothing and a hat when you are outdoors?
- How would you describe your usual diet? What kinds of things do you usually eat for breakfast? Lunch? Dinner? Snacks?
- What type of exercise do you get each week?
- Have there been any major changes recently related to your family or job?

Continued on page 10

- Full blood count and white cell differential to assess for hematologic abnormalities, such as anemia and lymphocytopenia, that can be predictive of flares. Some hematologic abnormalities can be due to concomitant therapy.
- Erythrocyte sedimentation rate (ESR) paired with C-reactive protein (CRP) to help distinguish flares from infections. Raised ESR with normal CRP may occur during a flare; raised ESR and CRP in combination may indicate an infection.
- Urea and serum creatinine testing. A rapidly rising urea and/or creatinine may indicate that renal activity is occurring. Urinalysis will evaluate red and white cells, protein, and cellular casts that may indicate clinically silent renal disease. Urine protein creatinine ratio is considered the gold standard for monitoring renal function in SLE and identifying silent renal disease.
- Liver function testing for patients being treating with some DMARDs.
- Serologic tests to detect the presence of antinuclear antibodies. Anti-double-stranded DNA antibodies (dsDNA) are positive in approximately 60% of patients with SLE and may fluctuate with disease activity. A flare may be imminent in patients with rising antibodies to dsDNA, even in the absence of other clinical signs. A positive dsDNA indicates that an organ may be at risk of damage.
- Assessment of complement. Falling C3 and C4 levels may indicate that a flare is imminent, or a steroid taper should be slowed temporarily. If these levels are falling and the patient also has rising antibodies to dsDNA, the medical team should schedule more surveillance in anticipation of a possible flare.
- Assessment of cardiovascular problems, especially if the patient complains of chest pain. Chest pain when lying down can be a symptom of pericardial effusion.

Treatment Options for Disease Flares

Therapy during a flare depends on whether the activity is considered mild, moderate, or severe. Symptoms of a mild flare may be managed with agents to control pain, swelling, and fever, such as NSAIDs or topical

"What to Ask..." continued

- Do you think you are having any issues with depression, stress, and anxiety?
- What have you done for fun recently?
- (For women of childbearing age only) Have there been any changes in your plans to become/prevent pregnancy?
- Do you have any sores in your mouth or nose? Have you noticed dryness in your eyes or mouth?
- Are your vaccinations up to date? Did you get the flu shot this year?
- Do you know the warning signs of a lupus flare?
- Have you noticed that you are losing more hair than usual?

Here are targeted questions that may be used specifically with teenaged patients:

- What activities or sports are you involved in these days? What else do you do for fun?
- Do you always remember to take your medications?
- Do you have a reminder system in your cellphone to prompt you to take your medications?
- Do you always remember to take your medications with you if you go out of town or spend the night at a friend's house?
- Have you found a good way to carry your pills with you?
- Do you often nap after school? How many hours of sleep do you get on a school night?
- Do you remember to use sunscreen and limit your time in the sun?
- Are you sexually active? If so, do you always use birth control?
- Is there anything that you wish your parents, family, or friends understood about your illness? Can you think of a way we could help with that?
- How do you think you are doing? Is there anything else you would like to discuss with me?

steroids for skin lesions. Glucocorticoids, particularly prednisone, are considered a mainstay of SLE treatment and management. They rapidly control disease activity in patients with both mild and severe SLE, and an increase in daily oral prednisone followed by rapid tapering or an intramuscular glucocorticoid injection has been found effective for controlling disease activity.[32,33]

Some research has reported a significant reduction in severe flares among patients treated with moderate-dose glucocorticoids when they present with increased C3a and anti-dsDNA antibodies. These agents have anti-inflammatory properties over the short term and immunosuppressive actions in the long term.[33,34]

A short course of moderate-dose corticosteroids can treat active disease and help prevent flares in clinically stable but serologically active patients with SLE. Use of corticosteroids, however, has been associated with dose-limiting toxicities and other side effects. In clinical practice, clinicians often stabilize corticosteroid doses in serologically active patients and await clinical manifestations before increasing dose levels due to potential toxicity.[34]

Increasing the glucocorticoid dose has been shown to be effective in pregnant women who SLE who flare. Because glucocorticoids increase the risk of gestational diabetes, infections, and premature rupture of membranes, the dose should be kept as low as possible. For resistant flares, immunosuppressive drugs and intravenous immunoglobulins can be considered in pregnant women, although they should be used with extreme caution due to possible side effects.[33]

Hydroxychloroquine (HCQ), an anti-malarial drug, has anti-inflammatory and immunomodulatory properties and can have a significant effect on long-term SLE outcome by modifying the course of the illness by reducing low-grade flares. The therapy is most effective in managing mucocutaneous and musculoskeletal symptoms, as well as for fatigue and fever.[34] A systematic literature review published in 2013 found that use of HCQ in patients with SLE was associated with less damage at 3 years after diagnosis when regular dose adjustments were made based on disease activity, steroid dose, and calendar year of diagnosis.[35]

Immunosuppressive agents are also indicated to reduce flares or relapses and can be prescribed with high-dose corticosteroids to control or reduce flares. Belimumab, a biologic therapy approved for use in adult patients with active, autoantibody-positive SLE who are receiving standard therapy, has been shown to be effective for reducing risk of severe flares and for extending time to first flare.

Nonpharmacologic therapy can also be recommended to help prevent flares, including the following: [37]

- Sun avoidance or, when not possible, frequent application of sunscreen
- Avoidance of cigarette smoke
- Eating a healthy diet of fruits, vegetables, and whole grains
- Exercising regularly
- Getting enough rest
- Reducing stress
- Using corticosteroid skin creams as needed
- Learning to recognize symptoms of flares

Summary

SLE flares not only affect a patient's quality of life but also indicate an increase in disease activity and potential organ damage. Depending on their severity, flares should signal the need for a temporary increase or change in therapy. Patients should be informed about the reasons for diagnostic testing when a flare occurs. Discussions with patients can help identify potential flare triggers in their environment, ways to diminish exposure to some of these triggers, and the importance of adherence to therapy to control disease activity.

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The Power of the Team

by Linda Grinnell-Merrick, MS, NP-BC

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Lupus is a chronic disease that requires long-term care, which can pose a significant challenge to the patient and health care team. There are many epidemiologic, socioeconomic, and psychosocial factors that come into play that make this a complicated disease to manage.

Patients with lupus are often young women, with some ethnic groups such as African-Americans and Hispanics being prone to more severe disease.[1] Quality of life is often severely reduced by unpredictable and fluctuating disease flares, along with side effects related to lifelong treatment. Socioeconomically, we have many patients with lupus who are unable to maintain regular employment and pay for health insurance. Frequently, these patients will only seek health care when they visit a hospital emergency room in a crisis.

Recognizing these persistent issues, my hospital recently created an initiative that identifies lupus patients at high risk of nonadherence and enrolls them in a quality improvement program. This program matches the patient with a comprehensive team that includes a rheumatologist, a registered nurse, and a social worker, among other professionals. The goal of this program is to decrease hospital admission rates, improve overall adherence, increase family involvement, get patients more involved in social activities, improve patient and family knowledge on lupus, and provide educational sessions on lupus to community providers. This program was made possible through a foundation grant that focuses on at-risk populations and will run for 2 years.

One of the first patients enrolled in this program was MG, a 20-year-old African-American female. MG presented to our office with general fatigue, multiple arthralgias, and facial rash. Her initial lab results, which are shown in Table 1 (values in yellow fields indicate abnormal results), were highly concerning, leading us to expediate her care and admit her to the hospital. There, MG received a renal biopsy and was started on high-dose corticosteroids.

Initially, MG was adherent to her follow-up care, coming to our office as scheduled for the first few months. Tapering her off corticosteroids, however, proved to be challenging—MG rarely remembered to bring her medications to her clinic visits and she could seldom recall the dose she was taking.

Things really began spiraling downhill about 6 months after we began seeing MG when her mother, who suffered from alcohol and drug addiction issues, kicked MG out of her house, forcing her into homelessness. Suddenly,

Table 1

Lab Test	Reference Range	Result
Anti-RNP	Latest Ref Range: 0.0 - 0.9 AI	>8.0
Anti-Smith	Latest Ref Range: 0.0 - 0.9 AI	>8.0
ANA Screen	Latest Ref Range: Negative	Positive
ANA Pattern	Unknown	Homogeneous
ANA Titer	Unknown	320
dsDNA Ab	Latest Ref Range: 0 - 4 IU/mL	492 IU/mL
Blood,UA	Latest Ref Range: Negative	3+
Protein,UA	Latest Ref Range: Negative mg/dL	100
Creatinine,UR	Latest Ref Range: 20 - 300 mg/dL	116
Protein,UR	Latest Ref Range: 0 - 11 mg/dL	393
TP Creatinine ratio,UR	Unknown	3.39

MG had no money, no home, and no phone. Not surprisingly, she stopped taking her medications soon after.

For the next 2 years, MG lived a topsy-turvy life, bouncing between her sisters' houses and homeless shelters. There were many missed appointments at our clinic; we often only received updates on MG's condition when she ended up in crisis in the emergency room. MG was extremely difficult to contact due to her lack of engagement and rare ownership of a cell phone. She frequently ended up in the emergency room, yet often left against medical advice once she felt she was better. She would tell us her medications "tasted bad" or that she "couldn't find them" when we asked why she was nonadherent to her treatment plan.

MG was one of the first patients we enrolled in our QI initiative. At the time we rolled out our plan, her life was beginning, at least to a small degree, to stabilize. She had recently turned 22 years old and was living in a private room at the YMCA (though she sometimes spent several days or weeks at her sisters' houses). She had obtained a free "Obama phone," although it was rarely functional. She had also enrolled in Medicaid and was receiving both Social Security disability as well as food stamps, although her status was tenuous due to missed classes and follow-up appointments with social workers.

We met with MG during one of her emergency department visits and explained to her what the QI program entailed. We knew that MG had made a previous connection with an RN in her primary care provider's office, so we recruited that nurse to be part of our initial connection team. We never pressured MG to participate in the QI program, but delicately explained during several subsequent emergency department visits how this new program would be beneficial to her well-being.

Working hand-in-hand with a social worker who was part of the QI team, we engaged MG to determine her specific needs. We began to set up calls and texts both to MG and her sisters for appointment reminders as well as reminders for pre-appointment lab draws. We placed a call to a local food pantry and set up appointments for MG to go grocery shopping. Our nursing team provided education regarding MG's medications and the dangers of medication nonadherence. Our team also coordinated with our hospital pharmacy so that MG could fill her prescriptions on site and leave with her medications in hand. We maintained close contact with MG's primary care office through our electronic medical record and phone calls. In sum, our program was designed so that MG would no longer fall through the cracks.

It's only been approximately 4 months since we put the latest plans in place. Since then, MG has shown up for 2 follow-up appointments—1 with her rheumatologist and 1 with her primary care physician—and has gotten her labs test drawn as scheduled. Her medication adherence is still a bit spotty, but it's much improved from where we began. Slowly, it seems as if MG is gaining trust in our team and is becoming more engaged in her care.

Building communication between the primary care and rheumatology offices has been vital to our success. It's not always an easy bridge to build, but we have found that exchanging messages in the electronic medical record and quick phone calls when necessary can make all the difference in a patient like MG. Seeing improvements not only in our patients' health but their faith and trust in their healthcare team is one of the most rewarding parts of our job. We're hopeful to have more patients like MG in the future who buy into our new team-based system.

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The Intersection of Lupus and End-Stage Renal Disease

by Jacqueline Fritz, RN, MSN, CNS, RN-BC

It's a question that I have considered from time to time in my nursing career—in a patient with severe systemic lupus erythematosus (SLE) who also develops end-stage renal disease (ESRD), can the patient's SLE ever go into remission? As with many clinical questions, experience gave me my answer.

In 2009, I met LW, a 37-year-old patient who had been diagnosed with SLE about a decade ago. Upon his initial diagnosis, he had a positive anti-ribosomal P titer (165 U; normal 1-20, borderline between 20 and 25), mildly elevated anti-chromatin antibodies (119 U; normal 0-99), and a mildly elevated serum creatinine level (1.58 mg/dl, normal 0-1.3). A renal biopsy showed grade V glomerulonephritis, which unfortunately is indicative of near-certain future ESRD requiring eventual dialysis and likely renal transplant.[1]

LW was started on daily prednisone 60 mg, which helped control his SLE symptoms, along with furosemide 40 mg, potassium chloride 20 mEq, daily isosorbide mononitrate ER 30 mg, daily labetalol hydrochloride 200 mg QD, daily lisinopril 20 mg, and daily mycophenolate mofetil 1 g.

Not long after his diagnosis, LW's begin spilling 3-4 g of protein in urine each day, which led to him being placed on cyclophosphamide. He received 7 doses over the subsequent 10 months, which unfortunately was ineffective in alleviating his lupus-related symptoms. Even more significantly, he soon developed lupus cerebritis, possibly caused by the prolonged use of prednisone and uremia (his serum creatinine was now up to 6.82 mg/dL).

LW was hospitalized on several occasions with many of the more common symptoms associated with lupus cerebritis, including polyneuropathy, severe headaches, hallucinations, psychosis, and seizures.[2] He was treated with additional prednisone and cyclophosphamide, which led to the common push/pull we often see with our patients. His cerebritis-related symptoms became better controlled, but LW suffered numerous side effects of prednisone, including weight gain, agitation, and large swings in his blood sugar levels.

LW would come into our office wearing beach sandals, the only footwear that would accommodate his grossly swollen and cumbersome feet. Trained as an engineer, LW was totally disabled when he first started coming to our office, barely surviving between frequent hospitalizations.

There were, not surprisingly, good and bad days. LW began to suffer from colitis, again possibly due to the prednisone but also potentially due to his SLE. Thankfully, he did not develop a pleural or pericardial effusion.

Due to his renal disease and loss of erythropoietin, LW's hemoglobin was perpetually low, regularly hovering around 7.7 g/dL, and he had a hematocrit of 23.5%. After many months of coaxing, LW finally agreed to see a nephrologist, who added epoetin alfa to his medication regimen in an effort to stabilize his blood counts. This led to resolution of his proteinuria as his renal output was reduced to <400 cc over a 24-hour span, which is considered oliguric renal failure.[3] In addition, his serum creatinine spiked to 13 mg/dL.

We felt that the root cause of many of LW's issues was renal failure and so, during visits when he seemed most mentally alert, we suggested to him that dialysis would likely have a positive impact on many of his symptoms. Knowing how difficult it would be for him to accept being dependent on dialysis for the rest of his life, I emphasized the very real danger of succumbing to his medical issues if they were not addressed soon.

Perhaps because of his engineering background and the science behind the technology, LW finally agreed to consider ambulatory peritoneal dialysis. I made sure that he understood that

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"While the reasons for their improvements are not well understood, I am always grateful when we can help patients get their lives back."

this would require daily treatment, but that, if all went well, he might be able to return to work and be in better control of his disease.

A few weeks later, LW came into our office wearing a dress shirt and tie. He was beaming as he told us, "I am working again!" While his lab results were still concerning—his serum creatinine was down, albeit slightly, to 9.0 mg/dL—his enthusiasm was something we had never before seen. While LW admitted needing to nap at lunch everyday due to fatigue, that was still a vast improvement from monthly hospitalizations, seizures, and psychoses.

So is LW's SLE truly gone? The medical literature shows it is not uncommon for patients on extended dialysis to see their lupus symptoms improve significantly. One recent review found that the percentage of lupus patients with clinical activity after the initiation of dialysis decreased to 55% after 1 year of dialysis, to 6.5% after 5 years, and to 0% after 10 years. There was a corresponding decrease in serologic activity and disease activity scores. There is even a term—lupus burn-out—to describe this phenomenon.[4] The underlying mechanisms behind these remarkable improvements in SLE-related symptoms is not entirely clear.

Of course, providers should note the treatment of ESRD does not always result in complete or even partial resolution of lupus-related manifestations as seen with LW. I have, however, had a second patient with a similar story after a renal transplant.

While the reasons for their improvements are not well understood, I am always grateful when we can help patients like LW get their lives back. Patients with severe lupus often suffer on the precipice of health-related calamities, and our options for treatment, while improving, still offer only moderate hope for some of our patients. All we can do is try our best for our patients and to maintain our optimism on the darkest of days with the hope that something will work as far down the line as our patients need to travel.

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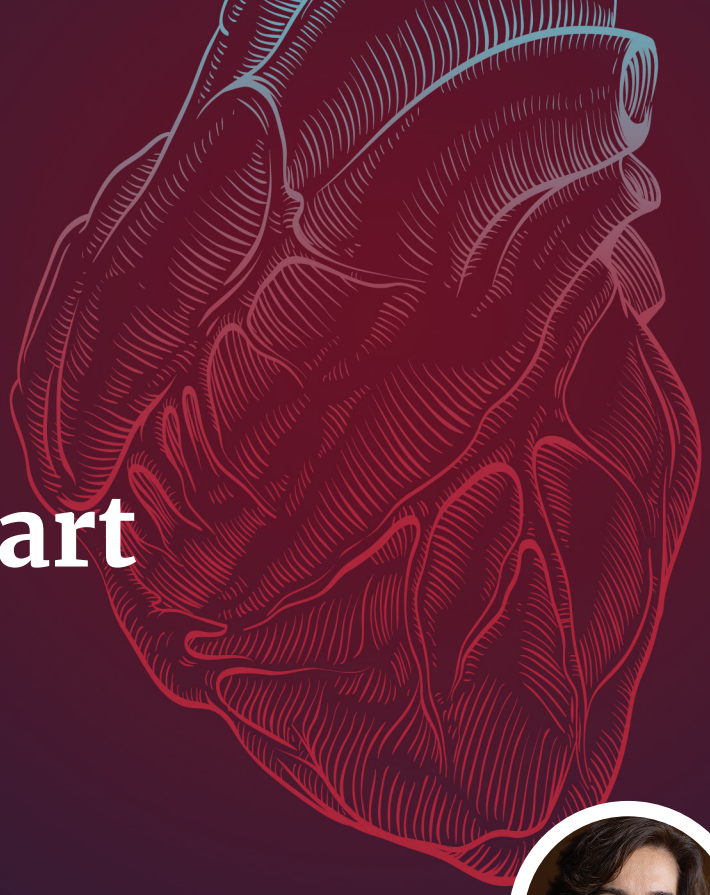
Never Forget About the Heart

by Monica Richey, NP, MSN, ANP-CP/GNP



I still vividly remember the day we received the news that WS had passed away at the age of 32.

A regular patient we had been seeing for more than a decade, it had been less than 1 month since WS' last appointment at our clinic. We had gotten to know her family—including her husband and children—very well as we managed the ups and down of her aggressive systemic lupus erythematosus (SLE).

WS was a model patient, always taking her medications as prescribed, showing up for every scheduled appointment, and getting pregnant only when her physician told her it was safe to do so.

That's what made the news of her untimely death such a shock to us all. There was nothing in her most recent workup that indicated an emergent medical condition. A massive, fatal heart attack? At only 32 years old? What are the odds?

Actually (and unfortunately), the odds are rather high.

Women aged 18–44 years who have been diagnosed with SLE have approximately 9 times higher risk than the general population of hospitalization due to heart attack or stroke. The risk of death due to cardiovascular disease (CVD) is 17 times higher in patients with SLE and heart attacks occur 52 times more frequently.[1]

In a nutshell, while the risk of CVD in the general female population increases after the age of 55, in women with SLE, it's early adulthood where the risk of CVD is highest.

It's well documented that the mere presence of SLE serves as an independent risk factor for CVD. Therefore, even if you screen both for modifiable risk factors such as smoking, hypertension, and obesity, as well as nonmodifiable risk factors such as age, gender, and family history, in determining a patient's risk of a CVD event, you still must also factor the presence of SLE into the equation. Let's also not forget that many of our SLE patients are being treated with corticosteroids, which further increases the risk of CVD.

While the reasons for the increased risk of CVD in younger women with SLE are not entirely clear, it is thought to be related to the interplay of inflammatory mediators such as leukocytes, cytokines, chemokines, adhesion molecules, complement, and antibodies that result in the formation of atherosclerotic plaques.[1]

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So what can the nursing community do to mitigate this risk? In 2010, the European League Against Rheumatism (EULAR) published recommendations for the management of CVD in patients with various rheumatic conditions, but unfortunately left SLE out. Consequently, there are no clear guidelines to pull from to help guide screening and management of CVD in patients with SLE. And with all of the other complications that often arise with our SLE patients, it can be easy to forget about our patients' hearts.

I was involved in one of the first CVD prevention programs in the country specifically targeted at patients with SLE. Beginning in March 2009, we enrolled 121 patients who were regularly assessed for modifiable and nonmodifiable CVD risk factors and educated on risk reduction strategies. Although we noted positive changes in diet and exercise—along with mean high-density lipoprotein and mean triglyceride levels—at the end of 3 years, there were no significant changes in blood glucose or body mass index levels.[2]

Nonetheless, this prevention program was well received by our lupus patients, who are always asking for information and guidance regarding diet and exercise. Anecdotally, there seemed to be a tremendous difference in patients' diets based upon their socioeconomic status. Clinic/Medicaid patients were more likely to have a poor diet and less access to fresh food and vegetables, and often could not afford a gym membership.

Participating in this program taught me that changes must be introduced gradually for most of our lupus patients—too many changes suggested at once were seen as an impossible challenge. I felt victorious when patients told me they had stopped drinking soda. Walking 30 minutes, even just once a week, was cause for celebration. There were times when we took two steps forward and then two steps back, such as when a patient spent months losing 5 or 10 pounds before suffering a major disease flare which put him back on prednisone and reversed the previous weight loss. Some patients lost interest in making lifestyle changes as their day-to-day struggles with their disease became too difficult, while others continued to make changes despite many obstacles. Overall, I would say that patients loved the 1-on-1 attention they got through this program, and I made many new friends.

Unquestionably, we need more research to help us understand why our patients with SLE, and especially young women, are at such an increased risk of CVD, but it is important for us all to be cognizant of the facts and inform our patients of the challenges that may lie ahead for them.

One recent study emphasized the need to order an electrocardiogram (ECG) in patients with SLE on a periodic basis. This study of nearly 500 patients with SLE found a high overall prevalence of ECG abnormalities (21.4%), including ST-segment and/or T-wave abnormalities, left ventricular hypertrophy, left axis deviation, left bundle branch block, right bundle branch block, and q wave variations.[3]

Finding out news like we did with WS can be a jolt. While we know that there are significant health risks among our patients with SLE, it's never an easy day to get the ultimate bad news about any of our patients. It's incumbent upon the rheumatology nursing community not only to teach our patients but to provide them with support to make lifestyle changes to impact their health. So "have a heart" and challenge your patients—you may be surprised at how much you are able to accomplish together.

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The Value of Collaboration in a Small Practice

by Cathy Patty-Resk, MSN, RN, CPNP-PC

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Cathy Patty-Resk, MSN, RN, CPNP-PC is a certified pediatric nurse practitioner in the Division of Rheumatology at Children's Hospital of Michigan in Detroit, MI, where she provides medical services to inpatient and outpatient pediatric rheumatology patients.

Pediatric rheumatology not only requires close attention to detail when it comes to direct patient care but also effective collaboration with a variety of other sub-specialists. We commonly consult specialists in nephrology, gastrointestinal disease, ophthalmology, dermatology, immunology, infectious disease, ear, nose, and throat, and hematology/oncology, just to name a few. The diagnosis of some of our more complicated patients requires a couplet or sometimes even a triad of sub-specialists. Collaboration isn't always easy, and building a trusted relationship takes time and patience, but cementing professional coalitions is often in our patients' best interests.

Of course, not all academic practices are created equal. In Michigan, we have three pediatric rheumatology practices spread throughout southern Michigan, located in Grand Rapids, Ann Arbor, and Detroit. Our practice in Detroit is on the east side of the state, with just a single rheumatologist. The Ann Arbor center has the most pediatric rheumatologists on its staff, although they perform a considerable amount of research in addition to clinical care. The Grand Rapids center has multiple pediatric rheumatologists on its staff as well and also performs research in addition to its patient care.

In our small academic practice, we have our one pediatric rheumatologist, one nurse practitioner (that's me), one registered nurse (RN), and two administrative staff that we share with other specialty departments. Each week, we have 22 slots in our schedule for new patients and 51 slots for follow-up appointments. We also dedicate 1.5 days per week to make phone calls, deal with insurance appeals, complete patient charts, and handle other administrative duties that come up. As with many of you reading this, we are thoroughly immersed in patient care every minute of the working day.

With such a small team and a busy schedule, effective communication and collaboration are vital to maximizing our time and best serving our patients. We are fortunate within our electronic medical record (EMR) that we can send and receive messages from other sub-specialists involved in a patient's care. This method is far more convenient than email. Prior to the recent coordination within our EMR, we would often have to check with individual providers to see which email address they preferred (work or home) or if they relied more on another form of communication (believe it or not, there are still some physicians who don't use email at all). It was also next to impossible to follow an email chain that included multiple providers. All of these problems led to fragmented, suboptimal care.

The transition to messaging within the EMR allows our team to have group communications that may be saved within the patient chart. We can read the most recent sub-specialist updates and easily toggle back and forth between information related to lab results, medication changes, or other information without going from screen to screen.

That isn't to say that this transition has been entirely seamless. It's important to remain respectful of the time and demands of other providers, and some of us had difficulty putting boundaries on the sheer number of messages we could send or reply to. Our RN has learned to be ready on non-clinic days or days when the clinic is slower to triage items that

need attention from our provider team. These may involve documents that need a formal signature, new records that require review, medication refill requests, insurance denials, and other materials. The RN also serves as a crucial link between each member of our team, making sure the sickest patients and the most difficult families get the attention they need. She also stays on top of new testing codes or other items that insurance companies need for prior authorizations. It's often a thankless job, but I know that I could not see and manage as many kids as I do without such an organized, top-notch RN.

While we have learned to utilize the messaging functions within our EMR successfully, our sub-specialty teams have recognized that we still need periodic face-to-face time with each other to discuss our most challenging patients. These face-to-face meetings, albeit infrequent, serve as a good opportunity to build social bridges as well, ensuring that there is a "face" behind the EMR messages.

Within our small department, our rheumatologist and I are in clinic together daily, so we'll often take advantage of brief openings when a patient is running late or there are unexpected lulls in the schedule to discuss our current cases. As I've become more experienced in pediatric rheumatology, I find fewer cases where I need our rheumatologist's guidance, so our discussions have morphed into a give-and-take regarding our most fragile patients or the current hospital inpatients who will soon become new patients in our clinic.

The improvements that have been made in communication within our hospital over the last few years are significant, but we are still working on improving relationships with those sub-specialists who work outside of our hospital and are involved in the care of our pediatric rheumatology patients.

We work, for example, with external ophthalmologists, dermatologists, and hematologists quite regularly on the co-management of patients. Opening a dialogue with these providers has taken time, patience, and persistence, but I find that once I do manage to speak directly with other sub-specialists, they are so happy that we have reached out to them to coordinate care that the conversation often ends with the exchange of cell phone numbers and the gracious, "Call me or text anytime."

I've also found that once parents know I've spoken directly with a collaborating sub-specialist—particularly when they are from outside our hospital—they seem less stressed and tend to be more agreeable with suggested changes in a treatment plan. In the past, I noted parents would often be a little on edge worrying about what their other sub-specialists would think of our suggestions. I don't see the struggle of them feeling like they are stuck in the middle any more.

Of course, the circle of communication could not be complete if I didn't talk about parents. Parents are empowered these days through remote access to our EMR, which allows them to view test results, read clinic notes, and send emails directly to their providers. It's true that I've had to spend more time than I might like explaining to the concerned parent that a lab value just barely outside of the normal range is not at all worrisome, reassuring them that nothing bad is going to happen. However, I'll almost always add, "I'm happy to re-check (the value) next week if it means you will be able to sleep at night."

Advances in telecommunication and EMR systems are allowing providers to collaborate and communicate better than ever, while at the same time allowing more parents to sleep better at night knowing their children are in good hands. It's a win-win-win situation all around.

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