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### A Step-by-Step Guide to Diagnosing SLE in the Primary Care Setting

Dr. Birnholz: The timely diagnosis and treatment of systemic lupus erythematosus is essential to preventing, or at least diminishing, the autoinflammatory multisystem damage associated with this disease. Unfortunately, diagnosis is often delayed—sometimes for years—due to nonspecific signs and symptoms, so how can we effectively assess our patients and get them to treatment faster?

From the ReachMD Studios in Fort Washington, Pennsylvania, this is CME on ReachMD. I'm Dr. Matt Birnholz, and here with me to discuss lupus diagnosis and treatment planning from the perspectives of both primary and specialty care are Dr. Louis Kuritzky and Dr. Robin Dore. Dr. Kuritzky is a family physician and Assistant Professor Emeritus at the University of Florida School of Medicine, and Dr. Dore is a practicing rheumatologist and professor of medicine at UCLA.

Doctors, it's great to have you both on the program today.

Dr. Dore: Thank you very much for having us today. I'm looking forward to the discussion.

Dr. Kuritzky: It's great to be here, thank you.

Dr. Birnholz: So let's start with a case study involving a patient named Felicia. So, Felicia is an uninsured, 31-year-old, African-American, mother of two who works as an Uber driver. She comes to Dr. Kuritzky's office today reporting fatigue, arthralgia in multiple joints, muscle aches, painless oral ulcers, and moderately swollen cervical lymph nodes for the last three or four days. She has had two similar episodes over the past two years, but lab tests showed nothing remarkable at that time, and both resolved on their own with the assistance of over-the-counter NSAIDs. She has been able to continue working as an Uber driver, but says the fatigue, in particular, makes it pretty tough, as she puts it, and she wonders why the symptoms keep coming back. So, with that background, Dr. Kuritzky, would you say Felicia's symptoms are consistent with lupus? And, if so, what are the most important laboratory tests you'll order to confirm or rule out SLE?

Dr. Kuritzky: Well, thank you for that introduction. I think what you've described is a fairly typical initial presentation of lupus. But the reason, however, that there is often a delay in diagnosis of lupus is it's not always so straightforward that tells us right from the outset that it's lupus unless the patient is presenting with a very predominant rash on their face, consistent with the lupus facial lesions, or a family history, or evidence of nephritis, or some combination of those. So, while the lupus is indeed the most likely diagnosis, there are plenty of reasons for people to have fatigue; a diverse number of issues can cause arthralgias and oral ulcers and lymph nodes. So we can't just make a diagnosis based upon these clinical criteria.

There are several sets of criteria that are used, and we're going to rely upon agencies like the Systemic Lupus International Collaborating Clinics; they have a set of criteria, or the American College of Rheumatology. Either one of those are respected guides for how a clinician should come to the diagnosis of lupus erythematosus at this time. Now, the patient is uninsured, so we need to be respectful of her resources, and try to be choosing wisely in which things we want to get. Certainly it would be appropriate to consider a CBC and an ANA, and possibly a C3 and C4 at this point, also as our initial tests with the ANA. We have this initial panel that includes the CBC, probably a CMP, and ANA, maybe a complement level, and I think that would be a reasonable place to get started. But I guess what I'd like help on, Dr. Dore, is do you feel like that's enough initial information to get us going? Should I have done more, do you think, at this initial encounter? Or do you want to have more of the tests at your hands when you see her and make a further

decision? Please advise me about that.

Dr. Dore: Two things I want to mention is one, I would want a urinalysis to make certain that she doesn't have any proteinuria or red cell or white cell casts. If the urine was abnormal, then probably when I saw her I would order a double-stranded DNA antibody because that is typically positive in patients who have lupus nephritis. But if her urinalysis was normal, and her comprehensive metabolic panel showed that she had normal renal function, the fact that she's uninsured, I would not order that double-stranded DNA antibody.

The other thing that's really important is if she is told that she has a positive ANA and I'm sure you would just mention to her that the ANA is a screening test for autoimmune diseases, but that some medications in some patients have a positive ANA. So until she sees the rheumatologist, not to go online and assume she has lupus because she has a positive ANA, but try to tell her that she might not, and certainly that the rheumatologist should be able to tell her if it is lupus, the severity, and what the prognosis might be. So many patients are so concerned that they have lupus when they only have a positive ANA, and most of the ANAs I see in referral, the patients don't end up having lupus.

Dr. Kuritzky: Well, thanks for that advice. I do see the situation you're describing more often in primary care since we're more commonly using, for instance, medications like hydralazine, and then their ANA comes back positive and they worry they have a dread disease. So I'm glad you are reminding me that we need to pay attention to the propriety of the ANA, and not jump to conclusions based solely upon that.

Dr. Dore: The other thing I didn't mention was, you need to have an ANA titer because otherwise the ANA will just come back positive or negative. And for when the rheumatologist sees the patient, she wants to know what the ANA titer is; whether it's 1-to 80, which is minimally positive, 1-to 160, which is more positive, and that again can give me an idea of if I need to order any other tests or to help with talking about the prognosis.

Dr. Kuritzky: What about the ANA pattern? The only thing I've seen in the literature consistently is the centromere pattern that is one that's least likely to be associated with lupus. Does the ANA pattern type really make a difference to us?

Dr. Dore: To me it does. And again, if it's a homogeneous pattern, that is much more likely to be lupus. If it's a speckled pattern, that's more likely to be some other autoimmune disease. And when they give you that titer, they will give you the pattern that counts as the highest titer. So let's say at 1-to 80 it was homogeneous, but at 1-160 it was speckled, then it's the highest titer pattern that tends to tell us more about the disease.

Dr. Birnholz: And you both bring up interesting points. Dr. Dore, I want to come back to you on this concept of the tests that we need to consider. What about tests that are unnecessary or unhelpful from your vantage point?

Dr. Dore: As I mentioned, I would not order, again because she doesn't have insurance, I wouldn't order that double-stranded DNA antibody to begin with. I would wait and see what her GFR and urinalysis showed. The other thing that can really add up costs is if you ask for a lupus panel, and then they will be testing for scleroderma, for Sjogren's, for rheumatoid arthritis, for mixed connective tissue disease, and that initial ANA. The cost might triple or quadruple by getting that whole panel. So for someone that doesn't have insurance, I want the basic things that we talked about, and then if necessary, I can pick and choose.

Dr. Birnholz: That's excellent. Thank you. So, let's advance Felicia's case now and say that her lab tests show the following. So her ANA comes back positive, C4 is low at 10 mg/dL, and her white blood cell count is also low. All other test are normal, however. So, Dr. Kuritzky, starting with you again, do these findings satisfy a diagnosis of lupus, and would you look to consult with a rheumatologist, such as Dr. Dore, to confirm the diagnosis in this case?

Dr. Kuritzky: Well, I still would refer to Dr. Dore for confirmation, even though the patient I think should be, at this point, given a provisional diagnosis of lupus because she fulfills either the SLICC or the ACR criteria. According to SLICC, you need to have 4 of 17 criteria of which at least one has to be clinical criteria, and another has to be immunologic criteria. The immunologic criteria are really laboratory markers; the ANA, double-stranded DNA, and a phospholipid antibody, complement levels, Coomb's test. Clinical criteria are the things that a patient came reporting to us; cutaneous changes, hair loss, ulcers in the oropharynx or nose, joint pain, serositis, evidence of renal disease, as Dr. Dore mentioned earlier, neurologic findings, anemia, low white count, thrombocytopenia. So the patient already does fulfill criteria by the SLICC criteria, and the ACR are fairly similar. Based upon that, what I would say as the primary care clinician is, 'You do fulfill the criteria for the diagnosis and it's going to be a diagnosis that you will have to wrestle with for much of the rest of your life from time to time. I want to have an expert in this disease see you, also for the advanced therapies beyond the things I can initially give you. You're going to need to have a strong relationship with your rheumatologist, as well as me.'

Dr. Birnholz: Excellent. Dr. Dore, would you want to weigh in on that regarding a referral, the diagnosis, what might be preferable or necessary from your vantage point?

Dr. Dore: Definitely from my vantage point, since she meets the criteria both through the SLICC criteria and the ACR, she should be seen and evaluated by the rheumatologist so that the rheumatologist would be able to discuss the diagnosis of lupus, talk about what the different therapies are, and then really with the shared decision-making between the primary care clinician, myself, and the patient, talk about what we feel would be the best starting therapies for her, and then deciding the follow-up between the primary care clinician and the rheumatologist. So, ultimately she has lupus and we need to put her on low doses of prednisone and some hydroxychloroquine. And it's important that I communicate with Dr. Kuritzky because I think the patient is going to say, 'Dr. Kuritzky, I've never seen Dr. Dore before, and I don't really know her, but I trust that you sent me to her, which is good, but I want to make certain that you agree with this treatment plan.' So it's very important that there's a buy-in because if the patient, the primary care clinician, and the rheumatologist all agree on the treatment plan, that the adherence and compliance is going to be so much better.

Dr. Kuritzky: Dr. Dore mentioned something that I think is really important for this patient's future. Prior to the internet, it wasn't easy for patients to access information, then we were really handing the information in one direction from clinician to patient. But now it's clearly bi- or tri-directional; sometimes the information being inaccurate, and sometimes accurate. I think an important element for this patient, because it's frightening to get a diagnosis of lupus, and many people have heard of it and they know that it is definitely not good news, is to instill reassurance and hopefulness. Because what we can tell this patient, and what I would say to her the outset is, 'We're going to have a long-term relationship, you and I, you and your rheumatologist, and the good news is that we have a diversity of treatments that usually make patients much better.'

Dr. Birnholz: For those just tuning in, you're listening to CME on ReachMD. I'm Dr. Matt Birnholz, and I'm speaking with Dr. Louis Kuritzky and Dr. Robin Dore about diagnostic and management considerations for SLE.

So, we've been focusing on our patient, Felicia, here, who has just been diagnosed with SLE, but I want to come back to Dr. Kuritzky on the question whether her presentation is fairly typical from your point of view.

Dr. Kuritzky: I think her presentation was fairly typical. She is a young person and she is female and she is African-American. And those are all signals that increase the likelihood of, with similar symptoms, that her diagnosis is going to be lupus. It's not always so obvious, and people can sometimes mask their symptoms, sometimes they'll live with them for awhile and, when symptoms go away, they say, 'Oh, it was nothing.' So she may have written off what her symptoms are, thinking that they're not something important, but they are, and we don't want disease progression to occur. I think most primary care clinicians would recognize this presentation as lupus. But that's a gift that we don't always get; we get persons who are lesser likely to have it; males are less likely. We get persons who are absent of any skin manifestations, and we get persons like, for instance Asian-Americans or Hispanic-Americans, in whom the disease is not nearly so obvious.

Dr. Birnholz: Well, you bring up a great opening here to maybe bring Dr. Dore into this subject, and maybe ask about some of the less typical patients where the diagnosis might actually get missed. Dr. Dore, what are your thoughts?

Dr. Dore: Well, certainly, from my point of view, Felicia is really a classic case. I am most concerned if I see an African-American male who has lupus because typically their disease is more severe. We frequently see central nervous system involvement or renal involvement, and unfortunately the thinking is, 'Well, lupus doesn't occur in men.' Lupus can certainly occur in men, and we don't want to miss that because we want to aggressively treat all the patients. The other place I think is very important is in the older patients. There are over 100 different medications that can cause drug-induced lupus. If I see an older patient, and let's say they are having the fatigue that Dr. Kuritzky mentioned, and they have some joint pain and they're on one of the many, or maybe more than one of the many medications that can cause drug-induced lupus; most of the antihypertensive medicines, the anti-arrhythmic medicines, the statins can all cause positive ANA. That's going to be a person that I am going to order an anti-histone antibody on, because that should be positive in somebody that has drug-induced lupus. And then the most important thing we want to do at that point is again work with the primary care clinician and see if the patient can be tapered off that medication that caused the drug-induced lupus.

Dr. Birnholz: So if we come back to our patient, Felicia, whose presentation is more fairly typical and representative of lupus. Dr. Kuritzky, what are the next steps then? Would you start treatment right away?

Dr. Kuritzky: Well, I think our patient is suffering a very substantial symptom burden. I would actually probably, at this point, call the rheumatologist and say my intention is, unless you can see this patient within the next week, to give her prednisone of a small dose of no more than 10 to 20 mg a day until she is able to see you. There are some clinicians who feel comfortable based upon the provisional diagnosis to use hydroxychloroquine, but in most environments in the United States, we'd rather see the rheumatologist first, and make sure the treatment intervention by the rheumatologist is in such short order that she doesn't have to take more than the briefest course of steroids; certainly no more than one to two weeks at the most. Because likely she's going to be encountering steroids on a repetitive basis throughout her lifespan, and we want to minimize the toxicity to the bone if we can by getting more appropriate first-line therapies engaged as soon as possible.

Dr. Birnholz: So, I'd like to close our conversation with a question about monitoring patients like Felicia at this point going forward. So once she started treatment, how would you both assess her response to that treatment and monitor her symptoms? And would you see her at defined intervals of checking for particular signs? So, Dr. Dore, let me start with you on that.

Dr. Dore: I would agree that in this patient, unless she can get in to see the rheumatologist very quickly, that she does need to be started on the steroids because of that low complement. And that means that her lupus is probably more active than someone who has a normal serum complement level. So, when I would see her, I would probably add hydroxychloroquine to make it so we can try to limit the dose of the steroids because they not only affect the bone, but also increase cardiovascular risk in these patients. At that point, I would want to see the patient back in probably three months. I would ask their primary care clinician to see the patient back in four to six weeks, again trying to be careful with the insurance, but checking the CBC, the CMP, the urinalysis, and at this time checking that serum complement. And if it's still low, then I'm going to need to see the patient again and probably talk about adding something like azathioprine or mycophenylate to try to make certain that the complement level becomes normal, because that low complement is associated with more aggressive disease.

Dr. Birnholz: Dr. Kuritzky, what about you? What's your vantage point from the primary care side of things?

Dr. Kuritzky: The patient is going to need to be seen on an ongoing basis. When her disease is quiescent, unless it's for other health maintenance issues, probably every six months is going to be fine. But until her disease is stabilized, she is going to need to be seen at a minimum of every three months. Now, many of the decisions are going to be in the hands of the rheumatologist, but I want to give her ongoing education about her disease. Make sure that she reduces her cardiovascular risk—because it's elevated from baseline with exercise, appropriate diet, and avoidance of toxins like cigarette smoking. The primary care clinicians may be interested to learn that sunscreen is an important intervention, because sunlight exacerbates the disease. And we want to be sure this patient is appropriately vaccinated. So sometimes patients, when they get a major primary diagnosis, that seems to be the loudest noise in the clinical room. Her lupus is important, but she has other life issues that are just as important that require the same maintenance as we provide for any other patient without lupus, as well as the areas of heightened risk for cardiovascular and renal disease.

Dr. Birnholz: Well, with those takeaways in mind, I very much want to thank my guests, Dr. Robin Dore, and Dr. Louis Kuritzky, for helping walk through this patient case and, in the process, defining best practices in diagnostic and treatment planning for lupus patients. Doctors, it was fantastic speaking with both of you today.

Dr. Dore: Thank you very much for asking us to present this information.

Dr. Kuritzky: Thank you for allowing me to participate in this educational experience.