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Practice and Educational Gaps in Lupus, dermatomyositis, morphea

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Part 1

Lupus Erythematosus – Best Practices

Clinical and laboratory assessment—Best practices in cutaneous lupus erythematosus include recognizing the clinical presentations of specific subsets of cutaneous lupus (localized and generalized discoid LE, subacute cutaneous LE, acute cutaneous LE). There should be recognition that more than one subset may be seen in a given patient, assessment of whether the disease is active, and documentation of the degree of skin disease activity. Also, recognition of the extent of cutaneous damage is important, particularly if there is ongoing activity that needs aggressive treatment because of continued risk for scarring and dyspigmentation. The impact on quality of life should be assessed. There should be recognition of the importance of considering other potential diagnoses and the frequent need for initial documentation by skin biopsy to rule out mimickers and assure the need for proper long-term management. It should be recognized that subacute cutaneous lupus is frequently associated with use of certain medications, and these should be reviewed and stopped if potentially a trigger. The patient should also be carefully assessed by history for systemic symptoms, and laboratory tests assessed to rule out systemic disease. These initial labs should include ANA, CBC, and urinalysis. Proteinuria, if present, should be further quantified with spot protein: creatinine ratios, and if abnormal, then 24-hour a urine for protein quantification. If there is an indication of systemic features, then dsDNA, C3, C4, anticardiolipin antibodies, and complete metabolic panel should be evaluated. Anti-Smith

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antibodies can be obtained if the diagnosis of lupus is unclear, since it is a specific, but not sensitive antibody for diagnosing lupus erythematosus. If patients are contemplating pregnancy, determining the presence of anti-SSA and anti-SSB antibodies is done, since pregnant patients are managed differently because of the risk of neonatal lupus if they have these antibodies. Dermatologists should evaluate patients with CLE for the presence of significant systemic disease and manage patients with skin-predominant disease. They should provide ongoing assessments for development of systemic disease, refer patients when indicated because of serious systemic findings, but assume responsibility for long-term care or refer patients to centers with dermatologists who care for such patients. Dermatologists provide optimal skin care and preventive assessment of patients with skin-predominant disease.

Patient education and treatment—Patients should receive counseling about broad-spectrum sunscreen use, sun avoidance, and use of sun protective clothing. Smoking cessation should be strongly recommended and patients given suggestions on how best to pursue this. If disease is mild, topical therapy should be instituted. If there is evidence that skin disease is severe or progressive, systemic treatment should be recommended with antimalarials, immunosuppressives, thalidomide, and lenalidomide. Patients with depression or anxiety may need referral to psychiatry.

Longitudinal care—Disease activity and damage should be monitored with photographs and skin disease severity scores such as the Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI) to assess disease progression (1). Patients should be counseled that CLE can progress to systemic disease. They need monitoring based on the type of CLE, evidence of SLE, and length of stable CLE {Okon, 2013 #7459}. Many patients meet SLE criteria but have predominantly skin findings. These patients need to be reassured that their disease is mild, but they need to be monitored to make sure their disease doesn't progress to organ-threatening disease. This monitoring should be done by dermatologists, and includes CBC, urinalysis, and when indicated, dsDNA and complement levels. This testing should be done at least yearly, even for patients with localized DLE, since they can progress to organ-threatening SLE including glomerulonephritis at any point. Patients with SLE tendencies should be monitored more frequently. Eye exams should be done at the most yearly if patients are on hydroxychloroquine (2). Patients on chloroquine should be monitored every 4–6 months. Patients with evidence of systemic disease may require ongoing care with antimalarials to prevent disease progression (3).

CLE – Estimate of Current Practice

There is no systematic collection of data related to current practice, but many patients are referred to dermatologists at academic centers, frequently by rheumatologists who have been initially referred the patients from dermatologists for their skin-predominant disease. This referral to rheumatologists is not necessary or in the best interest of the patient in terms of managing their CLE. Routine monitoring of labs is not always done when patients are followed by dermatologists for localized disease. Frequently there is no organized method for monitoring disease progression/improvement, and patients do not get systemic therapy when indicated for their CLE.

Lupus Erythematosus – Gaps

CLE does not automatically progress to SLE, but patients need baseline and ongoing monitoring. In addition, objective skin disease monitoring and evaluation of QoL is needed. This can be done by dermatologists, but frequently these patients are referred to rheumatologists when a diagnosis of cutaneous lupus is made prior to any further evaluation or systemic treatment. The practice gaps related to lack of complete work-up for systemic disease and systemic treatment of skin disease includes knowledge and skill gaps that are addressable. There are also attitude gaps, such as a dermatologist's hesitancy to screen for systemic disease and/or objectively assess and manage LE skin disease with systemic agents. As discussed, many dermatologists reflexively refer these patients to a rheumatologist, which is also an attitude gap.

Although recommendations for how to monitor patients with SLE exist, recommendations for monitoring patients with CLE or skin-predominant disease are less clear and must be individualized at the current time based on the severity of disease and systemic findings, with recognition that routine lab monitoring is required for skin-predominant patients (4,5). Treatment recommendations for CLE are eminence-based, rather than Delphi-derived recommendations (5–8). However, increasingly controlled trials or observational studies utilizing the CLASI are now providing evidence for approaches to treatment that can begin to address information and knowledge gaps (9–12). Much work is needed to improve the evidence for current approaches to evaluation and treatment, as well as to develop consensus guidelines that determine “best practices” for CLE.

The barriers related to attitude gaps are best addressed by changes in education of residents and practitioners by increasing interaction with rheumatology, a cultural/attitude change in dermatology related to providing ongoing care for sick patients with skin-predominant CLE, and an awareness of the presence specialized dermatologists in centers that frequently care for such patients where such patients can be referred.

Dermatomyositis – Best Practices

Clinical and Laboratory Assessment—A vision of best practices in the management of dermatomyositis (DM) entails both improving specific knowledge and skills of the average practicing dermatologist as well as the development evidence-based guidelines that will help guide even the most seasoned experts. At a minimum, dermatologists should play the major role in recognition, assessment, interpretation, and management of the cutaneous signs of the disease. Dermatologists should consider the diagnosis in cases not only with classic cutaneous findings but also in those with more atypical presentations that mimic hand dermatitis, psoriasis, or lupus. Dermoscopy should be used regularly to detect periungual telangiectasias, a sensitive marker for dermatomyositis. It should be common knowledge for a dermatologist that a significant proportion of DM patients will never develop detectable muscle inflammation. Our specialty needs to also appreciate the spectrum of histologic changes in DM skin biopsies and not rule out the diagnosis if interface dermatitis is not reported (unpublished data (13)), while at the same time advocating for the importance of biopsies for ruling out mimickers of DM. The dermatologist should understand how to use serologic testing to diagnose DM and order these tests on all patients

with any skin eruption that could be consistent with DM—this includes not depending on a positive ANA for diagnosis as well as routinely ordering the recently described DM-specific autoantibodies (14). All dermatologists should be able to identify typical signs and symptoms associated with active (versus damaged) skin disease in order to optimize therapy and prevent un-necessary use of immunosuppressants.

Screening for Systemic Disease—In a best practice scenario, the dermatologist would always appropriately screen their DM patients for internal malignancy. This includes an understanding of the time (within 1–2 years of first DM symptom) and patient population (clinical and lab risk factors) that are most appropriate to conduct such a search. Dermatologists should understand how advanced age, male sex, cutaneous necrosis, and autoantibodies inform risk for internal malignancy (16). The dermatologist should be prepared to employ not only the history and physical examination and “age appropriate” cancer screening but also more aggressive testing (such as computed tomography or traditional cancer screening tests out of the “appropriate age window”) in order to identify the patients with internal malignancy.

In addition, all dermatologists should at least be able to effectively screen patients for evidence of extracutaneous (systemic) disease in DM. Myositis should be assessed by evaluating not only for weakness but also asking about myalgia, dysphagia, and dysphonia, as well as performing basic muscle strength testing in the clinic. All patients should be regularly screened with muscle enzymes (CK, aldolase, LDH, AST, ALT) every 3–6 months; however, every physician should recognize that significant muscle inflammation can occur even if these tests are normal, and should understand that electromyography and/or magnetic resonance imaging should be performed in this setting in the patient with clinically significant weakness or dysphagia. Dermatologists should regularly (perhaps annually) screen their patients for interstitial lung disease by ordering and interpreting annual pulmonary function testing. They should be able to identify clinical (mechanic hands, Raynaud’s, cutaneous ulcerations) and laboratory (antisynthetase or anti-MDA5 autoantibodies) risk factors for interstitial lung disease, and, in high risk patients, consider more frequent or aggressive screening.

Patient Counseling and Treatment—Optimal patient management by a dermatologist would include both counseling and effective therapy for their skin disease. Patients should be counseled regarding risk of internal malignancy, extracutaneous disease (especially muscle and lung involvement), and risk of specific skin complications such as ulceration and calcinosis. Ideally each provider would use a simple DM-specific skin activity instrument (such as the Cutaneous Dermatomyositis Activity and Severity Index) with their patients in order to quantify skin disease activity and rationally evaluate the treatment regimen (15). Every dermatologist should assess how DM skin disease is impacting patient quality of life (QoL) in order to avoid the pitfall of undertreating skin disease and should appreciate that the topical therapies alone will not be sufficient skin therapy for the majority of patients. Thus, they would be comfortable prescribing and safely monitoring traditional and biologic systemic agents (including antimalarials, methotrexate, mycophenolate mofetil, leflunomide, and IVIG) or, be willing and able to refer patients to local **dermatologists** that can do so.

Dermatologists should take the role as primary provider for those DM patients with skin-predominant disease but understand when referral to other subspecialties (e.g. rheumatology, pulmonology, neurology) is warranted if the patient has evidence of clinically significant extracutaneous disease.

Dermatomyositis – Estimate Current Practice

No data exist to reflect the current practice of how dermatologists manage the DM patient. Based on our experience, many dermatologists do not consider the diagnosis of DM in cases that either do not present with classic clinical (such as Gottron's papules) or histologic (interface dermatitis) changes, and thus patients without obvious muscle weakness may not be diagnosed in a timely manner. These patients are also missed because many dermatologists are not familiar with the fact that DM-specific antibodies have now been described that can characterize 80–90% of all patients. There is a lack of awareness of when and how patients should be evaluated for cancer, as well as lung disease and myositis. Many dermatologists do not know how to interpret autoantibody testing with regards to prognosis and how that can, at a minimum, aid in the counseling of our patients. Many patients are not offered systemic therapy for their skin disease, and, even those with skin predominant disease are often referred to other specialties (especially rheumatologists), when optimal skin care and systemic screening should be performed by the medical dermatologist.

Dermatomyositis Gaps

The gaps in practice that exist in managing DM patients fall into two categories: first, there are gaps that can be addressed via professional education, practice skills enhancement, and attitude; the second type of gap exists due to a paucity of data or commercially available testing to help guide even the most experienced of practitioners. For example, many dermatologists are simply not aware of the clinical and histologic heterogeneity that can be seen in DM. In addition, new information regarding novel antibodies continues to be published at a rapid rate over the past several years. These are quantitative knowledge gaps that are addressable and more accurately represent communication gaps. There are also attitude gaps, such as a dermatologist's hesitancy to screen for systemic disease and/or manage DM skin disease with systemic agents. In addition, many dermatologists reflexively refer these patients to a rheumatologist, as discussed above, which is also an attitude gap. A major practice gap that needs to be closed is the underutilization of autoantibody testing in the clinical practice—this partly stems from a lack of communication to the practicing dermatologist, but also results from unavailability of testing in the clinical setting (see below).

There are several barriers that need to be addressed before these gaps can be closed. First, as a specialty we need to clarify and communicate with our dermatology colleagues what a consensus vision of "best practice" specifically looks like. This would be best achieved by establishing consensus among recognized international dermatology experts regarding cutaneous diagnostic criteria for DM, timing and methodology for screening for malignancy and extracutaneous disease, and a management algorithm for skin disease. Initially these conferences can provide tentative guidelines based on experience and opinion, but ultimately should specifically define what research studies or data are needed to better inform future

guidelines in the future. This would include creating and validating cutaneous diagnostic criteria for DM, identifying which malignancy screening tests should be used for which specific populations, and performing comparative efficacy assessments of systemic therapies for skin disease. One current barrier that exists is the lack of a reliable, quantitative, commercially available manner in which to test a patient for autoantibody against DM-specific targets--these tests are largely described in a research setting, and the few commercially available options have unknown sensitivity and specificity. This will require a large effort, likely international, to harmonize and validate consensus methodologies for each of these antibodies and make them commercially available to the practicing clinician.

Morphea – Best Practices—To date, no best practices have been established for morphea. The Childhood Arthritis and Rheumatology Research Alliance (CARRA) has published a consensus treatment and assessment plan for children with morphea (17). CARRA's hope is that all pediatric rheumatologists will treat and assess childhood morphea the same, which will allow for comparative effectiveness studies and ultimately the establishment of best practices. The Morphea Research Alliance (MRA) is striving to establish consensus treatment and assessment plans for adults with morphea.

The following practices are recommended by experts in the field of morphea. The patient history should document time of onset, rapidity of lesion growth/new lesion onset, possible triggers (trauma, radiation), symptoms associated with the morphea lesions, the impact of the disease on quality of life, a history assessing for other co-morbid autoimmune diseases and a family history assessing for autoimmunity.

A full body skin exam (including the oral and genital mucosa given the co-occurrence of mucosal involvement) of patients with morphea is helpful to assess the extent of involvement and to categorize the patient's morphea subtype (circumscribed, generalized, linear, mixed, pansclerotic) (18) and depth of involvement (dermal, deep dermal, subcutaneous fat, muscle, bone). The subtype of morphea and depth of involvement are important factors in therapeutic decisions and prognosis counseling. The physical exam, along with history, can be used to establish whether the disease is in the active phase (i.e. the patient is developing new morphea lesions, having expansion of existing lesions, or has erythema or violaceous discoloration surrounding the lesions) (19). Active disease is treatable with immunosuppression. Morphea in the damage phase (stable number of plaques, no expansion of the plaques, hyperpigmented or sclerotic plaques without signs of inflammation) will not improve with immunosuppression. Physical exam should also be used to assess for range of motion in all involved joints. Any limitations should be documented for clinical monitoring and patients with range of motion limitations referred to physical therapy and/or occupational therapy. Physical exam should also be used to help rule out systemic sclerosis (sclerodactyly, nailfold capillary changes, and Raynaud's phenomenon are early signs of systemic sclerosis that are not present in patients with morphea).

There are no autoantibodies that are specific to morphea and therefore autoantibody tests are not helpful in the diagnosis or prognosis of morphea. Assessment of autoantibodies specific to systemic sclerosis (anti-centromere, anti-topoisomerase, anti RNA polymerase III) is unnecessary in the evaluation of a patient with morphea. The diagnosis of systemic sclerosis

requires a combination of physical exam findings, Raynaud's phenomenon and autoantibodies. Positive systemic sclerosis specific antibodies in isolation (i.e. in a patient without sclerodactyly, Raynaud's phenomenon, and nailfold capillary changes) are not helpful diagnostically and are most likely to be false positive results. Biopsy is not required to make a diagnosis of classic morphea; however, biopsy may be helpful in non-traditional presentations.

Objective measurements of disease activity (full body photographs or body maps with locations and measurements, or the Localized Scleroderma Assessment Tool (LoSCAT) (19)) should occur at each visit, to monitor response to therapy. Children with head and neck morphea should be evaluated by ophthalmology to assess for asymptomatic ocular involvement (which is treatable and may prevent vision loss) (20). Although linear morphea of the head and neck has been associated with involvement of the central nervous system, MRI findings have not been correlated with symptoms and signs of CNS involvement (21). Therefore, only children with signs or symptoms of CNS involvement should undergo MRI.

Morphea treatment decisions need to take into account the disease subtype, depth of involvement, and quality of life impairments. In general, patients with active deep linear morphea, active deep mixed variant morphea, active deep generalized morphea and active pansclerotic morphea should be treated with systemic steroids (a 3 month taper) and methotrexate (ideally for a duration of 2 years) in an attempt to prevent new lesion formation and functional impairment (22–25). Patients with active superficial circumscribed morphea may be treated with topical therapy or localized phototherapy (24,25). Patients with active superficial generalized morphea may be treated with topical therapy, phototherapy or a combination of systemic steroids and methotrexate, based on patient preference and comorbidities (24,25). Patients with morphea should also be counseled that treatment is not always necessary, particularly for asymptomatic disease that is not causing or at risk of causing functional limitations. Patients with morphea in the non-active phase should be educated on signs of disease recurrence and offered clinical monitoring.

Lastly, but perhaps most importantly, patients need to leave their dermatologist's office understanding that morphea is not systemic sclerosis and will not increase their chances of developing systemic sclerosis in the future. Patient educational materials on morphea may be found through JAMA Dermatology (27).

Morphea – Estimate Current Practice—The Morphea Research Alliance (MRA) has surveyed the Medical Dermatology Society (MDS) and Rheumatologic Dermatology Society (RDS) on their current practices in regards to the evaluation and treatment of morphea (unpublished data). History, physical exam, and referrals to PT and OT were regularly performed. A majority of respondents ordered autoantibodies in the evaluation of morphea, particularly anti-Scl70 and anti-centromere antibodies (antibodies specific for systemic sclerosis). Respondents did not routinely have an organized and objective way of monitoring disease activity and response to therapy. Topical steroids are being routinely prescribed for subtypes of morphea that are unlikely to respond to topical therapy (linear disease, deep generalized disease). Methotrexate is not routinely prescribed for those patients with active linear morphea.

Additionally, it is estimated that very few clinicians assess quality of life in their morphea patients, perform exams of the oral and genital mucosa (28), rigorously assess for range of motion limitations, nor educate patients on the difference between morphea and systemic sclerosis. Patients in the damage (i.e. non-active) phase of morphea may also be unnecessarily treated with immunosuppressants.

Morphea Gaps—The gaps that exist in the care of a patient with morphea result from lack of knowledge, specific practice patterns, the need for development of new skills, and the need for a change in attitudes. Development of defined best practices and guidelines for the evaluation and treatment of patients with morphea would help to close many of our knowledge, skill, specific practice and attitude gaps. Lack of defined best practices contributes to the overuse of laboratory testing for autoantibodies, the lack of patient education on the differences between morphea and systemic sclerosis, the lack of objective disease monitoring, the lack of aggressive treatment for morphea that impairs quality of life and physical functioning, the lack of quantification of quality of life issues in clinical practice, the lack of oral and genital mucosal examination to exclude mucosal involvement, and the inappropriate treatment of patients with morphea in the damage phase with immunosuppressant medications.

There are many barriers to developing best practices. The first step in developing guidelines for the assessment and treatment of a condition is to describe the current practices. Revealing how physicians are treating a rare condition to other physicians establishes treatment norms and therefore consensus. Consensus can then be used to establish guidelines. When studying rare diseases, data needs to be pooled across practices in a standardized fashion to enable comparative effectiveness studies, which can then be used to establish best practices. Establishing best practices for a rare disease requires an international effort, a lot of protected time and funding.

Part 2

Best Practices in Medical Education

Residents are adult learners. Evidence has shown that adults learn best when the learning experience is interactive, is pertinent to the skills they need to acquire to best perform their jobs, allows for the ability to practice the skills that they learn, and provides constructive feedback on their performance (29). Additional research on the science of learning has elucidated several strategies for learners and educators to use to maximize knowledge acquisition and application. These include testing and retesting (30,31), interspersed practice and spaced retrieval (31–33), reflection and metacognition (34–37), providing and receiving corrective feedback (29,38) and cultivating a growth mindset (39).

Best practices in educating our residents on the diagnosis and management of dermatomyositis, cutaneous lupus erythematosus and morphea rely on involving residents in multi-disciplinary and sub-specialty clinics devoted to the care of patients with these relatively rare conditions. Knowledge retention is increased by making the resident an integral team member in the evaluation and treatment of these patients, and an active participant in their learning. Residents should be involved in reviewing outside records,

compiling outside clinical data, interviewing and examining the patient, efficiently presenting the patient by including pertinent information and leaving out extraneous information, and developing a differential diagnosis with supporting and refuting facts for each diagnosis. Prior to presenting the patient, the resident should also develop a plan for additional work up with reasoning as to why each test is important and how it will impact care, develop a plan for treatment with reasoning as to why two to three additional treatment options are less desirable, develop a plan for monitoring for medication toxicities and deciding on an appropriate follow up interval. If knowledge or skill gaps are revealed during a patient encounter, the resident should be involved in developing a plan for the acquisition of any knowledge or skill that was lacking. Knowledge retention is also increased by taking time at the end of clinic to reflect with the residents on what went well, what they can improve upon and their plan for improvement.

Ideally residents would rotate through multiple sub-specialty clinics over months (i.e. specific days assigned for each sub-specialty clinic over months, instead of working in one sub-specialty clinic for one month), to increase spaced retrieval. Studies have shown that massed practice (i.e. rotating within the same sub-specialty clinic for a month) increases the *perception* of knowledge gained, but that spaced retrieval (i.e. rotating in a sub-specialty clinic one day a week over a long period of time) increases the actual knowledge gained (32,33).

Prior to didactic sessions, educational materials should be shared with learners to help them to prepare for the session. Didactic sessions on dermatomyositis, cutaneous lupus erythematosus, and morphea should start with pre-tests, include interactive cases that allow for dialog and working through problems, and a posttest.

Estimate Current Practice—The vast majority of resident education occurs during clinical encounters and didactics. Clinical teaching tools, such as the 1-minute preceptor and reflective questioning, are likely not used with regularity. Feedback on resident performance in clinic may be infrequent or inadequate (40,41). Residents and faculty may approach clinical reasoning with an entity theory (i.e. believing that intelligence and the ability to clinically reason is a fixed characteristic), and not with a growth mindset (i.e. realizing that knowledge and clinical reasoning improve with effort and practice), limiting knowledge acquisition and growth (39). Didactics are generally hour long, noninteractive sessions.

The gaps that exist in cutaneous lupus erythematosus, dermatomyositis, and morphea resident education are largely rooted in knowledge, interpersonal communication and professionalism. Resident education in the care of cutaneous lupus erythematosus, dermatomyositis, and morphea is lacking in adult learning theory and effective corrective feedback. Additionally, the rarity of these conditions and the rarity of sub-specialty clinics devoted to the care of these patients results in infrequent patient care opportunities for residents.

Strategies to increase the efficacy of teaching and learning about cutaneous lupus erythematosus, dermatomyositis, and morphea include incorporating adult learning theory into medical education and increasing effective feedback. Faculty may need educational

sessions through their program director, their local graduate medical education department, the AAD or the ACGME. Additionally, these resources may also be used by faculty to sharpen feedback skills. Residents may be afforded the opportunity to rotate at centers with dedicated sub-specialty clinics, or offered rotations with other medical specialists who care for these patients. Simulated patients and on-line teaching may be an avenue to increase exposure to these conditions over time (i.e. increasing delayed retrieval). Lastly, an increase awareness of combined internal medicine-dermatology training and med-derm fellowships may create more experts in these fields to teach in dermatology at academic centers.

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Key Points

- Patients with skin-predominant lupus erythematosus, dermatomyositis, and morphea should be evaluated, treated, and followed by dermatologists who can take primary responsibility for their care
- Many academic centers have specialized centers with dermatologists who care for these patients
- Patients with skin-predominant lupus erythematosus should be followed regularly with labs, including urinalysis, to detect significant systemic disease.
- Patients with dermatomyositis should be appropriately evaluated for the presence of lung, muscle disease, and cancer. Antibody tests can help determine the risks for individual patients.
- Patients with morphea rarely progress to systemic disease, but therapies can be very helpful in treating and preventing progression of disease.