International Journal of Women's Dermatology 7 (2021) 583–587

Contents lists available at ScienceDirect

International Journal of Women's Dermatology

Review

Importance of collaboration of dermatology and rheumatology to advance the field for lupus and dermatomyositis ☆,☆☆,★

Victoria P. Werth MD [a,b,*], Anca D. Askanase MD [c], Ingrid E. Lundberg MD [d]

[a] Department of Dermatology, University of Pennsylvania, Philadelphia, Pennsylvania
[b] Medical Research, Corporal Michael J. Crescenz VA Medical Center, Philadelphia, Pennsylvania
[c] Division of Rheumatology, Columbia University Irving Medical Center, New York, New York
[d] Division of Rheumatology, Department of Medicine, Solna, Karolinska Institutet and Rheumatology, Karolinska University Hospital, Stockholm, Sweden

ARTICLE INFO

Article history:
Received 21 July 2021
Revised 12 September 2021
Accepted 13 September 2021

Keywords:
Cutaneous lupus erythematosus
systemic lupus erythematosus
dermatomyositis
multidisciplinary collaboration

ABSTRACT

There have been a number of advances in the clinical and translational understanding of cutaneous lupus and dermatomyositis, which both disproportionately affect women. These advances have involved ongoing collaborations between dermatology and rheumatology that highlight the importance of the skin in these disorders, with improvement in the education of trainees and clinical management of these complex multisystem diseases. In addition, a new disease classification has allowed inclusion of patients with skin-predominant dermatomyositis, frequently associated with systemic findings, in the spectrum of idiopathic inflammatory myopathies. Validated outcome measures allow translational research and facilitate progress toward better and more targeted therapeutics. Clinical trials using disease severity tools, such as the Cutaneous Lupus Erythematosus Area and Severity Index and the Cutaneous Dermatomyositis Disease Area and Severity Index, allow measurement of improvement in the skin. Recent results of phase 2 and 3 trials clearly show that patients will benefit from collaborative interactions and studies between dermatology and rheumatology.

Contents

☆ Conflicts of Interest: Dr Victoria P. Werth has received research grants from Pfizer, Corbus, Celgene, Amgen, Janssen, Biogen, Gilead, and Viela and has served as a consultant for Pfizer, Janssen, Neovacs, Idera, Octapharma, CSL Behring, Corbus, AstraZeneca, Pfizer, Biogen, Celgene, Resolve, Janssen, Gilead, Lilly, BMS, Nektar, Abbvie, Akira, Viela, GSK, EMD Serona, and Sanofi. She has also developed and validated outcome measures for cutaneous lupus (Cutaneous Lupus Erythematosus Area and Severity Index) and dermatomyositis (Cutaneous Dermatomyositis Disease Area and Severity Index). Dr Anca D. Askanase has served as an investigator/consultant for Abbvie, Amgen, AstraZeneca, Aurinia, BMS, Celgene, Eli Lilly, Idorsia, Janssen, Genentech, GSK, Mallinckrodt, Pfizer, and UCB. Dr Ingrid E. Lundberg has received research grants from AstraZeneca and served as consultant for Argenx, Corbus Pharmaceuticals Inc., Janssen, Kezaar, Octapharma, Orphazyme, EMD Serono Research & Development Institute, and Sobi. Dr. Lundberg also holds stock shares in Roche and Novartis.

☆☆ Funding: This project is supported by the Department of Veterans Affairs Veterans Health Administration, Office of Research and Development, Biomedical Laboratory Research and Development, National Institutes of Health (National Institute of Arthritis and Musculoskeletal and Skin Diseases) to Dr Victoria P. Werth (numbers R01AR071653 and R01AR076766). Dr Ingrid E. Lundberg received funding from the Swedish Research Council (Vetenskapsrådet), King Gustaf V 80 Year Foundation (Stiftelsen Konung Gustaf V:s 80-årsfond), Swedish Rheumatism Association (Reumatikerförbundet), and Region Stockholm (ALF project).

★ Study approval: The author(s) confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies.

* Corresponding Author:
E-mail address: werth@pennmedicine.upenn.edu (V.P. Werth).

https://doi.org/10.1016/j.ijwd.2021.09.002

What is known about this subject in regard to women and their families?

- Overall, patients with cutaneous lupus erythematosus, with or without systemic lupus, as well as those with dermatomyositis, demonstrate negative impacts on both emotional and symptom components of quality of life. Women are predominantly affected by virtue of getting these diseases more frequently.
- The families of patients with these autoimmune diseases are impacted by the severe effects of the disease, the medication regimens that suppress immunity, and the cost of the diseases in terms of ability to work and cost of medications.

What is new from this article as messages for women and their families?

- Interactions between medical specialties are critical to advance both the scientific understanding of and therapeutic options for autoimmune diseases such as lupus erythematosus and dermatomyositis.

Introduction

Lupus erythematosus (LE) and dermatomyositis (DM) are spectrum diseases that frequently affect women. With regard to cutaneous manifestations, there are patients with skin-predominant disease and those with systemic disease and minimal or no skin findings. The identification of patients with primarily skin autoimmune disease frequently requires collaboration between dermatologists and rheumatologists. Often, patients who present to dermatologists have somewhat different manifestations than those seeing rheumatologists. Only when both fields work together does the bigger picture of the disease emerge. These collaborations allow for the optimal development of disease diagnostic and classification criteria, agreement on how to measure outcomes, as well as the design, performance, and interpretation of clinical studies, and in the clinical arena assist in the diagnosis and management of various disease manifestations. The clinic and education, as well as the basic, translational, and clinical research needs in these multisystem diseases, benefit from close communication and collaboration.

The concepts described here for LE and DM apply equally to other conditions, including scleroderma/morphea, vasculitis, Behcet's disease, Sjogren's, and psoriasis. We will review how collaborations have benefitted progress in both LE and DM, with the goal to further expand these initiatives in the future.

Lupus erythematosus

Lupus criteria

Defining a disease is fundamental to clinical management and can assist in case selection for studies. In the case of systemic LE (SLE), there have been several important initiatives to define SLE. The components of the 1997 American College of Rheumatology (ACR) criteria, *Systemic Lupus International Collaborating Clinics* (SLICC), and the newest 2019 European League against Rheumatism (EULAR)/ACR criteria evolved to identify those patients with sufficient manifestations of lupus as having SLE. The initial 1997 ACR criteria included four skin symptoms that were somewhat overlapping and allowed patients with primarily skin disease to easily meet the criteria for SLE (Albrecht et al., 2004). The SLICC group thoughtfully included dermatologists in the next iteration of SLE criteria, to become known as the SLICC criteria (Petri et al., 2012). These were developed through strong collaboration between dermatology and rheumatology, included more cutaneous LE (CLE) subtypes, and removed photosensitivity, which is a very nonspecific finding in other skin diseases (Petri et al., 2012). Although the inclusion of alopecia was an area of discordance between the dermatologists and rheumatologists involved in developing these criteria, it began a series of essential dialogues. The new EULAR/ACR criteria lacked dermatology input, and requiring antinuclear antibody positivity excluded 7.5% of patients with CLE previously diagnosed with SLE, some of whom had inflammatory arthritis, cytopenias, and/or proteinuria (Tarazi et al., 2019).

Education and clinical management (local, national, and international)

Many dermatology departments and rheumatology divisions hold joint conferences for their faculty and trainees. Combined rheumatology–dermatology clinics with both dermatologists and rheumatologists in attendance and clinics where trainees in one discipline attend clinics held by the other discipline are more and more common. This cross-fertilization is also accomplished through combined internal medicine–dermatology residency programs, as well as for rheumatology fellows rotating in rheumatology–dermatology clinics, where cross-training facilitates cohesive educational opportunities. At the national and international level, there are many conferences, such as the Rheumatologic Dermatology Society where dermatologists and rheumatologists give lectures on relevant topics for dermatologists managing patients with SLE and DM, among other rheumatologic conditions (Sontheimer et al., 2019). Similarly, dermatologists routinely participate in sessions at ACR and regional rheumatology meetings. A yearly meeting for rheumatology fellows, the ACR-sponsored State of the Art symposium, frequently includes dermatology presentations.

Measuring outcomes in lupus erythematosus

With a better understanding of the pathogenesis of SLE and CLE, a need to measure meaningful improvement or worsening of disease activity became important. There have been many initiatives involving rheumatology and dermatology to discuss how to define lupus disease activity, severity, flares (Ruperto et al., 2011), remission (van Vollenhoven et al., 2017), and study design. This collaborative approach has led to important advances in measuring the skin as an outcome in both CLE and SLE.

The development of the Cutaneous Lupus Erythematosus Area and Severity Index (CLASI) included input from the dermatology and rheumatology community, as well as patients. This outcome instrument measures disease activity and damage separately, with a focus on evaluating skin body areas prone to lupus skin lesions. Activity is measured as erythema and scale, mucosal lesions, and inflammatory alopecia. Damage evaluates dyspigmentation and scarring. Inter- and intrarater validation studies showed excellent results, and further validation showed a correlation between improvement of activity and improvement in quality of life, as well as biomarkers (e.g., MxA, a type I IFN-upregulated protein; Albrecht et al., 2005; Furie et al., 2019). Measuring skin disease severity in CLE and SLE included validation studies comparing the results of evaluating disease severity in the same patients by both adult dermatologists and rheumatologists (Krathen et al., 2008), as well as pediatric dermatologists and rheumatologists (Kushner et al., 2019). This extension of validation to both the dermatology and rheumatology communities has allowed skin manifestations of lupus to become more clearly defined as outcomes included in lupus trials; this is particularly important because skin manifestations are present in approximately 50% of patients with lupus in clinical trials. The conspicuousness of skin improvement and the option of including high-quality photographs in studies facilitates their use in proof-of-concept trials for novel therapeutic approaches and can often capture meaningful changes with a shorter timeline (Furie et al., 2019; Karnell, 2021).

Additional interactions with the U.S. Food and Drug Administration in collaboration with the Lupus Foundation have led to a consensus among dermatologists, rheumatologists, and nephrologists about how best to measure outcomes and improve the design of studies that affect patients with CLE, with or without SLE (Merrill et al., 2018). The international community of dermatologists and rheumatologists has worked with EULAR to provide recommendations for monitoring patients with SLE in clinical practice and in observational studies (Mosca et al., 2010; 2011).

Clinical studies in lupus erythematosus

The renewed interest in LE has led to many collaborative studies, promising new therapies, and many novel therapeutics in development. There are more frequent interactions between rheumatologists and dermatologists as studies are planned and performed, data discussed, results presented, and publications prepared. The synergism between dermatology and rheumatology is clearly important and leading to therapeutic advances in the field (Furie et al., 2017; 2019; Karnell, 2021; Khamashta et al., 2016; Presto et al., 2018; van Vollenhoven et al., 2018; Werth, 2021; Werth et al., 2017). The community of pediatric rheumatologists and dermatologists, working through the Childhood Arthritis and Rheumatology Research Alliance, has annual meetings that provide opportunities to launch clinical research studies looking at pediatric-onset discoid LE, as well as differences in work-up and therapy between the two pediatric specialties (Arkin et al., 2015; 2019). This rich interaction highlights both differences in patients presenting to the two specialties and differences in evaluation and treatment. Such interactions are important to begin to understand how to design better studies and improve outcomes.

Research meetings

The collaboration between dermatology and rheumatology in hosting research meetings has facilitated interactions that further contribute to advancements in the field. This is prominently displayed at the Society of Investigative Dermatology, ACR, and EULAR meetings, where both dermatologists and rheumatologists present their work on an annual basis. Dermatologists and rheumatologists with interest in dermatology–rheumatology have held four international meetings planned around the International Investigative Dermatology meeting at 4- to 5-year intervals. This international conference on CLE, which provides a venue for collaboration between the international dermatology and rheumatology communities, has led to several ongoing collaborative projects (Concha et al., 2019; Schultz et al., 2015). There are frequent meetings of the international lupus community that include rheumatologists and dermatologists, such as Lupus 21st Century, Skin Rheumatism and Autoimmunity, and the International Congress on SLE, which are moving the field forward.

Diseased-focused foundations

Groups such as the Lupus Foundation of America and Lupus Research Alliance recognize the multidisciplinary approach required for the management and advancement of clinical and translational research in lupus. They routinely involve dermatology and rheumatology in many aspects of lupus research, in patient educational programs, and in the advocacy efforts they lead (Tse et al., 2021).

Grants

The Lupus Foundation of America, Lupus Research Alliance, and Rheumatology Research Foundation, in addition to the National Institutes of Health (National Institute of Arthritis and Musculoskeletal and Skin Diseases) and U.S. Department of Defense, have all sponsored collaborative research conferences and projects that have helped advance lupus research.

Dermatomyositis

DM, like lupus, is a disorder that is more common in women than in men, and similar to lupus, management of patients with DM benefits from collaborative clinics. This is also true for education and research as described above. Many centers worldwide have combined dermatology–rheumatology clinics where patients with skin diseases are assessed, and these clinics are also excellent educational platforms for rheumatologists and dermatologists in training. Notably, patients with DM with mild muscle involvement, including those with amyopathic or hypomyopathic DM, may still develop severe and life-threatening interstitial lung disease or have an underlying malignant, cancer-associated DM.

Dermatomyositis criteria

The Bohan and Peter DM criteria have been the classification criteria for DM for >4 decades. These criteria require patients with myositis to be defined as possibly having DM (Bohan and Peter, 1975a; 1975b). Dermatologists have developed their own nomenclature to define DM in patients who do not have muscle disease: sine myositis (Euwer and Sontheimer, 1991; Ghazi et al., 2013; Sontheimer, 2002). Patients with amyopathic DM develop interstitial lung disease at the same rate as those with classic DM (George et al., 2017; Morganroth et al., 2010).

Dr Ingrid Lundberg led a EULAR/ACR effort to redefine idiopathic inflammatory myopathies (IIMs) including the subgroup. Dr Victoria Werth joined the steering committee, and it quickly became apparent that patients with amyopathic DM were rarely seen by the rheumatologists on the steering committee. The interdisciplinary interaction was pivotal in recognizing the differences between patients who presented with DM to dermatology versus rheumatology. These initial conversations prompted a study to determine the number of patients with DM who initially presented to dermatology relative to rheumatology at the University of Pennsylvania over a 3-year period. The results were somewhat surprising: Only one patient with amyopathic DM presented to rheumatology, compared with 33 to dermatology (Quain et al., 2007), suggesting that either patients with amyopathic DM were presenting primarily to dermatology or were misdiagnosed. The different perceptions of the prevalence of amyopathic DM between dermatologists and rheumatologists is likely due to both of these potential reasons. A more recent study showed that just 44% of patients with DM were diagnosed as such prior to referral, and patients with DM were frequently classified as having SLE, CLE, or undifferentiated connective tissue disease (Da Silva et al., 2018).

The importance of classification criteria, frequently used as diagnostic criteria, in determining who has DM has profound implications for patients, who can go as long as 10 to 15 years before receiving their correct diagnosis of DM. The leadership of Dr. Lundberg in bringing together a multidisciplinary team and discussing different perspectives of the disease was critical in advancing to more all-encompassing classification criteria for IIM, including DM (Lundberg et al., 2017; 2018). These new EULAR/ACR DM criteria allow the classification of 74% of patients with amyopathic DM as being on the spectrum of DM (Patel et al., 2018), and further efforts to refine the skin criteria, in collaboration with the International Myositis Classification Criteria Project group led by Dr Lundberg, are ongoing (Concha et al., 2019; 2020). Of note, these collaborative efforts between dermatology and rheumatology have been ongoing for more than a decade, and the results are critical to correct classification of patients and the inclusion of patients with amyopathic DM in clinical and translational studies.

Measuring outcomes in dermatomyositis

Similar to what has occurred in lupus, there have been efforts to develop outcomes measures for DM that can be used to evaluate disease activity and response to therapy. The International Myositis Assessment and Clinical Studies (IMACS) Group has yearly multidisciplinary conferences around the ACR meeting and includes dermatologists and rheumatologists, among other subspecialties. IMACS has facilitated the collaborative development of criteria for minimal, moderate, and major clinical response in adult DM and polymyositis (Aggarwal et al., 2017). A recent review of outcome measures for DM included multiple specialties (Rider et al., 2011). Measuring skin disease activity in adult and juvenile DM included validation studies of the Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI), with participation of both adult and pediatric dermatologists, rheumatologists, and neurologists (Tiao et al., 2017a; 2017b). Additional multidisciplinary efforts to define antisynthetase syndrome, another subgroup of IIM where skin manifestations may be present, are ongoing. A recent meta-analysis of approaches for cancer screening in DM was published, certainly a problem that crosses between fields that manage DM (Oldroyd et al., 2021).

Clinical studies in dermatomyositis

As with lupus, the expanding interest in DM has led to collaborative studies and therapeutic advances. The opportunities for interactions between dermatology and rheumatology are expanding. A recent successful phase 2 trial of lenabasum, a nonpsychoactive CB2 agonist, for skin predominant DM (Werth et al., 2018) led to a phase 3 trial for classic DM that was recently completed. The ability to measure skin outcomes in classic DM is critical to demonstrate efficacy in such trials. A recent therapeutic trial in DM used the CDASI, a validated skin disease severity tool, to evaluate the skin (Aggarwal et al., 2020). There is an ongoing phase 2 study of anti-interferon-β antibody that initially targeted skin disease in DM and has now expanded to include classic myositis with active muscle disease.

Research meetings

Dermatologists and rheumatologists work collaboratively in a number of research organizations, including IMACS and the Global Conference on Myositis, and these have been important opportunities to bring research communities together to present work and advance the field.

Disease-focused foundations

The Myositis Association includes a multidisciplinary medical advisory board composed of dermatologists, rheumatologists, and neurologists (another important discipline in the management of patients with DM). The Myositis Association provides patient support and educational meetings, as well as funds research projects in DM. Such disease-focused organizations are vital for the advancement of science and care in the field and assist patients in navigating through an often confusing and debilitating chronic disease.

Conclusions

For patients with systemic inflammatory diseases, such as SLE or DM, multidisciplinary team management is critical to improve diagnosis, optimize management, and improve outcomes. Rheumatologists and dermatologists who specialize in these disorders, neurologists, pulmonologists, pathologists, physical and occupational therapists, cardiologists, nephrologists, and ophthalmologists need to be an integral part of the care team.

References

Aggarwal R, Charles-Schoeman CC, Schessl J, Bata-Csorgo Z, Dimachkie MM, Griger Z, et al. Efficacy and safety of IVIg (octagam 10%) in patients with active dermatomyositis. Results of a randomized, double-blind, placebo-controlled phase III trial (ProDERM study). Arthritis Rheumatol 2020;72(suppl 10) abstract 955.

Aggarwal R, Rider LG, Ruperto N, Bayat N, Erman B, Feldman BM, et al. 2016 American College of Rheumatology/European League Against Rheumatism criteria for minimal, moderate, and major clinical response in adult dermatomyositis and polymyositis: An International Myositis Assessment and Clinical Studies Group/Paediatric Rheumatology International Trials Organisation collaborative initiative. Ann Rheumat Dis 2017;76:792–801.

Albrecht JA, Braverman IM, Callen JP, Connolly MK, Costner M, Fivenson D, et al. Dermatology position paper on the revision of the 1982 ACR criteria for SLE. Lupus 2004;13:839.

Albrecht J, Taylor L, Berlin JA, Dulay S, Ang G, Fakhazardeh S, et al. The CLASI (Cutaneous LE Disease Area and Severity Index): An outcome instrument for cutaneous lupus erythematosus. J Invest Dermatol 2005;125:889–94.

Arkin L, Buhr K, Brandling-Bennett H, Chiu Y, Chong B, Curran M, et al. Practice-based differences in paediatric discoid lupus erythematosus. Br J Dermatol 2019;181:805–10.

Arkin LM, Ansell L, Rademaker A, Curran ML, Miller ML, Wagner A, et al. The natural history of pediatric-onset discoid lupus erythematosus. J Am Acad Dermatol 2015;72:628–33.

Bohan A, Peter JB. Polymyositis and dermatomyositis (first of two parts). N Engl J Med 1975a;292(7):344–7.

Bohan A, Peter JB. Polymyositis and dermatomyositis (second of two parts). N Engl J Med 1975b;292(8):403–7.

Concha JSS, Patsatsi A, Marshak-Rothstein A, Liu ML, Sinha AA, Lee LA, et al. Advances in cutaneous lupus erythematosus and dermatomyositis: A report from the 4th International Conference on Cutaneous Lupus Erythematosus-An ongoing need for international consensus and collaborations. J Invest Dermatol 2019;139(2):270–6.

Concha JSS, Pena S, Gaffney RG, Patel B, Tarazi M, Kushner CJ, et al. Developing classification criteria for skin-predominant dermatomyositis. Br J Dermatol 2020;182:410–17.

Concha JSS, Tarazi M, Kushner CJ, Gaffney RG, Werth VP. The diagnosis and classification of amyopathic dermatomyositis: A historical review and assessment of existing criteria. Br J Dermatol 2019;180:1001–8.

Da Silva DM, Patel B, Werth VP. Dermatomyositis: A diagnostic dilemma. J Am Acad Dermatol 2018;79(2):371–3.

Euwer R, Sontheimer R. Amyopathic dermatomyositis (dermatomyositis sine myositis). Presentation of six new cases and review of the literature. J Am Acad Dermatol 1991;24:959.

Furie R, Khamashta M, Merrill JT, Werth VP, Kalunian K, Brohawn P, et al. Anifrolumab, an anti-interferon-alpha receptor monoclonal antibody, in moderate-to–severe systemic lupus erythematosus. Arthritis Rheumatol 2017;69(2):376–86.

Furie R, Werth VP, Merola JF, Stevenson L, Reynolds TL, Naik H, et al. Monoclonal antibody targeting BDCA2 ameliorates skin lesions in systemic lupus erythematosus. J Clin Invest 2019;129:1359–71.

George MD, Shah R, Kreider M, Miller Jr WT, Merkel PA, Werth VP. Pulmonary function tests, interstitial lung disease and lung function decline in outpatients with classic and clinically amyopathic dermatomyositis. Br J Dermatol 2017;176(1):262–4.

Ghazi E, Sontheimer RD, Werth VP. The importance of including amyopathic dermatomyositis in the idiopathic inflammatory myositis spectrum. Clin Exp Rheumatol 2013;31(1):128–34.

Karnell JL, Wu Y, Mittereder N, Smith MA, Gunsior M, Yan L, et al. Depleting plasmacytoid dendritic cells reduces local type I interferon responses and disease activity in patients with cutaneous lupus. Sci Transl Med 2021;13(595):eabf8442.

Khamashta M, Merrill JT, Werth VP, Furie R, Kalunian K, Illei GG, et al. Sifalimumab, an anti-interferon-alpha monoclonal antibody, in moderate to severe systemic lupus erythematosus: A randomised, double-blind, placebo-controlled study. Ann Rheumat Dis 2016;75:1899–901.

Krathen MS, Dunham J, Gaines E, Junkins-Hopkins J, Kim E, Kolasinski SL, et al. The Cutaneous Lupus Erythematosus Disease Activity and Severity Index: Expansion for rheumatology and dermatology. Arthritis Rheum 2008;59(3):338–44.

Kushner CJ, Tarazi M, Gaffney RG, Feng R, Ardalan K, Brandling-Bennett HA, et al. Evaluation of the reliability and validity of the Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI) in paediatric cutaneous lupus among paediatric dermatologists and rheumatologists. Br J Dermatol 2019;180(1):165–71.

Lundberg IE, de Visser M, Werth VP. Classification of myositis. Nat Rev Rheumatol 2018;14(5):269–78.

Lundberg IE, Tjarnlund A, Bottai M, Werth VP, Pilkington C, de Visser M, et al. 2017 European League Against Rheumatism/American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups. Arthritis Rheumatol 2017;69(12):2271–82.

Merrill JT, Manzi S, Aranow C, Askenase A, Bruce I, Chakravarty E, et al. Lupus community panel proposals for optimising clinical trials: 2018. Lupus Sci Med 2018;5(1).

Morganroth PA, Kreider ME, Okawa J, Taylor L, Werth VP. Interstitial lung disease in classic and clinically amyopathic dermatomyositis: A retrospective study with screening recommendations. Arch Dermatol 2010;146:729–38.

Mosca M, Tani C, Aringer M, Bombardieri S, Boumpas D, Brey R, et al. EULAR recommendations for monitoring systemic lupus erythematosus patients in clinical practice and in observational studies. Ann Rheumat Dis 2010;69(7):1269–74.

Mosca M, Tani C, Aringer M, Bombardieri S, Boumpas D, Cervera R, et al. Development of quality indicators to evaluate the monitoring of SLE patients in routine clinical practice. Autoimmun Rev 2011;10(7):383–8.

Oldroyd AGS, Allard AB, Callen JP, Chinoy H, Chung L, Fiorentino D, et al. A systematic review and meta-analysis to inform cancer screening guidelines in idiopathic inflammatory myopathies. Rheumatology (Oxford) 2021;60:2615–28.

Patel B, Khan N, Werth VP. Applicability of EULAR/ACR classification criteria for dermatomyositis to amyopathic disease. J Am Acad Dermatol 2018;79(1) 77–83.e1.

Petri M, Orbai AM, Alarcon GS, Gordon C, Merrill JT, Fortin PR, et al. Derivation and validation of systemic lupus international collaborating clinics classification criteria for systemic lupus erythematosus. Arthritis Rheum 2012;64(8):2677–86.

Presto JK, Okon LG, Feng R, Wallace DJ, Furie R, Fiorentino D, et al. Computerized planimetry to assess clinical responsiveness in a phase II randomized trial of topical R333 for discoid lupus erythematosus. Br J Dermatol 2018;178(6):1308–14.

Quain RD, Teal V, Taylor L, Werth VP. Number, characteristics, & classification of dermatomyositis patients seen by dermatology & rheumatology at a tertiary medical center. J Am Acad Dermatol 2007;57:937.

Rider LG, Werth VP, Huber AM, Alexanderson H, Rao AP, Ruperto N, et al. Measures of adult and juvenile dermatomyositis, polymyositis, and inclusion body myositis: Physician and Patient/Parent Global Activity, Manual Muscle Testing (MMT), Health Assessment Questionnaire (HAQ)/Childhood Health Assessment Questionnaire (C-HAQ), Childhood Myositis Assessment Scale (CMAS), Myositis Disease Activity Assessment Tool (MDAAT), Disease Activity Score (DAS), Short Form 36 (SF-36), Child Health Questionnaire (CHQ), physician global damage, Myositis Damage Index (MDI), Quantitative Muscle Testing (QMT), Myositis Functional Index-2 (FI-2), Myositis Activities Profile (MAP), Inclusion Body Myositis Functional Rating Scale (IBMFRS), Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI), Cutaneous Assessment Tool (CAT), Dermatomyositis Skin Severity Index (DSSI), Skindex, and Dermatology Life Quality Index (DLQI). Arthritis Care Res (Hoboken) 2011;63(Suppl 11):S118–57.

Ruperto N, Hanrahan LM, Alarcon GS, Belmont M, Brunetta P, Buyon JP, et al. International consensus to define disease flare in lupus. Lupus 2011;20(5):453–62.

Schultz HY, Dutz JP, Furukawa F, Goodfield MJ, Kuhn A, Lee LA, et al. From pathogenesis, epidemiology, and genetics to definitions, diagnosis, and treatments of cutaneous lupus erythematosus and dermatomyositis: A report from the 3rd International Conference on Cutaneous Lupus Erythematosus (ICCLE) 2013. J Invest Dermatol 2015;135(1):7–12.

Sontheimer RD. Would a new name hasten the acceptance of amyopathic dermatomyositis (dermatomyositis sine myositis) as a distinctive subset within the idiopathic inflammatory dermatomyopathies spectrum of clinical illness? J Am Acad Dermatol 2002;46:626.

Sontheimer RD, Werth VP, Connolly MK. 10th Annual Meeting of the Rheumatologic Dermatology Society. Ann Transl Med 2019;7(Suppl 4):S159.

Tarazi M, Gaffney RG, Kushner CJ, Chakka S, Werth VP. Cutaneous lupus erythematosus patients with a negative antinuclear antibody meeting the American College of Rheumatology and/or Systemic Lupus International Collaborating Clinics Criteria for systemic lupus erythematosus. Arthritis Care Res (Hoboken) 2019;71:1404–9.

Tiao J, Feng R, Berger EM, Brandsema JF, Coughlin CC, Khan N, et al. Evaluation of the reliability of the Cutaneous Dermatomyositis Disease Area and Severity Index and the Cutaneous Assessment Tool-Binary Method in juvenile dermatomyositis among paediatric dermatologists, rheumatologists and neurologists. Br J Dermatol 2017a;177(4):1086–92.

Tiao J, Feng R, Bird S, Choi JK, Dunham J, George M, et al. The reliability of the Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) among dermatologists, rheumatologists and neurologists. Br J Dermatol 2017b;176(2):423–30.

Tse K, Sangodkar S, Bloch L, Arntsen K, Bae SC, Bruce IN, et al. The ALPHA Project: Establishing consensus and prioritisation of global community recommendations to address major challenges in lupus diagnosis, care, treatment and research. Lupus Sci Med 2021;8.

van Vollenhoven R, Voskuyl A, Bertsias G, Aranow C, Aringer M, Arnaud L, et al. A framework for remission in SLE: Consensus findings from a large international task force on definitions of remission in SLE (DORIS). Ann Rheumat Dis 2017;76(3):554–61.

van Vollenhoven RF, Hahn BH, Tsokos GC, Wagner CL, Lipsky P, Touma Z, et al. Efficacy and safety of ustekinumab, an IL-12 and IL-23 inhibitor, in patients with active systemic lupus erythematosus: Results of a multicentre, double-blind, phase 2, randomised, controlled study. Lancet 2018;392(10155):1330–9.

Werth V, Furie R, Gaudy A, Ye Y, Korish S, Delev N, et al. CC-220 decreases B-cell subsets and plasmacytoid dendritic cells in systemic lupus erythematosus (SLE) patients and is associated with skin improvement: pharmacodynamics results from a phase IIa proof of concept study. Arthritis Rheumatol 2017;69 abstract #886.

Werth V, Merrill J, Furie R, Dörner T, Vollenhoven RV, Lipsky P, et al. Effect of iberdomide on cutaneous manifestations in systemic lupus erythematosus: Results of a 24-week, placebo-controlled, phase 2 study. Ann Rheum Dis 2021;80(1):132.

Werth VP, Patel B, Concha JS, Okawa J, Pearson D, Hejazi EZ, et al. Safety and efficacy of lenabasum in refractory skin-predominant dermatomyositis subjects treated in an open label extension of trial jbt101-dm-001. Ann Rheumat Dis 2018;77(Suppl 2):1111–12.