for a medical centre with expertise in diagnosing and treating ECD and to refer the patient there. Careful readers of this article will probably not overlook dermatological signs of this extraordinary disease.1

C. Rose
Dermatopathology Laboratory, Maria-Goeppert-Strasse 5, 23562 Lübeck, Germany
E-mail: rose@dermatohistologie-luebeck.de

References
1 Kobic A, Shah KK, Schnitt AR et al. Erdheim–Chester disease: expanding the spectrum of cutaneous manifestations. Br J Dermatol 2020; 182:405–9.
2 Rush W. William Chester, M.D., 1903–1974. Dermatol Pract Concept 2001; 7:255–9.
3 Diamond EL, Dagna L, Hyman DM et al. Consensus guidelines for the diagnosis and clinical management of Erdheim–Chester disease. Blood 2014; 124:483–92.
4 Ozkaya N, Rosenblum MK, Durham BH et al. The histopathology of Erdheim-Chester disease: a comprehensive review of a molecularly characterized cohort. Mod Pathol 2018; 31:581–97.
5 Su HH, Wu W, Guo Y et al. Paediatric Erdheim-Chester disease with aggressive skin manifestations. Br J Dermatol 2018; 178:261–4.
6 Chasset F, Barete S, Charlotte F et al. Cutaneous manifestations of Erdheim-Chester disease (ECD): clinical, pathological, and molecular features in a monocentric series of 40 patients. J Am Acad Dermatol 2016; 74:513–20.

Conflicts of interest: none to declare.

The skin myositis Delphi group puts the details in ‘skin-predominant’ dermatomyositis

DOI: 10.1111/bjd.18790

In this issue of the BJD, the Skin Myositis Delphi Group describes the efforts of physicians who have united to resolve an important dermatological question – what really defines skin-predominant dermatomyositis?1 Acceptance of skin-predominant/amyopathic dermatomyositis (ADM) as a true dermatomyositis (DM) subtype has taken a considerable amount of time and effort on the part of dermatologists.2 However, the perseverance of a dedicated group of dermatologists has culminated in ADM being included as a recognized DM subtype in the recently validated 2017 European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria for idiopathic inflammatory myopathies (IIM).3 The 2017 EULAR/ACR criteria include three DM-associated skin manifestations to classify ADM: Göttron sign, Göttron papules and heliotrope rash. By using the EULAR/ACR classification criteria, patients with these pathognomonic skin manifestations of juvenile and adult DM were accurately classified without including muscle biopsy data.

Although the inclusion of ADM is a huge victory for dermatologists and other specialists who frequently diagnose and/or treat these patients, the 2017 EULAR/ACR IIM criteria for ADM are not perfect. Recent work by Dr Victoria Werth, has shown that up to 26% of patients with ADM still fail to meet the diagnostic criteria set forth in the 2017 EULAR/ACR IIM classification.4 Additional work has also shown a significant percentage of patients with skin-predominant ADM are misdiagnosed as having cutaneous lupus erythematosus.5,6 Failure to appropriately diagnose these patients as having DM is concerning for several reasons. For one, a misdiagnosis may lead to significant delays in screening appropriate patients for associated underlying comorbid conditions, including interstitial lung disease or visceral malignancies. Secondly, misdiagnosis implies that more than 25% of patients with skin-predominant DM may fail to be included in research efforts, including clinical trials, aiming to better understand and treat patients with DM.

To improve the classification criteria for skin-predominant DM and minimize the chances of misdiagnosis, the Skin Myositis Delphi Group, led by Dr Werth and several others, developed an extensive list of items seen in patients with DM in terms of distribution, morphology, symptoms, antibodies, histology and contextual factors, and invited 50 dermatologists and rheumatologists with expertise in DM from around the world to participate in several Delphi rounds to determine a consensus concerning those factors which are thought to be most important for a classification criteria in diagnosing skin-predominant DM. These efforts, which are detailed in this issue of the BJD, have resulted in a list of 25 potential criteria in the above categories that will be subjected to further validation in prospective testing.1

The findings in this manuscript represent a tremendous amount of work over several years by Dr Werth and colleagues, all in the name of ensuring that this subset of patients with DM have the same opportunities for screening, therapeutic intervention and enrolment in clinical trials testing cutting-edge medications as patients with classic DM. Importantly, the goal is not to replace the 2017 EULAR/ACR criteria, which are adequate in classifying ADM, but, if prospectively validated, these classification criteria may be complementary and utilized if there is a high index of suspicion of DM in a patient who does not meet the EULAR/ACR criteria for ADM. This work is yet another example of the positive impact that dermatologists from around the world can have on patients with systemic diseases that may be seen and treated by a variety of medical specialists. These efforts not only should be of interest to those who treat patients with DM, but also will hopefully stimulate similar work by dermatologists for other diseases that may be systemic in nature, but have skin-predominant subtypes.
Real-world evidence vs. randomized control trials

DOI: 10.1111/bjd.18791

Real-world evidence (RWE) and randomized control trial (RCT) data are considered mutually complementary. When a new drug is licensed for use it has passed through rigorous testing in clinical trials to determine side-effects, risks and efficacy. However, the outcomes of RWE are often regarded to be of different credibility; the true test of a drug is when it meets the everyday clinical setting outside the well-defined framework of the trials.

The advantages and disadvantages of RWE must be described clearly, and then the proper protocol can be planned.1,2

RWE includes patients with health issues that would lead to exclusion from RCT, but these patients are also in need of efficient treatment for their disease. Therefore, it is of the utmost importance also to gain treatment outcome experience with patients suffering from comorbidities to their disease and to clarify benefits and challenges when other treatments are given concomitantly.

In the paper by deWijs et al.3 in this issue of the BJD, the first-year clinical real-life experience with dupilumab, an anti-interleukin (IL)-13/IL-4R antibody, in 95 patients with atopic dermatitis (AD) is described. Patients were recruited from two academic dermatology centres in the Netherlands among patients suffering from moderate-to-severe AD, of whom 62 were already on active systemic immune suppressive therapy.

Dupilumab is the first biological treatment to be licensed for the treatment of moderate-to-severe AD.4

This study demonstrates that dupilumab is an effective treatment that results in a decrease of disease activity and burden, as demonstrated through an average decrease of the Eczema Activity Score Index of more than 50% as well as a reduction in the Investigators Global Assessment to 0 or 1 in 38% of the patients included in the study. However, it did also demonstrate that five patients discontinued the treatment due to side-effects or ineffectiveness and that 62% had ocular side-effects, which is somewhat more than expected from the SOLO studies where 3–5% of patients experienced conjunctivitis.4

This study is exemplary in registering data on patients undergoing new treatments. The use of outcome measures described by the Harmonizing Outcome Measures in Eczema initiative regarding how and which data to register facilitates comparison of data using the same validated tools.5 Further, as described by the TREAT-registry task force, it enables new studies, which will undoubtedly follow this study, to be easily compared and combined in meta-analyses.6 This will shed new light on the potentials of new treatment for AD.

M. Deleuran and C. Vestergaard

Department of Dermatology, Aarhus University Hospital, Aarhus, Denmark

E-mail: mettlede@rm.dk and chrivist@rm.dk

References

1 Kim HS, Lee S, Kim JH. Real-world evidence versus randomized controlled trial: clinical research based on electronic medical records. J Korean Med Sci 2018; 33:e213.
2 Suvarna VR. Real world evidence (RWE) – are we (RWE) ready? Perspect Clin Res 2018; 9:61–3.
3 de Wijs LEM, Bosma AL, Erler NS et al. Effectiveness of dupilumab treatment in 95 patients with atopic dermatitis: daily practice data. Br J Dermatol 2020; 182:418–26.
4 Simpson EL, Bieber T, Guttmann-Yassky E et al. Two phase 3 trials of dupilumab versus placebo in atopic dermatitis. N Engl J Med 2016; 375:2335–48.