reveals various clinical course and therapeutic responsiveness according to the clinical and serological subsets. Some myositis-specific autoantibodies (MSAs) are useful markers for the classification of ILD in myositis and give useful information for predicting the prognosis and determining treatment.

Anti-aminoacyl-tRNA synthetases are closely associated with a common clinical manifestation, termed "anti-synthetase syndrome" including high prevalence of ILD. ILD in patients with anti-synthetases shows a similar clinical course with a favourable response to therapy but frequent recurrences. Therefore, the concomitant use of glucocorticoids and immunosuppressive drugs is recommended from early stage of the disease.

Anti-MDA5 antibody was reported to be associated with clinically amyopathic dermatomyositis (CADM) with rapidly progressive ILD, especially in eastern Asian population. Because of a very poor life prognosis, the intensive immunosuppressive therapy against ILD from early stage is recommended using the combination of high dose glucocorticoids, calcineurin inhibitors and intravenous cyclophosphamide. We have experienced the effectiveness of plasmapheresis in some anti-MDA-positive patients with intractable ILD, suggesting a possible pathogenicity of anti-MDA5 antibody.

Thus, ILD in myositis is dependent on the autoantibodies, therefore it is important to know the profiles of MSAs in PMDM patients. Recently, we established the ELISA systems for anti-synthetase and anti-MDA5 antibodies, which are as efficient as the standard immunoprecipitation assays. These systems enable easier and wider use in the diagnosis and therapeutic decision of patients suspected to have myositis and ILD.

27 THE INTERNATIONAL CONSENSUS ON STANDARDIZED NOMENCLATURE OF ANTINUCLEAR ANTIBODY HEP-2 CELL PATTERNS (ICAP) INITIATIVE – ITS IMPACT AND UPDATE FROM 3RD ICAP

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10.1136/lupus-2017-000215.27

Background and Aims The indirect immunofluorescence (IIF) pattern in ANA test provides initial assessment of autoantibody responses in candidate patients at risk of developing SLE. The International Consensus on ANA Patterns (ICAP) Initiative workshop originated in 2014, followed by the 2nd and 3rd workshops in 2015 and 2016. The goal of ICAP is to provide guidelines for ANA interpretation, thereby optimising adoption in diagnostic laboratories and extended to patient care.

Methods A working committee addressed collective issues on ANA nomenclature that are raised by participants representing research, clinical, and diagnostic laboratories.

Results ANA patterns are separated into three major categories (nuclear, cytoplasmic, and mitotic patterns). A total of 28

patterns were defined, described in detail, designated with alpha-numeric codes (AC-1 to AC-28), and summarised under a nomenclature and classification tree (ANApatterns.org). ICAP initiatives include translation of the website into other languages, establishing guidelines in ANA reporting, and programs for continuing education. The translation initiative promotes the establishment and dynamic engagement of a worldwide network. To date, the website displays its content in English, Spanish, Portuguese, Italian and German, while Chinese, Japanese, French, and Greek translations are ongoing. ICAP has provided a common platform to address issues that are of great interest to the scientific community. The establishment of a consensus on ANA reporting will require interaction with committees in charge of establishing disease classification and diagnostic criteria.

Conclusions Future goals include building collaborative data on ANA patterns, establishing new consensus patterns, and developing an interpretative description for each pattern for their widespread clinical use.

28 DRIVERS OF THE SLE RESPONDER INDEX (SRI-5) ENDPOINT IN CLINICAL TRIALS OF SLE

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10.1136/lupus-2017-000215.28

Background and Aims SRI is a composite endpoint used in many SLE trials. The current investigation sought to identify those SRI components that drive response.

Methods This study evaluated data from two large multinational trials that used SRI-5 as primary endpoint (n=2262 patients).

Results The overall SRI-5 response rate was 32.8%. Non-response due to lack of a 5-point SLEDAI improvement, comed violation or dropout were observed in 31%, 16.5% and 19.1%, respectively. In contrast, only 0.5% (11/2262) of patients failed to respond due to deterioration by BILAG or PGA, once achieving the first three components.

The most common SLEDAI organ systems involved at baseline were mucocutaneous (90.6%), musculoskeletal (82.9%) and immunologic (71.6%). The 6 other SLEDAI-defined organ systems were active in ≤11% of patients.

At least one SLEDAI manifestation with low prevalence and high score (≥4 pts) was present in 18.1% and 17.2% of patients in Trials 1 and 2, respectively. SRI-5 response rates for these patients were higher, regardless of treatment assignment.

Conclusions The SRI-5 response is driven by SLEDAI improvement, concomitant medication adherence and trial completion. Patients with less frequent, more severe manifestations had high placebo response rates. A simple dichotomous improvement in SLEDAI score, coupled with successful trial completion and medication stability, provides a more efficient, clinically relevant approach to assess outcome.