AB0696

DETECTION OF COEXISTING MYOSITIS-SPECIFIC AUTOANTIBODIES WITH LINE AND DOT IMMUNOASSAYS IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

Jean-Baptiste Vulsteke^{1,2}, Xavier Bossuyt^{3,4}, Kristl G. Claeys^{5,6}, Doreen Dillaerts⁴, Nele Vanhorebeek⁷, Koen Poesen^{3,8}, Jan Lenaerts¹, Rene Westhovens^{1,2}, Philip Van Damme^{5,9}, Daniel Blockmans¹⁰, Petra De Haes¹¹, Ellen De Langhe^{1,2}.

¹University Hospitals Leuven, Rheumatology, Leuven, Belgium; ²KU Leuven, Skeletal Biology and Engineering Research Center, Leuven, Belgium; ³University Hospitals Leuven, Laboratory Medicine, Leuven, Belgium; ⁴KU Leuven, Clinical and Diagnostic Immunology, Leuven, Belgium; ⁵University Hospitals Leuven, Neurology, Leuven, Belgium; ⁶KU Leuven, Laboratory for Muscle Diseases and Neuropathies, Leuven, Belgium; ⁷KU Leuven, Leuven, Belgium; ⁸KU Leuven, Laboratory for Molecular Neurobiomarker Research, Leuven, Belgium; ⁹KU Leuven, Department of Neurosciences, Experimental Neurology, VIB Center for Brain and Disease Research, Laboratory of Neurobiology, Leuven, Belgium; ¹⁰University Hospitals Leuven, General Internal Medicine, Leuven, Belgium; ¹¹University Hospitals Leuven, Dermatology, Leuven, Belgium

Background: Myositis-specific autoantibodies (MSAs) can be identified in up to 60% of patients with idiopathic inflammatory myopathies (IIM) (1). Based on previous immunoprecipitation studies, MSAs are considered to be mutually exclusive (1), but detection of coexisting MSAs with line or dot immunoassay (LIA/DIA) has been described (2-3).

Objectives: To determine the prevalence of detection of coexisting MSAs with different LIAs/DIAs in patients with IIM, assess the concordance between different assays and describe the clinical phenotype of patients with coexisting MSAs.

Methods: Cross-sectional assessment of prevalence of coexisting MSAs as detected by two LIAs (Euroline Autoimmune Inflammatory Myopathies, Euroimmun, Lübeck, Germany; and ImmcoStripe Myositis Advanced LIA, Trinity Biotech, Buffalo, USA) and 1 DIA (12 IgG Dot, Alphadia, Mons, Belgium), concordance between these assays and clinical phenotype of patients with coexisting MSAs in a single-center cohort of patients with a diagnosis of one of the subtypes of IIM as diagnosed by the treating physician.

Results: Nineteen of 145 patients (12%) had coexisting MSAs on at least one assay: 5 on the DIA (3,5%) and 5 (3,5%) and 11 (7,6%) on the LIA of Euroimmun and Trinity Biotech respectively. The results were concordant between 2 assays for more than one MSA in only 3 patients. The three combinations of these patients were anti-Jo-1 and anti-NXP-2, anti-Jo-1 and anti-TIF1-gamma, and anti-SAE and anti-NXP2 autoantibodies. The first two patients had an antisynthetase syndrome and the last patient an overlap myositis phenotype. All assays combined, anti-TIF1-gamma (9/19), anti-Jo-1 (7/19) and anti-NXP2 (6/19) autoantibodies were the most detected MSAs with concurrent detection of another MSA, though concordance between assays for these MSAs was low to moderate.

Conclusion: Detection of coexisting MSAs with LIA or DIA occurs in a minority of patients with IIM with varying prevalence between assays from different manufacturers. The combination of more than 1 MSA is not concordant between LIAs/DIAs in the vast majority of patients, suggesting that in most patients detection of coexisting MSAs with LIAs/DIAs reflects a problem of specificity of the assays for the involved autoantibodies.

REFERENCES

- [1] McHugh NJ, Tansley SL. Autoantibodies in myositis. *Nature Reviews Rheumatology*. 2018.
- [2] Brouwer R, Hengstman GJD, Egberts V, Ehrfeld H, Bozic B, Ghirardello A, et al. Autoantibody profiles in the sera of European patients with myositis. *Ann Rheum Dis*. 2001;60:116–23.
- [3] Cavazzana I, Fredi M, Ceribelli A, Mordenti C, Ferrari F, Carabellese N, et al. Testing for myositis specific autoantibodies: Comparison between line blot and immunoprecipitation assays in 57 myositis sera. *J Immunol Methods*. 2016;

Acknowledgement: The study was funded by Alphadia, D-tek, Trinity Biotech and Euroimmun.

Disclosure of Interests: Jean-Baptiste Vulsteke Grant/research support from: SB PhD Fellow at FWO, grant from the Fund Joël Hurlet, Xavier Bossuyt Consultant for: Inova Diagnostics, Kristl G Claeys: None declared, Doreen Dillaerts: None declared, Nele Vanhorebeek: None declared, Koen Poesen: None declared, Jan Lenaerts: None declared, Rene Westhovens Grant/research support from: Bristol-Myers Squibb, Consultant for: Celltrion, Galapagos-Gilead, Philip Van Damme Grant/research support from: Senior clinical investigatorship at FWO-Vlaanderen, Daniel

Blockmans: None declared, Petra De Haes: None declared, Ellen De Langhe: None declared

DOI: 10.1136/annrheumdis-2019-eular.4171

AB0697

LOW DOSE CYCLOPHOSPHAMIDE AND PIRFENIDONE MIGHT WORK IN SYNERGY TO RELIEVE INTERSTITIAL LUNG DISEASE WITH CONNECTIVE TISSUE DISEASE: A PRELIMINARY OBSERVATIONAL STUDY

Qingran Yan, Lichong Shen, Xiaoxiang Chen. Renji Hospital, Shanghai Jiaotong University School of Medicine, Department of Rheumatology, Shanghai, China

Background: There are anti-inflammation and anti-fibrosis agents available for connective tissue disease associated interstitial lung disease (CTD-ILD). A clinical study has been initiated to assess the combination of the two (NCT03221257), but whether they can spare dose for each other is unknown.

Objectives: This preliminary study is aimed to observe outcomes of CTD-ILD patients receiving cyclophosphamide plus pirfenidone as a rescue therapy, with each agent at about one third of routine dosage.

Methods: We enrolled CTD-ILD patients who did not improve their symptoms (dyspnea or cough) after at least one-month steroids treatment (prednisone $\geq 1\text{mg/kg}$ daily). Patients who had adjusted immunosuppressive agents other than steroids or had received anti-fibrotic medications within three months before enrollment were ruled out. We switched the treatment into pulse cyclophosphamide (0.4g/m² monthly) combined with pirfenidone (300mg twice per day). Besides, we reduced the steroids to prednisone 0.5mg/kg daily and then tapered routinely. All the patients were followed up for 12 months.

Results: We enrolled seven patients, of whom two had anti-synthetase syndrome, two had Sjögren syndrome, two had scleroderma and one had mixed connective tissue disease. The media DLCO was 51% of prediction (range 47.7% to 63%) and media FVC was 72.3% of prediction (range 39% to 81%). The media 6MWD was 275m (range 202 to 324m). At the end of 12-month follow-up, all the patients regained functional independence with a media 52.7% increase of 6-minute walk distance (range 34.4% - 86.3%). Pulmonary function tests showed improved forced vital capacity (median improvement 13.4%, range 0-35.9%) and DLCO (median improvement 6.3%, range 1.7%-16%). The HRCT score had a median decrease of 20.1% (range 11.7% to 29.6%). For quality of life assessment, the SGRQ total score had a median improvement of 53.3% (range 19.5% to 61.7%). Of note, no adverse events were observed during the 12-month follow-up.

Conclusion: Our Study provided preliminary but promising clinical evidence for a new strategy in treating CTD-ILD, that cyclophosphamide and pirfenidone might work in synergy and spare dose for each other. A well-designed controlled study is needed to further establish its safety and efficacy.

REFERENCES

None.

Disclosure of Interests None declared

DOI: 10.1136/annrheumdis-2019-eular.2257

AB0697B

VALIDATION OF 2017 CLASSIFICATION CRITERIA FOR ADULT AND JUVENILE IDIOPATHIC INFLAMMATORY MYOPATHIES PROPOSED BY EULAR/ACR IN CHINESE PATIENTS

Xiaohui Zhang, Xinlei Yang, Lanlan Ji, Zhuoli Zhang. Peking University First Hospital, Rheumatology and Clinical Immunology Department, Beijing, China

Background: Idiopathic inflammatory myopathies(IIMs) are heterogeneous disorders characterized by muscle weakness and muscle inflammation. Although the Bohan and Peter criteria proposed in 1975 are most widely used^[1,2], there are some limitations. Firstly, they did not clearly specify how to exclude other forms of myopathy disease. Secondly, each criterion is not well defined either. EULAR and ACR jointly proposed the classification criteria for adult and juvenile IIMs and their major subgroups in 2017. The data-driven criteria exhibited high sensitivity and specificity. But most of the patients (62.6%) in the data were Caucasians, the performance of the criteria in Asian patients is unknown, which was one of the important limitations^[6,7].

Objectives: To evaluate the ability of 2017 EULAR/ACR Classification Criteria for Adult and Juvenile Idiopathic Inflammatory Myopathies (IIM) to