OP0101-HPR

QUALITY OF LIFE IN PEOPLE WITH SYSTEMIC SCLEROSIS WITH DIFFERENT DEGREES OF LUNG DISEASE - A CROSS-SECTIONAL STUDY

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Background: There are few studies evaluating different aspects of quality of life including depressive symptoms and physical capacity and physical activity in patients with systemic sclerosis (SSc) with different degrees of lung disease.

Objectives: The aim of this study was to evaluate differences in self-reported disability, physical capacity and activity, depressive symptoms and quality of life, between patients with SSc with no-mild lung disease and those with moderate-endstage lung disease.

Methods: In this cross-sectional study, 279 patients with SSc fulfilling the 2013 ACR/EULAR criteria for SSc (84% limited and 16% diffuse SSc) were included. Medsger disease severity scale was used to subgroup the patients into no-mild (n=156) or moderate-endstage lung disease (n=115). Disability was measured with Health Assessment Questionnaire-Disability Index (HAQ-DI); physical capacity (ability to walk, jog/run); and physical activity (different intensities) was measured with three single questions; depressive symptoms with Hospital Anxiety and Depression-scale (HADs); and quality of life was measured with The Short Form (36) Health Survey (SF-36).

Results: Patients with moderate-endstage lung disease reported higher scores on HAQ-DI ($p<0.001$) and lower scores on SF-36 physical component ($p<0.0001$) than patients with no-mild lung disease. Patients with moderate-endstage lung disease reported lower physical capacity ($p<0.0001$), less physical activity on low to moderate intensity the past 6 months ($p<0.016$) and less exercise on moderate to high intensity the past year ($p=0.022$) compared to those with no-mild lung disease. There was no difference between the two subgroups when it comes to the mental component in SF-36 ($p=0.2$), however patients with moderate-endstage lung disease had lower scores on the subscales vitality ($p=0.003$), social function ($p=0.002$) and emotional role function ($p=0.005$) as well as higher scores on the HADs depressive symptoms scale ($p=0.024$), than the patients with no-mild lung disease.

Conclusion: Patients with SSc with moderate-endstage lung disease report more disability, lower physical capacity and activity, are more depressed and the physical aspects of quality of life is lower, as well as vitality, social function and emotional role function, compared to patients with no-mild lung disease. Studies evaluating whether increased physical activity and exercise may improve depressive symptoms and aspects of quality of life in patients with moderate-endstage lung disease are needed.

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Immunodeficiency and autoimmune disease

OP0102

GENE SCREENING OF PRIMARY IMMUNODEFICIENCY DISEASES IN PATIENTS WITH JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Systemic lupus erythematosus (SLE) is one of the most common auto-immune diseases in childhood. Primary immunodeficiency disease (PID) patients may present or combine with autoimmune diseases.

Objectives: This study aimed to perform gene sequencing via high-throughput sequencing technology in a series of Chinese pediatric SLE patients, and investigate the concomitant situation of PIDs and SLE. Gene sequencing results may help clarify the pathogenesis of SLE.

Methods: This was a retrospective case series of SLE children who referred to the Peking Union Medical College Hospital between 01/2016 and 09/2019. Genetic tests were performed in patients who met the inclusion criteria. We

then collected demographic, clinical, and treatment information of all involved patients. Descriptive statistics were used.

Results: Seventy-one patients were finally included (eighteen boys and fifty-three girls). The median age at the time of disease onset was 9.5 (range, 3-15) years. It is notable that five patients experienced their first attack before the age of five. Twenty-seven patients showed a persistent increase in ESR during treatment, while thirteen cases presented with repeated CMV infection, thirty-four cases with persistent low complement levels, seven with basal ganglia calcification showed in skull CT or MRI, four with special type of rash (i.e., frostbite-like rash, discoid erythema, reticular erythema), two with obvious hepatosplenomegaly, and one case with type I diabetes. Gene sequencing results showed that about ten patients combine with primary immunodeficiency disease, including Aicardi-Goutières Syndrome (AGS) (n=4), Spondyloenchondro-dysplasia with immune dysregulation (SPENCDI) (n=1), STING-associated vasculopathy with onset in infancy (SAVI) (n=1), lysinuric protein intolerance (LPI) (n=1), Ras-associated autoimmune leukoproliferative disorder (RALD) (n=2).

Conclusion: SLE patients who present atypical or refractory manifestations should attach importance to the existence of primary immunodeficiency disease. Genetic tests are recommended for patients with early-onset SLE, especially those with recurrent frostbite-like rash or persistent CMV infection since childhood.

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Spondyloarthritis - treatment

OP0103

DOES GENDER, AGE OR SUBPOPULATION INFLUENCE THE MAINTENANCE OF CLINICAL REMISSION IN AXIAL SPONDYLOARTHRITIS FOLLOWING CERTOLIZUMAB PEGOL DOSE REDUCTION?

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Background: Previous studies have shown that withdrawing tumour necrosis factor inhibitors (TNFi) in patients (pts) with axial spondyloarthritis (axSpA) who have achieved sustained remission often leads to relapse.¹ However, none have formally tested TNFi dose reduction strategies in a broad axSpA population or evaluated whether relapse following TNFi dose reduction and withdrawal is associated with a specific demographic subgroup.

Objectives: C-OPTIMISE evaluated the percentage of pts without flare after TNFi dose continuation, reduction or withdrawal in adults with early axSpA treated with the Fc-free, PEGylated TNFi certolizumab pegol (CZP). Here, we analyse whether responses to reduced maintenance dose were comparable in pts stratified by axSpA subpopulation, gender and age.

Methods: C-OPTIMISE (NCT02505542) was a multicentre, two-part phase 3b study in adults with early (<5 years' symptom duration) active axSpA (stratified for radiographic [r]- and non-radiographic [nr]- axSpA). Pts received CZP 200 mg every 2 weeks (wks) (Q2W; 400 mg loading dose at Wks 0, 2 and 4) during the open-label induction period. At Wk 48, pts in sustained remission (Ankylosing Spondylitis Disease Activity Score [ASDAS] <1.3 at Wk 32 or 36 [if ASDAS <1.3 at Wk 32, it must be <2.1 at Wk 36, or vice versa] and at Wk 48) were randomised to double-blind full maintenance dose (CZP 200 mg Q2W); reduced maintenance dose (CZP 200 mg every 4 wks [Q4W]) or placebo (PBO) for a further 48 wks (maintenance period). The primary endpoint was the percentage of pts not experiencing a flare (ASDAS ≥2.1 at two consecutive visits or ASDAS >3.5 at any timepoint) during Wks 48-96. Analyses were conducted on subgroups according