skin disease during follow-up. Thus, performance of HRM already in very early disease stages can support individual risk-stratification of SSc patients.

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FRI0469

ACUTE AND CHRONIC EFFECTS OF TWO DIFFERENT INTRAVENOUS ILOPROST REGIMENS IN SYSTEMIC SCLEROSIS: A SINGLE CENTRE PRAGMATIC NON-RANDOMISED TRIAL

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Background: In systemic sclerosis (SSc) vascular involvement plays a pivotal role in the pathogenesis and clinical manifestations.¹ Intravenous (IV) iloprost (ILO), a prostacyclin analogue, is administered in SSc, according to EULAR recommendations, to improve Raynaud's phenomenon (RP) and to achieve digital ulcers (DU) healing and prevention,² even though no further indications about the regimen are specified.

Objectives: The aim of this study was to evaluate acute and chronic effects of IV ILO administered with two different regimens as assessed by power Doppler ultrasound (PDUS) and nailfold videocapillaroscopy (NVC) and in a group of patients not treated with ILO.

Methods: This was a single centre pragmatic non-randomised trial on 96 patients with SSc divided in 3 groups: no ILO (group A), ILO once monthly (group B) and ILO for 5 consecutive days every 3 month (group C). At each evaluation patients underwent PDUS and NVC. Group A patients were examined at baseline and after 3 months. Group B every month before and after IV ILO monthly therapy for 3 months. Group C before and after the 1 and 5 days of therapy at baseline and after 3 months. PDUS parameters included resistivity index (RI), finger pulp blood flow (FPBF) and periungual vascularisation. The sum of capillaries apex width in one millimetre was assessed through NVC. Results were analysed considering the average outdoor temperature at the place of residence.

Results: An acute IV ILO effect was observed for FPBF in group B and C ($p<0.001$ and 0.005 respectively). An acute effect was observed for RI and periungual vascularisation only in group B. A progressive increase was observed for the other parameters without being statistically different. On the contrary IV ILO effects were not observed any longer before the following infusion. Moreover, some parameters (FPBF in group B and RI in group C) showed a statistically higher increase as low as the outdoor temperature was.

Conclusions: IV ILO therapy was able to cause an acute effect with respect to PDUS parameters, especially in group B. The acute effect was not any longer maintained until the following infusion. Future studies are needed to assess time for re-treating.

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FRI0470

A PHASE 2 STUDY OF SAFETY AND EFFICACY OF LENABASUM (JBT-101), A CANNABINOID RECEPTOR TYPE 2 AGONIST, IN REFRACTORY SKIN-PREDOMINANT DERMATOMYOSITIS

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Background: Effective treatment options are limited for refractory skin disease in dermatomyositis (DM). Lenabasum is a non-immunosuppressive, synthetic, oral preferential CB2 agonist that triggers resolution of innate immune responses and reduces cytokine production by PBMC from DM patients.

Objectives: The purpose of this study was to test safety and efficacy of lenabasum (aka JBT-101, anabasum) in DM subjects with refractory, moderate-to-severely active skin disease.

Methods: A double-blind, randomised placebo-controlled 16 week Phase 2 trial (JBT101-DM-001; NCT02466243) enrolled adults with documented DM and a Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) activity score ≥ 14 , minimal active muscle involvement and failure or intolerance to hydroxychloroquine and stable DM medications including immunosuppressants. Subjects received 2 escalating dose levels of lenabasum (20 mg QD X 4 weeks, then 20 mg BID X 8 weeks) or PBO X 12 weeks. Subjects were followed off study drug X 4 weeks. Safety and efficacy outcomes were assessed from Day 1 through end of study at Week 16. The primary efficacy objective was to assess efficacy of lenabasum using CDASI activity score.

Results: 11 adults each received lenabasum and PBO (n=22). Demographic and disease characteristics were similar in both cohorts. Both cohorts had mean CDASI activity scores in the severe range^{33–35} despite immunosuppressants (19/22 subjects). Lenabasum subjects had clinically meaningful improvement in CDASI activity scores with mean reduction ≥ 5 points at all visits after 4 weeks. Improvement had statistical significance at end of study (figure 1, $p=0.02$, 2-sided MMRM) that first became apparent after 4 weeks. Lenabasum provided greater improvement than placebo in CDASI damage index, patient-reported global skin disease and overall disease assessments, skin symptoms including photosensitivity and itch, fatigue, sleep, pain interference with activities, pain, and physical function (examples in figure 1). Improvements in secondary efficacy outcomes reached statistical significance ($p\leq 0.1$, 1-sided MMRM) at multiple visits after week 4 (figure 1). There were no serious, severe or unexpected adverse events (AEs) related to lenabasum. Tolerability of lenabasum was excellent with no study drop-outs. Subjects in the lenabasum cohort had numerically more mild AEs than placebo subjects (29 vs. 19) and fewer moderate AEs (4 vs. 7). AEs in ≥ 3 subjects in any cohort were diarrhoea, dizziness (lightheadedness), fatigue and dry mouth.

Abstract FRI0470 – Figure 1. Effects of Lenabasum on Efficacy Outcomes in Refractory, Skin-Predominant DM

Conclusions: Lenabasum demonstrated consistent evidence of clinical benefit across multiple efficacy outcomes and had acceptable safety and tolerability in this Phase 2 trial in refractory skin disease in DM. Further evaluation of lenabasum in the treatment of DM is warranted.

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