Inflammatory markers were significantly elevated in 41 (82.0%) patients when the vena cava involvement developed, the mean ESR was 34.0±29.2mm/hr, and the median CRP level was 19.9(0.2-177.0) mg/L. The mean BDCAF2006 score of all patients was 4.6±1.6. Glucocorticoid was used in 47 (94.0%) patients after vena cava involvement was diagnosed, and cyclophosphamide was the first-choice immunosuppressant. Forty-one (82.0%) patients received anticoagulation treatment. Five patients had received placement of IVC filter, and 3 patients had taken balloon dilation of IVC. With a mean follow up of 4.1±3.8 years, 45 patients (90.0%) achieved clinical improvements, 6 patients (12.0%) had relapse of vascular involvement, 5 patients (10.0%) died. The respective estimated cumulative 1- and 5- years relapse-free rates were 90.9% and 83.1%, and the respective estimated 1- and 5- years survival rates were 95.9% and 90.1%.

Conclusion: Vena cava involvement is a rare complication in BD patients. The prognosis of these patients is relatively optimistic after proper treatment. To the best of our knowledge, our study is the largest cohort of BD patients with vena cava involvement.

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FRIDAY, 05 JUNE 2020

Scleroderma, myositis and related syndromes

FRI0226 OPTICAL COHERENCE TOMOGRAPHY OF THE SKIN DETECTS SCLERODERMA CHANGES IN CLINICALLY UNAFFECTED SKIN: AN OPPORTUNITY FOR EARLY DETECTION OF SYSTEMIC SCLEROSIS

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Background: The Very Early Diagnosis Of Systemic Sclerosis (VEDOSS) study has shown that 82% of patients with Raynaud's Phenomenon, specific ANA positivity and scleroderma pattern at nail fold videocapillaroscopy will fulfil classification criteria within 5 years. This is suggesting that there is a subclinical window of opportunity to diagnose systemic sclerosis (SSc) before clinical manifestations occur. In this scenario, a non-invasive tool to diagnose SSc in clinically unaffected skin might improve the early detection of disease in at risk-patients. Optical coherence tomography (OCT) of the skin has been shown to be a sensitive and accurate biomarker of skin fibrosis in SSc.

Objectives: Here we aimed to assess the ability of skin OCT to "detect" SSc in clinically unaffected skin from a multicentre cohort.

Methods: Dorsal forearm skin of SSc patients and matched-healthy controls (HC) was evaluated using VivoSight scanner (Michelson Diagnostics). Mean A-scans (mean OCT signal plotted against depth-in-tissue) were derived as previously described. Minimum Optical Density (MinOD), Maximum OD (MaxOD) and OD at 300 micron-depth (OD300) were calculated. Clinical involvement was assessed by an operator blinded to OCT findings using the mRSS. Receiver-operating characteristic (ROC) curve analysis was carried out for MinOD, MaxOD, and OD300 to evaluate their ability to discriminate between SSc and HC. Statistical analysis was performed using GraphPad Prism software V.7.0.

Results: One hundred seventy four OCT images were collected from 87 subjects [43 SSc (39 Female, mean age 49.7±9.1 years) and 44 gender/age-matched healthy controls (HC) (36 Female, mean age 50.2±8.3 years)] in two different SSc centres. All patients fulfilled classification criteria for SSc. OCT measures demonstrated discriminative ability in SSc skin detection with any clinical skin involvement (0-3 at site of analysis) with an AUC of 0.73 (MinOD, 95%CI 0.64-0.81), 0.77 (MaxOD, 95%CI 0.7-0.85) and 0.82 (OD300, 95%CI 0.76-0.89); p<0.0001 for all as previously indicated. Most importantly, all three measures showed comparable performance in detecting scleroderma also in clinically unaffected skin (mRss=0 at site of analysis), with an AUC of 0.7 (95%CI 0.6-0.81, p=0.001), 0.72 (95%CI 0.61-0.83, p=0.0003) and 0.72 (95%CI 0.61-0.83, p=0.0003) for MinOD, MaxOD and OD300 respectively.

Conclusion: Virtual biopsy by OCT recognises clinically unaffected skin of SSc patients from the HC skin. This is consistent with gene array data showing that scleroderma specific signatures are consistent in affected and clinically unaffected skin. These results inform future studies on at risk patients with clinically unaffected skin which may define a role for OCT in detecting subclinical SSc.

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FRI0227 A USABILITY SURVEY OF WRIST MOUNTED DISPOSABLE HEAT PAD ON RAYNAUD'S PHENOMENON IN PATIENTS WITH CONNECTIVE TISSUE DISEASES

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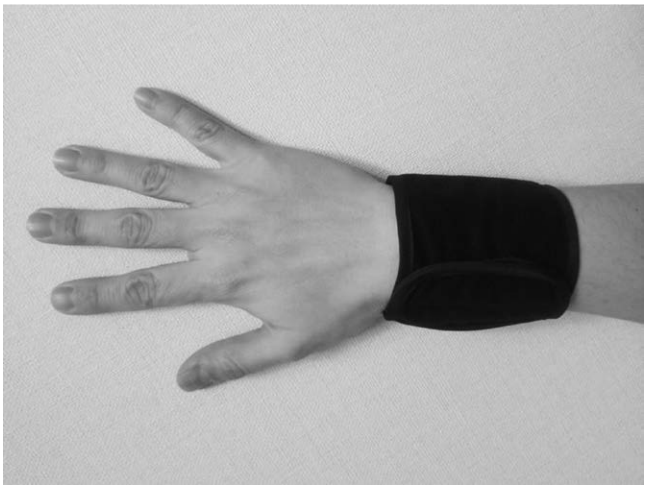
Background: For patients with connective tissue diseases (CTD), vasodilators are used to treat Raynaud's phenomenon (RP), they are difficult to control only by medication. Although physicians recommend the use of a portable handwarmer or gloves to patients with CTD presenting with RP, sustained heat-retention effects cannot be obtained from them because the patients' daily life-related activities prevent their continued use. Since the wrist mounted disposable heat pad maintains the degrees of freedom of the hands and fingers and can remain usable during the daily activities, we considered this heat pad as a useful and highly practical heating method for CTD patients presenting with RP.

Objectives: To investigate the usability and changes in symptoms resulting from the use of the wrist mounted disposable heat pad in CTD patients presenting with RP.

Methods: Subjects were 23 outpatients with CTD presenting with RP (23 females; mean age 62.6 years; mean duration following the onset of RP 10.3 years; 12 systemic sclerosis, 5 mixed connective tissue disease, 5 Sjögren's syndrome, and 1 systemic lupus erythematosus) who had used the wrist mounted disposable heat pad (put the pad in a specifically designed holder and wrap it around wrist joint (max. temperature 42 degrees Celsius, heat-retention time 6 hours)). We investigated through interviews with them the use situations, usability, and changes in RP. During their using the heat pad, medication and daily life-related precautions against RP continued to be implemented as before.

Results: Many patients had no knowledge of the heat pad (n=17, 73.9%). The most common wearing time of the heat pad was 5–6 hours (n=8, 34.8%). As for scenes of wearing the heat pad, patients who wore the pad when being out of the home accounted for the highest proportion (n=16, 69.6%), and as follows: at home (n=6, 26.1%), during kitchen work (n=3, 13.0%), and during housework (n=2, 8.7%). 17 patients (73.9%) replied that usability was "good", and 18 (78.3%) replied that usability was "better" compared with conventional measures. Moreover, many patients (n=16, 69.6%) replied that RP and associated symptoms had become reduced or alleviated. No patients replied that RP and associated symptoms had become exacerbated or severer. In terms of advantages of using the heat pad, patients who replied that the site on which the pad was mounted was felt to be warm accounted for the highest proportion (n=8, 34.8%), and those who replied that sites other than where the pad was mounted (such as fingertips, hands, and arms) were also warmed accounted for virtually the same proportion (n=7, 30.4%). Over 60% of the patients (n=14, 60.9%) replied that symptoms associated with RP (skin color, cold sensation, and pain) had become reduced or disappeared. In terms of disadvantages of using the heat pad, patients who replied that it was bothersome to use the pad accounted for the highest proportion while other patients made replies referring to cost and bad appearance. No significant accident occurred and as many as 17 patients (73.9%) replied that they would like to continue to use the heat pad in the future.



Conclusion: There have been few reports evaluating the usefulness of a heat pad for RP. The wrist mounted disposable heat pad was thought to be a heating method having the potential to achieve high levels of usability and practicality on CTD patients presenting with RP. Given that the heat pad alleviated RP or caused sites other than where the pad was mounted to be felt warm even though it did not directly heat the hands and fingers, the pad seemed to have usefulness attributed to the heating of the wrist. Although the heat pad seems to be an excellent method for addressing RP in patients' daily lives, we hope that this heat pad will be evaluated on a larger number of patients with the addition of objective indices.

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FRI0228

TOFACITINIB IN THE TREATMENT OF SKIN AND MUSCULOSKELETAL INVOLVEMENT IN ADULT PATIENTS WITH EARLY SYSTEMIC SCLEROSIS, EVALUATED BY ULTRASOUND

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Background: The whole management of systemic sclerosis (SSc) remains to be defined while trials mainly focus on the treatment of different organ involvement and disease-modifying treatments are still not available.

Objectives: To assess the safety and efficacy of tofacitinib (TOF) treatment on skin and musculoskeletal involvement as compared to methotrexate (MTX) treatment in patients with early SSc.

Methods: In this 52-week, prospective, investigator-initiated, open-label, single-centre study, 66 patients with SSc were enrolled. Thirty-three patients received 5mg of oral TOF twice a day; and thirty-three received 7.5-10 mg of MTX weekly. The primary outcome measures were: skin fibrosis improvement at week 26, assessed by the reduction in skin thickness - evaluated by the modified Rodnan skin score (mRSS) and the ultrasound (US) measured skin thickness; improvement in the musculoskeletal involvement, assessed by the reduction in the joint and tendon score (US10SSc score); and adverse events from baseline to week 26. The dynamics in the outcome measures within each group were examined through Wilcoxon tests and between-group comparisons were performed through Mann-Whitney U and Chi-square tests.

Results: At baseline, both groups of patients had similar median scores with no significant differences on all measures: mRSS (p = 0.589), US measured skin thickness (p = 0.822), and US10SSc score (p = 0.918). At week 26, significant differences were observed between the two treatment groups as the TOF treated patients showed a greater reduction in mRSS and musculoskeletal manifestations. In the TOF group, the median mRSS score decreased by 50% from 24 to 12 (IQR = 7.50) versus a smaller decrease of 8.70% in the MTX group, from 23 to 21 (IQR = 8.00), p < 0.001. The median US measured skin thickness in the TOF treated patients decreased by 12.87% from 1.71 to 1.49 (IQR =0.31) versus a decrease of 4.73%, from 1.69 to 1.61 (IQR =0.52) in the MTX group, p = 0.040. The US10SSc median score in the TOF group decreased by 56.25% from 16 to 7 (IQR = 6.50) versus a decrease of 12.5% in MTX group from 16 to 14 (IQR =10.50), p < 0.001. There was no significant difference between the groups in the number of adverse events from baseline to week 26. No cases of herpes zoster and deep vein thrombosis were observed in the TOF group.

Conclusion: The data show that in early SSc TOF may lead to a significant improvement of skin thickness, measured with the mRSS and US, and of the musculoskeletal involvement, measured by the US10SSc score. TOF has also shown a satisfactory safety profile.

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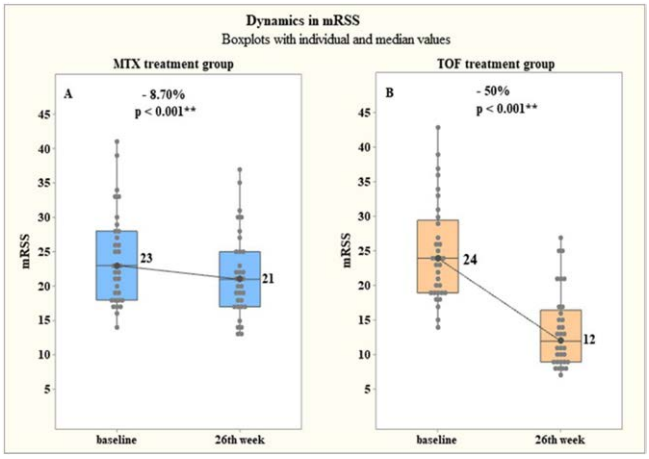

Figure 1. Panel A - Dynamics in mRSS between baseline and week 26 in the MTX group; **Panel B** - Dynamics in mRSS between baseline and week 26 in the TOF group.

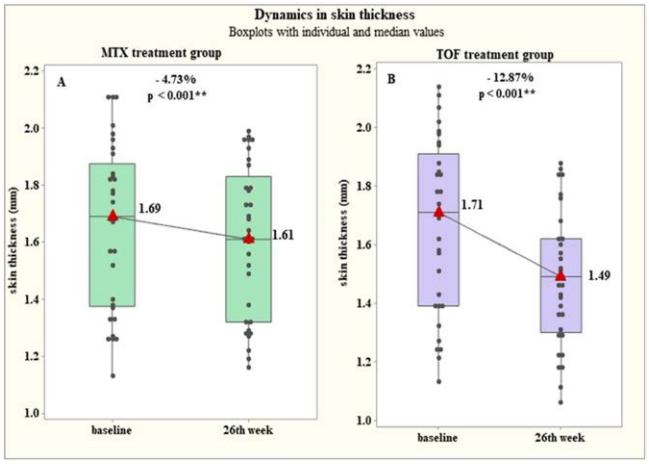

Figure 2. Panel A - Dynamics in skin thickness between baseline and week 26 in the MTX group; **Panel B** - Dynamics in skin thickness between baseline and week 26 in the TOF group.

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