Objectives: The general aim of this study was to determine the prognostic value between PRP and SRP (1,2). Although Raynaud’s phenomenon (RP) is very common in childhood, studies on diagnostic methods to differentiate between primary RP (PRP) and secondary RP (SRP) at a young age are scarce (3,4).

Methods: This was a case-control study, in which 83 patients diagnosed with RP and having undergone NCM in childhood were retrospectively included. Based on whether they were diagnosed with a connective tissue disease (CTD) during follow-up, they were classified as PRP or SRP. PRP and SRP patients were compared on demographics, NCM and ANA positivity. Variables associated with SRP were included in a multivariate logistic regression model. Predictive values were calculated for NCM, ANA positivity and the combination of NCM and ANA positivity.

Results: At the time of the baseline NCM, the mean age of the RP patients was 15.4±2.3 years. Averagely 6.4±3.2 years after the baseline NCM, 65 of the 83 patients were classified as PRP and 18 as SRP. The most common CTDs were MCTD and undifferentiated CTD. ANA positivity was associated with SRP (p=0.001). Of the NCM parameters, only capillary loss was associated with SRP (p=0.01). Abnormal numbers of dilated capillaries, giant capillaries and haemorrhages were not significantly associated with SRP. In a multivariate logistic regression model, only ANA positivity was predictive for SRP (OR 11.19, CI 3.07 - 43.68; p=0.001). Disease duration negatively correlated with BMI (R2=0.10, p<0.001). Disease activity negatively correlated with ANA (R2=0.22, p<0.001) and positively correlated with CRP (R2=0.26, p<0.001). Disease activity positively correlated with NCM (R2=0.34, p<0.001) and negatively correlated with ANA/NM (R2=0.26, p<0.001). Disease activity negatively correlated with NCM (R2=0.34, p<0.001) and positively correlated with ANA/NM (R2=0.26, p<0.001). Disease activity negatively correlated with NCM (R2=0.34, p<0.001) and positively correlated with ANA/NM (R2=0.26, p<0.001).

Conclusion: This study demonstrates that childhood RP is primary in most cases. Whereas RP in adulthood is mostly strongly associated with SSC, children with RP seem to be at risk of developing other CTDs with less apparent NCM abnormalities. Dilated capillaries, giant capillaries and haemorrhages on NCM are not associated with the spectrum of CTDs that children are at risk for, and do not differentiate between primary and secondary RP. Although capillary loss on NCM is associated with SRP, capillary loss may add little to the predictive value of serology. To clarify which NCM parameters are helpful for early detection of SSC-like CTDs, additional research is required.

References:
Background: Lower urinary tract symptoms (LUTS) are an underdiagnosed but frequent manifestation in systemic sclerosis (SSc) [1]. LUTS pathogenesis in SSc is undetermined, mainly involving dysautonomia, fibrosis and a possible antibody-mediated damage [2]. Divergently from general population, female sex and advanced age are not reported to significantly impact LUTS in SSc [2].

Objectives: To evaluate the potential influence of gender and hormone-related factors in LUTS prevalence and severity among SSc patients (Pts).

Methods: A population of 42 SSc Pts and 50 age- and sex-matched healthy subjects (HSs) was evaluated. SSc diagnosis was based on 2013 ACR/EULAR criteria. Demographic data, medications interfering with pelvic floor dynamics and general comorbidities commonly associated with LUTS – diabetes mellitus, chronic heart failure, chronic obstructive pulmonary disease, peripheral neuropathy, pelvic organ prolapse, fecal incontinence – were recorded. Validated self-reported questionnaires derived from the International Conference on Incontinence were used to assess prevalence and severity of LUTS, namely of urinary incontinence (UI) and overactive bladder (OAB) [2]. Data were analysed using non-parametric tests. A p value <0.05 and a confidence interval (CI) of 95% were considered statistically significant.

Results: There were no significant differences in main demographic data between SSc Pts and HSs. Specifically, median age was 61 years (IQR 21-85) vs 57 years (IQR 28-93) and female prevalent, 83% vs 84% in SSc Pts vs HSs, respectively. Amongst the female population, 83% of SSc Pts vs 84% of HSs was in post-menopausal state, with a median of 1 (IQR 0-3) vs 1 (IQR 0-4) pregnancy by natural route, respectively. No woman of the study had received hormone replacement therapy or local hormonal therapies prior to the study. Similarly, there were not any significant differences in analysed comorbidities, while ongoing treatment was significantly different between the two populations. SSc patients more frequently receiving calcium channel blockers and glucocorticoids than healthy subjects (p<0.01).

Conclusion: This study confirms the absence of pathogenic female-gender participation in LUTS prevalence among SSc Pts. However, consistently with findings on general population, a significant increased prevalence of urinary symptoms, particularly of stress UI, in SSc female Pts has emerged [4]. It is therefore conceivable that hormonal factors may act as a catalytic circumstance rather than pathogenic players in LUTS progression during SSc disease.

References: