patients (75%), presented by polyarthrities in 7/9 (77%), and oligoarthritis in 2/9 (22%). Two patients (16.7%) had cataract, one (8.3%) had bilateral uveitis, and one (8.3%) had optic atrophy. Sensorineural hearing loss was observed only in 3/12 (25%). Hydrocephalus was detected in 3/12 (25%). Delayed mental and psycho-speech development was observed in 6/12 (50%) patients. In 3/12 (25%), the development of MAS was recorded.

All patients had nucleotide variants in NLRP3 gene. According to NGS results and clinical characteristics, 8/12 (67.7%) patients were diagnosed with MWS and 4/12 (33.3%) had CINCA/NOMID syndrome. In children with MWS, heterozygous variant c.2113C>A in NLRP3 gene was the most common (5/8 (62.5%) patients). One of 8 (12.5%) patients with novel heterozygous variant c.2861C>T was detected; also one child (12.5%) have heterozygous variant c.943A>G. Four patients with CINCA/NOMID syndrome also had heterozygous variants in NLRP3 gene: c.5980G>A, c.2173C>A, c.1917T>C and c.769C>T.

Prior to genetic testing, 12/12 (100%) patients received NSAIDs; 6/12 (50%) were black, 10.2% were 'other' races and no patients were Asian. Median (range) of 246 patients in the analysis, 74.0% were female; 87.8% were white, 2% were female (80%). Mean disease duration at time of inclusion was 3.5 years. Mean age onset of Raynaud’s was 8.8 years and mean age of onset at the first non-Raynaud symptom before the age of 16 and were under the age of 18 at the time of inclusion. Patients were followed prospectively every 6 months with a standardized assessment.

Results: 39 patients in the JSSC cohort had 36 months follow up. 80% had a diffuse subtype. 96% of the patients were Caucasian origin. 31 of the patients were female (80%). Mean disease duration at time of inclusion was 3.5 years. Mean age onset of Raynaud’s was 8.8 years and mean age of onset at the first non-Raynaud’s was 9.5 years. Around 30% of the patients were anti-Scl70 positive and none of them show anti-centromere positive. The MRSS dropped from the time point of the inclusion into the cohort from 13.9 to 11.8 after 36 months. Pattern of organ involvement did not show any significant change, beside the increase of only mild to moderate changes from 49% to 73% (p=0.037). No renal crisis occurred. No mortality was observed. They were positive significant changes in the patient related outcomes. The physician global disease activity decreased from 40.0 to 22.1 assessed on a VAS scale of 0 to 100 (p <0.001).

Patients global disease activity decreased from 43.3 to 20.4 and patients global disease damage from 45.0 to 21.7 assessed on a VAS scale of 0 to 100 (p <0.001).

Conclusion: After 36 months follow up, we could observe a significant improvement of patient related outcomes and only significant change in organ pattern involvement. In a mostly diffuse subset patient population this is a very promising result regarding outcome.

Disclosure of Interests: Supported by the "Joachim-Stiftung".