research support from: The IRCCS Istituto Giannina Gaslini (IGG), where NR works as full-time public employee has received contributions (>10,000 USD each) from the following industries in the last 3 years: Bristol Myers Squibb, Eli Lilly, F Hoffmann-La Roche, GlaxoSmithKline, Janssen, Novartis, Pfizer, Sobi. This funding has been reinvested for the research activities of the hospital in a fully independent manner, without any commitment with third parties., Grant Schuert Speakers bureau: Novartis, Consultant of: SOBI, Alyssa Sproles: None declared, Sherry Thornton: None declared, Gabriel Vega Cornejo Speakers bureau: AbbVie, Grant/research support from: Bristol Myers Squibb, Eli Lilly, Novartis, P. Roche, UCB. Paid instructor for: AbbVie, Novartis, P. Roche, Consultant of: AbbVie, Bristol Myers Squibb, GlaxoSmithKline, Lilly, Novartis, Pfizer, Roche, UCB. Paid instructor for: AbbVie, Amgen, Gebro, GlaxoSmithKline, Lilly, Novartis, P. Roche, Roche, UCB. Paid instructor for: AbbVie, Novartis, P. Roche, Consultant of: AbbVie, Bristol Myers Squibb, GlaxoSmithKline, Lilly, Novartis, Pfizer, Roche, UCB, Michael Henrickson: None declared, Ivan Foeldvari Consultant of: Bristol Myers Squibb, Gilead, Hexal, MEDAC, Novartis, Pfizer, Sanofi, Daniel Kingsbury Consultant of: Pfizer, Margarita Askelson Consultant of: Currently working for Syneos Health providing services to Bristol Myers Squibb, Jin Qi Liu Consultant of: Bristol Myers Squibb, Employee of: Bristol Myers Squibb, Sanumta Mukherjee Consultant of: Bristol Myers Squibb, GlaxoSmithKline, Employee of: Bristol Myers Squibb, GlaxoSmithKline, Employee of: Bristol Myers Squibb, GlaxoSmithKline, Employee of: Bristol Myers Squibb, GlaxoSmithKline, Employee of: Bristol Myers Squibb, GlaxoSmithKline, Employee of: Bristol Myers Squibb, GlaxoSmithKline, Robert Wong Consultant of: Bristol Myers Squibb, Employee of: Bristol Myers Squibb, Daniel J Lovell Consultants bureau: Genentech, Wyeth Pharm, Consultant of: Abbott, Amgen, AstraZeneca, Boehringer Ingelheim, Celgene, GlaxoSmithKline, Hoffman-La Roche, Novartis, Pfizer, Regeneron, Takeda, Wyeth Pharm, Xoma, Alberto Martinis Consultants bureau: AbbVie, Novartis, Consultant of: AbbVie, Eli Lilly, EMD Serono, Idioma, Janssen, Novartis, Pfizer, Alexi Grom Consultant of: AbbVie, Novartis, NovoNpharm, Grant/research support from: AbbVie, Novartis, SoBiNpharm, Hermine Brunner Consultants bureau: GlaxoSmithKline, Novartis, Pfizer, Roche, Paid instructor for: Novartis, Pfizer (funds go to CCHMC/Employer), Consultant of: Boehringer Ingelheim, Bristol Myers Squibb, GlaxoSmithKline, Janssen, Merck, Novartis, Pfizer, Roche, UCB (funds go to CCHMC/Employer), Grant/research support from: Bristol Myers Squibb, Pfizer (funds go to CCHMC/Employer).

**REFERENCES:**


Acknowledgements: The National Paediatric Rheumatological Database has been funded by AbbVie, Chugai, Novartis and GSK.

Disclosure of Interests: Florian Milatiz: None declared, Martina Newrther: None declared, Jens Klotsche: None declared, Jana Hörstmann: None declared, Sandra Hansmann: None declared, Timmarn Kallinion: None declared, Christoph Rietschel: None declared, Ralf Trauzeddel: None declared, Joachim Peitz: None declared, Matthias Hartmann: None declared, Hermann Girschick: None declared, Kirsten Minden Speakers bureau: Pfizer, AbbVie, Consultant of: Novartis.

**DOIs:** 10.1136/annrheumdis-2021-eular.1081, 10.1136/annrheumdis-2021-eular.2356.
while 87.5% and 70% of patients had ANA positivity and dsDNA antibodies at the onset. The comparison between the groups with increased and normal IFN I-signature is presented in Table 1.

<table>
<thead>
<tr>
<th>Parameters</th>
<th>N=175</th>
<th>N=128</th>
<th>N=47</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age of onset of Raynaud’s years</td>
<td>10.0 (±3.8)</td>
<td>9.8 (±3.6)</td>
<td>10.6 (±4.3)</td>
<td>0.219</td>
</tr>
<tr>
<td>Mean age of onset of non-Raynaud’s years</td>
<td>10.2 (±3.9)</td>
<td>10.0 (±3.7)</td>
<td>10.9 (±4.3)</td>
<td>0.173</td>
</tr>
<tr>
<td>Disease modifying drugs</td>
<td>88% (154)</td>
<td>89% (114)</td>
<td>85% (40)</td>
<td>0.446</td>
</tr>
</tbody>
</table>

Conclusion: High IFN I-signature correlated with kidney involvement, ANA and RF-positivity, feritinemia, proteinuria and hematuria. Patients with high IFN I-signature received more aggressive treatment and needed longer glucocorticosteroid (GCS) treatment. More meticulous dynamic evaluation of IFN-signature is needed to clarify its role as a predictive and prognostic marker.

Acknowledgements: This work was supported by the RSF grant No 20-45-01005.

Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2021-eular.1956

Table 1. Comparison of demographic data and significant differences between dcJSSc and lcJSSc at time of inclusion.