Results: RTX therapy resulted in significant decrease of disease activity index, which statistically significantly correlated with decrease of mRSS - the main indicator of the severity of skin fibrosis (r=−0.39; p<0.001). Changes in parameters by follow-up periods are presented in the Table 1.

Table 1. Changes in clinical and instrumental parameters at RTX treatment (delta; median; lower quartile; upper quartile).

<table>
<thead>
<tr>
<th>Parameters</th>
<th>12-18 mo</th>
<th>24-30 mo</th>
<th>36-42 mo</th>
<th>48-54 mo</th>
<th>60-72 mo</th>
<th>SEMRSS</th>
<th>SEMQSS</th>
<th>SEMCCSS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Delta</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>mRSS</td>
<td>5.32 [4.0; 6.8]</td>
<td>5.4 [5.0; 6.8]</td>
<td>6.3 [5.0; 6.8]</td>
<td>8.3 [5.0; 6.8]</td>
<td>13.1 [11.0; 14]</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>p=0.001</td>
<td>p=0.001</td>
<td>p=0.001</td>
<td>p=0.001</td>
<td>p=0.001</td>
<td>p=0.001</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Activity index</td>
<td>0.19 [0; 0.3]</td>
<td>0.37 [0; 0.6]</td>
<td>0.39 [0; 0.6]</td>
<td>0.39 [0; 0.6]</td>
<td>0.39 [0; 0.6]</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>p=0.0009</td>
<td>p=0.0006</td>
<td>p=0.004</td>
<td>p=0.006</td>
<td>p=0.009</td>
<td>p=0.009</td>
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</tr>
</tbody>
</table>

Decreasing of mRSS statistically significantly correlated with increasing cumulative dose of RTX (r=−0.29; p<0.01). Decreasing disease activity index correlated with increasing cumulative dose of RTX (r=−0.37; p<0.01). IOS improvement was documented at all assessment time periods, although statistically insignificant.

Conclusion: The results of this study confirm reported positive effect of RTX on the reduction of skin fibrosis in SSc. Long-term follow-up demonstrated steadily decreasing skin fibrosis and improvement of microstructure with increasing oral aperture in parallel with a decrease of the disease activity index and increasing cumulative dose of RTX.

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OP0174 SUBCLINICAL INTERSTITIAL LUNG DISEASE IS FREQUENT AND PROGRESSES ACROSS DIFFERENT CONNECTIVE TISSUE DISEASES

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Background: Based on the argument that symptoms define disease, physicians commonly apply the terms “preclinical” or “subclinical” to describe patients with disease-related findings but no accompanying symptoms for connective tissue disease associated interstitial lung disease (CTD-ILD). The term subclinical frequently applies to patients with mild ILD changes on high resolution chest tomography (HRCT), normal forced vital capacity (FVC), and without respiratory symptoms. Previous work in systemic sclerosis (SSc)-ILD did show that patients with even minor extent of ILD at baseline often progressed and had increased mortality risk, suggesting that it is not appropriate to define these patients as “subclinical.”

Objectives: To identify the prevalence of subclinical ILD across CTD diagnoses, and assess the rate of progression of lung fibrosis compared to CTD without ILD and with CTD ILD.

Methods: All CTD patients, including SSc, anti-synthetase syndrome (ASS) and mixed connective tissue disease (MCTD) from the Oslo University Hospital diagnosed before 2015 and assessed for the presence of ILD by HRCT were included. The year 2015 was chosen to secure an observation time of at least five years from ILD diagnosis to study end on 01.01.2021 or time of death. All patients fulfilled the respective CTD classification criteria. Subclinical ILD was defined as an ILD extent ≤5% by semi-quantitative assessment of baseline HRCT, preserved lung function with FVC>80% predicted and without respiratory symptoms. Clinical ILD was defined as >5% extent of ILD or <5% extent of ILD on HRCT with respiratory symptoms or FVC<80%. The outcome was ILD progression, defined as increasing extent of ILD as >5% extent of ILD or <5% extent of ILD on HRCT with respiratory symptoms or FVC<80%. The outcome was ILD progression, defined as increasing extent of ILD as >5% extent of ILD or <5% extent of ILD on HRCT with respiratory symptoms or FVC<80%.

The results of this study confirmed positive effect of RTX on the reduction of skin fibrosis in SSc. Long-term follow-up demonstrated steadily decreasing skin fibrosis and improvement of microstructure with increasing oral aperture in parallel with a decrease of the disease activity index and increasing cumulative dose of RTX.

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OP0173 REDUCTION OF SKIN FIBROSIS IN SYSTEMIC SCLEROSIS ON RITUXIMAB TREATMENT. LONG-TERM FOLLOW-UP

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Background: A sound experience has been accumulated up to date with the use of rituximab (RTX) for treatment of systemic sclerosis (SSc). Some studies reported improvement of skin fibrosis following treatment with RTX, but long-term follow-ups are rarely focused on real-time follow-ups. The methods of this prospective study included 71 pts aged 46 years (17-66) on average, 59 (83%) pts were female, mean disease duration was 5.6±4.4 years, and mean follow-up - 42 months (12-72) (mo). Diffuse SSc was established in 42 (59%) pts. All pts received glucocorticoids in low doses. 40% of pts were receiving immunosuppressants at study entry. The following parameters were evaluated: Rodnan skin score (mRSS), interdigital space (IDS) (the distance between the tips of 1 and 5 fingers at maximum extension), oral aperture (OAp) and activity index (EScSG-Al) over the periods: 12-18 mo, 24-30 mo, 36-42 mo, 48-54 mo and 60-72 mo after initiation of RTX therapy. The results are presented as: mean values, delta (Δ), median, upper and lower quartiles.

Conclusion: Brentuximab vedotin already achieved the primary endpoint at 24 weeks, after half of the intended recruitment sample reached this landmark. A comparison with CRSG controls showed that mRSS only decreased significantly in patients treated with brentuximab. This interim report suggests that brentuximab vedotin might effectively improve skin involvement in patients with diffuse SSc and severe skin involvement.

REFERENCES:

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