Background:

The “Health Related Quality of Life” (HRQoL) or Quality of Life (QoL) is a term that became increasingly important during the last 15 years. It is not yet known however if children with Marfan syndrome are particularly impaired in different aspects of their QoL. We hypothesize that the QoL of patients with MFS is age-dependent due to increasing clinical features, and, compared to healthy children, reduced due to skeletal abnormalities, aortic root dilatation with consequent need for drug therapy and regular medical checkup.

Understanding the relationship between subjective QoL in children with MFS has important implications for preventive care strategies that may impact the outcome of HRQoL.

Patients and Methods:

220 children between 4-16 years were included in this study. 46 patients (4-16 years, mean age 10.8 years) completed the KINDL-R survey to assess QoL. Patients were included if MFS was determined genetically or clinically according to Ghent criteria, 39.1% were female, 60.8 % male. Additionally data of 174 kindergarten and school children were collected (4-16 years, mean age 10.9 years), 48.8% were female, 51.1% male. There was no significant difference between boys and girls. Samples were divided into 4 age groups and subjective QoL was compared. Additionally we created subgroups based on the main diagnostic criteria of the revised Ghent nosology.

Results:

<table>
<thead>
<tr>
<th>Age years (y)</th>
<th>MFS</th>
<th>Control</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>4-7 y</td>
<td>77.6 (95% ci 70.9-84.3)</td>
<td>77 (95% ci 73.1-80.8)</td>
<td>&gt;0.05</td>
</tr>
<tr>
<td>8-11 y</td>
<td>75.3 (95% ci 68.5-82)</td>
<td>74 (95% ci 70.8-77.1)</td>
<td>&gt;0.05</td>
</tr>
<tr>
<td>12-16 y</td>
<td>75 (95% ci 70.9-79.1)</td>
<td>68.4 (95% ci 66.2-70.6)</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>8-16 y</td>
<td>75.1 (95% ci 71.5-78.8)</td>
<td>70.4 (95% ci 68.6-72.3)</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>8-16 y</td>
<td>patients with lens luxation 81.6 (95% ci 75.2-88)</td>
<td>70.4 (95% ci 68.6-72.3)</td>
<td>&lt;0.05</td>
</tr>
</tbody>
</table>

Table 1. Quality of life Total Score in 220 children

Statistically significant differences could also be shown in test components “friends” and “school” (p<0.05) and subgroup analysis of MFS children with lens luxation (p<0.05).

Summary:

Surprisingly pediatric MFS patients showed partially higher HRQoL than children in the control group. MFS patients aged 12-16 years showed significant higher QoL than children in the control group although clinical features develop gradually.

QoL of Pediatric Patients with MFS is not reduced compared to healthy children.

Adolescent MFS patients showed significant higher QoL than children in the control group although clinical features impair gradually.

Children with MFS may be schooled through their environment to engage in more positive coping skills that increases their subjective QoL experience.

Conclusion:

Pediatric Patients with Marfan’s syndrome show normal or even higher QoL compared to our control group. Although symptomes of MFS patients develop over time, especially older MFS patients show significant higher QoL. A stable family and social environment may play an important role. Children with MFS may be schooled through their environment to engage in more positive coping skills that increases their subjective QoL experience.

References: