Repeated intra-articular GC injections were needed from 2012 etanercept was added for treatment with variable result. Inactive status of the infection function was recorded in 50% of cases, hypolacrimia – in one case. Recur- rent parotitis was present in only one case (7%). ANA were detected in 100% pts, anti-dsDNA – in 10 pts (71.4%), anti- Sm – in 7 pts (50.0%), anti-Ro – in 10 (71.4%), anti-La – in 7 (50%), RNP-70 – in 5 pts (35.7%), RF – in 6 pts (42.9%), hypocom- plementemia – in 3 pts (21.4%). The most common was the combination of positive ANA, anti-dsDNA, antiRo with acute cutaneous lupus, polyarthritids, generalized lymphopenath and expressed constitutional disorders – 8 pts (57%). 4 pts (28.6%) had polycional hypergammaglobulinemia. 3 pts (21.4%) had concomitant autoimmune non-rheumatic disease; 1 - autoimmune hepatitis, 1 - type 1 diabetes mellitus, 1 - autoimmune thyroiditis. Median disease activity by SLEDAI at the time of JSE verification was 15.9 scores [9,25;15,7].

Conclusion: According to our results, the frequency of detection of secondary SS in JLE was higher than the literature data. The clinical features include a high frequency of constitutional disorders, lymphopenath, skin manifestations, high frequency of antiRo with a significantly lower incidence of kidney involvement, sero- tipsis without SS. In pts with a diagnosis of SSE, the possibility of developing secondary SS should be considered (specially in girls with antiRo pos- itive). The early detection which affects the choice of therapy and prognosis. References: [1]Malagon C, Gomez M, Mosquera C et al. Juvenile polyauto- immunity in a rheumatology setting. Autoimmunity Reviews, Volume 18, Issue 4, 2019, p 369-381. https://doi.org/10.1016/j.autrev.2018.11.006.

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AB0097  IS HEIGHT ADJUSTMENT NECESSARY IN PEDIATRIC DENSITOMETRY IN ALL CHILDREN?

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Background: The current guidelines of the International Society for Clinical Densi- metry (1) recommend that in children with linear growth or maturational delay, Z score results should be adjusted. Height for age Z score (HAZ) adjustment is valid and can be calculated using the formula the formula proposed by Zemmel et al(2). It is possible that pediatric populations without linear growth or maturational delay, also benefit from HAZ, to prevent bone size from influencing the final Z score.

Objective: To evaluate Z score variability adjusted and without adjusting for height for age.

Methods: We analysed data from densitometry performed on patients 2-20 years of age, from 2016 to 2018, assessed in the pediatric rheumatology office of our hospital for presenting risk factors for low bone mass/osteoporosis. The HAZ was calculated according to Zemmel’s formula.

Results: Data from 103 patients are presented. Its characteristics are summa- rized in Table 1

Table 1.

<table>
<thead>
<tr>
<th>Mean age</th>
<th>9.8 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female</td>
<td>52.4%</td>
</tr>
<tr>
<td>Height Percentile ≤ 3</td>
<td>6.8%</td>
</tr>
<tr>
<td>Height Percentile ≥ 97</td>
<td>4.9%</td>
</tr>
<tr>
<td>LBM (Z score ≤ -2) spine</td>
<td>8.2%</td>
</tr>
<tr>
<td>LBM HAZ spine</td>
<td>6.4%</td>
</tr>
<tr>
<td>LBM whole body</td>
<td>10.5%</td>
</tr>
<tr>
<td>LBM HAZ whole body</td>
<td>7.2%</td>
</tr>
</tbody>
</table>

The table shows that the proportion of patients with BMD decreases in both the spine region and the whole body when adjusting for HAZ. When evaluating the relationship between densitometric measurements we found that spine Z score (ZsS) and whole body Z score (ZsWB) had a correlation coefficient of 0.73 (p<0.001). There were no differences between their averages (p=0.170).

At the LBM cut-off point (Z score ≤ -2) there were discrepancies in 7%, where 5% presented LBM in ZsWB but not in ZsS. The concordance index at this point was 0.557.

When comparing these measures with their HAZ adjusted equivalents, we observe:

HAZ adjusted ZsS vs ZsS without adjusting: There were no differences between their averages (p=0.913) with a correlation coefficient of 0.78 (p=0.001). Concordance index at cut-off point for LBM was 0.498, with a discrepancy of 7%, where 2% had HAZ adjusted to ZsS, but not to ZsS without adjusting.

HAZ adjusted ZsWB vs ZsWB without adjusting: There were no differences between their averages (p=0.367) with a correlation coefficient of 0.82 (p<0.001). Concordance index at cut-off point for LBM was 0.557, with a discrepancy of 7%, where 2% had LBM according to HAZ adjusted ZsWB, but not to ZsWB without adjusting.

Conclusion: There are discrepancies at the LBM cut-off point depending on the HAZ adjustment.

The pediatric population without linear growth or maturational delay, can also benefit from HAZ adjustment, especially those with high height percentiles in which their size can hide a diagnosis of LBM.

The youngest brother has started his therapy by methotrexate. It should be noted that the family has the eldest brother (20 y.o.), who has been suffering from arthritis since an early childhood with similar clinical picture. We are going to perform genetic analysis of the NOD2/CARD15 gene for the eldest brother.

Conclusion: Our clinical case shows that extremely rare BS may be misdiagnosed as JIA. Lack of efficacy of the etanercept therapy and uveitis de novo developing may be caused by genetic (non-idopathic) nature of disease. Classic triad of bogggy-arthritis, granulomatous uveitis and/or skin lesions without acute phase markers is required to perform genetic assay for the detection of a pathogenic mutation of the NOD2/CARD15 gene. This case is remarkable by the presence of BS in two (or 3) children of the family.

References: