Bone health index in children and adolescents with congenital adrenal hyperplasia.

H. Alsaffar, J. Reed, R. Davies, U. Das, S. Senniappan, M. Didi, J. Blair
Department of Endocrinology, Alder Hey Children's Hospital, Liverpool

Introduction
Patients with congenital adrenal hyperplasia (CAH) require life long glucocorticoid (GC) therapy. In CAH, the adverse effect of GC on bone health (BH) may be counteracted by the effect of modest elevations in adrenal androgens.

Aim
To examine relationships between BH index (BHI) standard deviation score (SDS), calculated by BoneXpert on bone age (BA) x-rays, BA, hydrocortisone (HC) dose (mg/m²/day), and mean 17-hydroxyprogesterone (17-OHP) concentration on 24 hour blood spot profile.

Method
Retrospective study of data collected during annual review. Data were analysed in two age groups: <10 years and ≥10 years. BA was reported according to Tanner Whitehouse II reference data.

Results
Data were available for 22 (11M) patients, age 10.7±3.4yrs. Results are reported in the following table.

<table>
<thead>
<tr>
<th>Age Range</th>
<th>Patients(gender) (episodes)</th>
<th>Age (yrs)</th>
<th>BA (TWII score)</th>
<th>BA-chronological age (yrs)</th>
<th>BHI SDS</th>
<th>Mean 17OHP (nmol/L)</th>
<th>Hydrocortisone (mg/m²/day)</th>
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</thead>
<tbody>
<tr>
<td>&lt;10yrs</td>
<td>12(7M:5F)(20)</td>
<td>6.9(3.15-9.93)</td>
<td>11.1(6.39-14.3)</td>
<td>3.47(-1.9 to +6.82)</td>
<td>-0.65(-4 to +2.3)</td>
<td>30.2(4-147.2)</td>
<td>10.4(7.5-16.6)</td>
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<tr>
<td>≥10yrs</td>
<td>15(8M,7F)(31)</td>
<td>12.75(10.2-17.5)</td>
<td>14.9(9.22-18.1)</td>
<td>2.06(-1.6 to +5.14)</td>
<td>-0.3(-2.9 to 2.5)</td>
<td>23.9(0-230.3)</td>
<td>10.0(5.8-17.3)</td>
</tr>
</tbody>
</table>

Data are reported as median (range)

BA was advanced in both age groups. BHI-SDS was < -1.5 in four subjects (18%). BHI-SDS was not related to either HC dose or 17OHP concentrations. BHI-SDS correlated positively with chronological age and BA advance (BA – chronological age) in age <10yrs, r=0.48 and r=0.35 respectively, and with BA for all patients (r=0.55, p<0.0001).

Conclusion
As anticipated, we observed modest BA advance, consistent with modestly elevated 17OHP concentrations. BHI SDS was < -1.5 SDS in 18% of patients, suggesting a negative effect of HC persists despite the protective effect of elevated androgens. These relationships may be more evident in a larger cohort of patients. The clinical significance of this observation, if any, is unknown. However, osteoporosis in adult patients with CAH is reported and this observation deserves further evaluation in a larger cohort, studied prospectively.