Dr Seino comments:

As previously stated, the mean plasma levels of 1,25(OH)$_2$D are significantly low in hypophosphataemic vitamin D-resistant rickets and increase after treatment, but remain relatively low in spite of large doses of 1α-OHD$_3$ (0.4–2.0 μg/kg per day). These results suggest that the metabolism of 1,25(OH)$_2$D$_3$ is accelerated in such patients. The relatively low level of 1,25(OH)$_2$D$_3$, even after administration of a massive dose of 1α-OHD$_3$, indicates that large doses are necessary in these patients. Moreover, the low level of 1,25(OH)$_2$D$_3$ before treatment showed a positive correlation with the low level of serum phosphate and TmP/GFR. This finding suggests that massive doses of 1α-OHD$_3$ are necessary in severe cases. I suppose that the case referred to by Kristiansen and Pedersen was mild, as commonly occurs in females. The risk of hypercalcaemia is always present during treatment with vitamin D metabolites. However, hypercalcaemia or hypercalciuria (urinary calcium/creatinine appears to be the best indicator) is easily controlled by reducing the dose of 1α-OHD$_3$ because it has a short half-life.

References


Somatomedin-C in the Beckwith-Wiedemann syndrome

Sir,

We refer to the paper by Spencer et al. We have obtained fasting sera for growth hormone and somatomedin-C on 3 unrelated patients with classical features of the Beckwith-Wiedemann syndrome. The results are shown in the Table.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Growth hormone</th>
<th>Somatomedin-C* (U/ml)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Patient</td>
<td>Normal</td>
</tr>
<tr>
<td>1</td>
<td>3 weeks</td>
<td>F</td>
<td>—</td>
<td>0.1</td>
</tr>
<tr>
<td>2</td>
<td>1 year</td>
<td>F</td>
<td>2</td>
<td>0.19</td>
</tr>
<tr>
<td>3</td>
<td>8 years</td>
<td>F</td>
<td>12</td>
<td>0.26</td>
</tr>
</tbody>
</table>

*By radioimmunoassay.

The growth hormone determinations were performed by Bio-Science Laboratories, Van Nuys, California, and the Somatomedin-C determinations by Nichols Institute, San Pedro, California, which also supplied the normal range. The very low values of Somatomedin-C in our patients do not support the concept proposed by Spencer et al. No explanation for the conflicting findings is offered.

Reference


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Dr Spencer comments:

The results presented by Robinow and Shafer are interesting but it is difficult to explain why they conflict with ours. However, since submitting our article, I have been able to test somatomedin activity in another patient with Beckwith-Wiedemann syndrome. The patient was 7 years old and had a bioassayable plasma somatomedin activity of 1·6 U/ml. This value, although not as high as that in the patient we reported earlier, is still raised compared with normal age-matched children; our range...