



² Seino Y, Shimotsuji T, Ishida M, Ishii T, Yamaoka K, Yabuuchi H. Vitamin D metabolism in hypophosphatemic vitamin D-resistant rickets. *Contrib Nephrol* 1980; **22**: 101-6.

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Somatomedin-C in the Beckwith-Wiedemann syndrome

Sir,

We refer to the paper by Spencer *et al.*¹ 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.

Case	Age	Sex	Growth hormone	Somatomedin-C* (U/ml)	
				Patient	Normal
1	3 weeks	F	—	0.1	0.2-2.0
2	1 year	F	2	0.19	0.2-2.0
3	8 years	F	12	0.26	0.4-2.0

*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.*¹ No explanation for the conflicting findings is offered.

Reference

- Spencer G S G, Schabel F, Frisch H. Raised somatomedin associated with normal growth hormone. A cause of Beckwith-Wiedemann syndrome? *Arch Dis Child* 1980; **55**: 151-3.

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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,¹ 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

Dr Seino comments:

As previously stated, the mean plasma levels of $1,25(\text{OH})_2\text{D}$ are significantly low in hypophosphatemic vitamin D-resistant rickets and increase after treatment, but remain relatively low in spite of large doses of $1\alpha\text{OHD}_3$ ($0.4-2.0 \mu\text{g}/\text{kg}$ per day).¹⁻² These results suggest that the metabolism of $1,25(\text{OH})_2\text{D}_3$ is accelerated in such patients. The relatively low level of $1,25(\text{OH})_2\text{D}$, even after administration of a massive dose of $1\alpha\text{OHD}_3$, indicates that large doses are necessary in these patients. Moreover, the low level of $1,25(\text{OH})_2\text{D}$ before treatment showed a positive correlation with the low level of serum phosphate and TmP/GFR.² This finding suggests that massive doses of $1\alpha\text{OHD}_3$ 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 $1\alpha\text{OHD}_3$ because it has a short half-life.

References

- Seino Y, Shimotsuji T, Ishii T, *et al.* Treatment of hypophosphatemic vitamin D-resistant rickets with massive doses of 1α -hydroxy-vitamin D_3 during childhood. *Arch Dis Child* 1980; **55**: 49-53.