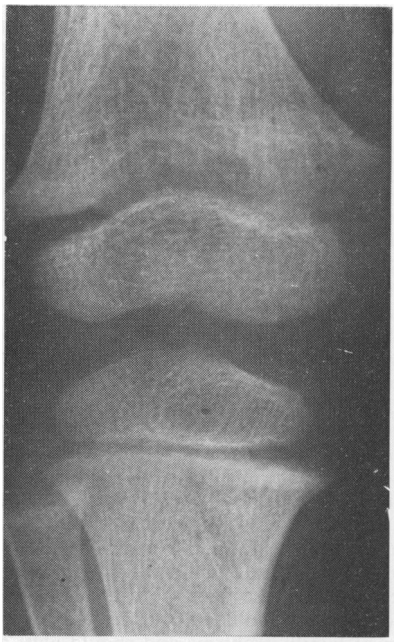

(b)

Dr Seino comments:

As previously stated, the mean plasma levels of $1,25(OH)_2D$ are significantly low in hypophosphataemic vitamin D-resistant rickets and increase after treatment, but remain relatively low in spite of large doses of $1\alpha OHD_3$ (0·4–2·0 μg/kg per day).[1–2] These results suggest that the metabolism of $1,25(OH)_2D_3$ is accelerated in such patients. The relatively low level of $1,25(OH)_2D$, even after administration of a massive dose of $1\alpha OHD_3$, indicates that large doses are necessary in these patients. Moreover, the low level of $1,25(OH)_2D$ before treatment showed a positive correlation with the low level of serum phosphate and TmP/GFR.[2] This finding suggests that massive doses of $1\alpha OHD_3$ 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 $1\alpha OHD_3$ because it has a short half-life.

References

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

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.

YOSHIKI SEINO
Department of Paediatrics,
Osaka University Hospital,
Fukushima-ku,
Osaka 553, Japan

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

1 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.

MEINHARD ROBINOW AND ALAN D SHAFER
Department of Pediatrics and Surgery,
Wright State University School of Medicine,
Children's Medical Center,
1735 Chapel Street,
Dayton, Ohio 45404, USA

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