Letters to the Editor

More on the Beckwith-Wiedemann syndrome

Sir,

We read with interest the article on Beckwith-Wiedemann syndrome: a quantitative, immunohistochemical study of pancreatic islet cell populations by Stefan et al. [1].

The authors suggest that the Beckwith-Wiedemann (BW) syndrome is a separate entity of neonatal hyperinsulinaemic hypoglycaemia characterized by a massive increase in insulin and glucagon cells together with a defect in glucagon and PP-cell segregation to distinct pancreatic regions.

We recently delivered a baby with the BW syndrome, already prenatally suspected by a combination of polyhydramnios, omphalocele and pronounced oedema of the placenta.

In order to obtain information on lung maturity, an amniocentesis was performed at 34 weeks of gestation. C-peptide was 0.25 pmol/ml and insulin 5 μ U/ml of amniotic fluid, both values being low. The mother went into spontaneous labour at 35 weeks of gestation. Polyhydramnios of more than 3 l was present. The birthweight was 3,500 g. The baby, a boy, survived and had the typical aspect of the BW syndrome. During the first day of life, blood Dextrostix values were around 1.8 mmol/l under a continuous glucose infusion of about 4 mg $\cdot$ kg⁻¹ $\cdot$ min⁻¹. Plasma insulin was 8 μ U per ml and C-peptide 0.47 pmol per ml.

Although our patient had the typical aspect of the BW syndrome with marked macrosomia, amniotic and plasma insulin and C-peptide

levels were not excessive during fetal and neonatal life respectively. There was also no hypoglycaemia under administration of glucose in usual dosages [1].

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References

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Vitamin D deficiency and the endocrine pancreas

Sir,

Gedik and Akalin [1] have demonstrated that insulin release following glucose challenge is impaired in patients with vitamin-D deficiency, and that it is restored to normal following vitamin-D treatment. This phenomenon is well established [2], but it is not established that vitamin D deficiency has no effect on glucagon release.

There are a few unusual features about the patients studied by Gedik and Akalin [1] which should be highlighted. All of their vitamin-D deficient patients had plasma calcium concentrations in the normal range. This can occur in vitamin-D deficiency, but is usually associated with secondary hyperparathyroidism. Parathyroid hormone (PTH) concentrations in sera of these patients were not elevated. This is particularly surprising since the patients had severe vitamin-D deficiency according to clinical and radiological criteria. Furthermore, PTH concentrations did not alter following treatment; this is most unusual. We have previously described the persistence of PTH hypersecretion following vitamin D therapy in osteomalacia, but even in this select group of patients there was some fall in PTH [1]. We have recently demonstrated a significant inverse correlation between 1,25 dihydroxyvitamin D [1,25(OH)₂D] and PTH concentrations (Fig. 1) in patients with vitamin D deficiency [4]. It is surprising, therefore, not to find elevated PTH concentrations in association with the low concentrations of 1,25(OH)₂D described. It is also relevant to mention that we have shown a significant inverse relationship between bone density and PTH concentrations in patients with vita-

Fig. 1. Geriatric study