Brittle or battered?

Sir,

My attention has been drawn to correspondence about this subject. There is reference to an article in which I reported study of a series of children with osteogenesis imperfecta with special reference to the occurrence of metaphyseal fractures.1

In his letter Dr Blumenthal states that 'metaphyseal fractures are a feature of both abuse and brittle bones'.2 This is true but needs qualification. In non-accidental injury the bones appear grossly normal and the metaphyseal fractures are often numerous. In osteogenesis imperfecta fractures of this type are infrequent and rarely number more than one or two. They were only seen in my series where there was gross abnormality of the skeleton and the diagnosis of the presence of a systemic bone disease was apparent at a glance.

To quote my last sentence, 'Confusion with non-accidental injury did not occur'.

References


Somatostatin analogue in short term management of hyperinsulinism

Sir,

We are interested by the report of Kirk et al of the use of somatostatin analogue for the short term management of hyperinsulinism in which a fall in glucose requirement of 4-6 mg/kg/minute was seen.3 We have previously reported similar success, in the short term control of hyperinsulinism in an infant with nesidoblastosis, using growth hormone twice daily by subcutaneous injection.2 A fall in glucose requirement of 5-5 mg/kg/minute was seen. The effect of growth hormone in the treatment of hypoglycaemia is reported elsewhere.3,4

At the time of our report further studies of this use of growth hormone were precluded by the withdrawal of pituitary growth hormone after reports of Creutzfeldt-Jakob disease. Now that a growth hormone produced by recombinant DNA technology is available, however, the choice of growth hormone over a somatostatin analogue has advantages.

Somatostatin has a very broad range of endocrine activity,1 with unwanted effects upon other endocrine axes that do not appear to be produced by growth hormone. Furthermore, on simply practical grounds growth hormone, unlike somatostatin, is available in most regional centres.

In the infant with hyperinsulinism, when it is necessary to temporise before surgery, we believe that growth hormone may be preferable, and its use certainly merits further study.

References


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Disialotransferrin developmental deficiency syndrome and olivopontocerebellar atrophy

Sir,

I read with great interest the paper of Kristianssen et al about disialotransferrin developmental deficiency syndrome.1 I would like to draw attention to the remarkable similarity between this syndrome and a condition recently reported in two siblings as olivopontocerebellar atrophy with neonatal onset.2 Common features included failure to thrive, hypotonia, developmental delay, joint restrictions, pericardial effusions, mild non-progressive liver disease, retinal dystrophy, and cerebellar hypoplasia.

Moreover there was thyroxine binding globulin deficiency in the patients reported by Harding et al,2 and although serum concentrations of thyroxine binding