Correspondence

hormone response to provocative stimuli of more than 15 mU/l would exclude a positive response to long term treatment and could result in treatment being withheld from patients who might benefit from it.

References

J M Wit
University Hospital for Children and Youth,
Het Wilhelmina Kinderziekenhuis,
Utrecht,
The Netherlands

Dr Forsyth and co-workers comment:

Dr Wit in his letter makes several points requiring an answer.
(1) He wishes clarification of the investigation of growth hormone deficiency. The purpose of the two papers on exercise tests from Dundee and Newcastle was to emphasise the usefulness of exercise, a physiological stimulus, in the early investigation of growth hormone deficiency, provided the exercise is carried out in a scientific manner. The Dundee group mentioned only insulin stimulation for those later diagnosed as growth hormone deficient, as this was true for the nine children concerned. The most recent of these, however, was diagnosed in 1979 and thereafter opinion moved away from the use of insulin towards sleep, clonidine, L-dopa, arginine, and glucagon tests in the various United Kingdom growth centres.
(2) He challenges the classic definition of growth hormone deficiency, which, allowing for the different biochemical tests now in use, is at present the definition accepted by the Human Growth Hormone Health Services Committee. In addition, height prediction studies in relation to mid-parental height and assessment of the hypothalamo-pituitary axis are taken into consideration.
(3) He favours the use of 24 hour growth hormone profiles in children with short stature or low growth velocity, or both, and growth hormone responses greater than 15 mU/l, in order to confirm or exclude growth hormone deficiency and he suggests a definition based on this test.

With regard to confirmation of growth hormone deficiency, or at least of the benefit of treatment, the Human Growth hormone Health Services Committee follows up all children on growth hormone in the United Kingdom through a coordinator, and these children have to show a satisfactory increase in height velocity during the first year of treatment in comparison with that during the previous year.
The use of a 24 hour sleep test as the final method of excluding growth hormone deficiency would create difficulties. Such a test is not easy to perform, especially during the night phase, when nervous children do not sleep deeply with an indwelling cannula in situ and, if used as a final definitive test, it would require routine electroencephalographic monitoring for the accurate assessment of the sleep pattern.

(4) He suggests that a therapeutic trial of growth hormone therapy for short, slowly growing children with normal peak growth hormone responses would be valuable. Such a trial is in progress in the United Kingdom as outlined by Professor Milner, Chairman of the Human Growth Hormone Health Services Committee, in his article.1

Dr Brook comments:

I agree with Dr Wit that a 24 hour profile is likely to produce the most clinically relevant information on short children growing slowly. The performance of such a profile is unfortunately beyond the capacity of the majority of departments of paediatrics in this country. Since it is very difficult to justify the performance of profiles on normal patients, it may be quite difficult to assess the results from a single child.

Where there is the slightest doubt about the possible use of growth hormone, a centre specialising in the management of such cases should be consulted.

Complications of diazoxide in the treatment of nesidioblastosis

Sir,

McGraw and Price attribute the problem of cardiac failure in a patient with nesidioblastosis to a side effect of diazoxide.1 Like the author, this unit recently encountered a patient who developed cardiac failure while being treated for nesidioblastosis. A girl, birthweight 3.9 kg had a convulsion at 6 hours of age attributable to a blood sugar value of less than 1 mmol/l. Despite treatment with intravenous glucose and steroids it proved difficult to maintain normoglycaemia. Insulin concentrations were grossly raised at 90 mU/l. Medical treatment of the hyperinsulinism consisted of diazoxide, hydrocortisone, and glucagon. At the age of 2 weeks she had the first of many episodes of cardiac failure. Cardiac echo showed a structurally normal heart with poor myocardial contractility as seen in a cardiomyopathy. Treatment with digoxin and diuretics was begun but the hypoglycaemia and cardiac failure remained difficult to control and at age 5 weeks the child underwent subtotal pancreatectomy. Histology confirmed nesidioblastosis. Unfortunately, despite being able to maintain normoglycaemia relatively easily after the operation, the baby developed pseudomonas septicaemia and died.

The above case illustrates some of the problems that can arise when treating a patient with nesidioblastosis. Although the cardiac failure encountered in these patients may be partly attributable to diazoxide as McGraw and
Price suggest, the influence of fluid overload given in an effort to maintain normoglycaemia might be important, as might corticosteroid administration causing water and salt retention. Another important aspect is the poor myocardial function, perhaps secondary to continued hyperinsulinism as seen in infants of diabetic mothers. Appropriate diuretic treatment in patients with nesidioblastosis might reasonably include a thiazide diuretic which, as well as producing a diuresis, may increase the blood sugar value. I would therefore like to suggest a thiazide diuretic might reasonably be used in conjunction with diazoxide when this is required for the treatment of hyperinsulinism.

References


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General Infirmary at Leeds,
Belmont Grove,
Leeds LS2 9NS
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D R Gillies

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