values make interpretation of coagulation tests unreliable, but our diagnosis of dysfibrinogenaemia was based on the grossly prolonged reptilase time compared with the thrombin time. If simple hypofibrinogenaemia were responsible, we would have expected both times to have been similarly prolonged. Furthermore, the coagulation factor assays are independent of the patient's fibrinogen value. As regards the diagnosis of congenital or hereditary tyrosinaemia, the record needs to be put right. It is correct to say that tyrosinaemia (with abnormal excretion of phenolic acids) and hypermethioninemia alone, do not indicate the diagnosis. In all three infants reported, not only was there evidence of acute liver necrosis but also of severe proximal renal tubular disease characterised by proteinuria, gross generalised aminoaciduria, phosphaturia, hypophosphataemia, and early rickets accompanied by considerably raised serum alkaline phosphatase, mainly of bone origin. The galactose-1-phosphate uridyl transferase deficiency galactosaemia was excluded in all, and none of the infants received fructose in the diet or in medication.

These observations strongly favour the diagnosis of congenital or hereditary tyrosinaemia of acute variety; unfortunately necropsy in the first two patients was not available. As for galactokinase deficiency galactosaemia, liver disease does not occur in this disorder.

References

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Lest we forget

Sir,

Your editorial was a splendid reminder to us to be primarily people-oriented. Your question, 'How often do we in our postgraduate and in-service training discuss the problems of children in hospital, their psychological needs, and the needs and the problems of unrestricted access for parents?' is timely.

Now we are in the third decade of 'deinstitutionalisation' of the severely mentally handicapped, do we need to ask a similar question? 'How often do we in our postgraduate and in-service training discuss the problems of severely subnormal children living at home, the physical and psychological needs, and all the problems of parents continuing to care for over grown toddlers who are extraordinarily slow to learn?'

How often do we reflect on the needs of the mothers of the severely handicapped? How often do we consider her fourfold role as: (a) Mother of the handicapped child; (b) Mother of the other child/children; (c) Wife; (d) Needing time/space to be 'herself'.

If we are to support the severely subnormal in the community then we must be acutely aware of the needs and stresses of those who provide day and night cover, too often, without a break.

Reference
1 Anonymous. Lest we forget. [Editorial]. Arch Dis Child 1985;60:93.

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Who should get growth hormone?

Sir,

The four papers on the assessment of potential candidates for growth hormone treatment contain some contradictions that may lead to misunderstandings among readers. These contradictions originate mainly from different definitions of growth hormone deficiency.

The two papers on exercise tests emphasise the latter half of the classic definition, that is the combination of a growth velocity below the 25th centile for age or bone age and a growth hormone response to adequate insulin hypoglycaemia under 15 mU/l. The commentary stresses the first half of this definition. Low growth velocity is considered as an absolute prerequisite for the diagnosis, but no indication is given as to which growth velocity is abnormally low. If the classic 25th centile were used, 25% of the children would have to be tested.

There is increasing evidence that the classic definition has lost its usefulness. Studies on 24 hour growth hormone profiles in short children have shown that the results of provocation tests are poor parameters of growth hormone secretory status and poor predictors of growth response to long term treatment.

In my opinion, growth hormone deficiency should be defined as the condition in which either the total 24 hours' production of growth hormone, or the frequency or amplitude of peaks, or both, is below normal. This does not imply that the classic criteria (growth velocity and biochemical tests) would lose their role in the diagnostic process. In case of low growth velocity and low responses to provocative stimuli, growth hormone deficiency can be diagnosed without 24 hour profiles. Such profiles, however, could be made 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. In case of normal growth hormone production a therapeutic trial might still be indicated, as children with abnormal growth hormone molecules or partial receptor defects seem to respond favourably to treatment.

In conclusion, in one issue of the Archives growth hormone deficiency is defined in the classic way as well as in a more updated way. Suggestions that growth...