Paediatric Research Society

Spring Meeting, Radcliffe Infirmary, Oxford, 30 and 31 March 1973

Pituitary-adrenal function in thalassaemia major. N. McIntosh. Whittington Hospital, London N.19.

Thalassaemia major is an iron-loading disease produced by gastrointestinal iron absorption and by the iron load of repeated transfusions. The distribution of iron is similar to that seen in haemochromatosis with the notable absence of iron deposition in the skin, despite the invariable slate grey skin colour seen in the condition. An investigation was undertaken of the effect of iron deposition in the pituitary and adrenal glands in 9 children with thalassaemia major. The age range of the children was 6 to 13 years. 8 were treated by a high transfusion regimen, the 9th has only been transfused once; 6 of the 9 have been chelated with their transfusions, and 5 of these are chelated daily. All chelation is with desferrioxamine B.

Pituitary function tests have shown no evidence of decreased secretion; in all the children high levels for 9 a.m. immunoreactive adrenocorticotrophic hormone have been found.

24-hour 17-ketosteroid and 17-hydroxycorticoid excretion was normal. Plasma cortisol levels taken after stimulation with 40 units Acthar Gel showed a normal response in 6 patients and a diminished response in 3. The physiological tetracosactrin stimulation test (Landon et al., 1967) showed impaired cortisol response in all subjects after administration of 31-25 and 62-5 ng, and in 5 out of 8 subjects after 125 ng. All children had a normal response to 0-25 mg.

These results suggest that in children with thalassaemia major there is partial adrenal insufficiency with high circulating adrenocorticotrophic hormone levels. This probably accounts for the skin pigmentation in this disease.

Reference


Adrenocortical function in normal, precocious, and delayed puberty. C. Forsythe, D. C. L. Savage, J. Cameron, and E. McCafferty. Department of Child Health, University of Dundee, Dundee.

There is a steady rise in the 24-hour urinary excretion of the 17-hydroxycorticosteroids and, more specifically, of the a-ketolic metabolites of cortisol throughout infancy, childhood, and adolescence which is directly related to body weight.

In contrast, there is a steep rise in the 24-hour urinary excretion of the 17-oxosteroids and more specifically, of dehydroepiandrosterone, androsterone and androsterone in relation to puberty. This increase is much greater than could be explained on a body weight basis, and it occurs late in delayed puberty and early in precocious puberty.

The metabolic degradation, which takes place chiefly in the liver, of the adrenal androgens alters at puberty, favouring the production of 5a-steroids rather than 5b-steroids. The rise in the 5a/5b ratio with age for the 17-oxosteroids measured by our method is statistically significant, and the rise in this 5a/5b ratio occurs early in precocious puberty and late in delayed puberty.

Data were presented from 62 normal infants, children, and adolescents and, in addition, from 10 patients with early or precocious puberty and from 7 patients with delayed puberty on which the above conclusions are based.

The changes in adrenocortical function at puberty are thought to be associated with activity of the gonadal axis, but the precise relation is not fully understood.

Plasma and urine testosterone responses to human chorionic gonadotrophin (HCG) in children with delayed puberty. B. T. Rudd, P. H. W. Rayner, and S. K. M. Jivani. Department of Paediatrics, the Children's Hospital, Birmingham 16.

The plasma testosterone and/or urine testosterone levels after 1500 units HCG for 4 days were measured in 29 male children with clinical evidence of delayed puberty due to the following causes. 10 children with constitutional delayed puberty (11 years 11 months to 16 years); 10 with cryptorchidism (6 bilateral) (9 years 8 months to 16 years); 4 with Prader-Labhart-Willi syndrome (11 to 13 years 6 months); and 5 with primary testicular atrophy (8 years 6 months to 16 years 2 months). 2 of the children in this group were anorchid on surgical exploration. The 10 children with constitutional delayed puberty and all but 3 of the 10 children with cryptorchidism had similar mean incremental rises in testosterone (range for plasma testosterone 33-298 ng/100 ml, urine 1-1-28-3 μg/24 hr).

Of 4 children with Prader-Labhart-Willi syndrome, 3 had a poor urine testosterone response to HCG (incremental rise 0-1-2-1 μg/24 hr). The 5 children with primary testicular atrophy failed to respond to HCG and were clearly separated from the other three groups. There was a small but positive correlation between the
Pituitary-adrenal function in thalassaemia major.

N McIntosh

Arch Dis Child 1973 48: 653
doi: 10.1136/adc.48.8.653