Patients with Turner's syndrome enter adult life with an overall deficiency of compact bone. Further studies are needed to assess the long-term significance of this and the possible influence on it of oestrogen therapy.

P. H. CHAPMAN. Royal Hospital for Sick Children, Glasgow. 'Prognostic significance of androgen excretion as measured by testicular function test.' In this investigation Leydig cell function was assessed by measuring plasma testosterone, urinary testosterone, and urinary androgens before and during stimulation with human chorionic gonadotrophin (6000 IU/day intramuscularly) for 3 days. From experience it was found that several of the measured androgens correlate well with the phenotype of the patient, and may have prognostic value. Representative cases will be shown to demonstrate these points, which may be stated biochemically thus: 5α-Androstan-3α, 17β-diol (5α-A-diol) is a hepatic metabolite of both testosterone and 5α-dihydrotestosterone and at puberty the urinary excretion of 5α-A-diol increases rapidly, the rise being related to an increased utilization of testosterone by testosterone-dependent tissues. Thus, a good androgenic status means adequate testosterone production in association with a high urinary excretion of 5α-A-diol. 5β-Androstan-3α, 17β-diol (5β A-diol) is associated with the development of the external genitalia. Good genital status is indicated by a high urinary excretion of 5β A-diol. When 5β A-diol is high, even in association with small external genitalia, the indication is that the external genitalia will develop provided there is an adequate supply of testosterone. 5β A-diol then may have prognostic value.

5α Androstan 3βol, 17-one (epiandrosterone) is a 17-oxosteroid metabolite of dehydroepiandrosterone (DHA) having retained the 3β-hydroxyl group. If the general tissue metabolism cannot utilize testosterone, as an alternative DHA becomes the principal anabolic hormone and the urinary excretion of epiandrosterone then increases. When testosterone is utilized as the anabolic hormone urinary epiandrosterone is low. Thus, a low urinary excretion of epiandrosterone indicates good somatic status.

D. B. GRANT. The Hospital for Sick Children, Great Ormond Street, London. 'Two cases of micropenis with rudimentary testes.' Two patients, aged 3 months and 1 month, with the syndrome of rudimentary testes and micropenis (Bergada et al., 1962) were presented. In both patients an extreme degree of micropenis was associated with an empty, hypoplastic scrotum. Both showed a normal male karyotype (XY).

HCG stimulation (5000 units x 3 days) was carried out in one patient. There was no significant change in either plasma testosterone or urinary steroids after HCG. At laparotomy tests could not be identified in either patient. In view of the extreme micropenis it was decided to rear both patients as girls and vulvoplasty with division of the scrotum was carried out by Mr. D. I. Williams.

British Paediatric Association

REFERENCE


G. M. Komrower. Royal Manchester Children's Hospital. 'Precocious puberty in association with pineal seminoma.' Case history of sexual precocity in a male child of 9 years 5 months. Duration of symptoms 4 months. Features: great increase of height and muscle bulk—deepened voice—pubic and facial hair—considerable penile enlargement without corresponding testicular growth. No behavioural or neurological symptoms or signs. Initial investigations revealed a significant increase of urinary 17-ketosteroids, testosterone, and gonadotrophins; a marked rise of plasma testosterone and alkaline phosphatase. Initial bone age was 9½ years but within 2½ months this advanced to 13 years with a growth spurt of 5·6 cm. X-ray of skull showed calcification in the pineal region and detailed studies showed a clearly defined and isolated pineal tumour. Suprarenal and thyroid function was normal but high levels of human growth hormone were determined. Cyproterone acetate therapy was started but after one month the boy complained of headache, and on ophthalmoscopy ven-triculography there was evidence of encroachment on the aqueduct of Sylvius and the tumour was removed—apparently intact. It was a pinealoma of the malignant seminoma type. After the operation the boy developed a homonymous hemianopia which subsequently has improved.

Measurements of luteinizing hormone and testosterone have been made before and during cyproterone therapy and also after the removal of the tumour. Further studies were initiated to determine whether the pinealoma was acting as an autonomous tumour or whether its effect was produced by hypothalamic disturbance. The boy has had a course of deep x-ray treatment.

D. C. L. Savage. Department of Child Health, Dundee. 'Excretion of individual adrenocortical steroids in obese children.' (To be published.)

P. H. W. Rayner and J. M. Court. Institute of Child Health, Birmingham. 'Effect of dietary restriction and anorectic drugs on linear growth in childhood obesity.' Simple obesity in childhood is associated with advancement of linear growth. The effect of a reduced calorie intake and anorectic drugs on the growth of obese children has received less attention.

The growth of 26 obese children (17 girls aged 3 years 9 months to 10 years 3 months, and 9 boys aged 3 years 10 months to 12 years 1 month) has been studied over periods of at least one complete year. Growth velocity, expressed as a percentage of the 50th centile velocity for age, has been analysed in terms of sex, weight change, skinfold thickness change, and therapeutic regimen. Three therapeutic regimens were assessed: diet (1000 cal) alone, or diet plus amphetamine derivatives (chlorphentermine, diethyl propion), or diet plus fenfluramine.
Children who lost weight, regardless of therapeutic regimen, showed a reduced growth velocity (90·1±22·2%) compared to those who gained weight (99·6±24·5%) but the difference was not statistically significant and did not correlate with the amount of weight lost. Children who received diet plus fenfluramine showed a mean growth velocity of 82·3 compared to 99·5±23·2% for diet alone and 99·9±26·4% for diet plus chlorphentermine or diethylpropion. The difference was only significant at the 10% level.

In view of the doubtful value and possible dangers of anorectic drugs in childhood, careful monitoring of the growth of obese children treated with these agents is indicated.

British Association of Paediatric Nephrology

C. CHANTLER, J. S. CAMERON, R. H. R. WHITE, and C. S. OGG. Guy’s Hospital, London and Birmingham Children’s Hospital. ‘Long-term stability of remission in the nephrotic syndrome after treatment with cyclophosphamide’. 57 children with the nephrotic syndrome and minimal changes on renal biopsy, who had relapsed repeatedly and showed corticosteroid-induced toxicity, were treated with cyclophosphamide from 1966 to 1969. At that time an initial dose of 5 mg/kg per day was used, and leucopenia maintained for an average of 12 weeks. All these children have now been followed for more than 4 years since the end of their treatment, some for 7 years.

At present, 18 children still remain in remission, 35 have relapsed, and 4 are dead, 2 in relapse. One child died of measles pneumonia shortly after treatment with cyclophosphamide, and another died of cerebral tumour in 1973. One other child developed Hodgkin’s disease also in 1973, aged 19.

The rate of relapse has been exponential over the first 5 years after treatment with a half-time of 3 years. This compares favourably with the near 100% relapse rate in similar children treated with corticosteroid withdrawal alone, but it is no better over the first 2 years than cyclophosphamide treatment at 3 mg/kg per day for 8 weeks. There was no suggestion that remissions were more stable in those treated for longer periods, nor did stability of remission relate to age or the duration of disease before treatment. These data permit a better assessment of benefit versus toxicity in the treatment of relapsing nephrotic children.

S. R. MEADOW. Department of Paediatrics and Child Health, Leeds. ‘Poststreptococcal nephritis—a rare disease?’. (To be published in full in the Archives.)

M. J. DILLON and JENNIFER RYNESS. The Hospital for Sick Children and Institute of Child Health, London. ‘Plasma renin activity and aldosterone concentration in children’. The central role of the renin angiotensin aldosterone system in the control of salt balance and blood pressure is well established. However, only limited data are available in children because of the large volumes of blood hitherto required for the estimation of plasma renin activity and aldosterone concentration.

For this reason, semimicro methods for the measurement of plasma renin activity (PRA) by radioimmunoassay (angiotensin I generation rate) and plasma aldosterone concentration (PAldo) by radioimmunoassay have been developed utilizing 0·5 ml and 1·0 ml plasma, respectively.

It was found that in healthy children on free diets the PRA varied inversely with age. In infants the mean value was 1392 pgAl/ml per hr with a progressive decrease through childhood to the mean adult value of 87 pgAl/ml per hr. There was some evidence of a negative correlation between PRA and sodium turnover, estimated from the urinary sodium/creatinine ratio. The mean value for PAldo in children over the age of 1 year was 6·2 ng/100 ml, but in infants was 20·2 ng/100 ml.

PRA was in the range from 1000–200 pgAl/ml per hr in several hypertensive children, without PAldo necessarily being above the upper limit of normal. On the other hand, in children with salt-losing states PAldo was much greater, usually over 10,000 pgAl/ml per hr, and in the majority of these children PAldo was over 30 ng/100 ml.

S. MELLER. Queen Mary’s Hospital for Children, Carshalton. ‘Significance of bacteriuria in Cardiff schoolgirls’. Over a 2-year period all infant and junior schools in the City of Cardiff were visited by a mobile bacteriuria screening laboratory. 11,939 girls aged 5 to 11 (89% of the total population) were screened, at an estimated cost of 75 pence per child. Bacteriuria was confirmed in 207 girls, a prevalence of 1·7%. Full clinical and radiological data were obtained in 180 children who are participating in a randomized controlled trial of treatment.

Although some urinary symptoms were common, notably incomplete bladder control and offensive urine, few children had a history suggestive of serious infection past or present. As a group, their general health was good and they were of normal stature. However radiological evidence of pyelonephritis was found in 26% and vesicoureteric reflux in 34%. 74 children have completed the first year of follow-up. In the control group, 24% had a spontaneous bacteriological remission without treatment. In the treatment group, 19% had a recurrence within 6 months and 54% within 12 months of a successful short course of appropriate antibiotic: 11% continued to have bacteriuria despite repeated courses of treatment.

Although screening for bacteriuria in this age group identifies one child with pyelonephritis out of every 260 girls examined, it has yet to be shown that a treatment programme can influence the natural history of the condition.

Anna MurpHy. Royal Hospital for Sick Children, Glasgow. ‘Renal venous thrombosis in hypertonic dehydration’. Renal venous thrombosis is a well-recognized clinical entity of early infancy. The typical case is characterized by renal enlargement, haematuria, uraemia, and thrombocytopenia. In the Royal Hospital
Proceedings: Effect of dietary restriction and anorectic drugs on linear growth in childhood obesity.

P H Rayner and J M Court

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