Correspondence

Archives of Disease in Childhood, 1973, 48, 86.

Hydroxyproline excretion in idiopathic, congenital, and paralytic scoliosis

Sir,

We are glad that Dr. P. F. Benson wrote to you (Archives, 1972, 47, 476) drawing attention to the work on this subject which he published in 1965 (Benson, 1965). Had he been less modest, he might have added that it was the exciting discovery which he made—that urinary hydroxyproline excretion is increased in scoliotic children—which led to the much larger study, subsequently made here by us, which you published (Zorab et al., 1971) confirming his results. We ought to have referred to his earlier work in our paper. In the study of total hydroxyproline excretion in 168 French scoliotic children, we found the higher levels were mostly in those undergoing treatment.

STEPHANIE CLARK and P. A. ZORAB
Paediatric Research Unit,
Institute of Diseases of the Chest,
London S.W.3.

REFERENCES

Syndrome of growth resistance, obesity, and intellectual impairment with precocious puberty

Sir,

The paper under this title by MacMillan, Kim, and Weisskopf in the Archives, February 1972, p. 119 prompts me to report a similar patient.

H.G. is the second daughter of parents of normal stature (midparental height 172 cm) and was referred at the age of 4½ years with obesity and delay in speech development. Her birthweight had been 2·8 kg and her motor development had been slow; she had not walked unaided until the age of 2½ years. On examination her weight was over the 75th centile while her height was below the 10th. The obesity was most marked on the trunk; her facies were normal, but her hands and feet were noted to be tiny. Her IQ (Stanford Binet) was about 80. A diagnosis of Prader Willi syndrome was made and some dietary restriction advised.

Her subsequent progress is shown in Fig. 1 and 2. Her weight and height have increased together; at the age of 7½ years pubic hair and breast development were

FIG. 1.—Height chart showing progress of patient (●).

FIG. 2.—Weight chart showing progress of patient (●).
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<table>
<thead>
<tr>
<th>Investigations</th>
<th>Normal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chromosomes</td>
<td>Normal</td>
</tr>
<tr>
<td>Maximum plasma growth hormone</td>
<td>&gt; 50.0 IU/ml</td>
</tr>
<tr>
<td>during Bovril test</td>
<td>400 mIU/100 ml</td>
</tr>
<tr>
<td>Plasma FSH</td>
<td>440 mIU/100 ml</td>
</tr>
<tr>
<td>Plasma LH</td>
<td></td>
</tr>
<tr>
<td>Plasma 11 hydroxycorticosteroids</td>
<td></td>
</tr>
<tr>
<td>Midnight</td>
<td>1.7 μg/100 ml</td>
</tr>
<tr>
<td>9 a.m.</td>
<td>12.9 μg/100 ml</td>
</tr>
<tr>
<td>Plasma TSH</td>
<td>2.7 μU/ml</td>
</tr>
<tr>
<td>Serum PBI</td>
<td>6.0 μg/100 ml</td>
</tr>
<tr>
<td>24 hr urinary oestrone</td>
<td></td>
</tr>
<tr>
<td>oestradiol</td>
<td>1.5 μg</td>
</tr>
<tr>
<td>oestriol</td>
<td>1.3 μg</td>
</tr>
<tr>
<td>24 hr urinary oestrone</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3.6 μg</td>
</tr>
</tbody>
</table>

FSH, follicle-stimulating hormone; LH, luteinizing hormone; TSH, thyroid-stimulating hormone; PBI, protein-bound iodine.

noticed and regular menstruation began at 8½ years. Since the age of 7½ years her bone age has been significantly advanced and therefore her predicted adult height is below normal. At the age of 8½ years she was admitted for some investigations the results of which are given in the Table. In other ways she has remained well and she has continued to attend a normal school.

The clinical picture and laboratory findings in this girl support the suggestion that some patients with features of the Prader Willi syndrome develop precocious puberty as a result of disturbed hypothalamic function.

J. M. PARKIN
The Royal Victoria Infirmary, Children's Department, Victoria Road, Newcastle upon Tyne NE1 4LP.
Syndrome of growth resistance, obesity and intellectual impairment with precocious puberty.

J M Parkin

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