**LETTERS TO THE EDITOR**

Prevalence of asthma and wheeze in the Highlands of Scotland

**EDITOR,—**In their study on asthma and wheeze in the Highlands of Scotland Austin et al performed exercise tests on children in six geographical regions between April and June 1992.¹ The prevalence of exercise induced bronchospasm was particularly high on the Isle of Skye which was one of the most rural of the areas studied. However, data from the weekly returns service of the Royal College of General Practitioners showed that new episodes of asthma in 5-14 year olds were almost three times greater in June compared with April in 1992. Both asthma and hay fever show annual and seasonal variation² and are partly affected by the severity of the prevailing pollen season. Therefore, unless the exercise testing was carried out simultaneously in each region, comparisons are likely to be unreliable.

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**Dr Austin and Russell comment:**

Dr Ross and Fleming raise an interesting point in relation to the exercise testing which we used to support our questionnaire survey carried out in March and April 1992. We know of no epidemiological study which has correlated the prevalence of bronchial hyperactivity with new asthma episodes, but it is reasonable to suppose that seasonal variations in general practitioner returns will be more closely related to variations in the prevalence of various trigger factors, including pollens, than to variations in underlying bronchial hyperactivity. Nevertheless, allergens are known to affect bronchial hyperactivity, and the prevalence of exercise induced bronchoconstriction might thus be influenced by the date of study.

We have therefore reanalysed our data to look at temporal effects. If exercise tests performed in June are omitted, the overall prevalence of exercise induced bronchial hyperactivity rises to 9-6% compared with 8-9% when data for June tests are included.

Although weekly general practitioner returns for new asthma episodes in school aged children show a rise in late June which is small compared with the peak rise in September.¹ In Highland Region, figures from one general practice suggest that there is little difference for out of hours consultations for childhood asthma between April and June. Again although the numbers are low (four calls in April, one in May, and two in June) (figures courtesy of Ardurlie Medical Practice, Inverness).

We believe it is unlikely that pollen levels had any major influence on our results. In the north of Scotland the pollen count is because of cold climate (pollen tends not to hang in the air of the prevailing winds, and pollen production is lower and occurs later than elsewhere in UK) and the relatively sparse vegetation, especially in Skye. In any case, the tests in Skye were performed on 8th May 1992, too soon to be influenced by the seasonal effects proposed by Ross and Fleming.

¹ Asthma Information Agency. Seasonal variations in asthma. Factsheet 93/4. Available from Department of Medical Sciences, St George’s Hospital Medical School, London. (Data 1987/92 quoted from Royal College of General Practitioners weekly returns service.)

Cardiovascular malformations in Turner’s syndrome

**EDITOR,—**We were interested to read the findings of Goztsche et al who reported the prevalence of cardiovascular malformations and karyotype associated with Turner’s syndrome.¹ We have recently reviewed the karyotype and phenotypes of 63 Turner’s syndrome patients who have been referred to our department. Cardiovascular malformations have only been reported in seven of them (2/14 45 X, 2/45 45X/47XXX, 2/24 45X/46XY, and 1/3 45 X 46Xr(X) patients). Like Goztsche et al we did not see cardiovascular malformations in any patient without a 45 X cell line and the abnormalities accounted for most of the cases. If, as Goztsche suggests, around 26% of Turner patients have cardiovascular abnormalities then it is likely that some of our older patients who have not undergone routine echocardiography have as yet unrecognised cardiac problems. This is an important point, as minor aortic abnormalities may be associated with dissecting an aorta in Turner’s syndrome adults² and this complication could be screened for in susceptible individuals.

Goztsche et al concluded that ‘no patient with structural abnormalities of the X chromosome had cardiovascular malformations’. This is true for their 13 non-mosaic patients but in fact 4/46 of their patients with cardiovascular problems did have a cell line containing a ring or iso(X). It is likely that these four individuals began life with a non-mosaic karyotype and that a 45 X cell line arose later as the abnormal X chromosomes were lost during cell division. It seems, therefore, that it is the presence of a 45 X cell line at an early stage of development that might influence whether a cardiovascular abnormality is likely to be present or not.

Why should this be so? It has been suggested that a gene present on both the X and the Y chromosome and which escapes X inactivation may be responsible for the somatic features of Turner’s syndrome.³ An individual must possess two functioning copies of this gene in order not to manifest Turner’s syndrome characteristics. Structurally abnormal X chromosomes may still express this gene which is believed to lie on the long arm of the X at Xq13 close to the centromere and the X inactivation centre. Only a single copy of this gene can be expressed in cells which contain a 45 X cell line. The role of non-mosaic karyotype in the causation of cardiovascular malformations or a secondary role because its absence gives rise to lymphoedema.⁴ In our study lymphoedema was documented in 79% patients with only a 45 X cell line and in 33% patients with structural X chromosome abnormalities.

We are in agreement with Goztsche and other authors who have suggested an increased prevalence of cardiovascular malformations in Turner’s syndrome patients with a 45 X karyotype. However, although the incidence of cardiovascular abnormalities is low in patients with non-mosaic structural abnormalities, patients who are mosaics for ring (X) or iso (X) with a 45 X cell line or who have X chromosomes or markers lacking the Xq13 region may still be at increased risk of heart problems and should be screened for these.

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Growth hormone and hypophosphataemic rickets

**EDITOR,—**There has been some interest in the use of growth hormone in children with hypophosphataemic rickets who have short stature as a consequence of their metabolic bone disease. In addition to its growth promoting role there is the theoretical advantage that growth hormone by increasing the tubular reabsorption of phosphate may lead to a reduction in phosphate supplement requirements which have been linked to the development of nephrocalcinosis.

We would like to report our observations on the relationship of bone phosphate handling and markers of bone turnover in a 7 year old girl with hypophosphataemic rickets treated with growth hormone over an initial seven month period. She was originally referred for an endocrine assessment because of her short stature. Although she was exceedingly short for her age (height SD score 3.9), there was no evidence of deformity of her legs and no rickets. An insulin tolerance test with adequate hypoglycaemia demonstrated a peak growth hormone response of 7·5 μg/l (15 mU/l). In view of this and her short stature it was decided to give her a therapeutic trial of growth hormone in a dose of 0·6 IU/kg/week which increased to 0·75 IU/kg/week after three months given as six daily injections/week.

Over a seven month period she gained 4·4 cm in height, her height SD score improved from 3·9 to 3·7 and height velocity SD score from 1·8 to 0·1. On the same doses of phosphate and vitamin D her plasma phosphate rose from 0·99 mmol/l to 1·37 mmol/l in three months and then dropped to 1·02 mmol/l at seven months. The level of TmPO4/giomerular filtration rate increased...
from 0·20 mmol/l at baseline to 0·55 mmol/l at three months and then dropped to 0·4 mmol/l at seven months. The changes in markers of bone turnover are as shown in the table with a more than twofold rise in serum osteocalcin and the carboxyterminal propeptide of type 1 collagen (PICP) as markers of bone formation and a 1·5 fold rise in the level of the carboxyterminal cross linked telopeptide of type 1 collagen (ICTP) and urinary hydroxyproline/creatinine ratio (OHP/Cr) as markers of bone resorption. These reflect the general increase in bone turnover induced by growth hormone treatment.

**Growth hormone in hypophosphataemic rickets**

<table>
<thead>
<tr>
<th>Months of treatment</th>
<th>Osteocalcin (ng/ml)</th>
<th>PICP (ng/ml)</th>
<th>ICTP (ng/ml)</th>
<th>OHP/Cr (umol/mmol)</th>
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<tbody>
<tr>
<td>0</td>
<td>30</td>
<td>241</td>
<td>22·0</td>
<td>178</td>
</tr>
<tr>
<td>3</td>
<td>78</td>
<td>542</td>
<td>30·2</td>
<td>261</td>
</tr>
<tr>
<td>7</td>
<td>85</td>
<td>598</td>
<td>30·3</td>
<td>179</td>
</tr>
</tbody>
</table>

These initial results are encouraging and suggest a potential therapeutic role for growth hormone in this condition, although it remains to be seen whether the effect on the tubular reabsorption of phosphate can be maintained long term.

**Treatment of fragile X syndrome**

**EDITOR**—In Slaney et al's otherwise impressive and important paper on DNA testing for fragile X syndrome in schools for learning difficulties,1 the authors state that 'treatment is not possible at the present time'. Although a cure is not possible at the present time there is good evidence for benefits of medical interventions (for example, the use of stimulants or folic acid for the frequently associated attentional deficits and hyperactivity2,3), and specific educational approaches focusing on the profile of cognitive, social, and behavioural deficits frequently witnessed in young people with fragile X syndrome.4,5 In addition, behavioural psychotherapy techniques can be invaluable in tackling such maladaptive behaviours as hyperactivity, self-injury, and obsessionality. Families are entitled to know that while the condition cannot be cured there is much that can be done to maximise affected individuals' potential and to minimise their handicaps.

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**Perianal infection with β haemolytic streptococcus**

**EDITOR**—I am writing in reference to the article by Wright and Butt on perianal infection with β haemolytic streptococcus.1 In the discussion of the article the author notes, 'the disorder has been mistaken for child abuse leading to unnecessary social investigation and family distress'. It needs to be stressed that genital trauma and anal trauma can predispose to infection and that I have seen one case where a child sustained anal sexual trauma and subsequently developed perianal β haemolytic streptococcus infection. The β haemolytic streptococcus would find a traumatised anus an easier target than an untraumatised anus.

In my opinion, any child who presents with a perianal infection has to have sexual abuse considered as a possible diagnosis by the examining doctor.

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Growth hormone and hypophosphataemic rickets.

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http://adc.bmj.com/content/72/6/543.4.citation

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