Letters

Effects of dexamethasone in myotonic muscular dystrophy

Sir: Myotonic muscular dystrophy may shorten life especially when the function of cardiac, respiratory and deglutition muscles is severely impaired. It is difficult to evaluate the separate roles of myotonia and/or dystrophy in muscle dysfunction; however, it seems that weakness is due to both. Certain therapeutic trials have found some relief of myotonia with quinine propranolol, phenytion, ACTH and prednisolone but these and other drugs which were used, showed no beneficial effects on the progressive weakness. We report a case in which coincidental use of high dose dexamethasone resulted in a marked improvement on both weakness and myotonia in a patient with myotonic muscular dystrophy.

A 64 year old male was evaluated for progressive ptosis, dysarthria and dysphagia. Two years prior to admission, he noticed the gradual onset of limb weakness with difficulties in speech and swallowing, all of the signs becoming quite prominent in the last four months. Past medical history was significant for cataracts, known for 12 years. Several members of the family are known to suffer from myotonic muscular dystrophy.

Typical clinical features of myotonic dystrophy were present including marked grip and percussion myotonia of distal limb muscles and tongue; however, mild left hemiparesis including the homolateral lower face was also noted. Needle electromyography was characteristic for myotonic dystrophy with the presence of “dive-bomber” myotonia. Sodium phenytion (300 mg) daily failed to have any beneficial effect. On the 14th hospital day, the patient developed a sudden dense left hemiparesis due to a large space-occupying lesion in the right parietal area, shown on CT scan. Treatment with 24 mg daily of oral dexamethasone resulted in mild improvement in the hemiparesis, while a dramatic change occurred in the patient’s motor functions. Clinical and electrical myotonia practically disappeared and the patient could eat, talk and grip as easily “as when he was healthy”.

The beneficial effect of systemic corticosteroids (cortisone, prednisone) and ACTH on myotonic muscular dystrophy and “myotonia congenita” have been known since 1950. Although some beneficial effect on muscle weakness was seen in boys with Duchenne muscular dystrophy, it is generally agreed that in myotonic dystrophy steroids may improve myotonia only, possibly by its membrane stabilising properties. It is reasonable not to use steroids routinely in myotonic muscular dystrophy. However, steroids should be considered for short term use especially when swallowing and respiration are severely impaired.

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References

Heat stress and neuroleptic drugs

Sir: Several lines of evidence point to an involvement of hypothalamic and nigral dopaminergic systems in thermoregulation.1 Intolerance to heat has been described as one symptom of Parkinson’s disease,2 and blockade of hypothalamic and striatal dopamine receptors seems to be the pathophysiological mechanism of the “neuroleptic malignant syndrome” which is characterised by hyperpyrexia and extrapyramidal signs.3 In addition to the interference with central thermoregulation, neuroleptic drugs also suppress sweating by their peripheral anticholinergic effect. The potentially dangerous impairment of heat dissipation in neuroleptic-treated patients is illustrated by the report of fatal heat strokes in connection with unusually high summer temperatures.4 On the other hand, phenothiazenes, by causing vasodilatation and abolishing shivering, may at low ambient temperatures lead to hypothermia.5 Thus, neuroleptics markedly impair temperature regulation, producing a poikilohtermic state rather than the normal isothermic state.

Heat stress in the sauna bath is part of everyday life in Finland, where psychiatric in-patients also have the opportunity of at least one bath per week. Since sauna bathing has become increasingly popular also in other countries during the last decades, the number of patients on neuroleptic drugs enjoying this habit may be growing. Against this background we found it worthwhile to explore, whether our patients on neuroleptic drugs run a recognisable risk by exposing themselves to the high temperatures encountered in the Finnish sauna.

The study was done in a mental hospital in Eastern Finland which serves the needs for psychiatric care of a population of about 250,000. Sixteen in-patients (10 men and 6 women, mean age 30-5 ± 6-6 years) were chosen for the study and their informed consent was obtained. They were in good physical health and had been on one or two neuroleptic drugs for at least 5 months prior to the study. The drugs used were haloperidol, perphenazine and thoridazine (five patients each), chlorpromazine, chlorprothixene and fluphenazine (four, two and two patients respectively). As controls (N = 14) we used age-matched male and female nurses. Oral temperature, heart rate and blood pressure were taken four times: (1) immediately before entering the sauna (2) after an initial period of 10 minutes in the sauna (64.3 ± 6.6 °C, dry atmosphere), (3) after a second period of 12 minutes duration in the heat, and (4) after a 30 minute cooling off period. Measurements were taken outside the heated unit at an ambient temperature of 25–28°C.

Rise and normalisation of the body temperature was almost identical in patients and controls (table). In both groups heart rate increased and diastolic blood pressure decreased in response to the heat stress. One patient on heavy medication (chlorprothixene 400 mg/day) reported a sensation of fainting during the trial. His blood pressure fell to 78/60 mmHg, but rise in temp-

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<th>Measurements</th>
<th>Oral temperature (°C) mean ± SD</th>
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<td>1</td>
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<tr>
<td>Neuroleptic-treated patients</td>
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<td>± 0.6</td>
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<tr>
<td>Controls</td>
<td>36.9</td>
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