Matters arising

Successful treatment of refractory Sydenham's chorea with pimozide

Sir: We read with interest the paper *The treatment of severe dystonia in children and adults* by Marsden *et al.*¹ We describe the successful use of pimozide in a case of refractory Sydenham's chorea.

The patient was a 14-year-old right-handed boy admitted to hospital for review. He presented two years earlier with right-sided hemichoreiform movements and facial grimacing which had persisted. He had a normal birth with normal motor milestones, and had had all the normal childhood illnesses including measles. There was no family history of chorea. He attended school and was able to play games but he wrote with difficulty. A subsequent assessment by a clinical psychologist revealed average intellectual ability. Physical examination revealed coarse choreiform movements of his right arm and leg; his cardiovascular system and the rest of the examination was normal. A two-year history of Sydenham's chorea is an unusual feature and so investigations were made to exclude the diagnosis.

His CSF protein was 0.16 g/l with three lymphocytes. His CSF serum IgG was 0.0007 (within normal range). His serum measles CFT was 1 in 160; his CSF measles CFT was less than 1 in 10. He was ANF negative. His serum copper was 15.6 (normal range 11-25.1 mmol/l). His serum ceruloplasmin was normal at 0.41 units (normal range 0.2-0.55). His urinary copper levels were normal. His EEG showed no obvious focal or paroxysmal features and his CT scan was normal. His ASO titre was normal.

This patient had been tried on numerous drugs including tetrabenazine and most recently clonazepam, all without benefit. His only other drug was prophylactic penicillin V 250 mg tds. He was started on pimozide at a dose of 2 mg bd. Within 48 hours his chorea had all but disappeared. No involuntary movements were detected at clinic follow-up. There was no evidence of a tardive dystonia six months after treatment.

The important feature was the rapidity and apparent specificity of action in this case. Tetrabenazine has been described as of use in Sydenham's chorea but was not effective in our patient.^{2,3} Pimozide has not previously been described as a treatment option in Sydenham's chorea. It may be that its action as a selective post-synaptic

dopamine receptor antagonist is important in treating this condition.⁴

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Hypercalcaemia complicating polyneuropathy

Sir: We were interested to read the paper on *Immobilisation hypercalcaemia complicating polyneuropathy in adolescent boys*.¹ We have recently reported two cases of marked hypercalcaemia associated with immobilisation due to polyneuropathy in adults.²

The first patient was a 43-year-old woman with severe Guillain-Barré syndrome who required assisted ventilation for 23 weeks. Five months later whilst remobilising during a prolonged convalescence she developed symptomatic hypercalcaemia (serum calcium 3.92 mmol/l). The second patient was a 32-year-old woman who, 3 months following a cadaver renal transplant, developed cytomegalovirus infection with respiratory involvement followed by a motor neuropathy. Hypercalcaemia (serum calcium 3.54 mmol/l) developed during the immobilisation period, but while the patient was recovering.

The finding of hypercalcaemia of immobilisation in patients with polyneuropathies is a relatively unrecognised complication of these disorders. In our patients hypercalcaemia developed during recovery of the polyneuropathy, and hypercalcaemia may be missed, therefore, unless serum calcium is monitored throughout the recovery phase. This may

be particularly important if hypercalcaemia aggravates or prolongs muscle weakness.

Whereas hypercalcaemia is rare, hypercalciuria is commonly observed following immobilisation for whatever reason.³ The mechanisms include a combination of suppressed bone formation and increased bone resorption which, if prolonged, result in considerable bone loss and increased risk of fracture, renal stones and hypercalcaemia.^{4,5} It is not certain whether trabecular bone loss is ever fully recovered following re-mobilisation. For all these reasons an aggressive approach to treatment may be worthwhile, possibly based on the extent of hypercalciuria. Whereas corticosteroids may suppress hypercalcaemia in immobilisation, it is likely that they aggravate bone loss by depressing bone formation. Prolonged steroid treatment increases bone resorption and hypercalciuria. Thus, it seems more appropriate to treat immobilisation hypercalciuria and hypercalcaemia with specific inhibitors of bone resorption such as calcitonin, or, as in our cases, a diphosphonate.⁶

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