Letters to the Editor

Hereditary essential tremor and restless legs syndrome

Sir Essential tremor is usually considered to be a monosymptomatic disorder with variable clinical expression, about half of all cases demonstrating the familial dominant pattern of inheritance. Possible associations with other movement disorders have been described, including Parkinson's disease. We report a patient with hereditary essential tremor and restless legs syndrome.

A 71-year-old woman complained of a tremor in her right upper limb. This had affected her writing since her teens but had become worse in recent months, also affecting the left arm. The tremor was exacerbated by anxiety and fatigue; alcohol had no relieving effect. Tremulousness of the voice had made it increasingly difficult for her to sing in the church, and restless legs were also noted. Both her arms were comfortably at rest, but she could not relax because of an uncontrollable urge to move her legs when at rest. Although present throughout the day, this was particularly evident at night and often prevented her from falling asleep. This symptom had also been present since childhood when she had been labelled a 'fidget' at school because of her restlessness in class. A significant exacerbation of these symptoms occurred during pregnancy. Both her father and paternal grandfather had been troubled by a similar tremor and restless legs syndrome. Moreover, her father had always been 'fidgety' and unable to settle. Her only sibling, a non-identical twin sister, was unaffected, likewise her only son (aged 47).

On examination, the patient had a distal tremor of the upper limbs, accentuated by posture. Writing showed intrusion of tremor and drawing of a spiral was impaired. Head titubation ('no-no') was also evident, and hodling a single note revealed vocal tremor. Other than a distal tremor, examination was entirely normal; in particular, there were no dystonic features nor evidence of a peripheral neuropathy. Investigations (urea, creatinine, total calcium, creatinine, tests, full blood count, serum vitamin B12, and red cell folate) were normal. Treatment with pranopanol (40 mg bid) marginally improved her postural tremor, but had no effect on her restless legs.

Clinically this patient had unequivocal hereditary essential tremor. Furthermore, she fulfilled the suggested diagnostic criteria for restless legs syndrome. Previous accounts of an association between hereditary essential tremor and restless legs syndrome are limited. Stein et al reported a pedigree in which restless legs and hereditary tremor coincided in three patients over two generations, with a similar time of onset in each patient, and opined that the syndrome was associated. Bain et al found restless legs in three of 93 patients with hereditary essential tremor; restless legs and tremor were found to co-segregate. Jankovic has reported encountering several patients with the restless legs syndrome and an essential tremor-like tremor but gives no further details.5

The association between essential tremor and restless legs in our patient could be the chance concurrence of two relatively common movement disorders. Alternatively, it may be indicative of an underlying link. The differing pharmacological responsiveness of the two disorders argues against a shared pathophysiological mechanism. A linkage at the genetic level would therefore seem less likely. Like hereditary tremor, restless legs syndrome has been reported to present in childhood and adolescence as an hereditary condition with probable autosomal dominant transmission. It may be that in this family, genes for an essential tremor and restless legs have co-segregated. In such families, identification of the genetic locus for one condition may thus facilitate delineation of the other.

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4 Borstein B. Restless legs. Psychiat Neurol (Basel) 1961; 141: 165–201


Hoekworms to treat haemachromatosis?

Sir Biological control strategies can be used to tackle ecological problems and plant diseases. For example, hydrocarbon-metabolizing bacteria are used to degrade oil spills and gardeners use parasitic organisms like the wasp Encarsia formosa, which preys on greenhouse whitefly. However, living organisms are rarely used for treatment of human disease, although Wagner von Jan-reyg won a Nobel prize in 1927 for his work with malaria therapy for syphils, now superseded by penicillin, and parasites are still valued by plastic surgeons. We should like to propose hoekworms for the treatment of haemachromatosis.

Idiopathic haemachromatosis is an autosomal recessive inborn error of iron metabolism with accumulation of 20–40 g of iron resulting in multi-organ damage and death from cardiac failure or liver disease (normal body iron content is 3–4 g). Removal of excess iron reverses many of the biochemical and functional abnormalities. Treatment includes weekly venesection of 450 ml of whole blood (equal to 0.2 g of iron) until iron stores return to normal, which may take two to three years. Although some countries accept haemachromatosis sufferers as paid blood donors, asymptomatic patients in the UK may not perceive any tangible benefit from their weekly hospital visits and are excluded from blood donations. Vitamin therapy is inconvenient and noncompliance can be a problem.

Angiostrongylus cantonensis is a nematode parasite of the human gut. The adult worms are about 11 mm long, and attach to the mucosa of the upper small intestine with a set of cutting mouthparts. They live on blood and plasma protein sucked from the lamina propria, each worm consuming 0.2 ml of blood (about 0.09 mg of iron) per day. Worldwide, 700–900 million people harbour hoekworms, making this infection second only to menstruation as the major global cause of iron deficiency.

The therapeutic potential of hoekworms for haemachromatosis is obvious. To attain a daily blood loss equivalent to 450 ml per week, 300–400 worms would be required, depending on the amount of iron resorbed. A hoekworm lives for only one week, and autoinfection does not occur so blood loss is predictable. A three- or six-monthly blood count, albumin and ferritin would be used to monitor therapy. Commonly prescribed drugs would not kill the worms while they reside but once iron stores had returned to normal a two-day course of mebendazole should eradiclate them. Hoekworms are well adapted and, apart from anaemia, morbidity from natural worm infection is minimal. The practicalities of experimental human infection have been studied previously. Side-effects include pruritus at the site of cutaneous penetration, mild laryngeal and pulmonary symptoms during pulmonary passage and occasionally, upper gastrointestinal discomfort. In the UK there is no danger of transmission to others (except in the special case of miners working in damp tunnels) because the soil dwelling stage requires a high ambient temperature. The main problem is likely to be patient consent. There is a natural aversion to the idea of intestinal parasites and, although we harbour them they could cause substantial perception of abdominal symptoms. For long-term maintenance therapy only a little blood loss is required, so biannual venesection would be used. In conclusion, we feel that despite current enthusiasm for alternative and 'natural' medicine, the possibilities of hoekworm therapy for haemachromatosis have been overlooked.

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1 Finlayson NDC. Hereditary (primary) haemachromatosis. BMJ 1990; 301: 350–1

Sir Obtaining an accurate drug history from a patient admitted to hospital requires reliable information during a three-week period. Information gathered by the general practitioner’s referral letter, the patient and their medicine bottles, relatives, or carers, and records held by general practitioners or community pharmacists. The accuracy of the drug history obtained during a three-week period in hospital has been shown to be low for the age group of patients examined by us.1 In our study, 40% of patients had at least one unintentional difference between the drugs prescribed for them in hospital and those prescribed by their general practitioner. Such inaccuracies can compromise patient care by hindering the identification of drug-related problems and further therapeutic management. Methods to improve drug history taking should involve improving information flow between primary and secondary care, and improving the skills of doctors or pharmacists taking the drug history. The clinical significance of the unintentional differences deserves further study.

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Incarceration of the penis by a metallic ring

Sir,

Incarceration or strangulation of the penis by an encircling object is an uncommon clinical event. This paper describes a case of long-standing penile incarceration by a metallic ring and its management.

A 35-year-old man presented in a distressed and embarrassed state with a heavy metal ring (outer diameter 3.2 cm, inner diameter 1.4 cm), encircling the penis at its root. The ring had been in place for a month, during which the patient had tried several manoeuvres to remove it, without success, although he was habituated to using a ring in this way. The patient had no urinary complaints. Movement of the ring was not possible and the penis distal to the ring was swollen, ulcerated and harbouring maggots (figure). Attempts to remove the ring using lubricants and by multiple punctures and aspiration of blood from the engorged penis failed. Division of the ring was not easy. Under spinal anaesthesia, the distal penis was degloved to the level of cavernous tissue and the iron ring was removed. Subsequent split-thickness skin grafting from the medial side of the right thigh on the denuded penis (5 x 3 cm) yielded good results.

A variety of constrictive bands (‘cock-rings’), rubber bands, etc have been used to increase sexual gratification and prolong erections.1 Patients usually present at a late stage when the penis is grossly swollen and attempts to remove the objects have already failed. When these constrictive bands are left in place for too long, the penile skin and shaft becomes oedematous, blood flow is compromised, and rupture of the urethra with extravasation may ensue. Browning et al described the use of string to compress the distal penis, making it elongated and narrow. However, other authors have reported ‘de-gloving’ the distal penis to the level of cavernous tissue before the foreign object could be removed.2,3 Sinha4 suggested that multiple punctures and aspiration of blood from the engorged penis is a simple and safe method and potentially dangerous and mutating methods of removal of the constricting object should only be used if this treatment fails.

In the present case, penile degloving and subsequent skin grafting did not affect erectile power and the cosmetic appearance was excellent. Even after long-standing penile incarceration, the patient did not have urethral stricture.

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Hookworms to treat haemachromatosis?

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