References

1. Hoenig J, Hamilton CM. Epilepsy and sexual orgasm. Acta Psychiatr Neurol Scand 1960;35:448–456
2. Penfield W, Rasmussen T. The cerebral cortex of man. New York: Macmillan, 1955:26–27
3. Bancaud J, Favel P, Bonis A, et al. Manifestations sexuelles paroxystiques et épilepsie temporale. Etude clinique, EEG at SEEG d'une épilepsie d'origine tumorale. Rev Neurol (Paris) 1970;123:217–230
4. Ruff RL. Orgasmic epilepsy (letter). Neurology 1980;30:1252
5. Bachman DS, Rossel CW. Orgasmic epilepsy (letter). Neurology 1984;34:559–560

Severe Myotonia Relieved by Nifedipine

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We report a case of incapacitating myotonia which remitted after institution of therapy with the calcium channel blocker nifedipine. A 24-year-old housewife complained of severe stiffness in her hands of approximately 10 months' duration. On gripping an object tightly she was often unable to release her grasp for some minutes unless she manually straightened her fingers using her free hand. She did not complain of weakness, and there was no family history of myotonia or cataracts.

Examination revealed gross, widespread voluntary and percussion myotonia. Finger extension, following fist clenching, took over 2 minutes. Apart from mild neck flexion weakness she was strong, but reflexes were generally depressed. Diffuse posterior capsular polychromatic lens opacities were seen on slit lamp examination. Urea and electrolytes, creatine kinase, and thyroid function measurements were normal. Electromyography demonstrated markedly prolonged myotonic after-discharges on needle insertion and percussion. Voluntary motor unit potentials were difficult to distinguish separately because of continuous myotonia; however long-duration slurred triphasic potentials with a reduced interference pattern were noted. Needle muscle biopsy of the left vastus lateralis showed normal muscle architecture and muscle fiber distribution.

Nifedipine was begun, 10 mg 3 times daily. However, the patient had increased the dosage, in view of its successful effect, to 90 mg per day in divided doses. One month later there was barely any clinical evidence of myotonia. Finger extension time recorded by dynamometry took less than 2 seconds. An asymptomatic postural drop in blood pressure was demonstrated (105/70 supine; 90/60 erect). The dosage of nifedipine was reduced to that previously recommended, and a month later finger extension times of 5 to 6 seconds were recorded.

Although it is not known how nifedipine works to relieve myotonia, it may prevent calcium transport at the surface membrane or prevent signal transmission at the tubulosarcoplasmic reticulum junction. Phenytoin also has calcium channel blocking properties which may account for this membrane stabilizing action [1]. Nifedipine has been found to be of benefit in treating exercise-induced muscle pain syndromes and muscle cramps related to hemodialysis [2, 3], and we suggest that nifedipine be considered initially in patients with troublesome myotonia. In recommended dosage it does not have any effect on the cardiac conduction system in man [4].

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References

1. Study RE. Phenytoin inhibition of cyclic guanosine 3′:5′-monophosphate (cGMP) accumulation in neuroblastoma cells by calcium channel blockade. J Pharmacol Exp Ther 1980;251:575–581
2. Sufit RL, Peters HA. Nifedipine relieves exercise related myalgias. Muscle Nerve 1984;7:647–649
3. Peer G, Blum M, Aviram A. Relief of haemodialysis induced muscle cramps by Nifedipine. Dialysis Transplant 1983;12:180–181
4. Kenny J. Calcium channel blocking agents and the heart. Br Med J 1985;291:1150–1151

Oculomasticatory Myorhythmia

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Schwartz and colleagues [1] have drawn attention to oculomasticatory myorhythmia (OMM) as a pathognomonic sign of central nervous system (CNS) Whipple's disease. Approximately one month before the publication of their paper we diagnosed an identical case.

Our patient is a 31-year-old man from Louisiana who was in excellent general health except for a 10-year history of arthralgias. He first noted blurred and double vision in January 1986 and then had spells of limited awareness, decreased responsiveness, and automatisms lasting up to an hour. In February 1986 he lost all vertical gaze and by May 1986 he developed rhythmic contractions of his tongue and pharynx. During this interval an electroencephalogram (EEG), cerebrospinal fluid (CSF) analysis, visual evoked responses, and computed tomography scan of the brain were unremarkable. He was referred to us in November 1986.

Mental status fluctuated between normal and orientation only to name. He had frequent periods of somnolence but no seizure activity. His speech was dysarthric, fundi and fields were normal, and there was no Kayser-Fleischer ring. There was no volitional gaze but reflex eye movements were intact in all directions. There were 2-cycle-per-second convergent movements of the eyes in synchrony with palatal and mandibular movements, and intorsion and extorsion movements of the right foot not in exact synchrony with the facial movements. The remainder of the neurological examination was normal.

Routine laboratory studies including liver functions, colla-