Objective: To report the successful treatment of primary orthostatic tremor in a 76-year-old man.

Background: Primary orthostatic tremor is a rare condition, with few reports describing therapies. Established therapies had previously failed in our patient.

Methods: Using an evidence-based evaluation of treatments via MEDLINE’s GRATEFUL MED search engine, a therapeutic option was determined during the first consultation with the patient, and pramipexole therapy was initiated.

Results: The therapy proved effective, and the patient had relief from his symptoms for the first time in 6 years.

Conclusion: Pramipexole is a potential therapy for primary orthostatic tremor.

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The consensus statement of the Movement Disorder Society on tremor defines primary orthostatic tremor (POT) as a unique tremor syndrome that can be identified by the following characteristics:

• A subjective feeling of unsteadiness during stance but only in severe cases during gait; patients rarely fall. None of the patients have problems when sitting or lying down.

• Sparse clinical findings that are mostly limited to a visible and, occasionally, palpable fine-amplitude rippling of the leg (quadriceps or gastrocnemius) muscles when the patient is standing.

• The diagnosis can be confirmed only by electromyographic recordings (eg, from the quadriceps muscle) with a typical 13- to 18-Hz pattern. All leg, trunk, and arm muscles can show this tremor, which is typically absent during tonic activation while the patient is sitting or lying down.1

The world medical literature contains few reports that describe cases of POT, and usually very few patients per article.2–8 Reported effective therapies include benzodiazepines, notably clonazepam,9,6,8 gabapentin,9,10 and levodopa.11 The reports disagree as to the effectiveness of primidone, barbiturates, or β-blockers.7,8

The following case report illustrates the effectiveness of a dopamine D3 receptor agonist in the treatment of 1 patient, and why this was a logical choice.

Report of a Case

A 76-year-old man developed leg aches in 1993. He related that his legs would quiver when he would stand. He found that he had a wider based gait, but that he could still support himself when walking. He could not stand in lines or attend any social events that required him to stand in 1 place and talk to people. After the patient was evaluated at an academic center in 1993, the treating neurologists concluded that his history and electrophysiologic test results were consistent with POT. The electrodiagnostic tests showed no abnormal activities when the patient was at rest. He had immediate short-duration 18- to 19-Hz bursts in his leg muscles and extremely low-amplitude bursts in his paraspinal muscles. When he leaned forward, bursts of activity occurred in his arms, and short-duration bursts were present with tonic contraction. The consulting physicians believed that these findings were typical of POT, and prescribed clonazepam and propranolol. The patient stopped taking the medications because he experienced intolerable adverse affects, without benefits.
The patient came to our clinic in April 1999 for a re-evaluation of the continuing problems with his legs. He had no head or upper-extremity tremors at any time, and he had no tremors in his lower extremities while he was sitting. He could drive an automobile without difficulty.

Physical examination confirmed that the patient’s legs were quiet when he was sitting or recumbent. He had no increase in tone in any extremity. He had no motor weakness, coordination difficulties on cerebellar testing, or reflex changes. When he stood up to walk, he quickly became unsteady owing to a pronounced rapid tremoring in his lower extremities that caused imbalance and discomfort. The tremors were visible and involved proximal and distal muscles. The clinical appearance and frequency of the movements were varied, because the patient could voluntarily inhibit some of the tremors. When he started to walk, he had a slightly widened gait. He could not walk on his heels or toes or perform the tandem walk. He could not stand still for the Romberg test. His upper extremities were normal.

Gabapentin has been reported to be of benefit in treating this syndrome.5,10 Clonazepam is noted to be an effective medication for another lower-extremity movement disorder: restless leg syndrome.12-14 A decision was made to treat our patient with the dopamine D3 receptor agonist pramipexole. This choice was based on 3 reports in the world literature that discuss the use of pramipexole for restless legs syndrome.15-17

The patient began therapy with 0.125 mg of pramipexole 3 times a day, and the dosage was increased by the same amount weekly up to 0.375 mg 3 times a day. He noted a remarkable improvement in his POT, and found that he responded best at a dosage of 0.25 mg 3 times a day.

Physical examination of the patient during the pramipexole therapy revealed that he could stand without the previously debilitating tremor. When he walked, the wide-based gait was improved, and he was able to perform the tandem walk and the Romberg test. He noted no adverse effects from the medicine. The therapeutic effect has been sustained for 2 months at the same dosage (0.25 mg 3 times a day).

The world medical literature since 1966 contains few reports that address POT. The literature notes that the symptoms are due to high-frequency (13- to 18-Hz) burst firing in the weight-bearing muscles.1,2,5,7,8 The cause of POT is unknown. Several investigators have examined the relationship between POT and essential tremor, and all conclude that both conditions are clinically and electrophysiologically different.1,2,5,7,8 The differences in pharmacological responses are also noted.5,7 To our knowledge, there are no published studies that have investigated whether there is a relationship between POT and restless legs syndrome.

The leg aches that our patient experienced are unusual for POT. However, 1 additional subjective symptom in 1 patient is difficult to assess in a rare disease, so any conclusions about it, and whether or not it represents a new disorder, can only remain in the realm of speculation.

Our patient’s response to pramipexole therapy indicates that, besides the established therapies for POT, there is an additional, and potentially less sedating, treatment for this rare condition. The observation that dopamine D3 receptor agonists can be effective for 2 distinct movement disorders involving the lower extremities suggests an area of future investigation into the neurologic basis of both conditions. However, the clinical results in 1 case cannot provide therapeutic conclusions. Ideally, an investigation would have to include enough patients to permit a double-blinded study involving pramipexole and placebo, along with electrophysiologic monitoring. Such an investigation may not be possible or practical if more cases do not become clinically recognized.

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Corresponding author: Michael F. Finkel, MD, Department of Neurology, Cleveland Clinic Naples, 6101 Pine Ridge Rd, Naples, FL 34119 (e-mail: finkelm@ccf.org).

REFERENCES


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