In general, dyskinesia refers to any neurological disease manifested by difficulty with volitional movements of the head, limbs, or body. There are many forms of dyskinesias, both primary and secondary, inherited and sporadic.

The group of paroxysmal dyskinesias is a heterogeneous and rare class of hyperkinetic movement disorders. This grouping has been further subclassified by various systems into several distinct pathological entities. All paroxysmal dyskinesias tend to be characterized by attacks of involuntary movements, which may take the form of chorea, athetosis, dystonia, or ballismus.

The mainstay of treatment for most paroxysmal dyskinesias has been medical management with carbamazepine and other AEDs. However, one of the subtypes, PNKD has been noted to be refractory to medical management; until recently, patients suffering from PNKD were left with minimal effective treatment options for this debilitating condition. Although DBS has been reported to be safe and effective in treating some of the more common types of primary dystonias, there are only a few reports of its use in PNKD.

We present the unique case of a 26-year-old man who had signs and symptoms consistent with PNKD, as well as significant psychiatric comorbidities. He was severely debilitated by his disease, lived in a group home, and suffered from frequent falls, necessitating the wearing of a protective helmet and face mask at all times.

The patient underwent implantation of bilateral deep brain stimulation quadripolar electrodes in the globus pallidus internus with the aid of image-guided stereotactic neurosurgery and microelectrode recording without complication.

At his 1-month postoperative follow-up, the patient reported a subjective 90% improvement in his symptoms; the only notable side effect was a slight increased slurring in his baseline dysarthria. Objective reporting and recording forms maintained by the patient’s caretakers over the following 18 months suggested a significant and sustained improvement in his overall balance, ambulation, and gross motor function with a substantial decrease in the incidence of reported falls.

The authors conclude that pallidal deep brain stimulation may be successfully applied to patients suffering from refractory paroxysmal nonkinesigenic dyskinesia with promising results. This treatment strategy deserves further prospective investigation, clinical consideration, and refinement. (DOI: 10.3171/2009.9.JNS09454)

**KEY WORDS** • paroxysmal nonkinesigenic dyskinesia • movement disorder • deep brain stimulation
movements of his extremities and face that were nearly constant in nature, but not particularly bothersome or disabling. Distinct from this were bilateral episodes of moderate to severe flexion and jerking movements of the legs, arms, neck, and face. The dystonic movements were often preceded by a tingling sensation over the right or left hemibody, followed by curling of the toes, and then generalized dystonic movements. The episodes would last from a few seconds to a minute and were varied in frequency, but they could occur as frequently as 20 times per day. He never lost consciousness during these events. These sudden episodes of dystonic movements were very distressing to the patient and caused him much anxiety. On frequent occasions, he had fallen and injured himself, suffering from numerous scalp lacerations, abrasions, concussions, and closed head injuries, which necessitated his wearing a hockey helmet with a face mask at all times. The patient felt that there was a general trend of increasing severity, frequency, and duration of his dystonic fits over the past several years. The episodes were known to be precipitated or exacerbated by caffeine, coffee, food dyes, heat, anxiety, and stress. His baseline chorea, however, did not fluctuate with these stressors.

The patient’s medical history was notable for anxiety disorder, depression, impulse control disorder, and suicidal ideation. Additionally, he suffered from frequent throbbing frontal headaches associated with nausea, which frequently followed the dystonic episodes. He also had some difficulty sleeping as these movements occasionally occurred during sleep. According to the patient, and based on prior medical records obtained from other institutions, the patient had no family history of seizures, headaches, mental retardation, learning disability, attention or hyperactivity, neurodegenerative disease, tics, or other neurological problems.

On examination, he was noted to have significant dysarthria. Choreiform and athetoid movements were noted in all limbs, as well as periodic dystonic spasms of the face, neck, and extremities. A mild bilateral esotropia was also noted. His muscle tone, bulk, and strength were noted to be grossly normal in both upper and lower extremities. His deep tendon reflexes were graded 2+ in his upper extremities and 3+ in his lower extremities. His plantar reflexes were down-going bilaterally. His gait was unsteady; he was noted to hold his legs stiffly when walking. His dystonic movements were noted to be more severe on the right side, which became more pronounced with walking or action.

Formal neuropsychological evaluation was limited due to the patient’s mental fatigue and frustration with several of the tests. On the Mini-Mental State Examination, he scored 16 of 23 and showed significant memory impairment (1 of 15) for recall of words. His cognitive findings were consistent with his diagnosis of mild mental retardation. He was found to have intact basic auditory attention and comprehension and was not believed to be suffering from significant depression at the time.

Previous workups throughout his life included numerous imaging studies, which failed to demonstrate any structural lesions accounting for his symptoms. No seizure activity was ever noted on multiple electroencephalography studies, even during his involuntary episodic movements. Laboratory tests for a number of inherited disorders, such as Niemann-Pick disease, were also performed, with no identifiable metabolic basis for his symptoms. The patient’s history and physical examination were most consistent with a diagnosis of PNKD.

For many years, the patient had been placed on numerous AED regimens and other alternative medications commonly used to treat movement disorders in an attempt to conservatively treat his PNKD, including carbamazepine, diphenylhydantoin, lamotrigine, Depakene, levetiracetam, clonazepam, mephobarbital, diphenhydramine, acetazolamide, haloperidol, chlorpromazine, clonazepate, sertraline, lorazepam, gabapentin, quinidine, trihexyphenidyl, and risperidone. None demonstrated any significant benefit, aside from some short-lived improvement with clonazepam. He remained severely debilitated by his condition due to the frequent falls and the significant anxiety they generated. The patient was both unwilling and unable to participate in many activities due to his condition.

Due to the refractory nature of his disease, we elected to implant bilateral pallidal DBS electrodes.

Operative Course. The patient underwent a 2-stage procedure. During the initial phase, he underwent placement of a stereotactic head frame and subsequent CT scanning. The images obtained were then imported into BrainLAB IPlan stereotactic server, version 2.5 (BrainLAB AG), where they were merged with the patient’s prior MR image. This fused image set was then used to plan the operative targets, entry points, and trajectories. The patient was then placed on the operating room table and fixed in place via the head frame. The procedure was performed under conscious sedation. Microelectrode recording, as well as clinical testing, was used intraoperatively to optimize the electrode placement (3387, Medtronic, Inc.) for maximal benefit. During the second stage, the electrodes were tunneled subcutaneously and attached to a Kineta dual channel internal pulse generator (IPG) (Medtronic, Inc.), which was then implanted in the patient’s subclavicular region.

Postoperative Course. At the initial postoperative visit, the patient reported a 90% subjective improvement overall in both the severity and frequency of his dystonic fits. The only principal side effect that the patient mentioned was some worsening of his baseline dysarthria. At the 2-year follow-up, the patient reported excellent results from the surgery in control of his dystonia. Although not absent, the dystonic fits were much less severe, occurring 0–3 times a day, although periods of anxiety could still increase their frequency. This allowed him to be significantly more functional in the course of his days. The frequency of his falls greatly diminished.

As of his last clinic visit, the patient’s current stimulator settings are as follows. The left stimulator is set at...
Deep brain stimulation for PNKD

3.7 V, with a pulse width of 90 μsec and rate of 150 Hz, with lead 1 negative and 3 positive. The right electrode is set at 3.7 V, with pulse width of 120 microseconds and rate of 150 Hz with lead 5 negative and 7 positive.

Postoperative MR images were morphed into Talairach space and analyzed using BrainLAB IPlan stereotactic software, version 2.5 (Fig. 1). Electrode locations relative to the midcommissural point can be found in Table 1. The anterior commissure–posterior commissure distance was 26.9 mm.

**Discussion**

Paroxysmal dyskinesias, although rare, were first recognized over 50 years ago. Several classification schemes have been suggested over the years, the most recent subdivides them into the following 3 broad categories: PKD, PNKD, and paroxysmal exercise-induced dyskinesia.

Paroxysmal nonkinesigenic dyskinesia, also known as paroxysmal choreiform dystonia, is distinguished from PKD by less frequent, but longer-lasting attacks, which are not induced by sudden movement, unlike PKD. The frequency of attacks may vary, although they are generally less often than in PKD. Patients may often lose the ability to communicate during attacks, but they do not lose consciousness. Attacks are often precipitated by anxiety, fatigue, caffeine, or alcohol, and typically begin in one limb, but often spread to all limbs and the face. Patient symptoms often may manifest as either predominantly choreoathetosis or dystonia.

Like PKD, PNKD exists in both primary and secondary forms, the latter most commonly due to demyelinating diseases such as multiple sclerosis. Other noted associations with PNKD are cerebral palsy, stroke, trauma, and metabolic disorders, most specifically, hypothyroidism. Recent genetic studies have shown that primary PNKD may have multiple heterogeneous causes, as

**TABLE 1:** Electrode locations relative to the midcommissural point in Talairach space from post hoc analysis using BrainLAB IPlan stereotactic software

<table>
<thead>
<tr>
<th>Electrode</th>
<th>Lateral</th>
<th>AP</th>
<th>Vertical</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>17 lt</td>
<td>3 pst</td>
<td>7 inf</td>
</tr>
<tr>
<td>1</td>
<td>17 lt</td>
<td>1 pst</td>
<td>4 inf</td>
</tr>
<tr>
<td>2</td>
<td>17 lt</td>
<td>1 ant</td>
<td>2 inf</td>
</tr>
<tr>
<td>3</td>
<td>17 lt</td>
<td>2 ant</td>
<td>1 sup</td>
</tr>
<tr>
<td>4</td>
<td>16 rt</td>
<td>2 ant</td>
<td>4 inf</td>
</tr>
<tr>
<td>5</td>
<td>16 rt</td>
<td>4 ant</td>
<td>1 inf</td>
</tr>
<tr>
<td>6</td>
<td>17 rt</td>
<td>5 ant</td>
<td>1 sup</td>
</tr>
<tr>
<td>7</td>
<td>17 rt</td>
<td>7 ant</td>
<td>4 sup</td>
</tr>
</tbody>
</table>

* ant = anterior; AP = anteroposterior; inf = inferior; pst = posterior; sup = superior.
associations between cases of PNKD exist with mutations of the *MR-1* gene on chromosome 2, and with a mutation on chromosome 10q22, involving a calcium-sensitive potassium (BK) channel.

Although there has been a case report in the literature of a patient with secondary PNKD from idiopathic parahypothyroidism who responded well to levetiracetam, most patients with PNKD, like ours, are refractory to AEDs and other medications, such as baclofen, used in treatment of dystonias. Some have meager responses to benzodiazepines such as diazepam and clonazepam. Until recently, patients with PNKD had few, if any, effective treatment options.

Deep brain stimulation has been successfully implemented with varying success in medically refractory dystonias, but with greater successes in patients with primary, generalized, hereditary dystonias. Only 2 case reports of the use of DBS in PNKD have been reported. Loher and colleagues reported successful thalamic nucleus ventralis intermedius DBS in a patient who developed PNKD after brachial plexitis at the age of 29 years, and Yamada and colleagues reported a case of posttraumatic PNKD in a 59-year-old patient who was successfully treated with bilateral globus pallidus internus DBS. In both cases, a single limb was affected. As far as we can tell, this is the first case of successful DBS for childhood-onset PNKD affecting all 4 limbs, head, and trunk.

Deep brain stimulation is a safe and promising treatment option for medically refractory PNKD.

**Disclaimer**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**References**


Portions of this work were presented in poster form at the 3rd Annual Neurology/Neurosurgery Resident & Fellow Research Symposium at the University of Rochester in 2007 and as an abstract at The Movement Disorder Society’s 12th International Congress of Parkinson’s Disease and Movement Disorders in Chicago, Illinois, in 2008.

Please include this information when citing this paper: published online October 2, 2009; DOI: 10.3171/2009.9.JNS09454.

Address correspondence to: Jason M. Schwalb, M.D., Director of Movement Disorder and Behavioral Neurosurgery, Henry Ford Health System, 2799 West Grand Boulevard, K-11, Detroit, Michigan 48202, email: JSCHWAL1@hfhs.org.