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Clinical Observations

Successful Treatment of Paroxysmal Movement Disorders of Infancy With Dimenhydrinate and Diphenhydramine

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ABSTRACT

BACKGROUND: Paroxysmal movement disorders including paroxysmal tonic upward gaze of infancy and paroxysmal dystonia of infancy are benign but uncommon movement disorders seen in young children. Although symptoms are intermittent and resolve spontaneously, they can cause discomfort and distress for the child. Current treatment options are limited to dopaminergic agents or anticonvulsants with limited efficacy. **PATIENT DESCRIPTION:** The authors present a child with paroxysmal tonic upward gaze of infancy and another with paroxysmal dystonia of infancy, both of whom responded successfully to treatment with low-dose dimenhydrinate or diphenhydramine, respectively. **DISCUSSION:** Dimenhydrinate and diphenhydramine both exert anticholinergic activity and have limited toxicity at low doses. This property makes either compound an attractive therapeutic option for paroxysmal movement disorders in infancy. These agents are generally well tolerated.

Keywords: benign paroxysmal tonic upward gaze of infancy, transient paroxysmal dystonia of infancy, dimenhydrinate, diphenhydramine

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Introduction

Paroxysmal movement disorders including paroxysmal tonic upward gaze of infancy and paroxysmal dystonia of infancy are benign but uncommon movement disorders seen in young children. Diagnosis is often by exclusion as other disorders such as epilepsy or structural brain abnormalities need to be excluded. Symptoms typically resolve spontaneously; however, attacks when they occur can cause discomfort and distress for children and their families. Treatment options are limited to dopaminergic agents or anticonvulsants with limited efficacy.[1,2]

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Patient 1

A 12-month-old boy presented with a 2-month history of episodic sustained forceful conjugate upward gaze that was increasing in frequency. Each episode persisted for 30-45 minutes and varied in severity. During mild episodes, the upward gaze was not complete and the child was still able to see. He compensated for the mild deficit by leaning his head forward in an “eyes up, chin down” position. In severe episodes, the child’s irises and pupils were covered and the child was unable to see causing him emotional distress. The episodes were worse during fatigue and relieved by sleep. At the time of presentation the episodes were occurring 2 to 3 times per day. Associated symptoms included mild pallor during some of the episodes and occasional nystagmoid eye movements. The child had no nausea or vomiting, ataxia, decreased level of consciousness, weakness, or abnormal movements during or in between these episodes.

His past medical history including prenatal and neonatal histories were noncontributory. Developmental history was normal with the exception of mild expressive language delay. There was a family history of migraine headaches, but no other neurological problems including movement disorders.

The boy was seen in clinic in between episodes. During examination he was alert and acting appropriately. He was in the 92nd percentile for

height, 74th percentile for weight, and 62nd percentile for head circumference. Cranial nerve examination including extraocular movements, pupillary response to light, visual fields on direct confrontation, and funduscopy were normal. No nystagmus or abnormal eye movements were seen. Truncal tone was normal. In the extremities, tone, strength, tendon reflexes, and cerebellar function were normal and symmetrical. Both plantars were down-going and gait was normal. The parents provided a video of a mild episode (Figure and Video).

The patient was hospitalized for further assessment. Blood tests and cranial magnetic resonance imaging were normal. He underwent a lumbar puncture with normal cerebrospinal fluid (CSF) cell count, glucose, protein, and amino acid analysis. CSF neurotransmitter analysis showed homovanillic acid and 5-hydroxyindoacetic acid below the reference ranges (Table 1). An electroencephalogram (EEG) was normal and exhibited no epileptiform discharges, including during an episode of upward gaze. A diagnosis of paroxysmal tonic upward gaze of infancy was made.

While in hospital he was administered a low dose of dimenhydrinate (12.5 mg once daily), resulting in immediate resolution of symptoms.

The patient demonstrated a marked and immediate improvement with the administration of dimenhydrinate. After treatment began he experienced only very mild episodes of residual upward deviation of one eye when fatigued or when a dose of dimenhydrinate was missed. No side effects were reported with the administration of dimenhydrinate. The same dose of dimenhydrinate was continued for 10 months as the patient continued to be episode free. The parents stopped the dimenhydrinate with no recurrence of symptoms.

In a follow-up visit, 17 months after initial presentation, his neurological examination was normal. Normal development was also reported with minor concerns regarding difficulties with expressive speech. The child was living in a dual language home that was felt to explain his expressive speech delay given his normal receptive language development.

Patient 2

A 14-month-old boy presented with episodic torticollis associated with truncal posturing followed by recurrent vomiting. The events started at age 10 months and gradually increased in frequency from once a week to 2 to 3 times per week lasting between a few hours to 3 days.

His parents noticed facial pallor before the events, and on a few occasions he also had a preceding conjugate upward gaze that was followed by torticollis, forceful posturing of his right side of his body, and vomiting. There was no change in the level of consciousness or involuntary limb movements during the episodes. He was able to communicate and attempted sitting and playing during that time. The child gained weight appropriately despite the episodic vomiting.

FIGURE.
Episode of paroxysmal tonic upward gaze in Patient 1. Supplementary video related to this figure can be found at http://dx.doi.org/10.1016/j.pediatrneurol.2015.10.019. (The color version of this figure is available in the online edition.)

TABLE 1.
Cerebrospinal Fluid Neurotransmitter Metabolite Values and Reference Ranges for Patient 1

Neurotransmitter Metabolite	Value (nmol/L)	Reference Range (nmol/L)
Homovanillic acid (dopa metabolite)	237	294-1115
5-Hydroxyindoacetic acid (serotonin metabolite)	127	129-520
3-0 Methyldopa (L-dopa metabolite)	40	<300

His past medical history, including prenatal and neonatal, was unremarkable and his development was normal. His family history was remarkable for migraine on both sides of the family with no history of other neurological problems including movement disorders.

The boy was seen in clinic between episodes. His head circumference was at the 50th percentile. Cranial nerve examination was normal, including extraocular movements, pupillary response to light and funduscopy. Truncal tone was normal. In the extremities, tone, strength, tendon reflexes, and cerebellar function were normal and symmetrical. Both plantar responses were down-going and gait was normal. The child demonstrated smooth motor transitions with no evidence of ataxia, or abnormal movement. The parents showed us a video demonstrating torticollis and a dystonic body posturing.

Subsequent investigations including blood tests and magnetic resonance imaging of the brain and cervical spinal cord were normal. An EEG was completed after an episode and was normal. An EEG during an event was not recorded. The patient also underwent a complete evaluation for gastroesophageal reflux, which was unremarkable.

Based on the severity of truncal and limb dystonia during attacks, a diagnosis of transient paroxysmal dystonia of infancy was made. The child was given diphenhydramine hydrochloride 12.5 mg three times per day, leading to a complete resolution of symptoms within 5 weeks. The child was administered diphenhydramine for a total of 3 months.

Discussion

Paroxysmal tonic upward gaze of infancy is an uncommon benign syndrome of childhood and infancy characterized by sustained episodes of conjugate upward deviation of the eyes, which is accompanied by neck flexion to compensate for abnormal eye positioning. The pathophysiology of paroxysmal tonic upward gaze remains obscure, although it could be due to a functional disturbance of the upper dorsal brainstem secondary to immaturity or depletion of neurotransmitter pathways or channelopathy.[1] Like dopa-responsive dystonia, paroxysmal tonic upward gaze has diurnal fluctuation of symptoms and is often relieved by sleep. Roubertie et al.[3] found mutations of the *CACNA1A* gene in a child with paroxysmal tonic upward gaze of infancy whose family members also had episodic ataxia and benign paroxysmal torticollis.

Although care needs to be taken to exclude structural brainstem abnormalities and other disorders that can cause the child to have upward gaze deviation such as epilepsy, oculogyric crisis, tics, and voluntary eye movements, patients with paroxysmal tonic upward gaze have negative investigations including neuroimaging and EEG.[2] Paroxysmal tonic upward gaze has varying degrees of improvement; however, most often spontaneously resolve over 1 to 4 years. Although the neurodevelopmental outcome is normal in half of the children with paroxysmal tonic upward gaze, it can be associated with ataxia and cognitive

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