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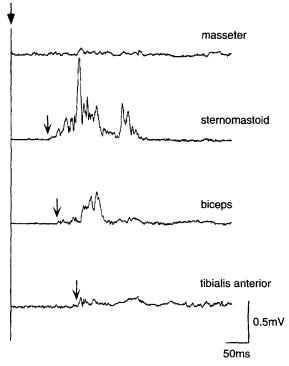
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# Symptomatic Hyperekplexia Occurring as a Result of Pontine Infarction



Hyperekplexia, or "startle disease," is characterized by a pathologic enhancement of the auditory startle reaction (1,2). Unexpected auditory stimuli or taps to the upper trunk and face elicit a brief, generalized jerk of the upper body. This is produced by a stereotyped sequence of muscle activation commencing in the sternomastoid and spreading to muscles innervated by cranial nerve nuclei in the brainstem and caudally to muscles innervated by motoneurons of the spinal cord. The pattern of muscle activation is identical to that seen in the normal auditory startle response. In both normal and abnormal auditory startle responses, the intervals between the onset of electromyographic (EMG) activity in upper-limb and lower-limb muscles are longer than would be expected for conduction down the corticospinal tract (2). This feature suggests that the efferent pathways of the auditory startle

#### stimulate ipsilateral supraorbital nerve



**FIG. 1.** Surface electromyographic (EMG) recordings following electrical stimulation of the ipsilateral supraorbital nerve. Stimulus intensity is twice the threshold intensity for patient perception of the stimulus. The *arrows* denote the onset of EMG activity in the sternomastoid (74 ms), biceps (91 ms), and tibialis anterior (132 ms). The *tracings* represent the average of five successive recordings.

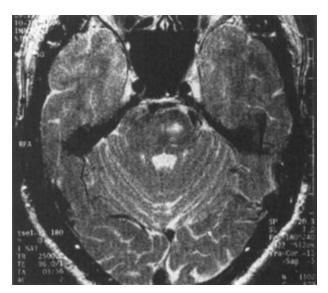


FIG. 2. Axial T2-weighted magnetic resonance image showing two areas of high signal typical of infarction within the left pons. The larger lesion is located in the ventral pons whereas the smaller, more dorsal slit-shaped lesion in the pontine tegmentum probably lies within the reticular nuclei (centralis caudalis and reticularis tegmenti or papillioformis).

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response may utilize a spinobulbospinal reflex system in which slowly conducting pathways descend into the spinal cord (3).

#### Case Report

A 70-year-old man developed the sudden onset of vertigo while at rest in bed. This was associated with an unsteady gait but he experienced no other neurologic symptoms at that time. The vertigo gradually improved over the next 9 months, but continued to occur after sudden head movement. At 9 months after the onset of vertigo, he began to experience "uncomfortable sensations" in response to sudden unexpected stimuli, particularly noise. He described these as "electric-shock-like jolts" in the arms, chest, and occasionally the legs, resulting in a brief jerking movement of the upper half of his body. They had become a source of increasing embarrassment to him. He had also recently noted that several episodes occurred in response to thoughts and perhaps dreams when he was lying quietly in bed. There was no family history of neurologic disease.

On examination, any unexpected sound and taps to the nose, chin, or forehead elicited a brief bilateral jerking movement of the head (neck flexion), shoulders (abduction), and arms (elbow flexion) with variable flexion of the trunk. This reaction did not appear to habituate after repeated stimuli. The findings on neurologic examination were otherwise normal-in particular there was a full range of ocular movement without nystagmus, facial sensation was normal, and there was no facial asymmetry. Tendon reflexes, plantar responses, gait, and coordination were normal. Reflex EMG activity was recorded in left masseter, left sternomastoid, left biceps, and left tibialis anterior in response to electrical stimulation of the left supraorbital nerve. Stimuli were delivered at twice threshold for perception of the supraorbital nerve stimulus and elicited a generalized myoclonic jerk of the upper body identical to that seen following a loud noise (see the videotape). The earliest reflex EMG activity occurred in sternomastoid, followed by masseter, biceps, and tibialis anterior (Fig. 1). The interval between the onset of reflex EMG activity in biceps and tibialis anterior was 41 ms.

A magnetic resonance image of the brain (Fig. 2) revealed a lesion consistent with infarction in the left ventral pons. A smaller lesion consistent with infarction was situated more dorsally in the pontine tegmentum, in the region of the left pontine reticular nuclei. The patient was treated with clonaze-pam, 1 mg daily, which mildly improved the severity of his symptoms.

## Discussion

In the present case, the brief generalized myoclonic jerk of the upper body in response to unexpected noise and other stimuli to the mantle area, the onset of the generalized myoclonus in sternomastoid, and the interval between muscle activity in biceps and tibialis anterior in each myoclonic jerk are typical of hyperekplexia. Hereditary, sporadic, and symptomatic forms of hyperekplexia are recognized (2). Symptomatic hyperekplexia has been described in the setting of infarction of the brainstem (4–6) and thalamic–subthalamic area (7) and in inflammatory lesions of the pons (2). In the present case, hyperekplexia followed a vascular event involving the pons with lesions placed adjacent to and involving the tegmental pontine reticular nuclei (centralis caudalis and reticularis teg-

menti or papillioformis). It is presumed that these lesions interrupted afferent or efferent connections of the pontine tegmental reticular nuclei mediating the startle response. Absence of the startle response in the Steele–Richardson–Olszewski syndrome has been attributed to the degeneration of nucleus pontis centralis caudalis of the reticular formation in that condition (8).

In the present case, the lesions evident in the vicinity of this nuclear complex have presumably altered the normal control of the startle response. The delay of 9 months or so between the pontine stroke and the appearance of hyperekplexia is reminiscent of the delay in appearance after stroke of posthemiplegic dystonia (9) and palatal myoclonus (10). In these conditions, neural sprouting and changes in synaptic sensitivity have been proposed during the interval between brain insult and the appearance of the movement disorder. Similar mechanisms may have occurred in relation to the pontine reticular nuclei in the present case, thus accounting for the delay between the stroke and subsequent exaggeration of the startle response.

Acknowledgment: T.K. is supported by the National Health and Medical Research Council of Australia.

## Legend to the Videotape

A 70-year-old man who exhibits an exaggerated auditory startle reflex (hyperekplexia). Symptoms commenced 9 months after a pontine infarction, demonstrated on magnetic resonance imaging (see Fig. 2).

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## Periodic Bursts of Rhythmic Dyskinesia Associated with Spinal Anesthesia



Neurologic complications associated with spinal or epidural anesthesia are very rare. Cytotoxic properties or excessive concentration of the local anesthetic may cause immediate neurologic complications. Another possible cause of immediate neurologic complications is direct trauma to the nerve root during lumbar puncture. Arachnoiditis is the most common cause of delayed neurologic complications after spinal anesthesia (1).

Several cases of movement disorders associated with spinal or epidural anesthesia have been reported (Table 1) (2–7): all of the patients developed sudden onset of transient dyskinesias during the operation or in the recovery room. They seemed to have a periodic single burst or bursts of jerks, jerky spasms, or prolonged spasms of the lower-limb muscles, frequently causing stereotyped triple flexion of the hip, knee, and ankle.

A 57-year-old woman developed continuous pain in her left knee joint after a traffic accident. One year after the accident, magnetic resonance imaging of the left knee showed a tear in the posterior horn of the medial meniscus, so arthroscopic examination and meniscectomy of the left knee were carried out, with intramuscular midazolam and atropine sulfate premedication. A vial containing 20 mg of tetracaine powder was reconstituted with 2 ml of 10% dextrose in water and 2 ml of normal saline. She was placed in a left lateral decubitus position, and a lumbar puncture with paramedian approach was performed at the L4-5 intervertebral space. Through a 25-gauge inside needle, a mixture of 1.8 ml of prepared tetracaine solution (corresponding to 9 mg of tetracaine) and 0.2 ml of 1:1,000

TABLE 1. Clinical features of the patients who developed movement disorders associated with spinal or epidural anesthesia

References	Type of anesthesia	Drugs used	Character of involuntary movements described by the authors	Intervals <sup>a</sup>	Clinical courses
Fox et al., 1979 (2)	Spinal	Tetracaine	Strong, irregular, very brief flexion and extension of the thighs and knees	~5 h	Spontaneous remission, responded to intravenous diazepam
Nadkarni et al., 1980 (3)	Spinal	Lidocaine	Bilateral plantar flexion followed by a burst of clonic contraction of the muscles of the abdomen, low back, thighs, and legs; duration, 5–7 s (interval, 2–3 min)	~2 h	Spontaneous remission, responded to intravenous diazepam
Watanabe et al., 1987 (4)	Spinal	Dibucaine	Consecutive cogwheel stereotyped slow movements (same as sleep-related periodic leg movements); each movement lasted 3-4 s (interval, 20-23 s)	3–4 min	Spontaneous remission after 70 min
Parkinson et al., 1990 (case 1) (5)	Spinal	Hydromorphine	Rhythmic jerking of the legs	A few minutes	Spontaneous remission after 1–2 h
Watanabe et al., 1990 (6)	Spinal	Lidocaine	Synchronous periodic jerky spasms causing extension of the great toe and partial flexion of the ankle, knee, and hip; each movement lasted 3–5 s (interval, 20–30 s)	20-30 min	Spontaneous remission after 2 h
Hachisuka et al., 1991 (7)	Epidural	Mepivacaine	Intermittent abdominal muscle spasms: spinal myoclonus	5 months	Responded to clonazepam

<sup>&</sup>lt;sup>a</sup> Time interval between the spinal anesthesia and the onset of movement disorders.

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