

abnormalities. Neither common anticonvulsants nor imipramine had any appreciable effect on the akinetic seizure or the generalized seizures that occurred later. The youngest sister of this patient began having frequent gelastic akinetic seizures at 5 years of age. A sleep EEG a few months later was essentially normal, with no evidence of sleep-onset REM. In our last patient with Niemann-Pick disease group C, a Mexican boy, gelastic akinetic seizures began at 3 years of age. The EEG was generally slow with occasional focal spike and slow-wave discharges. An all-night sleep recording revealed mild disturbances caused by partial obstruction but no evidence of sleep-onset REM. Now, at 5 years of age, the patient has fewer gelastic seizures, with brief unresponsiveness, and frequent generalized seizures that have been partly controlled with phenobarbital.

Kandt and colleagues [2] make a good case for cataplexy. Laughing is the only identified precipitating factor in all the cases associated with Niemann-Pick disease. As pointed out, cataplexy cannot always be differentiated easily from simultaneously occurring epilepsy. In our cases minimal giggling was the trigger — not strong emotion, as reported by these authors. In our patients, imipramine, the most commonly used medication for treatment of cataplexy, had no effect. We had no opportunity to try protriptyline.

Gelastic seizures or cataplexy has never been associated with Niemann-Pick disease group A or B, conditions characterized by a profound generalized sphingomyelinase deficiency. Therefore, such manifestations are a unique clue to the existence of Niemann-Pick disease group C [3] or one of its closely related milder forms with later onset, called group D and E. All affected patients have a number of common biochemical and ultrastructural abnormalities in the absence of an identified enzyme defect [4].

Departments of *Psychiatry, †Neurology, ‡Pediatrics,
and §Anatomy
University of California, Los Angeles
Los Angeles, CA 90024

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Reply

Raymond S. Kandt, MD

I agree with Drs Philippart, Engel, and Zimmerman that differentiation between akinetic seizures and cataplexy may be difficult. The telling feature in cases of cataplexy is the *provocation* of the attacks by laughter and the emotion of humor or, less commonly, other emotions. This pattern is in contrast to that in some akinetic seizures, which may be associated with,

rather than brought on by, laughter. In “gelastic epilepsy” the laughter may occur prior to the other seizure manifestations, during the seizures, or following the seizures [2]. The laughter is not characteristic of a specific seizure type or abnormality and may be associated with massive spasms and generalized electroencephalographic abnormalities or with epileptiform foci or abnormalities in the temporal or frontal lobe. Characteristically, epileptic laughter does not have a precipitant and is often emotionless, incongruous, or disagreeable [2]. Our first patient recognized that his cataplectic attacks were precipitated by the emotional concomitants of laughter and maintained full awareness during the telemetered episode. During that episode he had neither epileptiform discharges nor flexion or extension spasms. The later polysomnographic findings were characteristic of narcolepsy, and his cataplectic episodes were abolished by protriptyline. Therefore, we felt that cataplexy was confirmed and epilepsy excluded.

In an individual with cataplexy, absence of sleep-onset rapid eye movement periods on a routine electroencephalogram is not adequate evidence to exclude the diagnosis of narcolepsy. The polygraphic features of narcolepsy have been reviewed, and it is clear that the diagnosis can be missed easily if strict recording conditions are not met [3]. Furthermore, as noted in our report [1], cataplexy may occur long before other manifestations of the narcolepsy-cataplexy tetrad, or may even occur in isolation.

The cases discussed by Philippart and colleagues highlight the diagnostic difficulties that may be encountered when trying to distinguish cataplexy from akinetic seizures, especially those seizures that are preceded by laughter.

Departments of Neurology and Pediatrics
University of Michigan Medical Center
Ann Arbor, MI 48109

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Sensory Loss from Whole Sural Nerve Biopsy

J. Clarke Stevens, MD, and Peter James Dyck, MD

In the article entitled “Comparison between fascicular and whole sural nerve biopsy,” Pollock and coworkers [1] report no significant difference in symptoms and sensory deficit between fascicular and whole nerve biopsy; they conclude that “whole nerve biopsy should be recommended in preference to fascicular biopsy since it is simpler, has greater diagnostic potential, and allows for a more complete morphological evaluation.”