

TITLE: Treatment of Landau-Kleffner Syndrome with Pulse Dose Prednisone: Behavioral and Language Outcomes

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Rationale: Landau-Kleffner Syndrome (LKS) poses a challenge for treatment because language and behavioral impairment can persist even when seizures are fully controlled with antiepileptic drugs (AEDs). Although daily dose prednisone has been found to improve cognitive and behavioral functioning, significant side effects preclude more than a brief treatment regimen. This study was aimed at evaluating the effect of one or two day per week treatment with prednisone on aphasia and behavior in a small sample of LKS patients.

Methods: One female and three male patients, ages 7 to 12 years at baseline assessment, underwent neuropsychological testing prior to and during treatment with one or two day per week prednisone (2 mg/kg/day, divided into two doses over 24 hours). All four patients had EEG abnormalities characterized by frequent focal, multifocal, or generalized spike and wave activity while awake that increased in sleep. During sleep these discharges occurred in prolonged runs or continuous discharges (CSWS). All patients were of normal estimated nonverbal intelligence and had either a history of language regression or frank paraphasias on baseline testing. On average, patients were retested three months following baseline assessment. Patients' scores on measures of expressive/receptive language and verbal intelligence during baseline and treatment conditions were compared. Behavioral observations were also documented at each testing interval.

Results: All patients evidenced improvement from the moderate/severe to mildly impaired range in at least one language modality (confrontation naming, receptive language, or verbal intelligence). Even when a language function remained severely impaired, absolute level of ability was measurably improved. One patient remained highly volatile and distractible. Three patients displayed resolution of extreme behavioral difficulties that included aggression, tearfulness, or rigidity (i.e. patient would comply only when allowed to perform a task as he wanted). Distractibility was notably improved, but not resolved, in these three patients.

Conclusion: One to two day per week prednisone yields marked improvement in language and behavioral functioning while minimizing side effects of daily steroid treatment. Although some language and behavioral impairment appears to persist during treatment, the improvements are sufficient to allow the LKS patient to attend school and engage in other routine activities.