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Language and behavioral outcomes of treatment with pulse-dose prednisone for electrical status epilepticus in sleep (ESES)

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ABSTRACT

Few studies have examined treatment response in electrical status epilepticus in sleep (ESES), and fewer still have evaluated the effect of corticosteroid treatment employing a pulse-dose regimen. The aim of this study was to examine the effectiveness of pulse-dose prednisone in treating language and behavioral disturbances that often accompany ESES. The sample included 17 patients age 5 to 10 years at time of baseline electroencephalogram (EEG) and neuropsychological assessments. For all patients, focal, multifocal, or generalized spike and wave activity occupied greater than 50% of the nonrapid eye movement (REM) sleep record. Patients were seen for follow-up EEG recording and neuropsychological testing with an average of 10 months following initiation of pulse-dose prednisone. Improvement in language or behavior was examined in relation to resolution of ESES on EEG, age at seizure onset and treatment, duration of ESES, duration of treatment, lesional versus nonlesional epilepsy, history of language or behavioral regression, seizure control at follow-up, and intelligence quotient (IQ). With the exception of a greater likelihood of patients with low IQ to demonstrate improvement in language or behavior, improvement was seen in most patients, irrespective of ESES or other factors.

1. Introduction

"Electrical status epilepticus" was a term proposed by Patry, Lyagoubi, and Tassinari for the phenomenon of continuous spike and wave discharges provoked by sleep [1]. The term "epilepsy with continuous spike–waves during sleep" (CSWS) was supported by the Commission on Classification and Terminology of the International League Against Epilepsy in 1989 and implied an accompanying neuropsychological disorder [2]. Although electrical status epilepticus in sleep (ESES) and CSWS are terms that are often used interchangeably, some have preferred to use ESES to refer to the electrographic pattern and CSWS to refer to the syndrome of neuropsychological regression and electroencephalogram (EEG) pattern [3].

It is difficult to overstate the challenges to daily functioning posed by behavioral and language regressions related to ESES. The behavioral disturbances often exceed those seen in attention-deficit/hyperactivity disorder (ADHD) alone, and can be manifested by aggression, violent temper tantrums, compulsive behaviors, stereotypies, and highly oppositional behavior [4]. These types of behaviors can make it difficult or impossible for a child to have a productive day at school and pose significant stress for parents at home. ESES-related aphasia can range from a near absence of speech and failure to comprehend even the most elemental aspects of speech and environmental sounds [4], to much more subtle manifestations, such as increased false starts and hesitations in speech [5]. Regardless of severity, language disturbance related to ESES has the potential to constrain a child's academic learning and impede a child's ability to communicate with others during the active phase of the condition.

Corticosteroids, in addition to benzodiazepines and valproate, are among the common treatments for ESES [6–8]. In a large meta-analysis, steroids and surgery were found to be the most effective treatment approaches for ESES [9]. Improvement in language and behavior in response to daily corticosteroid treatment has been documented, usually in association with improved EEG [10,11]. Daily corticosteroid treatment can result in a Cushing-like syndrome with longer than short-term use [10–12], and tapering to a regimen of 1 mg/kg on alternate days was found to maintain improvements in EEG and cognitive-behavioral functioning in one small sample [10]. Use of pulse dosing appears to reduce the risk of side effects, and has been found to result in improved language and symptoms of autism spectrum disorder (ASD) [5,13], making it an alternative to daily prednisone that can potentially be used for a longer duration.

The aim of this study was to further investigate the efficacy of pulse-dose prednisone for ESES-related language and behavioral abnormalities. Patient data were analyzed to examine the concordance between improvement on EEG and neuropsychological functioning, and differential

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Table 1
Patient characteristics.

Patient number	Etiology/MRI	Seizure types	ESES at diagnosis	Age ESES detected (years)	Age at treatment onset (years)	Age at follow-up (years)	Comorbidities at baseline	Baseline IQ
1	Nonlesional	Focal	Diffuse, multifocal	8.6	8.6	8.9	ASD	VIQ 65 PIQ 84
2	IVH	Focal	Multifocal	10.3	10.3	10.8	ADHD	VIQ 73 PIQ 69
3	TSC	Myoclonic	Generalized	8.0	8.0	8.4	ADHD/ID	VIQ 58 PIQ 46
4	Neuroepithelial cysts	Focal	Unilateral	7.0	7.0	7.1	ASD	VIQ 50
5	Cortical dysplasia	Focal	Temporal–parietal and generalized	5.3	5.3	7.6		VIQ 93 PIQ 100
6	PVL	Atypical absence	Generalized	9.7	9.7	10.9	ADHD/ID	VIQ 57 PIQ 47
7	Nonlesional	Focal	Central–temporal bilateral	8.1	8.1	9.1	ID	VIQ 77 PIQ 65
8	PVL	Focal	Unilateral	9.1	10.9	11.4	ASD/ID	VIQ 67 PIQ 73
9	Nonlesional	Focal myoclonic tonic	Unilateral and generalized	7.2	7.2	8.3	ASD/ID	
10	BECTS	Focal	Bilateral frontal–central–temporal	5.5	8.3	8.9	ADHD	VIQ 95 PIQ 110
11	PVL	Focal	Unilateral	7.2	7.3	7.8	ADHD	VIQ 88
12	Perinatal ischemia	Focal	Generalized	5.8	7.2	8.0	ADHD/ID	VIQ 57 PIQ 63
13	Congenital hydrocephalus	Focal	Generalized	3.1	10.0	10.5	ADHD/ID	VIQ 74
14	Intrauterine stroke	Focal	Unilateral	5.0	5.5	6.6	ID	VIQ 66 PIQ 51
15	Nonlesional	Focal	Bicentral	6.2	8.3	9.3	ADHD	VIQ 103 PIQ 97
16	Polymicrogyria	Focal	Unilateral central–temporal	7.0	7.0	7.3	ASD/ID	VIQ 65 PIQ 74
17	Neonatal embolic stroke	Focal	Unilateral central–temporal	8.9	8.9	10.0	ASD/ADHD/NLD	VIQ 89 PIQ 66

Legend: ASD – autism spectrum disorder; ADHD – attention-deficit hyperactivity disorder; ID – intellectual disability; IVH – intraventricular hemorrhage; NLD – nonverbal learning disability; BECTS – benign epilepsy with centrotemporal spikes; PVL – periventricular leukomalacia; TSC – tuberous sclerosis complex.

treatment response based on presence or absence of a history of regression, lesional versus nonlesional epilepsy, baseline intelligence quotient (IQ), age at treatment, duration of ESES at the time of treatment initiation, duration of treatment, and seizure control at follow-up.

2. Methods

2.1. Participants

The study protocol was approved by the Institutional Review Board of Children's Hospitals and Clinics of Minnesota. In a retrospective review of the records of 32 patients treated with prednisone for ESES, those patients who underwent both pretreatment and follow-up EEG and neuropsychological testing were identified. Patients who did not undergo EEG and neuropsychological testing in close temporal proximity at both pretreatment baseline and follow-up (n = 13) were excluded. Two patients were unable to continue the pulse-dose regimen because of behavioral side effects: one patient experienced aggressive and rambunctious behavior, and a second patient experienced increased emotionality. The patient sample (Table 1) included 10 males and seven females age 5 to 10 years ($\bar{x} = 8$ years) at the time of baseline neuropsychological assessment that was conducted within two months of baseline EEG recording. Fifteen patients experienced focal onset seizures, one patient experienced primary generalized epilepsy (myoclonic seizures), and one patient experienced both focal and generalized (myoclonic tonic) seizures. At baseline, only 7 of 17 patients were seizure-free. Patients evidenced EEG abnormalities characterized by frequent focal, multifocal, or generalized spike and wave activity while awake that increased in sleep and occupied greater than 50% of nonrapid eye movement (REM) sleep during overnight or long-term video-EEG recording (Figs. 1 and 2) [14].

Mean verbal IQ (range: 50–103; $\bar{x} = 75$) and performance IQ (range: 46–110; $\bar{x} = 73$) at baseline fell in the mildly impaired range for the sample as a whole. The majority of patients evidenced structural abnormality on magnetic resonance imaging (MRI). A large proportion (47%) had a congenital or perinatal etiology, such as vascular complications of preterm birth, ischemic insult, stroke, or hydrocephalus. Seizure onset ranged from 0 to 7.4 years ($\bar{x} = 2.8$ years), and age at diagnosis of ESES ranged from 3.1 to 10.3 years ($\bar{x} = 7.1$ years). Many patients met criteria for one or more diagnoses of a developmental condition, including ADHD (47%), ASD (35%), and intellectual disability (47%) at the time of baseline neuropsychological assessment.

2.2. Procedure

The initial neuropsychological assessment included a parent interview to clarify the nature of any language or behavior changes the child had experienced, an informal speech sample that was elicited by having the child describe absurdities in pictures [15], and measures of intellectual development, confrontation naming, and comprehension of oral directions that were administered as part of a comprehensive neuropsychological test battery at baseline assessment [16–21]. The psychological diagnosis (ASD, ADHD, intellectual disability) was also verified as part of the standard comprehensive neuropsychological evaluation.

Patients were treated with oral prednisone once per week at a goal dosage of 2 mg/kg/day divided into two doses over 24 h. In six patients, treatment was initiated as a pulse-dosing regimen, and in 11 patients, treatment was initiated with a 5–7 day burst (2 mg/kg/day) followed by taper and then transition to pulse dosing. All patients were treated with other antiepileptic drugs (AEDs) simultaneous with prednisone treatment. Two patients (13 and 15) were also treated with high-dose diazepam (20 mg and 15 mg, respectively) at the time of follow-up, and seven were treated with low-dose clonazepam (0.25–1.5 mg) concomitant with prednisone treatment. No patient had undergone epilepsy surgery prior to medication treatment for ESES. No significant Cushingoid phenomena were noted in any patient.[1] There were no cases of increased infection rate or significant weight gain.

Patients were seen for follow-up neuropsychological assessment and EEG recording one to 27 months ($\bar{x} = 10$ months) following initiation of pulse-dose prednisone and prior to discontinuation of their regimen. The EEG was classified as improved if there was resolution of ESES at follow-up. The follow-up neuropsychological assessment included a parent interview, language sample to gauge qualitative changes in expressive language, and readministration of selected language tests. Some patients' limited capacity for cooperation precluded administration of the full test battery during either baseline or follow-up assessment.

On the basis of baseline testing, patients were classified as having experienced a distinct language regression if they evidenced a dysphasia (e.g., phonemic paraphasias, marked dysfluency) on formal testing or were reported by parents to be displaying a distinct change from premorbid language functioning. Patients were classified as having experienced a behavioral regression if parents reported a distinct

[1] No patients demonstrated excessive abdominal or facial fat deposition (increased adiposity), increased blood pressure, impaired growth, or hirsutism.

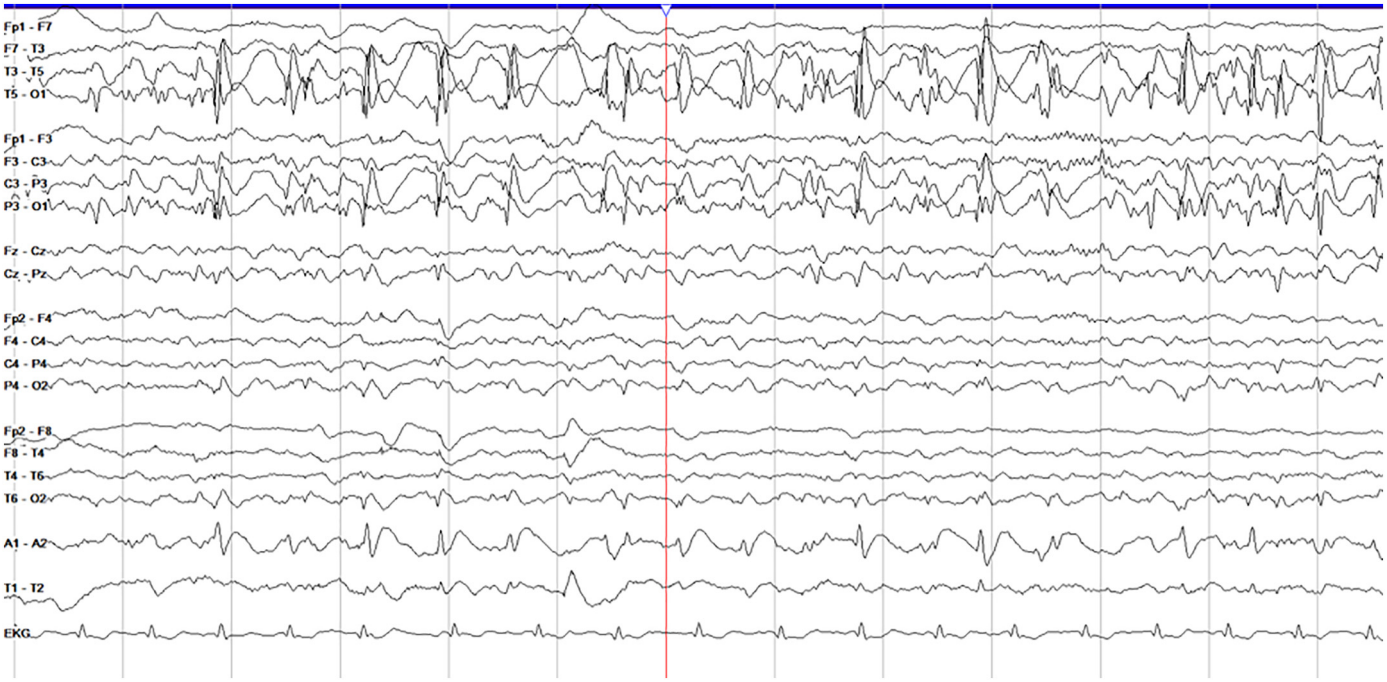

Fig. 1. Focal discharge which is present >50% of the sleep recording.

change from premorbid functioning, such as increased aggression, decreased social interest, or decreased judgment and impulse control.

Patients were classified as having improved language functioning if they displayed significant qualitative improvement in dysphasia on a speech sample or improved language test performance that exceeded expected maturational gains (i.e., improvement in standard score of greater than one standard deviation). Patients were classified as having experienced improvement in behavior if reported history or observations during testing suggested a distinct change from baseline functioning (e.g., markedly improved behavioral compliance, improvement from frequent "meltdowns" to mostly positive demeanor).

Patients who experienced improvement in language or behavioral functioning were compared with those who did not on variables that included resolution of ESES on EEG, history of regression, lesional versus nonlesional etiology, duration of ESES at the time of treatment initiation, duration of treatment, baseline IQ, age at time of treatment, age at time of diagnosis of ESES, and seizure control at the time of follow-up assessment. Differences between groups on these variables were examined with Fisher's exact test, chi-square, or paired samples *t*-test.

3. Results

For the sample as a whole, 10 patients (59%) experienced improvement in either language or behavior. Eight patients experienced improved language, and six of 12 patients with disordered behavior at baseline experienced improvement in behavior.

3.1. Factors affecting language outcome

Forty-seven percent of the sample experienced distinct improvement in language, primarily based on review of qualitative language samples and reported history. Cognitive test scores were relatively insensitive to medication effects (Table 2). Only two patients experienced improvement in standard scores that exceeded expected maturational gains. In one patient, verbal IQ improved from 58 to 89, and performance IQ improved from 46 to 73. In the other patient, verbal IQ improved from 65 to 86, and comprehension of oral directions improved from the mildly impaired to average range.

As indicated in Fig. 3, improvement in language was only slightly, but not significantly, more likely in those evidencing resolution of ESES on EEG. Percentage of patients experiencing language improvement was not significantly related to presence of a structural abnormality, acquired versus developmental language abnormalities, or degree of seizure control at follow-up. As indicated in Table 3, those who experienced improved language did not differ from those whose language was unimproved based on age at time of testing, age at seizure onset, or duration of treatment. Those with lower verbal and performance IQs were more likely to experience meaningful improvement in language, and there was a trend toward those with shorter duration of ESES at time of treatment to experience improved language.

A description of qualitative changes in language is presented in Table 4. Although features of dysphasia did not resolve completely, patients tended to experience improvement in length of utterance, intelligibility of speech, and frequency of paraphasic errors.

3.2. Factors affecting behavioral outcome

There was improvement in behavior in 50% of patients who displayed disordered behavior at baseline. There was improved behavior in three of four patients for whom ESES resolved and three of eight patients for whom ESES did not resolve (Fig. 4), a finding that fell short of statistical significance by Fisher exact test. Trends toward a more favorable response to treatment were also noted in those with lesional epilepsy and good seizure control at follow-up, but these differences were not statistically significant. As for those experiencing language improvement, those possessing lower IQ at baseline were more likely to experience behavioral improvement (Table 5), whereas age, age of seizure onset, duration of ESES, and duration of treatment were unrelated to behavioral improvement. Table 6 presents descriptions of qualitative changes in behavior in those experiencing a favorable response to treatment. Agitation and poor behavioral compliance remained problematic for some patients, but patients were often better able to devote

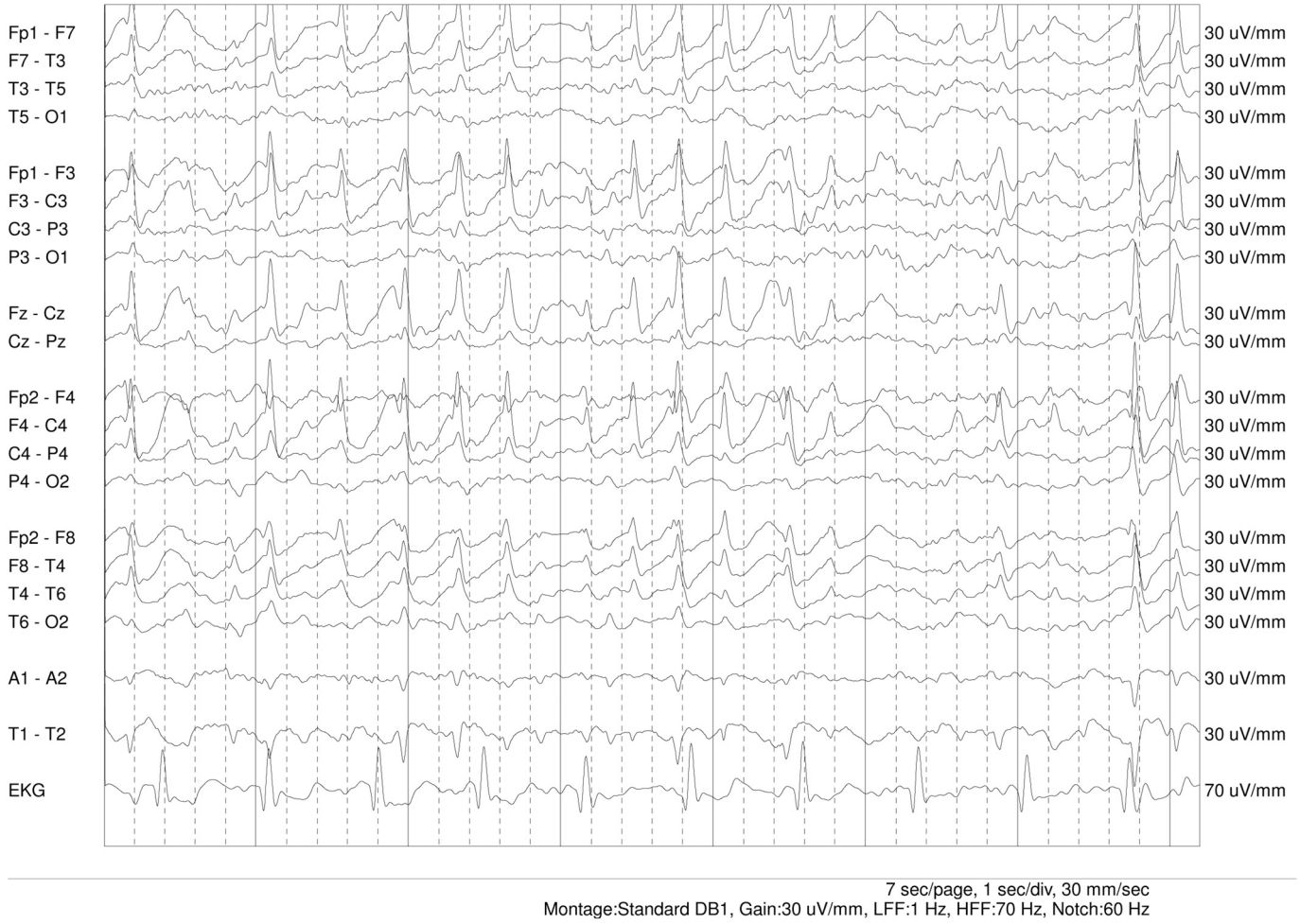

Fig. 2. Generalized activity in sleep >50% of the sleep recording.

cooperation and effort during testing, and were reported to be better able to function appropriately in school and home settings.

3.3. Other results

Two of three patients with congenital cortical malformations (polymicrogyria, tuberous sclerosis, cortical dysplasia) displayed significant improvement in behavior or language in response to treatment, which is comparable to rate of improvement in those with other etiologies. Those with a perinatal etiology (63%) were also as likely as other patients to experience language or behavioral improvement in response to treatment. There was no difference in language or behavioral treatment response between those with unilateral and bilateral ESES.

With the exception of one patient who no longer met criteria for a diagnosis of an intellectual disability, all patients maintained diagnoses of intellectual disability, ADHD, or ASD that had been established at baseline.

Table 2
Cognitive test scores prior to and during treatment.

	Baseline	Treatment	p
VIQ (n = 11)	74.5	75.5	ns
PIQ (n = 7)	71.1	72.4	ns
Naming[a] (n = 16)	79.6	81.6	ns
Comprehension[b] (n = 15)	8.5	5.5	ns

[a] Results are represented as standard scores.
[b] Results are represented as scaled scores.

There was no difference in improvement between those whose pulse-dose regimen was preceded by a burst-and-taper of daily prednisone (6 of 11 improved) and those who were treated directly with pulse dosing (4 of 6 improved). Those who were treated with a benzodiazepine in any amount were less likely to experience language or behavioral improvement than those only treated with other forms of AEDs (Fisher exact test: p = .05).

4. Discussion

This study examined the effectiveness of pulse-dose prednisone in treating language and behavioral disturbances associated with ESES. The majority (59%) of the sample experienced a meaningful improvement in language or behavior during a treatment period that extended up to 27 months without experiencing complications of treatment.

4.1. Relationship between treatment-related changes in EEG and language/behavior

Unlike several previous studies [10,11,22,23], we found that improvement in language and behavior was not dependent on resolution of ESES during treatment. There was a trend toward those experiencing resolution of ESES to be more likely to experience improvement, but a large minority of patients experienced improvement without improved EEG. This is consistent with a study by van den Munckhof et al. who also reported cognitive improvement in many patients whose spike–wave

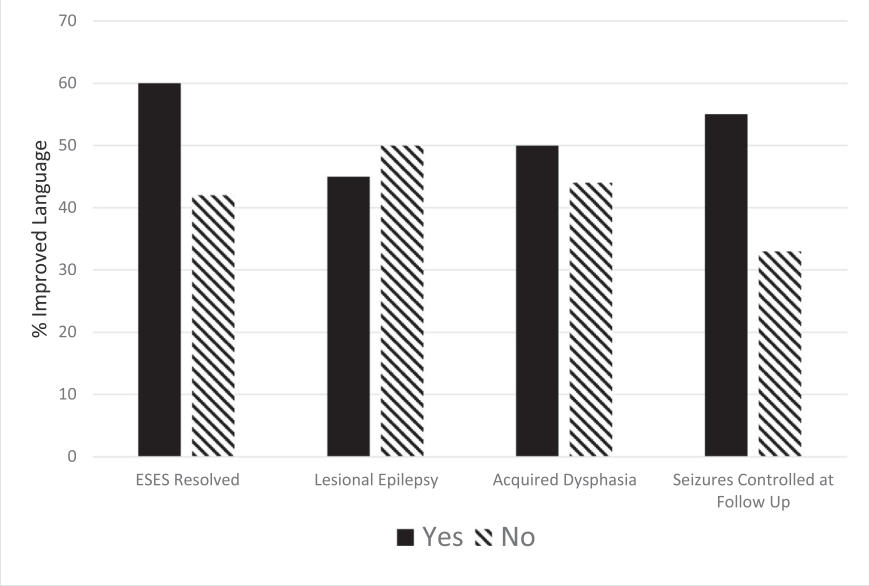

Fig. 3. Percent of patients with improved language by ESES resolution, lesional vs. nonlesional epilepsy, acquired vs. developmental language problems, and seizure control at follow-up.

index remained above 50% [24]. One explanation for the difference in current and previous findings may be that the resolution of ESES with steroid treatment may be dose dependent, with greater improvement in nocturnal epileptiform activity occurring during daily rather than pulse dosing. Our findings suggest that the basis of improvement in language and behavior may be due to factors other than resolution of ESES specifically. For example, van den Munckhof et al. have hypothesized that the positive effect of corticosteroids and intravenous immunoglobulin (IVIG) treatment in patients with ESES may be due to a reduction in the inflammatory effects of cytokines and other inflammatory proteins. They reported higher levels of interleukin-6 among patients with ESES relative to controls, and found a treatment-related reduction in cytokine levels in association with improvement in ESES [25].

Our findings also suggest that the decision to continue or discontinue pulse-dose prednisone therapy in any given patient should be made on the basis of improvement in language and behavior rather than sleep EEG alone. This is in agreement with others who have hypothesized that epileptiform activity on EEG may represent an accompanying feature rather than the principal cause of language or behavioral disturbance in ESES [26,27]. Our results are also compatible with others who have not found a strong relationship between EEG and changes in function [28,29]. A related issue is whether to reinstate treatment in a patient displaying dysphasia or behavioral regression when he/she has a history of ESES and good response to prednisone, but whose current sleep EEG does not currently demonstrate ESES. One might argue that it is the impairment, not the EEG, that one should treat in these patients, as our data suggest no direct correlation between ESES and a patient's functioning. This is true, in particular, when we consider the potentially high variability in spike–wave percentage over the natural course of disease prior to complete resolution of ESES [30].

Table 3
Characteristics of patients by language treatment response.

	Language improved	Language unimproved	p
Age (years)	7.9	8	ns
Age at seizure onset (years)	3.5	2.3	ns
Duration of ESES (months)	2.4	17.7	.09
Duration of treatment (months)	8.1	12.4	ns
Baseline verbal IQ (n = 16)	62	86	p < .01
Baseline performance IQ (n = 12)	62	84	p = .05

4.2. Relationship between history of regression and treatment-related changes

Improvements were not seen exclusively in patients with a history of distinct regression in language or behavior, although there was a trend for those who had experienced a clear decline in functioning to be more likely than those with only developmental language or behavioral disturbance to display a favorable response to treatment. The finding that improvement was seen even in those whose impairment was of developmental origin is consistent with findings of Altunel et al. who found that adrenocorticotropic hormone (ACTH) treatment resulted in some improvement in symptom severity in those with ADHD, ASD, and stuttering, conditions that are usually considered developmental in origin [31]. As has been reported elsewhere [10], significant language or behavioral limitations persisted despite an otherwise favorable response to treatment. In addition, nearly all patients demonstrated the developmental conditions (ADHD, intellectual disability, ASD) at follow-up that had been documented at baseline. This suggests that a realistic expectation for treatment should not be the complete

Table 4
Treatment-related changes in speech and language functions among those displaying a favorable response to treatment.

Patient	Baseline	Follow-up
1	Paraphasic errors and nonsensical utterances, marked impairment in comprehension and word finding	Still aphasic, but sometimes uttered some good quality sentences
2	Mildly disordered syntax	Well-organized complex sentences and increased length of utterance
3	Loquacious but halting speech	Improvement in length of utterance, fluency, and VIQ and PIQ
4	Spoke in nonsensical rhymes, paraphasic errors, decline in receptive vocabulary, but speech often fluent	Some subtle paraphasic errors and rare nonsensical utterances
6	Robotic speech and repetitive phrases	Some stammering, improvement from moderately to mildly impaired VIQ and receptive vocabulary
9	Single word utterances, speech often unintelligible	Speech immature only
13	Nonsensical speech, phonemic paraphasias, speech sometimes unintelligible	Resolution of paraphasias, speech intelligible
16	No aphasia observed	Greater than expected maturational gains on tests of VIQ and language comprehension

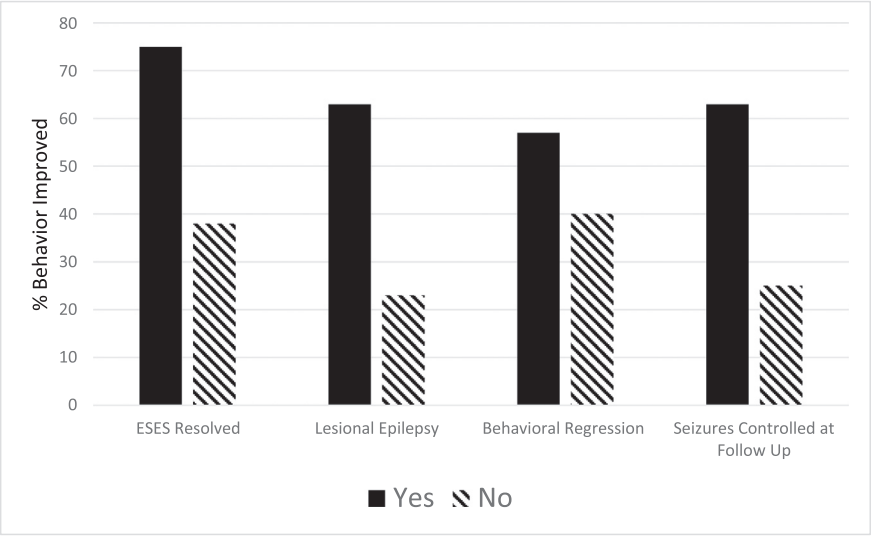

Fig. 4. Percent of patients displaying behavioral disturbance at baseline (n = 12) who experienced behavioral improvement, by ESES resolution, lesional vs. nonlesional epilepsy, history of behavioral regression, and seizure control at follow-up.

normalization of language or behavior, but meaningful improvement in daily functioning. More appropriate goals of treatment might include sufficient improvement in frustration tolerance to permit a patient to attend school, or improvement in intelligibility of speech such that a child's relative can finally understand what he/she says.

4.3. Changes in cognitive test scores

Formal test measures were relatively insensitive to improvements in language and behavior, a finding that is compatible with previously reported insensitivity of IQ scores to treatment-related changes [24]. Only two patients displayed significant improvement in test scores. A thorough parent interview, behavioral observations during testing, and a qualitative language sample were much more likely to reveal changes in response to treatment. This suggests that the decision to discontinue treatment should not be made exclusively based on unchanged cognitive test scores.

4.4. Other findings

Unlike two previous studies [11,23], we found patients with lower IQ to be more likely to respond favorably to treatment. As in a study by Liukkonen et al. [23], we found that those with structural abnormalities could be positive responders to steroid treatment. Treatment response was not significantly related to degree of seizure control during prednisone treatment, age, age at seizure onset, or duration of treatment. Though not a central aim of the study, we found that those whose regimen was preceded by a week-long interval of daily dosing were not more likely than those who were directly treated with a pulse-dosing regimen to experience improvement in language or behavior. Though not statistically significant, those who had a shorter duration of ESES at the time of treatment onset were more likely to experience language improvement. Patients with congenital malformations and those with a history of perinatal injury were as likely as the rest of the patient sample to experience significant improvement in either language or behavior. While the current study did not identify variables that predict a good treatment response, it does indicate that the presence of ESES, and verification through neuropsychological evaluation of some impairment in language or behavior, whether acquired or developmental, may be sufficient to indicate a patient is an appropriate candidate for treatment.

Table 5
Characteristics of patients by behavioral treatment response among those presenting with behavioral disturbance at baseline (n = 12).

	Behavior improved	Behavior unimproved	p
Age (years)	8.3	7.3	ns
Age of seizure onset (years)	3.2	2.9	ns
Duration of ESES (months)	14.5	10.7	ns
Duration of treatment (months)	8.4	13.8	ns
Baseline verbal IQ (n = 11)	63	85	p = .01
Baseline performance IQ (n = 9)	64	88	p = .09

4.5. Limitations

One limitation of the present study is that the sample included only patients who tolerated weekly prednisone treatment until follow-up neuropsychological testing could be conducted. Infrequently, some parents discontinued prednisone treatment when they did not observe a distinctly favorable response, which might have slightly increased our proportion of favorable responders. Small sample size may have contributed to statistically nonsignificant findings in comparisons where trends seemed evident, such as the appearance of a positive relationship between ESES resolution and improved behavioral functioning.

The largely qualitative and sometimes subjective nature of observed changes in language and behavior could be also considered a shortcoming of the current study. A more formal quantification of language variables,

Table 6
Treatment-related changes in behavior among those displaying a favorable treatment response.

Patient	Baseline	Follow-up
1	Increased aggression and emotionality, could not attend school because of disordered behavior	Still easily agitated but could attend school part days
4	Highly noncompliant behavior	Some improvement in behavioral compliance, could devote deliberate effort to testing
6	Quickness to agitation, social immaturity	Still some quickness to agitation and rigidity, but less problematic
8	Behavioral "meltdowns," decreased social interest, increased aggression toward parent	Decreased aggression, increased social engagement, more cheerful and cooperative in testing
13	Highly distractible and impulsive such that the patient was unable to fully comply with testing	Good attention to testing tasks, not distractible
14	Frequent redirection needed during testing, often cried or gave up	Increased compliance and attention during testing, cheerful

such as systematic measurement of length of utterance and errors in syntax, might yield more objectively discernable changes in language. Formal measures of ASD symptom severity, social communication, and aberrant behavior might be helpful to quantify the degree of behavior change in response to treatment.

The present study does not directly address the potential for relapse in language and behavior following discontinuation of pulse-dose prednisone. Anecdotally, we have observed maintenance of treatment-related improvements in some patients and relapses in others, the latter requiring reinstatement of therapy, sometimes for years, but without obvious detrimental outcomes. Chez et al. [13] reported that patients did not experience Cushing-like side effects with treatment even after 18 months of treatment.

We did not include a no-treatment control group, increasing the likelihood of improvements due to maturation alone. However, we did not find longer treatment interval (and hence increased opportunity for maturational gains) to be associated with greater improvement. Because we did not employ a no-treatment control group, one cannot rule out the possibility that some improvements that were observed at the time of follow-up testing might have been related to natural fluctuations in the course of the condition as has been reported in Landau–Kleffner syndrome [32].

4.6. Conclusion

This study found that, among patients who are able to tolerate treatment with pulse-dose prednisone for ESES without problematic side effects, the majority experienced a meaningful improvement in language or behavior. Improvements were most evident on qualitative measures, such as a qualitative language sample, reported history, or informal behavioral observation rather than formal cognitive test measures. A favorable response to treatment was not dependent on any single factor, suggesting that those presenting with ESES and either developmental or acquired disturbance in language or behavior, may be appropriate candidates for a trial of pulse-dose prednisone.

Disclosures

None of the authors have conflicts of interest to disclose.

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