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Case Report

Serial neuropsychological testing before and after hemispherectomy in a child with electrical status epilepticus in slow wave sleep

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ABSTRACT

Electrical status epilepticus in slow wave sleep (ESES) is typically characterized by neuropsychological deterioration to varying extents. While the syndrome is self-limiting for some, for others it has a swiftly declining course that requires aggressive neurosurgical intervention. Here, we present a patient with ESES secondary to a large left middle cerebral artery stroke in utero who experienced behavioral and neuropsychological deterioration that rapidly progressed. Collectively, her neuropsychological status, EEGs, MRIs, and nature of her ESES warranted a left hemispherectomy to prevent further decline. The patient was seen for four neuropsychological evaluations. Over time, neuropsychological test scores did not adequately capture her degree of impairment and change. Behavioral observations during evaluations and parental reports played a critical role in determining her initial decline and mild post-surgical improvement. For all practitioners, this report illustrates one extreme of ESES. It also illustrates, particularly for neurologists and neurosurgeons, the importance of behavioral observations relative to test performances.

1. Introduction

Encephalopathy with electrical status epilepticus in slow wave sleep (ESES) represents a large spectrum of disorders and is characterized by an EEG pattern consisting of sleep-related activation and diffusion of spike-wave discharges that usually occupy ≥85% of non-rapid eye movement sleep. Onset is typically between ages two and 14 years, and the condition is often self-limiting by adolescence [1–3]. However, a sizable portion of patients with ESES does not experience spontaneous remission, instead suffering rapid irreversible or minimally reversible developmental, behavioral, and neuropsychological deterioration [2]. Mechanisms underlying the presence and severity of neuropsychological impairment remain inadequately understood because the condition is rare, resulting in published research studies with small, heterogeneous samples. Some children require aggressive surgical intervention for seizure control and cessation of cognitive deterioration. A unilateral MRI lesion of structural etiology is among the top factors that produces favorable surgical outcomes [4,5]. The variability in course and presentation of ESES constrains prescription of a precise gold standard for treatment.

This case report describes a girl (BX) who developed ESES secondary to a large in utero stroke. While she initially presented to Neurology with hemiparesis and delay in achieving early developmental motor and language milestones, she subsequently developed clinical seizures and then ESES accompanied by significant neuropsychological and behavioral changes that worsened rapidly. This case illustrates how serial neuropsychological evaluations reflected marked behavioral changes over time that were poorly captured by test scores and that were not fully appreciated initially in surgical decision-making by either the treatment team or patient's family. Thus, the role of the neuropsychologist in advocating for surgical intervention to prevent further cognitive decline also is highlighted.

2. Materials and methods

2.1. The study subject

For deidentification, patient pseudo-initials were created using a random letter generator. All dates were fabricated, although relationships among dates were maintained because of their importance in understanding when events occurred relative to one

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another. All applicable items on the CARE (CAse REports) guidelines checklist[1] are reported in this paper.

2.2. Neuropsychological evaluations, measures, and scores

Neuropsychological evaluations were conducted once prior to neurosurgery and three times post-surgically (i.e., two weeks, two months, and six months). Table 1 contains all neuropsychological measures at each evaluation. Neuropsychological data (performance-based and parent-report scores) are presented in a simplified format longitudinally, grouped by domain and presented as percentiles, in Tables 2 and 3. Scores described as "average" are between the 16th and 84th percentiles. Because the neuropsychological evaluation two weeks post-surgically was brief, BX's parents did not complete questionnaires.

3. Results

3.1. Case presentation

Patient BX was born at 41-weeks gestation after an uncomplicated delivery. She initially presented to Neurology at age three years (i.e., mid-2015) with right hemiplegia and episodic behavior concerning for focal seizures. Ensuing EEG was abnormal, consistent with a epileptiform discharges over the left central area reflecting an increased tendency for the expression of focal seizures. Brain MRI several weeks later showed cystic encephalomalacia of the left cerebral hemisphere in the middle cerebral artery distribution (consistent with remote stroke), smaller left middle cerebral artery than the right, smaller left basal ganglia than on the right, and evidence of Wallerian degeneration in the left cerebral peduncle. Pre- and post-surgical MRI slices are presented in Fig. 1. She started oxcarbazepine but continued to have focal seizures.

A subsequent series of speech-language, physical, and occupational therapy evaluations over the next four months identified mild speech impairment characterized by articulation errors that reduced speech intelligibility, age-appropriate receptive and expressive language skills, balance impairment, fine motor manipulation difficulty, and sensory concerns (e.g., hypersensitivity to lights, need for increased tactile input). Additional parental concerns included inability to toilet independently and excessive appetite.

3.1.1. Clinical course

About one year after her initial EEG (i.e., mid-2016), video-EEG (vEEG) was performed and was abnormal due to the presence of left centro-parieto occipital spikes and low amplitude slowing over the left hemisphere. She presented to the emergency room for numerous episodes of status epilepticus (SE) between mid-2016 and mid-2017. Episodes of SE were characterized as focal onset (left hemisphere) evolving into bilateral convulsive SE of childhood due to remote brain injury from stroke. Seizures were 3–7 min in duration, and required rectal diazepam.

BX was educated in a developmental preschool in 2017 with an Individualized Education Program where she reportedly performed at or close to grade level. However, seizure frequency and severity peaked about 1.5 years after her initial presentation (i.e., shortly after beginning kindergarten). This coincided with abrupt slowing in academic achievement and significant behavioral changes including increased disengagement, repetitive hand motions, drooling, and behavioral outbursts. Upon returning to school after a holiday break, her clinical presentation had deteriorated (e.g., increased gait disturbance, falling out of chair, classroom wandering, talking to herself, inability to engage in schoolwork despite one-on-one assistance at all times). BX was amnestic for most events over an approximately three-month period (i.e., fall 2017 to early 2018).

vEEG repeated in early 2018 was consistent with nonconvulsive ESES with an active epileptiform discharge in the left frontal region associated with voltage attenuation over the left hemisphere. Fig. 2 shows pre- and post-surgical sleep EEGs. Follow-up MRI findings immediately after vEEG were the same as her first MRI but also included surrounding gliosis and patchy T2/FLAIR hyperintensity in the dorsal thalamus, mild malrotation of the left hippocampus, and a pineal cyst. No right hemisphere abnormalities were identified.

Over 2.5 years since starting oxcarbazepine (i.e., mid-2015 to early 2018), she had converted to trials of lamotrigine, levetiracetam, clobazam, zonisamide, and divalproex without success. By this point, her treatment team was discussing surgical options. In conjunction with poor control with antiseizure medications and abnormal vEEG and MRI findings, her significant cognitive decline and behavioral changes were a major impetus to presenting BX's parents with the option of an anatomical left hemispherectomy instead of less aggressive surgery. The prevailing opinion among her care team was that a modified hemispherectomy would be insufficient for seizure control and prevention of further cognitive decline and neurosurgical intervention was performed in mid-2018.

3.2. Neuropsychological assessment

3.2.1. Pre-surgical assessment

Because of continued deterioration, BX underwent neuropsychological evaluation in early 2018 (i.e., approximately two months prior to surgery; age six years, eight months). BX performed in the average range on measures of verbal intelligence, language comprehension, expressive vocabulary, auditory attention, and verbal memory delayed recognition. In contrast, her performances on measures of nonverbal reasoning, visuospatial skills, word reading, auditory working memory, and most motor skills (even on her side without hemiparesis) were below average. However, many behaviors (also seen at school) negatively influenced test performances including constantly asking to see her mother, complaining of task difficulty, oppositionality, extreme inattentiveness (in contrast to average performance on an attention task) and impulsivity, and requiring multiple repetitions of instructions (a product of inattention rather than insufficient language comprehension). The test battery was shortened significantly relative to what had been planned because of her inattentiveness and impulsivity. In the short period between the neuropsychological evaluation and report completion, another vEEG confirmed ESES.

3.2.2. Post-surgical inpatient assessment 1

BX underwent a complete anatomical left hemispherectomy in mid-2018 and subsequently participated in intensive inpatient and outpatient therapies for two months. A brief neuropsychological evaluation was performed two weeks after surgery during inpatient rehabilitation at age six years, 10 months. She was seizure-free during that interval of time. In contrast to pre-surgical oppositionality, BX was extremely friendly and interactive. While she remained quite inattentive, this had improved mildly. She remained profoundly impulsive but in a qualitatively different way than pre-surgically. Her impulsivity could be better described as "stimulus bound" in that she would grab any object she saw, attempt to use it properly, and then put it in her mouth. The evaluation was moved to another room with fewer distractions, but the

[1] Checklist available at https://www.equator-network.org/reporting-guidelines/care/.

Table 1
List of neuropsychological and parent-report measures used at each evaluation.

Test Domain	Test Name	Evaluation			
Patient Performance					
			Post-Surgical		
		Pre-Surgical	2 weeks	2 months	6 months
Verbal Intelligence	KBIT-2 Verbal Intelligence	x			x
Nonverbal Reasoning	KBIT-2[a] Matrices	x			x
	WISC-V[b] Matrix Reasoning			x	
Word Reading	WJ-IV[c] Letter-Word Identification	x		x	
	WRAT-5[d] Word Reading				x
Comprehension	NEPSY-II Comprehension of Instructions	x	x		x
Expressive Vocabulary	EOWPVT-4[e]	x			
	Expressive Vocabulary Test-2nd edition		x		
Verbal Fluency (Semantic)	NEPSY-II Word Generation-Semantic			x	x
Visuospatial	VMI-6[f] Visual Perception	x		x	x
	NEPSY-II Arrows		x		
	NESPY-II Block Construction	x		x	x
Fine Motor Coordination	Grooved Pegboard (Dominant Hand)	x		x	x
	VMI-6 Motor Coordination (Tracing)	x		x	x
	VMI-6 Visual Motor Integration (Drawing)	x		x	x
Grip Strength	Dynamometer (Dominant Hand)	x		x	x
Gross Motor	MSCA[g] Leg Coordination	x		x	
Auditory Attention	NEPSY-II Auditory Attention Combined	x		x	x
Auditory Working Memory	CMS[h] Numbers	x			
	NEPSY-II Sentence Repetition		x		
	WISC-V Digit Span				x
Verbal Memory	CMS Stories Delayed Free Recall	x			
	CMS Stories Delayed Recognition	x			
	NEPSY-II Narrative Memory Free Recall			x	x
	NEPSY-II Narrative Memory Recognition			x	x
Visual Memory	CMS Faces Immediate Recognition	x			
	CMS Faces Delayed Recognition	x			
	NEPSY-II Faces Immediate Recognition				x
	NEPSY-II Faces Delayed Recognition				x
Visual-Verbal Memory	NEPSY-II Names Immediate Recall		x		x
	NEPSY-II Names Delayed Recall		x		x
Parent Report					
Adaptive Functioning	Adaptive Behavior Assessment System-3rd edition	x		x	x
Executive Functioning	Behavior Rating Inventory of Executive Function-2nd edition	x			x
Behavior	Child Behavior Checklist				x
	Conners-3rd edition			x	x

[a] Kaufman Brief Intelligence Test-2nd edition (KBIT-2).
[b] Wechsler Intelligence Scale for Children-5th edition (WISC-V).
[c] Woodcock-Johnson Tests of Achievment-4th edition (WJ-IV).
[d] Wide Range Achievement Test-5th edition (WRAT-5).
[e] Expressive One-Word Picture Vocabulary Test-4th edition (EOWPVT-4).
[f] Beery-Buktenica Test of Visual Motor Integration-6th edition (VMI-6).
[g] McCarthy Scales of Children's Abilities (MSCA).
[h] Children's Memory Scale (CMS).

behavior persisted. Despite these behaviors, scores in language comprehension and auditory working memory improved mildly. While her expressive vocabulary score declined, this was largely a product of perseveration, as she repeated prior responses from another test with varying vocal inflections. Expressive language was markedly qualitatively better (in contrast to her score and relative to the pre-surgical evaluation), characterized by improved intelligibility, adequate expression of her wants, and intact ability to repeat short sentences. Formerly characterized as right hemiplegia, she demonstrated post-surgical hemiparesis. It had been anticipated that motor skills would remain unchanged post-surgically due to motor planning believed to be subserved by her right hemisphere.

3.2.3. Post-surgical outpatient assessment 2

BX's next outpatient neuropsychological reevaluation was two months after surgery, prior to returning to school at seven years of age and during this time she remained seizure-free. BX was highly impulsive, grabbing test materials as soon as they were presented and choosing answers before considering all response options, as well as being inappropriately affectionate at times with faculty/staff, which represented a change in her behavior. Word reading remained significantly suppressed, comparable to a preschooler who had not entered kindergarten. Right-sided hemiparesis remained prominent, although her dominant hand grip strength and grooved pegboard scores improved.

While no other neuropsychological scores improved, BX's responses on questionnaires greatly contrasted with past and present scores by indicating fewer concerns regarding emotional and behavioral functioning and significant improvement in all areas of adaptive functioning. She also reported several qualitative behavioral improvements including a mild reduction in mouthing objects, increased responsiveness to redirection, consistent independent toileting, and mildly decreased impulsivity (which continued to greatly contrast with what was observed during the evaluation). She denied observing any worsening of behaviors or cognition; however, social immaturity was becoming more apparent due to her lack of progress. An anticipated right homonymous hemianopia post-surgically was confirmed but BX compensated well by turning her head to the right and moving her eyes to the left to scan the right visual field.

Table 2
Neuropsychological test scores (percentiles).

Age (Years:Months)	6:8	6:10	7:0	7:4
		Post-Surgical		
	Pre-Surgical	2 weeks	2 months	6 months
Verbal Intelligence	21			19
Nonverbal Reasoning	<1		<1	4
Word Reading	1		<1	7
Language				
Comprehension	25	63		9
Expressive Vocabulary	68	10		
Verbal Fluency (Semantic)			5	16
Visuospatial				
Visual Perception	7	9	5	10
Block Construction	Invalid		5	9
Motor				
Fine Motor Coordination				
Grooved Pegboard (Dominant Hand)	3		19	16
Tracing	<1		<1	<1
Drawing	13		6	8
Grip Strength (Dominant Hand)	73		95	79
Leg Coordination	2		<1	
Attention & Working Memory				
Auditory Attention	25		Attempted	Attempted
Auditory Working Memory	5	16		9
Memory				
Verbal Memory				
Delayed Free Recall	5		16	16
Delayed Recognition	37		2–5	<2
Visual Memory				
Immediate Recognition	5			<1
Delayed Recognition	Invalid			16
Visual-Verbal Memory				
Immediate Recall		16		5
Delayed Recall		5		2

Table 3
Parent-report scores (percentiles).

Age (Years:Months)	6:8	7:0	7:4
		Post-Surgical	
	Pre-Surgical	2 months	6 months
Adaptive Functioning*			
Communication	5	50	37
Functional Academics	5	9	9
Self-Direction	9	25	16
Leisure	9	63	16
Social	16	50	37
Community Use	25	25	9
Home Living	16	75	9
Health and Safety	5	37	25
Self-Care	5	25	9
Executive Functioning**			
Inhibition	99		69
Initiation	95		86
Self-Monitoring	97		97
Task-Monitoring	99		76
Shifting	86		46
Emotional Control	66		16
Working Memory	97		79
Planning and Organizing	42		34
Organization of Materials	86		76
Behavior**			
Inattention	99	97	95
Hyperactivity/Impulsivity	99	96	82
Learning Problems	99	99	99
Anxiety/Depression	50		50
Withdrawal	84		58
Somatic Complaints	99		50
Social Problems	99		66
Rule-Breaking	69		58
Aggression	65		50

*Higher percentile indicates better adaptive functioning.
**Higher percentile indicates greater cognitive or behavioral concern.

3.2.4. Post-surgical outpatient assessment 3

BX's final neuropsychological evaluation was six months after surgery (i.e., age seven years, four months). Updated vEEG between neuropsychological evaluations was abnormal (consistent with history of hemispherectomy) but without evidence of epileptiform activity including ESES and she remained seizure-free. Because of continued limited use of her right arm and hand, she was scheduled to begin receiving botulinum toxin A injections three weeks after this evaluation.

BX denied new concerns and continued to report qualitative improvements. However, her responses on the adaptive functioning questionnaire indicated a number of mild declines compared to four months prior. Higher scores two months post-surgically likely were over-estimations of BX's abilities in the context of seeing several improvements in her daughter's functioning, while by six months post-surgically, her perception of adaptive functioning had become more realistic. She signified improvement in executive function on questionnaires comparing her pre-surgical evaluation to her 6-month post-surgical evaluation that was not readily apparent during the assessment. However, many of the improvements identified still reflected impairment relative to peers.

While BX remained distractible and impulsive, there was progressive improvement. Mouthing objects rarely occurred. Peer relations had improved, and she had developed several friendships at school. At the evaluation, motor skills and appearance were notable for right hemiparesis, wide-based gait during ambulation, right leg bracing, and chronic contracture of her right hand. She initially approached the neuropsychologist as if she was going to hug him immediately, but she self-adjusted her behavior appropriately, a notable behavioral change and demonstration of self-inhibition. Overall, behavioral observations and test scores were most consistent with one another compared to prior evaluations. BX was able to complete the evaluation without breaks, something she previ-

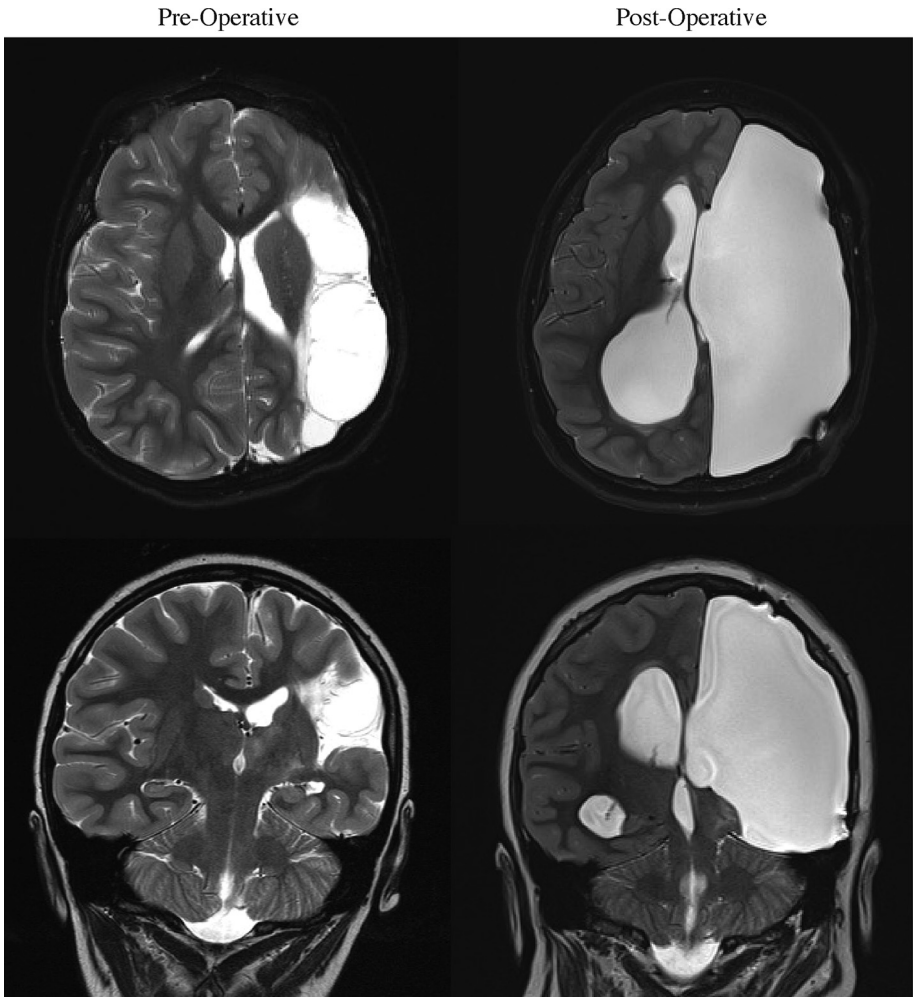

Fig. 1. T2-weighted MRIs (axial on top, coronal on bottom) demonstrating pre-operative lesion (left column) and post-operative left hemispherectomy (right column).

ously had been unable to do. Other behavioral improvements included generally understanding test directions, appropriately requesting assistance as necessary (e.g., asking the examiner to hold paper in place while she drew), and obtaining averages scores in the areas of verbal intelligence, semantic verbal fluency, visual memory delayed recognition, grooved pegboard, and grip strength. In addition to surgery, therapies, and orthopedic bracing, behavioral and cognitive improvements may have reflected reduced dosages of clobazam and zonisamide, both of which were treatments at the the time of pre-surgical and third post-surgical evaluations.

4. Discussion

We described the clinical and neuropsychological trajectory of a girl with a severe case of ESES that required left hemispherectomy to alter the course of behavioral and cognitive decline. Our case is in contrast to a recent case report in Epilepsy & Behavior Reports of a child with ESES who demonstrated a different course. In that case, neuropsychological scores showed clear decline and then demonstrated improvement and a favorable outcome with ESES that resolved with treatment before requiring surgery [6]. In our case, although neuropsychological scores remained quite suppressed through serial assessments, qualitative behavioral observations during assessments and parental reports of BX's behaviors captured critical information that otherwise would not have been obtained. While this issue is clear to neuropsychologists, it is not necessarily apparent to practitioners in other fields. As is always the case with neuropsychological evaluations, test score interpretation must occur in the context of patient-specific medical factors and behavioral observations. The present case findings are consistent with those reported in a study with children who underwent hemispherectomy due to ESES in which postoperative neuropsychological testing demonstrated only slight improvement or mixed results. No patients showed significant improvement on testing, but parents generally reported improvements and no further regression [7]. Meanwhile, Kalscheur, Farias-Moeller, and Koop's (2021) case report illustrated a vastly different trajectory of decline followed by significant improvement without surgery.

The precise mechanisms underlying cognitive deterioration in ESES are unknown, but there are several published hypotheses. Among the more compelling hypothesis is that seizures and interictal increases in cerebral blood flow create a self-perpetuating cycle of damage to neuro-vascular coupling [8,9]. Peng et al. demonstrated that the spike wave index from interictal epileptiform discharges during non-rapid eye movement sleep and cerebral blood flow velocity oscillations were at least partially responsible for neuropsychological damage in ESES [2]. BX's abnormal left hemispheric anatomy and neurovasculature may have served as contributors to ESES and lead to her underlying neuropsychological decline.

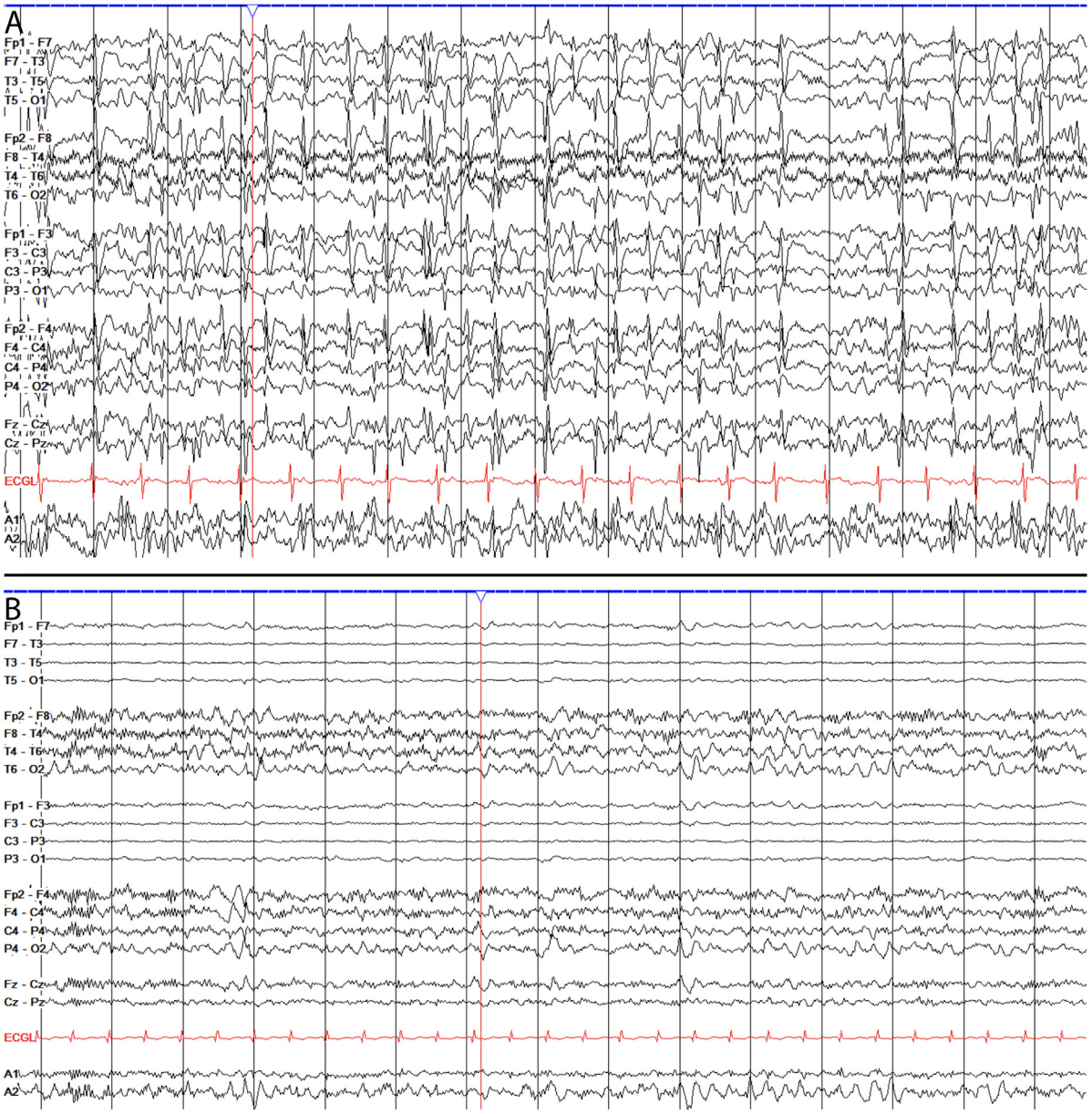

Fig. 2. Pre- and post-surgical sleep EEGs. A. 1 month prior to surgery: As soon as the patient drifted into sleep, a nearly continuous pattern of spikes, spike-slow wave, and sharps was seen. Discharges were noted up to three times per second with a slight variation in frequency as the patient was sleeping. ESES comprised 90–100% of the first two hours of sleep. B. 4 months after surgery: ESES was not seen. EEG settings: Bipolar montage; LFF 1 Hz, HFF 70 Hz, 10 μV/mm, 15 s/page.

One of the most apparent limitations of this case report was the lack of consistency in the exact neuropsychological measures utilized at each evaluation. The rationale for test selection at each evaluation was guided by the principle that, because of the relatively short intervals between evaluations, we hoped to mitigate practice effects as not to give a false sense of neuropsychological improvement. However, BX's behavioral manifestations of cognitive impairment, particularly her inattention and impulsivity, likely would have resulted in minimal, if any, practice effects that would have skewed interpretation, a point highlighted by van Iterson et al. [10].

5. Conclusions

This case report exemplifies the unique but vital contribution neuropsychology can make to understanding the impact of ESES on the developing brain. Anecdotally, our experiences support prior suggestions that all children with ESES should undergo at least two neuropsychological evaluations to assess impairment [11], although the precise neuropsychological tests and time intervals at which assessments occur (e.g., every six months) remain to be determined. Nevertheless, serial neuropsychological assessment, particularly in close proximity to EEG changes or after initiating other treatments such as steroids, may be among the most informative prognostic data utilized in surgical decision-making. Because of the rarity and heterogeneity in presentation of ESES, multi-center reports with common data elements are needed to identify factors predictive of successful outcomes to establish more consistent treatment standards.

Ethical statement

This case was submitted to Akron Children's Hospital IRB who determined that this did not constitute research because it con-

tained presentation of a single subject's retrospective data who no longer receives care through the same institutional service. A data use agreement (DUA) was obtained and executed between the two authors' current institutions. Per the DUA, the manuscript was reviewed and approved by Akron Children's Hospital.

Author contributions

DP was the primary neuropsychologist responsible for the patient's care. DP consulted with LS throughout BX's care regarding data interpretation and treatment recommendations. DP and LS participated in all multi-disciplinary team discussion regarding the patient, and LS also observed the patient's behavior at one neuropsychological evaluation. DP wrote most of the original draft of the manuscript, created preliminary versions of the tables and figure, and formatted the manuscript to journal requirements. LS wrote portions of the original draft, performed editing on all subsequent drafts of the text, edited tables, and acquired images for the figure.

Declaration of competing interest

Authors have nothing to disclose.

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