Case Report

Acquired visual agnosia as an uncommon presentation of epileptic encephalopathy in a 6-year-old boy with CSWS

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ABSTRACT

Background: Acquired visual agnosia in the context of continuous spikes and waves during slow sleep (CSWS) is rarely described. We present a case of an almost 7-year-old boy who lost his ability to name pictures and recognize familiar faces. Initial encephalography (EEG) revealed sleep induced epileptiform activity with a spike-wave index (SWI) of 100%, predominanting in the left posterior head region.
Methods: Serial neuropsychological testing with concomitant EEG was done during the first 18 months of treatment with intravenous methylprednisolone. We administered intelligence scales, verbal tasks (memory, fluency), visual tasks (drawings, search, face recognition), and tasks requiring visual-verbal integration (picture naming, visual closure).
Analyses: Neuropsychological recovery studied with reliable cognitive change cut-offs and 95% confidence intervals.
Results: With treatment, there was an improvement of the EEG pattern (SWI reduction to 45%), followed by a relapse (SWI 82%). Neuropsychological measures in part synchronized with improvement, stability, and fluctuating values. Significant increases were seen on Verbal Comprehension Index and semantic memory. Visual Spatial Index remained unchanged (67 to 73). Naming pictures showed only limited change. Interpreting degraded pictures remained extremely difficult.
Discussion: Acquired visual agnosia may be seen in children with CSWS. Early recognition, prompt accurate treatment and tailored neuropsychological assessment remain crucial.

Introduction

Visual agnosia in children – namely the inability to recognize objects in the absence of an ophthalmological disorder – is considered a cerebral visual impairment (CVI) associated with deficits in the ventral visual stream.

Recognizing an object goes beyond visual analyses and is a complex hierarchical process. Object recognition depends on the visual ventral stream, that is, the occipito-temporal association fibres that run bilaterally and connect various brain areas from posterior to anterior [1–4]. These brain areas respond differentially (or preferentially) to specific visual exposure, like living organisms versus non-living objects, man-made objects like tools, objects having features shared by many or only by some objects, but overlap is large [1,5,6]. With increasing age and with experience, localization and lateralization of visual functions show increased specificity [7–11]. For example, imaging studies on children in the early school years (ages 5–8 years) have shown that distinct temporal areas develop a preference for tools earlier than for animals [10], and selectivity for letters and words versus faces develops as a function of learning to read [11]. With age, there is an increasing lateralization, with visual search relying more on the right, and object naming more on the left hemisphere, but the integrity of both hemispheres often remains important for visual recognition [7–9].

Visual agnosia is rare, but may be seen in children born preterm or with perinatal brain injuries. In these children, visual agnosia is considered a developmental disorder that becomes evident when the child becomes older and may appear as the difficulty to recognize quite specific objects, letters, or people [12,13]. Visual agnosia

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in children, acquired after a period of normal development, is not often reported [14].

Acquired agnosia in the context of epilepsy, is most often described as *auditory* agnosia. The subtype of auditory agnosia is relevant in Landau-Kleffner syndrome and is characterized by agnosia for familiar sounds and loss of already acquired language. It is similar to the syndrome of spikes and waves in slow sleep (CSWS) due to Electrical Status Epilepticus in Sleep (ESES) on EEG, as an epileptic encephalopathy associated with cognitive loss. On the sleep EEG, children with CSWS show almost continuous spike-wave discharges during slow sleep, with spike-wave indexes (SWI) higher than 85%. SWIs between 50% and 85% [15,16] are now also being considered part of the CSWS spectrum., with lower SWI percentages partly associated with milder phenotypes of [17]. The epileptic activity is most often seen in the perisylvian region and cingulate gyrus, but can also present in the frontal or parietal cortex [18]. While CSWS often resolves spontaneously during puberty, variable long-term neuropsychological outcomes are seen, ranging from normal outcome to severe developmental deficits [19,20]. Encephalopathy with CSWS may or may not be preceded or accompanied by clinical seizures [20].

Visual agnosia in the context of epileptic encephalopathy with CSWS, is seldom described. To increase awareness of this atypical manifestation of encephalopathy with CSWS, we report a rare case of acquired visual agnosia. We hereby provide the electroclinical and detailed neuropsychological course over 18 months following diagnosis.

Case

A six-year-old, right-handed boy was referred to the Child Epilepsy Center (KEC) of SEIN by his pediatric neurologist. Six months prior to referral, the boy had started experiencing visual complaints like difficulties reading letters and numbers, with progressive difficulty recognizing familiar faces. He was born after a full term, uncomplicated pregnancy and delivery. He had suffered a single typical febrile seizure at age two years but had never experienced an afebrile clinical seizure. The child had shown an unremarkable development up to age 5½ years. He appeared as a

bright child who taught himself to read. He then gradually lost interest in reading and he was unable to recognize his mother when she picked him up from school. On evaluation, he was unable to name most of the figures on the eye chart.

Neurological examination showed no focal signs. Ophthalmological examination, visual evoked potentials and brain imaging (MRI), were normal. A first electroencephalography (EEG) showed focal occipital epileptiform discharges, which were considered to have a causal relation with the CVI and he was treated with levetiracetam followed by oxcarbazepine. The initial treatment was ineffective in improving his visual functioning, and therefore, ASM was discontinued.

At SEIN, 24-hour CCTV-EEGs were performed in the Epilepsy Monitoring Unit (EMU) using Micromed EEG system (Micromed Mogliano Veneto, Italy). The first EEG, at T1 (Table 1), showed a low-voltage (30–50 μv) reactive posterior dominant alpha rhythm intermixed with irregular theta and delta. In wakefulness, the EEG demonstrated frequent (1 per 15-second epoch) isolated spikes and (poly-)spike waves at the temporo-parieto-occipital region with left sided predominance. During NREM (stage I-IV) sleep it displayed an increase of regional spike wave activity up to 100% (SWI = 100%). During REM-sleep the index decreased (no index was determined). The child evidenced no clinical signs during the EEGs.

No abnormalities were seen on neuroimaging. 3-Tesla MRI was performed repeatedly, and included 3D T_1-weighted sequences, 3D FLAIR, 3D T_2-weighted sequences, diffusion tensor imaging, and susceptibility weighted imaging. Postprocessing software (MAP 18) was applied, which also showed no structural lesion. Array-based comparative genomic hybridization (array CGH) showed a 15q11.2 duplication, inherited from the asymptomatic mother and considered a familial polymorphism with uncertain relation to the clinical symptoms, because of reported low penetrance [21]. Whole exome sequencing showed no abnormalities.

An unusual neuropsychological phenotype was observed at his first assessment (T1), when the boy insisted he was unable to name common coloured pictures on a card. His presenting difficulties confirmed the diagnosis visual agnosia.

Table 1
Timeline of treatment with Clobazam (CLB), Ethosuximide (ESM) and Methylprednisolone (MP), cumulative number of MP pulses, results on EEG and results on neuropsychological measures. Index scores and age-adjusted asymmetrical 95% Confidence Intervals [in square brackets] on the Intelligence Tests (WISC-V[NL]/WISC-III[NL]), Peabody Picture Vocabulary Test (PPVT) and Story Learning Test iter-SEIN, at serial assessments T1 to T5.

	T1	T2	T3	T4.1	T4.2	T5
Timeline (months)	0	3	6	12	14	18
Neurology						
Antiseizure medication		CLB start/stop	ESM start	ESM stop		
Methylprednisolone pulses	start MP	MP (3)	MP (6)		MP (9)	MP (12)
EEG (Spike Wave Index)	100%		49%	60%		82%
Neuropsychology						
Intelligence	WISC-V	WISC-V	WISC-V	WISC-III / WISC-V		
FS-IQ	76 [71;83]	100 [94;106] *	99 [93;105] *	96 [89;104] **		
Indexes						
General Ability	81 [75;89]	92 [86;99] *	101 [94;108] *			
Cognitive Proficiency	64 [61;76]	102 [95;109] *	78 [72;87] *			
Nonverbal	63 [59;71]	83 [78;90] *	80 [75;87] *			
Verbal Comprehension	100 [91;109]	116 [106;123] *	116 [106;123] *	120 [110;126] **		
Visual Spatial	67 [62;79]	67 [62;79]	86 [79;96] *	73 [66;83]		
Processing Speed	49 [46;62]	83 [77;93]*	56 [52;68]	72 [66;83]		
Fluid Reasoning	76 [70;86]	85 [79;94]	91 [84;99] *			
Working Memory	88 [81;98]	120 [110;126] *	107 [98;115] *	88 [81;98]		
PPVT	76 [71;85]	68 [64;78]	78 [73;87]	110 [102;116] *		
iter-SEIN Story Telling	Story A	Story B	Story D	Story C	Story B	Story A
Learning Index	106 [94;118]	117 [107;127]	115	112	106 [96;116]	135 [123;147] *
Retention Day 1	109	123	117	106	117	117
Retention Day 2/3	112	109	103	114		112

Note. * = value falling outside of 95% CI of T1; ** = Reliable cognitive change.

Methods

After the first electroclinical and neuropsychological assessment (T1), he was given ASM and methylprednisolone pulse (MP) treatment. To monitor the treatment effects, several 24-hour EEGs and neuropsychological evaluations followed (T2 to T5) at the EMU over 18 months. Table 1 and Figs. 1 and 2 show the timeline, the treatment, and the neuropsychological results. To obtain a comprehensive picture of the child's neuropsychological functioning, the intelligence scale was supplemented with selected measures that assessed: (a) predominantly verbal abilities, (b) predominantly visual-motor abilities, and (c) the ability to integrate visual and verbal processing. Over time, tests were repeated, wherever possible, using alternate versions. At T2, no EEG was conducted. Given the absence of an evaluation by the neuropsychologist (LvI) at T4 (T4.1), the boy returned to see her five weeks later (T4.2).

Neuropsychological measures

The reader should be referred to supplemental information (Table S1 and Figure S1) for details on the neuropsychological testing. The assessment featured the WISC-V^NL, the Dutch Wechsler Intelligence Scale for Children, 5th Edition [22]. Ten subtests lead to five primary indexes; two ancillary indexes known as the General Ability Index (GAI) and Cognitive Proficiency Index (CPI); a Non-Verbal Index (NVI); and a Full-Scale IQ (FS-IQ; Table 1). For the T4 assessment, the child was administered an alternate form to the WISC-V^NL, namely subtests from the earlier 3rd edition of Wechsler's children's scales, the WISC-III^NL [23].

Verbal neuropsychological tasks administered included: (a) alternate versions of the iter-SEIN story telling test, leading to a Learning Index and Retention Indexes the same day and at Day 2 or 3 [24,25]; (b) verbal semantic fluency and letter fluency. Tasks requiring both verbal and visual processing included (a) pointing at pictures named by the examiner, from the Peabody Picture Vocabulary Test, PPVT-II^NL [26]; (b) naming pictures of 15 different common coloured objects from the Lindeboom card; (c) rapid naming of drawings of animals or objects which appeared

repeatedly on a sheet [27,28]; (d) naming of visual degraded pictures [29]; (e) mentally reassembling of drawings of pieces of a puzzle [30]; and (f) telling the stories depicted on two coloured cards, known as the Picture Story subtest [29]. The predominantly visual or visual-motor tasks were: (a) copying geometrical drawings [31]; (b) recognizing the face of a previously presented photograph from the subtest Faces [32]. The ability to display visual selective, auditory sustained attention and combined visual-auditory divided attention was measured with subtests from the TEA-ch [33].

Parents provided oral information on their child's development and filled in questionnaires tapping their perception of the child's attention, autism, social behaviour, anxiety/depression [34] and executive functioning [35]. Parents gave written informed consent for the case report.

Analyses

Normal variation, regression to the mean and practice effects may all account for changes observed during serial assessment [36]. Practice effects are most clearly seen at second testing, diminish thereafter [37], and are overall smaller in children with epilepsy than in typically developing children [38]. To establish cognitive change, earlier studies [39,40] have used confidence intervals (90% or 95% CIs), and measures based on standard errors (SE), or on standard deviations (SD). Wise [41] proposed that changes of + 2SD should be termed "recovered", +1 SD "positive response", and + 0.5 SD "minimal response". We applied the criterion of 95% CI (Table 1) and ± 2 SDs (Supplementary Table S2) to establish meaningful changes (loss or recovery). First, wherever provided by test makers, 95% confidence intervals were used to determine whether any subsequent test scores were significantly different from earlier scores [42]. In addition, to establish reliable cognitive change from T1 (WISC-V^NL) to T4 (WISC-III^NL), cut-off values were determined for 95% CIs after twelve months [39]. This procedure yielded cut-off scores of 17 IQ points for the verbal abilities, 22 for the non-verbal abilities, and 17 for FS-IQ. Second, for standardized subtests with a mean = 10, SD = 3, a difference of 2 SD (6 scaled score points)
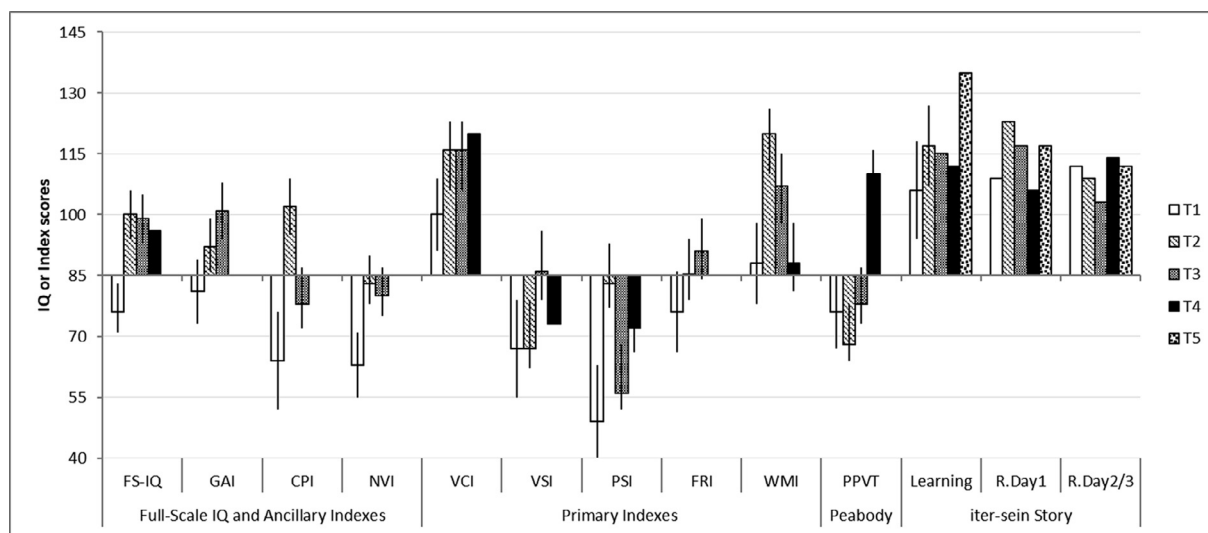
Fig. 1. Results (index scores and 95% age-dependent asymmetrical confidence intervals) on the WISC-V^NL Intelligence Scale, the Peabody Picture Vocabulary, and iter-SEIN Story telling at serial testing T1 to T5. FS-IQ = Full Scale IQ. GAI = General Ability Index. CPI = Cognitive Proficiency Index. NVI = Nonverbal Index. VCI = Verbal Comprehension Index. VSI = Visual Spatial Index. PSI = Processing Speed Index. WMI = Working Memory Index. PPVT = Peabody Picture Vocabulary Test. Learning = iter-SEIN Learning Index. R. Day 1 = iter-SEIN Retention Index Day 1. R.Day 2/3 = iter-SEIN Retention Index Day 2 or Day 3. The absence of 95% CI, (as for the iter-SEIN Retention Index), indicates that no confidence intervals were reported in the manuals. Note that mean = 100, SD = 15 and that the × axis is set at 85. Values above the axis are considered average. Index score 85 = -1SD = 16th percentile; score 70 = -2SD = 2nd percentile; score 55 = -3 SD = 1st percentile.
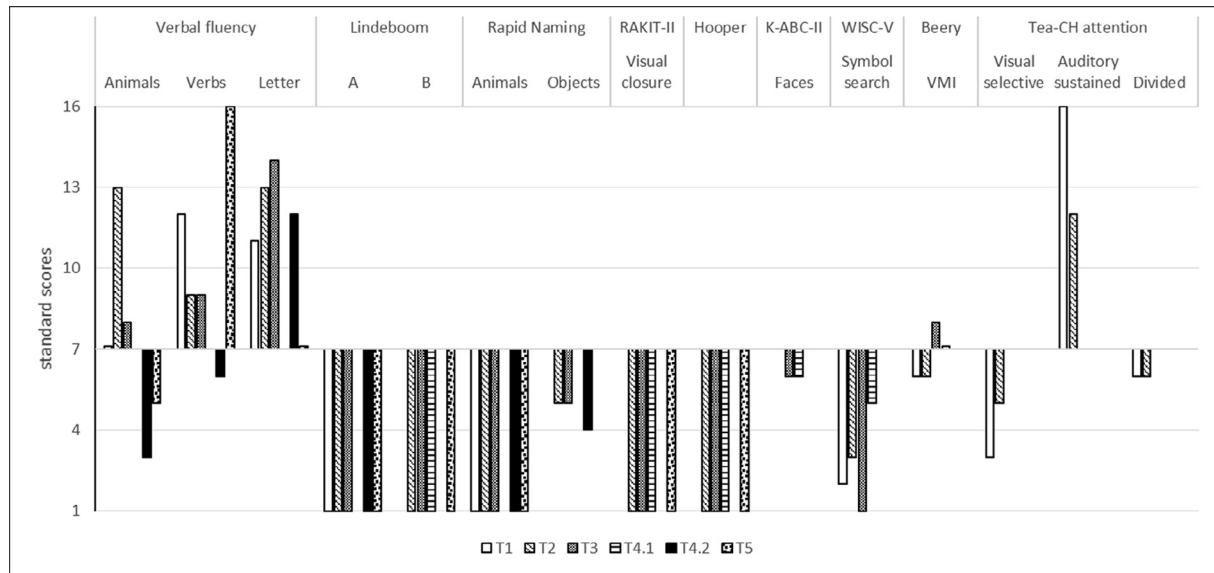
Fig. 2. Results (standard scores with mean = 10, *SD* = 3) on the neuropsychological tests at T1 to T5. For rapid naming animals, only results on card A are depicted. See Supplements for test descriptions and raw scores. Note that the × axis is set at 7, with all values above the axis considered average. Scaled score 7 = -1*SD* = 16th percentile; score 4 = -2*SD* = 2nd percentile; score 1 = -3 *SD* = 1st percentile.

from the earlier measure was interpreted as significant change (loss or recovery).

Results

Electroclinical measures

The child was treated with intravenous methylprednisolone (MP, 3 days 20 mg/kg/d), once a month for a period of six months. After clinical evaluation halfway this treatment period (T2) oral clobazam (0,8 mg/kg/d) was added. Clobazam was withdrawn after one month, because of adverse behavioural events. The second overnight video-EEG (T3), after six MP pulse treatments were given, showed a significant improvement (SWI reduction to 49%). Ethosuximide was started (titrated up to 30 mg/kg/day) as standard ASM for CSWS. The third (T4) and fourth EEG (T5), performed 12 and 18 months after the T1, progressively worsened with SWI scores of 60% and 82%, despite adjunctive MP pulse administration. With some bilateral and generalized involvement, epileptic activity showed predominantly left posterior spreading. No clinical seizures were seen over the treatment period. Table 1 shows the timeline, ASM, the cumulative number of MP pulse treatments, and the SWIs after MP treatment.

Neuropsychological measures

For neuropsychological tests with a mean = 100 and *SD* = 15, results are provided in Table 1 and Fig. 1. Note that the × axis was set at 85 (- 1 *SD*, 16th percentile). For the additional subtests, raw scores are provided at Supplementary Table S2; Fig. 2 shows the standardized scores (mean = 10 and *SD* = 3), with the × axis again set at −1*SD* (standard score 7).

Intelligence

During the course of treatment, no further cognitive loss was observed. Overall, the child improved on FS-IQ over time (Table 1). His gain of 20 points in FS-IQ between T1 and T4 – after a change in test version – was indicative of reliable cognitive change (recovery) during the treatment period. Reliable cognitive change was also

seen for the verbal abilities (VCI). Visual Spatial/Perceptual Index remained unchanged (67 to 73).

The child also made gains on GAI, reflecting improvement on its components, most clearly the indexes Verbal Comprehension (VCI) and Fluid Reasoning (FRI). The Cognitive Proficiency index (CPI) fluctuated significantly. CPI comprises the Processing Speed Index (PSI), which relies on visual and motor abilities, and the Working Memory Index (WMI); the boy's performance fluctuated on both of these indexes. The ancillary Non-Verbal Index (NVI) changed from 63 at T1 to 83 at T2 and 80 at T3. While the child's gain in NVI was also suggestive of recovery, his scores remained low.

Verbal versus non-verbal abilities. At T1, the child's difference between his verbal and non-verbal abilities, (i.e., VCI vs. VSI), was 33-points. A difference of that magnitude is considered very rare (base rate for the difference < 0.01%). At T4, the difference between verbal and non-verbal (performance) abilities had increased to 47 points. This meant that the boy's clearest gains as a result of treatment were in the areas that he was already proficient; by contrast, his test scores on areas of deficit largely remained low during the intervention.

Verbal tasks

The child performed well on verbal tasks and showed meaningful gains over time (Figs. 1 and 2, Tables 1 and S2).

Semantic memory. Overall, the child performed well on the iter-SEIN. Indexes for Learning and Retention, ranged from average to very high.

Verbal semantic fluency and letter fluency: Variable performance, with low scores to high average scores, was seen. Fluency for animals decreased, fluency for verbs increased.

Visual – Verbal tasks

The child's severe picture naming difficulties seen at the first testing remained over time.

Pointing to pictures of spoken words. At T1, the boy's low standard score on the PPVT (76) indicated that in the light of his average verbal comprehension (VCI = 100), his ability to identify pictures named by the examiner was impaired. At T4, recovery was seen.

Lindeboom colored pictures. At T1, the child indicated that he was not able to identify the pictures presented on card A. He skipped items, and after three minutes the was able to produce only one correct response out of 15: a *question mark*. By way of comparison, 6 year-olds are expected to get 14 or 15 pictures correct, with an average completion time of 24.8 seconds (*SD* = 6.9; range = 14–39). Noteworthy, the item he knew was not an object, but a punctuation mark. Pictures were numbered and he identified the numbers. Given a new try at naming the pictures, he diligently counted the pits of a *strawberry* and concluded it must be a *ladybug*. In a third round, he gave different names to the already (albeit wrongly) identified objects. He described the colors of the pictures. Both his completion time and number of correctly named pictures fell far below the lowest scores of the 6-year-old age norms. From T2 onward, the boy improved in the sense that he was able to name a few pictures (Supplementary Table S2), but he continued to fall 3 *SD* below the norm on both number of pictures correctly named and completion time. While he performed somewhat better on card B, standard scores were similarly low.

Rapid Animal Naming. At T1, the boy totally failed Animal card A. The pictures repeated themselves often, and with great effort, he "discovered" that a picture appeared more than once. At T2, the examiner provided the picture names. With progressively more difficulty and longer times at each row, he completed the task (Table S2). Animal card B was skipped to lessen the child's distress. From T4 onwards, he was able to do both cards, but his time to complete remained extremely long.

Rapid Object Naming. At T2, he named two pictures correctly, and also gave two "globally" correct responses (e.g., bench for chair); the examiner provided the missing names. Whereas the child performed better on Object naming than on Animal Naming – and results were scorable – he demonstrated no major improvement over time.

Visual closure and Visual Integration. At T2, he was unable to identify any picture on any of the tasks (see Table S2). He displayed either small increments or no improvement at all on later assessments, suggesting that these more complex visual tasks remained virtually impossible for him to solve.

Picture Stories. His Picture Story performance was very poor at T1, though he improved at later assessments.

Visual and visual motor tests

Overall, little improvement was seen on visual and visual-motor tasks.

Visual-motor Integration. At T1, the boy copied the simple line drawings (e.g., circle) of the Beery but failed on the composite pictures; no changes were seen at subsequent sessions. At T1, he drew only a dot and a stick to represent a person engaging in an activity; later on, however, he was able to draw stereotyped (but recognizable) human figures.

Face recognition. The child scored low even when compared to norms for younger children; no progress was observed.

Parental information

Scholastic achievement: With special aid for children with CVI, the child progressed from grade 1 to grades 2 and 3 in regular education. His reading speed and mathematical abilities were lower than that of his classmates'. He engaged in ball sports and music. He distinguished teachers and schoolmates by their voices. At T5, his mother reported overall improvements and stable mood, but also the persistence of the visual problems.

Parental appraisal of behavior. T1, his parents' ratings classified him in the clinical range on Attention Deficit Disorder and General Anxiety, and in the subclinical range on the Autism scale. Over time, the parents showed no particular concerns in their ratings, except for a subclinical score on the Autism scale. The parents expressed no concerns about the child's executive functioning at any measurement. Overall, the child's problems appeared moderate and transient to his parents, and reflected adaptation problems. On all measurements, the parents rated the child's health and quality of life as excellent.

Discussion

In the present paper, we described the electroclinical and neuropsychological trajectory of a boy with a highly atypical presentation of epileptic encephalopathy with posterior (predominantly left occipito-temporal) CSWS and with disabling acquired visual agnosia.

During the first months of treatment, the EEG improvements were largely mirrored by the neuropsychological evaluations. The last assessment showed that the boy continued to make small gains on neuropsychological tasks in spite of the increased EEG abnormalities.

On the neuropsychological level, the child was able to read letters and numbers; he progressed at school. Improvement was seen mostly on verbal tasks. Reliable cognitive change suggestive of recovery was seen on Full-Scale IQ, verbal abilities, and semantic memory. On the other hand, the child showed low scores and a fluctuating course on tasks with higher reliance on visual and motor abilities and visual motor speed. He evidenced gains across reassessments on the composite Non-Verbal Index, but his scores remained low.

The child's core neuropsychological deficit resided in his inability to visually interpret and, therefore, name common objects. With treatment, he displayed limited progress. For example, he passed from non-quantifiable performance to quantifiable albeit extremely low scores. He remained virtually unable to interpret degraded visual information and scored low on visual tasks requiring search, matching, integration, or closure. He remained largely unresponsive to faces, which may have contributed to parental judgement that he showed subclinical autistic-like behaviours. Within the verbal tasks, it was noteworthy that he showed no progress in semantic fluency for animals, possibly reflecting privation of meaningful experience.

On the electroclinical level, changes were seen during the course of eighteen months. At first, these changes suggested adequate response to methylprednisolone, with SWIs decreasing from virtually 100% to 45%. Thereafter, SWI rose again to 82%.

These results are in line with previous studies on CSWS, particularly on acquired auditory agnosia, suggesting that recovery of cognitive functions, lost after early normal development, may be complete in some cases and limited in others, while fluctuations over time may also be seen [20]. As in the present study, retrospective cohort studies on the treatment of encephalopathy with CSWS had shown that anti-seizure medication (ASM) is often found ineffective, while highest efficacy in terms of the improvement of electroclinical picture may be obtained with corticosteroids [43].

The profile of preserved, developing, and afflicted areas of performance in visual tasks is consistent with the results of previous studies, which suggest both specialisation and overlap in the brain areas responsible for different kinds of object recognition [1,5,6,10–12]. The present findings, however, are noteworthy. For example, while young children show less specialization for visual information [7,9], this child first tested at the age of 6 years already showed highly specific deficits (naming animals) together with equally specific preserved areas (naming letters and numbers). Also, depending on the stage of visual processing – earlier or later in the ventral stream – different disorders may be found [44–46]. In the presence of CSWS, which spreads over the brain, it could be expected that early or later processing of visual information

(or both) could be affected. We suggest that the difficulties experienced by the child with drawing, with visual matching, and naming common objects, could best be understood as a disorder in early visual processing, referred to as *apperceptive* visual agnosia. Apperceptive agnosia is characterized by the inability to recognize objects, to draw a figure, or to copy a figure. Disorders later in the ventral stream, on the other hand, are referred to as *associative* visual agnosia. In associative agnosia, the person can successfully copy a drawing, but cannot recognize or name what is pictured [44–46].

The child showed major visual problems, but walked around freely and was able to engage in ball sports. This pattern is consistent with the occipito-temporal dysfunction associated with the ventral stream of visual information processing ("knowing what"), with spared functioning of the dorsal, occipito-parietal stream ("knowing where"). Impairments in the dorsal visual stream may lead to impaired perception of moving objects [12,47]. These were not observed in the present case.

The case presented here bears a large resemblance to a previous case [48]. The authors followed, for two years, an 8-year-old child with predominantly left occipito-temporal CSWS. As in our case, the child presented with object naming disabilities, disorders in the identification of complex figures, and difficulties copying; verbal abilities and semantic memory were preserved. The persistent low scores on visual integration in our case suggest a more severe picture.

Our case also bears resemblance to a report of a 12-year-old girl with chronic bilateral occipito-temporal epileptic activity [49] and severe deficits in face recognition and naming. Similar to our case, the child had relatively spared verbal abilities compared to the non-verbal deficits and had no difficulties moving around in space. Different from our case, the girl had intellectual, reading and psychiatric disorders. Based on functional neuroimaging, the authors provided an example of the dissociation between the ventral and dorsal streams of visual information processing in their case. The girl failed to show category specific-brain activation that was typically seen in control children. The intact dorsal stream allowed the girl to move around normally, but the impaired ventral stream prevented her from properly interpreting faces, tools, and common objects.

Limitations and assets of the study

In an extensive study pooling CSWS-case studies [43], the authors contended that quantitative neuropsychological data were often lacking in these studies or were limited to changes in IQ. In the present study, with some variability and additions, we followed the child's neuropsychological performance systematically throughout eighteen months. Virtually all measures were quantitative. Beyond data on general intellectual development, we included specific data on the major neurodevelopmental deficits.

Given the severity of the clinical picture seen at presentation, treatment effects were monitored closely. Repeated testing, however, poses methodological challenges and limits interpretability, given that practice and learning effects may influence the later scores. To deal with these problems in interpretation, parallel or alternate forms were used wherever available. Also, criteria for gains or losses were set to evaluate the meaningfulness of observed changes.

Beyond practice effects, positive changes seen in verbal abilities were considered an indicator of recovery. They exceeded the criteria for reliable cognitive change, and they were mirrored on other verbal tasks, tested with alternate forms, such as semantic memory. On the other hand, the absence of major changes in the core problems – as in the visual spatial tasks or picture naming tasks – was brought into the light. This lack of progress in areas of severe deficiency occurred despite numerous repetitions after short intervals.

A drawback is that neuropsychological tests that tap specific functions often lack appropriate norms (i.e., Dutch). The lack of appropriate norms, together with the lack of alternate versions of most specific neuropsychological tests, have prompted the authors to co-norm existing tests and to develop alternate forms. These alternate forms enrich the possibilities of assessment and enable within-subject comparisons, but lead to limitations in international comparisons.

Conclusions

In rare cases, children may present with posterior CSWS, associated with acquired visual agnosia. As in acquired auditory agnosia, acquired visual agnosia my present without clinical seizures.

The visual disorders as well as spared areas of visual performance may be highly specific. The detection of the disorders may be especially challenging due to the dichotomy between apparent visual integrity and discrepant integration and interpretation of information.

EEG results and results of neuropsychological evaluation may show only partial synchronicity, suggesting that SWI is likely not the only neurophysiological marker of encephalopathy.

Recommendations

As with acquired auditory epileptic agnosia in the absence of clinical seizures, the presence of acquired visual epileptic agnosia should be acknowledged whenever a child with normal eyesight presents with first symptoms of deterioration in visual perception.

In neuropsychological assessment – even without a running EEG – beyond global cognitive deterioration, unusual patterns of high and lower abilities, atypically large variability in performance should alert the clinician as to the possibility of underlying nighttime epileptiform activity.

We suggest that in concert with early treatment, electroclinical evaluations together with tailored serial neuropsychological testing, should be used to demonstrate improvement over time.

Ethical Statements

The authors state that they read the ethical standards of the Journal and comply with them

The authors state that they obtained written informed consent by the parents for the case report

The authors state that the article has not been published elsewhere

The authors state that the article is not submitted for consideration elsewhere.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ebr.2021.100465.

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