Cognitive and Behavioral Disorders in Rolandic Epilepsies and Variants*

Summary

This article reviews the data on the frequent temporary cognitive-behavioral disorders encountered in BPERS (Benign Partial Epilepsy with Rolandic Spikes) with emphasis on their probable direct epileptic origin. Although behavioral and school problems had been mentioned in initial descriptions of the syndrome, BPERS was for a long time seen as a benign epilepsy syndrome and other problems tended to be dismissed and not envisaged as a possible "cognitive" manifestation of epilepsy. It was gradually realized that there was a close relationship between BPERS and acquired epileptic aphasia (Landau-Kleffner syndrome), which was the first example of a mainly "cognitive" epilepsy in children. Prolonged reversible oromotor deficits were subsequently recognized during the active epilepsy phase in some children with an otherwise typical syndrome and good final prognosis. These cases showed that this epilepsy syndrome could cause prolonged "epileptic" deficits. Several neuropsychological studies confirmed the clinical experience that children with BPERS had normal intelligence but that a proportion showed variable attentional or selective (language, visuospatial etc.) deficits as compared to normal controls. This was thought to possibly explain some of the learning and school problems that many of these children experienced in the active phase of the syndrome. Rare longitudinal correlative EEG-neuropsychological studies have recently shown that acquired temporary cognitive-behavioral problems correlate with epileptic activity (EEG) in some children. The open question is now whether this epilepsy can cause a specific developmental learning or more general cognitive disability if the onset is severe, early and affects brain areas other than the strictly "rolandic". BPERS is a model for the study of the cognitive manifestations of focal epileptic discharges in a developing brain, although the prolonged fluctuating and cognitive manifestations and their dynamics of onset and recovery can not be explained in terms of simple ictal-postictal symptoms and suggest that several different mechanisms are probably involved. The practical clinical implications of these cognitive manifestations are also discussed.

Troubles cognitifs et du comportement dans les épilepsies partielles bénignes (BPERS) et variantes

Cet article résume les données récentes concernant les troubles cognitifs et comportementaux qui accompagnent souvent l'épilepsie partielle bénigne de l'enfant, et les arguments suggérant une origine épiléptique directe à ces troubles. Les premières descriptions du syndrome mentionnaient déjà des difficultés d'apprentissage et du comportement, mais cet aspect a été longtemps méconnu, l'accent étant mis sur la bénignité de l'affection et de son pronostic sur le plan épiléptique, sans étudier s'il pourrait s'agir là d'une manifestation "cognitive" de l'épilepsie. On a ensuite reconnu la parenté de ce syndrome avec l'aphasie acquise avec épilepsie (syndrome de Landau-Kleffner), qui fut le premier exemple d'épilepsie "cognitive" de l'enfant. Plus tard, des troubles oromoteurs prolongés réversibles ont été décrits pendant la phase active du syndrome chez des enfants qui avaient par ailleurs tous les signes typiques du syndrome, y compris l'évolution favorable. Ce syndrome pouvait donc entraîner des déficits neurologiques prolongés de nature "épileptique". L'impression clinique selon laquelle ces enfants ont une intelligence normale, mais avec, dans une proportion variable, des difficultés attentionnelles ou spécifiques (langage, visuo-spatiales, etc.) a été confirmée par plusieurs études neuropsychologiques avec sujets-contrôles. Ces résultats pouvaient expliquer certains problèmes d'apprentissage ou scolaires dans la phase aiguë du syndrome. Plus récemment, de rares études longitudinales ont montré la corrélation des troubles neuropsychologiques transitoires avec l'activité paroxystique à l'EEG. La question est ouverte de savoir si cette épilepsie peut causer des difficultés spécifiques d'apprentissage ou des problèmes cognitifs plus globaux, lorsqu'elle débute tôt, est sévère et touche d'autres régions cérébrales que l'aire strictement rolandique. L'épilepsie partielle bénigne de l'enfant est un modèle pour l'étude des manifestations cognitives générées par des décharges épileptiques focales dans un cerveau en plein développement. Cependant, les manifestations prolongées et fluctuantes, ainsi que leur dynamique d'installation et de récupération, ne s'expliquent pas simplement en termes de symptômes critiques ou post-critiques, et suggèrent que des mécanismes physiopathologiques propres à ces épilepsies entrent probablement en jeu. Enfin, l'attitude pratique à adopter en présence de manifestations cognitives ou comportementales est discutée.
Kognitive Störungen und Verhaltensstörungen bei Rolando-Epilepsien und anderen idiopathischen Partial-epilepsien des Kindesalters


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Introduction

BPERS is the most frequent type of idiopathic epilepsy in children. It is a genetically determined epilepsy syndrome. Focal cortical brain regions (mainly the lower perisylvian area) develop an abnormal local electrical activity with corresponding occasional clinical seizures. The bioelectrical disturbance may last for a brief or a more extended period of childhood, but normalizes at or before puberty. It manifests on the EEG as focal sharp waves (FSW) mainly in the centrotemporal ("rolandic") areas. Seizures are simple partial motor and sensory seizures involving the lower face and the pharyngeal region, the so-called "sylvian" seizures, and tend to occur during sleep (after falling asleep or before arousal in the morning) sometime with extension to the hemibody or with generalization. The seizures are usually rare, disappear before puberty and the children have no mental or neurological handicap. Rolandic epilepsy was long considered the prototype of a specific epileptic syndrome with sharp boundaries.

Over the years, it became apparent that some children with the typical syndrome had epileptic foci in other locations either at the time of initial diagnosis (as an additional focus or as the only focus) or later in the course. This could occur in the same or the contralateral hemisphere sometime with a changing preponderance of left or right sided discharges. Rarely, children develop atypical features such as frequent or difficult to treat seizures, other types of seizures (myoclonic-astatic) or acquired neurological or cognitive deficits. It was also increasingly realized that a significant proportion had learning and/or behavioral problems. Family studies showed that not all children with these EEG abnormalities had clinical seizures\[1-2\].

All these data have gradually brought the concept that BPERS is only the most typical and frequent manifestation of a spectrum of genetically determined partial epilepsies of childhood with focal sharp waves on the EEG (FSW). See table 1 and table 2.

Cognitive and behavioral problems in BPERS and variants (table 3)

In the initial descriptions of the syndrome, it was recognized that some children with rolandic epilepsy had behavioral, learning or school problems. This tended to be explained by the psychological consequences of the disease or side-effects of drugs\[1-4\]. An influential view on the cause of the associated cognitive and behavioral problems in children with BPERS was later put forward by Doose\[1-2\]: After extensive clinical and EEG studies of affected children and their siblings, he proposed that children with FSW had a hereditary impairment of brain maturation (HIBM) which was manifested either by seizures or intellectual impairment or both. However, most children with this syndrome, even those with many seizures and/or frequent EEG discharges do not show any intellectual decline, learning or behavioral problems. This would suggest that the hereditary impairment manifests itself either as epilepsy or as a developmental problem in a totally dissociated manner which would be surprising if both were manifestations of the same basic disorder. Doose did not think that the paroxysmal EEG abnormalities could by themselves interfere directly with cognitive function, a possibility which has proved difficult to accept and to demonstrate; however which appears now very likely if not evident and will be further discussed. Quantitative neuropsychological studies with normal controls are now being increasingly published and show mild but significant differences in either attentional abilities, language or visuospatial competences as well as school problems (despite normal intelligence) and behavior disturbances in a proportion of a quarter to half of the children\[5-7\]. However such cross-sectional data do not allow to tell if the deficits are due to the basic brain dysfunction responsible for the epilepsy or to other factors, for
Table 1: Prototypical features importance of syndrome and evolution of ideas.

<table>
<thead>
<tr>
<th>Prototypical features</th>
<th>Importance</th>
<th>Evolution of ideas</th>
</tr>
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<tbody>
<tr>
<td>School-age, seizures mainly in sleep</td>
<td>Most frequent form of non-symptomatic childhood epilepsy</td>
<td>Too narrowly defined electroclinical syndrome</td>
</tr>
<tr>
<td>Sylvian, hemispheric +/- generalization</td>
<td>Concept of epileptic syndromes</td>
<td>Variability in clinical expression, location of discharges and associated problems</td>
</tr>
<tr>
<td>EEG with centrotemporal spikes</td>
<td>Focal (multifocal) hyperexcitability during maturation without lesion</td>
<td>Rolandic epilepsy: only one manifestation of a spectrum (see table 2)</td>
</tr>
<tr>
<td>Neurologically and cognitively normal children</td>
<td>Model of cognitive effects of focal epilepsy/epileptic discharges</td>
<td>Minor cognitive, behavioral problems frequent in active phase of syndrome</td>
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<tr>
<td>Remission before adolescence</td>
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<tr>
<td>Genetically determined epilepsy</td>
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</tbody>
</table>

Table 2: The spectrum of idiopathic (genetic) partial childhood epilepsy with FSW.

<table>
<thead>
<tr>
<th>Clinical epilepsy syndrome</th>
<th>Major features</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign partial with rolandic spikes</td>
<td>“sylvian” seizures</td>
<td>Most frequent, benign, the prototype</td>
</tr>
<tr>
<td>Benign partial epilepsy with acquired neurologic “cognitive” disorders</td>
<td>Oromotor (anterior opercular syndrome), graphomotor, gait disturbances</td>
<td>Prolonged deficits related to upper or lower rolandic area</td>
</tr>
<tr>
<td>Acquired epileptic aphasia</td>
<td>Acquired auditory agnosia</td>
<td>May start or be followed by more typical benign rolandic epilepsy</td>
</tr>
<tr>
<td>Landau-Kleffner</td>
<td>Myodonic-astatic seizures in addition to partial seizures with FSW</td>
<td>Severe epilepsy phases with variable cognitive stagnation, deterioration, sometimes none</td>
</tr>
<tr>
<td>Atypical partial epilepsy of childhood (&quot;pseudo-Lennox&quot;)</td>
<td>Cognitive arrest-dementia, psychiatric disturbances</td>
<td>Link with above syndromes less clear. Can be symptomatic of various focal cortical lesions with thalamic involvement (?)</td>
</tr>
<tr>
<td>Partial epilepsy with CSWS **</td>
<td></td>
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</tr>
</tbody>
</table>

**Childhood epilepsy with CSWS (Continuous Spike-Waves during Slow Wave Sleep). Some cases of partial childhood epilepsies within the rolandic spectrum develop a marked increase of the epileptic activity on the EEG during sleep (the so-called continuous spike-waves on the EEG, CSWS) for instance in the Landau-Kleffner syndrome. In some cases, there is a catastrophic, rapid or insidious stagnation or regression of some cognitive functions and a behavioral deterioration [34, 35, 15, 36]. Convergent clinical data and new evidence drawn from electrophysiological studies and functional imaging [37] suggest that the cognitive and behavioral dysfunction is directly related to the particular role that the affected cortical area plays when the epileptic process becomes active. The mechanisms however are ill-understood [35]. The CSWS phenomenon is probably due to a disturbance of corticothalamic oscillatory mechanisms at work during slow sleep and which seem to play a role in consolidation of material acquired during waking [35]. One especially dramatic example of how partial epilepsy can lead to a progressive dementia and/or a massive behavioral regression in children is the “acquired epileptic frontal syndrome” [35]. Cases of partial epilepsy with CSWS are increasingly described in a variety of developmental and acquired focal cortical pathologies in children (also in association with thalamic lesions) [39].
example the effect of the epileptic activity on cognitive functions. In this respect, “benign partial epilepsy with rolandic spikes” (BPERS) is a model for the study of the cognitive manifestations of focal epileptic discharges in a developing brain. The syndrome is frequent, there is no obvious brain lesion, the focal EEG abnormalities are usually numerous, the epilepsy most often starts at a relatively advanced stage of brain maturation and the children are normal and easily testable. The study of this syndrome may also help us to understand what can happen with other focal epilepsies in which presence of a lesion and other confounding factors complicate the issue.

Table 3: Cognitive and behavioral disorders in BPERS. Historical perspective.

- Behavioral-school problems mentioned in initial descriptions of the syndrome.
- Rolandic epilepsy: benign epilepsy syndrome, normal children; other problems dismissed.
- Close relationship between BPERS and acquired epileptic aphasia (Landau-Kleffner syndrome) recognized.
- Prolonged reversible oromotor deficits in some children during active epilepsy phase reported in some children with otherwise typical BPERS.
- Neuropsychological studies confirmed normal intelligence but showed variable attentional or selective (language, others) deficits in a proportion of children as compared to normal controls.
- Longitudinal correlative EEG-neuropsychological studies with acquired temporary cognitive-behavioral problems correlate with epileptic activity (EEG) in some children.
- (Transient cognitive impairment during EEG discharges suspected, but not largely confirmed.)

Evidence for a direct link between epilepsy (or paroxysmal epileptic EEG abnormalities) and cognitive-behavioral disturbances in BPERS and variants

Acquired prolonged reversible “neurological” deficits in BPERS

Long-lasting, fluctuant but fully reversible oromotor deficits have been demonstrated in longitudinal studies of individual cases which can be related to a dysfunction of the perisylvian region. These deficits can be either mild such as drooling or oromotor apraxia [8, 9] or more severe with facial lingual and pharyngeal motor dysfunction resulting in an opercular syndrome [10, 11, 12]. Some of these children also have phonological impairments or word finding difficulties indicating that neural circuits specific to speech and language can also be involved [13]. The EEG shows abundant focal, often bilateral focal “rolandic” discharges with marked increase and generalization during sleep, sometime amounting to “Continuous Spike-Waves during Sleep” (CSWS). The clinical deficit and its recovery correlates with the onset and disappearance or decrease of the paroxysmal EEG. The dysfunction however in most cases does not seem to be a typical focal status epilepticus or repeated seizures with prolonged postictal states, and has been called paraictal [14]. It is suspected that a strong local inhibition around the epileptic focus is what determines the dysfunction, not clearly in relation to the clinical seizures [14].

These cases offer a very striking demonstration that the intense focal epileptic activity occurring in this epilepsy syndrome can interfere in a prolonged sometime
insidious way with a very specific aspect of a cortical function. In some cases the neurological deficit happens without recognized seizures so that the diagnosis of epilepsy is delayed.

Importantly, these children have clinical and EEG findings otherwise similar to the other cases of BPERS with a same final benign course of the epilepsy, and no evidence of an underlying focal pathology on MRI.

From such cases, one can suppose that location of the epileptic activity (focus or foci) and the specific role that this area plays at the time the epileptic disorder becomes active is what will determine the nature and severity of the deficit and the occurrence (or not) of cognitive or behavioral consequences. For instance if the main epileptic activity is situated in the superior temporal area (Heschel’s gyrus) or spreads to it, it will impact on the decoding of sounds causing an acquired auditory agnosia that is the Landau-Kleffner syndrome [15]. In this frame of thinking, one can expect a variety of possible cognitive/behavioral problems in BPERS and variants, knowing that there is a great variability in the age at onset and localization of these focal sharp waves (FSW) [16].

Example of selective “epileptic” deficit in BPERS. Acquired reversible graphomotor dysfunction

An example of very selective “epileptic” deficit in an otherwise typical BPERS is illustrated by the following privileged case in whom the graphomotor function was selectively involved [17] and its dynamics of recovery over a prolonged period could be studied quantitatively (figure 1). SC, an 11 year old boy with typical BPERS from the age of 7 years, was seen because of progressive deterioration of writing that could not be explained by a motor, praxic or sensory deficit. This could be documented by computerized analysis of the graphic act and compared to the results in normal age-matched children. His initial very low score improved spectacularly and very rapidly (within one to 2 weeks) in correlation with the reduction of the epileptic activity, probably due to withdrawal of carbamazepine, but very far from becoming normal. We thought that he had reached his best level, reasoning that his focal epilepsy which involved cortical areas concerned with fine motor control, had been active for a long time and had caused a permanent “weakness” of this particular function.

Surprisingly, follow-up during the next 2 years showed a continuous regular improvement until normalization of almost all writing parameters. We could thus document a prolonged recovery period. Interestingly, his epilepsy had started at the age of 7 years and by history, writing had become difficult from that time on. We concluded that the acute reversible

Figure 1. Acquired reversible isolated graphomotor deficit in BPERS. An example of selective “epileptic” developmental deficit (see text).
Acquired cognitive deficits during the active epilepsy phase of BPERS

Most children with BPERS and moderately active epileptic disease do not show easily measurable cognitive changes, even though they have clinically difficulties in school, attentional or behavior problems. A direct chronological relationship between the activity of the epileptic disease (measured by the intensity of epileptic discharges on the EEG and sometime also clinical seizures) is difficult to document and can only be done in longitudinal studies. A few prospective studies have suggested a relationship [21] between the general activity of the epileptic disease at successive periods and the quality of cognitive functioning. We have performed a neuropsychological and EEG follow-up study of children with typical BPERS from the time of diagnosis. Out of 22 children, 8 showed sectorial weaknesses in visuospatial or verbal short term memory or in attention which disappeared on follow-up concomitant with improvement or normalization of the EEG [18]. These transient difficulties were interpreted as a subclinical effect of epileptic EEG activity on cognitive functions which reflected also on the school results. We felt that our data had to be taken with caution, as the differences were minor and could have had other explanations.

Baglietto et al. [14] made a longitudinal controlled study of 9 children with BPERS. These children were probably a selected group of the more severe cases of this syndrome, because they had an intense EEG activity during sleep which could be quantified at different time periods of the disease and was coupled with cognitive evaluations. They performed a sleep EEG every 3 months until remission of the discharges and performed detailed neuropsychological testing at the peak of the disorder (T0) and at remission (T1) as determined by the disappearance or the marked decrease of the discharges during sleep, 6 to 24 months later. They showed a significantly lower IQ and other test results (although still in the normal range) at T0 in the patients as compared to the controls. There was also marked improvement at T1 in the patients who performed this time as well as the controls. These findings support the view that a cognitive dysfunction occurs during the time of active epilepsy (the marker here being the EEG activity during sleep), but this recovered in that study without evidence of a persistent problem.

BPERS as a cause of developmental learning disability?

If BPERS can be the cause of stagnation or regression of an acquired cognitive skill, could it also cause a specific learning disability if the onset of epilepsy coincides with the period when the skill in question is just starting to be acquired and affects the relevant neuronal networks (for instance reading)? To demonstrate it, such a situation must be followed prospectively with EEG correlation. One must show that epilepsy interferes with that skill directly and exclude that this is not due to predisposing or other general cognitive, psychological or medical factors (drugs), which all could prevent progress in that skill. This exceptional opportunity occurred in a case initially seen in our prospective study of 22 cases with BPERS [18: case 8] which was regularly followed after publication for a further 4 years with an isolated specific difficulty in learning to read. The decisive data came only after a long follow-up with stagnation, regression and rapid improvement in correlation with the paroxysmal EEG activity (Mayor, in preparation 2004).

Transient cognitive impairment (TCI) during EEG discharges

Studies looking for transient cognitive impairment during the focal EEG discharges in BPERS have been very rare, which is surprising given the importance of the problem and the availability of cases. After the initially often quoted study of Binnie [20] who found evidence of TCI in typical cases of BPERS, no clear confirmatory studies have been published. It is not clear whether this is due to the difficulty to demonstrate minor changes during these brief discharges or whether there are in fact no cognitive correlates of this particular type of EEG epileptic anomaly. In a detailed longitudinal case study, De Saint-Martin [12] described a child with BPERS who had a prolonged oromotor deficit and facial myoclonias. Long-lasting drooling, dysarthria and dysphagia occurred in the periods when interictal abnormalities were abundant and bilateral, but not during severe or repeated seizure episodes. The authors suggested that the deficits therefore did not represent ictal or postictal manifestations but powerful enhanced inhibition around these “interictal” spike foci.

In summary, it is still not completely clear and well understood whether subtle on-line brief epileptic dysfunction during the interictal EEG discharges do occur and can be an important cause of cognitive or behavioral dysfunction observed in this syndrome [21].

deterioration of his handwriting at 11 years was due to a sudden worsening of his epilepsy, but that the learning and automatization of this skill had been interfered with for a long time previously and needed a prolonged period to reach an almost normal function. Whatever the mechanisms for this, it has important implications for other dysfunctions of a more direct cognitive nature and which can occur in the course of these epilepsies.
Other frequent problems of uncertain but possible relationship to the epileptic activity

Behavioral problems in BPERS

Behavioral problems have been found in a proportion of children with BPERS, as compared to normal controls, mainly using questionnaires [17]. As in other epilepsies they could be explained as a reaction to the diagnosis with all its psychological and social implications or as possible side-effects of medication. However, when it occurs even before the diagnosis is made or in cases with only one or few nocturnal seizures, which are untreated and in which all attempts have been made to dedramatize the situation, one can suspect that they may have a physiological origin more directly related to the epileptic disease. Recently some cases of idiopathic partial epilepsy of frontal origin with many characteristics of BPERS and transient but severe behavior and attentional problems have been observed [23]. It may be that within the spectrum of BPERS there are cases with more anterior epileptic foci and “frontal” epileptic dysfunction, mainly or exclusively in the behavioral-attentional domain, but this needs to be further studied.

Attentional deficits in BPERS

The different abilities which one refers to under the global term of attention (alerting responses, sustained attention, divided attention, inhibition) are the most complex and also the most vulnerable of the human brain functions. It is thus not surprising that “inattention” is frequently found in children with epilepsy for many different reasons other than the epileptic activity itself. In an inattentive child with BPERS who has school problems, one has to distinguish the deficits which can be attributed to a localized cortical epileptic dysfunction in the perisylvian region and which implicate a specific instrumental capacity (i.e. graphomotor skill, verbal expression, language comprehension, reading) from those due to a more general disturbance in the domain of attention [23]. This could be due to a diffuse effect of the epileptic dysfunction or an involvement of the prefrontal regions. There is no clear demonstration that an isolated attention-hyperactivity disorder can be an acquired reversible manifestation of BPERS, although this can in theory occur, especially in the rare cases in which the epileptic focus is located or spreads to the prefrontal region [22]. The recent findings of a higher frequency of FSW in children with ADD-H (Attention Deficit Disorder with Hyperactivity) needs to be further explored [26]. At the present time, there is no evidence of a close link between ADD-H and BPERS.

Specific developmental disorders, mental retardation with co-occurrence of rolandic epilepsy or epileptic abnormalities (FSW) on the electroencephalogram

Children with simple developmental delay, developmental language disabilities or autistic spectrum disorders occasionally have seizures with clinical and EEG characteristics of BPERS. More often, epileptic EEG abnormalities including FSW are sometime found during the etiological work-up of such children who never had clinical epilepsy. This can be the either the association of two unrelated disorders or the FSW (which is not specific to BPERS) or the marker of an other disorder, typically fragile-X-syndrome or Rett syndrome in which very similar EEG characteristics can be found [25, 26]. The possibility of a direct link between FSW and the cognitive problems must be envisaged with caution. When a child is diagnosed as having rolandic epilepsy he may have paroxysmal EEG discharges for a long time and possibly also previous several unrecognized seizures (typically during sleep). The recognized active epilepsy phase is not necessarily that of the basic epileptic disorder which could have interfered with developing cognitive functions for a long time already.

A particularly early and prolonged epileptic activity in developing and « specializing » neuronal networks could induce a permanent structural dysfunction, that is an abnormal connectivity of the involved structures, particularly if it occurs at the time of active learning and synaptic stabilisation. Thus, a same child could have developmental retardation for these reasons but at a later age show a stagnation or even a regression in a given domain during a new phase of activity of his epilepsy as a more direct consequence of the ongoing epileptic disease. It is a challenge to demonstrate that this really does happen.

It is a priori difficult to envisage that some children within the BPERS spectrum could have mental delay or specific developmental problems when the huge majority of children have normal (and sometime superior) cognitive abilities and suffer no decline during the active epilepsy phase. It can be hypothesized that school-aged children with typical BPERS (i.e. with sylvian seizures) are those least at risk of cognitive dysfunction because the epileptic disorder is located in a zone with early maturation and no direct involvement in cognition. In others, possibly when the epileptic activity is more anteriorly situated in the frontal area or starts exceptionally early and is long-lasting it could have severe developmental consequences. So far, it remains unproven [18].
It should be stressed that the majority of children with BPERS will have no significant short and long term consequences of their epilepsy (figure 2), but one can not really predict at onset those who may have an atypical course of their epilepsy. In a very large study of children with BPERS, 26/378 children (7%) had atypical evolutions (other seizures: 11; Landau-Kleffner: 3; operculum syndrome: 7, mixed: 5) [27]. However, minor cognitive disturbances, as discussed in this review, would certainly pass unrecognized in a large transversal study since they are not the cause of a major scholastic handicap and since the clinical epilepsy is mild. One may even wonder why so much importance should be given to them. One reason is that a child whose cognitive competences fluctuate or are not in keeping with his potential or who develops behavioral problems during most important and prolonged periods of his childhood may suffer, as well as his family, more than one can imagine, especially when these are misunderstood.

In a newly diagnosed case who has no behavioral or cognitive problems, one would recommend a good documentation of his school and cognitive abilities, but there is no rational to start antiepileptic therapy (unless there are frequent seizures or other special reasons) in such a situation even if the EEG is very active. The children should be followed even if they have no new seizure episodes, so that possible acquired cognitive-behavioral problems might be evaluated and their possible relationship to the epilepsy considered [28]. When there are already school or behavior problems it may be very difficult to know if the epileptic activity is the cause of the problems and if it is justified to attempt to suppress the EEG discharges unless these were clearly acquired, or are markedly fluctuating. Unfortunately, most antiepileptic drugs, with the exception of sulthiame, do not suppress EEG discharges in BPERS and can sometimes markedly aggravate the situation. The significance and reality of these transient cognitive deficits in BPERS could be better asserted if it could be shown that effectively suppressing the EEG discharges with antiepileptic drugs clearly improves the cognitive function concerned. Paradoxically, more has been learned about the direct effect of epilepsy on cognitive function in BPERS by those cases who markedly aggravate their epilepsy with AEDS (especially Carbamazepine) and produce temporary cognitive

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**Figure 2.** Evolution of cognitive functions in BPERS. This figure shows the evolution of cognitive functions, learning abilities and school progress in children with BPERS and the possible role of epilepsy. Most children will overcome the active period without problems, although some will have fluctuations, relative stagnations or even regressions, even though they may remain statistically in the “normal” range. Age at onset of the epileptic disease, age at diagnosis (which may be much later) and location, extent, and activity of epilepsy (not shown on figure) will all influence the course.
EEG discharges but this is sometimes only transitory also been shown to have a clear suppressing effect on epilepsy in childhood. A study a great variety of cognitive and behavioral effects more severe variants, offer exceptional opportunities to fluctuating problems of epileptic origin, BPERS and its possible developmental effects or subtle prolonged and extreme examples.

Despite the great difficulties to evaluate the possible developmental effects or subtle prolonged and fluctuating problems of epileptic origin, BPERS and its more severe variants, offer exceptional opportunities to study a great variety of cognitive and behavioral effects of epilepsy in childhood.

References


34. Boed M, Casaer P. Continuous spikes and waves during slow wave sleep: a 30 months follow-up study of neuropsychological recovery and EEG findings. Neuropediatrics 1989; 20: 176-180


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