COGNITIVE PROFILES OF CSWS SYNDROME

Eliane Roulet Perez
CHUV, Neuropediatric Unit
Rue du Bugnon 46, 1011 Lausanne
Switzerland

1. FROM AN EEC PATTERN TO AN EPILEPTIC SYNDROME

1.1. The EEG Pattern of CSWS

Continuous spike-waves during sleep (CSWS), also described under the term of “electrical status epilepticus during sleep” (ESES),\(^5,14\) refers to a striking electroencephalographical (EEG) abnormality consisting of bilateral discharges of spike-waves (1.5–5 Hz) that almost completely replace the physiological non-REM slow-wave sleep activity (85% of the tracing is a recognized but still debated criterion). This EEG pattern can be associated with different seizure types (i.e., atypical absences, atonic episodes, nocturnal partial and generalized seizures) of variable severity. Although the underlying mechanism is still uncertain, CSWS are now considered to be a result of “secondary bilateral synchrony,” i.e., rapid secondary generalization from one or several cortical epileptic foci. Such foci can be found in the waking state or during periods of REM sleep when the amount of discharges tends to decrease, and they are often located in the frontal or central regions of the cerebral cortex.

1.2. The Syndrome of CSWS

As a syndrome, epilepsy with CSWS is defined as a combination of this particular EEG pattern with epilepsy and acquired neuropsychological disorders. The age of diagnosis lies usually between 2 and 10 years, but the precise onset of CSWS is often difficult to determine since a sleep EEG is not always recorded after a first seizure. CSWS can persist for months and years, but usually remit progressively during puberty. The etiology of the epilepsy is either a static focal (sometimes multifocal) brain lesion (sequelae of meningitis, prenatal-perinatal ischemic damage, cortical dysplasia) or unknown (cryptogenic/idiopathic).
A still unsolved question is whether the syndrome of CSWS and acquired epileptic aphasia (AEA; or Landau Kleffner syndrome), which also consists of an acquired neuropsychological disorder accompanied by an abnormal sleep EEG with nearly continuous generalized or bitemporal epileptic discharges, are really two different syndromes or should be merged into one global entity. Discussion of this problem is beyond the scope of this chapter. However, it should be kept in mind that these syndromes are classified on the basis of EEG findings in epilepsy with CSWS and of a clinical symptom in AEA, which is quite confusing. This separation would be valid only if CSWS were never found in AEA or aphasia would never be a manifestation of epilepsy with CSWS, which is definitely not the case. Since AEA is discussed in detail in the previous chapter, we shall focus here on data concerning children with CSWS who do not exhibit isolated or predominant language deterioration.

2. NEUROPSYCHOLOGICAL FINDINGS

As stated before, acquired neuropsychological impairments are a crucial finding. In fact, they are the hallmark of the CSWS syndrome. It should be pointed out, however, that there is no unique cognitive and behavioral profile associated with CSWS. Rather, there are different possible clinical pictures which will be discussed below. In most cases, deterioration is reported, but the spectrum of the severity of the disorder is quite variable. Dementia, aphasia, apraxia and psychotic behavior have all been reported, although they are rarely described in detail. Before onset of CSWS, there may be a history of normal or mildly delayed development.

2.1. Acquired Frontal Syndrome with CSWS

In a longitudinal study of four boys with CSWS, we attempted to better understand the nature and evolution of the behavioral and cognitive disorder in correlation with the epilepsy. In these children, a severe neuropsychological regression appeared between 3.5 and 8 years, which was associated with a frontal focus (on the right side in three, on the left in one). Previous development was unremarkable and no brain lesion was found. Clinical seizures were of mild to moderate severity. Deterioration occurred between one and two years after the first seizures and was quite rapid in two cases (after a few weeks) but insidious in the two others (after more than 6 months).

2.1.1. Behavioral Changes. Behavioral changes were the most disturbing symptoms that first alarmed the parents. Initially, attention deficit (distractibility and inability to focus attention), hyperactivity and impulsiveness were noticed. Aggressiveness, mood swings and disinhibition appeared later. Normal play was replaced by repetitive activities and mouthing of objects and clothing. The sense of danger was lost. In our cases, we did not observe complex ritualistic behaviors, fascinations or aberrant perceptions. Behavioral disorders were often so severe in the worst phase of the disease that a formal neuropsychological examination was impossible and assessment had to be limited to observation and administration of a few subtests of various batteries (McCarthy or Wechsler scales).

2.1.2. Subtle Cognitive Changes. In one of our patients, unexpected learning difficulties in mathematics and French with well-preserved graphomotor skills were noticed by the