West syndrome associated with Down syndrome: case report and literature review
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ABSTRACT
West syndrome is the most frequent cause of epilepsy in Down syndrome. This syndrome is often associated with poor long-term prognosis in most of children. We report a girl with West syndrome associated with Down syndrome revealed at 8 months of age for repetitive flexor spasms and electroencephalography (EEG) showed hypsarrhythmia. She had Down syndrome facies, microcephaly, psychomotor development delay and axial hypotonia. Computed tomography of the brain was normal. Her karyotype was 47, XX, + 21. Phenobarbital therapy was immediately effective with good clinical control of seizures, while the EEG monitored after one month was unchanged. At 2 years of age, the patient had hypertonic status epilepticus following a lung infection. The EEG showed a persistence of hypsarrhythmia. Sodium valproate and hydrocortisone therapy was effective with good seizure control but her psychomotor development was severely impaired. After a follow-up of 7 years, the patient presents growth retardation, microcephaly, severe psychomotor development delay, generalized hypotonia and tetraparesis. Knowledge of West syndrome in Down syndrome allows the early detection and prompt management of this neurological complication in order to optimize psychomotor development and improve the quality of life of these children.

Keywords: West syndrome ; Down syndrome ; hypsarrhythmia ; Prognosis