EPILEPSY IN CHILDREN WITH CEREBRAL PALSY

ABSTRACT:

Epilepsy is known to have a higher association with cerebral palsy. It has been observed that seizures in children with epilepsy and cerebraal palsy tend to have an earlier onset, necessitating the use of more than one antiepileptic drug with high risk of seizure relapse after antiepileptic therapy discontinuation The objective of the paper was to introduce with the relationship between cerebral palsy and epilepsy, and to determine the occurrence, associated factors, nature and prognosis of epilepsy in children with cerebral palsy.

Key words: children, cerebral palsy, epilepsy, antiepileptic therapy

Cerebral palsy (CP) is a group of non-progresive disorders caused by stationary developing brain damage of heterogeneous etiology. CP includes mostly disorder that affects muscle tone, movement, and motor skills. It can also lead to other health issues, including vision, hearing, speech, behavior and/or communication problems and learning disabilities, psychomotor retardation, and epilepsy (1,2,3,4).

Epilepsy and CP appear together in 15-60% of children. Dimitrios et all stady fom 1999. in patients deriven from a total population of 493 children with CP, described an overall prevalence of epilepsy in CP patients of 36,1% (5).

All types of seizures can be seen in patients with CP. Complex partial and secondary generalized tonic-clonic are perhaps the most frequent seizure types. Generalized tonic and tonic-clonic seizures, myoclonic seizures, and atonic seizures are also common. Epileptic syndromes, such as West syndrome and Lennox-Gastaut syndrome, are particularly frequent in children with CP. Typical absence seizures are observed less frequently in children with CP. Children with CP do have symptomatic, localisation related, epileptic seizures in 85,4% and idiopathic in only 14.6% (6).

Classification of the type of epilepsy is often difficult in children with CP for many reasons: firstly the partial onset prior to generalization may not be apparent or witnessed; impairment of consciousness during ictal period may be difficult to detect in a child with severe handicaps; lastly, the differentiation between myoclonic, brief tonic and atonic seizures could be difficult without ictal electroencephalography (EEG) or video EEG.

The seizure disorder is the consequence of the brain abnormalities associated with the CP, but genetic factors are also important in the development of epileptic seizures in these children (6,7). Among the perinatal factors, structural and developmental defects of the brain, chromosomal defects, intrauterine infections and hypoxic ischemic brain injuries are the more obvious causes that may result in seizures in children with CP. Brain imaging may provide a clue regarding the timing and nature of the brain insult in these children. In up to 94.6% of the children with CP significant radiological abnormalities are described, with 32% showing significant brain volume reduction without any other obvious pathology. Cortical, central or combined atrophy and an infarct picture are frequent demonstrated neuroimaging findings in children with epilepsy and CP.

It is difficult to explain why some children with CP do not have seizures in spite of significant radiological abnormalities. One possibility is that seizures may occasionally be missed in children with severe handicaps. If EEG recording is routinely performed on this group of children with CP with no seizures, presence of epileptic discharges may be evident indicating occult or missed seizures.

Whether seizures in early life produce more neuronal damage is not clear, but clinical studies indicate that early seizures are associated with more cognitive deficiencies. Severe seizures per se are responsible for progressive cognitive deterioration in children with CP (8).

The epileptic disorder might start at any age, but the first epileptic seizures typically are seen during infancy. Neonatal seizures preceded epilepsy more often in children with CP (19.7-42,9% %) than in children with epilepsy without CP (7.3-28,6%) (6). Gururaj found out 78.6% and Dimitros 71.3% children with CP developed seizures in the first year of life (5,6).

In persons with CP and MR, the diagnosis of epilepsy presents unique difficulties. The patients

generally are not able to describe the epileptic events, and the physician or someone trained in epilepsy observe the events only rarely. Patients with CP and MR frequently present with behaviors that resemble epilepsy. Generalized tonic extension crisis in children with severe spasticity, resembling generalized tonic seizures, is observed frequently in response to external stimuli. Gastroesophageal reflux might produce generalized tonic extension (Sandifer syndrome) in some cases. In children with severe quadriplegia, a similar clinical response can be seen as a consequence of chronic constipation or pain. Episodes of unresponsiveness frequently are seen in individuals with mental retardation. The patient's behaviors closely resemble absence seizures. Stereotyped movements (eg, nodding the head, odd hand postures, complex mannerisms, rocking, spinning, waving or flapping hands) are also a potential cause of misdiagnosis. Patients with mental retardation often take psychotropic medications, and some of the adverse effects, such as oculogyric crisis and dystonias, can be confused with epileptic events. Self-injurious behavior is common in children who already have an active epileptic disorder. These cases often are referred to the neurologist to rule out seizures of frontal or temporal origin. Nonepileptic seizures (pseudoseizures) should always be considered in any individual with drug-resistant epileptic behaviors.

Epilepsy is more common in certain types of CP and in turn, might be a reflection of the severity of damage to the brain. Walace report, spastic tetraplegia was the commonest type of CP complicated by seizures, and spastic diplegia the commonest type of CP in the control group of CP without seizures (9). Gurses et al. reported that 47% of children with periventricular leucomalacia (PVL) had epilepsy, 78% of which was intractable. PVL was mostly associated with spastic diplegia (10). There are only seldom observed seizures in children with extra pyramidal type and pure cerebellar type of CP. In the hemiplegic variety, partial seizures were more common representing a unilateral, focal lesion, such as infarct or a porencephaly (69- 73%) (11).

The overall outcome of seizures in children with CP is poor, needing prolonged course of anticonvulsant medications, polytherapy with higher incidence of refractory seizures and admissions for status epilepticus.

The ultimate prognosis depends on the etiology of the CP. Studies done in children with CP showed that the disorder evolves naturally. In some cases, seizures remit by the second decade of life. The causes of remission are not clear but might be associated with developmental changes in neurotransmitter systems. As a group, children with CP have a remission rate of 30% (10). However, epilepsy is not a lifelong condition in all patients. A total of 60% to 70% of patients will experience a 5-year remission on medication. In a seizure-free patient, the issue may arise about whether medication is still needed, given the adverse effects, cost, and inconvenience associated with antiepileptic drug therapy. In contrast, the decision to withdraw antiepileptic medication has implications for patient safety, driving privileges, employment, and liability. Indeed, the decision to stop treatment is in many ways more difficult than the decision to start it. There is no consensus on how long the duration of antiepileptic drug withdrawal after seizure control should bee (13). In many instances, discontinuing antiepileptic medications is possible after a seizure-free period. The time varies among physicians, but discontinuing medications after 2 years without seizures is usually considered safe. However, in the author's practice, medications may be safely discontinued after 4-5 years without seizures in persons with brain damage. Discontinue antiepileptic medications slowly. In children treated with multiple medications, the author recommends tapering one medication at a time, allowing approximately 6-12 months to discontinue each medication. Withdrawal seizures are not unusual and are not necessarily an indication that the medication being tapered is needed. However, in most instances delaying the tapering of the medications is safer. If a patient remains seizure-free throughout the withdrawal phase, the chances for success are encouraging but not ensured. If a patient does not relapse during the first year, the prognosis is more optimistic.

The role of EEG in antiepileptic drug withdrawal is controversial. Although an abnormal EEG before drug withdrawalwas a negative prognostic factor in many studies, the predictive value of EEG has not been confirmed universally (14). Patients with an abnormal EEG before drug withdrawal are twice more likely to relapse than were patients with a normal EEG. Activity on the EEG did not correlate with seizure relapse, although multifocal epileptiform discharges had a higher tendency to relapse (15). However, these results have not been replicated in other studies. This inherent limitation in the sensitivity of EEG affects its predictive value in selecting patients for antiepileptic drug withdrawal (16).

It has been suggested that neurologic deficit and mental retardation are poor prognostic factors for seizure relapse after AED discontinuation. AED treatment was discontinued in 65 children with CP and histories of epilepsy after 2 seizure free years. Patients with spastic hemiparesis had the highest relapse rate (61.5%), and those with spastic diplegia had the lowest rate (14.3%).) No other factor correlated significantly with the risk of seizure relapse (17) .

Discontinuation of AEDs in children with CP can, and should, be practiced when possible after patients have been seizure-free for at least 2 years. AED discontinuation in patients with spastic hemiparesis is significantly more likely to lead to seizure relapse than in patients with other CP types, but no other factor is yet known to increase the chance of relapse. However, when the analysis was limited to those patients who had either spastic diplegia or spastic hemiparesis forms of CP, a significant difference was found between the effects of those two forms on relapse time (P = .0485). The coefficient in the model medicated that those with spastic hemiparesis had a 6-fold greater chance of having relapses at any point in time than those with spastic diplegia (17).

It is common practice to wait at least 2 seizure-free years before AED discontinuation is attempted. An abnormal neurologic examination result has been proposed as a sign of poor prognosis. All of our patients had abnormal neurologic examination results, but nearly two thirds did not have seizure relapses. Therefore, the presence of a neurologic deficit, as well as CP, does not necessarily mean a poor prognosis after AEDs are discontinued.

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