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Epileptic Encephalopathies

There are many types of epilepsy syndromes based on the symptoms, age, causes of the seizures, and the severity of the episodes.

Developmental and Epileptic Encephalopathy (DEE) refers to severe epilepsy syndromes distinguished by seizures, often drug-resistant, and severe cognitive and behavioral disturbances. Encephalopathic patients often exhibit attention and behavioral problems, autistic-like behavior, loss of language, and psychiatric and sleep problems related to epileptic conditions.

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An epileptic encephalopathy may cause progressive academic, behavioral, and even motor problems. The epileptic activity itself contributes to intellectual and behavioral issues.

An epileptic encephalopathy frequently affects children but may occur in adults. DEE seizures are often lifelong, although, with specific syndromes, episodes can subside with time. A diagnosis of epileptic encephalopathies should include an EEG consisting of both the sleep and wake states. A magnetic resonance imaging (MRI) of the brain to look for structural flaws. A metabolic profile includes arterial blood gas, ammonia, tandem mass spectrometry, and lactate. Obtaining a urine analysis to check for inherited metabolic defects is often recommended.

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Treatments

In clinical practice, the ability to discern between epileptiform activity and an epileptic disorder may be challenging, as variability may be seen within each epilepsy syndrome and a given child over time.

Those with DEE typically have epilepsy that is often challenging to treat. Seizures are usually persistent and respond poorly to most medications.

The treatment goals are not only to control seizures but also to prevent or reverse the neurologic loss of function. Once a diagnosis is presented, an experienced specialists should promptly design a treatment plan. In many cases, hormonal and immune therapies are at the forefront of treatment.

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Treatments

More traditional antiepileptic drugs and surgery (when an identifiable lesion is present) play a limited but useful role.

Treatments are often targeted based on seizure type(s) or epilepsy syndrome. However, therapies can target a specific cause.

For example, some children with a structural cause can be good candidates for epilepsy surgery. Having surgery early helps to prevent further neurological regression. Some genetic disorders may respond very well to a specific medication but significantly worsen with another agent. Metabolic disorders may respond well to a particular supplement or diet. Increasingly focused “precision” treatments involving enzyme replacement or genetic therapies have proven successful.