Infantile Spasms are often a precursor of the LGS, although not formally classified as symptomatic generalized epilepsy. By definition, infantile spasms begin during the first year of life. They affect approximately 1 in 5000 children. The epileptic spasms manifest as flexor or extensor tonus, myoclonus, or a mixed pattern. The spasms last 1 to 20 seconds each and occur in clusters for up to 20 minutes. *West Syndrome* is the combination of epileptic spasms, hypsarrhythmia (a chaotic, disorganized epileptiform EEG pattern), and mental retardation. It is common for the term infantile spasms to be used synonymously with West syndrome. Infantile spasms have a poor prognosis with over 90% developing mental retardation and most progressing to symptomatic generalized epilepsy; a small percent of cryptogenic cases recover. There are many causes of infantile spasms including perinatal insults, cerebral malformations, CNS infections, tuberous sclerosis, and inborn errors of metabolism.

Other Seizure Conditions

Febrile seizures affect between 3% and 5% of children younger than the age of 6 years. About 30% of children have more than one episode; the likelihood of recurrence is greater if the first seizure occurs before 1 year of age or there is a family history of febrile seizures. There are a number of genes that, when mutated, predispose to febrile seizures. Although most affected children have no long-term consequences, febrile seizures increase the risk of developing epilepsy later in life. This risk is low for most children (2% to 3%), but is 10% to 15% in those who had prolonged or focal febrile seizures *(complicated febrile seizures)*, a family history of nonfebrile seizures, or neurological abnormalities before the first febrile seizure.

DIAGNOSIS

Accurate diagnosis is the cornerstone of epilepsy treatment. The diagnostic evaluation has three objectives: to determine that the events are epileptic seizures, to identify a specific underlying cause, and to establish if the seizures are symptomatic and isolated or if epilepsy is present and, if so, to determine the specific epilepsy syndrome.

History and Examination

The patient's and witnesses' descriptions of the events are central to diagnosis. Extra attention should be given to exploring details of the behavior during the seizure. The setting of the seizure can suggest acute causes such as drug withdrawal, central nervous system infection, trauma, or stroke. Recent-onset seizures in an adult suggest a new intracranial lesion. A remote history of seizures suggests epilepsy. Any focal feature before, during, or after the seizure suggests a structural brain lesion requiring appropriate investigation. The pattern of the seizures and the patient's age are often important clues to the seizure and epilepsy type.

The physical examination is normal in most patients with epilepsy. Examination should seek overt or subtle focal neurological signs: slight unilateral lower facial paresis, clumsy fine finger movements, or mild hyperreflexia. These can be present in focal epilepsy with a contralateral seizure focus. Careful skin examination is indicated to detect features of neurocutaneous syndromes such as a facial port-wine stain involving the upper eyelid in Sturge-Weber syndrome, hypopigmented macules (ash-leaf spots), shagreen patch, facial angiofibromas in tuberous sclerosis, and café-au-lait spots and axillary freckling in neurofibromatosis. Asymmetry in the size of the hands, feet, or face signifies a long-standing abnormality of the cerebral hemisphere contralateral to the smaller side. Absence seizures can be triggered in untreated children with hyperventilation for 2 to 3 minutes.

Laboratory Tests—EEG

Electroencephalography (EEG) is the most helpful diagnostic test for seizures and epilepsy. EEG findings help establish the diagnosis, classify the seizures correctly, identify the epilepsy syndrome, and make therapeutic decisions. In combination with suitable clinical findings, *epileptiform* EEG discharges, termed *spikes* or *sharp waves,* strongly support a diagnosis of epilepsy. In patients with recurrent seizures, focal epileptiform discharges are consistent with focal epilepsy, whereas generalized epileptiform activity usually indicates a generalized form of epilepsy. However, most EEGs are obtained between seizures, and interictal abnormalities alone cannot prove or disprove a diagnosis of epilepsy. Up to 50% of patients with epilepsy show epileptiform abnormalities on their initial EEG. The chance of capturing epileptiform activity is enhanced by sleep deprivation the night before the test so that the patient sleeps during a portion of the EEG recording. Serial EEGs increase the yield of positive tracings. A small proportion of patients with epilepsy have normal interictal EEGs despite all efforts to record an abnormality.

The interpretation of the interictal EEG is confounded by two factors. Epileptiform discharges occur in about 2% of normal people; many of these may be asymptomatic markers of a genetic trait, especially in children. Also, the interpretation of the EEG is subjective. Normal benign variant waveforms and artifacts can be misinterpreted as epileptiform activity and erroneously considered to be evidence of seizure susceptibility.

Epilepsy can be definitively established by recording a characteristic ictal discharge during a representative clinical attack. This is uncommon during routine EEG recordings, but can be accomplished with *inpatient video–EEG long-term monitoring* performed at many epilepsy centers throughout the world. Inpatient video-EEG monitoring is indicated in people who have ongoing seizures despite being treated with appropriate antiseizure medications. About one third of patients admitted for long-term monitoring are found to not have epilepsy; these patients have psychogenic nonepileptic attacks. In the 25 to 30% of people with focal epilepsy who continue to have disabling seizures despite trials with multiple AEDs, inpatient video-EEG monitoring to define the seizure focus is a critical test for determining candidacy for resective epilepsy surgery.

Laboratory Tests—Neuroimaging

Brain MRI complements EEG findings by identifying structural pathology that is causally related to the development of epilepsy. MRI is the best test to detect epileptogenic cerebral lesions including hippocampal sclerosis, neuronal migration disorders, tumors, focal atrophy, arteriovenous malformations, and cavernous malformations. It is important to obtain a complete imaging study that includes T1-weighted, T2-weighted, and inversion-recovery sequences in coronal and axial planes with and without contrast. Imaging in the coronal plane perpendicular