Complex partial status epilepticus manifests as a sustained state of confusion often associated with motor and autonomic automatisms. Some attacks produce bizarre behavior or stupor. Patients may resist assistance due to their abnormal state, which can last for hours or even days. The EEG usually shows nearly continuous discharging activity predominating in one or both temporal regions.

Absence status epilepticus (petit mal status) resembles complex partial status and consists of a confused state with some automatic behaviors. The EEG is characteristic with continuous runs of generalized 3 to 4 Hz spike and slow wave activity. The condition occurs in children or young adults with known absence epilepsy. Rarely, absence status occurs as the first manifestation of epilepsy in adults with no history of seizures. *Atypical absence status* with fluctuating confusion lasting for hours or longer occurs in patients with symptomatic generalized epilepsy (e.g., Lennox-Gastaut syndrome) and is accompanied by a generalized spike and slow wave pattern of 2.5 Hz or slower on the EEG.

Partial motor status, also known as *epilepsy partialis continua,* ranges from highly focal, clonic movements of the face or hand to jerks that involve most of the limb or half the body. The clonus frequency can vary from one every three seconds to three per second. It is relatively uncommon. Its causes include stroke, trauma, neoplasms, encephalitis, and hyperglycemia; sometimes the cause never becomes clear. Epilepsy partialis continua often resist all efforts at treatment and neurosurgical removal of the causative lesion is sometimes required.

Physicians infrequently witness seizures; most of the time they learn about the semiology during the history. Merely observing a patient in the midst of a seizure does not indicate that the patient should be treated for status epilepticus. However, once status epilepticus is diagnosed, treatment is urgent. The longer status epilepticus lasts, the more difficult it is to terminate and the more likely it is to cause brain damage. Aggressive therapy is mandatory for convulsive status epilepticus (Table 118-5). If initial therapy is not rapidly effective, anesthetic agents requiring intubation and ventilation should be used within an hour of onset. Complex partial status can also result in permanent neuronal injury and should similarly be treated aggressively, although therapeutic decisions are often made to try to stop complex partial status with agents that do not cause respiratory depression to avoid intubation. Absence status is unlikely to result in permanent sequelae and usually responds promptly to benzodiazepine treatment. Investigation of the cause of the status epilepticus should be undertaken during the treatment and continued after the seizures stop. Severe hyperglycemia can produce refractory partial motor and complex partial status; seizures stop once hyperglycemia is corrected.

Postanoxic status myoclonus is accompanied by generalized polyspike epileptiform discharges on the EEG but is usually unresponsive to treatment for status epilepticus. It typically indicates an irreversible condition with a poor prognosis.

TABLE 118-5 TREATMENT OF CONVULSIVE STATUS EPILEPTICUS

TIME (MIN)	STEPS
0-5 (ABCs)	Give O_2; ensure adequate ventilation Monitor: vital signs, ECG, oximetry Establish intravenous access; obtain blood samples for glucose level, complete blood cell count, electrolytes, Ca, Mg, toxins, and AED levels
6-9 (glucose) (benzodiazepine)	Give glucose if blood glucose level is low or unavailable. In adults give 100 mg thiamine as well. Intravenously administer 1-2 mg of lorazepam or midazolam or 5-10 mg of diazepam as initial therapy. Alternatively use rectal diazepam gel 0.2 mg/kg.
10-20	If the initial dose of benzodiazepine is not effective, then continue to intravenously administer either lorazepam 1-2 mg every 5 minutes up to a maximum of 0.1 mg/kg or diazepam 5-10 mg every 5 minutes up to a maximum of 30 mg in patients over 12 years old. If diazepam or midazolam is used and the status epilepticus stops, phenytoin (or another AED) should be administered promptly to prevent recurrence of seizures as diazepam and midazolam's duration of action against seizures can be less than 30 minutes.
21-40 (phenytoin)	If status epilepticus persists, administer 20 mg/kg of fosphenytoin* intravenously no faster than 3 mg/kg/min up to 150 mg/min in adults. (Alternatively use phenytoin at maximum 1mg/kg/min up to 50 mg/min in adults in a proximal IV.) Monitor carefully for hypotension, arrhythmia, local extravasation.
>40 (phenobarbital) (intubate)	If the seizures do not stop after fosphenytoin/phenytoin, give 20 mg/kg of phenobarbital intravenously at maximum 100 mg/min. When phenobarbital is given after a benzodiazepine, ventilatory assistance is usually required.
(general anesthesia)	If status epilepticus persists, give general anesthesia (e.g., propofol, midazolam, or lorazepam drip) to induce a burst suppression pattern (EEG monitoring, if available, should be instituted). Vasopressors or supplemental IV fluids are often necessary.
(alternatives)	Alternative/additional treatments include IV valproate (30 mg/kg) load, levetiracetam, and lacosamide.

ABCs, Airway, breathing, and circulation; AED, antiepileptic drug.
*Always dosed in phenytoin-equivalents (PE).

GENETIC COUNSELING AND PREGNANCY

Heredity

Patients with epilepsy should be advised about the hereditary risks to their offspring, although in most people with epilepsy it does not influence their decision about having children. The idiopathic epilepsies have complex inheritance with about 10% of children of an affected parent developing seizures. There are over 200 Mendelian-inherited syndromes with epilepsy, all rare.

Teratogenicity

Children of mothers taking antiseizure medications have a birth defect rate of 6% to 9%, which is two to three times that of the general population. Convulsions, however, pose a substantial risk to the mother and fetus. Thus, AEDs should not be stopped during pregnancy. Two AEDs, valproate and carbamazepine, have been incriminated in neural tube closure defects. Since the neural tube closes by 28 days of fetal development, this defect develops before the mother is aware she is pregnant. Phenytoin, phenobarbital, and primidone have been associated with a spectrum of neurodevelopmental abnormalities. All five of these older AEDs are classified as pregnancy category D by the FDA and should be avoided if possible. Large registries suggest that the newer AEDs have less teratogenicity, but the data are