

The NORSE (New-Onset Refractory Status Epilepticus) syndrome : Defining a new disease entity

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We describe a distinct clinical syndrome of new-onset refractory status epilepticus.

After noticing three similar cases with resistant status epilepticus, with no discernible cause who died within three months without abolition of seizures despite multiple medical interventions, a retrospective review of all status epilepticus seen 2000-2002 at our hospital was done. Four more cases were prospectively ascertained 2003-4. Status epilepticus was defined as persistent clinical generalised epileptic activity lasting more than 30 minutes, without restoration of consciousness.

In total we identified seven females, age 20 to 52, (mean 33), six Chinese, one Malay with no significant past medical history. Five had preceding fever. All required ventilation during prolonged stays. There were no localising signs and extensive work-ups were essentially negative apart from MRI changes seen in temporal lobes and leptomeninges. Initial EEG showed frontal ictal activity, subsequently developing into multifocal discharges. All received standardised maximal doses of intravenous antiepileptics, with subsequent intravenous anesthesia. Five out of seven patients died on intensive care. Two survived in a vegetative state, with frequent seizures. Neuropathology in two showed no inflammatory response and apart from diffuse patchy neuronal cell loss, with reactive gliosis, no other abnormalities were detected.

We describe a new clinical syndrome of primary new-onset refractory status epilepticus (abbreviated to NORSE). Unifying features include female gender, young age and catastrophic outcome. Unlike in other cases of resistant status epilepticus, extensive workup including neuropathology did not reveal any underlying cause. Increased recognition of this clinical entity will help delineate its underlying etiology.