

key points

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Epilepsy usually presents with seizures occurring during

both wakefulness and sleep. However, there are some epilepsies in which seizures appear mainly or exclusively during sleep. Nocturnal epilepsies account for 10-15% of all epilepsies, and 80% of nocturnal epilepsies in adults are focal. They present a diagnostic challenge as they can be difficult to differentiate from normal movements and behaviour during sleep and also from several non-epileptic, sleep-related, motor and behavioural disorders. Sleep-related hypermotor epilepsy (SHE) is often idiopathic, although there can be a positive family history in up to 40% of cases, and the genetic form shows an autosomal dominant inheritance pattern. Although the median age of onset is 14 years, this type of epilepsy can occur at any age.

More than 90% of the seizures in SHE occur during

sleep. Seizures occur during the N2 stage of NREM sleep so are particularly likely to occur within 30 minutes of falling asleep (even during daytime naps), and shortly before waking. Seizures in SHE are simple partial seizures which easily wake the patient so that they will usually be able to recall the seizures, often describing auras of somatic sensations or feeling unable to breathe.

SHE seizures have a rapid onset and offset, a short

duration (usually < 2 min), and a stereotyped motor pattern for that individual. Several semi-purposeful hyperkinetic movements may be seen, including bipedal or bimanual movements (running, cycling, or kicking), and axial movements (rocking, sitting or twisting). There can be prominent repetitive vocalisations that include unintelligible shouting or swearing. Tonic posturing may occur and includes head turning to the side of the seizure activity and fencing postures. Additionally, patients tend to experience brief fragments of epileptic events that look like periods of arousal without dramatic motor features throughout NREM sleep.

In parasomnias complex motor or behavioural events

occur during sleep in association with abnormal intrusions of one stage of sleep or wakefulness into another. Sleepwalking, confusional arousals, and sleep terrors are the most common NREM parasomnias. The most common REM parasomnia is REM sleep behaviour disorder, in which patients act out their dreams, normally in violent ways such as punching, kicking, or falling out of bed. It can be very difficult to differentiate between SHE and parasomnias but this can be facilitated by asking seven key questions including 'When do the events occur?' and 'How many events occur during one night?' SHE is characterised by a high frequency of attacks (3-8 events or more per night), and occurrence in clusters.

Many of the sleep-related motor and behavioural

disorders are diagnoses that can be made on clinical grounds, and often investigation results will be normal. Half of patients with SHE have a normal EEG both when awake and asleep, and as few as 14% of patients will have an abnormal MRI scan.