

## Non-convulsive status epilepticus

The recognition of convulsive (tonic-clonic) status epilepticus is clinically obvious and easily diagnosed. However, the recognition and diagnosis of non-convulsive status epilepticus (NCSE) is often more difficult - and not uncommonly because people (including doctors) either do not know that it can occur - or fail to consider it at the right time.

As with all epilepsies, the classification of non-convulsive status epilepticus is somewhat complicated but can be simply divided into the following four types:

- **complex partial status epilepticus (CPSE)**, also sometimes called psychomotor status
- **absence status**, also sometimes called 'spike and wave stupor'; it may be split into typical and atypical
- **hypsarrhythmia** which is the characteristic finding in children with infantile spasms or West syndrome. Many, including the author, but not all epilepsy experts regard this as a form of non-convulsive status
- **electrical status epilepticus of slow-wave sleep (ESESS)** also sometimes called continuous spike wave in slow sleep (CSWS). This is a rare but probably under-diagnosed type of non-convulsive status epilepticus.

The term 'electrical status epilepticus' is often used to mean the same as non-convulsive status epilepticus. This is simply to make the point that the EEG (the electroencephalogram), if recorded at the same time as the episode of non-convulsive status, will show continuous abnormal brain activity arising from either the whole brain (in which case it is called absence status epilepticus) or from just one part of the brain (when it is called complex partial status epilepticus). The term also emphasises the fact that the clinical, in contrast to the EEG, manifestations of non-convulsive status may not be that obvious and may even be very subtle. Children who develop complex partial status epilepticus may show one or more of the following features:

- a variation in conscious level, from drowsiness to confusion or even stupor; children may appear half-asleep, 'drugged' or even like a 'zombie'. Sometimes the children can appear to respond a little to what is being said to them but they almost never respond fully.
- a change or fluctuation in behaviour for no apparent reason; these fluctuations may occur over hours, days or even weeks; the child's parents and other carers are usually the first (and therefore most important) people to recognise this particular feature of CPSE.

- semi-purposeful movements and actions; these actions may be quite strange or bizarre but importantly are clearly not the child's usual actions, and must therefore be regarded suspiciously.
- a change in speech or even a loss of speech, again for no obvious reason. This is quite commonly accompanied by dribbling or drooling from one or both corners of the mouth.
- motor automatisms - involuntary and uncontrolled jerky or twitchy movements of the face, mouth or limbs. Sometimes these jerks or twitches are very subtle, may 'flit' or move from one limb to another and may last for many hours or days.

Because of these often subtle manifestations, complex partial status epilepticus is more difficult to diagnose in children who have pre-existing learning difficulties or behaviour problems. This also explains why non-convulsive status may not be diagnosed immediately in these children - a change in behaviour or speech may simply be blamed on the child's 'usual' behaviour or speech problems! Not uncommonly complex partial status epilepticus may also be misdiagnosed as drug intoxication (too high a dose of or too many anti-epileptic drugs), as an acute psychiatric illness (called a psychosis) or an infection/illness.

Whenever a child is thought to be in NCSE, an EEG must be obtained. The EEG will be able to either confirm or exclude the diagnosis of non-convulsive status epilepticus. Ideally an EEG should be recorded before the child receives any treatment for possible non-convulsive status. The EEG may also be used to monitor the response to treatment.

Non-convulsive status epilepticus is more likely to occur in children who have a severe epilepsy which is difficult to control although it may also occur in children who have never had seizures before. NCSE, and particularly CPSE, is also more likely to occur in children who have neurological problems other than epilepsy - including learning difficulties and cerebral palsy. The types of epilepsy where non-convulsive status, and particularly atypical absence and complex partial status epilepticus, is more likely to occur are the following:

- the Lennox-Gastaut syndrome.
- severe myoclonic epilepsy in infancy (sometimes referred to as Dravet syndrome)
- symptomatic generalised or partial epilepsies (symptomatic means that there is a cause for epilepsy - for example, tuberous sclerosis complex or following brain damage such as meningitis or encephalitis).

- the Landau-Kleffner syndrome (a rare type of epilepsy which is also called acquired epileptic aphasia and similar to a condition called electrical status epilepticus of slow wave sleep)
- children with a genetic or chromosome abnormality, particularly Rett syndrome, Angelman syndrome and one called Ring Chromosome 20 may also have frequent episodes of NCSE and these may either be atypical absence status or CPSE.

In addition, it is quite common for children with these severe and often difficult-to-treat epilepsies to have frequent and repeated episodes of non-convulsive status epilepticus.

Although NCSE is not usually a dangerous or life-threatening condition it may have an adverse effect on a child's memory and intellectual abilities, particularly if it is not recognised for many weeks or months. This is true of all types of NCSE, including electrical status epilepticus of slow-wave sleep.

The treatment of atypical absence and complex partial status epilepticus is similar to the treatment of convulsive status epilepticus. The drugs which are commonly used to treat NCSE include diazepam, midazolam, phenytoin or phenobarbitone. Diazepam can be given rectally (into the child's back passage) or intravenously (through a drip); phenytoin and phenobarbitone are given intravenously. The benzodiazepine group of drugs – diazepam, lorazepam (also called Ativan) or clobazam (Frisium) may also be

effective when given by mouth – either swallowed or placed under the child's tongue if swallowing is difficult. Another benzodiazepine drug, midazolam (Hypnovel and Epistatus), when given into the buccal or cheek cavity may also be very helpful. Buccal midazolam is now replacing rectal diazepam as the emergency treatment of seizures, including NCSE. This is because evidence has shown that it appears to be more effective than rectal diazepam. It is also less unpleasant and more acceptable for parents and nurses to give, and children and adults to receive this emergency medication. Very rarely, NCSE may be converted to convulsive status epilepticus after giving a benzodiazepine. Other drugs that may be helpful in treating hypersarrhythmia in West syndrome include vigabatrin, steroids (prednisolone or tetracosactide) and nitrazepam. The treatment of Landau-Kleffner syndrome and electrical status epilepticus of slow-wave sleep is often more difficult but drugs including sodium valproate (Epilim), prednisolone, clobazam (Frisium), ethosuximide (Zarontin) and sulthiame (Opsolot) may be effective.

Non-convulsive status epilepticus is frequently more difficult to treat than convulsive status epilepticus. One of the reasons for this is because NCSE is often diagnosed late and may have been occurring for some days or even weeks, unlike convulsive status epilepticus which is usually diagnosed and treated within 30-60 minutes. This again emphasises the point that NCSE needs to be thought about and diagnosed in children who are known to be at risk from developing this condition.

Dr Richard E. Appleton  
 Consultant Paediatric Neurologist at the Roald  
 Dahl EEG Department, Paediatric Neurosciences  
 Foundation, Alder Hey Children's Hospital,  
 Liverpool  
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 Tuberous Sclerosis Association can be obtained from our  
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