Non Convulsive Status Epilepticus In Children

Status epilepticus (SE) is a major clinical problem, often occurring in childhood, with a high potential for morbidity. The condition is subdivided into convulsive and nonconvulsive forms (CSE and NCSE, respectively). Non convulsive status epilepticus denotes electrographic seizures without convulsive activity and often manifests as altered mental status or coma. It may be difficult to diagnose in paediatric patients in whom changes of behaviour and consciousness may not be as easily recognised as in adults.

History
One of the first detailed accounts of nonconvulsive status epilepticus (or ‘automatisme ambulatoire’), was by Charcot in 1888, who described a patient who got into trouble with the law because he had boarded a train without a valid ticket (Charcot,1888).1 Lennox in 1949 described S, a boy aged 11 years who had occasional days of being confused, when he was able to eat but not to converse, i.e. periods of what he then termed ‘petit mal’ status which were successfully treated with Tridione.2

Clinical presentation
Children with non convulsive status epilepticus usually present with altered levels of consciousness, without convulsive activity it is often a difficult diagnosis to make as there are many other conditions which can cause altered mental status in childhood. It is important to rule out other causes of altered consciousness in children, some of which are life threatening, before considering NCSE.

Differential diagnosis of NCSE.
1. Head injury: Often a clear history and other signs of head injury.
2. Meningitis and encephalitis: There is often a history of high temperatures, irritability and headaches. The child is systemically unwell.
3. Raised intra-cranial pressure: There is often a history of fluctuating level of consciousness, headache, vomiting and focal neurological deficits.
4. Toxic causes: It is important to ask if there is a history of drug abuse or access to medication. Accidental overdose is more common in younger children or toddlers. Pupil size can give an indication of aetiology.
5. Metabolic causes: Inborn errors of metabolism may present with altered consciousness. For example, MCADD (medium chain acyl CoA dehydrogenase deficiency) can cause acute episodes of altered consciousness. These are triggered by prolonged fasting and hypoglycaemia is usually present.
6. Altered consciousness following syncope.
7. Prolonged post ictal confusion.
8. Specific epilepsy syndromes: for example, Panayiotopoulos syndrome.

A thorough history and examination is mandatory.

History of NCSE
1. Preceding symptoms: Most children with NCSE will have had preceding seizures in the acute setting, most of which are isolated brief convulsions rather than convulsive status epilepticus.3 Autonomic symptoms prior to the episode, particularly vomiting would suggest Panayiotopoulos syndrome.
2. Classify epilepsy: In a child who has previously been diagnosed with epilepsy it is important to take a full history and classify the epilepsy according to the ILAE classification.
3. Current anti-epileptic medication history: Erroneous classification of idiopathic generalised epilepsy and inappropriate treatment with narrow spectrum anti-epileptic drugs like carbamazepine can precipitate NCSE.4 It is also important to note whether the epilepsy was well controlled on medication and also verify compliance with medication.
4. Observation by carers: In a review of children presenting with NCSE, 32 out of 50 children presented with ‘change in behaviour’.5 This consisted essentially of a reduction in activity, slowness, and impairment of consciousness to a varying degree constituting a confusional state. Poor balance or incoordination usually associated with intermittent bilateral jerks of the limbs was reported as an additional prominent feature in eight of the group. Table 1 lists some descriptions by parents, teachers, or hospital staff of children during NCSE and on remission.

Table 1: Non-convulsive status epilepticus: examples of Psychological features as described by observers6

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Acknowledgements:
Thanks to Dr Anna Maw for help with the editing.

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References
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3. Prolonged post ictal confusion.
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Clinical examination specific to NCSE
The counting test: In a child with ‘absence status epilepticus’, it is worth asking the child to count from 1 to 100. If the child has absences during the counting process, you are able to see it, as the child will suddenly stop counting, stare or have automatisms and start counting again.¹

Investigation
1. EEG is diagnostic in non convulsive status epilepticus.
2. Consider intracranial imaging if you are concerned about an underlying serious cause like head injury or raised intracranial pressure.
3. Blood tests which may be useful to diagnose metabolic disorders include liver function tests, urea and electrolytes, ammonia and lactate and do not forget glucose.

Classification
NCSE may be classified as generalised non convulsive status epilepticus.

Generalised non convulsive status epilepticus.
Absence status epilepticus:

a. Typical absence status epilepticus: The main clinical feature of typical absence status epilepticus is altered state of consciousness, but changes in behaviour have also been reported. Children with typical absence status epilepticus may be able to eat and drink, withdraw from pain, walk about and respond to simple commands. The counting test (see above) may be useful. The duration may last minutes to days or weeks. An EEG during typical absence status epilepticus shows generalised spike wave discharges that occur at a frequency of 3Hz.

Only 3 out of 50 children in the series described by Stores et al. had typical absence status epilepticus. Typical absence status epilepticus occurs in children with idiopathic generalised epilepsies, particularly in children with absence epilepsy and juvenile myoclonic epilepsy. It may be triggered by inappropriate antiepileptic drugs like carbamazepine, fever, excitement or fatigue.

b. Atypical absence status epilepticus: On clinical grounds alone the distinction between typical and atypical absence status epilepticus may be difficult.

In the series by Stores et al., 31 out of 50 children were classified as having atypical absence status epilepticus, EEG revealed 4-7Hz spike wave activity with a variable proportion of spikes to waves from one child to another. 18 out of 31 were classified as having Lennox-Gestaut syndrome and 13 out of 31 were classified as having myoclonic atonic seizures.

Focal non convulsive status epilepticus.
Focal non convulsive status epilepticus may be classified on the basis of which lobe the seizures arise from though this may be difficult.

Focal non convulsive status epilepticus arising from the temporal lobe may present with confusion, strange behaviour, oral or manual automatisms. EEG may be variable with focal spike waves or spike wave or sharp wave discharges with inconsistent localisation.

Management
Treatment of NCSE will be guided by the precise syndromic diagnosis and the underlying cause. As most clinical forms of NCSE are not associated with systemic and chronic neurological complications, a less aggressive pharmacological management is suggested. However Stores et al reported convincing levels of intellectual or educational deterioration in 27 out of 50 children over the course of the seizure disorder of which NCSE had been a part. Detailed prospective study with neuropsychological follow up is required to clarify these issues. However in the meantime, it is important to recognise episodes of NCSE at an early stage and instigate prompt treatment.

Treatment approaches
1. Focal and generalised non convulsive status epilepticus may be treated with administration of buccal midazolam, based on the child’s weight and age. If seizures continue for more than 10 minutes the dose may be repeated or intravenous lorazepam may be given.³

2. Another approach to treatment may be to give a short course of oral benzodiazepines. This may be considered in children who present with recurrent NCSE, whose parents are able to recognise the signs and symptoms of NCSE and initiate management at home.³

3. Typical absence status epilepticus precipitated by inappropriate antiepileptic drugs will require discontinuation of the medication.

4. Adjustments to the child’s continuous medication may be undertaken. One example of this may be to increase the dosage according to the child’s weight, as children may have had a growth spurt. Compliance issues with the medication should also be discussed.³

5. Atypical absence status epilepticus may be difficult to diagnose in children as it can present with altered mental status and change in behaviour. It is important to rule out other serious differential diagnoses. EEG can be helpful in establishing a diagnosis. Management approaches are generally less aggressive than with convulsive status epilepticus. ◆

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REFERENCES
2. Lennox W. The petit mal epilepsies: their treatment with phenobarbitone. JAMA 1945;129:1069–73. [Abstract/FREE Full text]
5. Dr Abbie Reeve – Consultant Paediatrics Kings Lynn, Personal correspondence.
8. Personal practice.