

# Non-convulsive status epilepticus

D J MANNING AND L ROSENBLOOM

Child Development Centre, Alder Hey Children's Hospital, Liverpool

**SUMMARY** Status epilepticus can complicate any type of seizure activity. A group of 13 children with non-convulsive status has been studied. Five presented with chronic fluctuating neurological features, while eight had intermittent episodes of their atypical status, although each of these lasted for several days. The clinical features, treatment, and outcome for these groups of children are described. Most of the children in both groups are mentally retarded after regressing at the time of their status epilepticus. The recognition and aggressive treatment of atypical status is important in reducing the risk of subsequent mental handicap.

Status epilepticus is one of the common medical emergencies of childhood. The term is usually used to describe repeated tonic-clonic seizures with intervening loss of consciousness. In this form recognition is easy and without prompt and vigorous treatment considerable morbidity and mortality may ensue.<sup>1</sup>

Status epilepticus can complicate any type of seizure activity. The familiar 'convulsive' status is but one manifestation of a range of illness. Other more insidious presentations, which are by no means rare in childhood, have been recognised. This paper documents and gives case histories of less common forms of status epilepticus with a view to highlighting aspects of presentation, treatment, and prognosis.

## Patients and methods

The children studied were those admitted to the regional paediatric neurology unit at our hospital between 1971 and 1984 with status epilepticus other than tonic-clonic status. All children had repetitive or fluctuating neurological symptoms coinciding with continuous spikes or slow waves, or both, on the electroencephalogram (EEG). Seizures were classified according to the proposal for revised clinical and electroencephalographic classification of epileptic seizures.<sup>2</sup> In some children the neurological symptoms lasted from weeks to several months. These children had extensive metabolic, neurophysiological, and, where appropriate, neuroradiological investigations performed to exclude other possible causes of their symptoms.

Thirteen children (six boys and seven girls) were studied. Twelve children were known to have

epilepsy and one had never suffered classical seizures. The children were divided into two groups by mode of presentation, periodicity of symptoms, and difficulty of seizure control.

**Group 1.** Chronic, fluctuating neurological symptoms with electrical status (Table 1).

## Clinical features

These children presented insidiously with fluctuating symptoms and very prolonged episodes of status, lasting from one to 24 months. Two children presented with intermittent ataxia but minimal impairment of consciousness. One of these had never suffered classical seizures. Two further children had intermittent receptive and expressive dysphasia, in one case associated with a prolonged fugue state. The fifth child showed fluctuating unresponsiveness and developmental regression. In spite of relevant and extensive neurological investigations<sup>3</sup> no underlying causes for the children's symptoms were ascertained. Clinical relapses were always associated with electrical status.

Table 1 Characteristics and epilepsy type in the two groups: Group 1, children with chronic, fluctuating symptoms; Group 2, children with episodic, acute status

	Group 1 (n=5)	Group 2 (n=8)	
Male:female	1:4	4:4	
Age range (median)	10 months-10 years (14 months)	7 months-11 years (7 years)	
Seizure type			
Atypical absences:	3	Myoclonic:	5
Complex partial:	1	Complex partial:	2
No previous fits:	1	Unilateral:	1

*Treatment*

Table 2(a) gives the response to treatment with drugs. Because of the insidious symptoms no child was given intravenous anticonvulsants at presentation. In the three children given adrenocorticotrophic hormone EEG improvement accompanied clinical remission, but relapse occurred on withdrawal and repeated courses were required. Treatment with sodium valproate in one child with intermittent ataxia resulted in sustained clinical and EEG improvement. In a girl with complex partial status treatment with clonazepam was effective after carbamazepine, phenytoin, and phenobarbitone had been unsuccessful.

*Outcome*

Seizures were well controlled in all cases (<1 per month) and in two children anticonvulsants were eventually stopped without ill effect. The outlook for intellectual development, however, was not so favourable. Mental regression occurred in four of the five children during the period of their electrical status, and in each case deterioration halted but did not reverse when the tendency to status abated.

**Group 2.** Acute, recurrent episodes of status with poorly controlled epilepsy (Table 1).

*Clinical features*

Among these children episodes of status were characterised by increasing frequency of their usual seizure type, most commonly myoclonic jerks, atonic attacks, and automatisms. There was variable impairment of responsiveness, but in no case was there loss of consciousness. Drooling, ataxia, dis-

orientation, and aggressive behaviour were often noted. The duration of status ranged from three days to four weeks. In several children there was a history of deteriorating seizure control for weeks before presentation. In only one child was a potential precipitating factor (reduction in phenobarbitone dosage) identified. Again investigations failed to identify underlying diseases.

*Treatment*

Table 2(b) indicates the poor response to intravenous diazepam, particularly among children with myoclonic status. Altogether, diazepam was only effective in two cases. Other drugs, notably clonazepam and chlormethiazole, were usually required. Stopping of intravenous treatment resulted in relapse in four cases of myoclonic status.

*Outcome*

All of these children had poorly controlled epilepsy (>1 seizure per month). At the time of writing four children continued to suffer daily seizures, and three of these have suffered further episodes of myoclonic status. The outlook for intellectual development was poor. Regression in association with their status epilepticus occurred in four cases. Extensive investigation revealed no cause other than recurrent status for this regression. Four children had computed tomograms taken two months after the episodes of status. These yielded normal results in two, but the other two had evidence of cerebral atrophy. Again control of status did not result in intellectual improvement in any case but merely halted further deterioration.

Table 2 *Response to treatment with drugs in the two groups*

Case no	Seizure type	Treatment with drugs	
		Ineffective	Effective
<i>(a) Group 1, children with chronic, fluctuating status</i>			
1	No seizures		Sodium valproate
2	Complex partial	Carbamazepine	Clonazepam
3	Atypical absences	Sodium valproate; clonazepam	ACTH
4	Atypical absences		ACTH
5	Atypical absences		ACTH
<i>(b) Group 2, children with episodic, acute status</i>			
6	Myoclonic	Diazepam	Clonazepam
7	Myoclonic	Diazepam	Chlormethiazole
8	Myoclonic	Diazepam	Chlormethiazole
9	Myoclonic	Diazepam; chlormethiazole	Clobazam
10	Myoclonic		Diazepam
11	Complex partial	Diazepam; phenytoin	*
12	Complex partial	Clonazepam	Diazepam
13	Unilateral	Diazepam	Clonazepam

\*Spontaneous resolution.

ACTH=Adrenocorticotrophic hormone.

### **Illustrative case histories**

**Case 1.** A 3 year old girl had a six month history of fluctuating gait disturbance. She had suffered an episode of ataxia one year previously. There were no perinatal or developmental problems nor a history of seizures. On examination she was hypotonic with an ataxic, broadbased gait. Metabolic and virologic studies yielded negative results and a computed tomogram was normal. The EEG showed continuous, generalised, high amplitude spike and slow wave discharges. She was treated with sodium valproate. The ataxia improved during the next few months, coinciding with EEG improvement. Intellectual development was normal.

**Case 2.** This 10 year old girl had a one year history of deteriorating school performance and personality change. She had become progressively more withdrawn and had prolonged episodes of vacant staring. There was variable attention deficit and she constantly answered 'Pardon?' when asked questions. Writing ability deteriorated and there was expressive dysphasia with persistent word confusion. She had suffered tonic-clonic seizures since the age of 18 months, which were well controlled by sodium valproate. On examination she was in a fugue state but was cooperative. Psychometric testing showed poor auditory memory and verbal coding and a reading age of 8.1 years. The EEG showed continuous high amplitude spike and slow wave activity with left temporal preponderance. Carbamazepine was introduced, but the fugue state persisted virtually unabated for a year with persistent expressive dysphasia and EEG evidence of status. At 11 years her reading age was 7 years. After introduction of clonazepam there was a gradual improvement with resolution of the personality change and improved word finding. At 13 years the reading age was 11.4 years and speech was fluent. Full scale intelligence quotient (Wechsler intelligence scale for children) was 77, the EEG was normal, and clonazepam was successfully withdrawn.

**Case 8.** This boy presented at 4 years with a three day history of myoclonic and atonic attacks and intervening lethargy, ataxia, and aggressive behaviour. Developmental progress was normal until 3 years when he suffered an episode of tonic-clonic status epilepticus during an upper respiratory tract infection. He subsequently developed myoclonic-atic epilepsy, which was poorly controlled by anticonvulsants. On examination he was lethargic, ataxic, and dysarthric. Frequent myoclonic jerks and atonic attacks were observed. The EEG showed continuous, generalised, high amplitude, irregular

polyspike and slow wave discharges. Intravenous diazepam produced some EEG improvement but no clinical change. Infusion of chlormethiazole ended the episode but relapse occurred on withdrawal and intravenous clonazepam was subsequently effective. The total duration of status was four weeks. Two further episodes occurred and were terminated by clonazepam. At the time of writing he was continuing to have daily seizures while receiving a combination of phenytoin and nitrazepam and was mildly mentally retarded. A computed tomogram taken two months after the initial episode of status showed mild cortical atrophy.

### **Discussion**

In their usual forms myoclonic, absence, partial, and unilateral status share many similarities with tonic-clonic status. The epilepsy of affected children is typically resistant to treatment and deteriorating seizure control culminates in continuous seizures of the type usually suffered by the child. In contrast to tonic-clonic status impairment of consciousness is marginal and the ictal state is associated with dulled responses, automatisms, drooling, ataxia, and behaviour disturbance. Episodes are often prolonged and resistant to treatment with intravenous diazepam.

Of perhaps greater interest is a group of children who present more insidiously with chronic neurological symptoms, such as ataxia, dysphasia, and unresponsiveness. Symptoms, while fluctuating, are not clearly episodic and may persist with varying degrees of severity for years. Alterations in responsiveness are subtle and impairment of consciousness minimal. Most of these children have a past history of epilepsy (complex partial or atypical absences), but this has usually been well controlled and remission is common. It is not surprising, therefore, that the inherent 'epileptic' nature of the symptoms may not be initially recognised. Given the periodicity of the symptoms, the EEG evidence of status, the exclusion of other potential causes, and the response (albeit temporary) to anticonvulsants, however, there is no doubt that these children are in status epilepticus and should be treated accordingly. An index of suspicion is crucial as diagnosis (by EEG) is straightforward.

Brett first drew attention to ataxia as a symptom of minor epileptic status.<sup>4</sup> He considered that this 'pseudoataxia' was due to the intrusion of repeated myoclonic jerks. Ataxia may result from disruption of cortical connections as well as reflecting cerebellar or peripheral nerve disease. It may, therefore, be a symptom of diffuse cortical disease. The two children presented here with intermittent ataxia

showed several of the features outlined by Brett, notably prolonged fluctuations in symptoms, similar EEG changes, and in one case temporary improvement with adrenocorticotrophic hormone but ultimate dementia.

Dysphasia is a well documented manifestation of non-convulsive status, particularly complex partial status.<sup>5-8</sup> De Pasquet *et al* reported dysphasia lasting three weeks in a 17 year old boy with complex partial status.<sup>5</sup> Kellerman described recurrent aphasia and personality disorder in a 6 year old boy with sleep induced electrical status.<sup>7</sup> In both cases language difficulty persisted after status had resolved. Dysphasia in the two cases reported here was much more prolonged (lasting one and two years, respectively) and was less clearly episodic. Although there was gradual resolution in both cases, there were residual difficulties with language comprehension and mild mental retardation.

The most worrying feature of the children presented in this series is the poor prognosis for intellectual function. Ten children were ultimately retarded, whereas only four were before the episodes of status. Investigations failed to elucidate other possible causes of regression. The implication is that 'minor' epileptic status, no less than generalised tonic-clonic status, can cause brain damage. Several authors provide evidence in support of this. Brett reported that 10 of 22 children with minor epileptic status showed intellectual regression.<sup>4</sup> Doose and Volzke, in a review of 117 children with myoclonic-astatic epilepsy, found a close correlation between status and dementia.<sup>9</sup> Fifty two per cent of children who had suffered status were severely retarded compared with 24% of those who had not. Ohtahara *et al* reported that eight of 25 children with non-convulsive status regressed.<sup>10</sup>

There is no evidence that successful treatment can improve intellectual function. The aim is to prevent further deterioration, and treatment, therefore, should be just as prompt and vigorous as for convulsive status. Given the potentially prolonged nature of episodes and the poor response to diazepam, the use of other drugs, such as clonazepam or chlormethiazole, should be considered early in the course of presentation. In young children with chronic symptoms and electrical status adrenocorticotrophic hormone may be effective. As the effect is

often transient perhaps treatment should be continued for prolonged periods.

The nomenclature of status epilepticus has been confusing.<sup>10-12</sup> Terms such as 'non-convulsive' and 'minor motor' status have been used to embrace types of status other than generalised tonic-clonic status and may imply that these types are less severe. Gastaut has stated that there are 'as many types of status epilepticus as there are types of epileptic seizure'.<sup>13</sup> Acknowledgement of this fact facilitates earlier recognition of less common types of status, emphasises the potential for brain damage, and provides a rational basis for aggressive treatment.

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Correspondence to Dr D J Manning, Department of Physiology, London Hospital Medical College, Turner Street, London E1 2AD.

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