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Aetiology, course and outcome of children admitted to paediatric intensive care with convulsive status epilepticus: A retrospective 5-year review

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Summary A retrospective case note study of the aetiology and course of children in convulsive status epilepticus (CSE) admitted to a large paediatric intensive care unit (PICU) was undertaken between January 1999 and April 2004. Status epilepticus was defined as a prolonged (>30 min) tonic–clonic seizure irrespective of whether the seizure had stopped prior to admission to PICU. During this period, 137 (74 male) children aged 1 month to 15 years were admitted to PICU with 147 episodes of status epilepticus. Forty-seven of the 137 children (34%) were admitted following a prolonged febrile seizure. Thirty-eight of the 137 children (28%) had a remote symptomatic cause for the CSE, 24 (18%) were admitted for an acute symptomatic cause and 15 (11%) were admitted with an acute exacerbation of a pre-existing idiopathic/cryptogenic epilepsy. Six children had a progressive encephalopathy and no cause was identified in the remaining 7 of the 137 children (5%). Forty-nine (36%) of the 137 children had pre-existing epilepsy. The mean duration of CSE was 44 min. Forty-nine (36%) children admitted to PICU who had received a benzodiazepine with either phenobarbital or phenytoin, required further treatment to terminate the presenting episode of CSE. Forty-two of these 49 were treated with thiopentone anaesthesia and the remaining 7 with a continuous infusion of midazolam, successfully terminating status in all. No child died. Of the 70 children considered to be previously neurologically and developmentally normal prior to admission, only 1 child demonstrated a new gross neurological abnormality at the time of latest follow-up. Seven patients (5%) developed new or de novo epilepsy.

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Convulsive (tonic–clonic) status epilepticus (CSE) is currently defined as a generalised convulsion lasting 30 min or longer, or repeated tonic–clonic convulsions recurring over 30 min without recovery of consciousness between each convulsion.[1–4] It is the most common neurological emergency in childhood[5] and is life-threatening with risk of neurological sequelae.[2,4,6]

There is no precise figure of the incidence of CSE in children, which in part, relates to the definition of status and the population studied. Epidemiological studies have suggested that four to eight children per 1000 may be expected to experience an episode of CSE before the age of 15 years,[7] and in children with first seizures, 12% present with CSE as their first unprovoked seizure.[8] A recent systematic review of the epidemiology of status epilepticus suggested an incidence of 18–20/10,0000 children per year.[9] Approximately 10–25% of children with epilepsy will have at least 1 episode of CSE.[10] Mortality and morbidity due to CSE have declined during the last two decades. Studies in the 1970s suggested a mortality rate of 3–11%,[11,12] compared with zero in the last few years.[13,14] More recently, Chin et al. reported a mortality of 3% in 91 children admitted with confirmed status epilepticus; all deaths were in children with an acute symptomatic or progressive cause of CSE.[15] The overall reduced mortality may be explained by a change in the definition of CSE (1 h duration in the 1970s, in contrast to 30 min more recently) and more aggressive treatment of seizures occurring outside hospital and stricter hospital management of CSE with clear and time-defined protocols. Neurological sequelae of CSE (epilepsy, motor deficits, learning difficulties and behaviour problems) tend to be age-dependent, occurring in 6% of those over 3 years but in 29% of those under 1 year of age.[11] Important factors that influence neurological outcome are reported to include aetiology, duration of CSE and age.[6] Children under 1 year of age more commonly have an acute symptomatic aetiology, which may explain the relatively poor outcome in this age group. Although the outcome of CSE is primarily determined by its cause, the duration of CSE may also be critical, with longer durations being associated with a higher risk of morbidity and mortality.[16,17] Delayed treatment of an initial seizure may also increase the frequency of CSE.[6,18] Early seizure termination is therefore important and this is reflected in the recently published protocols for treating CSE in children.[19–22] The current approach is therefore to treat every child who is in a tonic–clonic convulsion for more than 5 min as if they were in 'established CSE', to try and prevent the development of persistent status. Early treatment may consequently contribute to a lower mortality and morbidity.

Children in CSE who are admitted to an intensive care unit (ICU) are likely to reflect the severe end of the spectrum; this may reflect the underlying cause or the treatment of the CSE, or both. Consequently, data from admissions to paediatric intensive care units (PICUs) for status epilepticus may clarify some of these issues and may facilitate the development of strategies to try and reduce the duration of CSE and its complications. There are few data on children with CSE admitted to PICU; two reports published over the last 11 years have reported mortality rates of 6% in 147[23] and zero in 186 children;[13] however, a recent study found the case fatality of CSE to be 3% (2–7%).[15]

The purpose of this retrospective and observational study was to explore the possible roles of aetiology, course and short-term neurological outcome of children with CSE admitted to an ICU. It was not within the remit of this review to assess the longer-term developmental, cognitive and behavioural outcome of this population.

Patients and methods

The Royal Liverpool Children's Hospital (Alder Hey) (RLCH) is one of the largest children's hospitals in the UK, serving both a large local population of Liverpool (300,000) and a tertiary population with a total population of approximately four million. The hospital has a 20-bed paediatric intensive care unit. The diagnosis and demographic characteristics of all patients admitted to Alder Hey PICU are entered into a computerised database that includes information on diagnosis, duration of admission, duration of ventilatory support, condition at discharge, survival and morbidity. The admission database and discharge summaries during the period 1st January 1999 to 1st April 2004 were searched to identify all children admitted with diagnoses of 'seizure', 'epilepsy', 'convulsions', 'febrile convulsions', 'status epilepticus', 'encephalopathy' and 'coma'. The final criterion for defining the population-base of this study was all children under the age of 16 years admitted to PICU at RLCH, between 1st January 1999 and 1st April 2004 with CSE lasting at least 30 min. All available data, including medical records and intensive care unit charts were reviewed by hand; this included all paramedic (ambulance) and accident and emergency (A + E) records. The review included age, sex, any pre-existing medical diagnosis, the most reliable estimate of the actual duration of CSE, including the time before entering the hospital, and also the aetiology of status, the treatment received both before and on admission to hospital, duration of admission on PICU, and neurological sequelae at