Epidemiology of Status Epilepticus

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1. INTRODUCTION

Although prolonged seizure states have been recognized since ancient times, the frequency with which status epilepticus (SE) occurs was not fully appreciated until the last decade. Even in the 21st century, SE continues to challenge clinicians and investigators. Despite recent advances in its diagnosis and treatment, and the advent of sophisticated intensive care units, SE is associated with a persistently high mortality rate. Inpatient medical costs relating to SE have been estimated at $4 billion annually in the US alone (1).

The study of SE presents some methodologic challenges. Patients with SE are not a homogenous population. While SE commonly occurs in those with an established diagnosis of epilepsy, it can also present as the initial manifestation of epilepsy. In addition, it frequently occurs de novo in the setting of other systemic and neurologic conditions that may influence its clinical course. The mortality associated with SE has varied according to the reporting site, with lower mortality at an epilepsy center, and higher rates at a university hospital (2). Therefore, the study of SE requires analysis of large populations in order to assess accurately its causes and outcomes. While several epidemiologic studies have looked at its incidence, a comprehensive study of the epidemiology of SE has been conducted in Richmond, Virginia, where SE cases have been identified prospectively for over a decade. This chapter reviews the literature regarding the epidemiology of SE, with emphasis on data from the Richmond SE database.

2. EARLY STUDIES OF SE

2.1. Frequency

Early studies attempting to assess the frequency and other characteristics of SE were hampered by the lack of a standard definition and classification. Most early studies focused on generalized tonic-clonic SE, because it is easily recognized clinically (3). In 1907 Turner reported that 5% of his 380 patients had SE (4). Lennox reported that 10% of 1271 patients he had seen before 1940 had had at least one
episode of SE (5). A number of retrospective chart reviews assessed SE as a proportion of hospital admissions (6). These calculations range from 0.01% of all admissions over a 20-yr period (7), to 0.13% of all casualty visits to a Helsinki university hospital over 1 yr (8), to 3.5% of all admissions to two neurologic intensive care units over an 8-yr period (9).

Not surprisingly, when epilepsy admissions, rather than general admissions, were considered, the rate of SE was higher, ranging from 1.3% (10) to 5.4% (8). Rates of SE among all epilepsy patients ranged from 2.3% (11) to 10% (5,12,13). Several studies have documented that rates of SE among children with epilepsy are higher than in adults, ranging from 13 to 24% (14–16).

Hauser estimated the incidence of SE in the general population based on a number of factors (16). By summing the following estimates—the number of patients with newly diagnosed epilepsy who present with SE, the number of patients with established epilepsy who develop SE, the annual incidence of febrile SE, and the incidence of SE relating to acute symptomatic seizures—he arrived at an estimate of 180 to 280 persons with convulsive SE per 1 million population per year (16). Shorvon augmented this tally by adding estimates of absence SE, complex partial SE, neonatal SE, nonconvulsive SE, and other SE syndromes, and calculated the estimated total annual incidence of all SE to be about 500 (441 to 646) cases per million in the general population (3).

2.2. Mortality

Early studies of SE mortality focused on convulsive SE, and were limited by problems with case ascertainment, SE definition, and selection bias (3). Series from the 19 and early 20 centuries came from specialized hospital settings, which likely skewed the results toward higher mortality rates. Mortality rates ranging from 10 to 50% were reported (4,10,17,18). SE was a significant cause of death in children with epilepsy and in institutionalized patients (3). Shorvon reviewed 12 case series published between 1970 and 1989 and found overall SE mortality rates ranging from 3 to 11% in children, and 14 to 59% in adults. Totaling the cases in the various studies, the pediatric mortality following SE was 7%, the adult mortality 28%, and the total mortality for adults and children 18% (3). The majority of deaths were attributed to the underlying cause of the SE.

3. DEFINITION

The heterogeneity of cases labeled as SE in the early studies emphasized the need to establish a standard definition of SE. This was the goal of the Working Group on Status Epilepticus convened by the Epilepsy Foundation of America in 1993 (19). The definition agreed upon is “more than 30 minutes of continuous seizure activity, or two or more sequential seizures without full recovery of consciousness between seizures.” This definition is used by all the recent epidemiologic studies listed in Table 1.

While this definition has been accepted generally, some argue that the definition of SE should incorporate a shorter duration of seizure activity. These arguments are