Benign partial epilepsy of childhood with centrotemporal spikes (BECT), (benign rolandic epilepsy) accounts for 6% to 16% of childhood epilepsy.1,2 Partial-onset seizures, typically nocturnal, begin between 3 and 13 years of age with remission by 16 years.3 The EEG shows a characteristic spike over the rolandic area.4 Not all children with BECT require antiepileptic drug (AED) treatment.5–9 We studied a population-based cohort with BECT comparing daily treatment with AED versus no medication.

Methods. We assembled an incidence cohort of children who fulfilled the International League Against Epilepsy criteria for BECT4 who had seizure onset from 1985 to 1995. Patients were selected by reviewing all EEG reports at the IWK Health Center for features suggestive of BECT, followed by confirmation of the diagnosis from physician and hospital records. Because we read all pediatric EEG for Nova Scotia, our cohort is likely population based.10 Families completed a 30-minute, semistructured telephone interview. The study was approved by the Research Ethics Board of the IWK Health Center.

Results. Demographics. Eighty-three patients with BECT were identified and 79 (95%) interviewed. There were 46 (58%) boys. Mean (±SD) age at first seizure was 89 ± 26 months (range, 33 to 148), and mean age at follow-up was 16 years (range, 8 to 26). Family arrangements were mostly traditional—69 (87%) caregivers were married. Forty-four families (56%) were rural and 35 (44%) urban. Social class was greater than class 3 in 26%, equal to class 3 in 23%, and lower than class 3 in 52% (Hollingshead scale).

Clinical history at diagnosis. A child neurologist assessed all 79 children, 39% after their first seizure and 22% after the second. Before diagnosis, 34 patients had 1 to 18 simple partial seizures and 34 had 1 to 10 secondarily generalized seizures. Ten children had both. In 50 children, seizures before diagnosis were nocturnal only, in 14, diurnal, and in 13, both. In two cases, the nocturnal/diurnal information was unclear. All children were neurologically normal and six (8%) required school resource help.

Choosing antiepileptic drug treatment versus nontreatment. Overall, 43 (54%) were treated with AED, and 36 (46%) were not. By the third seizure, 67% of the non-AED group and 75% of the AED group had received information about the benign nature of BECT (p = not significant). Before diagnosis, time from first to second seizure, number of seizures, nocturnal/diurnal occurrence, and types and average length of seizures did not differ in treated and untreated children. Rates of treatment were unrelated to social class or urban/rural habitation.

Patients diagnosed in 1991 to 1995 were treated less often than those diagnosed earlier (8/28 [29%] versus 35/51 [69%]; p = 0.0003). Girls were more likely to receive AED than boys (70% versus 43%; p = 0.03).

Treatment with AED started at diagnosis in 19 children and was delayed until further seizures in 24. Parents elect-
ing AED treatment were more likely to fear physical or brain damage from seizures (19/43 versus 8/36; \( p = 0.04 \)). Other reasons to choose medication were parental anxiety (58%), seizure severity or frequency (51%), fear of social consequences (42%), concerns about death during a seizure (33%), daytime seizures (32%), and activity restrictions (14%).

Parents in the non-AED group feared medication side effects (50%) and perceived their child’s seizures to be of low frequency or severity (64%). Both groups indicated that a major determinant for their decision was the physician’s advice—72% in the treated group versus 83% in the untreated group.

Most (72%) presented to an emergency department, more often in the treated group (84% versus 58%; \( p < 0.01 \)) and usually for the first seizure (49 of 57 emergency department visits). Overall, 58% of parents had a personal acquaintance with epilepsy, with no difference between groups.

Clinical course in treated versus not treated. For the AED group, the first drugs prescribed were carbamazepine (82%), phenobarbital (11%), and clobazam (7%). In 28%, the initial AED was changed, usually because of side effects. Treatment duration averaged 28.5 months (range, 1 to 71). AED treatment reduced the number of secondarily generalized tonic-clonic seizures compared with the untreated group (\( p = 0.0002 \)), although there was no effect on partial seizures (\( p = 0.2 \)). After starting AED, 19 (44%) were seizure free, and only 7 (16%) had additional generalized tonic-clonic seizures. Recurrent partial seizures occurred despite treatment in 22 (51%), with 7 children (19%) having more than 10. All recurrences in the AED group were short (<5 to 10 minutes).

In the non-AED group, only 4 (11%) were seizure free after diagnosis. Eighteen (50%) had at least one secondarily generalized seizure, 7 had more than six generalized seizures, and one had status epilepticus but a normal outcome. Twenty-three (64%) had additional simple partial seizures, with 8 (22%) having more than 10.

Overall, patients had more than 900 seizures with no significant injuries. One untreated child fell down stairs during a nocturnal simple partial seizure while running frightened to her parents’ bedroom.

At final follow-up, all AED-treated patients had stopped medications and were seizure free for an average of 7.4 years (range, 1 to 13; median, 8). For the non-AED patients, all were seizure free at follow-up for an average of 5.4 years (range, 1 to 13; median, 5). In the AED group, 20% of parents indicated that they would have selected no treatment if they had to choose again. None of the non-AED parents regretted their decision (\( p = 0.003 \)).

Social impact of treatment versus nontreatment strategy. Nearly 75% of parents reported extreme fear with the first seizure, and 35 (44%) feared the child might die. These fears were unrelated to the treatment decision. After diagnosis, 74% of the AED group and 83% of non-AED group indicated decreased anxiety about seizures.

There was no difference between groups for frequency of night monitoring or activity restrictions. Overall, 39 families provided night monitoring—an audio bedroom monitor, frequent night-time checks, or someone to sleep in the child’s room. Restrictions for 37% of the children included limiting sleepovers, swimming, climbing, sports, unsupervised time in the bathtub, or staying up late. Parents reported that 22 (28%) children were treated differently by relatives, peers, or teachers because of the epilepsy—the same in both groups.

Educational achievement did not differ between groups. For the 55 still in school, 86% had satisfactory school progress and 85% were thought to have at least an average number of close friendships. At final follow-up, 24 patients were older than 18 years (18 treated, 6 untreated). Fifty percent of both groups were married, none divorced. Only three were no longer in school and unemployed (two treated, one untreated). None abused alcohol and only one had a “drug” problem. Three had been convicted of a crime (one treated) and three others (two treated) were viewed as socially isolated.

Discussion. Our study represents the “real-life” experiences of children and families with BECT in a regional population. Many families in urban and rural settings from all social classes chose non-AED treatment. Other families chose AED, but they appeared more anxious about injuries from seizures and were more likely to have visited an emergency department. Physician recommendation seemed to play a pivotal role in the treatment decision. We have no explanation for why girls were more likely to be treated than boys. In the last 3 to 4 years of case ascertainment, the rate of AED treatment dropped from 70% to 30%, presumably as a result of physician advice. None of the non-AED families regretted their decision, compared with 20% in the AED treatment group.

Children treated with AED had fewer secondarily generalized seizures than those with no AED. This was not a randomized trial with checks for compliance; however, simple partial seizures appeared more resistant to treatment. If AED are prescribed for BECT, families should know that >50% will have additional seizures. Without AED, nearly 90% had further seizures, but significant injuries did not occur. Because of the very small risk of status epilepticus, home use of benzodiazepines (e.g., rectal diazepam) might occasionally be considered.

From a strictly epilepsy remission point of view, AED treatment was of no advantage because all patients achieved the expected remission. Treatment also did not improve educational achievement, school satisfaction, or social adjustment. Our sample of young adults at final follow-up was limited (18 treated, 6 not treated); however, there was no trend in social outcome favoring either treatment group.

Loiseau’s personal experience with 168 AED-treated patients followed to at least 20 years of age led to his statement “antiepileptic medication is advisable in about half the patients.” Lerman and Kivity reported their personal impression that the long-term social outcome is excellent regardless of treatment strategy provided that the family understands the benign nature of BECT. Nearly all their patients were treated with AED. Ambrosetto and Tassinari reported on 30 patients followed until at least 17 years of age. Ten were not treated but had
similar numbers of seizures as the 20 treated patients. All had remission. Recently, Fejerman offered a personal impression that only 30% required treatment.9 Our population-based study validates and quantifies each of these impressions.

References

Narcolepsy associated with other central nervous system disorders

The vast majority of cases of narcolepsy are idiopathic without recognizable brain pathology. However, cases purporting to be symptomatic narcolepsy caused by another disorder of the brain have been reported since the early 1900s.1 Described associations include neoplasms, localized most frequently to the diencephalon or brainstem,2 other diencephalic lesions,3 MS,4 and head injury.5 Problems with ascribing a causative relationship include imprecise definitions of narcolepsy6 and difficulty ruling out a coincidental occurrence of two unrelated disorders. Cases may be reported because the nature or location of the pathology may conform to preconceived concepts, whereas cases with other less accepted associations may remain undescribed.

To avoid the biases potentially present in such reports, we used a computerized record system to identify all patients with a diagnosis of narcolepsy as well as any other disorder of the CNS seen at our institution over a 13-year period. We determined the nature of the other disorder and the time relationship between its onset and the onset of narcolepsy. We hypothesized that lesions of the diencephalon or brainstem commencing in close temporal proximity to that of narcolepsy would be disproportionately represented in the sample.

Methods. The Mayo Clinic computerized record system codes the diagnoses at each visit of all patients seen. The coding system is based on the International Classification of Disease system. All patients seen at the Mayo Clinic (Rochester, MN) between January 1, 1975, and December 31, 1998, with both a diagnosis of narcolepsy as well as a diagnosis of any other CNS disorder were identified and their charts reviewed. Narcolepsy was defined as excessive daytime sleepiness and either a history of cataplexy, two or more sleep-onset REM (SOREM) periods on a multiple sleep latency test (MSLT), or both. Cataplexy was defined as transient bilateral weakness of the body, legs, neck, or face.

Article abstract—The authors identified patients with the coexistence of narcolepsy and another CNS disorder seen between 1975 and 1998 at their institution. Eighteen patients were identified, nine with narcolepsy commencing within 1 year before or after the other disorder. Seven patients (39%) had hypothalamic–pituitary syndromes. When they occur together, narcolepsy and other CNS disorders frequently emerge at about the same time, suggesting a causative relationship. Hypothalamic–pituitary pathology was the most common association.

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