

BENIGN EPILEPSY OF CHILDHOOD WITH CENTROTEMPORAL SPIKES

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SINGAPORE MED J 1994; Vol 35: 241-242

Epidemiology

Benign epilepsy of childhood with centrotemporal spikes (BECT) is the most common epileptic syndrome in school aged children. It represents 24% of epilepsies in children aged 5 to 11 years(1). Though less well-known than childhood absence (petit mal) epilepsy, it is in fact three to four times more frequent. Yet in one series of 448 children consecutively seen in a specialised private practice, BECT represented only 10.7% of the cases. In contrast, absence epilepsy represented 13.4% of the sample. These divergent results are a reflection of differences in study population(2). BECT is usually easily controlled, and patients are less likely to be referred for specialist care. This probably accounts for the relatively low rate observed in the study of Malaysian patients seen at the University Hospital(3).

BECT is probably as common amongst Asian children as in Caucasians. In a personal series of 439 children who developed afebrile seizures before the age of 10, 18% had BECT. Of the 190 children whose afebrile seizure started between 3 and 10 years of age, 34% had BECT. In the Malaysian study, published in this issue of the journal(3), BECT was seen in Chinese, Malays and Indians in approximately the same proportion as the general patient population of the University Hospital, suggesting that BECT occurs with similar frequency in all races and ethnic differences do not account for the low prevalence.

BECT may be underdiagnosed because the ictal manifestations eg gurgling noises, choking, drooling, twitching of the face, deviation of the mouth to one side and vomiting may not be recognised as seizures either by the parent or the doctor. As the seizures usually occur in sleep, the focal onset may be missed and the child is only noticed to be seizing when the seizure has become generalised. The diagnostic features on electroencephalography (EEG) may only be present during sleep, and this may not be routinely obtained in a busy EEG laboratory. Even if epileptiform discharges are seen on the EEG, BECT may not be diagnosed if a central and/or mid-temporal focus is insisted on as a diagnostic criteria as in the Malaysian study(3).

Most studies have reported a slight but extremely consistent predominance of males (male-female ratio 6:4), and the ratio of 12 boys and 9 girls in the Malaysian study is in accord with this(3).

Aetiology

The EEG trait in BECT is probably autosomal dominantly transmitted with age-dependent penetrance. The liability to seizures may be influenced by other genetic and environmental

factors, and focal cerebral pathology may interact synergistically with the hereditary factor and participate in the generation of focal sharp waves or seizures.

Clinical criteria

Based on the work of Dalla-Bernadina et al(4), the following clinical criteria for BECT have been proposed: 1) absence of clinical or neuroimaging evidence of brain damage; 2) family history of idiopathic epilepsy (not essential); 3) onset of seizures between 18 months and 13 years of age; 4) variable symptoms, but never generalised tonic or atonic seizures; 5) seizures ending during childhood or adolescence; 6) no long-term behavioural or cognitive impairment. EEG criteria include: 1) normal background activity; 2) normal sleep organisation; 3) characteristic interictal focal sharp waves, with variable location; 4) possible appearance of similar multifocal paroxysms; 5) possible brief bursts of generalised spike waves; 6) increased frequency of focal abnormalities during slow wave sleep, without morphologic changes and with a spike-wave index of less than 80%.

Seizures

In BECT, seizures typically occur while the patient is asleep or shortly after awakening. In 51-80% of cases, seizures occur only in sleep. Approximately 30% of patients have seizures in waking only and 5-25% in both waking and sleep(5). The findings in the Malaysian study are in accord with the world literature.

In most studies of BECT, 70-80% of the fits are partial seizures. They may be the only type of attack, or they may alternate with generalised fits, which occur in 20-25% of patients. The high percentage of generalised seizures observed in the Malaysian study may be due to the seizures being noticed only after it has become generalised or the initial focal manifestations were not recognised to be an ictal manifestation.

Partial seizures often involve preferentially one side of the face (37%), the oropharyngeal muscle (53%), and to a lesser extent the upper limb (20%). The lower limb is affected in only 8% of seizures. Facial seizures consist of a tonic contraction of one side of the face and/or clonic jerks of the cheek and eyelids. Oropharyngeal signs include guttural sounds, choking, retching, movement of the mouth, feeling of suffocation, profuse salivation and sensation inside or about the mouth described as "a dry throat" or "my tongue prickled". Inability to speak is common, although the children know what they want to say and may utter inarticulate sounds. When the arm is involved, it is mostly by clonic jerks, but a jacksonian march is rare. Strictly localised seizures are very brief (few seconds to a minute); they occur more often in children over 5 years of age. In younger children, the seizures tend to be less localised and not uncommonly involve a complete half of the body, generally without jacksonian spread. They are often longer than the more localised attacks, lasting several minutes or even half an hour. Long attacks are at times followed by a Todd's paralysis, although this is uncommon. Generalised seizures may be the only ictal manifestation of BECT. However, generalised fits are mainly nocturnal, and a focal onset may easily go unrecognised.

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EEG

Only 9% of patients with the typical epileptiform discharges of BECT on EEG actually have clinical seizures⁽⁵⁾. Hence if these discharges are incidentally observed in patients referred for EEG because of headaches, learning disabilities or other complaints unrelated to seizures, the patients should not be diagnosed or treated as having epilepsy.

The pathognomonic features of the epileptiform discharges on EEG are the typical morphology and field of the sharp waves, not their location. Younger patients tend to have sharp waves in the posterior temporal-occipital regions. With maturation, the sharp waves "migrate" into the temporocentral or vertex region and less frequently also into the frontal areas. In patients with benign focal epilepsies of childhood, sharp wave in the different locations have almost identical morphologies and are affected similarly by physiological parameters; with maturation, sharp waves may appear in different locations; not infrequently, sharp waves of identical morphology occur simultaneously in different locations.

The finding of EEG features typical of BECT in patients with petit mal reported in the Malaysian study is not unexpected. In 13% to 20% of patients with BECT, bursts of generalised spike-waves or polyspike-waves are observed⁽⁶⁾, and cases of typical petit mal absences with both generalised 3 Hz spike-waves as well as central spikes have been reported⁽⁷⁾.

Relationship of BECT with febrile seizures

There seems to be a close relationship between BECT and febrile seizures. There is a high frequency of epileptiform discharges typical of BECT on the EEG of children who have had febrile seizures⁽⁸⁾, and there is a relatively high frequency of febrile seizures in patients with BECT⁽⁹⁾. The 14.3% rate of febrile seizures in patients in the Malaysian study is not unexpected.

Treatment and prognosis

The seizures in BECT are usually easily controlled with carbamazepine, phenytoin, valproate or phenobarbital monotherapy. Approximately 20% of patients may have persistent seizures in spite of optimal treatment. However, regardless of the response to treatment and the severity of the seizure disorder, almost all patients will outgrow their seizures and intelligence is not affected. Only about 2% of patients experience seizures after

recovery from BECT. Very rare partial seizures or generalised tonic-clonic seizures, usually isolated events, have been reported to occur during adolescence and sometimes later. These do not constitute a relapse of BECT but may represent another form of idiopathic epilepsy for which BECT is a risk factor.

Failure to recognise this condition may lead to overtreatment resulting in adverse effects of medication. In addition, if the uniformly excellent outcome is not explained to the parents and the patient, emotional disturbances may result from anxiety, anguish, overprotection, or rejection.

Whether all patients with this condition should be treated is controversial. Twenty-five percent of children will have only a single seizure, and approximately 50% of the patients will have less than 5 seizures. In addition, seizures usually occur in sleep, and when the seizures occur during waking, they tend to be focal so they are unlikely to endanger the patient. Many parents in Singapore choose not to start anticonvulsants when the situation is fully explained to them. However, if the seizures recur frequently and are distressing to the patient or parents, monotherapy with either carbamazepine, valproate or phenytoin may be started and continued until 2 years from the time of the last seizure.

Though EEG is important in the diagnosis of the condition, there is no correlation between epileptiform discharges on the EEG and seizure frequency or severity. Normalisation of the EEG usually occurs later than clinical remission, the average interval being 2 years. Hence, EEG should not be used to guide decision regarding anticonvulsant therapy.

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