MALAYSIAN CHILDREN WITH: "BENIGN EPILEPSY OF CHILDHOOD WITH CENTROTEMPORAL SPIKES"

V Manonmani, C T Tan

ABSTRACT

Benign epilepsy of childhood with centrotemporal spikes (BECT) was studied in Malaysian children, and was observed in Chinese, Malay and Indian children in the ratio 10:6:5. There were 12 boys and 9 girls. Fit frequency varied from almost daily to a single fit. The age of onset ranged from 2 – 13 years and BECT was not noted in any child over 13 years old. There was a strong circadian rhythm and fits occurred mainly in sleep. Generalised seizures were more common than partial seizures. During the 3-year study from April 1989 to April 1992, 21 children with BECT were identified from the EEG records done at the University Hospital and it was found that this genetic epilepsy which is autosomal dominant with age dependent penetrance[1] occurs in approximately 4.8% of our epileptic children. In addition there were 3 children in whom petit mal co-existed with a BECT EEG trait.

Keywords: *benign partial epilepsy, electroencephalography, centrotemporal spikes*

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INTRODUCTION

Benign epilepsy of childhood with centrotemporal spikes (BECT) is the commonest type of partial motor epilepsy in childhood[2], but there have not been many reports of BECT outside the Western community. BECT is characterised by the onset of paresthesias/twitching of one side of the face, tongue or palate, which often produces a 'glugging' sound, speech arrest and drooling. The seizure may spread to involve one half of the body and then become generalised. The seizures usually occur during sleep. In the awake state, the child is conscious during the partial seizure, and can subsequently describe the onset of the seizure on recovery from the speech arrest. The incidence is 21/100,00 and it accounts for about 25% of childhood epilepsy. The age of onset is 2 – 13 years and recovery is the rule irrespective of treatment[1,3].

In our study, we studied the clinical features and occurrence of BECT in Malaysian children.

MATERIALS AND METHODS

The cases were identified from the EEG records done at the University Hospital from April 1989 to April 1992. EEG Criteria: High amplitude unifocal or bifocal slow spikes or sharp waves (occurring synchronously or side-shifting; occurring singly or in clusters; with a stereotype morphology of the sharp waves; sometimes followed by a slow wave and with activation by drowsiness and sleep) localised over C3, C4, T3, T4. The EEG background activity is normal[1,4]. The EEG records chosen were based on the above criteria and all our EEG's had wakeful and sleep tracing. Only children with a normal neurodevelopmental outcome are included. A total of 440 children with epilepsy, aged 1 – 15 years, had EEG's during the study period and 24 BECT EEG tracings were identified. Most of the EEG's were done for the first time in children recently diagnosed to have epilepsy. The medical records of the 24 children who met these criteria were then reviewed, and data were recorded from the available information, and from subsequent follow-up of these children. Description of the seizures (or seizures if the child had more than one type of seizure) was obtained from the child's/parents' recall of the event.

Department of Paediatrics
University Hospital
Lembah Pantai
59100 Kuala Lumpur
Malaysia

V Manonmani, MRCP (UK)
Lecturer

Department of Medicine
University Hospital

C T Tan, MD
Professor and Consultant Neurologist

Correspondence to: Prof C T Tan

RESULTS

A total of 21 children with the electroclinical syndrome of BECT were identified. In addition 3 children had petit mal co-existing with a BECT EEG trait. The findings are shown in Tables I, II and III.

We found that BECT represented approximately 4.8% (21/440 x 100) of the epileptic children, in Malaysia, and there were 10 Chinese, 6 Malay and 5 Indian children with BECT. (Malaysia is a multiracial community composed mainly of Malays, Chinese and Indians and the University Hospital patients consist of Chinese 40%, Malays 33% and Indians 26%). The age of onset ranged from 2 - 13 years. BECT was not noted in any child over 13 years old. A circadian rhythm was observed and fits occurred mainly in sleep. Fifteen children (71.4%) had seizures during sleep only, 3 children (14.3%) had seizures while awake only, and 3 children (14.3%) had seizures both in sleep and wakefulness. When seizures occurred in the wakeful state, the children were drowsy and about to go to sleep or had just awakened. There were 12 boys and 9 girls. The type of seizure observed was partial in 6 children (28.6%), generalised in 11 children (52.4%) and both partial and generalised in 4 children (19%).

A history of febrile fits was obtained in 3 children (14.3%). A family history of fits in siblings (mean sibling size was 3.5 siblings/family) or parents was obtained in 4 children (19%). Intelligence, behaviour and school performance was satisfactory in most patients as reported by parents, except Case 6.

Interestingly, petit mal co-existed with a BECT EEG trait in three boys[5], two of whom were a set of twins, whose mother had childhood epilepsy. One of the twins had a past history of febrile fits. These 3 boys who clinically did not manifest BECT seizures, but rather absence seizures, are excluded in the analysis as BECT is an electroclinical syndrome based on clinical manifestations as well as EEG findings. EEG photoconvulsive response was not noted in any of our patients.

Computerised tomogram (CT) scans of the brain done in 5 children were normal.

DISCUSSION

The EEG features of BECT was first described by Y Gastaut in 1952, and he noted that the Rolandic spikes had no focal

Table I – The subject characteristics

Case	Sex	Present Age (yr)	Seizures: Onset age (yr)	Seizures: During sleep	Seizures: Awake	EEG: Type P	EEG: Type G	EEG: RD Right	EEG: RD Left		Therapy
1	F	12	10	+	–		+		+		C
2	M	12	8	+	+	+	+	+	+		V
3	F	10	7.5	+	–		+	+	+		C
4	M	6	4.5	–	+	+		+	+		–
5	M	9.5	since small		Petit mal			+	+	GSWD	C
6	M	12	4	–	+	+		+	+		D/P
7	M	12.5	11	+	–	+		+			C
8	M	10	7		Petit mal				+	GSWD	V
9	M	10	7		Petit mal			+		GSWD	V
10	F	8.5	7.5	+	–		+	+			C
11	M	7	6.5	+	+	+	+	+			C
12	M	11	7	+	+	+	+	+	+		V/C
13	M	4.5	2	+	–		+		+	GSWD	D
14	F	10	9	+	–		+	+			C
15	F	9.5	8	+	–	+		+	+	GSWD	–
16	F	11.5	11	+	–		+	+	+		V
17	M	13	11	+	–		+	+			C
18	M	13	13	+	–		+	+	+		C
19	M	10	9	+	–		+	+			C
20	M	10	9	+	–	+	+	+	+		C
21	F	12	11	+	–	–	+	–	+		–
22	F	9	6	–	+	+	–	+	+		P
23	F	11	7	+	–	+	–	+	+		P
24	M	10	10	+	–	–	+	+	+		–

P: Partial seizure only
G: Generalised seizures only
C: Carbamazepine P: Phenobarbitone V: Sodium valproate
GSWD: Generalised spike & wave discharges

RD: Rolandic discharges
M: Male F: Female
D: Phenytoin

The three children with petit mal were excluded when analysing the data

significance. Nayrac and Beaussart in 1958 described the salient clinical features. Bancaud et al in 1958, confirmed the disappearance of such discharges before puberty generally[5].

In our study, BECT roughly accounted for 4.8% of the children with epilepsy which is rather low. This is most likely due to a lower genetic risk and the underrepresentation of a mild condition such as BECT, at our centre, which is a tertiary institution. Other contributing factors are that children with a single fit often do not have an EEG and may be missed, as will children with mild partial seizures which are not diagnosed. That genetic risks for seizures differ in different populations, has previously been shown by the high incidence of febrile seizures in the Japanese[6]. BECT was observed in all the 3 major races in the country.

It was not observed in anyone over 13 years old and this confirms its benign nature in our population and indicates a good prognosis for our children with BECT.

We found that generalised convulsions were the most commonly reported seizure type, and it always occurred in sleep. They were most likely secondarily generalised as the child's parents being awakened by the 'glugging' noises made at the onset of the seizure seldom saw the start of the seizure.

One child (Case 6) who had poor school performance had an 8-year duration of epilepsy from 4 years to 12 years old. He had been on prolonged treatment with phenytoin, and had also been treated with phenobarbitone earlier. He had gingival hyperplasia secondary to phenytoin. His school performance may be related to his medication.

Table II – Clinical features

Events reported during seizures of children with BECT	No. of patients (n = 21)
– Seizures during sleep	18
– Generalised convulsion	15
– Seizures woke the patient	14
– Hypersalivation	13
– Deviation of mouth	10
– Guttural sounds	9
– Speech impairment	7
– Fully conscious throughout the seizure	6
– Unilateral jerks of whole side	3
– Hemifacial jerks	3
– Paresthesias of the mouth	2
– Brachiofacial jerks	2
– Jerks of an arm	1

The three children with absence seizures were excluded.
Four children had both partial and secondary generalised fits on different occasions, and three children had seizures during sleep as well as when awake.

Treatment of seizures in BECT depends on several factors, including parental anxiety. Generally, infrequent noctural seizures require no treatment, whereas frequent fits or diurnal fits may be treated[5,7]. Overmedication and polypharmacy should be avoided. We tend to treat most of our patients, as parents would otherwise have sought treatment elsewhere, since multiple doctor consultations are very prevalent here. The parents were repeatedly informed of the good prognosis. Control of the seizures was

easily achieved, and all drugs seemed effective. However one child (Case 12) who developed obesity on sodium valproate was switched over to carbamazepine with possible better control of the fits.

Table III – Seizure frequency

No. of fits or seizure frequency	No. of children (n = 21)
1 fit	2
2-3 fits	7
1-2 episodes of fits occurring in a cluster	3
2-12 monthly fits	5
More frequently than 2 monthly	4

CT scans of the brain are not necessary in BECT and the 5 children with CT scans had no significant abnormalities.

In the 3 children with petit mal, who had complaints of staring spells and 3/sec spike and wave discharge, the centrotemporal spikes may just be an incidental finding[8].

A photoconvulsive response was not observed in any child, and so the inheritance of this trait is possibly on a different gene from that of BECT.

CONCLUSION

BECT is a genetic epileptic syndrome which occurs in all major races in Malaysia.

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