Benign partial epilepsy in infancy

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Abstract

The aim was to examine the occurrence of benign partial epilepsy in infancy (BPEI). BPEI was defined as epilepsies with complex partial seizures (CPS) or secondary generalised seizures (SGS), or both, compatible with the following characteristics: normal development before and after onset, no underlying disorders, normal interictal electroencephalograms (EEGs), and good response to treatment. All 75 patients who developed epilepsy within the first 2 years of age between 1987 and 1993 were evaluated: 22 patients fulfilled the definition completely; eight had CPS only, four SGS only, and 10 had both CPS and SGS; 17 had clusters of seizures. Eight patients had a positive family history. The average age of onset of seizures was 5.9 months. Interictal EEGs were all normal. Response to treatment was excellent and the average period of seizure persistence was 3.0 months. All had normal psychomotor development. Patients with BPEI were more common in this study than previously reported.

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It is generally accepted that most of the epilepsies which develop in infancy, especially partial epilepsies, are symptomatic of brain damage or metabolic disorder and their prognosis for development and seizures is poor.^{1–5} But infantile partial epilepsy with favourable outcome may be more common than suspected because previous studies on infantile epilepsies have been from highly specialised centres, with a selection bias towards more severe cases.

One of us, using ictal electroencephalograph and VTR recordings, reported partial epilepsies with good outcome in infancy.^{6,7} These are characterised by partial seizures or secondary generalised seizures, often occurring in clusters, a high incidence of benign seizures in the family, normal interictal electroencephalograms (EEGs), normal development before and after the onset, and good response to treatment.

The purpose of this study was to examine the occurrence of such benign partial epilepsies in infancy (BPEI) in a first line general hospital, using mainly clinical criteria.

Methods

We evaluated all 75 children referred to the department of paediatrics of Anjo Kosei Hospital, who developed epilepsy in the first

two years of life between 1987 and 1993. We included only patients with more than two afebrile seizures and excluded patients with occasional seizures due to acute insults to the central nervous system. Patients with seizures associated with mild diarrhoea were also excluded. There were some patients with neonatal seizures, but patients who had seizures only within the first four weeks of life were excluded.

We defined BPEI as epilepsies meeting the following criteria: (1) complex partial seizures (CPS) or secondary generalised seizures (SGS), or both; (2) normal development before and after onset; (3) no underlying disorders or neurological abnormalities; (4) normal interictal EEGs; (5) good response to treatment with antiepileptic agents.

Complex partial seizures were defined clinically on the basis of our previous study,⁶ as seizures characterised by motion arrest, decreased responsiveness, staring or blank eyes, or mild convulsive movements, such as eye deviation, head rotation, clonic movements, or increased limb tone. Oral automatisms were observed in some patients. A secondary generalised seizure was one presenting with CPS followed by a generalised tonic-clonic seizure. Duration of the seizures was mostly 1–5 min in both types, although it was not measured precisely.

Patients with only neonatal seizures and those with prolonged seizures lasting more than 30 min were excluded. A cluster of seizures was defined as more than two seizures within 24 h. The family history was regarded as positive only when a parent or a sibling had more than two afebrile seizures with favourable outcome.

Our 75 patients included 25 patients with West's syndrome. Thirty six patients had clear evidence of symptomatic aetiology.

Results

Twenty two (29%) of 75 patients completely fulfilled our definition of BPEI. There were 12 males and 10 females. Eight patients had CPS only, four SGS only, and 10 had both CPS and SGS.

Seventeen patients (77%) had clusters of seizures. Eight (36%) had a positive family history of benign infantile afebrile seizures. Six patients were sibling cases of three families and two of them were a pair of identical twins.⁸ Fathers of two patients had afebrile seizures with favourable outcome in infancy, but details were unclear. Nineteen patients developed epilepsy between 3 and 10 months of age, and average age of onset was 5.9 months (range 2 to 12 months). No patients had delay in psychomotor development before onset.

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Computed tomography or magnetic resonance imaging of the head, or both, was performed in 19 patients. None was abnormal. Interictal EEGs were normal in all patients. Ictal EEGs were recorded in three patients. The foci of paroxysmal discharge were occipital in all three. Paroxysmal discharges showed secondary generalisation in two patients but remained localised in one.

Nineteen patients were treated with antiepileptic drugs (11 with zonisamide, six with carbamazepine, one with phenobarbitone, and one with valproate), and spontaneous remission of seizures was observed in three patients without treatment. The serum concentrations of zonisamide and carbamazepine were 2.8–11.9 µg/ml (average 6.7 µg/ml) and 2.6–8.4 µg/ml (average 4.9 µg/ml), respectively; the concentrations of phenobarbitone and valproate were 7.5 µg/ml and 54 µg/ml, respectively. The average age at the last seizure was 8.9 months and the average period of seizure persistence was 3.0 months (ranged 0 to 12 months). Seizures disappeared within two months from the onset in 13 patients. Four patients experienced recurrence after treatment began, but seizure control was achieved after an increase in dosage or only transient administration of phenobarbitone. All patients showed normal development at the end of follow up, though the average age at last follow up was 54.3 months (range 17 to 99 months) and the follow up period was insufficient in some patients. Antiepileptic treatment was withdrawn or discontinued voluntarily after 1–1.5 years in 17 patients and none of them experienced recurrence of seizures.

Discussion

Many investigators have stated that epilepsies beginning in infancy have a poor prognosis and are frequently associated with neurological and intellectual impairment.^{1–5} Benign partial epilepsy is considered rare in infancy. Most of these reports were from specialised hospitals, and there may have been bias toward more severe cases. Recently, however, there have been several reports on partial epilepsies or seizures with favourable outcome that began in early infancy,^{6–10} and in some of these the authors considered that benign partial epilepsies were not rare in infancy.^{7,9} In our study, 29% of epilepsies beginning in the first two years completely fulfilled the definition of benign partial epilepsy. When West's syndrome was excluded, nearly half of the patients had BPEI. This indicated that benign partial epilepsy was not uncommon in early infancy among the general population. As Anjo Kosei Hospital has a neonatal intensive care unit and we routinely follow up the patients who are admitted there, our patients are more likely to be symptomatic and severe than those from the general population. BPEI must be more common than was seen in our study.

The clinical manifestations of patients with BPEI are very similar to those of patients with

benign infantile familial convulsions reported by Vigevano *et al.*¹⁰ In fact, eight of our patients who had family history of benign infantile seizures also fitted the definition of benign infantile familial convulsions. We consider that these two epileptic syndromes may overlap.

The diagnosis of complex partial seizures in infancy is not easy without ictal EEG recordings, because epileptiform discharges were not seen in the interictal EEG. The initial seizure manifestations of all our patients included impaired consciousness or decreased responsiveness, suggesting absence seizure or complex partial seizure. Absence seizures are extremely rare in infants. The duration of absence seizures is mostly less than one minute but complex partial seizures lasting more than one minute are common, especially in young children.¹¹ Cyanosis, which was common in our patients, is not observed during absence seizures but was usually seen during complex partial seizures. For these reasons we considered the seizures in our patients to be of the complex partial type.

Secondary generalised seizures, or even complex partial seizures, are also easily mistaken for primary generalised seizures. Yamamoto *et al.*,¹³ in a study using ictal EEG/VTR recordings, suggested that complex partial seizures in children may be confused with generalised tonic or tonic-clonic convulsions because of concomitant convulsive movements. It is not always easy to recognise the presence of partial seizures preceding secondary generalised seizures, because partial seizures are usually subtle and short. The patients with BPEI and secondary generalised seizures previously reported by us presented with apparently generalised tonic-clonic convulsions. Some had seizures at night, precluding the observation of initial partial seizure manifestation. Tsurui *et al.*¹² also reported that apparently generalised seizures in patients with benign infantile convulsions proved to be secondary generalised partial seizures. Most of the patients with BPEI with secondary generalised seizures are likely to be diagnosed as having generalised seizures unless an ictal EEG recording is done or unless the initial seizure manifestation was specially sought. Thus the incidence of BPEI with secondary generalised seizures may be even higher than our present study suggests. BPEI with complex partial seizures and secondary generalised seizure is the easiest syndrome to diagnose because the epileptic nature is suggested by the secondary generalised seizure, while the diagnosis of the epilepsy syndrome is suggested by the co-occurrence of complex partial seizures.

In conclusion, the present study has confirmed our previous impression that benign partial epilepsy is a common condition in infancy. We should not be pessimistic about the prognosis of children with features consistent with BPEI, although it is impossible to predict the outcome with certainty at the first presentation. Excessive and prolonged treatment should be avoided in such patients.

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