Benign Infantile Seizures

DEVENDRA MISHRA, NIRAJ KUMAR NIKUNJ, MONICA JUNEJA AND *BIBEK TALUKDAR

*From Departments of Pediatrics, Lok Nayak Hospital and *Chacha Nehru Bal Chikitsalaya, Maulana Azad Medical College, Delhi, India.*

Correspondence to: Dr Devendra Mishra, Department of Pediatrics, Maulana Azad Medical College and associated Lok Nayak Hospital, Delhi 110 002, India. drdmishra@gmail.com
Received: July 15, 2014; Initial review: August 21, 2014; Accepted: November 26, 2014.

Background: Benign infantile seizures are a common form of idiopathic seizures in infants, but infrequently reported. **Case characteristics**: Four cases identified over a 9-month period. **Observation**: All had a cluster of focal seizures, normal development and no abnormality on hematological and biochemical work-up. **Outcome**: No recurrence of seizures over a follow-up of 5 to 9 months. **Message**: Identification of this syndrome has important therapeutic and prognostic implications.

Keywords: *Epilepsy syndrome, Idiopathic, Infant.*

Benign infantile seizures (BIS) are characterized by onset of seizures in the first 2 years of life, with no known cause and excellent outcome [1]. Infants with similar features but with a positive family history for infantile epilepsy are labelled as Benign familial infantile seizures (BFIS), which has an autosomal dominant inheritance [3]. Caraballo, *et al.* [4] reported BIS as the third most common form of epilepsy in the first two years of life. However, not much information is available on this syndrome among Indian infants, although previously it has been pointed out that benign epilepsies are under-represented in Indian studies on epilepsy [5]. We herein report four children with BIS identified prospectively.

CASE REPORTS

These four children were identified during a 9-month period (April-December, 2012) wherein 75 infants with first seizure were prospectively studied. The various clinical characteristics of these four children are detailed in ***Table* I**. All had a cluster of focal seizures (secondary generalization in two), had normal development, normal neuroimaging and inter-ictal electroencephalography (EEG), and did not have recurrence of seizures. Ictal EEG was not possible.

DISCUSSION

In a well-infant presenting with the first afebrile seizure, the initial concern is the possibility of a metabolic cause or an idiopathic epilepsy syndrome. Other than West syndrome, BIS and BFIS are the commonest epileptic syndromes reported in this age [4]. Onset is mostly within the first year of life in both syndromes and seizures occur in clusters, have focal features including behavioral arrest, cyanosis, head/eye version, tonic stiffening of the limbs, and bilateral clonus [3]. The cluster can last 1 to 3 days. Interictal EEG is normal in the majority; ictal abnormalities have been described in a few patients. Initial genetic studies reported specific chromosomal-linkages, though later studies have suggested genetic heterogeneity. More recent data supports the hypothesis that this disease may be a channelopathy [2,6]. Another

TABLE I SALIENT FEATURES OF SEIZURES IN THE STUDY SUBJECTS

	Case 1	*Case 2*	*Case 3*[#]	*Case 4*
Age (mo/Sex	9/F	1.5/M	3/M	3/M
No. of seizures	3 in 48 hr	4 in 24 hr	3 in 48 hr	2 in 24 hr
Further seizures in hospital	No	Yes, 9	No	Yes, 3
CSF	Not done	Normal	Not done	Not done
DQ* (motor, mental)	104, 96	Normal[$]	79, 93	79, 88
Follow-up	7 mo	5.5 mo	9 mo	5 mo
Outcome	2 seizure more, no further seizures off-AED^	Discharged in 3 d on AED, tapered in 3 mo, No seizures	No seizures off-AED	No seizures off-AED

[*]Developmental quotient using DASII, 3mo after discharge; [#]Father and paternal uncle had history of seizures in infancy; [$]by non-formal assessment; ^Antiepileptic drugs discontinued by parents on their own.

subgroup with associated paroxysmal choreoathetosis has also been described [6]. As per ILAE classification [7], both BFIS and BIS are considered similar, except for the family history.

Generally, no further seizures are observed in cases treated pharmacologically. In untreated cases, there can be isolated or brief clusters within one year of age. Treatment with antiepileptic medication is not mandatory [8]. We started antiepileptic drugs in first two patients, but with increasing confidence about this diagnosis, we discharged the next two infants without drugs, and they did not have any seizure recurrence over the next 5 to 9 months.

The most characteristic feature of the syndrome is the occurrence of a cluster of few brief seizures, lasting for 1-3 days, with the child being well inter-ictally [9]. Recognition of this syndrome helps in avoiding long term anti-epileptic therapy.

Contributors: DM: diagnosed the cases, prepared the manuscript, and will be the guarantor; NKN: managed the patients, searched the literature, and helped in manuscript preparation. MJ, BT: Provided important intellectual inputs during patient management and manuscript preparation; BT: reported EEG findings. All authors approved the final manuscript.

Funding: None; *Competing interests*: None stated.

REFERENCES

1. Watanabe K, Okumura A. Benign partial epilepsies in infancy. Brain Dev. 2000; 22:296-300.
2. Specchio N, Vigevano F. The spectrum of benign infantile seizures. Epilepsy Res. 2006;70 Suppl 1:S156-67.
3. Vigevano F, Fusco L, Di Capua, Ricci S, Sebastianelli R, Lucchini P. Benign infantile familial convulsions. Eur J Pediatr. 1992;151:608-12.
4. Caraballo RH, Cersósimo RO, Espeche A, Fejerman N. Benign familial and non-familial infantile seizures: A study of 64 patients. Epileptic Disord. 2003;5:45-9.
5. Udani V. Pediatric epilepsy – An Indian perspective. Indian J Pediatr. 2005;72:309-13.
6. Vigevano F. Benign familial infantile seizures. Brain Dev. 2005; 27:172-7.
7. Engel J, Jr. Report of the ILAE Classification Core Group. Epilepsia. 2006; 47:1558-68.
8. Caraballo RH, Cersósimo RO, Amartino H, Szepetowski P, Fejerman N. Benign familial infantile seizures: Further delineation of the syndrome. J Child Neurol. 2002;17: 696-9.

Recurrent Kawasaki Disease

PRAMILA VERMA, NEETI AGARWAL AND MAHESH MAHESHWARI

From Department of Pediatrics, People's College of Medical Sciences & RC, Bhanpur, Bhopal, India.

Correspondence to: Dr Pramila Verma, Associate Professor
Department of Pediatrics, Sr MIG-B-10, Peoples campus, Bhanpur, Bhopal India.
drpramilav@yahoo.com
Received: July 14, 2014;
Initial review: August 21, 2014;
Accepted: December 06, 2014.

Background: Recurrent Kawasaki disease is rare. **Case characteristics**: An eight-month old infant had classic Kawasaki disease with transient coronary artery dilatation. **Observations:** Recurrence of incomplete Kawasaki disease after two years of initial diagnosis. **Outcome:** The index episode of Kawasaki disease was resistant to single infusion of immunoglobulin, while repeat episode responded within 24 hours of institution of therapy. **Message:** Early recognition of recurrent Kawasaki disease requires a high index of suspicion.

Keywords: *Incomplete Kawasaki disease, Outcome, Recurrence, Resistant.*

Kawasaki disease (KD) is an acute, self-limiting, medium-size vessel vasculitis of unknown etiology that predominantly involves the skin, mucous membranes, lymph nodes and coronary arteries. Standard therapy of KD is with a single intravenous infusion of immunoglobulins (IVIG) and high-dose aspirin until the acute phase reactants normalize. IVIG-resistant KD, which occurs in approximately 15% of children, can be defined as the persistence of fever beyond 36 hours of the initial IVIG infusion, and mandates a 2nd or even 3rd dose of IVIG [1-3]. Recurrent KD is mostly reported in Japan and the US, occurring in 3-4% and 0.8% of cases, respectively [4], but is only rarely reported from India [5].

CASE REPORT

An 8-month-old boy was referred to us with cough and cold for 15 days, fever for 5 days, and loose motions for 2 days. On examination, he was irritable and febrile. He had tachypnea and tachycardia with normal blood pressure.