Benign Childhood Epilepsy

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Updated: May 17, 2011

Benign Epilepsy of Childhood With Centrotemporal Spikes

A benign partial epilepsy of childhood, this condition, benign epilepsy of childhood with centrotemporal spikes (BECCT), is defined within the International League Against Epilepsy (ILAE) classification scheme as an idiopathic age- and localization-related epileptic syndrome with a combination of clinical and EEG characteristics used for diagnosis.

This epileptic syndrome is characterized by brief, simple partial and hemifacial motor seizures with associated somatosensory symptoms, which have a tendency to evolve into generalized tonic-clonic seizures. EEG shows high-voltage centrotemporal spikes often followed by a slow wave. BECCT is also known as lingual epilepsy, sylvian seizures, benign centrotemporal epilepsy, and benign rolandic epilepsy.

If the patient has the typical clinical history and EEG findings and has normal findings on neurologic examination, further workup is not indicated. However, in the presence of atypical features or abnormal examination findings, the use of magnetic resonance imaging (MRI) is indicated.

Benign rolandic epilepsy has been reported to occur in the presence of CNS pathology. However, in most of these reported instances, the BECCT was probably coincidental.

Epidemiology

BECCT is the most common epilepsy syndrome in childhood. In Connecticut, USA, it represents 9.6% of all epilepsies in children aged 0-15 years. However, BECCT and its variants may represent 20-25% of epilepsy cases diagnosed in patients aged 5-15 years.

Studies from other countries show that BECCT accounts for 6.5-16% of all childhood epilepsy. In a study from India, however, it represented only 1.6% of epilepsies in children younger than age 16 years. In a study from Italy, epilepsies with rolandic foci accounted for up to 23.9% of epilepsies in children aged 4-15 years.

Reported incidence of seizures with central temporal spikes ranges from 10.7-21 per 100,000. Age of onset ranges from 2-13 years but usually is between 4 and 11 years, and frequency of onset peaks at 5-9 years. The disorder occurs more commonly in boys; the boy-to-girl ratio is 6:4. A study by Kramer et al found no gender difference in incidence.[27]

Clinical manifestations of seizures

The syndrome is termed rolandic epilepsy because of the characteristic features of partial seizures involving the

region around the lower portion of the central gyrus of Rolando.

Common characteristic features include the following:

- Unilateral somatosensory involvement, mostly of the tongue (occasionally of the inner cheeks, lips, gums, or even a single tooth)
- Speech arrest
- Preservation of consciousness in most cases
- Pooling of saliva
- Tonic or tonic-clonic spread to face

Less often, sensory spread to the face or arms occurs; very rarely, a typical jacksonian march occurs. Other features include (1) absence of psychic symptoms, (2) rarity of complex automatisms, and (3) absence of amnesia and postictal confusion states.

BECCT is associated with the following 3 types of nocturnal seizures:

- Typical brief hemifacial seizures associated with speech arrest, drooling, and preservation of consciousness (identical to diurnal seizures)
- Seizures similar to those described above but with gurgling/grunting noises, loss of consciousness, and, at times, vomiting at the termination of the seizure
- Generalized convulsions (often secondarily generalized)

Although the somatosensory aura is common, it often is missed, because the child is young and the symptoms usually occur at night.

The expression of seizures appears to be age dependent. In older children, pure hemifacial seizures are more common, whereas in younger children hemispheric and generalized convulsions are more frequent.

Seizures can occur during the day or night, although in most children, seizures occur most often during sleep. Seizures occur only during sleep in 51-80% of cases, during both sleep and wakefulness in 5-40% of cases, and only during wakefulness in 0-32% of cases.

Frequency of seizures is usually low. Approximately 10-13% of patients have only a single seizure during the entire course, regardless of AED therapy; 66% have infrequent seizures; and 20% have frequent seizures (sometimes multiple seizures per day).

Occurrence of seizures in clusters is common. Duration of seizures is usually quite brief, ranging from 3-60 minutes; diurnal seizures tend to be shorter, especially the sensory ones. However, associated status epilepticus may be resistant to standard AEDs. One case manifested as an anterior operculum syndrome. Status epilepticus may occur in as many as 11% of patients. Postictal paralysis may occur in 7-21%.

Other clinical features

Headache and migraine occur commonly in patients with BECCT. In one series, recurrent headache was present in 67% of patients with BECCT upon presentation, and up to 80% had migraine reported on follow-up care.

A case-controlled study, however, found no significant difference in migraine incidence between cases and controls.

Febrile seizure history is not uncommon in BECCT.

Neuropsychological assessment

Children with BECCT usually show normal development and intelligence and have normal neurologic examination findings. Considering its prevalence, BECCT may be present in developmentally or neurologically abnormal children. The presence of developmental abnormality does not rule out the diagnosis of BECCT, nor does it necessarily worsen the prognosis. Behavioral and learning problems do occur.

Children with BECCT may have problems with visuomotor skills, visuospatial memory and skills, language, and attention; neuropsychological abnormalities are usually transient.

A systematic review on attention impairment in rolandic epilepsy, in which Kavros et al evaluated 14 studies, found

that the weight of evidence, defined as the majority of studies evaluated, indicated that attention systems are impaired in children with active centrotemporal spikes. The impairments resolve upon EEG remission.[28]

A study by Connolly et al found that the quality of life in children with BECCT may be compromised.[29] The compromise, which affects domains such as competence and psychosocial function, may be related to a cognitive variable and the emotional impact of the child's epilepsy on the parent.

No evidence exists to suggest that BECCT causes neurologic or behavioral abnormalities.

Genetics

BECCT is considered to be of genetic origin. Some patients have significant family history of epilepsy or centrotemporal spikes, although the exact frequencies vary, with a range of 9-59%.

Isolated EEG abnormalities (including rolandic spikes) are common in families of patients with BECCT. One study reported that at least 1 close relative had a temporal spike or SW discharge in up to 30% of the families. In another study, 15% of siblings of probands had seizures and rolandic discharges, whereas 19% had rolandic discharges alone.

Centrotemporal sharp and spike activity on EEG has been proposed to be an autosomal dominant trait with age-dependent penetrance. Only 12% of affected individuals have even had a seizure. Penetrance is low during the first 5 years of life, approximates 50% between ages 5 and 15 years, and then drops off to a low value after age 20 years. Whether a given child with the EEG trait develops epilepsy depends on a variety of inherited factors. Therefore, BECCT is inherited multifactorially rather than in an autosomal dominant fashion.

Some individuals with benign neonatal seizures have later developed BECCT. Linkage studies failed to establish a relationship between BECCT and benign familial neonatal convulsions. Two loci previously thought to be linked with BECCT, the human leukocyte antigen (HLA) region on arm 6p and the fragile X site, have been excluded.

In a study, Neubauer et al found evidence for linkage of BECCT to a region on band 15q14.[30]

In 1995, a new autosomal dominant syndrome was characterized by rolandic epilepsy, oral and speech dyspraxia, and cognitive dysfunction, with electroclinical features that resembled BECCT. Clinical anticipation was found in the family described in the study, suggesting that the genetic mechanism could be an expansion of an unstable triplet repeat.

Etiology

BECCT arises from the lower portion of the central gyrus of Rolando. Because BECCT is age dependent, has a strong genetic predisposition and an excellent prognosis, and occurs in structurally normal brains, it most likely represents hereditary brain maturation.

Many children with the EEG trait never develop seizures. Whether a child develops seizures depends on many factors, which may be hereditary. There may be an inhibitory factor that is capable of preventing seizures but can be broken through by external or internal elements.

Electroencephalography

EEG findings in BECCT are distinctive. The typical interictal EEG shows centrotemporal spikes or SW, which are either unifocal or bifocal. The spikes are usually slow, high voltage, and diphasic. Typical findings include a negative SW with a blunted peak preceded by a small positive wave and followed by a prominent positive wave with amplitude frequently up to 50% of the preceding negative SW.

When the SWs are unilateral, they are always synchronous in the central and midtemporal areas, although sometimes of different amplitudes. When bilaterally asynchronous, the spikes vary in frequency and amplitude from side to side. They can occur singly or in clusters. In about 40% of patients, the spikes are bilateral on initial or subsequent EEG records.

Sleep and drowsiness activate the spikes. Centrotemporal spikes are present only during sleep in as many as 30% of patients. Obtain a sleep recording if BECCT is suspected on clinical grounds when the awake EEG is not revealing. Spike discharges are not altered significantly by photic stimulation or hyperventilation.

Rolandic spikes usually occur on a normal background. When the spikes occur frequently, however, a focal pseudoslowing occurs that is secondary to the slow-wave component of the spikes.

Typically, the centrotemporal spikes have a horizontal dipole, with maximal electronegativity in the centrotemporal region and electropositivity in the frontal region. This suggests that the spikes are the result of a generator located in the lower rolandic region where the zero potential exists—between the frontal positivity and centrotemporal negativity.

Rolandic discharges having the same dipole field can be seen in children without clinical seizures and in children with epilepsy who do not have typical benign rolandic seizures. The spikes included with BECCT may be located in many areas other than the typical central-midtemporal areas.

The morphology of the spikes (rather than the location) is the distinctive factor in identifying the discharge in association with the benign rolandic epilepsy. Insistence on a centrotemporal location for the EEG may lead to a misclassification of the type of epilepsy. The term benign focal epilepsy of childhood also has been used, and when the discharge is located in the centrotemporal region, it is called benign focal epilepsy of childhood with a centrotemporal (or rolandic) location.

Differential diagnosis

Benign rolandic epilepsy must be differentiated from the following:

- Rolandic spikes and no seizures (often with behavior problems, headaches, or autonomic dysfunction)
- Rolandic spikes and a history of antecedent brain damage, cerebral palsy, or active local pathology
- Central spikes occurring commonly in Rett syndrome and fragile X syndrome
- Malignant rolandic epilepsy
- Psychomotor seizures and evolving temporal lobe epilepsy
- The aphasia-convulsion (Landau-Kleffner) syndrome and massive midtemporal spikes

Treatment

In view of the benign nature of the condition, intensive therapy is unnecessary. In the case of infrequent nocturnal partial seizures, withholding AEDs is reasonable if the child and family are comfortable with this approach.

One study found that in treated patients with BECCT, the frequency and duration seizures and the prevalence of active epilepsy were no different from those in untreated patients with the syndrome.

If treatment is indicated, carbamazepine is often the first medication to be tried, and seizures usually are well controlled. Once-a-day administration may be the only regimen needed to control seizures. Other reportedly effective AEDs include phenobarbital, phenytoin, valproic acid, clonazepam, clobazam, gabapentin, and sulthiame. Levetiracetam may also be effective and well tolerated.[31]

Although most patients respond to a low dose of a single drug, a few have seizures that are highly drug resistant. No correlation is known between resistance to AEDs and final outcome.

Duration of treatment may be shorter in some cases than epilepsy in general, and AEDs may be discontinued successfully in patients with normal EEG findings who have been seizure free for more than 2 years.

Prognosis

By general consensus, the prognosis of BECCT is excellent, as almost all patients achieve remission by adolescence. This includes patients whose seizures have been drug resistant.

A meta-analysis study on the course of BECCT found that 50% of patients were in remission by age 6 years; by age 18 years, 99.8% of the patients were in remission. Rarely, BECCT can relapse in adulthood; about 2% of patients in BECCT remission experience other seizure types.

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Disclosure: Nothing to disclose.

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Disclosure: Nothing to disclose.

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Disclosure: Nothing to disclose.

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