Editorial

Benign childhood partial epilepsies: benign childhood seizure susceptibility syndromes

I often wonder how “epilepsy”, which has been associated with demonic possession and magic, can still be used as a medical diagnosis. Social prejudice against “epilepsy”, accumulated over hundreds of years, is impossible to change. There is no other field in medicine in which a symptom (seizure) or a broad, unqualified term (epilepsy) are used as a diagnostic label for numerous disorders of different aetiology, prognosis and response to treatment.

The most important milestone in recent epileptology has been the recognition of epileptic syndromes and diseases, most of which are well-defined and easy to diagnose. The International League Against Epilepsy (ILAE) has published a proposal for the classification of epilepsies and epileptic syndromes, which, despite imperfections, should be considered as a significant step to improve diagnostic precision.

The benign childhood partial epilepsies (BCPE) exemplify the importance of the syndromic classification of epilepsies. They are common, comprising about one quarter of all epilepsies with onset under 13 years of age and have an excellent prognosis. BCPE are classified by the ILAE among “age and localisation-related idiopathic epilepsies”. The epileptic seizures and the EEG abnormalities are focal (localisation-related). They only occur in children (age-related). Physical, mental and laboratory examinations other than EEG are normal (idiopathic). The combination of a normal child with infrequent seizures and an EEG with disproportionately severe focal epileptogenic activity is highly suggestive of benign childhood partial epilepsy. The prevalent practice of not requesting an EEG after a first seizure may result in underestimation of the prevalence of BCPE, as 10–40% of children with BCPE may have only a single fit.

Two syndromes of BCPE are currently recognised by the ILAE: benign childhood epilepsy with centro-temporal spikes (BECTS) and childhood epilepsy with occipital paroxysms (BCEO). BECTS has been well-described but the clinical variants of BCEO are still controversial. There are also syndromes of benign childhood partial epilepsies with EEG spikes in other cortical regions which await recognition, such as benign childhood epilepsy with somatosensory evoked spikes, with frontal-parietal-temporal spikes and affective symptomatology, and with frontal, or midline spikes.

Benign childhood epilepsy with centro-temporal spikes

Benign childhood epilepsy with centro-temporal spikes is (previously known as Rolando's epilepsy) comprises three quarters of benign childhood partial epilepsies, and is characterised by striking ictal clinical manifestations and EEG abnormalities. A typical case is of a boy of 7–10 years of age found by his parents at night salivating and gurgling, attempting unsuccessfully to speak and apparently conscious. Unilateral motor seizures, mainly of face and hand, may ensue, progressing to a secondary generalised tonic-clonic seizure. Such seizures may occur once only, or the patient may suffer several within 1–5 years. Status epilepticus has been reported. In a few children atomic and/or myoclonic seizures may also occur either spontaneously or as a rare adverse effect to treatment with carbamazepine. They also have a good prognosis. The interictal EEG shows frequent, very high amplitude, sharp and slow wave complexes, unilateral or independently bilateral in the central and midtemporal areas, which are exaggerated by sleep. The morphological characteristics of centrotemporal spikes and dipolar topography of their electrical field (focal negativity in the centrotemporal regions with simultaneous positivity in the frontal regions) are more frequently associated with BECTS than symptomatic epilepsies. Ten to 20% of patients show sharp-slow wave complexes in other cortical locations. Seizures resolve within 1–3 years of onset and no later than 16 years of age. Rare cases (no more than 1–2%) persist into adult life and probably represent coincidence with other causes of seizures or misdiagnosis. Recurrences after the age of 16 years are so rare that they have been published as case reports. The benign character of this syndrome has been demonstrated in long follow up studies: 10–25 years after the last seizure only 1–2% of the patients over the age of 16 have solitary or infrequent seizures. This common epileptic syndrome with remarkable clinical and EEG features was first described in 1957 but was only generally recognised 10–15 years later.

Benign childhood epilepsy with occipital paroxysms

It would be surprising if an age-related epileptogenic susceptibility was confined only to the centro-temporal area of the cortex. Evidence precluding the description of BECTS indicated that spikes occurring in the occipital and other cortical locations may disappear with age or could be found in normal children. It was only recently, however, that the clinical manifestations of the BCEO were described.

Benign childhood epilepsy with occipital paroxysms is
less common than BCECTS, with a prevalence of 20–25% of benign childhood partial epilepsies. There are two clinical variants: an early- and late-onset BCEOP. A typical case of the early-onset BCEOP is a five-year-old child who wakes at night vomiting. There is deviation of the eyes to one side, and consciousness is often impaired. This state may last a few minutes to hours (simple or complex partial status), terminating with hemi- or generalised convulsions. Less common but better known is late-onset BCEOP which is the only variant recognised by the ILAE. It occurs in older children and may be misinterpreted as basilar migraine. The seizures are frequent, mainly diurnal and last between a few seconds and 2–3 minutes. They are characterised by visual illusions and/or blindness, often followed by post-ictal headache. The visual illusions of occipital epileptic seizures are usually multicoloured circles and spots in contrast with the predominantly black and white linear pattern of migraine. Consciousness is usually preserved but episodes of loss of consciousness may occur with hemi-, generalised seizures or without convulsions. The interictal EEG in both variants is characterised by occipital paroxysms, consisting of long runs of high amplitude sharp and slow wave complexes, often bilateral and attenuating when the eyes are open. Shape and amplitude are similar to centro-temporal spikes of BCECTS. Fixation-off sensitivity, that is, elimination of the discharges by visual fixation, is common.

Benign childhood epilepsy with frontal and benign childhood epilepsy with midline spikes

Benign childhood epilepsy with frontal and midline spikes have been described and long follow-up reports have confirmed a benign course, but no systematic studies have been published. Clinical manifestations are similar to those described in BCECTS and early onset BCEOP, but EEG sharp and slow wave complexes are located in the frontal and midline electrodes.

Unified concept

Benign childhood epilepsies with partial seizures and focal EEG sharp-slow wave complexes are a group of syndromes, which, in my opinion, share common clinical and EEG characteristics.

Seizures are infrequent, usually nocturnal and remit within 1–3 years from onset. Brief or prolonged seizures, even status epilepticus, may be the only clinical event of the patient’s lifetime. Ictal hypersalivation, vomiting, headache, pallor or sweating, unusual in other epileptic syndromes, are frequent and may occasionally appear in isolation. Childhood EEG characteristics of one may evolve into or simultaneously develop features of another form of benign childhood partial epilepsy. Febrile convulsions are common.

Neurological examination and intellect are normal, but some children may experience behavioural problems at the active stage of the disorder. Brain imaging is normal.

There are severe EEG abnormalities which are disproportionate to seizure infrequency. Epileptogenic foci, irrespective of their location, manifest abundant, high amplitude sharp-slow wave complexes, mainly in clusters. They are often bilateral, independent or synchronous, frequently combined with foci from other cortical areas or brief generalised discharges, and are exaggerated in stage I-IV of sleep. A normal EEG is exceptional and should provoke a sleep EEG study. Similar EEG features resolving with age are frequently found in normal school age children (1%), and children having an EEG for reasons other than seizures.

There is no reason to believe that all these syndromes differ from each other merely because an “epileptogenic” focus is a little anterior or posterior, lateral or medial to the centrotemporal regions. Studies of large cohorts of children with EEG spikes and sharp-slow wave foci have shown that spike prevalence is far higher in the centrotemporal (63%) than in the occipital (29%) or frontal (8%) locations. There is an age-related prevalence of such EEG foci with a peak at age eight to ten years for...
centrotemporal and four to five years for occipital spikes, followed by a sharp decline and disappearance by adolescence. 42 This age-dependent prevalence of different EEG foci corresponds exactly to that of the different types of benign childhood epilepsy and strengthens the concepts that BCEP constitute a single nosological entity with phenotypic variations. A unified concept of benign childhood partial epilepsies is also suggested by the frequency of more than one type of BCPE in an affected child and siblings. 19-22 37 38 50-55

The syndromes of benign childhood partial epilepsies are distinguished by clinical features dependent on location of EEG foci, but may represent phenotypic variations of the same genetic trait. Because of clinical and genetic studies of benign childhood partial epilepsies 56-59 some authors have suggested an overlap or genetic link between BCECTS and other syndromes of idiopathic generalised epilepsies or reading epilepsy. 56 Similar arguments have been raised over a putative relationship between BCECTS and a proportion of patients with the syndrome of acquired epileptic aphasia (Landau-Kleffner syndrome) 59 or a similar condition “epilepsy with continuous spike-waves during slow wave sleep”. 60 61 It is possible that idiopathic, not symptomatic or cryptogenic, forms of some of the above syndromes may represent more severe and extreme variations of the same genetic trait. Results and views, however, are often conflicting and beyond the scope of this editorial.

Treatment
There is no consensus on whether to treat these children with antiepileptic drugs. Most authors have recommended a 1–2 year course of treatment, mainly with carbamazepine, after a second documented seizure, but treatment may not be needed. 62 All major antiepileptic drugs have been reported successful even in small doses. Epileptogenesis secondary to kindling has been feared by some authors; 63 but the benign course of BCPE is reassuring.

Conclusions
All agree that BCPE or, at least, BCECTS (the major representative of the group) has a benign course with remission within a few years of onset and no later than 16 years of age. Unfortunately, current textbooks and even journals of paediatrics, medicine and neurology pay scant attention to these syndromes, often considering childhood seizures under the heading “epilepsy” without considering aetiology and prognosis. There is still a considerable social stigma attached to the diagnosis of epilepsy, and such seizures currently exclude patients from lifelong employment in certain fields. Benign childhood partial epilepsies, like febrile convulsions, 64,65,66 are age related, show genetic predisposition, may be manifested by a single seizure only, remit within a few years of onset, and may or may not require a short course of antiepileptic medication. The risk of recurrent seizures in adult life is less (1–2%) than in febrile convulsions (4%). As benign childhood partial epilepsy remits after the age of 16 years, legislation and employment guidelines should be altered to acknowledge a prognosis similar to that of febrile convulsions. Social prejudice, legislation and medical attitude cannot be expected to change rapidly. I propose removing the label “epilepsy” from patients with benign childhood partial epilepsies. This has already been achieved for febrile convulsions, and avoids inappropriate use of a term which implies “chronic brain disorder” as defined by the World Health Organisation dictionary of epilepsy. 67 Although the use of the term “benign childhood convulsions” would draw the parallel with febrile convulsions better, some of these children never convulse, others may have only one seizure occurrence in their life and others may have the EEG marker of BCPE without the companion seizures. The term benign childhood seizure susceptibility syndromes (BCSSS) is preferable, denoting an idiopathic propensity of cortical neurons to cause well defined focal seizures and/or EEG manifestations which are limited to childhood.

CP PANAVIOTOPoulos
Department of Clinical Neurophysiology and Epilepsy,
St Thomas’ Hospital,
London, UK

29 Beaumanoir A. Semiology of occipital seizures in infants and children. In:

Neurological stamp

Benjamin Franklin (1706–90)

Denied a formal education beyond the age of 10 because of his family’s poverty, Benjamin Franklin nevertheless, had many careers—including those of printer, author, philosopher, diplomat, scientist and inventor. He was fortunate not to die prematurely in 1752 when he performed his famous, but hazardous, experiment with a kite during a thunderstorm.

Franklin became the first person to identify lightning as an electrical discharge and as a result of his invention of lightning rods, he saved countless buildings from destruction. The simple terms charge, battery, plus, minus, negative, positive, armature and conductor were invented by him.

His contributions to medicine included serving on a committee with Lavoisier to investigate mesmerism, and inventing bifocal lenses and the flexible catheter. In his letters he discussed lead poisoning, deafness, and the infective nature of colds, and infections from corpses. He wrote a famous discourse on gout, a disease from which he suffered for 41 years. His medical knowledge and rules on health were first published in Poor Richard’s Almanac, in 1732. He also carried out research on the physiology of circulation and respiration, wrote extensively on the dilatation of the cardiac ventricles, a cure for yaws and the cause of fevers. Among his large number of inventions are the rocking chair, and an efficient stove.

For a time Franklin was Postmaster General of Philadelphia. He was instrumental in forming the academy that later became the University of Pennsylvania and was the principal founder and first president of the Pennsylvania Hospital (1751), the oldest independent hospital of the American Colonies.

Franklin has been frequently portrayed on postage stamps but in 1976 he was shown with a map of North America on a stamp commemorating the USA’s bicentennial. (Stanley Gibbons 1667, Scott 1690).

L F HAAS

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