2

Editorial

The classification of epilepsy – it's clinical applications

Jagath C Wijesekera*

Journal of the Ceylon College of Physicians, 1998, **31**;1 & 2, 2-4

Introduction

Attempts to classify the epilepsies were first made over one hundred years ago. Classifications based upon the clinical manifestations of epileptic seizures was the forerunner of today's classification of seizure types. The introduction of the Electroencephalogram (EEG) in the early 1930s had a major impact on classification providing a method of measuring and visualizing the physiological correlates of the clinical seizures. The issue of classification was taken up by the International League Against Epilepsy (ILAE) in the 1960s and in the past 20yrs the ILAE has formulated systems of classification that have become widely accepted.

The ILAE classification of seizure type

This method of classification which is now most widely used, is a descriptive scheme based on the clinical and EEG manifestations of seizures devised by the ILAE and officially adopted in 1982 (Table 1). In this scheme EEG data are taken into account but aetiology, age and anatomical site are ignored[1]. In this scheme seizures are divided into three groups: generalised, partial (focal) and unclassifiable. Generalised seizures are further divided into tonic-clonic (grand mal), absence (petit mal), myoclonic, atonic and clonic seizures. Partial seizures are subdivided into simple partial and complex partial categories according to the preservation or alteration of consciousness (simple and complex respectively). Generalised seizures are those in which epileptic discharges involve both hemispheres widely and simultaneously from the onset of the seizure, whereas partial seizures are those in which epileptic activity is confined to a focal area of the brain. The epileptic activity of partial seizures (simple or complex) may spread to become generalised, in which case the seizure is said to be secondarily generalised.

* *Consultant Neurologist, Institute of Neurology, National Hospital of Sri Lanka, Colombo.*

Table 1

International classification of seizure type (1981)

1. **Partial seizures**
   - (A). Simple Partial Seizures
     - (1) With motor symptoms
     - (2) With somato-sensory or special sensory hallucinations
     - (3) With autonomic symptoms and signs
     - (4) With psychic symptoms
   - (B). **Complex partial seizures**
     - (1) simple partial onset followed by impairment of conciousness
     - (2) with impaired conciouseness at onset
   - (C). **Partial seizures evolving to secondary generalised seizures**
     - (1) simple partial evolving to generalised
     - (2) complex partial evolving to generalised
     - (3) simple partial evolving to complex partial evolving to generalised
2. **Generalised seizures**
   - A. (1) absence seizures
     (2) atypical absences
   - B. myoclonic seizures
   - C. clonic seizures
   - D. tonic seizures
   - E. tonic-clonic seizures
   - F. atonic seizures
3. **Unclassifiable epileptic seizures**

The ILAE classification of the epilepsies and epileptic syndromes and related disorders

More recently in recognition of the fact that a seizure type classification does not account for the aspects of the heterogenicity of epilepsy, the ILAE devised a new scheme in 1986, and adopted it in 1989 (Table 2.) The classification takes into account seizure type, EEG, Prognostic, Pathophysiological and aetiological data[2]. It retains the division of the epilepsies in to generalised and partial (now called localization – related) categories, with each category subdivided in to symptomatic and idiopathic. Two new categories are added.

(a) Epilepsies and syndromes undetermined whether focal or generalised.

(b) Special syndromes.

This scheme is complex and may confuse those not familiar with it. However, it is a serious attempt to incorporate more than simple seizure type data into a more comprehensive classification.

The advantages of a correct syndrome diagnosis will be illustrated by reviewing two epileptic syndromes and emphasising the additional information of Prognosis and Long-term outcome such a diagnosis provides in comparison to a seizure type diagnosis.

1. Roland Epilepsy – (Benign Childhood Epilepsy with Centro–Temporal Spikes)

Rolandic Epilepsy is a well-defined epileptic syndrome[3,4] that starts between the ages of 3 to 13yrs. The child has simple partial seizures with sensory and motor symptoms, originating from the throat and often there is secondary generalisation. The seizures occur almost exclusively at night after a few hours sleep. The EEG shows a high voltage Centro-temporal spike focus, which may shift from side to side. Despite the clear-cut partial nature of the seizures and pronounced focal EEG changes, the aetiology of this syndrome is idiopathic with a genetic predisposition.

Consequently, in typical cases, there is no need for neuro-radiological examination, which would otherwise be indicated by the focal nature of the epilepsy. In addition despite the strict focal nature of the seizures, the response to anti-epileptic treatment is very sucessful. Treatment can be discontinued after a few years of freedom from seizures and in some children seizures are so infrequent that treatment is not indicated.

A diagnosis of this syndrome therefore offers important information about therapy, out-come and prognosis.

Table 2

International classification of epilepsies and epileptic syndromes and related seizure disorders (1989)

(NB – Abbreviated with important epileptic syndromes only)

1. **Localisation related (focal, partial)**

   1.1 Idiopathic
   Rolandic Epilepsy (Benign childhood epilepsy with Centro–Temporal spikes)

   1.2 Symptomatic
   Temporal Lobe Epilepsy
   Occipital Lobe Epilepsy
   Frontal Lobe Epilepsy

2. Generlised Syndromes

   2.1 Idiopathic
   Childhood absence epilepsy
   Juvenile absence epilepsy

   2.2 Symptomatic or Cryptogenic
   Infantile spasms (West Syndrome)
   Lennox Gastaut Syndrome (myoclonic astatic epilepsy)

3. **Syndromes undetermined whether focal or generalised**

   Neonatal seizures
   Epilepsy with continous spike waves during sleep

4. **Special syndromes**

   Febrile convulsions

---

2. Juvenile myoclonic epilepsy

In this syndrome seizures start around puberty[5]. The aetiology is idiopathic, with a pronounced hereditary element. It accounts for about 5% of all cases of epilepsy[6]. There are three-seizure types –

(a) Myoclonic jerks occurring in the mornings

(b) Generalised tonic-clonic convulsions also occurring in the morning or at least before noon.

(c) Absences – (rarely)

Myoclonic jerks may be present years before the onset generalised convulsions. Unless questioned specifically, patients very seldom describe the jerks, although they may greatly interfere with everyday life. Sleep deprivation and alcohol are well established *seizure provoking factors* in general in all epilepsies but in this syndrome may provoke seizures to such an extent that it may be necessary for the patient to abstain completely from alcohol and adopt a right life-style with regard to sleep.

An EEG obtained at the correct time (i.e in the morning after sleep deprivation) may show polyspike – wave paroxysms. Characteristic EEG changes are sometimes seen in family members without seizures. The drug of choice this syndrome is *Sodium Valproate*[7]. Despite complete seizure control for many years, withdrawal of treatment is often associated with recurrence; so extended periods of treatment are called for. To detect this syndrome any young person who present with convulsions should be questioned about myoclonic jerks, in isolation or preceding a seizure.

The syndrome of Juvenile Myoclonic Epilepsy therefore has implications with regard to *diagnostic measures, seizure presenting behavior, choice of treatment and duration of therapy*[8].

Conclusion

The syndromes of Rolandic epilepsy and Juvenile Myoclonic epilepsy are thus good examples of the advantages the syndrome classification has over the seizure type classification. Another advantage of the former is it's flexibility and the potential for change and expansion. There are however a number of profound disadvantages of the syndrome classification. They are –

(a) it is a highly complex system with clumsy terminology.

(b) in the attempt to be all-inclusive the classification becomes unwieldy.

(c) common syndromes are mixed with those which are extremely rare.

However, these disadvantages are outweighed by the many advantages of the syndrome classification and one has to acknowledge that it is a serious attempt to incorporate more than simple seizure type data into a more comprehensive classification with greater clinical implications.

References

1. The Commission on Classification and Terminology of the International League Against Epilepsy. Proposal for revised clinical and electroencephalographic classification of epileptic seizures. *Epilepsia* 1981; **22**: 489-501.

2. The Commission on Classification and Terminology of the International League Against Epilepsy. Proposal for revised classification of epilepsies and epileptic syndromes *Epilepsia* 1989; **30**: 389-98.

3. Lombroso CT. Sylvian seizures and mid-temporal spike foci in children. *Arch. Neurology* 1967; **17**: 52-59.

4. Beaussart M. Benign epilepsy in children with Rolandic (centro-temporal) paroxysmal foci. *Epilepsia* 1972; **13**: 795-811.

5. Janz D, Christian W. Impulsiv-Petit mal. *Dtsch z Nervenheilk* 1957; **176**: 346-86.

6. Janz D. Epilepsy with impulsive petit mal (juvenile myoclonic epilepsy) *Acta Neurol Scand* 1985; **72**: 449-59.

7. Covanis A, Gupta AK, Jeavons PM. Sodium valproate monotherapy and polytherapy. *Epilepsia* 1983; **23**: 693-720.

8. Gram L. Epilepsy Octet. Epileptic seizures and syndromes. *Lancet* 1990; **336**: 161-63.