

EDITORIAL

# Intractable or refractory epilepsy

Neil Gordon

Retired, The Children's Hospitals, Manchester, United Kingdom

---

There can be no doubt that the management of intractable or refractory epilepsy is one of the major problems that neurologists have to face, whatever the age of the patient. It can be considered under at least five headings; causes, lifestyle, drugs, alternative therapy, and surgery.

## Causes

The prognosis for those suffering from frequent seizures will be closely linked to their etiology (1). If there is extensive brain damage, whether this results from trauma, disease, or brain tumors, there is likely to be a poor response to whatever treatment is given. The more diffuse the lesions the worse this will be: and localisation is also a factor, for example mesial temporal sclerosis causing complex partial seizures. For these reasons patients who are mentally handicapped often have the additional handicap of epilepsy (2). The best chance of controlling the fits lies in making an accurate diagnosis and applying appropriate treatment, whether this is surgery to remove a tumor or the identification of a metabolic disorder which can be controlled, such as pyridoxine dependency or biotinidase deficiency (3). It is important to identify epileptic syndromes, often genetically determined, so that at least an accurate prognosis can be given, even if this does not effect treatment. For example the prognosis for infantile spasms and the Lennox-Gastaut syndrome are well known (4). Tuberous sclerosis is another diagnosis to be viewed with concern. Even within the field of epilepsy there are important diagnostic issues; for instance differentiating minor seizures from reflex anoxic seizures, and absence epilepsy and pseudoseizures from complex partial seizures (5).

## Life style

Epilepsy may respond badly to treatment for reasons unrelated to medication. This applies especially to adolescents. Lack of sleep, fatigue, and stress can be factors, as can compliance. Abuse of alcohol, solvents, and drugs will inevitably have adverse effects. If significant depression or anxiety are present these should be treated as a matter of urgency, and, even at the risk of the occasional seizure, too many restrictions should not be imposed. The establishment of a well-regulated life-style can often work wonders (6).

## Drugs

The choice of the appropriate drug for the individual patient cannot be over-stressed, nor can the use of one drug only and in an adequate dose; not too little or not too much (7). If, in order to control the seizures, more than one drug has to be given, then a close watch must be made for side effects and the influence of one drug on another. There is no doubt that some anti-epileptic drugs make some types of epilepsy worse, for example excessive drowsiness from phenobarbitone, the toxic effects of phenytoin, or giving carbamazepine for the multiple seizure types which occur in the Lennox-Gastaut syndrome (8). In this context it must surely be important for one doctor, acceptable to the patient and family, to assume the responsibility for a patient's anti-epileptic drug treatment in the long-term.

The problems related to the drug treatment of children with epilepsy are well illustrated in the paper by Saneto et al. (9) in this issue of the Journal of Pediatric Neurology.

## Alternative therapy

In view of the complexity of anti-epileptic drug treatment for epilepsy no wonder alternatives have been sought. One method that has been successful, particularly in patients with absence epilepsy, is the use of the ketogenic diet. It has to be administered with some skill, but in spite of its expense and unpalatability it can be well tolerated, especially

---

**Correspondence:** Neil Gordon, M.D.FRCP, HonFRCPCH.

Huntlywood  
3 Styal Road  
Wilmslow  
SK9 4AE

United Kingdom

Tel: 0 16 25 525437.

E-mail: neil-gordon@doctors.org.uk

Received: July 17, 2004.

Accepted: July 19, 2004.

when using medium-chain triglycerides (10). Alternative drugs can be considered in certain conditions, for example steroids for treating infantile spasms.

Behavioural methods of seizure control have been tried, but with limited success (11). These include reward management, self-control, and psychotherapy, and also biofeedback training which aims at reinforcing certain rhythms in the electroencephalography. In the case of reflex epilepsy avoidance of the trigger mechanism may be the best approach, but deconditioning can be tried, such as repetitive monocular stimulation in visually evoked seizures.

### Surgery

If there is no doubt about the intractability of a patient's epilepsy the main point to be made is; sooner rather than later. If this treatment is successful in childhood it may mean that years of lost schooling are avoided, quite apart from the other benefits to the patient and family. There are a wide variety of operations available, the excision of an epileptic focus being the most obvious one, but even generalized epilepsy can be treated in this way by procedures such as callosotomy, and multiple subpial transactions. The secret of success seems to be in the meticulous preoperative assessment of the patient (12).

### Conclusions

In view of the many possibilities for improving the lot of these often severely handicapped patients a holistic approach is essential so that all aspects of the patient's condition are considered. A doctor

alone is unlikely to be able to do this, and a team will be needed which includes medical specialists, psychologists, social workers, educationalists, and others when there are special needs.

### References

1. Aicardi J. Clinical approach to the management of intractable epilepsy. *Dev Med Child Neurol* 1988; **30**: 429-440.
2. Hauser WA, Nelson KB. Epidemiology of epilepsy in children. *Cleve Clin J Med* 1989; **56**: S185-S194.
3. Wolf B, Grier RE, Allen RJ, et al. Phenotypic variation in biotinidase deficiency. *J Pediatr* 1983; **103**: 233-237.
4. Roger J, Dravet C, Bureau M. The Lennox-Gastaut syndrome. *Cleve Clin J Med* 1989; **56**: S172-S180.
5. Finlayson RE, Lucas AR. Pseudoepileptic seizures in children and adolescents. *Mayo Clin Proc* 1979; **54**: 83-87.
6. Messing RO, Closson RG, Simon RP. Drug-induced seizures: a 10-year experience. *Neurology* 1984; **34**: 1582-1586.
7. Gordon N. Intractable epilepsy. *Dev Med Child Neurol* 1988; **30**: 830.
8. Blume WT. Uncontrolled epilepsy in children. *Epilepsy Res* 1992; **5 Suppl**: 19-24.
9. Saneto RP, Kotagal P, Rothner AD, Baker J, Kotagal LL. Valproic acid use in pediatric partial epilepsy after initial medication failure. *J Pediatr Neurol* 2004; **2**: 199-203.
10. Gordon N. Medium-chain triglycerides in a ketogenic diet. *Dev Med Child Neurol* 1977; **19**: 535-538.
11. Fenwick P. Behavioural therapy of epilepsy. In: Pedley TA, Meldrum BS (eds). *Recent Advances in Epilepsy* (5th ed). Edinburgh: Churchill-Livingstone, 1992, pp 75-92.
12. Palmini A, Andermann F, Oliver A, Tampieri D, Robitaille Y. focal neurological migration disorders and intractable partial epilepsy: results of surgical treatment. *Ann Neurol* 1991; **30**: 750-757.