

Intractable Epilepsy: The Invisible Disability

W. McIntyre Burnham

WHAT YOU WILL LEARN

- The nature of epilepsy
- The nature of *intractable* epilepsy
- The cognitive, psychiatric, behavioral and reproductive problems associated with intractable epilepsy
- The resources available for people with epilepsy

WHAT ARE THE EPILEPSIES?

Definitions

The “epilepsies” are a group of neurological disorders, characterized by spontaneous, recurring seizures. The epilepsies are also called “seizure disorders”.

Except for headache, epilepsy is the most common condition treated by neurologists. About 4% of the population will have epilepsy at some time during their lives. About 1% of the population has epilepsy at any given time.

“Seizures” are periods of neural hyperactivity, caused by an imbalance between excitation and inhibition in the central nervous system. During a seizure, the neurons in the brain cease their normal activities and fire in massive, synchronized bursts. After some seconds or minutes, the inhibitory mechanisms of the brain regain control, and the seizure ends.

During a seizure, the epileptic activity in the brain can be seen as a series of “spikes” or “spikes and waves” in electroencephalographic (EEG) recordings. These “spikes” or “spikes and waves” are called the “electrographic” seizure. The *behavior* of the patient during the epileptic attack - which may or may not involve convulsions - is called the “clinical” seizure. If the clinical seizure involves muscle spasms, it is called a “convulsion”.

Seizures are also called “attacks”, which is an acceptable word. The term “fit” is acceptable in Britain, but not in North America. (For more discussion, see Burnham, 2002; Guberman & Bruni, 1999; Engel & Pedley, 1997.)

Onset of epilepsy

The onset of epilepsy may occur at any time during life. In many individuals, however, seizure onset occurs in childhood, before the age of 15. During young adulthood or in middle age, seizure onset is less likely. There is an increased incidence of seizure onsets, however, after the age of 60. These late-onset epilepsies may be the result of small strokes (Guberman & Bruni, 1999).

Frequency of seizures

The frequency of seizures varies enormously. Some individuals experience only a few seizures during their whole lives. Other people experience many seizures every day.

Classes of epileptic seizure

There are a number of different types of epileptic seizure, some of which occur primarily in children. A description of all of the seizure types is beyond the scope of the present discussion. Some of the more common types are described in Table 1.

Epileptic syndromes

Recently, there has been an attempt to fit epileptic seizures into larger entities known as “epileptic syndromes”. An epileptic syndrome consists of a seizure type (or types), a prediction about the time of seizure onset and (possibly) offset, a possible cause, a prognosis, and a likely response to medication.

Well known - and very serious - epileptic syndromes include West syndrome and Lennox-Gastaut syndrome. West syndrome usually has its onset during the first year of life, whereas Lennox-Gastaut syndrome usually begins between ages 1 and 8. Both syndromes involve drug-resistant seizures, an abnormal EEG between seizures, and, in most cases, mild to severe developmental delay (see Guberman & Bruni, 1999).

Causes of epilepsy

In some individuals with seizures (about 40%), there is a clear-cut abnormality in the brain, such as a scar or a tumor. These people are said to have “symptomatic” epilepsy. In such patients, the seizures are caused by the abnormality in their brain. In other individuals with seizures (about 60%), the brain appears to be completely normal. These people are said to have “idiopathic” epilepsy. In patients with idiopathic epilepsy, the seizures are thought to be caused by genetic factors.

Actually, there is probably a genetic contribution to most cases of epilepsy. If one of a pair of identical twins has seizures, there is a 95% chance that the other twin will also have seizures. Even in cases of symptomatic epilepsy, involving a scar or tumor, some patients develop seizures, whereas others do not - suggesting a genetic predisposition in those that do. In some types of epilepsy the genetic factor is strong, and inheritance follows simple Mendelian rules. In most cases of epilepsy, however, inheritance is “multifactorial” - meaning that several genes are involved. In these cases, inheritance does not follow simple Mendelian rules (see Burnham 2002; Guberman & Bruni, 1999).

Epilepsy: Relation to other disabilities

A number of common disabilities are caused by brain damage or dysfunction. People with these disabilities often also have seizures as well. Table 2 presents a list of common disabilities, and indicates the percentage of individuals that also have epilepsy.

Treatment of epilepsy: Drug therapy

The most common therapy for epilepsy is treatment with anticonvulsant drugs. The anticonvulsant medications are also called “antiepileptic drugs”, “AED’s” or “antiseizure drugs”.

A wide variety of anticonvulsant drugs are available. Among the most commonly prescribed are: 1) ethosuximide, which is used for absence seizures; 2) phenytoin and carbamazepine, which are used for tonic-clonic and partial seizures; and 3) valproic acid, which is a wide-spectrum anticonvulsant, effective against many types of seizures. Phenobarbital is an older drug that is still in use, most often in children.

A number of new drugs have been introduced within the past few years. These include gabapentin, lamotrigine, levetiracetam, tiagabine, topiramate, vigabatrin, and zonisamide. The new drugs are generally thought to have fewer side-effects than the older drugs. It is not clear, however, that they are better at stopping seizures, and they are considerably more expensive.

The anticonvulsant medications do not cure epilepsy. They simply suppress seizures on a temporary basis. People with seizures must continue to take their medications, once, twice or three times daily, sometimes for the rest of their lives (for discussion, see Bialer et al., 2002; Guberman & Bruni, 1999; Burnham, 1998).

WHAT IS INTRACTABLE EPILEPSY?

Successes and failures of drug therapy

People who have seizures in public experience both social and economic discrimination. The impact that seizures have on life, therefore, really relates to whether they can be controlled

with anticonvulsant medications. If the seizures are responsive to medication - as they are in about 60% of cases - the patient may never have another seizure. He or she will have to take medication regularly, but epilepsy will not prevent a full, productive life.

If the seizures are only partly responsive to medications - as they are in about 20% of cases - the individual will continue to have some attacks. If the attacks occur in public, he or she may face social and economic discrimination. Moreover, if an individual has even one seizure a year, he or she may never be able to legally drive again (see below). Partially resistant seizures, therefore, have a clear, negative effect on life. Still, many people with partially resistant seizures also live productive, successful lives.

If seizures are fully drug resistant - as they are in about 20% of patients - the individual will continue to experience seizures – which may be frequent – despite the best medications. Drug-resistant seizures are called “intractable” or “refractory”. Patients with intractable seizures cannot legally drive, and often have major problems at home, at school and at work. (For more discussion, see Burnham, Carlen, & Hwang, 2002; Johannessen, Gram, Sillanpaa, & Tomson, 1995.)

Patients with intractable epilepsy are frequently treated with two or three anticonvulsants at the same time. Even such “polypharmacy”, however, does not stop their attacks. These are the people who are referred for non-pharmacological therapies, such as surgery, the ketogenic diet or vagal nerve stimulation.

Treatment of intractable epilepsy: Non-drug therapies

If seizures prove intractable to drug therapy - or if drug side-effects cannot be tolerated – several sorts of non-drug therapy are available.

If the individual experiences partial seizures – and the seizures always arise from the same part of the brain (the “focus”) - he or she may be a candidate for seizure surgery. In adults, the most common sort of seizure surgery involves removal of the part of the brain that contains the focus, most often the anterior part of one of the temporal lobes. Surgery may stop the seizures, or make them more controllable with medication (see Luders, 1992).

Another non-drug therapy is the ketogenic diet. This is a high-fat diet, with adequate protein, and very little carbohydrate. Many people find the diet unpalatable, and it is hard to maintain, since all of the food must be weighed and measured. The ketogenic diet, however, stops seizures in about a third of people who have failed drug therapy, and decreases seizures in another third. Traditionally, the diet has been used only in children. Recent reports, however, suggest that it is effective in adults as well. Unfortunately, due to the unbalanced nature of the diet, individuals can only stay on it for two or three years. (For discussion, see Wheless, 2001; 1995.)

A third treatment for drug-resistant seizures is vagal nerve stimulation. A device similar to a cardiac pacemaker is implanted into the chest muscle, and a wire is connected to the vagus nerve, a nerve that originates in the brain. Intermittent stimulation is used to control seizures.

Vagal nerve stimulation is not as effective as surgery or the ketogenic diet, but it is an alternative to consider when the other therapies fail. Vagal nerve stimulation, or “VNS”, is now the second most common type of non-drug therapy in the United States. (For discussion, see McLachlan, 1997.)

Unfortunately, many people with intractable seizures are not good candidates for seizure surgery. The ketogenic diet does not offer long-term control, and less than half the people who try vagal stimulation show significant benefits. Many individuals with intractable epilepsy face a life of uncontrolled seizures, despite the best attempts at therapeutic intervention.

Intractable epilepsy as a disability

Epilepsy is sometimes called the “invisible disability”. Is this a reasonable assessment? If one thinks in terms of people with drug-responsive seizures, the term “disability” is probably too strong. These people usually lead normal lives, and reach their full potential for accomplishment.

Intractable epilepsy, however, is clearly a disability. Part of the problem is the seizures themselves. Public seizures lead to stigma, and to economic and social discrimination. There is the possibility of head injury due to the falls, and people with uncontrolled seizures have to be careful about climbing ladders, or standing near the edge of subway platforms. They are warned against swimming alone, and told to take showers rather than baths. They cannot drive. Life becomes limited and circumscribed.

The seizures themselves, however, are only part of the problem. In addition to seizures, people with intractable epilepsy suffer from a number of related non-physical disabilities. These are called the “co-morbidities” of intractable epilepsy (below). These are less well known than the seizures, and less understood. Unfortunately, they are seldom treated.

THE CO-MORBIDITIES OF INTRACTABLE EPILEPSY

Both parents and educators agree that the co-morbidities of intractable childhood epilepsy are often more serious than the seizures themselves. These co-morbidities are summarized in Table 3. If seizures cannot be stopped by drug or non-drug therapy, the co-morbidities must be addressed, and strategies for dealing with them must be evolved.

Intractable epilepsy and cognitive impairment

Intractable epilepsy is often associated with cognitive impairment. We have already discussed the profound impairments seen in children with the West and Lennox-Gastaut syndromes. Impairment, however, may be associated with many forms of intractable seizures. In children, this often becomes evident during the school years. Children with intractable seizures

have lower IQ's, often in the "low normal" range of 80-85. Studies have also found a significant correlation between low IQ and the duration of the child's seizure disorder.

Even when IQ's are in the normal range, children with uncontrolled seizures do less well in school than non-epileptic children. A number of factors may contribute to this:

1) **Frequent Absence Attacks** One cause of learning problems is frequent absence seizures. These mild, non-convulsive attacks consist only of brief lapses of consciousness. Some children have hundreds per day, however, and clusters of dozens may occur within a few minutes. During these periods, the child cannot follow what is going on around him or her. Children with absence epilepsy, therefore, may give the appearance of being "slow learners". Actually, their intelligence is normal.

2) **After-effects of Seizures** Severe seizures, such as tonic-clonic attacks, cause a major perturbation in the brain. The after-effects of such seizures last for hours. If a child has had one or more seizures during the night, he or she may show excessive fatigue during the following day, and may appear to have forgotten things he or she knew the day before.

3) **Interictal Spikes** Some children - particularly those with complex partial seizures - have isolated epileptic "spikes" in their EEG between seizures. These are called "interictal spikes". Interictal spikes produce no outward manifestation, but they slow the child's ability to process and retrieve information, causing transient cognitive impairments.

4) **Site-Related Deficits** Children with an epileptic focus in particular parts of the brain may show *selective* deficits related to that area. Children with a focus in the left hemisphere (dominant for language), for example, often have trouble with finding and remembering words. Children with a focus in the right hemisphere may have problems with visual memory.

5) **General Memory Defects** One of the most common complaints in people with intractable epilepsy is a general defect in memory. Children will complain that they have to have their lessons repeated over and over before they can remember them. They know that their classmates don't have this problem.

The reasons for memory impairment are not completely clear. In some cases it may relate to the side-effects of anticonvulsant drugs (below). In others, it may relate to changes in the brain. Individuals with long-standing intractable seizures arising in the temporal lobes begin to lose neurons in the hippocampus, a subcortical forebrain structure. This loss of neurons is called "hippocampal sclerosis" or "mesial temporal sclerosis". The hippocampus is involved in memory formation, and hippocampal sclerosis, when severe, is associated with memory problems.

6) **Side-effects of Medication** In addition to the cognitive impairments associated with intractable seizures, there are impairments associated with the sedative (sleep inducing) side-effects of the anticonvulsant drugs. These side-effects are most serious at the start of therapy. They improve as tolerance develops, but they don't entirely disappear. They are worst with the older drugs, such as phenobarbital, but they may be seen with almost any of the anticonvulsants.

They are more of a problem in children taking multiple drugs. Table 4 presents the commonly used anticonvulsants, and indicates whether they are likely to significantly impair cognition.

7) Sleep Disorders / Daytime Drowsiness People with uncontrolled seizures often suffer from sleep disturbances. These are believed to be caused both by their seizures, and by some of the anticonvulsant drugs. This disturbed sleep leads to daytime drowsiness, and poor cognitive performance. (For discussion, see Bazil, 2003; Foldvary-Schaefer, 2002.)

As children grow, the co-morbidities of intractable epilepsy grow with them. The focus of trouble simply shifts from the school to the work place. Cognitive deficits, once acquired, are likely to remain. The after-effects of seizures still occur in adults, as do the sedative side-effects of anticonvulsant medications. (For further discussion, see Bortz, 2003; Motamedi & Meador, 2003; Williams, 2003; Viskontas, McAndrews, & Moscovitch, 2000).

Intractable epilepsy and psychosocial impairment

Emotional and psychosocial difficulties are disproportionately high in people with intractable epilepsy.

In one large study, about 50% of the children with intractable epilepsy were identified as having serious psychosocial problems. In another study, clear-cut psychiatric disorders were identified in 33% of children with epilepsy, as compared to 7% in the general population and 12% in children with other chronic illnesses.

Some of the more common problems include anxiety, depression, irritability, aggression, and irrational periods of rage. In children at risk for suicide, there is a fifteen-fold over-representation of children with epilepsy.

Emotional disturbances, especially anxiety and depression, are also common in adults with uncontrolled epilepsy. Self-confidence is low. Often there is social isolation and withdrawal. Adult children may continue to live with their parents. The suicide rate is five times higher than in the general adult population. People with seizures are also over-represented in the prison population.

In addition to anxiety and depression, some people with intractable seizures develop a schizophreniform psychosis. This occurs in about 5-10% of people with intractable epilepsy. Usually the patient has complex partial seizures with a focus in one of the temporal lobes, and usually he or she has had uncontrolled epilepsy for at least 10 years. In occasional patients, the psychosis will clear after a seizure, and then gradually reappear.

A recent study on adults with uncontrolled seizures - and normal IQ's - reported that about 30% had psychiatric disorders, including psychosis, antisocial personality disorders, anxiety and depression.

Both in children and adults, these psychiatric problems are responsive to therapy -

including therapy with antidepressant medications. Unfortunately, these psychiatric problems are seldom diagnosed or treated. Therapy focuses on seizure control, and emotional problems are neglected. (For discussion, see Boro & Haut, 2003; Bortz, 2003; Gilliam, Hecimovic, & Sheline, 2003; Vazquez & Devinsky, 2003; Blumer, 2002; Harden & Goldstein, 2002; Leonard & George, 1999; Jahnukainen, 1995; Sahlholdt, 1995).

Intractable epilepsy and ADHD

In addition to emotional problems, children with uncontrolled seizures may have problems with hyperactivity. It is estimated that 20-30% of children with epilepsy suffer from concurrent attention deficit/hyperactivity disorder (ADHD). A still larger number of children with seizures show deficits in attention or in impulse control, without showing the full ADHD syndrome.

The emotional and behavior problems noted above may be compounded by the effects of the anticonvulsant drugs. ADHD is exacerbated by sedatives, and many of the anticonvulsants have sedative side-effects. Children on anticonvulsants, therefore, may suffer a change of personality. They may become impulsive, hyperactive and irritable - and they may engage in both verbal and physical aggression. These problems disappear when the drug is stopped.

Anecdotally, neurologists report that most of the anticonvulsants can produce behavioral side effects. The worst problems, however, are probably associated with clonazepam (Rivotril). A related drug, clobazam (Frisium), causes fewer problems. In children, phenobarbital also sometimes causes a hyperactive syndrome. One of the new drugs, vigabatrin (Sabril), is associated with depression or outright psychosis in a small number (2-4%) of patients. Vigabatrin is not prescribed much any more, however, since it also tends to cause visual field defects.

Such "paradoxical excitability" may also be seen in elderly patients, but it is less likely to occur in adults.

The connection between anticonvulsants and behavior problems is often overlooked. If a patient has a change in behavior after the initiation of a new drug, the possibility of behavioral side-effects should be considered. A patient with severe behavior problems on one drug may be quite normal on another. (For more discussion, see Sanchez-Carpintero & Neville, 2003; Williams, 2003.)

Intractable epilepsy and reproductive problems

Reproductive and hormonal disorders are common in both men and women with intractable epilepsy. This is particularly true if the epilepsy is of temporal-lobe origin.

In women, menstrual disorders are seen, such as irregular or missed menstrual cycles, or cycles in which there is no ovulation. Fertility is reduced to 70-80% of normal. Hormonal disorders include hypogonadism (with too little estrogen) and polycystic ovaries (with too much estrogen). Anticonvulsant drugs, and particularly valproate, may contribute to these disorders.

In men with intractable epilepsy, there is an increased risk of erectile dysfunction. Over 90% of men with epilepsy have abnormal semen analyses, including decreased sperm count and impaired sperm motility.

In both sexes, diminished sexual desire and responsiveness have been described. (For discussion, see Boro & Haut, 2003; Isojarvi, 2003; Morrell, 2003; 1998; 1991; Edwards et al., 2000).

Intractable epilepsy and the family unit

Intractable seizures, and the emotional, cognitive and behavior problems which accompany them, are a burden not just for the child, but also for the family.

Parents - and particularly mothers - often blame themselves for their child's epilepsy. In some cases, shame is added to guilt as the grandparents blame the parents for their child's condition. Many parents go through the stages of denial, anger and depression, usually seen when a child dies. Some parents develop psychosomatic reactions, such as sleep disturbances, headaches and loss of appetite.

It appears to be the episodic, unpredictable nature of epilepsy that makes it harder to live with than other childhood disabilities. Both mothers and fathers live in fear of the next attack. It is common in newly diagnosed cases that one parent will stay up all night for fear that the child will die in its sleep.

Eventually, most parents come to terms with their child's epilepsy. Some, however, remain "stuck" in a state of constant crisis. Many parents of children with chronic epilepsy would benefit from short-term psychotherapy. They seldom seek it or receive it, however.

Support groups consisting of other parents living with childhood epilepsy are of considerable help. These are usually organized by the regional epilepsy associations. In the absence of therapy or support, divorce is a very real possibility.

Siblings of epileptic children suffer as well. They may develop both emotional complaints (fear of becoming sick or dying, nightmares) and physical complaints (headache, vomiting). There may be sibling rivalry, as parents are perceived as "favoring" the child with seizures. A large study found that about 25% of the siblings of children with chronic epilepsy were perceived as "disturbed" by their teachers. Siblings may also require therapy - in addition to children with seizures and their parents. (For discussion, see Wada et al., 2001; Ellis et al., 2000).

Intractable epilepsy and the schools

Children with intractable epilepsy often have major problems at school.

These may be due in part to the cognitive problems associated with seizures. Because the children often have *selective* learning difficulties, it is important that the child undergo psychological or psychoeducational assessments to identify areas of weakness and strength. Once it is established that the child has specific learning difficulties, an education plan can be identified to improve academic performance in that area.

From the school's point of view, however, most difficulties relate not to cognitive problems, but to behavior problems. Teachers receive no education in epilepsy, and often do not understand the connection between uncontrolled seizures and behavior problems. When behavior problems occur, the child is simply perceived as a "bad apple". Children with epilepsy are frequently sent to the principal, and are often suspended from school. The children themselves sometimes express hatred of their teachers, and their families often end up "at war" with the school system.

The stigma of epilepsy, and the insensitivity of other children - and sometimes teachers - negatively affect the emotional and behavioral status of the children. Many children with seizures are excluded from school and recreational activities by teachers, and teased and bullied by their peers.

The school situation would be much improved if both the schools and the parents understood the co-morbidities associated with intractable epilepsy. What is needed is a partnership between health care providers, the parents and the school. It is important: 1) that parents and health care providers tell the school about the child's seizures and how they are to be managed; 2) that the co-morbidities of intractable epilepsy are explained and dealt with; 3) that an education plan to improve academic success is evolved; 4) that supports, such as an educational assistant, are put in place; 5) that the importance of the child's participation in recreational and class activities is accepted, and 6) that social interactions with peers are encouraged. (For discussion, see Tidman, Saravanan, & Gibbs, 2003; Hightower, Carmon, & Minick, 2002).

Intractable epilepsy, work and driving

Perhaps the biggest problems for adults center around work. The majority of adults with uncontrolled seizures are unemployed or underemployed - that is, employed in jobs well below their level of competence.

Part of the problem is driving. In most countries, driver's licenses are revoked after the first seizure. They can be reinstated if the driver is seizure free for a period of time - often a year. People with intractable epilepsy, however, are seldom seizure free for a period as long as a year. Most of them will never legally drive again. Therefore, they cannot take any job which requires driving, or which can only be reached by car.

A second part of the problem is public seizures. People are often fired if they experience seizures at work. Even though the law forbids firing for epilepsy, employers eliminate people with public seizures, using some other pretext.

A final problem is disclosure. People with seizures fear that they will not be hired if they disclose their problem to potential employers. They also fear that they will be fired later if they do not.

There are no simple solutions to these problems. Work - and a stable income - are major concerns for adults with intractable epilepsy. (For discussion, see Bishop & Allen, 2001; Wada et al. 2001).

SUMMARY

In addition to seizures, children with intractable epilepsy often have a lowered IQ, and real problems with learning and memory. They also have significant psychosocial problems, some of them related to the anticonvulsant drugs. People who work with these children must learn patience, and try to evolve new strategies for the promotion of learning. Adults with uncontrollable seizures also have cognitive and psychosocial problems. They cannot legally drive, and they may develop reproductive disorders. They face discrimination in the work place, and are frequently unemployed or underemployed. Work - and a stable income - are major concerns. These people need sympathy and support. Intractable epilepsy is a true – if invisible – disability.

ACKNOWLEDGEMENTS

The author would like to thank Dr. Paul Hwang, Irene Elliott, and the staffs of Epilepsy Ontario and Epilepsy Toronto for help in preparing this chapter.

QUESTIONS FOR FURTHER THOUGHT AND DISCUSSION

1. Much of the stigma associated with epilepsy arises from the fact that seizures are frightening to watch. Should the public be shown what seizures look like? Should they become accustomed to seizures? Should they be taught first aid for seizures?
2. People with seizures who continue to drive have more accidents than the general population. They do not have more accidents than people with diabetes or heart disease, however. People with diabetes and heart disease *are* allowed to drive. None of these disease groups has as many accidents as disease-free young adult males. Should people with seizures be allowed to drive?
3. What are the economic and social costs of intractable epilepsy, a disorder that often starts in childhood, often lasts through life, and is found in 1 in every 500 people?

4. Teachers receive little or no training related to the disabilities they will meet in the classroom. Should education in disabilities be a part of teacher training?

SPECIAL TERMS

Absence seizures (formerly called a “petit mal” seizures) - Non-convulsive seizures that consist only of a few seconds of blank staring and immobility. The “three per second spike and wave” activity is seen in the EEG.

Anticonvulsant drug - A medication used to treat epileptic seizures. The term is something of a misnomer, since many seizures do not involve convulsions.

Complex partial seizures (formerly called a “temporal lobe” or “psychomotor” seizures) - Seizures in which epileptic “spiking” is often seen on both sides of the patient’s brain in the temporal-lobe areas. The patient does not convulse, and does not lose consciousness, but is out of contact with his environment. Consciousness is said to be “impaired”.

Epilepsies - A group of neurological disorders, characterized by spontaneous, recurrent seizures.

Intractable seizures - Seizures that resist control by anticonvulsant drugs.

Ketogenic diet - A high-fat diet, with adequate protein and very limited carbohydrate.

Seizure surgery - Therapy that involves brain surgery.

Seizures - Periods of self-sustained neural hyperexcitation.

Simple partial seizures (formerly called “cortical focal” seizures) - Seizures in which “spiking” is limited to one part of the patient’s brain. The clinical symptoms depend on the brain area activated by the epileptic discharge, and are very variable. Consciousness is retained.

Tonic-clonic seizures (formerly called “grand mal” seizures) - Dramatic convulsive seizures which involve a loss of consciousness and a convulsion which consists first of stiffening of the whole body (“tonus”) and then jerking of the whole body (“clonus”).

Vagal stimulation - Therapy that involves the implantation of an electrical stimulator that stimulates the patient’s vagal nerve.

MORE RESOURCES

Books and articles

American Journal of Medical Genetics (2001) 106. (This whole volume is devoted to the genetics of epilepsy.)

Burnham, W.M. (2002). *Epilepsy*. In L. Nadel, (Ed.), *Encyclopedia of Cognitive Neuroscience*. London: Nature Publishing Group.

Burnham, W.M. (1998). *Antiseizure Drugs*. In H. Kalant and W. Roschlau, (Eds.), *Principles of Medical Pharmacology* (pp. 250-261). New York: Oxford University Press.

Burnham, W.M., Carlen, P.L., and Hwang, P.A. (Eds.). (2002). *Intractable Seizures: Diagnosis, Treatment and Prevention*. New York: Plenum Press.

Dodson, W.E. and Pellock, J.M. (Eds.). (1993). *Pediatric Epilepsy Diagnosis and Therapy*. New York: Demos Publications.

Engel, J. and Pedley, T.A. (Eds.). (1997). *Epilepsy: A Comprehensive Textbook*. New York, Philadelphia: Lippincott-Raven.

Guberman, A. and Bruni, J. (1999). *Essentials of Clinical Epilepsy*. Woburn, MA: Butterworth-Heinemann.

Johannessen, S., Gram, L., Sillanpaa, M. and Tomson, T. (Eds.). (1995). *Intractable Epilepsy*. Bristol, PA: Wrightson Biomedical Publishing Ltd.

Levy, R.H., Dreifuss F.E., Mattson, R.H., Meldrum, B.J. and Penny J.K. (Eds.). (1989). *Antiepileptic Drugs*. New York: Raven Press.

Luders, H.O. (Ed.). (1992). *Epilepsy Surgery*. New York: Raven Press.

McLachlan, R. (1997). Vagus Nerve Stimulation For Intractable Epilepsy: A Review. *J Clin Neurophysiol*, 14, 358-368.

Rowan, A.J. and Ramsay, E.E. (1997). *Seizures and Epilepsy in the Elderly*. Boston: Butterworth-Heinemann.

Wallace, S. (Ed.). (1996). *Epilepsy in Children*. London, New York: Chapman and Hall Medical.

Internet Information

A number of websites provide useful information:

- 1) Epilepsy Ontario - <http://epilepsyontario.org>
- 2) Epilepsy Toronto – <http://info@epilepsytoronto.org>

- 3) Epilepsy Canada – <http://www.epilepsy.ca>
- 4) Epilepsy Foundation of America – www.efa.org

REFERENCES

Bazil, C.W. (2003). Epilepsy and Sleep Disturbance. *Epilepsy & Behavior*, 4, S39-S45.

Bialer, M., Johannessen, S.I., Kupferberg, H.J., Levy, R.H., Loiseau, P., and Perucca, E. (2002). Progress Report on New Antiepileptic Drugs: A Summary of the Sixth Eilat Conference (EILAT VI). *Epilepsy Research*, 51, 31-71.

Bishop, M.L. and Allen, C. (2001). Employment Concerns of People with Epilepsy and the Question of Disclosure: Report of a Survey of the Epilepsy Foundation. *Epilepsy & Behavior*, 2, 490-495.

Blumer, D. (2002). *Psychiatric Aspects of Intractable Epilepsy*. In W.M. Burnham, P.L. Carlen, & P.A. Hwang, (Eds.), *Intractable Seizures*. (pp. 133-147). New York: Plenum Press.

Boro, A. and Haut, S. (2003). Medical Comorbidities in the Treatment of Epilepsy. *Epilepsy & Behavior*, 4, S2-S12.

Bortz, J. (2003). Neuropsychiatric and Memory Issues in Epilepsy. *Mayo Clin Proc.*, 78, 781-787.

Burnham, W.M. (2002). *Epilepsy*. In L. Nadel, (Ed.), *Encyclopedia of Cognitive Neuroscience*. London: Nature Publishing Group.

Burnham, W.M. (1998). *Antiseizure Drugs*. In H. Kalant and W. Roschlau, (Eds.), *Principles of Medical Pharmacology*. New York: Oxford University Press.

Burnham, W.M., Carlen, P.L., and Hwang, P.A. (Eds.). (2002). *Intractable Seizures: Diagnosis, Treatment and Prevention*. New York: Plenum Press.

Edwards, H.E., MacLusky, N.J., and Burnham, W.M. (2000). The Effect of Seizures and Kindling on Reproductive Hormones in the Rat. *Neurosci Biobehav Rev.*, 24(7), 753-62.

Ellis, N., Upton, D., and Thompson, P. (2000). Epilepsy and the Family: A Review of Current Literature. *Seizure*, 9, 22-30.

Engel, J. and Pedley, T.A. (Eds.). (1997). *Epilepsy: A Comprehensive Textbook*. New York, Philadelphia: Lippincott-Raven.

Foldvary-Schaefer, N. (2002). Sleep Complaints and Epilepsy: The Role of Seizures, Antiepileptic Drugs and Sleep Disorders. *J Clin Neurophysiol*, 19(6), 514-521.

Gilliam, F., Hecimovic, H., and Sheline, Y. (2003). Psychiatric comorbidity, health, and function in epilepsy. *Epilepsy & Behavior*, 4, S26-S30.

Guberman, A. and Bruni, J. (1999). *Essentials of Clinical Epilepsy*. Woburn, MA: Butterworth-Heinemann.

Harden, C.L. and Goldstein, M.A. (2002). Mood Disorders in Patients with Epilepsy. *CNS Drugs*, 16(5), 291-302.

Hightower, S., Carmon, M., and Minick, P. (2002). A Qualitative Descriptive Study of the Lived Experiences of School-aged Children with Epilepsy. *J Pediatr Health Care*, 16, 131-137.

Isojarvi, J.I.T. (2003). Reproductive Dysfunction in Women with Epilepsy. *Neurology*, 61(S2), S27-S34.

Jahnukainen, H. (1995). *Psychosocial consequences of intractable epilepsy in adults*. In S. Johannessen, L. Gram, M. Sillanpaa, and T. Tomson, (Eds.), *Intractable Epilepsy*. (pp. 165-169). Bristol, PA: Wrightson Biomedical Publishing Ltd.

Johannessen, S., Gram, L., Sillanpaa, M. and Tomson, T. (Eds.). (1995). *Intractable Epilepsy*. Bristol, PA: Wrightson Biomedical Publishing Ltd.

Leonard, E.L. and George, M.R.M. (1999). Psychosocial and Neuropsychological Function in Children with Epilepsy. *Pediatric Rehabilitation*, 3(3), 73-80.

Luders, H.O. (Ed.). (1992). *Epilepsy Surgery*. New York: Raven Press.

McLachlan, R. (1997). Vagus Nerve Stimulation For Intractable Epilepsy: A Review. *J Clin Neurophysiol*, 14, 358-368.

Morrell, M.J. (2003). Reproductive and Metabolic Disorders in Women with Epilepsy. *Epilepsia*, 44(S4), S11-S20.

Morrell, M.J. (1998). Effects of Epilepsy on Women's Reproductive Health. *Epilepsia*, 39(S8), S32-S37.

Morrell, M.J. (1991). Sexual Dysfunction in Epilepsy. *Epilepsia*, 32(S6), S38-S45.

Motamedi, G. and Meador, K. (2003). Epilepsy and Cognition. *Epilepsy & Behavior*, 4, S25-S38.

Salholdt, L. (1995) Psychosocial consequences of intractable epilepsy in children. In S. Johannessen, L. Gram, M. Sillanpaa, and T. Tomson, (Eds.), *Intractable Epilepsy*. (pp. 153-163). Bristol, PA: Wrightson Biomedical Publishing Ltd.

Sanchez-Carpintero, R. and Neville, B.G.R. (2003). Attentional Ability in Children with Epilepsy. *Epilepsia*, 44(10), 1340-1349.

Tidman, L., Saravanan, K., and Gibbs, J. (2003). Epilepsy in Mainstream and Special Educational Primary School Settings. *Seizure*, 12, 47-51.

Vazquez, B. and Devinsky, O. (2003). Epilepsy and Anxiety. *Epilepsy & Behavior*, 4, S20-S25.

Viskontas, I., McAndrews, M.P., and Moscovitch, M. (2000). Remote Episodic Memory Deficits in Patients with Unilateral Temporal Lobe Epilepsy and Excisions. *J Neurosci*, 20(15), 5853-5857.

Wada, K., Kawata, Y., Murakami, T. et al. (2001). Sociomedical Aspects of Epileptic Patients: Their Employment and Marital Status. *Psychiatry and Clinical Neurosciences*, 55, 141-146.

Wheless, J.W. (2001). The Ketogenic Diet: An Effective Medical Therapy with Side Effects. *J Child Neurol.*, 16(9), 633-635.

Wheless, J.W. (1995). The Ketogenic Diet: Fa(c)t or Fiction. *J Child Neurol.*, 10(6), 419-423.

Williams, J. (2003). Learning and Behavior in Children with Epilepsy. *Epilepsy & Behavior*, 4, 107-111.

Table 1. Common seizure types (old names in parentheses)*

GENERALIZED SEIZURES (involve the whole brain)

Absence Seizures (Petit Mal)

Non-convulsive seizures which consist only of a few seconds of unconsciousness, blank staring and immobility. The eyelids may flutter. The EEG shows 3/second “spike and wave” activity all over the brain. The individual has no memory for the period of the attack.

Tonic-Clonic Seizures (Grand Mal)

Dramatic seizures which involve a loss of consciousness plus whole-body convulsions, which consist first of stiffening (tonus) and then of jerking (clonus). The EEG shows constant “spiking” all over the brain. The individual has no memory for the period of the attack.

PARTIAL SEIZURES (involve only part of the brain)

Simple Partial Seizures (Focal Cortical)

Sensory or emotional experiences, or contralateral jerking on one side of the body. Sensory experiences relate to the part of the brain involved, and may be auditory, visual, etc. The EEG shows “spiking” limited to one part of the brain. The individual is conscious, and will remember the period of the attack.

Complex Partial Seizures (Psychomotor)

The individual is conscious, but is out of touch with the surrounding world. There may be automatic movements, such as lip smacking and fumbling with the clothes. The EEG shows “spiking” in the temporal lobe. The individual has no memory for the period of the attack.

*modified from Burnham (1998)

Table 2. Disabilities often associated with seizures (percent of affected individuals likely to have seizures indicated in parentheses)*

GENETIC SYNDROMES

Tuberous sclerosis (>80%)
Sturge Weber syndrome (70-90%)
Fragile X syndrome (20-40%)
Rett syndrome (70-80%)
Down syndrome (2-15%)
Huntington's disease (5-10%)

NON-GENETIC SYNDROMES

Cerebral palsy (frequent, varies with type)
AIDS (13%)
Multiple sclerosis (5-10%)
Stroke (5-10% embolic, 2.5-25%, hemorrhagic)
Alzheimer's disease (15%)

* modified from Guberman and Bruni (1999)

Table 3. Co-Morbidities of Intractable Epilepsy*

COGNITIVE

- Lowered IQ
- Developmental delay (West and Lennox-Gastaut Syndromes)
- Selective cognitive deficits (related to partial seizures)
- Global memory problems due to seizures
- Global memory problems due to the sedative side-effects of anticonvulsant drugs

PSYCHOSOCIAL

- Low self esteem
- Psychiatric disturbances (found in at least 30%)
- AD/HD (20-30% of children)
- Personality changes due to the side-effects of anticonvulsant drugs

REPRODUCTIVE (ADULTS)

- Lowered fertility
- Lessened desire and responsiveness

* All of these vary from individual to individual. They will be sometimes present and sometimes absent.

Table 4. Anticonvulsant drugs which are more and less likely to cause cognitive impairment (have “sedative” side-effects)*

MORE LIKELY**

Phenytoin (high doses) (Dilantin)
Phenobarbital (Luminal)
Primidone (Mysoline)
Clonazepam (Rivotril)
Topiramate (Topamax)

LESS LIKELY**

Carbamazepine (Tegretol)
Valproate (Depakene, Epival)
Clobazam (Frisium)
Vigabatrin (Sabril)
Gabapentin (Neurontin)
Lamotrigine (Lamictal)

*modified from Guberman and Bruni (1999)

**Effects vary from individual to individual.