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Looking ahead

For many centuries, seizures and epilepsy were thought of in terms of magic and mysticism, demons and dread. Were the seizure events a divine blessing or a demonic curse? Even into the mid twentieth century, epilepsy, idiocy, and insanity were often spoken of as interrelated, for managements often dulled the mind and disturbed the functioning. Epilepsy, mental illness, and falling IQs were thought to be the product of a deteriorating brain, possibly inherited. The prejudices gave rise to such terms for epilepsy as “demoniac” and “lunacy” (moonstruck).

Advocates responded with denial that epilepsy was anything more than a brief interruption of normal function due to a sudden uncontrolled burst of abnormal electrical activity in the neural network of the brain, occurring in otherwise normal people who had no related intellectual or emotional disturbances.

The dawn of the modern age came with the findings of the Federal Commission on Epilepsy and Its Consequences of 1977. This exhaustive study spearheaded the emerging awareness that epilepsy is seldom “just seizures.” Roughly half of the individuals with epilepsy experience significant emotional problems and nearly half experience learning difficulties. Such associated problems often lead to later problems in employment and socialization, thus becoming more handicapping than just seizures, spikes, and medications.

The medical world now approaches epilepsy in terms of electroencephalograph (EEG) interpretations, medications, and antiepileptic blood levels, to which have been added surgery, stimulators, and diets. This is the medical model of epilepsy. The educational model portrays a child with epilepsy as one with academic struggles, epilepsy-related absenteeism, and class disruptions. The psychiatric model debates a picture of epileptic personalities and psychotic diagnoses. The employer model presents an insurance-risk, underproductive, and too often absent employee with epilepsy. Little is said of a language model of epilepsy. Few speak of the whole-person view of epilepsy and even less of the child model. Yet comprehensive epilepsy care needs to address the language, learning, emotional and later employment problems as well as the medical aspects of the condition.

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Problems associated with epilepsy in children

The brain is a dynamically growing, changing, and developing organ, especially in the early years of life. Seizures can inhibit, alter, or distort brain development as well as the related functions. The child's inherent potentials as well as handicaps can be altered dynamically by early interventions if given the chance (Svoboda, 1979; Svoboda, 1989).

The developing child

Seizures and seizure therapy interfere with brain functions by overactivation, interruption, inhibition, and destruction of vital functional pathways. Long or frequent attacks may alter neural circuits and neurotransmitter balances. This is seen especially in the limbic system.

In childhood, the brain and brain functions are developing over the first decades of life. Seizures and seizure care may interfere with the essential emerging skills at the age of epilepsy onset and thereafter. The immature brain has the capability of reorganizing around insulted areas, thus rendering the damage less impairing. This diminishes with maturation.

Epilepsy dysfunctions

Language

The incidence of speech and language problems in epilepsy is not known, for this area is often overlooked. Language problems are most apt to develop in individuals with epilepsy involving the left hemisphere and especially the temporal-frontal areas. Such problems may precede, occur simultaneously with, or follow seizures. The problems may be episodic or ongoing; if ongoing, they may result in a gradual loss of language abilities. The manifestations of such problems depend on the location of the seizure disturbance. Such manifestations may present as difficulties in recognizing speech sounds, in understanding what is being said, in expressing ideas, in remembering key words, or in speaking clearly and smoothly. Communication impairments often result in behavioral reactions. The manifestations may be subtle or may appear only under stress.

Learning

Roughly half of children with epilepsy have learning difficulties. Learning problems may show up as retardation, regression, specific learning disabilities, or underachievement. Retardation is most apt to appear in individuals with symptomatic generalized or multifocal seizures. Intellectual regression is most apt to be seen with medication reactions, or in symptomatic epilepsy, especially in epileptic syndromes, with progressive causes, or with overlooked seizures, such as with nighttime

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events. Specific learning disabilities are more apt to be seen with partial seizures and medication reactions, showing up as delays and distortions in reading, spelling, math, writing, and other academic areas. Attention and memory are most vulnerable, as is academic performance. Underachievement may result from overlooked problems, missed seizures, and adverse attitudes of those around the individual. Attention problems often relate to small subclinical discharges interfering with attention.

Behavior

One in every four children with epilepsy is reported to have behavior problems. Some estimates are at least twice that. Emotional and behavior problems may relate to both extrinsic factors and intrinsic factors. Extrinsic factors include the attitudes of the parents and siblings, the teachers and peers, and the resultant self-concept of the individual, which often reflects the experiences with other individuals. Intrinsic factors may result from the nature and location-lateralization of the epilepsy and related complications. Problems in understanding may result in any of autistic, avoidant, attention, or activity problems, whereas problems in expression may result in withdrawal, anger, and parental overprotectiveness. These may interfere with the development of socialization skills.

Vocation may be impaired more by the lack of adequate remediation of language, learning, and emotional factors than by the seizure stigma, the epilepsy restrictions, or the medications.

Factors and modifiers

Seizure type and location

The location and degree of brain involvement of the seizure pertains to the dysfunctions seen.

Generalized seizures involve the entire brain and all brain functions at the onset. Usually the individual is of relatively normal IQ, but often recurring generalized seizures or prolonged attacks may lead to some deterioration or depression of intelligence. Little bursts may interrupt attention.

Partial seizures are more often related to learning disabilities and language impairments than to retardation, although the majority of individuals with this group of seizures are relatively normal. Frequently recurring or prolonged partial seizures can be damaging to the nerve cells, leading to loss of specific functions, a dropping of intellectual processes, and emotional problems.

The laterality of the partial seizure discharge is important. Dominant-hemisphere discharges can impede language processing, while non-dominant-hemispheric discharges may impede non-verbal functions such as perceptual motor skills. Frontal involvement, especially early, may interfere with planning and organization skills

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and the development of executive functioning. Limbic involvement impairs memory and distorts emotional reactions.

Epileptic syndromes may present in idiopathic and symptomatic forms, all with some degree of related impairment. The symptomatic syndromes such as West's syndrome and Lennox–Gastaut syndrome are usually associated with severe impairments of language and learning with related severe behavior problems. Even the benign syndromes have been associated with subtle interruptions of function by special testing procedures.

Reflex epilepsies are those seizures triggered by specific sensory stimuli to the brain, such as flashing lights, video games, hearing speech, reading, concentration, etc. Academic efforts may occasionally trigger such events, resulting in brief seizures that interrupt the academic effort.

Transient cognitive impairments may result from epileptiform discharges in vital brain areas that may be neither large enough nor long enough to produce clinical symptoms but can interrupt, distort, or obliterate cognitive and emotional functions. The result may be interrupted attention, impaired recall, or distorted behaviors.

Appearance

Little seizures, like stares or small jerks, may easily be overlooked. Nocturnal events may be missed. By comparison, grand mal attacks are the center of attention, aversion, and fear by curious yet anxious onlookers. The stigma over the seizure spreads to include the child and all functioning attempts thereafter, with failures often blamed on the epilepsy if not on the medications used to treat the seizures.

Idiopathic versus symptomatic

All seizures are subject to the adverse attitudes and interactions of their environment. Consequently, language, learning, and behavior problems may be seen with all seizure types. However, people with symptomatic seizures are more apt to have problems and more apt to have more severe difficulties due to both the seizures and the underlying damage. Those with idiopathic epilepsy have only the effects of the seizures to cope with.

Gender

The male brain tends to be more lateralized in brain functions and more vulnerable to the insults that result in cognitive difficulties and language impairments. Gender differences appear to play a major role in how emotions are expressed, such that boys tend to act out and girls tend to withdraw. Teachers and parents display differences in expectations depending on the gender.

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Age at onset

A major difference between childhood epilepsy and adulthood epilepsy is the factor of brain (and brain function) development. Children are not just little adults. The earlier-onset seizures interfere more with the developing brain and brain functions, as well as perhaps distorting reparative efforts of plasticity. Yet earlier seizure insults may allow more recovery by utilizing alternative brain sites. This effect lessens with age.

To fully understand pediatric epilepsy is to understand developmental differences in children. This entails understanding the developing brain, the developing child, the developing language skills, the developing learning abilities, and the developmental stages of emotions and behaviors. The seizures, the medications, and the approaches differ according to the stage of development, as do the parents, the school, and the peers.

The common age divisions of childhood are divided into neonatal (0–2 months), infancy (2 months–2 years), toddlers (2–5 years), childhood (5–12 years), and adolescence (13–18/21 years of age). Adolescence is often alluded to as the typical onset of puberty. Infancy may be subdivided into early infancy and late infancy, correlating with the first and second years of life. Similarly, adolescence has been divided into early adolescence, implying the first three or four years, and late adolescence. There is some effort to carve out a preadolescence period of 9–12 years of age, representing the hormonal preparation for puberty. These stages parallel the development surges of the brain and the maturation of the neurotransmitter system, as they are manifest in seizure presentation, in cognitive approaches, and in related emotional reactions.

The International Classification of the Epilepsies movement in syndrome classification, speaking of neonatal seizures, early and late infantile-onset seizures, early childhood seizures (toddler period on into childhood), childhood seizures, and adolescent (juvenile) seizures, follows these same age periods. Some seizures, such as atypical absence, may emerge in the preadolescent period. Infantile spasms represent the infant brain's style of reacting to a variety of insults. Landau–Kleffner syndrome begins after infancy, when language has become sufficiently established to be noticeably lost. This syndrome is an early-childhood emerging seizure type occurring when language is being established. The various juvenile epilepsies tend to reflect the hormonal changes of puberty as well as the continuing maturation of the brain.

These periods also characterize major periods of development of the brain and of brain functions. They relate to stages of emotional development in children. Infancy is a perceptual-motor period, late infancy to the toddler age is one of language acquisition and establishment, childhood is a concrete age, and adolescence marks the onset of abstract thinking. Once begun, the developmental processes continue

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on into the next age. Similarly, one can see stages of emotional developmental challenges at each age. The trusting infant changes into the independent young toddler (“terrible twos”), to the friendly but hero-worshipping early childhood, to the more social-centered later childhood stage. In adolescence, there is a repetition of this evolution, including the terrible twos, as the child moves away from family dependence to self and society dependence.

The epileptic factors may distort developmental processes in the developing brain, yet the developing brain may be able to overcome such distortions by alternate development processes. The developed brain is essentially matured and thus not apt to experience developmental distortions. However, any damage done at this age is a loss, since the mature brain is limited in its ability to recover functions.

The brain is especially sensitive to insults in the earliest years of life. Recovery may be gained with age but in part this may be more of compensation and rewiring of the developing nervous system. Children with severe seizures in infancy are often more functionally handicapped than those with seizures of onset later in childhood and those with early-childhood-onset seizures may be more handicapped than those with adolescent-onset seizures. This is seen with other medical conditions; for example, children under three to five years of age with leukemia and other cancers are more sensitive to chemotherapy or radiation therapy than children over the age of seven.

Anticonvulsants can be affected by age. Metabolism can be more erratic in younger children. The so-called “safe” anticonvulsants like phenobarbital may impair nerve cell growth in infancy, although this is more in the balance centers (cerebellum) than in the thinking brain. Phenytoin can also have adverse effects in the younger child. The treating physician needs to weigh the risks of treatment versus the risks of non-treatment; if treatment is the choice, the physician must monitor the risks carefully and, if needed, provide early interventions.

Timing

Seizures and even seizure bursts during the performance of important tasks may result in failures and frustrations. Seizures at night may go unnoticed but render the child sleep-deprived the next day, thus impaired in function and behavior.

Frequency

Frequent seizures do not allow time for the brain to recover between episodes. Frequent seizures over a period of time may lead to changes in the neuronal circuitry and neurotransmitter balance, resulting in functional losses and delayed emergence of some psychiatric complications.

7 The whole-child model

Length

Lengthy seizures, as with status epilepticus, through the metabolic and circulatory changes in the brain as well as the release of neural excitatory amino acids, can be destructive to vital learning areas of the brain. Changes in cognitive functions (especially memory) and in behavior may be seen following a lengthy bout of epileptic status. The child appears less susceptible than the adult to the devastation of status.

Therapy

All antiepileptic drugs may help or hinder brain functions and emotions. This can occur if the blood–brain levels of the medication are excessive, if multiple drugs are used, or if the patient is overly sensitive to the drug. In children, the effect is not only on the brain but also on brain development. With new medications and seizure surgery, the seizures may be controlled but the associated language, learning, and behavior problems often remain.

Attitudes and interactions

Children do not experience seizures; they experience the reactions to the seizures. This is what shapes their self-concept, their learning experience, their communicative efforts, their social skills, and their preparation for adulthood. It is the experiences with parents and peers, educators and employers. From the time of the diagnosing of the epilepsy, the child's parents go through an evolution of their feelings and responses to the seizures, which affects their parenting as well as their view of the child. Teachers tend to expect less and blame all on the medications. Reduced expectations may result in reduced help for problems, which are then often overlooked or blamed on the medication. Children with epilepsy are still expected not to learn as well, not to tolerate as much stress, and not to be able to participate as fully as their classmates. Medical personnel interact with the young patient as an entity to be poked, prodded, punctured and pounded upon, and then ignored, as the physician addresses all questions and explanations to the worried parents, with the back often turned to the child. Whose seizures are they anyway? The parents'?

The whole-child model

Epilepsy in modern times is no longer only the control of seizures; it is the helping of the individual with epilepsy to gain his or her whole potential and abilities in terms of learning, socialization, communicating, earning, and enjoying. The need is to look beyond the epilepsy and see not only the seizure problems but also the child's potential.

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The physician expert in epilepsy treats the whole child, detecting early signs of emerging problems in education, behavior, learning preparation, and socialization; such a physician is very vigilant to clues of potential problems and responds rapidly. The treating team is not just the medical personnel, but also the parents and family, the educators, the community, and others involved in remediative services. The true management approach draws together all aspects of functional living into a unified therapy to help the child and ultimately to help the child help themselves. Treat the child, not the EEG, not the spikes, not the pills, not the seizure classification.

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Part I

Speech and language problems

2

Language: the challenge

Communication is the exchange of ideas and feelings. The main avenue of communication is language, in many forms, both verbal and non-verbal. Verbal language is the code of learning and underlies much of emotional development. If there is no means of communication, there exists limited learning and scant socialization. Language is among the highest of evolutionary skills. The developing brain may sacrifice other higher cognitive abilities to preserve the ability to communicate.

So intricate and fragile is this ability that many worldly woes emanate from gender differences, age dissimilarities, societal variants, and national differences in mother tongues. Far greater is the betrayal of an ability that is distorted or lost due to a brain insult or to epilepsy. The individual is frustrated, as are the parents, teachers, and social contacts.

Are speech and language problems a part of epilepsy?

Dr Frank M. C. Besag, noted, “If children have seizures, then they are particularly liable to language deficits.” He asked, “What is it about having seizures that should determine that language deficits specifically occur?” In the same discussion, Dr Bruce Hermann commented, “There is not much work on language” (Besag & Hermann, 2001).

Failure to communicate accurately or adequately underlies many of the problems experienced in families, in school, in social interactions, and in employment. Some children with epilepsy do not communicate well. They experience subtle and often variable language problems. They may misunderstand or they may be misunderstood. Such misunderstandings distort emotional development, educational efforts, and later earning. Some try to communicate but not clearly or smoothly, resulting in taunts and frustrations.

Amid the shock and confusion following the occurrence of a seizure, the turmoil of EEGs, scans, and other tests, the follow-up pills and blood tests, problems of speech and language are overlooked, although they may well be a part of the whole seizure problem.

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Speech–language problems occur in the population of children with epilepsy. In one clinic study, 20% were found to have language problems and 4% were referred for further evaluation. (Williams *et al.*, 1992). Is brain damage causing both the seizures and the speech–language problems? Do recurrent seizures lead to brain damage and subsequent language problems? Does the epilepsy directly cause the language dysfunction? The types of speech–language problems may be temporary, ictal or postictal, paroxysmal or temporary that may become permanent.

The international experience

One of every four children with epilepsy may have speech–language problems, according to a presentation on speech and language dysfunctions of epilepsy given at the International Epilepsy Conference in Oslo, Norway in July 1993. It was found that 51 of 200 children with seizures had speech–language problems in screening. The types of problems seen included loss of speech, receptive or expressive language problems, problems in recall, motor speech disturbances, and, most commonly, mixed problems.

The national experience

Language problems of epilepsy include word-finding difficulties, memory disturbances, as well as ictal and interictal language problems (Gordon, 1991). These may be of lateralizing and localizing value, especially in epilepsy surgery evaluations. Word-finding problems (43%) and forgetting names (31%) are the most common language findings in individuals with temporal lobe epilepsy. These problems may relate to the cause or the interactivity of non-traditional language areas in the anterior and inferior temporal lobe. The incidence is more than twice that found in a control population. Such individuals should at least be screened if not tested formally, for they can be helped.

The Kansas Epilepsy Center experience

At the St Francis (Wichita, KS) Epilepsy Symposium of 1987, David Henry and Marilyn Brown reviewed the findings of language and auditory processing deficits in mostly children referred to the Kansas Epilepsy Center. The referral population was 96% children, seen by a team of multidisciplinary specialists especially trained in epilepsy. The patients comprised 11% preschoolers, 54% grade scholars, 31% adolescents, and 4% adults. Although initially 85–89% were felt to have generalized seizures in referral, the evaluation showed that the incidence was reduced to 60% by adolescence. Partial seizures were diagnosed in 36–40% from the onset.

The types of speech–language problems included problems in language processing (53%), problems in articulation (3%), auditory memory problems (especially in short-term memory) in (61%), and problems other than memory (16%).

13 Overall conclusions

Fluency and voice prosody problems were detected in this group. The problems were subtle to profound in presentation. Relationships to seizure frequency, age of onset, and seizure intensity were noted. Alternations in the intelligibility of the speech (80%) and vowel prolongations (20%) were noted. Errors bore some relationship to the lateralization and localization of the seizure and the discernment of the physician examiners. Problems observed in these children included abnormal speech discrimination (23%), which was usually bilateral (18%). Other difficulties included problems in auditory processing (41%), which could be unilateral (20%) or bilateral (13%). The most common auditory-processing problems included peripheral auditory deficits of hearing and articulation, auditory attention, auditory memory, language-processing problems, and native cognitive abilities.

Audiometry examinations were performed for those with the slightest indication for such by history or by screening. Thus, new testing was performed most often in school-age children and least often in preschoolers. Middle-ear infections were common. In those tested, problems found included flat tympanogram in one or both ears (14%), and the placement of ear tubes in one or both ears (8%). Of the 60% of the entire group tested, 23% had reduced hearing in one or both ears, and 14% had moderate reduction in both ears. High-frequency problems were common and appeared to contribute to some reduction in understanding of speech in 70%; problems in discrimination in a noisy background were seen in 24% who had problems with one ear and in 9% who had problems with both ears.

Overall conclusions

The following section is based on Svoboda (1979).

Speech–language problems are more common than suspected yet often overlooked

Language problems may be seen with nearly all types of seizures but especially those involving the frontal-temporal lobes and adjacent areas.

Following a generalized tonic–clonic seizure, speech deficiencies and distortions may be part of the postictal confusion state. This gradually clears. Occasionally, children with uncontrolled frequent absence seizures, especially atypical forms, may present with blocks, halts, and jerky speech efforts. They may exhibit humming or they may get stuck on words. There are omissions in understanding. Rarely, their speech efforts may seem confused, slurred, or even deficient. This can be seen more commonly with excess stress. Jerky speech with momentary halts or inflections may be seen with minor motor seizures (the akinetic drops or myoclonic jerks).

With some simple partial seizures, blocks, halts, utterances, or distortions of pronunciation may be seen. Complex partial seizures, especially those involving the dominant temporal lobe, are more apt to disturb language processing with

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problems of anomia and auditory memory noted. Learning disabilities involving the auditory sphere, often with secondary perceptual motor problems, may be seen. Aphasias, arrests, and anomias may be noted, as well as problems in word fluency. There may be a deficiency or a loss of language. Speech utterances may occur ictally. Some individuals have a flat, emotionless style of speech, with a loss of prosody.

Reflex seizures may occur, such as in individuals who experience seizures when they try to read aloud. There may be a history of stuttering when reading as a child.

Epileptiform discharges of a subclinical nature involving language-processing areas may contribute to language-processing impairments.

All aspects of language have been associated with epilepsy-related problems

The five most common problems are (1) the loss of a smooth flow of spoken ideas, (2) a slow, slurred, labored, tick-tock speech pattern, (3) trouble with understanding others, (4) difficulties expressing ideas clearly, leading to misunderstandings by others, and (5) problems with remembering the names of people or items. However, still other problems have been seen in children with epilepsy.

There are multiple causes for language problems in epilepsy

With seizures, there are four ways in which language can be impaired, including (1) the underlying brain insult that causes both the seizures and language disturbance, (2) the seizures themselves, (3) episodic, epileptic short-circuits disturbing language processing, and (4) medication reactions. A negative environment in which the child with epilepsy struggles to develop may aggravate these.

The underlying brain insult may produce both seizures and language problems. The insult may be an underlying brain irritant, such as a focal chronic virus infection, an autoimmune problem, a tumor, or a vascular malformation. If the child has an active process of the brain, the speech may continue to deteriorate. However, if the seizures eventually improve or become controlled, the language function also may then improve or recover. Occasionally, the speech problem may develop before the seizure difficulties.

The seizures, especially if prolonged or repeated, may cause a loss of a language skill (especially in an adult) or a distortion or depression of language development (in a child) with or following the seizure. Speech and language problems may be seen with obvious seizures or with a left-hemispheric disturbance, especially involving the left frontal-temporal area, with or without any epileptic symptoms.

The seizure discharges themselves may short-circuit language processes and, if larger or longer in duration, may produce more recognizable seizure symptoms. The electrochemical seizure potential discharges, not great enough to produce seizures, still short-circuit language processes without a history suggestive of epilepsy.

15 Overall conclusions

The antiepileptic drugs used to control the seizures may impair speech and sometimes also language, especially if used at higher doses or if more than one drug is used. Some patients are sensitive at therapeutic blood levels. Sometimes, the seizures appear to be controlled but the speech problem persists. Occasionally, an increase or an alteration of the anticonvulsant within the normal therapeutic range may overcome the speech difficulties.

The course of the language problem may be variable

Speech and language problems may precede the onset of seizures, may develop along with the seizures, or may develop some time later after the onset of the seizures. Such problems may be ongoing or they may occur in episodes, often linked with the appearance of seizures clusters. If they are an ongoing problem, the severity may wax and wane. Speech and language problems, like learning problems and emotional problems, can be brought out by stress, by poor seizure control, or by inappropriate use of anticonvulsant medications. Nocturnal seizures may result in daytime struggles. Of all these, stress is the most apt to bring out a speech or language problem. The child with a seizure coming from the left half of the brain is at the most risk for such problems.

The onset may be before, during, or following the onset of the seizure disorder. The symptoms may be temporary, lasting hours to weeks, may recur as brief episodes, or may present as a progressive problem. Symptoms may present as an aura to seizure, as part of seizure, or as an aftereffect of seizure, or they may emerge as a problem between seizures.

A group of special language-regression syndromes exist as a special challenge

There are a group of seizure syndromes that are especially prone to emerging in the first decade of life. The onset of seizures, sometimes fairly resistant to medication, may be accompanied by the loss, often severe, of language. The frustrated patient often develops major behavior problems. The seizures may be outgrown, after which language may or may not return. Specific epilepsy language combination syndromes include the acquired agnosia/aphasia with epilepsy syndrome of Landau-Kleffner, the sleep spike-wave status syndrome, absence/minor motor status, protracted partial complex status, and Rasmussen's encephalitis, the latter often having a dire outcome.

Language problems may underlie some learning and behavior problems of epilepsy

A variety of emotional reactions to the frustration of language problems are noted. Children with a receptive aphasia may be described as autistic, as having attention deficit/hyperactivity disorder (ADHD), or as withdrawing, whereas children with expressive aphasia are more apt to be shy or to display temper reactions. Similar

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tendencies are seen in elderly people after strokes. Problems of communication may underlie social difficulties and learning problems (Cantwell *et al.*, 1980, 1981; Gordon, 1991).

The challenge

Considering three aspects, i.e. language as the base for learning, language as related intimately to emotions, and epilepsy as a potential distorter of language, the physician, teacher, or therapist should consider the relationships between speech–language and epilepsy.

This leads to three questions to be considered in approaching the child with epilepsy: how does epilepsy interfere with language, what type(s) of epileptic language problems are potentially present, and how may abnormal language functioning affect overall functioning as well as seizure control?

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