

Syndrome of Acquired Aphasia with Convulsive Disorder in Children

William M. Landau, M.D. and Frank R. Kleffner, Ph.D.

CERTAIN OF THE CHILDREN who are evaluated and taught at the Central Institute for the Deaf are classified as aphasic because they present a specific deficit in the ability to use speech and language.¹ For most of these children the deficit seems to be congenital, since they have failed to acquire the ability to use speech and language normally.² Some of them, however, have acquired the ability to use language in an apparently normal fashion and have subsequently lost it. Six such children have been seen in the last two years. One of the six became aphasic and hemiplegic after a severe head injury; the remaining five developed aphasia in relation to a convulsive disorder. These five cases are reported here.

CASE HISTORIES

Case 1. A white male, third of four children, was born in April 1948. Pregnancy and birth history were unremarkable. He was a healthy infant, sat at about six months, and walked before he was a year old. He learned to talk normally at 15 to 18 months. Further behavioral development was also unremarkable. In 1952 when the boy was four, he had one nocturnal generalized seizure. When he was five (July 1953), he developed several furuncles over his face. A few days later he fell in the yard and was found in a semiconscious condition. He staggered into the house and vomited several times. Following treatment with penicillin, it became apparent to the family that the child was having difficulty understanding what was said to him, a defect interpreted as "plain stubbornness." His speech also became garbled.

He was hospitalized in August 1953. His electroencephalogram (figure 1A) showed a generalized spike dysrhythmia, most prominent in the temporal leads bilaterally. Neurologic examination was not remarkable except for the deficit in speech. He was considered to have an aphasic disorder, primarily receptive. Three lumbar punc-

tures revealed two white cells and one red cell per cu. mm.; glucose, 47 mg. per cent; protein, 16 mg. per cent; chloride, 112 mEq. per cent; Lange, negative; Wassermann, negative. The patient was given Dilantin and phenobarbital medication and the seizures were controlled.

When he was first seen at the Central Institute for the Deaf in October 1953, he showed very little use or understanding of speech. His hearing seemed unimpaired. Formal auditory tests were not given since he responded quickly and consistently to a variety of soft sounds. He behaved in a stable and intelligent fashion, although he was clearly frustrated by his difficulties in understanding and using speech. Arrangements were made to have him attend the speech clinic; however, two weeks later the mother called and reported that the child was speaking and understanding normally and that he had been admitted to kindergarten. A repeat electroencephalogram at this time (figure 1B) showed no significant change except that the temporal spikes were somewhat more prominent on the left.

Five months later (March 1954) he had a seizure, with unconsciousness and with clonic jerking in the right hand and arm. In July 1954 he had twitching in his right face, looked pale, and went to sleep. During the rest of the summer he had several episodes of twitching of his right face. He would grab at his right cheek and then sit and rest. On one occasion he said that he was nervous. On other occasions he would take off his right shoe to see if his foot were injured and say, "It feels like my nerves." That fall, during a period of about a week, his comprehension and production of speech diminished rapidly to a level worse than that in 1953. There were no obvious seizures at this time.

In December 1954 he was seen in the pediatric clinic. An electroencephalogram again showed a

From the division of neurology and the Beaumont-May Institute of Neurology, Washington University School of Medicine, and the division of speech pathology and speech correction, Central Institute for the Deaf, St. Louis, Missouri.

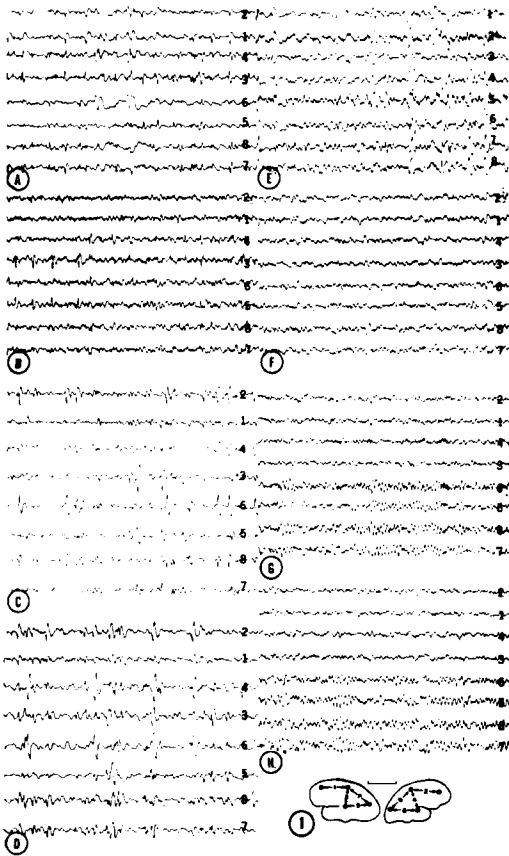


FIG. 1. Serial electroencephalograms in case 1. A. August 10, 1953. B. October 28, 1953. C. December 11, 1953. D. January 7, 1955. E. March 14, 1956. F. April 18, 1956. G. May 23, 1956. H. August 1, 1956. Descriptions in text.

generalized spike dysrhythmia (figure 1C). Physical examination was essentially unremarkable. Skull films were normal. Roentgenograms of the extremities showed "mild bone retardation." Fasting blood sugar was 74 mg. per cent, nonprotein nitrogen 25 mg. per cent, calcium 12.2 mg. per cent, and phosphorus 4.8 mg. per cent. Hemogram and Kahn test were normal. Urinalysis with examination for porphogens and phenylpyruvic acid was normal. In January 1955 he was hospitalized. Lumbar puncture revealed an opening pressure of 210 mm. of water. There were one polymorphonuclear leukocyte and six lymphocytes per cu. mm. Cerebrospinal fluid protein was 27 mg. per cent, glucose 52 mg. per cent, and Wassermann negative. Psychological examination indicated that the child was not mentally deficient. His electroencephalogram was unchanged (figure 1D).

Later in January 1955, he was seen again at the Central Institute for the Deaf. His speech was primarily jargon, although occasionally he uttered appropriate words when he was angry or excited.

His voice quality and inflection were normal. He could imitate voice pitch and quality changes in the voice with facility, indicating that he had no significant loss of hearing. He was enrolled in the speech clinic but the lessons were discontinued again, this time because of transportation difficulties. His speech did not change during the next year, although there were no seizures. He had increasing difficulty getting along with his siblings and other children.

Neurologic examination in February 1956 showed, in addition to the speech disturbance, some flattening of the lower right face. This was not noted at subsequent examinations. The child was right-handed and right-eyed. His electroencephalogram in March 1956 was unchanged (figure 1E). He was given Paraldone, in addition to Dilantin and phenobarbital. An electroencephalogram in April (Figure 1F) showed a mild fast and slow dysrhythmia but without paroxysmal spikes. His mother reported improvement in behavior and some better understanding of speech. The electroencephalogram was even better ordered in May and August (figure 1G and H). Speech continued to improve with special instruction and he was admitted to the first grade of public school on a trial basis in September 1956. He was generally well behaved except when there were changes in his daily routine, such as might be occasioned by visitors or a trip away from home.

The patient's eldest brother (case 2) had a similar disturbance when he was younger. The patient's father had had two nocturnal grand mal seizures following a herniorrhaphy at age 47 and another seizure two years later. The father's electroencephalogram showed a mild degree of fast activity without focalization. Neurologic examination has been normal. The mother aged 41 has had no seizures. Her electroencephalogram showed a mild fast dysrhythmia. Her brother has had some type of blackout spells. An 11 year old brother of the patient had a mild mixed slow and fast dysrhythmia without focalization, but he has shown no clinical symptoms. A 15-month-old sister had a normal electroencephalogram. No other relatives have had convulsive or speech disorders.

Summary of Case 1. An eight year old boy with a family history of convulsive disorder developed aphasia on two occasions, once at age five following a series of convulsions, and once without apparent relationship to clinical seizures at age six. Electroencephalograms showed a generalized spike dysrhythmia. In the first episode, speech recovered shortly after anticonvulsant therapy was started. On the second occasion, speech improved with speech therapy as the electroencephalogram improved with increased drug therapy.

Case 2. The first born sibling of case 1 was born in November 1940. Birth, early development, and learning of speech were entirely normal. There were no febrile seizures in infancy. On two

or three occasions at about age eight, usually when he had a sore throat and fever, there were brief episodes of unawareness and eye-rolling. When he was nine years old he fell from his bicycle and bumped the back of his head. Soon after this he was struck on the right side of his head by a swing. On neither occasion was he unconscious or ill. A few weeks after these accidents the parents noticed that he seemed to have trouble with his memory and hearing. He had trouble producing speech, and his mother stated that he could describe some things he couldn't name properly. There was some inconsistency in his response to sounds. The boy, now 15, states that at the time he understood what was said to him but couldn't think of the words he wanted to say.

In August 1950 he was hospitalized and a report at that time stated: "The patient seemed to be attentive to his environment, but would not respond to any spoken command. However, if a loud noise were made to the side or behind the patient, blinking of the lids was noted. There was no tendency to turn his head to the site of the noise. He did not respond to written commands, although he could mimic acts done by the examiner, such as squinting the eyes, protruding the tongue, and so on." Lumbar puncture showed initial pressure of about 100 mm. of water and chemical constituents were normal. Routine laboratory tests including hemogram, urinalysis, and serum calcium, potassium, and sodium were normal. An electroencephalogram showed right hemispherical spike discharges, more prominent in the central leads (figure 2A). Without specific therapy the patient's speech returned in a week or two while he was in the hospital.

The parents neglected to bring the boy back for further examinations until he developed seizures 15 months later. While going to sleep one night he had a frightening dream in which he was being chased. During this time there was a loud buzzing in both ears; he was frightened and ran to his parents' bedroom. There he fell into a generalized convulsion. Two similar attacks occurred in the next few weeks. He was started on Dilantin and phenobarbital medication and since then has had no convulsions. Occasionally he still has a buzzing in his ears about the time he is going to sleep.

In November 1951 (figure 2B) the electroencephalogram showed a mild definite slow dysrhythmia, without focalization or spike discharges. The electroencephalogram was improved in June 1952 (figure 2C). Further improvement was noted in October 1953 and March 1956 (figure 2D and E). Neurologic examination in March 1956 was entirely within normal limits. He was right-handed; eyedness was indeterminate. He was then 15 years old and doing well in the tenth grade.

Summary of Case 2. A 15 year old boy with a family history of convulsive disorder had some minor seizures at age eight. At age nine, following some minor head injuries without

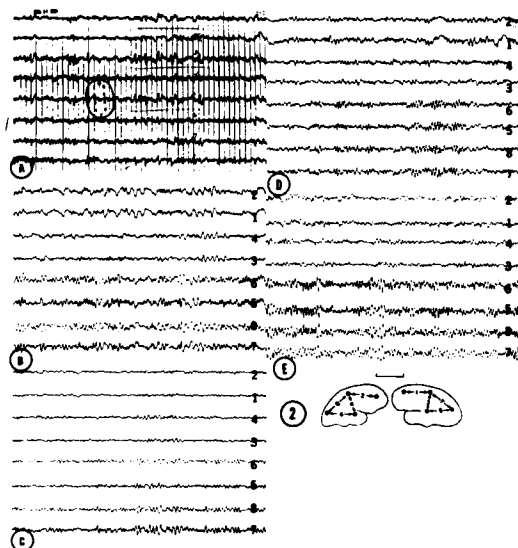


FIG. 2. Serial electroencephalograms in case 2. A. August 14, 1950. B. November 12, 1951. C. June 19, 1951. D. October 28, 1953. E. March 29, 1956. Monopolar leads as indicated in A. Bipolar leads as in diagram in other tracings.

serious clinical signs or seizures, he became aphasic for a period of several weeks. At the time there were spike discharges over the right hemisphere in the electroencephalogram. Over a year later he had three generalized seizures, with aura of fear and tinnitus. His clinical condition has remained good without seizures on anticonvulsant medication.

Case 3. A white female, the second of two children, was born in February 1950. There was no family history of convulsive or communication disorder. Pregnancy and labor were uneventful. The baby breathed well at birth and developed normally. She gained weight well and was quite active. She sat at six months, walked at about a year, and was talking between a year and two years. By the age of three she was speaking in full sentences. During the winter of 1953-54 the mother noticed that the child had rather regular involuntary jerkings of her left arm. In May 1954 there was a nocturnal grand mal seizure. Several months later some involuntary jerkings of the left side of the face without loss of consciousness were noted. Five months later there was another nocturnal seizure. In November 1954 the child was seen by a neurologist. Skull roentgenograms and neurologic examination at that time were normal. The electroencephalogram in December (figure 3A) showed a diffuse spike dysrhythmia. Dilantin therapy was begun and no generalized seizures have occurred since then.

By early 1955 (age five) the mother became aware that the patient did not comprehend stories when they were read to her and was only interested in

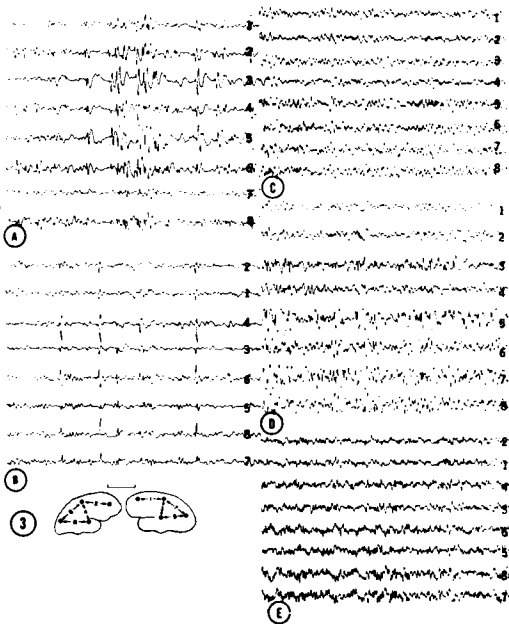


FIG. 3. Serial electroencephalograms in case 3. A. November 22, 1954. B. March 15, 1956. C. May 25, 1956. D. September 18, 1956. E. December 18, 1956.

the pictures. As her comprehension diminished her speech production became progressively worse until there was practically no speech or understanding when she was first seen at the Central Institute for the Deaf in November 1955. The mother reported that the girl seemed to understand or use an occasional word at home. Formal auditory testing was not done, but her hearing seemed well within the normal range. She imitated silent mouth actions by the examiner and added sound to the actions when the examiner added sound. She turned to a low whistle, to her name, and to voices and noises in the hall outside the examiner's office. The mother reported that when the girl was playing outside she could be called by voice or by whistle. Any attempt to speak resulted in jargon, with normal vocal quality and inflection but without words. She printed several letters on the blackboard spontaneously. The mother reported that about a year previously the girl could print her name and her brother's name correctly. During the examination she could not do this on oral request, but when the examiner printed these names on the blackboard she very readily "copied" them both correctly, though she actually paid little attention to the names printed by the examiner. Psychometric examination at this time using the Advanced Performance Test yielded an IQ of 117.³ Special speech instructions were begun at this time.

During the neurologic examination in March 1956 the child was alert, friendly, and cooperative. She was right-handed and right-eyed. Although she could imitate movements of the examiner, she

could not imitate simple sounds such as "Ah" unless her hand was placed on the examiner's larynx. She could sound consonants and vowels properly from letter symbols in her speech book. The remainder of the examination was normal. Fasting blood sugar was 79 mg. per cent, calcium 10.6 mg. per cent, and phosphorus 4.7 mg. per cent. The electroencephalogram in March 1956 continued to show generalized spike discharges (figure 3B). During a clinical examination ten days later, some twitchings about the left side of the mouth were noted. In addition, there were several spells of blank expression with lip smacking. There was slight flattening of the lower left face at rest. Dilantin medication was increased and phenobarbital was added.

In May the child was much improved. No minor seizures were observed during the interval; she had about 50 useful words. An electroencephalogram showed a moderate degree of slowing but no spike discharges (figure 3C). There was only slight improvement in speech over the summer, and the electroencephalogram showed frequent spike discharges in September (figure 3D). Paraldone was started.

By December 1956 her speech had improved further, she could understand some short sentences, and she was more attentive in the teaching process. Behavior was good, there were no seizures, and the electroencephalogram (figure 3E) showed fast and slow activity but no paroxysmal spikes.

Summary of Case 3. A six year old girl had the onset of myoclonic jerks at age three and grand mal seizures at age four. After successful medical treatment for the seizures had been instituted, she gradually developed aphasia. Improvement in speech occurred with special instruction and additional medication, and seemed to be inversely correlated with the presence of generalized spike discharges in the electroencephalogram.

Case 4. A white female, the first of two children, was born in July 1948. There was no family history of convulsive or communication disorder. The pregnancy was uneventful. The labor was not considered abnormal, although there was a face presentation. Labor lasted 17 hours, with two hours of hard labor. The infant's general condition was good, although there was a slight swelling of the forehead.

She ate and gained weight well. She sat at six months and walked at 13 months. She was toilet-trained at two years. Speech developed during her second year, and she was able to say short sentences and said her prayers at night by the time she was three. The child suffered a fall without unconsciousness or evidence of serious injury when she was three and a half years old. A few months later in June 1952 the parents noted that she ceased to respond to calls, although she still talked well. Her ears were examined and were reported to be normal. During the summer her speech

gradually disappeared; understanding seemed to go first. For several days she was quiet and withdrawn, but there were no seizures of any kind.

In October 1952 she was hospitalized. Except for her difficulty in communication, physical examination showed no apparent abnormality. Hemogram and urinalysis were normal as was the skull roentgenogram. Electroencephalograms on several occasions revealed generalized spike wave discharges from all leads during most of the recording.* The child scored an IQ of 137 on the Merrill-Palmer test. It was felt that she displayed some negativism but that she was generally warm and affectionate and the examiners were impressed that she was well integrated in motor performance. It was noted that at times she appeared quite alert and at others somewhat confused. Some facial grimacing was also noted. At discharge from the hospital she was given Paradione for a trial period of two weeks.

The parents report that at the time of the patient's discharge from the hospital she said a few words but these disappeared again in a week or so. During the winter of 1952-53 the child seemed frustrated and was hard to manage, but did not withdraw from social contact. There was disagreement among many physicians as to whether she was deaf.

An electroencephalogram in June 1953 (figure 4A) again showed generalized spike wave discharges most prominent in the temporal leads. During 1953-54 she went to a school for the deaf. She did very poorly but there was no behavior difficulty. In 1954 she was seen by a neurosurgeon. Her electroencephalogram was unchanged except that the spike activity occurred "predominantly in the temporal lobes and more so on the right." Spinal fluid examination and pneumoencephalogram were entirely normal. Psychiatric evaluation was summarized: "The behavior would suggest psychogenic regression but its severity would equal childhood schizophrenia while her retention of warm affect is against that diagnosis."

Some time between 1952 and 1954 the child was placed on Dilantin medication possibly with some phenobarbital. For five or six days while she took these drugs she had spells during which her head shook toward the left and she was unsteady; afterward she was drowsy for an hour or so. The medication was stopped and except for the trial of Paradione she has taken no other anticonvulsant drugs.

The patient was first seen at the Central Institute for the Deaf in November 1954 at age six. She was exceptionally alert and friendly during the entire examination but showed no understanding of speech. She responded to spoken commands with a puzzled expression and a shrug of her shoulders or she ignored them completely. Little that was going on about her escaped attention.

*These records were shown by Dr. B. K. Bagchi at the meeting of the American Electroencephalographic Society, Atlantic City, N. J., 1956.

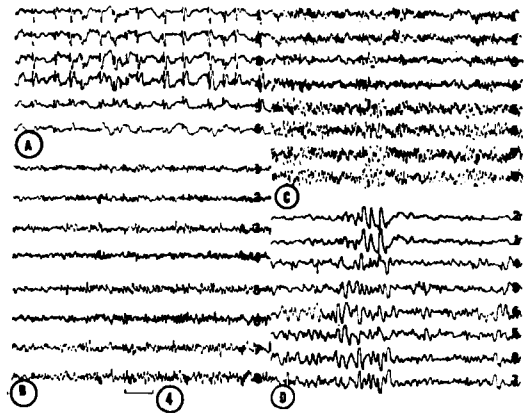


FIG. 4. Serial electroencephalograms in case 4 (A, B, and C) and single record in case 5 (D). A. June 24, 1953. B. April 18, 1956. C. September 19, 1956. D. April 1, 1955. Monopolar leads in A: 1, 3, and 5 are left frontal, vertex, and occipital leads; 2, 4, and 6 are homologous leads on the right. Other records are derived bipolarly, as in other figures.

She seemed to respond to an occasional sound but such responses were never repeated when attempts were made by the examiner to verify them. She played with noisemaker toys and seemed to enjoy them but she did not respond to their sounds when she was engaged in some other activity. On several occasions during the examination she seemed to respond to slight incidental noises, though most of the time she appeared completely oblivious to any sound including extremely loud noises. Because of this inconsistency of response, routine audiometric tests were not attempted. Indirect measurement of hearing was attempted through the conditioning of electrodermal responses to sound (EDR or PGSR), but it was virtually impossible to establish the conditioning on which this test depends. She named a few objects or pictures with poorly articulated words, although she showed no ability to understand them through hearing or lipreading. Her voice was high pitched and lacked natural quality and inflection, resembling the tone which is characteristic of some deaf children. Psychometric examination in November 1954 with the Advanced Performance Test yielded an IQ of 164.

She was enrolled in the Central Institute for the Deaf full-time classes for aphasic children in September 1955. By the end of her first semester in school she had begun to respond to sound quite consistently. She could write the letters for any individual speech sound spoken softly some distance behind her. By the end of her first school year of class work she had a speaking and understanding vocabulary of approximately 200 words. She could read and quickly memorize paragraphs of six or seven simple sentences and could answer simple questions on such paragraphs through both hearing and lipreading. Audiometric tests given in February 1956 showed normal threshold for

pure tones in both ears. In May 1956 near the end of her first year at the Central Institute for the Deaf she was given the Hiskey non-verbal test of learning aptitude and her LQ (learning quotient) was 102.4⁶.

Neurologic examination in March 1956 revealed a bright, friendly, hyperkinetic but cooperative child who was right-handed and left-eyed. She showed no neurologic deficit beyond her difficulty in communication. Vestibular response to caloric stimulation (40" at 18° C) was within normal limits. An electroencephalogram taken in April 1956 showed a mild mixed fast and slow dysrhythmia without any spike discharges (figure 4B). This pattern persisted in September 1956 (figure 4C).

Summary of Case 4. An eight year old girl developed normally until several months following a minor head injury at age three and a half. She gradually became aphasic at age four, and during the same period of many months her electroencephalogram contained many generalized spike wave discharges. A few minor left-sided seizures occurred while she was taking Dilantin. Her communication has improved with intensive training in speech and language and with the spontaneous disappearance of the paroxysmal electroencephalographic discharges.

Case 5. A white female, the first of three children, was born in December 1943. There was no family history of convulsive or communication disorder. The pregnancy and labor were uneventful. The child fed well and gained weight properly. She sat at six months and walked at 13 months. By the time she was two or two and a half years old she was talking in short sentences. Her general development was good. Although her speech is said to have been "babyish" and one teacher suggested the possibility of a hearing deficiency, the first grade teacher stated that the patient had the best vocabulary in the class. There were no serious behavior difficulties and the child got along well with her contemporaries.

At the end of the first grade in 1951, about a month after recovery from the mumps, she lost the ability to respond to speech and noises. This occurred over a two-week period during which speech practically disappeared. She was given some intramuscular injections of vitamins. In two weeks speech and understanding had recovered. No seizure of any kind was noted at the time.

⁶This represents a considerable difference from the I.Q. of 164 on the Advanced Performance Test administered earlier. There seems to be a rather consistent discrepancy between the scores on the Advanced Performance and the Hiskey for aphasic children (both congenital and acquired), whereas deaf children score more nearly the same on both tests. The major difference between the tests is that the Hiskey test is heavily weighted with memory items, particularly items requiring memory of sequences. Since the Hiskey test has been administered infrequently at the Central Institute for the Deaf, this observation has not been subjected to statistical evaluation.

Many consultants examined the child and most of them felt that she suffered from an "emotional block;" psychotherapy for both parents was undertaken on several occasions. The child's intellectual capacity was observed to be above average.

In the fall of 1951 the patient entered the second grade and did well until the end of that school year. Speech and understanding were lost again, more rapidly this time, in a day or so. She appeared generally well, although it was obvious that she was quite frustrated by her inability to produce anything other than "jabber noises." During this summer enuresis occurred.

She started in the third grade in the autumn of 1952, but she had little speech and hearing and her academic work was poor. She was not a disciplinary problem, however.

In the spring of 1953 she had a grand mal convulsion early one morning. Except for tiredness she was unaffected when she awoke. In May 1953 her physical examination was unremarkable and she seemed to hear. Ear, nose, and throat examinations and skull roentgenograms were normal. Several examiners were impressed with the possibility of a psychiatric disturbance. A generalized spike-wave dysrhythmia was observed in the electroencephalogram. After some difficulty in conditioning electrodermal responses to sound, it was possible to elicit responses within the limits of normal threshold. Mebaral medication was instituted and no more seizures have occurred.

The patient was first seen at the Central Institute for the Deaf in February 1954 at age ten. She did not speak or understand speech. There was no evidence of hearing during the entire examination. She behaved in an intelligent fashion and it was possible to communicate with her fairly well through writing. She read and followed correctly a number of simple written instructions, and was able to write answers to a number of simple written queries. She used one or two very poorly articulated words during the examination but clearly preferred writing to speaking. Near the end of the interview, she was sitting in a classroom where other aphasic children were being taught and appeared extremely interested in the class work. Her mother motioned to her that it was time to leave and the patient responded "you go, I stay." This was her only good speech and her only utterance of more than one word during the entire initial interview.

A few days later she entered the Central Institute for the Deaf classes for aphasic children. During her first few months in the school she relied heavily on writing for communication. She would often write for her teacher "I cannot hear" or "Talk is hard. I will never learn to talk." Occasionally she would write the question, "Why can't I hear?"

A conventional audiometric test was attempted two weeks after she began her schooling at the Central Institute for the Deaf, but her responses were too inconsistent for reliable evaluation.

In April 1954 she scored an IQ of 115 on the Advanced Performance Test. In December, after slightly less than two semesters in the school, a successful audiometric test showed normal thresholds of hearing. The test had to be administered carefully, however, for if two stimuli were presented in quick succession she became confused and her responses became erratic.

By January 1955 she had begun to progress rapidly. She was highly motivated and eager to improve her vocabulary and language. Those words which she knew could be spoken to her in a whisper. Those words which she did not know, she seemed not to hear at all. Her own report concerning words and phrases which she did not understand was that she did not "hear" them. Later she reported that when she first entered the Central Institute for the Deaf she had been able to hear music and noises but that when people talked she sounded like "blah, blah, blah."

Neurologic examination in March 1955 revealed a bright cooperative child, right-handed and right-eyed, without evidence of neurologic deficit other than the communication disorder. Vestibular responses to caloric stimulation (40" at 18° C) were somewhat diminished. In April 1955 an electroencephalogram showed a mild slow dysrhythmia with paroxysmal bursts and temporal spikes during and after hyperventilation (figure 4D).

At the end of the 1955-56 school year, she could read, write, and converse with minimal difficulty. Her major remaining difficulty was in unfamiliar and abstract vocabulary.

During her fifth semester of special instruction at age 12 (March 1956), she was given the American Standard School Achievement Test and performed at the grade equivalent of 6.1. She entered grade seven in a public school in September 1956 and has done well.

Summary of Case 5. A 13 year old girl developed normally until age seven when she became aphasic for a period of two weeks. She was subsequently well for about a year when she became aphasic again, this time without immediate recovery. Her condition remained unchanged for another year when she had a single generalized convulsion. Since then, with special speech training and anticonvulsant medication, her recovery has been nearly complete.

DISCUSSION

After apparently normal acquisition of speech and language, all of these children developed aphasia for periods ranging from a day to several months. The symptoms then persisted from two weeks to several years. In addition, the children had a variety of convulsive manifestations: grand mal, partial, petit mal, and myoclonic seizures.

It does not seem reasonable to infer that this

language disturbance is simply a variety of postictal Todd's paralysis, since there was poor correlation between the incidence of clinical seizures and the onset of the disorder of communication. Moreover, the convulsive phenomena that did occur before, during, or after the language disturbance were readily controlled medically or required no therapy (case 4). The degree of paroxysmal disorder in the electroencephalograms of these patients, however, did tend to correspond with the severity of language disturbance, although this relationship was not perfect. (For example, the electroencephalogram in case 1 was unchanged over the period of recovery from the first episode of aphasia.) The electroencephalographic disturbances were usually bilateral, often more prominent in the temporal leads. Temporal localization was also suggested by the focal character of many of the seizures and by the transient facial pareses seen in two patients.

Where there was lateralization of paroxysmal brain activity, the side more involved was not necessarily the same as that inferred to be "dominant" on the basis of handedness and eyedness. Roberts⁵ has shown extensive data in adults that handedness and dominance for speech are, indeed, practically independent variables.

Although our patients show no evidence of structural brain lesions,⁶ it may be suggested that persistent convulsive discharge in brain tissue largely concerned with linguistic communication results in the functional ablation of these areas for normal linguistic behavior. This thesis is supported by the good performances on non-verbal intellectual tests. No behavior similar to the clouded state of consciousness defined as "petit mal status" was seen. Further support is gained from the favorable course of the aphasic symptoms in these children. Although we believe that intensive speech training and anticonvulsant medication are both important in this improvement, there has been improvement from some episodes of aphasia with either form of therapy or with none.

The maintenance of normal or superior intellectual capacity in our cases helps to distinguish them from those with more malignant organic brain disease. The latter have probably not been seen in our series because our case finding agency is a school. Where language defect is an island of behavioral deficit, the distinction of acquired aphasia from a "primary" psychotic disorder is obvious. However, when the child has a regressive behav-

ioral reaction to the loss of communication, this separation may be difficult. The electroencephalographic disturbances among this group of aphasics are a useful differential cue.

Another problem is the differential diagnosis from peripheral deafness. Conventional audiometry is often impossible since aphasic children may respond to sounds in an inconsistent manner. Derbyshire's⁷ method of electroencephalographic audiometry may be a more reliable testing procedure. In our experience, the presence or amount of hearing has often become clear only after a period of observation in the class room, playground, and dormitory.

SUMMARY

Five children are described who show a syndrome of acquired, largely receptive aphasia, sometimes recurrent, in association with some manifestations of convulsive disorder. Convulsions may be generalized, partial, myoclonic, or petit mal. The seizure manifestations have been readily controlled medically and are not closely correlated with the aphasic symptoms. In all cases a severe paroxysmal electroencephalographic abnormality, usually diffuse, is observed; electroencephalographic improvement tends to parallel improvement in speech re-education. General prognosis for these children is good with appropriate treatment.

ADDENDUM

A sixth case has come to our attention while this paper was in press.

Case 6. A nine year old girl without family history of convulsive disorder developed normally and was in the third grade. At age four there had been a single spell in which she fell limply unconscious and drooled. She was well until November 1956 when she complained of headaches and a vague visual disability. She could see but seemed to grope about. Examinations by physician and ophthalmologist were negative. One evening there were four brief tonic seizures 15 minutes apart. She was confused between spells and somewhat drowsy afterward. There were no residual symptoms immediately and she returned to school. During the next few weeks she gradually withdrew, had nightmares and temper tantrums, and

had progressive difficulty "like stuttering" leading to absence of speech. Psychotherapy was instituted at this time; a variety of psychodynamic explanations for the symptoms were suggested.

In early January 1957 there was a single convulsive seizure involving the right face and also the right side of the body. She was then hospitalized. Physical examination, lumbar puncture, skull roentgenograms, and pneumoencephalogram were normal. Psychologic tests showed an IQ of 80 on nonverbal material. Psychiatric and psychologic consultants believed that the symptoms were psychogenic. Two electroencephalograms a week apart showed a prominent left temporal focus of high voltage 1 to 7 per second activity.

Examination at the Central Institute for the Deaf revealed a bright, friendly, cooperative child who seemed to enjoy the examination. She responded to both oral and written commands and could give reliable answers to questions requiring "yes" or "no" answers. There was no spontaneous speech. There was perseverative repetition of the syllable "um-m-m" on attempting to name pictures and objects. Her attempts to imitate words and syllables spoken by the examiner resulted in the same perseverative syllable. She could write a number of single words in response to questions such as "what color is your dress?" In some written words there was perseverative repetition of individual letters. She was discharged from the hospital without medication.

A month later her parents reported considerable improvement. At this examination she used speech, but in a telegraphic, poorly enunciated style. Often she could not say a word which she could write. Perseveration was still evident in both speech and writing. Imitation of sounds and words was still poor in spite of obvious effort. The electroencephalogram again showed the left temporal focus, but there were also generalized paroxysms of 3 to 6 per second waves, in some of which the voltage was higher on the right side. Shortly afterward anticonvulsant medication was instituted and improvement continued. The mother wrote in April 1957 that speech was as good as it had ever been and that the child had returned to school.

We are indebted to all those who generously shared their observations and records with us: Dr. Walter Moore, St. Louis University Medical School; Dr. Donald Thurston, Dr. Jean Holowach, and Dr. Irwin Levy, Washington University Medical School; Dr. James L. Wilson and Dr. B. K. Bagchi, University of Michigan Medical School; Dr. A. J. Derbyshire, Harper Hospital, Detroit; Dr. Robert S. Knighton, Henry Ford Hospital, Detroit; Dr. Samuel Livingston, Johns Hopkins Hospital, Baltimore.

REFERENCES

1. MCGINNIS, M. A., KLEFFNER, F. R., and GOLDSTEIN, R.: Teaching aphasic children. *Volta Review* 8:239, 1956.
2. EWING, A. W. G.: *Aphasia in Children*. London, Oxford University Press, 1930.
3. LANE, H. S., and SCHNEIDER, J. L.: Performance test for school-age deaf children. *Am. Ann. Deaf* 86:441, 1941.
4. MACPHERSON, J. G., and LANE, H. S.: Comparison of deaf and hearing on the Hiskey test and on performance scales. *Am. Ann. Deaf* 93:178, 1948.
5. ROBERTS, L.: Handedness and cerebral dominance. *Tr. Am. Neurol. A. pp.* 143-148, 1955.
6. GUTTMANN, E.: Aphasia in children. *Brain* 65:205, 1942.
7. DERBYSHIRE, A. J.: Personal communication.

Neurology[®]

Syndrome of Acquired Aphasia with Convulsive Disorder in Children

William M. Landau and Frank R. Kleffner

Neurology 1957;7:523

DOI 10.1212/WNL.7.8.523

This information is current as of August 1, 1957

| | |
|---|---|
| Updated Information & Services | including high resolution figures, can be found at: http://n.neurology.org/content/7/8/523.citation.full |
| Citations | This article has been cited by 9 HighWire-hosted articles: http://n.neurology.org/content/7/8/523.citation.full##otherarticles |
| Permissions & Licensing | Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions |
| Reprints | Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise |

Neurology [®] is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 1957 by the American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

