PROGRESS IN PEDIATRICS

Convulsions in Childhood*

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Seizure Patterns

Convulsions occur very frequently in childhood; different hospitals for children unanimously indicate that 6 to 12% of their hospitalized patients are admitted for convulsions[1]. The younger the child the more frequent is the occurrence of convulsive disorders. In the newborn period they are 20 times more frequent than in the kindergarten and early school age and 80 times more frequent than in the age group between 10 and 16 years[2]. In most instances the convulsion represents a symptom of another disease, rarely it is the only symptom of the disease, e. g. epilepsy. It is therefore justified to endeavor a classification of the diseases in which convulsions occur (see Table 2). However, before classifying the convulsive disorders it is necessary to define what is understood as a convulsion and to describe the different types or patterns of them. Jackson, nearly one hundred years ago, defined a convulsion as a sudden violent discharge; modern electroencephalography has yielded the scientific proof of this statement and has shown that a convulsion or seizure can manifest itself in many different ways according to the part of the brain which is discharging. Seizures originating in the spinal cord (tetanus, strychnine intoxication) and others said to be due to hyper-excitability of the peripheral nerves will not be discussed in this paper. Seizures originating in the brain are shown in Table 1.

A convulsive discharge can start in any part of the cerebrum or brain stem and can spread from there to other parts. The initial focal discharge is sometimes called aura; its exact analysis is important for the localization of the focal origin of a convulsion.

* Read before the First Pediatric Seminar at the Research Institute of Child Health, Ankara, April, 1957.

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TABLE : 1

SEIZURE PATTERNS AND THEIR LOCALIZATION

TYPE	LOCALIZATION
Centrencephalic Seizures	
Grand mal	ant. brain stem
Petit mal	
Akinetic	
Myoclonic fits	
Hypsarrhythmia	
Psychomotor?	
Tonic, postural	post. brain stem
Breath-holding spells	
Focal Seizures	
Generalized motor	complete motor area
Jackson (partial or total)	prerolandic
Masticatory (smack, salivation, chew, swallow)	Lowe prerolandic
Simple adversive	frontal
Higher organized movements	temporal
Sensory (tingling, numbness sense of movement etc.)	postrolandic
Visual	occipital
Auditory (buzzing, drumming)	temporal
Vertiginous (dizziness)	temporal
Olfactory	temporal (uncinate)
Autonomic	different locations
Dreamy states, visions, hallucinations	post temporal

Table 1 is mostly self-explanatory. Autonomic and psychomotor seizures can arise from different locations (cortex and diencephalon). In diencephalic autonomic seizures sudden vasodilatation, rise of blood pressure, lacrimation, narrowing or widening of the pupils, salivation, or shivering (pilomotor reflex) may be seen. Autonomic auras preceding focal cortical seizures may consist in nausea, epigastric pain, chilly sensation, or precordial palpitation. A few examples of psychomotor attacks follow:

A 12-year-old boy in recumbent position became slightly confused after volitional hyperventilation, sat up, became cyanotic, smacked, complained of epigastric pains, was nauseated, stepped from the examination table, made a few steps, went back to the table, and lay down; then the attack was over.

A 13-year-old girl becomes slightly confused after volitional hyperventilation, pronates and supinates alternatingly her left hand, then tries to unbutton her blouse, and becomes cyanotic; then the attack is over. EEG revealed seizure discharge in right temporo-occipital lead.

A 9-year-old boy (114786) plays with his sisters, suddenly he falls to the ground, is stiff and cyanotic for a few minutes, but not unconscious, then gets up and walks —supported by 2 persons— to his bedroom. There he experiences auditory hallucinations, hears a terrible noise, though it is absolutely quiet in the house, vomits and falls asleep. There is no amnesia. EEG shows a severe dysrhythmia with frequent atypical spike-wave formation in left temporo-occipital lead.

The origin of so called akinetic or drop seizures is not clear in all cases. Usually these seizures are considered as belonging to the petit mal triad with bilateral spikes and waves. We saw recently a 5-year-old boy (114377) with right frontal adversive and drop seizures. During the latter he was thrown to the ground with such vehemence that he injured himself on several occasions quite considerably. EEG showed focal seizure discharges in the right frontal region. This type is probably not a true akinetic seizure, but a flexor spasm and deserves a separate clinical classification (Penfield).

The EEG is of great help in the evaluation of convulsive disorders, though it is not rare to find no convulsive pattern in the interseizure EEG (Livingston in 46%, Penfield in 5% of their patients with convulsions). On the other hand a convulsive pattern may be found in normal individuals without a manifest convulsive disorder. Whether such persons are potential epileptics or not remains an open question. The interpretation is particularly difficult if convulsive discharges are found in the EEG of patients who are suffering from non-epileptic conditions appearing in attacks, as for instance migraine or narcolepsy. We saw recently 2 such cases, a case of migraine with generalized synchronous bursts of 4/sec high voltage sharp waves in a 7-year-old boy and a 9-year-old girl with narcolepsy and typical 3/sec spikes and waves[5]. Neither case reacted to antiepileptic treatment, which speaks against these attacks being of epileptic origin, though both conditions may occur in epilepsy.

Whenever nature is pressed into a classification, incongruities are inevitable, as in Table 2, though it has proven to be of didactic and practical value. Some of the most frequent and typical forms of childhood convulsions will be discussed on the following pages.

TABLE : 2

ETIOLOGICAL CLASSIFICATION OF CONVULSIVE DISORDERS

A) *Occasional*

1. Non-nervous Diseases
   - Fever
   - Shock-acidosis
   - Hypoglycemia
   - Uremia
   - Hypocalcemia
   - Acetonemia
   - Hypoxia and hyperkapnia
     - Breath-holding spells, cyanotic congenital heart
   - Alkalosis
   - Pyridoxin deficiency
   - Exogenous toxins, poisons
   - Overdosage of drugs
2. Diseases of the Nervous System (Convulsion during acute disease)
   - Meningitis
   - Encephalitis, encephalopathy (Kernicterus)
   - Intracranial hemorrhages and other space-occupying lesions
   - Trauma of skull

B) *Chronic diseases of Nervous System with Convulsions as Accessory Sign*

1. Congenital, prenatal brain lesions
   - Malformations
   - Infections : Toxoplasma
     - Rubella
     - Syphilis
2. Birth-traumatic lesions
3. Postnatal brain lesions
   - Trauma
   - Hemorrhage
   - Infections
   - Space-occupying lesions (tuberous sclerosis, neurofibromatosis)
   - Vascular accidents
   - Neuroallergies
4. Chronic demyelinating diseases
5. Storage diseases

C) *Chronic diseases of Nervous System with Convulsions as Predominant or Exclusive Signs*

1. Focal epilepsy
2. Cryptogenic or centrencephalic epilepsy

FEBRILE CONVULSIONS

About one third to one half of all the convulsions in childhood accompany an acute febrile disease and are called therefore febrile or infectious convulsions. Whether it is justified to postulate such an entity will be discussed at the end of this chapter.

Age and sex incidence: The majority of these convulsions occur between the ages of 6 months and 4 years (88% according to Zellweger, 92% according to Herlitz); they are rare between the 4th and 9th year and exceptional after the 9th year. Boys and girls are affected with the same frequency, yet we found that girls more frequently than boys experience more than one febrile convulsion; this phenomenon remains unexplained so far and requires further study.

Diseases complicated by febrile convulsions: Nearly 50% of all febrile convulsions occur during an upper respiratory infection, which is not astonishing if we consider the frequent incidence of respiratory infections including tonsillitis and acute throat infections in childhood, but in any other *acute* febrile condition febrile convulsions may occur. However, it is noted that some acute infections, such as infectious diarrhea, roseola infantum, measles and smallpox vaccination, are more frequently accompanied by febrile convulsions than other infectious diseases. Kramar and Blaszo[6] found in the brains of patients with shigellosis an ictogenic substance. The spinal fluid of such patients, injected into dogs, produced convulsions. During the vaccination campaign in Lebanon of January-February 1957 we saw several children with convulsions on the first day of their vaccinia reaction. These febrile convulsions have to be distinguished from postvaccinal encephalitis. The former occurs during the rise of fever of the vaccinia reaction (3rd to 5th day after vaccination), is followed by a short period of postictal sleep, then the child is all right again. Postvaccinal encephalitis occurs after the vaccinia reaction, on the 7th-14th day, and lasts longer; convulsions did not occur in all cases. In all our cases of postvaccinal encephalitis there was a pleocytosis and an increase of protein in the C. S. F.; the C. S. F. during febrile convulsions is normal. Measles and roseola infantum are frequently accompanied by febrile convulsions. It may be that a tendency to neurotropic complications inheres in these conditions, yet convulsions may be due partly to a sudden high rise of fever. The age incidence of roseola infantum (1-3 years) may also bear some responsibility (low convulsive threshold). Febrile convulsions do not occur in chronic conditions such as typhoid, tuberculosis, subacute bacterial endocarditis, etc.; convulsions complicating such diseases have a different etiopathogenetic origin (encephalitis in typhoid fever, meningitis or brain tubercle in tuberculosis, embolism in subacute bacterial endocarditis).

Day and height of fever during convulsions: Nearly all febrile convulsions occur during the first day of fever. A febrile convulsion during the second day of the fever is rare, and on later days is exceptional. We analyzed the degree of temperature rise in 180 cases of febrile convulsion. In 122 cases it was between 39 and 41, and in ten cases above 41, but 22 children convulsed when the temperature was between 38 and 38.9 and in two cases convulsions occurred when the temperature was below 38 degrees but rose within the next few hours. In animal experiments overheating of the animals can provoke convulsions, but for febrile convulsions this can not be the only mechanism involved in its production.

Convulsive pattern and EEG: In more than half of the cases febrile convulsions are generalized, and in the remaining patients different types of focal convulsions are seen; in other words, if a seizure is focal, localized organic brain damage may be assumed. In Beirut we saw a good number of patients with organic brain lesions and febrile convulsions. Abnormal EEG patterns were found in 55% of more than 100 cases studied by M. Lennox.

Family history of children with febrile convulsions: Most authors found a high incidence of convulsive disorders in other members of the family. We found a family history of convulsive disorders in 25 % of our cases; Ounsted[8] and Lennox[9] even reported as many as 40% of their patients as having a positive family history. This incidence is considerably higher than in epileptics, where convulsive disorders in relatives are found in 7% of the cases, yet Pache[11] did not confirm this high incidence. In other families a high incidence of vasomotor lability (migraine, faintings), neurasthenia, nervousness and neuroses is found.

Other physical findings in children with febrile convulsions and follow-up:

A good number of the patients themselves reveal signs of imbalance of the vegetative nervous system. Also signs of increased nervousness are found; some children were extremely sensitive, ill-adjusted, or unstable with abnormal or excessive reactions during their period of negativism, etc. Similar observations were made during follow-up; vasomotor lability, acrocyanosis, increased dermographism, faintings (orthostatic), and migraine were frequently seen. Other convulsions, breath-holding spells, grand mal, petit mal or focal epilepsy were found in the follow-ups of 20% of our series; in the experience of other authors the incidence of other convulsive disorders following febrile convulsions varies between 2.4% (Herlitz), 5% (Faxen), 12.5% (Melin), and 69% (Peterman). On the other

hand the occurrence of febrile convulsions in the past of known epileptics varies between 1.9% (Melin) and 39% (Bridge).

Synthesis. Are febrile convulsions a separate clinical entity?

Summarizing the facts related above, it appears as if several factors contribute to or are responsible for the occurrence of febrile convulsions, which however are not all present in every case.

1. Neurotropic tendency of certain acute infections.
2. Familial convulsive tendency.
3. Low convulsive threshold in early childhood.
4. Hyperpyrexia.
5. Organic brain damage.
6. Epilepsy.
7. Imbalance of the neurovegetative nervous system.

The last point probably needs some explanation. It is known that an acute infection stimulates the sympathico-adrenal defense mechanisms (Frey). In view of the lability of the neurovegetative system of these children, it is conceivable that the sympatico-adrenal defense mechanisms react so excessively (sudden violent vasoconstriction?) that a convulsion is precipitated.

In view of the multiplicity of causes involved in the pathogenesis of febrile convulsions it is quite understandable that febrile convulsions are no longer considered as a separate clinical entity distinct from epilepsy. According to Bridge "both belong in a single group, best described with the name of convulsive disorders. The differences are not of a fundamental nature but only of type and degree. In one there is a single and obvious important precipitating factor in the infection; in the other there are usually numerous precipitating factors including infection. In febrile convulsions of the simplest kind a child's basic susceptibility to convulsive forms of reaction may not be low, but the physical changes induced by the infection are sufficiently marked to bring about a seizure. In epilepsy, on the other hand, a more pronounced and enduring tendency toward convulsive forms of reaction is present, and a less intense precipitating stimulus is required." In spite of this the term febrile convulsions is still widely used mainly for practical purposes. There is nothing to say against it, if the physician realizes its restrictions.

HYPOGLYCEMIC CONVULSIONS

Hypoglycemic convulsions are rare events in children; we saw 3 cases among 550 children with convulsive disorders. (Two were due to the injection of too high a dose of insulin in diabetics). They may occur in the newborn period if the mother is diabetic. This

reactive hyperinsulinism of infants born from diabetic mothers corrects itself within a few days. Hypoglycemic convulsions do not start as suddenly as febrile convulsions or as epileptic seizures, but occur after a premonitory period of fatigue, feeling of weakness, tiredness, perspiration, and flushing. Headache, irritability, and hunger may be experienced by older children. A convulsion may be termed of hypoglycemic origin when the blood sugar is found to be low before and during the seizure, when prolonged fasting initiates an attack and when the attack can be terminated by the administration of glucose. A reactive hypoglycemia may occur also a few hours after a meal or after heavy muscular exercises (relative hyperinsulinism). Hypoglycemia is a symptom which has many different causes (see Table 3),

TABLE: 3

ETIOLOGY OF HYPOGLYCEMIA

1. Transient in newborn, especially when mother diabetic.
2. Glycogen storage of liver, Type 1
3. Galactosemia
4. Adrenocortical insufficiency
5. Hypopituitarism and diencephalic disturbance
6. Hypothyroidism
7. Hyperinsulinism
   a) functional, postprandial
   b) organic
8. Defficiency of alpha-cells
9. Infantile idiopathic hypoglycemosis
10. Gastrointestinal disease (malabsorption)
11. Liver disease (defficient glycogenolysis)
12. Renal glycosuria

but by far the most frequent cause, particularly in early childhood, is infantile familial idiopathic hypoglycemosis (McQuarrie).

This condition has an early onset, mostly before the age of two years, and shows a familial tendency. No abnormal functions of the liver, suprarenals, thyroid or pituitary are found and no peculiar stigmata are present; according to McQuarrie this hypoglycemia responds well to ACTH, but a spontaneous recovery with increasing age is usual.

Repeated hypoglycemic attacks may damage the brain as shown by the slow waves appearing in the EEG, and even epilepsy may ensue. We saw one case with an organic brain lesion: spastic hemiplegia and convulsions accompanied by hypoglycemia. The hypolgyc-

emia disappeared after a treatment with alloxan; the convulsions however persisted.

Hypoglycemic convulsions are treated by administration of glucose; afterward the cause of the hypoglycemia has to be investigated and treated accordingly.

BREATH-HOLDING SPELLS

If embryonic cats, whose development of the motor functions has reached the reticular substance of the mesencephalon, are placed in a container saturated with CO_2, a tonic convulsion with opisthotonus occurs; sometimes the hindlegs move in the same manner as those of a standing horse do, when it wants to move on. The pattern of a breath-holding spell is exactly alike. A young child begins to cry for some reason or another, stops breathing, becomes cyanotic, loses consciousness, falls to the ground, and becomes stiff or opisthotonic, occasionally with a few movements of one or both legs. After a few seconds respiration begins again, cyanosis disappears, consciousness returns, and the convulsion is over; postictal sleep is rare. This type of convulsion can occur as a "functional" disorder, or it can be the expression of an organic lesion in the mesencephalon as well (mesencephalic seizure).

A) FUNCTIONAL TYPE OF BREATH-HOLDING SPELLS :

Incidence, onset, duration: An occasional single breath-holding spell is a common event when a child gets unexpectately frightened or injured or after experiencing a sudden pain. In other children a conditioned reflex develops and they go into a breath-holding spell whenever things are not going as they want, when they are frightened, angry, or displeased, or when they want attention. The onset of breath-holding spells coincides with the period of physiological negativism and occurs in the last months of the first year or in the second, third or fourth year. Their occurrence may last for several months or even years. After the age of 6 years no functional breath-holding spells have been observed. In many cases these spells occur only in the presence of one particular person, for instance the mother or the grandmother.

Physical findings and EEG: Physically these children are normal; the EEG is normal in the majority of cases; Livingston found seizure discharges in only 2% of his cases.

Family history and follow-up: We found in the families of such children a relatively high incidence of convulsive disorders, including breath-holding spells, and also of nervous disorders, neurasthenia, etc. A follow-up of our cases revealed mechanical irritability of the

nerves (Chvostek, etc.,+) in all cases, increased irritability of muscles and hyperreflexia in many cases. All were very nervous individuals, hypersensitive, easily excited with a labile vasomotor system; some had orthostatic faintings, others sympaticotonic glucose tolerance tests, tendency to post prandial hypoglycemia and to hunger diabetes, nervous polydipsia and polyuria, etc. Abnormal behavior such as enuresis, head banging, and temper tantrum are not rare. Occasionally other convulsions occurred; epilepsy was described by Heuyer and Dublineau. We found tonic convulsions during pertussis in one child, and tetany and orthostatic convulsions in another child, a 15-year-old girl, who had been hospitalized for breath-holding spells in her toddler age. Livingston found the incidence of other convulsive disorders among patients with former breath-holding spells about as high as in the average population.

Treatment: A single breath-holding spell does not require any treatment. In cases where more than one spell ocurs, it might be possible to stop them if the parents create a strict but loving home situation and ignore the spells. In more serious cases a change of environment may be necessary. Anticonvulsive drugs are of no benefit.

B) ORGANIC TYPE OF BREATH-HOLDING SPELLS :

In 1948 I described the case of a 7-week-old infant with a mild grade of microhydrocephaly, muscular hypertonus and delayed development. This infant experienced daily severe convulsions which had exactly the pattern of breath-holding spells.

These convulsions occured at any moment, when the infant was crying, when the bed was touched, when the light was turned on. The autopsy at the age of $3\frac{1}{2}$ months revealed severe degenerative changes in the globus pallidus, Corpus Luysi and Ammon's horn. Recently a 6-month-old baby was admitted for multiple breath-holding spells (70282); in his family cases of epilepsy were known; the convulsions started already during the first weeks of life. A prolonged neonatal jaundice was present, but had not been investigated. At 6 months these convulsions were still present, the mental and physical development was retarded, and neurologically there was muscular hypertonia. EEG showed no convulsive pattern, PEG showed dilatation of the anterior horns of the two lateral and particularly of the third ventricle [14]. A case with the early intractable breath-holding spells was reported by Wiedemann. After the age of one year the child died and a tumor in the mesencephalon was found. Up till now we have found this type of convulsion only in

young infants with signs of organic brain lesions. Penfield described under the heading of mesencephalic seizures a type of convulsion which is exactly like the pattern discussed here. He saw it in older children and adults with suprasellar, interpeduncular, pontine and cerebellar tumors as well as in cases of syphilis in the midbrain and traumatic lesions of the hypothalamus.

CONVULSIONS DUE TO PYRIDOXINE (VIT. B_6) DEFICIENCY

Suckling rats fed a vitamin B_6 deficient milk or nursed by vitamin B_6 deficient adult rats develop seizures, whereas the adult animals develop other signs of pyridoxine deficiency. Some time ago it was found that infants around 2 months of age developed convulsive seizures when they were fed with a special powdered milk — SMA. (This particular series of the product was withdrawn from the market, and the SMA product presently on the market does not show any untoward effects). It was then found that the convulsive disorder could be cured either by adding pyridoxine to the milk or by changing the milk [15]. Roberts and others [16] found a decrease of glutamic acid decarboxylase activity in Vitamin B_6 deficient animals. Glutamic acid, on the other hand, given to Vitamin B_6 deficient rats raises the electroshock threshold of the brain. However, the pathogenesis of these convulsions has still not been discovered fully; since the organism is probably able to produce pyridoxine, it is not yet clear how the vitamin B_6 deficiency occurs. Inadequate intake, failure of intestinal synthesis, faulty absorption or increased consumption are theoretically possible. Though pyridoxine deficiency may cause convulsions, pyridoxine therapy does not control seizures of other origin [17].

We do not discuss in this connection other types of occasional convulsions; it also would lead too far to speak about those diseases which are accompanied occasionally by convulsions (Table 2 B). A few problems of focal epilepsy have been discussed in the introductory remarks. From the many problems of centrencephalic epilepsy only two may be mentioned which are of particular importance in childhood.

Petit mal and pyknoepilepsy:

The question whether petit mal and pyknolepsy are different entities or not is still unanswered. Like other observers, I have seen a number of cases with frequent petit mal attacks, which appeared between 4 and 10 years of age and disappeared around puberty or

later. Both have generalized 3/sec spikes and waves in their EEG, normal PEG, normal neurological findings, and normal intelligence; both react well to (sometimes) benzedrine, hydantoin or also as a last refuge tridione or paradione; the diones do not only control the clinical seizures, they improve the spike and wave pattern in the EEG as well. The staring spells disappear frequently in puberty or adolescence; in other cases grand mal seizures may occur. I might mention a 7-year-old boy from Algeria with daily multiple attacks. We could control them with tridione. But when the seizures were controlled he behaved terribly, like a little devil, spitting in the face of the nurse, beating her, biting other children, kissing them thereafter, etc. The situation was so terrible that finally we had to discontinue the antiepileptic treatment. The result was a good boy with many attacks. Unfortunately, we have no follow-up, but I still feel a little uneasy thinking that the seizures could have caused an ictogenic brain damage; however cases are known with 50 and more petit mal attacks every day for years without any brain damage.

SALAAM AND HEAD-NODDING SPELLS, INFANTILE SPASMS

Synonymous are infantile myoclonic seizures, myoclonic seizures, and propulsive petit mal epilepsy[18]. Recently the term hypsarrhythmia is used, though hypsarrhythmia—being an electroencephalographic term (meaning mountainous arrhythmia)—is not fully identical with infantile spasms. About 2/3 of the infants with hypsarrhythmia in the EEG have infantile spasm, the remaining children suffer from petit mal, grand mal, halfsided tonic clonic convulsions or have no convulsions at all [19]. On the other hand a child may have infantile spasms and no hypsarrhythmia in the EEG.

Convulsive pattern: Clinically three types of seizures occur. Myoclonic jerks consist of a lightning-like quiver or sudden jerk, which goes through the body or parts of it. In the first few months of life it is not always easy to distinguish them from normal jerky mass movements, which are physiological at this age. Head-nodding and salaam spells occur frequently in series after the child awakens. In the former the child nods his head, in the latter he raises the whole upper part of his body, the legs flex in the hip, the arms are raised anteriorly or anterolaterally. Head-nodding spells have to be differentiated from spasmus nutans and jactatio capitis. The EEG reveals generalized, synchronous, high voltage, random, slow waves and spikes.

A) IDIOPATHIC OR CRYPTOGENIC INFANTILE SPASMS :

Usually the infants develop normally till the onset of the convulsive disorder, which takes place between the fourth and eighth months of life. With the onset of the convulsions the mental and physical development stops in the majority of cases, and already-acquired functions may be lost again. The neurological examination reveals a considerable muscular hypotonia, but no other findings besides the severe mental and physical retardation. The PEG is normal or reveals only a slightly dilated third ventricle. Rarely more than one child is affected, yet convulsive disorders in other family members occur in about 18% of the cases [19]. The infantile spasms disappear around the age of 4-6 years; other fits, mainly grand mal, may appear; the severest forms of idiocy are common in the vast majority of cases. Of 73 cases followed beyond the age of 5 years (Woehler, Zellweger), 38 were idiots, 14 were oligophrenic, 13 had died at an early age, 5 were epileptics and only 3 were able to earn their living. The treatment is difficult; drugs control the seizures unsatisfactorily. According to Baird a combination of dilantin (8-10 mg/kg) and phenobarbital (3 mg/kg) yields some good results whereas Janz recommends dilantin and tridione. It may be worth while to combine miltown with the previous drugs. Mysoline and milontin as well as diamox were without benefit. Stamps et al. had some good results with long-continued high doses of aureomycin, terramycin or chloromycetin [22].

B) SYMPTOMATIC INFANTILE SPASMS :

Some years ago [7] we described infantile myoclonic seizures in a case of porencephaly and in a case of aplasia of the corpus callosum. Gibbs et al [31] later described them in a case with a cysterna interventricularis (hyopolasia of the callosum). Since then many cases of this symptomatic form of infantile spasms have been observed. Among 243 cases observed by Gibbs and Gibbs[21], 105 revealed a presumptive etiology such as birth injury, anoxia, hemorrhage, kernicterus, meningitis and recently Baird and Borofsky stressed particularly the occurrence of infantile spasms after smallpox and DPT vaccination. In such cases convulsions and EEG changes similar to those in the idiopathic form are seen. The onset in some cases may be earlier, as in the 4th month, in others they may appear in late infancy. Besides the convulsions and the defect of intelligence, neurologic findings such as quadriplegia, hemiplegia, paraplegia or atonic diplegia may be present. The therapy does not differ from that of the idiopathic form. The prognosis depends on the cause and the underlying brain pathology.

TREATMENT OF ACUTE CONVULSIONS

What is the physician's approach when he is called to a convulsing child? A short history will inform him whether this is the first convulsion of a particular child or if the child is a known case of a known convulsive disorder. In the latter case phenobarbital, chloral hydrate or paraldehyde will be given to overcome the attack or, in case these drugs do not prove to be potent enough, general anesthesia is indicated 1-2 hours after a repeated dose of the above drugs. If the attack is the first one in a previously healthy child, the approach is somewhat different. In the vast majority of case the convulsion will probably enter one of the following categories: febrile convulsion, an acute disease of the central nervous system (in particular meningitis, encephalitis, or trauma), tetany, hypoglycemic convulsion or first epileptic fit. Take the temperature of the patient; if there is a fever a hypoglycemic or epileptic seizure is already improbable. It can still be a febrile convulsion, a tetany (Vit. D deficiency tetany begins very often during an infectious disease) or a CNS disease. Check for signs of rickets and tetany; the Chvostek and peroneal signs may be negative during the convulsion, but then a carpopedal or laryngospasm may be present; however in tetany, particularly in Vitamin D deficiency tetany, generalized convulsions may occur. On such occasions, particularly in the presence of rachitic bone changes, it is not easy to rule out tetany. Determination of serum calcium or, if this is not possible, determination of the Q-T interval in the EKG, may settle the diagnosis; in the meantime phenobarbital or paraldehyde may be given.

A higher fever speaks more in favor of a febrile convulsion, however as previously quoted febrile convulsions occur also with low fever. A quick examination may allow detection of an acute infectious disease such as tonsillitis, otitis media, measles (Koplik spots) etc. The possibility of a meningitis has always to be kept in mind in infancy even in the absence of meningitic signs or bulging fontanel. A lumbar puncture is therefore mandatory and should be done in every case of probable febrile convulsion. The diagnosis of hypoglycemic convulsions was discussed previously. For the treatment of a febrile convulsion as well as for convulsions of another cause (except hypoglycemia), phenobarbital, chloral hydrate or paraldehyde is given. For vitamin D deficiency tetany, 5-10 cc of calcium 10% I. M. or I. V. and 600,000 units of vitamin D are given as soon as the convulsion has stopped. Then the calcium treatment has to be continued by mouth (10% calcium chloride, 5 times daily 5-10 cc) for 1-3 days. Hypoglycemia requires intravenous or rectal glucose solution.

The next problem is the further management of a child with febrile convulsions. In our institution we follow somewhat arbitrarily the following rules.

In case of normal EEG and only one convulsion, no anticonvulsive treatment is given, the mother is advised to give the child 50-100 mg of phenobarbital by mouth in case the child is showing signs of a begining infection or acute fever.

If the child has had repeated (5-6) febrile convulsions and a negative EEG, anticonvulsive treatment is given for 1 year. In cases of organic brain lesion, epileptic character changes or seizure discharges in the EEG, anticonvulsive treatment is given for 3 years. If during this period no other seizures occur the treatment is discontinued.

TREATMENT OF CHRONIC CONVULSIVE DISORDERS

The treatment of chronic convulsive disorders would be wrong if it consisted only in the administration of anticonvulsive drugs.

The establishment of a good relationship between the treating physician and the parents is important. The parents have to be told that the therapeutic efforts of the physician depend also on the cooperation of the parents. They have to observe the child, and report to the doctor the occurrence of further fits or changes in behavior, character and performance. The parents are to be convinced that the child should lead as normal a life as possible, in order to prevent the patient from feeling that he is different from his fellow playmates or classmates. By all means he has to go to school. Particularly in Middle Eastern countries the child with a convulsive disorder is frequently kept at home. The family considers a convulsive disorder as a discrediting disease or a dishonour, which has to be hidden. The child should also participate in extracurricular activities as much as possible. He is allowed to go to swim in a pool and even in the sea provided someone is near him. In cases with frequent, not fully controlled attacks it might be wise to prevent him from climbing trees, from riding a tricycle etc. But if we have to forbid a particular activity it should be done in a way that the child does not resent it. That does not mean that the child should lead a hectic life; on the contrary a normal life is mandatory with a *regular schedule* and without additional stimulation such as result from extra parties, movies etc. Protection from excessive isolation is indicated.

There are epileptic children who are difficult to guide, who are turbulent, obstinate and unruly, who have difficulties in adjusting themselves. These behavior disorders frequently represent a mani-

festation of the disease and are encountered preferably with understanding love rather than with punishments. The parents have to understand that it is "something in these children which forces them to behave badly" and the parents have to assure their children that they want to help them to fight the tendencies of this "something."

As a part of the treatment the parents have to prepare a harmonious environment for their child with a convulsive disorder. Two examples may illustrate what is meant.

A 6-year-old girl was having daily convulsions in spite of 3 antisacer compositum pro infantibus per day (150 mg diphenylhydantoin sodium, potassium bromide 0.6, phenobartial 0.075). An investigation by social service revealed that the child was living in a very unsatisfactory environment; the child was living in the house of the mother, who was divorced and a prostitute. During a 3 months' stay in the hospital the child did not have one single convulsion, even after discontinuing the anticonvulsive treatment fully.

Neglect and an unhappy home situation therefore were probably to some extent responsible for the occurrence of seizures. Overprotection may increase the convulsive tendency as well, as the following example shows.

A girl 9 years of age had petit mal, 50 and more attacks daily, in addition she had signs of normocalcemic tetany. PEG showed slight dilatation of the lateral ventricles. EEG showed 3/sec spikes and waves. Under tridione and antisacer the petit mal did not disappear, but twice the child was for several months in an institution (a children's home directed by a physician). No attacks occurred there. An analysis of the environment revealed that the mother, not very educated, thought of nothing but her child's convulsions, talked always about it in the presence of the child and pampered her too much. Furthermore the child was better dressed (a rich child living in a poor residential section) than the other school children, which exposed her to a lot of teasing. Away from from home and school she was happy, literally bloomed, and had no convulsions. These two examples show that psychological elements are of importance and require just as much consideration as drug therapy.

The value of ketogenic diets is beyond doubt particularly in younger children. But we do not use it, because it is quite expensive and difficult to prepare and it again places the child in a special position of being forced to eat what others do not eat and not to eat what others are allowed to eat.

However, other authors[3] report excellent results with ketogenic diet in all forms of epilepsy, particularly in minor motor seizures of

the akinetic or myoclonic variety. Livingston selects children between 2 and 5 years of age for this therapy. To achieve a sufficient ketosis a diet has to be given on the basis of only 1 gm carbohydrate and protein together for every 4 gm of fat. 4 gm fat plus 1 gm CHO+protein (36 cal+4 cal) are 40 calories. This is called one unit. The caloric requirement of a given patient is then calculated and the number of required units determined.

Example: The patient's body weight is 16 kg. The caloric requirement is 16×70 cal=1120 cal. 1120 divided by 40 equals 28. The child's diet consists of 28 units. That is 28×4 gm fat=112 gm fat and 28×1 CHO+ Protein together = 28 gm CHO + Protein.

On the other hand acidosis can be produced by the commercially available inhibitors of carbonic anhydrase, 2 acetylamino 1, 3, 4, thiazole-5-sulphonamide, or diamox. Ketosis is claimed to have good effects, particularly in petit mal epilepsy. We have not yet been forced to use it and we are glad of it, because however effectual this drug may be, it does not act without inducing damage to the kidney tubules. In a few adults it worked for 4 months and then lost its effect.

For the institution of anticonvulsive drug therapy a few general rules have to be considered. It is wise to begin with one drug at medium doses and increase the dosage till seizures are controlled. If the maximum dose is reached before the seizures are controlled, then a combination of two drugs or a known combination of drugs, for example antisacer compositum * may be used. According to some authors some drugs influence grand mal better, for instance, bromides, barbiturates, and also hydantoin derivatives. For psychomotor fits, hydantoins, oxazolidin diones including mysoline and peganon, methylphenylsuccinimides (milontin) and urea derivatives (phenuron) are suggested. For petit mal benzedrines and diones are particularly recommended. In our epilepsy clinic we start in petit mal with benzedrines, go then over to antisacer compositum and give paradione only when high doses of antisacer compositum do not control the seizures. In most cases of focal epilepsy and grand mal, antisacer compositum controls the fits.

* Antisacer compositum pro infantibus contains: Atropin 0.0002, Caffein citric. 0.05, Phenobarbital 0.025, Sodium diphenylhydantion 0.05, Potassium bromide 0.2, 1-4 tablets for children below 5 years of age. Antisacer compositum for adults contains Atropin sulf. 0.00025, Caffein 0.0125, Phenobarbital 0.025, Sodium diphenylhydantoin 0.1, Potassium bromide 0.4, for children over 5 years 1-6 tablets daily.

A good dose of bromides (sodium bromide 20.0, potassium bromide 20.0, syrup 40.0, water ad 200, twice daily 5 cc equals 2 gm of bromides) is given to epileptic children with difficult character, destructiveness, agressiveness, etc. Salt intake is reduced when bromides are given.

TABLE: 4

DOSAGE OF ANTICONVULSIVES FOR THE TREATMENT OF ACUTE CONVULSIVE STATES

	Phenobarbital in mg	*Chloral hydrate in mg*
0 — 1 year	25 — 75	0.25 — 1.00
1 — 2 years	75 — 150	1. 0 — 1. 5
2 — 5 years	150 — 250	1. 0 — 2. 0
> 5 years	200 — 300	1. 0 — 3. 0

Paraldehyde (by rectum) 0.3 cc/kg body weight, to be repeated every 4-6 hours.

TABLE: 5

TOXIC EFFECTS OF ANTICONVULSANTS

Bromides	Drowsiness
	Acne
Phenobarbital	Drowsiness
	Excitability
	Rash (rare)
Dilantin	Ataxia
	Diplopia
	Nystagmus
	Rash
	Hyperplastic gums
	Hirsutism
	Rarely granulocytopenia
Benzedrine	Irritability
	Insomnia
	Tremor
	Anorexia
	Weight loss
Paradione	Photophobia
	Rash
	Pancytopenia
	Agranulocytosis

Whenever anticonvulsive drugs are given the child has to be watched for toxic effects, which are summarized in Table 5.

In cases of focal or centrencephalic epilepsy drug treatment is discontinued when a child is free of seizures for 3 years.

The EEG is not readily influenced by anticonvulsive therapy except that spikes and waves may disappear under dione treatment. The treatment is discontinued when the patient is for 3 years free of seizures. The discontinuation should not be abrupt, but the doses should decrease gradually.

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