The Significance of Epileptic Seizures in Infancy and Childhood

A. MOSOVICH

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A disruption of the normal functioning of the cells which compose the central nervous system will give way to a disorder in function. This can be expressed by abnormal activity at all levels: motor, sensory, sensorial, behavioural, and when, by its extent, it reaches higher levels, it can also include psychic and intellectual functions.

It can manifest itself by uncontrollable and unnecessary activities. It can vary from simple automatisms to more complex and organised automatic behaviour. Consciousness may be present with uncontrollable reactions. It can be clouded or may be completely absent.

Most of these expressions of dysfunction can be episodic or accidental. They can be caused by trauma, infections and inflammations. They can be sudden, brief, momentary, or they can last minutes, hours or days. They can be single or they can repeat themselves in time. The main feature of this tendency to recur is the repetition of the same motor, sensory or psychological pattern which, associated with disturbances of consciousness complete the most significant traits of the epileptic syndrome.

Department of Neuromuscular and Convulsive Disorders, Alemán Hospital, Buenos Aires, Argentina
A. MOSOVICH

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There is not, as yet, one word to define the polymorphous manifestations of cellular derangement. In the English language words such as 'spells', 'seizures', 'attacks', 'lapses' are used when referring to it. In Spanish we say *'convulsiones'* or we speak of *'ataques'*. But, both *'ataques'* or *'convulsiones'* require the addition of an adjective to qualify the nature of it. We say *'ataque epiléptico'* because there are many other types of *'ataques'*. And we say *'convulsión epiléptica'* because there are many other types of *'convulsiones'*.

Seizures are not in themselves a disease. They are an expression of an altered physiological state. They can appear as an inhibition of the activity or as a sudden and paroxysmal excitation, and the discharges can start spontaneously in diseased neurons. They can be provoked in normal neurons by stimuli of different nature: electrical, pharmacological, or physiological.

The hypersensivity of neurons which will react to hyperthermia, overhydration, hyponatraemia, hypoxia and hypoglycaemia, cerebral ischaemia, changes in carbon dioxide tension and cellular overhydration, is a peculiar property which depends upon as yet not completely clarified factors. To speak about a threshold would be an oversimplification even though the abovementioned conditions modify the convulsive threshold of neurons. Foerster referred to a

predisposition or '*krampfbereitschaft*'. Rodin, following the German school, speaks about a specific 'seizure propensity' and states that while a convulsive seizure is a symptom of a temporarily lowered threshold, it will remain isolated or infrequent unless the individual also has the necessary mechanisms for propensity towards spontaneous recurring attacks. *'Il n'y a pas qui veut, mais qui peut'.*

From what has been defined and described it can be assumed that an abnormal discharge can clinically be disguised in the most unexpected ways. A child who is playing happily in the courtyard and suddenly, with a tremendous expression of fear in his eyes, pallor, his heart pounding, runs to get hold of the skirt of his teacher or his mother, or a child who may start with episodes of rage and temper tantrums without any known cause, are only two examples. But no matter how complex the manifestations of abnormal discharges are, when it comes to a convulsive seizure, we are facing a dramatic event in the life of a family. It may come like lightning, out of a clear blue sky. Or it can be anticipated, with fear and guilt, when one of the parents or his relatives has the stigma of epilepsy, thus creating tensions and neurotic trends in a family. It is a prologue of what can become drama or an ensémble of medical facts which the family must endure and the physician may contribute to the improvement thereof.

The physician when summoned to assist a child with convulsions is confronted with the immediate care of the patient as well as establishing a diagnosis and being the recipient of the family's worries. The child might be running a temperature and the convulsion might be due to the rapid rise of fever at the onset of some upper respiratory infection or middle ear disease. The physician must concentrate on lowering the temperature and treat its cause, taking steps to prevent a probable recurrence of convulsions in the course of this sickness.

A propensity to convulsions in the course of a febrile illness in a child always requires antipyretics. This precaution should be continued until convalescence, and as soon as the child is able to get up and about, he should be sent for an electro-encephalographic examination.

There are some significant features that might favour a provisional prognosis:

(a) Duration of the seizures; the shorter the better. It is obvious that there is less chance of brain damage as a result of a convulsion which lasts 5 or even 10 minutes than one that persists beyond 20 minutes.

(b) A family history of epilepsy even though there might be a family history of febrile convulsions in childhood. A family history of epilepsy qualifies the episode as an epileptic attack and the treatment therefore planned accordingly in spite of a normal EEG.

(c) Normal birth and delivery.

(d) Normal EEG. An abnormal EEG, either with paroxysmal activity, asymmetries or focal signs, obtained in the convalescence of the febrile illness, is indicative of brain reactivity, and the persistence of the anomalies for more than 3 months after the febrile convulsion will be an indication to treat the child as epileptic.

Even though these requisites are fulfilled, we prefer to warn the family to be on the alert, avoiding a high rise of temperature in any infectious disease. In addition to antipyretics and/or cold pads, the child should be submitted to anticonvulsive medication. According to various statistics, 5% of the general population suffer febrile convulsions at some time or other.

But it is also true that the vast majority of epileptics have their first seizures before the age of 10 years. We shall confine ourselves to this group of patients, whose physical and neurological examinations are essentially normal; without apparent impairment of mental activity and intelligence, at least during the initial phases of the disease; whose EEGs are mostly, not all and not always, abnormal; and where the propensity is evidenced by a spontaneous, recurrent tendency to seizures.

Petit mal comprises, in its pure form, less than 3% of the total number of epileptic children. They range from simple lapses of consciousness, accompanied by some motor movements, to more complex manifestations. In addition to the lapses of consciousness, there are several other minor misleading symptoms which belong to temporal lobe epilepsy.

Spells of unmotivated fear and anxiety, confusional states, hyperkinesis and/or unmotivated paroxysmal rage outbursts, form part of the difficulties encountered in clinical diagnosis between temporal lobe or petit mal manifestations. The coexistence of both types of seizures in the same patient is not rare. The EEG is of utmost importance in the diagnosis, no typical 3/sec spike and wave is observed in the latter whereas an anterior temporal negative spike might occasionally be present.

I have always been puzzled by the peculiar association of photogenic epilepsy with compulsive neurotic trends which I call 'compulsive epilepsy'. I remember a case of a child who would stand at a window facing the sun rapidly waving her hand in front of her eyes until she got petit mal spells with associated sexual pleasure derived from it. Precipitating petit mal blank spells would be an alternative to masturbation with identical subjective sensations.

Petit mal may continue throughout childhood, adolescence and in some cases into adulthood. I have seen a case of general paresis in a 60-year-old man with clinical absences and spike and wave EEG pattern. It may remit in adolescence, but one-third of children with petit mal develop some other type of seizure, particularly grand mal.

Age plays an important role in the incidence of a peculiar type of attack usually discussed under the heading of 'minor motor seizures', 'infantile epilepsy', 'infantile myoclonic encephalopathy'; 'massive myoclonus', 'salaamtics', 'propulsive epilepsy', 'lightning major seizures' among other names. There is at present a universal tendency to call it West's Syndrome, in the memory of Dr West, an English paediatrician who described this condition in his own son in 1841. The seizures start as early as the first week of life up to the age of 5 - 6 years, although the preponderant age of onset is in the first year of life—as much as 85%.

They consist of sudden, massive spasms of the body musculature. There is flexion of the trunk and lower limbs with abduction and flexion of the arms in front of the body. Extensor spasms may also be observed. They occur several times a day and the child is apathetic after the attacks. It is important to state that focal or generalised convulsions may be combined with the spasms.

Aetiology cannot be confined only to symptomatic cases, although they are described in the course of leucodystrophy, lipoidosis, encephalitis, tuberous sclerosis, inborn errors of metabolism such as phenylketonuria and other processes. The fact that the majority of cases are of unknown cause makes it difficult to present an adequate interpretation as to its pathophysiology.

Known of old, more frequent than petit mal, minor motor seizures were fragmentarily described and a variety of synonyms were coined to denominate them. Electroencephalography and the report of the therapeutic value of ACTH in the treatment of those children by Sorel and Dusoucy-Boulye in 1957 attracted renewed interest, permitting grouping its symptoms as a syndrome complex whose main features are: the type of seizures; EEG pattern; age of onset.

This syndrome complex can be defined as a form of epilepsy which generally begins in early infancy, with a peculiar seizure pattern and is associated with a typical EEG pattern. The diagnosis of this syndrome carries a disheartening prognosis as to the mental development of the patient. In the same way as other empirical treatments in medicine, the use of ACTH and cortisone has proved to be of benefit. Neither the dose nor the duration of treatment follows any logical scheme.

However, the earlier the child is started on this treatment the better the chances of improvement. The least we can say is that upon the child's maturation the seizures gradually remit, with or without treatment. Unfortunately, we are faced with the appalling fact of his mental retardation.

Having outlined the physician's attitude towards the infant or child in the course of his first seizure, we can easily deduct that the prognosis will depend upon adequate information as to severity, duration, type and precipitating causes.

Information should always be obtained when possible from the child himself (if he is old enough) and from the family or household. This requires several interviews and patience. The parents could have overlooked non-convulsive symptoms or they can use misleading terminology.

A complete neurological examination as well as an EEG is essential in each case. Additional auxiliary tests will depend upon the physician's schooling and clinical judgement. Simple X-ray films of the skull might show calcifications, asymmetries, microcephaly, etc. Fundi examination is rich in information, such as cherry spots as seen in Warren Tay's disease, facomata in tuberous sclerosis, scars as in toxoplasmosis, and vascular malformations in Sturge-Weber's disease. A lumbar puncture may be correctly indicated when the convulsion occurs in the course of a febrile illness with neurological signs pointing to a meningeal complication. Laboratory tests for hypoglycaemia, hypocalcaemia or other investigations will contribute to integrate an adequate diagnosis.

Any convulsion should be considered an important event. We have seen that it can be a symptom of congenital, heredodegenerative or metabolic neurological disease, or a complication or a repercussion upon the nervous system of general pathology. The prognosis will therefore depend upon correct diagnosis, and our ideal should be, knowing the aetiology, to suppress the determining cause. If this cannot be done, we should at least control the recurrence of seizures by medical, pharmacological and psychological treatments, in order to diminish the risk of complications. Better results will be achieved through medical research.

A better attitude towards a patient with seizures will be achieved through medical education as well as education of the general public.