WEST'S SYNDROME (INFANTILE SPASMS)— A POLYMYOGRAPHIC STUDY

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A syndrome of brief repetitive massive spasms involving the muscles of the neck, trunk, and limbs, accompanied by either regression or loss of motor and mental skills, was described by Dr. W. J. West, a practitioner in Tonbridge, in his own son (1841). Dr. West had taken the child to London for a consultation with Sir Charles Clarke who had already seen four cases of what he called the 'salaam convulsions'. Sir Astley Cooper, who also saw Dr. West's child, apparently had never seen or heard of such a case: in his opinion, 'it either arose from disease of the brain, and the child would not recover, or it proceeds merely from teething, and when the child cuts all his teeth, may probably get well'.

In more recent times the original observations of Vazquez and Turner (1951) and those of Gibbs and Gibbs (1952), that a gross electroencephalographic (EEG) abnormality is common in this syndrome, have been confirmed by many workers. As the sudden, brief, massive spasms are the most dramatic feature, this syndrome (including also mental and motor regression and gross EEG abnormality) has been described under various names (lightning fits, infantile myoclonus, infantile spasms, salaam attacks, jack-knife attacks, massive spasms, etc.); even the term 'hypsarrhythmia', at first introduced by Gibbs to describe the grossly irregular appearance of the EEG, has often been misapplied by other authors to describe the clinical syndrome. In order to avoid further confusion it is proposed that the condition be named 'West's syndrome', at least until more is known about either the aetiological factors or the underlying pathological processes. It would be difficult to improve upon this author's original description of the clinical features and early course of the illness.

Patients with West's syndrome have been referred to the Neurophysiological Department at Great Ormond Street at the rate of about 20 to 40 a year from 1957 to 1963 inclusive, representing a figure of

the order of nearly 2% of all new referrals. Though in the published material both clinical and EEG studies in this syndrome are numerous, little attention has been paid to the study of the motor phenomena occurring during the brief attacks.

This neglect may be explained by the following considerations: (1) that in the majority of patients the factors precipitating the attacks are unknown; (2) that the series of brief spasms may last only a few minutes and recur perhaps only once or twice a day; and (3) that a variety of stimuli may stop the series of brief spasms, sometimes for hours. For example, attempts to film the attacks are often wasted because the floodlights may inhibit the beginning of a series of attacks, or occasionally even interrupt them once they have started.

In a study of the co-ordination of voluntary and involuntary movements of the four limbs (Pampiglione, 1959, 1961) a polymyographic method was developed which could be applied to record simultaneously the electrical activity of extensor and flexor muscles of arms and legs in babies even during violent movements.

In the present paper the timing, duration, and sequence of the electrical events in muscles, which underlie the motor phenomena, were documented during one or more series of spasms in 25 patients. Often the EEG was recorded together with the activity of the groups of muscles with mostly antagonistic action (biceps and triceps, quadriceps and hamstring).

Method

The electrodes were silver/silver chloride discs stuck to the skin with a rim of collodion, the conductor being a saline jelly. The contact resistance was lowered to less than 5 kOhms. The interelectrode distance was 1.5 to 2.5 cm. on the skin overlying each muscle. The poly-electromyograms were recorded on an 8-channel ink-writing oscillograph (Offner Type T), the time constant

FIG. 1.—This boy had prolonged series of spasms at intervals of 10-20 seconds. The spasms at first lasted nearly 4 seconds with a very abrupt onset as seen here (the time scale is in $\frac{1}{2}$ -second intervals). The spasms became very short in duration towards the end of a series showing, however, a similar distribution.

usually being reduced to 0.1 second, and occasionally to 0.03. The high-frequency cut of the apparatus was approximately 10% at 70 c/s. The amplification was of the order of 20-30 micro-volts per millimetre pen deflection in the original trace, but occasionally had to be reduced further. The EEG was usually recorded at the same time with similar electrodes placed on the scalp in a predetermined arrangement according to surface landmarks and measurements (Pampiglione, 1956).

Results and Discussion

Although clinically the attacks appeared to be more often in flexion than in extension, one of the most constant findings was that in each attack of a series a considerable amount of electrical activity (muscle action potentials) was recorded both from the flexor and from the extensor muscles. Often the total duration of electrical phenomena in both flexor and extensor muscles lasted for a similar time. Biceps and triceps as well as both quadriceps and the hamstring groups of muscles became electrically active (see Figs. 1, 2, and 3), whether each attack lasted a tenth of a second or much longer. When the duration of motor phenomena was extremely short it was possible to observe a slight delay in the appearance of action potentials in the muscles of the thigh in comparison with those of the arm, suggesting

that the central events were affecting the arms just before the legs (Fig. 2).

Usually the muscle action potentials recorded from the biceps were of slightly higher voltage and often more numerous than those recorded from the triceps when the spasms were in flexion in a given patient. The reverse would occur in other patients with prevalent extensor spasms (Fig. 3). The electrical phenomena in the muscles of the thigh seemed to follow similar trends, but differences between quadriceps and hamstring (as extensor and flexor muscles) appeared more difficult to recognize than between biceps and triceps. It seems probable that when either extension or flexion of the arms occurred, the activity of groups of muscles in the neck and trunk might change distribution, thus determining whether the spasms were either in extension or flexion. The muscles of the thighs appeared to contract simultaneously (both extensor and flexor groups). Should these observations be representative, the legs would just stiffen up in the same way whether the spasms appeared in flexion or extension in each patient.

It was also noticed that in the same patient the duration of each spasm would vary considerably during a series of attacks. The change in duration of each spasm of a series did not appear to be related

FIG. 2.—The same patient as in Fig. 1. This is one of the few patients who showed an extremely brief jerk apparently with a single muscle action potential in both flexors and extensors of arms and thighs (time scale in $\frac{1}{2}$ -second intervals) towards the end of a series of spasms.

FIG. 3.—This baby always had very brief spasms but in each case a group of muscle action potentials rather than a single volley occurred. The spasms were in extension in the arms and neck and the muscle action potentials in the triceps during each spasm appear larger than those over the biceps, while there is no great difference in the brief activity seen in quadriceps and hamstring (the time scale is in 1-second intervals).

FIG. 4.—Considerable variability in the time relations between the electrical events in the EEG and in the muscle during a series of frequent spasms. At first the muscle action potentials appear after the beginning of the discharges in the EEG, but later the muscle action potentials appear to precede the discharges at cerebral level (the time scale is in 1-second intervals).

to the type and amount of apparently voluntary activity carried out by the baby in between the spasms. In general, the most common duration of each spasm in various patients was of the order of $\frac{1}{4}$ to 1 second. Considering the over-all shape of the burst of muscle action potentials, the onset was fairly rapid and the muscle action potentials increased to a maximum over a very short period, and then tended to tail off much more slowly, disappearing altogether during relaxation. While sometimes prolonged spasms lasting 3 to 4 seconds were seen,

only rarely did single muscle action potentials appear. This, therefore, differs from the electrical events occurring in 'myoclonic seizures' (Dawson, 1946, among others). In our observations even the briefest jerks were usually accompanied by a group of muscle action potentials and only exceptionally by a highly synchronized single volley (Fig. 2), bearing in mind, moreover, that the records were taken through a chopper amplifier.

The relation of the EEG phenomena to the myographic events was more variable than one might

perhaps have expected (Fig. 4). In fact, though in particular patients after an initial discharge in the EEG lower amplitude faster elements might be seen for a second or two during the main part of the spasms, on other occasions in the same patient the EEG changes might be much less marked, and there might even be occasions during which, in the presence of fairly marked generalized spasms, no gross EEG changes would appear.

From these polymyographic observations carried out during the spasms in 25 children with West's syndrome, it seems that there is a particular qualitative difference between these attacks and all the other seizures that can be observed in infants of this age-group. The sudden brief involvement of both agonist and antagonist groups of muscles in the four limbs regardless of whether flexor or extensor muscles predominate is not commonly seen in any other form of seizure, suggesting that in West's syndrome the events at central levels may have some peculiarities as yet to be studied and understood.

The motor events at peripheral levels that may be studied by polymyography and the central events recorded with the EEG show a much more variable relation than the somewhat stereotyped clinical

features of the repetitive spasms might lead one to suspect.

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