

**Paramyoclonus multiplex : report of a new case, with further history of a case reported in 1896, which has since recovered / by F.W. Langdon.**

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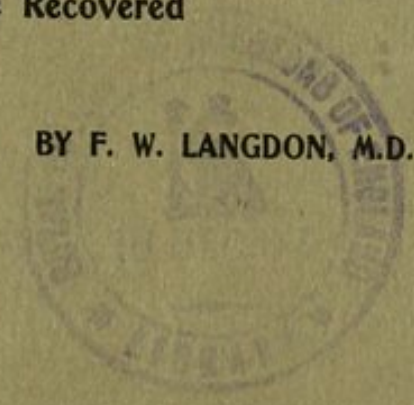
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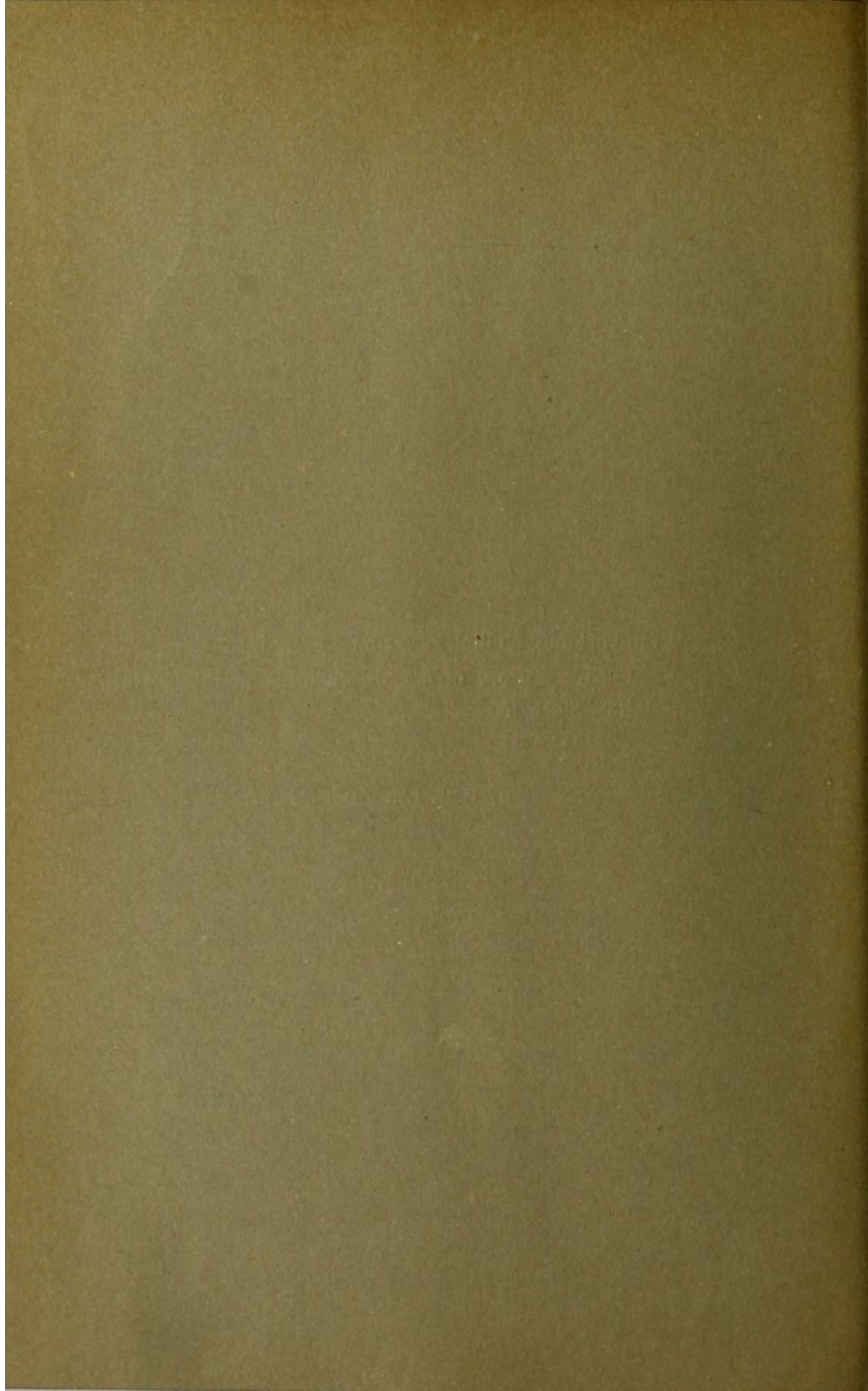
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**Paramyoclonus Multiplex: Report of a New Case, With  
Further History of a Case Reported in 1896, Which  
Has Since Recovered**

**BY F. W. LANGDON, M.D.**

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PARAMYOCLONUS MULTIPLEX: REPORT OF A NEW CASE,  
WITH FURTHER HISTORY OF A CASE REPORTED  
IN 1896, WHICH HAS SINCE RECOVERED.<sup>1</sup>

By F. W. LANGDON, M.D.,  
CINCINNATI, O.

The patient, a bright little girl of thirteen, was admitted to my service at the Presbyterian Hospital, February 9, 1902.

*Complaint*—Muscular spasms, occurring in paroxysms, which began six days previous to admission, and were preceded by pain in the back. This pain followed shortly after a shock from being frightened in play by her brother who was fond of teasing her.

*Family History*—The father was said to be a dissipated man, who had been separated from his family for some years. The mother, who accompanied the child, was in good health, and not subject to any nervous disorder. She gave a history of seven pregnancies, of which two terminated prematurely, one before and one after the birth of our patient. Three died within a few months of birth.

The brother, the only living child, except the patient, is eighteen, well physically and bright mentally.

*Personal History*—The patient at birth, appeared strong and healthy and weighed ten pounds, according to the mother's statement. The mother was unable to nurse her at the breast. She had "spinal meningitis" at six months of age, measles at four years, and peritonitis at ten. Had an illness at eleven which was diagnosed St. Vitus' dance, and, which, from the mother's account, somewhat resembled the present attack, but was milder and lasted a few weeks.

*Present State*—Patient is underdeveloped physically; has defective teeth, suggesting the Hutchinsonian type, but not conforming to it.

Skin clear, no corneal scars.

Temperature, 98.4; pulse, 108, low tension, small volume.

Heart and lungs clear.

Appetite and digestion good, bowels constipated.

Urine passed in bed or with stool, so that it was some days before a specimen was procured for examination. This re-

<sup>1</sup>Read at the annual meeting of the American Neurological Association, June 5, 6 and 7, 1902.

vealed no albumin or other abnormal constituent and was normal in appearance.

The total amount passed, was probably less than normal.

Menstruation has never appeared.

*Nervous System*—The mental state appears to indicate excessive fatigue. The patient answers questions logically, but very slowly and with great apparent effort. She complains of no pain or discomfort of subjective character, but says "don't hurt me," "don't press so hard," when gently moved about during the examination. This hyperesthesia to ordinary touch seems universal and not confined to localities or zones.

In disposition she is good-tempered and with an unusual degree of patience for a child, not complaining except when touched, and then flinching rather than resisting.

*Cranial Nerves*—The violence of the muscular spasms, which are brought on by questioning and handling, prevent any detailed tests.

There is no contraction of visual fields to rough tests; no defect of hearing, excepting possibly a hyperesthesia.

Smell and taste are apparently normal.

*Trunk and Extremities*—Patient lies on her left side, but assumes the dorsal decubitus readily upon request. There seems to be no loss of any voluntary movement but excessive fatigue appears to accompany the slightest exertion.

The grasp is so feeble that it cannot be registered on an ordinary dynamometer.

The muscles of the trunk generally, both dorsal and ventral, and those connecting the trunk and extremities, are the seat of frequently repeated violent clonic contractions. These clonic spasms closely resemble the effect of closure of a galvanic circuit in the muscles.

While these clonic spasms are bilateral, and generally, though not always, synchronous on the two sides, they are not strictly symmetrical in power, being usually much more vigorous on the left side as regards the muscles moving the limbs. At times the difference in activity between the two sides was scarcely noticeable.

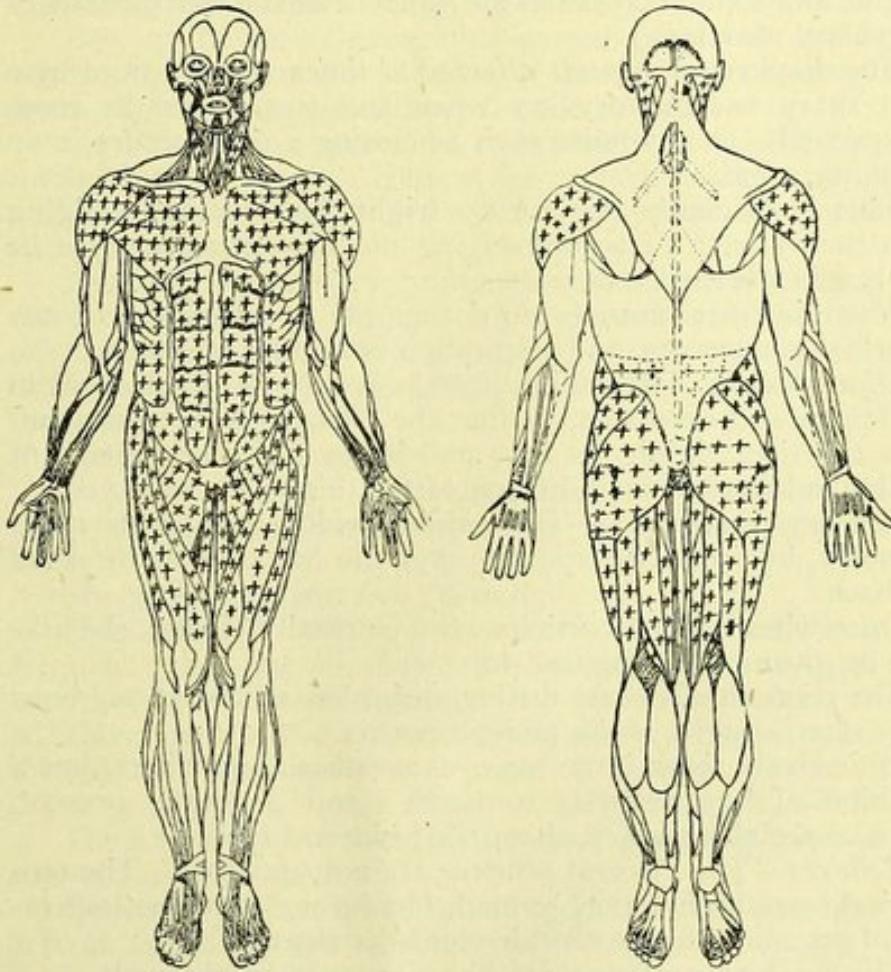
The contractions in the *erectores spinae* are apparently about equal on the two sides.

A transient *opisthotonus* is produced by them at times, and on three separate occasions during her first day in the hospital they were so violent as to throw her out of bed on to the floor, which accident was attended with some contusions.

The spasms, which are practically continuous during waking hours, are rendered more violent and more frequent by touching the skin, testing the tendon reflexes, or entrance of another

person into the room; and by attempts of the patient to answer questions.

As regards distribution of the spasms, it is shown fairly well on the accompanying illustration which is reproduced from that made for the first case reported by the writer<sup>2</sup>.



Distribution of myoclonic spasm indicated by plus marks.

The chief difference is in the marked involvement of the *erectores spinæ* in the present case, and their escape in the former one.

The biceps and triceps in the present case are the seat of occasional spasms, while the muscles of the face and neck, the forearms and hands, and the legs and feet seem to escape entirely. The wrists, fingers, ankles and toes remain flaccid, even during the violent paroxysms; their only movements appearing

<sup>2</sup>Cincinnati Lancet-Clinic, February 8, 1896.

to be transmitted from the shoulder and upper arm contractions.

The tongue can be protruded and remain so during the paroxysms; conversation can be carried on, facial movements made at will, without apparent effect, other than the production of fatigue, and some increase in the violence of the contractions of the truncal muscles.

The diaphragm appears affected at times, as evidenced by a short sharp exclamatory cry when any one enters the room unexpectedly, or any noise such as closing a door occurs in an adjoining room.

This is evidently not due to fright, but appears, judging from the absence of accompanying mental disturbance, to be simply a heightened reflex act.

The intercostal muscles do not appear to be the seat of any abnormal movements, and respiration is regular and free.

Contraction of the thigh-muscle groups is about equal in the flexors and extensors, so that the effect is to produce simply a transient rigidity at hips and knees. No movements of rotation, adduction, or abduction of the thighs are observed.

*Sensory Symptoms*—The patient seldom complains spontaneously, but when questioned says she has pain in the head and back.

Once when the contractions were unusually violent, she asked to be given something to "stop me."

The contractions cease during sleep, but return at any hour of the day or night, if the patient awakes.

Objectively there is no area of anesthesia anywhere, but a condition of hyperesthesia to touch seems generally present. This is especially marked about the head and face.

*Reflexes*—The visceral reflexes are not impaired. The tendon jerks are highly exaggerated, but do not give the impression of organic disease. Ankle-clonus is absent.

The cutaneous reflexes are likewise much heightened.

The Babinski sign is absent. Testing either the tendon or cutaneous reflexes, brings on the spasms when absent, and increases their activity when present.

The following notes made by the writer at various intervals, show the range of variation in the muscular movements and mental state during her two months' stay in the hospital.

Feb. 8 to 15, contractions chiefly, sometimes entirely, limited to left side. Frequency usually about 55 or 60 per minute, but varying at times between 27 and 98. Occasional intervals of freedom from contractions for from 5 to 10 minutes by the watch.

During this period, hyoscine hydrobromate in 1-200 grain

doses was given every three hours, excepting during sleep. Also potassium iodide in 5 to 10 grain doses.

Previous to her admission to the hospital, chloral and potassium bromide had been given in full doses, with the effect, according to the mother, who had been a professional nurse, of increasing the violence of the spasms.

Feb. 21 to 26. During this period potassium iodide was continued, but Fowler's solution of arsenic in increasing doses up to 12 drops was substituted for the hyoscine.

On Feb. 22, the spasmodic contractions numbered 84 to the minute by two separate counts; were shock-like as usual, bilateral, but rather stronger in the left side; practically continuous except during sleep.

Feb. 26. Spasms irregular in character; first a group of 7 to 10 contractions, rythmical and at the rate of about 60 to the minute; then a change to a group of 7 to 10, much quicker and more violent. Chiefly the glutei and erector spinæ groups are involved.

Mental condition good.

Feb. 27 to March 1. During this period sulphonal in 10 grain doses was given every three hours excepting during sleep.

She received about five doses daily.

Note March 1, 5 P.M. (by writer). Individual shocks less frequent (32 to the minute). At intervals of one to three minutes the shocks occur in groups of 7 or 8, very quickly repeated; then a pause, and a return to the slower rate of 30 or 32 to the minute. Then seven very rapid and violent shocks (sometimes 14 or 15). Then a return to the 32-rate per minute.

The nurse reports that this mode of grouping has been observed for three days.

Patient is delirious, and talkative for the first time since her arrival, and complains of dizziness in head.

Pulse ranging from 72 to 94.

Sulphonal omitted March 1, and no antispasmodic remedies given for three days.

March 3. Pulse 60, low tension, small volume. Spasms in groups of 8 to 15 shocks (rate about 60 to the minute). Between these groups of shocks are intervals of freedom, varying from a half hour to an hour.

Has been singing and elated much of the time; constructs doggerel. Seems good-humored and logical when questioned.

Fluid extract cactus grandiflora in three-drop doses was ordered to improve the circulatory depression.

March 5. Spasms as last noted, but with shorter intervals



of rest ( $\frac{1}{2}$  minute). Rate 30 to minute with groups of 8 to 15 quicker shocks occasionally.

Mentally somewhat stupid or slow of comprehension.

Asks to be given something to "stop me."

Hyoscine given one dose 1-100 grain and patient slept quietly all night without awakening and without spasms.

Other remedies tried in turn were thyroid desiccated in 3-grain doses; sodium bromide 20-grain doses; conium, Squibb's fluid extract, 3-drop doses; potassium bromide, 10-grain doses; solution bimeconate of morphia, 10 to 15-drop doses.

Each of these drugs was given separately for some days in connection with some tonic medication, but no appreciable improvement could be traced to their use, with the exception of the morphia salt, which lessened the frequency and violence of the individual spasms as well as secured short intervals of entire freedom from them during the waking hours.

The effect on the mental state, however, was bad, as was the effect on appetite, the patient developing an aversion to food of any kind, while before this, all food offered was acceptable and enjoyed.

The spasms continued less violent, 25 to 30 per minute for some days after cessation of the morphine salt, and the mother insisted on removing her from the hospital to her home, where she was placed, by my advice, in the care of Dr. E. M. Keefe, to whose courtesy I am indebted for the following notes:

For the most part the treatment pursued was a tonic one, comprising iron, arsenic, manganese and strychnia in small doses. Occasional doses of hyoscine or bromides were given when the violence of the paroxysms seemed to demand relief but with little apparent effect.

The patient was better and worse by turns very much as when in the hospital. On the whole, however, a gradual improvement took place after removal to her home, so that by May 16 she was free from the muscular shocks for a whole day.

This period corresponded to the one hundred and second day after the onset of the disease.

On the following day, however, the excitement produced by a neighbor in the same house chastising a child, brought back the spasms in full force for 24 hours, after which they have gradually lessened up to date (May 24), when there have been none for 48 hours.

Her general condition is better, as regards color and flesh; she is mentally cheerful and happy, but still so weak as to be unable to sit up.

Further reports of progress and termination are to be expected.

*Notes on the Further Progress of a Case of Paramyoclonus Reported in 1896.* (Referred to as case one in text.)

Summary.—P. H., male, aged 48, of Irish birth. Onset shortly after a period of business worry, and a mild attack of la grippe.

Distribution of shock-like contractions as shown in accompanying illustration. Frequency of contractions about 70 per minute usually, but varying at times between 50 and 250. Duration of paroxysms from  $\frac{1}{2}$  minute to ten minutes.

Number of paroxysms or attacks in a day, varying between 20 at first and two at the latter part of the disease.

Intervals of rest between paroxysms from a few minutes to several hours.

Paroxysms brought on at any time by questioning patient, exposure of surface of body to the air, and by ordinary voluntary movements of the patient.

*Remedies used:* quinine and arsenic in moderate doses, which appeared at first to lessen the number of paroxysms markedly, but later had no observable effect.

Solution bimeconate of morphia, given in ten-drop doses, at four hour intervals, for a day was followed by almost continuous spasms for 24 hours.

Total time in hospital about two months during which period he went home for eleven days, returning much worse than before.

*Hitherto Unpublished Notes of Further Progress of Case I. P. H., January, 1896.*

During the eleven days' sojourn at home above noted, the paroxysms of clonic spasms became much more frequent than when in the hospital, and the individual contractions were said to be much more severe. Friends who came with him to the hospital at the time of re-admittance, stated that on one day he was "almost maniacal" in his actions.

A brother-in-law states that he was always a "very impatient" man; hard to control, and likely to over-exert himself at work. During the six weeks following his re-admission, the paroxysms varied in frequency from 8 to 15 for the most part, once going as low as four, and one day (March 20), the spasms were almost continuous night and day, so that separation into definite paroxysms was impossible. •

On one day he complained of a "burning sensation in stomach," and on another day, of a "cracking sensation" in the head, but these subjective conditions were not accompanied by any exacerbation in the frequency of paroxysms or the violence of the contractions.

Patient vomited about  $\frac{1}{2}$  hour after meals for some days,

and also vomited his medicine, which at this time was sodium phosphate. On one occasion he had a moderately severe pain in the abdomen, for which solution of morphia bimeconate was given in small doses. This, while apparently benefiting the abdominal pain, was followed by a pain in the "back of the head," and by considerable increase in the frequency and severity of the spasms. He left the hospital little if any improved as regards the spasmodic contractions and disappeared from view for nearly two years.

On Jan. 25, 1898, he called at my office apparently in perfect health, having gained 25 lbs. in weight, and been free from spasms of any kind for eleven months.

He stated that after leaving the hospital, March 1896, he continued treatment at various places for about a year, but with no benefit to the spasmodic contractions.

He ascribes his recovery to his return to hard laboring work (ditch digging), which he resumed a year ago (1897), after discarding all medical treatment.

Since this note, I have seen him in May 1900, and again in January 1902, and he has had no relapse nor ill health of any kind during these intervals.