

table is given of the relative frequency of psychical disorders in association with tumours of different areas of the brain, the highest proportion being shown in tumours of the corpus callosum, and next of the frontal region.

Book VII., by Roubinovitch, treats of mental disorders in congenital or infantile cerebral lesions (idiocy and imbecility), and is as complete and well illustrated as the rest of this work.

Dr. Roubinovitch is also responsible for Chapter I. of Book VIII., treating of myxœdema and cretinism, while Dr. Dutil discourses on mental disorders from over-function of the thyroid body (exophthalmic goitre).

Dr. Anglade writes fully in Book IX. of the treatment of mental disorders, and Book X., which completes the work, is by Dr. Vallon on administration, lunacy law, asylums, experts, civil capacity, responsibilities, crimes, life insurance, traumas, feigned insanity, and deontology.

There appears to be only one adverse criticism to make on this monumental work, which is of encyclopædic importance, and redounds in the highest degree to the credit of our French colleagues, and must ever remain a most valuable work of reference.

The criticism is directed against the index, which is merely a table of contents of the different chapters. For such a large work a full index should have been given so as to facilitate the labour of searching through the different chapters for any special subject to which reference is desired. Considering the size and weight of the book, it would have been better in two volumes, but its extraordinarily low price (26 fr.) is an inducement to everyone engaged in psychiatry to purchase it.

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*Myoclonus Multiplex.* By CHARLES L. DANA. (*Journal of Nervous and Mental Disease*, August, 1903).

*Pathology of Paramyoclonus Multiplex* (Friedreichs Type).

By J. RAMSAY HUNT. (*Journal of Nervous and Mental Disease*, July, 1903).

*Die Progressive Myoklonus-Epilepsia* (Unverricht's Myoklonie). By Dr. HERMANN LUNDBORG.

THE above recent monographs on myoclonus may well be reviewed together, for they raise the important question as to what is meant by the name "myoclonus." Is "myoclonus" a

disease—an entity—or is it only a symptom? As originally used by Friedreich, it certainly indicates a disease having definite clinical symptoms. Unverricht attached the name to a class of cases differing entirely from that described by Friedreich. How, then, should the term “myoclonus” be used?

Dana starts by the statement that myoclonus is a general term covering all the diseases in which muscle-twitching is a prominent symptom. The term here is used as indicating a symptom, and as such he proceeds to classify the diseases in which the symptom occurs; the classification he arrives at is as follows:—

(1) Paramyoclonus multiplex, of Friedreich; astasic myoclonia, of Vanlair; multiple spinal myoclonus, of Lowenfeld; fibrillary chorea, of Morvan; fibrillary myoclonus, of Kny.

(2) Functional or hysterical myoclonus multiplex, chorea major, chorea electrica, of Henoeh.

(3) Myospasms; memory-spasms, of Friedreich; habit chorea; chorea variable des degenerés; convulsive or spasmodic tic; tic general; Tourette's disease, myriadrit, palmus; tic neurosis, of Collins.

(4) Degenerative chorea, hereditary chorea, Huntingdon's chorea; myoclonus-epilepsy; myoclonus, of Unverricht, of “familial” type; myoclonia congenita, of Seeligmüller(?); hereditary degenerative chorea of Sachs.

(5) Infectious chorea, chorea minor, Sydenham's chorea; chorea electrica, of Dubini(?); chorea electrica, of Bergeron; senile chorea, of Gowers(?).

It is difficult to see what advantage can be derived from a classification which puts under one head diseases which are essentially different in type.

The “tics” form a most useful clinical group, and, as a type, are easily distinguished from Sydenham's chorea. If the term “myoclonus” is to include all diseases in which spasmodic muscular movements occur, which neither clinically nor pathologically—so far as pathology goes—have anything in common, then the term loses its significance and has but little value.

It would be well that the term “paramyoclonus multiplex” or “myoclonus multiplex” should be reserved for that form of myospasms characterised by multiple isolated contraction of individual muscles.

Hunt goes still further, and would distinguish contractions of this type from the contractions of a cerebral type which are characterised by movements of a more or less co-ordinate nature.



FIG. 1.  
Animal at rest ; ears horizontal.



FIG. 2.  
Infra-orbital vibrissæ touched with finger-tip ; homolateral pinna rising.



FIG. 3.

Front view of elevation of homolateral pinna only when infra-orbital region touched.



FIG. 4.

Tonic maintenance of the elevation of the pinna for a few seconds after the withdrawal of the stimulation.



FIG. 5.  
*Both ears pricked up as the animal is excited and escapes.*



FIG. 6.  
Ears horizontal, although ray of direct sunlight is flashed on to eye; example of absence of reaction of the pinna to certain other kinds of stimuli.





It would be well if the term "myoclonus" could be reserved for the two conditions above mentioned, viz., the paramyoclonus of Friedreich and the myoclonus of Unverricht.

Hunt has had the opportunity of investigating pathologically one case of myoclonus of Friedreich type. He finds no abnormality of the nervous system, except some slight change in the vessels of the gray matter of the spinal cord. The muscles, on transverse section, have a diameter of unusual size, measuring in some instances 175 micromillimetres. In contrast to this are very small fibres having crescentic forms and flattened out on the periphery of the large fibres. Instead of finding the nuclei of the sarcolemma sheath confined to the periphery of the muscle fibre and just beneath this membrane, they are found scattered as well between the sarcous elements. Sometimes three or four nuclei occupied a central position in a single fibre. The nuclei of the sarcolemma sheath are increased in number.

Lundborg, in an excellent monograph on *Die Progressive Myoklonus-Epilepsia*, divides the myoclonias as follows:—

- (1) Myoklonia symptomatice.
- (2) Myoklonia simplex, or myoklonus multiplex.
- (3) Myoklonus-epilepsia, divided into two forms: (a) a progressive form (Unverricht's family type); (b) an intermittent sporadic form.

The tics, Huntingdon's chorea, should not be confused with the myoclonia, from which they are easily distinguished.

This classification would commend itself to the writer of this review, not only on account of its simplicity, but also because it allows of a wide use of the term as a symptom in association with other diseases, and recognises two well-marked types of disease which may be considered as entities.

The monograph deals more especially with the one form of myoclonus epilepsy, and the following description of the disease is given:—

The disease begins in later childhood and lasts during life, so that some of the patients between 60 and 70 years old have suffered from fifty to sixty years.

Most patients, however, die about middle age. Three stages are described: the first, characterised as the epileptic-tetaniform stage, is of comparatively short duration. The attacks occur mostly at night and during sleep, at first with a long interval, but always tending to become more frequent.

The patient awakes out of sleep with painful tonic or clonic and tonic cramp in various parts of the body. The attacks are

very different in their intensity. In the mildest attacks rapid muscular contraction takes place in the extremities. In the severest cases the attack resembles tetanus, and the contractions follow one another so quickly that the muscles never become flaccid. These contractions do not produce a locomotor effect, owing to the fact that the antagonistic muscles are in like contraction. Every manipulation tends to increase the spasm. Consciousness is preserved, and the patient has pain and distress, and may cry out with pain. Urine and fæces are not passed incontinently.

The second stage, so-called myoclonic epileptiform stage, lasts for several years (ten or more). The disease is now more pronounced during the day. Tremor—fibrillary and fascicular muscular contractions occur—beginning usually in the upper extremities, then in the buttock muscles, the throat, the face, and, lastly, the diaphragm, the pharynx and larynx, and, finally, the ocular muscles and sphincters of the bladder and rectus—in a word, all the voluntary muscles.

The patient experiences good and bad days.

During this stage psychical phenomena present themselves and the intelligence falls off; the patients exhibit changes between depression and exaltation.

The third—terminal—stage is one of cachexia. This may begin at almost any time, but generally shows itself after the first decade of the disease.

The muscular movements gradually increase in the course of years, whilst the epileptic attacks become less frequent, and eventually disappear. The periodicity of the attack becomes irregular, and the intermediate periods of rest shorter.

Rigidity of muscles now manifests itself, and the patient assumes a fixed and bent position. Any attempt to alter the position produces a definite resistance. The patient sweats and salivates profusely, passes into a somnolent half-stupor stage. Urine and fæces are often passed incontinently; the cachexia increases, and the patient dies of inanition or of some complication.

Two factors in the etiology of the disease came into prominence; the first of these is the hereditary and family nature of the disease, and secondly the influence of alcohol.

In two families there were five cases; in one family four cases; in seven families three cases; and in nine families two cases.

The occurrence of paralysis agitans in other members of the



same family is a striking feature brought out in the cases recorded, for in one family in which five cases of myoclonic epilepsy occurred five members of the family were affected with paralysis agitans.

The author next deals with the differential diagnosis of the disease.

The important diagnostic points are :—

It is a family disease—starting with nocturnal epilepsy—associated in childhood with attacks resembling tetanus, and after a few years lightning-like involuntary muscular contractions which gradually affect all voluntary muscles. The patient has good and bad days. The muscular contraction frequently affects only a part of a muscle. The contractions occur irregularly and are increased by voluntary movement ; during sleep they stop or are very much diminished. As time goes on a definite muscular rigidity occurs. The knee-jerks are exaggerated. No trophic disturbance of muscles or abnormality of sensation is present. Sweating and salivation are marked features. After some years failure of mental powers, sometimes associated with mania and hallucinations occur.

The author next contrasts the disease with chronic progressive chorea (Huntingdon's chorea). Both diseases arise from a degenerative basis, have a progressive course, and end in dementia. The involuntary contraction of muscles is present in both cases.

The points of distinction are :—

*In Progressive Myoclonus Epilepsy.*

- (1) Direct heredity unusual.
- (2) Disease begins in childhood, with nocturnal epilepsy—and later on the muscular contractions appear—the muscular contraction and the attacks stand in relation to one another.
- (3) Periodicity dependent on various sources of reflex irritation.
- (4) Sensory clonic reaction present.
- (5) The muscle contractions are myoclonic—  
Sharp contractions affecting single muscles.
- (6) On intention movement the contractions increase.
- (7) The patient soon loses the power of walking.

*Huntingdon's Chorea.*

- (1) Direct heredity usual.
- (2) The disease affects persons between 30 to 40 years of age and is rarely combined with epilepsy.
- (3) No periodicity.
- (4) Sensory clonic reaction absent.
- (5) The muscle contractions are choreic—  
Slow contractions affecting groups of muscles.
- (6) On intention movement the chorea becomes less.
- (7) The power of walking, though difficult, is not lost till the final stage of the disease.

The differential diagnosis between this disease and Koschewnikow's epilepsy and Dubini's chorea electrica is then

discussed, and the similarity of myotonia (Thomsen's disease) to myoclonia is pointed out.

#### PATHOLOGICAL ANATOMY.

In the few cases which have been examined, the changes found have been slight and often attributable to some inter-current condition.

In Clark and Prout's cases changes were found in the second layer of cells of the cortex, and also in the large pyramidal cells of the third layer—and these authors suggest that these changes underlie the myoclonic spasm.

It would seem, therefore, probable that myoclonus is dependent on changes in the Rolandic region of the cortex.

The possible relation of the disease to changes in the thyroid and parathyroid is discussed, for it has been shown experimentally that clonic spasms occur after removal of the thyroid.

Ramsay Hunt, in the examination of a case of paramyoclonus multiplex, which would come under Lundborg's second division, could find no change in the nervous system, but found changes in the muscles, the fibres being of an unusually large size—with an increase of nuclei in the muscle fibre and also in the sheath of the muscles.

In conclusion of this short review, it would seem that but little advantage is gained by placing under the term myoclonus all the various conditions enumerated by Dana, and it would be far better to adopt the classification given by Lundborg:—

(1) Myoklonia symptomatice—using the term myoclonus simply as a symptom indicating a form of myospasm characterised by multiple spontaneous, and isolated contraction of individual muscles.

(2) Myoclonus multiplex.

(3) Myoclonus epilepsy—(a) A progressive form. (b) An intermittent form.

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