

DISSEMINATED OR INSULAR SCLEROSIS.

The disease thus named consists in the formation of scattered islets of sclerosis in various parts of the brain and spinal cord, sometimes also in the cranial nerves. Our knowledge of the affection is recent. The lesion was, indeed, long since figured by Carswell and Cruveilhier, and it was described, thirty years ago, by Frerichs, Rindfleisch, &c., but the malady was not generally recognised until re-investigated by Vulpian, and by Charcot and his pupils, and described by Charcot in his widely circulated 'Lectures.' It is called by the French "Sclérose en Plaques Disseminées," by the Germans "Multiple Sclerosis," and, in this country, the French designation has been modified into "Disseminated Sclerosis;" but it is also called "Insular Sclerosis," a convenient term, proposed by the late Dr. Moxon.

James S. et al ^{married, with healthy children, was} admitted ^{to} the National Hospital
May 18 1904. He was left-handed, except for writing
in which he used the right hand. He thinks he had
some injury to the right hand when about 10 which
compelled him to use the left. He had served in the Navy
for 20 years & during that time he had many blows
on the head. At 16 he had gonorrhea but denies
ever having a venereal sore although he admits many
risks. Some time in 1902 he ~~had~~ was struck
on the head by a piece of coal & in November,
how long after the blow he does not know, he had
a severe fit, ~~appeared~~ & after it he layed in bed
five days. He remembers, during this time, seeing
a bright white light, on the left, which moved
only with the eyes. ~~when he moved them. Other~~
~~were it was constant in position.~~ Afterwards, for
a month or two, he had slight attacks in which
his head turned to the left ~~with~~ there was brief
loss of consciousness.

In March 1904, ~~he~~ on one day, he had three
severe attacks, ^{in one day} with loss of consciousness, & spasms
on the left side, ~~and~~ ^{and} colored lights to the left
~~they returned afterwards~~ ^{recurred} ~~every~~ ^{after} every few days
until his admission, & afterwards. ~~There~~ ^{attacks} ~~that were~~
^{There that were} severe seemed to make the left limbs weak

Many ^{more severe} attacks were seen, & very carefully observed by Dr Gordon Holmes, who was then Reader Medical Officer. They were heralded by the appearance of ^{several coloured} balls just to the left of the preying-point. These were white in the middle, red outside, & blue between; keeping the same size, always bright & conspicuous, the visible whether the eyes were closed or open. ~~if the eyes were moved they would move with them.~~ ^{From near the preying-point} Hence they moved to the left in a series of bounds, & disappeared about 90° from the preying point. They were visible 5 to 7 minutes & during this time the eyes were constantly jerking towards the left & occasionally the head did the same.

Soon after the appearance of the balls the ~~fields were tested by the pingers.~~ At first there was only slight "dimness" in the left half of ^{each} field, but this rapidly deepened to complete loss, up to the middle line, except just around the preying point. It remained complete until 5 minutes after the coloured objects had disappeared. Then the loss ~~became~~ ^{became} ~~relatively~~ very slowly lessening but at first he could see ^{to the left} ~~against~~ its round objects the size & shape of the ^{previous} coloured balls, ~~in the left.~~ The recovery from the hemianopia appeared to ^{proceed} from the medial part of the field towards the left; but it

passed away so slowly that ⁱⁿ half an hour
a little dimness still remained

In another fit; the first thing was
jerking movement of the eyes to the extreme left,
& of the head by tonic spasm. After three
seconds the spasm ceased, presently to recommence.
Such attacks continued for 20 minutes & a
sound was made with the mouth every few
seconds as if the patient were eating. The left
arm was rigid but without movement ^{other than pronator} of the humerus was
held across the palm of the left hand. There was
complete loss of sensation to both pain & touch
up to two inches above the elbow joint. ~~Considerable~~
~~force was necessary to bend the joints.~~ In the left
^{there was some spasm, abduction of}
~~leg, knee was~~ ^{is} ~~rigidly~~. The right limbs were
quite normal. During this period the patient
saw the coloured balls to the left, ^{staring at} ~~staring at~~
during ten minutes & when they disappeared
the eyes ^{head} could be moved to the right, but the
tremor of the arm ceases, but was replaced by
irregular motion comparable to athetosis. In a
few minutes the loss of sensation was found to
extend only a few inches above the wrist. There
was no impairment of consciousness but he
frequently used wrong words in speaking, which
bore no resemblance in sound to them he
intended to use

He was well educated for a man of his social
standing & ^{it was noted that he} fully understands the nature of objects
& can easily express his ideas & wants.
He speaks correctly & without any mis-
take or need to correct himself. He understands
everything said to him even when spoken to
quickly or in complicated sentences. He can
read fairly well but says it is much more
difficult than it was before he became lame
with fits. ~~He~~ The effort tires him, & he often
has to lay a newspaper down. He can see the
words quite well but has to read it again & again
before he can understand the meaning. His right
hand being unaffected, he ~~can~~ can form the
letters quite well & legibly, but complains of great
difficulty in expressing his thoughts in writing.
He says "I either leave out letters or put in
wrong letters & sometimes I put in wrong
words." When asked to write a letter he begins
deliberately & slowly, stopping frequently
to read over what he had just written &
~~then~~ often stopping to think how to write
certain words. ~~A letter~~ A letter that I had
he wrote contained the mistakes he mentioned
a letter omitted here & there & words placed
wrongly. He can read what he has writ-
ten fairly well & recognizes his mistakes
but cannot correct them. He makes similar
mistakes in writing from dictation. He can
copy writing very well or print into writing.

~~He~~ ~~at~~ ~~an~~ ~~examination~~ he seemed a healthy
man in general; smell, sight, & taste were
normal; ~~and so were the~~ ^{the} ~~after disc~~ ^{eye-movements} ~~presented no abnorm~~
~~ality.~~ ^{the} ~~ocular movements were normal~~
The visual fields were repeatedly taken ^{with} by the
perimeter & in general were of quite normal
The grasp was R 90, L 75. Sensation was
thought to be very slightly deficient ^{on the} ~~throughout the left~~
~~side.~~ The left knee-jerk was the more active,
but there was no clonus; the left plantar reflex
was extensor, the ~~right~~ right flexor.

He was evidently an intelligent man,
but after his attacks he was apt to use
wrong ~~words~~ words & to make mistakes in
reading & still more in writing, as will be
described. The attacks occurred almost daily
at first. They varied much in severity.

In the slightest the eyes became fixed,
without deviation; the left side of the face became fixed
in ^{slight} ~~conv~~ spasm; the head turned slowly to the left,
consciousness seemed dull for about a minute,
but he kept his seat on a chair; ~~nothing~~ ^{rather} more was noticed. ~~Then~~ In attacks
~~slightly~~ more severe, the left arm became rigid
& the elbow flexed; without movement in the
wrist or fingers. Any movement of the lips
terminated each slight attack, ~~but~~ no sensation
of small cords be ascertained, & ~~only~~ ^{but} sometimes by
taste of an unpleasant bitter taste in the mouth.

Turner

Bevor

arousen

Mittleship

Sanders

Amor

South and chewing movements aneurism
destroyer aneurism

Bazard look turn t

Myr Turner

Sapp

Quake man t small

Westphal t. alfalfa bulb

Gower t.

t

Sopening

Colman

aneurism

dreamy

smoking

meningitis

Hamilton

aneurism

smoke or petio.

NARCOLEPSY.—The term "narcolepsy" has been used in several senses, but is best applied to a condition, for which some name is needed, in which there is a tendency to fall into sound sleep for a short time, ~~usually for a few minutes, rarely for an hour or more.~~ The condition is distinguished from trance by the brevity of the attacks ~~of sleep~~, by their ~~strong~~ tendency to recur, and by the fact that the patient can commonly be roused ~~at any time from the condition.~~ In some cases, the malady has been apparently the result of some peculiarity of the nervous system. Thus, in one recorded case, a man had been liable, all through his life, to such attacks of sleep, which were induced by most varied influences. He had a nasal fistula, and whenever a probe was passed down this, he fell asleep. ~~In other cases~~ The affection comes on in adult life, and ~~passes away~~ after lasting for a few months or years. Thus in one girl, the attacks commenced at sixteen, and continued till she came under treatment at twenty-two. She would suddenly feel drowsy, her eyelids drooped, in a moment she was sound asleep, and after about five minutes woke up quite fresh. Intensely vivid dreams accompanied the sleep, but she never could recall their details; sometimes she would speak aloud in the dream. ~~There was no change in the colour of the face.~~ After deficient sleep at night, the sleep attacks lasted longer, sometimes even half an hour. She could be readily waked at any time, and by an effort could keep off the attack, but then could not help yawning continually, and felt uncomfortable. ~~There was never headache.~~ Such attacks occurred four or five times a day, when she was engaged in sedentary occupations or studies, but when she led an active life, as on a holiday, they would be absent for two or three weeks.

The characters that separate ^{is distinct from} this affection from ordinary trance have been already mentioned. It is most likely to be confounded with minor epilepsy, but from this it is sharply distinguished by the perfect resemblance of the attacks to ordinary sleep in their onset and character. When a feeling of somnolence is the warning of an attack of true *petit mal*, the unconsciousness lasts only a few moments. An active life with change of scene is probably the most important element in treatment. ~~In the case mentioned above, no drugs did so much good as a combination of caffeine and nitro-glycerine, which almost entirely arrested the attacks.~~

* The term "narcolepsy" has been unwisely applied also to such epileptic attacks.

† Some forms of lethargic sleep have been distinguished as "Gayet's disease" (a rare form, fatal in a few months, somewhat like African lethargy), "Gerlier's disease" (summer somnolence among the Swiss), and "Wernicke's disease," in which sleep has been the result of acute inflammation of the nuclear grey matter beneath the corpora quadrigemina. The fact that gradually deepening coma may be due to such an organic lesion does not justify the ascription of all forms of lethargy to such a process (Mauthner, 'Wien. Gesellsch. der Aerzte,' May, 1890).

*a nervous idiosyncrasy
& has existed throughout life*

*and nervous stimulants
seem useful*

as/

AFRICAN LETHARGY.—A curious malady, presenting similar somnolence but of very different nature, is the "sleeping sickness" of the West Coast of Africa, met with chiefly in the Congo and Sierra Leone regions. It is said to prevail less at the coast than inland, and to be especially severe in the valley of the Congo. It affects exclusively negroes (by whom it is much dreaded), and occurs in both sexes and at all ages, but is most frequent in males between twelve and twenty. Except that depressing emotions seem to predispose to it, the proximate causes are entirely unknown. Europeans living in the same localities are exempt. Swelling of the cervical glands sometimes occurs at the onset, and they are excised by the native doctors as a remedial measure; but the condition is not invariable, and its influence is doubtful. The general health at first is good. There is a gradually increasing tendency to somnolence, and the patient will fall asleep at various and unusual times, at his work or over his meals. A tendency to persistent drooping of the upper lids sometimes occurs at the onset. At first he can be roused, and if treated by cutaneous stimulation and purging, the symptoms may be removed for a little time, but they soon recur and increase in spite of treatment; the periods of sleep grow longer and more frequent until the patient is always asleep, is reduced to a merely "vegetative" existence, and ultimately cannot be induced to take food. He gradually emaciates, and dies at the end of three, six, or twelve months from the onset of the symptoms. Just before death the disposition for sleep often ceases. The disease is extremely fatal. Guérin met with 148 cases, all of which died. The observations of Gore and others place the mortality somewhat lower—at about 80 per cent. Post-mortem examination has revealed only hyperæmia of the arachnoid, slight signs of chronic meningitis, but no considerable excess of fluid within the ventricles or outside the brain. The cerebral substance is usually pale. *Filaria sanguinis* has once been found in the blood,* but apparently only as a coincidence. In some cases with organic brain disease, it would seem as if the symptoms had acquired a special character from the cause of the lethargy, whatever it is. Poisoning by a fungus growing on cereals has been suggested, but the malady, once established, progresses steadily in spite of change of residence and of diet. No treatment appears to influence the symptoms. Only one observer (McCarthy) has seen good from excision of the cervical glands. This mysterious affection—mysterious in more respects than are recognised by those who have theorised about it—clearly needs more systematic investigation than it has yet received.†

It is clear, moreover, that a tendency to somnolence, increasing gradually to lethargy and coma, is the expression of morbid blood-states more

* S. Mackenzie, 'Lancet,' 1890.

† The most recent account of the disease is by Junker v. Langeegg, 'Wien. med. Wochenschr.,' 1891.

often in tropical countries than in temperate regions. Such cases have been observed in South America, attended with pyrexia, with only trifling secondary changes in the brain, but with the enlargement of the spleen that shows a morbid state of the blood. The symptoms of these cases differ, in essential particulars, from the African malady.

HYPOCHONDRIASIS.

Hypochondriasis is a morbid state of the nervous system in which there is mental depression due to erroneous ideas of such bodily ailments as might conceivably be present. This limitation is necessary to distinguish the condition from those forms of actual insanity in which there is a delusion of the existence of some impossible ailment—impossible either in its nature or else by reason of its incompatibility with life. A patient, for instance, who thinks that his spinal cord has become detached from the brain, or who believes that his throat is hermetically closed, is insane, not hypochondriacal.* The term is, ~~moreover, almost exclusively~~ ^{chiefly} applied to the condition above defined when this is met with in men. A similar state in females is regarded as forming part of "hysteria." The name has come down to us from the times of Hippocrates and Galen, and, like "melancholy," had its origin in the idea that the conscious life was largely influenced by "vapours," &c., produced in the abdominal organs. It was not until the early part of the present century that the disease was generally admitted to be one of the brain.†

CAUSES.—The affection ~~consists in a particular disposition of the brain, which leads to habitual anxiety without adequate cause, and to the concentration of this anxiety on the individual's own health.~~ ^{generally rests on an undue tendency to} ~~This morbid tendency is often a matter of individual temperament,~~ ^{personnel} which may be the result of neurotic inheritance; the most severe forms occur in persons whose family presents a tendency to insanity. In other cases there is no morbid heredity, but the state is acquired through persistent ill-health, which sets up the morbid mental habit of self-attention and concern. These two causal varieties may be termed the "acquired" and the "temperamental" forms. The age at which hypochondriasis comes on varies according to its causes. It is scarcely known in childhood, but when due to inherited tempera-

* It is unfortunate that alienists sometimes apply the term to the cases of insanity with somatic delusions; this is contrary to its customary use, and productive of confusion.

† An interesting sketch of the history of hypochondriasis is given in Gull and Anstie's article on the disease in Reynolds' 'System of Medicine,' vol. ii.

ment it often becomes established quite early in adult life, ~~and sometimes is even distinct in youth.~~ It may, however, commence at any period, ~~and the acquired form often commences in or after middle age.~~

Hypochondriasis seldom develops without some exciting cause in the shape of a distinct bodily ailment, although this is often very trifling in degree. It suffices, ~~however,~~ to give rise to unpleasant sensations, which excite attention and ~~give rise to concern.~~ Dyspeptic troubles are the most common ~~excitants;~~ others are palpitation of the heart, and ~~weakness~~ of the nervous system; ~~but there is hardly any part of the body in which functional disturbance may not be the starting point of the malady.~~ When the attention has once become fixed on a bodily sensation, and this has become the basis of a fancied malady, other feelings elsewhere are soon perceived; corresponding ailments are imagined, and any trifling disorder is magnified into a serious disease.

undue sensibility

which may promote Conscious sensations from any part or organ; and the sequence of imagined disease grows more and more usually fostered by the study of medical writings

SYMPTOMS.—In hypochondriasis there are three chief elements: excessive anxiety regarding ~~the~~ personal health; undue attention to any ~~indications of derangement, especially to the various sensations emanating from the organs of the body; and lastly,~~ as the consequence of these, ~~there is~~ some false idea that disease is present which does not exist. Occasionally the last is absent; the patient may know and realise that he is free from any serious malady, but he is rendered miserable by the continuous discomfort he experiences.

This discomfort, as already stated, often has its origin in some actual disorder, but it is always vastly increased by the habitual attention paid to the sensory impressions proceeding from the organ deranged, or from other parts when there is no local disorder. The influence of attention has been already mentioned in the account of "cephalic sensations" (p. 863), which often form part of the symptoms of hypochondriasis, and the statements there made are applicable to all the various sensory impressions from other organs that disturb the hypochondriac's life. It is this continuous attention that perpetuates the morbid state, and renders its treatment so difficult. Not only are sensations attended to until consciousness is continually under their influence, but the patient is always searching for indications of the ill-health he dreads. ~~He scrutinises his tongue before breakfast and his evacuations after breakfast, and notes, during the day, the influence of each meal on his abdominal feelings, of exertion on his pulse, and of mental work upon his head.~~ It is easy for ~~the~~ ^{him} hypochondriac thus to collect each day a series of "symptoms" which he ponders over, endeavours to interpret, and sometimes records, ~~in minute detail, in a diary.~~ As an instance ~~of this morbid self-observation and attentive regard to every real or fancied sensation,~~ I may quote *verbatim* the description given to me by one of these unfortunate sufferers, in whose physical condition no other flaw could be found

than trifling occasional indigestion. "I do not breathe free; I do not breathe clear. After I did my work yesterday there was a pain in my temples and in the back of my head. There is a little pain in the heel when I press upon it. I have a sensation of tightness round the sides of the chest. I have also felt slight tightness about the knees. The appetite is not the thing at all. There is slight distension; I have not found it to-day, but I did yesterday. Last night I felt the food in my throat, and a noise in the chest such as you feel in the ear. My head is hot on the top now. Talking even for a few minutes seems to affect the eyes, and an uncomfortable feeling comes in them. This morning in the train, too, after it stopped, I seemed to feel for a moment as if I was going backwards and forwards. My forehead gets hot when I talk. Some days ago I had an uncomfortable feeling in the loins, and afterwards in the bowels, and a week ago I had some pain in the armpit." ~~This patient had no false idea of local illness; his symptoms were too vague and varying to permit him to entertain the idea, but it was impossible to convince him that there was not some grave general derangement of his health.~~ In the subjects of the temperamental form, with neurotic heredity, there is often slight real derangement of the nervous system, headaches, neuralgic pain, vague giddiness, noises in the ears, sensations of weakness of the legs, &c., which keep up the mental state. Similar troubles occasionally characterise also the acquired form, especially in gouty subjects.

~~In many cases, moreover, slight derangement of the functions of some one organ constitutes an anchor for anxiety; attention is focussed on the local sensations so that they attain an ever-growing intensity, and the conception of a definite ailment becomes fixed, sometimes ineradicably. In some instances the idea has its origin not so much in the sensations of the patient himself, as in the fact that circumstances have brought the disease prominently under his notice. This influence is, indeed, often operative when there is no persistent hypochondriasis. Most medical students imagine, at some period of their career, that they are the subjects of heart disease. But the study of medicine provides the antidote, as well as the bane, to all healthy minds. With mental instability, however, an idea often becomes firmly rooted. An elderly man believed himself to be the subject of diabetes, from which a friend had died, and he persisted in his belief, in spite of the assurance that there was no sugar in his urine.~~

The subjects of this disorder may have the appearance of perfect health, but ~~those in whom the condition is of long duration~~ are often careworn and depressed in aspect, and are preoccupied in manner. Their sufferings are described in language that is ^{seems} obviously exaggerated. The amount of depression is often very great, but in cases of pure hypochondriasis there is scarcely ever the suicidal tendency common

with no discoverable disease, the unexplained sensations may attain a degree that makes life seem unendurable.

such as the death of a friend,

and may lead to suicide & even in pure hypochondriasis

grave. Occasionally, a temperamental hypochondriac may be met with who describes life as shorn of all pleasure by his sufferings, and asserts that he has suicidal impulses; but if the latter really exist, there is generally more than pure hypochondriasis.

To describe in further detail the symptoms of these cases would be merely to multiply particular illustrations of the general facts above stated. There is hardly a malady that is not at times the subject of the morbid fancy. The general facts are true of all; it is only the applications that vary. One class of cases, however, commonly included in this category, deserves special mention, the cases of sexual hypochondriasis. Most of those who wrongly imagine that they have some ailment of the sexual organs do not really suffer from hypochondriasis. This erroneous belief is justified because it is founded on evidence which is adequate to cause the belief in those who have no means of estimating its value. The idea of spermatorrhœa, however, sometimes does concentrate the attention on sensations from those parts, and may set up true hypochondriasis, by the tendency to attribute to the supposed disease any discomfort that may be experienced. But there is no such disease as spermatorrhœa. The slow escape of the continuous secretion from the testicles is a normal and necessary event; neither it, nor the process of secretion, has the slightest influence on the nervous system.

DIAGNOSIS.—The diagnosis of hypochondriasis depends, in the first place, on the exclusion of the disease from which the patient imagines that he is suffering. ~~This of course involves an accurate knowledge of the symptoms of each malady, and it is necessary to exclude real disease before the obvious mental state can be allowed to influence the diagnosis.~~ The danger that a mistake of this character should be made depends on two circumstances. The first is the neglect of the consideration just mentioned; the manifest temperament leads to a hasty diagnosis, and deters from thorough examination. It is obvious that hypochondriasis does not exclude organic disease, and that hypochondriacs not only may, but at some time must, suffer from such disease. Secondly, error is sometimes due to the fact that the patient suffers from some uncommon malady, the symptoms of which are unfamiliar or unexpected. Thus a man with very distinct symptoms of diphtheritic paralysis had been told that these were all due to nervousness and fancy, because the patient was a member of the medical profession, the diphtheria had been doubtful, and it had been acquired in an unusual way—by inoculation.

The other diagnostic question is the distinction of hypochondriasis from insanity. ~~It is in the permanent hypochondriac, with a family history of insanity, that this difficulty arises.~~ The distinction depends chiefly on the reasonableness of the false ideas. A patient who has unpleasant sensations in the abdomen, and believes that he is suffering from cancer, in spite of the assurance that he is not, cannot on this

Regarding this, it need only be said that the patient's mental state should not preclude a thorough examination. A hypochondriac's fancies are occasionally true.

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account be regarded as insane. But a man who believes that his brain has been converted into sawdust is, by that belief, proved insane. Of course pure hypochondriasis may co-exist with other independent evidence of insanity, just as it may with any other malady. Many cases, however, are on the border line between soundness and unsoundness of mind—cases in which the patient believes that he is suffering from some disease, not in itself unreasonable, but still out of the question because the evidence of it would be tangible, and does not exist. ~~Such patients are often actually insane, although they cannot, and ought not to be, treated as such, unless the mind is unsound on other points.~~

PROGNOSIS.—The distinction between the two forms of hypochondriasis, the acquired and the temperamental, is important in regard to prognosis. The subjects of the latter form seldom recover; if one false idea is removed, another takes its place. The acquired form, on the other hand, ~~sometimes~~ ^{may} pass away altogether, and the prospect of recovery ~~from it~~ is good in proportion to the shortness of time it has existed, and to the degree of derangement of health in which it has arisen.

TREATMENT.—The details of the treatment of hypochondriasis have to be varied to suit the exigencies of each case; the principles are the same in all. The first thing is to correct whatever is actually wrong, ~~both in the patient's general health, and also in the organs to which his attention is directed.~~ In acquired cases, especially, there is often some weakness of the nervous system that may be lessened ~~by nerve-tonics and by an improvement in the patient's mode of life.~~ The second element is ~~the removal of the false ideas and the withdrawal of the attention of the patient from his physical condition.~~ The statements made regarding the treatment of "cephalic sensations" (p. 863) are applicable to all forms of hypochondriasis, and it is unnecessary to repeat them, or to apply them in detail to similar symptoms in other parts. ~~It is essential to make the patient realise how misleading bodily sensations often are regarding the actual condition of the parts from which the feelings seem to proceed, and how essential it is that the sensations should be disregarded. He should also be made to understand that his efforts to neglect them will not be at once successful, and that perseverance for a long time will be necessary. A grave responsibility rests upon physicians who are consulted by these patients. The mere consultation, even when the advice is wise, helps to perpetuate the morbid state, and it is certain that the mere consideration of "What have I to tell the doctor?" often does more in the way of harm than the "doctor" can do in the way of good. When all that can be done to remove actual disorder has been accomplished, it is often right to refuse to be any longer a passive party to the perpetuation of the symptoms. Sometimes such a~~

an effort to
banish

must be made to

refusal, if the grounds for it are made clear, will do more real good to the sufferer than can be achieved by any other means.

NERVOUS WEAKNESS; NEURASTHENIA.

It has become fashionable to apply the term "Neurasthenia"* to conditions of weakness of the nervous system. The use of the word has brought with it a tendency to regard the condition thus denoted as a definite disease. Books have been written about it, and it has been divided into numerous classes according to the character of the symptoms that are present, and the causes that give rise to it. It is convenient to be able to designate the condition by one word instead of two, but there is no more justification for regarding neurasthenia as a definite malady, as a disease due to a definite morbid process, or even as an affection characterised by a well-marked group of symptoms, than there is for adopting a similar course with regard to "debility" among general diseases. "Neurasthenia" underlies and may cause a large proportion of the functional diseases of the nervous system, and when "neurasthenic" patients seek advice, there are, in many cases, symptoms of nerve disturbance sufficiently definite in character to bring the case into some special category. Moreover, the conception has furnished material for the exercise of an unlimited amount of intellectual ingenuity, in a quasi-scientific process of elaborate description and multiple classification. Like other general terms, it has a tendency to hinder investigation, and to prevent the thorough study, and the due discrimination, of the nature of maladies, essential alike for the progress of medicine and the treatment of disease.

But the term has taken possession of medical thought in every country to an extent which compels its adoption. The cause which has rendered it so effective is obvious. While the undue extension of the use of the word, conspicuous in many quarters, is to be regretted and deprecated, there are many cases for which it is a convenient designation, and to which it may be applied without other disadvantage than the indirect sanction that may seem to be given to its unwise use. These are the cases in which there are symptoms of disturbance, or of impaired function of the nervous system, more or less persistent, almost infinitely various in their features, but which have in common the negative characteristic that they do not conform to any of the types that have received definite names. To include the latter under the term is for many reasons unwise. Hypochondriacal, hysterical, and many other varieties of "neurasthenia" have been described; but if the fact that impaired nutrition and strength of

or rather to all the symptoms which such depletion strength may cause. The word means only "depletion of nerve strength" but it is used

* Invented by an American physician, Dr. Beard.

The "empiricist" is effective in every department of art which acts on the human mind for its subject, and the term "neurasthenia" has become dominant for what it may be made to denote, by neglecting its connotation is ignored by the user, & is a vast nebula to its subjects.

the nervous system is to be made the ground for including under the term all the maladies which result from the condition, half the functional and nutritional affections of the nervous system should be described in one huge chapter under this designation. Equally unwise is it to distinguish classes according to the cause of the condition, when its manifestations present no peculiarities corresponding to the particular causes. Such varieties as "syphilitic neurasthenia," "influenzal neurasthenia," "malarial neurasthenia," have no scientific justification.

Even among the cases of nerve disturbance which cannot be placed in any recognised category, there are some that ought not to be included in this, or, indeed, in any other class of actual disease. Many persons possess a congenital peculiarity of nerve-function, for which they may seek medical advice, but which is not really morbid. Such is the persistence of the "shyness" of early life, or a tendency, life-long, to look on the darker side of things, or the vaso-motor activity which causes so many persons, all through the first half of life, to blush at the least emotion, and flush under every favorable physical influence.

When these, and the cases of a definite, long-recognised type, are excluded, those to which the terms "neurasthenia" and "neurasthenic" may be applied with convenience and without disadvantage are sufficiently numerous. In many the condition is distinctly "constitutional;" that is, the defect in the nervous system is inherent in the individual, and a similar ancestral tendency can often be traced. It dates, in some, from childhood; in others, it comes on after puberty or in early adult life, without any discoverable cause. Of the latter, a large proportion are females, who are unable to bear even the average strain of life, and break down in various ways. They may be raised to a little higher level of nervous health, but cannot be made really strong. Males sometimes suffer in the same way, but in them depressing influences can more often be traced. In both sexes the state often results from definite diseases of the nervous or general system or of other organs, which leave lasting impairment of nerve strength. Various influences, of occupation, amusement, dissipation, excess, may induce the condition; to enumerate these in detail is scarcely necessary.

The deficient strength implied in the name, persistent as it is in the cases thus designated, is always the expression of some degree of impairment of nutrition in the nerve-centres, brain or spinal cord, or both. It generally involves many parts, and is expressed by symptoms of great diversity; those of a certain class preponderate in some cases, but in many, various derangements of function alternate or co-exist. As a rule, deficient capacity for functional activity is associated with an undue readiness for slight action, which may even occur without its usual causes.

Sensory symptoms are the most common, but are confined to those

of "irritative" character. Loss of sensation does not exist in simple "neurasthenia," but subjective sensations are almost constant. Fatigue may be produced with undue readiness by muscular exertion and by mental effort. Headache is common, and so also are the varied cephalic sensations described at a preceding page. Pains in the back are frequent, sometimes felt throughout the spine, sometimes chiefly in the sacral region; they are usually dull and "aching" in character. Spinal tenderness, often at more than one place, is frequent in women; walking and standing readily increase the discomfort in the back, sometimes so as almost to incapacitate the sufferer. In the limbs, slight fugitive pains are often coupled with varied sensations of tingling, numbness, and the like, which may be greater on one side than on the other. *Muscae volitantes* annoy and alarm the patient, and when there is hypermetropia, the ciliary muscle is unable to maintain its usual contraction, so that "asthenopia" is added, and the use of the eyes begets uncomfortable sensations in them, superficial or deep, seldom painful, although actual pain may be felt behind the eyes or in the forehead, after their prolonged use. Auditory hyperæsthesia may cause pulsating tinnitus, or, in its central form, may render every loud sound distressing.

Muscular strength is only lessened in the severer degrees of nervous weakness, but the power of sustained exertion is generally reduced; fatigue is not only sooner felt, but is often a more unpleasant sensation than the fatigue of health, and whatever pain or discomfort to which the sufferer is liable is apt to be induced. Even talking may quickly cause a feeling of weariness and any cephalic sensations to which the patient is liable. Many of these sufferers habitually talk in a low voice, as if every sentence involved an exertion almost beyond their strength. A sense of muscular inertia and powerlessness is very frequent, especially in the earlier part of the day, when there is no real lack of strength; the effort needed for any exertion may seem beyond that which is possible.

Symptoms of motor irritation or over-action are very rare except in trifling form. The sense of stiffness in the limbs is not accompanied by any real rigidity. Some sufferers are very liable to cramp in the calves or feet. Especially common is the quivering contraction of some muscle, or part of a muscle; the orbiculares, glutei, thigh or calf muscles are frequent seats. It seems due to the intermitting, instead of continuous, release of the nerve-force which causes muscular tone, and has no other significance than a slight general impairment of nutrition and function, although it often causes grave anxiety. Restlessness is common, and sometimes an unendurable sense of "fidgets" causes movements almost choreic in their character and constancy, accompanied even by actual starts of the limbs.

When there are sensory symptoms in the legs, or undue readiness and degree of fatigue on standing and walking, the knee-jerk is often increased, perhaps from the diminished resistance in the reflex centres,

due to the fall of nutrition below the normal standard. No foot-clonus can be obtained. The state of the sexual functions, in the male, depends chiefly on antecedent influences, but the mental pre-occupation, engendered by the varied sensations which absorb the mind, and the sense of disability, deepened by a fear of impending calamity, often cause an apparent failure of desire and power, superficial only, and ceasing with its cause. In women, the state of the menstrual functions depends more on the general health than on the state of the nervous system, although the latter may be depressed by mental suffering, or by the apprehension of grave results of irregularity.

But as the most special, elaborate, and complex part of any system is that most prone to suffer, and exhibits in greatest degree the effects of general impairment, the functions of the brain seldom escape disorder in these cases, and often the cerebral disturbance is the most pronounced manifestation of the state. All the functions are apt to suffer; mental power is lessened, the ability to concentrate the attention fails, memory is imperfect, and the influence of the will upon it, in "recollection," is weakened in especial degree. There is as much conscious difficulty in mental as in physical exertion, and, in proportion as the sufferer shrinks from the unpleasant effort, the resistance that has to be overcome seems to increase. All cerebral processes that depend on the will quickly cause fatigue, which is expressed by varied sensations, psychical or cephalic. The subjective discomfort in the limbs and back depress the brain, on which, often, the effects of the varied sensory symptoms seem to be focussed. A large proportion of these, indeed, arise in the sensory centres of the brain, and the interaction of all its parts is such that the disturbance of one region influences the whole. The purely mental functions suffer with the rest; mental depression is conjoined with irritability, and every annoyance is magnified into a disaster. While actual giddiness is uncommon, a sense of impending giddiness sometimes causes much distress, and may even, by some indirect mechanism, induce uncertainty of movement.

Visceral symptoms, pain, discomfort of many kinds, and even definite disorders of subordinate functions are frequent, but those of the digestive system are of especial importance. They are to be recognised, as distinctly augmenting other symptoms, in about two thirds of the sufferers, and a close relation between gastric disorder and cephalic discomfort is especially conspicuous. A feeling of shortness of breath, or oppression at the chest, is often complained of. Secondary disturbance of the action of the heart is frequent, and the readiness with which the vaso-motor system responds to central and visceral derangement, is the source of many symptoms, flushing, pallor, sense of heat and cold, chills, and imaginary fever, are common complaints. Sleep is seldom good, restlessness and insomnia increase the nerve prostration, and often, when the patient does sleep, he wakes un-

refreshed, weary, and languid. Starts often hinder the advent of sleep, and this, when it comes, is disturbed by dreams or interrupted by sensations of tingling in the hands, sometimes with a feeling of contraction in them when there is none, sometimes with an actual contraction such as that described at p. 704 as "nocturnal tetany."

The further development of such effects of malnutrition usually takes the form of one of the definite functional ailments already described, many of which, as neuralgia, hypochondriasis, hysteria, really consist in a high degree and elaborate development of the effects of the condition now under consideration. The description of the symptoms might be multiplied indefinitely, and yet, so various are they that no description, however lengthy, would embrace half the varied manifestations of mere nervous weakness.

PROGNOSIS.—The prospect that the condition will pass away must depend on its duration, and especially on the degree to which it is the effect of influences that can be arrested, or, on the other hand, is the manifestation of an inherent inability of the nervous system to reach or maintain a proper level of nutrition and function. Each case must be made the subject of separate study, and the causal influences must be compared with the extent to which the interaction of the symptoms tends to keep them up, and with the possibility of emancipating the sufferer from the pathogenic influences, external to and within himself. When patients have been freed from the acquired state, it is extremely prone to return on any impairment of the general health or strain upon the nervous system.

TREATMENT.—The advice that has been given regarding the treatment of hysteria, hypochondriasis, neuralgia, headache, and cephalic sensations, embraces most that can be done to remove the condition of "neurasthenia." All influences conducive to physical strength should be secured, and all causes of depression and exhaustion of the nervous system should be sought for and eradicated. Powerful causes (as sexual excess) are sometimes quite unrecognised as such by their victims. A sea voyage and mountain air afford the most potent means of restoring the strength of the nervous system, if this has been lately lost. In all cases, rest from work that is obviously depressing should be secured, and the rest made more effective by such mental and physical influence as can be secured by change of personal and local surroundings. In many, however, some mental occupation is better than perfect freedom to devote attention to the sensory discomfort. Tonics should be given of such form as is indicated by the symptoms, and drugs that lessen the instability of the sensory structures, without depressing the system, are often of service, such as small doses of Indian hemp, given with bromide, belladonna, or chloride of aluminium. The effect of nitro-glycerine, in augmenting the arterial blood-supply, and thus facilitating improvement in nutrition

and the influence of tonics, is often very conspicuous. The physical influence of rest and massage are often useful on account of the complete repose given to the motor nervous system. Hydropathic treatment is of service in many cases, not only for its direct effect, but also for the conditions which it indirectly secures. The details of the treatment that may be adopted with advantage in various cases, need not be enumerated because they chiefly consist in the application, to the special state, of the measures that are within the familiar knowledge and capacity of every well-trained practitioner.

MOTOR NERVES OF THE EYEBALL.

(THIRD, FOURTH, AND SIXTH NERVES.)

The motor nerves of the eyeball are the third, fourth, sixth, and sympathetic. The external muscles are supplied by the three former, the internal muscles by the third nerve and the sympathetic. ~~It will be convenient to reserve for special description the affections of the internal muscles, and to consider first the general symptoms of paralysis of the external muscles, the special symptoms of disease of each nerve, and the causes and treatment of such disease.~~

When there is no muscular contraction the eyeball is in the position of rest, ~~and in this position, under normal circumstances, the line of vision is at right angles to the plane of the face.~~ This is termed its "primary position." ~~Any deviation from the primary position is due to muscular action.~~ Many movements are produced by more than

other than the normal tone,
The upper eyelid is raised by the levator supplied by the third nerve, and a slight influence is exerted by the plain fibres which descend from the fascia to the tarsal cartilage & are innervated by the sympathetic. The upper lid is lowered, ~~by~~ & the lower raised, by the orbicularis but considerable effect on them is produced by the convexity of the eyeball, which presses on the tarsal cartilages. The orbicularis is probably supplied by fibres of the facial nerve which arise from the highest cells of the nucleus *mid-ventr.*

Their action & combination are described in books on physiology

one muscle. Each of the straight muscles turns the eyeball in the direction indicated by its special name, but the superior and inferior recti, in consequence of their origin being nearer the middle line of the body than is their attachment to the eyeball, tend to turn the eyeball inwards and to rotate it, so as to incline the vertical axis of the globe. The upper end of this vertical axis (which it is convenient to indicate by *S.*) is inclined inwards by the superior rectus, outwards by the inferior rectus. This tendency to rotation is counteracted by the simultaneous contraction of the oblique muscles; the inferior, which rotates *S.* outwards, acts with the superior rectus; the superior oblique, which rotates *S.* inwards, acts with the inferior rectus. The oblique muscles also counteract the inward movement of the globe on its vertical axis, caused by the superior and inferior recti, and

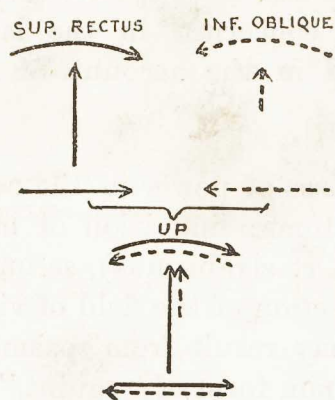


FIG. 86.

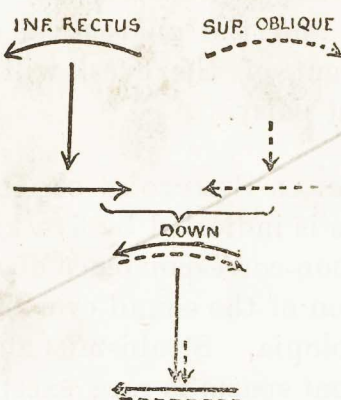


FIG. 87.

FIG. 86.—Action of elevators of right eye; the arrow-heads indicate the direction of movement produced by the muscles named. The curved lines represent the rotation on the antero-posterior axis, the vertical lines that on the transverse axis, and the horizontal lines that on the vertical axis of the eyeball.

FIG. 87.—Action of depressors of right eye. See last fig. and description.

they also assist the recti in moving the eyeball upwards and downwards, having a slight elevating and depressing action (see diagrams, Figs. 86 and 87). When the eyeball is moved diagonally the lateral rectus acquires a rotating influence, but this is counteracted by the opposite tendency of the associated superior or inferior rectus. In the case of slight inward diagonal movements the rotating tendency of the superior and inferior recti corresponds with that of the lateral muscle concerned, and must therefore be counteracted by an oblique muscle.

~~The origin of the eyeball nerves has been already described (p. 52).~~ The tract of grey matter beneath the aqueduct of Sylvius, which gives origin to the third nerves, is continuous behind (*i. e.* below) with that of the nuclei of the fourth nerves, and is connected with the apparently separate nucleus of the sixth by the posterior horizontal fibres. We have as yet no complete evidence of the precise distribution of function in the nucleus of the third nerve in man. The conclusion of

Hensen and Voelcker,* ^{is nearly that here indicated} from their experiments on dogs, has been stated at p. 53. A Mid-Line.

case investigated by Kahler and Pick† suggests that the arrangement of centres is not simply from before back, but that there is also a lateral difference of function. They suggest the accompanying modification of the plan of Hensen and Voelcker.

Ciliary muscle.
Sphincter iridis.
Lev. palp.
Rect. Int. Rect. sup.
Rect. infer. Obliq. inf.
(Obliq. sup., 4th n.)

The evidence is strong that the anterior part of the nucleus controls accommodation, and the next the action of the iris to light, and that these functions are subserved by the anterior fasciculi of the roots of the nerve,‡ while a small hæmorrhage behind these centres has paralysed the two internal recti.§

The relation between the internal rectus and the opposite external rectus, and the anatomical arrangement concerned in the lateral movement of the eyes, will be described in the account of their conjoint palsy.

GENERAL SYMPTOMS OF PARALYSIS.—Loss of power in the ocular muscles is indicated by five kinds of symptoms: limitation of movement, non-correspondence of visual axes (*i. e.* strabismus), secondary deviation of the sound eye, erroneous projection of the field of vision, and diplopia. Strabismus and diplopia may result from spasm, but persistent spasm is rare except as the common form of "squint," and in this diplopia is absent, for a reason that will be presently mentioned. An abnormal state of the eyeball muscles must not be assumed from the posture during comatose states. In sleep stupor, as in common sleep, the globes may diverge and be directed upwards; one may be higher than the other, and on the patient being roused they may at once assume a normal position. Moreover, a congenital difference of position is far from rare, and may readily mislead; it may also result from old and forgotten injury.

Limitation of Movement.—If a muscle is paralysed, the eyeball cannot be moved so far as normal in the direction of action of that muscle. In complete palsy the defect is absolute, ~~the eyeball cannot be moved beyond mid-position unless slight movement by other muscles is possible.~~ After a time the globe is fixed in the opposite position by contraction of the unopposed antagonist of the paralysed

But there is an increasing suspicion that the arrangement is more complex and that fibres of each third nerve arise from the opposite nucleus, & that the groups of cells correspond to movements rather than to muscles

there occurs

* 'Arch. f. Ophth.,' Bd. xxiv, 1878, p. 1. See also p. 53.

† 'Prag. Zeitsch. f. Heilk.,' 1881, p. 301, and 'Cent. f. Augenh.,' 1883. The chief evidence is that in two cases of nuclear disease and palsy of some muscles there was a marked difference in the affection of the medial and lateral fibres of some fasciculi. The observed grouping of palsy in disease is seldom significant.

‡ It is not probable that the position of these centres in man is as far forwards beneath the floor of the third ventricle as it is in dogs, and as it has been assumed to be in man by some writers. The nuclei of the third nerves cannot be traced farther forwards than the position of the posterior commissure.

§ Bouchard, 'Rev. de Méd.,' 1891.

~~muscle.~~ Thus in paralysis of the external rectus the eyeball cannot be moved outwards, and after a time is turned inwards by the contracture of the internal rectus. If the paralysis is incomplete, movement is deficient in proportion to the amount of palsy, ~~and towards the limit of movement the motion is often jerky ("paralytic nystagmus") from remissions in the contraction, analogous to tremor in a weak limb.~~ When both eyes are open, and acting together, the extent of movement is usually less than when the other eye is closed and the patient uses the affected eye only, ~~and is compelled to strive to "fix" the object with it.~~

Strabismus and Secondary Deviation.—In consequence of the defect in movement, the axes of the two eyes do not correspond in positions that necessitate the action of the paralysed muscle. If a lateral rectus is affected, the axes converge or diverge, producing convergent or divergent "strabismus," which increases, the greater the degree of movement of the sound eye in the direction of action of the paralysed muscle. Paralysis of the external rectus causes convergent strabismus, that of the internal rectus divergent strabismus. ~~The deviation of the axis of the paralysed eye from parallelism with that of the sound eye is termed the "primary deviation."~~

This is ~~If the sound eye is prevented from seeing the object, and the patient looks at this (is made to "fix" it) only with the affected eye, the sound eye is moved still farther in that direction, and hence the deviation of the visual axes is increased. This is called the "secondary deviation." Its existence and amount may be best ascertained by subsequently covering the paralysed eye, and making the patient "fix" with the unaffected eye, which has then to move back to its former position. The hand, or a piece of paper, may be so placed as to intercept the vision of the one eye, while leaving it exposed to observation. A piece of ground glass placed over the eye answers the same purpose, and permits observation.~~

~~The occurrence of secondary deviation depends on the fact that normally two muscles which act together are equally innervated for a given movement. If one is weak, and an effort is made to contract it (as in fixing with that eye), the increased innervation influences also the other muscle, and causes an undue contraction. It is as if a rein acted equally on a hard-mouthed and a tender-mouthed horse yoked together; the effort to make the former deviate would cause an excessive deviation of the latter. The secondary deviation is a delicate test for weakness of an ocular muscle, and may reveal its existence when the primary deviation is too slight to be observed.~~

overaction ~~Moreover these deviations afford a distinction from strabismus due to muscular spasm, such as ordinary squint. In the latter the deviation exists in all movements, in paralytic strabismus, only in those movements that call into action the paralysed muscle. In ordinary strabismus, the deviation is the same whether the patient fixes with one eye or with the other; in paralytic strabismus fixation with the normal eye evokes the primary, and that with the affected eye the~~

too

its occurrence

weakness

which looks alike

secondary deviation: these two are similar in direction, but opposite in character, the primary being a defect, and the secondary an excess of movement.

Erroneous Projection.—An interesting and important effect of the increased innervation that causes the secondary deviation, is erroneous projection of the visual field. We judge of the relation of external objects to each other by the relation of their images on the retina; but we judge of their relation to our own body by the position of the eyeball as indicated to us by the innervation we give to the ocular muscles. When there is no muscular effort, and the eyes are at rest in mid-position, we know, to use popular language, that an object at which we are looking is opposite our face; that is, that a line from the centre of the field of vision to the macula lutea is perpendicular to the facial plane. If we turn the eyes towards one side, we know that an object in the middle of the field is to the side of its former position; how far to that side we estimate by the degree of movement of the eyes indicated by the amount of innervation; and if we want to touch the object, the knowledge thus gained enables us to so contract the muscles of the arm as to effect the desired movement with precision. But the increased innervation needed by a weak ocular muscle gives the impression of a greater movement of the eye than has really taken place, and suggests that the objects seen are further from the middle line than they really are. If the patient then attempts to touch the object, the finger goes beyond it. The erroneous projection exists, of course, only in those movements of the eyes for which the weak muscle is necessary. It is greatest when the affected eye is used alone; it disappears altogether when this is closed. The erroneous projection is always in the direction of action of the affected muscle. The knowledge of the relation of external objects to the body, gained from the contraction (i. e. innervation) of the eyeball muscles, is one of the most important sources of guidance to the centres that regulate the muscular contractions for maintaining the equilibrium of the body. The erroneous projection of the field destroys the harmony between the visual impressions and the others that are correct; the resulting discord and its consequences may affect consciousness as the sensation of "giddiness." This giddiness depending on paralysis of an ocular muscle, is termed "ocular vertigo."

Double Vision.—We have seen that the field of vision of the paralysed eye is erroneously projected. Each object in the field is referred to a position other than that which it actually occupies, and corresponding to the degree of ineffectual innervation. But if the patient looks with both eyes, the field of the unaffected eye, being normally projected, does not correspond with the field of the affected eye; the images formed in the two eyes are mentally referred to different positions; objects are seen double. The patient is conscious of the doubling only of that object the image of which is formed at the macula lutea of one eye or the other, that is of the object which is

found if an attempt is made to touch the obj

This erroneous "projection" is in fact does not agree with other impressions & sometimes the discord causes giddiness "ocular vertigo".

un/

which is looked at

~~fixed by one eye, for which the eye is accommodated, and to which the attention is directed.~~ The image formed in the unaffected eye, being referred to its correct position, is termed the "true image," the other is termed the "false image." The distance between the two images ~~depends on the amount of the error of projection, and~~ corresponds to the degree of deviation of the eyeball from the position which it should occupy with that amount of innervation.

~~In consequence of this deviation of the two visual axes, the retinal image is formed at the macula lutea only in one eye (the fixing eye). In the other eye it falls upon a part of the retina away from the macula, at a distance from it which corresponds with the degree of deviation, and therefore with the amount of erroneous projection.~~ It was formerly thought that the circumstance that the two images are formed upon non-corresponding parts of the two retinae affords an adequate explanation of the diplopia.

but But the diplopia which occurs in secondary contracture of a muscle cannot be explained by the simple theory of non-correspondence of retinal areas. A muscle—the right external rectus, for instance—is paralysed, and when all the muscles are at rest and each eye should be directed straight forwards, the right eye is turned inwards by the permanent shortening of the unopposed internal rectus. The image of an object opposite the eye is formed on the macula

lutea of the left eye, but to the inner side of the macula lutea of the right. That this is no reason why there should be diplopia will be evident if we suppose for a moment that the patient has only the right eye, that this is normal and voluntarily turned to the left. It is clear that the object in the position supposed will be seen (not distinctly, because its image falls on an excentric part of the retina) in its actual place. So, too, if we suppose the patient to have only the left eye, and that this is directed towards the object; this will also be seen in its normal place. Thus there is no reason in the mere deviation of the eyes, and the non-correspondence in position of the retinal images, why the object should be seen in this

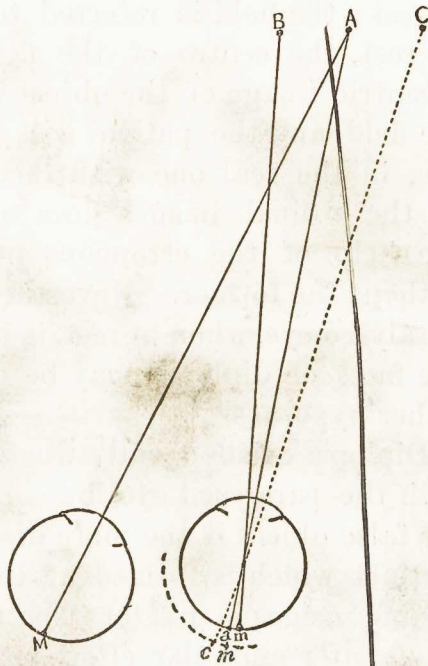


FIG. 88.—Diagram to illustrate the production of diplopia by the erroneous projection. In consequence of weakness of the right external rectus the image of A formed at the macula, M, of the left eye, falls within the macula of the right eye at a. But the effort corresponds to that necessary to bring the macula to m (in the broken arc). Hence B, whose image is at m, seems to occupy the position of A, and the image formed at a to the left of m is projected to the other side of A, and seems to occupy the position C, as if it were formed at c to the left of m.

eye in an erroneous position,—why there should be double vision. The explanation is that in the case supposed—of a normal eye being turned to the left—the field is mentally projected to the left, in correspondence with the degree of active muscular innervation; the centre of the field lies thus to the left, and the object straight in front is referred to its proper excentric position in the field. But in the case of the contracted eye all the muscles are at rest: the field is referred to the position normally corresponding to rest, the centre of the field is projected directly in front, the excentric image of the object is referred to an excentric position in the field, and the patient sees the object in a false position with this eye, in the real one with the other. Thus the non-correspondence of the retinal images does not explain the diplopia, except with the help of the erroneous projection. The latter does explain it without the former. Nevertheless, if there is no contracture, and the paralysed eye, when at rest, is in mid-position (as in recent paralysis), the facts of diplopia may be explained, and its laws formulated, on either hypothesis.

Diplopia exists equally whether the patient fixes with the normal or with the paralysed eye, but in the former case the true, in the latter the false object is the more distinct, that image being always the more distinct which is formed at the macula. The distance between the double images is greater when fixation is by the paralysed eye, because the greater muscular effort results in a wider error of projection; of this greater effort the secondary deviation of the non-paralysed eye is evidence. As Graefe has pointed out, if the patient moves his finger from the fixing (paralysed) eye to the apparent image, in trying to touch it, the line taken by the finger corresponds in direction with the visual line of the other (secondary deviating) eye; each represents the excessive amount of muscular innervation, and the degree of excess is a measure of the erroneous projection of the field of the paralysed eye.

In recent paralysis, diplopia exists only when the eyeball is in a position that needs the action of the paralysed muscle. In old cases with secondary contracture and permanent deviation, diplopia may occur during rest, or even during other movements. The distance between the double images increases, the farther the object is moved in the line of action of the affected muscle. As a rule it is the false image which appears to move from the other, and the direction of its movement is that of the action of the muscle.

The false image may be on the same side of the other as the eye by which it is seen (*homonymous or simple diplopia*), or it may be on the other side (*crossed diplopia*). When the muscle which is paralysed is an abductor, and the eyes therefore converge, the diplopia is simple; when the muscle is an adductor, and the eyes diverge in paralysis, the diplopia is crossed. This depends on the fact that, in the former case (of convergent strabismus from loss of abduction), the false image is

*in spasmodic strabismus
+ secondary contracture
of an ocular muscle*

*the principle is explained more fully in treatise
on ocular disease*

*+ the visual axes cross,
and the visual
axes do not cross*

formed on the side of the retina towards the sound eye, and is therefore projected on the other side of the real object. In the case of divergent strabismus from paralysis of an adductor the false image is formed on the side of the retina farthest from the sound eye, and is therefore projected on the side of the real object towards the sound eye. The student may remember the relation by the rule that when the visual lines (prolonged ocular axes) cross, the diplopia is *not* crossed.

In diagonal positions of the eyeballs the two images are not parallel; the false one appears inclined. ~~On the theory that the diplopia is due to the non-correspondence of the parts of the retinae on which the images are formed, the obliquity is explained by the fact that an abnormal rotation occurs in these diagonal positions. This rotation is due to the action of the oblique muscle, which should prevent rotation by opposing this tendency of the paralysed muscle. The obliquity is explicable also on the theory that the diplopia is due to erroneous projection: as v. Graefe has shown, the same principles apply to an abnormal rotation as to a defect in lateral movement.~~

The total field of vision which is brought into view by various movements of the eyes, the head being still, may be represented as a circle, the centre of which is at the fixing-point when the eyes are at rest, while the radii represent the various movements, horizontal, vertical, and diagonal. The circle is termed the "motor field." In paralysis of one muscle without secondary contracture there is diplopia only in one part of this field, that corresponding to the movement of the muscle. Thus in paralysis of the right external rectus there is diplopia in the right half of the field. The line which separates the area of single or double vision varies in direction with each muscle, and corresponds with the position of the axis of the eyeball around which the globe moves when the muscle is in action; but the correspondence is seldom exact, on account of the co-operation of other muscles in certain positions. To lessen the double vision the patient often holds his head in such a position as to call the paralysed muscle into action as little as possible. The movement of the head is on an axis which corresponds to the line of separation of single from double vision, and thus to the axis on which the eyeball is moved by the affected muscle.

Patients are not always conscious of double vision. If the muscular weakness is slight the two images may nearly correspond, and the result may be an indistinct outline to the apparently single image. Again, if the images are far apart the less distinct excentric image may be neglected, and the patient may be conscious ~~only of that which is seen by the normal eye. This is generally the case in old palsies. The detection of diplopia is facilitated by placing a red glass before one eye, preferably the sound eye, because the more distinct image seen with this eye is thus rendered less preponderant.~~ A strip of white paper is then held up in various parts of the motor field; the patient looks at it without moving his head: one image is

in consequence of the combined action of muscles of which only one is paralysed. The same is true when one of the straight muscles is affected, but in less degree: this is because an oblique muscle normally corrects the inclination its direction would cause & is increased with it.

especially

coloured

& using a strip of white paper to find the effect of the image & thus facilitate the detection of the double image.

seen red, the other white. ~~The two are thus more easily distinguished, and their relative position can be ascertained. If the red glass alone does not at first suffice, the view from each eye may be alternately obstructed (as by the hand held before the eye); and if there is double vision the position of the object will seem to alter as it changes from red to white. Such obstruction will, alone, often elicit unnoticed diplopia.~~

In employing diplopia in diagnosis we must first prove that it is not monocular, by ascertaining that it ceases when one eye only is used.* We then learn by the coloured glass whether the double vision is simple or crossed, whether the two images are on the same level or not, whether parallel, and the direction in which movement of the object increases the distance between them. When the images are side by side, and the diplopia is simple, the paralysed muscle is of the eye on the side towards which movement increases the distance between the images. If the diplopia is crossed, it is a muscle of the other eye. If the images are one above the other, the paralysed eye is that of which the image ascends from the other in looking up, descends in looking down. If there is secondary contracture of the antagonist of the paralysed muscle, diplopia exists through the whole range of movement in the plane of action of the muscles, *i. e.* during the action not only of the paralysed muscle, but also of its antagonist; but the distance between the images during movement remains the same on the side of the contracted antagonist, while it alters on the side of the paralysed muscle. It must be remembered that the vertical action of the superior and inferior recti is greatest in abduction of the eye, and that during adduction the oblique muscles also elevate and depress. Vertical movements are therefore in the line of action of two sets of muscles, and to discriminate between them we must move the object up and down both during adduction and abduction. Vertical diplopia in abduction is due to the superior or inferior rectus; if in adduction only, it may be due to paralysis of an oblique muscle.

Aid in diagnosis is also to be obtained by prisms, on the principle that a prism so placed that its base corresponds with the direction of action of the paralysed muscle, increases the distance between the double images; one in the opposite position tends to blend them. They may also be brought together by a prism placed before the unaffected eye in the position in which before the affected eye the diplopia is increased. Fusion by a prism before the affected eye removes the erroneous projection; fusion by a prism before the normal eye brings the projection of this field into the same error as the other.

~~PARALYSIS OF INDIVIDUAL MUSCLES.—We may now consider the symptoms of paralysis of each muscle. It must be remembered that~~

* The cause of monocular diplopia is still unknown. It is sometimes well marked when there is no discoverable abnormality in the refraction or in the fundus of the eye.

more than one muscle is often affected, and the symptoms are correspondingly combined; and that in diseases of the central nervous system, palsy and spasm are sometimes so associated as to render an exact diagnosis difficult and occasionally impossible. ~~This difficulty is often due to the circumstance that a given palsy was preceded by some other derangement of movement, as another slight paralysis and secondary contracture.~~

The paralyses described and figured are of the muscles of the right eye, except that of the external rectus, which is of the left eye for comparison with the paralysis of the opposite internal rectus.*

External Rectus, Left (Fig. 89).—The defect of movement is outwards, with convergent strabismus; secondary deviation is by the opposite internal rectus. The diplopia is simple, in the left half of the motor field; the two images are on the same level in the horizontal movement, and the distance between them increases as the object is moved to the left. In diagonal positions the false image is inclined, the images diverging from the horizontal plane. Towards the limit of movement there are often slight spasmodic rotatory movements by the unopposed oblique muscle. The head is inclined to the left.

Internal Rectus, Right (Fig. 90).—The defect of movement is inwards; the secondary deviation is by the opposite external rectus; the strabismus is divergent. Diplopia is crossed, and exists in the left half of the motor field.

* The diagrams of double vision are after those of Woinow ('Das Verh. der Doppelbilder,' Wien, 1870), modified a little by actual observation. The objects are figured as described by a patient when they are held between him and the observer. ~~If figured as seen by the patient, the position of the object is reversed.~~

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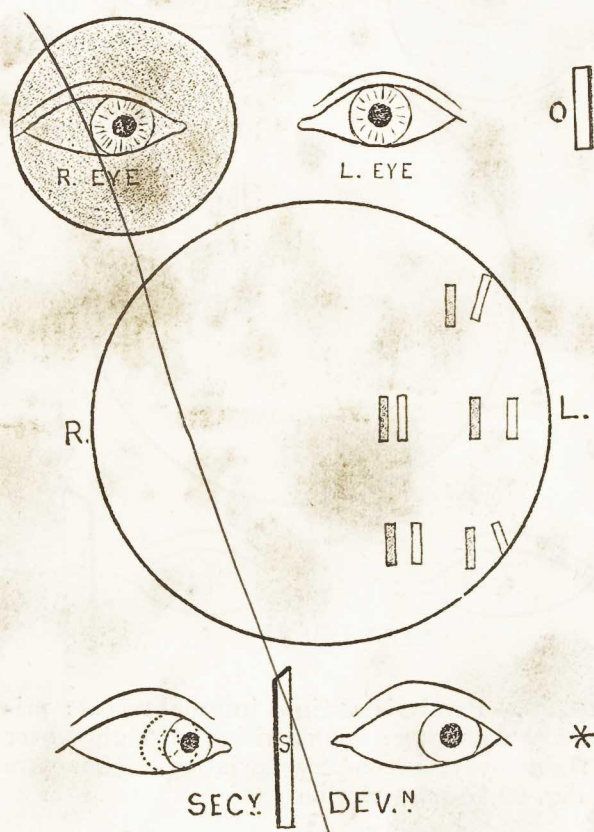


FIG. 89.—Paralysis of the left external rectus; coloured glass over right eye; primary deviation on looking towards an object (o) on the left; position of double images; secondary deviation of the right eye when the screen, s, obstructing the fixation of * by this eye, compels fixation by the weak muscle. When the screens are removed, the right eye, in fixing, moves back to the position of the dotted outline of the cornea. (In the diagram of diplopia the highest white [false] image should have been represented a little lower than the true image.)

In diagonal movements the images diverge towards the horizontal plane. The head is inclined towards the left.

Superior Rectus (Fig. 91).—Movement is defective upwards, especially upwards and outwards, and in attempting the movement the eyeball is rotated, the upper end of the vertical meridian outwards, by

the inferior oblique. Secondary deviation is by the opposite superior rectus. The diplopia is crossed and exists in the upper half of the motor field; the images are one above the other, the upper image being the false one, and receding from the other as the object is moved upwards. The false image is inclined. The difference in height is greatest in abduction, while the inclination is greatest in adduction. These differences are due to the influence of the inferior oblique, which rotates as well as elevates. The head is held backwards and inclined towards the sound side.

Inferior Rectus (Fig. 92).—Movement is defective downwards, especially downwards and outwards, because in the

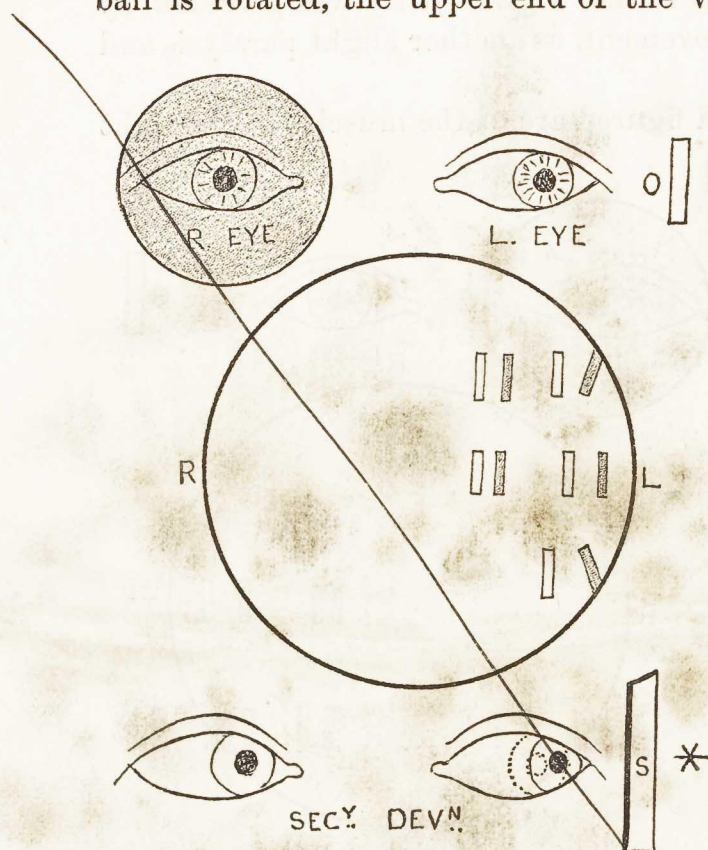


FIG. 90.—Paralysis of right internal rectus; primary deviation; double vision (red glass over right eye); secondary deviation. (Compare Fig. 89 and description.)

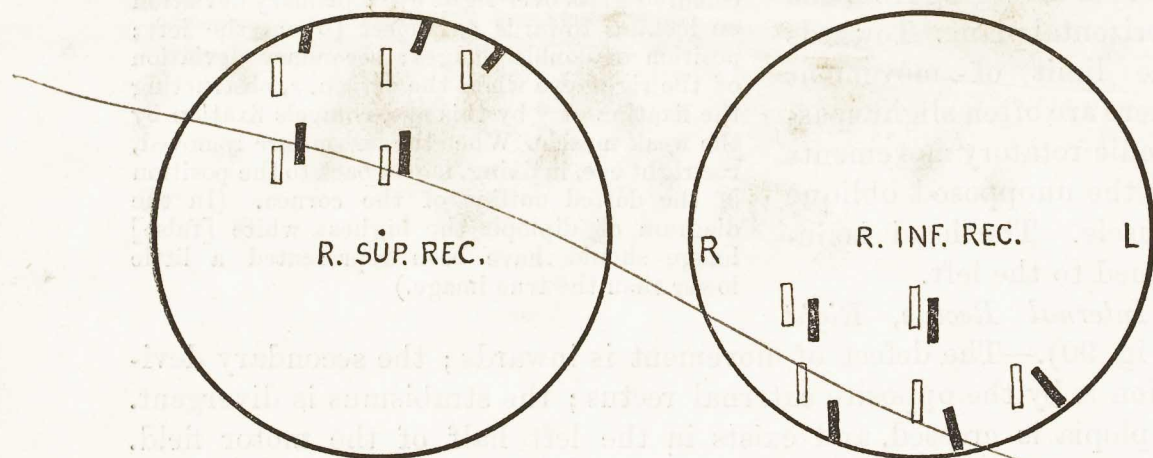


FIG. 91.

FIG. 92.

FIG. 91.—Diplopia in paralysis of right superior rectus. (The black image is that of affected eye.)

FIG. 92.—Diplopia in paralysis of the right inferior rectus. (Right image black.)

downward and inward movement the superior oblique is best able to supplement the paralysed rectus. The upper end of the vertical meridian is inclined inwards by the oblique, from the loss of the rotating action of the inferior rectus. Diplopia exists in the lower half of the field, especially in the outer portion; it is crossed, the images being near together laterally, but the false image is below the other, and recedes with the downward movement. The two images (if the object is held vertically) diverge from the horizontal plane. The difference of height is greatest on looking inwards; that of obliquity on looking outwards. Secondary deviation is produced by the opposite inferior rectus. The head is inclined downwards and towards the affected side. In an attempt to look down, the upper lid does not descend, but remains raised in correspondence with the position of the eye (see Fig. 94, and "Facial Paralysis").

Superior Oblique (Fig. 93).—Defect of movement chiefly downwards and inwards, since in this position the muscle exerts most depression, but it is often scarcely recognisable. Strabismus exists only below the horizontal plane, and is convergent by the unopposed inferior rectus, causing deviation inwards. Secondary deviation is chiefly by the opposite superior oblique and inferior rectus, &c., turning the eye down and in. Diplopia exists in the inner lower part of the motor field; it is simple; the false image

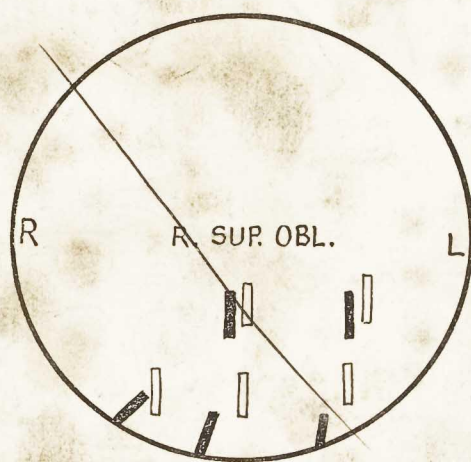


FIG. 93.—Diplopia in paralysis of right superior oblique. (Right image black.)

is the lower, and the images diverge towards the horizontal plane. The difference in height is greatest in adduction, the obliquity of the false image is greatest in abduction. The lateral distance is greatest in the middle line, and lessens when the object is moved either inwards or outwards. The head is inclined forwards and towards the sound side. The diplopia is chiefly noticed when the patient has to look down, as in descending stairs, which appear double. The detection of this palsy is difficult, and the diplopia is the most important indication; if the other eye cannot be moved downwards the detection of the palsy may be impossible.

Inferior Oblique.—Defect of movement inwards and upwards, the effect being the converse of that of paralysis of the superior oblique. The secondary deviation is inwards and upwards by the inferior oblique and superior rectus, &c., of the sound eye. Diplopia exists in the upper inner part of the field; it is simple, the false image above the other, especially in adduction, and oblique (the images converging towards the horizontal plane), especially in abduction. The head is inclined backwards and towards the affected side.

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Congenital Anomalies are not rare, and sometimes entail diagnostic difficulty. One eyeball & ocular fovea may be a little higher than the other, & it is very common for the upward movement to be unequal in degree. Sometimes some ~~one~~ movement is oblique instead of straight in one eye; the left eye for instance ~~on looking to the right~~ ^{upwards} moves obliquely ~~to the right~~ & on looking ~~to the~~ horizontally to the right, moves upwards as well as inwards. In these cases there is often also a difference in level. The fact that in Congenital cases double vision can never be found is an important aid to their recognition.

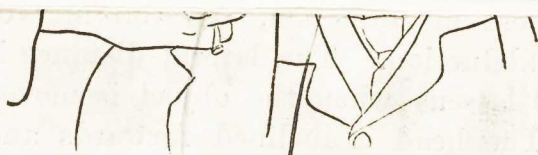


FIG. 94.—Partial paralysis of left third nerve. Defective movement upwards, from weakness of the superior rectus, and downwards, from weakness of inferior rectus.

external muscles of the eye are paralysed, with the exception of the superior oblique and external rectus; the levator palpebræ, sphincter iridis, and ciliary muscle are also affected. The upper eyelid droops and cannot be raised; the eye can be moved only outwards, and a little downwards and inwards by the superior oblique. The pupil is of medium size, and does not contract; power of accommodation is lost.

* The third nerve is commonly termed in Germany the "oculo-motorius," a name which has the grave practical inconvenience of preventing the use of the term "oculo-motor" in its exact and much-needed general significance.

figures given above show, it is a frequent ocular palsy, perhaps because the nerves have so long and exposed a course.

Third Nerve.*—When the whole of the third nerve is affected, all the

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~~Of isolated palsies, that of the external rectus is incomparably the most~~

frequent, and the order or frequency of 145 cases was found by A. Graefe to be—external rectus 105, superior oblique 52, inferior rectus 10, superior rectus 9, internal rectus 4, inferior oblique 2. The frequency of isolated palsies of the first two muscles is due to their separate innervation. In the estimate, the frequency of loss of some of the associated palsies is probably not included. They were not then adequately recognised.

AFFECTIONS OF SPECIAL NERVES.

Sixth Nerve, Abducens Oculi.—Only one muscle, the external rectus, being supplied by the sixth nerve, the disease of the nerve causes only paralysis of this muscle. As the figures given above show, it is a frequent ocular palsy, perhaps because the nerves have so long and exposed a course.

*Third Nerve.**—When the whole of the third nerve is affected, all the

external muscles of the eye are paralysed, with the exception of the superior oblique and external rectus; the levator palpebræ, sphincter iridis, and ciliary muscle are also affected. The upper eyelid droops and cannot be raised; the eye can be moved only outwards, and a little downwards and inwards by the superior oblique. The pupil is of medium size, and does not contract; power of accommodation is lost.

* The third nerve is commonly termed in Germany the "oculo-motorius," a name which has the grave practical inconvenience of preventing the use of the term "oculo-motor" in its exact and much-needed general significance.



FIG. 94.—Partial paralysis of left third nerve. Defective movement upwards, from weakness of the superior rectus, and downwards, from weakness of inferior rectus.

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In an attempt to raise the eyelid the frontal muscle contracts strongly (see next page). After a time the deviation outwards of the eye increases, in consequence of contracture in the external rectus, and the pupil becomes larger than it was at first by a similar contracture in the radiating fibres.

Paralysis of the third nerve is often partial; the various muscles supplied by it are affected in different degrees, and some may escape altogether. The muscles to the eyeball may be involved, and the levator may escape, but the paralysis of the former is then seldom complete. The levator and superior rectus may suffer apart or together; in bilateral nuclear disease they are generally paralysed together. The external muscles may suffer, and those within the globe may be unaffected; the converse relation is only seen in central cases.

Both third nerves may be affected, but it is rare for both to be paralysed completely, as in the case shown in Fig. 95. There is then complete double ptosis, and if the lids are raised the eyeballs are seen to be turned outwards, and to be almost immovable. In most cases of bilateral disease of these nerves the paralysis is partial. In some cases the distribution of the palsy is determined by functional association; thus each internal rectus may be affected and no other muscles: such cases are central and nuclear in nature, involving this nerve merely because its functions are so extensive; they will be separately considered.

Ptosis.—The drooping of the eyelid may be complete, so that it cannot be raised, or incomplete, being raised to a certain point but not higher. There is usually a conspicuous over-action in the corresponding half of the frontalis, so that the eyebrow is always higher than it should be. The frontalis normally acts with the levator when the eyes are directed much above the horizontal plane. This is an



FIG. 95.—Paralysis of both third nerves in a child two years old. Complete double ptosis, and divergence of both eyes by the external recti when the eyelids are raised. The child was of a tubercular family, and presented also double optic neuritis and weakness of the limbs of both sides. No other cranial nerves were affected. The probable cause of the symptoms was a tubercular tumour in the interpeduncular space. Under tonic treatment all the symptoms passed away, and a year later the child appeared quite well.

instance of secondary over-action in an associated muscle, analogous to the secondary deviation in an ocular muscle (p. 171). The frontalis responds to the increased innervation, which has no influence on the paralysed levator. It appears as if the patient were trying to raise the lid by the frontal muscle, but this appearance is, at least chiefly, fallacious, and the real explanation is that just given. But in some persons the frontalis can produce a slight elevation of the lid; hence to ascertain whether a slight voluntary elevation of the lid is due to the levator or frontalis, it is necessary to fix the eyebrow by pressing it against the bone.



FIG. 96.—Paralysis of the left third nerve; ptosis; over-action of frontalis.

Fourth Nerve.—The symptom of paralysis of the fourth nerve is paralysis of the superior oblique, which has been already described. It is seldom met with alone, except in cases of nuclear disease.*

Sympathetic.—Disease of the sympathetic causes paralysis of the radiating fibres of the iris, with consequent loss of the dilatation of the pupil. There are also slight prominence of the eye and slight ptosis, supposed to be due to atony of the involuntary fibres of Müller in the fascia of the orbit.

INTERNAL OCULAR PALSY.—Ciliary Muscle: Cycloplegia; Loss of Accommodation.—The evidence of this in normal and hypermetropic eyes is that near vision fails and distant vision remains good, while near vision can be restored by a convex glass. Usually, though not invariably, the contraction of the iris which normally occurs on accommodation is absent also. In myopia and presbyopia the absence of the power of accommodation makes little difference to sight, and the diagnosis of the condition is difficult. Accommodation depends on the third nerve, and is lost in complete, and often in partial, damage to the trunk of the nerve, and in disease of the anterior fibres by which it arises, or of the anterior part of its nucleus. The path from the lenticular nerve is through the ganglion and ciliary nerves. Loss of accommodation in one eye only, without the affection of other branches of the third nerve, is possibly due to disease of this ganglion or of the nervous ganglia within the eye, but we have no evidence at present of the effect of disease of these structures.† Bilateral cycloplegia, occurring alone, usually depends on disease of the centres in the nuclei of the third nerves. It occurs in diphtheritic paralysis, of which it is one of the earliest and most constant symptoms. It occurs also as an associated symptom in degenerative disease of the spinal cord, especially in tabes. Its relation to such disease is similar to that of the loss of the reflex action, immediately to be described.

* See Remak, 'Neur. Cent.', 1888, p. 5.

† See Hesch, 'Neur. Cent.', 1889, p. 592.

which may indeed have a trifling influence.

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degenerations such as are
associated with tabes

Iris: Iridoplegia.—The pupil may be unduly large (*mydriasis*), from palsy of the sphincter (third nerve) or spasm of the radiating fibres (sympathetic), or it may be unduly small (*myosis*) from the opposite conditions. The iris has three actions, two reflex and one associated: (1) Reflex contraction of the sphincter on exposure of the eye to light. (2) Reflex dilatation by the radiating fibres on stimulation of some cutaneous nerve. (3) Contraction on accommodation, usually, but not necessarily, associated with convergence.

(1) *Loss of the Associated Action: Accommodation Iridoplegia.*—The pupil does not lessen in size when an effort to accommodate is made. To examine this action it is only necessary to make the patient look first at a distant and then at a near object; the two should be in the same line of vision, so as to avoid any change in the amount of light that enters the eye. There is usually paralysis of accommodation, but the ciliary muscle may be competent, and yet the associated action of the iris may be lost, and *vice versa*. This loss is less common than is that of reflex action; it is due to the same causes as cycloplegia.

(2) *Loss of the Light Reflex: Reflex Iridoplegia.*—In examining the action of the iris to light, care must be taken to test each eye separately, keeping the other eye covered. ~~Light entering one eye acts on both pupils, and the contraction of each is the result of the total amount of light that enters both eyes. If one eye be covered and the action of the other pupil noted, it will be found to contract still more when the first eye is uncovered.~~ A good light is important; a slight action, ~~distinct on a bright day, is often imperceptible in a dull light.~~ If artificial light is used, it is best to make the patient look at an object in a distant dark part of the room, and then bring a light suddenly in front of the eye. If the patient looks at a near light, he will accommodate for it, and the pupil may then contract when there is no action to light. ~~Hence, if the light is looked at, it must be at least four feet from the patient, and should be bright.~~ To avoid the accommodation it is desirable, if a light is brought in front of the eye, that the other eye should be shielded from the light but not closed, so that, by continuing to fix a distant object, accommodation may be kept relaxed. ~~Reflection of light into the eye by an ophthalmoscopic mirror is not a good test, because the light falls only on a very small area, and may miss the macula, the most sensitive part.~~ Reflex iridoplegia is usually double; ~~when one-sided, as in a case recorded by Eales,* its cause is usually exceptional.~~ But I have observed the loss to be unilateral in tabes, although this is very rare.

The path through which this reflex action is produced is the optic nerve, both optic tracts, probably the corpora quadrigemina, the anterior part of the nucleus of the third nerve behind the centre for accommodation, probably the second fasciculus of origin of the third nerve, its trunk, the ciliary ganglion, and the ciliary nerves.

* 'Ophth. Rev.,' ii, p. 225.

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but occasionally

(3) *Loss of the Skin Reflex: Cutaneous Iridoplegia.*—The dilatation on cutaneous stimulation occurs in most persons under normal circumstances. The skin of the neck is a convenient place for the stimulation. ~~The faradic current applied with a wire brush, so as to produce a sharp painful sensation, has been usually employed, but in most persons~~ The prick of a needle or pin, of a quill point, and even a pinch, is sufficient.* The motor path for this action lies in the cervical sympathetic, and the fibres connecting this with the cord at ~~the lowest part of the cervical region.~~ The centre on which it depends is said to be beneath the corpora quadrigemina, to the outer side of that for the light reflex. If so, both motor and sensory paths must traverse the cervical region of the spinal cord.

Each reaction is lost when disease interrupts its path or damages the centre on which it depends. Thus the light reflex is impaired or lost in disease of either the optic nerve (including the retina) or in disease of the trunk of the third nerve. Disease of one optic tract does not lessen the action, unless the light falls only on the blind half of the retina (*see p. 163*), no doubt because the fibres from the most sensitive (central) region of each retina pass by both optic tracts, and so disease of one does not abolish the reflex. So, too, the skin reflex is lost in disease of the cervical sympathetic, and in some affections of the cervical spinal cord, especially in those that impair sensibility. In any case we infer the seat of the disease from the other symptoms that are associated with the loss.

When the loss of reflex action occurs without such accompanying symptoms, and is thus isolated, so far as immediate associations are concerned, it is generally due to disease of the centres, degenerative in character.† It occurs in association with disease of the spinal cord, chiefly with locomotor ataxy, ~~in which it is a common and early symptom. The loss is frequent also in general paralysis of the insane, and is occasionally met with in other degenerative diseases of less definite type.~~ It may occur also without other nerve symptoms. In most of the cases I have seen, in which it existed alone, the patients have had constitutional syphilis many years before, and the same ~~statement is true of at least one disease (locomotor ataxy) with which the symptom is often associated.~~

*and of this the symptom
is an important
suggestive indication*

* In some persons in whom the reflex action is readily produced, it occurs on slighter cutaneous impressions. It may occur, for instance, when the palm of the hand is tickled, as schoolboys know who pretend to dilate the pupil by an act of the will.

† This has not yet been demonstrated, but hardly admits of doubt because the loss of reflex action occurs under the same conditions as another affection to be presently described, progressive paralysis of the external muscles, the central and degenerative nature of which has been proved.

The frequency of loss of the light reflex without loss of the associated action of the iris was first pointed out by Argyll-Robertson. The fact that loss of reflex dilatation is usually associated with that of reflex contraction was discovered by Erb.

lwh The two palsies are often, but not always, conjoined. There may be loss of reflex action to light, when the pupil still dilates on stimulation of the skin. The pupils are often small, reduced to two, one and a half, or one millimetre in diameter. The association of these small pupils with spinal disease was noticed before the more significant loss of the light reflex was discovered, and the condition was called "spinal myosis." But the pupils are not always small; they may be three, four, or five millimetres in diameter, although there is no trace of reaction to light. I think that it will generally be found that when the pupils are not small the skin reflex persists, and when they are small it is lost. ~~The small pupils may be the result of the loss of tone in the radiating fibres accompanying the loss of the skin-reflex, and preserved when the pupils are large.~~ In cases of disease of the third nerve the ciliary muscle is paralysed as well as the iris. When the reflex loss is due to central degeneration the ciliary muscle is usually unaffected, and the pupils contract during accommodation. In some cases, however, there is also cycloplegia. ~~All the internal muscles of the eyeball are then paralysed, a condition which we shall again consider.~~

ly COMPOUND OCULAR PALSIES.—*Loss of Convergence and of Accommodation.*—The power of accommodation may be lost although the eyes can still be converged, but in many cases the two actions, habitually associated, are lost together. The internal recti may contract in a normal manner in lateral movements of the eyes, but ~~cannot be made to contract together.~~ Doubtless there is a separate centre for the converging action, as we shall presently see there is for the action of one with the opposite external rectus in lateral movements. ~~The former must be closely connected with the accommodation centre, since we only converge when we accommodate. A remarkable example of this loss in a child has been published by Eales.* There was absolute loss of convergence, of accommodation, and of the associated action of the iris, while all other movements of the eyes, and the reflex action of the pupil, were perfect. The loss developed gradually without recognisable cause, and, after lasting for about a year, passed away.~~

Paralysis of the upward movement of the eyes has often been observed in cases of central disease, associated with paralysis of the levators. It may result from disease of the posterior part of the third nerve nuclei (Kahler and Pick). ~~When due to such a focal lesion it is usually unilateral.~~ It is possible that there is also a higher centre, disease of which may paralyse the upward movement without the lid, since this isolated symptom may be met with. I have recorded one such case in which the symptom was well marked.†

* 'Trans. Ophth. Soc.,' iv, 1884, p. 300, and oral communication.

† 'Trans. Ophth. Soc.,' i, 81, p. 117. The patient has died since the account of her case was published. A very small tumour was found in the middle line

*on one side,
or, in degeneration
lesions on both.
Paralysis of the
upward movement
without the levator
has been met with,
even bilateral*

Paralysis of both upward and downward movements of the eyes, without impairment of the lateral movements, has also been observed, and is probably also due to a lesion in the same position. The paralysis has been bilateral and accompanied with ptosis. But it has also been caused by disease of the cerebral hemisphere, situated in the corpus striatum and optic thalamus.*

Paralysis of Lateral Movement: Conjugate Deviation of the Eyes.—The conjugate deviation of the eyes, towards the side of a lesion in the cerebral hemisphere, the result of a loss of the power of moving them to the other side, has been already described (p. 78). The deviation is occasionally due to spasm, and is then from the side of the cerebral lesion. We infer to which of these two mechanisms it is due, by the associated symptoms in the limbs, ~~which are paralysed and flaccid when the eyes deviate from them, rigid with spasm when the deviation is towards them, and from the side of the lesion.~~ These are merely an extension to the eyes of the effects of the disease manifest in the limbs. Their occurrence and degree seem to be related to the seat of the cerebral lesion. But in disease of the pons the condition is reversed. A paralyzing lesion on one side causes a loss of the movement of both eyes towards that side.† The seat of the disease that has this effect is the tegmental region of the pons, at or above the nucleus of the sixth,‡ and the facts show that the path for this combined movement descends in the same side of the pons as that towards which the eyes are moved, and that the movement is effected through the nucleus of the sixth. The sixth nucleus must therefore act on the the opposite third nucleus and internal rectus, and the path by which the influence is exerted is almost certainly the posterior horizontal fibres. It has even been thought that through these fibres, those of the third nerve for the internal rectus actually arise from behind the posterior quadrigeminal bodies, damaging these slightly, the velum, and the adjacent part of the inferior vermiciform process of the cerebellum.

It must be remembered that disease of the nerves or their roots may chance to affect only the fibres for the superior recti. This was apparently the case in a patient with an interpeduncular syphiloma (Thomsen, 'Berlin. Gesellsch. f. Psych.,' June 7th, 1886). One superior rectus was more affected than the other, a character that is probably of diagnostic importance.

* Wernicke, 'Berlin. kl. Wochenschr.,' 1876, p. 394, and 1878, p. 154. A similar loss, associated with hemianopia, was present in a case recorded by Lang and Fitzgerald, 'Trans. Ophth. Soc.,' vol. ii, p. 230.

† Much attention has been lately given to this palsy. The most important writings on the subject are those of Broadbent, 'Med. Times and Gaz.,' 1872, vol. i; Duval and Laborde, 'Journ. de l'Anat. et Phys.,' 1879; Bernhardt, 'Gehirn-geschwülste,' Wernicke, 'Gehirnkrankheiten,' Bd. i; Mierzejewski and Rosenbach, 'Neurolog. Centralbl.,' 1885, p. 363; and Bleuler, 'Deut. Arch. f. kl. Med.,' 1886, Bde. xxxvii and xxxviii, in whose paper abstracts of most writings on the subject will be found, and an important case by Hughes Bennet ('Brain,' 1889), in which there was permanent conjugate deviation of head and eyes, the result of a patch of softening limited to the sixth nucleus. Power of convergence was retained.

‡ Very seldom below, and then reaching up to the nucleus. See note on next page.

is connected with through

~~the sixth nucleus; this assumption is opposed by many facts, and it seems that the horizontal fibres merely connect the two nuclei.*~~ Other signs of disease of the pons are often present, paralysis either of the facial or of the fifth nerve. Nystagmus is often associated (see p. 207).†

Since the movement is excited from the opposite cerebral hemisphere, the path must cross the middle line above the middle of the pons, and it probably does so at the corpora quadrigemina. ~~If so the arrangement must be that shown in Fig. 97.~~ We do not know, however, whether the pons contains merely the path to the sixth nucleus, or whether there is a special centre for the movement distinct from this nucleus. The latter is probable on account of the relation of the movement to sensory impressions, especially to those of the auditory

on movement to the other side

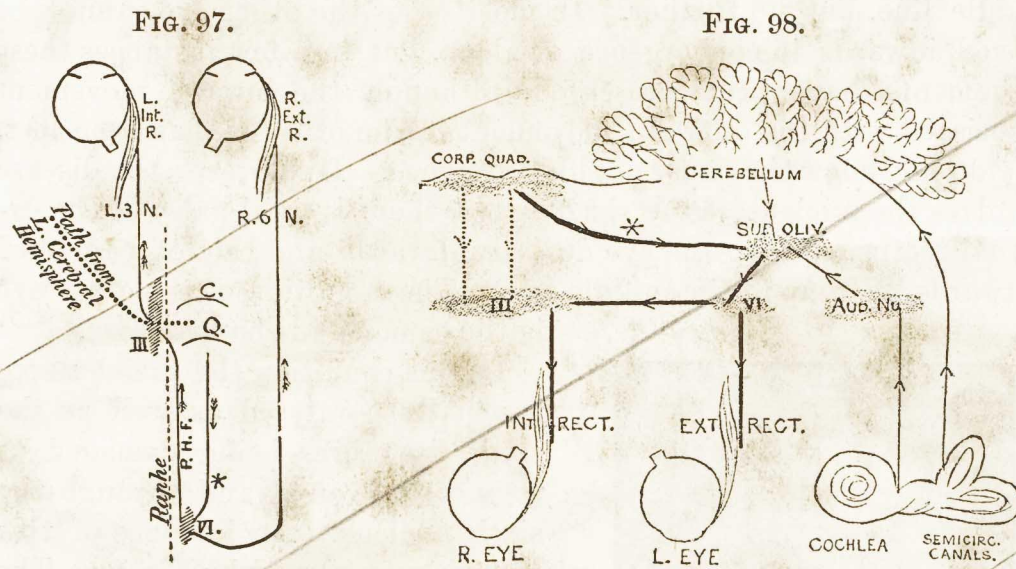


FIG. 97.—Diagram of the mechanism for the associated lateral movement.

FIG. 98.—Diagram of probable relation of the superior olivary body to the associated lateral movement. The asterisk in each figure indicates the downward path.

nerve, combined with the fact that a movement of the head is often associated with that of the eyes, so that a connection of the auditory nerve with the sixth nucleus would scarcely suffice. The connections of the superior olivary body, according to Bechterew, are precisely

* The evidence is that the internal rectus is not always totally paralysed; it has been known to act in convergence although not in the lateral movement, and also inwards, in a normal manner, if the other eye was covered, although there was no movement when the eye with the paralysed external rectus was uncovered. Moreover, when the sixth nucleus has been totally destroyed, no degenerated fibres could be found in the opposite third nerve.

† Experiments suggest that deviation towards the side of a lesion of the pons means irritation and imperfect destruction. The opposite deviation follows subsidence of the irritation, each being attended by nystagmus (Laborde, 'Gaz. méd. de Paris,' 1878).

those that such a centre would have. By some fibres of the fillet, which degenerate downwards, this body is connected with the corpora quadrigemina, and fibres pass between it and the sixth nucleus, the auditory nuclei, the cerebellum, and the lateral column of the spinal cord; the last may subserve the movement of the head.* If this hypothesis is correct, the mechanism must be such as is shown, in side view, in the diagram Fig. 98. The arrow-heads indicate the direction of conduction; and the thicker lines indicate the path by which the movement is excited by the will.

According as the disease does or does not involve the nucleus of the sixth nerve, the symptoms present some variation. If the disease is above the nucleus, there is a loss of the power of moving both eyes beyond the middle line towards the side of the lesion. If they are moved towards the other side, they can be moved back as far as the middle line, but no further. ~~In most cases the other eye cannot be moved inwards in convergence or alone, but in a few instances these movements have been preserved, although the inward movement associated with the outward movement of the other eye has been lost. We do not know the cause of this difference.~~ If, however, the disease involves the nucleus of the sixth nerve, there is total palsy of the external rectus, so that the eye deviates inwards and cannot be moved outwards, even up to the middle line. The condition of the other eye

is the same as in the first case.



FIG. 99.—Complete paralysis of the left sixth and facial nerves (with inability to move the right eye towards the left) from an acute lesion on the left side of the pons. (From a photograph.)

In this condition the facial nerve is usually paralysed as well as the sixth, its fibres being damaged as they course round and through the sixth nucleus. An instance of this palsy is represented by Fig. 99. The onset was sudden, and slight hemiplegia was present at first. The left sixth and facial nerves were completely paralysed; the left eye deviated inwards, and no outward movement was possible; the right eye could not be moved inwards; the facial muscles presented the reaction of degeneration. If a lesion in the pons damages the fibres of the sixth nerve away from their nucleus, the paralysis of the external rectus is complete but exists alone, ~~there is no deficiency of movement~~

* In one recorded case the lesion was below the level of the sixth nucleus, the lower end of this nucleus being near the highest part of the lesion. It is probable that the superior olivary body was involved (Senator, 'Berl. Psych. Gesellsch.,' July 9th, 1883).

of the opposite internal rectus. The lesion that causes these symptoms may be either a chronic process, such as a small tumour, or an acute lesion, hæmorrhage, or, more frequently, softening. Other symptoms of disease on one side of the pons are often present, especially in the region of the fifth nerve on the side of the lesion, sometimes in the limbs on the opposite side.

Total Ophthalmoplegia.—Paralysis of all the muscles of both eyes, internal and external, is a very rare consequence of disease at the neighbourhood of the orbital fissure and optic foramen on each side, ~~either in the orbit or within the skull, such as thrombosis in each cavernous sinus.*~~ But it is met with chiefly in cases of nuclear disease, and will be described in the account of this variety.

CAUSES AND CAUSAL VARIETIES.—Paralysis of the ocular muscles may be due to disease of the nerves in the orbit or at the base of the brain, to lesions of the fibres of origin between the surface of the brain and the nerve-nuclei, and to disease of the nuclei themselves. ~~A defect in certain ocular movements may also be produced by disease of the cortex of the brain and of the path from the cortex to the nuclei, but the loss that is usually so produced is that of the conjugate lateral movement of both eyes, already described in the account of hemiplegia, with slight ptosis on the side opposite to the lesion. It is possible, however, that a defect of associated movements (e.g. of the upward or downward movement) is also sometimes due to disease of the hemisphere, but our knowledge of this cause is imperfect. Of the remaining lesions those of the nuclei (constituting the important group of nuclear palsies) will be separately described.~~ *may cause*

~~Injury is a common cause, and may damage the nerves in any part of their course, especially at the orbital fissure or in the orbit, but sometimes at their origin. Some of the roots of one third nerve have been found torn.† One eye is usually alone affected. The injury is usually a blow on the side of the head, which causes an extravasation and immediate paralysis, or excites an inflammation and palsy after a few days. All or only some of the nerves may suffer. Recovery is slow and often imperfect. Sometimes, indeed, there is a late extension, perhaps by processes outside the nerves, or cicatricial contraction of adjacent tissue. Thus a blow on the right temple was followed at once by palsy of all parts of this third nerve. Slow improvement was followed by a relapse, with palsy of the external rectus.~~

Neuritis in the orbit or at the base of the brain, involving the trunks or branches, is not rare. It may be syphilitic in either situation. In the orbit it is sometimes primary and "rheumatic," but ~~it~~ is occasionally secondary to adjacent cellulitis. Within the skull it is generally ^{due to local} secondary to meningitis. The nerves in the orbit are damaged by slow pressure only in rare cases of orbital growths, but within the

* Coupland, 'Trans. Oph. Soc.' vol. v.

† Hutchinson, Astley Cooper Prize, May 1st, 1865.

skull they often suffer compression ~~by growths or aneurisms~~. All the nerves to one eye may be compressed in the wall of the cavernous sinus by thrombosis in it, by an aneurism of the internal carotid, or by an adjacent growth,* and both third nerves may be damaged by a growth between the cerebral peduncles, or by an aneurism of the posterior cerebral artery. Tumours of the nerves themselves are rare; neuromata are almost unknown, but nodular syphilitic growths sometimes occur, and may be symmetrical—~~situated, for instance, on the two sixth nerves near their origin (Barlow)~~. The root-fibres suffer, in their deep course, from various lesions of the brain,—hæmorrhage, softening, tumours, and islets of disseminated sclerosis. Such lesions, ~~involving the sixth or third nerve-fibres~~, often also implicate the motor tract, ~~and there results hemiplegia associated with palsy of one of these nerves~~. ~~In consequence of the decussation of the motor tract below the origin of the nerve~~, The two symptoms are always on the opposite sides, "alternate hemiplegia;" if on the same side there must be two lesions. Very rarely a small focus of softening in the crus may damage only some of the roots of the third nerve.† Paralysis ~~of all the ocular muscles~~ ^{also} has been caused by the fumes of charcoal, by lead, and that of the third nerve has been met with in Graves' disease.‡ ~~Other toxic states, as that from alcohol, may have a similar effect. In some, the mechanism is a peripheral neuritis, but an action on the centre is more frequent.~~

toxic agents

Diphtheritic Paralysis.—A rare cause of palsy of the external muscles is diphtheria. The external rectus has been occasionally affected, and both internal recti have been weakened in association with the loss of accommodation so common in this disease. In one case all the muscles supplied by one third nerve were affected (Vadelot), and in two others there was paralysis of all the external muscles of both eyes.§

paralysed

Tabetic Paralysis.—In locomotor ataxy it is very common for some of the ocular muscles to be ~~weakened~~. The loss of power is often transient, and then may recur; sometimes it is permanent. ~~Each form may occur at any stage of the disease, and is sometimes an early symptom, even preceding those in the limbs.~~ The external rectus, or the levator and superior rectus, are the muscles most frequently affected; occasionally all the muscles supplied by the third nerve suffer. Loss of the light-reflex is extremely common, and ~~other internal palsies are not infrequent~~. External palsy is generally on one side only, but sometimes both eyes are affected (see Figure, vol. i, p. 408). We have scarcely any evidence at present of the

often for a time only,

H

cycloplegia is occasionally added

* Many instances of such palsy are on record. A typical one is recorded by Nettleship, 'Trans. Ophth. Soc.,' vol. i, p. 186.

† Kahler and Pick, 'Arch. f. Psych.,' x, 334. It has been termed "fascicular palsy." "Root palsy," internal or external, would be a better name.

‡ Knapp, Buzzard, Finlayson.

§ Uhthoff, 'Neur. Centralbl.,' 1885, p. 125; and Mendel, ib., p. 128.

Myasthenia. — In this mysterious
malady (^{gr}weakness of the ocular muscles is
not rare. The superior recti suffer most, in
association with the levators ^{obturator} ~~and~~ the inferior
recti least. The affection of the lateral mus-
cles varies much, & differs even in those that
act together.

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nerves. Thus the mere fact that paralysis is ^{preceded by} ~~due to~~ syphilis is only the first step in the diagnostic ~~problem of the nature of the disease.~~ ^{& diagnosis}

Relapsing Palsy.—One form of paralysis of the third nerves, which occurs chiefly in the subjects of syphilis, presents a peculiar tendency to relapse ^{and persist}. One third nerve becomes paralysed, improves under treatment, and then, perhaps while the treatment is being continued, the other third nerve suffers, and afterwards the affection of the first returns. An instance of the palsy is shown in Fig. 100. Syphilis was contracted four years previously, but the sudden onset of the palsy of the left nerve occurred during a severe cold.



FIG. 100.—Double ptosis from paralysis of the third nerves, complete on the left side, partial on the right. Over-action of frontales, greater on the side of the complete palsy.

The patient was treated ^{duly} with iodide and mercury, but the course of the paralysis was that described above, and the permanent condition was that the left eye could only be moved outwards and a little upwards. There was extreme ptosis of the left eye and slight ptosis of the right. The internal muscles were also paralysed. In another case, similar in course but less complete, the interval after the syphilis was thirty years, so that its influence is doubtful. The nature of the lesion in these cases is uncertain. The course and limitation of the symptoms seems inconsistent with nuclear disease.

Recurring or Periodical Palsy.—

The last variety to be described is a rare form of transient palsy, usually on one side only, which comes on at intervals during many years.* It has been miscalled "relapsing palsy," but typical attacks are recurrences, not relapses, and, in most instances, have been periodical. Both sexes suffer, but women ~~are~~ more subject to the disease than men. It often dates from early childhood, and ~~has been known to begin at eleven months,† at fifteen months,‡ at five years of age, and later;§ it continues up to at least middle life.~~ An attack may occur at the same time each year, or at intervals of about six months. A much longer interval usually separates the early attacks. Sometimes the intervals have been long and irregular, amounting to several years,||

* Cases have been recorded in this country by Saundby, 'Lancet,' 1882, ii, 345, and 1885, i, 57; and by Snell, 'Trans. Ophth. Soc.,' v, 193.

† Möbius, 'Neur. Cent.,' 1884, p. 307.

‡ Snell, loc. cit.

§ Thomsen, 'Charité-Annalen,' 1885, p. 562.

|| Five to nine years in a case recorded by Camuset, 'L'Union méd.,' 1876, p. 906.

while in other cases they have been short, and attacks have even occurred at each menstrual period.* As a rule the paroxysm begins with severe pain in the eye, and often with headache and vomiting. These symptoms last two or three days; the palsy may accompany them or may come on as they lessen. In many cases only the third nerve has been affected, but in some the external rectus was also involved. The palsy may be complete or incomplete; the internal muscles are often, but not always, affected; there is usually ptosis. The loss of power lasts for a few days or weeks, and then gradually passes away. When the attacks are frequent they are usually brief. In many cases a slight defect of power has persisted during the intervals. ~~In one case there was concentric contraction of both fields of vision, greatest in the paralysed eye, and varying in degree in proportion to the motor palsy (Thomsen).~~ The attacks may last for several weeks, from three to eight; and, as a general rule, when the intervals are long, so are the attacks. When the paroxysms recur frequently—as, for instance, every month—they last for a few days only. A child of three and a half, with some indications of congenital syphilis, had ptosis of the right eye. Four months later it had ptosis of the left eye, and a second attack in the same eye ten months afterwards. Each time the attack lasted a few days, and passed off without leaving any weakness. In several cases the headache was unilateral. In patients subject to the affection, an attack has been brought on by mental shock.

The nature of this disease is mysterious. It has been compared to migraine, and is perhaps ~~more~~ more closely allied to this ~~than to any other disease.~~ Charcot has termed it "migraine ophthalmique," and in one case there was a strong family history of migraine (Snell). But the long duration of the attacks, the motor character of the chief symptoms, their long duration, and the occasional persistence of slight defect of movement, are marked differences from purely migrainous and neuralgic affections.† It has been suggested that the cause is organic disease with periods of activity, but this theory is scarcely tenable. Vaso-motor disturbance has, of course, been invoked to explain it, but this is only putting the difficulty farther back. The few post-mortem examinations so far obtained do not clear up the mystery. With permanent

* Hasner, 'Prag. med. Wochenschr.', 1883, No. 10.

† A case recorded by Buzzard ('Clin. Lect.', p. 164) is perhaps a connecting link between these cases and pure neuralgias. A woman had been subject for many years to fortnightly paroxysms of neuralgia of the first division of the fifth nerve, and for two years before she came under observation each attack was followed by partial palsy of the third nerve lasting for a few days. Complete palsy has occurred during each attack of migraine (Anderson, 'Trans. Oph. Soc.', 1892).

In a case recorded by Pflüger there were periodical attacks of paralysis of variable seat; of the left third and facial nerves at eighteen, of the nerves on the right side at twenty, of the left third nerve again at twenty-two, and six months later of the left sixth and facial. Each attack lasted from one to two months, and was preceded by pain, which before the last was about the left mastoid process.

has been considered

indeed it has been termed

Yet a connection is indicated by the fact that ptosis sometimes occurs during ordinary migraine, on the side of the headache apparently inhibitory in origin.

weakness there have been changes in the peripheral nerves and not in the nuclei. ~~A woman died of phthisis at thirty who had suffered, since childhood, from periodical attacks of palsy of the left third nerve. This appeared grey, and its roots were surrounded by small grey granulations which contained tubercular bacilli, which did not extend into the crus. Some fatty degeneration was found in the muscles supplied by it, the other muscles being normal.* In other three examinations the lesion was peripheral and the nuclei intact.†~~

NUCLEAR OCULAR PALSY.—Disease of their nuclei is a frequent cause of paralysis of the eye-muscles, internal and external, and certain forms may be due to disease of the corpora quadrigemina which does not penetrate the nuclei. The symptoms vary much according to the position and character of the lesion, which may be various, and either acute or chronic; ~~but they are, as a rule, bilateral, except when there is the isolated affection of the sixth nucleus, already considered in the account of such conjugate palsy.‡~~

The chief forms of nuclear palsy may be classified according to the character of the symptoms and probable nature of the morbid process. The most common is (1) chronic nuclear palsy, due usually to degenerative processes, very rarely to such chronic disease as a tumour. A rare form is (2) acute nuclear palsy, in which the symptoms come on in a few days or a few weeks. The onset suggests inflammation, and the form has been termed (by Wernicke) "polio-encephalitis superior," ~~§ from its analogy with polio-myelitis~~, and to distinguish it from a similar affection of the lower nuclei in the medulla, "polio-encephalitis inferior." ~~But~~ It may apparently also be excited by toxic influences. (3) In *sudden nuclear palsy* the onset occupies a few minutes, or at most an hour or two. These cases are often miscalled "acute," but this term is needed for class 2. Such an onset always indicates a vascular lesion, ^{especially obstruction} which probably is the common cause of this form.

The characteristic feature of all forms is the paralysis of muscles of both eyes, internal or external, symmetrical or irregular. Limitation to one eye occurs so seldom that it should not suggest nuclear disease. On the other hand, both eyes may suffer from disease of the

* It should be remembered that disturbance of function may cause organic changes or determine disease of the nerve, as well as result from such disease.

† 'Neur. Cent.', 1888, p. 88.

‡ Nuclear ocular palsy is also termed ophthalmoplegia, a term first used by Brunner in 1850. The nature of the cases was pointed out by M. Graefe in 1856 ('Arch. f. Oph.'), and, in 1868, compared with labio-glossal palsy. Förster correctly localised the lesion in 1878 for external palsies; and internal ophthalmoplegia was well described by Hutchinson in 1878 ('Med.-Chir. Trans.'), and the external ophthalmoplegia, with post-mortem proof of its nature, in 1879 (ibid.).

§ A name open to many objections, both theoretical and practical. Nuclear palsy of all the external muscles has, indeed, been associated with polio-myelitis in a man of forty, but the cord affection was acute, and that of the eyes was chronic (Sachs, 'American Journ. Med. Science,' 1889).

nerves (trunks, branches, or endings), but such bilateral disease is rare, and, without other evidence of a nerve lesion, should suggest nuclear mischief.

(1) *Chronic nuclear palsy, chronic ophthalmoplegia*, includes the isolated loss of the reflex action of the iris, isolated palsy of the ciliary muscles, palsy of all the internal muscles, of all or many of the external muscles, and lastly, of both the external and internal muscles. It also includes loss of the upward or downward movements of the eyes, ptosis, and the conjugate lateral palsy already described. The combinations met with are extremely numerous and varied.

Later observations have shown that we cannot separate most forms of chronic palsy. They occur under similar conditions; they may be variously combined, and the dependence of one of them on nuclear disease has been conclusively proved, so that it is probably true of most. Each form is met with in association with locomotor ataxy; the simple loss of reflex action is indeed present in the majority of cases of this disease. It has been mentioned that when this loss exists alone the subjects of it have frequently suffered from constitutional syphilis, and the same fact is true of the other forms of ophthalmoplegia.* They sometimes occur, however, without this antecedent, which may or may not develop later. They occasionally affect young persons, and are met with more frequently in males than in females, resembling in this their congener, tabes. It is seldom that any immediate cause can be traced; in one instance the symptoms are said to have come on after a wetting. I have seen the condition as a late and permanent sequel of diphtheria.

Some other features deserve mention. The first muscles to suffer may be those that are associated in action—the superior recti and levators, the two internal recti, or the internal rectus on one side and the external rectus on the other; or it may be quite irregular. The loss of power, at first slight, slowly increases; often a strong effort shows more power than is habitually exerted, and the weakness may be less in the morning than in the evening. The disease slowly extends, until at last, often after several years, only one or two of the muscles of both eyes retain power, and sometimes every muscle is paralysed. The levators, however, seldom become completely paralysed, and may be unaffected. The aspect of the patients is peculiar; when there is partial ptosis, this gives them a sleepy expression, and in total palsy without ptosis a staring look results from the immobility of the eyes, which are fixed in mid-position. Sometimes, although rarely, they are slightly prominent. Occasionally one eye is much

* As in so many of these degenerative diseases, the facts that can be ascertained probably under-represent the relation to syphilis. The case of Mr. Hutchinson, examined by me (see p. 197), is an illustration of this. The man persistently denied any venereal sore, and no trace or history of syphilitic disease could be found. But some years later a child of this man was brought to Mr. Hutchinson with characteristic notched teeth and interstitial keratitis.

tabes and other post-syphilitic degenerative maladies

the combination

Bork

more affected than the other; ~~on one side there may be total palsy, on the other only ptosis, or loss of the light-reflex.~~ The internal muscles are often unaffected; ~~when this is the case the diagnosis of nuclear palsy can be made with confidence, because the escape of the internal muscles in bilateral disease of the nerve-trunks is scarcely possible.~~ ~~This feature was thought to be a characteristic of progressive ophthalmoplegia by von Graefe, but it is not so; the affection of the external muscles may be combined with loss of reflex action of the iris, with eyelid ptosis, or with both as "total ophthalmoplegia."~~ In the case presently to be mentioned, in which the nature of the lesion was ascertained, the internal muscles were affected. Double vision may trouble the patient in the early stage of the affection, but it generally passes away as the disease progresses, and is sometimes absent from the first, perhaps when the early loss is of associated movements. The duration of the malady is long, ^{and} ~~in one of Hutchinson's cases the symptoms continued for seventeen years. In one of Mauthner's the affection is said to have been limited to one eye for twenty years.~~ It is common ^{may} ~~for~~ arrest to occur when a certain degree of palsy has been attained. The symptoms may exist alone, but they are frequently associated with ^{degenerative} ~~indications~~ of other diseases of the nervous system—with optic nerve atrophy, with affection of the bulbar nerves* (rarely amounting to typical bulbar palsy), with general paralysis of the insane, and especially with locomotor ataxia. It may attain a considerable degree before there are any symptoms of tabes, even when this is ultimately severe. A similar condition is perhaps sometimes congenital, and even hereditary.

The nature of these cases is not a matter of speculation. In a case of Mr. Hutchinson's, in which I examined the brain (1879), the state of the nuclei of the ocular nerves was nearly the same as that of the grey matter of the spinal cord in progressive muscular atrophy. ^{anterior} ~~The patient was a man, aged fifty-five, who had had syphilis. The symptoms began gradually, seven years before death, with palsy of the internal recti and ciliary muscles. Ultimately all the ocular muscles became very feeble, the optic nerves atrophied, there was mental excitement, and some palsy developed in the limbs, the condition resembling that of some forms of general paralysis of the insane. The roots of the ocular nerves, outside and inside the brain, were grey, small, and contained scarcely any normal fibres. In the nuclei a few nerve-cells of normal size were seen, but these had, for the most part, lost their processes, and a large number of the cells were reduced to small angular bodies or had disappeared (Fig. 101).~~

* This combination existed in the case that was a late sequel of diphtheria.

† Thus Hirschberg ('Berlin. Gesellsch. f. Psych.,' June 8th, 1885) has described the case of a man with congenital double ptosis and paralysis of all the ocular muscles, incomplete in the superior oblique and the internal muscles, whose mother presented a similar condition, while his son had congenital ptosis and paralysis of the superior recti.

some cases

clear evidence

tabes

or may be

presents the same change &

In a typical case, ~~presented~~
(Med. Clin. Trans.
1879)

nuclei

The causation of the degeneration is mysterious but must be regarded as a variety of the myopathy of tabes, under which it is discussed

~~In the intervening tissue there were many connective-tissue nuclei. The change involved the sixth, fourth, and the whole of the third nuclei. The other nerve-roots and nuclei were normal. The spinal cord was not obtained. Thus the lesion, in a fairly characteristic case, was a degeneration, limited to structures having a common function. The internal palsies occur under similar conditions, and their frequent limitation to a single function in the two eyes points to a like change of more restricted seat. It is only by toxæmic influences or degeneration that structures of common function are selected, as it were, for isolated disease, and it is probable that all degenerations are often the effect of a toxæmia (see "Tabes," vol. i), perhaps a chemical "toxine," produced by organisms. In other and younger subjects they seem to be due to deficient vital endurance of the structure, varying in degree so as to be manifested at birth or not until adult life.~~

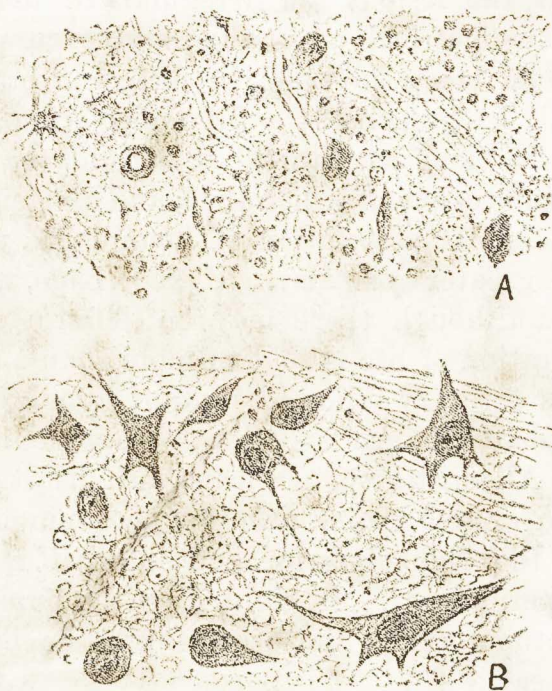


FIG. 101.—A, part of sixth nucleus in progressive ophthalmoplegia; B, cells of the normal nucleus, for comparison.

But in all degenerations destruction begins as slight and slow changes in nutrition, doubtless sometimes capable of recovery, as when internal palsy passes away after it has lasted for many months, and perhaps for more than a year. ~~I have known the light-reflex to return, in stationary tabes, after it had been lost for at least two years, and the remarkable case of internal ophthalmoplegia recorded by Eales (see p. 185) is an instance of the way in which such a palsy, limited and complete, may pass away. Such recovery is especially intelligible if the changes are due to a toxæmia.~~

Infantile oculo-facial palsy is a rare variety of the chronic form, which may be congenital, or begin at some period of childhood, rarely in early adult life. Palsy of the external ocular muscles, all or many, is very commonly accompanied by paralysis of the face, sometimes chiefly of the zygomatics. Ptosis is common, but the internal muscles do not suffer. It usually becomes stationary, and remains so through life. Nothing definite is known of its causation, but its history suggests defective vital endurance in the nuclear structures.

Sudden Nuclear Palsy.—The vascular lesion which causes sudden palsy, and the onset of which is survived, is usually obstruction of the

* Möbius (1892) has collected forty-four cases ('Munch. med. Wochenschr.').

"atrophy"

arterial branches which the basilar gives to this part. The obstruction is usually bilateral, because disease in the wall of the basilar, whether atheroma or syphilitic, affects the branches given off at the spot on each side. Hence small foci of softening are often found in each ~~third nucleus.~~ ~~The same conditions determine the bilateral character of the lesion and symptoms in the rare cases of embolic obstruction.~~ But the lesions are irregular in distribution, and the symptoms unsymmetrical as a rule, and even partial, ~~different parts of the nuclei being supplied by different branches, according to Heubner.~~ This irregularity is a difference from the chronic form, while other distinctions are furnished not only by the onset, but by the tendency to improvement which ~~they, and all similar lesions, present,~~ and by the occasional association of hemiplegia, usually on the side opposite to the greater eye-palsy. Hæmorrhage, when it occurs, is apt to spread, and although there may be definite ocular palsy at the onset, the ~~duration of life is seldom more than a few hours.~~ Only in rare cases has recovery followed irritant apoplexy, that has made hæmorrhage probable. ~~An instructive example of the embolic form is recorded by Allen Starry.† A man with heart disease had sudden giddiness and double vision, followed by somnolence, unequal pupils, paralysis of the right inferior and internal recti, and the left inferior and superior recti. In eight days the pupils were equal, and the internal rectus normal; the other muscles remained paralysed at the end of a year.~~

An example of a much slighter lesion, probably due to syphilitic vascular disease, and characteristic in its course and irregularity, was that of a man who, one day, suddenly found both upper lids drooping, and then that both eyes deviated outwards, and could not be moved. In a few days he could move the right eye fairly well, but six months later he presented complete internal ophthalmoplegia in both eyes, with, in the left, slight ptosis and complete palsy of the other muscles supplied by the third nerve, but not of the external rectus, and the superior oblique possessed a little power.

Acute and subacute nuclear palsy includes the cases which reach their height in a few days or a few weeks, an onset that suggests one of two morbid processes—*inflammation*, or the action on the centre of some poison circulating in the blood, to which, indeed, actual inflammation may sometimes be due. ~~But such a poison may abolish function without causing visible changes; hence the name proposed by Wernicke† and widely used, "polio-encephalitis superior," is not always correct. Even the less objectionable "acute nuclear palsy" may prove to be sometimes inaccurate, since the symptoms may be~~

* See Sturge, 'Trans. Oph. Soc.,' vol. i.

† 'Journ. Nerv. and Mental Dis.,' 1888.

‡ Who first accurately distinguished this form ('Gehirnkrankheiten,' ii, 233). It was also well described by Thomsen ('Arch. f. Psych.,' 1888). Good descriptions have been given by Mauthner ('Vorträge').

So too

Symptoms

and is rapidly fatal

† sudden onset, usually with giddiness, in a patient with heart disease justifies a suspicion of embolism.

due to peripheral neuritis and not to nuclear disease. Hitherto, however, most post-mortem examinations have shown evidence of inflammation in the nuclei beneath the corpora quadrigemina, softening, minute extravasations, &c. Thus in two cases of external palsy, complete except that the levator escaped, Thomsen found great hyperæmia of the nuclei, third, fourth, and sixth (and of the hypoglossal), with degenerative changes in the nerve-elements. But the malady is rare, pathological observations are very few, and our knowledge of the cause of this form is therefore meagre. Of three cases described by Wernicke, one followed poisoning by sulphuric acid, and the other two in the subjects of chronic alcoholism. Injury

occurred

has been supposed to be an occasional cause. Eye-symptoms usually exist alone, the combination with bulbar palsy being seldom met with except in the chronic form. In a few cases, however, acute imperfect bulbar palsy has spread to the upper part of the face, and then to the eye muscles, a form that has been termed "ascending inferior polio-encephalitis."

In the usual form of acute nuclear palsy the eye muscles suffer in the same irregular manner as in the sudden variety. It is common for the internal muscles to escape: the levator may or may not be involved. In fatal cases the motor influence spreads, becomes extensive, both in the centres of other nerves and in the cortex, and the cerebral functions suffer so widely as to cause death in one or two weeks. In the cases that survive there is some improvement in the ocular palsy, which may be considerable, so that there is only slight residual impairment, or considerable palsy is limited to one eye. This is explained by the random character of inflammatory damage.

ultimately

In the rare cases in which a lesion is limited to the grey matter of the corpora quadrigemina, the chief eye symptom appears to be a loss of certain conjugate movements, especially of that upwards. But acute lesions usually produce other effects which mask those in the ocular muscles.

upward

Peripheral Ophthalmoplegia.—Palsy of the muscles of both eyes, resembling the nuclear form, may be due to peripheral neuritis. How relatively frequent this form is we cannot say, since the chief cause of peripheral neuritis, alcoholism, may also cause inflammation of the nuclei. Neuritis may be suspected when many other nerves suffer at the same time, or alcoholic multiple neuritis co-exists in the limbs,* although the latter cannot be regarded as proof, since alcohol may affect the nuclei. A case of neuritic ophthalmoplegia, which may be regarded as typical, is that of a man of sixty-two in whom all the eye muscles became weak as well as the levator palpebræ. The fifth nerve suffered, swallowing became difficult, and death followed. The ocular nuclei were normal, but there was wide-spread peripheral neuritis of both cranial and spinal nerves.†

It

* Suckling, 'Brit. Med. Journ.,' 1888.

† Meyer, 'Near. Cent.,' 1888.

with

It has

while the hypoglossal nucleus has been involved in the superior form. Very rarely a universal inflammation of the motor grey matter in the spinal cord + cerebral nuclei has caused universal fatal paralysis, chronic in course but proved after death

in rare cases

OTHER FORMS OF PTOSIS.—*Reflex Ptoſis*.—In very rare cases irritation of the fifth nerve has been observed to cause transient drooping of the eyelid, which must be referred to inhibition of the centre for the muscle. A physiological connection doubtless underlies this effect; spasmodic closure of the lids frequently results from irritation of the fifth nerve; for closure of the lids there is always relaxation of the levator (as is shown by the slight amount of orbicular contraction which will suffice), and hence irritation of the fifth nerve has a tendency to cause relaxation of the levator. Reflex ptoſis has been observed to follow division of the fifth nerve, no doubt from the irritation of the fibres by the section (Longet). It may result from the extraction of a tooth, as the following case shows. An upper right molar, decayed and causing much pain, was extracted from a woman aged fifty-five. The tooth was firm, and the patient had no anæsthetic. A few hours afterwards right-sided ptoſis existed, varied by occasional attacks of clonic spasm in the levator, each lasting a few seconds. Both symptoms continued, in slighter degree, on the following day, and gradually improved, so that the condition of the eyelid by the fifth day was natural. On the sixth day there was some pain referred to all the branches of the fifth nerve, but this soon passed off, and there was no recurrence of nervous symptoms.*

Ptoſis from Paralysis of the Sympathetic.—The unstriated muscular fibres (fibres of Müller), which exist in the fascia of the orbit, and are innervated from the sympathetic, act indirectly on the tarsal cartilages by the connection of these with the fascia. They probably aid to a slight extent in maintaining the upper lid in its normal position. When the cervical sympathetic is paralysed, the upper lid on that side is a little lower than on the other. Its movements are unimpaired. Ptoſis from this cause is distinguished by the presence of other symptoms of paralysis of the sympathetic, such as contraction of the pupil, and sometimes dilatation of the vessels of the surface or altered secretion of sweat.

Congenital ptoſis, not due to malformation of the lid, is usually bilateral and partial. It is generally associated with defective power of elevation of the globes, and is probably due to a congenital central defect. In one case there was also a defect in the outward movement of the eye, associated with an inward movement of the other, but absent when the eye was moved alone. Congenital ptoſis may be hereditary, and is probably allied to the more extensive infantile nuclear palsy already described. Slight double ptoſis sometimes runs in families, and may (as I have seen) affect chiefly the female members, sometimes coming on after puberty, and occasionally after middle life. The characteristic over-action of the frontal muscles causes the anxious aspect already mentioned, and this may actually be more conspicuous than the slight drooping of the lids to which it is secondary.

In another rare form the ptoſis is accompanied by wasting of all the subcutaneous tissues of the lid, which become almost translucent.

* Communicated to me by Mr. H. R. Gooding, by whom the case was observed.

Ptoſis on the same side

related

It

caused by
Right ptoſis followed
the occurrence of a
few hours after the
extraction of an

passed away slowly
in five days

Its occurrence in migraine has been
already mentioned

in the superior rectus
and other muscles. It
is sometimes hereditary
& part of the same

Both forms of
nuclear palsy may
only develop after
puberty from abio-
trophic failure—
defect of vital en-
durance

the ptoſis is accompanied by
also chiefly a woman

Allied to this is a form which may be termed *morning ptosis*. During sleep the levator is relaxed, to permit closure of the eyes by a gentle contraction of the palpebral orbicularis. Many sound sleepers find a difficulty in opening the eyes on being first roused. In weakly women this difficulty is sometimes increased; after waking, it may be impossible for them to raise the eyelids for ten, fifteen, or thirty minutes. During the rest of the day they have no difficulty. ~~This condition is usually recovered from.~~

*due to relaxation
of the levator
and slight spasm*

Hysterical ptosis is occasionally met with, single or double. It is generally accompanied by a slight spasm in the orbicularis; this can readily be proved by making the patient look upwards, when the ~~spasm~~ ^{contraction} of the orbicularis becomes much greater to prevent the lid from moving with the eyeball. When double* (Fig. 102) both eyelids droop, and the patient puts her head back when she is told to look up. If the head is held in the attempt to look up, both orbiculares contract, and prevent the lids rising. This contraction of the orbiculares proves that there is no true paralysis of the levators. There is sometimes over-action of the frontales associated with the ptosis, as if from a struggle to overcome the contraction of the orbiculares.



FIG. 102. — Hysterical ptosis, bilateral. Over-action of frontales.

TREATMENT.—The most important element in the treatment of paralysis of the ocular muscles is that of the morbid process that causes them, and this is described elsewhere. It is only necessary here to mention those measures that the special effect of the disease renders necessary. Whenever the onset is acute or subacute, or if other symptoms suggest inflammation, counter-irritation should be employed. A blister may be placed behind the ear or at the occiput when the disease is probably at the base of the brain, on the temple if it is in the orbit. The blister is often followed by a striking increase in the power of the affected muscle. In acute rheumatic cases hot fomentations should be applied to the orbit, forehead, and temple, frequently repeated. ~~Leeches may be applied to the temple, at the onset if the patient's general condition does not contra-indicate their use.~~ In syphilitic cases appropriate remedies should be freely employed. In most cases of local neuritis, indeed, whether syphilitic or not, mercury is probably the most efficient agent we possess. Defective general health ^{must} be corrected by tonics, of which iron and strychnia are the most useful.

and salicylates given

* A case is recorded by Debove (*La Semaine médicale*, 1890) of paralysis of both third nerves with double divergent strabismus, said to be hysterical. The condition came on after an accident.

For nuclear palsy, in syphilitic subjects, mercury and iodide have seemed useful only when the onset was acute or subacute. It is probable that in some instances these drugs have cured the patient, but in other cases it is doubtful whether the improvement has been more than the tendency to recovery which usually follows an acute process that has spent its force. In cases that are chronic from the beginning this treatment is as powerless as ~~it commonly is in locomotor ataxy~~, and the most effective measures are those suited to primary nerve degenerations elsewhere—arsenic, quinine, strychnine, and small tonic doses of mercury in syphilitic patients. ~~Strychnine may be given with advantage by hypodermic injection, as in progressive muscular atrophy (see vol. i, p. 497).~~ Such treatment as is here suggested appeared distinctly beneficial in one case, the sequel of which renders it peculiarly instructive. A gentleman aged forty, who had probably not had syphilis, presented entire loss of the reflex action of each iris, with large pupils; accommodation was normal, and the iris contracted with it. There was also slight double ptosis, weakness of both internal, of the left superior, and of the right inferior recti, and great diminution in the right knee-jerk, all of gradual onset and from six to eighteen months' duration. At the end of a three months' treatment the external ocular muscles had become normal, although the reflex action was still lost, and the right knee-jerk had increased to the same degree as the left. He continued without relapse for nearly two years, and then suddenly presented the acute mental derangement of general paralysis of the insane, from which he quickly died. But we do not yet possess the means of combating with success the toxæmic influences that are probably the most frequent cause.

Recorded cases of the recurrent or periodical palsy do not show that any treatment has a marked influence on the attacks. Early counter-irritation ^{may} probably afford most prospect of modifying their course, and tonic treatment may be adopted before an expected attack. In cases in which the attacks recur frequently, the treatment adopted should be that for migraine.

Direct Treatment.—Electricity has been recommended and employed in the treatment of these paralyses, but its influence is not great. Direct application to the affected muscle is scarcely practicable. It has, indeed, been carried out by a very small electrode, or by a wet camel-hair brush in the centre of which is a conducting wire. The extreme sensitiveness of the conjunctiva renders the application most painful, and although the pain may be lessened by cocain, the application of a voltaic current, sufficiently strong to stimulate the muscle, in such proximity to the retina and on such a delicate structure as the conjunctiva, is scarcely safe. In most ocular palsies the muscles do not respond to faradisation. ~~The voltaic current may be applied through the eyelid, in the case of a superior or inferior muscle, the eye being turned in the opposite direction. The considerable diffusive power of this current makes it probable that the~~

agents seemed to

take

especially by hypodermic injection

electricity would reach the muscle, although even thus a current cannot be safely used of sufficient strength to produce evidence of stimulation. The application of a faradic current through the eyelid is useless, even if the muscle would respond to it, because a thin layer of contractile muscle prevents any stimulation of the subjacent tissues.

For these reasons, most authorities are agreed in deprecating any direct application. A method of indirect electrification has been recommended (by Benedikt and others) which consists in placing one pole (anode) on the forehead, the other (kathode) on the margin of the orbit near the affected muscle. If the faradic current is employed, the orbital pole is kept still; if the voltaic current is used, this pole is stroked along the skin, or the circuit is alternately made and broken by an interrupter. A slight increase of power in the muscle may be observed after the current has been so applied for a few minutes, but it quickly passes off, and in cases free from sources of fallacy I have never been able to trace any permanent effect. It is true that works on electro-therapeutics contain cases in which improvement or cure was ascribed to this agent, but in most instances the authors have ignored the tendency to spontaneous improvement in recent cases, and the influence of drugs, as iodide of potassium, which were given at the same time in many instances.

The diplopia due to partial paralysis of a muscle may be removed by the use of a prism. One strong enough to completely fuse the images is undesirable, because it tends to prevent the strengthening influence of effort. A weak prism, strong enough so to approximate the images as to permit their fusion by muscular action, sometimes does good. It may be used for an hour daily as a gymnastic exercise. The giddiness due to erroneous projection can only be removed by putting the eye out of action by an opaque glass; this glass may be in a pair of spectacles, the other glass being merely tinted; the opacity is then inconspicuous to an observer. But this has the same disadvantage of withdrawing the stimulus of exertion. An opaque glass over the sound eye is useless for the vertigo, and increases the amount of the secondary deviation.

Operative interference can do little in cases of ocular palsy. The only condition in which it is admissible is that in which antagonistic contracture has been developed, and the paralysed muscle has regained power, but cannot overcome the opposing contracture. In such a case the tendon of the contracted muscle may be divided without disturbing its other connections; it forms a fresh attachment a few millimetres further back, and the result on the movement of the eye is often very satisfactory.

The treatment of paralytic ptosis is that of disease of the third nerve. The muscle itself is not accessible to electrical stimulation. In the double ptosis of nervous debility, nervine tonics, quinine and strychnia, are useful, and their influence is aided by local treatment, which stimulates the fifth nerve, and so produces a reflex action on

— 28 —
at 50

is not true

But the effect is
trifling and transient

is a question for the
Ophthalmic Surgeon

the centre of the levator. Electricity may be used, ~~either the voltaic or induced current~~ applied to the skin about the orbit, but a stimulating liniment which stings the skin (as chloroform liniment on spongiopiline) usually answers as well. The morning ptosis can always be quickly relieved in this manner. The hysterical form is often a very obstinate affection. Blisters to the temple, faradism, and (in the unilateral form) tying up the other eye, are the most efficient means, and usually in time remove the symptoms.

SPASM OF THE OCULAR MUSCLES.

Few forms

The varieties of spasm of the ocular muscles are numerous, but only a few are of medical significance. Two classes may be dismissed with a mere mention. (1) Those that are connected with disorder of the ocular visual process, as the convergent strabismus of hypermetropia, the divergence of myopia, the adaptive deviation that occurs when there is partial opacity of the media, and the irregular position that often accompanies absence of sight. (2) The secondary deviation in one eye, consequent on palsy of a muscle of the other eye, and the antagonistic contracture in the same eye. The second class has been already described; those of the first do not come within the scope of this work. Of the remaining forms, one of great importance—nystagmus—will be separately described. The remainder may be grouped into five classes.

*are unimportant;
those due to disorders
of refraction, and
secondary deviation
from weakness of an
antagonist.*

(1) *Associated spasm from central disease, is not uncommon.* Its character depends on the position of its cause: both eyes are affected in disease of the hemispheres; one eye only in some cases where the lesion is in the nuclei or the eye muscles. In a paralysing lesion of one hemisphere the eyes deviate towards the diseased side, but the deviation is merely due to the unopposed influence of the opposite hemisphere. An irritating lesion of one hemisphere causes conjugate deviation towards the opposite side, the result of spasm; this occurs also at the onset of unilateral convulsions, and is often associated with muscular rigidity of the limbs on the side towards which the eyes are directed. ~~In one interesting case, deviation to the right was, during four months, a persistent symptom of a depressed fracture below the left parietal eminence, and it ceased when the depressed bone was raised by trephining.*~~

non

Since a lesion in one side of the pons causes a loss of the conjugate movement towards the side of the lesion, the muscles being excited from structures in this position (see p. 186), irritating disease may

* Thompson, 'Brain,' April, 1883, p. 99. It may be noted that the fracture was behind the region in which experiment places the centres for the lateral movement, which is in front of the central convolutions (see p. 13). But the structures that overact are often only near a primary lesion, the influence of which is exerted on the neighbourhood, and sometimes only in one direction.

be expected to cause spasmodic deviation towards that side. But such spasm is seldom observed, apparently because the structures concerned are readily damaged and rendered inactive. ~~An acute lesion which causes the conjugate paralysis may, however, produce a distinctly spasmodic deviation towards the opposite side, probably by an indirect influence on the corresponding centre of the other side of the pons.*~~ Between the two there must be a close functional relation, indispensable for their normal action, and throwing light on many phenomena of disease (see "Nystagmus," p. 209).

be attended by a
with occasional clonic
spasm

(2) *Irregular spasm from brain disease.*—In irritating diseases of the base of the brain, especially in meningitis, spasm occurs in one or more of the ocular muscles, causing slight irregular deviation of the visual axes, comparable to the variable rigidity that occurs in the limbs, and may change in seat and degree from time to time. It is ~~probably due to irritation of the motor nerve-trunks.~~ Care must be taken not to mistake for spasm the deviation from loss of power which is so common from the same cause. In spasm there is deviation when the eyes are at rest in the mid-position; in recent paralysis there is not. It must be remembered that paralysis and spasm are often conjoined. Such irregular spasm is generally tonic.

always peculiar in
features and variable

Slight irregular spasm of the ocular muscles sometimes occurs in chorea. It is rarely sufficient to be seen, but causes transient diplopia, which may be thought to indicate organic brain disease if the dependence on chorea is not known. If patients are questioned on the point, this diplopia will be found to be by no means rare.

(3) *Chronic spasm in individual muscles* is extremely rare, apart from the secondary deviation already mentioned. ~~In the best marked cases the spasm has not been continuous, but has occurred on certain movements of the eyes, sometimes associated with pain.~~ Thus, of two cases recorded by Hock,† in one, as soon as an object was moved to the right of the middle line, spasmodic contraction occurred in the right internal rectus (with great pain), bringing the eye into extreme adduction, and as soon as the object was moved to the left beyond the middle line the spasm relaxed, and the right eye moved outwards into the fixing position. In the other case there was slight weakness of the right internal rectus, and as soon as the object was moved to the left of the middle line, spasm in the left external rectus moved the eye into extreme abduction. The spasm thus affected the muscle that would be the seat of secondary deviation, but was distinguished from this by its extreme degree. This case is interesting because the spasm occurred with slight left hemiplegia, and, from some accompanying disturbance of taste, may have been due to disease in

Usually
in the muscle which is
followed by spasm
in it. There may be
other evidence that
the cause is disease
in the upper part
of the pons.

* As in a case I have recorded of acute anæmia of the right half of the pons (due to arterial obstruction) which caused deviation of the eyes to the left. It was increased from time to time, with violent nystagmus ('Trans. Ophth. Soc.,' 1884, p. 308).

† 'Wiener Klinik,' April, 1876, p. 116.

the upper part of the pons. Sometimes two muscles ^{normally} acting together are involved in the spasm, as, in one case, the superior oblique and inferior rectus, which are normally associated (Stilling).

(4) *Hysterical Spasm*.—In hysterical fits the eyes are ^{often} usually directed either upwards and to one side, often so as to almost entirely conceal the cornea, or they are directed inwards in strong convergence—a common symptom. They never diverge as they do in organic disease. Sometimes the convergence persists during the intervals, and is then usually associated with spasm of accommodation.

(5) *Paroxysmal Spasm*.—In convulsive attacks in which the convulsion is unequal on the two sides, the eyes constantly deviate (with the head) towards the side most convulsed, and if the second side is affected in greater degree after the first, the eyes subsequently deviate towards that side. When the fit is over there may also be a deviation from the side most convulsed, accompanying the transient weakness common in that side. Such spasm is seldom unilateral, but during brief attacks of clonic torticollis (in a child of eleven months) drawing the head to the right, there was strong deviation of the left eye in the same direction. A single muscle may be the seat of brief spasm, resembling in miniature an epileptoid seizure, and sometimes attended with transient obscuration of consciousness. During the attack there is diplopia, and often giddiness from the erroneous projection. Clonic spasm in the orbicularis may occur at the same time. Such attacks, for instance, occurred in a man aged forty-seven; there was very slight permanent defect of power in the left external and both internal recti. Occasionally, without any exciting movement of the eye, tonic spasm in the external rectus would draw the left eye strongly outwards for about thirty seconds. During this time there was constant winking with both eyelids, which seemed to the patient to be due to an effort to get the eye right, but could not be prevented. The deviation ^{suddenly} ceased, but for a few minutes afterwards the left eyelid was about one twelfth of an inch lower than the other, and then both were alike. There was no history of venereal disease, and syphilis was for other reasons very improbable.

Another patient, aged thirty-six, also without history of syphilis, suffered from frequent attacks (lasting only a few seconds), two of which I witnessed. There was a sudden sensation of heat, spreading from the left inner canthus and extending over the eye and temple, accompanied with impairment of sight of both eyes, varying from slight dimness to absolute loss of sight, and with abduction of the left eye from mid-position to about halfway to the outer canthus, the right eye being still. If walking, he deviated to the left during the attack, probably from erroneous projection of the left field. In the intervals, movements of the eyes were perfectly normal. The attacks consisted of tonic and then clonic spasm, and resembled perfectly miniature epileptic convulsions. Some years later this spasm had ceased, but

be the prelude to nuclear palsy

in which there was abduction of the left eye only, half way from the mid position to the canthus. There was post

They proved to

Supporting

it develops when there is a condition lessening the amount of light which enters the eye, such as superficial disease of any kind. It also occurs in albinism, ^{the amount of light} when ~~an~~ ^{is} in excess. Thus there is an abnormal amount of light during the time when the child acquires the power of fixing a light, a process in which a reflex action ~~from~~ on the muscles takes an important share. This form consists of an alternate action of the same ~~rate~~ ^{rate}, there is not the quicker motion in the direction of vision.

Miners' nystagmus develops in those who have worked for years in getting coal, lying in a constrained position with the head inclined. It occurs especially in ~~the~~ badly lighted mines & is disposed to by defects of general health & alcoholism. It is often rotatory.

The common form, results from organic disease or degeneration, near ^{or in} the mid-brain, pons, or cerebellum. It is not met with in disease of the cerebral hemisphere, ~~away~~ ^{above} the central ~~nuclei~~ ganglia; rarely from disease in these, chiefly the optic thalamus. It may be caused by affections of the labyrinth of the ear which induce vertigo, an important fact in connection with the influence of cerebellar disease in causing it.

Pathology. ^{The slightest} All degrees of nystagmus may develop into those that are ~~more~~ considerable, and in these there is a deliberate alternation of the opposing muscles. This is apparently the result of an excessive activity of the muscle-reflex action between reciprocal muscles.

We cannot doubt that there is a ~~centre~~ for structures that observe this action in the vicinity of the nuclei of the ocular nerves, analogous to those that exist in the spinal cord. Sherrington has obtained, in the latter, a similar insubordination by cutting off the voluntary impulse. The same alternate movement occurs, and he has proved that the arrest of action in one muscle, followed by a contraction in the opponent, is due to an afferent ^{nerve} impulse produced by the extension of the latter, which inhibits the spinal centre for the former & is followed by activity of its own centre. Thus the same effect is produced, & the alternation goes on. ~~This is~~ the resemblance to metagamus is perfect. In this the acting muscles, say those moving the eyes to the right, contract under the will, but just before they reach the limit of movement, they suddenly relax from central inhibition; their opponents, the left-sided muscles, contract ~~in turn~~, & the eyes move back, rather more slowly, but just before reaching the mid-position, they are in turn arrested & a ~~quite~~ quicker action of the right-sided muscles again move the eyes to the right.

The structures (muscle-spindles) ~~are not found~~ which are believed to be sensitive to tension do not present the same aspect in the ocular muscles as in others, but their tendons present sensitive neural structures, and the fact that a twig from each muscle nerve passes to the fifth nerve, shows that afferent impulses must arise in them; they doubtless pass to the highest

cells of the fifth nucleus, adjacent to the motor nuclei. But the nuch-reflex alternation ~~is~~ not produced by a defect in the volitional impulse, as in the spinal centres. Its causes are such as may disturb the balance of the mid-brain structures, which subserve all ocular reflex action, ~~including~~ and also the association of the two eyes. The structural arrangement for the alternation of opposing ocular muscles must be rendered insubordinate with reading, as ^{is shown by the reading} ~~its excessive activity~~ with which it becomes excessive in infantile nystagmus, from a mere disproportion in the amount of light. This must act on the same centre, so far as concerns its influence on the ocular muscles, & enables us to understand that dim lighting is an element in causing the nuch's form. Nystagmus, ^{when} in moderate degree, only occurs on voluntary movement of the eyes, it is absent at rest, in the midposition. The reflex alternation is present only when the centre is energised by the volitional impulse. ~~This shows that the alternation arises in the lower centre.~~ But the latter causes the movement to be quicker in the direction of volition. At times, the nystagmus in a certain direction may attain ^{such} a degree that it continues when the will is not acting, and even in opposition to the will; e.g., the quick motion to the right may persist in the midposition, on upward & downward movements, and even during half the movement of the eyes to the left. These features show clearly that the symptom depends on the lower midbrain structures, & we can understand that the derangement may be slight, so as to cause the

nystagmus to have only the semblance of a slight jerking,
~~to~~ which it is reasonable to ascribe to the same mechanism
~~the slight jerking~~ until there is evidence to the contrary. Moreover the
 tendency to alternate action seems ^{sometimes} to ~~involve~~ be so partial
 as to involve ~~single~~ ^{individual} muscles and, in the oblique movements,
 in which two muscles take part, the movements may corres-
 pond to only one of them, with its opponent. The two eyes
 act alike, showing that the disorder is of structures above
 those for the binocular connection. We can perceive the
 complexity of the arrangement, but ~~the~~ anatomy ^{has} ~~does~~ not yet
 enabled us to localize or unravel it.

Among the influences that take part in balancing the
 lower centre are those exerted by the labyrinth, doubtless the
 semicircular canals & their relation to equilibrium. Increased
 pressure in an exposed tympanic cavity & therefore ^{the} labyrinth
 causes nystagmus, the quicker movement being towards ^{the other} ~~that~~ side;
 diminished pressure also causes it, quick to the same side.
 Brief nystagmus follows rotation of the body, and the injection
 of hot or cold water, or probing. Disease on one side of the
 pons, abolishing conjugate movements towards the side diseased,
 causes ~~energetic~~ nystagmus on movement to the other side,
 no doubt by ~~the~~ loss of balance of the reflex structures.

The slight jerking Nystagmus accompanies partial
 paralysis only when this has lasted for a time. It must be
 remembered that the ~~jerking~~ ^{the} impressions from the weakened
 muscles are lessened & that they exert an influence on the centre

The subjective symptoms of are often absent. The oscillations of the eyeballs must cause the image to move to and fro on the retina but an apparent movement is confined to cases in which the oscillation is wide and then only in the direction of the quicker motion. It is indeed ^{hardly} ~~not~~ more than a tendency to move, ^{in the same direction as the eyes,} as may be perceived by a voluntary quick movement of the eyes, to & fro. Perhaps this tendency to move is only the result of an after image, which disappears in the direction of motion. ^{There is no reason why there should be apparent movement} A movement of the eye, with an opposite & equal movement of the image on the retina, gives the impression that the object is still. Enduring infantile nystagmus is never attended by any sense of movement. On the other hand, it is frequent in the nystagmus ^{it is not easy to explain} of miners. The "dancing" of objects renders this form peculiarly disabling.

The practical importance of definite nystagmus is the evidence it affords of organic disease, visible or degenerative. In such a malady as disseminated sclerosis, ^{for instance,} with symptoms easily misunderstood, ~~the~~ its indication is often of great service. But it is of small localising value except in showing disease in the cerebellum or medial structures, and ~~the disease~~ a lesion outside the pons is probably on the side towards which the chief nystagmic movement takes place. Slight nystagmus may be left by an old paralysis of an ocular nerve which has quite passed away, doubtless from an enduring partial disturbance of balance in the reflex centre. ~~Very slight~~ A trifling nystagmus, on certain strained movements, may be congenital but too rarely to be important.

it; a motor sensation which, if intense, goes on to movement in which the eyes and head lead the way. Hence involuntary movement of the eyes is not surprising, the cause of the intermittence being, as already stated, a separate problem. In the nystagmus of miners, which may occur only in the recumbent posture, we may also trace, as one factor, an influence of the equilibrial centre (and probably of the canal impressions) on the centre for the movement of the eyes.

The wide variation in the position of organic disease that may cause nystagmus is scarcely surprising when we consider how wide and various must be the total connections of the functions of vision, of movement of the eyes, and of the maintenance of equilibrium; and that we can trace a connection between nystagmus and the derangement of each of these, including some causes of vertigo. But it is also common in various degenerative diseases in which we cannot trace its source, but which show how easily deranged must be the mechanism by which in health the muscular contractions are kept uniform. It would even seem as if the nuclear centres have a tendency to rhythmical or intermittent action, which is normally counteracted, possibly by the influence of the corresponding centres on the other side. When deranged, spasm that would be continuous in the limbs becomes intermittent in the eyes. This view is suggested by the case of arterial obstruction in the pons already mentioned. Nystagmus was clearly due to the over-action of the centres on the undamaged side, under the influence of loss of function of those on the other.* But this part of the subject needs far more systematic study than it has yet received.

The practical significance of nystagmus in diagnosis is extremely great, not from any distinct indication of the seat or precise nature of the disease, but because it shows the presence of more than merely functional disturbance. It is often marked at the early stage of degenerative disease when other symptoms are equivocal, and a search for it should never be omitted, and should always include the upward movement of the eyes. It may be trusted without hesitation, and prevents a mistake in diagnosis in a very large number of cases.

Spasm of the levator is occasionally met with as an isolated symptom, and also as one of the features of exophthalmic goitre. The isolated form (sometimes termed *lagophthalmos*, or "hare-eye") is usually due to some inhibitory irritation in the region of the fifth nerve. Late in life it may come on as an independent affection, analogous to other muscular spasms, as torticollis and facial spasm. The contraction is usually tonic, an excessive degree of the normal

* In continuous contraction the muscular fibres contract alternately; in clonus all seem to contract together, and the contraction is brief but renewed. Such a common brief contraction would explain the intermissions of nystagmus, the opponents being always ready to contract when the original action ceases. This readiness prevents the assumption that the common action of all the fibres is due to any other cause than the condition of the centre of the acting muscles.

tonic contraction of the levator which keeps the eyelid raised; the muscle is imperfectly relaxed when the eye is directed downwards or the lids are closed. Hence the eyelid is a little higher than the other when the eyes are directed straight forwards; and on looking down, the lid does not descend, so that a wide extent of sclerotic is exposed above the cornea. When the lids are closed, the upper lid may be brought down by the orbicularis, but not to the same extent as on the other side, and hence the lids do not descend equally. The exposure of the sclerotic above the cornea gives the impression of slight prominence of the ball. A long-continued spasm in the levator seems, indeed, to cause the eyeball to be actually a little more prominent than the other, in consequence of the origin of the levator being below the level of the upper part of the eyeball; but the apparent prominence due to the exposure of the sclerotic is much greater than the real prominence. The retraction of the upper lid may cause a slightly greater fulness of its tissues than exists on the other side. Unless the affection is due to removable irritation of the fifth nerve it is extremely obstinate.

Clonic spasm is also rare. It occurred from time to time in the case of reflex ptosis mentioned on p. 200. A very singular case of congenital spasm of the levator was exhibited at the Ophthalmological Society by Mr. Marcus Gunn. There was slight ptosis and slight myosis on one side, and the eyelid was raised by slight contraction of the levator whenever the external pterygoid of the same side was put in action.*

Spasmodic elevation of the upper eyelid also occurs from irritation of the cervical sympathetic. We have seen that this nerve supplies the plain muscular fibres of the orbit, which are indirectly connected with the eyelids; and that, as their paralysis may cause slight drooping of the lid, so their stimulation may produce slight elevation. It is probable that this spasm is the cause of the increase in the normal elevation, and of the defective descent of the lid in looking down, in many cases of exophthalmic goitre (Graves' disease). But this form will be considered in the account of that disease. I have once seen a similar slight retraction and defective descent, on the left side only, in a girl of seventeen, who had distinct myxœdematous swelling of the breasts, neck, and face, bilateral, but not elsewhere, and with no symptom of Graves' disease, and also, as a transient symptom on one side only (after anxiety), associated with cardio-thyroid disturbance so slight that alone it would scarcely have been noticed. Similar spasm is said to have been met with in pregnancy as a reflex symptom.

* 'Ophthalmological Transactions,' vol. iii, p. 283. As one of a committee appointed to report on the case, I had an opportunity of carefully examining it. The simplest explanation for the condition seemed to be that some of the levator fibres of the third nerve arose from the motor nucleus of the fifth nerve. The small size of the pupil might be due to the influence of those cells of the third nerve-nucleus which ought to have been, but were not, connected with the levator.

in Graves disease,
and more frequently
hindered descent. It is
ascribed to overaction,
perhaps with hypertrophy,
of the plain levator. It
is met with also in cases
of uncertain nature, pro-
bably partial forms
of this disease. In these
it may be unilateral

Various ^{central} ~~congenital~~
connections of the
levator have been
met with, congenital
in origin e.g. with the
external pterygoid. When
ever this muscle con-
tracts so did the
levator

Treatment.—Little can be done for the treatment of these ocular spasms beyond the removal of their cause as far as this can be effected. ~~Hook's second case was apparently cured by specific remedies, his first case by tenotomy.~~ Hysterical convergence can generally be removed by a small blister to each temple, if the potent influence of neglect is ineffective. The paroxysmal epileptoid form of ocular spasm is very obstinate. Bromides have little influence upon it, and tonics do more good than sedatives. Counter-irritation to the temple sometimes seems to produce a beneficial effect.

In the treatment of nystagmus, the improvement of vision, if this is defective, and the removal of any cause that can be discovered, are the chief measures. The form that occurs in miners needs, as a rule, a change of occupation, or at least of habitual position, so as to relieve the eyes from this postural strain, together with an improvement in the light by which the work is done. ~~For other forms very little can be effected. The application of a feeble voltaic current, from the mastoid process to the closed eyelids, was recommended by Svetlin, but has failed in the hands of others.~~ But the symptom, in many cases, scarcely calls for treatment.

Spasm of the levator is a most intractable affection, unless there is irritation of the fifth nerve or any other cause which can be removed. When no cause can be traced, the most varied treatment by counter-irritation, sedatives, and electricity usually fails. Associated with any symptoms of exophthalmic goitre, the treatment is that of the latter disease.

*only be removed by treatment
of this cause, if this is
possible*

the lenticular nucleus, within it, or in the caudate nucleus. The extravasations outside the nucleus are restrained externally by the grey cortex of the insula and its ^{adj.} adjacent layer of white substance; they may tear up or displace inwards the central ganglia. Blood within the ganglia often extends upwards and outwards in the white centrum ovale, and may attain a ~~very~~ large size. The posterior hæmorrhages are due to the rupture of the lenticulo-optic arteries of the middle cerebral, which pass through the lenticular nucleus into the anterior part of the optic thalamus. The extravasation usually commences in the thalamus, or between it and the corpus striatum, and the blood is apt to escape between the two into the lateral ventricle. ~~They often damage the posterior (sensory) part of the internal capsule.~~ The small arteries to the inner portion of the thalamus, which pass from the posterior cerebral, or posterior communicating, sometimes ~~give way and~~ cause small extravasations near the surface (internal thalamic hæmorrhages), which are very prone to rupture into the ventricle. The branches of the posterior cerebral to the hinder part of the thalamus may give rise to extravasations in this situation (posterior thalamic hæmorrhage), which may either rupture into the lateral ventricle or may extend down into the crus and even into the pons.

Centrum Ovale.—Large hæmorrhages ^{into the corpus striatum} usually spread into the centrum ovale ~~from the corpus striatum.~~ The vessels in the white substance itself are small, and give rise only to small hæmorrhages, rarely larger than a walnut, and often oval, with the long axis in the direction of the fibres. Larger extravasations are ~~sometimes found~~ in the white substance of the occipital lobe. ~~These are due to the rupture of branches of the calcarine division of the posterior cerebral artery.~~ ^{may occur}

Cortex.—Hæmorrhages limited to the cortex may occur in almost any position, but they are rare and are usually small, ^{unless} although they ~~sometimes~~ extend into the white substance, and attain in it a larger size. ~~This occurs chiefly when the membranes have been thickened by chronic inflammation.~~ ^{in consequence of}

Crura Cerebri.—Hæmorrhages into either the corpus striatum or optic thalamus may extend down into the crus. Those which commence in the crus are usually small, and oval in form. They may descend into the pons, but do not pass up into the central ganglia. ~~They may be situated in the inner part of the crus, or in the outer part beneath the corpora geniculata, or in the upper part beneath the corpora quadrigemina.~~

Pons.—Hæmorrhages are most frequent near the middle line, from the rupture of the median branches of the basilar; ~~and the raphe usually prevents their extension to the other side.~~ The extravasation ^{is} often ~~has a spherical shape, and may be kept from the fourth ventricle only by a thin layer of tissue (Fig. 117), which may give way, and the blood may escape into the cavity.~~ Sometimes small

^{the blood may burst into transverse}

wounded

or be kept from this

extravasations may be
~~hæmorrhages extend in a transverse direction, and these are due to the rupture of the small lateral branches of the median arteries. Hæmorrhages in the lateral portions of the pons, from rupture of the radicular arteries, are rare. The vessel that most frequently gives way is the branch to the root of the fifth nerve.~~

but may
 Cerebellum.—Large hæmorrhages are most frequently due to the rupture of a branch that the superior cerebellar artery gives to the dentate nucleus. ~~A small extravasation may occupy the interior of the dentate nucleus, arising from a branch of the same artery. Hæmorrhage into the inner and hinder part of the hemisphere may occur from rupture of a branch of the posterior cerebral. Extravasations near the fourth ventricle readily burst into it. The~~

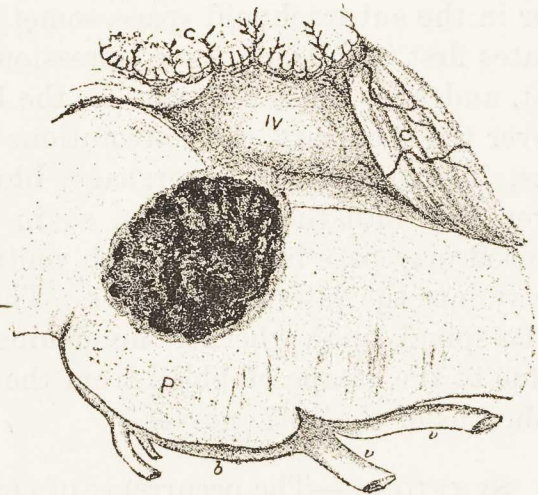


FIG. 117.—Hæmorrhage into the pons, on the left of the middle line. (After Carswell.)

cortex of the cerebellum offers much less resistance to hæmorrhage than does the grey matter of the cerebral convolutions. ~~The superior and inferior cerebellar peduncles are rarely the seat of hæmorrhage, but an Extravasation, sometimes occurs in the middle cerebellar peduncle, and, more frequently, a hæmorrhage in one half of the pons may extend a short distance into the peduncle, passing in the direction of its fibres.~~

into the peduncles are rare
 In ventricular hæmorrhage, whether primary or secondary (by rupture of an extravasation in the substance of the brain), the blood fills all the ventricles in a third of the cases. If slowly effused, it may occupy only one lateral ventricle, and such limitation is met with in about one quarter of the cases of each variety. It occupies both lateral ventricles alone (not extending to the third and fourth), more frequently in primary than in secondary hæmorrhage. It is rarely confined to the fourth, still more rarely to the third. The blood is usually coagulated, ~~and if small in amount the clot may correspond in form to the ventricular cavity.~~ If the blood is slowly effused, the distension of the lateral ventricles may be less than that of the third and fourth; in the latter the accumulation of blood is aided by the influence of gravitation in the recumbent posture. In primary hæmorrhage the walls of the ventricles may be intact, the choroid plexuses are anæmic from pressure, and it may be difficult to discover the source of the blood. Superficial lacerations of the wall of the distended ventricle may be of secondary origin, as is shown by the fact that there are many of similar aspect. Sometimes it is difficult to say whether laceration is the source of the hæmorrhage or is produced by it. Occa-

It parallels to preceding paragraph

sionally there are also separate extravasations into the substance of the brain; or a meningeal hæmorrhage may co-exist, even when the ventricular extravasation is small.

The appearances in *meningeal hæmorrhage* differ according to its seat and amount. There is a layer of blood either upon the arachnoid or in the subarachnoid space, sometimes in both. The blood accumulates first in the sulci and depressions, and is generally most abundant at, and sometimes confined to, the base. If considerable in quantity over the convexity, the convolutions may be ~~distinctly~~ flattened. Just as, in ventricular hæmorrhage, blood may escape from the fourth ventricle into the meninges, so in extensive meningeal hæmorrhage blood may pass into the fourth ventricle, and a little may even find its way into the lateral ventricles. The blood may also pass down into the spinal canal, whether the meningeal hæmorrhage is primary, or is due to the escape of blood from the ventricles. It often distends the sheaths of the optic nerves.

SYMPTOMS.—The occurrence of cerebral hæmorrhage is indicated by symptoms of two classes, the one general and transient, the other local and more or less permanent. Both sets of symptoms are usually present, but either may be so slight as to be inconspicuous. In addition, the onset may be preceded by what are termed premonitory symptoms, and followed by symptoms outside the cerebral functions, alterations of pulse, temperature, urine, &c.

The premonitory symptoms, so called, are on the whole rare. They consist of headache, slight vertigo, weakness or tingling in the limbs, slight mental changes, or slight affection of speech. These symptoms may be continuous or ~~paroxysmal, coming on suddenly and passing away after a few hours or days.~~ *transient* It is doubtful how far it is correct to speak of them as prodromata of hæmorrhage. The miliary aneurisms, however numerous, cause no symptoms until they burst. *latter* The more severe antecedent symptoms are sometimes due to the occurrence of actual small hæmorrhages, or to the commencement of a hæmorrhage that is at first slight and afterwards ^{rarely slow} suddenly becomes considerable.* Slighter "prodromata" may be due to the atheroma that often co-exists with miliary aneurisms, and they are the result of interference with the blood-supply to certain parts of the brain. *but slight antecedent symptoms, &c.* They thus indicate merely an associated condition. Hence they are far less frequent before attacks of hæmorrhage than before attacks of thrombotic softening, and they are similar in the two cases.

Of the general cerebral symptoms the most common is loss of consciousness; of the local symptoms, hemiplegia. The loss of

* Thus a young man, who had suffered from lead-poisoning, was attacked with severe pain in the right side of the head, and after this had continued for ten days, left hemiplegia suddenly came on, without loss of consciousness. He subsequently died, and a hæmorrhage was found in the right hemisphere, which had damaged the whole thickness of the internal capsule, but had apparently occurred in two stages (Hardy, 'Gaz. méd. de Paris,' June 11, 1880).

the hæmorrhage is in the cortex, although hæmorrhage into the corpus striatum or elsewhere now and then causes a general convulsion. Vomiting occasionally occurs, more frequently when the hæmorrhage is into the cerebellum than when it is in other parts of the brain. Its occurrence is probably influenced in part by the condition of the stomach; if a meal has been taken shortly before the onset, the process of digestion may be arrested, and the contents of the stomach may be vomited.

The temperature usually falls within an hour after the onset, and may be only 97° or 96° ; it may even reach 94.4° in the rectum. In cases fatal during the first twelve hours, the fall may continue until death; in other cases the temperature may become normal at the end of the first day, and remain so in slight cases, or rise, in more severe forms, to two, three, or four degrees above the normal (Bourneville). But there is one apparent exception to the rule of initial depression. In cases of hæmorrhage into the pons or medulla, the temperature may rise above the normal within an hour of the onset, and during the next hour may attain a height of 104° or 106° , and the rise may go on until death. It may also rise above the normal when the hæmorrhage is into the grey matter of the central ganglia, as Hale White has shown, by experiment and observation †

The duration of the apoplectic state varies much in cases that recover from it. At the end of from half an hour to six hours reflex action returns in the limbs, the patient gives some signs of returning consciousness, and moves those parts that are not paralysed. Difficulty of swallowing and impairment of articulation may exist for a day or two. Full general headache is usually felt as consciousness returns, and there is occasionally some delirium.

Even during the state of apoplexy, some indications of a unilateral lesion may be present; reflex action, if lost, returns on one side only; on the other it remains absent in the foot, cremaster, or abdomen: the muscular tone is different in the two arms, as already mentioned; and sometimes the affected side becomes rigid. The head and eyes may be directed to one side in "conjugate deviation," usually from the side on which the limbs are affected, sometimes towards it, according to the laws already stated (p. 78). As the general cerebral symptoms lessen, the local symptoms, persisting, become more prominent. The return of movement in the unaffected limbs defines the paralysis, which is usually hemiplegic. When the loss of consciousness is slight or absent, the loss of power is conspicuous from the first, and the subjective symptoms, already described as occasionally attending the onset of apoplexy, rise into greater prominence.

The attack often occurs during sleep. The patient is either found

* In a case of extensive ventricular hæmorrhage, due to the rupture of an aneurism, recorded by Bastian ('Trans. Clin. Soc.,' 1883, p. 18).

† A gradual rise to 107.5° during the two days life endured, occurred in a case of hæmorrhage into the lenticular nucleus, observed by Pasteur ('Lancet,' 1889).

has been known from

presence of food in the

94.4°

has been met with

even in slight cases, with delay

unconscious in the morning, or wakes unaware of what has happened, and only discovers his paralysis when he attempts to stand. The state of sleep has been thought to favour the occurrence of hæmorrhage, but it is not proved that the number of cases occurring during sleep is larger than corresponds to the proportion of existence spent in that condition.

there is In all except the slightest cases, there occurs, two or three days after the onset, slight general febrile disturbance, due to inflammatory changes about the cerebral lesion. There may be merely headache, loss of appetite, quickening (sometimes slowing) of the pulse, and a rise of temperature of one or two degrees, lasting a few days. In other cases the rise of temperature is greater, consciousness may be dulled or even again lost, or there may be some delirium. If consciousness has not been regained, the coma deepens, there is a tendency to sloughing and vesication of the skin, swallowing is impossible, mucus accumulates in the chest, and the patient usually dies. During this stage of inflammatory reaction, rigidity ("early rigidity") often develops in the paralysed limbs. The urine becomes less abundant, and may lose its acidity.

Before the initial coma has passed away, it may be suddenly deepened, or consciousness, after being regained, may be again suddenly lost, in consequence of the extravasation bursting into the ventricles. The symptoms of this occurrence are described further on.

As the general cerebral symptoms subside, only those remain that are due to the local effect of the lesion, and consist in the loss of the functions subserved by the structures destroyed. Since the hæmorrhage is usually in one side of the brain, the persisting symptoms are commonly unilateral in distribution. But the unilateral symptoms that can be at first recognised are much wider in extent than those that ultimately remain. There may be at first an apparent loss of function of almost all parts of one hemisphere, and even of the whole brain. There is not only loss of motor power, but there is often loss of sensibility, and this in cases in which the ultimate loss is purely motor. Indications of hemianopia may often be found in the early stage; if the finger is brought suddenly before the eye, first from one side and then from the other, it will be found that the eyelids blink when the finger comes from the unparalysed side, and not when it comes from the affected side. The early conjugate deviation of the head and eyes, already mentioned, is sometimes a symptom of the same character. These wide initial symptoms show that the interference with the function of the hemisphere extends at first far beyond the limits of the destruction wrought by the hæmorrhage. The effect is probably partly due to the influence of the pressure on the adjacent nerve-elements, partly to anæmia produced by that pressure, partly to inhibition by the irritative influence of the acute lesion. These symptoms gradually lessen; some, such as the hemianopia, usually pass away in a few days, others in a few weeks, until there remain only

*body hemispheres
may be*

often

*structures
early*

those that are the direct effect of the destruction; these persist for months, and, unless the loss is of such a character that it can be supplemented by the action of the other hemisphere, they continue for the rest of life. ~~The pressure is, of course, greatest on the structures nearest to the hæmorrhage, and the symptoms thus produced last longer than do those that result from the slighter pressure on distant parts.~~ The symptoms produced by these two mechanisms have been distinguished as "direct" and "indirect" symptoms. ~~But the two are the same in character, and can only be distinguished in practice by the gradual disappearance of the one and the persistence of the other.~~

first few days In some cases there is a slight increase of the local symptoms during the period of "inflammatory reaction," due, no doubt, to the damage to adjacent structures by the inflammation around the lesion.

usually **Chronic Stage.**—The enduring symptoms, which persist after the initial stage is over, are due to the local interference with the functions of the damaged part of the brain, and are determined by the situation of the lesion. Persistent general cerebral symptoms, such for instance, as are so conspicuous in cases of tumour, are for the most part absent in hæmorrhage. Headache is trifling; optic neuritis is practically unknown. Convulsions are rare. Some mental change may be present, slight or considerable, and evidenced chiefly by defective memory, irritability, and emotional instability, but is less than in atheromatous softening.

Persistent Local symptoms, persistent in most cases, are absent when the lesion is so placed as to spare the structures concerned directly in motion, sensation, &c.

The most common symptom is motor hemiplegia, because hæmorrhage is most frequent in the region of the corpus striatum and internal capsule, and the anterior part of the hinder line containing the fibres of the motor path rarely escapes laceration or compression.

When When due to damage to the motor centres or path, the hemiplegia gives the characters already described, and is attended with rigidity, excess of wrist- and knee-jerk, and with the foot-clonus that indicates secondary degeneration in the pyramidal fibres. Hemianæsthesia is less common, because hæmorrhage is frequent near the posterior part of the capsule, in which the sensory tract is contained. But it is not uncommon to have some blunting of sensibility, especially on the extremities of the limbs on the hemiplegic side. Increased sensitiveness to pain is sometimes present, and may co-exist with tactile anæsthesia, and be accompanied by spontaneous pains, but these are less common than in cerebral softening. Trophic disturbances in the hemiplegic side vary in frequency and degree; there may be persistent

* A not very happy distinction, since the pressure effects are as much the direct effects of the hæmorrhage as is the laceration. Only the inhibitory symptoms are, strictly speaking, indirect, but these cannot be separated practically from those that are due to pressure.

compensated

Both are due to loss of function of the nerve structures, & whether this is due to slight compression or to destruction we can only be ascertained

due to destruction of tissue depend on

are rare, such as headache or general convulsions, & optic neuritis is unknown as a secondary effect. It may be due to as an effect of renal disease, or when the hæmorrhage has been in a soft glomerulus

paralysis

not there is often

elevation of temperature, the muscular wasting, and the joint inflammation described in the chapter on hemiplegia. The atrophy of the muscles varies in degree in different cases, but it is moderate, and is not attended with any considerable change in electrical excitability, or any destructive degeneration of the ganglion-cells of the cord.

The symptoms produced in each locality are for the most part those that have been described in the chapter on "Localisation," but a few additional facts, relating especially to hæmorrhage, may be here mentioned.

Cortex.—Hæmorrhage into the ^{cortex} substance of the cortex is very rare, but occurs sometimes from aneurisms about which there is sufficient inflammatory thickening to prevent external rupture. This may be produced when embolism is the cause of the aneurism, and the inflammation in the wall of the artery spreads to the tissues over it. The onset is often attended by convulsions, which begin locally, and may be one-sided if the motor area is affected. Subsequent paralysis may affect only part of one side. Hæmorrhage into the centrum ovale causes symptoms similar to those due to a cortical lesion in the corresponding situation, but without symptoms of irritation of equal degree, unless the lesion is just beneath the motor cortex, when, for some reason, irritation symptoms (convulsions and rigidity) may be severe and lasting (Mills).

Crus Cerebri.—A limited extravasation may cause the characteristic crossed palsy of third nerve and limbs; but often the hæmorrhage passes beyond the limits of the crus, either upwards into the foot of the internal capsule (and then causes well-marked hemianæsthesia as well as hemiplegia) or downwards into the pons, and may then paralyse both third nerves, and the limbs on both sides. The corpora quadrigemina are never the seat of limited hæmorrhage.

Pons Varolii.—Initial loss of consciousness may be present or absent, just as in hæmorrhage elsewhere. Initial convulsions are especially frequent, rarely unilateral, but usually general; sometimes affecting the legs only, a symptom almost unknown from disease in other parts. The convulsion is often irregular in type, sometimes tonic, or tonic varied with occasional clonic jerkings. The paralysis is often bilateral, but a small hæmorrhage may cause hemiplegia from the first. Bilateral palsy may affect mainly the legs or the arms. Anæsthesia often accompanies it, and may even preponderate. The pupils are often strongly contracted, so as to suggest opium poisoning, or they may be dilated and motionless,—the difference depending on the irritation or paralysis of the nuclei of the third nerves. The deviation of the head and eyes from the side of a unilateral lesion is occasionally observed (see pp. 318 and 186). Respiration often suffers early, and may present irregularities, sometimes almost convulsive in their character. Vomiting is frequent. The temperature often rises rapidly to a hyperpyrexial degree. Death is usually more speedy

Seldom Considerable

An occasional cause of the rupture of an aneurism is the meningitis thickening of the vessels, resulting palsy or convulsions. It is at first local; the latter is especially common & is also met with when the extravasation is beneath the cortex.

only either impaired

Centres

changed; however may & occurs the almost convulsive. A rapid rise of temperature

is a characteristic symptom, of fatal augury.

Sensation

Seldom

than in hæmorrhage into the cerebrum; it has occurred in ^{a few} ~~seven~~ minutes.

Medulla Oblongata.—A considerable hæmorrhage causes death very quickly, and even instantaneously. It is doubtful whether convulsions occur when the extravasation is confined to the medulla. The patient rarely survives the actual onset; if he does, the symptoms that persist resemble those of bulbar paralysis, but this is extremely rare; sudden ~~apoplectic~~ bulbar paralysis is due, in the vast majority of cases, to softening from vascular occlusion, not to hæmorrhage.

almost always,

Cerebellum.—Loss of consciousness is as common as in hæmorrhage elsewhere, and presents the same variations in its degree. There is often no initial paralysis; sometimes there is hemiplegia, due to pressure, and it may be on the same side as the hæmorrhage, or on the opposite side, according as the pressure is exerted on the pons or on the medulla. The absence of hemiplegia is more significant than its presence. The pressure may also cause various paralyses in the parts supplied by the cranial nerves that arise from the pons and medulla. The state of the pupils varies; vision is unaffected. Vomiting is more frequent than in hæmorrhage elsewhere, being met with in half the cases, and it is also more often persistent. It may occur without, as well as with, loss of consciousness. If recovery takes place, the pressure-effects, including the hemiplegia, pass away, and the only lasting symptom is cerebellar unsteadiness, which persists in some, but not in all the cases. Hæmorrhage into the cerebellum often bursts into the fourth ventricle, causing fatal depression of the functions of the medulla. When the middle cerebellar peduncle is the seat of hæmorrhage, the characteristic symptoms, forced lateral position of the head, difference in height of the eyes, and a tendency to lie on one side, and even to rotate, is usually distinct, even sometimes at the onset during the stage of apoplexy, and if the patient is conscious there is usually intense vertigo.

Ventricular Hæmorrhage.—The effusion of blood into the ventricles of the brain is indicated by severe apoplectic symptoms, the origin of which is easy to understand. When, as in the majority of cases, the rupture is into one lateral ventricle, the blood rapidly distends the other also, and the whole cortex is compressed. The blood soon passes into the third and fourth ventricles, and compresses the important structures that lie in the floor of the latter. In the rare cases in which the rupture is into the fourth ventricle the pons and medulla bear the full force of its pressure, and the ~~grave symptoms of interference with this part of the brain are conspicuous from the first, and resemble closely those produced by hæmorrhage into the substance of the pons.~~ This is also the case when the rupture is into the lateral ventricles, and occurs slowly; the fourth ventricle ~~may then be much more distended than the lateral ventricles (see p. 395).~~

the blood passes down into

which may

Hæmorrhage into the ventricles is sometimes primary, but much

* Mickle, 'British Med. Journal,' 1881, ii, 151.

one-sided / more frequently secondary to hæmorrhage into the cerebral substance, from which the blood tears its way into the cavities. In the former case the severe apoplexy is primary; in the latter it supervenes on the symptoms of the cerebral hæmorrhage already described. The primary apoplexy may have passed away or have lessened, or may be still at its height, when it suddenly returns or becomes deeper. The initial deviation of the head and eyes ceases, and ^{may be} is often replaced by a deviation in the opposite direction. The pulse is again slowed, and ~~may fall to 50 or 40 beats per minute.~~ The temperature ~~may fall to 97° or 96°.~~ The respiration becomes more laboured and stertorous. The reflex action is ^{often} lost in the limbs and eyes; the pupils are sometimes dilated, sometimes contracted; rigidity often appears in the limbs on the hemiplegic side, sometimes in those on the other side; there may be convulsions, sometimes general, sometimes affecting only the unparalysed side. Paroxysms of general and clonic spasm, with slight opisthotonos, have been observed.* After a few hours the pulse ~~often~~ becomes more frequent; the temperature may remain low or may rise, sometimes to 104° or higher. The chest becomes filled with râles, occasionally with extreme rapidity, even within a couple of hours of the onset; the face becomes livid, respiration is increasingly difficult, and the patient dies from the interference with breathing. In many cases the blood escapes from the fourth ventricle into the subarachnoid space at the base of the brain, and the symptoms may be in part due to this.

Primary ventricular hæmorrhage causes symptoms which may, from the first, closely resemble those of the secondary form, but more frequently the onset resembles that of hæmorrhage into the substance of the brain, in the presence at first of unilateral symptoms. Prodromata are rare, but headache is occasionally met with, very variable in seat, character, and duration. The onset may be—(1) By sudden apoplexy, deepening rapidly, death may occur in a few hours. (2) By apoplexy with hemiplegic symptoms, or with convulsions. (3) In the very rare slow hæmorrhage, hemiplegia first occurs alone, loss of consciousness only supervening after a few hours. Hemiplegia occurs (because the blood is effused first into one lateral ventricle) and causes paralysis on the opposite side by the compression of the motor path or centres. When the effusion is rapid and both lateral ventricles quickly become distended, the unilateral symptoms quickly give place to general relaxation of the muscles, and loss of all reflex action. Rigidity is often met with, but less frequently than in the secondary form; it is usually bilateral, sometimes one-sided, and occasionally involves only the muscles of mastication: it may be intermittent. Convulsions are also frequent, occurring in at least a third of the cases, sometimes general, sometimes affecting only the paralysed side, or a part of it. In cases of slow onset, speech is often lost before consciousness. The power of swallowing usually persists until the

* Bastian, 'Trans. Clin. Soc.' 1884, p. 22.

unilateral, or irregular

ful. and coma follows, as

and

apoplexy becomes profound. ~~The temperature resembles that of other forms of cerebral hæmorrhage.~~ The malady is usually fatal, ~~but recovery has occurred, shown by old and altered clot in the lateral ventricles; but it is only possible when the hæmorrhage is so small in quantity that the symptoms are slight and equivocal.~~

Meningeal Hæmorrhage.—The symptoms of meningeal hæmorrhage vary much according to its cause. The rupture of a large aneurism at the base causes severe apoplexy, rapidly deepening to lethal intensity. But since slight hæmorrhage sometimes occurs before the final rupture, the loss of consciousness may be preceded by definite prodromata, and is so preceded more frequently than is intra-cerebral hæmorrhage. These prodromata are severe headache (sometimes occipital), giddiness, and occasionally vomiting. The attack itself is attended by paralysis and atony of the limbs on both sides, and sometimes by palsy of cranial nerves or by convulsions.

When the hæmorrhage is of traumatic origin, the effect of the injury obscures the initial symptoms. In these and other cases in which the escape of blood is gradual, the patient may recover consciousness and continue his occupation for some hours or even for a day or two, complaining only of headache, and then gradually become somnolent and pass into a state of coma.

In some cases of meningeal hæmorrhage, convulsions are the most prominent symptom, and they may be either general or unilateral, and in the latter case may commence locally in the face or arm, or by deviation of the head. Rigidity of limb is comparatively rare, far more so than in meningitis. In some cases there is mental excitement or delirium, in others there is mental dulness. The state of the pupils is very variable; they may be contracted or dilated or unequal. Sometimes headache and giddiness are accompanied by tingling in the limbs and weakness on one or both sides. Initial apoplexy may be absent, and these symptoms, commencing suddenly, may increase until consciousness suffers. The variation presented by these cases is thus very great.

In some cases, in young adults, the occurrence of meningeal hæmorrhage is probably indicated by intense pain in the head at one spot, soon followed by coma and by convulsions. The cases are remarkable as occurring without other exciting cause than exertion in a hot room, sometimes after a meal. The patient may recover partial consciousness in a day or two, and present for about a week the symptoms of slight meningitis, but considerable mental dulness is apt to ensue, and to continue sometimes for months. Or in this, as in other forms of meningeal hæmorrhage, the patient may die in the state of coma, after a few hours; sometimes death seems due to the violence of the convulsions. Coma occasionally passes away and ~~returns~~ ^{reforms}. The frequency with which recovery occurs cannot well be estimated. ~~The~~ Meningeal hæmorrhage of newly born children is separately described.

during birth

PATHOLOGY.—While the rupture of the vessel is always the result of weakening of its wall, and of the pressure of blood within it, the actual conditions of rupture vary considerably. The variations relate to the degree of blood-pressure—the size of the artery, of the aneurism, and of the opening ~~in it~~—the freedom of exit of the blood, or the hindrance to its escape by clot within the dilated part, and the resistance in the tissue into which the blood passes, which is less in the grey than in the white substance. The precise conditions in any individual case can rarely be traced, but on them must depend the size of the hæmorrhage, and the rapidity with which the blood is effused. These two elements chiefly determine the symptoms that attend the onset. No doubt the occurrence of both aneurism and hæmorrhage is due largely to the slightness of the support that the cerebral tissue affords to its vessels. This is less in the old than in the young, on account of the larger size of the perivascular spaces, occupied only by mobile liquid.

It is a well-known law of hydrostatics that if a liquid passes into a closed chamber by a small opening, the total pressure within the chamber is that in the opening multiplied by the number of times the area of the wall of the chamber exceeds the opening in size. Thus the total pressure within the aneurism must be much greater than in the artery from which it springs, and, at the same time, the wall is weakened. After rupture, the blood in the cavity formed by the extravasation will be influenced in the same way; and although the actual condition and the result are doubtless modified, the general law must hold good in some degree. We are thus able to understand how so small a jet of blood may produce a cavity in the brain of so large a size. Doubtless the process is facilitated by the softening and disintegration of the adjacent tissue. On the other hand, the transmission of the pressure must be modified and retarded when the blood begins to coagulate.

The most common seat of hæmorrhage is the region of the corpus striatum, because the arteries that most frequently rupture are the branches that come off at right angles from the middle cerebral/ in the fissure of Sylvius, and pass upwards through the lenticular nucleus and the internal capsule to the caudate nucleus and optic thalamus (see Fig. 44, p. 62). A large artery which passes between the outer part of the lenticular nucleus and the external capsule, then through the former to the internal capsule, is so frequently the source of the extravasation that it has been termed by Charcot *par excellence* “the artery of cerebral hæmorrhage.” A small hæmorrhage from it simply separates the external capsule from the lenticular nucleus, occupying a narrow fissure thus formed, which may correspond to almost the whole outer aspect of the lenticular nucleus, but a large hæmorrhage displaces inwards and erodes all the central ganglia. All these branches of the middle cerebral pass to the internal capsule, and as this lies over the inner upper aspect of the lenticular nucleus, it rarely

escapes damage—by pressure if the hæmorrhage is small, by laceration if it is large. Hence hemiplegia, transient or permanent, is so common a symptom. The branches that have a posterior course, to the hinder part of the capsule, less frequently give way, and hence lasting hemianæsthesia is comparatively rare. The pressure in these vessels has been found to be little less than that in the carotid (Mendel),—it is supposed because they have no anastomoses; that in the arteries of the cortex is far lower.

The mechanism by which the loss of consciousness and the other symptoms of apoplexy are produced has been previously considered (pp. 99 *et seq.*). It is only necessary here to repeat that we must recognise a double mechanism, the mechanical effect of the pressure on the cortex, and the inhibition of its cells by the mechanical irritation of the torn nerve-fibres. Both of these effects are the greater the more rapidly the blood is poured out, and the larger its amount; while the inhibitory effect is doubtless influenced also by the position of the lesion, injury to structures (as the fibres in the pons) that have an extensive connection with the cortex being specially effective.

Thus consciousness is preserved at the onset, only when the hæmorrhage is small, or the blood escapes very slowly, or when the extravasation is so placed that the irritation has but a slight influence on the cortex. When the effusion occurs slowly but the conditions are unfavorable to the cessation of the flow of blood, consciousness may be lost, not suddenly at the onset, but gradually, as the hæmorrhage attains a considerable size, giving rise to the “ingravescent apoplexy.” As Broadbent has pointed out, this form is often due to rupture of the artery that passes outside the corpus striatum, although even in the case of this vessel there is sudden initial apoplexy much more frequently. The difference is probably due to the character of the rupture in the wall, whether it is large or small. Ingravescent apoplexy may also occur from rupture of a vein (as is often seen in traumatic hæmorrhage); the pressure in the vein is low, and the blood escapes slowly. It may even result from the rupture of a surface aneurism into the brain substance, the membranes being thickened and resistant.

DIAGNOSIS.—The diagnosis of the nature of the cerebral lesion has to be made under two conditions; first during the initial apoplexy, and secondly when this has passed, and only the enduring effects of the lesion remain. The first is incomparably the most important, since by it the treatment has to be determined. Unfortunately, it is often, of all the diagnostic problems presented to the physician, at once the most difficult and the most urgent.

The first question in diagnosis is whether an attack of apoplexy is of cerebral origin. The chief points in the differential diagnosis have been considered in the account of this condition. If cerebral symptoms, such as unilateral numbness or weakness, have preceded the

This

always

unconsciousness

(See Apoplexy)

loss of consciousness, or are to be traced in it, as by one-sided relaxation of limb or deviation of the head, the cerebral nature of the attack is clear. If these are absent, the question must be determined by the considerations mentioned at p. 103. An actually sudden onset limits the diagnosis to the distinction from syncope, and this should be easy. If the onset was gradual, or, as is often the case, its character cannot be ascertained, the distinction has to be made from poisoning (especially by chloral, opium, and alcohol) and from uræmia, by the indications already described ~~in the section on apoplexy~~.

The distinction from the apoplectiform attacks of cerebral congestion, from those of general paralysis of the insane, and from the so-called "simple apoplexy," in which there is no sign of congestion and after death no lesion of the brain, is more difficult. In all three, local symptoms are absent. In congestion the coma is rarely profound; the loss of consciousness is often imperfect, and is generally brief. The only symptoms are general; there is no local loss of power. There have usually been previous attacks of the same character, transient, leaving no after symptoms. In a first attack, or without a history, the diagnosis may be impossible. In general paralysis of the insane the preceding symptoms are almost always sufficiently pronounced to indicate the nature of the apoplectiform seizure. The attack lasts only a few hours, and the patient rapidly recovers his ordinary state. They are more readily confused with attacks of simple congestion than with hæmorrhage.

The attacks of "simple apoplexy" that occur in the old, mysterious in their nature, may resemble closely the apoplexy of cerebral hæmorrhage, and it is doubtful whether a distinction between the two is possible in practice. The so-called "serous apoplexy" is merely a variety of this form, ~~and is important only from ignorance, and significant chiefly of the unobserved facts that are close at hand~~.

~~If the presence or history of local symptoms makes it certain, or the intensity of the general symptoms renders it in high degree probable, that the attack is due to an organic cerebral lesion, the chief diagnostic question is whether the lesion is hæmorrhage, or softening from vascular occlusion.~~ If the patient is under forty, the presence of heart disease, or, in its absence, a suspicion of syphilis, renders vascular occlusion far more probable than hæmorrhage, provided the apoplectic symptoms are of moderate severity. But these causal indications do not absolutely exclude hæmorrhage, even in early adult life or in childhood, since both these conditions are undoubted causes of aneurism of the larger cerebral arteries. The rupture of an aneurism is probable if, under such circumstances, the apoplexy is intense in degree and the coma rapidly deepens. ^{such} In a case of heart disease, preceding symptoms, headache, &c., increase the probability of aneurism, *rather than of embolism*

In the second half of life, especially after forty-five, when miliary aneurisms become common, neither the presence of heart disease nor a history of syphilis affords the same strong presumption against

hæmorrhage that it does in early life, and their significance is subordinate. Thrombotic softening from atheroma of the arteries becomes common *pari passu* with hæmorrhage, and it is only when the symptoms make it probable that the lesion is vascular occlusion, and not vascular rupture, that the heart disease or syphilis raises a question as to the cause of the occlusion. The older the patient the less weight do these points deserve; because atheroma increases, and syphilitic disease lessens, ~~in frequency~~ ^{in frequency,} with advancing years, while the degenerative valvular disease of the old causes embolism far less frequently than do the endocarditic lesions of the young.

Between thrombotic softening and hæmorrhage the diagnosis is often difficult. The distinction is of great importance, because the treatment suitable to the two conditions is very different. A probable diagnosis can be made only by comparing the several symptoms, cerebral and general, and balancing their indications. Neither age nor sex gives help, except that in extreme old age, ~~over eighty~~, there is a probability in favour of softening rather than hæmorrhage. The conformation and nutrition of the patient are not of much significance. The most important indications are those drawn from the state of the heart and circulation, the presence of conditions favouring the bursting of an artery, especially a pulse of high tension, and a strongly acting hypertrophied heart. The opposite conditions favour the formation of a clot in a degenerated vessel, and therefore thrombotic softening; so also does considerable degeneration. Irregularity of the pulse is opposed to hæmorrhage if combined with feebleness, but is unimportant if there is deep coma and the pulse is strong. ~~Considerable degeneration of the arteries of the limbs is somewhat in favour of softening, as Nothnagel has pointed out.~~ In Bright's disease, especially the granular kidney, there is a slight, but only a slight, probability in favour of hæmorrhage, for atheroma and softening are also frequent consequences. Hæmorrhages in the retina in connection with albuminuric retinitis do not render cerebral hæmorrhage more probable, unless they are large. Visible aneurisms in the retinal arteries probably do constitute strong evidence of cerebral hæmorrhage, but they are very rare. If the attack was apparently induced immediately by strong mental excitement or physical exertion, the fact is in favour of hæmorrhage; if by prolonged grief, or ~~the exhaustion after physical exertion, it is in favour of softening.~~ ^{by more likely.}

Prodromata of some duration in the form of numbness, tingling, or weakness in the side afterwards paralysed are in favour of atheroma and thrombosis, and so also are previous headache or slight attacks of weakness ~~in other parts~~. Local convulsions at the onset of the attack ~~are also in favour of softening, because they occur chiefly in cortical disease, where softening is more common than hæmorrhage;~~ ^{lesions} a general convulsion suggests hæmorrhage. The significance of the occurrence of loss of consciousness at the onset depends on other associated conditions. As a general rule, loss of consciousness is

conspicuous arterial/

more frequent, greater in degree, longer in duration, and occurs with a smaller lesion, in hæmorrhage than in softening. Its significance depends, therefore, on the probable size of the lesion. If there is reason to think that this is small, loss of consciousness is in favour of hæmorrhage; if it is probably large, the absence of apoplexy is strong evidence of softening. ~~But it is unfortunately very difficult to judge of the extent of the lesion in the early stage.~~ Partial hemiplegia is, ~~however,~~ almost always due to a small lesion, and the occurrence of initial apoplexy in such a case is in favour of hæmorrhage. Deep and prolonged coma is always in favour of hæmorrhage, unless the general indications in favour of thrombosis are very strong and the symptoms bilateral, suggesting thrombosis in both hemispheres. A considerable initial fall of temperature, exceeding 1° , or a considerable rise within a few hours, is in favour of hæmorrhage, provided the symptoms do not indicate obstruction of the basilar, in which there may be an initial fall similar to that of hæmorrhage. Hyperpyrexia, with evidence of a lesion of the pons, may occur in either condition, but there is ~~then~~ seldom sudden apoplexy ^{in the} ~~except in~~ hæmorrhage. The greater the secondary inflammatory disturbance, the more probable is softening, especially if there are also secondary convulsions. In the subsequent chronic stage of hemiplegia, the most important indications are that mobile spasm (athetosis, post-hemiplegic chorea, &c.) and recurring convulsions beginning in the paralysed ^{side} ~~limb~~ are strong evidence that the lesion was softening, and not hæmorrhage. But in this stage the history and the state of the heart and circulation also usually afford trustworthy guidance. By careful attention to these several diagnostic indications, and observing their relative weight, and comparing the direction of their significance, the cases are few in which a diagnosis of adequate probability cannot be made. It will often be found that all the indications point in one direction, and, even if not strong, derive weight from their combination, or that one important indication is decided although the others are equivocal.

The diagnosis of secondary ventricular hæmorrhage rests on the occurrence of a second apoplectic seizure, or on the distinct intensification of primary coma, with the extension to the second side of the muscular relaxation that was at first unilateral. But the significance of this depends on the initial attack having the character of hæmorrhage. When there is thrombosis in an artery of one hemisphere, the formation of another clot in a large artery on the opposite side may be attended with exactly the same symptoms as the rupture of a hæmorrhage into the ventricles; hence the occurrence of such symptoms should not be allowed to influence a diagnosis of thrombosis if the evidence of this was distinct.

The diagnosis of a primary ventricular hæmorrhage from hæmorrhage into the substance of the brain is rarely possible. There is no symptom definitely distinctive from hæmorrhage into the substance of

the brain if the initial symptoms are on one side, or ~~into~~ the meninges if they are bilateral, unless it be, in the latter case, the suddenness and depth of coma.

In meningeal hæmorrhage the diagnostic problem differs according as there is or is not initial sudden apoplexy. If there is, the distinction is chiefly from intra-cerebral hæmorrhage, and this is often impossible from the symptoms alone. If there is no initial loss of consciousness, the symptoms are chiefly headache, delirium, and convulsions, and, developing gradually, may closely resemble those of meningitis. In all cases an important element in diagnosis is the condition under which the disease occurs. In the young, ~~in whom alone the difficulty is likely to occur,~~ meningeal hæmorrhage very seldom occurs except after injury. In the old, in whom spontaneous hæmorrhage may be met with, primary meningitis is very unlikely.

PROGNOSIS.—Two questions always present themselves: first, the risk of death; secondly, the prospect of recovery from the resulting paralysis. The initial danger is proportioned to the intensity and duration of the coma. If it has not begun to lessen at the end of twenty-four hours the probability is against recovery. Most cases die in which there is marked interference with respiration, indicated by its sighing character, Cheyne-Stokes rhythm, or by the accumulation of mucus in the lungs. Bilateral symptoms in the limbs are also of very grave significance, indicating, as they usually do, either hæmorrhage into the ventricles or into the pons. The lower the initial temperature the graver is the immediate prognosis. A considerable rise of temperature within a few hours of the onset is also very serious, and so is the early appearance of albumen ~~or sugar in~~ *the* urine. *hiccough* If the initial symptoms were severe, or the patient is old or feeble, the period of secondary inflammation is also attended with danger, the indications of which are considerable fever, delirium, or a tendency to the formation of sloughs and bedsores. Most cases die in which a slough appears on the buttock before the end of the first week. In ventricular hæmorrhage the prognosis is almost certainly fatal. In the few cases of the primary form that recover, the hæmorrhage is small in amount, and the diagnosis of the condition as a rule impossible. In meningeal hæmorrhage with coma the prognosis is exceedingly grave; but if consciousness is regained, there is a fair prospect of recovery, especially in traumatic cases, even when delirium succeeds. But there may be considerable mental impairment for weeks, or even for months.

The prognosis as regards the paralysis depends on whether it is due to direct destruction of the nerve-elements by a lesion involving the conducting path, or whether it is due to indirect damage by a lesion adjacent to it. It is usually necessary to wait for a diminution in the palsy before an opinion can be formed. The parts in which there is

some return of movement before the end of a month will probably recover useful power. Paralysis that is complete at the end of three months will probably remain considerable in degree for the rest of life, except in those parts in which the other hemisphere can compensate for the loss. Thus some power always returns in the leg, ~~except, often,~~ ^(least) in the flexors of the ankle, and it is usually at last sufficient to enable the patient to stand and walk. In the shoulder, also, there may be a return of power ~~while~~ ^{as} the hand remains paralysed. The development of late rigidity in the hand renders recovery of power unlikely.

Although TREATMENT.—~~Little can be done to prevent the occurrence of cerebral hæmorrhage when miliary aneurisms, its main cause, are once formed. Moreover, the degenerative changes incidental to age, or the result of an inherited tendency, cannot be arrested, and perhaps not even retarded. But when the conditions likely to lead to arterial dilatation can be recognised, something may be done to prevent its occurrence by lessening its main factor, the increased strain on the wall that comes from arterial tension, and cardiac over-action, constant or occasional. A tranquil life, as free as may be from severe exertion of body or excitement of the mind, with nutritious but light food, probably has some tendency to effect these objects, and the intra-arterial pressure can be to some extent influenced by occasional saline aperients and diuretics, and by the avoidance of alcoholic excess and "high living." We cannot recognise the good we may thus effect, which is probably often considerable, if the patient's life is under our control.~~

are beyond treatment,

their occurrence and to diminish the tendency to their rupture by lessening
little alcohol,

but cannot doubt that it is

At first, In the attack itself the aim of treatment must be to secure the conditions that favour the arrest of the bleeding, *i. e.* to reduce the blood-pressure within the arteries, especially that within the cerebral vessels, ~~and, of course, to avoid the conditions likely to increase the pressure.~~ *Perfect* Physical rest is of the utmost importance. The patient should be laid down, but with the shoulders and head well raised. The clothes about the neck should be loosened, and flexion of the neck avoided, ~~so as to prevent the hindrance to the return of blood from the head that compression of the veins would cause.~~ All muscular effort should be forbidden. It cannot be doubted that, in many cases, the amount of hæmorrhage has been greatly increased by ~~the patient walking about after the onset of the attack.~~ *essential* Even passive movement should be as little as possible. *exertion*

Venesection was for long regarded as the most important element in treatment; and ~~many~~ ^{some} authorities are still of the same opinion, although the disrepute into which the lancet has fallen leads to hesitation in its use. Formerly the surgeon did not scruple, in any case of apoplexy, to cut across the temporal artery, to obtain a free flow of blood. No other agent reduces so quickly and so considerably the tension of the blood, and patients occasionally regain consciousness while the blood is flowing. On the other hand, it has been objected

that the loss of blood may weaken the heart's action to a degree that ~~would increase the peril of an extensive hæmorrhage into the brain.~~ It is also said that, in most cases in which venesection is employed, no distinct beneficial influence can be traced. But the absence of immediate evidence of its utility is not a consideration of much weight, since the arrest of the bleeding does not, at the moment, lessen any of the conditions on which the loss of consciousness depends. But if venesection is used, it should be with discrimination.

~~The diagnosis of hæmorrhage should be reasonably certain, since in thrombosis loss of blood will do only harm, by weakening the heart and favouring the extension of the clot. This effect, to be desired in hæmorrhage, is to be dreaded in softening.~~ The indications for venesection are a regular, strongly acting heart, and an incompressible pulse. ~~They are strengthened by distinct hypertrophy of the heart, by full arteries, strongly pulsating carotids, and a turgid face.~~ The contra-indications are softness of the pulse, irregularity and dilation of the heart. If the apoplexy is so deep that the respiration and heart are suffering, it is very doubtful whether venesection can do any good. When it is indicated, the sooner the blood is drawn the better. A large opening should be made into the vein, and ten or twelve ounces allowed to flow as quickly as possible. As a means of arresting hæmorrhage from a branch of the middle cerebral, compression of the common carotid in the neck has been advocated by Spencer and Horsley,* on the ground of the observed effects, in monkeys, of a diminished flow of blood. Their observations certainly justify a trial of the method, but the diagnosis must be clear, since it would certainly favour an extension of the clot in thrombosis.

An important element is free purgation. This has a powerful influence on the cerebral circulation, by filling the capacious vessels of the intestinal canal. It should always be employed, but its slowness renders it an inefficient substitute for bleeding if the symptoms are urgent and venesection is distinctly indicated. Croton oil or calomel is the most convenient agent. With the same object a diuretic should be given.

To promote the contraction of the cerebral vessels, ice may be applied to the head, provided the patient is not collapsed, and a grain of ergotine may be injected under the skin. Mustard plasters to the nape of the neck may be employed for the same purpose, to induce reflex contraction of the arteries, if the patient's condition is one of pallor and collapse. The old practice of applying them to the soles and calves rests on an intelligible basis, since even from these distant parts an effect may be produced on the cerebral vessels, and the dilatation of the vessels of the surface must aid in attracting the blood from the brain.

Alcohol should be avoided unless the patient's collapse and cardiac depression are extreme. If a mild stimulant is indicated, ammonia

and increase its depression from the hæmorrhage. This danger may be disregarded if the venesection is only such as to distinctly lessen the abnormal tension. In absence of immediate improvement, it is no evidence that it is not useful and wise. But the

full

if the patient cannot swallow

Spencer Horsley

* 'Brit. Med. Journ.', 1889, i.

Or ether inhaled subcutaneously
~~may be given.~~ It is very desirable to arrest convulsions and vomiting, because these tend to increase the bleeding. *(by bromide or chloral)*
~~In one case, an injection of thirty grains of chloral into the rectum appeared to stop the convulsions.~~ *(by ice bromide or cocaine)*
~~which survive the onset~~

In all cases, ~~but especially if the coma is prolonged, or the secondary pyrexia considerable,~~ the danger of the occurrence of bedsores must be borne in mind, and should be obviated as far as possible by extreme cleanliness, by a water-bed, and by changing the position of the limbs from time to time, so as to prevent long-continued pressure on the same spot. Hot-water bottles should ~~also~~ be employed with great care, since a degree of warmth that can be borne with impunity in health ~~will~~ ^{may} cause a blister and ~~slough in early hemiplegia.~~

In the after-treatment of these cases, when the disturbance of the onset has passed away, and slowly lessening palsy remains, it is important, with the view of preventing a recurrence, to observe the rules of life described at the commencement of this section.

The treatment of ventricular and meningeal hæmorrhage does not differ from that of the intra-cerebral form, although little can be hoped for from any measures that can be adopted.

INFANTILE MENINGEAL HÆMORRHAGE (CEREBRAL BIRTH PALSY).

Meningeal hæmorrhage is occasionally produced during birth; the ~~accompanying~~ damage to the brain may cause permanent symptoms, weakness of the limbs and of the trunk, inco-ordination, spontaneous movements, convulsions, and mental defect. The cases vary much in degree, and ~~it is only in the more severe forms that all these symptoms are present.~~ *in character*
 In some cases the legs suffer chiefly, and the condition is then termed "congenital spastic paraplegia," ~~a malady that has been already described (vol. i, p. 441).~~ *See*
 When ~~the~~ spontaneous movements constitute the most conspicuous symptom, the state has been described as "congenital chorea."

The relation of these symptoms to ~~injury to the brain during birth~~ *thus cannot be proved*
 was suggested by Dr. Little,* and ~~has been clearly established by the researches of an American physician, Dr. Sarah McNutt.†~~
 The following account ~~of the symptoms~~ is based on an extensive series of consecutive cases that have come under my notice, ~~doubtful ones, and acquired lesions, being excluded.‡~~

The hæmorrhage that injures the brain is almost always due to

* 'Nature and Treatment of Deformities,' 1853; 'Obstetrical Trans.,' 1862.

† 'American Journal of Obstetrics,' January, 1885, and the 'Am. Journ. of Med. Science,' January, 1885. See also a valuable paper by Dr. H. R. Spencer, 'Obstet. Trans.,' vol. xxxiii, and Parrot, 'Clinique des Nouveau-nées.' ~~These cases have also been carefully studied, especially in their later stages, and in conjunction with the acquired cerebral palsies, by Osler ('Cerebral Palsies of Children,' 1889), and by Sachs ('Journ. Nerv. and Ment. Dis.,' 1890; 'New York Med. Journ.,' 1891).~~

‡ Later observations fully confirm the conclusions I stated in a lecture on "Birth Palsies," 'Lancet,' 1888.

*Dr Sarah
McNutt*

special difficulty in parturition. In some cases (about a fifth) the presentation is unnatural, and the head is born last; in such cases intense mechanical congestion accompanies the compression, and the occurrence of hæmorrhage is easy to understand. But the same symptoms follow cases of head presentation, when the labour has been long and difficult, and its immediate effects were shown by the difficulty in getting the child to live, or by distinct symptoms of convulsions, rigidity, immobility of some part, dysphagia, &c., present from birth or coming on within a few days. About half the cases are firstborn, but in almost all the others the labour was specially difficult. In some of the cases the forceps was used, but it is probable that, as a rule, the lesion has been the result not of the use of the instruments, but of the conditions that made them necessary. In rare cases the clumsy use of the instrument may have produced the injury, as in one instance in which a permanent indentation corresponded to the centre for the opposite arm, the part chiefly affected. But it is most important that the fact should be clearly recognised that the result is not usually due to the instrumental aid, because parents are only too ready to expand even a question to an assertion, that may do great and undeserved harm to the practitioner who attended. In one case, a fifth labour, the forceps was not used, although it had been employed in each of the preceding labours, and the children had escaped injury. But I have met with a few cases in which the condition was characteristic, but the labour unusually quick and easy, and even premature. It is conceivable that the rapid birth of a skull softer than normal may cause the same intra-cranial damage as the opposite conditions. The affection seems to be almost as frequent in one sex as in the other, the proportion of girls to boys being as five to six.

In the majority of cases nothing particular is noticed in the condition of the child during the first few days or weeks of life. In a few, however, conspicuous symptoms attract attention. There may be an evident difficulty in swallowing; one arm and leg are not moved; sometimes none of the limbs are moved, but this receives less notice than does one-sided immobility. Another frequent symptom is Convulsion, general or unilateral, and often accompanied by persistent rigidity of the limbs, and with inversion of the thumbs. Rarely the head is bent back. The convulsions usually cease at the end of a week or two after birth; deglutition, if at first impaired, remains so in some degree, and there may be a conspicuous difficulty in supporting the head, which falls to one side long after the age at which infants are usually able to support it. But these symptoms are often disregarded, and very often it is not until the child is four or five months old that a deficiency of movement attracts attention; while in others, suspicion is only excited when the child fails to stand and walk at the age at which it should begin to do so. Sometimes the first thing that is noted is strong adductor spasm in the legs, in

and males

In a few well-made cases,

or there may be

the hæmorrhage

ascribe the condition to the forceps and blame the practitioner without reason. In one fifth of the cases the head was born last, and intense mechanical congestion would result.

Boys suffer rather more frequently than girls, as six to five

is frequent & prolonged difficulty in supporting the head is common & is often associated with difficulty in swallowing

other cases it is the inability to effect ordered movements with the hands. In some instances convulsions are the most prominent symptom, and the difficulty in the use of the limbs is often ascribed to them.

The weakness, spasm, inco-ordination, and spontaneous movements usually increase during the first two years of life, being revealed by the structural and functional development of the nervous system. Their distribution varies, but all four limbs are affected in about two thirds of the cases (~~twenty-two out of thirty-six~~), and the trunk and neck muscles are also conspicuously involved in about one third (~~eleven cases~~). *Sometimes* In a small proportion the limbs of one side only are affected (~~seven cases~~), and in rather fewer (~~six cases~~) the legs alone suffer in considerable degree. ~~Very rarely the symptoms are confined to a single limb.~~

In the arms there is weakness and ^{irregular} ~~some~~ spasm, which varies in character, but is greatest towards the extremity of the limb, and interferes with voluntary movement. The elbow is generally flexed, sometimes extended. The wrist is at times bent at a right angle to the forearm, at other times it is over-extended. The persistent strong flexion may even lead to subluxation. The thumbs are inverted or extended; sometimes the metacarpal bone is extended and the phalanges flexed. The fingers are flexed at the metacarpo-phalangeal joints, and in many cases they are over-extended at the middle joint, which may be subluxated. Often, however, the fingers and wrist are in constant movement, now flexed and now irregularly extended. The movement is generally slow, like the post-hemiplegic athetoid spasm (p. 86); sometimes it is quick, and may bear considerable resemblance to the movements of chorea. In slight cases the amount of spasm is trifling, and its chief effect is the interference with voluntary movement, which always exists in some degree. The fingers are separated and moved irregularly in an attempt to take hold of an object. It is often impossible for the child to pick up an object, although it can grasp firmly but slowly. In a severe case, when the hand has at last closed, it may be very difficult for the child to relax the grasp. The condition of the legs is that described and figured at p. 446, vol. i. Any attempt to move them, and any peripheral impression, excites extensor spasm, which renders the limbs rigid, the heel being drawn up and the toes pointed down. There is commonly some persistent contracture in the calf muscles, but this can almost always be completely overcome by steady pressure on the sole. Often the feet are slightly inverted, and I have once seen such extreme talipes varus that each foot was turned in and fixed at a right angle to the leg. Occasionally there is some persistent contraction of the flexors of the knee. There is often also spasm in the adductors of the thighs, which may be sufficient to ^{bring} ~~carry~~ one leg in front of the other on an attempt to stand or walk. The knee-jerk is excessive when the relaxation of the spasm permits its state to be ascertained;

*and are flexed or extended
fingers vary in posture
& are often*

*Even in slight cases, it may be sufficient to prevent the heels from coming to the ground in walking.
Sometimes greatly.*

a foot-clonus ^{may} ~~can seldom~~ be obtained. Spontaneous movements of the toes and feet ^{are then common} ~~may occasionally~~ be observed, ^{may be} ~~and there are sometimes~~ constantly repeated movements of flexion and extension of the trunk or limbs, especially when the mind has suffered. ~~In very rare cases the arms suffer more than the legs.~~ The limbs that are affected are generally thin, but there is never extreme muscular wasting. ~~Diminished length of limb may be perceptible when the affection is one-sided, but if both sides are involved no difference can be found.~~

may be spontaneous

The muscles of the trunk are sometimes weakened, so that the child has a difficulty in sitting up, and, if allowed to do so, lateral curvature of the spine may be produced. There is never distinct paralysis of the face, but sometimes traces of spasm may be observed ~~about the mouth.~~ ^{may} ~~Occasionally~~ Difficulty in swallowing and defective articulation exist, ~~from paralytic weakness, and sometimes associated with~~ tremulous difficulty in moving the tongue. The defective articulation is due, in many cases, to another frequent symptom, mental defect. This exists in at least two thirds of the cases, from a ~~doubtful amount~~ to complete idiocy, ~~with permanent inability to speak.~~ In most of the cases in which mental power is low, the affection of the limbs is considerable, but the opposite rule does not hold good; the muscular affection may be severe and general, although the mind is nearly or quite normal. The difficulty in swallowing or mental defect ~~or both,~~ often cause dribbling of saliva, ~~and this seems sometimes to go on as a sort of habit.~~

slight degree

I have once seen considerable defect of sight (associated with great mental weakness); the ophthalmoscopic appearances and action of the pupils were normal. Loss of sensation in the skin is never to be found.

The affection of the two sides is seldom quite equal, and the two limbs most affected are always on the same side. Occasionally, as already stated, one side escapes, and, if the right limbs are those involved, the child is left-handed. When the legs seem to be alone affected, slight inco-ordination, or at least awkwardness of movement, may generally be observed in one arm or in both on careful examination.

?

Convulsive attacks occur only in a small proportion of cases, and they are rather more frequent when the affection is one-sided than when it is general. They seldom date from birth; initial convulsions rarely persist. Subsequent fits begin generally during the second half of the first year, sometimes later; they may cease after lasting for a short time, or may recur. The occurrence of fits bears no relation to the severity of the limb symptoms; and, indeed, the latter may be scarcely recognisable when the former are severe. If the limb symptoms are one-sided, so also, as a rule, are the convulsions. ~~In a case in which the legs only were affected, and the leg centre alone found damaged, the fits began in the great toe of one side or the other.*~~

* Ferguson, 'Amer. Journ. of Obstetrics,' 1891.

The subsequent course of these cases is one of slow improvement, which is greatest when age enables the will to be brought to bear on the defect; but it is only when symptoms are slight that they approximately disappear. A peculiar oscillation in walking is apt to be developed, and for a long time the patient may walk on the ^{front} ball of the foot, the toes being over-extended, or upon the toes chiefly, from the talipes equinus. The knees are sometimes imperfectly extended even in walking, and there may be persistent inversion of the feet. These conditions may lessen, but often remain in some degree, so that a peculiarity of gait may persist through life.

When the symptoms described are presented after birth and death occurs
 PATHOLOGY.—~~In many of these cases, as we have seen, convulsions, rigidity, and paralysis are observed immediately after birth. If children die, who present such symptoms, meningeal hæmorrhage is invariably found. The extravasation is sometimes over the convexity of the brain, sometimes at the base. In the former situation it is generally bilateral, and is most considerable over the central region and towards the middle line (Fig. 118); it is found also on the medial aspect of the hemisphere (Fig. 119); this is probably due to the effects of the overlapping of the edges of the sagittal sutures. Sometimes, however, it exists only over the posterior part of the convexity. Where the layer of blood is thicker, the convolutions are much compressed, and are sometimes much injured, the cerebral tissue being broken up and infiltrated with blood. The hæmorrhage at the base is chiefly in the posterior fossa beneath the tentorium, surrounding the pons medulla and cerebellum, and it generally proceeds from a laceration in the cerebellar hemisphere. Dr. McNutt found that the basal hæ-~~
 morrhage occurred in cases of head-presentation, while the most intense extravasation over the cerebral hemispheres was found in

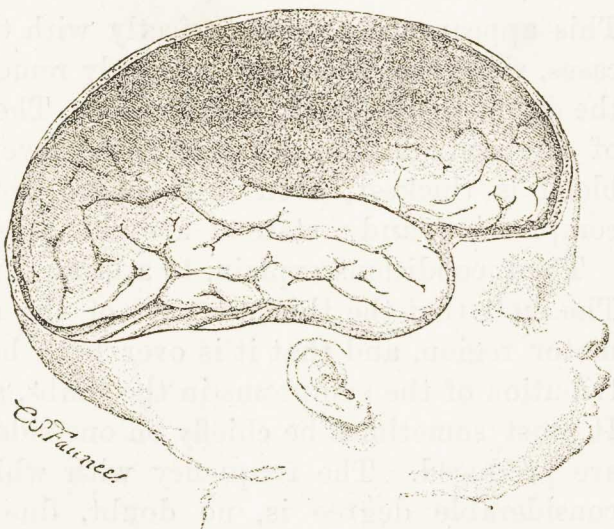


FIG. 118.—Meningeal hæmorrhage during birth. The shading represents the extravasation.

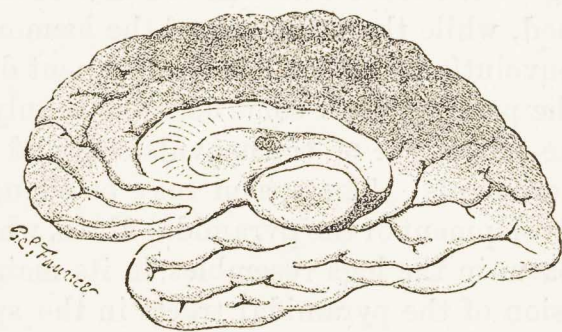


FIG. 119.—Medial aspect of the hemisphere; same case. (After McNutt.)

~~cases of foot-presentation. It is probable, however, that the cortical hæmorrhage also occurs in cases in which the head is born first.~~

~~In older children, who present the symptoms that have been described, the lesion found is atrophy of the convolutions in a certain region of the brain, generally the central convolutions, which may be small and indurated, and lie at the bottom of a considerable depression.~~

*If infancy is survived
by such sufferers, for*

FIG. 120.

FIG. 121.

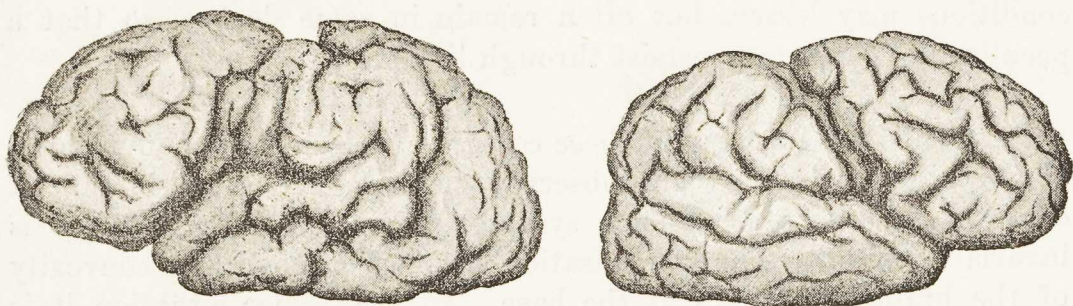


FIG. 120 left, and FIG. 121, right hemispheres of the brain of a child, the subject of bilateral birth-palsy. (After McNutt.)

This appearance agrees perfectly with the condition found in early cases, since the blood will be slowly removed, and only the damage to the cortex of the brain will remain. The long-continued compression of the convolutions hinders their development where the layer of blood is thickest, even if they are not actually damaged by the compression, and a state of atrophic sclerosis results at the spot.

These conditions explain, to a large extent, the symptoms observed. The facts that the thickest part of the extravasation is often at the motor region, and that it is over both hemispheres, explain the ~~distribution of the~~ symptoms in the limbs, and their bilateral character.

When ~~It must sometimes be chiefly on one side, and thus unilateral effects are produced.~~ The frequency with which the legs are involved in considerable degree is, no doubt, due to the position of the leg centres near and in the longitudinal fissure, and it is here that the extravasation is most abundant. ~~Experiment refers to the same~~

and ~~region the centres for the trunk muscles, which are also often weakened, while the extension of the hæmorrhage in front of the central convolutions may explain the frequent difficulty in supporting the head.~~

Structure ~~The persistence of some nerve-cells only slightly damaged may explain the occurrence of the convulsions, and perhaps also the spontaneous movements. The spasm may be connected in part with the arrested development of the pyramidal fibres, which are fewer than normal. The spasm in the legs resembles, in its main features, that which follows a lesion of the pyramidal tracts in the spinal cord, and any differences are no doubt due to the occurrence of the disease during the stage of development. Whether any more complex mechanism may be concerned in producing the inco-ordination, such as the deficient control and balance of other cerebral centres, we cannot say. The extent of the damage to the cortex of the brain is a sufficient explanation of the~~

This also explains the weakness of

mental defect that so often co-exists; and the fact that it is, in rare cases, chiefly over the occipital lobes, enables us to understand the impairment of sight occasionally observed.

We are not at present able to say what symptoms are due to hæmorrhage about the pons and medulla. It seems reasonable to ascribe to this the weakness of the neck and the symptoms in the region of the bulbar nerves. Nevertheless it is possible that these may be due to symmetrical lesions in the cerebral hemispheres in the lower parts of the central convolutions, and the prefrontal lobe adjacent to them, over which the hæmorrhage is apt to extend, as the figures show. It is unlikely that any limb symptoms are due to injury to the anterior pyramids of the medulla, because the symptoms are similar in all cases, and are certainly generally due to the cortical lesion. That they are the same, whether or not there is weakness of the neck, is a fact that suggests the cortical origin of the latter symptom also.

DIAGNOSIS.—In a well-marked case, in which the symptoms are general, the recognition of the disease is easy if its occurrence is known. Some difficulty is presented by cases in which the symptoms are one-sided, or in which the legs only are affected. The most important distinctions from other cerebral diseases are, first, that the condition is stationary and not progressive; and secondly, that there is no history of a distinct onset at ~~some~~ ^a period subsequent to birth. Symptoms of similar character are occasionally caused by a tumour of the brain, but ~~then gradual development and steady increase show the existence of a progressive lesion.~~ ^{an} Similar stationary symptoms may succeed an acute lesion; there is then a clear onset to be traced, often with severe symptoms, and ~~preceded by perfect freedom from any similar affection.~~ In the case of birth-palsy, if the derangement of movement was first noticed at a certain period, it is generally evident that attention had not been ~~previously~~ ^{previously} specially given to the state of the limbs. A distinct difficulty in birth, or an unnatural presentation, can generally be ascertained; and in many cases there is a history of convulsions, &c., immediately after birth, to confirm the diagnosis. When the legs suffer chiefly, the case is often thought to be disease of the spinal cord. ~~As already mentioned, however,~~ ^{As already mentioned, however,} slight derangement of movement in the hands can generally be detected, and it should be remembered that primary chronic disease of the cord in young children is almost unknown. In any case of spastic paraplegia in a child, in whom there is no bone disease, a cerebral cause is always probable.

The greatest diagnostic difficulty is the distinction of the cases from those in which the brain has suffered ~~before birth, either from disease or developmental defect.~~ ^{from pure} In such cases the birth is, as a rule, easy, although it is doubtful whether the distinction is absolute (see above). ~~The limbs do not present the inco-ordination or rigidity, and there~~

- symptoms are slighter & there are

*but it must be remembered
that a rapid premature
birth is sometimes as harmful*

^{while}
~~are~~ no acute symptoms immediately after birth, ~~or indication that the~~
~~child has suffered during the process.~~ On the other hand, there is
 usually grave mental defect, and often a history of a tendency to
~~idiocy or nervous maladies in the family.~~

the known causes of idiocy

PROGNOSIS.—As the damage to the brain is necessarily permanent, so are its effects. But when there is no considerable mental defect, the disorder of movement lessens in later childhood, because the will gradually acquires more power over the movement of the limbs, and is able, to some extent, to counteract the influence of the spasm. In all except the most severe cases the child ultimately acquires ~~some~~ ability to stand and walk, ~~although it may be only at the age of six or eight.~~ As already stated, the gait is often permanently peculiar. The mental condition also slowly improves. In every case, however, the prognosis must depend on the severity of the symptoms; what state the improvement will ultimately leave must depend upon the amount of defect, motor and mental, and upon the signs of improvement that can be discerned. ~~It must be remembered also that, as a rule, the greater the defect, the later does improvement occur.~~

character and

The condition is one on which treatment can have little
 TREATMENT.—~~Drugs have no influence on the morbid state, and but little on the symptoms.~~ Even in the early stage, soon after birth, ~~it is doubtful whether any treatment can influence the process by which the brain is damaged.~~ The meningeal hemorrhage probably occurs during birth only, and there is no evidence of any subsequent increase in the extravasation. No agents can be expected to facilitate the removal of the compressing blood, which is only slowly absorbed after permanent tissue-changes have occurred in the cortex. Doubtless it could be removed by a surgical operation, but this would ~~have~~ ^{be} to be severe, and the diagnosis would require a degree of certainty that could scarcely be obtained until changes had occurred in the brain, from which recovery would be doubtful. The slow diminution in the symptoms cannot be ascribed to the recovery of the damaged structures, since they must have passed into a stationary cicatricial condition long before improvement commences. It must be due rather to the functional education of those parts that are only slightly injured, and to the compensation effected by undamaged parts. Treatment, as far as it is possible, must consist in promoting this by training the motor power that remains, and increasing the use that it can afford, by a process of muscular education, and especially by rhythmical movements of the limbs, fingers, &c. When the inco-ordination is slight, and the child is old enough, a key type-writer is a useful means of muscular and mental education. As soon as the child becomes able to stand, the attempt to walk is, in itself, a valuable training. Instruments sometimes enable it to stand some time before this would otherwise be possible, and they may thus distinctly accelerate improvement. It is useless, however, to employ

mental

instruments until there is enough voluntary power and control to enable the child to stand alone with their aid. Electricity in all forms is useless; its prolonged use, ~~in cases under my care, has shown~~ that it has no influence over either the weakness, the spasm, or the inco-ordination. The treatment of convulsive attacks is similar to that of other forms of epilepsy, but they are less readily influenced by drugs.

*may coincide with the
natural improvement for
which it receives and deserves
Credit*

SOFTENING OF THE BRAIN.

Softening of the brain* is the common result of many morbid processes. The diminution of consistence is due to the breaking up of the nerve elements; the resulting particles become separated by serum, so that a soft pulp replaces the original firm tissue. It is the characteristic effect of inflammation of the brain,—and not long since all forms of softening were thought to be inflammatory. We now know that most cases of acute softening are due to a very different cause, to the arrest of the blood-supply by occlusion of an artery.† In the part supplied by the occluded vessel, the nerve elements, deprived of nutrition, at once lose their functional powers, and ~~quickly undergo degeneration.~~ In many parts of the brain there is ~~not~~ sufficient communication between the terminal arterial twigs to permit of the re-establishment of the circulation, ~~and~~ structural degeneration occurs in the delicate nerve elements with great rapidity; in other parts, anastomoses enable the blood-supply to be re-established, and the function returns after transient arrest. A similar result may follow occlusion of a vein: ~~if the venous communications are insufficient to allow the blood to escape by some other channel, the circulation in the part is at an end, and the nerve elements break up;~~ but the venous anastomoses are generally sufficient to prevent complete necrosis of the part. In rare instances there is softening that cannot be referred to either of these processes, but it is then always chronic, never acute. Thus we may distinguish, as pathological varieties of softening, Acute Softening

receding unless there is

The same is true of

* It should be remembered that there is a considerable difference between the popular and the medical uses of the term "softening of the brain." The general meaning attached to the term is chronic mental failure, and the disease most frequently thus designated is general paralysis of the insane. In medical use the term is restricted to the condition in which the physical state actually exists. Some care must therefore be exercised in using the term, lest it convey an erroneous impression of the nature and course of the disease.

† The relation of ~~senile softening to degeneration of the arteries~~ was first clearly recognised by Rostan in 1823, and soon afterwards by Abercrombie, Carswell, and others. That embolism is a frequent cause of softening was first pointed out by Virchow (in the first volume of his 'Archives'), and in this country by Kirkes, who ~~in an admirable paper~~ ('Medico-Chir. Trans.,' 1852, p. 281) brought our knowledge of the subject within a measurable distance of its present position, and he did so ~~apparently without being acquainted with the earlier paper of Virchow.~~

from (1) inflammation, (2) arterial occlusion, (3) venous occlusion, and Chronic Softening. The first of these varieties ~~will be~~ described in the chapter on inflammation of the brain, and the last among the degenerations. Softening also results from pressure, ~~but is a secondary and unimportant form,~~

as of a growth

SOFTENING FROM ARTERIAL OCCLUSION (NECROTIC SOFTENING; ENCEPHALOMALACIA).

Softening from arterial occlusion is not only far more common than all the other forms of softening put together, but it is one of the most frequent diseases of the brain, perhaps exceeding in frequency cerebral hæmorrhage*, although this, being more often fatal preponderates in hospital necropsies.

GENERAL ETIOLOGY AND PATHOLOGY.—Two pathological processes may cause the occlusion of an artery; a plug ~~coming from a distance~~ may be carried into it by the blood (embolism), or a plug may be formed in it by coagulation (thrombosis). There is an important difference in the pathological relations of these two processes. Embolism is the result of a morbid process elsewhere in the circulation, commonly in the heart. Thrombosis is usually the result of local disease of an artery of the brain, by which either its calibre is narrowed, or its inner surface is changed, so as to cause coagulation; often both these conditions coincide. The chief pathological processes that cause these changes are atheroma and syphilitic disease. A change in the constitution of the blood, rendering it prone to coagulate, and slower movement of the blood, from feebleness of the heart, often aid the effect of the arterial disease in producing thrombosis, and ~~sometimes~~ ^{may very rarely} cause it when the arteries are free from disease.

The distinction between

~~Although the primary processes of thrombosis and embolism are thus distinct, and usually occur under very different conditions, they are often conjoined in a secondary manner. If an artery is obstructed by embolism, the stagnant blood in it may clot, extending the occlusion. If a clot forms in an artery it may be detached (as Laborde has shown), and may obstruct the vessel further on by a process which is actually embolism. But the process is regarded according to its general and primary character, and the distinction between them should always be kept clearly in view.~~

space large al caps
EMBOLISM.—The source of the plug must be somewhere between the lungs and the brain,—in the pulmonary veins, the left side of the heart, the commencement of the aorta, or the large arteries of the neck. It is possible that septic material may pass through the lungs and lodge in the brain, but only capillaries can be obstructed by

* This is the common opinion, and is very likely correct, although the evidence is not conclusive. In the post-mortem room, hæmorrhage is the more frequent lesion, but hæmorrhage is more often quickly fatal than softening is.

is of the first importance, but it may be noted that clot forms on each side of an embolus, & a primary thrombus this

Embolism
Small caps across

~~particles that can pass through the capillaries of the lungs~~, and any local softening of the brain thus produced is usually purulent, a cerebral abscess or minute points of suppuration. When an artery has been plugged in consequence of septicæmic processes in the general system (as uterine phlebitis), there has usually been either a pulmonary abscess or endocarditis to furnish the embolus. In most cases the plug comes from the heart, and its source is either diseased valves, "vegetations" from which are separated and carried along with the blood, or a coagulum in the left auricle, a fragment of ~~the clot being detached~~. It is rather more common in cases of recent endocarditis, rheumatic or "ulcerative," than in chronic disease. A fresh attack of endocarditis, in valves previously diseased, is especially likely to give rise to embolism. The plug may come from either the aortic or mitral valves, but comes from the latter much more often than from the former. The most frequent cause is mitral constriction, in which several conditions probably favour the occurrence; the slow flow through the narrow orifice during diastole permits the aggregation of white corpuscles on the valve, and the quick flow during the auricular systole tends to detach the masses thus formed. Moreover, in this disease there is often great dilatation of the auricle, and clot forms in the auricular appendix; fragments of this clot are apt to be detached, or softening occurs within it, and the particles pass into the blood. The softened clot often contains organisms, and in ulcerative endocarditis, also, the particles detached (~~usually small~~) may carry infective micrococci into the cerebral arteries. Hence in both these conditions there is often plugging of minute vessels, and inflammation in the walls ~~of the vessels and the adjacent tissue~~.

which is

infective

Outside the heart, the plug comes, occasionally, from the lungs. Less rarely it comes from the aorta; when its wall is the seat of atheroma, ~~fragments of degenerated tissue or of clot may be carried away by the blood. I have once known the plug to come from the interior of an aneurism.~~ Echinococcal embolism has been once observed.

Embolism occurs with equal frequency in both sexes. It is met with at all periods of life, but is most frequent between later childhood and middle life. In the old it is less frequent, both absolutely and in comparison with thrombosis. The subjects of it have usually suffered from those diseases that are attended with endocarditis—rheumatic fever, chorea, scarlet fever, in this order of frequency, ~~it may occur in the course of these diseases, or succeed them at any interval of time.~~ At the time of the embolism, or soon afterwards, the signs of organic heart disease are almost always to be recognised. ~~It occurs also, although very rarely, as a result of other acute specific diseases, any one of which is occasionally attended with endocarditis.~~

but also almost any acute specific disease may be attended by endocarditis and this sequel.

An immediate exciting cause is rarely to be traced. ~~I have met with one instance in which the obstruction followed immediately a severe fright, which, by exciting the action of the heart, may have effected the detachment of the plug.~~ Increased coagulability of the

of severe emotion has been followed by this attack

blood sometimes co-operates by causing an increase of deposit on diseased valves, or the formation of a separate clot in a feeble heart. Hence embolism often occurs a week or two after childbirth. This is a point of some practical importance: conditions favouring thrombosis do not necessarily render embolism less probable.

span large distal cap
~~THROMBOSIS.~~ ~~As we have seen,~~ thrombosis may be due to atheroma, to syphilitic disease of the arteries, to disease of their wall by adjacent inflammation, compression, or invasion by new growth, or to a change in the blood.

may
 Atheroma.—The larger arteries at the base of the brain are very common seats of the thickening of the inner coat, called by Virchow "Endarteritis deformans," which, when fattily degenerated, constitutes "atheroma." Opaque yellow thickenings, sometimes calcified, are the result. ~~Only one or two of these may be present, but the~~ change usually involves many of the larger vessels at the base, and extends for a considerable distance along the chief branches. Its distribution may be symmetrical. ~~A similar change may exist in arteries elsewhere, or it may be confined to those of the brain. The exciting cause of this disease is probably the strain to which these arteries are exposed in consequence of their proximity to the heart and their deficient external support. It is not easy to explain their occasional freedom from atheroma when this is abundant elsewhere. The result of the degeneration is to abolish the elasticity of the vessel, to change its calibre, sometimes to lessen it or to close it altogether, sometimes to enlarge it. Alterations in the lining membrane lead to the formation of clot upon it, as on a foreign body. Where the calibre of the vessel is increased the current is retarded, and this also facilitates coagulation. The smaller arteries in the cerebral substance do not suffer in the same degree;*, but the orifices of branches coming off from an atheromatous vessel are often narrowed or closed, although the main trunk is pervious, and thrombosis may be confined to these.~~

Whatever increases strain on the arteries may induce it earlier.
 Atheroma is common after middle life, and increases in frequency with age. ~~It occasionally occurs before forty, chiefly in association with chronic Bright's disease, in which the arterial tension increases the strain on the vessels. Gout also induces early degeneration. Softening from atheroma shows a corresponding relation to age. It increases in frequency as life advances, and the increase continues, if due correction is made for the diminished population, to extreme senility, exhibiting in this respect a contrast to hæmorrhage (see p. 387). The occurrence of atheroma, and therefore of softening, is facilitated by chronic alcoholism. A cachectic state of the system, or some cause (such as prostrating illness or grief) that alters the constitution of the blood, and weakens the action of the heart, often concurs in producing the thrombosis. The gouty diathesis may thus co-operate, and even cause premonitory thrombosis elsewhere.~~

* Fatty degeneration of the cells lining the perivascular sheaths of the minute arteries is very common even in the young, but is without pathological significance.

may also facilitate embolism

and to alter its inner surface, — promoting

the flow through beyond the diminished orifice the lessened flow leads to blood is almost stagnant & readily coagulates on the altered wall, suddenly occluding the branch

Syphilitic disease is a common cause of softening.* It is chiefly a consequence of acquired syphilis, and so occurs ~~in adult life~~. ~~About half the sufferers are between thirty and forty at the time of the attack, one third between twenty and thirty, and most of the remainder between forty and fifty; under twenty and over fifty it is very rare.~~ The period after infection at which it is developed varies much, but in the majority of cases is from ^{two} one to twelve years. I have met with it as early as six months and as late as nineteen years after the primary sore. Fifty cases in which the diagnosis was practically certain, and the period after infection could be definitely ascertained, were distributed as follows:—In a quarter the onset was during the first two years after infection, and the remainder were distributed thus: 2—3, nine cases; 3—5, seven cases; 6—10, nine cases; 11—15, eleven cases; 16—20, four cases. The longest intervals were 18 and 19 years, the shortest 6 months (except a doubtful case at 3 months). No cases in married women are available for comparison because the date of infection cannot be ascertained; many other cases are not available because the individual has had more than one sore. The above twenty-six cases are all that could be employed for this purpose out of about a hundred of which I have notes, in which the diagnosis was reasonably certain. ~~It has been met with in characteristic form as a result of inherited syphilis, between birth and the tenth year.~~

As an isolated change (apart from massive syphilitic growths) it is almost confined to the larger arteries of the base. The vessels most commonly affected are the internal carotid, middle cerebral, vertebral, basilar, and posterior cerebral. One or many vessels may be affected. The distribution of the change is commonly irregular, but sometimes it is symmetrical. The change may proceed to the obliteration of the vessel, but more frequently when the lumen is considerably narrowed a clot forms within it and suddenly arrests the circulation, or forms in branches arising from the affected part. The disease consists in a fibro-nuclear growth in the wall, which causes a nodular projection on the exterior (Fig. 122), and diminishes ~~also~~ the calibre of the vessel. The projections are more nodular, irregular, and less opaque than in atheroma. The structure of the growth resembles that of syphilomata elsewhere. It generally begins by a nuclear proliferation between the inner coat and the elastic lamina, or outside the latter: in the former case the elastic lamina is pushed outwards; it may undergo degenerative softening, and a thrombus

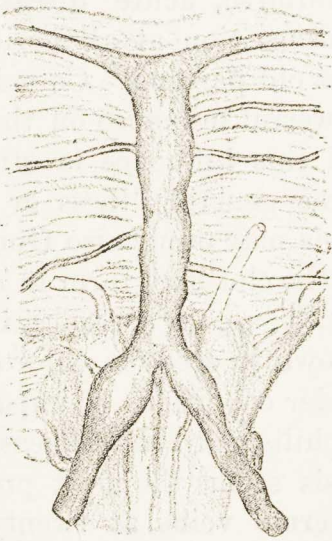


FIG. 122.—Syphilitic disease of the basilar and vertebral arteries; thrombosis of left vertebral (to the right.)

* See also the writer's 'Syphilis and the Nervous System,' 1892, Lect. II.

*One half between 30 & 40
between 20 and 50.9
chiefly in middle life. But
it occurs also from the in-
herited disease, from
early infancy up to the
10th year.*

*and it is almost evenly
distributed throughout
this period.*

may form on the altered inner surface. Antisyphilitic treatment usually induces a fibroid cicatricial change, which ^{causes} the nodular projection to be slight, and the aspect may resemble that of early atheroma. If ~~the~~ ^{may be} elastic and muscular elements are replaced by distensible fibroid tissue, the wall may bulge into an aneurism.

Other forms of arterial disease causing thrombosis are very rare. Occasionally pressure on an artery so narrows it that coagulation occurs. Secondary arteritis may result from inflammation of the membranes, especially from traumatic meningitis. The outer coat of the vessels becomes thickened, and degenerative changes may be set up in the middle and inner coats, and may lead to thrombosis months or years after the primary mischief, from which the patient may have apparently recovered. In the course of tubercular meningitis, a tubercular or inflammatory infiltration of the walls of the arteries has been met with, especially in small vessels, and has produced thrombosis. This occurs chiefly at the convexity, and may precede definite symptoms of inflammation, especially in adults suffering from phthisis. ~~The inhalation of carbonic oxide has caused arterial degeneration and softening, even in young adults.†~~ Lastly, ~~it may be mentioned that~~ ligature of the carotid occasionally, although rarely, causes necrotic softening of the brain. Usually the collateral circulation is sufficiently free to prevent this.

Blood-states; Simple Thrombosis.—In certain conditions of the blood, coagulation occurs with undue readiness. These states accompany childbirth, acute diseases, cancer, gout, tuberculosis, and general malnutrition at all periods of life, but especially in young children and old persons. Coinciding with disease of the arteries, it exerts a powerful influence on the occurrence of thrombosis in them. In the old this influence is often seen; the accident follows, for instance, some general illness or some depressing emotion. Arterial thrombosis may occur also from this cause when there is no arterial disease, or the clot may form in a vein or sinus. This occurs occasionally in adults, as, for instance, in the course of phthisis. It has even been known to occur in chlorosis.‡ Sudden hemiplegia sometimes occurs under conditions of impaired general health in young adults, in whom syphilis and heart disease can both be excluded; spontaneous thrombosis seems the most probable cause, since it has been observed in external veins at twenty-one, twenty-three, and twenty-four years (Angelucci). A sudden cerebral lesion frequently occurs in children, and is probably due in most cases to thrombosis in a vein or artery, but ~~it is occasionally the result of endocarditis and embolism. These cases are separately described.~~

Cardiac Weakness.—The slower the blood-current, the more readily does coagulation occur. Hence feebleness of the heart's action is a

* 'Med. Ophthalmoscopy,' Case 4, 2nd ed., p. 270.

† Poelchen, 'Virchow's Arch.,' Bd. cxii.

‡ Skerrett, Clin. Soc., 1885.

but may lessen the cavity still more, & even close it, by the contraction of the vessel near the point of occlusion.

Many intracranial aneurisms are thus caused

DEGENERATIONS OF THE BRAIN.

CYSTIC DEGENERATION.

due to THE shrunk brain of the old is occasionally riddled with small cavities, from the size of a pea downwards. The change is seen chiefly in the white substance, and is sometimes general, sometimes limited to certain parts. The cavities are the enlarged perivascular spaces, and the alteration seems to represent, in the substance of the brain, the senile atrophy which causes the surface to shrink, the sulci to be widened, and the subarachnoid fluid in them to be increased in quantity. It is also usually associated with a similar enlargement of the ventricles. As Charcot has pointed out, the most frequent seat of this cystic degeneration is a zone of the white substance between the region supplied by the cortical arteries and that of the central vessels, in which the terminal branches of each set are inadequate for the perfect maintenance of nutrition. It is not probable that these cavities, however numerous, give rise to symptoms, any more than does the general shrinkage of the brain, but the arteries within them have but little support, and dilatation and rupture may be facilitated, especially when the cavities exist outside the lenticular nucleus, where cerebral hæmorrhage so often takes place. They often contain débris of nerve-tissue, and hæmatoidin crystals, the remains of some small extravasation. Occasionally, especially in the pons, the cavities are met with at an earlier age, even in young adults, and are then sometimes thought to have a pathological significance which they do not really possess.

CHRONIC PROGRESSIVE SOFTENING.

The old view, that softening ^{is} may occur as a primary process, although wrong as regards the common ^{form} acute softening, seems to be justified by rare cases in which softening occurs without demonstrable association with disease of the arteries, or correspondence with their distribution, and is so gradual in its onset, and slowly progressive in its course, as to make a dependence on the vessels very improbable. Some cases of this kind have been collected by Wernicke,* and a

* 'Lehrbuch der Gehirn-Krankheiten,' Bd. ii, p. 149.

is uncertain

few other instances are to be found in medical literature, old and recent. The affection is almost ^{chiefly} always seated in the white substance of the hemisphere, a feature in which it differs from the acute softening of actual obstruction. But there is no reason to suppose that the grey matter ^{never} invariably escapes. The change consists of simple white softening, which presents, under the microscope, chiefly the products of degeneration. The affected area is often sharply limited from the adjacent normal substance. The extent of the lesion varies much in different cases: subcortical softening has even ^{been found} extended through the whole convexity of one hemisphere. Although the grey matter of the cortex has not been primarily affected in the cases examined, it may suffer secondarily when the softening of the white substance is extensive. The diffuent tissue is then limited externally by the pia mater, ^{also by} and sometimes there is a thin layer of cortical grey matter below this. Chronic softening may also affect the cerebellum, but it there differs from the cerebral softening, since the cortex suffers more than the white substance. I have seen the cortex of almost the whole of both hemispheres of the cerebellum uniformly softened and atrophied, while the white substance was unaffected. In this case there were also several areas of softening in the cerebral hemispheres, in the white substance and in the grey matter of the cerebral ganglia.

The recorded cases are still too few to afford much information regarding the causes of chronic progressive softening. All that can be said is that it occurs in both sexes, and is a disease of the second half of life, most frequent between sixty and eighty. The chief symptoms are hemiplegia—motor and sensory—of gradual onset. Weakness commences in one part and gradually spreads to the whole of one side, but without considerable sensory loss. Slight irritative symptoms occur, numbness, tingling, and formication in the limbs, and in some cases these have been the seat of paroxysmal pain ^{in them}. Rigidity may accompany the commencing paralysis, but ceases when it is fully developed. Convulsions are very rare. There may be occasional attacks of giddiness, but apoplectic seizures do not occur in the chronic form. The intellect may be little affected, or the patient may become dull and apathetic, and, towards the last, comatose. But the cerebral symptoms ^{after attaining} have usually reached a certain degree of severity, and then ^{in some cases} remained stationary. Death ultimately occurs from bedsores, pneumonia, or some intercurrent senile malady, perhaps favoured by the nervous state. The duration of recorded cases has varied between a few months and two years, but the onset cannot easily be fixed.

Although it is not probable that the condition is directly related to arterial disease, the latter is present in most senile cases, and may have an indirect influence in determining the lesion, its common seat, the white substance, ^{being} nourished partly by the central, partly by the cortical vessels, with the effect just mentioned in the account of cystic degeneration. Subacute and acute cases are met with, which present every gradation of course between the chronic form, the less

become
cerebral

have usually
some secondary effect

being in region
of

There is little is known of their precise pathology

INSULAR SCLEROSIS.

543

rapid forms of softening from arterial occlusion, and focal inflammatory softening. It is especially important to keep in view the cases, in the old, in which the symptoms suggest a focal chronic inflammation (mentioned on p. 469). Pathological facts are urgently needed regarding all these forms, and the subject is commended to the notice of those under whose observation, in workhouses for instance, the diseases of the old so often come.

It will be seen that the chronic symptoms bear a considerable resemblance to those of cerebral tumour. A difference is in the fact that headache is more often absent than present, and very rarely reaches the degree of intensity that is so characteristic of tumour. Optic neuritis has also been absent in the cases in which the eyes have been examined. The greatest diagnostic difficulty is presented by cases of very chronic course. If symptoms develop in a subacute manner, reach a considerable degree of intensity in six weeks or two months, and then remain stationary, the distinction from tumour is more easy, because a rapidly growing new formation rarely undergoes early arrest. The age of the patient is also of some significance; in the first half of life, tumour is more probable than softening. The occurrence of general convulsions makes softening unlikely, but ~~cortical~~ fits may be produced by softening situated at the motor region. *that which begins locally*

Little can be said regarding the treatment of these cases. Our knowledge of them depends only on the cases in which the nature of the disease has been ascertained by a post-mortem examination, and in these it does not appear that any treatment employed has had an appreciable influence on the course of the disease. The most suitable measures will probably be those recommended for the chronic stage of acute softening—rest, nutritious food, with general and cardiac tonics.

DISSEMINATED OR INSULAR SCLEROSIS.

The disease thus named consists in the formation of scattered islets of sclerosis in various parts of the brain and spinal cord, sometimes also in the cranial nerves. Our knowledge of the affection is recent. The lesion was, indeed, long since figured by Carswell and Cruveilhier, and it was described, thirty years ago, by Frerichs, Rindfleisch, &c., but the malady was not generally recognised until re-investigated by Vulpian, and by Charcot and his pupils, and described by Charcot in his widely circulated 'Lectures.' It is called by the French "Sclérose en Plaques Disseminées," by the Germans "Multiple Sclerosis," and, in this country, the French designation has been modified into "Disseminated Sclerosis;" but it is also called "Insular Sclerosis," a convenient term, proposed by the late Dr. Moxon.

*remember that some
which are only lately
only lately to be furnished
by those under whom*

primary

The pathological relations of the morbid process are still, to a large extent, obscure, and much remains to be done in elucidating its clinical history ~~and pathological relations~~. As regards the symptoms of the typical form, little can be added to the description given by Charcot, but there is much uncertainty regarding the untypical varieties, which are ~~by no means~~ ^{means} rare.

Three forms of the disease have been distinguished—cerebral, spinal, and cerebro-spinal, according to the situation of the islets of sclerosis, and the corresponding difference in the symptoms they produce. The cerebro-spinal variety must be regarded as the typical form, which gives rise to the complete series of symptoms; but the disease may be at first limited to ~~one part~~, and may afterwards become general; or it may remain limited, so far as symptoms are concerned, to the part first affected. Hence the nature of many cases is misunderstood, and other diseases have often been described as examples of this affection; its name having become familiar, it is apt to be suspected where it does not exist.

ETIOLOGY.—Insular sclerosis occurs in both sexes with nearly the same frequency. It is met with at almost all periods of life between childhood and old age, but especially in the first ~~half~~ ^{third} of adult life. The majority of the cases commence between twenty and thirty-five. In one verified case it commenced at seven, and its symptoms have been observed still earlier; in another case it began as late as sixty, but in old age it is extremely rare. Direct heredity, or the affection of two brothers or sisters, has been noted in a few instances, but is quite exceptional; indirect inheritance is more common, shown by a family history of insanity, epilepsy, or of some form of chronic paralysis. Often, however, no hereditary predisposition can be traced. In about half the cases the disease develops without any recognisable cause, near or distant. Among the influences to which it has been immediately ascribed, the most frequent are exposure to cold, mental distress, over-exertion, some acute disease, an actual lesion of the central nervous system of traumatic or other nature, or some specific febrile disease. But any single exciting influence is to be traced ~~only in a very small proportion of the cases, and the most noteworthy fact regarding them is the occurrence of the disease at a time of life when general influences have, as a rule, no late sequelæ.~~ ^{seldom} Among the acute diseases that have preceded it, typhoid fever and smallpox are the most frequent; it has also been known to follow diphtheria and erysipelas. The preceding morbid processes in the nervous system have been apparently acute or subacute inflammation (myelitis, &c.), primary, or secondary to some injury. Their symptoms have subsided, or have left some stationary residual effects; after an interval of a year or more, the indications of insular sclerosis have slowly developed. I have known it to begin during pregnancy, remain stationary until the next pregnancy, and then become progressive.

Preceding syphilis can scarcely ever be traced, so that it is doubtful whether the very rare cases in which symptoms of this disorder in syphilitic subjects have been removed by treatment were really due to sclerosis. (See "Pathology")

PATHOLOGICAL ANATOMY.—The islets of sclerosis that characterise the disease are always irregular in situation, scattered, as it were at random, through the central nervous system, subject only to the most general laws of distribution, and corresponding in locality in no two cases. The only law which can be traced is that they generally commence in the white substance. In the brain they seldom invade the grey matter of the cortex, and never seem to begin in it, however numerous they may be in the white substance of the centrum ovale, a favourite seat. They are irregular in shape, and vary in size from that of a pea (or less) to that of a walnut. In colour they are reddish grey, a little darker than the normal grey substance of the cortex, and rather more translucent. Their section is usually on the level of that of the adjacent cerebral tissue; occasionally it is a little depressed, but on

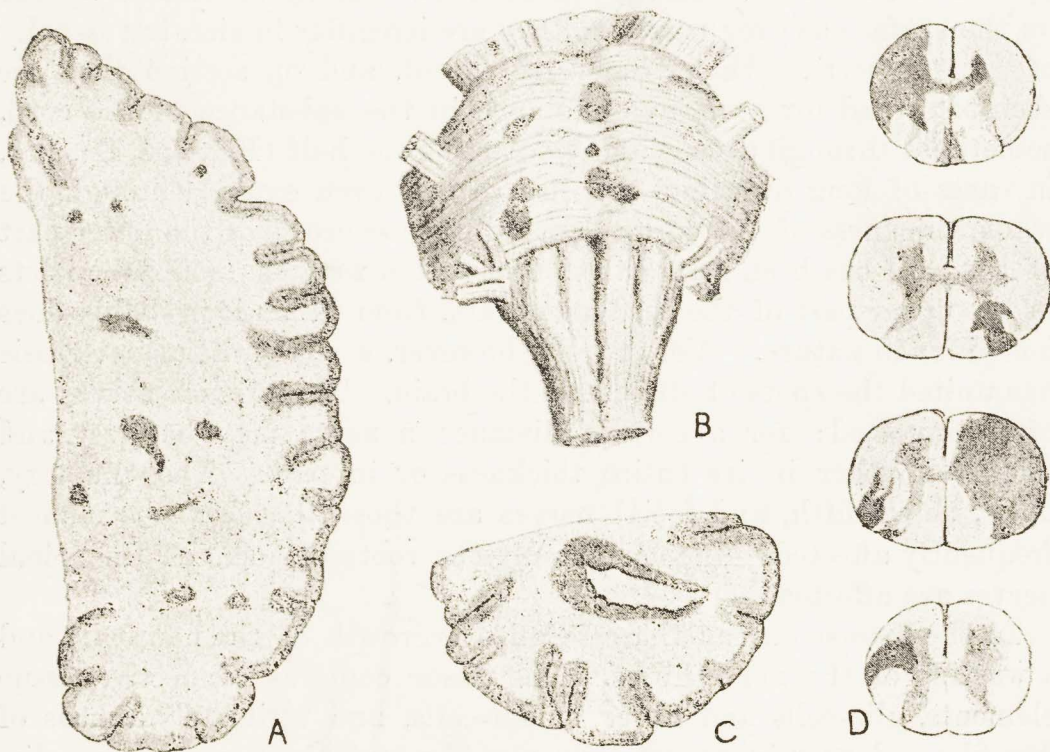


FIG. 144.—Insular sclerosis.

- A. Centrum ovale of right hemisphere.
- B. Pons and medulla.
- C. Peri-ependymal sclerosis around the descending cornu of the lateral ventricle. (A, B, and C are from an original case.)
- D. Sections of the spinal cord to show the varying distribution of the sclerosis at different regions. (After Leyden.)

* Some facts bearing on this question will be considered in the section on "Pathology."

the surface (as in the crus or pons or cord) they ^{may be} ~~are sometimes~~ slightly prominent. The sclerosis does not, however, increase perceptibly the volume of the part in which it occurs. To the naked eye the islets are sharply defined; ~~the transition to the normal tissue is sudden.~~ The consistence of the sclerosis is usually a little firmer than that of the brain-tissue, and ~~they are~~ ^{it is} sometimes distinctly firm; occasionally, in ^{more &} ~~old cases, some may be almost cartilaginous in their firmness.~~ Owing to the position of the islets in the cerebral hemisphere there is usually no indication of their presence on the surface of the brain, even when a large number are found in its interior. Although the cortex is seldom involved, the central ganglia are frequently invaded, and the sclerosis is sometimes abundant beneath the lining membrane of the lateral ventricle (Fig. 144, C) and in the corpus callosum. In the crus and pons the islets are often numerous; some may be conspicuous on the surface from their contrast with the white fibres which ~~constitute the superficial layer of these parts, while others are scattered through the interior of the pons or medulla.~~ They are met with also in the cerebellum, but are usually few, and ~~here also are~~ confined to the white substance. In the spinal cord the areas of disease appear on the surface as grey ^{spots} ~~patches~~; they are irregular in size, but usually of greater vertical than transverse extent, and on section they are seen to extend for a variable distance in the substance of the cord, sometimes through a considerable part of one half (Fig. 144, D), and, in cases of long duration, ~~the disease may even~~ extend through the whole thickness of the cord. Such diffuse sclerosis of the lower part of the cord has been met with in association with characteristic islets in the upper part of the cord and brain, from which it probably does not differ in nature. Very rarely, however, a slight diffuse sclerosis has united the spots of disease in the brain. The cranial nerves are often involved; for a certain distance a nerve may be grey and sclerosed either in its entire thickness or in part. The olfactory, optic, third, fifth, and facial nerves ~~are those that~~ have been most frequently affected. ^{involved} ~~Less commonly the roots of some of the spinal nerves are affected.~~

In the diseased areas there is an overgrowth of the neuroglia and a wasting of the nerve-fibres. The tissue contains connective-tissue elements, glia-cells and fibres, spider-cells, and ultimately a mass of fine fibres, denser in aspect than the fibres of the normal neuroglia. Among these tissue-elements there may be fatty granules and globules, the remains of the degenerated nerve-elements. The nerve-fibres that pass through are changed in various degrees. The chief alteration is a narrowing of the white substance, which becomes reduced (as seen in section) to a narrow, ~~scarcely appreciable~~ ring around the axis-cylinder, and even this may cease to be recognisable. The axis-cylinders persist longest, but ultimately they may disappear.* The

* The persistence of the axis-cylinders was first pointed out by Charcot, and has been since confirmed by Schultze, Déjérine, Babinski, and others.

the longer the duration

walls of the vessels become thickened, and the increase of the adjacent connective tissue may be very conspicuous. Should the sclerosis involve the grey substance, the nerve-cells become atrophied, and the intervening structures suffer changes like those of the white substance, but, from its nature, less readily perceived. Over affected areas that reach the surface there may be a slight thickening of the pia mater, but seldom sufficient to be recognised with the naked eye.

If an area of sclerosis occupies part of a tract of fibres ~~that is liable to undergo~~ secondary degeneration, such degeneration may or may not be found beyond the part diseased, ~~but it is usually present in some degree. The difference seems to depend on the condition of the axis-cylinders; as long as the change is confined to the white substance of the nerve-fibres there is no secondary degeneration, but as soon as~~ the axis-cylinders suffer, degeneration occurs. ^{always} ~~This is true of the optic~~

When the optic nerve undergoes general atrophy, the same tendency is seen for the axis-cylinders to persist and the medullary sheath to suffer. The connective tissue is much increased between the bundles and in the inner nerve-sheath, and the nuclei between the nerve-fibres are multiplied (Uhthoff). An islet of sclerosis in this or any nerve presents the conditions already described.

SYMPTOMS.—The wide variations in the position of the islets of sclerosis entails a corresponding difference in the symptoms that are produced. Loss of muscular power in the limbs, variously distributed, and often at first partial, is the most ^{common} constant manifestation of the disease. Since the morbid process usually involves the pons and spinal cord as well as the brain, some islets can hardly fail to occupy the motor tract. The deficiency of power in the arms is very variable in degree, but the characteristic feature of the weakness is its association with a peculiar symptom, jerky irregularity of movement, wide in range, and ~~ceasing on rest~~. It may exist also in the neck muscles, and in the legs; also in the latter it is often masked by the ~~general unsteadiness it causes~~, but ^{more the} it is sometimes conspicuous, even when the arms are little affected; ataxy may exist alone, but simple spasmodic paraplegia is still more common. Some of the cranial nerves are often also paralysed. Among other symptoms, nystagmus is exceedingly common, and so is a peculiar disturbance of articulation. These symptoms vary in occurrence and degree, in different cases, to an extent which ^{so as to make} makes it impossible to give a description of the disease alike accurate and comprehensive, and also gives rise to numerous difficulties in its diagnosis.

The variation in the character of the symptoms extends to their order and to their mode of onset. In some cases the peculiar inco-ordination in the hands, or in one alone, is the first indication of the disease. In other and more frequent cases the legs first become weak, or the patient is unsteady in standing or walking. Dull pains in the limbs may trouble the patient for a year or two before the

degree in which the fibres are ~~to~~ involved;

when as well as of other nerves

Slight at first and

cause on their movement in the recumbent position. There is no spontaneous tremor

much the variations also rare,

precede

motor symptom, or
 onset, or a sense of "numbness" in the hands and feet, occasionally with actual lessened sensibility, may occur and pass away, or even recur more than once. Less commonly some cranial nerve-palsy occurs at the onset, or speech is observed to be peculiar, but in the majority of cases the limb symptoms precede the others. It is important to remember that some of the preceding symptoms may be due to a morbid process connected with the cause of the disease.

most
 Of the individual symptoms, the jerky irregularity or tremor is the most peculiar and characteristic. It is, as just observed, manifested most frequently in the arms. At rest the limb is still, and presents no spontaneous tremor, but as soon as a movement is attempted, in the endeavour, for instance, to take hold of some object, the arm is thrown about in a wildly irregular manner, and moves in sudden quick jerks, now in one direction, now in another, until at last, by a great effort, and sometimes with a sudden dash, the patient succeeds in grasping the object. If he tries to raise a glass of water, the movements are so wild that the contents of the glass are probably spilled. In an attempt to write, the violent movements produce only irregular strokes upon the paper as the hand dashes to and fro, and often the paper is pierced or even the pen is broken in the attempt. In some instances the movements are slighter in range and more regular, even resembling simple tremor, although its relation to the malady may be proved by the presence of the typical form in some other part. It is said that in rare instances the movements have been known to continue during rest, but there is some doubt as to the nature of such exceptional cases. The movements are, as a rule, increased by effort, by motion, and by attention. The more the patient tries to overcome them, the more violent they are, but the variations in degree are not great, and the disorder of movement is unceasing.*

when
 The muscles of the neck often share the disorder of movement, and there is irregular oscillation of the head, and sometimes of the trunk, whenever the patient sits up. The derangement may even be greater in the muscles that support the head than elsewhere, and it may cause so much unsteadiness that the head has to be kept still by the hand to permit the patient to drink. When laid at rest the head at once becomes still. Although the affection of the movement of the legs usually causes unsteadiness, which may render walking impossible without assistance, and even standing difficult, they sometimes present the irregularity of movement above described. It will be seen that the distribution of this symptom varies much, and with it the general aspect of the cases. For instance, while it is usually most marked in

* Hence they are called by the Germans "Intentions-Zittern," a term which cannot be rendered in English without inaccuracy, on account of the meaning of "intention." "Movement-tremor" embraces all varieties, but fails to indicate the irregularity of the typical form. Perhaps "atactic tremor" would be, on the whole, the best term, because there is no ataxy without voluntary movement.

transient sensory or motor symptoms may occur during a year or so before permanent affection

termed intention-tremor, because by the Germans, because limited to voluntary movement. It is seen especially

but simple weakness with spasm is more common

the arms, one unquestionable case presented in these only the fine irregular tremor on sustained movement, great inco-ordination in the legs, and extreme jerky disorder in the muscles supporting the head. Rarely the limb symptoms exist on one side, alone or chiefly; one limb is then more affected than the other, as in a case in which the left leg was merely weak, while the left arm presented typical irregularity of movement. Such instances might be multiplied indefinitely; they are explained by the great variety in the lesions. *Seal of the process*

Some weakness usually accompanies this irregular inco-ordination; it may be at first slight, and indeed, at the very onset, it may be absent. On the other hand, weakness may exist alone, especially in the legs. Once established, the loss of power gradually increases.

The jerky irregularity has been ascribed by Charcot to an irregular resistance to conduction in the nerve-fibres at the sclerosed spot; it is supposed that the changes in the white substance, or in the axis-cylinder, interfere with conduction along the latter, and as this change is unequal in degree, even in adjacent fibres, there is a various degree of retardation of the voluntary influence on the muscles, and hence the irregularity of the contractions. This hypothesis derives some support from the fact that a precisely similar symptom is present in some cases of tumour of the brain, in which, for instance, a tubercular growth compresses the motor path, as in the case of tubercle in the crus shown at Fig. 142, p. 509. I have met with this symptom also in a case of tubercle of the pons. To some (Erb, &c.) the theory has appeared inadequate, and they incline to the alternative explanation, that the disorder is due to the occurrence of islets of sclerosis in some particular situation in which they derange the co-ordinating function. The symptom *has been* said to coincide with sclerosis of the pons, and to be absent in cases in which the lesion is *limited to* the spinal cord.

Simple weakness in the legs is common, and is often the earliest symptom of the disease, and may present a paroxysmal increase on exertion, so that the patient falls occasionally, and can only resume walking after a rest. When it is considerable it is usually accompanied by an excess of myotatic irritability (increased knee-jerk, foot-clonus, &c.), which goes on to spasm, so that a state of spastic paraplegia is established. This is indicative, as in other cases, of degeneration of the lower parts of the pyramidal tracts. Such degeneration in this disease is secondary, the consequence of the development of areas of sclerosis somewhere in the dorsal region of the cord, involving the pyramidal tracts. In the rare cases in which the lesion is limited to the spinal cord, such spastic paraplegia may be the only symptom of its presence, and many cases have been mistaken for primary lateral sclerosis. The ataxy of the legs, present in other cases, varies in its character, and probably also in its mechanism. There may be, as we have seen, jerky inco-ordination with which the myotatic irritability is usually increased. In other cases there is simple unsteadiness, sometimes like cerebellar, sometimes like spinal inco-ordination. It is

The variations in the distribution of this motor symptom is due great. This is

hence

Secondary

motor fibres

but the opposite error is far more common

Rarely probable that this is due to the interruption of the upward paths from the muscles, ~~as explained in vol. I.~~ In a rare group of cases the state of the legs is identical with that in true tabes, ~~perhaps in consequence of a patch of sclerosis in the posterior columns in the lumbar region; but it is also possible that the condition is true tabes, a system-degeneration in one part of the central nervous system accompanying the insular sclerosis in another part.~~ This association will be considered in the section on pathology.

The excess of the muscle-reflex action in the legs may increase to rigidity, the common ~~variable~~ ^{even to} extensor spasm, or, in rare cases, flexor contracture, so that the legs are permanently drawn up. ~~Occasionally a single group of muscles may become rigid and contracted in the arms as well as in the legs.~~ Muscular wasting is rare, but ~~local~~ ^{local} irregular atrophy may result from the invasion of the grey matter of the cord by some islet of sclerosis.

Cutaneous sensibility is normal in the early stage of the disease, but it may afterwards be impaired. ~~The loss varies much in its distribution according to the situation of the sclerosis on which it depends.~~ *in various parts* Rarely there is hemianæsthesia from the development of an islet in the sensory path within the brain. The loss is more often partial, involving only a small area on a limb, and it may or may not correspond to the motor palsy. When there is ~~diffuse sclerosis of the cord there may be extensive loss of sensibility on the legs.~~ *large an extensive area of slighter impairment* The condition of sensation is intelligible when we remember how slowly the axis-cylinders of the nerve-fibres suffer, and that a greater amount of damage is required to abolish sensory conduction than suffices to cause motor palsy. Irritative symptoms are occasionally present; a sense of vague numbness or ~~tombac~~ ^{tombac} in the hands, sometimes also in the feet, may be a ~~very early symptom; and may even pass away before other disturbance is developed, although it occasionally persists.~~ *be transient or* Dull aching pains in the limbs are not uncommon, especially in the legs, sometimes in the back, ~~and may occur from time to time throughout the course of the disease.~~ Sharp momentary pains also occur, but not often. A girdle pain may ~~occur~~ ^{be present} early or late, persistent or transient, complete or partial, but also is not frequent. These sensations are chiefly complained of when there is other evidence of the presence of sclerotic islets in the spinal cord, ~~but even then they may be absent, and the comfort of the patient may be undisturbed.~~

Eye symptoms are frequent ~~and important.~~ The optic nerve may suffer in various ways. There may be impairment of sight in one eye or in both, often with an irregular contraction in the field of vision, and without at first any visible changes in the optic nerve to account for it. This condition depends upon the development of an islet of sclerosis in one or both optic nerves, or in the optic chiasma. ~~It may progress to almost complete blindness of one eye. After a time, secondary atrophy supervenes, and can be seen with the ophthalmoscope.~~ *becomes visible to develop*

moscope. Occasionally the atrophy is preceded by slight neuritis, when the sclerosis is near the eyeball. ~~In some cases, again, there is a primary atrophy of the optic nerves, exactly like that which occurs in tabes, with a similar failure of sight, proportioned to the visible alteration in the nerve.~~ Both eyes usually suffer, but one is often affected earlier and more than the other. ~~This complication is, according to my own experience, rather more frequent than the form first described, and is of much pathological interest.~~

The internal ocular muscles are seldom involved, but each form of internal palsy is sometimes met with, loss of accommodation rather more frequently than loss of the light-reflex ~~alone~~. The pupils are sometimes unequal, or become so on accommodation.

On the other hand, the external muscles often present some abnormality. The most frequent disturbance is nystagmus, which is ~~an~~ ^{abnormal} exceedingly common symptom of the disease, although we cannot explain it. Both forms occur, spontaneous and on movement, but the former is uncommon; the latter is that usually present, and it does not differ in features from that which is met with in other central degenerations. It is generally on lateral or upward, seldom on downward ^{movement} rotation. The quick displacement of the globes is, as a rule, in the direction of the voluntary movement, but curious variations are sometimes met with, especially in association with loss of power. ~~One patient could only move the eyes to the left halfway to the canthus for a moment, but nystagmus only occurred on movement to the right. In another case, on movement to the right there was nystagmus with the quick movement to the right; on movement upwards there was rotatory nystagmus, and vertical oscillation when the eyes were directed to the left. In another the quick motion was to the right, whether the movement of the eyes was towards the right or the left side. These variations quite baffle our power of explanation.~~

The external palsies are more often of a conjugate movement than of a single nerve or muscle, ~~except the external recti (sixth nerves), which may be weakened on one or both sides; one superior rectus is occasionally feeble.~~ But convergence, and the movement of both eyes towards one side, are the motions most often impaired, and they are occasionally lost. In exceptional cases, however, there is considerable and increasing palsy of one of the ocular nerves, as the third, due to the development of sclerosis in the nerve-trunk. Transient diplopia is also occasionally complained of, even in the early stage of the disease.

Other cranial nerves ^{rarely} may also become paralysed by the same mechanism and with the same ^{rarity}, especially the facial, fifth, or hypoglossal nerves. In the affection of the first of these, all parts of the face suffer, and the degenerative reaction is well marked. ~~Very rarely there is clonic spasm in the face, which may even be the first symptom. In the fifth nerve, either one or both parts may be~~

More frequent is
and aspect of
the malady is less
progressive than in tabes
& seldom causes com-
plete blindness

disturbance
very

movement is

It may be greater on the
side of least palsy
& may be rotatory
or vertical, even on
lateral movement,
variations which we
cannot explain

Sometimes

especially the sixth,
less frequently the third,
probably from

has been surely observed

involved. The implication of the sensory part causes much less pain than is produced by most other lesions of this nerve-trunk. The sclerosis that thus damages these nerves may be either in the nerve itself, ~~near the surface of the brain, or in the pons or medulla, at the central course of the fibres.~~ Difficulty in swallowing may be present ~~when the pons or medulla is the seat of the morbid process.~~

Another ~~very~~ frequent symptom—so common as to be of diagnostic importance—is a peculiar change in articulation. Syllables (less frequently words) are unduly separated and ~~unduly~~ accentuated, in what has been termed “staccato,” or “syllabic,” or “scanning” utterance, often with a tendency to elide the ends of words. Some patients, indeed, present only elisive or slurring articulation. ~~The syllabic form suggests a difficulty in uttering the syllables, and there may be an actual consciousness of this difficulty.~~ The movement of the tongue is sometimes jerky and irregular, but the face is seldom involved; ~~and the peculiar affection of speech is independent of any visible disorder of the movement of the tongue.~~ ^{and} It has been met with when the pons and medulla were free from sclerosis; ~~and its source is thus obscure.~~

Slight mental change is common; considerable alteration is very rare. There may be failure of memory, but especially frequent is an undue complacency and contentment, which, under the increasing disability, is distinctly unnatural. ~~There is not often any mistaken idea of improvement, but the patient may seize on and exaggerate any distinct diminution in the symptoms.~~ This morbid complacency is rather more frequent in women ~~than in men, and in young adults than in later life.~~ ^{characteristic} Hysterical and emotional disturbance may also occur, even in men. ~~Very rarely mental disturbance is considerable enough to amount to chronic insanity.~~

Among other symptoms that are frequently met with are headache and giddiness; both may occur early in the disease, and are usually paroxysmal. The vertigo is especially pronounced, but its frequency has been over-estimated, and after a time it ceases to occur. The attacks may be very severe and come on apart from any apparent cause. As in other forms of severe giddiness, it may be accompanied by vomiting, and this may also occur in paroxysms without vertigo, often in association with headache.

Other paroxysmal disturbances are sometimes met with, especially of apoplectiform character, similar to those that occur in general paralysis of the insane. There may be attacks of simple coma, with pyrexia (even to 104°), or there may be unilateral convulsions, or the attack may be followed by transient hemiplegic weakness or rigidity. Occasionally the patient dies in such a coma, and, post mortem (as in general paralysis), no acute lesion can be found. Such paroxysmal disturbance, or some more trifling functional derangement, may precede the definite symptoms of the disease.

The general nutrition of the sufferer is often unimpaired, and some-

and articulation

*who may also present
symptoms of*

times there is an actual increase in the amount of subcutaneous fat. Trophic disturbances have been observed in rare cases,—local œdema, altered secretion of sweat, changes in the growth of hair or nails, chronic arthritis, and herpetic eruptions.

COURSE AND DURATION.—In some cases the course of the malady is uniformly progressive from the first to the last. More frequently periods of apparent arrest alternate with periods of progress, and its full development may be followed by arrest, actual or approximate, for years. The duration varies between two and fifteen years, the average being between three and six years. ~~The most rapid case I have known was fatal a year and three quarters from the onset.~~ The most common cause of early death is interference with the functions of the medulla oblongata, especially impairment of deglutition with its various consequences. In a few cases death occurs from failure of respiration. In the cases of longer duration the patients die from weakness, bedsores, ~~kidney disease, &c., and in not a few instances death results, not from any effect of the disease, but from some intercurrent malady.~~

PATHOLOGY.—Compared with many of the other forms of sclerosis in the central nervous system, this variety is sharply distinguished by its irregular distribution. The islets are, as it were, scattered at random ~~in seat~~, and they thus present a marked contrast to the "system-degenerations," in which ~~a tract of sclerosis extends through the whole of a group of fibres having a certain function, and is sharply limited to these, or is restricted to a certain area of grey matter, or even to related groups of cells.~~ The fact suggests that, whereas systemic sclerosis certainly commences by a wasting of the nerve-elements, to which the overgrowth of connective tissue is secondary (see vol. i, 2nd ed., p. 425), in disseminated sclerosis the primary change ~~must be~~ ^{is} in the interstitial tissue, and the damage to the nerve-fibres must be secondary. The conclusion is supported by all we know of the pathology of the disease, by its aspect and its histological features, by the gradual way in which the white substance of the fibres wastes before the overgrowth of tissue about them, and by the absence of secondary degenerations, unless the local process has a certain seat and a certain degree. The immediate ~~local~~ ^{local} determining causes of the overgrowth ~~cannot yet be even guessed at.~~ It has been conjectured that it proceeds from the walls of the vessels, but whenever there is an overgrowth of interstitial tissue there is always a special increase adjacent to the vessels, and these may take a secondary share in the process even when there is no primary vascular change (see vol. i, p. 426).

The mystery of the origin of insular sclerosis, of ^{the} mechanism of its causation, and the nature of its pathological relations are increased by the fact that, different from system-degeneration as it

but cases may run a more rapid course a year & a half or even less

but the most common cause is

degeneration, with secondary sclerosis, involves

we do not know

is in the main, the two are not separated at all points. System-degenerations may be combined with insular sclerosis. The most common instance of this is the occurrence of primary atrophy of the optic nerves in disseminated sclerosis. ~~This is a true system-degeneration of the nerves, similar to that which occurs in tabes. It is possible that in some cases there is also a system-sclerosis of the posterior columns of the cord in the lumbar region, so that tabes and insular sclerosis are combined, while some of the combined ocular palsies point to the same conclusion, and so also does the occurrence of the disease in families with a pure neurotic disposition, such as insanity indicates.~~ Hence, ^{them to} different as the two processes are in their immediate pathology, they ~~may~~ possess some common relations in ~~nature or causation~~, but our knowledge is still insufficient to enable us to make any definite statement on the subject. We may, however, bear in mind two very different sets of facts. One is the relation between the nutrition of the interstitial neuroglia and that of the nerve-fibres, especially of the medullary sheath, conspicuous in all processes of secondary degeneration, alike in general relations and in minute features. ~~It is a converse relation, the decay of the one involving the growth of the other (see vol. i, 2nd ed., p. 396), but is not therefore to be passed as without significance. It seems alike influential whichever element in the process leads the way, as insular sclerosis illustrates. The primary atrophy of the optic fibres causes interstitial growth, and the scattered islets of the latter cause the nerve-fibres to waste.~~

degeneration of the lumbar cord like that of simple tabes

whereby the neuroglia overgrows as the nerve fibres waste, by ~~an~~ nutritional energy which becomes independent

A second fact which ~~deserves to be borne in mind, although its significance is obscure, is that the only lesion which resembles insular sclerosis in general distribution, and sometimes in characters, is one that is apparently quite distant from it in etiology, the disseminated inflammation in the central nervous system produced by syphilis.~~ ^{which seems to be a result of} This is always distinguished by its subacute onset, and usually by the caseation that it presents; ~~but when such caseation is absent the aspect of the lesions is scarcely distinguishable from that of sclerosis.~~ The illustration of the latter (Fig. 144) may be compared with the caseating foci shown in Fig. 125. I have seen a disseminated myelitis, without caseation, the aspect of which could scarcely have been distinguished from insular sclerosis; but it developed in a few weeks (immediately after treatment had removed a syphilitic tumour of the brain), and caused death in three months. These specific inflammations usually resist treatment, and are therefore probably due to a "toxine" ~~produced by the organisms rather than to the latter—a fact which brings them into line with some systemic degenerations, such as tabes, although the latter begins in, and the former outside, the nerve-elements.~~ We have here processes that bear a close analogy to those of insular sclerosis. ~~But it is no more than an analogy, because the general history of this disease make it impossible for us to ascribe it to the acquired influence of either organisms or their~~

secondary

insular sclerosis

and the form is interstitial

products, unless in occasional and exceptional instances (as after an acute disease) which perhaps approach in nature, such forms of inflammation as that met with in the subjects of syphilis. The common absence of distinct causation, the early adult age of the subjects, and the frequent neuropathic heredity, all point to some inherent condition, some disseminated congenital tendency of the nutrition of the neuroglia. The system-degenerations of the same period of life (such as that of "hereditary ataxy") we have seen (vol. i.) must be ascribed to an inherent defect in the vital endurance of the tissues, and such degenerations may occur with insular sclerosis. Moreover, in connection with the apparent scattered tendency to an abnormal growth of the neuroglia we may remember, first, the facts already mentioned connecting the nutritional tendency of it and of the nerve-elements, which may involve similar liabilities to disease; secondly, the features of "gliomatosis" of the cord, and its active growth in early adult life; and thirdly, the modern theories (perhaps also facts) which ascribe tumours of many kinds to congenital tissue-elements, whose long quiescence may end without perceptible cause. In the present state of our knowledge it seems impossible to do more than offer such suggestions regarding the pathology of this disease. It may be noted that they include its chief, even different, features, its many varieties, and gradation into doubtful forms, and do not exclude the possible action, in rare cases, of acquired influences, such as those connected with acute specific maladies and general causes. Lastly, we can thus understand our inability to cure, and too often even to arrest the disease.

The chief general facts seem to be that insular sclerosis is a neuroglial disease, and that, apart from secondary degenerations, system-disease of the nerve-elements sometimes forms part of it. It is essentially a process of morbid growth, but may perhaps sometimes commence as an interstitial inflammation. Most morbid growths, however they may originate, when once set up, have a vital independence, and often a distinctly aggressive character, which carries them far beyond the range of their primary cause.

DIAGNOSIS.—The great characteristic of the disease is the combination of the jerky inco-ordination and progressive weakness in the limbs with nystagmus. In such cases the diagnosis is easy. The inco-ordination differs from that of locomotor ataxy in the wildly irregular movements or tremors which, as it were, break out on any attempt at voluntary motion. The only disease in which this symptom exists in precisely the same form as in disseminated sclerosis is cerebral tumour (see p. 510). But these cases are distinguished by other indications of a tumour of the brain, and real difficulty in diagnosis can only arise from ignorance of the occurrence of the symptom in such cases.

The diagnosis from paralysis agitans depends chiefly on the sym-

We may conjecture that from the occurrence of disseminated inflammation or to some acute disease that the ~~common~~ causal conditions of insular sclerosis may point to morbid tendency of

We may remember also the tendency of residual embryonal tissue to overgrow in early adult life (see vol. i.) as giving some plausibility to the theory that the seeds of sclerosis may start from some points of developmental origin. We can conceive that toxic agents may excite active processes in these & we

altered speed
and

the local limitation of the symptoms to as to one arm, and to

ptoms which exist in that disease in addition to the tremor, on the regularity of the latter, even when considerable in range, ~~and its locality, as will be specially described.~~ The distinction does not depend on the relation of slight tremor to movement, or on the affection or freedom of the muscles of the neck. Hesitation between the two should be rare.

In some cases of general paralysis of the insane, the twitching contractions of the muscles ~~occur in the limbs with unusual intensity,~~ and may disorder voluntary movement in a manner very similar to that seen in disseminated sclerosis. The chief difference is that the spasm is most intense in the lips and tongue in general paralysis, and that the speech presents a drawling character, with hesitating phonation, while considerable mental change ~~is almost invariable.~~ A similar difficulty, at least in aspect, is presented by some cases of infantile brain disease, involving the motor cortex of both hemispheres. There may be jerky inco-ordination of the arms, identical in aspect with that of sclerosis, inco-ordination of the legs, and even a staccato articulation. Birth palsy (meningeal hæmorrhage) or thrombosis in the veins and (incomplete) in the longitudinal sinus, are the probable causes. The latter may have been the lesion in a case in which the symptom dated from a brain illness at fifteen months and ultimately resembled, very closely, those of insular sclerosis. But however close the resemblance, the history of such cases at once distinguishes them. The difficulty of the diagnosis may seem great at the first glance, but the onset, or development and careful observation of the several symptoms always shows the nature of the case.

The spastic paraplegia that results from insular sclerosis in the dorsal region of the spinal cord, and secondary degeneration below is not distinguishable from that met with in other chronic lesions of the same part, including primary lateral sclerosis. But characteristic symptoms elsewhere usually appear. The distinction of this disseminated dorsal myelitis, described in the section on Pathology, depends on the same considerations, but especially on the difference in the course of the two diseases, the subacute development of the paralysis in the disseminated inflammation.

Cases have been described by Westphal,* Leyden, and Langer,† in which symptoms closely resembling those of insular sclerosis have existed during life without any post-mortem lesion to which they could be ascribed. There is much doubt as to the exact nature of such cases, but they are too rare to be of much practical importance.

~~Perhaps the greatest real difficulty is presented by the cases in which there is only fine tremor in the hands with some weakness in the legs. The fact is certain that fine tremor may be combined with the other symptoms (see p. 548), and that which may co-exist with more typical disorder may also exist alone. A careful and repeated~~

* 'Archiv f. Psych.,' xiv, pp. 87 and 767.

† 'Wien. med. Presse,' 1884, p. 698.

The fact that early
paralysis agitans, tremor
may occur only on
movement is no real
difficulty ~~because~~ of
the fact is known

Especially birth palsy
(meningeal hæmorrhage)

but without these
it is seldom an indication
of this disease. It must
be remembered that

search should be made for any unequivocal symptoms, and these will often be found in the movements of the eyes. These cases are most common in girls, even when genuine, and they furnish the most frequent examples of the inevitable diagnostic problem, the distinction from hysteria. The difficulty is occasioned by the fact that an irregular tremor, occurring on movement only, is sometimes due to hysteria. While resembling that of sclerosis in irregularity, it is much smaller in range, and it is always accompanied by a tardiness of muscular effort, if the patient is desired to exert force, and there is the contraction of antagonistic muscles, described in the section on hysteria, which is of so much diagnostic importance. The variations in the symptoms, and the other indications described in the chapter on "Hysteria," seldom leave real room for a mistake. ^{But} an error is especially easy when actual symptoms of hysteria preceded the onset of the disease, or occurred during its course. But hysterical tremor ^{a mistake} may generate a hopeless prognosis, and distress equally causeless and severe. The other error is the common one, and can only be avoided by the custom, which should be a habit, of ignoring any symptoms of hysteria until after a thorough search for those of organic nature, referred to above. Nystagmus may be safely accepted as evidence of more than hysteria, and this is one of the many maladies in which the symptom is of great diagnostic value.*

PROGNOSIS.—The prognosis of insular sclerosis is exceedingly grave. It depends chiefly on the observed tendency of the disease. Yet the exceptions met with in most maladies are not unknown even in this disease, and should prevent a prognosis absolutely bad. One patient, who seemed unlikely to live many months, had improved at the end of a year and a half so as to be able to walk several miles. In cases in which there are periods of apparent arrest, the duration of life is likely to be longer than in those in which the progress of the symptoms is uniform. In the latter it is not probable that life will be prolonged for more than two years from the time at which the symptoms are sufficiently distinct to permit the diagnosis to be made with confidence. In the former the patient may live for several years, but in all cases much depends on the amount of care that can be given. The symptoms of greatest gravity are those of damage to the nerves of the medulla oblongata, especially difficulty in swallowing. The earlier such symptoms come on, the shorter is likely to be the duration of life.

TREATMENT.—Insular sclerosis is a malady for which, as its history shows, little can be done. The primary overgrowth of the interstitial

* Nystagmus has been said to occur in hysteria, but the assertion is of very doubtful truth. In an extensive investigation into the distinction between these two diseases, Sougues could not discern it in any case of hysteria ('Nouv. Icon. Salpêtrière,' 1892).

tissue resists therapeutical influences more than does the primary degeneration of the nerve-elements. But it is probable that its causes vary, and, if so, some forms may prove more amenable than the majority. The treatment for the system-degenerations is that which most deserves a trial in the insular form, especially the nervine tonics, such as arsenic, ^{nitrate of silver}, quinine, &c. The hypodermic injection of arsenic has been recommended by Eulenburg, but the evidence that this method of administration has a special influence is insufficient.

Occasionally these drugs seem to retard the progress of the disease, but an arrest of its progress is seldom lasting. Solanine, in doses of $\frac{2}{3}$ gr., has been recommended by Grasset as lessening the tremor. Hydropathic treatment, and the use of electricity in various forms, have been advocated in Germany, but their influence is exceedingly doubtful. In the case of a peripheral palsy, such as that of the face, due to sclerosis of the nerve, the question presents itself whether the muscles should be subjected to electrical treatment. As a rule it is not worth while to give the patient the pain and inconvenience of the applications. The disease is so often progressive, that no recovery of the nerve can be anticipated; hence electrical treatment, however prolonged, can have no influence in modifying the condition, and there is not the same reason for applying it to the muscles that exists in the case of lesions from which the nerve-fibres may recover. It is of course desirable to maintain the patient's general health in as good condition as possible, and to avoid all depressing influences. In the case of women, pregnancy is especially undesirable; the malady is apt to make more rapid progress during the later period of pregnancy, and sometimes after delivery. When more is known of the causes and essential pathology of the disease in different cases, more rational methods may brighten the therapeutic prospect,

Hyoscyne, in doses of $\frac{1}{250}$ gr. has some influence on the finer tremor.

at present gloomy

DIFFUSE SCLEROSIS.

If the connective-tissue elements are increased through a considerable area in any part of the brain, the sclerosis is said to be "diffuse," in distinction from the small islets of the disseminated or insular sclerosis just described. Diffuse induration may involve either the white substance or the cortex, part or the whole of one hemisphere or of both, the cerebrum or the cerebellum. Sclerosis may also be secondary to some other lesion, such as softening, the evidence of which (as a cavity adjacent) is still conspicuous; but such forms are not here considered. Two varieties must, however, be distinguished as fundamentally distinct,—those in which there is, and those in which there is not, diminished bulk of the affected part. In the latter, as a rule, the change may involve the white substance only or the grey matter, or both, and the induration is primary; in the former the cortex is chiefly affected, sometimes with a secondary implication of the deeper structures. In these cases the diminu-

connective tissue change

bulk there is
 tion in ~~size~~ constitutes what is called "atrophy of the brain," general or partial, and it is often very difficult to say in which class the lesion should be placed, and by which term it should be named, "sclerosis" or "atrophy." But it is certain that, in most cases of partial disease, *as a rule* and in many in which the affection is general, unilateral or bilateral, the ~~influence~~ ^{pressure on} causing atrophy is primary and the induration is secondary, the result of the wasting of the nerve-elements which entails the overgrowth of the interstitial tissue. Such sclerosis is ~~very~~ common as a secondary change. Sclerotic induration, for instance, follows the ~~damage~~ ^{pressure on} to the cortex produced by the meningeal hæmorrhage that occurs during birth (see p. 413). It is a necessary consequence of interstitial inflammation, however produced, and so is found in the cortex, when this is diseased in cases of acquired infantile hemiplegia, whether the primary process is venous thrombosis or acute inflammation. If any ~~influence~~ ^{pressure on} causes a general arrest of the growth and development of the cortex of the brain, it induces this combination of atrophy and sclerosis, the cortex being the part on which such influences seem chiefly to act. This state may result from a concussion of the brain in childhood, from an actual focal lesion, or from the effect of severe and repeated epileptic fits. But in all such conditions we must regard the sclerosis as secondary and not primary, the first effect being produced on the nerve-elements; and the failure of their nutrition or development entails an overgrowth of the neuroglia, which ultimately becomes firmer than in some other conditions.

In the cases that may be regarded as primary sclerosis of the brain there is little or no diminution in the size of the part, and no indication of an affection of the nerve-elements to which the increase of connective tissue is secondary. They differ, however, so much in their features, causation, and symptoms, that it is difficult to classify them into definite forms until many more ~~carefully observed facts~~ ^{facts} have been collected. In one group of cases, met with almost exclusively in syphilitic children, there is induration of the grey cortex, usually through considerable areas, sometimes confined to a single convolution, rarely in small nodules.* The tint of the grey matter is but little altered, but the change in its consistence is very distinct to the touch. When this affection is of long duration there may be a trifling diminution in the bulk of the part. ~~There may be slight thickening of the superjacent pia mater, but there is no~~ *the pia mater may be thickened but there is no* arterial disease distinctly related to the affected region. Very rarely the central grey matter has been the seat of such diffuse sclerosis. The microscope shows an increase in the neuroglia, and wasting of the nerve-cells. ~~A similar change is seen in the sclerosis that attends cortical atrophy.~~ The interstitial tissue has been found to present the aspect of a finely granular network, and the axis-cylinders of the included nerve-fibres have had a peculiar irregular outline.†

* It is well described by Bury and Barlow, Tuke's 'Dict. Psych. Med.,' ii, 1261.

† Schmaus, 'Virchow's Archiv,' Bd. cxiii.

The white substance is not often the seat of sclerosis in congenital syphilis, except in very severe and extensive cases; and cortical sclerosis seems to be equally rare in the acquired disease of adults. But I have seen, in this affection, ^{observed in syphilis} a diffuse induration of extensive tracts of the centrum ovale of each hemisphere, ~~causing increased consistence, readily felt, but~~ with little change in aspect. ^{by preliminary} A diffuse, almost universal sclerosis has been observed in a subject of chronic alcoholism, associated with induration of the kidneys and other parts.* Lastly, areas of diffuse sclerosis have been found, in rare cases, between and connecting the islets of the disseminated form, † a condition that is perfectly consistent with the pathological suggestions ~~made on a preceding page regarding this variety.~~

regions of

already given

The symptoms of diffuse sclerosis have varied much in different cases, so much indeed that its existence can only be guessed at—except, perhaps, in some cases of congenital syphilis. In all cases of cortical sclerosis in childhood, mental defect exists in proportion to the extent ^{or} and, when limited, to the seat, of the lesion. Every grade of defect, down to profound idiocy, is observed. When the central region ^{cortex} is affected, motor symptoms become conspicuous, loss of power on one side or on both, convulsions, and especially contractures of the limbs and spontaneous movements. In adults, the symptoms have been, on the whole, similar, but have presented great variations. Hemiplegia, contracture, attacks of clonic spasm or tremor, headache, giddiness, vomiting, paroxysmal loss of consciousness, and slowness of speech, have been met with, ~~some frequently.~~ Among the latter, the attacks of ~~spontaneous movement have been thought to be the most characteristic~~ (Strümpell).

Much more must be ascertained regarding the affection before any prognostic data can be obtained, or rules for treatment laid down, beyond those indicated by its apparent causes, and such as are suited for any chronic cerebral affection.

MILIARY SCLEROSIS.

The term “miliary sclerosis” was first applied by Drs. Batty Tuke and Rutherford to a lesion found by them in the white substance of an atrophied cerebellar hemisphere in an insane patient. The lesion was chiefly microscopic, and was only discovered after the brain had been hardened. It consisted of minute spots of degeneration, in which there was little evidence of a process deserving to be called “sclerosis.” The term has been since used by others to designate another microscopic lesion, which was at first thought to be really pathological, but has since been abundantly proved to be of artificial origin—an

* Strümpell, ‘Arch. f. Psych.’ 1879.

† See Westphal, ‘Arch. f. Psych.’ viii; and Kelp, ‘Deut. Arch. f. kl. Med.’ x, 1872. Cases have also been recorded by Schule, Jolly, and F. Schulze.

"artefact," as such changes have been termed. The most common appearance is that in minute areas the tissue remains unstained by reagents, and presents a striking contrast to the well-stained vicinity. On close inspection, however, the tissue-elements can be traced in the unstained area; and it is evident that the difference between these spots and the adjacent tissue is due to the absence of staining. This appearance is chiefly seen when spirit has been employed in hardening. The central organs ought never to be placed in spirit, at any rate, after the first twenty-four hours. This is only one of many artificial changes produced by spirit, which are often very puzzling to the observer.

It is questionable whether the term "miliary" ought to be applied to a purely microscopic lesion; it is more accurately used to designate changes resembling in size "miliary" tubercle. ^{such} But that there is a form of sclerosis of the brain which is truly "miliary" is evident from one well-marked example that has come under my notice. In this case, throughout both hemispheres of the brain, and in all parts of them, the cortex contained minute reddish-grey spots at the junction of the grey and white substance. In tint they were a little darker than the darkest grey substance, and their aspect was exactly like that of the morbid tissue in insular sclerosis, and equally distinct in the recent state. In size they varied from that of a mustard seed, or a little larger, to the smallest point visible; large areas, one eighth of an inch in length, were formed here and there by the coalescence of the smaller spots. Their size and distribution are shown in Fig. 145, A. None could be seen in the white substance of the hemisphere, but they were found in the central ganglia, especially in the outer part of the lenticular nucleus (B) and in the claustrum (C). None were found elsewhere in the brain, except two beneath the corpora quadrigemina. Under the microscope the centre of the large areas consisted of a delicate fibrous tissue with abundant spider-cells and small round nucleated cells. In the fibrous tissue were small cavities, especially at the periphery of the larger spots and throughout the smaller ones, so that the tissue had a somewhat sponge-like aspect. No connection could be traced between ~~most of these areas and vessels, but in places~~ ^{except they} a similar change, of spongy aspect, could be seen along the side of a vessel, sometimes extending for a considerable distance. Thus there was a wasting of the nerve-elements, and an increase in the connective tissue in the affected areas.

I have not met with the record of any quite similar case, but an analogous lesion has been observed in a few cases of general paralysis of the insane, invisible, however, in the recent condition.*

In my case there was no mental derangement. The patient was a man aged fifty, and the first symptom was general weakness of the limbs, accompanied with some rigidity and a few left-sided convulsive

* See Grief, 'Archiv f. Psych.,' xiv, p. 287; and Simon, ib., Bd. ii.

attacks, beginning in the muscles of the shoulder. Then the speech became "mumbling" and unintelligible; coma set in, and after a few days of unconsciousness he died. He had also some cardiac

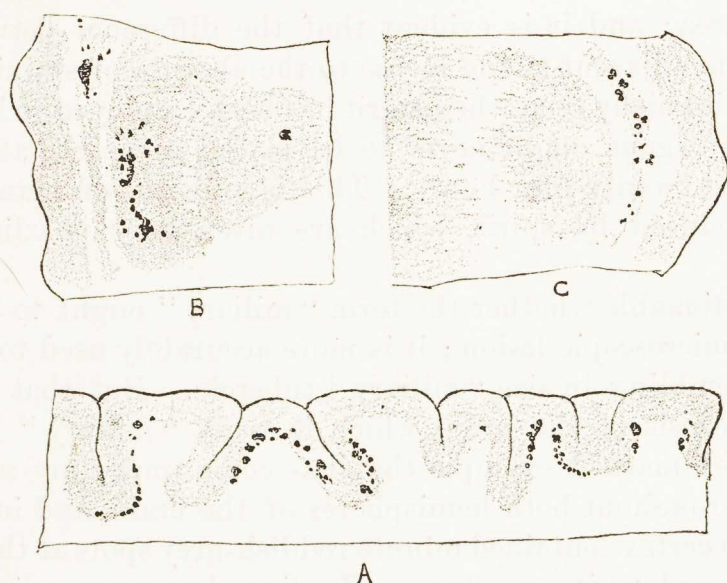


FIG. 145.—Miliary sclerosis of the brain. Sections, A, of the cortex, B, of the left, and C, of the right lenticular nucleus and claustrum. The black dots represent the size and distribution of the minute foci of sclerosis.

weakness, enlargement of the liver and spleen, and general anasarca without albuminuria. The duration of the nervous symptoms was about ten weeks. No cause for the disease could be traced, but he had had constitutional syphilis many years before.*

ASSOCIATED PALSY OF THE BULBAR NERVES.

It has been mentioned that, among the cranial nerves, we may recognise two special groups, subserving associated functions and liable to associated disease. Each group is motor: one comprehends ~~the nerves of the eyeball muscles~~, the associated palsy of which has been described at p. 194; the other includes the nerves to the complex series of muscles of the upper part and orifice of the respiratory and alimentary passages—the mouth, fauces, pharynx, and larynx. These arise from tracts of grey matter in the lowest part of the "middle brain" and the junction of this with the spinal cord, the "medulla oblongata," or "bulb." The associated palsy of these "bulbar" nerves has now to be considered. The most common form is due to a degenerative process similar to that which occurs in the spinal cord. But, as we have already seen, a similar paralysis may result from a sudden lesion in the same part—sudden or "apoplectic" bulbar paralysis. It is sometimes termed the "acute" form, but there is

the motor nerves of the eye

* Fuller details of this case will be found in the 'Lancet,' 1886, January 23rd, p. 145.

also a rare variety due to acute inflammation of these structures, for which the epithet "acute" is needed. On account of the general resemblance of the symptoms it will be convenient to mention the symptoms of the acute and sudden forms in connection with the common, chronic variety. As we shall see, disease of the nerve may cause a like effect, and so also (although by a very different mechanism) may disease of the two cerebral hemispheres. ~~These forms (especially the latter), from their superficial likeness to bulbar disease, but essential difference from it, have been called "pseudo-bulbar paralysis."~~

The nerves, in the distribution of which the symptoms occur, are the fibres of the facial for the lower part of the face, especially for the orbicularis oris, the hypoglossal for the tongue, the spinal accessory for the larynx, and ~~probably also for the palate, and the glosso-pharyngeal or vagus for the pharynx.~~ It deserves note that the facial nerve occupies, as it were, an intermediate position between the two nerve-groups above mentioned as liable to associated disease; it supplies, on the one hand, the muscle that closes the eyelids, and on the other, that which closes the mouth. ~~But the orbicularis palpebrarum does not seem to be very closely associated with the eyeball muscles, for it does not suffer with them in disease of their nuclei. Its functional association with the eyeball muscles is unimportant; it is only a small part of these relations, and does not apparently include its action in protecting the eyeball.~~ ^{eyeball muscle} But the orbicularis oris and the transverse muscles of the front of the tongue have a functional association closer, perhaps, than any other two muscles in the body equally distinct. Neither can be put in action without the other (see p. 50, note). The orbicularis is always involved in degenerative disease of the hypoglossal nucleus, and escapes in disease of the chief nucleus of the facial, as in a case of acute atrophic ("infantile") paralysis in which the face was involved (see pp. 231 and 240).

The central connection of the chief parts that are affected in the disease is well illustrated by the combination of paralysis of the tongue, palate, and vocal cord that results from disease of the nerve-roots on one side of the medulla (p. 307). In central disease of the medulla the paralysis has the same distribution as in a lesion of the nerve-roots, but it is ~~almost always~~ bilateral, and, in addition, the pharynx is often, and the lips are always, paralysed. These escape in disease of the roots, because the labial nerve-fibres, although they ~~seem to~~ arise from cells in the medulla, ascend to the lower part of the pons, which they leave by the facial trunk. The most characteristic and complete distribution is seen in the cases of chronic degeneration in which the nerve-cells suffer according to their functional ^{character} nature. The chief centres concerned lie, however, so near together in the medulla, that random processes, such as softening or hæmorrhage, may damage them on both sides, and cause palsy of the

- trunk

while essentially different

same distribution as that which results from primary degeneration. Hence all forms are included under the term "bulbar paralysis," but only the chronic cases and the inflammatory form can be regarded as constituting distinct maladies.

CHRONIC BULBAR PARALYSIS.

The chronic bulbar paralysis, the "labio-glosso-pharyngeal" (or "~~laryngeal~~") paralysis, was first described in 1859 by Du Mesnil, and systematically by Duchenne in 1860, while this, like so many of Duchenne's investigations, became widely known chiefly through the lectures of Trousseau. The awkwardness of the descriptive name ~~given by him~~, led Wachsmuth, in 1864, to propose to call the disease "progressive bulbar paralysis," a designation that has come into general use. The dependence of the symptoms on degeneration of the nuclei of the medulla was established especially by the labours of Charcot's pupils and of Leyden.

CAUSES.—The disease occurs chiefly in the second half of life, although it is sometimes met with in middle life, and in a few anomalous instances has developed in childhood, at seven, eight, and ten, and occasionally in several children of the same family.* ~~But~~ ^{Most} of the sufferers have been between fifty and seventy when the disease commenced. Males are rather more liable than females. Direct inheritance is practically limited to rare cases in which the affection forms part of progressive muscular atrophy,† but indirect inheritance, shown by other related neuroses, may often be traced. ~~Moreover, a strange family disposition has been observed in cases occurring in early life.~~ It does not seem to be a consequence of syphilis. Immediate causes can be traced in only a minority of the cases; the most frequent are mental depression and anxiety (sometimes combined with physical exhaustion, ~~as in nursing a relation through a grave and fatal illness~~), exposure to cold, debilitating influences, such as insufficient food, and injury, such as a blow upon the back of the neck. Over-use of the muscles has been supposed to be a rare cause; the symptoms developed rapidly in a man after playing on the clarinet for two whole nights, but the patient ~~at the same time~~ ^{also} had been exposed to cold (Stein). In a considerable proportion of the cases, no cause, immediate or remote, can be traced. ~~It is not unlikely that many of these will ultimately be found to depend on some toxæmic influence.~~ I have known the characteristic form to follow lead-poisoning in advanced life, and

* At seven and eight, two sisters and a brother, the offspring of intemperate cousins, Moty, 'Gaz. des Hôp.,' 1889; at eight, Remak, 'Berl. Ges. de Psych.,' 1890; at eleven, Hoffmann, 'Cent. f. Nervenkr.,' 1889; at twelve, Blumenthal, 'Inaug. Diss.,' Dorpat, 1884. Many supposed cases in early or middle life have been due to tumours.

† As in three cases in one family described by Bernhardt, an instance observed by Berger, and one in old age (mother and daughter) by myself.

also partial bulbar paralysis ^{has been} to be associated with partial nuclear ophthalmoplegia in early adult life as a late sequel to diphtheria.*

The relation to progressive muscular atrophy has been already mentioned. The bulbar disease is indeed part of the spinal affection, the grey matter of the medulla undergoing the same degenerative change as that of the spinal cord. Lastly, a limited acute nuclear inflammation may be followed by slow degenerative extension.†

SYMPTOMS.—The distribution of the symptoms, indicated by the name given to the disease by Duchenne, ~~has been already mentioned.~~ The lips, tongue, throat, and often the larynx, are paralysed on both sides. The symptoms are, so to speak, grouped about the tongue as a centre, and it is in this organ that the earliest weakness is usually manifested. In a few cases the distinct paralytic symptoms have been preceded by some discomfort or pain at the back of the head. The onset of definite symptoms is gradual; the cases in which there is a sudden or even acute onset are not examples of this form. The first symptom is generally a trifling indistinctness of speech, due to imperfect articulation of those sounds in which the tongue is chiefly concerned, the lingual consonants *l*, *r*, *n*, and *t*, and afterwards *s*. The commencing change in articulation is noticed only when the parts are fatigued by use, and the patient may be conscious that fatigue is produced with undue readiness. The early difficulty in articulation is often ascribed to some dental imperfection, or to dentists' procedures undertaken to lessen the imperfection. The tongue at first can still be protruded, although sometimes not so far as normal, and there may be a difficulty in bringing its tip firmly against the roof of the mouth for the articulation of the linguo-palatal consonants, as *t* and *d*, while the continuous sibilant *s* can be perfectly articulated. The lips become weak with or soon after the tongue, and some difficulty in swallowing is often added; the latter may, indeed, be the earliest symptom, or it may only come on after articulation has become much impaired. In consequence of the weakness of the lips the power of whistling is lost, and there develops a difficulty in the pronunciation of the sounds in which the lips are chiefly concerned, *o*, *u*, *p*, *b*, and *m*. The lips are not brought together so perfectly, or separated so promptly, as is necessary for the pronunciation of the labial explosives, and hence *b* and *p* become *m* and *v*. The power habitually exerted is generally less than the patient can exert, if he tries. A word can often be articulated perfectly by a deliberate effort when the habitual articulation is very imperfect. The

* In a case described by Stadthagen, diphtheria at four was followed by the common paralysis of the palate, and this by wider symptoms of bulbar palsy. When the patient came under observation, some years later, there still was paralysis of the lips and palate, accompanied by weakness and contracture in the limbs ('Archiv f. Kinderheilk.,' Bd. v).

† Reinholdt, 'Arch. f. klin. Med.,' 1889.

It has rarely

attempts to remedy it

on a deliberate effort

difficulty in speech is soon increased by the weakness of the palate, which ceases to shut off the nasal cavity. Swallowing gradually becomes difficult; the weakening of the tongue impairs the first part of the act of deglutition, that in which the food is rolled back into the pharynx by the application of the tongue to the roof of the mouth; the paralysis of the palate permits the regurgitation of liquids into the nose, and the weakening of the pharyngeal muscles further interferes with the ^{descent} ~~movement~~ of the food.

As the paralysis increases, closure of the mouth becomes impossible, the lower lip droops, and the muscles of the chin often become feeble. The elevator of the upper lip and zygomatics either escape, or are so much less affected than their labial opponents, that the zygomatic muscles undergo contracture, and a very pronounced naso-labial furrow becomes a characteristic feature of the disease. Saliva dribbles out of the open mouth, and the patient has to keep a handkerchief constantly in his hand.* The tongue can no longer be protruded; only the tip can be projected over the lower teeth. Mastication is difficult, because the tongue can no longer guide and keep the food between the teeth. The palate may hang motionless, or it may still be raised a little by the will. Articulate speech at last becomes impossible, and ~~the only expression remaining to the patient is~~ ^{an air} laryngeal phonation, slightly modulated, and broken into the rhythm of formless syllables, the meaning of which can be hardly more than guessed at. Swallowing becomes more and more difficult. Semi-solids are usually swallowed better than liquids, because there is less risk of their escape into the nose or larynx, and better than solids, particles of which are more apt to enter the larynx from the increasing weakness of the epiglottidean muscles, which fail to reflect the epiglottis during the act of deglutition. The muscles of the vocal cords generally suffer ^{now} ~~now, if not before~~; the voice is low-pitched from the defective approximation of the cords, and the cough is imperfect from the feebleness of closure. ~~At first the glottis may be closed only in one of several attempts to cough.~~ The laryngeal palsy rarely becomes complete, and it is still rarer for the power of abduction to be specially lost, common as abductor palsy is in some other forms of central degeneration. ~~I have, however, seen abduction completely lost on one side when it was preserved on the other.~~ Thus in some cases the pharynx suffers, in others the larynx, but both are usually involved in the later stage. The difference seems to correspond to a difference in the localisation of the affection, according to the functional relation of the parts; in some cases those for articulation and in others those for deglutition being apparently primarily involved. Since all the

* It is often thought that there is an increase in the secretion of saliva, and "salivation" is said to exist, but it is doubtful whether the amount is larger than the normal. A pathological increase has been asserted very strongly by some authors; Schulz supposed that, in one case under his care, the secretion was increased to six times the normal quantity.

*It is doubtful whether
the secretion is really
increased*

muscles concerned and the structures related to them are in great part the same in the two sets of cases, there is considerable general correspondence in the symptoms and lesion.

There is no loss of sensibility in any of the affected parts, and taste is unimpaired, but the reflex action in the throat, so active in health, ultimately fails from the motor changes. The palate or fauces, ~~and even the larynx, can be touched or tickled without exciting the spasm that is normally produced.~~ Sometimes this loss of reflex action precedes the other symptoms of the disease (Krishaber). It increases very much the risk of the entrance of food into the larynx. But reflex action is not invariably lost; Erb twice found it preserved in the palate and pharynx to a late stage of the disease.

Alterations of nutrition are chiefly visible in the tongue, ^{and} its condition varies much in different cases. In some the organ is large and broad throughout, although soft and flabby to the touch. In other cases it is conspicuously wasted, and the surface is deeply furrowed and wrinkled from the loss of the muscular substance (Fig. 146).

In some cases the lips retain their normal size; in others they are conspicuously thin. Electric irritability is seldom much changed; the muscles, even when greatly wasted, may still react to faradism with readiness. Rarely, in rapid cases, there is loss of faradic and preservation of voltaic irritability, and Erb has observed the increased and altered reaction to voltaism, when the faradic excitability is normal, of the mixed (or "middle") form of reaction (see vol. i, p. 26).

Symptoms of motor irritation, such as convulsion or spasm, are absent, but choreiform twitchings may occur on an attempt to speak, chiefly due apparently to the influence of emotion.

Thus there appear to be two forms, one with wasting and loss of the reflex action, the other with no wasting and sometimes with distinct evidence of excess of muscle reflex action, at least so far as the muscles of mastication are concerned. The occurrence of the latter form cannot be doubted even when allowance is made for the concealment of atrophy by fat. It is apparently analogous to the spastic paralysis met with in the limbs. Not only may the "jaw-jerk" be increased in this form, but I have once seen a distinct clonus in the muscles of mastication, obtained by depressing the jaw.

The intellectual faculties, as a rule, are unimpaired. The only alteration that is common is a curious emotional mobility; laughter or tears are evoked by trifling causes, and the immobile face and unmodulated tone may render the laughter strangely grotesque. But the unhappy sufferer is painfully conscious of his state; and his

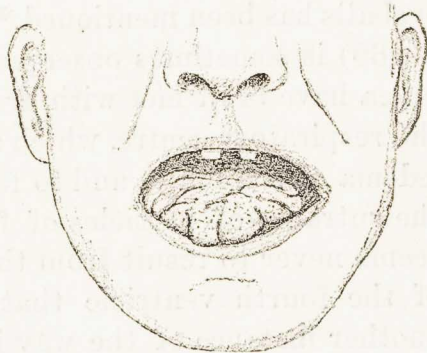


FIG. 146.—Wasting of the tongue, and maximum protrusion, in a case of chronic bulbar paralysis.

except such as distress induces

on

distress is increased by the inability to express it, except by the laryngeal gestures to which speech is reduced. ~~His consciousness of inability often renders him indisposed to make even an attempt to express himself.~~

^{the disease}
~~Progressive bulbar paralysis~~, as already mentioned, is often associated with progressive muscular atrophy in the limbs and trunk, accompanied, it may be, with the symptoms of lateral sclerosis of the cord. Either the bulbar or spinal symptoms may lead the way, and dominate the aspect of the case. Towards the close of a general progressive atrophy the mouth and throat muscles may suffer, or when these are first and considerably affected, wasting may ^{often and occur} ~~show itself in the muscles~~ of the limbs. Less commonly, bulbar paralysis is associated with spastic paralysis of the limbs and slight wasting; lateral sclerosis is then found after death, with only slight changes in the anterior horns.

Even in cases not accompanied by general atrophy, the paralysis may spread beyond the muscles of the lips, tongue, and throat. The rare implication of the muscles of the upper part of the face and of the eyeballs has been mentioned.* Excessive frequency of the pulse (150—160) is sometimes observed towards the close, and attacks of dyspnoea have been met with from the extension of the degeneration to the respiratory centre, which also, towards the close, seems to produce oedema of the lungs, and to facilitate the low pneumonia produced by the entrance of particles of food. ~~It is noteworthy that Glycosuria seems never to result from the degenerative changes beneath the floor of the fourth ventricle that constitute this disease. This may be another instance of the way in which functional relation determines the distribution of the morbid process.~~ Occasionally the muscles of mastication become weak, and the pterygoids, which ^{specialty} ~~move the jaw~~ laterally, are said to suffer first, and often only (Duchenne). Optic nerve atrophy has been noted in very rare cases.†

Among the indirect effects of the ~~disease~~ are the weakness and emaciation that result from the imperfect supply of nourishment ~~consequent on the difficulty of swallowing, and the bronchitis that is apt to be set up by the passage of particles of food into the air-passages.~~ ^{from}

^{a time}
The *Course* of the disease is progressive, although not uniformly so. From time to time its ^{course may be} ~~progress is retarded~~, and it may seem for weeks, or even for months (seldom for a year or so), to be at a standstill. The hopes thus excited are usually baseless, for the disease again ^{progresses} ~~increases~~, often at a more rapid rate. ~~An intercurrent~~ ^{malady} ~~malady, which lessens the patient's strength, accelerates the progress of the disease. But the cases which occur in the first half of life occasionally become arrested before reaching an extreme and dangerous degree, and the duration of this later stage may be indefinite.~~

The *Causes of Death* are gradual weakness from inanition; broncho-

* Attention has lately been especially directed to it by Guinon and Parmentier, 'Nouv. Icon. de la Salpêtr.', 1890 and 1891.

† Ritchie, 'Glasgow Med. Journ.', 1888; Robin; Galezowski (quoted by Ritchie).

with all its consequences, pneumonia, from the entrance of food into the air-passages; failure of respiratory power from associated atrophy of the muscles of the trunk, or the effects on lungs and heart from the extension of the degeneration to the pneumogastric centres.

PATHOLOGY.—In the affected muscles there are changes similar to those seen in progressive muscular atrophy of the limbs. The fibres may present extensive granular and fatty degeneration, or may be simply narrowed. Usually some fibres are much affected, and others but little, so that empty sarcolemma sheaths and normally striated fibres may lie side by side. There is an increase of the nuclei of the sheaths and of the interstitial tissue, and the latter may present an abnormal accumulation of fat. Masses of reddish pigment, the product of the degeneration of the fibres, lie between them. The nerve-endings in the muscle are also degenerated. The motor nerve-trunks are grey and soft, and the microscope shows degeneration of the nerve-fibres and increase of the interstitial tissue.

The medulla oblongata itself generally appears normal to the naked eye; rarely there is slight diminution in bulk. ~~The important morbid appearances are revealed only by~~ Microscopical examination. ~~There may be distinct atrophy of the fibres of the hypoglossal and other nerves within the medulla. In the motor nuclei, changes are found similar to those presented by the grey matter of the cord in progressive muscular atrophy. There is wasting of the nerve-cells, many of which lose their processes and shrink. The interstitial tissue is altered in various degree, and contains granule-corpuscles and other products of the degeneration of the nerve-elements in early cases; in some instances there is a conspicuous increase in the connective-tissue elements; the vessels may be increased in number and their walls thickened. But cases vary remarkably in this respect: in some, only a wasting of the nerve-cells can be discerned; while in others the aspect is that of an actual interstitial inflammation, even involving minute extravasations in the early stage,* and yet limited to the motor nuclei. The variations suggest that there are parenchymatous and interstitial forms.† This difference may explain some facts of limitation or irregularity of the symptoms.~~ shows disappearance of them neuroglia, & the walls of the vessels are

Degeneration is often found in the anterior pyramids, just as the pyramidal tracts are commonly degenerated in the corresponding affection of the anterior cornua of the spinal cord, which so often co-exists with the bulbar degeneration. In such combined cases the sclerosis of the pyramidal fibres may be traced through the pons and cerebral peduncles. When, as is commonly the case, there is muscular atrophy elsewhere, the spinal cord presents corresponding alterations in the grey and white substance. It is probable that, in some cases of bulbar paralysis, the disease is confined to the pyramidal

* Eisenlohr, Arch. f. Psych., 1879, p. 34.

† Compare "Acute Polio-myelitis," vol. i.

without wasting,

fibres, as in cases of lateral sclerosis of the spinal cord causing spastic paralysis.

The degeneration in the bulbar nuclei is constant and most considerable in the hypoglossal nucleus, next in the adjacent part of the nucleus of the spinal accessory nerve, and it is usually found in less degree in the nuclei of the vagus, seldom in the nucleus ambiguus or glossopharyngeal, or in the motor nucleus of the fifth nerve, or in the chief facial nucleus (fig. 36, p. 48). The collections of small cells in front of and behind the hypoglossal nucleus* usually escape. Degeneration of some of the fibres of the raphe has been occasionally noted at the level of the hypoglossal nucleus. Considerable degeneration has been found in the fibres of the "loop" of the facial nerve, even when the cells of the chief facial nucleus were but little affected—a mysterious fact. The posterior longitudinal fibres may be normal or partly degenerated† (in proportion, it is said, to the affection of the hypoglossal nucleus); especially the longitudinal fibres suffer in the inner part of the reticular formation which are supposed to continue the anterior ground fibres of the cord. The degeneration is always bilateral.

The grouping of the symptoms according to function makes it practically certain that the incidence of the lesion is determined by function, that it begins, as a rule, in the nerve-elements, and follows their functional relations in its extension. The exact degeneration on which the paralysis of the orbicularis depends is the most obscure point. Lockhart Clarke believed that the small cells above the hypoglossal were connected with the facial nerve, and that it was their disease that causes the paralysis of the lips. Later researches have failed to confirm this, but it is highly probable that the orbicular fibres do descend to the neighbourhood of the hypoglossal nucleus, and may even arise from its cells (see p. 50); although it is impossible to trace them among the many horizontal fibres of the reticular formation, in which they probably pass downwards, perhaps reaching it by the raphe, into this many fibres of the facial nerve may readily be traced at the upper part of the "loop."‡ The fibres of the anterior pyramids and pyramidal tracts in the pons and crura have been found degenerated, chiefly in the cases in which there has also been atrophy or palsy of the limbs; the degeneration of the pyramids is then the rule (see vol. i, p. 490).

Hence it seems The probability has been already mentioned that, as Duchenne originally maintained, there are two varieties, the atrophic and the paralytic; and that, as already mentioned, in some of the cases without obvious wasting, the primary disease is not in the motor cells, but in the fibres that connect these with the cerebral hemispheres, or in

* The former is often termed "Roller's nucleus."

† See Muratoff, 'Cent. f. Nervenk.', 1891, p. 514.

‡ The tongue and lips have been paralysed together and alone on one side, with the reaction of degeneration (Grugia and Mattencei, 'Arch. Ital. Mal. Nerv.', 1887).

Thus we can understand
~~the lower extremities of these fibres within the nuclei. We have seen that the pyramidal fibres may be degenerated, and these fibres are homologous with those that constitute the upward path from the nuclei to the cortex. In cases in which these fibres, and not the nuclei, are diseased, there should be no loss of reflex action, and we have seen that, in some cases, the reflex action is preserved. Moreover, in some of these cases there is marked excess of myotatic irritability in the muscles of mastication, even, as we have also seen, so great as to permit a jaw-convulsion to be obtained—distinct evidence of the degeneration of the fibres of the upper segment of the motor tract for these muscles (see vol. i, p. 201). The other parts specially paralysed do not permit this muscle reflex action to be tested. Thus these cases will constitute the bulbar homologue of primary lateral sclerosis of the cord.~~

Atrophic bulbar paralysis must be regarded as an affection practically identical with the progressive muscular atrophy that is so often associated with it. The difference between them depends only on the seat of the morbid process, and not at all on its nature.* The disease is a degeneration of the lower segment of the motor path for the muscles affected, and in some cases it may be a degeneration of the whole path, upper and lower segments, or of the upper segment alone, when there is no wasting. Although an affection of the cortical cells has not yet been found, it is highly probable, from the analogy of progressive muscular atrophy. In other cases, again, there may be a combination of the two conditions analogous to those in which atonic atrophy in the hands is associated with tonic palsy in the upper arms (see vol. i, p. 480).

the over
 The relation of the degenerative changes to the function of the affected cells, so conspicuous in the distribution of the symptoms, merely, of course, means that a certain common function is associated with a common special susceptibility, on the part of the cells, to suffer from a defect in nutrition, which is progressive in character, and tends to increase to destructive degeneration. It is essentially a defective vitality, and, as such, is associated with an exuberance of growth in the interstitial elements, analogous to that which is seen in the elements surrounding a degenerating nerve fibre. The real pathology of the disease is the cause which induces the nutritional changes, and of that we know almost nothing. The possible influence of toxic conditions should always be remembered.

DIAGNOSIS.—The distribution of the palsy, its bilateral character, its gradual onset and progressive course, separate the disease with sufficient sharpness from most other maladies. The slow onset distinguishes it from acute lesions of the medulla, which may cause the symptoms of similar character and distribution described in the

* The essential identity of the two diseases was first urged by Kussmaul (Volkmann's 'Clin. Lect.,' No. 54, 1873).

*spinal centres
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next section. The chief difficulty is presented by organic diseases of the medulla, which may also cause "bulbar palsy" of ^{slow} onset. ~~The most frequent of these is a tumour in or outside the medulla, or a narrowing of the foramen magnum by thickening of the bone or enlargement of the odontoid process, damaging the bulb by invasion or compression, or compressing the nerve-roots; but, the symptoms thus produced seldom present the perfect bilateral symmetry that characterises the degenerative affection. One side is affected first or most, the difficulty of swallowing preponderates over the other symptoms, and the lips usually escape altogether. Moreover, headache is usual, and convulsions are occasionally met with.~~ ^{such as} ^{medulla} ^{of}

Tumours within the medulla sometimes give rise to greater difficulty, but the cases in which a growth acts on both sides in such an equal manner as to cause perfectly symmetrical symptoms are ~~excessively rare, and there are usually other indications to help the diagnosis, either there are other indications of a tumour, or the patient is so young as to render the degenerative disease highly improbable.~~ ^{very} Insular sclerosis, involving the medulla, is seldom so symmetrical as to give rise to real difficulty, and there are always indications, in other parts, of the morbid process.

Chronic lesions in both cerebral hemispheres may cause symptoms resembling those of bulbar palsy, constituting a chronic form of "pseudo-bulbar paralysis." Although such an effect is far more rarely produced by chronic than by acute lesions, it has been caused by symmetrical sclerosis of the two hemispheres; * The chief distinctions are ~~afforded by the development of symptoms first on one side and then on the other~~ ^{and} the affection of the limbs, as well as the face, in distinct double hemiplegia, ~~and by other symptoms indicative of the nature of the morbid process.~~ Reflex action is preserved in the parts paralysed, and these are never wasted, but alone these points are not sufficient for the diagnosis, although they may be allowed weight in support of other indications.

PROGNOSIS.—In every case of gradual onset the prognosis is most grave. The affection is so ^{then} progressive ~~in its tendency~~, and the parts affected are so important for life, that the disease almost invariably leads to death. This is especially true of the senile form, and of that which supervenes on spinal muscular atrophy. When the affection begins in middle life, arrest may be regarded as a possible event, and it is not improbable in the rare cases that come on in late childhood, or follow some disease that has nerve-degeneration for its occasional sequel. The possible cause, as well as the ~~obvious~~ ^{obvious} course, must, therefore, be considered in the prognosis. ^{generally}

TREATMENT.—Although experience shows that we can scarcely expect, in any case, that our treatment will have an appreciable influ-

* Jolly, 'Arch. f. Psych.,' iii, 1872.

ence on the disease, we may strive at least to retard its progress, and employ such measures as, from their action on the nervous system, are most likely to produce this effect. ~~It is probable that cases will occasionally be met with in which the morbid tendency is less strong than it usually is, and in which treatment may have some influence.~~ Nervine tonics, quinine, strychnine, arsenic, phosphorus, or nitrate of silver may be given, ~~or hypodermic injections of strychnia ($\frac{1}{16}$ gr.) may be administered, as recommended for progressive muscular atrophy (vol. i, p. 497).~~ The latter is especially indicated in the non-senile forms. In advanced cases I have known transient improvement to follow the injection of strychnia combined with minute stimulant doses of morphia ($\frac{1}{36}$ — $\frac{1}{24}$ gr.). The power of swallowing has been increased by this treatment, but unfortunately the effect has not been permanent. To lessen the flow of saliva, belladonna or atropine has been given, but seldom with effect.

Electricity has been extensively used, ~~With the view of influencing the morbid process in the medulla, the voltaic current has been passed from one mastoid process to the other.~~ In order to maintain the nutrition of the muscles, either faradism or voltaism may be employed, for they respond to either in the majority of cases. It may be applied to the tongue, lips, or pharynx; to the last by placing the positive pole at the back of the neck, and moving the other along the side of the throat. Electrification of the sympathetic has also been used as a method of treatment, but is probably as destitute of rational foundation as it certainly is of practical effect. Indeed, the result of all electrical treatment is most unsatisfactory. For an hour or two after each application there may be a little more power, but the effect does not last. I have never observed distinct evidence that electricity had the slightest influence on the course of the disease in any one of many cases in which I have seen it used.

In all cases the feeding of the patient demands much care. Semi-solid pulpy food can usually be swallowed better than liquids or solids. When deglutition is impossible without constant risk of the entrance of particles of food into the air-passages, and of prostrating and distressing paroxysms of choking, liquid food may be given by an œsophageal tube, or by means of a catheter or soft india-rubber tube passed through the nose into the upper part of the œsophagus. The food may be slowly poured into the tube through a small funnel, the patient's head being bent back. The only alternatives are the injection of food by the stomach pump, or the less effectual method of rectal feeding with peptonised food. The larynx has been opened when the attacks of threatened suffocation were frequent and severe.

Chronic Bulbar Paralysis without Anatomical Changes, anomalous and mysterious, has been occasionally met with. The cases have conformed, in general features and course, to the common degenerative variety, but have deviated from this in some minor point. Palsy of

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(voltaism) or peripheral
(voltaic or faradic
current, according to the
response)

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Arthur Myashtun

lips, masseters, and pharynx was attended, in one case,* with only slight weakness of the tongue, but with an amount of paralysis of the intercostals that caused fatal dyspnœa. No changes could be discovered in the nerve-nuclei, and the hypothesis of chronic peripheral neuritis naturally presents itself to the mind, but this was excluded by Oppenheim† in a case in which the larynx, pharynx, and tongue were paralysed. Degenerative changes in the lower part of the cerebro-nuclear fibres may have escaped detection.‡ Moreover, function may be abolished by changes in the nutrition of the nerve-cells, that leave their aspect unaltered.

SUDDEN (APOPLECTIFORM) BULBAR PARALYSIS.

It has been already mentioned that various acute lesions of the medulla cause symptoms in the region of the nerves implicated in the degenerative form, and that these lesions generally differ from the latter in the less regular grouping of the symptoms produced. Occasionally a sudden lesion causes symptoms that are perfectly symmetrical, and correspond very closely to those of degeneration of the bulbar nuclei. Most of the sufferers have been advanced in life, —at the period in which ^{atheroma} arterial degeneration is common; but this form has also been met with in younger persons as a result of blows and falls on the head or neck,§ or of syphilis, influences which may cause disease in the vertebral artery, or lower end of the basilar, and narrow or occlude the branches. Such disease may cause successive sudden attacks, sometimes induced by depression of the general health or some influence that retards the circulation or promotes thrombosis. This is also true of another very rare mechanism, embolism, by which the vertebral may be closed (owing to the narrower calibre at its extremity), or its branches; the latter are commonly saved from separate occlusion by the angle at which they arise, which makes the entrance of a plug difficult, though not impossible. This arrangement, while it indisposes to embolism, promotes thrombosis. Among the ^{rare} many causes of thrombosis in this situation must be placed chronic basal meningitis and its effects on the arteries.¶ The onset of the paralysis is sudden and "apoplectiform," sometimes with giddiness and vomiting, usually without loss of consciousness. The initial symptoms may be of wider range; there

it is also met with in middle life from syphilis, & probably as a result of minute hemorrhage and adjacent inflammation. Arterial

* Dr. L. E. Shaw, 'Brain,' 1890.

† 'Virchow's Archiv,' Bd. cviii.

‡ Compare vol. i, p. 440.

§ As in the case of a boy aged twelve, recorded by Schulz ('Neurol. Centralbl.,' 1883, p. 99), in which bulbar paralysis followed immediately a blow on the back of the neck, and increased considerably a day or two afterwards. It is assumed that there was traumatic hæmorrhage, followed by secondary inflammation. Compare a case of fatal traumatic hæmorrhage recorded by Bochefontaine, 'Arch. de Phys.,' 1883, p. 160.

¶ The possibility that thrombosis, found after death, may have been only a terminal event must also be borne in mind.

may be weakness of the limbs or an affection of sensibility, usually in the form of subjective sensations, rarely actual anæsthesia. ~~Any of the symptoms of a lesion of the part may co-exist, often only for a time; the~~ ^{early} ~~wider~~ symptoms pass away, ~~and~~ ^{but} leave a permanent condition closely resembling that of the degenerative form, ~~but~~ differing in the absence of a progressive tendency. For a time, indeed, ~~improvement may be conspicuous; the symptoms lessen, and then the condition becomes stationary or nearly so, for a time which differs with the causal influences.~~ ^{there may be} As elsewhere, the symptoms may ~~increase~~ ^{last} for a few days, owing to inflammatory processes in the vicinity of the most affected area. Functional disturbance (as spasm in the masseters) has been observed to ~~precede or attend~~ ^{at} the onset. In ~~some~~ ^{rare} cases, after a time, degenerative changes supervene on the original acute lesion, and the case assumes a progressive character. ~~Not seldom the symptoms, although bilateral, are unsymmetrical; but it is very rare for them to be quite one-sided, as in a case recorded by Hirt, in which paralysis of one vocal cord and of the corresponding side of the tongue came on suddenly; there was wasting of the tongue with the reaction of degeneration.* Very often the symptoms, although bilateral, are more or less irregular in distribution and character, as we should expect from the nature of their cause. This feature is often prominent and important; there may be absolute inability to swallow, with no paralysis of the tongue or face. When the onset is by several successive attacks, each is usually slight, although one may be more severe than the others.~~

As an example of this sudden form may be mentioned the case of a man ^{who} ~~aged sixty-three.~~ ^{at} ~~fifty-five~~ ⁶³ he had a slight attack of right hemiplegia of ordinary type, without affection of speech or of swallowing, from which he recovered perfectly in the course of a few months. ~~Five weeks before being seen he suddenly became unable to articulate, and had great difficulty in swallowing. There was no loss of consciousness. No change in his condition had occurred when he came under observation. His condition then resembled perfectly that of the progressive degenerative form. His lower lip hung down; saliva constantly dribbled from the mouth. The tongue was broad, flabby, and almost motionless, only the tip could be brought over the lower teeth (see Fig. 147). The palate was flaccid, but could be raised a little. Swallowing was very difficult, and attacks of choking were frequent.~~



FIG. 147.—Bulbar paralysis of sudden onset, maximum protrusion of the tongue.

* Hirt, 'Berlin klin. Wochenschrift,' 1885, No. 26.

The irregularity of the symptoms marks

at 63

Sip, weeks later his

The vocal cords could be brought together, although with little force, so that no explosive cough was possible. He could still phonate, although with little modulation. There was not the slightest power of articulation; attempts to speak resulted only in "ah-ah-ab." Expression by writing was unimpaired. This patient was seen from time to time for five years after the onset, and his condition remained essentially unaltered. His power of swallowing varied; sometimes it became less, and then under treatment it improved again, but on the whole the loss of articulation and of swallowing, the paralysis of lips, tongue, and larynx, were neither better nor worse at the end of the five years than they were five weeks from the onset. There was no wasting and no loss of faradic irritability. As in this case, the muscles involved are usually those supplied from the medulla oblongata only, but a curious case has been recorded by Dixon Mann in which there was also some paralysis of the muscles of mastication.*

The pathology of these cases rests at present on little exact observation. It is probable that they depend on softening from vascular occlusion, situated in or near the middle line, and, at any rate, in the cases with muscular wasting, ~~damaging~~ ^{involving} the nuclei that are the seat of degeneration in the progressive form. The position of the lesion in cases such as that detailed above, in which there is no wasting and no loss of electrical irritability, i. e. no evidence of damage to the nuclei themselves, is uncertain. It is probable that, in many cases, the lesion is just above the nuclei, so as to damage the paths from the cerebral hemispheres, which, after descending with the pyramidal fibres, certainly decussate near the nuclei, and would therefore be liable to common damage from a single lesion in the middle line. Thus, in a case described by Leyden, there was an area of softening in the middle line, at the level of the olivary bodies.† When the one vertebral artery is much smaller than its fellow, disease of the latter may damage both sides of the medulla.‡ But there is evidence that bilateral symptoms sometimes result from a one-sided lesion. The functional connection between the nuclei of the two sides is exceedingly close; they habitually act together in perfect equality, and destruction of those on one side may for a time interfere with the function of their fellows on the other side. In most cases of the kind the duration of life has been short, and it may be that the muscles of the other side would ultimately have recovered. No opportunity should be lost of ascertaining the exact nature of all cases of sudden

* Dixon Mann, 'Brain,' July, 1884, p. 244. It is very difficult to explain this direct combination of symptoms except on the hypothesis of a double lesion. But an association of indirect nature is conceivable, since the functions of chewing and swallowing are so constantly united.

† Leyden, 'Arch. f. Psych.,' vii, p. 44.

‡ The basilar may be, in effect, a continuation of one vertebral only, the other ending in the posterior inferior cerebellar artery.

but-

multiple small lesions may cause symptoms of disease of the pons, such as

As a hemorrhage in this situation is scarcely compatible with survival of the actual onset, on account of the pressure of ecerts and the proximity of vital centres.

large

but a one-sided lesion may involve both the fibres that have crossed & also those about to cross & thus cause bilateral symptoms

During

the softening involves

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~~onset, whether a microscopical investigation is undertaken or not.~~

The *Diagnosis* of these cases presents little difficulty. The ~~symptoms are due to a sudden lesion of the medulla, and the cases in which they are symmetrical are not strictly separable from the others just mentioned, in which symptoms of similar character are of less regular and symmetrical arrangement.~~ The distinction from the degenerative form rests on the mode of onset, ~~sudden in the one case, gradual in the other.~~ If a sudden increase of the paralysis should occur in the slow form, the preceding symptoms distinguish it from the acute variety. ~~The nature of the lesion is shown by the indications that furnish guidance in other parts of the brain.~~ It is important to note that symptoms may result from ~~very slight vascular derangement, on account of the proximity of important parts, and that slight gradual derangement sometimes precedes the sudden attack,~~ *the closure of a very small vessel* as in softening from arterial disease in other parts.

(See p.) The chief diagnostic difficulty in connection with these cases is due to the fact that the "pseudo-bulbar paralysis" mentioned on p. 530 may simulate very closely the symptoms of a sudden lesion of the medulla. But, as a rule, the indications of a lesion in each cerebral hemisphere are clear. There are two distinct attacks, scarcely ever simultaneous, and it is the second attack that leaves the paralysis of the lips, tongue, and pharynx that simulates a lesion in the medulla. In most cases of the kind the disease has been in the central ganglia, especially in the lenticular nuclei; less commonly it has been in the white substance, or in the lower part of the motor cortex, or in this part on one side and in the central ganglia on the other. In cases of this nature, the diagnosis is not difficult if the observer is aware of the possibility of bulbar symptoms. A much greater difficulty exists if the bulbar symptoms follow a single apoplectiform seizure. In some instances, lesions occur simultaneously in both hemispheres; in others, a subsequent post-mortem examination has shown that only one hemisphere of the brain was diseased, and the lesion has sometimes been on one side, sometimes on the other. We have seen that an acute lesion may completely inhibit the function of the corresponding centre in the other hemisphere, even for days. It has been assumed that the lenticular nucleus has a special relation to the processes of articulation and deglutition,† an assumption which is scarcely consistent with the frequency with which it has been found diseased without the occurrence of bulbar (or other) symptoms. The implication of the internal capsule is generally possible in lesions of this nucleus. Another hypothesis is that in cases of one-sided lesion there is an exclusive relation of the affected functions to one hemisphere, owing to some congenital structural peculiarity.‡ On the

* Cases of this character have been described by T. Barlow, Joffroy, Lépine, Berger, Jolly, Ross, and others.

† Ross, 'Brain,' July, 1882.

‡ O. Berger, 'Breslauer Arztl. Zeitschr.,' 1884.

indicate disease of the medulla. the sudden onset shows that it is due to a vascular lesion; the mere fact of survival excludes hemorrhage

latter

vessel

symptoms may

due to disease in both hemispheres

in the very rare cases of simultaneous lesions in both hemispheres. But their rarity almost places them outside the practical diagnosis especially since the seat of the disease has no influence on the treatment that is needed. Moreover a lesion on one side may cause bilateral symptoms at first, by its inhibitory influence on the corresponding centres of the other hemisphere (see p.) but these pass away in a few days. In some rare mysterious cases of one-sided disease, bilateral bulbar symptoms have been enduring. No satisfactory explanation has been given of these cases.

other hand, it has been held that undetected microscopical lesions of the medulla are the real cause of the symptoms in these and many other cases of pseudo-bulbar paralysis.* The certain fact is that we have much still to learn by careful observation before these cases of unilateral disease can be comprehended.

Besides the distinction ~~often~~ afforded by the mode of onset, pseudo-bulbar paralysis is characterised by the slighter degree of the symptoms, and by the facts that the nutrition of the tongue is unimpaired, reflex action is undiminished, and there is no change in electrical irritability. The larynx is seldom paralysed. ^{By} These negative characteristics ^{are} sometimes present when the lesion is in the medulla, ^{so that} their diagnostic value is not absolute. The fact that a second lesion may occur in the medulla must also be borne in mind, ~~and must occasionally lessen even the moderate amount of confidence with which the diagnosis of pseudo-bulbar paralysis can be made.~~

Prognosis.—Sudden apoplectiform bulbar paralysis involves much danger to life in the early stage of the disease, but the subsequent prognosis is less grave than in the chronic degenerative form. ~~The tendency of the symptoms to progress is slight or absent. In many cases considerable improvement occurs; some recover entirely. In others, as in that described above, little or no power returns in the paralysed parts. The only guide as to the probable course of the symptoms is that afforded by the condition of the patient when the acute stage has passed. If, at the end of a month, there is no sign of improvement, and there is still a considerable degree of paralysis, it is improbable that much improvement will ensue. But the further forecast must depend on the nature of the lesion and on the indications available, which have been considered in the general account of such lesions. Then, if the evidence points, as it often does, to atheroma and thrombosis, the danger of a recurrence will be less, according to the absence of influences favouring thrombosis, or their limitation to such as may be obviated by suitable measures. The malady is only a local instance of the disease already considered.~~

The *Treatment* of this form illustrates the fact just mentioned; it is essentially that of acute softening from vascular occlusion, as described in detail at a previous page. ^{already} The general management of the case, in regard to feeding, &c., is the same as in the degenerative variety.

ACUTE INFLAMMATORY BULBAR PARALYSIS (POLIO-MYELITIS BULBI).

The term "acute bulbar paralysis" is commonly applied to the sudden form described in the last section, but is needed for a third variety, to which alone it is strictly applicable, in which the symptoms

* Oppenheim and Siemerling, 59th 'Versam. Deut. Aerzt.,' 1886.

develop, not suddenly, in a few minutes, but acutely, in a few days. The mode of onset is that which is characteristic of acute inflammation, and that this is actually the lesion has been proved by several cases. In one observed by Etter,* a boy, aged fifteen, was taken ill with headache and vomiting, discomfort in the throat, difficulty in swallowing, and fever. In the course of the first week there developed bilateral paralysis of the face, accompanied by palsy of the tongue, greatest on the left side, paralysis of the palate, and of the left sixth nerve.

Death resulted from pneumonia on the tenth day. The post-mortem and microscopical examinations revealed many myelitic foci in the medulla, symmetrical on the two sides, but greater on the left. They involved the left sixth nucleus, the left facial nerve within the pons, and the right facial nucleus, the left hypoglossal nucleus and the right hypoglossal fibres, and the accessory nucleus on each side, the motor part of the vagus nucleus on each side, and many cells and fibres in the tegmental region of the lower part of the pons. The foci of inflammation in the accessory nuclei could be traced down the cord as far as the fourth cervical nerves. Such a lesion is evidently closely analogous to that which, in the spinal cord, causes acute atrophic paralysis. We have seen (p. 194) that a similar inflammation may involve the nuclei of the ocular nerves in the upper part of the pons. The distribution in this case is that which has been found in the few others investigated, but the glosso-pharyngeal nuclei may also suffer. The inflammation is sometimes irregular in its distribution in the nuclei, and in one case it caused paralysis of all parts of the face. Hence this affection has been termed "acute inferior nuclear inflammation."

Its causes are as little known as are those of the similar inflammation of grey matter in other parts.

The character of the changes seen with the microscope depends on the acuteness of the inflammation, varying from destruction of the nerve-elements and leucocytal aggregations or minute cavities of disintegration, to mere change of form and aspect of the nerve-cells, "cloudy swelling," loss of the processes, and ultimate shrinkage into small angular bodies. The increased interstitial tissue may present conspicuous spider-cells and the other features described in the account of analogous affections.

It is possible that similar symptoms are sometimes due to a peripheral neuritis of the bulbar nerves. Disease elsewhere prepares us for a similar susceptibility of the motor cells and their nerve-endings to morbid influences, which may act generally on one, occasionally on the other, owing to some slight difference in nature. But such acute peripheral bulbar neuritis has hitherto only been met with as part of a more general affection of the cranial nerves, such as existed in a case described by Eisenlohr,† in which there were hæmorrhages in nearly

* 'Corresp.-Bl. f. Schweizer Aerzte,' 1882, No. 24.

† 'Arch. f. Psych.,' ix.

He died

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and

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A lesion of the same character have been found in

changes described in the account of poliomyelitis

I have seen, in poliomyelitis, affection of the nuclei of the thoracic and one side of the face

seen to be such as cause

all the cranial nerves. Bulbar palsy was the predominant symptom, ~~but included all parts of the face; the motor fifth was also weakened.~~

The treatment of this affection must be that of other forms of acute inflammation of the brain.

ATROPHY OF THE BRAIN.

The whole or part of the brain may be below the normal size. In general atrophy the texture of the brain is normal; the whole brain is small, and the skull is also small (microcephaly). Little is known of the causes of this condition, whether the small size of the brain is the cause of the small size of the skull, or the reverse. The condition is generally associated with a high degree of mental defect of long duration, ~~but an unexpected diminution in the size of one hemisphere is sometimes met with in chronic insanity, probably connected with a predisposition to the mental derangement.~~

In partial atrophy of the brain, ~~one part is unduly small in proportion to the rest. The part so affected is usually changed in structure. It is firmer than normal—less firm only in rare cases of uncertain nature. It contains more connective tissue and fewer nerve-elements. Such sclerotic induration involves a difficulty as to the classification of the lesion, as already mentioned (p. 555).~~ Almost any part of the brain may be thus smaller than normal—the whole of one cerebral hemisphere, or only part of it, seldom both hemispheres, the central ganglia on one side, the pons, or the cerebellum. Atrophy of the cerebellum may involve the whole or only one hemisphere, or both hemispheres—the middle lobe being normal. ~~When the whole of one cerebral hemisphere is atrophied, one part is usually more affected than the rest.~~ Several parts of the brain are frequently atrophied together, in a manner that shows ~~a causal relation, depending on a structural connection, in consequence of which the atrophy of one part entails that of the other.~~ Thus atrophy of the whole of one cerebral hemisphere is usually associated with atrophy of the opposite cerebellar hemisphere. The latter may be associated with atrophy of the opposite corpus striatum ^{alone} ~~without the rest of the hemisphere, and it is usually associated also with atrophy of the opposite olivary body of the medulla oblongata.~~ The parts concerned in vision may be thus marked out by separate wasting (see p. 71). The arteries ~~may share the diminution in size without presenting any special disease.~~ *are also lessened by*

Partial atrophy of the brain is probably in most cases acquired, ~~This is clear in many instances in which it is attended with symptoms that commenced during the first years of life. In some of these cases the lesion dates from birth, and the change in the brain is the~~ *as is shown by attendant*

result of its compression by local meningeal hæmorrhage ^(see p. 413). In other instances the lesion occurs during infancy, and is accompanied by "infantile hemiplegia," in the account of which (p. 456) will be found some evidence as to the nature of the disease. In some cases, again, it is probable that an attack of meningitis was the cause. Both hemispheres may suffer from this cause, and also in the cases of meningeal hæmorrhage during birth. In all varieties, the local atrophy is often accompanied by a general diminution in size of the hemisphere, no doubt due to the connection that exists between all parts, and to the effect of considerable damage to one part of the growing brain on the development of the rest. With only local atrophy of the superior parietal lobule, one hemisphere of an adult has been found one and a half inches shorter than the other.*

Partial atrophy of the brain may also develop during intra-uterine life. It may be found present at the time of birth. In some cases it is perhaps due to intra-uterine disease similar in character to that which, after birth, has similar consequences. More frequently the atrophy is such that it can scarcely be thus explained; Atrophy of the cerebellum is more frequent in these cases than is atrophy of one cerebral hemisphere. Sometimes both cerebellar hemispheres are very small, and the middle lobe is of normal size. It is very difficult to explain this condition on the supposition of any intra-uterine morbid process; it would seem rather to be connected with the entire absence of the cerebellum sometimes observed. This is apparently due to some perversion or defect of the process of development, the cause of this is practically unknown, but we are probably safe in ascribing it to a positive germinal defect.

The symptoms that attend atrophy of the brain vary much. Mental defect is the most constant, and usually amounts to idiocy. Hemiplegia and epileptic fits often accompany atrophy of one cerebral hemisphere. Bilateral weakness, with athetoid movements and inco-ordination, may attend bilateral atrophy, such as is due to difficult birth and meningeal hæmorrhage compressing the cortex. In many cases, however, it is certainly incorrect to regard these symptoms as the direct consequences of the diminished size of the convolutions; they are the result of the disease which has also caused the atrophy, the local atrophy directly, the slighter general atrophy of the hemisphere, which is often present, indirectly, by checking its growth. In atrophy of the whole of the cerebellum, unsteadiness has been observed similar to that which results from disease of the middle lobe. When only the hemispheres have been affected, there have sometimes been no motor symptoms; but in some cases there has been intellectual defect. Atrophy of one hemisphere of the cerebellum has also been found when no symptoms, that could be ascribed to it, were present during life.

Senile Atrophy.—In old age the brain wastes, like many other organs,

* Fraser, 'Glasgow Med. Journ.,' 1889.

already described considered together with the

may be which occurs

It is often

are usually rather than of the state itself

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has been

and becomes smaller and firmer. The amount of fluid in the ventricles and on the surface is increased in proportion to the lessened bulk of the brain. This natural increase of fluid was formerly regarded as pathological, and ^{found after} a mysterious death was ascribed to "serous apoplexy"—a purely imaginary lesion, not yet quite forgotten. But this wasting of the brain is commonly attended by no symptoms. Senile mental failure is often ascribed to it, but, since it may exist in considerable degree without the slightest mental defect, ^{any intellectual change} ~~caution should be observed in attributing to it any mental change that may co-exist.~~

HYPERTROPHY OF THE BRAIN.

Under the name "hypertrophy of the brain" a condition has been described in which the brain is of abnormal size. Sometimes it has been found also to be of distinctly abnormal weight, but the wide variations in the weight of the brain under normal conditions render this criterion decisive only in extreme cases.* The size of the brain in relation to the size of the skull is the feature that has attracted most attention. If the condition develops before the bones have united, the skull is said to undergo enlargement similar to that of hydrocephalus. If the sutures have been closed, the convolutions are compressed and pale; and when the brain has been removed, it cannot be replaced in the cranial cavity—a feature which, since the days of Morgagni, has attracted attention, perhaps more than it deserves. In all cases the ventricles are empty; their walls are pressed together, and the vessels of the brain contain little blood.

Decisive examples of this state are extremely rare; there are few satisfactory observations on the minute structure of the enlarged organ, and we do not therefore know whether this is the same in all cases. In some, an increase in the neuroglia has been found, and has been regarded as the cause of the enlargement (Virchow; Tuke, in ~~an unusual case in which the enlargement was confined to one hemisphere~~). In other cases no increase in the connective tissue could be found.

Such enlargement of the brain has been met with chiefly under three conditions. (1) In very young children soon after birth. It is in these that the hydrocephaloid enlargement of the head has been

* The weight and size of the brain differ according to age and stature, but even thus tested the variations are great. Proportion to absolute body-weight has also been taken as a guide, but it is not to be relied on, since the body-weight varies in each direction, that of obesity and emaciation, irrespective of stature. If the body-weight is taken as a term of comparison it should be the average for each age and stature.

described. (2) Towards the end of the first year of life, in association with rickets. It may be that, the closure of the skull being retarded, the brain attains an abnormal size in consequence of the deficiency of the mechanical restraint that should be furnished by the skull when its bones unite. It has been thought that the enlargement of the brain increases the size of the skull, but the shape of this is that characteristic of rickets, and the size may be due to the double influence of abnormal ossification, and the enlargement of the brain. (3) An increase in size has been occasionally observed in older children and even in adults, but of its nature very little is known.

Of the causes of this condition only two facts can be regarded as established: first, that it has been occasionally observed in families; secondly, the relation to rickets already described.

The symptoms that it produces are very uncertain. The form that occurs soon after birth and causes enlargement of the head is said to be attended by nervous symptoms almost identical with those of chronic hydrocephalus, so that the distinction of the two is practically impossible. It is doubtful whether the rickety enlargement causes any symptoms. We are not justified in referring to it the laryngeal spasm or general convulsions or mental backwardness of rickety children, since these may be present when there is no enlargement of the brain.

Even more doubt exists as to the symptoms of the form that has been met with at a later age. In some cases it has apparently caused none. In others, acute cerebral symptoms, resembling meningitis, have occurred, and have run a rapid course, ending in death. The evidence of a connection between the enlargement of the brain, ~~which was regarded as chronic, and the acute symptoms is not satisfactory, and no explanation of the supposed connection has been attempted.~~ The condition seems never to have been recognised during life, and nothing is known of its treatment.

The so-called hypertrophy of a small part of the brain, as of certain convolutions, or of the pons Varolii, has probably been always due to an infiltrating growth (see p. 495).

From what has been said, it is clear that the pathology of enlargement of the brain needs fresh study in the light of future investigation. Moreover, the common tendency to speak of the enlargement caused by overgrowth of connective tissue as "hypertrophy" of an organ, is an inexact use of the word, which, when applied to an organ without qualification, should designate only an enlargement chiefly due to an overgrowth of the elements on which the function of the organ depends.

HYDROCEPHALUS.

Hydrocephalus, or dropsy of the brain, consists in an accumulation of fluid within the skull, either in the subdural space (external hydrocephalus) or within the ventricles (internal hydrocephalus). It may be acute or chronic, the result of other morbid processes that are conspicuous ("secondary form"), or the consequence of a process that is chiefly manifested by this effect (so-called "primary form").

ACUTE HYDROCEPHALUS.

The only known cause of actually acute hydrocephalus is meningitis. This, in all forms, may be attended by effusion of fluid in the subdural space or into the ventricles. The latter, for instance, occurs in four fifths of the cases of tubercular meningitis. Hence the term "acute hydrocephalus" was for a long time the common designation for meningitis. ~~The~~ External effusion is the direct result of the inflammation of the pia mater, and the internal effusion is probably also the result of inflammation of the choroid plexuses and velum interpositum, perhaps also of the lining membrane of the ventricle.

In some cases, however, the effusion into the ventricles is the only pathological change in an affection of acute and febrile course. The external meninges are healthy; the choroid plexuses may be the seat of distinct inflammatory changes, and the lining membrane of the ventricles may be finely granular; the adjacent brain tissue is softened, and the brain-substance and convolutions are compressed. There is no indication of any processes of which the effusion could be a mechanical consequence, such as is an occasional cause of chronic hydrocephalus. The symptoms during life are those of an acute inflammatory disease, and resemble very closely those of tubercular meningitis. Hence it is commonly assumed that they are due to a ventricular meningitis, affecting chiefly the choroid plexuses, and ~~causing an abundant effusion of serum. Although the pathology of the cases is obscure, no better explanation has yet been given.~~

CHRONIC HYDROCEPHALUS.

EXTERNAL CHRONIC HYDROCEPHALUS.—Whenever there is wasting of the brain, there is increase of the subarachnoid fluid to occupy the space left. This is a common senile condition. Of the same character is the accumulation of fluid met with in some cases of arrested development, in which the brain is small and does not fill the cranial cavity.

In other cases, without the brain being smaller than normal, there is a congenital excess in the amount of fluid in the subdural space, and this expands the skull. The expansion may be so great as to prevent the child being born alive. If birth is survived, the head rapidly increases in size, and is of the same shape as in the internal hydrocephalus, to be immediately described. Post mortem, the excess of fluid is the only morbid condition. There is no change in the membranes, and the origin of the condition is not known. It is usually attended by symptoms similar to those of internal effusion, ^{and has} having a similar course. Caries of the bone of the skull, in such cases, has occasionally permitted the escape of the effusion.

External hydrocephalus is sometimes "sacculated," *i. e.* limited to a certain region by adhesions between the dura and pia mater. This condition is, in effect, a meningeal cyst. Its position may be over part of one cerebral hemisphere or beneath the tentorium. If there is enlargement of the skull, this is correspondingly unsymmetrical. The brain may be compressed opposite the seat of the effusion, and corresponding local symptoms may result. These cases doubtless arise from inflammation, and their course may be varied by intercurrent attacks of a meningitic character.

CHRONIC INTERNAL HYDROCEPHALUS may be either congenital or acquired.

Congenital Internal Hydrocephalus develops *in utero*, and may cause such enlargement of the head as to prevent birth until the fluid is let out; or it may be moderate at the time of birth, and afterwards rapidly increase. The causes and pathological mechanism are practically unknown. It has been ascribed to maternal ~~grief or other~~ emotion, but on no reasonable grounds, and also to injury of the foetus by falls, &c.,—an influence which is at least intelligible. A connection has been supposed to exist between uterine disease and hydrocephalus, and has been explained hypothetically, but it has yet to be proved. There is, however, no doubt that a tendency to the occurrence of foetal hydrocephalus sometimes runs in families, and that more than one child may be affected in succession.

Sometimes all the ventricles are distended; more often the fourth ventricle suffers little, even when there is no obstruction in the aqueduct of Sylvius. In such a case the aqueduct may be funnel-shaped, dilated towards the enlarged third ventricle. If the distension is confined to one or both lateral ventricles, there is some obstruction at the foramen of Monro (see further, under "Acquired Hydrocephalus"). The fluid is usually clear, of low specific gravity, 1001—1009, and contains a small and variable amount of albumen, some chloride of sodium, and sometimes urea, cholesterin, and other substances in small quantity. The amount of liquid varies according to the degree of the disease, and has been as much as twenty-seven pounds. In proportion to its quantity, the substance of the cerebral

hemispheres is compressed, and thinned by extension and wasting, especially the white substance, and, it is said, the callosal fibres.* ~~The corpus callosum is displaced upwards, and if the cranium is enlarged, the hemisphere extends further forwards, backwards, upwards, and outwards than normal.~~ The cerebral substance may be reduced to a layer only a few millimetres thick, constituting the thin wall of a vast cavity. In such a case, all traces of the convolutions and sulci may have disappeared, and even the basal ganglia may be almost unrecognisable. More often indications of the sulci can be traced, and the compressed basal ganglia lie at the bottom of the sac. The cranium is enlarged in proportion to the amount of effusion, and the bones of the skull are thin. The falx is necessarily stretched, and its edge ~~forms a larger curve than normal, the extension of its edge raises the anterior part of the tentorium.~~ Hence the subtentorial space is increased in size, and is ~~not completely filled by the cerebellum, the interval being~~ occupied by liquid, and sometimes by loose connective tissue. Otherwise the membranes are normal. The choroid plexus may be thickened, as if from old inflammation. The lining membrane of the ventricles is often finely granular ~~on the surface~~, and is sometimes thickened.

The external enlargement of the skull is very conspicuous and characteristic, and rapidly increases after birth. The fontanelles become very large, and bulge; at the sutures, the bones are widely separated. The cranium has a rounded shape, and ~~becomes~~ disproportionately large in comparison with the face. The disparity is increased by the projection of the frontal ~~portion of the skull.~~ ^{region} The orbital plates have an oblique direction, and the eyes are directed downwards and partially covered by the lower eyelids. If the child lives, the head may attain an enormous size; in one case, at sixteen months old, the circumference was 107.6 cm. (Klein). The symptoms vary much. There is usually considerable mental defect, ~~often amounting to idiocy.~~ The limbs are weak; convulsions and various contractures are frequent; ~~occasionally febrile attacks occur, attended by vomiting.~~ The head is supported with difficulty, on account both of its weight and of the muscular weakness. The eyeballs are often rolled from side to side, and their axes may not correspond. The skin of the head is thin, and the hair scanty. In extreme cases, blindness occurs, and the ophthalmoscope shows optic nerve atrophy, produced by the stretching of the nerve, or by the compression of the chiasma. In congenital cases, the rapid increase of the disease usually causes death in the second or third month of life, by marasmus, convulsions, or coma. Occasionally the morbid process becomes arrested, and the patient may live on to adult life ~~and even to old age.~~ It is said that the fluid is sometimes gradually absorbed. Ossification of the bones may progress, and is usually completed by the development of "Wormian bones" within the sutures. If the disease is considerable in degree,

* Anton, 'Wien. med. Jahrb.,' 1888.

the mental and motor symptoms usually continue in some degree through life—mental weakness, often with irritability of temper, epileptic fits, muscular weakness, and contractures.

Acquired Chronic Internal Hydrocephalus may be (1) secondary to a lesion that produces the effusion mechanically; (2) consecutive to an attack of meningitis; (3) of apparently primary origin.

Secondary Mechanical Form.—Whytt pointed out in the last century that any obstruction of the veins of Galen, hindering the return of blood from the intra-ventricular vessels, causes effusion into the ventricles, which may reach an extreme degree. The most common cause is an adjacent tumour. But an obstruction to the passage of fluid out of the ventricles is ^{in some cases} another mechanical cause, commonly co-existing with the pressure on the veins, and more certainly effective when it acts alone. The fluid normally escapes by the openings in the membrane closing the fourth ventricle, the foramen of Magendie in the middle line, and the openings, one on each side, behind the roots of the glosso-pharyngeal nerve, first described by Mierzejewski. These may be occluded by meningitis, and then all the ventricles become distended. If the passage from the third to the fourth ventricles is closed, only the ventricles above are distended, and from obstruction at the foramen of Monro the effusion is confined to the lateral ventricles. Progressive effusion is probably in most cases the result of one of the two mechanical processes just mentioned, chiefly of the hindered exit (see Hilton on "Rest and Pain"). A moderate amount of effusion may be associated with diseases (as abscess or tumour) which do not involve any distinct mechanical influence. Its mechanism is obscure.

Primary Hydrocephalus.—Internal effusion sometimes occurs without any of the above causes being traceable. In children with yielding skulls, it is supposed that the mechanical congestion of frequent cough, or the tendency to transudation present in anæmia, may lead to some effusion, but it is not probable that the amount from these causes is ever considerable. At any age, the condition has been known to develop and attain a considerable degree, without, it is said, any other pathological condition being discoverable than the slight traces of ventricular inflammation that are met with in the congenital cases. Dean Swift died from this disease at seventy-eight, after it had existed for three years. ^{But such cases are probably due to} obstruction of the foramina of exit

The symptoms in the acquired form are, on the whole, similar to those in the congenital variety, due allowance being made for differences of age. These are mental weakness, somnolence, coma, muscular weakness, convulsion, contractures, and loss of sight from pressure of the distended third ventricle on the optic chiasma. Temporal hemianopia may, therefore, precede the complete blindness. In young children, with incomplete closure of the sutures, the head readily enlarges, although rarely to the same extent as in the congenital form. In the adult, enlargement of the head is less common, although it

has been known to occur, and the sutures may, as I have seen, become separated, usually after the gradual thinning of the cranial bones described in the section on tumours of the brain (p. 503). It might be supposed that the intra-ocular circulation would show the effects of the increased pressure within the skull, but this is seldom the case, on account of the anastomoses of the ophthalmic vein. Even during the process of separation of the sutures, I have been unable to observe any marked increase in the size of the retinal veins. The course of the acquired form varies much. Death usually occurs at the end of a few months or years. The progress sometimes ceases, and if the affection is moderate in degree, recovery may occur. It is only in children, in whom the ready enlargement of the head enables a certain diagnosis to be made when the disease is still in an early stage, that recovery can be proved. The sac has been known, in very rare instances, to rupture into the subdural space.

DIAGNOSIS.—As just intimated, hydrocephalus can only be diagnosed with certainty when there is distinct progressive enlargement of the head. When the bones are united, internal effusion may be suspected if its indications slowly follow an attack of meningitic character, or accompany the symptoms of a tumour of the cerebellum. But primary hydrocephalus causes only symptoms that are not distinctive, and are much more frequently produced by other morbid processes. Hence, as a matter of fact, the existence of the disease, when it leads to no enlargement of the head, and when the causal indications just mentioned are absent, cannot be recognised.

In slight degree the enlargement of the head may be confounded with that produced by two other causes, rickets and thickening of the bone. In the former the head has a somewhat square form, and not the globular shape characteristic of hydrocephalus. Although the fontanelle may be large, it is not bulged. The other signs of rickets are present in high degree. Thickening of the cranial bones may simulate hydrocephalus at almost any age. I have seen it in a boy of ten, the subject of inherited syphilis, and in a man of fifty. In the latter, a slow progressive enlargement of the head during several years led to a diagnosis of hydrocephalus, but at the post-mortem examination the cranial bones were found to be three quarters of an inch in thickness, and the cavity was of normal size. It is doubtful whether the nature of these rare cases can be ascertained during life. A distinction of internal from external hydrocephalus can only be made by paracentesis, and not always with certainty even by this means, since a thin-walled sac may be readily opened by a comparatively superficial puncture.

The PROGNOSIS of hydrocephalus of any form is usually grave and always uncertain, unless the occurrence of arrest can be distinctly recognised.

TREATMENT.—Whatever lessens the volume of the blood diminishes for a time the amount of the effusion. Thus an attack of diarrhoea lessens the prominence of the fontanelle. But purgatives are inadmissible; to be effective they must be more vigorous than a hydrocephalic child can bear. Diuresis constitutes a safer, but unfortunately less effective, mode of attaining the same end. In no case in which these measures have been used has a permanent effect been produced. Agents that are supposed to promote absorption, as iodide of potassium and mercury, have been extensively tried, but are as a rule powerless, and are sometimes harmful. The most direct treatment, which is unfortunately the most dangerous, is evacuation by puncture with a trocar or aspirator, a small quantity being let out each time, and compression of the skull by elastic bandages kept up during and after the operation. This procedure is of course most suitable to external hydrocephalus, but it has been employed in ventricular effusion, occasionally without ill effects, but with absolute success only in rare instances. The place for puncture was formerly at the outer angle of the anterior fontanelle, but antiseptic measures have permitted trephining and puncture in a locality at which the lateral ventricle is most accessible. Continuous drainage has even been employed, but the result has seldom been satisfactory, whatever the nature of the case.* The first puncture has generally been well borne, and has relieved urgent symptoms of cerebral compression, but the second or third has usually had little effect, and has sometimes been fatal—in some instances apparently because too little time was allowed to elapse between the operations. Puncture of the spinal membranes below the cord has also been resorted to. It can, of course, relieve internal hydrocephalus only when there is no obstruction to the passage of the fluid from the ventricles, and this the operation ^{alone} ~~only~~ may decide. It has relieved urgent symptoms for a time, and has been thought capable of starting absorption in chronic cases, but its demonstrated service is limited to relief. On the other hand, it seems to do no harm.†

Good has been observed to follow simple compression of the skull, a mode of treatment first advocated and energetically employed fifty years ago by Barnard, of Bath. Trousseau employed strips of diachylon plaster, a third of an inch broad, in the following manner:‡—(1) From each mastoid process to the outer part of the orbit on the opposite side; (2) from the hair at the back of the neck, along the sagittal suture, to the root of the nose; (3) over the whole head in such a manner that the different strips shall cross each other at the

* See Keen, 'Phil. Med. News,' 1889, and 'Trans. Tenth Int. Med. Congress;' Robson, 'Brit. Med. Journ.,' 1890.

† See Quincke, 'Berlin. klin. Wochenschrift,' 1891, for the details of ten cases, five of whom were adults; also Wynter, 'Lancet,' 1891, i.

‡ 'Journal de Médecine,' April, 1843, quoted by West, 'Dis. of Infancy and Childhood,' 7th ed., 1884, p. 136.

cortex; (4) a long strip around the head three times, taken first above the ears and eyebrows, and a little below the occipital protuberance, so that the ends of all the other strips shall project below the circular strip; these ends are next to be doubled up on the circular strip, and its remaining two turns passed over them in the same direction as the first turn. It is necessary to watch the effect, and loosen the strips if there are any symptoms of compression. Dr. West advises a broad elastic band as safer and more manageable than the plasters. His experience of pressure in severe cases has not been encouraging, since it has not hindered the accumulation of fluid, and has increased the symptoms of compression of the brain.