 SENSORIMOTOR NEUROLOGY.


In contrast to the great number of clinical and pathological reports on the infantile (Tay-Sachs) type of amaurotic idiocy, reports on the juvenile and especially the so-called late infantile (Bielschowsky) types are scarce. This is probably due to the fact that the last two varieties are rare and do not present such a clear-cut clinical picture as the classic form of Tay-Sachs disease. The case described by Hassin occurred in one of twins, and presented clinical features of unusual interest. A Hungarian-American girl, age 7 years, was admitted to hospital with symptoms of more than three years' duration. She was unable to talk, walk or take nourishment, and presented signs of decerebrate rigidity. She lay motionless on her back, the head retracted in the median line, the eyes and mouth half open, and the hands and fingers markedly flexed; the legs were adducted and extended at the hip and knee joints the feet in a position of equinovarus. There was total absence of voluntary movement, but constant muscular twitching occurred in the eyelids and muscles of the lower extremities. On rotation of the head, with the chin toward the left shoulder, the left forearm after a few seconds became slowly extended at the elbow, the leg on the corresponding side becoming more rigid, while the opposite extremities became more flexed at the elbow and at the knee. Rotation of the head to the right caused extension of the right forearm and leg, with flexion of the contralateral extremities. The tendon reflexes were exaggerated, the abdominals and plantars absent, and the pupils sluggish in their reaction to light. Ophthalmoscopic examination revealed bilateral optic atrophy. The Wassermann reaction was negative.

Examination of the brain showed marked fronto-parietal atrophy, ventricular dilatation and atrophy of the cerebellum. The pathological changes included widespread cellular degeneration typical of amaurotic family idiocy; vacuolated cells containing amorphous deposits; atrophy of all cerebellar layers; rarefaction of medullary nerve fibres; marked degeneration of the deeper layers of the occipital lobe, and of the ganglion cells of the nucleus ruber; absence of secondary degeneration of the white substance of the brain and spinal cord; lipoid accumulation in the adventitial spaces of the blood vessels and in the subarachnoidal space and but little inflammatory phenomena.

The author concludes that the distinguishing features of the late infantile type of amaurotic family idiocy are the existence of marked cerebellar changes and the relatively slight involvement of the optic thalamus.

Decerebrate rigidity is a prominent feature in all types of this disease, and should therefore suggest this morbid condition in obscure cerebral lesions.

R. M. S.
ABSTRACTS


A case is fully described both as to its clinical findings and post-mortem appearances. Insistance is laid on the importance of the presence of eosinophils in the cerebrospinal fluid especially when found in conjunction with psychical deterioration and extrapyramidal lesions. Incidentally attention is drawn to the possibility of asymmetry of position occurring with an intact cerebellum as a result of pyramidal lesions, as in the present case. The author considers that the absence of Janischewski’s sign (turning inward of the palms when the arms are stretched vertically upwards with the palms forwards) is of importance in excluding cerebellar lesions.

R. G. G.


The authors describe the interesting case of a man of 48 who had had syphilis fifteen years previously and, later, two attacks suggestive of cerebral gumma, which were cured by specific remedies. He then exhibited signs of Schilder's encephalitis and subsequently died, antispécific remedies being unavailing. Multiple gummata in the organs were disclosed at autopsy. The pathology of the condition is discussed in relation to tumour and specific infection but no definite conclusion is reached.

R. G. G.


The now somewhat old conception of diffuse brain sclerosis is being dismembered by the separation therefrom of three different conditions, viz., the disease of Pelizaeus, Merzbacher, and Krabbe; the blastomatous variety described by Cassirer and Lewy; and Schilder's encephalitis periaxialis. Bielschowsky's new case belongs to the last of these categories.

The patient was a man who at the age of 50 for an unknown reason lost all his teeth and three years later suffered from a vague illness with rise of temperature for which no cause could be found. Four years later similar attacks of fever recurred. They were then associated with apraxic-aphasic phenomena lasting only a few hours, and with a slight left pyramidal disorder disappearing some ten days later when the temperature fell. Three such attacks occurred. In view of the likelihood of some infection careful blood and other tests were made, with negative result. When the patient was examined
Possibility.

They produce total side of cortico-cerebellar authors raise case, loudly. sometimes for a word single encephalitis. In view of evidence. It is uncertain whether it belonged in fact to the class of epidemic encephalitis. In addition to pronounced cerebellar and general symptoms, for a whole month the child remained conscious of the last attack no definite organic signs were found. Yet within a very short time the illness began once more, on this occasion with Jacksonian epilepsy of the left arm. A diagnosis of a typical disseminated sclerosis was made. The patient left hospital apparently very well, and appeared a month later with fever and soon unconsciousness set in. A cerebral tumour was thought a possibility. Coma of three days preceded death.

Pathological examination showed a general absence of myelin throughout the white matter of both hemispheres, with conservation of axones. The change stopped short of the cortex everywhere. The meninges were full of collections of cells (macrophages and lymphocytes); cells filled the vessel sheaths in meninges, cortex and spinal cord; perivascular gliosis-formation was seen in many places, especially in the frontal regions, on the boundary between grey and white matter, occasionally penetrating the former or occurring in it in a scattered fashion; among the demyelinated parts fibrous glia was prominent. These and other points substantiated the diagnosis of encephalitis periaxialis diffusa. The combination of inflammatory and degenerative tissue-reactions is seen in other states than Schilder’s encephalitis, e.g., in general paralysis. Bielschowsky says that the findings in meninges and cortex have not been remarked in this condition before, and that they closely resemble what has been observed by Spielmeyer in spotted fever. A useful collection of cases of diffuse sclerosis of infective origin is given in the article, and a discussion of their relation to disseminated sclerosis.

S. A. K. W.

[69] Abolition of speech (anarthria) during meningoencephalitis.—

The patient was a young girl of eight years, who suffered from an acute illness of an encephalitic type, in which meningeal symptoms were also much in evidence. It is uncertain whether it belonged in fact to the class of epidemic encephalitis. In addition to pronounced cerebellar and general symptoms, for a whole month the child did not make the least attempt to articulate a single word or letter, though the larynx was normal and she constantly cried, sometimes loudly. Recovery was slow and, up to the time of reporting the case, imperfect.

In view of the marked cerebellar symptomatology, suggesting cerebellar or cortico-cerebellar involvement (the pyramidal system was intact), the authors raise the question whether the cerebellar component of the motor side of speech is so necessary that a severe disturbance of the cerebellar apparatus produces total anarthria; that is, whether cerebellar anarthria is a clinical possibility. They do not, however, answer their own question.

S. A. K. W.

Although admitting that the cerebral blood vessels possess no vasomotor innervation, and that they are uninfluenced by adrenaline, the authors nevertheless believe that the cerebral arteries—like those of the retina—are subject to spasmodic occlusion. The causative factor probably depends upon the presence of mechanical or chemical irritants in the vicinity or in the circulation. An etiological and pathological classification of the various types of occlusion is suggested:

1. Those due to local changes in the vessel walls (atheroma or syphilitic arteritis); the commonest and best known.
2. Those associated with Raynaud’s disease.
3. Angiospasms dependent upon some exogenous poison (quinine, nicotine, lead).
4. Those dependent upon some endogenous poison (as in Bright’s disease, hyperpiesis, migraine and possibly idiopathic epilepsy).
5. Angiospasms associated with a neurosis.

The symptoms are those of cerebral ischaemia and are primarily irritative and later paralytic in nature; the symptoms of Dejerine’s intermittent claudication of the spinal cord are cited in illustration. Treatment is mainly symptomatic. Benzyl benzoate combined with luminal is advocated for the immediate relief of the spasmodic manifestations.

M. C.

Contribution to the symptomatology of cerebellar lesions (Contributo alla sintomatologia delle lesione cerebellare).—V. M. Buscaino. Riv. di pat. nerv. e ment., 1926, xxxi, 382.

A case of a cyst in the right cerebellum is described which was diagnosed during life. The author pays special attention to the three following signs;

1. asymmetry in the position of the head, which was inclined to the right side with rotation of the face to the opposite side.
2. Asymmetry of the arms held above the head with the palms facing forward, the right palm rotating inwards.
3. Asymmetry of the arms held horizontally forward, the right hand being at a lower level than the left.

R. G. G.

Segmental macroscopic syringomyelia, probably from ascending neuritis (Siringomielia segmentaria macrosomica del miembro superior derecho con pseudo hipertrofia muscular y grandes alteraciones osceas, de probable origen infeccioso neuro ascendente).—M. Soto and L. E. Ontaneda. Rev. Argentina. de neurol. y psiquiat., 1927, i, 290.

This interesting case raises afresh the question of whether an ascending neuritis of infective origin can invade the central nervous system locally, producing
a clinical syndrome of syringomyelia with pronounced trophic changes of corresponding distribution.

The patient, a man of 36, had had an operation on the right hand for a local ulceration caused by the deposition of some larva (nature not mentioned) under the skin. Some twenty days after he felt acute pains in the cicatrix, radiating upwards. After their disappearance an interval of some nine years passed ere the other symptoms, suggestive of syringomyelia, gradually made their appearance. The trophic signs were confined to the right arm, the sensory changes to the segments from C3 to D6 inclusive, also on the right. Among the earliest spinal or root symptoms should be mentioned muscular twitching of the flexors of the right hand and fingers, a rare symptom in syringomyelia, and thought by the authors to have resulted from toxic invasion of the anterior horns. In any case the strictly local development of all the symptoms is undoubtedly impressive, and lends support to the view that an ascending neuritis may reach the cord.

S. A. K. W.


The authors report a case of a barmaid of 40 who came under observation with a peripheral neuritis of alcoholic origin. Symptoms were present referable to the eighth nerve, viz., vertigo and deafness. Examination of the cochlear and vestibular apparatus revealed spontaneous nystagmus; Rinne positive, Weber negative, Schwabach negative; slight diminution of auditory acuity; bilateral labyrinthine hyperexcitability.

Eleven other cases collected from the literature are reviewed. Males are affected more often than females (10 to 1). The onset of symptoms is usually sudden. The cochlear division suffers more than the vestibular; the affection is always bilateral, though not necessarily to the same degree on the two sides. Deafness and a continuous non-pulsatile tinnitus are the two main symptoms. Rinne’s test is usually positive. Among the vestibular manifestations vertigo is the commonest; nystagmus is rare. The prognosis is variable; from the eleven cases quoted three were cured, three improved slightly and three became worse; two patients could not be traced. The authors cannot recommend any particular line of treatment other than abstention from tobacco and alcohol.

M. C.


The case was that of a youth of 20, with advanced postdiphtheritic palsies involving masticatory muscles, shoulder, arm and leg musculatures, as well as that of the trunk. In addition, the palate was paralysed, and the reverse
of the Argyll Robertson phenomenon was noted in the eye muscles. Severe sensory changes existed, implicating deep but not superficial sensibility. As in this case no serum treatment was administered, no connection between serum and paralysis was possible.

The case is unusual because of its progressive character.

S. A. K. W.

[75] Contribution to the knowledge of paraplegia in flexion (Babinski type) of cerebral origin (Contribution à la connaissance de la paralplégie en flexion, type Babinski, d'origine cérébrale).—Ludo van Bogaert and Rudolph Ley. Jour. de neurol. et de psychiat., 1926, xxvi, 547.

Among the causes of a cerebral variety of paraplegia in old age and arteriosclerosis, the type described by Marie and Foix is one of the more unusual. The case reported in the present paper was one of multiple vascular lesions, giving rise to a pseudobulbar syndrome, together with many of the features of Parkinsonism; the intellectual state was one of senile dementia. The lower extremities were paraplegic in flexion (type Babinski). The right plantar response was of the extensor type and the left one flexor. Autopsy revealed sclerosis of the basilar arteries; there was a progressive subependymal necrosis; numerous lacunes of disintegration were present in the basal ganglia of both sides. The left globus pallidus and caudate nucleus showed an état fibreux. The right pyramidal tract was degenerated in all its length.

M. C.


By hemiplegia in flexion is understood a condition in which the lower extremity exhibits flexion corresponding in minor degree to that of the upper. According to the author this peculiarity and rarity is always associated with the syndrome of Foville. If the latter is on the same side as the hemiplegia in flexion the lesion is in the opposite crus cerebri; if on the opposite side, the lesion is in the upper pons.

S. A. K. W.


This reflex consists in a contraction of the recti abdominis and the adductors of the thigh on striking the symphysis pubis. It is a true tendoperiosteal reflex and is not the same as the abdominal reflex, for the two may be found dissociated in conditions such as disseminated sclerosis and tabes. In hemiplegia both abdominal and medio-pubic reflexes are absent on the affected side, which is a paradoxical reaction.

R. G. G.
Corpus callosum and dyspraxia (Corpo calloso e disturbi disprassici).

—CARLO BERLUCCHI. Il Cervello, 1926, v, 1.

The first case here reported was one of tumour involving mainly the posterior and (in part) middle thirds of the corpus callosum, but dyspraxic or apraxic phenomena were not found during life. References are given to four or five recent callosal cases similarly characterised by absence of apraxia, though specially looked for. Accordingly, some revision of Liepmann's theory seems desirable, in the author's view.

His second case was also one of callosal tumour, which invaded much more than that structure in its posterior third, seeing that it passed outwards under the ascending parietal gyrus, involved the limbic area, and compressed also the caudate. In this case symptoms of the nature of tonic innervation were present, but no apraxia.

Some discussion of tonic innervation (Zwangsgreifen) is given, and of its possible pathogenesis, especially of the question whether it is of callosal origin.

S. A. K. W.

The reflex from the external malleolus and the phenomenon of Piotrowski
(Il riflesso del malleolo esterno e il fenomeno di Piotrowski).—K. SAGIN. Riv. di pat. nerv. e ment., 1926, xxxi, 264.

The phenomenon of Piotrowski consists of a contraction of the gastrocnemius with flexion of the foot when the lower half of the tibialis anticus is struck with a percussion hammer. A similar result from striking the external malleolus is held by Sagin to be an extension of this phenomenon when reflex activity is specially raised. Others have denied this on the ground that the Piotrowski reflex is a musculo-tendinous response while that from the external malleolus is a periosteal-tendinous response. The author, however, maintains that the external malleolus reflex is never found in the absence of that of Piotrowski.

R. G. G.

An associated thumb-movement as a sign of pyramidal disease
(Daumen-Mitbewegungsphanomen als Pyramidenzeichen).—R. WARTEMEBERG. Klin. Woch., 1927, vi,

In many cases of pyramidal affection, slight or severe, the author has noted that when the patient tries to flex his fingers against resistance the corresponding thumb flexes and opposes. The sign has been described before, but this method of eliciting it is simple and useful. It has been shown to occur even when other pyramidal signs are minimal.

S. A. K. W.
Subacute combined cord degeneration.—Walter F. Schaller. Trans.
Amer. Med. Assoc., 1926.

The case was that of a man of 58, who presented the usual symptoms of sub-
acute combined degeneration, though the sensory symptoms were minimal,
and in addition complained of occipital headaches and impaired hearing. The
heart action was weak and intermittent, and cold sweats occurred.

Considerable interest attaches to the pronounced changes found in the
brain cortex, which are distinctly unusual in the ordinary variety of the affection.
Cortical atrophy seemed to have arisen from a primary involvement of nerve
fibres, with secondary changes in nerve cells. Lipoid degenerations were
present in the white matter, and plaques also in subcortical tissues. There
was marked increase of glia without infiltrative or exudative reaction.
It is also noteworthy that no special degree of anæmia was present during life.

S. A. K. W.

Familial spinal spastic paraplegia (Paraplegia spinale spastica fami-

The author describes the two cases of a brother and sister with spastic para-
plegia. They were about 50 years of age, while another brother had died in
early childhood with the same disease.

After discussing the views of various authors he come to the conclusion
that congenital diplegias without mental symptoms and cases of family spastic
paraplegia are examples of the same primary condition, viz., a failure of the
proper development of the pyramidal tracts perhaps in the medulla which
either prevents them functioning at all (the congenital cases) or causes the
structures to die earlier than do the rest of the nervous structures.

R. G. G.

Observations and studies on the hereditary transmission of progressive
muscular atrophy of the Charcot-Marie type (Observazioni e studi
sulla transmissione ereditaria dell’ atrofia muscolare progressiva
tipo Charcot-Marie)—L. de Lisi. Riv. di pat. nerv. e ment., 1926,
xxxii, 390.

An elaborate study of family histories in this disease leads to the conclusion
that while Mendelian laws cannot be applied simply or dogmatically, trans-
mision may be recognised in this condition with the disease-process as a domi-
ant, a simple recessive or a cosexual recessive.

R. G. G.

A study of the vibratory sensation.—R. S. Ahrens. Arch. of Neurol.
and Psychiat., 1927, xiv, 793.

Present knowledge concedes that the vibratory sensation may be felt in the
soft parts, but that it is best recognised when contact is made with bone. As
in any test which depends on the subjective sensations of the patient, a con-
siderable degree of error is to be expected. Further, in this test we are not
dealing with a clear-cut cessation of sensation, but rather with a gradually diminishing intensity of sensation which fades into absence. A study of normal individuals shows that there is no predominating increased sensitivity in either right or left lower extremity or anterior sacral spine. In the upper extremity, however, the author found a definitely increased sensibility on the right side, a fact which suggests that deep sensibility is more acute in the extremity most used. In the field of neurology the most striking findings are encountered in tabes dorsalis, and there appear to be few exceptions to the rule that in tabes there always occurs a reduction in the time of appreciation of vibrations of fixed amplitude in the lower half of the body, and that this reduction in time is singularly constant over the sacrum.

Pernicious anaemia is another disease in which the quantitative estimation of the vibratory sensation promises to be of value. So far, no patient examined by Ahrens has shown a normal response to this test, the general tendency being for the sensation to be impaired or lost in the distal portions of the lower extremities with a gradual approach to normal in the upper extremities. In syphilis of the nervous system, other than tabes, changes in the vibratory sensation are not constant or characteristic, while in disseminated sclerosis the peculiar patchwork pathology may give any variety of sensory findings. In the early stages of posterolateral sclerosis a marked impairment is found in the lower limbs with an approach to normal as the upper extremities are reached, and as the disease progresses the curve approaches that of advanced tabes. Lastly, in peripheral neuritis there is a generalised impairment of the vibratory sensation, the upper and lower extremities being equally affected.

R. M. S.


This brief communication deals with a rare phenomenon of peculiar interest, though unfortunately the details are not so complete as might be desired.

The patient, a man of 58, is a typical case of tabes with optic atrophy, resulting in blindness; the symptoms have been observed for at least eight years. Some two years ago, for the first time, he became aware of a sensation of extraordinary lightness of his body, with absence of all sense of fatigue after prolonged muscular effort. At first this experience produced in him a feeling of euphoria and hopefulness as to recovery, but was soon after followed by anxiety as to its real nature.

According to the author’s account, examination shows no loss of muscular sense in arms or legs, but there is loss of pain sense and diminution of vibration sense in the latter limbs. However, Romberg’s sign is present, while the patient feels when walking as though he were stepping on rubber. When he gets out of bed he has a pleasant sensation as though he were rising into the air “like an indiarubber balloon.” This sensation recurs at intervals...
during the day; he often feels as if he were suspended or floating in the air. As a result, in the lower limbs he frequently exhibits the so-called dancing reflex, or reflexus saltatorius, commonly considered to be more or less exclusively a hysterical phenomenon, but not so in this case.

The illusion of levitation and of absence of any feeling of bodyweight deserves much closer examination than is here given it.

S. A. K. W.


Doubt has not infrequently been cast on the value of the pupillary reaction test of Wernicke as a means of differentiating between an anterior and a posterior hemianopia, and the author here adduces further evidence against its usefulness in this respect. Retrochiasmal lesions of one bandelette or tract by no means constantly result in symmetrical defects of the visual fields, and the various clinical combinations that may occur are examined at some length. Thus hemianopia of one eye with normal vision in the other is one possibility; a paracentral scotoma on one side and no defect on the other can also occur; a third possibility is homolateral integrity and loss of colour vision in part in the crossed eye. Other rarities: are enumerated.

S. A. K. W.


The author has usefully compiled an account of previously recorded cases of this condition and adds a new case of his own. In all bilateral cases there is a characteristic dissociation of the double actions of the internal rectus; other ocular and pupillary defects may or may not accompany it. The lesions are sometimes syphilitic, sometimes vascular and inflammatory, sometimes traumatic. Only one case (Spiller's) has been examined pathologically. Lutz' case was that of a little girl of ten, in whom the ocular symptoms supervened on mumps. It seems clear that the lesion must involve the posterior longitudinal fasciculus.

S. A. K. W.


The patient was a man of 40, who showed the following ocular symptoms. (1) Right eye.—On his looking to the left, the right internal rectus failed to contract and no movement of that eye took place; on convergence, however,
the muscle contracted normally. (2) Left eye.—With lateral deviation to the right, the left internal rectus contracted normally, but on convergence its reaction was nil.

The author explains that a single lesion will account for the phenomena; there are supranuclear paths to each internal rectus nucleus, which cross in the midline; if uncrossed fibres are responsible for movement of the eye to one side, and crossed fibres for convergence, then a lesion above the right internal rectus nucleus, implicating uncrossed fibres to it and crossing fibres to the corresponding nucleus of the other side, will produce the symptoms here described.

S. A. K. W.


The commonest annular opacity of the cornea is the well-known arcus senilis, due to degenerative changes in Bowman’s membrane and in the peripheral corneal parenchyma. A green-brown colouration of the peripheral margins of the cornea is an occasional finding in pseudosclerosis and in progressive lenticular degeneration. According to Bostroem it does not always take the form of a complete ring, and probably results from urobilin depositions in Descemet’s membrane and in the deepest layers of the cornea.

A still rarer form is here described, known as the keratitis annularis of Vossius, presumably of inflammatory origin.

S. A. K. W.


The characteristic features of Meniere’s disease are tinnitus auris, and reduction of auditory acuity and imperfect bone-conduction on the same side. To this combination is added the occurrence of vertiginous attacks, rudimentary, moderate, or severe. Actual loss of consciousness is a rarity. As a rule prodromata are wanting. During the attack nystagmus of a violent character is practically invariable; in ten out of twelve cases observed by the author the rapid movement of the nystagmus took place towards the affected side, being essentially of a rotatory type, and, in the majority of the cases, purely rotatory. Much less common is a horizontal nystagmus towards the sound ear.

Of much significance is the author’s finding that even in aggravated cases of the affection reactions to rotatory and caloric tests are normal. Moreover, he has shown that even in the course of an attack it is sometimes impossible to obtain any reduction of the reaction normally following caloric action on the peripheral apparatus.
He is convinced that Menière’s disease has nothing to do with the labyrinth itself, considered as a peripheral organ. His explanation is that it is caused by central disorder of the otolith nuclear complex in the medulla oblongata, and that it is akin, with migraine, to epilepsy, though not strictly either otic migraine or aural epilepsy. The supposition is that the diseased condition is preceded by toxic ear trouble and the existence of dominant irritation bombarding the medullary centres; when tension has reached a certain point, a sudden discharge occurs, viz., the Menière attack. Vomiting and the so-called ictus laryngeus are subsidiary phenomena.

S. A. K. W.

PROGNOSIS AND TREATMENT.


Dr. Bordier’s treatment consists of a combination of spinal radiotherapy with diathermy. After determination of the main spinal site or sites of the lesions of poliomyelitis appropriate X-ray radiation is applied; for children under two years the dose is about 200 units R per seance, for those who are older, an increase in proportion; for the adult, 1000 units R. The direction of the rays is at right angles to the vertebral lamina. After three successive irradiations an interval of about 25 days elapses ere a second series is undertaken.

The diathermy part of the treatment is intended to favour the nutrition of the tissues of the paralysed limb. It should be utilised from the outset, independently of the radiotherapy, and treatment should be performed daily, for about ten minutes. Dr. Bordier also employs the sinusoidal current. He gives figures derived from the results of this treatment in other hands than his own, according to which cures are claimed in 15 per cent. of cases, and amelioration in 75 per cent. (total number of cases treated, 65).

S. A. K. W.


The interest of this brief communication resides in the description of the treatment of acute cases of epidemic encephalitis by dye-therapy. A solution of neutral acriflavine is freshly prepared, dissolved in normal saline, boiled and cooled to body temperature and given preferably by syringe with a fine needle and injected slowly. The author begins with 10 c.c. of 0.5 per cent. solution of neutral acriflavine in normal saline, and repeats the intravenous injections daily with a gradual increase till 25 c.c. is reached. He gives details of several cases in which excellent results were promptly obtained.

S. A. K. W.